What does the neurootologist need to know about eye movements?

1) Functional classification of eye movement subtypes
2) How to examine at the bedside the different eye movement subtypes
3) Localization using eye movement abnormalities in classical syndromes that may present to the neurootologist

1) Syndromes of the posterior fossa
   1) Wallenbergs
   2) AICA
   3) INO
   4) Cerebellum
      1) FLOCCULUS
      2) NODULUS

2) Distinctive syndromes that present to the Neurootologist
   1) OPSOCLONUS
   2) PSP (Progressive Supranuclear Palsy
   3) WERNICKES
   4) UPBEAT NYSTAGMUS

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The dominance of the fovea

- G Westheimer
- Courtesy, Chris Kennard
Function and classification of eye movements

• Serve the needs of vision, and specifically those of the fovea where spatial acuity is best.

• Bring images onto the fovea (saccades and vergence)

• Keep images still on the fovea, for best spatial resolution (vestibular, pursuit and vergence)
Skew deviation, ptosis, head tilt  Wallenbergs

• Learn to do a simple alternative cover test
Saccadic dysmetria – overshoot to one side, undershoot to the other

WALLENBERG’S SYNDROME
Wallenberg’s Syndrome - PICA distribution infarct involving the dorsolateral medulla
Wallenberg's Syndrome - PICA distribution infarct involving the dorsolateral medulla

Restiform body (ICP)
Wallenberg's syndrome (PICA)

- Skew deviation (OTR), due to involvement of caudal vestibular nuclei mediating otolith-ocular responses
- Saccadic dysmetria (ipsipulsion) due to a functional lesion of the fastigial nucleus from interruption of olivary-cerebellar climbing fibers to the cerebellar cortex which increases Purkinje cell inhibition on underlying fastigial nucleus.
- REMEMBER distal (medial) PICA syndrome which can mimic an acute vestibular neuritis.
VOR head impulse sign in a patient with hearing loss, facial weakness and limb ataxia
Middle cerebellar peduncle lesion in AICA infarct
AICA syndrome

• Peripheral involvement: Internal Auditory Artery: Unilateral loss of vestibular function
  - Skew deviation (OTR), due to involvement of otolith-ocular responses
  - Impulse sign due to involvement of canal-ocular responses
  - Hearing loss

• Central involvement: Ataxia, due to involvement of cerebellar pathways. Other signs of lateral pons
Internuclear ophthalmoplegia (INO) with adduction failure and abducting nystagmus and spared adduction with convergence
INO Skew deviation and head tilt
MLF lesion
MLF syndrome (INO)

- Slowing or loss of (conjugate) ADduction of eye on side of lesion with sparing of vergence-mediated adduction.
- Contralateral eye nystagmus on ABduction
- OTR due to involvement of otolith-ocular pathways traveling in the lateral portion of the MLF.
Downbeat, gaze-evoked and rebound nystagmus in cerebellar atrophy
Pursuit and VOR Cancellation abnormalities in cerebellar atrophy
Cerebellar flocculus and paraflocculus (tonsils)
Abnormal VOR in cerebellar disease

Abnormal direction
Head-shaking nystagmus in cerebellar disease
downbeat after horizontal head shaking
Cerebellar floccular-parafloccular (tonsil) syndrome

- Gaze-evoked, rebound, downbeat nystagmus
- Loss of pursuit and cancellation of the VOR during combined eye-head tracking
- Abnormal ‘cross-coupling’ with vertical responses to horizontal stimulus
- Postsaccadic drift
Periodic Alternating Nystagmus (PAN)

Null every two minutes
Anatomical Locus of PAN
PAN: Pathogenesis and Treatment

• Two key normal mechanisms
  - **Central velocity-storage mechanism** located within the vestibular nuclei that improves the ability of the vestibular system to respond to low-frequency head motion by perseverating peripheral vestibular signals.
  - **Adaptation mechanism** that acts to null any sustained unidirectional nystagmus (which in natural circumstances is always due to a lesion)

• In PAN, instability in velocity storage is produced by loss of (gaba-mediated) inhibition from the Purkinje cells of the nodulus onto the vestibular nuclei.

• Short-term adaptation (which is working normally) causes reversals of nystagmus leading to sustained oscillation.

• **Baclofen (GABA-b)** provides the missing inhibition and stops the nystagmus.
  - Usually need only 10 mg PO TID.
  - Avoid precipitous discontinuation.
  - Memantine may be a useful adjunct
  - Medications do not work as well in congenital PAN.
Opsoclonus

Patient with a few days of vertigo then objects began jumping

Causes

Paraneoplastic

Metabolic-toxic

Post viral, immune mediated
Some normal subjects can voluntarily produce saccadic oscillations (this capability can run in families)
Progressive loss of balance with falls

PROGRESSIVE SUPRANUCLEAR PALSY (PSP)
Slow vertical saccades, saccadic intrusions
PSP  small midbrain, 'humming bird' or 'penguin' sign
Anatomical locus for vertical saccade commands
Top of the Basilar Syndrome
supranuclear vertical gaze palsy
predominantly down
Vertical (down) gaze problems
Lesions in riMLF and caudal thalamus
Artery of Pecheron (subthalamatic perforator from the posterior cerebral artery)
Recent onset of jumping vision and imbalance

Wernicke's syndrome (B1-thiamine)
Bariatric surgery
Chemotherapy
Alcoholism
Eating disorders
Hypermesis Gravidarum
Wernicke’s Disease – Absent VOR slow phases (acute horizontal bilateral vestibulopathy (Choi I))
Wernicke's Disease
(lesions in the medial vestibular nuclei)
Upbeat nystagmus
Upbeat nystagmus and the nucleus intercalatus and nucleus of Roller (perihypoglossal nuclei).

Minagar et al. Neurology 2001
Pierrot-Deseilligny and Milea, Brain, 2005
A Phone Call from an Acutely Vertiginous Patient: Key Diagnoses

**Stroke**
- Neurological symptoms?
- Headache or neck pain?
- Any previous episodes?
- Duration of the spell?
- Is it positional?
- Hearing symptoms?
- Age?
- Vascular risk factors?

**BPPV**
- Positional, Transient, Recent inciting events (dentist, hairdresser, trauma)

**Vestibular Neuritis**
- Sustained vertigo even when at rest though worsened by movement, Viral illness

**Vestibular Migraine**
- Headache, Family or personal history of migraine, Aura, Light sensitivity, Relieved by sleep

**Menieres**
- Aural symptoms (pain, pressure, fullness in ear, seashell tinnitus, hearing loss (low frequency, fluctuating)

**Headache, Neck pain**
- Other
- Neurological Symptoms (diplopia, numbness, weakness, hiccoughs, dysphagia, dysarthria, incoordination), Hearing loss, Risk Factors (age, hypertension, lipid disorder, heart disease, diabetes)
# The Acutely Vertiginous Patient: Key Findings

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>Stroke</strong></td>
<td>Spontaneous jerk nystagmus that does not suppress with fixation, Gaze-evoked, direction-changing nystagmus, Skew deviation, Absent head impulse sign (though if present does not r/o stroke) Other neurological signs</td>
</tr>
<tr>
<td><strong>BPPV</strong></td>
<td>Positional nystagmus, Transient, after a latency, usually mixed vertical torsional if posterior canal or horizontal if lateral canal B Posterior canal BPPV changes direction on sitting up</td>
</tr>
<tr>
<td><strong>Vestibular Neuritis</strong></td>
<td>Sustained, spontaneous mixed horizontal-torsional nystagmus in straight ahead gaze. Obeys Alexander’s law (more intense with gaze in the direction of quick phase) More intense when lying with bad ear down. Suppressed with fixation, Positive head impulse sign</td>
</tr>
<tr>
<td><strong>Vestibular Migraine</strong></td>
<td>May mimic BPPV or Vestibular Neuritis or have central ocular motor signs (gaze-evoked or vertical nystagmus)</td>
</tr>
<tr>
<td><strong>Menieres</strong></td>
<td>Spontaneous nystagmus similar to vestibular neuritis, Nystagmus may spontaneously change direction in the first hours after onset. Loss of hearing,</td>
</tr>
</tbody>
</table>
A Flow Chart For Classification Of Nystagmus

Is fixation impaired because of a slow drift, or an intrusive saccade, away from the fixation target?

If a slow drift is culprit

Jerk

Unidirectional

Changes direction with viewing eye = Latent Nystagmus

Vestibular (constant velocity)

Central

Peripheral

(Use effect of fixation and vector of nystagmus)

Changes direction with direction of gaze (gaze-evoked)

Pendular

Acquired (MS or OPT) or Congenital

Changes direction with direction of gaze (gaze-evoked)

Acquired (velocity decreasing e.g. cerebellar)

Congenital (velocity increasing)