**A Flow Chart For Classification Of Nystagmus**

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

If a *slow drift* is culprit

- **Jerk**
  - Unidirectional
    - Vestibular (constant velocity)
      - Central
      - Peripheral (Use effect of fixation and vector of nystagmus)
    - Changes direction with viewing eye = Latent Nystagmus
  - Changes direction with direction of gaze (gaze-evoked)
    - Acquired (velocity decreasing e.g. cerebellar)
    - Congenital (velocity increasing)

- **Pendular**
  - Acquired (MS or OPT) or Congenital
A Flow Chart For Classification Of Nystagmus

If a saccade away from the fixation target (saccadic intrusion) is the culprit?

- If a saccade away from the fixation target (saccadic intrusion) is the culprit?
  - With an intersaccadic interval (~200 ms) before a return saccade
    - Square-wave jerks (small saccades away from and back to fixation), macrosquare wave jerks (much larger saccades)
    - Macrosaccadic oscillations, Large saccades around fixation (extreme degree of saccade hypermetria)
  - Without an intersaccadic interval (back to back saccades)
    - Opsoclonus (multiaxis)
    - Flutter (one axis) Includes ‘voluntary nystagmus’
    - Ocular Bobbing Single vertical saccade away from fixation followed by slow drift back
Waveforms of Nystagmus

A: Jerk or linear (vestibular)

B: Velocity decreasing (acquired gaze-evoked, congenital latent)

C: Velocity increasing (congenital)

D: Pendular
SCC Organization: a guide to nystagmus

Arrows indicate direction of slow phase with stimulation

Central Patterns

Peripheral Pattern

Flourens
### Summary table of nystagmus treatment (Strupp et al, J Neurology, 2011)

<table>
<thead>
<tr>
<th>Direction of nystagmus (quick phase)</th>
<th>Downbeat nystagmus (DBN)</th>
<th>Upbeat nystagmus (UBN)</th>
<th>Acquired pendular nystagmus (APN)</th>
<th>Period alternating nystagmus (PAN)</th>
<th>Infantile (congenital) nystagmus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Downward, may be diagonal with lateral gaze</td>
<td>downward</td>
<td>upward</td>
<td>mainly horizontal, may have vertical and/or torsional components</td>
<td>horizontal</td>
<td>mainly horizontal; may have torsional and small vertical components</td>
</tr>
<tr>
<td>Upward</td>
<td>jerk, constant, increasing, or decreasing slow-phase velocity</td>
<td>jerk, constant, increasing, or decreasing slow-phase velocity</td>
<td>pendular, sinusoidal slow-phase</td>
<td>Jerk, mostly constant slow-phase velocity</td>
<td>accelerating slow-phases; foveation periods when the eye is transiently still</td>
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<tr>
<td>Special features</td>
<td>Increased intensity during lateral and downward gaze; sometimes suppressed by convergence</td>
<td>Increased intensity during upward gaze; may convert to DBN on convergence</td>
<td>Associated with other oscillations (e.g., palate) and with hypertrophic degeneration of the inferior olive</td>
<td>Changes direction every 90 to 120 sec</td>
<td>Null zone, in which nystagmus is minimal; often suppressed with convergence</td>
</tr>
<tr>
<td>Sites of lesion</td>
<td>Cerebellum (bilateral floccular hypofunction); rarely lower brainstem lesions</td>
<td>Medial medulla, pontomesencephalic junction, rarely cerebellum</td>
<td>Pontomedullary, probably affecting components of neural integrator for gaze holding</td>
<td>Cerebellum (nodulus, uvula)</td>
<td>Uncertain; some cases are associated with afferent visual system anomalies</td>
</tr>
<tr>
<td>Etiology</td>
<td>Cerebellar tumors, degenerations, and stroke; idiopathic; often associated with bilateral vestibulopathy and neuropathy</td>
<td>Brainstem or cerebellar stroke and tumors; Wernicke’s encephalopathy</td>
<td>Multiple sclerosis, oculopalatal tremor due to brainstem or cerebellar stroke</td>
<td>Cerebellar degenerations, cranio-cervical anomalies, multiple sclerosis, cerebellar tumors and stroke</td>
<td>Uncertain; may be associated with afferent visual system anomalies; hereditary in some patients (e.g., FRMD7 mutations)</td>
</tr>
<tr>
<td>Treatment</td>
<td>4-aminopyridine (5-10 mg tid), 3,4-diaminopyridine (10-20 mg tid), baclofen (5 mg tid) clonazepam (0.5 mg tid)</td>
<td>Often transient, treatment often not necessary; baclofen (5-10 mg tid)</td>
<td>Memantine (10 mg qid) gabapentin (300 mg qid)</td>
<td>Baclofen (5-10 mg tid) gabapentin (300 mg qid)</td>
<td>Gabapentin (300 mg qid) memantine (10 mg qid)</td>
</tr>
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</table>
A Phone Call from an Acutely Vertiginous Patient: Key Diagnoses

- **Stroke**
- **BPPV**
- **Vestibular Neuritis**
- **Vestibular Migraine**
- **Menieres**

**Headache, Neck pain, Other Neurological Symptoms** (diplopia, numbness, weakness, hiccoughs, dysphagia, dysarthria, incoordination), Hearing loss, Risk Factors (age, hypertension, lipid disorder, heart disease, diabetes)

- Positional, Usually turning over or getting in and out of bed,
- Transient, Recent inciting events (dentist, hairdresser, trauma, bed rest),
- Happened before?

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

- **Sustained vertigo even when at rest though worsened by movement,**
- **No hearing change,**
- **Viral illness**

- **Headache, Family or personal history of migraine,**
- **Headaches may only be remote,**
- **Triggers, aura Light and noise sensitivity,**
- **Relieved by sleep**

- **Aural symptoms** (pain, pressure, fullness in ear, seashell tinnitus, Fluctuating hearing loss (low frequency))
- **Otolithic crises of Tumarkin**
- **Reversals of direction of vertigo,**
- **Lermoyez (hearing improves as vertigo begins)**
<table>
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<th>The Acutely Vertiginous Patient: Key Findings</th>
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<tr>
<td><strong>Stroke</strong></td>
</tr>
<tr>
<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 including hearing loss</td>
</tr>
<tr>
<td><strong>BPPV</strong></td>
</tr>
<tr>
<td>Positioning nystagmus, Transient, after a latency, usually mixed vertical torsional if posterior canal or horizontal if lateral canal. Posterior canal BPPV changes direction on sitting up, Lateral canal may be apoapogeotropic (beats to sky) or geotropic (beats to ground) <strong>When most intense is beating toward involved ear</strong></td>
</tr>
<tr>
<td><strong>Vestibular Neuritis</strong></td>
</tr>
<tr>
<td>Spontaneous, sustained, mixed horizontal-torsional nystagmus in straight ahead gaze. Obey 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>
</tr>
<tr>
<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>
</tr>
<tr>
<td>Spontaneous nystagmus similar to vestibular neuritis, Nystagmus may spontaneously change direction in the first minutes and then hours after onset. Loss of hearing, especially low frequency, Henneberts sign (tragal compression produces nystagmus)</td>
</tr>
</tbody>
</table>
Epley

Step 1

Step 2

Step 3

Step 4

Step 5

Semont

Step 1

Step 2

Step 3

Step 4

Helminski, Zee et al.