Bilateral Internuclear Ophthalmoplegia
Eye Movements

- Bilateral Internuclear Ophthalmoplegia
- Acquired Pendular Nystagmus
- Lid Nystagmus
- Upbeat Nystagmus
Clinical Features

Medial rectus muscle weakness ipsilateral to the side of the lesion with paresis of adduction or adduction lag.

Abducting nystagmus of the eye contralateral to the lesion – *Dissociated nystagmus*
Clinical Features

Normal convergence

**Skew deviation** – **hypertropia** on the side of the lesion

**Dissociated** vertical nystagmus – downbeat with greater torsional component in the contralateral eye
Bilateral INO

Additional Signs:

- Gaze evoked vertical nystagmus
- Impaired vertical pursuit
- Decreased vertical vestibular response
- Small amplitude saccadic intrusions suggesting involvement of the brainstem adjacent to the MLF
Pathogenesis of Certain Signs in Internuclear Ophthalmoplegia

<table>
<thead>
<tr>
<th>Ocular Motor Deficit</th>
<th>Possible Pathophysiologic Substrate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ipsilateral hypertropia skew</td>
<td>Otolith imbalance</td>
</tr>
<tr>
<td>Vertical-gaze evoked nystagmus</td>
<td>Vertical saccades bring eye to target but vertical eye position signal is inadequate</td>
</tr>
<tr>
<td>Vertical vestibular and pursuit movements impaired</td>
<td>Bilateral interruption MLF axons carrying vertical vestibular and smooth pursuit signals</td>
</tr>
</tbody>
</table>
Weakness of adduction is due to impaired conduction in axons from the abducens internuclear neurons which project to the medial rectus motor neurons in the contralateral oculomotor (third nerve) nucleus.
INO

Adduction weakness is most evident during saccades and adduction lag is brought out by asking the patient to look all the way to the right and all the way to the left (i.e. make large saccades).
The speed of the adducting eye depends on a strong agonist contraction. The adducting saccade may be slow and hypometric.
In the abducting eye, abducting saccades are hypometric with centripetal drifts of the eye and slowing. A series of small saccades and drifts have the clinical appearance of abducting nystagmus - dissociated nystagmus.
Dissociated nystagmus may be due to: impaired ability to inhibit the affected medial rectus or

Dissociated nystagmus reflects the brain’s attempts to compensate for the adduction weakness.
Etiology

Multiple sclerosis (commonly bilateral)

Brainstem infarction (commonly unilateral), including vasculitis, complication of arteriography and hemorrhage

Brainstem and fourth ventricular tumors
Etiology

Arnold-Chiari malformation and associated hydrocephalus

Infection: bacterial, viral and other forms of meningoencephalitis and AIDS
Etiology

Wernicke’s encephalopathy

Metabolic disorders: hepatic encephalopathy

Drug intoxications: phenothiazines, tricyclic antidepressants, narcotics, propranolol, lithium, barbiturates.
Pendular Horizontal Oscillations

Relatively high frequency oscillations that dampen after a blink

PHO are partially suppressed following a saccade and on convergence.
Primary position upbeat nystagmus is attributable to a lesion(s) in the region of the

Nucleus intercalatus

Nucleus of Roller
Clinical Features of Acquired Pendular Nystagmus (APV)

May have horizontal, vertical and torsional components; their amplitude and phase relationship determines the trajectory of the nystagmus in each eye.

Phase shift between the eyes is common (horizontally and torsionally; seldom vertically) – may reach 180 degrees, so that the nystagmus becomes convergent-divergent or cyclovergent.
Clinical Features of APN

Amplitudes often differ, and nystagmus may appear monocular

Trajectories may be conjugate, but more often are dissimilar

Oscillations sometimes suppress momentarily in the wake of a saccade
Clinical Features of APN

In Association with Demyelinating Diseases

Frequency 2-8 Hz (typically 3-4Hz)

Generally greater amplitude in the eye with poorer vision

Internuclear ophthalmoplegia commonly associated

May have an associated upbeat component
Syndrome of Oculopalatal Tremor

Frequency 1-3 Hz (typically 2 Hz)

May be vertical (with bilateral lesions) or disconjugate vertical-torsional

Accentuated by eyelid closure

Movements of palate and other branchial muscles may be synchronized
Whipple’s Disease

Frequency typically about 1 Hz

Usually convergence-divergence, occasionally vertical; sometimes with associated oscillatory movements of the jaw, face or limbs (oculomasticatory myorhythmia)
APN: Whipple’s Disease

Vertical gaze palsy similar to the clinical picture of progressive supranuclear palsy is usually also present
