EYES THAT DANCE
AND
EYES THAT CAN’T

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NON-DANCERS

- Thyroid Myopathy
- Ocular Motor Apraxia
- Chronic Progressive External Ophthalmoplegia (CPEO)
- Parkinson’s Disease
- Progressive Supranuclear Palsy (PSP)

NYSTAGMUS BASICS

- Also known as Talantropia (rarely)
- Involuntary, rhythmic to and from oscillation of one or both eyes
- The slow phase is the abnormal movement
- The fast phase is the normal response (corrective)

DEFINITIONS

- Direction: Named by fast component if is one
- If pendular, named by Plane (e.g., horizontal)
- Form: Horizontal, vertical, rotatory
- Amplitude: the width of the swings
- Frequency: the number of oscillations per sec
- Intensity: amplitude X frequency

DEFINITIONS

- Symmetry: whether the waveform and intensity in the two eyes are the same
- Conjugacy: two eyes moving in the same direction (conjugate) or opposite (disconjugate)
- Position of gaze: pattern variation with different ocular gaze positions
- Spontaneity: present under normal viewing or must be induced (OKN, head position)
KEY CONCEPTS

- Oscillopsia: The perception of movement
- Strongly suggests acquired nystagmus
- "Null" point: A position where the nystagmus is reduced in amplitude and frequency, perhaps to stability

PENDULAR NYSTAGMUS

PENDULAR vs Jerk Nystagmus

PENDULAR

Jerk

FORMS OF NYSTAGMUS
INFANTILE (CONGENITAL) NYSTAGMUS

- 80% of all nystagmus cases
- Presents prior to age 6 months
- Beats in direction of gaze
- MAY BE PENDULAR OR JERK IN PRIMARY GAZE
- Jerk in side gaze
- Remains horizontal in upgaze (uniplaner)

ALEXANDER’S LAW

- Nystagmus is greater when looking in the direction of the fast phase
- Usually points to Vestibular cause

INFANTILE NYSTAGMUS

- Usually a null position
- Often a latent component
- Monocular; only when occluded
- OKN responses inverted
- Diagnostic of congenital nystagmus
- Frequently seen with albinism

ADULT “CONGENITAL” NYSTAGMUS

- MNEMONIC: CONGENITAL
  - C - convergence and eye closure dampen
  - O - Oscillopsia absent
  - N - null point possible
  - G - gaze position may change direction of nystagmus
  - E - equal amplitude in both eyes
  - N - Near acuity better than distance (?)
  - I - inverted OKN response
  - T - Torticollis possible if null point
  - A - Abolished in sleep
  - L - Latent nystagmus may be present
Null Point/Nystagmus Blockage

SPASMUS NUTANS

- High frequency, low amplitude nystagmus
- Bilateral but asymmetric
- May be unilateral

Onset 4-18 months; Resolves by 3-4 years of age
Triad: head tilt, head nodding, nystagmus
- Suggested the head nodding may compensate for nystagmus
- Check for chiasmal glioma
- May be associated with 3rd ventricle mass

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LATENT NYSTAGMUS

- One eye is occluded
- Drifts towards nose
- Fast phase away from covered eye
- Associated with strabismus or congenital nystagmus
- Marks condition as congenital

LATENT NYSTAGMUS

- Measure VA with either high plus lens over one eye or angled occluder
- Let light into occluded eye
LATENT NYSTAGMUS

Manifest Latent Nystagmus

- Oxymoron?
- Amblyopia or Suppression - essentially monocular; always “manifest”
- Always congenital, no CNS association

PERIODIC ALTERNATING NYSTAGMUS

- Horizontal
- Direction alternates periodically
- About every 2 minutes
- May briefly be stable before alternating
- Present in primary gaze
- Remains horizontal in upgaze (uniplanar)
- Same characteristics in light and dark
- Smooth pursuit and OKN impaired

SOME CAUSES OF PAN

- Cranio-cervical junction anomalies
- Arnold-Chiari
- MS
- Other causes that affect the cerebellum
- May follow progressive bilateral loss of vision from ocular causes

ARNOLD-CHIARI

- Oscillopsia
- Worse with head movement
- Valsalva induced dizziness, vertigo, HA
- Various types of nystagmus, downbeat, GEN, PAN
DOWNBEAT NYSTAGMUS

- Present in primary gaze
- Amplitude increases in gaze down and to the side
- Impaired pursuits in down gaze
- Convergence may increase, decrease, or convert to upbeat nystagmus

Some Causes Of Downbeat Nystagmus

- “Down” in the Brainstem
  - Lesions at cervico-medullary junction
  - MS
  - Arnold-Chiari malformation
  - Hydrocephalus
  - Drug intoxication: antidepressants, lithium
  - Spinocerebellar ataxias

UPBEAT NYSTAGMUS

- Present in primary gaze
- Not affected in lateral gaze
- Accentuated with up gaze
- May convert to downbeat with convergence
Some Causes Of Upbeat Nystagmus

- Diffuse or multifocal brainstem damage
- Anterior cerebellar vermis
- Infarct in medulla or superior cerebellar peduncle
- Wernicke’s encephalopathy
- MS
- Leber’s
- Transient in infants

TORSIONAL OR ROTARY NYSTAGMUS

- Best observed viewing conjunctival vessels
- Not suppressed by visual fixation, but may be by convergence
- May result from problem with central vestibular pathway

ROTARY/TORSIONAL NYSTAGMUS

- Arnold-Chiari
- Wallenberg Syndrome
- MS
- Brainstem tumor
- A-V malformation in cerebellar peduncle
- Congenital

Some Causes Of Torsional Nystagmus

Rebound Nystagmus

- Transient, rapid horizontal jerk
- Initiated by eccentric gaze
- First beats in direction of gaze, then reverses upon relocation in primary gaze
- Associated with cerebellar or posterior fossa lesions

Rebound Nystagmus
**Dissociated (Disconjugate) Nystagmus**

- Eyes different in amplitude
- Abducting nystagmus in INO
- May be prudent to give note for police
- Posterior fossa lesions

**Bino**

**Convergence-Retraction Nystagmus**

- Attempted upgaze yields jerk convergence and retraction
- Best seen with OKN
- Patients may adopt chin-up position
- Hallmark of Parinaud’s Syndrome
- Often pinealoma

**Convergence-Retraction Nystagmus**

**Age-Related Differential Diagnosis of Dorsal Midbrain Syndrome**

- Infant = congenital aqueductal stenosis
- 10 yr old = pinealoma
- 20 yr old = head trauma
- 50 yr old = brainstem AVM
- 40 yr old = multiple sclerosis
- 50 yr old = vertebrobasilar CVA

**Seesaw Nystagmus**

- Disjunctive vertical nystagmus
- One up, one down - alternating
- Pendular torsional nystagmus
- Elevation/intorsion, depression/extort
- May also have hemi-seesaw
- May be pendular or jerk
- May be congenital or central cause
**SEESAW NYSTAGMUS**

- Anterior tumors
- Often large parasellar tumors extending into 3rd ventricle
- Bitemporal hemianopsia
- Trauma
- May also occur with primary vision loss like RP

**VESTIBULAR NYSTAGMUS**

- Most probable cause of acquired, horizontal jerk nystagmus in primary gazes
- May be unilateral or bilateral vestibular loss
- Dysfunction of vestibular end organ, vestibular nerve or vestibular nuclear complex
- Unidirectional
- Invariable rotary component if peripheral
- Fast phase beats away from damaged end organ
- Usually associated with vertigo

**VESTIBULAR DYSFUNCTION**

- Peripheral (inner ear)
  - Recurring periods of vertigo
  - Always nystagmus
- Central (brainstem/cerebellar)
  - Damage to the vestibular nuclei or connections
  - Milder symptoms than peripheral

See Saw Nystagmus

Lesions in the vestibular pathway simulate head movement - results in vestibular nystagmus
PERIPHERAL VS CENTRAL

Peripheral
- Nystagmus is away from lesion
- Romberg - falls to side of lesion

Central
- Nystagmus direction may change with change in gaze
- Romberg fall does not vary with head position

HEAD SHAKING TO INDUCE NYSTAGMUS

VESTIBULAR NYSTAGMUS

Benign Paroxysmal Positional Vertigo (BPPV)
- True vertigo, always positional
- Lying down, rolling over, getting up, bending over, looking up
- Short duration - seconds
- No associated hearing loss
- Head trauma, post labyrinthitis, elderly
- Positive Hallpike maneuver

HALLPIKE MANEUVER
**BPPV POSTERIOR CANAL**

**BPPV Treatment**
- Time
- Particle repositioning maneuvers - Epley
- Cawthorne (habituation) exercises
- Occasionally medication
- Rarely surgery
- Rarely disabling (safety issues)

**EPLEY MANEUVER**

**VESTIBULO-OCULAR REFLEX (VOR)**

**VOR**
- Coordination of head movement and eye movement
- Involuntary
- Allows continued fixation with head movement
- Moves eyes in direction opposite to head
- Slow conjugate pursuit
- Fast, Corrective Movements up to 400-700°/Sec

**VOR GENERATION**

[Diagram of VOR generation process]
Potential Use For Testing

- Test of dynamic visual acuity
  - Normal subjects lose 1 line
  - Decreased vestibular function lose 5
  - Oscillopsia - brought on or increased by head motion

CLINICAL VOR

SUPPRESS VOR

INABILITY TO SUPPRESS VOR
LID NYSTAGMUS

- Yoked innervation of lid and eye elevators

MYASTHENIA AND LID NYSTAGMUS

VOLUNTARY NYSTAGMUS

- A "functional" condition
- Up to 8% of the population!
  - Often familial
- Usually tied with convergence
- High frequency, low amplitude
- All saccades - not true nystagmus

GAZE EVOKED NYSTAGMUS

- End point nystagmus common, poorly sustained
- Usually horizontal on upgaze (uniplanar)
- May be physiologic or pathologic
- "Endpoint nystagmus" is physiologic
- Remember: may need note for police
True GEN

- Greater amplitude
- Less eccentricity needed to elicit
- Jerk in direction of gaze (upbeat with upgaze, etc.)
- May result from brainstem or cerebellar disease
- Medications common (anticonvulsants, sedatives)

GEN

GEN - MS

TEST YOURSELF

- What type of eye movement or lack of movement is present?
- What condition may be associated?
- What else can you predict about the vision function or course of the causative condition?

CASE 1

CASE 2
SACCADIC INTRUSIONS

- Inappropriate, spontaneous, involuntary saccadic eye movements that interfere with steady fixation
- Rapid and brief
- Differentiate from saccadic dysmetria and nystagmus
SACCADIC INTRUSIONS

- Continuum of saccadic oscillations with or without an intersaccadic interval
- If horizontal only = ocular flutter
- If multidirectional = opsoclonus

OPSOCLONUS

- Involuntary bursts of saccades in all directions
- "Saccadomania"
- Often large amplitude - interferes with fixation, pursuits, or convergence
- Often associated with cerebellar signs

Other Causes

- Young adults - follows fever
- Children - parainfection or paraneoplastic - "Dancing Eyes, Dancing Feet"
- Opsoclonus-myoclonus syndrome
- Adults - visceral carcinoma

OCULAR FLUTTER

- Conjugate saccades in horizontal plane
- May be spectrum of opsoclonus
- Usually intermittent (3-5)
- Associated with voluntary saccades or fixation effort
- Common companion of dysmetria
- Also associated with cerebellar signs
Macrosaccadic Oscillations

SQUARE WAVE JERKS
- Small saccades away from fixation
- Return in 200 msec
- Patient may note "jumping"
- May be seen in normal patients
- 6-24%
- More prominent during pursuits
- Obvious during ophthalmoscopy

Pathological in cerebellar and basal ganglia disease, also cerebral
- PSP, Alzheimer’s
- Increased frequency compared to normals
- Est. 70% of cases closed head injury
- Sq wave jerks greater than 1 degree are pathological
- Cigarette smoking increases frequency

Square Wave Jerk Waveform

SQUARE WAVE JERK
SUPERIOR OBLIQUE MYOKYMIA

- Microtremor
- Jiggling or transient vertical/torsional diplopia
- Easily observed with slit lamp
- Monocular oscillopsia
- Precipitated by return from downgaze to primary
- Most cases are spontaneous, normal imaging

SO MYOKYMIA

MEDICAL TREATMENT OPTIONS

<table>
<thead>
<tr>
<th>Type</th>
<th>Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAN</td>
<td>baclofen</td>
</tr>
<tr>
<td>Pendular</td>
<td>gabapentin, memantine</td>
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<tr>
<td>Episodic</td>
<td>acetazolamide</td>
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<tr>
<td>Downbeat</td>
<td>baclofen, clonazepam, acetazolamide</td>
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<tr>
<td>Upbeat</td>
<td>baclofen</td>
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<tr>
<td>SO myokymia</td>
<td>gabapentin, carbamazepin</td>
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</tbody>
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OTHER TREATMENT OPTIONS

- Base-out prism if null point with convergence, may reduce nystagmus
- Rigid contact lenses
- Vision therapy / biofeedback
- BotTox

SURGICAL TREATMENT OPTIONS

- Kestenbaum - null zone shift
- Artificial Divergence Procedure
- Four Muscle Tenotomy
TEST CASE

EYES THAT CAN'T DANCE

- Thyroid Myopathy
- Ocular Motor Apraxia
- Chronic Progressive External Ophthalmoplegia (CPEO)
- Parkinson's Disease
- Progressive Supranuclear Palsy (PSP)

THYROID MYOPATHY

- Muscles usually affected in order: IR, MR, SR, LR
- Effect on IOP
- Restriction may be mistaken for paresis of ipsilateral antagonist
- Note extorsion on abduction

OCULAR MOTOR APRAXIA

- Deficiency of voluntary horizontal eye movements
- Vertical intact
- Head thrust uses VOR
- Congenital
- May be agenesis of corpus callosum
CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA (CPEO)

- Age of onset varies
  - Often early childhood
- Slowly progressive bilateral ptosis
- All EOMs affected
  - Reduced saccadic velocity
- Downgaze LEAST affected

CPEO

- Mitochondrial disease
- Rarely diplopia
- Kearns-Sayre Syndrome
  - Also has retinal and heart abnormalities

PARKINSON’S DISEASE

- Death of dopaminergic neurons in substantia nigra and pedunculopontine nucleus
- Hypokinetic disorder
- Rigidity, difficulty initiating movements, turning, stopping; resting tremor; stooped posture
- Onset chronic, ages 50-65, men > women,
  - Lifespan post-diagnosis 13 years
PARKINSON'S OCULAR SIGNS

- Fixation interrupted by square wave jerks
- Hypometric horizontal and vertical saccades
  - Starts with normal saccadic velocity
- Impaired smooth pursuit and convergence
- Intact vestibulo-ocular reflex

PROGRESSIVE SUPRANUCLEAR PALSY (PSP)

- Usually after age 50
- Degenerative, progressive
- Parkinson-like symptoms
- Dementia
- Supranuclear vertical gaze palsy
  - Especially downgaze

PSP APRAXIA LID OPENING

- Initial loss of vertical saccades and convergence
  - Vertical saccades slow, decreased range, slow initiation
- Difficulty with DOWNGAZE
  - Walking, seeing food on plate, reading
- Eventually all saccades and pursuits compromised
- Unable to use visual info to cancel VOR
- Apraxia of lid opening

PARKINSON'S

- Tendency to fall backward
- Lack of response to L-dopa
- Estimated 12% of Parkinsonian patients have PSP
- Median survival rate 5 to 7 years
COURSE GOALS

- Recognize and accurately describe the covered oculomotor conditions
- Raise the clinicians’ index of suspicion regarding the underlying cause of the observed eye movements
- This is a course in DIAGNOSIS (brief mention of management)