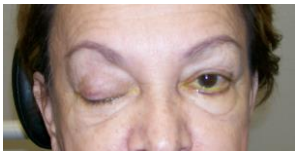
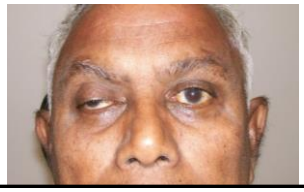
NEURO-OPHTHALMIC EMERGENCIES THAT YOU CAN NOT MISS

Rim Makhlouf, OD, FAAO
Joseph Sowka, OD, FAAO, Diplomate
Nova Southeastern University College of Optometry

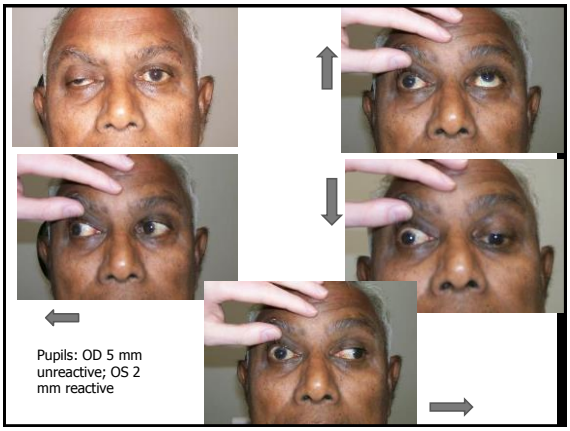
WHICH IS BETTER? ONE OR TWO?





63 YOM

- Long standing glaucoma patient
- Sudden onset of orbital pain x 3 days
- + DM; +HTN
- On coumadin
- Pacemaker
- No vision change
- Presents as walk-in emergency glaucoma eval



Pupils: OD 5 mm unreactive; OS 2 mm reactive

63 YOM

- Pupil involved CN III palsy
- 3 days duration at least
- Most likely cause: intracranial aneurysm
- Sent to ER with detailed notes and recommendations
- Endovascular therapy with coils
- Hospitalized 23 days

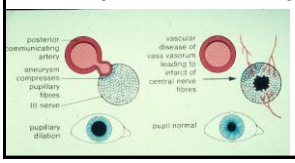
Secondary aberrant regeneration

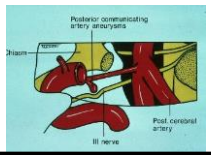
CN III PALSY CLINICAL PICTURE

- An eye that is down and out with a ptosis
- Adduction, elevation, depression deficits
- Isocoric or anisocoric

CN III ANATOMY

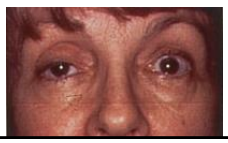
- Vulnerable to compression from aneurysm in subarachnoid space
  - Posterior communicating artery (PCOM)
  - Junction PCOM and ICA
  - Tip of basilar artery
- Pupillomotor fibers vulnerable to compression, relatively immune to ischemia

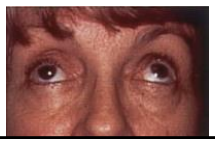




STILL MORE CLUES

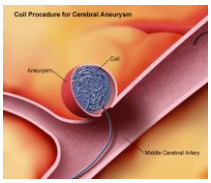
- Pupil involved CN III palsy is PCOM aneurysm until proven otherwise
- Incomplete palsy is PCOM aneurysm until proven otherwise- Regardless of pupil
- 30% of CN III palsy are caused by aneurysm
- Vasculopathic CN III will resolve in time
- Life threatening posterior communicating aneurysm will rupture in time

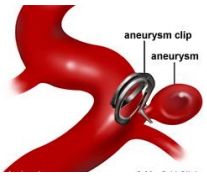




STILL MORE CLUES

- CN III palsy caused by aneurysm
  - 20% die within 48 hrs from rupture
  - 50% overall die
  - Average time from onset to rupture – 29 days
    - 80% rupture w/i 29 days
  - Many never make it to hospital





SUSPECT THE WORST

- Optometrist sees patient with CN III palsy
- Referred to ophthalmologist next day
- Pt dies from subarachnoid hemorrhage before consult

DOES PRESENCE OF VASCULOPATHIC RISK FACTORS HELP?

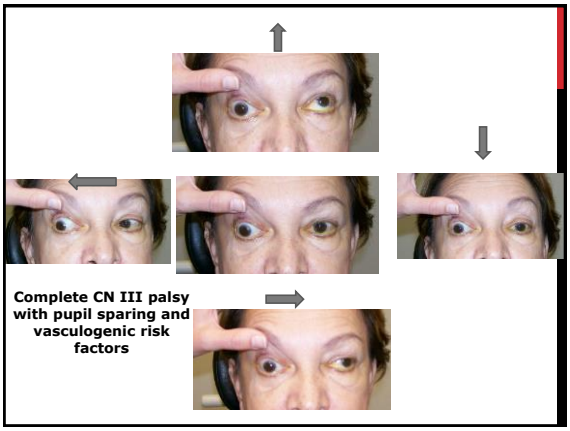
- Arteriosclerotic risk factors in elderly favors microvascular etiology but does not rule out aneurysm
- HTN, DM, atherosclerosis, hyercholesterol all common and don't protect against aneurysm
- Answer: no, but makes me very nervous when NOT present

ANEURYSM RISK ASSESSMENT: ISOLATED CN 3 PALSY

- Isolated dilated pupil — none
- Complete CN3-normal pupil — low
- Partial CN3 – normal pupil — high
- Pupil involved CN3 — emergency

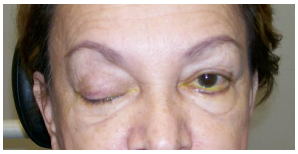
A DIFFERENT PATIENT AND PROGNOSIS

- 63 YOF
- Diabetes and HTN- poorly controlled
- Sudden onset retro-orbital pain



Complete CN III palsy with pupil sparing and vasculogenic risk factors

WHICH IS BETTER? ONE OR TWO?



Resolves over several weeks

Hospitalized 23 days with 2 neurosurgical procedures…but did live

NEVER OUT OF THE WOODS

- Pt develops CN III palsy from aneurysm
- Successfully treated with aneurysm clip
  - All endovascular coils are inert and MRI safe; not all clips are MRI safe
- Pt undergoes F/U MRI with non-MRI safe clip in major medical center
- Clip displaces during MRI
- Patient has fatal hemorrhage during procedure

ODE TO A THIRD NERVE

When the eye is down and out with ptosis,
You better hope for miosis.
If the palsy is total with pupil sparing,
In an Oldie it's vascular and not too daring.
A partial palsy calls for double duty,
Because it's probably an aneurysm going through puberty.
But if the pupil is dilated,
An aneurysm has violated.
No time for deferral and no time for referral.
Send to the ER without debate.
Remember, twenty percent will die within the first forty-eight

Joseph Sowka, OD

CASE HISTORY

- 72 yo Caucasian Male
- CC: routine eye exam + glaucoma follow-up

PERTINENT FINDINGS

- VA OD 20/20-2 OS 20/25
- PUPILS Round, reactive to light, no APD
  - Scotopic OD 6mm OS 4.5mm — Anisocoria
  - Photopic OD 5mm OS 3.5mm — Anisocoria
- MOTILITIES Full and smooth OU
- CF: FTFC OU

COMPARE ANISOCORIA

Anisocoria DIM > BRIGHT

- Faulty Pupil = Smaller Pupil
  - Horner's

Aniscoria BRIGHT > DIM

- Faulty Pupil = Larger Pupil
  - Adie's pupil, 3rd Cranial Nerve Palsy

PERTINENT FINDINGS

- VA OD 20/20-2 OS 20/25
- PUPILS Round, reactive to light, no APD
  - Scotopic OD 6mm OS 4.5mm — Anisocoria 1.5 mm
  - Photopic OD 5mm OS 3.5mm — Anisocoria 1.5mm
- MOTILITIES Full and smooth OU
- CF: FTFC OU

PHYSIOLOGICAL ANISOCORIA

- Chronicity
- Absence of symptoms or associated findings
- Normal pupillary light responses
- Difference of < 1 mm (usually < 0.4 mm) b/w pupil sizes

COMPARE ANISOCORIA

Anisocoria DIM > BRIGHT

- Faulty Pupil = Smaller Pupil
  - Horner's

Aniscoria BRIGHT > DIM

- Faulty Pupil = Larger Pupil
  - Adie's pupil, 3rd Cranial Nerve Palsy

DOES THIS LOOK LIKE HORNER'S SO FAR?

No but ...

Let's do some additional testing to confirm

What drug can we use?

DOES THIS LOOK LIKE HORNER'S SO FAR?

No but ...

Let's do some additional testing to confirm

What drug can we use?

- Aproclinidine 0.5%

In which eye?

DOES THIS LOOK LIKE HORNER'S SO FAR?

No but ...

Let's do some additional testing to confirm

What drug can we use?

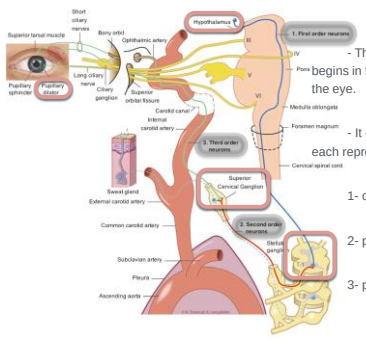
- Aproclinidine 0.5%

In which eye?

- Both

HORNER'S SYNDROME

- Syndrome of oculosympathetic paresis



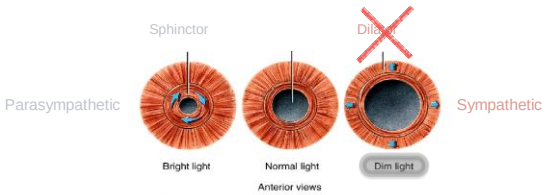
- The oculosympathetic pathway begins in the hypothalamus and ends in the eye.
- It consists of a 3-neuron arc, each representing an order of neuron:

1- central (or 1st-order),

2- preganglionic (or second-order)

3- postganglionic (or third-order)

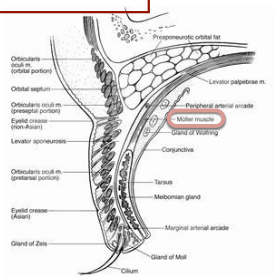
- Denervation of the dilator muscle results in a pupil that dilates more slowly than the normal one, leading to miosis



- Pupil does not dilate as well as in the contralateral eye, the resulting anisocoria is therefore accentuated in dim light

- Müller's muscle is also sympathetically innervated and acts as an accessory elevator of the upper eyelid by providing about 2 millimeters of elevation:

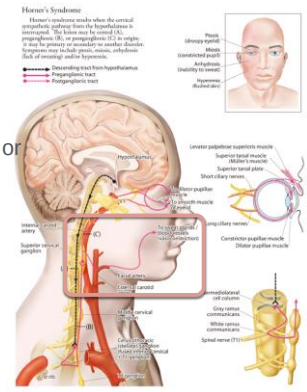
  - Subtle ptosis
  - Weakness of the inferior tarsal muscle results in reversed ptosis



The resulting smaller palpebral fissure may also result in the false impression of a sunken globe, or enophthalmos

Enopthalmos: posterior displacement of the eyeball within the orbit

Possible ipsilateral loss of facial sweating or hemifacial anhydrosis

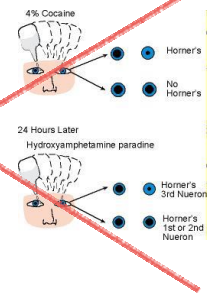


1. Testing:

   a) **Traditionally:**

   (1) Cocaine: to confirm diagnosis

   (2) Hydroxyamphetamine: can be used to distinguish central and preganglionic from post-ganglionic lesions



Iopidine testing:

- Widely available anti-glaucoma drop makes it an easy test

APROCLINIDINE TESTING

1 drop Aprolinidine 0.5% in each eye

↓

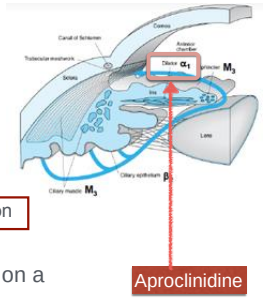
Wait 60 minutes

↓

Measure pupils

APROCLINIDINE

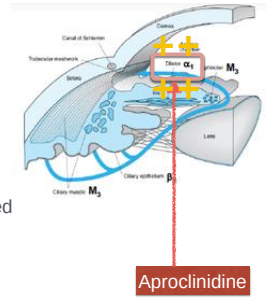
- Selective α2 agonist
- Used to reduce IOP
- Has only weak α1 action
- Little to no effect on a normal pupil.



APROCLINIDINE TESTING

Patients with Horner syndrome may develop denervation hypersensitivity of α1 receptors on the iris dilator muscle

- Mydriasis of the affected pupil in response to apraclonidine



APROCLINIDINE TESTING

If positive ... Reversal of miosis

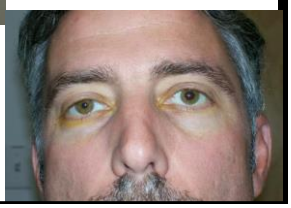
Horner

If negative ... No reversal

No Horner

Pre-Iopidine

Post-Iopidine



APROCLINIDINE TESTING: RESULTS

Results: Reversal of miosis

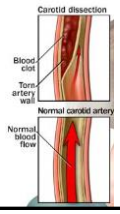
Horner

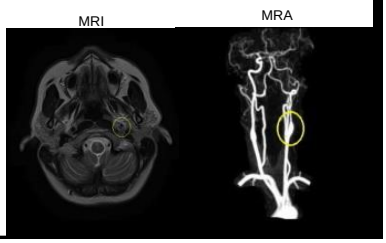
IMPRESSION/PLAN

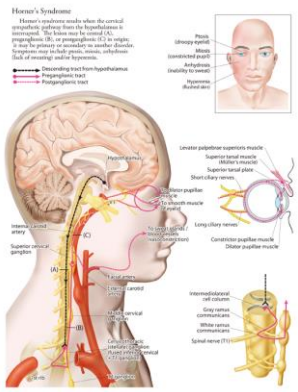
- **Horner's Syndrome**
  - Referred to neuro-ophthalmologist for further testing
    - MRI of chest, neck and brain ordered
    - Result: Pancoast tumor
      - Scheduled for surgery

INTERNAL CAROTID ARTERY DISSECTION (ICAD)

- Stretching of the internal carotid artery, causing the lamina intima to tear and blood to invade the resulting space







INTERNAL CAROTID ARTERY DISSECTION (ICAD)

- Stretching of the internal carotid artery, causing the lamina intima to tear and blood to invade the resulting space

- Result: ischemia, dissection, and/or thrombosis, etc …

- Cause: typically a trauma that resulted in hyperextension and rotation of the neck (eg, car accident, valsalva, sports, exercise/resistance training, etc)

INTERNAL CAROTID ARTERY DISSECTION (ICAD)

- Most common ***ocular*** finding: Horner Syndrome (36-58%)
  - Acute & painful (neck & up)- > suspect ICAD until proven otherwise
- Most common **symptom: Ipsilateral headache (68-92% of pts)**
- Other **symptoms*:* focal cerebral ischemia (49-67%), CN palsies (12-14%), amaurosis fugax (6-30%), ophthalmic/retinal arterial occlusion (5%), ischemic optic neuropathy (3.6%).**
- Workup: CT(CTA)/MRA, MRI … STAT!!
  - Prevent any ischemic insults to the brain and/or eye

TAKE HOME MESSAGE

- **Watch those pupils!**
- **When in doubt or when anisocoria is >= 1mm**
  - Aproclinidine 0.5% is widely available
    - Used as an antiglaucomatous drug to reduce IOP
- **Acute, painful Horner**
    - Emergent CTA/MRA of carotid artery system to R/O ICAD
    - Also Emergent MRI of oculosympathetic pathway if etiology unclear

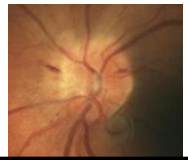
ODE TO HORNER' S SYNDROME

When the lid is low and the pupil small,
Check to see the sweat don't fall.
Cocaine is no longer universal,
Iopidine will cause reversal.
You have to scan head to chest,
And remember that MRA is best.
Pain in association, will surely cause commotion.
Send to the ER without correction,
Remember, it might be carotid dissection.

Joseph Sowka, OD

WHICH IS BETTER? ONE OR TWO



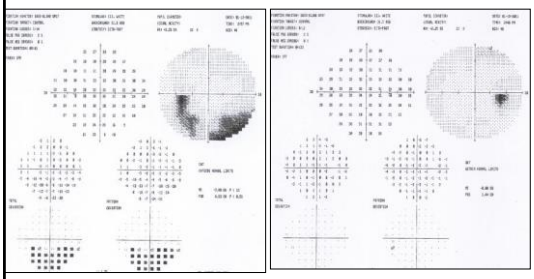



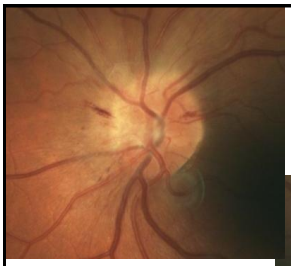
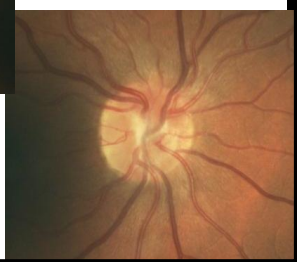

48 YOWM

Painless loss of visual field OS

- 20/20 OD, OS
- Noticed upon waking

Med Hx: Unremarkable, except for viral illness 3 weeks before



NAAION OS
Disc at risk OD

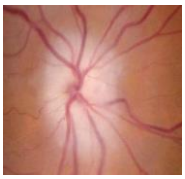
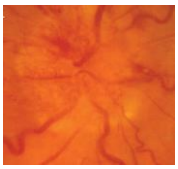
74 YOWM

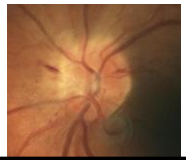
- **Presents with 'worst headache of his life'**
  - Sees: PA, ED physician; cardiologist; NP; 3 ODs
    - *3 week period*
  - Histories: Eye ache; jaw pain, scalp pain, facial pain, somnolence; malaise; jaw claudication
  - Diagnoses: TMJ; Lyme disease
  - *"vasculitis such as temporal arteritis highly unlikely", "Not giant cell arteritis"*
    - *However, ESR and CRP ordered and elevated- never reviewed*
  - End result?

ISCHEMIC OPTIC NEUROPATHY

- **Results from local infarction at the level of the optic nerve**
- **Unilateral presentation but high incidence of subsequent contralateral involvement**
- **May be arteritic (AAION) or non-arteritic (NAAION)**
- **Pale nerve (more so with AAION); extensive NFL edema; arteriolar constriction, peripapillary hemorrhages evident**

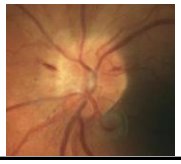
AAION VS NAAION



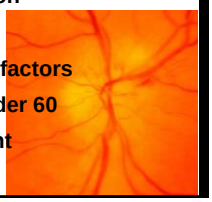
NAAION

- **Risk factors:**
  - Hypertension, diabetes, atherosclerotic disease, small optic nerves
- **Inferior field defects**
- **Hyperemic swollen nerve**
- **Progressive moderate vision loss with potential recovery**
- **Late 30s/ early 40s**
- **Painless**



AAION

- **Pallid optic nerve swelling with flame hemorrhages, arteriole attenuation and NFL infarcts**
- **Pain (of some sort)**
- **Severe optic nerve dysfunction**
- **Visual field defects**
- **Giant cell arteritis/ PMR- risk factors**
- **Typically 70s, uncommon under 60**
- **High risk bilateral involvement**



GIANT CELL ARTERITIS/ TEMPORAL ARTERITIS

- **A systemic disorder affecting primarily the elderly and characterized by granulomatous inflammation of large- and medium-sized arteries**
- **Most patients are over age 60 and the majority are over age 70**
- **Men and women are equally affected**
- **Inflammatory cells produce cytotoxic enzymes and reactive oxygen species that destroy vascular tissue and obstruct vessel lumen**

GIANT CELL ARTERITIS/ TEMPORAL ARTERITIS

- **Headache (present in over 90%)**
- **Scalp tenderness**
- **Jaw claudication (almost diagnostic)**
- **Ear pain**
- **Arthralgias**
- **Temple pain and/or tenderness**
- **Malaise**
- **Intermittent fevers**
- ***MUST ASK PATIENT DIRECTLY**

OCULAR MANIFESTATIONS

- **Loss of vision**
  - Transient (eg, amaurosis fugax; TIA)
  - Persistent (ischemic neuropathy; retinal artery occlusion)
- **Double vision**
  - Third nerve palsy (pupil-sparing most common)
  - Fourth nerve palsy
  - Sixth nerve palsy
- **Eye pain**
- **Red eye**
- **Retinal ischemia- cotton wool spots**

OCULAR MANIFESTATIONS

- **About 5% of cases of AION in patients over age 60 are due to GCA**
- **Visual acuity tends to be very poor (HM or less)**
- **Bilateral involvement not uncommon**
- **Disc not only swollen but also pale (infarct)**
- **Evidence of retinal ischemia not uncommon**
  - Isolated CWS in elderly

DIAGNOSIS

- **Careful history: Must ask about nonvisual symptoms**
- **Examination**
- **Laboratory studies**
  - Erythrocyte sedimentation rate
    - Lowered by statins and NSAIDS
  - C-reactive protein
    - Not affected by statins and NSAIDS
  - Elevated platelet count

AAION VERSUS NAAION

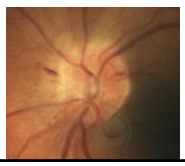
- **Think AAION>>NAAION**
  - Systemic symptoms of GCA
  - TVOs/amaurosis
  - Elevated
    - ESR/CRP/ Platelets
  - **Early massive vision loss**
  - **Chalky white optic disc edema**
- **Treatment: Prompt steroids and hydration**
- **Consider IV when vision loss present**
  - Very effective in prevention of second eye
  - Occasionally restores vision

Hayreh SS, Podhajsky PA, Zimmerman B. Occult giant cell arteritis: ocular manifestations. Am J Ophthalmol. 1998 Apr;125(4):521-6.

GT Liu et al. Ophthalmology 1994

WHICH IS BETTER? ONE OR TWO

Bilaterally blind



Residual field loss, but otherwise not bothered

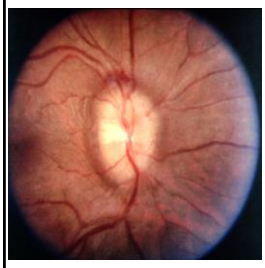
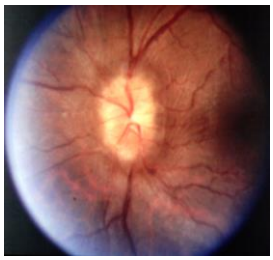
ODE TO AN ISCHEMIC NERVE

When your patient's optic nerve is ischemic
You better hope the disc is hyperemic.
In NAAION no treatment is needed
And life will not be impeded.
But if the disc is swollen and pale,
And vision is an epic fail
If the patient is sixties, seventies or eighties
You will feel heat like in Hades
ESR and CRP are required
And steroids must be acquired
Remember, when you see a choked disc
Always assess the giant cell risk

Joseph Sowka, OD

28 YOF

- Presents with intermittent blurred vision & visual "blackouts", intermittent diplopia, and chronic headache steadily worsening X 2 weeks
- MHx: "white coat hypertension", shoulder injury X 6 mos
- Meds: Flexeril® 10 mg BID PRN
- Height / weight: 5'3", 220 lbs.
- VA: OD 20/20, OS 20/20
- Pupils & motility: normal

DEFINITION:

PAPILLEDEMA: EDEMA OF THE OPTIC DISC, SPECIFICALLY RESULTING FROM ELEVATED INTRACRANIAL PRESSURE.

PATHOPHYSIOLOGY

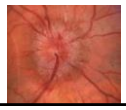
Increased CSF pressure is transmitted to the optic nerve

↓

Stasis of axoplasmic flow

↓

Intra-axonal edema



PAPILLEDEMA: SYMPTOMS

- Maybe asymptomatic
- Headache
- Nausea & Vomiting
- Dizziness
- Tinnitus (ringing in the ears)
- Diplopia
- Transient Visual Obscurations

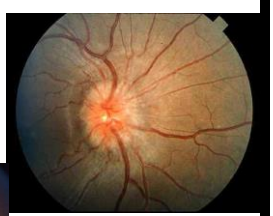
CN III, IV or VI palsy due to raised intracranial pressure, or due to brain lesion compression on the nerves

PAPILLEDEMA: SIGNS

- **Bilateral Disc Edema (Can be UNI, but rare)**
  - DDX Pseudo-papilledema
- **VA**
  - Usually not affected
- **CV**
  - Normal or reduced
- **Visual Field Defects**
  - Any pattern
  - NFL-related: arcuate and paracentral scotomas, nasal step defects
  - Enlarged blindspot: refractive scotoma vs axonal damage
- **APD?**

PAPILLEDEMA: SIGNS

- **Bilateral Disc Edema (Can be UNI, but rare)**
  - DDX Pseudo-papilledema
    - Blurred disc margins
    - Disc splinter hemorrhages
    - Dilated veins
    - Absence of SVP (±)
    - Vessel obscuration
    - Paton's folds
    - Optic nerve hyperemia




PAPILLEDEMA: SIGNS

- **Bilateral Disc Edema (Can be UNI, but rare)**
  - DDX Pseudo-papilledema
    - Optic Nerve Head Drusen

PAPILLEDEMA: SIGNS

- **Stages of papilledema**

Papilledema Grading System (Frisen Scale) Reference: Frisen L. Swelling of the optic nerve head: A staging scheme. J Neurol Neurosurg Psychiatry 1982; 45:13-18

Stage 0 - Normal Optic Disc Blurring of nasal, superior and inferior poles in inverse proportion to disc diameter. Radial nerve fiber layer (NFL) without NFL tortuosity. Rare obscuration of a major blood vessel, usually on the upper pole.

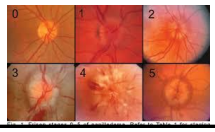
Stage 1 -Very Early Papilledema Obscuration of the nasal border of the disc. No elevation of the disc borders. Disruption of the normal radial NFL arrangement with grayish opacity accentuating nerve fiber layer bundles. Normal temporal disc margin. Subtle grayish halo with temporal gap (best seen with indirect ophthalmoscopy). Concentric or radial retrochoroidal folds.

Stage 2 - Early Papilledema Obscuration of all borders. Elevation of the nasal border. Complete peripapillary halo.

Stage 3 - Moderate Papilledema Obscurations of all borders. Increased diameter of optic nerve head. Obscuration of one or more segments of major blood vessels leaving the disc. Peripapillary halo-irregular outer fringe with finger-like extensions.

Stage 4 - Marked Papilledema Elevation of the entire nerve head. Obscuration of all borders. Peripapillary halo. Total obscuration on the disc of a segment of a major blood vessel.

Stage 5 - Severe Papilledema Dome-shaped protrusions representing anterior expansion of the optic nerve head. Peripapillary halo is narrow and smoothly demarcated. Total obscuration of a segment of a major blood vessel may or may not be present. Obliteration of the optic cup.



PAPILLEDEMA: SIGNS

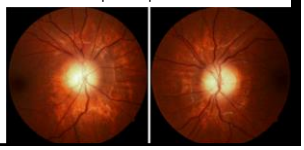
- **Stages of papilledema**

Early Papilledema

Established Papilledema

Chronic Papilledema

Atrophic Papilledema

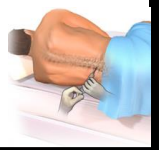


PAPILLEDEMA: CAUSES

- **Increased brain volume**
  - Eg, space-occupying lesions (brain tumor, hemorrhage or abcess)
- **Decreased skull volume**
- **Increased CSF production**
- **Decreased CSF drainage**
  - Eg, hydrocephalus, meningitis, dural venous sinus thrombosis, subarachnoid hemorrhage
- **Pseudotumor Cerebri (PTC)**

PAPILLEDEMA: CAUSES

- **Pseudotumor Cerebri (PTC)**
  - Diagnosis of exclusion
  - Normal MRI
  - Normal biochemical and cytological composition of CSF
  - Increased opening pressure on lumbar puncture (> 25 cm H2O)
  - No other neurological disease (except for cranial nerves)

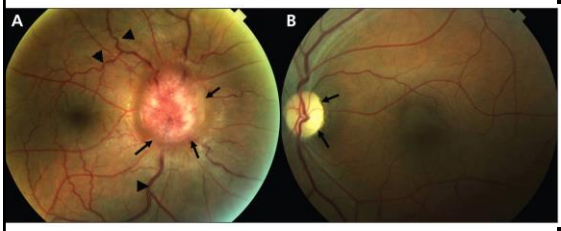


PAPILLEDEMA: CAUSES

- **Pseudotumor Cerebri (PTC)**
  - Most commonly in obese women of child-bearing age
    - Also called Idiopathic Intracranial Hypertension (IHH)
  - Other causes:
    - Tetracyclines, vitamin A, endocrine disease (hyper/hypothyroidism), sleep apnea, chronic respiratory disease

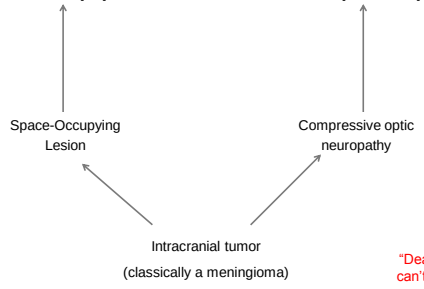
BEWARE: FOSTER-KENNEDY SYNDROME

- **Unilateral papilledema + Contralateral optic atrophy**



BEWARE: FOSTER-KENNEDY SYNDROME

- **Unilateral papilledema + Contralateral optic atrophy**



"Dead discs can't swell!!"

DDX WITH OTHER CAUSES OF DISC SWELLING

- No specific signs for DDx papilledema vs other causes of disc swelling

- Bilateral disc swelling ➔ Papilledema until proven otherwise

PAPILLEDEMA MANAGEMENT

- **Suspect papilledema -> Emergency**
  - Ancillary testing to R/O ONHD
  - If true disc swelling ➔ Order MRI of the brain STAT
  - MRV may be ordered to help R/O sinus venous thrombosis or stenosis
  - If MRI/MRV normal ➔ Order Lumbar puncture
    - (CSF pressure **AND** CSF composition)
  - If MRI/MRV normal, CSF composition normal, and CSF opening pressure elevated ➔ Diagnosis of PTC

PAPILLEDEMA MANAGEMENT

- **Confirmed Diagnosis**
  - Treat underlying cause
  - Regular ophthalmic examination
    - VA
    - Color Vision
    - VF
    - Optic Disc Evaluation

ODE TO A SWOLLEN DISC

When you think the disc is swollen
The vessels north and south will appear stolen.
Not all elevated nerves are edematous
Just like not all snakes are venomous.
Your thoughts should go to papilledema
But infection and inflammation should still be in your schema.
MRI, MRV and LP, are soon to be.
Remember, pseudotumor is a diagnosis of exclusion
Female and firm does not make PTC a forgone conclusion.
Brain tumors can exist when the PTC profile is classic.
Do the evaluation so they don't end in a casket.

Joseph Sowka, OD