Day 1

7:30 – 8:00  
Registration

8:15 – 10:00  
Anatomy and Physiology of the VOR and Oculomotor Systems

10:00 – 10:30  
Exhibitor Break

10:30 – 12:00  
Clinical Vignettes

12:00 – 12:30  
Lunch

12:30 – 1:45  
Distinguishing Central from Peripheral Causes of Vestibular Symptoms

1:45 – 2:30  
LAB: Oculomotor Exam – Central vs Peripheral

2:30 – 3:00  
Break

3:00 – 5:00  
Lecture/LAB: Vertical Canal (Posterior/Anterior) BPPV – Maneuvers based on RCT evidence

Day 2

8:00 – 9:30  
VOR Adaptation Exercises: Past, Present, and Future

9:30 – 10:00  
Break

10:45 – 12:00  
The VOR in Age and other Neurodegenerative Diseases

12:00 – 2:00  
Lunch/Business Meeting

2:00 – 3:30  
Lecture/LAB: Horizontal Canal BPPV – Maneuvers based on RCT evidence

3:30 – 3:45  
Break

3:45 – 4:30  
Wrap up QnA

Objectives

1. Identify crucial examination tools and treatment methods to identify pathology of the oculomotor systems
2. Describe the difference in central vs peripheral manifestations of vestibular disorders.
3. Describe evidence for abnormal vestibular function within two models of neurodegenerative disease (i.e. Diabetes, Multiple Sclerosis)
4. Describe/discuss the evidence for loss of vestibular function damage associated with healthy aging
Anatomy and Physiology

Michael Schubert PT PhD
Johns Hopkins University School of Medicine

Vestibular System inside the Temporal Bone of the Skull

What does the Vestibular System do?

Healthy Function
• Gaze Stability
• Postural Stability (Balance)
• Orientation in Space
• Cerebral perfusion (Autonomic NS)

Abnormal Function
Oscillopsia
Dysequilibrium
Abnormal sense of movement / orientation

Signs
Saccades after head rotation
Decreased visual acuity during head movements
Ataxia
Imbalance
Unique Anatomy and Innervation Pattern
6 Semicircular Canals (horizontal, anterior, posterior)
2 Otolith organs (utricle, saccule)

Superior Vestibular Nerve innervates aSCC, hSCC, utricle

Inferior Vestibular Nerve innervates pSCC, saccule

VOR Pathway
• MOVIE

VOR Physiology
• 3 neuron arc
• Produces compensatory eye movements in response to head movement
  • Equal amplitude and speed in the opposite direction
• Allows for stable gaze during head movements
  • Angular and Linear VOR
• Ocular Counter Roll
**Vestibular Neurophysiology 101**

1st Rule: Vestibular afferents encode angular head rotation via semicircular canals, and linear head acceleration and tilt from the otolith organs.

2nd Rule: Stimulation of a semicircular canal produces eye movements in the plane of that canal.

3rd Rule: For high accelerations, head rotation in the excitatory direction of a canal elicits a greater response than does the same rotation in the inhibitory direction.

4th Rule: Reverberating circuitry in the vestibular nuclei allows the brain to detect low-frequency VOR by preserving the nystagmus, known as "velocity storage."

5th Rule: Sudden changes in saccular activity evoke changes in postural tone.

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**Vestibulo-Ocular Reflex (VOR) Physiology**

- Deflection of the stereocilia toward the single kinocilia in each hair cell leads to excitation (depolarization), and deflection of the stereocilia away from the kinocilia leads to inhibition (hyperpolarization).
- Deflection of the stereocilia occurs by motion of the endolymph results in an opening (or closing) of the transduction channels of hair cells, which changes the membrane potential of the hair cells.
- In the horizontal SCC, hair cells are oriented so that endolymph motion toward the ampulla causes excitation.
- In the vertical SCCs (posterior and anterior) hair cells are oriented so that depolarization occurs when endolymph moves away from the ampulla.

---

**Vestibular Haircells – Morphologic Polarization**

The semicircular canal haircells are bi-directionally sensitive and always excited as the stereocilia move towards the kinocilia.

- The kinocilia of the otolith organ haircells are oriented w/ respect to striola:
  - Utricle – towards striola
  - Sacculus – away from striola
- Otolith organs sensitive to multiple directions.
Type I and Type II Hair Cell

Type I hair cells are goblet-shaped:
- Afferents terminals synapse via cup-like/calyx nerve terminal
- Higher proportion in central zone

Type II hair cells are more cylindrical in shape:
- Afferent terminals synapse via conventional button/bouton nerve terminal
- Higher proportion in peripheral zone

Engstrom et al 1979; Diksic et al 2005

Semicircular Canal Cupular Anatomy

Central and Peripheral Regions

Cupular Dynamics

- Deflection of cupula causes excitation or inhibition
**SCC Orientation**

- Canals roughly orthogonal with each other
- End-organ on one side can sense motion in all directions
- hSCC tilted up ~30°
- Pairs of canals are coplanar
- Push-pull arrangement – canals are mated
  - RALP, LARP, Yaw

**Gain of the VOR = Eye velocity/head velocity**

**Phase of the VOR = timing relationship between head and eye position**

**VOR Physiology – Excitation Inhibition Asymmetry**

Recording from vestibular nerve afferent - Excitatory stimuli cause greater response than inhibitory stimuli

**Inhibitory Cut-Off**

During rapid head rotation, the contra-rotational VOR is inhibited to Zero and can no longer detect the head rotation

SIX EXTRAOCULAR MUSCLES CONTROLLED BY THREE CRANIAL NERVES

<table>
<thead>
<tr>
<th>Sensory Route</th>
<th>Motoneuron</th>
<th>Muscle Action</th>
<th>Cranial Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Horizontal</td>
<td>Right abducens</td>
<td>Left abducens</td>
<td>CN III (oculomotor)</td>
</tr>
<tr>
<td>Anterior</td>
<td>Left abducens</td>
<td>Right abducens</td>
<td>CN VI (abducens)</td>
</tr>
<tr>
<td>Posterior</td>
<td>Left abducens</td>
<td>Right abducens</td>
<td>CN IV (trochlear)</td>
</tr>
</tbody>
</table>

Right angular acceleration, inertia causes endolymph to lag behind, cupula deflected to the left with inhibition of left hSCC and excitation of right hSCC.

Perception of rightward turn, right beat nystagmus.
CONSTANT VELOCITY ROTATION

• After @ 25 seconds: Endolymph and canal moving together, no relative difference in motion, cupula not deflected, no perception of rotation, no nystagmus

CHAIR STOPS FROM A RIGHTWARD YAW CONSTANT VELOCITY

• Now the chair stops, and so the SCC stops moving but the momentum of endolymph causes it to continue, cupula deflected right, perception of motion left, left beat nystagmus

Otolith Anatomy – Polar Morphism

The kinocilia of the utricular hair cells are oriented toward their striola.
The kinocilia of the saccular hair cells are oriented away from their striola.
Otolith Anatomy and Physiology

- Otolith organs = saccule and utricle
  - Respond to linear acceleration and static head tilt.
- Sensory hair cells project into a gelatinous material that has calcium carbonate crystalline-structure material (otoconia) embedded in it.
- Provides the otolith organs with an inertial mass.
- A central region exists within the utricle and the saccule known as the striola, which divides the otolith organs into two parts.
- The kinocilia of the utricular hair cells are oriented toward their striola, whereas the kinocilia of the saccular hair cells are oriented away from their striola.
- As with the SCC, motion toward the kinocilia causes excitation, while motion away leads to inhibition.
- Utricular excitation occurs during horizontal linear acceleration and/or static head tilt and saccular excitation occurs during vertical linear acceleration.

Otolith Physiology

VOR Central Projections to the Cerebellum
Central Projections

- Thalamic projections
- Cortical Projections
  - Parieto-Insular Vestibular Cortex
  - Supplemental Motor Cortex
  - Multimodal sensory areas

Vestibulo-Spinal Pathways

- Medial & Lateral VST – vestibulospinal tract
  - Connect receptors w/antigravity motor neurons in cervical & lumbar spinal cord
  - Maintain head and body posture

- MVST
- Vestibulocollic Reflex
- LVST
  - Ipsilateral increased tone in extensors
  - Reciprocal inhibition of flexors

The Vestibular and Visual Senses are linked

- Vestibular nuclei neurons respond to visual rotation independent of head rotation
- Same firing rate is increased when visual and vestibular stimuli are combined (Keller and Precht 1979)

E. Transient response of one VN to vestibular stimulation.
F. Same unit, response to rotation in room light.
Vestibular and Visual Systems are Linked

The inter‐dependency between vestibular and visual afference enables a means for vestibular compensation

• Unique type of saccade is substituted for a deficient vestibulo‐ocular reflex (VOR)
• The latency and amplitude of these unique saccade are dependent on light

The Vestibular and Somatosensory Senses are Linked

• 41% of vestibular nuclei neurons (cat) are modulated by hindlimb stimulation (cat) (McCall AA et al. 2016)
• In human, limb position, weight distribution, and ground compliance (standing) alter vestibulospinal reflexes (Grasso et al. 2011; Marsden et al. 2002; Welgampola and Colebatch 2001)

Oculomotor Exam
Michael C Schubert PT PhD
Johns Hopkins University
Departments of OHNS and PMR
**Patient Report** - *helps discern what rehab you will apply and if it will be useful*

- You must discern what the patient is feeling
- What do they mean by dizzy, imbalance, spinning?
  - Dizzy: global term implies lightheadedness
  - Vertigo: illusion of motion
- Are these symptoms coming from the vestibular system?
- How can we tell what is causing their dizziness?
- Can we treat their condition? How?

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**History – Consider the Tempo, Symptoms and Circumstance**

<table>
<thead>
<tr>
<th>Tempo</th>
<th>Symptoms</th>
<th>Circumstance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic</td>
<td>Vertigo</td>
<td>Spontaneous</td>
</tr>
<tr>
<td>Acute</td>
<td>Dysequilibrium</td>
<td>Motion provoked</td>
</tr>
<tr>
<td>Spells</td>
<td>Oscillopsia, Lightheaded/Dizzy</td>
<td>Position provoked, Standing / Walking</td>
</tr>
</tbody>
</table>

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**Ocular Motor Examination** *Adapted from Leigh 2006, Kallman 2009, Schubert 2010*

<table>
<thead>
<tr>
<th>Assessment</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Observation</td>
<td>Head Thr, Ocular Thr Reaction, Posis, etc</td>
</tr>
<tr>
<td>Visual Confrontation Testing</td>
<td>Field cuts</td>
</tr>
<tr>
<td>Papillary light reflex</td>
<td>Optic N. or Ocul N III</td>
</tr>
<tr>
<td>Extraocular Movements/ROM</td>
<td>ROM - monocular and binocular</td>
</tr>
<tr>
<td>Ocular Alignment</td>
<td>Cover Tests; Alternate Cover Tests; Maddox Rod</td>
</tr>
<tr>
<td>Gaze Holding</td>
<td>Ability to maintain stable gaze without generation of other eye movements in 3 cardinal planes</td>
</tr>
<tr>
<td>Smooth Pursuits</td>
<td>Ability to maintain dually moving target on fovea of retina</td>
</tr>
<tr>
<td>Optokinetic Nystagmus</td>
<td>Reflexive jerk nystagmus occurring w/ visual flow</td>
</tr>
<tr>
<td>Saccades</td>
<td>Ability to make single rapid eye movement to refocus image on fovea of retina</td>
</tr>
<tr>
<td>VOR</td>
<td>Ability to stabilize gaze while head moves</td>
</tr>
</tbody>
</table>
Observation:

Abnormal eye alignment due to a right cranial nerve III palsy secondary to TBI.

Abnormal/compensatory head posture to alleviate diplopia caused by a vertical misalignment of the eyes due to a right CN IV palsy due to TBI.

Clinical Exam – Room Light

- Ocular ROM
- Spontaneous nystagmus
- Gaze evoked nystagmus
- Smooth Pursuit
- Saccade
- VOR head impulse test
- VOR cancellation
- Vergence
- Dynamic Visual Acuity
- Cover Cross-Cover Test

Clinical Exam – Frenzel Lens (fixation removed)

- Spontaneous nystagmus
- Gaze evoked nystagmus
- Head shaking and/or Mastoid Vibration
- Pressure Tests (Valsalva, tragal compression, pinched-nose Valsalva)
- Hyperventilation
- Positioning Tests (Dix Hallpike, roll test, etc...) – to be covered in BPPV Lecture
- Cover Cross-Cover Test – to be covered in Central Lecture/Lab
Ocular ROM

**WHAT TO DO:**
Assess alignment with both eyes viewing

Assess alignment with only one eye viewing (fusion prevented)

Spontaneous and Gaze Evoked Nystagmus

**WHAT TO DO:**
* May need to hold head still to ensure VOR is not engaged if person moves head when asked to move eyes

Gaze-Evoked Nystagmus
Do you think this is Peripheral or Central?
HEAD SHAKING NYSTAGMUS (HSN) TEST

WHAT TO DO:
- Horizontal 2Hz for 20 reps
  - Abnormal is horizontal nystagmus > 3 beats, or increase of an existing nystagmus
  - Vertical nystagmus after horizontal head shaking (cross coupling) – SUGGESTS CENTRAL
- Vertical - 2Hz for 10 reps
  - In UVH, post vertical HSN may beat in the opposite direction

Clinical Oculomotor Exam – Pressure/Labyrinthine Integrity Tests

Pressure tests assess integrity of the labyrinth

WHAT TO DO:
- Tragal Pressure – firmly close the flap over the external auditory canal with finger,
- Closed glottis Valsalva – ask patient to bare down (as if lifting heavy object)
- Pinched nose Valsalva – have patient attempt to breath through pinched nostrils

Pressure/Labyrinthine Integrity
Closed Glottis Valsalva
Head Impulse Test – each semicircular canal

**WHAT TO DO:**
Rapid head rotation that is of a:
- High acceleration
- Small amplitude
- Unpredictable timing and direction

Sensitivity / Specificity
- Vestibular nerve section 100% / 100%
- Report of dizziness 35% / 96%
- UVN - 71% / 82%
- BVH - 64% / 82%
- Data suggests at least 40% caloric asymmetry for a +HIT

Which VOR is Abnormal?

What VOR is Abnormal
Vertical Canal Head Impulse Test (HIT) LARP/RALP Neck Neutral

Vertical Canal HIT LARP/RALP 45 head rotate 1st

Clinical Exam - Vertical Canal HIT
Dynamic Visual Acuity Test – Clinical Version

DVA is scored as the difference between static visual acuity and visual acuity during 2 Hz head oscillations
-2-line or less difference is considered normal
-3-line or greater difference is considered abnormal and suggests a vestibular deficit
-should improve with compensation

www.i-see.org/eyecharts.html

WHAT TO DO:
Passively move head side to side at 2 Hz while patient reads the chart
Subtract acuity during head motion from acuity head still

Hyperventilation Test

WHAT TO DO:
• Have patient attempt to ‘blow out a candle from across the room’
• Do this for 60 second using deep, rapid breaths at the rate of 1 breath / sec
• Nystagmus (either new or a reversal of an existing) is a positive test and suggestive of demyelinating disease process on the vestibular nerves.
• Multiple Sclerosis, Neurofibromatosis, Vestibular Schwannoma, etc.

Clinical Oculomotor Exam: Saccades

WHAT TO DO
Ask your patient to make 30deg up/down/left/right saccades to discrete target
Target should be 18-24" from face (~50cm)
• Accuracy (overshoot always abnormal, 2-3 undershoots is OK)
• Velocity – saccades should be very rapid
• Timing – is there a delayed initiation?
Abnormal Laboratory Pursuit Test in 20 y.o. after TBI.

**WHAT TO DO**
Ask your patient to follow your slowly moving target up/down/left/right
Target should be 18-24" from face (~50cm)
Examine for
- Velocity – be careful of target speed
- Abnormal if there are corrective, or catch-up saccades

---

VOR Cancellation

**WHAT TO DO**
- Two methods:
A – Patient holds arms straight out in front with hands together and thumbs up. Examiner rotates head and trunk together while patient looks at their thumbnail
B – Examiner holds patient head at the side, asks patient to look at nose. Examiner then rotates the entire head side to side
- Do not move any faster than 1 Hz!
- Why?
- Abnormal if there are corrective saccades during the head rotation
Vergence: Adjusts eyes for different viewing distances

- Region of cortical control depends upon type of associated eye movement (pursuit, saccades etc)
- Located in the midbrain
- Provides depth perception when making associated eye movements
- Influences the VOR during near target head rotation

Positional Testing

- We will cover the Dix Hallpike and Sidelye tests in BPPV lecture
- We will cover Ocular Alignment in the Oculomotor Lecture and lab

Leftward gaze  Rightward gaze
What Peripheral Lesion Might Cause upbeating nystagmus with Mastoid Vibration?

• 2 years before - A 59 year old female originally presented with sound and pressure induced dizziness, pulsatile tinnitus, and autophony.
• Under infrared (IR) goggles, downbeat nystagmus was observed in the plane of either superior canal after ipsilateral application of a 2 kHz tone at 100 decibels (dB HL).
• Ocular VEMP testing with 500 Hz tonebursts at 125 dB nHL showed abnormally high amplitudes of the evoked potential: 23.8 microvolts on the right and 18.4 microvolts on the left (normal range 0-17 microvolts).
• CT scan was performed, which showed bilateral dehiscence of both superior canals.
• Right SCD repair 1st (middle fossa craniotomy), with plugging and resurfacing of a 3.0 by 0.8 mm defect of the superior canal.
• The surgery resolved the right sided symptoms, but she had persistent left sided symptoms of pulsatile tinnitus, autophony, and sound induced vertigo.
• 1 year later, she underwent a left middle fossa craniotomy, with plugging and resurfacing of a 4.5 by 0.3 mm defect of the left superior canal.

What stimulation can cause this result – Upbeating Nystagmus with Mastoid Vibration?

• In Bilateral Superior Canal Dehiscent plugging and mastoid vibration.....
  • The Superior canals are deactivated
  • The Horizontal canals negate each other
  • The Posterior canals are functioning normal
    • Torsion from each pSCC negates itself
    • Downward excitation from each pSCC is summed residual
    • Result is Upbeating Nystagmus

What is this?

hSCC BPPV? – Unlikely
• Canalithiasis
  • Geotropic
  • Short lived, though some studies report >60sec and 1 reported 90sec duration but nystagmus always stopped (de la Meilleure et al 1996)
• Cupulolithiasis
  • Apogeotropic – long duration (adherent to cupula)
  • Apogeotropic – short duration (free floating within ampullary segment of the hSCC)
What is this?

• Migraine? - Unlikely, based on
  1. Our patients age (May and Schulte 2016; Morgante et al 2015)
  2. Our patient did not meet the criteria for vestibular migraine (Lempert et al 2012)
  3. Mean peak slow phase velocities reported in those with definite migraine is much slower (~12-20d/s) compared with our 62-74d/s peak slow phase velocity (Lechner et al 2014)

What is this?

Light Cupula? Appears to be the best fit

• Persistent direction-changing geotropic nystagmus with vertigo that stops when the patient lies supine and has their head rotated in yaw towards the affected ear (known as the null point).

<table>
<thead>
<tr>
<th>Nystagmus Characteristics</th>
<th>Light Cupula</th>
<th>hSCC</th>
<th>Our Case</th>
</tr>
</thead>
<tbody>
<tr>
<td>Geotropic Nystagmus (GN)</td>
<td>YES</td>
<td>YES</td>
<td>YES</td>
</tr>
<tr>
<td>Persistent</td>
<td>YES</td>
<td>NO</td>
<td>YES</td>
</tr>
<tr>
<td>Prone reversal</td>
<td>YES</td>
<td>NO</td>
<td>YES</td>
</tr>
<tr>
<td>Fatigability</td>
<td>NO</td>
<td>YES/NO</td>
<td>NO</td>
</tr>
<tr>
<td>Null plane</td>
<td>YES</td>
<td>YES/NO</td>
<td>N/A</td>
</tr>
<tr>
<td>Latency</td>
<td>NO</td>
<td>YES</td>
<td>NO</td>
</tr>
</tbody>
</table>

hSCC – horizontal semicircular canal; Fatigability – reduced velocity with repeat positional testing. *Transit requires ear and direction of roll can vary. N/A not performed. Adapted from Kim et al 2014

VPT treatment likely did NOT work

• After treatment, repeat roll testing revealed the nystagmus and vertigo were still present and intense with no fatigue.
• The patient was given instructions to avoid the supine position for the remainder of the day only, and to continue with his normal activities
• Patient returned 1 week later with no nystagmus or vertigo, resolved
• DHI score 4 out of 100
• ABC rated at 98 percent.
• No evidence of geotropic nystagmus in either the Dix-Hallpike or the roll test positions
Conclusion

- Repositioning maneuvers not effective in Light Cupula (Ban et al 2016)
- Treatment has also included vestibular suppressants for symptomatic relief
  - Positional vertigo and nystagmus were improved within 1 week in 74% and lasted for 3 or 4 weeks in the other 26% (Kim et al 2014)
- If you see persistent geotropic (horizontal) nystagmus in roll/Dix Hallpike positions
  1. Wait at least 90sec to ensure the nystagmus does not stop
  2. Test for fatigue by repeating the positional test
  3. Attempt to identify a null point by rotating the head towards either side slowly perhaps as much as 40°

Nystagmus associated w/ Migraine

N=26 patients captured during an acute vestibular migraine
- **100% had positional nystagmus (fixation blocked)**
- 19% had Spontaneous & Gaze Evoked nystagmus
  - Downbeating, Upbeating, Torsional & combinations
  - NOT visualized without fixation blocked
- 35% had abnormal horizontal headshaking nystagmus

(Polensek & Tusa, 2009)
Oculopalatal Tremor

- Acquired pendular nystagmus with tremor of the soft palate
- Can be unilateral or bilateral
- Tremor Hz is ~2Hz
- Due to disruption of dentate-red-olivary pathway; the inferior olive generates a normal signal that is no longer inhibited by the cerebellum

If you see spontaneous pendular vertical or roll nystagmus, check the soft palate

---

**Bow and Lean test may help lateralize hSCC BPPV**

Have the patient bend head down (bow) and up (lean), wait in each position and check for nystagmus

Location of the otococia within the lumen of the SCC determines direction of nystagmus

**MUST CONSIDER RESULTS OF BLT WITH THE RESULTS FROM SUPINE ROLL TEST**

- Long Arm of hSCC
  - Bow: Otoconia will move towards the cupula (ampullopetal) and cause nystagmus that will beat towards the affected ear
  - Lean: Reverse with pitch up.
- Short Arm of hSCC
  - Opposite nystagmus pattern from long arm
ASSESSMENT AND MANAGEMENT OF CENTRAL VESTIBULAR DYSFUNCTION

Michael C Schubert PT PhD
Johns Hopkins University
Department of OHNS

OUTLINE

• Anatomy of central vestibular pathways
• Distinguish central from peripheral vestibular lesions
• Common causes
• Evidence based practice
WHAT IS CENTRAL VS PERIPHERAL?

Peripheral vestibular system:
- Vestibular end organs (3 semicircular canals and 2 otolith organs)
- Vestibular portion of the VIIIth cranial nerve

Central vestibular system:
- Vestibular nuclei
- Vestibuloocular pathway
- Vestibulospinal pathway
- Vestibulocollic pathway
- Vestibulocerebellum
- Vestibulocerebral pathways
- Primary and secondary cortical areas

CENTRAL VESTIBULAR ANATOMY

Secondary vestibular afferents synapse with extraocular motor nuclei, the spinal cord, or the flocculus of the cerebellum.

a. Central vestibular neurons that project to the extraocular motor nuclei (Gaze Stability) receive a majority of their monosynaptic inputs from regular afferents

b. Central vestibular neurons that project to the spinal cord (Postural Stability) receive a majority of their inputs from irregular afferents

c. Central vestibular neurons projecting to the flocculus of the cerebellum receive relatively mutual contributions from regular and irregular afferents.

VESTIBULAR PATHWAYS FOR POSTURAL CONTROL OF THE HEAD AND BODY (VSR)

Descending tracts:
Medial vestibulospinal tract travels bilaterally via the medial longitudinal fasciculus to the cervical cord:
- head movements and integrating head and eye movements

Lateral vestibulospinal tract travels to the thoracic spinal cord:
- head and body position in space, for walking upright
Secondary neurons also have extensive connections with the reticular formation, thalamus, parietal and insular lobes (PIVC)…….

- These connections contribute to the integration of arousal and conscious awareness of the body and to discriminate between movement of self and the environment.

Central Vestibular Anatomy

CEREBELLUM

Compares intended to actual movement

- Compares info from cortex to peripheral sensory (GTO, proprioception, vestibular apparatus, eyes, ears)
- Error correcting mechanism
- Critical for motor skill adaptation

CENTRAL VESTIBULAR ANATOMY – FUNCTION OF THE CEREBELLUM

Secondary VOR neurons also have extensive connections with the cerebellum.

- Cerebellum has inhibitory influence on vestibular nuclei
- Cerebellum maintains calibration of the VOR, contributes to posture during static and dynamic activities, and influences the coordination of limb movements

In damage, the VOR becomes miscalibrated

Flocculus: adjusts and maintains gain of VOR (Chiari malformation; cerebellar degeneration)
Nodulus: adjusts duration of VOR responses and mediates otolith input (medulloblastoma)
Vermis: responds to vestibular stimulation

DIFFERENTIAL DIAGNOSIS
PERIPHERAL V. CENTRAL DYSFUNCTION – THE OCULOMOTOR EXAM

OCULOMOTOR CONTROL
6 neuronal control systems that provide two main roles in oculomotor control:
1. Hold Image on the Retina via
   • Visual Fixation
   • Vestibular Ocular Reflex
   • Optokinetic Reflex
2. Change the Angle of Gaze via
   • Smooth Pursuit – Hold images of a moving target on the retina
   • Saccades – Rapid conjugate movements of the eyes to place the object of interest on the fovea
   • Vergence – Adjust the eyes for different viewing distances

HOLD IMAGE ON RETINA AND CHANGE ANGLE OF GAZE
• Moving the eye requires overcoming the viscous drag imposed by supporting structures/tissues.
  • Occurs via a powerful contraction of the extraocular muscles via a burst of activity in the ocular motor nuclei.
  • Visco-elastic properties of the eye want to restore the eyeball to its resting position in midline
• Maintaining the eye in the eccentric position requires a tonic level of neural activity to achieve a steady contraction of the extraocular muscles
Saccade Generation
Oculomotor Neurons Generate a Pulse-Step

**Pulse of Innervation - Velocity Command**
- burst cells generate a burst of neural activity required to overcome viscous drag and initiate eye movement

**Step of Innervation – Position Command**
- tonic level of neural activity required to maintain the eye in the eccentric position

---

**NEURAL INTEGRATOR - NI**
The NI translates velocity (pulse) signals into position (step) signals
Involves a network of interconnected neurons

- **Horizontal**
  - Nucleus prepositus hypoglossi (medulla)
  - Medial vestibular nucleus (medulla)
  - Vestibulo-cerebellum

- **Vertical**
  - Interstitial nucleus of Cajal (midbrain)
  - Vestibulo-cerebellum

Lesions in the pons typically affect the generation and holding of horizontal saccades
Lesions in the midbrain typically affect vertical saccades

---

**MORE COMMON SIGNS/SYMPTOMS IN CVD**

A. Downbeat nystagmus and the role of the VOR to help distinguish central from peripheral

B. Abnormal Subjective Visual Vertical (SVV)
  - Bucket test

C. Lateropulsion

D. Ocular Tilt Reaction (OTR)

E. Oculomotor abnormalities/HINTS

F. Ocular misalignment/HINTS
A. NYSTAGMUS (VERTICAL) IN CENTRAL DISORDERS

- Spontaneous nystagmus in central vestibular disorder (CVD) is commonly due to cerebellar mal-position, degeneration (Brandt and Dieterich 1995), or ischemia (Wagner 2007).
- Downbeat nystagmus (DBN) most common type of nystagmus in patients with CVD.
  - Chiari malformation (mal-position)
  - Bilateral brainstem lesions (where pathways cross midline [pons] between the vestibular nuclei and floor of 4th ventricle), bilateral floccular lesions, (Brandt and Dieterich 1995) (Buttner 1995), or lesions of the vestibulocerebellum (Wagner 2007).
- Upbeat nystagmus (UBN) more common with brainstem lesions (bilateral lesions of the vertical VOR pathway in the medulla, midbrain).
  - Also possible with anterior cerebellar vermis lesions (Lee 2009; Buttner 1995)

B. SUBJECTIVE VISUAL VERTICAL

Quantified test that determines the individuals’ perception of vertical:
- Pathologic SVV is the most sensitive sign of vestibular tone imbalance in the roll plane (Dieterich and Brandt 1993).
- It can result from lesions of central and peripheral vestibular pathways.
- SVV tilt is observed in 94% of patients with acute unilateral brainstem lesions that affect central graviceptive pathways. This figure considerably exceeds the diagnostic sensitivity of current brain imaging devices (Zwergal et al 2008; Dieterich and Brandt 1993).

Procedure:
- Patients sit upright looking into a translucent plastic bucket so that the bucket rims prevent any gravitational orientation clues.
- On the bottom inside the bucket, there is a dark, straight, diametric line. The examiner rotates the bucket clockwise or counterclockwise to an end position and then slowly rotates it back toward the zero degree position. Patients indicate stop at the position where they estimate the inside bottom line to be truly vertical. The examiner reads off the degrees on the outside scale.
- Ten repetitions have to be performed. Monocular or binocular testing is valid. In a group of 30 healthy subjects, the range of absolute deviations of binocular subjective visual vertical from true vertical was 1.1 ± 1.98.

SVV TEST – BUCKET TEST
C. LATEROPULSION

Tendency to fall or lean sideways
- Direction of lateropulsion depends on site and extent of lesion (same as SVV)
- Lateropulsion rarely the only presenting sign (Thomke 2005).
- 66% of 134 isolated pons CVA had lateropulsion as their main presenting symptom, while 133 had contraversive tilting of the SVV (Yi 2006)

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Moderate head &amp; body tilt w/o considerable imbalance</td>
</tr>
<tr>
<td>II</td>
<td>Head &amp; body tilt w/ considerable imbalance, no falls</td>
</tr>
<tr>
<td>III</td>
<td>Head &amp; body tilt and falls w/ eyes closed</td>
</tr>
<tr>
<td>IV</td>
<td>Head &amp; body tilt and falls w/ eyes open</td>
</tr>
</tbody>
</table>

D. PATHOLOGIC OCULAR TILT REACTION (OTR)

Triad of Signs
- Lateral head tilt
- Skew deviation with depression of 1 eye and elevation of opposite eye
- Torsional rotation of the eyes (superior pole) towards the down ear

Central cause - persists (often)
Peripheral cause - recovers (often)

E. OTHER OCULOMOTOR ABNORMALITIES

- Saccadic abnormalities
  - Ipsipulsive saccades
    - Do vertical saccades ‘drift’ to one side?
  - Dysmetric saccades
- Impaired VOR Cancellation
  - Saccadic?
  - Direction changing nystagmus
OTHER OCULOMOTOR ABNORMALITIES

- Saccadic abnormalities
- Impaired VOR Cancellation
- Direction changing nystagmus
- Down beating nystagmus
- Saccadic smooth pursuit

DIFFERENTIAL DX
CENTRAL VS. PERIPHERAL CAUSES OF ‘VESTIBULAR-LIKE’ SYMPTOMS?

Dizziness in the Emergency Room

9472 cases surveyed

- Otologic/vestibular (32.9%)
- Cardiovascular (21.1%)
- Respiratory (11.5%)
- Neurologic (11.2%, including 4% cerebrovascular)
- Metabolic (11.0%)
- Injury/poisoning (10.6%)
- Psychiatric (7.2%)
- Digestive (7.0%)
- Genitourinary (5.1%)
- Infectious (2.9%)
Overview of Differences between Central vs. Peripheral Vestibular Pathology

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Nystagmus*</th>
<th>Vertigo*</th>
<th>Other Symptoms</th>
<th>Other Signs</th>
<th>Fixation- Suppressed Nystagmus?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central</td>
<td>Pure vertical (often DBN*)</td>
<td>Less Common</td>
<td>Diplopia (other “D”s), Lateropulsion, Aural Fullness, Infrequent Hearing Loss (except AICA)</td>
<td>Abnormal pursuit or saccades, Persistent ocular tilt reaction</td>
<td>No</td>
</tr>
<tr>
<td>Peripheral</td>
<td>Mixed horizontal and torsional</td>
<td>More Common</td>
<td>Hearing loss, fullness in ears, tinnitus</td>
<td>Positive HIT; pursuit and saccades usually normal</td>
<td>Yes</td>
</tr>
</tbody>
</table>

* Central DBN vs SCC BPP

---

**Central vs Peripheral – Pseudo vestibular neuritis**

*Use the Head Impulse Test (HIT)*

- The cerebellar infarction territory most commonly involved was the PICA territory (24/25: 96%).

- 10% acute cerebellar CVA had abnormal head impulse test but these patients had other abnormal ‘central’ findings i.e. headache, truncal instability (Newman-Toker 2008).

- Conclusion: Patients with stroke should be evaluated with the head impulse test – HIT (Savitz 2007)

---

**HINTS:**

**HEAD IMPULSE NYSTAGMUS TEST OF SKEW**

3 step bedside examination superior to MRI within 1st 72 hours of lesion

Stroke suspected if any of the following exist:

- Normal head impulse test
- Direction changing nystagmus in eccentric gaze
- Skew deviation (vertical ocular misalignment)

In cases where there was a false positive on the head impulse, the coexisting presence of skew deviation correctly identified all those w/ stroke

The presence of normal horizontal head impulse test, direction-changing nystagmus in eccentric gaze, or skew deviation (vertical ocular misalignment) was 100% sensitive and 96% specific for stroke.

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Kattah 2009
Another acronym to distinguish Central from Peripheral vestibular pathology: STANDING

Bedside Algorithm for isolated Vertigo (Vanni et al 2015)
Used as a Diff Dx for patients presenting to the ED with complaints of isolated vertigo

- SponTaneous Nystagmus – present?
- Direction - if SN is present, what direction?
- Head Impulse test – normal or not?
- Standing – ataxic, unable?

Vascular supply and AICA versus PICA CVA

- The two most common vascular causes of acute vertigo are the AICA and PICA CVA.
- Vertebral artery supplies the posterior inferior cerebellar artery (PICA)
  - PICA supplies inferior cerebellar hemispheres and dorsolateral medulla
- Basilar artery supplies the Pons and the anterior inferior cerebellar artery (AICA)
  - AICA supplies peripheral vestibular labyrinth via the labyrinthine artery, the ventrolateral cerebellum as well as the lower 2/3 pons including the rostral 1/3 of vestibular nuclei.
- The major difference between the two is the AICA CVA may have auditory signs/symptoms from cochlear injury (92%).

PICA INFARCT

- PICA is the largest branch of the vertebral artery
  - Supplies the dorsal lateral medulla and portions of the cerebellum (uvula, nodulus and paraflocculus).
- When the PICA is occluded at its origin (vertebral artery), Wallenberg's results
- Not all PICA infarcts cause a Wallenberg syndrome
- Kim et al report 79% PICA infarcts had abnormal SVV, 100% had lateropulsion (2009)
WALLENBERG SYNDROME

Typical Signs:
• Horner syndrome (ipsilateral ptosis, miosis, facial anhidrosis)
• Ipsilateral OTR, SVV, and Lateropulsion
• Vertical saccades may deviate toward the side of the lesion
• Spontaneous nystagmus often similar with peripheral vestibular disorder
• Ocular lateropulsion is pathognomonic for PICA infarct causing a dorsal lateral medullary lesion
  • Spontaneous nystagmus with quick phase beating towards the lesioned side
• Dysphagia

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OCULAR LATEROPULSION

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Kathleen B. Digre MD
http://medstat.med.utah.edu/

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Treatment for CVD

Exercise Guidelines and Suggestions

• Incorporate exam findings: Focus on challenging balance exercises and reducing symptoms
• Sort through which symptoms are treatable. Fearful individuals more likely to be depressed (Kressig 2001).
• For mTBI and CVD in general, gentle habituation exercises often an excellent starting point (Shepard 95, MSQ)
  – Usually best to start with lower intensity exercises and fewer repetitions
  – Progression using habituation is slower, but effective (Shepard 95)
  – DVA and the DHI found to be reliable outcome measures in evaluating the progress of patients with balance disorders associated with TBI (Gottshall et al 2003).

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Treatment for CVD
Inappropriate Use of Rehabilitation

- TIA’s may present with sudden vertigo or complaints of hearing loss that lasts minutes.
- Orthostatic hypotension (≥ 20 mm Hg systolic drop supine to stand) as a cause of dizziness
- Pre-syncopal lightheadedness may be from poor blood perfusion in the brain
- Vomiting as the sole presenting symptom
- Pure vertical nystagmus
  - Velocity may be reduced/eliminated with 4 amino-pyridine (Strupp 2009)

Treatment Goals in Central Vestibular Pathology

- Reduce dizziness, start slowly
- Appropriate use of Ankle and Hip strategies
- Establish an independent home exercise program
  - Walking program
- Improve interaction among visual – vestibular – somatosensory inputs

Expectations for Recovery

- Generally, prognosis is not as good the recovery course is longer
- Lesions of compensatory pathways
- Lesions of other sensory and motor systems
  - May be more difficult to coordinate complex balance tasks
  - Multi-step exercises may be too difficult
Central Oculomotor Lab
Ocular Misalignment - Symptoms

If Severe:
• Diplopia - sensation of seeing an object at 2 different locations in space
  • determine if the diplopia is horizontal or vertical
  • determine if the diplopia is binocular or monocular. Monocular diplopia very rare.
  • Head tilt will be present (vertical misalignment)

If Subtle:
• Difficulty maintaining focus
• Ocular soreness
• Headaches
• Mental dullness

CENTRAL OCULOMOTOR LAB
STRABISMUS
Misalignment of visual axes;
• Exo – outward (laterally)
• Eso – inward (medially)
• Hyper – upward
• Hypo – downward
• Excyclo – torsional deviation, upper pole templeward
• Incyclo – torsional deviation, upper pole nasaward

Ocular Misalignment/Strabismus
Always named by the resting position

A. Tropia:
• Deviation of visual axes during binocular viewing of a single target
• Manifest (static) deviation: present in all circumstances
  • fusional vergence cannot correct (i.e., focusing on target)
  • deviation is readily observable

B. Phoria: Always named by the resting position
• Deviation of visual axes during monocular viewing of a single target
• Latent deviation: deviation is not always apparent
  • With fatigue, or when fusion is broken, a phoria can be observed
  • Need to break fusion of eyes to test: eg: Maddox Rod, Alternate Cover Tests
Assessing Ocular Alignment

**Cover Uncover Test**
- While focusing on target, one eye is covered
- Look for "movement of redress" of uncovered eye
- Identifies tropia of uncovered eye (eso/exo/hyper/hypo)

**Cover Cross Cover/Alternate Cover Tests**
- Observe for movement of occluded eye once uncovered
- In practice, cover and uncover tests are done together ("Cover/Uncover" Test)
- Identifies phoria, if the Cover test is negative

---

**Cover/Uncover: Tropia**

- Illustration of tropia:
  - Left exotropia
  - Adducts to focus
  - Returns to preferred resting position

---

**Cover/Uncover: Phoria**

- Illustrated is an exophoria:
  - Moves to preferred resting position
  - Returns to regain fixation on target
Cover/Uncover Tests

Alternate (Cross) Cover Test
- Occluder quickly transferred from eye to eye
- Prevents binocular viewing
- Do multiple times – you may see the deviation increase over time

CONVERGENCE SPASM

Clinical Presentation
- Spontaneous convergence
- Pupillary constriction (miosis)
- Inability to abduct eye(s) with gaze testing

Clinical testing
- Usually patient describes dizziness associated with spasm
- Can be positional induced (with Dix Hallpike, etc)
- Can appear like bilateral cranial nerve VI palsy – perform gaze testing monocular to differentiate (Furman 2010, Knapp 2002, Chan 2002)
CONVERGENCE SPASM - ETIOLOGY

- Head Trauma
- Functional (hysterical)
- Thalamic hemorrhage
- Pineal tumor
- Encephalitis
- Wernicke-Korsakoff syndrome
- Verteobasilar insufficiency
- Chiari malformation
- Multiple Sclerosis
- Metabolic encephalopathy
- Phenytoin intoxication
- Idiopathic

SYMPTOMS OF VERGENCE DEFICITS:

- Trouble reading, focusing
- Trouble focusing from far to near (eg.: taking notes in class)
- Blurred vision
- Headaches
- Eye strain
- Sensitivity to light (w/ excessive vergence response)
- Pulling sensation around eyes
- Avoidance of reading
- Comprehension deficits over time

Treatment of Convergence Spasm

For severe cases:
- Neuro-ophthalmology consult warranted
- Cycloplegic drops
- Botox
- Lens corrections/prism
- Convergence exercises?
- It is not known if convergence exercises can improve convergence spasm
Rehab Treatment of Convergence Insufficiency & Binocular Vision Abnormalities

Example Exercises:
- Brock’s string
- Pencil pushups
- Dot card
- Popsicle sticks/Two Targets
- Useful website: http://www.eyecanlearn.com/

Clinical Vergence Testing

WHAT TO DO:
Ask patient to fixate on target that is brought SLOWLY towards the bridge of the nose
- Normal diplopia no > than 5 cm from tip of nose
  (Scheiman 2003)
- Test at least 3X – to assess fatigue of system

Brock string convergence vision therapy. Often used in treating convergence insufficiency.

Dot card (similar to Barrel card) used for vision therapy for convergence insufficiency.
**Brock String**

**Materials:** Two targets

**Procedure:**
- Hold staggered targets in front of patient at eye level
- Have patient focus eyes on front target
- Shift gaze to second, rear target
- Repeat x 30 - 60 seconds or to symptom tolerance (Issue as HEP)

*Be careful not to over stimulate pt – symptoms often accumulate throughout the day*

**PRACTICE**

- **Cover/Uncover Test**
- **Alternate Cover Test**
- **Vergence Exercises**
TESTS PERFORMED IN ROOM LIGHT

NOTE: Many of these tests in room light can be combined

1. **Spontaneous nystagmus**: If patient unable to keep head still, hold the patient's head with one hand (on top or under the patient’s chin), have the patient look straight ahead and observe for nystagmus (slow phase/fast phase) or any abnormal eye movement.

2. **Gaze-holding nystagmus**: Continue to hold the patient's head stationary. Using a point target (e.g. the end of your finger pointed at the patient or the end of a pen pointed at the patient), have the patient follow your finger while looking 20 - 30 degrees to the **right, left, center, then up and down**.

   Pause in each of those positions to observe for nystagmus. Note the direction of the nystagmus in each position. Be sure to keep your finger 18 - 24 inches away from the patient's face throughout the entire test.

3. **Eye Movement range**: Move your finger out further to examine if the patient has full ocular range of motion. Ask the patient to follow a moving object (the end of your finger pointed at the patient) that is held 18 – 24 inches in front of the patient's face (to avoid convergence of eyes). It is usually sufficient to test for full vertical and full horizontal eye movements however, some people do an “X” or “box” combination and that is also acceptable. You are check if the eyes move conjugately. You can ask the patient if there is double vision at the end range. If the patient indicates that there is, you need to determine if it is horizontal or vertical diplopia e.g. "do you see two fingers side by side or two fingers, one above the other?

   Note #1: A small amount of “end-point” nystagmus may be observed at the point of full ocular range in all directions. This should be minimal in younger individuals but becomes more obvious with increased age e.g. > 65 years.

4. **Vergence**: 1 of two methods:
   a. Hold the patient's forefinger in your hand about 2 feet away from the patient's face.
   b. Provide a discrete target ~ 2 feet away
Next, Ask the patient to focus on the finger/target while you move it toward the bridge of the patient's nose. Ask the patient to tell you when the target becomes double. The eyes should converge and the pupils should constrict. You must verbally encourage the person to “stay on the target” if they have difficulty.

Normal near point of convergence is any distance < 10 cm (approximately 4 inches). If the distance at which the patient sees double or one eye jumps out of alignment is > 10 cm, vergence is considered to be abnormal.

5. Smooth pursuit eye movements: Note: in the above test for ocular range of motion, you can observe if the person’s eyes are smoothly following your moving finger or not. Just be sure you are not moving your finger too fast. If the eye movement appears to be saccadic, slow the speed of the target (finger) movement down to see if the person can follow smoothly.

Hold the patient's head stationary, if needed (hand placed on top of the patient's head or under the patient’s chin), and have the patient follow your slowly moving finger horizontally (from center to 30 degrees right and then to 30 degrees left), and then come back to center and test for vertical smooth pursuit (center to 30 degrees up to 30 degrees down). The test can be repeated; you may have to hold the eyelids up in order to see the downward eye movement clearly.

You are looking for a smooth conjugate eye movement. The key is to move your finger at the correct speed (~ 20 degrees/second). If you move your finger too fast, the eye movement will become saccadic (jerky). Also, do not test more than 30 degrees R/L/U/D.

Abnormal smooth pursuit would be jerky (saccadic) eye movements that are not the tester's fault. Note the direction of pursuit when it occurred. It is important to realize that as we get older, the peak velocity of smooth pursuit eye movement decreases and pursuit becomes more saccadic. Additionally, vertical eye pursuit is often interrupted by a saccade even in younger individuals.

Note: A small jump in the eye movement as the eyes cross the midline is OK

Note: One can combine the above tests into one test by pausing at the appropriate positions to check for spontaneous and gaze holding nystagmus, and by observing the eye movements as the patient follows the target from one point
to another. If the eye movements appear to be saccadic, slow down the target movement to see if the person can track smoothly. You may need to repeat the smooth pursuit eye movements several times to get an accurate assessment of smooth pursuit.

6. **Saccadic eye movements**: Continue to hold the patient's head stationary. Hold your finger tip about 15 degrees to one side of your nose. Ask the patient to look at your nose, then at your finger, repeating several times. Do this from the center to right to center, center to left to center, center to up to center and center to down to center.

   You are looking for the number of eye movements it takes for the patient's eyes to reach the target so you have to get the person to do it when you tell them. Normal is < 2. You may have to ask the patient specifically to make only one eye movement.

7. **VOR Cancellation**: Grasp the patient's head firmly with both hands on the side of their head. Instruct the patient to look at your nose. Slowly move the patient's head from side to side approximately 30° while you move in the same direction that you move the patient's head with your face remaining directly in front of the patient's face. Observe if the patient can maintain visual fixation and/or if the patient makes saccadic eye movements.

8. **Head Impulse Test**: Grasp the patient's head firmly with both hands on the side of their head. Instruct the patient to look at your nose. Move the patient's head slowly back and forth being to ensure the patient is relaxed. **It is imperative that the patient be informed that you will be moving their head quickly, but only through a small range.** They should be instructed to relax and not to blink. If you noted that the patient had pain or significant restriction in cervical spine mobility, this test should be performed with extreme caution or should be deferred.

   When you move the patient’s head in one direction, the head movement should be through a small amplitude with the position held at the end. Don't immediately bring the person's head back the other direction. Observe for the patient's ability to maintain visual fixation. You should note if the patient makes corrective saccades to re-fixate your nose and the direction of head movement that caused the re-fixation saccades. You should consider repeating this procedure with the patient visually fixating a distant target if you find that they make corrective saccades with near target fixation.
Note: If you are uncomfortable moving the person’s head from center to an eccentric position, try moving the person’s head from an eccentric position to center. The test is a bit more predictable that way but may be easier to perform.

9. HORIZONTAL HIT
10. **VERTICAL HIT – Two methods:**
Assessing Ocular Alignment:

**Cover Uncover Test**
- While focusing on target, one eye is covered
- Look for “movement of redress” of uncovered eye
- Identifies tropia of uncovered eye (eso/exo/hyper/hypo)

**Cover Cross Cover/Alternate Cover Tests**
- Observe for movement of occluded eye once uncovered
- In practice, cover and uncover tests are done together (“Cover/Uncover” Test)
- Identifies phoria, if the Cover test is negative

**Cover/Uncover: Tropia**

![Diagram showing left exotropia, adducting to focus, and returning to preferred resting position](image)
Cover/Uncover: Phoria

Illustrated is an exophoria

Eye moved out when covered

Eye moved back when uncovered

Moves to preferred resting position

Returns to regain fixation on target

R L

Brock string convergence vision therapy. Often used in treating convergence insufficiency.

Dot card (similar to Barrel card) used for vision therapy for convergence insufficiency.
11. **Optokinetic Nystagmus:** If you have access to an optokinetic drum, have the patient follow the striped lines with their eyes while you slowly move the drum in one direction. Repeat this procedure rotating the drum in the opposite direction. You should observe for optokinetic nystagmus (slow phase eye movements in the direction of drum rotation). Be careful to not rotate the drum too quickly. You should note if the patient does not produce slow phase eye movements or if slow phase eye movements are saccadic in nature. Additionally, you should note the direction of drum movement in which this occurs.

You can use striped fabric as well, for OKN testing.

12. **Static and Dynamic Visual Acuity:** Have the patient wear their glasses if they need distance correction. Depending on the type of acuity chart being utilized, have the patient sit, or stand, the appropriate distance from the chart. (The ETDRS charts are designed to be viewed from a distance of either 2 or 4 meters and provide Snellen equivalent acuity ratios or LogMAR values as noted on the chart). Have the patient read to the lowest line that they can until they cannot correctly identify all the letters on a given line. Note the line where this occurs and/or the number of optotypes the patient incorrectly identifies.

Now, standing behind the patient, grasp the patient's head firmly with both hands.
on the side of their head and move their head side to side at a frequency of 2Hz (2 complete side to side cycles per second). Ask the patient to read to the lowest line that they can until they are unable to correctly identify all the letters on a given line. Note the line where this occurs and/or the number of optotypes the patient incorrectly identifies.

Keep the range of motion of the head movements small so as to not restrict the visual field, which may occur with patients who wear glasses.

“DVA” is the decrement in visual acuity that occurs with head movement so it is the difference between visual acuity with the head stationary and visual acuity with the head moving. There are no normal values by age for this clinical test. DVA is considered to be abnormal if acuity degrades by three or more lines.

TESTS PERFORMED WITH FIXATION BLOCKED (FRENZEL LENSES, IR SYSTEM)

13. **Spontaneous Nystagmus:** Holding the patient's head stationary with one hand placed on top of the patient's head, have the patient look straight ahead and observe for nystagmus.

14. **Gaze-holding Nystagmus:** Continue to hold the patient's head stationary. Have the patient move their eyes so that they look 30° to the right, left, and up pausing briefly at the endpoint to observe for nystagmus noting the direction of the fast phases. NOTE: patients tend to look all the way to the right or left when you ask them to do this rather than within the 20° or 30° within which gaze-holding nystagmus should be tested so you will need to correct them if you are observing nystagmus at end range.

15. **Head-Shaking Nystagmus:** It is important that you inform the patient that you will be moving their head side to side and to clear them for any cervical spine problems prior to performing this test. It is also important to maintain the patient's head position at the end of the head shaking while observing for nystagmus. Grasp the patient's head firmly with both hands on the side of their head. Note the direction of the nystagmus. One or two beats is not considered significant.

If there is persistent nystagmus induced by horizontal head shaking, this procedure should be repeated vertically but only moving the patient's head ten times. Again, observe for nystagmus noting the direction of the quick phases. In
the case of unilateral peripheral vestibular hypofunction, there may be no nystagmus induced by vertical head-shaking or the nystagmus may be horizontal but in the direction opposite to that induced by horizontal head shaking.

We recently showed that tilting the head after 1st identifying an increase in nystagmus should suppress the nystagmus in peripheral vestibular pathology (Maia et al 2017)

16. **Pressure-Induced Nystagmus:** Observe the patient’s eyes for nystagmus or drift when
   a. you occlude the external auditory canal by applying pressure to the tragus or
   b. they perform a valsalva maneuver (Valsalva-induced nystagmus) or
   c. they attempt to blow air out through closed nostrils

17. **Hyperventilation Induced Nystagmus:** Have the patient hyperventilate for 60 seconds (one breath per second). As the patient nears the minute point, place the Frenzel lenses or infrared goggle system on the patient’s face and observe for nystagmus that was not present before beginning the test, or is a reversal of the pre-hyperventilation nystagmus. If this test is positive, it raises the possibility of demyelinating process (such as an acoustic neuroma) along the 8th cranial nerve.
BPPV of the pSCC and aSCC

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Benign Paroxysmal Positional Vertigo

- Mechanical disorder of the inner ear caused by abnormal stimulation of 1 or more of the 3 semicircular canals within the ear.
- When eye movements have been recorded with video-oculography, the distribution of BPPV suggests:
  - 41-65% unilateral PC-BPPV
  - 20% multi-canal BPPV
  - 21-33% LC-BPPV
  - 17% AC – BPPV


Semicircular Canals – Crista Ampullaris and Cupula

The fluid filled canals normally act to detect rotation of the head.

Rotation of the head causes deflection of the sensory hair cells located within the crista ampullaris.

The hair cells are embedded within a gelatinous membrane – the cupula.
Activation of the pSCC Sensory Afferent

In the vertical canals, flow of endolymp away from the ampulla excites the nerve while flow towards the ampulla inhibits the nerve.

Maculae are Weighted Sensory Membranes

Otoconia weights the sensory membrane. The weighted sensory membrane acts to detect gravitational forces on the head.

Cupulolithiasis – Schuknecht, 1969

• 1st theory to describe BPPV involved cupulolithiasis - dislodged otoconia may directly attach to the cupula weighting the membrane.

• Reorientation of the canal relative to gravity deflects the cupula causing activation of the sensory organ.
**Cupulolithiasis – Nystagmus**

- Mathematical models incorporating the fluid dynamics of BPPV support the theory of cupulolithiasis.
- No latency before onset of nystagmus because of weight of the particles on the pSCC cupula (Rajguru et al., 2004).
- Maintained activation of pSCC when the orientation of the head is changed relative to gravity (Rajguru et al., 2004).
- The effect builds up gradually over time (nystagmus in theory would crescendo) (Hain, et. al., 2005).
- Any reduction in afferent input would be the result of adaptation by the hair cell afferent complexes (Rajguru et al., 2004) – but this is incomplete/debatable as the cupula is still deflected yet the nystagmus persists.
- Compared with canalithiasis, similar amounts of adherent otoconia only produce 1/3 as much nystagmus (Rajguru et al., 2004; Hain, et. al., 2005).

**Canalithiasis**

- 2nd theory of BPPV was postulated by Hall et al in 1979, stating free-floating otoconia debris settle within the long arm of the canal.
- Reorientation of the canal relative to gravity causes otoconia (arrow) to move to the lowest part of the canals.
- Movement of the otoconia creates a drag on the endolymph.
- Drag results in fluid pressure on the cupula activating the primary afferent.

**Comparison of Human Canal Planes in Reid Stereotaxic Coordinate System**

- Reid horizontal plane defined by the center of the external auditory canal and the inferior margin of the bony orbits.
  - PC is placed at 56° from the sagittal plane
  - AC is placed at 41° from the sagittal plane

Afferents from pSCC Project to Oculomotor Nuclei

- Excitation of CN VIII activates:
  - Ipsilateral superior oblique
  - Contralateral inferior rectus

- Inhibition of CN VIII activates:
  - Ipsilateral inferior oblique
  - Contralateral superior rectus

- Therefore stimulation of the pSCC results in upbeating and torsional nystagmus (torsion towards the affected ear).

Afferents from aSCC Project to CN Nuclei

- Excitation of CN VIII activates:
  - Ipsilateral Superior Rectus
  - Contralateral Inferior Oblique

- Inhibition of CN VIII activates:
  - Ipsilateral Inferior Rectus
  - Contralateral Superior Oblique

- Therefore stimulation of aSCC results in downbeating and torsional nystagmus (torsion towards the affected ear)

Positional Tests

- Positive findings on positional tests confirm the diagnosis suspected from history and identifies the canal(s) involved. Positional tests place the plane of the canal being tested into the plane of gravity.
Dix-Hallpike Test (DHT)

- DHT is the standard from which the diagnosis of PC-BPPV is made (Bhattacharyya et al., 2008; Fife et al., 2008).
- Performed with the patient wearing Frenzel goggles or using video-oculography to prevent visual suppression of the nystagmus.
- Sensitive to detecting PC and AC BPPV.
  - Series of positions. Each maintained for 45 seconds.
  - To identify the canal involved, note direction of nystagmus.

Vertical Canal Tests – Dix Hallpike and side lie Dix Hallpike

Which semicircular has otoconia in it?

Plunger effect of otoconia
In the vertical canals, torsion during excitation identifies the affected side

Don’t forget to check for Nystagmus when return to sit

Diagnostic Criteria for pSCC BPPV

- Posterior Semicircular Canal Nystagmus
  - Vector of nystagmus primarily upward and torsional (towards the lowermost ear) (Aw et al., 2005).
  - Characteristics of Nystagmus
    - 1- to 40-second latency before the onset of vertigo and nystagmus (Brandt et al., 1980; Epley, 1980; Herdman, 1990).
    - Vertigo and nystagmus < 60 seconds in duration (Balogh et al., 1993).
    - Fatigues with repeated positioning (Balogh et al., 1993).
Video of pSCC CRT – which ear is being treated?

Liberatory or Semont Maneuver for pSCC BPPV (Semont, Freyss et al. 1988)

• Designed to treat cupulolithiasis or canalithiasis
• The linear acceleration at the peak of the swing acts in the opposite direction to gravity (Faldon and Bronstein, 2008).
• Reversed nystagmus – 2nd position flow of endolymph away from ampulla and 3rd position flow of endolymph towards the ampulla resulting in a reversed direction of nystagmus.
• The duration of the 180 degree whole-body swing needs to be less than 1.5 seconds (Faldon and Bronstein, 2008).
Liberatory or Semont Maneuver for pSCC BPPV

Illustrated for treatment of the left PC (Radtke et al., 2004)

Semont which ear is being treated?

Diagnostic Criteria for aSCC-BPPV

- Vector of Nystagmus
  - Primarily downward directed
  - Small torsional component or no torsional component (50% of patients). If torsional component, may be either
    - Torsional towards the lowermost or uppermost ear depending on affected side.
- In both the head right and head left position, the open end of the ampullary segment points downwards at about 40° off vertical (Bertholon et al. 2002).
- Therefore, positive findings may be evoked in both the head right and head left positions of the DHT.

The straight head hanging position (30° of extension) of the DHT adds an extra 20° of cervical extension (Bertholon et al. 2002).

This enables the otoconia to clear the curvature of the long arm of the aSCC.

Characteristics of aSCC BPPV

Nystagmus (Bertholon et al., 2002)

• 1- to 5-second latency before the onset of vertigo and nystagmus.
• Vertigo and nystagmus < 60 seconds in duration.
• Fatigues with repeated positioning.

Video of Deep Head Hang for aSCC BPPV
What about vibration of the Mastoid?

• Vibration in theory prevents debris from ‘marginating’/collecting along the canal wall.
• Vibration has **NOT been shown** to improve the short or long-term outcome of BPPV (Hain et al., 2000; Macias et al., 2004; Motamed et al., 2004).
• You might consider its use for recalcitrant cases of BPPV
  • Kim et al suggest it is useful in combination with a repositioning maneuver for hSCC BPPV (2017)

(2010 data) Comparison of CRP versus Liberatory Maneuver

<table>
<thead>
<tr>
<th>Study/Intervention Groups</th>
<th>Number resolved/ Number in study</th>
<th>Percent Resolve</th>
<th>Odds Ratio - Confidence Intervals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Massoud et al, 1996**</td>
<td>CRP 43/46 LM 46/50</td>
<td>93/92</td>
<td>0.80(0.17-3.79)</td>
</tr>
<tr>
<td>Soto Varela et al, 2001*</td>
<td>CRP 30/42 LM 26/35</td>
<td>71/74</td>
<td>1.16(0.42-3.18)</td>
</tr>
<tr>
<td>Radtke et al., 2004**</td>
<td>Self-admin. CRP 35/37 LM 19/31</td>
<td>95/58</td>
<td>0.08(0.02-0.38)</td>
</tr>
<tr>
<td>Tanimoto et al., 2005*</td>
<td>CRP only 28/39 CRP+Self-admin.CRP 36/40</td>
<td>72/90</td>
<td>3.54(1.02-12.30)</td>
</tr>
</tbody>
</table>

* Standard treatment CRP.
** Standard treatment self-administered CRP.

Self Administered CRP (Radtke et al., 1999)

- Head extended over edge of pillow (Position 2 and 3).
- When roll onto side, rest head in lower hand keeping neck slightly side bent away from the supporting surface to prevent canal conversion to AC (Rajguru et al., 2004).
- Hold each position for a minimum of 30 seconds to allow debris to settle (Hain et al., 2004).
- 3 cycles of exercise 3 times per day.
- Stop exercises when symptom-free with routine and exercises for 2 consecutive days.
Self Administered Liberatory Maneuver

- Self treatment
  - 3 cycles of exercise 3 times per day.
  - Stop exercises symptom-free with routine and exercises for 2 consecutive days.

Illustrated for treatment of the left PC (Radtke et al., 2004)

Number of Cycles Required within a Treatment Session to Optimize Outcome of CRP

Repeating the CRP more than once is significantly more effective than performing the CRP once within a treatment session. The success rate of the group receiving more than 1 cycle of the CRP within a session was 21.4% higher than the group receiving 1 cycle (Korn, Dorigueto et al., 2007).

<table>
<thead>
<tr>
<th># of Sessions</th>
<th>Group I (1 Cycle)</th>
<th>Group II (4 Cycles)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>51 (68%)</td>
<td>42 (89%)</td>
<td>0.039</td>
</tr>
<tr>
<td>2</td>
<td>17 (23%)</td>
<td>3 (6%)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>3 (4%)</td>
<td>2 (4%)</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2 (3%)</td>
<td>0 (0%)</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>2 (3%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Comparison of the Epley, Semont, and sham maneuvers for the treatment of pSCC BPPV (Lee et al. Audiol Neurootol. 2014)

- Epley (36 patients); Semont (32 patients); Sham (Epley maneuver for the unaffected side, 31 patients)
- N-14 institutes
- Each maneuver was repeated twice
- After the first maneuver, the Epley group showed a significantly higher resolution rate of positional nystagmus than the Semont or sham groups (63.9, 37.5, and 38.7%, respectively).
- After the second maneuver, the resolution rate (83.3%) of the Epley group was significantly higher than that (51.6%) of the sham group.
- Conclusion: The Epley maneuver was significantly more effective per maneuver than Semont or sham maneuvers for the short-term treatment of posterior canal BPPV.

- Epley and Semont maneuvers for posterior canal benign paroxysmal positional vertigo: A network meta-analysis.
- Meta-analysis: RCT that used an Epley or Semont maneuver in posterior canal benign paroxysmal positional vertigo patients were analyzed in this project. Efficacy outcomes included 1-week recovery rate and end of study recovery rate.
- Of 589 articles, 12 studies that enrolled 999 posterior canal benign paroxysmal positional vertigo patients were selected. The pooled analysis revealed that the Epley maneuver was as efficacious as the Semont maneuver, in both the 1-week recovery rate and end of study recovery rate.
- Conclusion: These two techniques were both better than sham-controlled treatment in the two efficacy indicators. No difference was observed in recurrence rate for treatments.

Average Short Term Success Rate of CRP

Average short term success rate of Canalith Repositioning Procedure in RCT(*) and quasi RCT (Helminski, et. al., 2010).

<table>
<thead>
<tr>
<th>Study</th>
<th>Number resolved/ Number in study</th>
<th>Percent Resolve</th>
<th>Odds Ratio - Confidence Intervals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lynn et al., 1995*</td>
<td>16/18</td>
<td>89</td>
<td>22.0(3.41-141.73)</td>
</tr>
<tr>
<td>Von Brevern et al., 2006*</td>
<td>28/35</td>
<td>80</td>
<td>37.33(8.75-159.22)</td>
</tr>
<tr>
<td>Froehling et al., 2000</td>
<td>16/24</td>
<td>67</td>
<td>3.20(1.00-10.20)</td>
</tr>
<tr>
<td>Sherman et al., 2001</td>
<td>27/33</td>
<td>82</td>
<td>24.75(4.31-142.02)</td>
</tr>
<tr>
<td>Average</td>
<td>87/110</td>
<td>80 &lt; 9</td>
<td></td>
</tr>
</tbody>
</table>

Note: Variability in study by Froehling et al., 2000 may be due to clinical expertise of the study personnel.

BPPV Recurrence

- Of patients treated successfully
  - 25% redevelop BPPV within 1 year
  - 44% redevelop BPPV within 2 years
  - 36% recurrence rate per Cochrane meta-analysis (9-11 studies of 745 patients)

(Cochrane Database Syst Rev. 2014, The Epley (canalith repositioning) manoeuvre for benign paroxysmal positional vertigo)
Daily CRP or BD exercises does not reduce recurrence rates

Daily routine of Brandt-Daroff exercises (Helminski, et. al., 2005) or self-administered CRP (Helminski, et. al., 2008) does not affect the time to recurrence or rate of recurrence of PC-BPPV.

Case 1 – Differentiate between PC and AC-BPPV

- Patient History
  - 40 year old female with a 7 year history of episodic positional vertigo. Most recent episode began 7 days prior to initial physical therapy evaluation.

- Chief complaints
  - Vertigo evoked by
    - Positional changes of the head relative to gravity (roll in bed towards right, get in and out of bed, bend forward, and look-up).
    - Rapid movements of the head.
  - Nausea and vomiting
  - Anxiety

Examination

Left Dix-Hallpike Test – downbeating with left torsion; little/no nystagmus at return to upright.

Survival Analysis – Time to Recurrence

Survival Analysis – Time to Recurrence (Helminski, et. al., 2005)

Survival Analysis – Time to Recurrence (Helminski, et. al., 2005)
Examination
Right Dix-Hallpike Test - downbeating and left torsion; return to upright no nystagmus.

Examination
Supine Roll Test – Head Right – (right beating), Center – slight downbeating, and Head Left.....

Examination
• Dix-Hallpike Test – Head Hanging transient burst of upbeating/right torsion nystagmus followed by downbeating/left torsion nystagmus
• Return to sitting – downbeating/left torsion.
Clinical Impression

Initial DHP test suggests left aSCC;
Repeat DHP test suggests right pSCC.
Supine roll test suggest hSCC too

What might explain this?
Multi SCC BPPV
Inhibited Right pSCC - consider location of the initial otoconia
Repositioning Maneuvers for Benign Paroxysmal Positional Vertigo

Advanced Practice in Vestibular Physical Therapy
BPPV Treatment Lab

Tests for Positional Vertigo

**Dix-Hallpike Test** (A) the patient sits on the examination table and turns his/her head 45° horizontally (1). The head and trunk are brought straight back "en bloc" so that the head is hanging over the edge of the examination table by 20° (2). Nystagmus is looked for and the patient is asked if they have vertigo. Although not shown in the figure, the patient then slowly returns to a sitting position with the head still turned 45° and nystagmus is looked for again. This test then is repeated with the head turned 45° in the other direction. The figure below shows movement of otoconia (canalithiasis) in the right posterior SCC (large black arrows) during the Dix-Hallpike test. In this example, the patient would have nystagmus and vertigo when the test is done on the right side, but not likely when the test is done on the left side.

In the **Sidelying Test** (B), the patient sits on the bed with the legs over the side and turns his/her head 45° horizontally to the left (1). The patient then quickly lies down on their right side (2). Nystagmus is looked for and the patient is asked to report any vertigo. Although not shown in the figure, the patient then returns to a sitting position with the head still turned 45° to the left, and nystagmus and vertigo is rechecked. The patient then turns his/her head 45° horizontally to the right and lies down quickly on their left side. Nystagmus and vertigo is checked. The patient then sits up and nystagmus and vertigo is checked. This figure also shows movement of mobile otoconia in the right posterior SCC (large black arrows) during the test. In this example, the patient would have nystagmus and vertigo when the test is done on the right side, but not likely on the left side.
1a. Canalith Repositioning Treatment/procedure (CRT/CRP)  
Illustrated below for Right BPPV canolithiasis

**INDICATION:** Paroxysmal up/Down beating nystagmus with a torsional component (pSCC/aSCC BPPV, canolithiasis variant)

**PROCEDURE:** Lay down head 45 degrees to the involved side, head extended, wait for 20---30 seconds or until dizziness/nystagmus resolves, slowly roll 45 degrees to uninvolved wait 20---30 seconds or until dizziness/nystagmus resolves, slowly roll 45 degrees nose down on the uninvolved side and wait 20---30 seconds or until all dizziness nystagmus resolves

**SUCCESS:** Absence of nystagmus/vertigo on repeated maneuver. You may see a vertical burst of nystagmus upon sitting up (does not always mean failure)
Liberatory Maneuver for Posterior Canal (PC) or “Semont”
Illustrated for Right pSCC.

INDICATION: Persistent upbeating with torsional nystagmus consistent with PC BPPV especially if more persistent or resistant to treatment with CRT/CRP, primary treatment for cupulolithiasis

PROCEDURE: Move patient quickly onto involved side (Brandt---Daroff BD position), nose (head) rotated up 45 degrees and stay in that position for 1---2 minutes (Step 2). Quickly, in one quick motion, sit up and go into opposite side (good side) with nose (head) now rotated down 45 degrees and stay in this position for 1---2 minutes (Step 3). Remember that the flip to be effective needs to be completed in <1.5 seconds. If secondary orthotropic burst, return slowly to sitting, if reversal of nystagmus or unsure, repeat Steps 2& 3 for up to 3 repetitions. Maneuver can be augmented by starting by laying slowly step 3 then quickly going to Step 2 wait 1---2 minutes and then complete flip to Step 3. (Cohen, H. 2007)

SUCCESS: observing a second vertical burst of nystagmus in the nose down position and/or resolve of all nystagmus/vertigo on final repetitions. If unsuccessful, can be prescribed as home exercise routine 3x/day 3x in a row till 2 days asymptomatic.
Sleep maneuver for pSCC. Demonstrated for the Right pSCC

Sleep Maneuver for pSCC BPPV

N=81 confirmed pSCC BPPV
88.9% success within 2 weeks
Not controlled
Maintain Position B for 3 minutes
Sleep in Position C

Shih and Wang 2012
Alternate test for aSCC BPPV: Deep Head hang

Deep Head Hang (30° of extension)
Place the head in a straight head hanging position to enable the otoconia to clear the curvature of the canal (Bertholon 2002).

• A minimum of 60° of extension is needed to clear the curvature of the AC (Kim and Amedee, 2002; Crevits, 2005).
Deep head hang treatment for aSCC Canalithiasis

Indication: Short duration, paroxysmal Down beating with/without Torsion consistent with AC BPPV Canalithiasis, especially when you can’t determine the side.

Procedure: Hold position 2 and 3 for 3 minutes or less depending on nystagmus and vertigo (Jacovino et al. J Neurol 2009;256:1851-1855.)

Success: Resolved nystagmus/vertigo on repeated maneuver
Liberatory Maneuver (LM) modified for the Left Anterior Canal (AC) or “Modified Semont Maneuver”

**Indication:** Persistent (or short duration) downbeating nystagmus with torsional component consistent with AC BPPV cupulolithiasis

**SUCCESS:** observing a second vertical burst of nystagmus in the nose down position and/or resolve of all nystagmus/vertigo on final repetitions. If unsuccessful, can be prescribed as home exercise routine 3x/day 3x in a row till 2 days asymptomatic.
LOCALIZING LATERAL CANAL BPPV

Supine Roll Test (SRT, pictured for Left side involvement)

GEOTROPIC (Canalithiasis) versus APOGEOTROPIC (Cupulolithiasis or debris in the ampullary or anterior arm)

(Schubert MC: Vestibular Special Interest Group Special Edition Newsletter on BPPV Dec 2012)
GEOTROPIC/CANALITHIASIS: A. lay supine (during transition you may see right beating nystagmus), B. rapid roll to the involved side (Left) will produce strong symptomatic paroxysmal geotropic (left) beating nystagmus as the crystals/debris moves towards the cupula, you may see a spontaneous reversal of the nystagmus (slight right beat may be persistent), C. rapid roll to the uninvolved side (right) will result in a less intense paroxysmal geotropic (right) beating nystagmus as now the crystals/debris are moving away from the cupula.

APOGEOTROPIC/CUPULOLITHIASIS: A. lay supine (during transition you may see left beating nystagmus), B. rapid roll to the involved side (left) will produce a mild persistent apogeotropic (right) beating nystagmus as the crystals/debris are deflecting the cupula in an inhibitory direction, C. rapid roll to the uninvolved side (right) will result in a more intense persistent apogeotropic (left) beating nystagmus when on the right side as the cupula is now being deflected in an excitatory direction.

Fundamental FACT:
In hSCC BPPV, the nystagmus is always more intense when it is beating toward the affected ear (regardless of geotropic or apogeotropic).
**Bow and Lean Localizing Test:**

Choung YH et al. ‘Bow and Lean Test’ to determine the affected ear of horizontal canal benign paroxysmal positional vertigo. Laryngoscope 2006; 16: 1776

**Geotropic/Canalithiasis:** When bow forward debris will migrate toward the ampulla/cupula, exciting the specific canal involved, i.e. if right side canalithiasis (geotropic) then when bow will see right beating nystagmus.
Apogeotropic/Cupulolithiasis: When bow forward, cupula will deflect inhibit the canal involved, i.e. if right side cupulolithiasis (apogeotropic) then when bow you will see left beating.
3a. Canalith Repositioning Treatment (CRT/CRP) for the geotropic BPPV or “Barbeque roll”. Illustrated for Right side hSCC Canalithiasis

**INDICATION:** Geotropic Direction Changing Nystagmus (DCN)

**PROCEDURE:** Start on involved side (most symptomatic side on roll testing, direction of the nystagmus in the bow test and or side that demonstrated a spontaneous reversal), slowly roll to nose up wait 20---30 seconds, roll to the uninvolved side and wait, roll until nose down into the bed (doesn’t need to be body prone if flexible enough), and slowly sit up keeping orientation. 1---3 repetitions, Augment success by sending home with self-repositioning and/or forced prolonged positioning on uninvolved side. You may continue to roll the patient over onto their side (always roll away from affected ear) if easier for them to sit up.

**SUCCESS:** Nystagmus stays orthotropic once you start the 270 degree roll with full resolve when nose down and on repeat testing.
APPIANI or GUFONI Maneuver for hSCC canalithiasis (published by Appiani in US, by GUFONI in Italian, aka Modified Canalith Repositioning (CRT) for Geotropic BPPV)

**Indication:** Symptomatic, paroxysmal Geotropic DCN on roll testing consistent with LC BPPV, localize side by the most symptomatic/intense nystagmus, side of spontaneous reversal if seen, and/or direction of nystagmus on the bow test

![Image of the maneuver]

**PROCEDURE:** Quickly onto the UNINVOLVED side and wait for one minute then turn head down as close to 90 degrees as possible and stay for two minutes. This was established by RCT by Kim et al (2012).

**SUCCESS:** Geotropic paroxysmal bursts that resolve nose down with resolve on repeat testing

GUFONI Maneuver for hSCC Apogeotropic nystagmus

**INDICATION**: Apogeotropic DCN, localize side by the least symptomatic/intense side, direction of nystagmus in the lean test, and/or direction of psuedonystagmus. (Pictured for Left left hSCC cupulolithiasis)


**PROCEDURE**: Quickly lay on the affected side and stay for 2 minutes, quickly tilt head upward 45 degrees and stay for 2 minutes, return to sitting. This is the maneuver established via Randomized controlled trial by Kim et al 2011.

**SUCCESS**: Resolution of or conversion of nystagmus to geotropic (must then reposition)

**Head Shaking:**
For head---shaking maneuver, in a sitting position, pitch the head forward by approximately 30°. Now move the head sideways in a sinusoidal fashion at an approximate rate of 3 Hz (3 times back and forth in 1 second – basically as fast as possible) for 15 seconds. This also successfully treated hSCC cupulolithiasis (Kim et al 2011)
Mastoid Oscillation

Kim et al. 2017 Efficacy of mastoid oscillation and the Gufoni maneuver for treating apogeotropic horizontal benign positional vertigo: a randomized controlled study.

**Success Rate**: Immediate Success after 1 treatment 48% (32/67). Next day 72% resolved.
A Vestibulo-Ocular Reflex and the Need for Speed

Activities of daily life (such as running) can have head velocities of up to 550 d/s, head accelerations of up to 6,000 d/s², and frequency content of head motion from 1 to 20 Hz.

For an Astronaut, their ADL includes traveling at 4.76 miles/s or 7,150 miles per hour.
Only the vestibular system can detect and appropriately respond across these broad ranges of head velocity and Hz

One result of an abnormal vestibular system...
Poor Gaze Stability

Eyes are not stable during rapid head rotation and patients perceive blurred visual targets - gaze instability
The Past: What treatments exist for Gaze Instability?

Vestibular Rehabilitation

Exercises designed to improve VOR function via VOR Adaptation or Substitution

Q: Where did Gaze Stability (VOR Adaptation) Exercises Originate?

A: The clinic, evidence came later

Terence Cawthorne MD (1945) makes a few critical clinical observations

- Noted improved performance (function) and reduced symptoms in 120 patients with Meniere’s disease after surgical ablation AND 58 patients with traumatic brain injury (both groups suffered similar symptoms)
- Described the catch up saccade: “…following sudden head turning it is not often possible to observe any involuntary eye movements, though sometimes a fleeting flicker may be present.”

The Past: Cawthorne Focuses on the Vestibular System as Cause for Symptoms and Basis of Rehab

Cawthorne (1945) further

- Hypothesized the root cause of similar symptoms in vestibular hypofunction and mTBI is the vestibular system “…in both it may be the end-organ in the labyrinth that is the seat of the damage…”

- Established the value of "Prehab" (Magnusson 2007), when he “takes his cases of Meniere’s syndrome into hospital a week or two before operation….., so that they begin to learn the exercises and are encouraged by seeing the progressive recovery of these other patients.”

Q: Who developed the original Vestibular Rehabilitation Program?

A: Cawthorne and Cooksey

In 1941, Cawthorne asked FS Cooksey to develop “the principles governing the restoration of fitness after injury to the vestibule and sought my help to develop a system of rehabilitation for these cases.”

1946 FS Cooksey’s published his Rehabilitation Program

He described 4 principals:

1. Clinician must "gain confidence and co-operation by explaining the nature of the symptoms and purpose of treatment with special emphasis on the need to make regular and gradually increasing efforts to do just those things which they find is stressing.”

2. Anxiety must be relieved early on.

3. A single member of the rehabilitation team should be in charge and be present whenever they are seen by the surgeon.

4. The rehab should include physical, mental, occupationally relevant exercises “planned to occupy the whole day and at the same time allow adequate periods for rest…”
Cawthorne-Cooksey Exercises

Non-specific
- ROM head/trunk
- Pursuit and vergence exercises
- VOR exercises not explicitly mentioned
- Posture and gait

Took a lot of time

Should VPT be providing patients with vestibular hypofunction, pursuit, vergence, or saccade exercises?
NO

The Vestibular and Visual Senses are linked

Vestibular neurons respond to visual rotation independent of head rotation

Same firing rate is increased when visual and vestibular stimuli are combined (Jüker and Precht 1979)

Vestibular neuron response to (A) visual alone, (B) vestibular alone, (C) visual and vestibular stimuli. (D) is the visual stimulus

E. Transient response of one VN to vestibular stimulation. F. Same unit, response to rotation in light room.

Vestibular and Visual Systems are Linked

The interdependency between vestibular and visual afference enables a means for vestibular compensation
- Unique type of saccade is substituted for a deficient vestibulo-ocular reflex (VOR)
- The latency and amplitude of these unique saccade are dependent on light

Schubert et al. 2008
Mills et al. 2016
The Vestibular and Somatosensory Senses are Linked

41% of vestibular neurons (cat) are modulated by hindlimb stimulation (cat) (McCall AA et al. 2016)

In human, limb position, weight distribution, and ground compliance (standing) alter vestibulospinal reflexes (Grasso et al. 2011; Manden et al. 2002; Welgampola and Colebatch 2003)

Unique neuronal response

The Past: Cawthorne Cooksey Exercises Worked

Earliest studies from 1970’s show the value of Cawthorne/Cooksey (CC) exercises (Hecker et al 1974; Dix 1979)

Many of the early CC studies included broad patient diagnoses with clearly described BPPV, migraine, TBI, and vestibular hypofunction

The CC form of vestibular rehabilitation predominated into the late 80’s and 90’s as studies began to objectify better their results (Norre 1979; Norre and DeWeert 1981; Norre and Beckers 1988; 1989; Shumway Cook and Horak 1990; Horak et al 1992)

In the mid 90’s, Herdman uses VOR adaptation exercises vs. pursuit-only eye movements to show improved posture, gait and perception of imbalance (Herdman et al OHNS 1995)

Pursuit group showed no change... and vestibular rehabilitation begins to differentiate

The Present:

Purpose: Develop training regimes and methods to improve gaze stability as measured by the vestibulo-ocular reflex

Current best practice (Vestibular Rehabilitation)

- Lengthy (~30 minutes), repetitive (4-5 times per day), imprecise (affects both sides – unnecessary? in the case of 1 sided loss)
- Gait and gaze stability (dynamic visual acuity) improve, and dizziness reduces but the time to benefit is long (6-12 weeks), the required effort is large, and the physiological vestibular reflex response (passive head rotation) improves little or at all (Herdman 2003, 2007; Scherer et al 2008)
- Poor compliance
  - 50% were under-compliant on a majority of the days (performed fewer exercises or for shorter durations) (Huang 2014)
  - The other 50% were ‘generally above-compliant’ (did many more yaw exercises than prescribed)
- Drop out rates increase for unsupervised training (55% vs 10%) (Pavlou 2013)
- Additionally, up to 25% of patients seen in vestibular rehabilitation clinics do not make much improvement in various outcome measures (Brown et al., 2001; Schubert 2006 et al; Herdman et al 2012)

Herdman et al 1994
The Present: Treatment Options for Gaze Instability

Based on models of VOR adaptation – retinal slip & head movements
- x1 & x2 viewing exercises
- Gaze shift/2-target exercise

Progression
- Duration
- Velocity
- Full field vs foveal stimulus
- Position
- Target distance

The Present: Gaze stability exercises help – but can we be more direct/quicker/more efficient with greater outcomes?

Gaze Stability Exercise improve DVA
Increase in VOR gain during active head rotation of ‘high’ velocities
(Schubert et al 2008)
VOR gain improved <5% passive head rotation in a few patients
(Scherer et al 2008)
Remembered target exercises increased VOR gain in some patients
(Scherer et al 2010)
In chronic unilateral loss (Schubert et al 2008)
- Increased number of compensatory saccades (overt and covert)
- Decreased amplitude of compensatory saccades (overt and covert)
- Compensatory saccades (overt and covert) occurred more frequently when trying to identify an optotype

So, VPT can improve DVA, how people feel, but VOR does not seem to change much (passive head rotation)……

What is VOR Adaptation
Adaptation - change in neural response to a stimulus that remains constant
For the VOR, change in magnitude of the slow phase response related to a head rotation
A PT correlate – functional improvement in dynamic visual acuity (gaze stability) from constant retinal slip error signal

Goals of VOR Adaptation:
- decreasing retinal slip
- improving visual acuity during head rotation
- improved postural stability
- decreased symptoms of imbalance, dizziness
What is VOR Adaptation

A. Normal VOR with a still visual target (i.e. X1 exercise)
B. Enhanced VOR – visual target moving opposite direction of head (i.e. X2 VOR exercise) - note the increase eye velocity and fire rate
C. VOR suppression with the visual target moving with the head - note the flat eye velocity
D. Reversed VOR with the drum moving at a velocity twice that of head velocity. Note the reversal peak eye velocity compared with panel B.

Properties of VOR Adaptation

- Context specificity
  - VOR learning occurs best at the training Hz and training position
  - Adaptation best when tested in the same head position of the training stimulus
  - This has important clinical implications

Schubert et al 2004

Degree of adaptation may be related to overall activity level within the vestibular system
Experimentally induced adaptation of gain
- Healthy young
- Young following UVH
- Elderly individuals
Summary of VOR gain adaptation

Retinal slip occurs when images move on retina and appears to be strongest error signal to drive change.

Retinal slip during head rotation for minutes to hours increases VOR gain 10%–35% (Tilikete 1993; Tilikete 1994; Shelhamer 1994; Yakushin 2003; Schubert 2008a).
- Most robust learning appears < 4Hz (Raymond 1998; Broussard 1999; Clendaniel 2001).

Retinal slip during head rotation for hours to days changes VOR gain > 60% (Melvill-Jones et al., 1988; Istl-Lenz et al., 1985; Miles and Elighmy, 1980; Miles and Braitman, 1980; Clendaniel et al., 2001; Kuki et al., 2004).
- Learned low gains are more resilient and required hours of up training for restoration to baseline.
- In the light, learned high gains decay spontaneously, only requiring minutes of down training to return to baseline (Boyden and Raymond, 2003; Kuki et al., 2004).

The Role of Activity in Vestibular Compensation

What is Vestibular Compensation? - Global term that refers to state of change in both static and dynamic behavior in response to a permanent vestibular lesion.

Static Compensation
- Occurs when tonic symmetry is established between the vestibular nuclei.
- Does not require input from the contralateral vestibular nuclei.
- Involves the intrinsic property of vestibular neurons:
  - neurochemical influences
  - increased excitatory efficiency of dorsal root fiber
  - release from cerebellar inhibition
  - synaptic changes
- Occurs in the absence of vision.

Dynamic Compensation
- Requires head motion.
- Requires retinal slip.
- Requires exposure to light.
- Sensory Integration/Reweighting
  - symmetric VOR gains during low velocity head rotations.
  - improved visual acuity during head rotation.
  - reduced postural instability in stance and gait.
  - reduced fall risk.

Complexity of Motor Learning in General

1. There are multiple time scales of learning - with different rates of learning and forgetting (seconds, minutes, hours, days, and months).
2. The pattern of training influences learning and forgetting, and rest periods between training sessions influence the retention of learning (consolidation).
3. Different contexts require different motor behaviors – VOR gain can be adapted up when eyes look up and adapted down looking down.
4. Deciding where blame resides when motor performance is impaired is difficult (the credit assignment problem). For example, am I the problem or is the environment different? Are my eye muscles weak? Has there been a change in how I perceive the world?
5. Learning takes place at multiple levels within the nervous system, from alterations in ion channel and membrane properties on single neurons, to more complex changes in neural circuit behavior, to higher-level cognitive processes including prediction and development of purposeful strategies to optimize motor performance.
Prior VOR Adaptation studies have limitations

Most studies
a) Done in complete dark
b) Using whole body head rotation or ambulating while wearing strong magnifying/minimizing lenses
c) With slow head velocities and/or low frequencies
d) Sinuoidal rotation

If VOR motor learning is context specific then how do these properties translate into functional activity in the world?

The Future
Is VOR adaptation to head impulses possible (relevant velocities of head rotation)? YES

Adaptation is robust during impulse head rotation (Schubert 2008). Adaptation appears better retained when incrementally applied (Schubert 2008; Migliaccio and Schubert 2013; 2014).

<table>
<thead>
<tr>
<th></th>
<th>Incremental</th>
<th>2</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>control</td>
<td>17.3 ± 4%</td>
<td>11 ± 9%</td>
<td>0.029</td>
</tr>
<tr>
<td>passive</td>
<td>14.2 ± 3%</td>
<td>6 ± 8%</td>
<td>0.018</td>
</tr>
<tr>
<td>UHV</td>
<td>18.2 ± 9%</td>
<td>6 ± 4%</td>
<td>0.008</td>
</tr>
</tbody>
</table>

Must provide a unique stimulus (Schubert 2008; Migliaccio and Schubert 2013; 2014).

The Future
Can we adapt the VOR to one side in healthy control? YES

Unilateral VOR gain adaptation is possible!

Migliaccio AA and Schubert MC 2013

20% ↑ VOR Gain
10% ↑ VOR Gain
~10% VOR gain increase, but 70% less than adapting side
No VOR gain increase

Migliaccio AA, Schubert MC 2014
The Future: Can we adapt the ipsilesional VOR gain in UVH?

Yes.

Pre-adaptation VOR gain 0.59 ± 0.22
Post-adaptation VOR gain 0.75 ± 0.30
(Mean of 29% improvement)
We continue to work on this

The Future: Can patients with BVH have VOR gain Adaptation?

Yes

Schubert et al 2008
We have developed a method to increase the VOR using a graded exposure to retinal slip –

The Future: Incremental VOR training

Healthy Control
Increased VOR gain (mean 17%) AND recruitment of compensatory saccades
Unilateral Vestibular Hypofunction
Unilateral VOR gain change possible (mean 23%)
Migliaccio AA and Schubert MC 2013
Migliaccio AA and Schubert MC 2014

Incremental Stimulus
Times 2 Stimulus

39% increase
Can we adapt the VOR in lighted conditions that can be replicated at home?

**Aim:** Determine the effect of background light level and laser target intensity on magnitude of VOR gain adaptation.

N=12 healthy subjects tested over 10 separate sessions.

Session 1-8, the background light level during adaptation training was: dark, 0.1, 0.2, 0.3, 0.5, 0.7, 1 and 290 lux. For sessions 9-10 the laser target intensity was halved with background at 0.1 and 0.3 lux.

Adaptation using left/right active head impulses for 15 minutes. VOR gain started at 1.0 and driven up by 0.1 every 90 seconds for rotations to one side (adapting) with fixed (at 1.0) towards the non-adapting side.

We measured active and passive impulse VOR gains before and after training.

<table>
<thead>
<tr>
<th>Source of Light</th>
<th>Lux</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 candle of light at 1m</td>
<td>1</td>
</tr>
<tr>
<td>Full moonlight</td>
<td>1</td>
</tr>
<tr>
<td>1 watt white LED</td>
<td>25–120</td>
</tr>
<tr>
<td>Overcast Day</td>
<td>100</td>
</tr>
<tr>
<td>40 watt light bulb</td>
<td>325</td>
</tr>
</tbody>
</table>

**How dark must the room be? Not completely - < 8 lux!**

1 lux = the illumination of a surface from a candle 1 meter away.

Active training leads to adaptation in less light (8 Lx) than testing with passive rotation (16 Lx).

Our data suggest incremental adaptation training increases the VOR gain when performed ≤ 8 lux

The active VOR gain % increases towards the adapting side were greatest at the lower ambient light levels of 0 (dark) and 0 lux.

**How bright must the target be?**

Target brightness during both 0 (dark) and 0.1 lux ambient light had no significant impact on VOR gain adaptation for both the Active and Passive VOR (ANOVA: active, p = 0.143, passive, p = 0.948).

Mahfuz et al 2016
Conclusion for influence of light during incremental VOR adaptation training with Relevance to VPT

The Adapting Side has greater % increase in VOR gain (p=2.7x10⁻²¹)
The LUX level significantly affects VOR gain adaptation (p=0.0001)
Intensity had no effect on % VOR Gain change, though some subject reported it easier to track the laser at 50% intensity
The type of head impulse test (active vs passive) after training did not have an effect on the amount of VOR gain change (p=0.97)
Our data suggest unilateral incremental VOR gain adaptation training is best when performed at or below 1 lux

Clinical Relevance of Incremental VOR Adaptation

It is currently unknown what the minimal clinically significant difference in VOR gain is to have a functionally relevant improvement in gaze stability or gait stability

We have shown that DVA (dynamic visual acuity) improves as VOR gain improves – and at those values we can reach with 15 minutes of incremental training.

Schubert et al 2008

Incremental aVOR Gain Training

Active VOR Training
Left Gain 1.0
Right Gain 0.5
Thank you for Listening
The Beginning – Anatomic Maturation

- At 4 weeks gestation, the otic vesicle has formed from invagination/cup to pit to vesicle (Bruska et al., 2009).
- The otic/auditory vesicle has two pouches, one dorsal, the other ventral.
- By 5 WG, the dorsal pouch enlarges to form, in chronological order, the anterior, posterior, and horizontal semicircular canals and each otolith macula (Streeter, 1906).
- By 7 WG, the ampulla swelling now contains the crista ampullaris for each semicircular canal.
- By 8-9 WG, otocystic rings are present near the tips of newly developing hair bundles (Sanes and Dechesne, 1985).
- By 10 WG, the cochlea has formed a full 2.5 turn coil.
- Between 9 and 18 WG there is a 3x increase in labyrinth length.
- By 17-19 WG, labyrinth size is presumed comparable to adult form.
- By 22-24 WG, the otocystic membrane is near maturity with otocystic ranging in size from ~2 to 5 μm (Sanchez-Fernandez and Rivera-Romar, 1984).
- Adult average 10 μm.

Vestibular End-Organ Anatomic Development

- Isolated, partial inner ear preparations of left vestibular end-organs from 13 WG human fetus.
- Vestibular “triad,” consisting of a horizontal and anterior ampullae and their associated cristae, joined with the utricle and remnants of the VIIIth cranial nerve. (Lim et al. 2014)
Healthy Aging

**Maculae have Greater # of Hair Cells vs Cristae**

**Fetal**
- Saccular macula: ~19,100 hair cell counts (14–23 WG)

**Adult**
- Utricular macula: ~29,500 to 39,200 hair cells
- ~2000 – 8000 in the central striola region (Rosenhall, 1972)
- Saccular macula: ~16,000 - 21,300 hair cells (~ half those estimated in the utricule)
- Semicircular canal cristae: ~6700 – 8300 hair cells (Lopez et al., 2005; Rosenhall, 1972; Watanuki and Schuknecht, 1976)
- No difference across 3 semicircular canals

**Type I and Type II HC Densities Decline**

Significant decline in type I HC number with age (Lopez et al 2005)
- Peripheral zone has significant decline in Type I and II hair cells
- Central zone has more Type II hair cells
- No change in intermediate Zone
- No change of Type I HC in the Central zone
- No Significant Effect of Age on support cells

**Hair Cell Degradation**

(A) Crista ampullaris from a 67-year-old individual. 
(B) Crista ampullaris from a 95-year
(C) Macula utricle from a 55-year-old
(D) Macula utricle from a 90-year-old

Less sensory hair cells, basement membrane thickening (BM), and lipofuscin deposition (thick arrowheads)

Ishiyama et al 2009
Healthy Aging
Decline in Vestibular Neurons

21,000 – 25,812 vestibular ganglion neurons (Park et al., 2001; Tang 2002)

Decline in total ganglion neuron starts ~ 35-40 y.o.
Largest loss stabilizes ~ 60 years of age
(Park et al 2001)

> 70 y.o., 21% hair cell loss in the utricular maculae and ~ 24% on the saccular maculae and ~ 40% cristae ampullaris
(Richter 1980; Rosenhall 1973)

Review of Vestibular Testing
Clinic vs Laboratory methods

- Semicircular canal (n=6)
  - Head Impulse – all six canals (clinic and laboratory methods)
  - Dynamic visual acuity – all six canals (clinic and laboratory)
  - VNG/ENG – horizontal canal only (laboratory only)

- Otolith end organs (n=4)
  - VEMP – ocular and cervical (laboratory only)
  - SVV/Bucket (clinic and laboratory)
  - Head tilt test (clinic)

Subjective Head Tilt Test – suggested utricular test

- VOG with position sensor (arrow) attached to goggle, n=43 patients with vestibular schwannoma
- Static Head Tilt: Eyes open in dark, 30 s wait, measure head position
- Subjective Head Vertical: Subject makes 3, 30–40° head tilts towards each shoulder (alternate) in total darkness, wait ~ 15 s then return to head straight position and waits another 15 s. Measure final head position

Control (healthy subjects) show head tilting <2° is normal; 2–3° moderately abnormal; >3° definitely abnormal (Hørsen et al 2011; Jutila 2014)

<table>
<thead>
<tr>
<th>Measure</th>
<th>Pre-operative</th>
<th>Post-operative</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Ipsilateral %</td>
<td>Contralateral %</td>
</tr>
<tr>
<td>Ipsilateral %</td>
<td>Ipsilateral %</td>
<td>Contralateral %</td>
</tr>
<tr>
<td>Static Head Tilt</td>
<td>56 (24)</td>
<td>19 (8)</td>
</tr>
<tr>
<td>Subjective Head Vertical</td>
<td>63 (27)</td>
<td>32 (14)</td>
</tr>
</tbody>
</table>
Concomitant decline in cochlear and saccular function associated with aging may reflect the common embryologic origin of both structures.

Noise exposure may contribute to both saccular and cochlear dysfunction.

Screening older individuals with high-frequency hearing loss for saccular/other vestibular hypofunction may help identify those at risk for fall.

Zuniga et al. 2012

Healthy Aging
Correlation between Hearing and Vestibular Loss

Worse Dynamic Visual Acuity during Passive Head Impulses

Agrawal et al. 2012
Healthy Aging
Otolith function worsens

Age-dependent changes in amplitude and latency of oVEMPs to bone-conducted vibration

Reduced n10 oVEMP amplitude with age (left panel).
Increased n10 oVEMP latency with age (right panel).
The box and whisker plots show the median and quartiles (Iwasaki 2013)

Semicircular canal and otolith end organ dysfunction

- Every end organ (each ear) affected
- The semicircular canals seem to be more commonly affected
- Utricular function (oVEMP) ‘relatively’ spared

Agrawal et al 2012
Healthy Aging
CANAL Function degrades in those >80 years

- VOR gain declined at a rate of 0.012/year (p = 0.033).
- Individuals aged 80 years or older had a nearly 8-fold increased odds of yaw impulse VOR gain less than 0.80 relative to those aged less than 80 years in multivariate models (prevalence of 13.2% vs. 2.8%; OR 7.79, 95% CI: 1.04-58.38).

Li et al 2015

CANAL Function: Head Impulse
- healthy elderly >70 years

- The vertical canal VOR gains are more variable than the horizontal canals
- The vertical SCC VOR gains tend to be lower (>0.7) than the horizontal canal gains (>0.8)
- The mean posterior SCC VOR gains tend to be the lowest (vs. mean horizontal/anterior SCC)
- Perhaps relates to falling based on reduced pitch head motion detection?

McGarvie et al 2015

CANAL Function: Impulse VOR gain in healthy elderly with no complaints of dizziness/imbalance
Emerging evidence suggests the presence of a compensatory saccade may be better marker for VOR deficiency, irrespective of VOR gain

Anson et al 2016
VOR deficits in Healthy Elderly Despite Normal DHI

- 36-44% of 70-95 y.o. have evidence of hSCC VOR hypofunction based on vHIT
- 65% of > 80 y.o. showed VOR deficits
- DHI scores were normal (5.6 ± 11.2)

CANAL Function: VOR Gain in Diabetes

Head velocity specific
- Lower velocity - no difference in VOR gain (active or passive, whole body 100d/s rotations) (Gamron 2002)
- Abnormal impulse gain all canals (Cardenos-Robledo et al 2016)

CANAL Function: Dynamic Visual Acuity in Diabetes

DVA using passive impulses within the individual SCC plane can identify acute isolated semicircular canal lesions (Schubert et al 2006)
Superior vestibular nerve distribution for canal innervation (horizontal/superior SCC) appears selectively damaged in DM (posterior canal appears spared)
CANAL Function: Dynamic visual acuity in each semicircular canal and otolith end-organ dysfunction in diabetes

- Abnormal DV A – semicircular canal
  > 0.18 LogMar (~2 lines of visual acuity)

- Abnormal or absent o/c VEMP – otolith
  Threshold/magnitude

21/25 (84%) had dysfunction of at least 1 vestibular end-organ
72% had at least 2 affected end-organs

OTOLITH Function: Diabetes

- Cervical VEMP
  38% of DM group had an absent cVEMP on one side, compared to 18% of age-matched (p=0.01)

- Ocular VEMP
  28% of DM group had an absent oVEMP on one side, compared to 13% age-matched (p=0.01)

Ward et al. 2015

Horizonal Semicircular Canal Function in Multiple Sclerosis

- Reduced gain of the VOR and presence of compensatory saccades in subjects with MS

Garg et al. In Review
CANAL Function in Multiple Sclerosis

- As a group, VOR gain in MS was not different than controls.
- HOWEVER, patients with MS use more compensatory saccades/head rotation.
- Both patient with MS and age-matched controls used compensatory saccades, patients tended were earlier (p=0.05).

Garg et al. In Review.

OTOLITH Function: Subjective Visual Vertical (SVV) in Elderly, MS and DM

Healthy Elderly
- > 3d in 5/51 elderly (70-79 yrs)
- Subjective percept thus appears normal

Multiple Sclerosis
- 48% abnormal (mean 0.82° ± 2.32) vs. healthy (mean 0.22° ± 1) (Crevits 2007)
- 36% abnormal SVV > 5deg (Serra et al. 2003)

Diabetes
- Unknown

<table>
<thead>
<tr>
<th>MS Oculomotor signs</th>
<th>MS patients with SVV &gt; 5deg</th>
<th>MS patient with SVV &lt; 5deg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Misalignment of visual axes</td>
<td>2(29%)</td>
<td>2(10%)</td>
</tr>
<tr>
<td>Gaze-evoked nystagmus</td>
<td>4(57%)</td>
<td>3(27%)</td>
</tr>
<tr>
<td>Nystagmus in central position</td>
<td>0(0%)</td>
<td>0(0%)</td>
</tr>
<tr>
<td>Saccadic disinsertion</td>
<td>6(86%) (in both planes)</td>
<td>5(41%) (4 in both planes)</td>
</tr>
<tr>
<td>Intersaccadic saccadic latency</td>
<td>4.07% (2 majority bilateral)</td>
<td>1.89% (1 majority bilateral)</td>
</tr>
<tr>
<td>Smooth pursuit impaired</td>
<td>5(71%) (5 majority bilateral)</td>
<td>4(31%) (4 majority bilateral)</td>
</tr>
<tr>
<td>VOR impaired</td>
<td>2(29%) (2 majority bilateral)</td>
<td>1(27%) (1 majority bilateral)</td>
</tr>
<tr>
<td>Vertigo impaired</td>
<td>2(29%)</td>
<td>2(27%)</td>
</tr>
</tbody>
</table>

OTOLITH Function: VEMPS in MS

- 70% patients with MS have abnormal oVEMPs compared with cVEMPs (8%)
- Ocular VEMPs were absent in 50% tested ears, delayed in 20% cases
- cVEMP seems to be less affected than oVEMP

(Rosengren 2011; Oh 2015)
VOR in cerebellar pathology

- Acute vestibular syndrome can be due to cerebellar lesion with the expected mixed central/peripheral vestibular signs
  - cVEMPs appear unaffected by at least certain cerebellar peduncle and cerebellar hemispheric lesions. (Papacostas 2015; Yacovino 2017)
  - Normal VEMPs in recent CANVAS patient (Rust 2017)
- Head shaking does not suppress with fixation (Maia 2017)

**VOR in cerebellar pathology - Acute Floccular Syndrome**

- Lesion restricted to the right flocculus
- Normal c-VEMP, o-VEMP initially diminished on the left side. Recovers 4 wks
- Abnormal vHIT. Poor VOR cancellation
- The right ear showed 42% caloric weakness, normal at 4 weeks
- Contraversive, large SVV tilt, normal at 4 wks

**Conclusion**

- Type I and II vestibular SCC hair cells decline with age but variable depending on their location within the crista
- Saccular end organ normally has only ½ the HC compared with the utricle
  - With age, the saccule appears to show degraded function sooner than the utricle
  - The utricle appears to be relatively spared in healthy aging
- Age 35/40, the vestibular afferents begin a 15-20% decline that appears to stabilize by age 60/65
- Both diabetes and MS show significant vestibular end-organ damage beyond healthy age-matched controls
  - Critical to consider the VOR when treating these patients
Your 1st patient at your next clinic

- 72 y.o. female with chief report of progressive imbalance
- No vertigo
- Describes being dizzy with head motion
- Reports oscillopsia during rapid head motion
- Reports some mild numbness in left lower leg

Room Light Exam

Oculomotor Room Light
Poor Pursuit or Poor VOR or both?

Video Head Impulse Test

Cerebellar Ataxia with Neuropathy and Vestibular Areflexia Syndrome (CANVAS)

- Multisystem ataxia with cerebellar, vestibular, and sensory involvement
- Slowly progressive
- Pathologically, CANVAS is defined by a multiple cranial nerve and dorsal root neuropathy and a consistent pattern of cerebellar atrophy.
- May also include dysphagia, cough, autonomic dysfunction (such as postural hypertension), somatic allodynia, and dysesthesia (Szmulewicz 2011, 2014)

In CANVAS, saccades during SLOW head rotation (poor bilateral VOR and pursuit) are a common clinical finding.
What Treatment Will You Do?

- Gait and Balance
- VOR gaze stability
- Habituation
- Safety/Fall risk management

Thank You for Listening
Vestibulo-ocular Reflex (VOR) Physiology 101

- Deflection of the stereocilia toward the single kinocilia in each hair cell leads to excitation (depolarization), and deflection of the stereocilia away from the kinocilia leads to inhibition (hyperpolarization).
- Deflection of the stereocilia occurs by motion of the endolymph results in an opening (or closing) of the transduction channels of hair cells, which changes the membrane potential of the hair cells.
- In the horizontal SCC, hair cells are oriented so that endolymph motion toward the ampulla causes excitation.
- In the vertical SCCs (posterior and anterior) hair cells are oriented so that depolarization occurs when endolymph moves away from the ampulla.

Otolith Anatomy

- Otolith organs = saccule and utricle and respond to linear acceleration and static head tilt.
- Sensory hair cells project into a gelatinous material that has calcium carbonate crystalline-structure material (otoconia) embedded in it; this provides the otolith organs with an inertial mass.
- A central region exists within the utricle and the saccule known as the striola, which divides the otolith organs into two parts.
- The kinocilia of the utricular hair cells are oriented toward their striola, whereas the kinocilia of the saccular hair cells are oriented away from their striola.
- As with the SCC, motion toward the kinocilia causes excitation, while motion away leads to inhibition.
- Utricular excitation occurs during horizontal linear acceleration and/or static head tilt and saccular excitation occurs during vertical linear acceleration.
Six Extraocular Muscles Controlled by Three Cranial Nerves

<table>
<thead>
<tr>
<th>Horizontal</th>
<th>Lateral</th>
<th>Medial</th>
<th>Movements</th>
</tr>
</thead>
<tbody>
<tr>
<td>MVN</td>
<td>Right ocular right abducens</td>
<td>Right SR</td>
<td>Right SR</td>
</tr>
<tr>
<td>anterior</td>
<td>L/N</td>
<td>Left ocular left IL</td>
<td>Left SR</td>
</tr>
<tr>
<td>posterior</td>
<td>MVN</td>
<td>Left trochlear left IL</td>
<td>Right SO</td>
</tr>
</tbody>
</table>

Dx of hSCC BPPV

- Supine Roll Test
- Lie Down test
- Bow and Lean Test

hSCC BPPV

- Geotropic - nystagmus beats toward the ground and lasts < 60s. Indicates canalithiasis
- Apogeotropic – nystagmus beats away from the ground and lasts > 60s. Indicates cupulolithiasis
- In hSCC BPPV, nystagmus is always more intense when it is beating toward the affected ear (regardless of apogeotropic or geotropic).
Lie Down test: hSCC BPPV - Left Side

Small arrows denote fast phase of nystagmus

BPPV of the left hSCC. Passing from the sitting (A) to the supine position (B).

For geotropic form, the movement of the debris causes an inhibition of the cupula producing a nystagmus directed towards the healthy side (Right beat).

For apogeotropic form, lying down will cause beating towards the affected side (Left beat).

Help Diagnosis for hSCC

Possible Seated Nystagmus (apogeotropic or geotropic)

Possible Sit to Supine Nystagmus (apogeotropic or geotropic)
Bow and Lean Test for hSCC Nystagmus

Apogeotropic hSCC canalolithiasis and Geotropic hSCC canalolithiasis

**NOSE DOWN**

Bow Nystagmus

Ampulla

Posterior arm hSCC

Otoliths

Lean Nystagmus

**NOSE UP**

left hSCC: lateral view

---

Bow and Lean Test (BLT) 

hSCC Canalithiasis

Choung et al 2006

---

BLT- hSCC Cupulolithiasis

Choung et al 2006

---

The affected ear is determined as the same direction as that of bowing nystagmus and the opposite direction to that of leaning nystagmus.
Direction of nystagmus?

Direction of nystagmus?

Bow and Lean
Gufoni for Canalithiasis

RCT (Kim et al 2011)
- Side lie healthy side (weaker nystagmus) 2 min
- Then rotate head down, hold 2 min

hSCC Canalithiasis

BBQ Roll
- hSCC canalithiasis
- Illustrated for treating the Right Side
Gufoni for Cupulolithiasis

• **Gufoni** – always lie down on side of weaker beat nystagmus (affected side)
  – Cupulolithiasis – lie down on affected side 2 min, then head up, wait 2 min (RCT Kim et al 2012)

hSCC Cupulolithiasis

Kim et al 2017 Cupulolithiasis Affecting either side of the Utricle
Time for Lab