



**WARREN  
NEUROSCIENCE  
INSTITUTE**  
*at Saint Francis*

**Neuro-Ophthalmic  
Emergencies**

7<sup>th</sup> Annual Trauma and Stroke Symposium

September 12, 2024

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# DISCLOSURES



I have relevant financial relationships with Alexion and Argenx as a speaker on their speaker bureau's. I have mitigated these relationships with Saint Francis Education. I will not be promoting any healthcare goods or products from Alexion nor Argenx during my presentation.

# OBJECTIVES

- To identify the clinical features of ischemic optic neuropathy.
- To recognize features differentiating arteritic and non-arteritic ischemic optic neuropathy.
- To review stroke-related causes of diplopia.

# EVALUATION OF “ACUTE” VISION LOSS



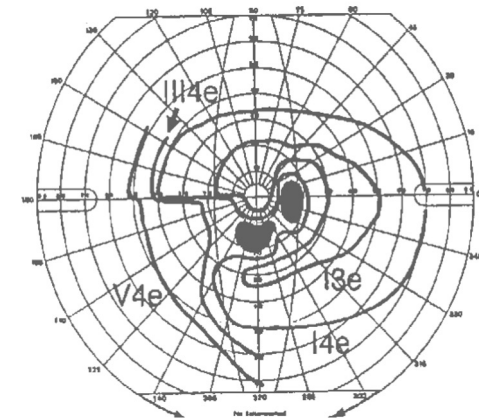
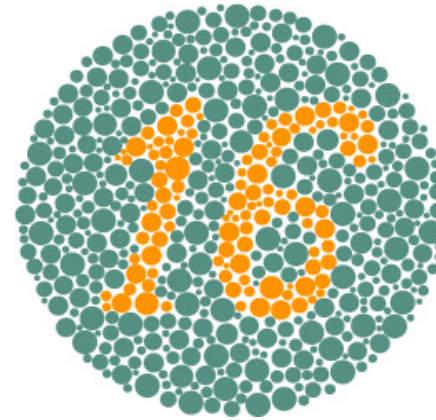
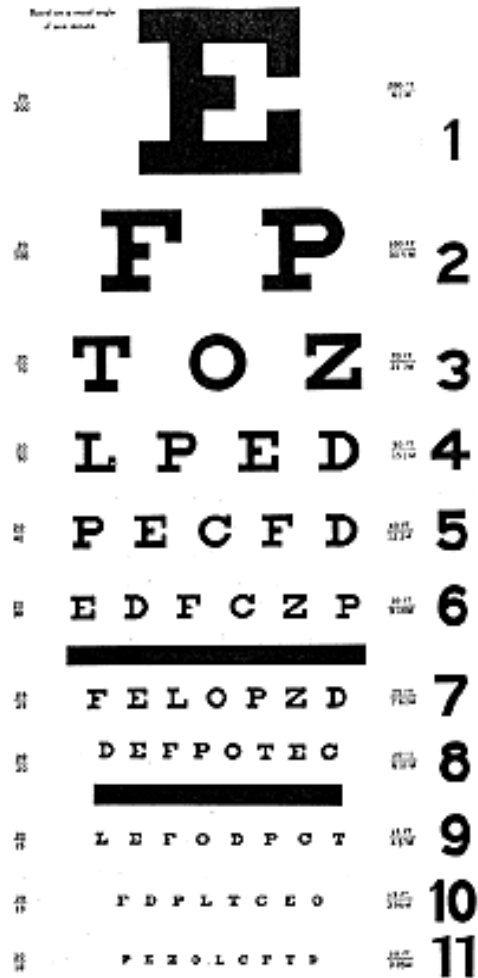
**R<sub>x</sub>**

- Optometrist
- Ophthalmologist

A white rectangular note with a red pushpin at the top right corner. The note contains the text 'R<sub>x</sub>' in a large, bold, serif font, followed by a list of two professions: 'Optometrist' and 'Ophthalmologist'.

# LOCALIZATION TO THE OPTIC NERVE

- Specific functions are easily measured
- Can be easily assessed *in vivo*
- Injury not often improved with treatment



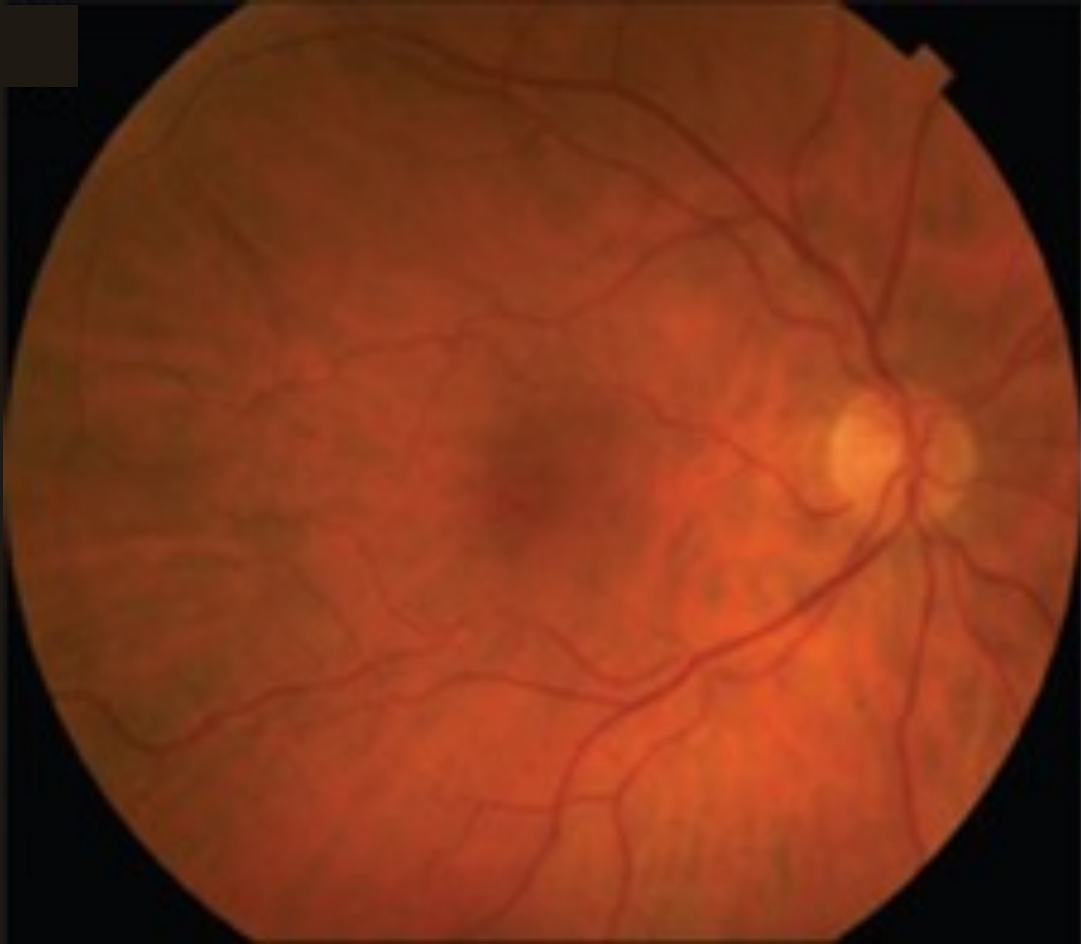
# 65-YEAR-OLD MAN:

**CCC:** “Shade being pulled over my LEFT eye”

**HPI:** Noted shortly after awakening today  
No associated pain, headache, other sx

**PMH:** HTN, dyslipidemia, obstructive sleep apnea

**PE:** VA: 20/25 OD, 20/60 OS  
Color (HRR): 9/10 OD, 3/10 OS  
CVF: superior visual field constriction OS  
Pupils: afferent pupillary defect OS



**“disc-at-risk”**

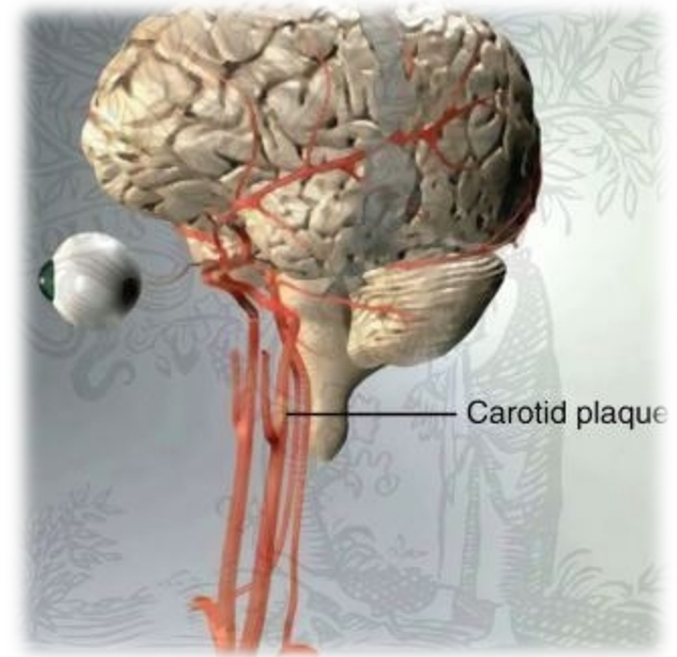
# N-AION:

## NON-ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY:

- most common unilateral optic neuropathy > age 50
- hours-days; pain is universally absent
- variable visual acuity loss at onset
- disc edema (segmental, +/- hemorrhage)
- 15-20% risk of fellow eye involvement w/i 5 yrs

### • Risk Factors:

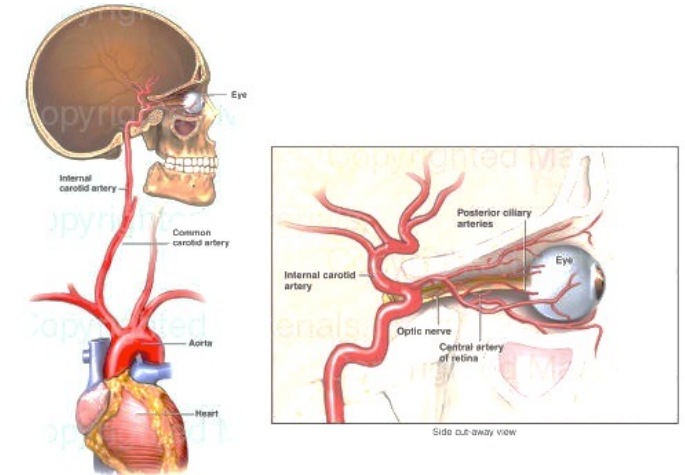
- HTN, DM
- **carotid stenosis**
- nocturnal hypotension (? meds)
- sleep apnea





# Recognizing Ocular Ischemia

- Approximately 15% of patients will have a disabling stroke within 3 months following a TIA
  - 50% will occur within 48 hours after resolution of the TIA symptoms
  - hemispheric TIA from severe carotid artery stenosis is associated with the highest risk of stroke (20% at 3 months)
- “Rapid-access” TIA clinics:
  - SOS-TIA study (France)
  - EXPRESS study (GB)
  - Reduce 90 day risk of stroke by 80%
  - Reduction in overall hospital bed days and associated health-care costs
- US: ED-based protocols and/or observation units



# YES, IT'S A STROKE...

- AHA and NSA: “all TIA patients should be evaluated emergently in a specialized unit” (inpatient or outpatient).
- Risk of stroke and vascular death is increased after retinal TIA (ANY transient vision loss of presumed vascular cause).
- TIA and stroke exist on a continuum; TIA offers an opportunity to initiate treatment that can reduce the risk of permanent disability.
- Risk of stroke, cardiac events and death after a true retinal TIA is likely as high as for patients with cerebral TIA (Biouesse and Trobe, *Am J Ophthalmology* 2005).



### Class I recommendations

1. Patients with TIA should preferably undergo neuroimaging evaluation within 24 hours of symptom onset. MRI, including DWI, is the preferred brain diagnostic imaging modality. If MRI is not available, head CT should be performed (Class I, Level of Evidence B).  
*suspected TIAs (Class I, Level of Evidence A).*
3. Noninvasive testing of the intracranial vasculature reliably excludes the presence of intracranial stenosis (Class I, Level of Evidence A) and is reasonable to obtain when knowledge of intracranial steno-occlusive disease will alter management. Reliable diagnosis of the presence and degree of intracranial stenosis requires the performance of catheter angiography to confirm abnormalities detected with noninvasive testing.
4. Patients with suspected TIA should be evaluated as soon as possible after an event (Class I, Level of Evidence B).

**\*\*Ocular TIA/Infarct INCLUDED...**

# N-AION: TREATMENT

## **Treatment:**

- daily antiplatelet
- risk factor modification

## **Outcome:**

- minimal improvement in VA
- visual field defects usually persist

## **Risk:** recurrent ischemia (fellow eye or stroke)

- CT angiography of head and neck
- echocardiogram
- sleep study (if sleep apnea suspected)

# THE “6 H’S” OF N-AION

- **H**ypertension (or **H**ypotension)
- **H**yperglycemia (diabetes)
- **H**yperlipidemia
- **H**ypersomnia (obstructive sleep apnea)
- **H**igh intraocular pressure (post-cataract, LASIK)

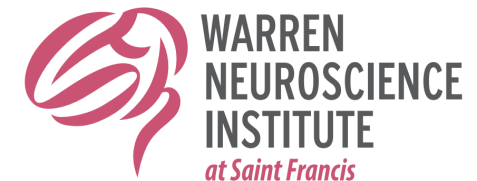
# NAION: PDE-5 Inhibitors



Meta-analysis of peer-reviewed publications\*

- 39 cases of NAION associated with ED med use
- reports vary significantly:
  - frequency of use prior to NAION
  - dose of drug used
  - time interval between last use and onset of NAION
  - many (not all) w/ cerebrovascular risk factors

**Pfizer's case-crossover study:  
2X increased risk w/ED med**



# 75-YEAR-OLD WOMAN:

**CC:** “My RIGHT eye went blind”

**HPI:** unilateral temporal headache  
cannot recall time of onset of vision loss  
recent ~5 pound weight loss

**PMH:** DM, polymyalgia rheumatica (PMR)

**PE:** VA: Bare HM central OD, 20/30 OS  
Color (HRR): 0/10 OD, 7.5/10 OS  
Pupils: APD OD





## **ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY AKA: GIANT CELL OR TEMPORAL ARTERITIS**

- RARE < 60; prevalence increases with age
- acute onset; vision loss is severe (VA <20/200)
- fundus: pallid, “chalky” edema +/- cotton-wool spots
- **Risk Factors:**
  - age (mean 70)
  - associated with PMR



# GCA: Symptom Profile

Headache	58%
Jaw claudication	53%
Weight loss	31%
Malaise	22%
PMR	22%
Anorexia	20%
Fever	11%
Neck pain	11%
Scalp tenderness	11%



# GCA: DEMOGRAPHICS

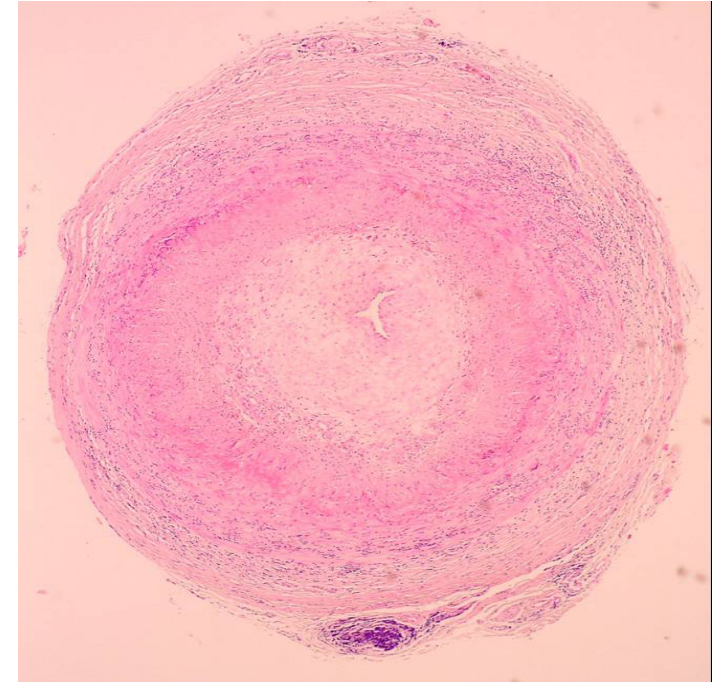
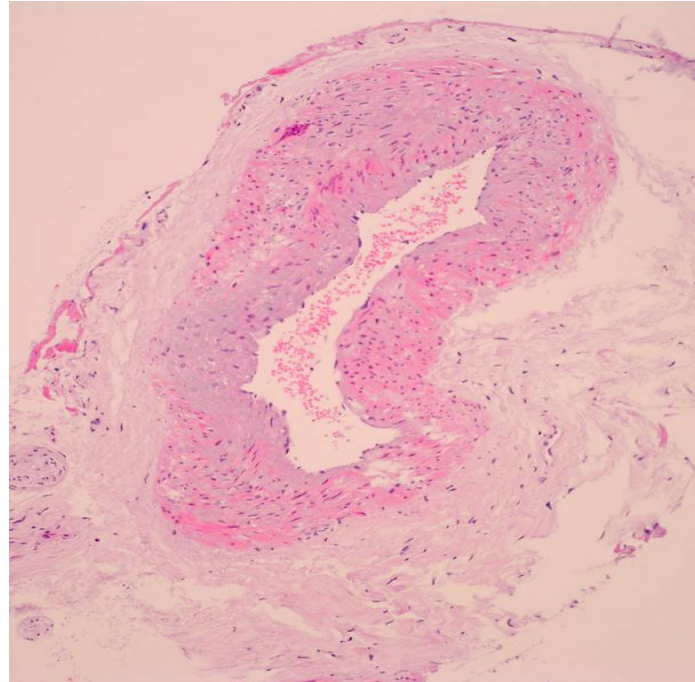
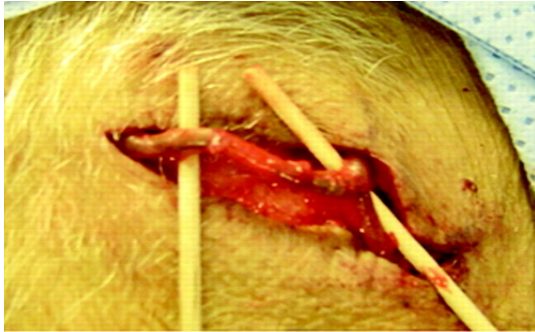
## AGE IS IMPORTANT!

- consider GCA > age 50
  - age 50: 11-17/100,000
  - age 80: 28/100,000
  - >80: ~1/1000
- 3:1 F:M



# GCA: Diagnosis

- **Clinical Presentation**
- Labs: ESR, CRP may be elevated
- Temporal artery ultrasound: helpful only if POSITIVE
- **Temporal Artery Biopsy (\*\*)**



# GCA is a vascular emergency...

## Goals:

1. Decrease active inflammation
2. Improve vision in the affected eye (uncommon)
3. Prevent involvement of fellow eye
4. Decrease systemic complications of vasculitis

## If GCA suspected:

1. Treat first; ask questions later...
2. Begin steroids immediately (min 80 mg/d)
3. Admit to hospital (with internist co-management)
4. IV steroids 250 mg every 6 hours x 3-5 days
5. Temporal artery biopsy, best if bilateral



## GCA Myths:

1. Diagnosis requires systemic symptoms.
  - **21% present ONLY with vision loss (occult)**
2. Diagnosis requires elevated ESR.
  - **Normal ESR does not exclude GCA.** →
3. Steroid therapy can be tapered by protocol.
  - **Each patient must be treated individually.**
  - **Serial monitoring w/ ESR and CRP most useful.**
4. GCA always “burns itself out.”
  - **Long-term (9-12 months) is required.**
  - **May need life-long tx to prevent vision loss.**



ESR,  
CRP,  
fibrinogen  
and  
platelet  
count  
increase  
diagnostic  
yield

## N-AION

## A-AION

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Age:	> 50	> 70 (typically)
Onset:	hours-days	acute/rapid
Pain:	absent	absent + systemic sx.
Nerve:	edema + heme “disc-at-risk”	pallid edema
Outcome:	minimal improvement	no improvement *fellow eye
Mgmt:	urgent (?)/risk factors	<b>EMERGENT/TAB</b>

# Diplopia: Vascular Causes



- **ALWAYS acute (emergency?)**
- Key questions:
  - monocular/binocular
  - intermittent/constant
  - $\Delta$  with direction of gaze
  - $\Delta$  with distance/near



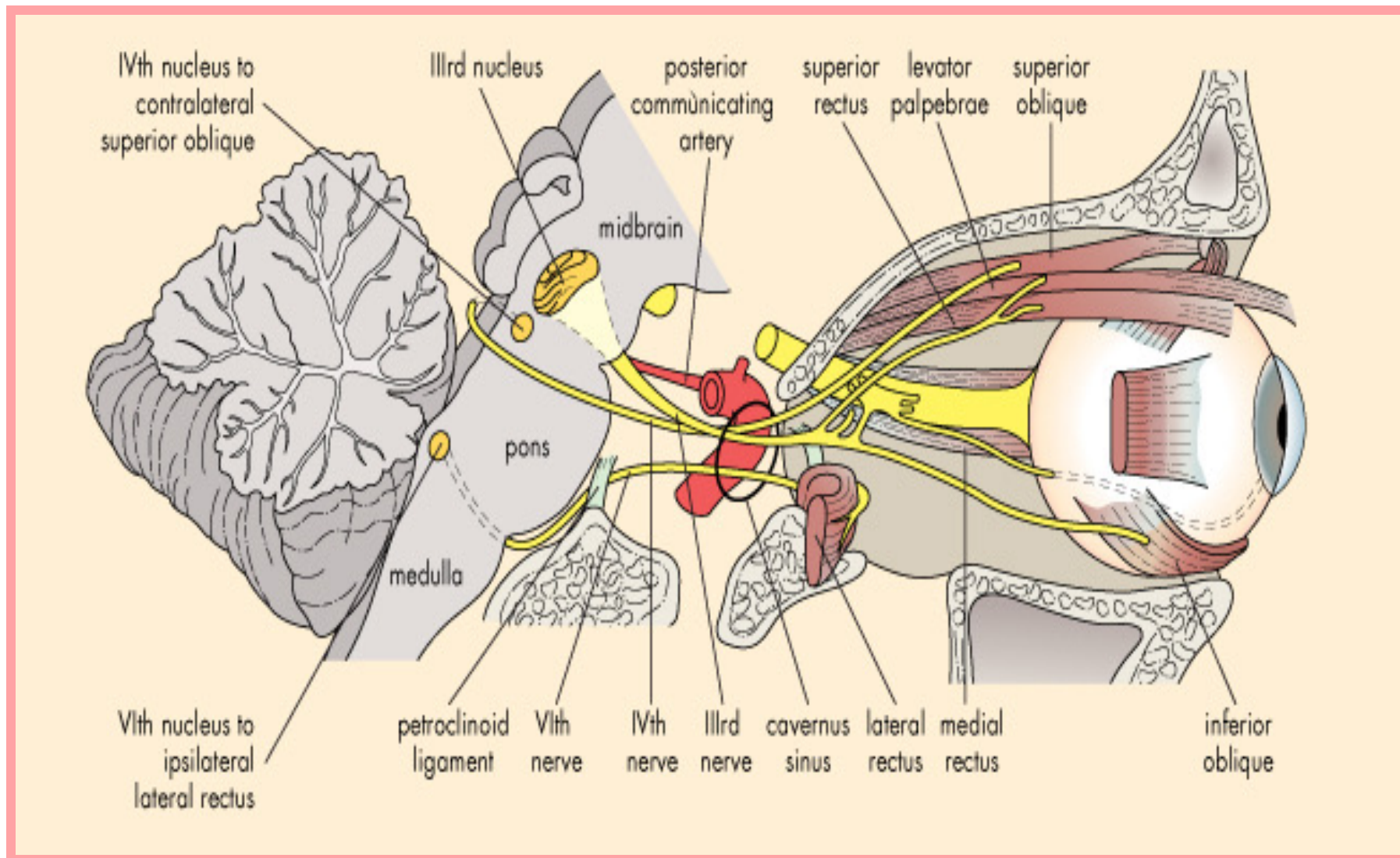
# Monocular Diplopia

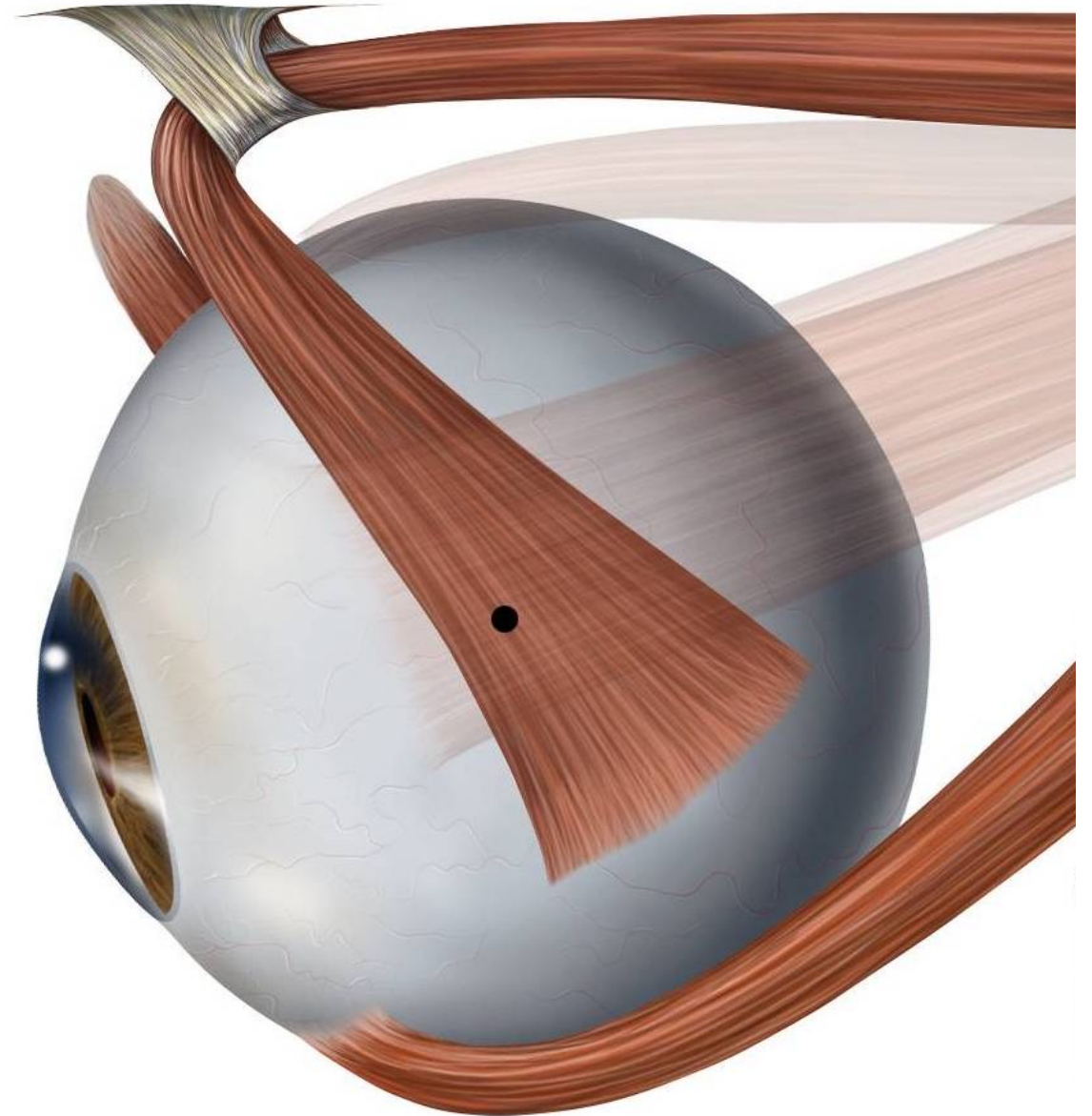
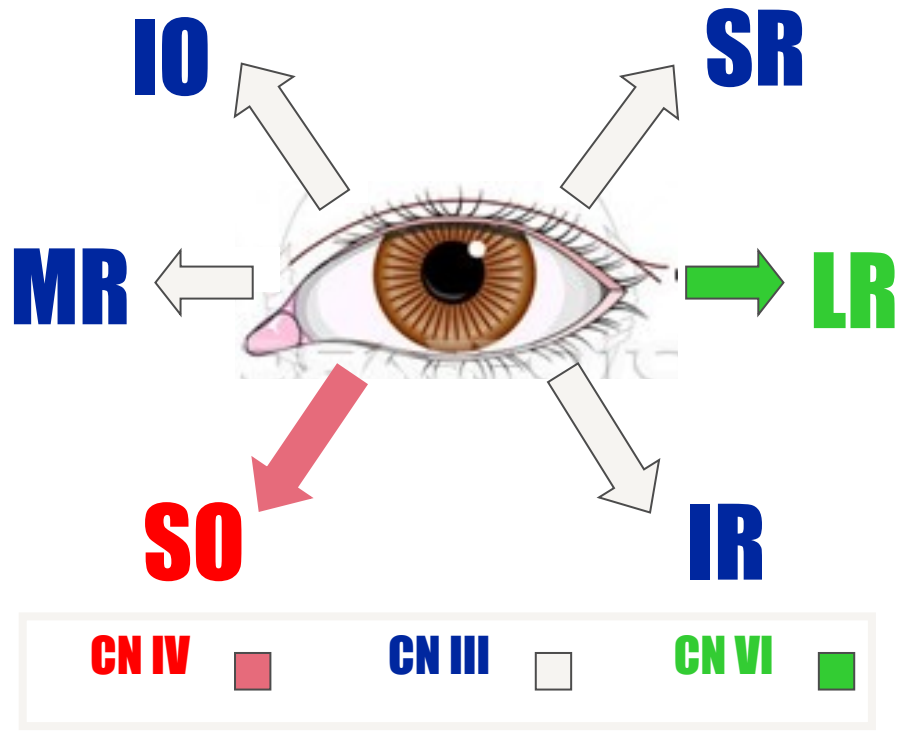


- Need for/inadequate refraction
- Dry eye
- Cataract or IOL
- Corneal irregularity
- (almost) NEVER neurologic



# CN III, IV, and VI: Lateral View





# 58-YEAR-OLD MAN:

## 04/2024 ED: acute “double vision”

- binocular, horizontal, worse in L gaze

**PMH:** HTN, DM, hyperlipidemia, NPDR (stable)

**Exam:** BP 141/74

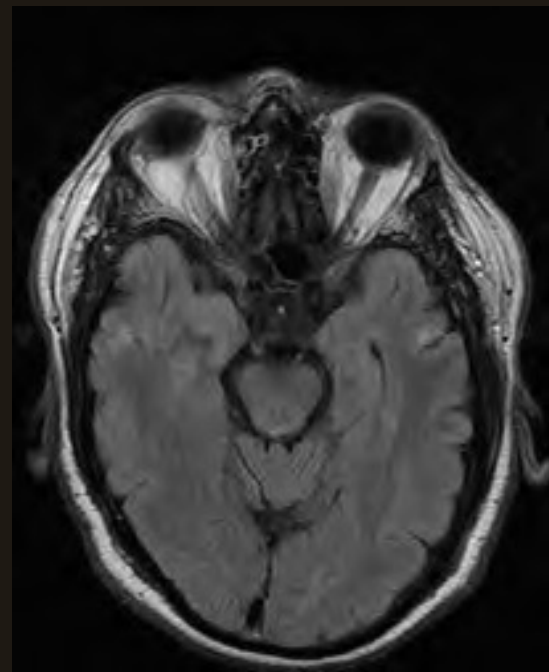
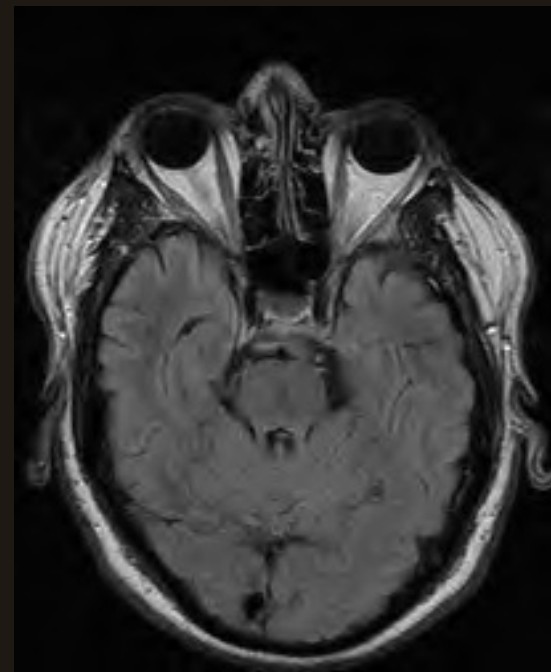
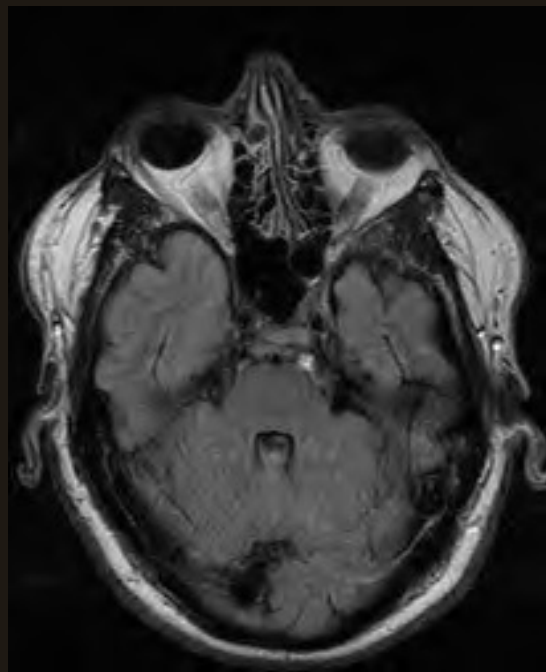
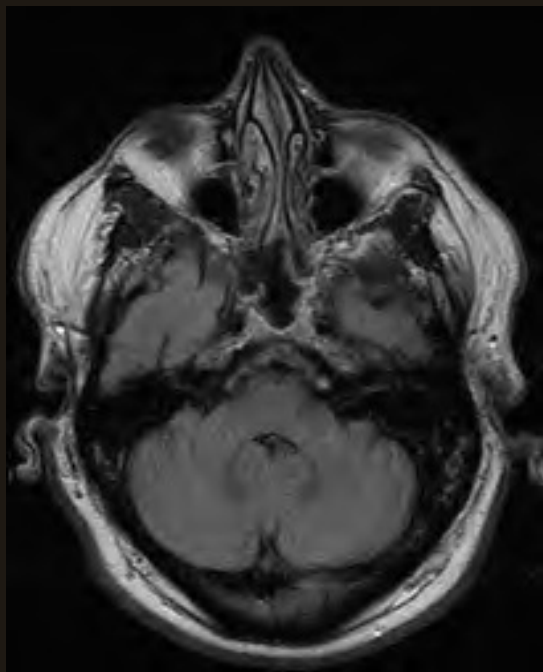
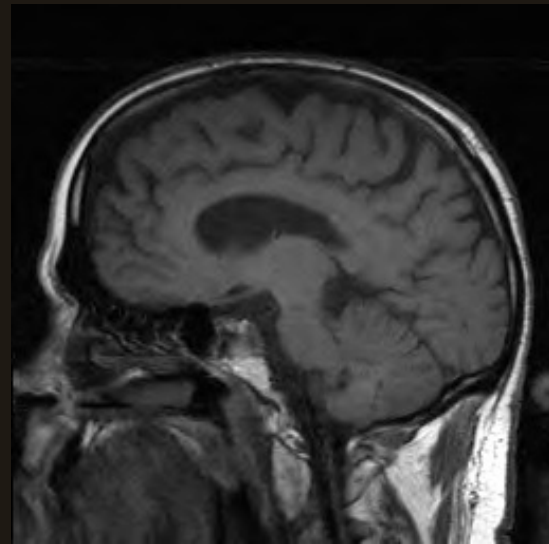
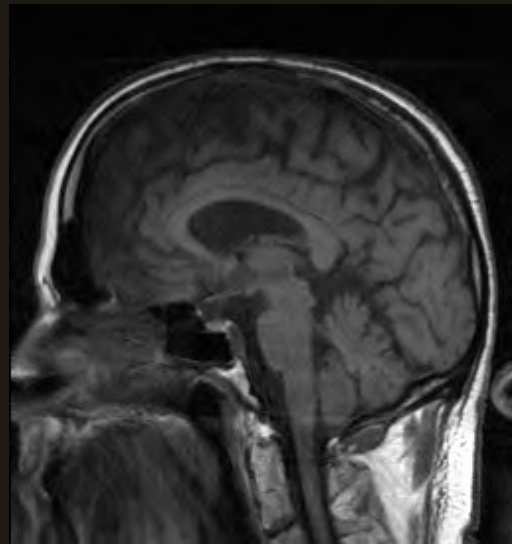
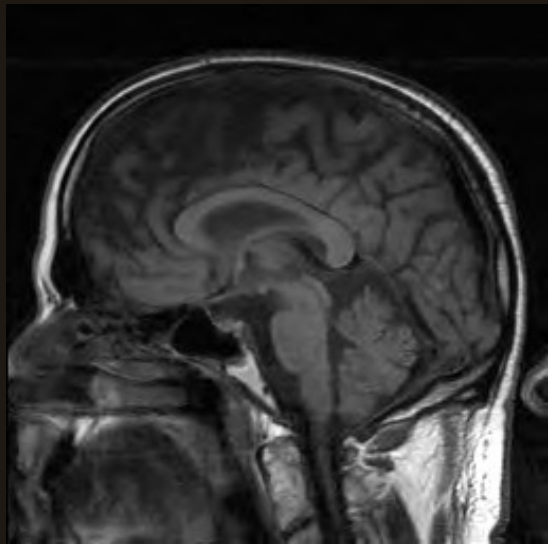
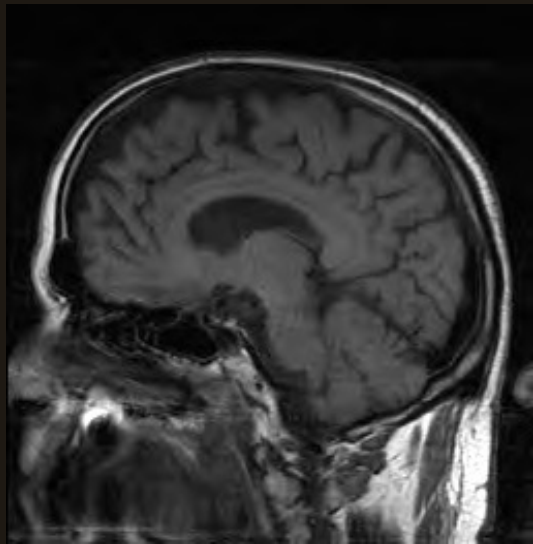
BCVA 20/15 OU

Color: Full OU

VF: Full OU

Pupils: 5mm OU, no APD





## 09/2008 F/U:

- “Things are much better...”
- diplopia completely resolved
- EOM and ocular alignment normal

**Dx:** microvascular (“diabetic”) CN IV palsy



# 72-YEAR-OLD MAN:



# CN III PALSY

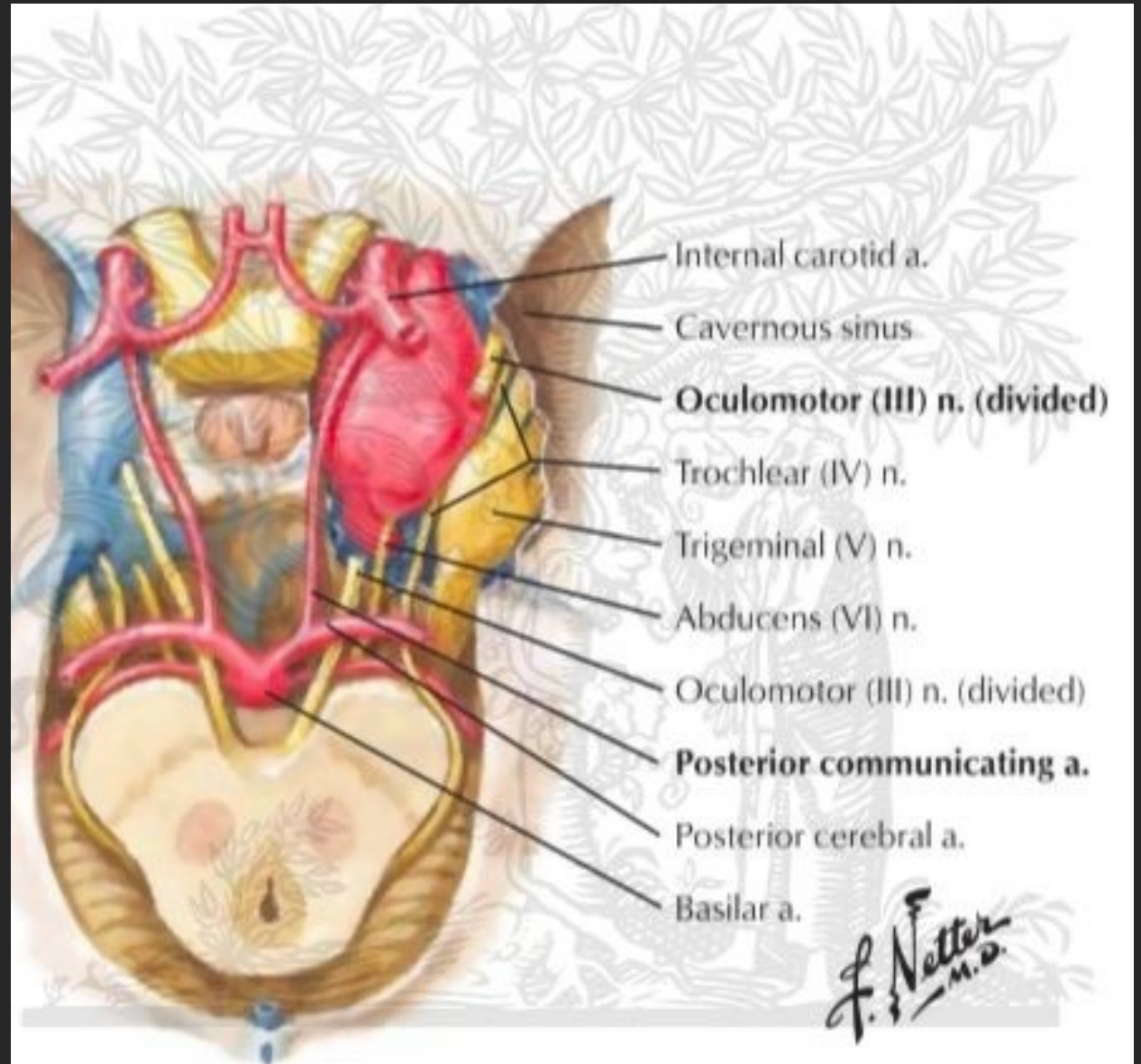
- eye “down and out”
- complete vs incomplete
- pupil-involving vs pupil-sparing



- DDX:
  - microvascular
  - aneurysm (P-comm)



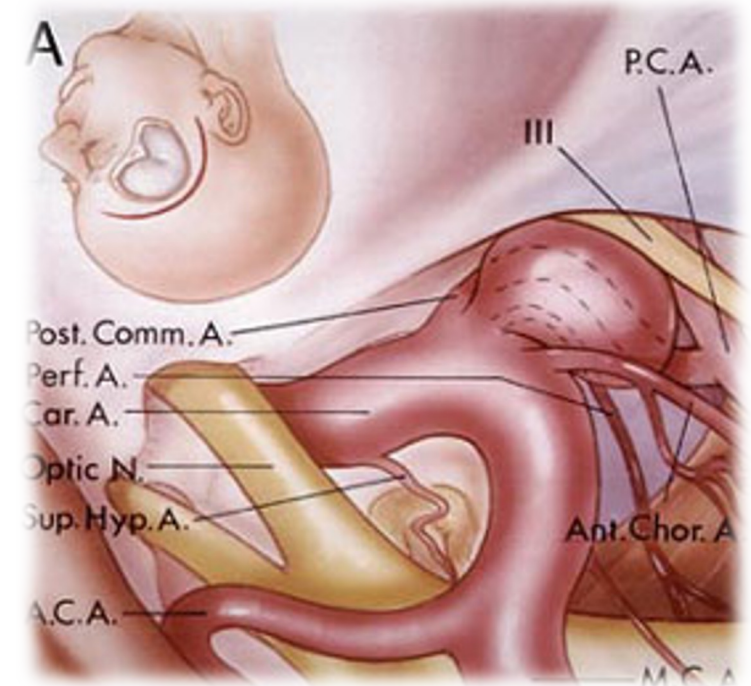




# IMAGING STRATEGIES IN ISOLATED THIRD NERVE PALSY

## ELMALEM ET AL, NANOS 2010

- 17 cases of aneurysmal TNP at Emory (2001-2010)
  - 4/17 SAH on CT (easily diagnosed)
  - 5/17 correct diagnosis (MRA or CTA)
  - 8/17 diagnosis missed



# ANEURYSMAL TNP: MISSED DIAGNOSIS

- 1/8 study not available for direct review
- 7/7 study was adequate, but MISREAD by radiologist
  - 6/7 wrong/incomplete information provided
    - **CLINICAL FINDINGS more important than TECHNIQUE USED**
    - **“EXPERT interpretation of non-invasive imaging should detect nearly all aneurysms responsible for isolated 3<sup>rd</sup> nerve palsy”**



**CLINICAN + RADIOLOGIST = IDEAL**



# INTERMITTENT (“STUTTERING”) DIPLOPIA



## **BEWARE:**

- impending brainstem ischemia
- giant cell (temporal) arteritis

# SUMMARY

- **NAION (or "ocular TIA"):**

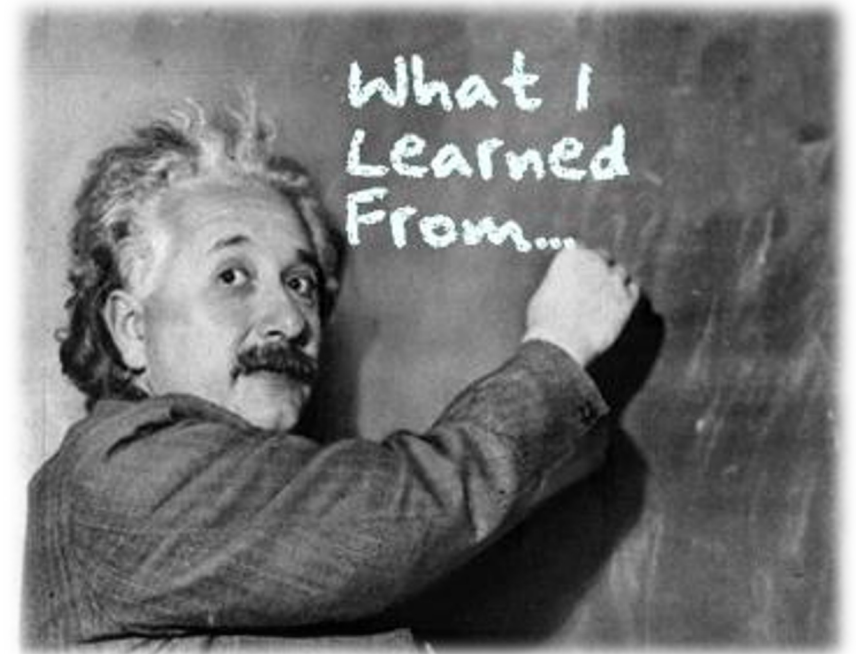
- should be managed as ACUTE STROKE if within 48 hours
- mandates complete cerebrovascular evaluation
- manage or modify risk factors

- **AION/Giant Cell Arteritis:**

- a vascular emergency: TREAT FIRST
- consider in appropriate patient populations
- age, labs, temporal artery biopsy

- **Diplopia:**

- isolated/microvascular can often be managed conservatively
- caution w/ CN III (aneurysm) and intermittent diplopia (GCA)





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