Acquired Nystagmus

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  - Gaze holding systems
- Acquired pendular nystagmus
- Acquired jerk nystagmus
- Saccadic oscillations
- Management
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Nystagmus

- Is an involuntary rhythmic oscillation of one or both eyes
- Direction: Horizontal, vertical, torsional or mixed
- Conjugate or Disconjugate
- Dissociated
Gaze Holding System

- Nystagmus → by failure to hold the eyes steady
- Mechanisms to hold eyes steady:
  - Vestibular ocular reflex (VOR)
  - Optokinetic & smooth pursuit systems
  - Separate ‘fixation system’
  - Neural integrator (holds the eyes steady in eccentric gaze)
Physiological Nystagmus

- Induced by
  - self-rotation
  - instilling water into ear (vestibular nystagmus)

- Fine end-point nystagmus
  - Differentiate from pathological gaze-evoked nystagmus
Waveforms

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Acquired Pendular Nystagmus

- To-&-fro sinusoidal oscillation of the eyes
- **Direction:**
  - Horizontal, vertical or torsional
- **Binocular:**
  - disconjugate & dissociated
- **Uniocular:**
  - Heimann-Bielschowsky Phenomenon
    - Optic pathway glioma (children with onset <2 years)
- **C/o oscillopsia**
- Associated with anterior visual pathway defect or brainstem lesion
Acquired Pendular Nystagmus

- **Visual loss**
  - Aetiology: trauma, retinal or optic nerve disease
  - Large amplitude, often vertical, maybe uniocular

- **Multiple Sclerosis (MS)**
  - Disconjugate & dissociated
  - C/o disabling oscillopsia

- **See-saw nystagmus**
  - Aetiology: lesion of optic chiasm, brainstem disease, advanced retinitis pigmentosa
  - 1 eye rise & intort while the other eye falls & extort
Acquired Pendular Nystagmus

○ Spasmus Nutans
  ● Aetiology unknown
  ● Onset before 18 months old
  ● Triad of nystagmus, head nodding & torticollis
  ● Nystagmus: rapid, pendular dissociated low amp oscillation
  ● Nystagmus may ↓ with head nodding
  ● Resolves clinically by ~5 years

○ Oculopalatal tremor (myoclonus)
  ● Aetiology: pontomedullary infarct/ haemorrhage
  ● Synchronised ocular & palatal oscillations
Acquired Jerk Nystagmus

Slow phase → corrective fast phase

Induced by Vestibular System Disease

1. Peripheral Imbalance
   - Aetiology: disease affecting vestibular organ e.g. labyrinthitis → R-L imbalance
   - Mixed horizontal-torsional nystagmus
   - Slow phase towards affected side
   - Fall towards affected side
   - VOR abnormal/ absent
   - C/o oscillopsia, nausea, vertigo, dizziness
Acquired Jerk Nystagmus

Induced by vestibular system disease

2. Central Imbalance
   - Vestibular imbalance → by cerebellar or medulla lesions

Down-beat Nystagmus
   - Fast phase down in all positions
   - ↑ on down-gaze & often lateral gaze
   - Aetiology: cerebellar degeneration, cerebellar ischaemia, Arnold-Chiari malformation, drug intoxication (e.g. anticonvulsants & lithium)
   - Other mechanisms:
     - Imbalance of vertical smooth-pursuit → spontaneous upward drift
     - Mismatch for vertical saccade generation
Acquired Jerk Nystagmus

2. Central Imbalance
   - Cerebellar or medulla lesions

Up-beat Nystagmus
   - Fast phase up in all positions
   - ↑ on up-gaze
   - Associated with impaired vertical smooth pursuit
   - Aetiology: MS, brainstem tumour/ stroke, Wernicke’s encephalopathy, cerebellar degeneration, drug intoxication
   - Other mechanisms:
     - Imbalance of vertical VOR
     - Mismatch of saccadic generation & velocity-to-position integration
2. Central Imbalance

- Periodic Alternating Nystagmus (PAN)
  - Horizontal jerk nystagmus
  - Periodically reverses direction of fast phase
  - Cycle: range from few seconds to 4 min
  - Diff Diagnosis: congenital vs acquired
  - Aetiology: craniocervical anomalies, cerebellar degeneration/tumour, brainstem infarct, MS, bilateral visual loss
Acquired Jerk Nystagmus

Induced by dysfunction of gaze-holding mechanism

- **Gaze-evoked nystagmus**
  - Nystagmus in eccentric gaze but absent in P.P.
  - Jerk nystagmus with fast phase to the right on right gaze
  - Jerk nystagmus with fast phase to the left on left gaze
  - Upbeat nystagmus in upgaze
  - Downbeat nystagmus in downgaze
- **Aetiology**
  - Cerebellar disease, medication (anticonvulants or sedatives), alcohol
- **Rebound nystagmus**
  - Patients with gaze-evoked nystagmus return eyes to P.P., observe transient nystagmus with slow phase to previous gaze position (*opposite*)
Saccadic Oscillations

- **Saccadic intrusions**
  - Inappropriate saccades that intrude on steady fixation

- **Saccadic oscillations**
  - Dysfunction of saccadic system → saccades take eyes away from target & result in unsteady eye movements

- **Differential Diagnosis**
  - From nystagmus
Saccadic Oscillations

- **Square wave jerks (SWJ’s)**
  - Small conjugate back-to-back saccades
  - Inter-saccadic interval (200 msec)
  - <5° in amp & frequency <10/min
  - May be seen in healthy subjects
  - Aetiology: progressive supranuclear palsy, cerebellar disease, Parkinson’s disease, MS, schizophrenia

- **Macro SWJ’s**
  - >5° in amp
  - Inter-saccadic interval (80 msec)
  - Aetiology: cerebellar disease
Saccadic Oscillations

- **Ocular Flutter**
  - Episodic, horizontal back-to-back saccades
  - No inter-saccadic intervals

- **Opsoclonus (saccodomania)**
  - Back-to-back saccades in all directions
  - Aetiology: infants with neuroblastoma, cerebellar encephalitis, cancer, idiopathic

- **Ocular bobbing**
  - From P.P. fast, conjugate eye movement down
  - After few sec slow drift back to P.P.
  - Aetiology: severe pontine dysfunction
Miscellaneous Oscillations

- **Voluntary Nystagmus**
  - 10 rapid back-to-back saccades
  - May be initiated by convergence
  - Seen in hysterical pt’s or malingers
Self-directed activity

Read p.1-6

Management

- **Aim**
  - Treat any underlying disorder
  - Reduce nystagmus & oscillopsia

- **Options**
  - Medication
  - Prisms
  - Optical stabilisation
  - Surgery
Management

Medication

- **Baclofen**
  - May ↓ down-beat, up-beat, see-saw & PAN

- **Clonazepan**
  - May ↓ down-beat, see-saw & acquired pendular

- **3,4-diaminiopyridine / 4-aminopyridine**
  - Randomised, controlled studies showed successful ↓ in down-beat nystagmus
  - Effective in some with upbeat nystagmus
Management

Medication

- **Gabapentin**
  - Successful ↓ in acquired pendular nystagmus caused by MS, oculopalatal tremor

- **Memantine**
  - Successful ↓ in acquired pendular nystagmus caused by MS, oculopalatal tremor
  - May be effective in upbeat nystagmus

- **Carbamazepine**
  - Successfully ↓ saccadic oscillations in some

- **Immunoglobulins or prednisolone**
  - Occasionally ↓ ocular flutter & opsoclonus
Management

- **Prisms**
  - BDΔ may ↓ oscillopsia in down-beat
  - BOΔ induce convergence & may ↑ VA if nystagmus dampen on near fixation
  - BIΔ may ↓ nystagmus & oscillopsia if symptoms worsen on near fixation

- **Optical Stabilisation**
  - Aim: ↓ effect of eye movement on retinal image
  - Principle: combine high + lenses with high – CL
  - 100% image stabilisation not needed to o/c oscillopsia
Management

○ Botulinum Toxin
  - Aim: ↓amplitude of nystagmus by targeting all EOM
  - Retrobulbar injection (usually the eye with better VA)
  - Result in ↑VA & ↓ c/o oscillopsia
Management and the future

- **Medication**
  - Clinical trials using memantine, gabapentin and 3,4-diaminopyridine/aminopyridine
  - New drugs

- **Surgery**
  - 4 large horizontal rectus recessions
  - 4 large vertical rectus recessions (Spielman, 2009)
  - Vertical Anderson Procedure (bilateral SR recessions & IR tenotomy)
  - Combine Kestenbaum surgery with gabapentin

- **Further research**
  - Understanding of underlying mechanisms
  - Virtual reality
4 large vertical rectus recessions

(Spielman, 2009)
Summary

- Overview
  - Acquired pendular nystagmus
  - Acquired jerk nystagmus
  - Saccadic oscillations

- Management options
  - Medication
  - Prisms
  - Optical stabilisation
  - Surgery
Preparation: nystagmus lectures

- Read handouts
- Read review article