Ptosis and Pupils

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How to Examine the Nervous System

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Ptosis

- Bilateral, longstanding, familial:
  - CPEO (chronic progressive external ophthalmoplegia)
- D/D: Acute, recent, unilateral

Palpebral fissure

- Palpebral fissure
  - 7–12 mm in the vertical midline
- Cornea
  - about 10.5 mm in vertical diameter
  - upper lid usually covers the top 0.5–1 mm
  - bottom lid touches the lower limbus

Blepharospasm or ptosis?

- Blepharospasm
  - eye closure resulting from a contraction of the orbicularis oculi muscle
- Ptosis
  - partial or complete eye closure caused by paresis or paralysis of
    - levator palpebrae muscle or
    - superior tarsal muscle

Blepharospasm

- May be a result of some painful or irritating ocular disease
- May be voluntary to abolish the false image of diplopia
- May be a dystonia (involuntary movement because of organic disease), in this case always bilateral
**Blepharospasm or ptosis**
- **Blepharospasm**
  - Eyebrow is pulled down below the superior orbital margin
- **Ptosis**
  - Eyebrow is on the margin or above
  - If above, the forehead is wrinkled as the patient uses his frontalis to compensate for the drooping lid

**Ptosis: CN3 palsy or Horner’s?**
- **CN3 palsy**
  - Big pupil on the side of the ptosis, with or without the appropriate EOM palsies
  - Complete ptosis
  - Ptosis does not change on upward gaze
- **Horner’s syndrome**
  - Small pupil on the same side
  - Incomplete ptosis
  - Ptosis will diminish on upward gaze
  - The lower lid is higher than the normal lower lid (covers more of the limbus at 6 o’clock, due to paresis of the inferior tarsal muscle)

**Horner syndrome**
- Refers to a constellation of signs produced when sympathetic innervation to the eye is interrupted
  1. Miosis
  2. Dilation lag
  3. Mild-to-moderate ptosis*
  4. Slight elevation of the lower lid (upside-down ptosis or reverse ptosis*)
     * 3 & 4 are due to denervation of the sympathetically controlled Müller muscle
  5. Impaired flushing ipsilaterally
  6. Impaired sweating (anhidrosis) ipsilaterally

**Bilateral ptosis following stroke**
- Bilateral ptosis has been seen following basilar artery infarction and ischemia
- There are no other signs of either third nerve or sympathetic lesions
  - Pupils and eye movements were normal
  - Patients were alert
The Pupils

Prime physical signs
- Pupil size
- Pupil equality between right and left
- Pupil responses to various stimuli
- Conscious vs. comatose

Pupil size (1)
- 2 to 5 mm in diameter
- Depends on
  - Light
  - Near reaction
  - Sympathetic tone
  - Parasympathetic tone

Pupil size (2)
- Smaller pupils
  - Newborn and elderly (than youths)
  - Bigger pupils
    - Schizophrenic patients and frightened people
    - Blue-eyed people (than brown-eyed)
    - Myopic people
  - Pupils in young people
    - Normally constrict and dilate in a repetitive rhythmic way (hippus); its absence is also normal

Pupil equality
- 15 to 20% of patients have anisocoria that cannot be explained, but the difference is usually not more than 1 mm
- Normal inequality:
  - the difference in size remains constant in both bright and dim illumination
- Pathologic inequality:
  - the difference in size changes with the illumination

Anisocoria
- Anisocoria that changes with illumination
  - One should measure pupil sizes in average room light, dimness, and bright light
- Horner’s syndrome
  - the small pupil will not dilate in dim light as much as the pupil of the other normal eye
- CN3 palsy
  - the larger pupil will not constrict as much as the normal pupil in bright light
Horner’s syndrome

- The small pupil of will not dilate in dim light as much as the pupil of the other normal eye

CN3 palsy

- The larger pupil will not constrict as much as the normal pupil in bright light
- All patients with a new-onset CN3 palsy with pupillary involvement should be presumed to have a cerebral artery aneurysm until proved otherwise => require an expeditious MRI angiogram or four-vessel angiography (emergently if a headache is also present)

Pupil equality

- The followings do not cause anisocoria
  - Unilateral or bilateral diminished or absent visual acuity or peripheral visual field defects
  - Differences in refractive errors between the two eyes

How to examine the Pupils

Light Reaction

- Sitting, moderate background illumination
- Flashlight held vertically, pointing up, below the eye, and just in front of the cheek
- Ask patient to look at the far, illuminate each eye in isolation
- Afferent: CN2
- Efferent: CN3

Direct / consensual light reaction

- Direct
  - Ipsilateral pupillary constriction in response to bright light exposure
- Consensual
  - Contralateral pupillary constriction in response to bright light exposure
Abnormal light reaction
- Q: Lesion at?
  1. Right eye has a normal direct response and an abnormal consensual response
  2. Right eye has an abnormal direct response and a normal consensual response
  3. Right eye has neither a direct nor a consensual response

Near reaction
- Light reflex ok → near reflex ok (test can be skipped)
- Near reflex: ask patient to look at the far wall and then at the tip of your pencil or tip of his own finger (5 to 10 cm in front nose)
- Look for two things:
  - bilateral constriction of pupils
  - convergence eyes
- Sluggish direct light response but brisk near response → lesion at right afferent limb of light reflex

Abnormal pupils
- Marcus Gunn pupil
- Argyll-Robertson pupils
- Horner’s syndrome
- Oculomotor nerve lesion (unilateral)
- Adie’s syndrome
- Benign anisocoria
- The factitious big pupil
- Parinaud’s syndrome
- Carotid artery occlusion (unilateral)
- Pontine miosis (bilateral)

Marcus Gunn Pupil
- Implies afferent pupil defect, e.g. retrobulbar neuritis of optic nerve
- Swinging flashlight test (e.g. right afferent defect):
  - Examine his pupil responses in dim illumination
  - Shine strong light into left eye → both pupils constrict
  - Shine into right eye → right pupil seems to constrict momentarily and then dilates widely, as does the left
  - Swing light back to left eye → both constrict
- Defected pupil:
  - normal consensual light reaction → constriction
  - reduced direct light reaction → relative dilatation

Argyll-Robertson pupils
- Result from
  - Tertiary syphilis of the nervous system
  - Diabetes
  - Late signs of bilateral tonic pupils
- Pupils (usually bilateral)
  - small, irregular, and unequal
  - light response (may be absent) much less evident than near response
  - do not dilate in the dark
  - respond poorly to mydriatics
  - can be made to constrict even more by miotics
- Visual acuity not impaired

Horner’s syndrome
- AKA oculosympathetic palsy (unilateral)
  - lesion of the sympathetic fibers can be in the brain stem, cervical cord, apex of the lung, carotid sheath, or orbit
- Pupil
  - small and round
  - good response to light and near
  - anisocoria more prominent in the dark (affected pupil dilates later and less)
- Additional signs
  - ptosis (incomplete)
  - apparent enophthalmos
  - warm, dry, nonsweating, ipsilateral face
- Painful Horner’s syndrome
  - carotid artery dissection
  - cluster headaches (normal carotid artery)
Oculomotor nerve lesion (unilateral)

- Structural oculomotor nerve lesion
  - Ipsilateral mid-dilated pupil
  - Pupil not respond to light or near
  - Difference in pupil size is greater in the light (in contrast with Horner’s syndrome)
  - Mydriatics and miotics are both effective
- Diabetic oculomotor nerve palsies
  - Usually have normal pupils
  - Can be painful

Adie’s syndrome

- Adie’s syndrome, or tonic pupil or “the big, slow pupil”
  - Enlarged pupil may be unilateral or asymmetrically bilateral
  - Does not react to light or eventually constricts after being exposed to very bright light for 15-20 min
  - Eventually constricts for near after a similarly long effort
  - Reactivation is just as protracted
  - Difference in the pupil sizes is best seen in the light
  - Pupils respond to mydriatics and miotics and demonstrate denervation supersensitivity (pupil will constrict from 2.5% solution of Mecholyl or 0.125% pilocarpine)
  - Accommodation is slow and may be the presenting complaint
- Holmes-Adie syndrome
  - Adie’s synd + absent / diminished deep tendon reflexes
  - Cause is unknown

Benign anisocoria

- Usually a young adult
- Chronic → less important
- Examine old photographs
- Response to light and near in both eyes is normal
- Difference in pupil size is no greater in dimness or light
- Mydriatics and miotics have a normal response, and there is no diagnosis

The factitious big pupil

- Causes
  - Eyedrops containing impurities with atropine-like properties
  - Ointments with atropine-like properties inadvertently introduced into eye
  - Occasionally a deliberate atropine abuser
- S/S
  - The biggest pupil you have ever seen
  - No response to light or near
  - Difference greatest in bright light
  - Neither mydriatics nor miotics change the pupil
  - (Pilocarpine will not constrict an atropinized pupil; it will, however, constrict an Adie’s pupil)
  - Reexamine the eye daily for 3 consecutive days

Parinaud's syndrome

- Parinaud's syndrome, also known as dorsal midbrain syndrome
  - Paralysis of upgaze
  - Accommodative paresis
  - Pupils mid-dilated with light-near dissociation
  - Convergence-Retraction nystagmus
  - Eyelid retraction (Collier’s sign)
  - Conjugate down gaze: "setting-sun sign"

Carotid artery occlusion (unilateral)

- An enlarged pupil ipsilateral to the occlusion has been reported in atheroma and Takayasu’s disease
- The pupil reacts poorly to light (direct and indirect) and near
- The explanation is probably ischemic atrophy of the iris, rather than nerve disease
Pontine miosis (bilateral)
- The classic sign of pontine infarction or hemorrhage is small (1 to 1.5 mm) pupils
- They will constrict to light if a bright enough stimulus is used and if examined through a magnifying glass

Accommodation & convergence
- For near vision
  - the eyes converge (ie, turn toward the midline),
  - the pupils constrict, and
  - the lenses thicken
- Convergence: simultaneous contractions of the two medial recti
- Failing accommodation is most commonly related to aging, as the lens becomes less resilient

Accommodation & convergence
- Complete peripheral CN3 palsy: parasympathetic nerve fibers subserving accommodation, as well as those subserving the pupillary near response, will be interrupted
- The diabetic, out of control or of recent onset, can have a sudden improvement in near vision if symptom reversed
- Anticholinergic drugs commonly produce a complaint of blurred vision from diminished accommodation if taken in large enough doses

Near reaction
- Sudden onset of diplopia, with eyes divergent
- Full monocular eye movements
- E.g. periaqueduct syndrome
- Convergence paresis
- Patient staring at the tip of his nose
- Looks like unilateral or bilateral lateral rectus palsy
- Often a hysterical disease - when you ask the patient to look laterally, which he says he cannot do, his pupils constrict, proving that he is overconverging
- Can be organic following head injury and can be part of the periaqueduct syndrome

Near reaction
- Toxin of Clostridium botulinum
  - large, nonreacting pupils
  - paralysis of accommodation
  - Ptosis
  - extracocular muscle palsies
  - patient awake, with progressing respiratory distress
  - may have vomited but usually constipated
- Diphtheria
  - paralysis of accommodation
  - affects bulbar originating nerves and cardiac rhythm
Myasthenia gravis

- Ocular complaints often diagnosed late
- Weakness of one muscle or one eye or any combination of muscles
- The essence of the disease is excessive fatigability, that is, the patient cannot sustain upward gaze or sustain the upper lids in a fixed open position
- Of all the possible combinations of myasthenic muscle weakness (eg, ocular, pharyngeal, or limb), bilateral fluctuating undulating ptosis is probably the most common

Myasthenia gravis

- When the eye opening muscles are weak and the eye closing muscles are also weak, the diagnosis is almost always either MG or one of the CPEOs
- Consider MG when a young woman or an old man says,
  - "My eyelids are drooping" or
  - "I see double when I’m watching the late news on TV"

CPEO

- **Chronic Progressive External Ophthalmoplegia**
- Progressive, very chronic
- Restricted range of EOM and ptosis
- Usually do not cause diplopia
- May have ptosis only (patient may unaware)
- Most family members look the same
- CPEO is actually a number of diseases
- May be associated with oculopharyngeal dystrophy, myotonic dystrophy, thyroid ocular myopathy
Thanks for your attention