



**The Pupil**

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**Disclosure Statement:**

**The Pupil**

**Leonard V. Messner, OD**

- King Devick Technologies (scientific advisory board)
- Heidelberg Engineering (scientific advisory board)

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**"The evaluation of pupils is vastly  
overrated...unless it's not."**

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**“Optometry students both under and overcall pupillary abnormalities with equal frequency and exuberance.”**

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### Key Points

- Pupil testing
- Common pathologic pupillary abnormalities:
  - Afferent pupillary defects
  - Tonic pupils
  - Horner syndrome
  - CNIII palsy

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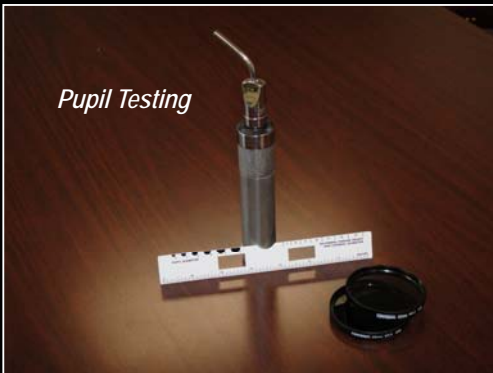
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### Modified Upgaze Technique for Pupil Examination

- Instruct the patient to "look up" at a point on the ceiling (approx 40 degrees above the midline)
- Introduce the light source toward the lower limbus
- Advantages:
  - Reduction of blink reflex (engagement of levator)
  - Pupil is not obscured by the Purkinje-1 image
  - Magnification of pupil

Hsu JL, et al. J Neuro-ophthalmol 2010

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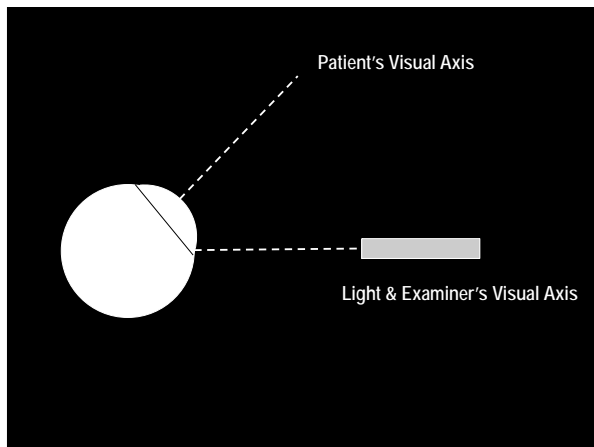
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### The Relative Afferent Pupillary Defect (Marcus Gunn Pupil)

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## Epidemiology and Pathology

- Damage to afferent visual system
- Asymmetric disease
- No anisocoria
- *Relatively unaffected* by media opacities
- *Signature of optic neuropathy*

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## The Swinging Flashlight Test

H. Stanley Thompson - 1976

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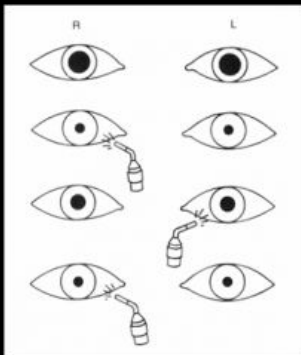
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Thompson HS. Arch Ophthalmol 1976

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The consensual response induced by the "good" eye is greater than the direct response produced by the "diseased" eye.

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28 y/o woman

- Acute vision loss OS
- Pain on eye movement OS
- BVA:
  - 20/20 OD
  - 20/400 OS
- Afferent pupillary defect OS

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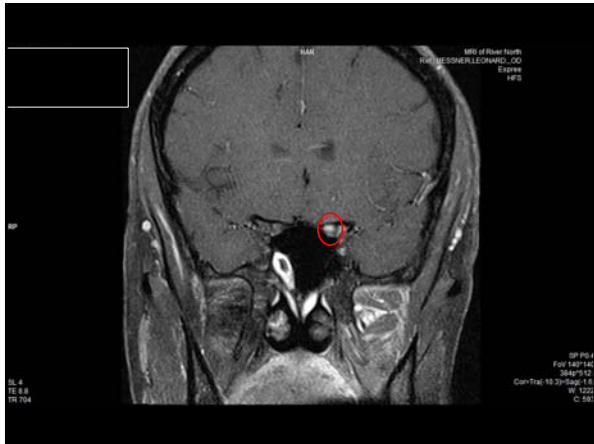
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### Etiology of Afferent Pupillary Defect

- Signature of unilateral/asymmetric optic neuropathy
  - Demyelination/optic neuritis
  - Infarction/ischemic optic neuropathy
  - Compressive optic neuropathy
  - Traumatic optic neuropathy
  - Glaucoma (asymmetric)
  - Papilledema (asymmetric)
  - Retinal disease
    - Central retinal artery occlusion
    - Central retinal vein occlusion (non-perfused)

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### Polling Question #1

What is the most common cause of an afferent pupillary defect?

- a. Optic neuropathy
- b. Macular degeneration
- c. Cataract
- d. Diabetic retinopathy

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# Tonic Pupils

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- ## Causes of Tonic Pupil
- local (orbit)
  - infection
  - inflammation
  - ischemia
  - tumor
  - anesthesia (R-B block)
  - s/p surgery
  - toxicity (quinine)
  - s/p laser
  - trauma
  - neuropathic (diabetes)
  - s/p CN III palsy
  - *Adie's syndrome*
- Loewenfeld - 1967 / Thompson - 1979

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## Adie's Syndrome

• Prevalence	2/1000
• Mean age	32
• Female/Male	2.6:1
• Reduced deep tendons	89%
• Sector palsy	100%
• Accommodative paresis	66%
• Denervation supersensitivity	80%
• Decreased corneal sensitivity	90%
• Unilateral	80%
• Bilateral	4%/yr (miosis)

Lee & Braze, 1998

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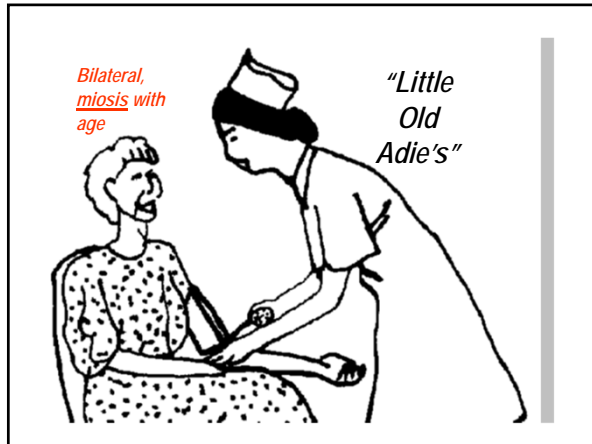
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### Clinical Features of Adie's Tonic Pupil

- "Flat" edges
- "Vermiform" iris movement
- Poor response to light & near or LND
- "Dilation lag" following prolonged near effort
- "Paradoxical Pupil" - anisocoria greater in light & dim



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### 28 y/O Woman

- Anisocoria that reverses from bright to dim illumination
  - Right pupil larger in light and smaller in dim



Pupil testing

Testing for corneal sensitivity

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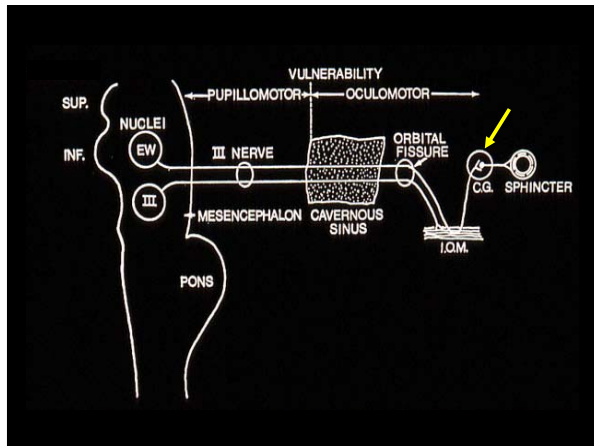
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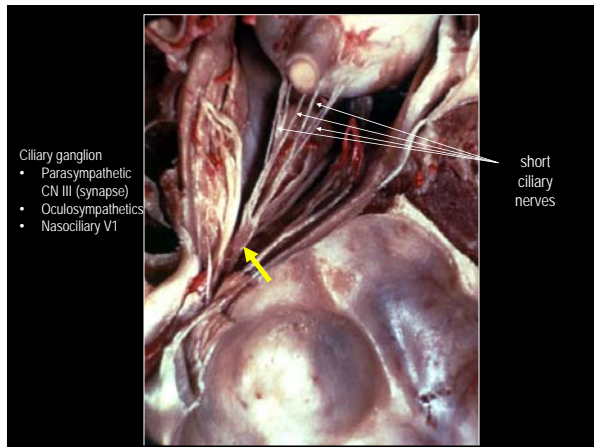
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### Pathogenesis of Adie's Tonic Pupil

- Ciliary ganglion
  - 90% CB
  - 3% iris
- Aberrant regeneration of CB fibers to iris sphincter (light-near/gaze pupil dissociation)

Adie WJ. Brain 1932

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### Pharmacologic Testing for Adie's Tonic Pupil

- Weak (1/8 or 1/10) pilocarpine
- Miosis owing to "denervation supersensitivity"
- *Acquired phenomenon*

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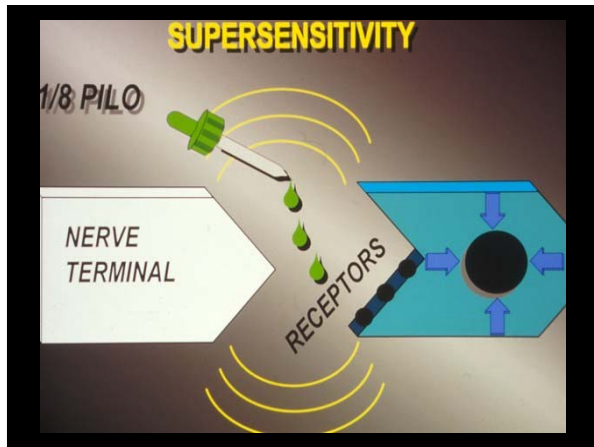
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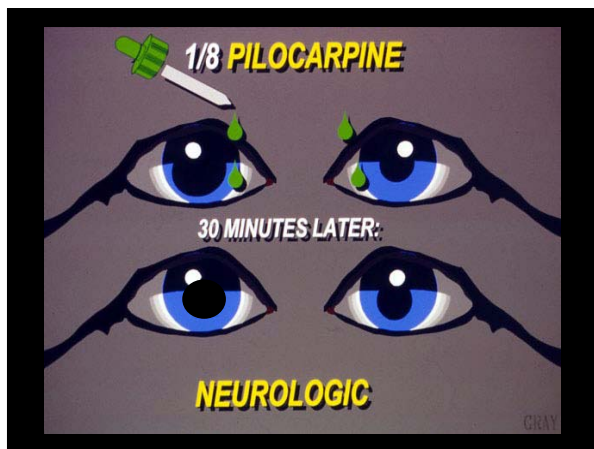
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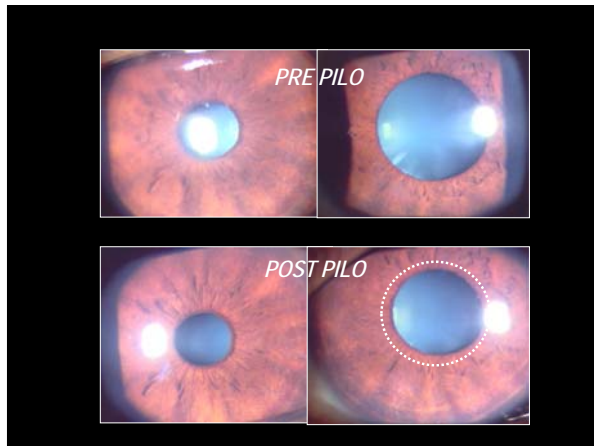
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### Management of Adie's Tonic Pupil

- Cholinergic maintenance therapy
- Reading glasses
- Reassurance

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### Adie's "Masquerades"

- Syphilis
- Giant-cell Arteritis

*FTA-Abs & ESR/C-reactive protein for atypical cases!!!*

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### 72 y/o Woman

- Recent observation of tonic pupils
- BVA:
  - 20/30 OD
  - 20/20 OS



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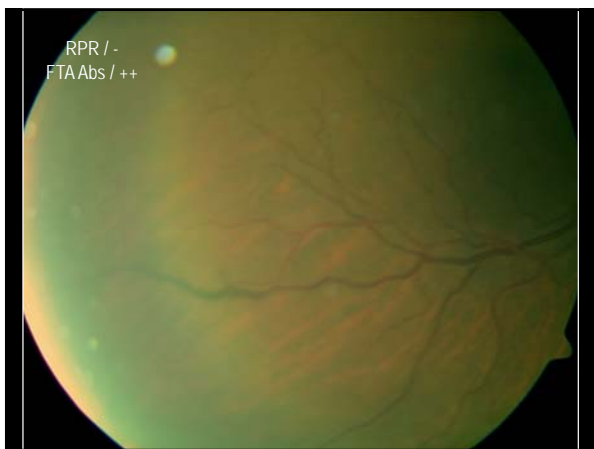
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### Polling Question #2

What is the anatomical location of the lesion responsible for Adie's tonic pupil?

- a. CN III nucleus
- b. Cavernous sinus
- c. Ciliary ganglion
- d. Trigeminal ganglion

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### Oculosympathetic Paresis (Horner Syndrome)

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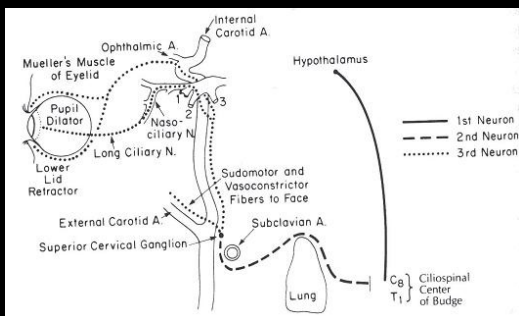
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### Efferent Oculosympathetic Pathways



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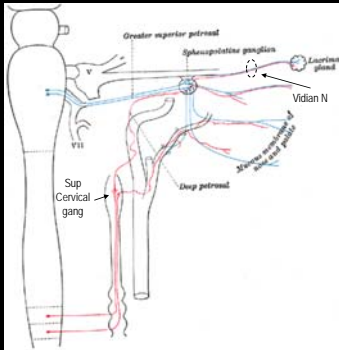
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Sympathetic (deep petrosal N) & parasympathetic (greater superficial petrosal N) from sphenopalatine ganglion to lacrimal gland (Vidian N)



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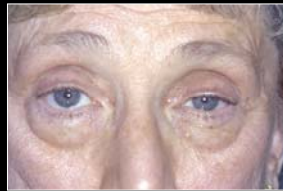
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### Clinical Features of Horner Syndrome

- Ptosis
- "Pseudo" enophthalmos
- Miosis
- Dilation lag
- Heterochromia irides (congenital/pediatric)
- Decreased lacrimation (Vidian nerve)
- Hemi-facial anhidrosis
  - preganglionic



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### Oculosympathetic Lesions

1. First order neuron (central)
2. Second order neuron (pre-ganglionic)
3. Third order neuron (post-ganglionic)

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### Common Causes of Horner Syndrome (Neuron & Frequency)

	Common	Uncommon
First-order (central) neuron (10-15%)	Lateral medullary stroke Spinal cord lesion	Hypothalamic, midbrain, or pontine injury
Second-order (preganglionic) neuron (40-45%)	Pancoast tumor Brachial plexus injury Iatrogenic trauma Neuroblastoma	Cervical disc disease
Third-order (postganglionic) neuron (40-45%)	Carotid dissection Carotid thrombosis Cluster headache Cavernous sinus lesion "Small vessel ischemia"	Intraoral trauma

Liu G. *ANNALS* 2015

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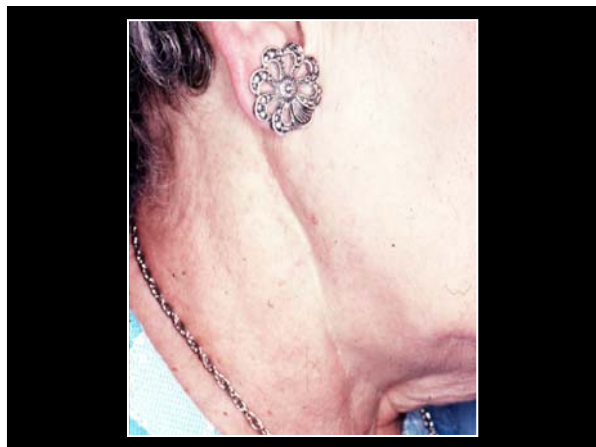
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**37 y/o man**

- Hx of recent auto accident with whiplash injury
- Transient monocular blindness, OD
- Right side neck pain with intracranial noise

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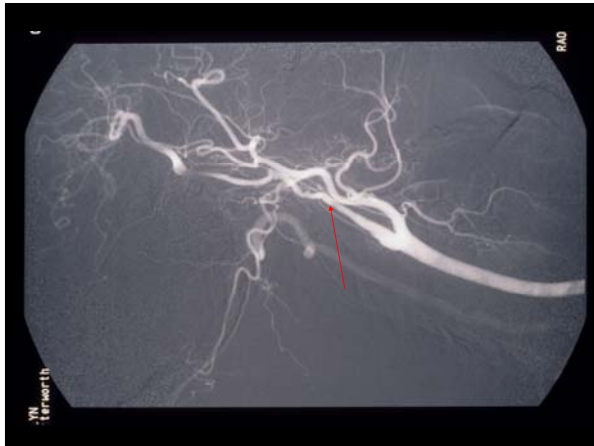
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### Emory University Longitudinal Assessment of 146 Cases of Carotid Artery Dissection (1972-1997)

- 91/146 (62%): eye signs/symptoms
- 76/146 (52%): eye signs/symptoms as presenting feature
- 65/146 (44%): painful Horner syndrome
- 41/146 (28%): TMB (31 with pain)
- 27/76 (36%) with initial presenting eye signs suffered a retinal or hemispheric stroke (average 6.2 days)

Blouise V, et al. Am J Ophthalmol 1998

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### Pharmacologic Testing for Horner Syndrome



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### 0.5% Apraclonidine in the diagnosis of Horner's Syndrome

- Weak alpha-1 agonist /strong alpha-2 agonist
- No dilation / constriction of normal pupil (alpha-2 agonist)
- Dilation of Horner's pupil (alpha-1 agonist - adrenergic denervation supersensitivity)
- "reversal of anisocoria"
- Contraindicated in kids < 2 yrs.

Morales J, et al. Arch Ophthalmol 2000

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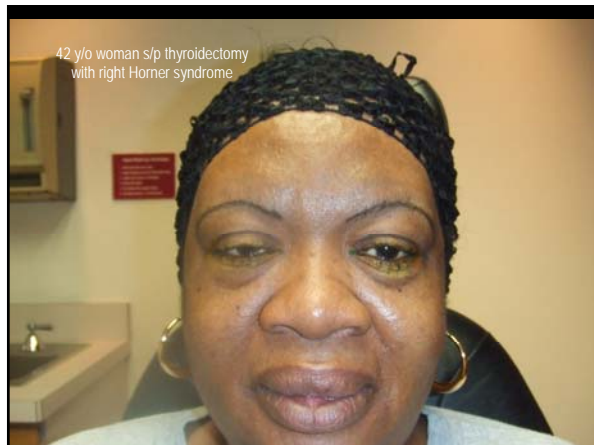
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### Work-up & Imaging for Horner Syndrome

- Adults:
  - If known cause / long-standing: periodic observation
  - If <1 yr. without known etiology:
    - MRI of head, neck, chest
    - MRA of carotids and intracranial vessels

Liu G. NANOS 2015

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### Work-up & Imaging for Horner Syndrome

- Kids:
  - Rule out neuroblastoma (< 24 months)
  - MRI of head, neck, chest
  - VMA-HVA levels (spot urine testing)

Liu G. NANOS 2015

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Infant with Opsoclonus from Neuroblastoma



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### Polling Question #3

All of the following are clinical features of Horner syndrome EXCEPT:

- a. Pupillary miosis
- b. Upper lid ptosis
- c. Dilation lag
- d. Anisocoria greater in bright vs. dim illumination

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### Third Nerve Palsy

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### Signature of CN III Palsy

- Hyperdeviation which increases in up-gaze and reverses in down-gaze
- Exo which increases across from the vertically limited eye

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### Pupil Involved CN III Palsy

- 52 y/o man
- Sudden onset painful diplopia
  - Horizontal & vertical
  - Distance & near



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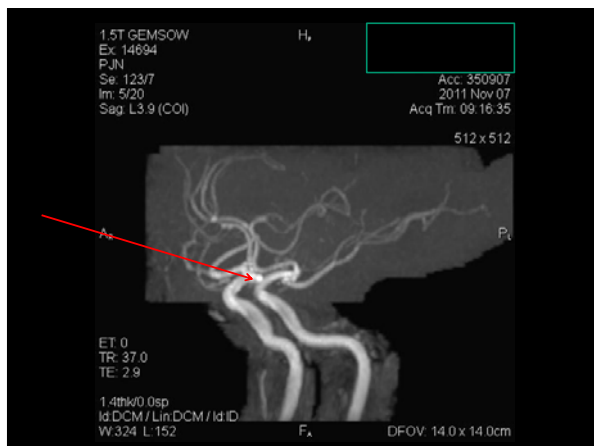
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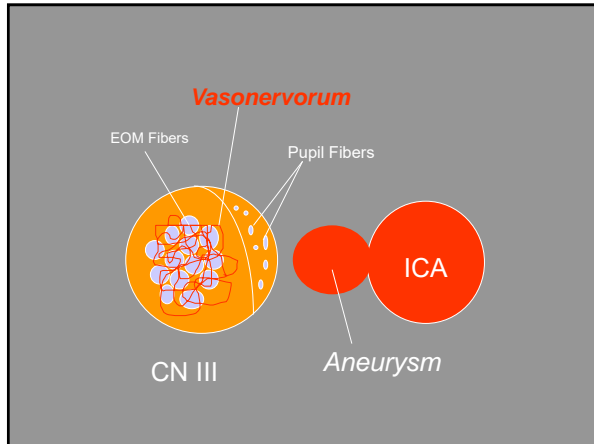
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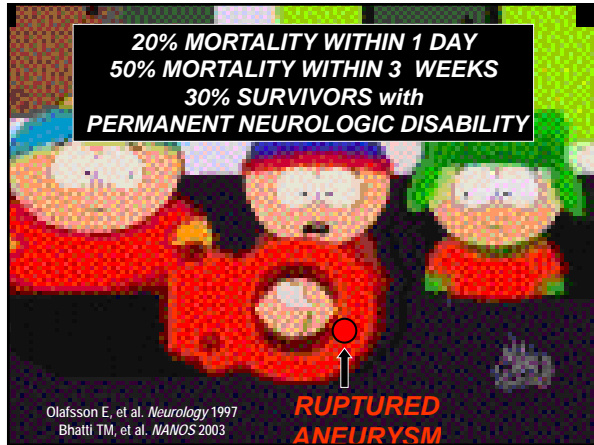
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**"Rule of the Pupil"**

	Pupil Involved	Pupil Spared
<b>Aneurysm</b>	86%	14%
<b>Ischemic / Vascular</b>	23%	77%

Kissel JT, et al. *Ann Neurol* 1983  
Goldstein JE, et al. *Arch Ophthalmol* 1960

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## Neuroimaging for CNIII Palsy (Pupil-Spared)

- MRI / CT of brain
- Cerebral angiography
  - CT Angiography
  - MR Angiography

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## Aberrant Regeneration of CN III Palsy

1. Eyelid synkinesia
2. Pseudo Graefe sign
3. Light-near dissociated pupil



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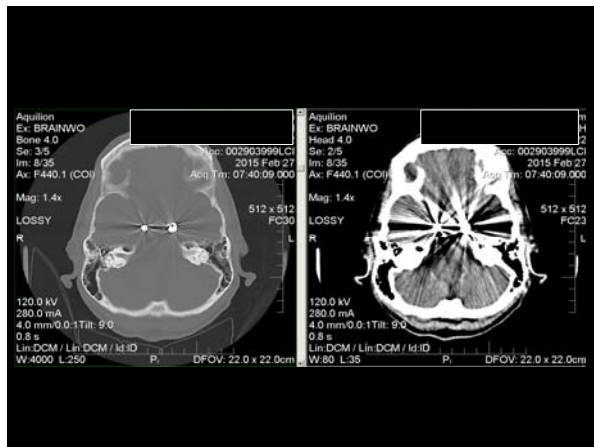
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## Key Points

- Pupil testing
- Common pathologic pupillary abnormalities:
  - Afferent pupillary defects
  - Tonic pupils
  - Horner syndrome
  - CNIII palsy

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Thank you!



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