Neuro-ophthalmology emergencies

Clinical history

- How, when, how long ago, for how long…
- Document
- Guide questioning appropriately
- Avoid leading questions, listen to the patient
- Elderly patients ++
- Pupillary reflexes, color vision, confrontation fields

- A. Optic neuropathies
- B. Transient vision loss/ acute arterial ischemia
- C. Papilloedema
- D. Double vision
- E. Non-explained visual loss
- F. Pupils – urgent causes of anisocoria

- **A. Optic neuropathies**
- B. Transient visual loss/ Acute arterial ischemia
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- D. Double vision
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- F. Pupils

Optic neuropathies

- Optic neuritis & inflammatory optic neuropathies
- Ischemic optic neuropathies
  - Non arteritic
  - Arteritic (Giant cell arteritis)
- Other optic neuropathies
  - Compressive
  - Infiltrative
  - Infectious
  - Toxic/nutritional
  - Traumatic
  - .....

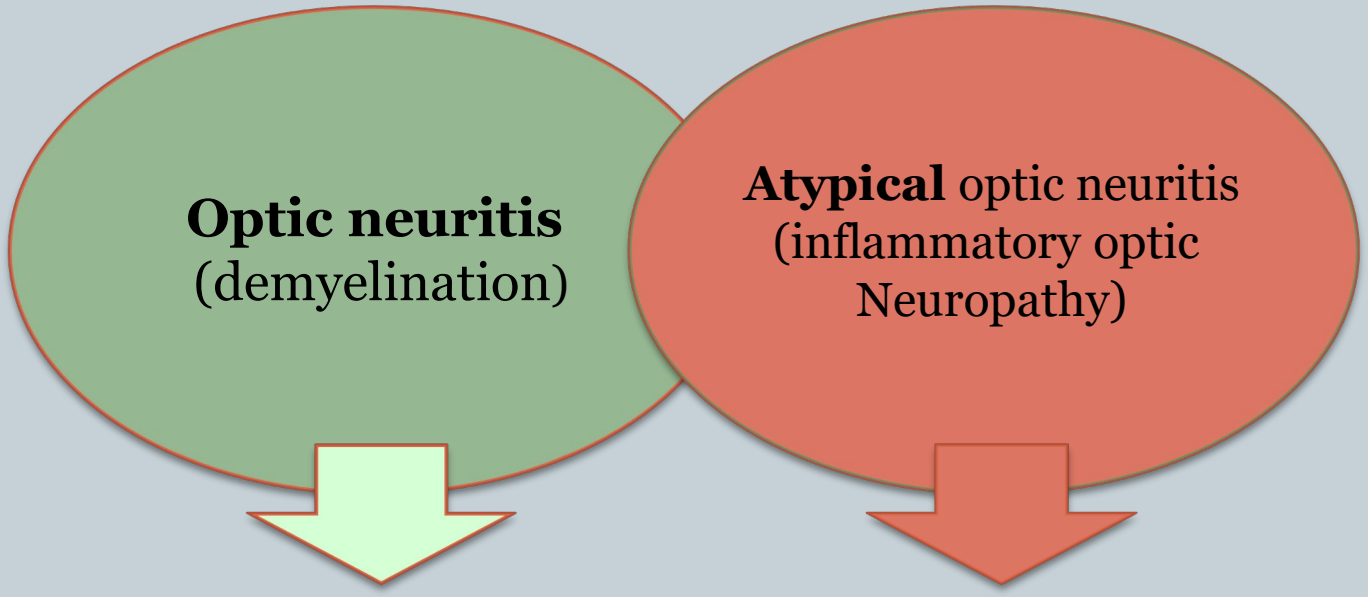
Acute optic neuropathies

?

Presentation, accompanying signs and clinical history

Optic neuritis

→ *Subacute reduction of vision preceded/ accompanied by **pain.***
Dyschromatopsia
Relative afferent pupillary defect
Normal (2/3) or swollen disc (1/3)



Neuro-ophthalmology referral

Urgent investigations
Urgent neuro-ophthalmology referral

Atypical optic neuritis (inflammatory optic Neuropathy)

VA below 6/60 (counting fingers or less)

No pain or intense pain/intense headaches

Pale discs with no previous known history of demyelintion/MS

Age > 45 years or < 15 years

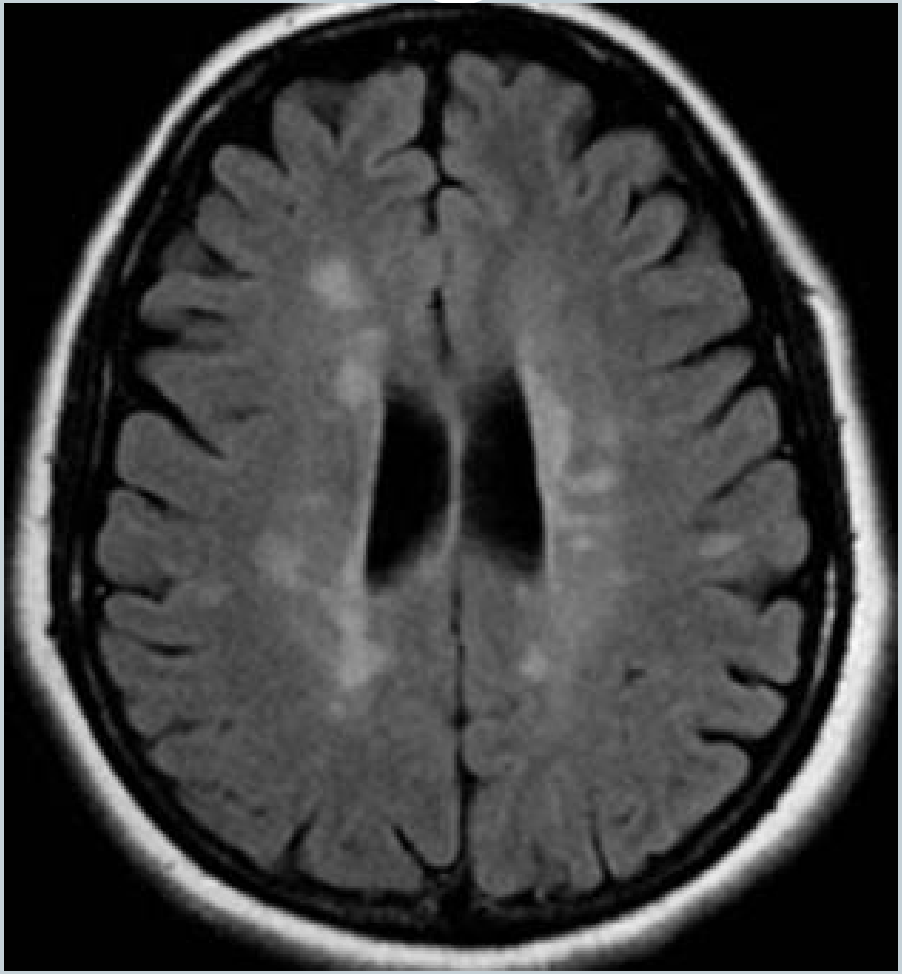
Bilateral

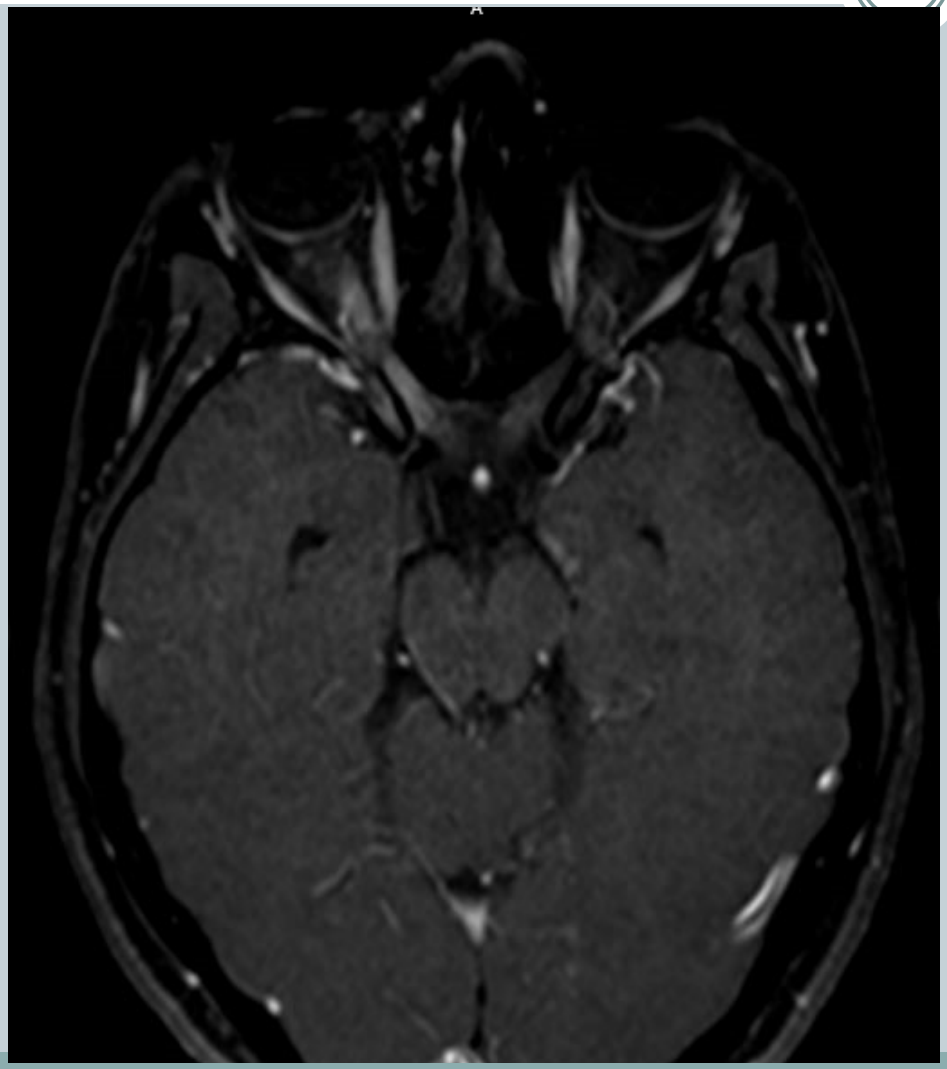
Marked papillitis with presence of haemorrhages, exudates or pallor *

Positive history of immunosuppression or autoimmune disease, ongoing systemic symptoms suggesting Inflammation, current or recent infection, other acute neurological symptoms

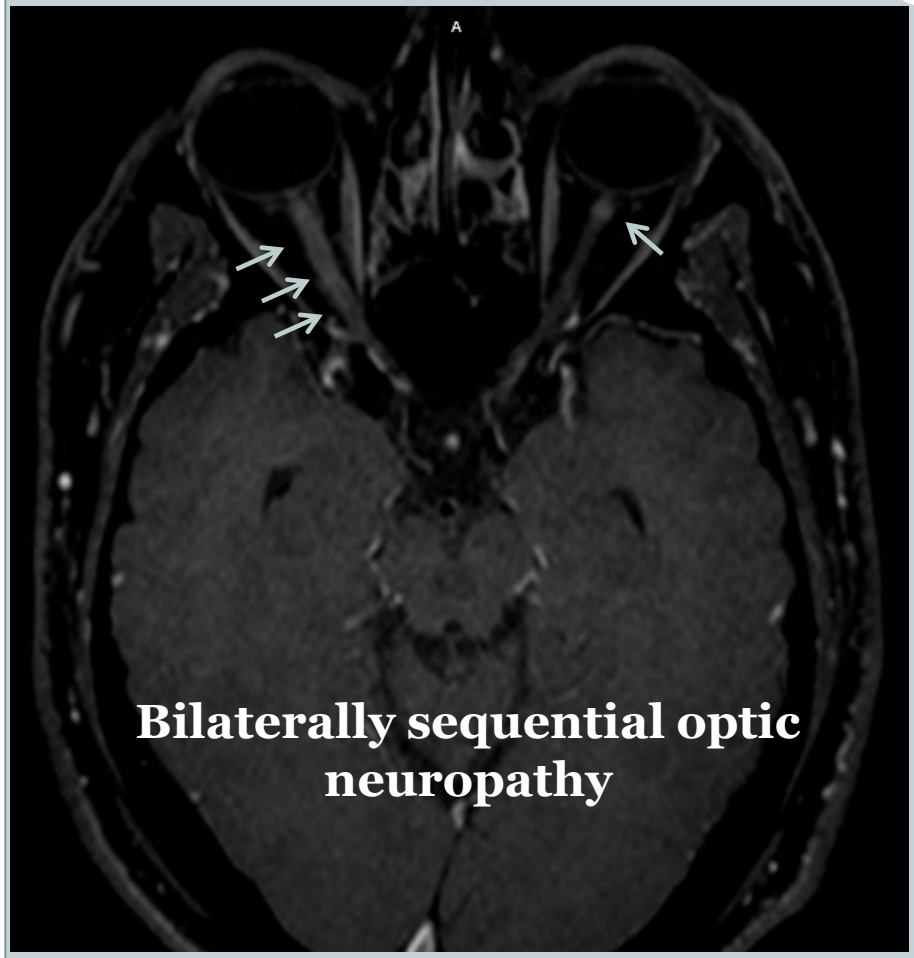
Pain persisting > 2/52

Vision loss progressing over 2 weeks since onset



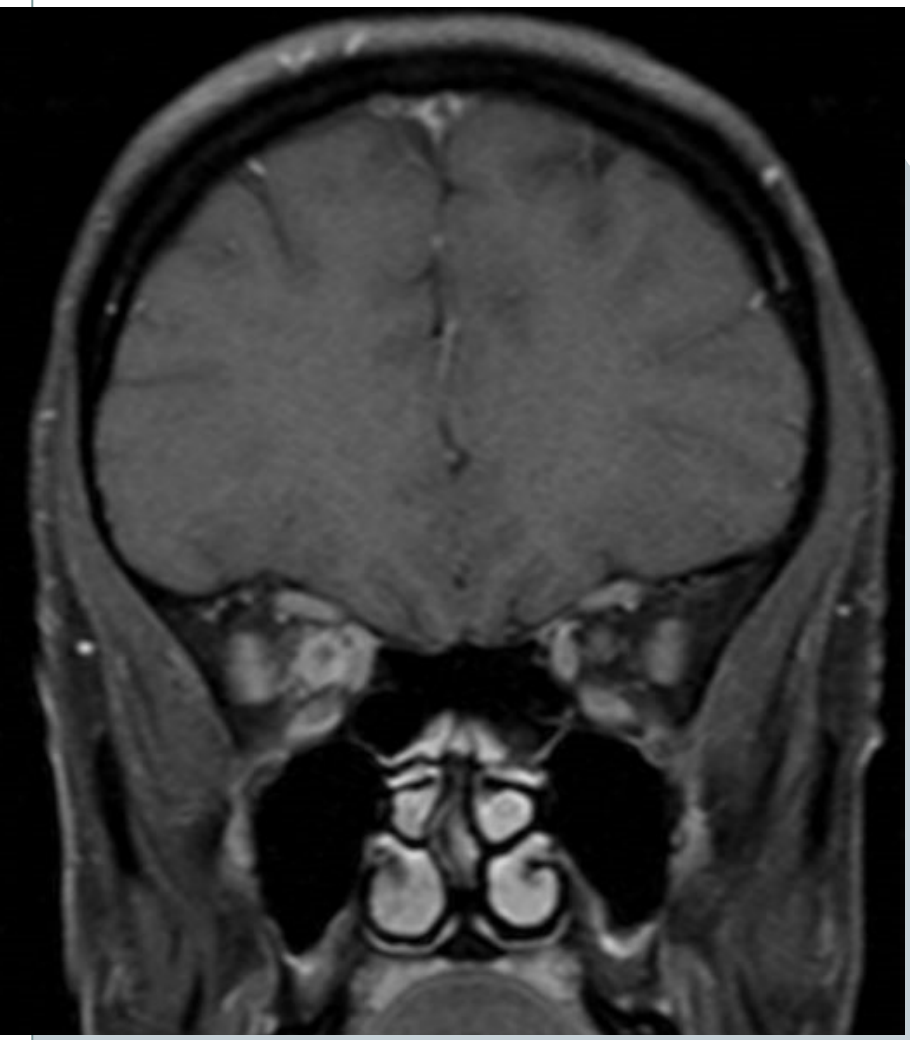


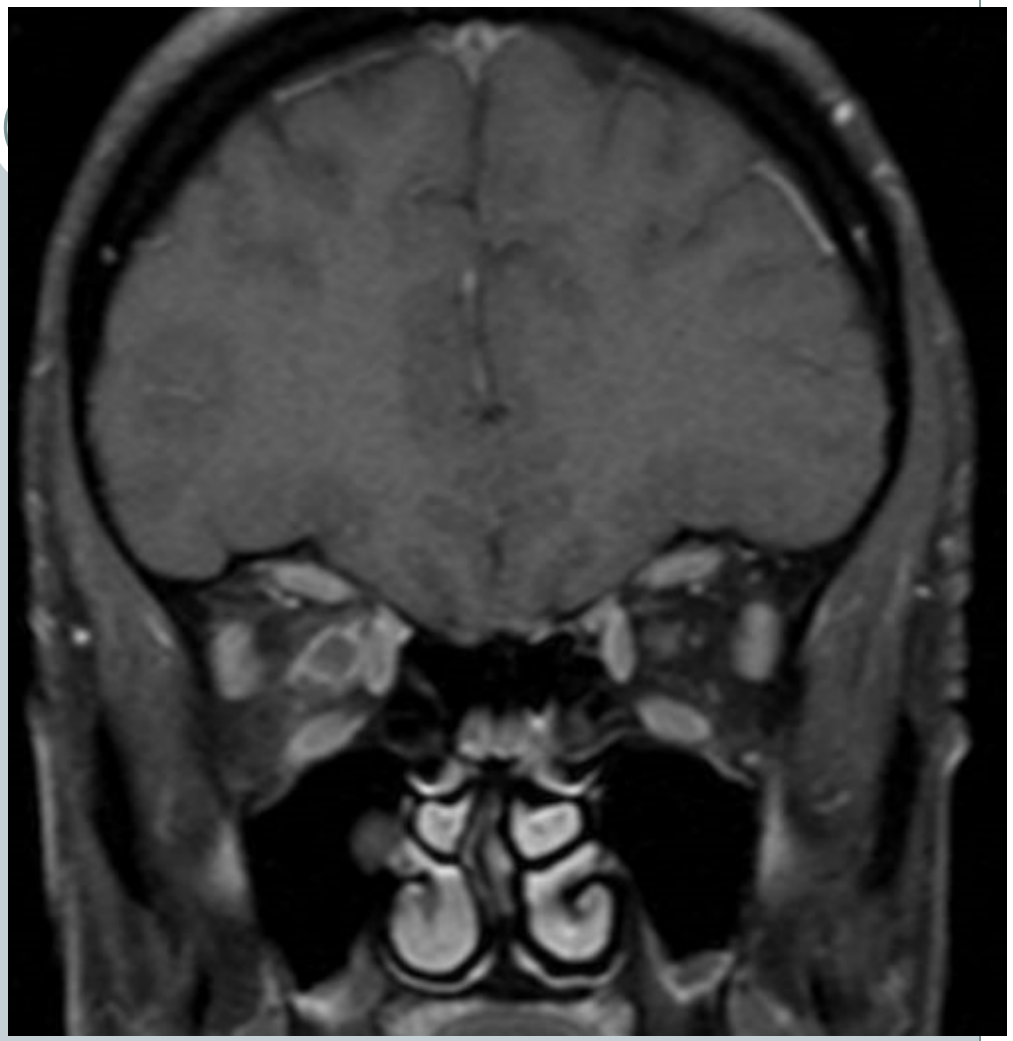
- 24 y old woman
- 4/7 history of rapidly progressive right eye visual loss
- Pain ++
- LONG SEGMENT OF ENHANCEMENT
- Aquaporin 4 +
- Prompt treatment IV steroids + AZA



- **Antibody mediated neuro-inflammation (neuro-myelitis optica spectrum)**
  - **Aquaporin-4 antibody (astrocyte)**
  - **Anti-MOG antibody (oligodendrocyte)**
- **Post infectious**
- **Other**

- **Neuro-ophthalmic emergency +++ (time=vision)**
- **Urgent investigations**
- **High dose steroids +/- plasma exchange**





Granulomatous/infiltrative radiological pattern

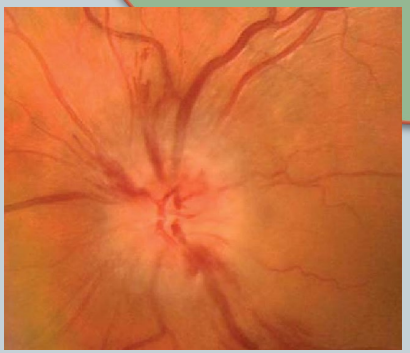
Malignancy, sarcoid, TB, ANCA vasculitis..

LP, CT TAP...

Ischemic optic neuropathy

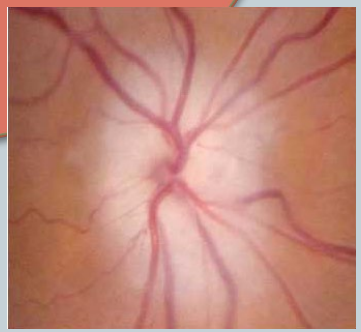
Non arteritic

Arteritic = Giant Cell Arteritis



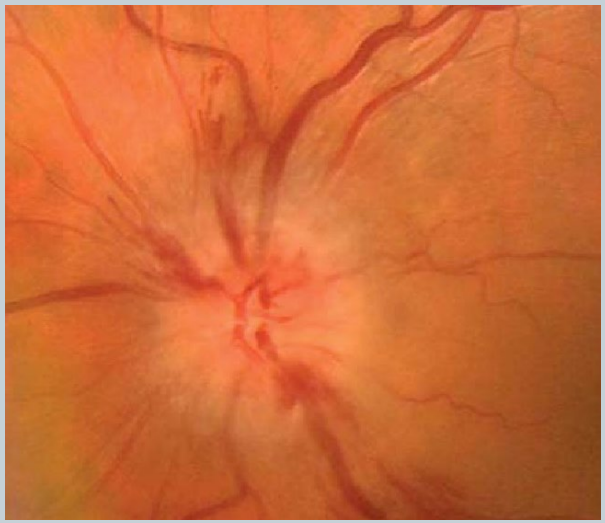
Absence of red flags
Relative good vision
Normal inflammatory makers

Urgent
CRP, ESR, FBC
Clinical assessment : GCA??

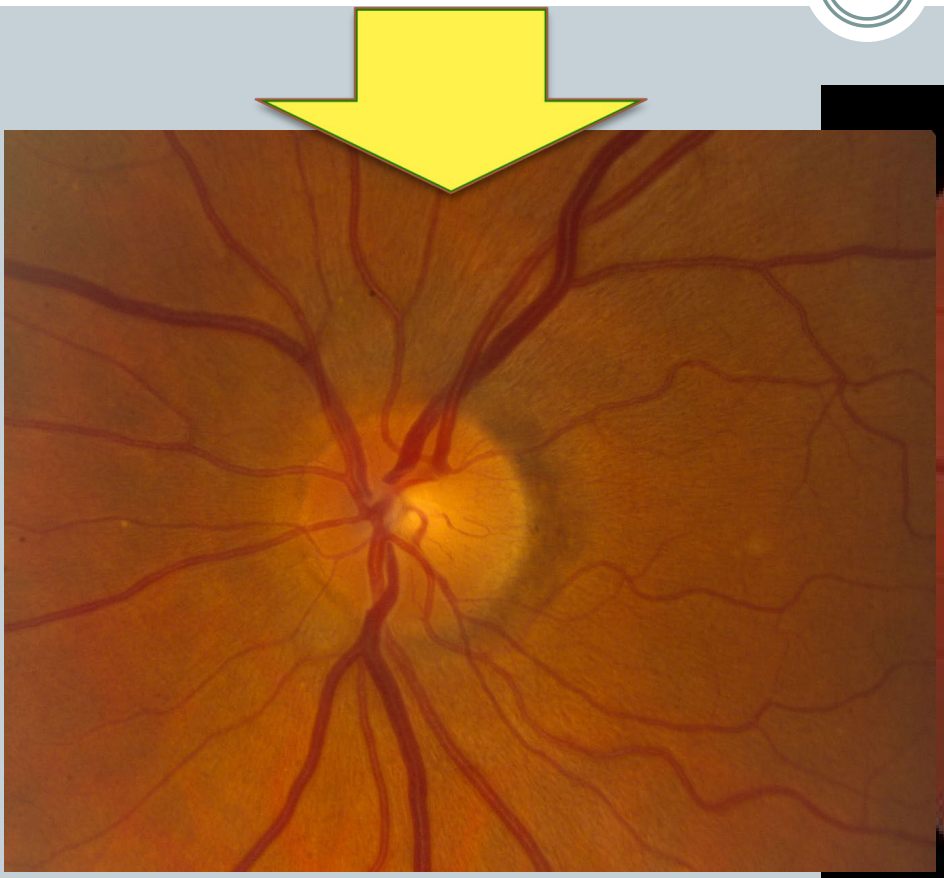


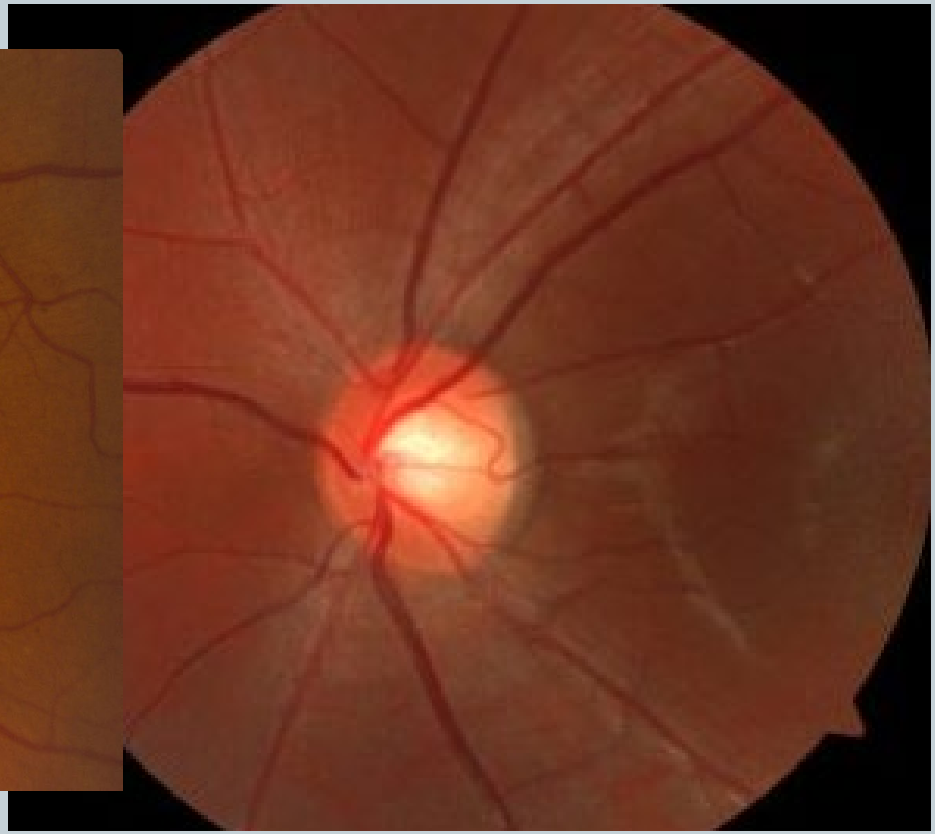
Visual prodromes
Variable vision
Systemic signs
Raised inflammatory markers

Non arteritic optic neuropathy

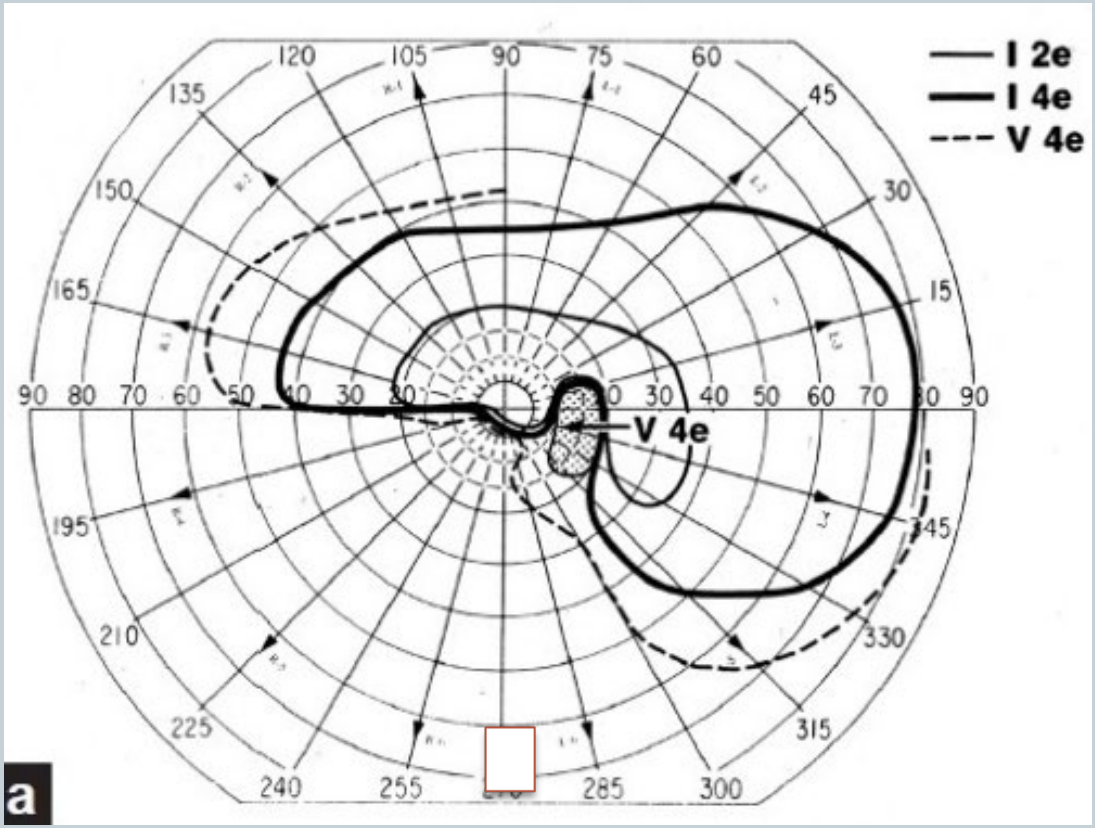


Hyperemic swelling
Small disc
Painless
Altitudinal field defect often (other optic neuropathies can also have altitudinal defects)
Sudden onset and non progressive
Often central vision preserved
No dyschromatopsia if vision relatively preserved





Small disc
(small cup/disc ratio)
…. but other optic neuropathies happen with small cups



Altitudinal defect

Unilateral optic nerve swelling

Lidia Alvarez
2017

Complete medical & system history & visual function (*including acuity, colour vision and fields with CONFRONTATION*)

Most senior doctor in casualty to assess discs and history!!!!

Blood pressure

Disc photos or OCT optic nerve fiver layer: Document appearance

Asymptomatic and *normal* visual function (incidental)

Is it really swollen?

Yes → Formal field s & Neuro-ophthalmology opinion

No → A Small crowded discs
B Tilted discs?
C Optic nerve head drusen?
D Other pseudo-swelling?

→ As per bilateral swollen disc guideline

Symptomatic : reduced vision or field defect

Age <45

Compatible with a typical optic neuritis? * see guideline

Yes → Review in 2-3 weeks to confirm recovery signs + Neuro-ophthalmology referral

No → **Urgent neuro-ophth opinion**
+ /- Optic neuropathy bloods
+/- MRI orbits

Age > 50

URGENT CRP, ESR , FBC + urine dipstick *in A+E*

Raised inflammatory markers

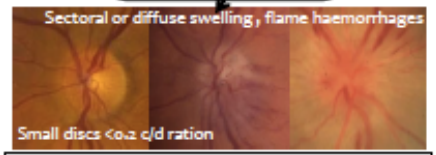
GCA compatible systemic symptoms? (sometimes absent)

URGENT senior A+E Opinion : GCA?

Normal inflammatory markers

In keeping with NA-AION? → (Painless + Acute onset + Small discs with unilateral hyperemic swelling) and no SYSTEMIC RED FLAGS? (see below)

No → Urgent neuro-ophth opinion



Hyperemic swelling (no pallor acutely), altitudinal field defect (often) ; relatively preserved colour vision and visual acuity if fixation spared (often), swelling resolves in 6-10 weeks

→ Neuro-ophthalmology referral

Systemic red flags : immunosuppression, autoimmune disesase, acute disease process(severe anemia/ blood loss, severe hypotension, infection…), new onset headache/ severe headache, neurological focality.

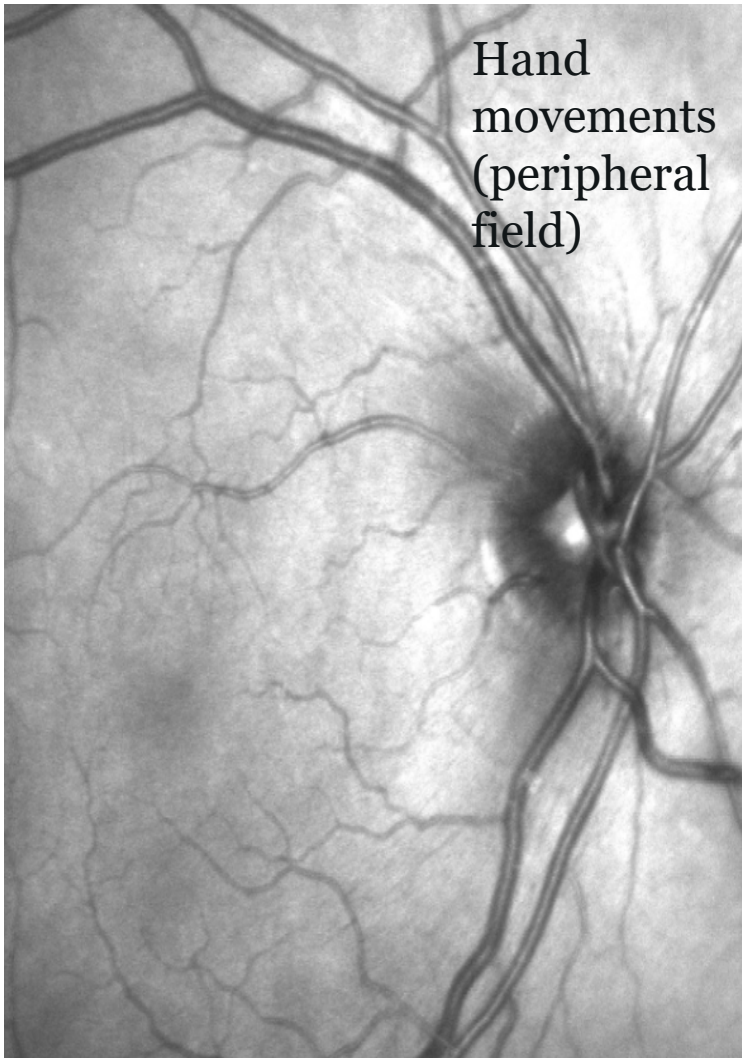
Other optic neuropathies

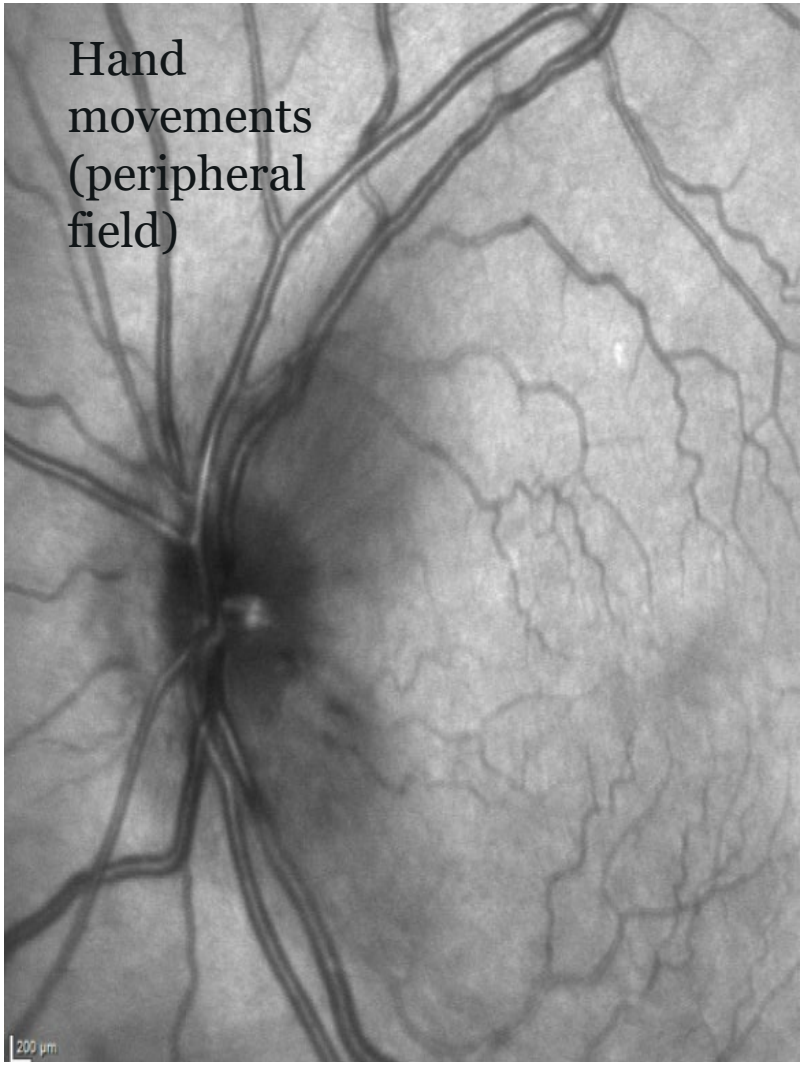
Toxic /nutritional : alcohol, medications (ETHAMBUTOL)→ reversible if discontinued

Traumatic

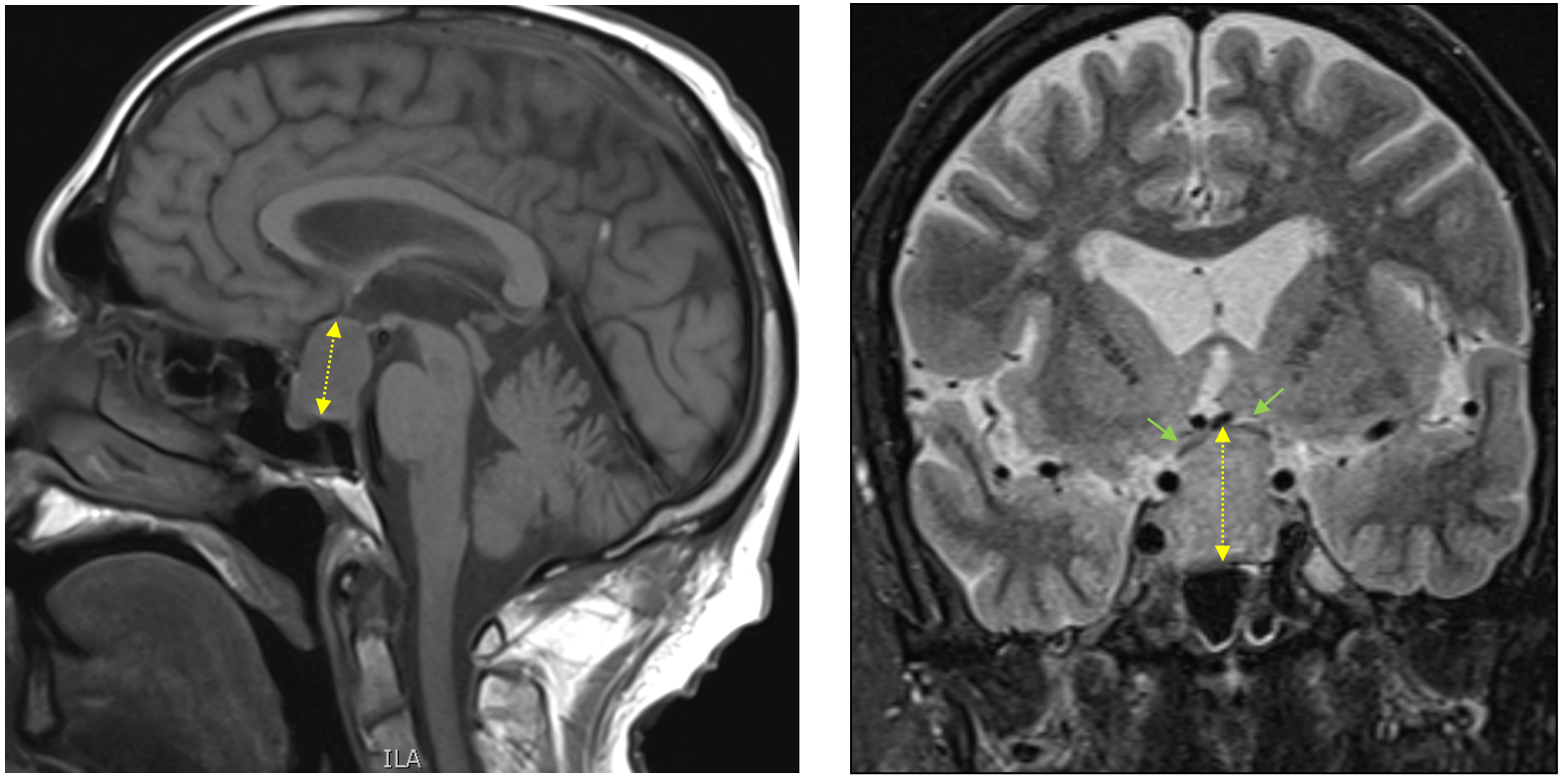
Infiltrative

Compressive

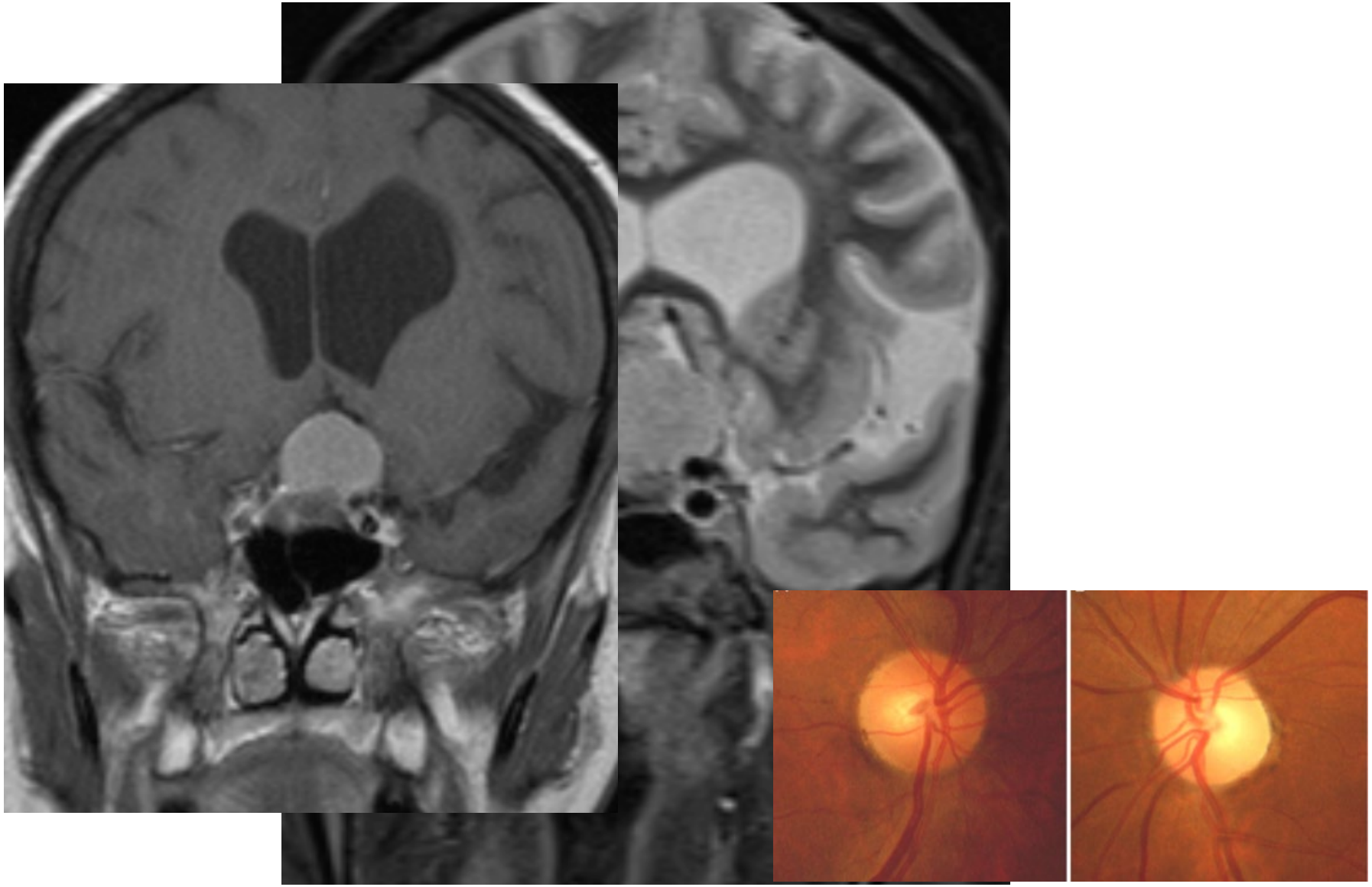




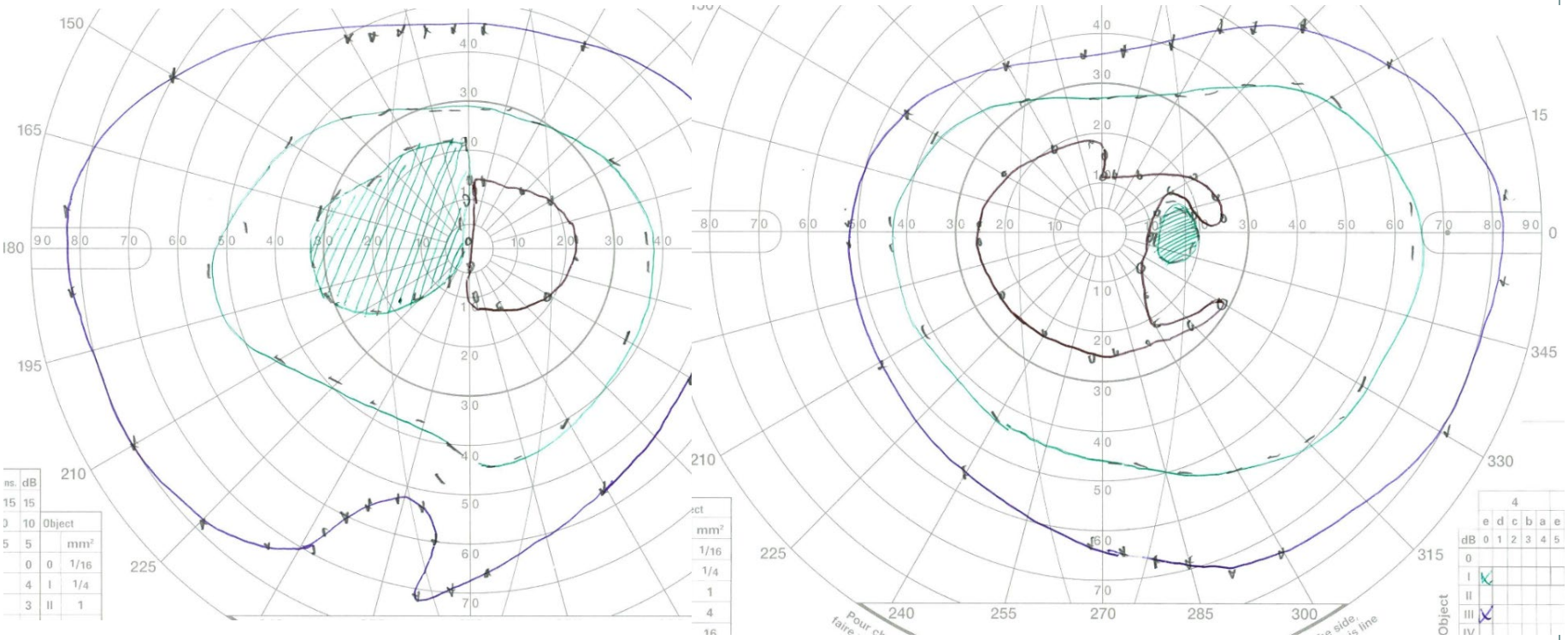
Alcohol dependent patient attending A+E with **acute** visual loss
Admit for investigations



Pituitary adenoma

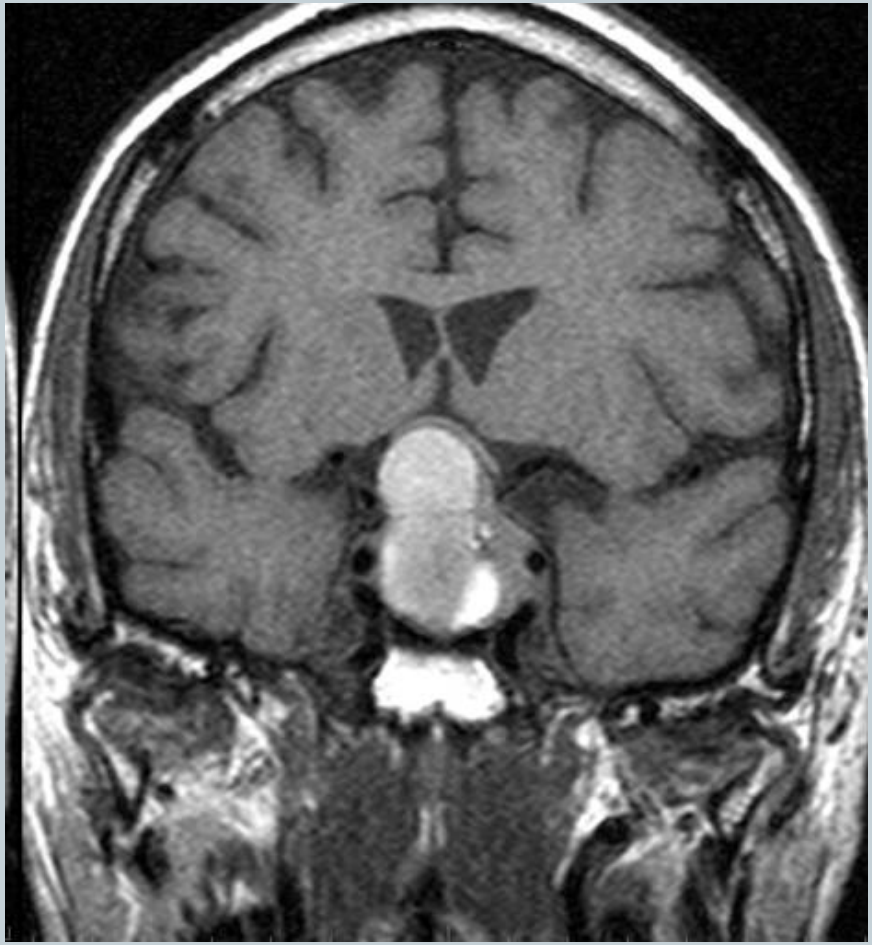


2/12 history of progressive visual loss in the left eye with mild reduction in acuity LE and mild temporal pallor



Bitemporal hemianopia L>R

Pituitary apoplexy



Acute haemorrhage/infarction pituitary gland (harbouring adenoma)

Rapid visual loss

Hypopituitarism

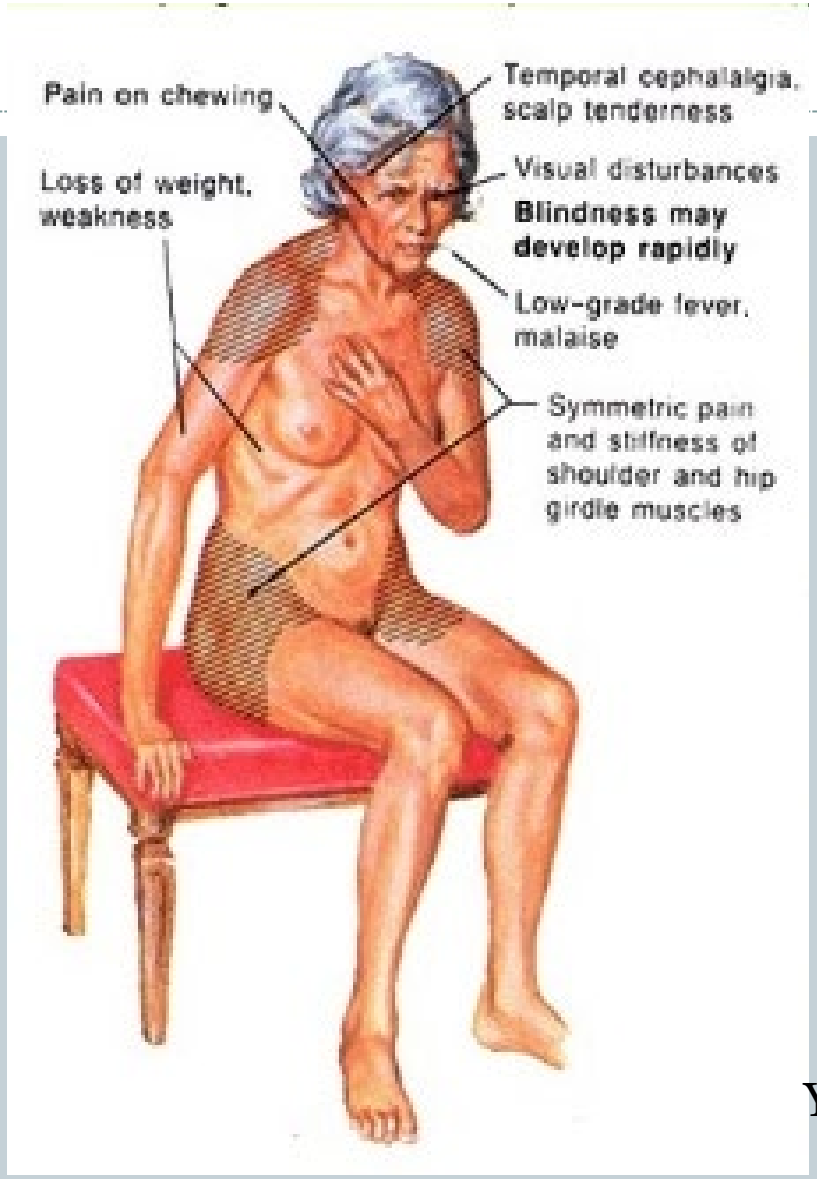
Adrenal insufficiency→ fatigue++, nausea/vomiting/ GI, hypotension, hypoglycemia..

Sudden+
Headache ++++

Giant Cell Arteritis

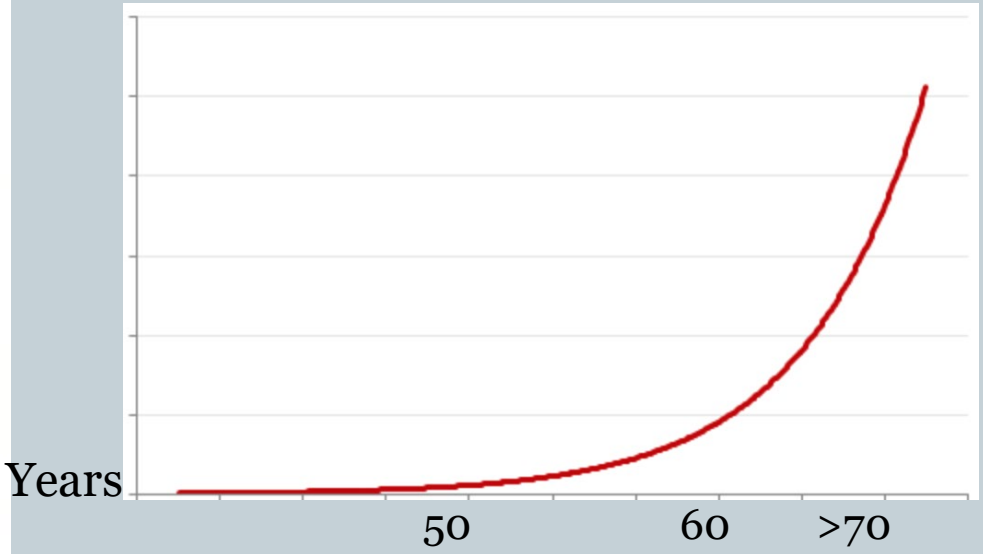
- 1st disease as cause of litigation in ophthalmology

- Why?
  - Loss of vision **preventable** if not established
  - SECOND EYE can follow quickly
  - Missing the diagnosis → blindness, stroke, death



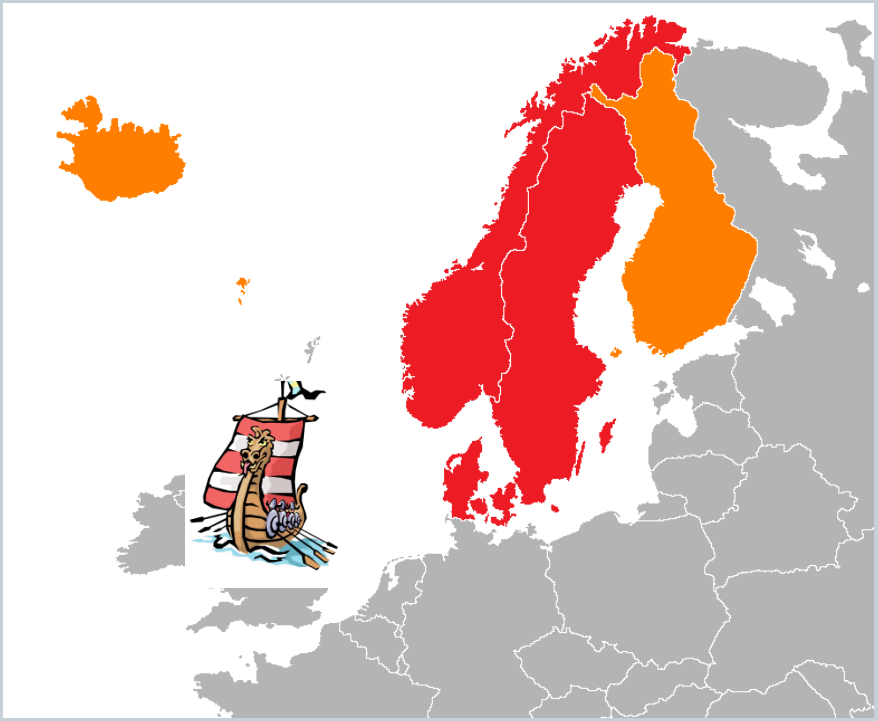
PROTOTYPE PATIENT

- Over 50 (most often over 60)
- White caucasian >>> other ethnicities
- Exponential increase with age



High incidence in the UK

- Highest incidence Scandinavian countries and **populations of Scandinavian descent** (linked to certain HLA phenotypes)



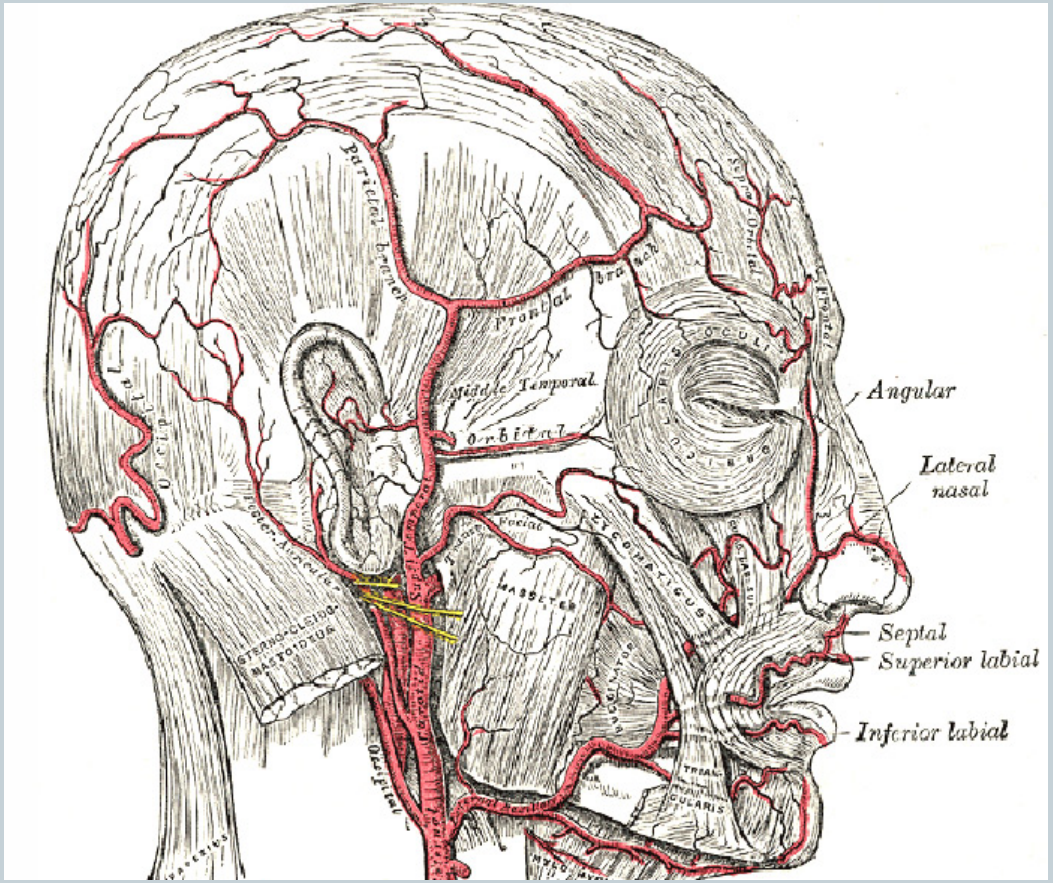
Incidence in studies:

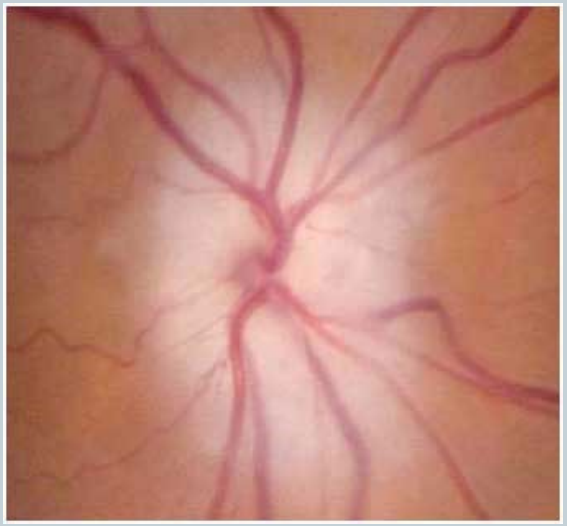
- Norway **33 per 100.000**
- Iceland **30 per 100.000**
- **United Kingdom** **22 per 100.000**
- North of Spain 10 per 100.000
- African American population studies → **0.4 per 100.000**

Very low and unknown incidence → Asia, African continent ...??

Clinical presentations / subtypes

- **Cranial arteritis** (temporal, facial, ophthalmic (posterior ciliary), vertebral…)

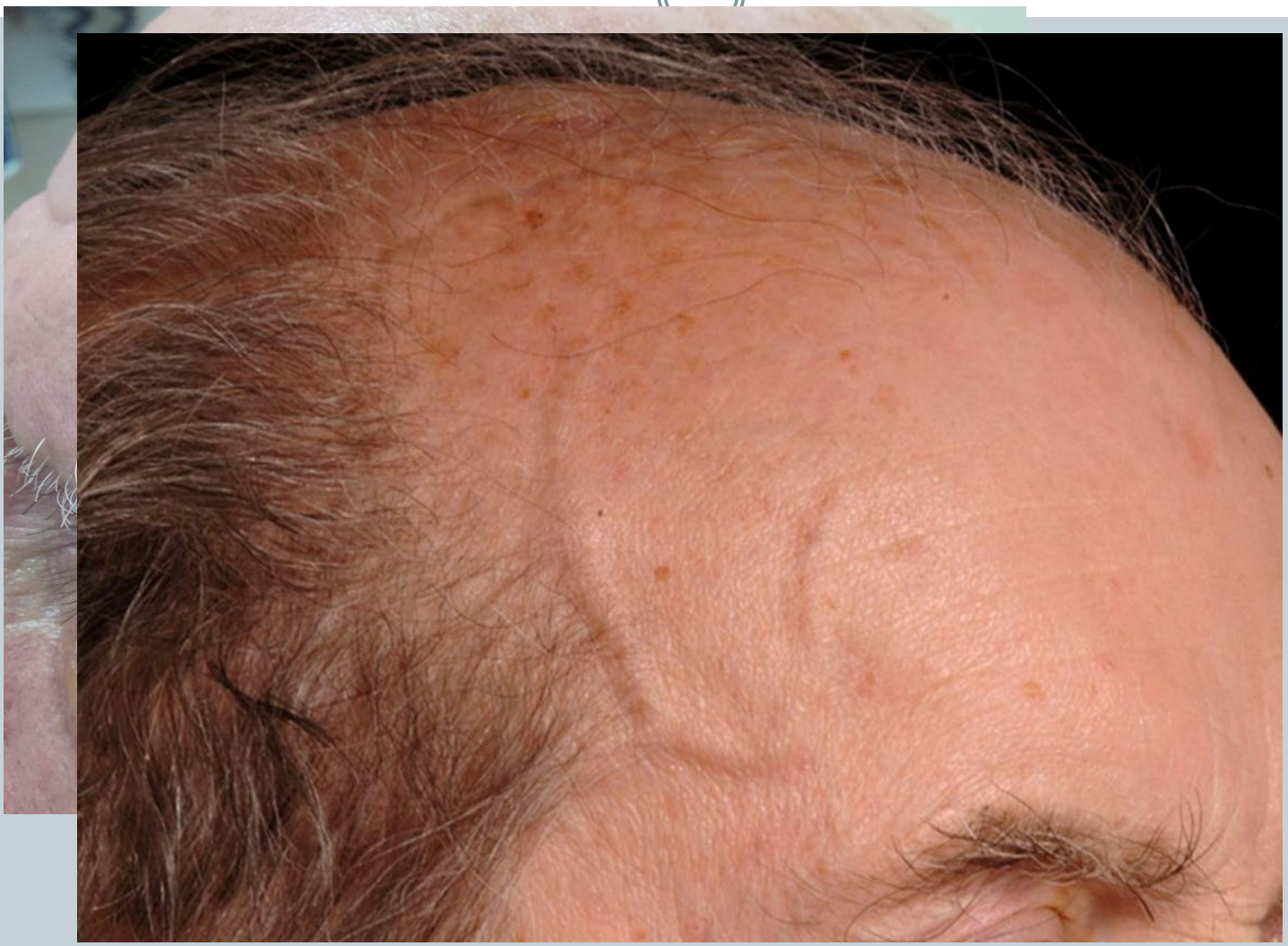




Jaw claudication, temporal headache/ pain, loss of vision…

Abnormal temporal artery

thickenned/normal lumen but hardened (lost pulse in areas..)/ kinks, nodules



Often **abnormal (not always, and not always evident)**

Clinical presentations / subtypes

- **Silent** /great vessel disease →fatigue, weight loss, fever of unknown origin
- **Occult** (only ophthalmic symptoms)→ up to 20%[1]
- **Normal inflammatory markers** (very infrequent 3-5%)[2,3]

1. Occult giant cell arteritis: ocular manifestations. Am J Ophthalmol.Apr; 1998 125(4):521-526.
2. Giant cell arteritis with normal ESR and/or CRP is rare, but not unique!. *Eye* 2013; 27: 1418–1419.
3. Giant cell arteritis with normal inflammatory markers. *Acta Ophthalmol Scand* 2007; 85: 460.

Inflammatory makers

- Very useful .. as ***almost always*** elevated
- CRP more sensitive

- Often CRP > 50 and ESR > 60
- But… a significant proportion only discretely elevated markers
- CRP 18, 20, 30 … happens and is not uncommon

Ocular signs and symptoms

Visual disturbance/ visual loss → ophthalmic artery >> short posterior ciliary arteries

- Amaurosis fugax
- Peripheral visual loss
- "Blurred vision"
- Marked visual loss –PL- NPL

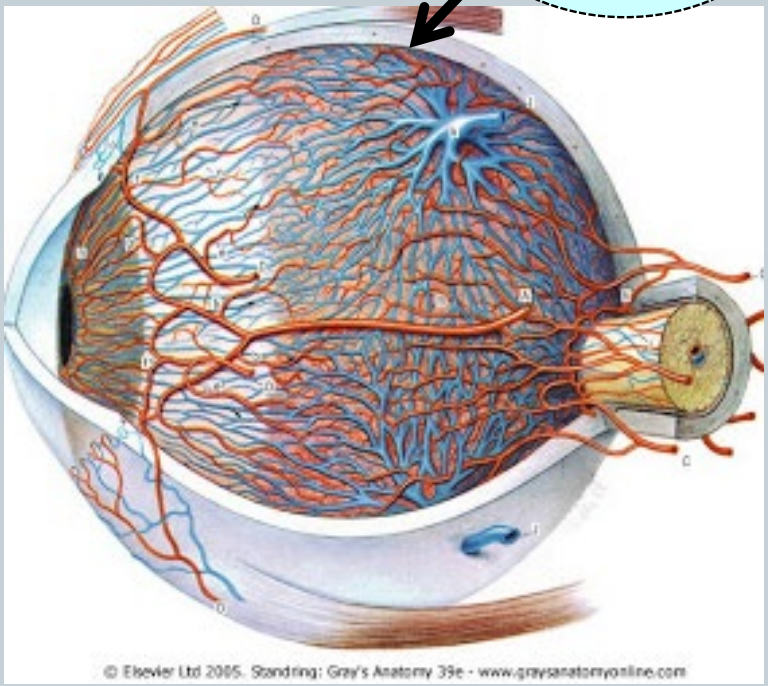
Diplopia

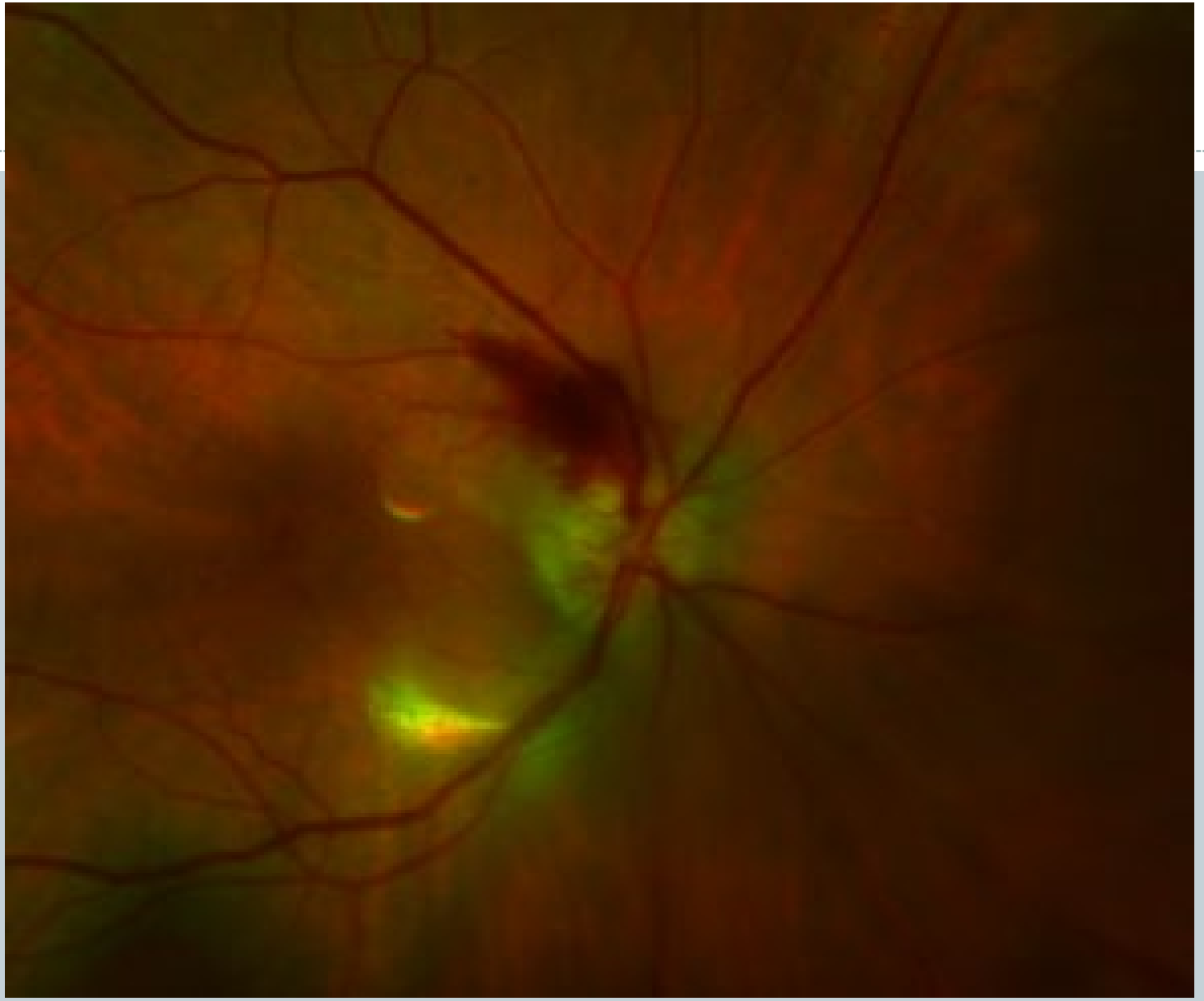
- Intermittent diplopia

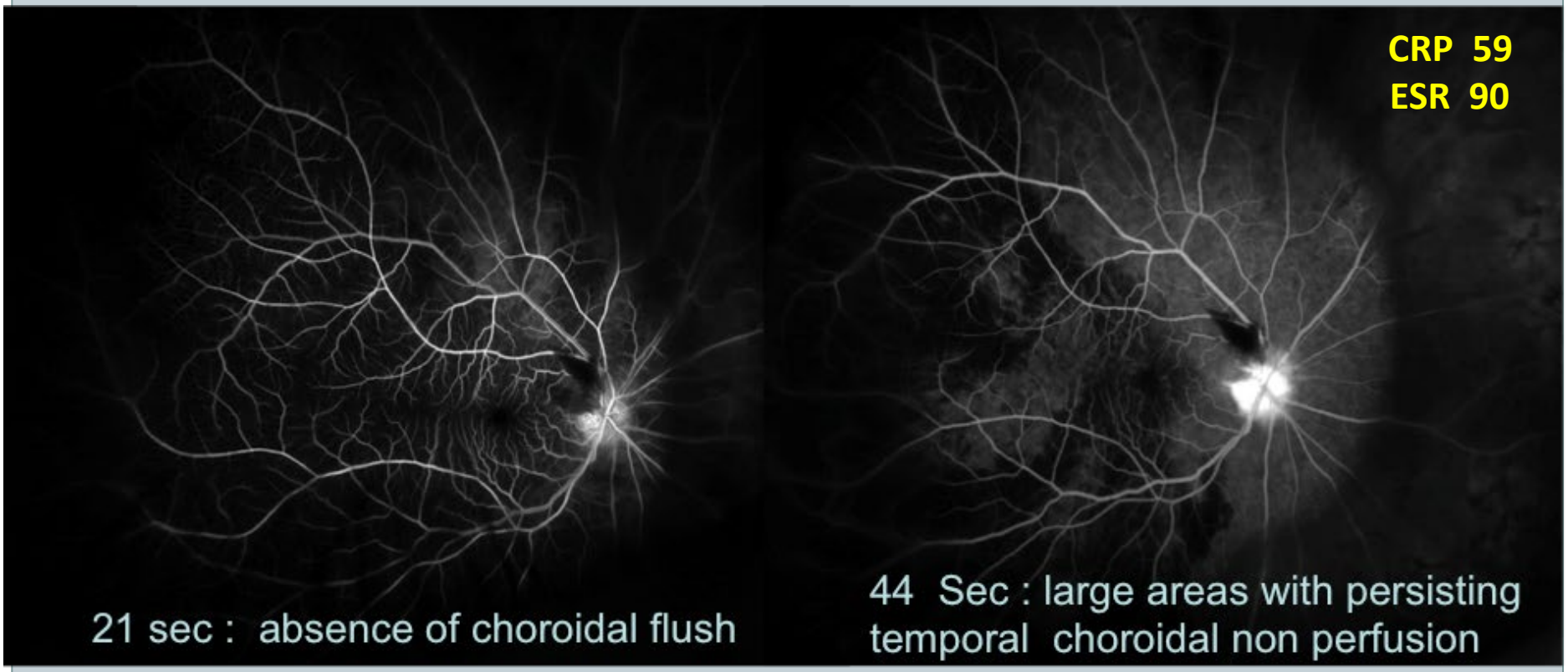
Ciliary arteries

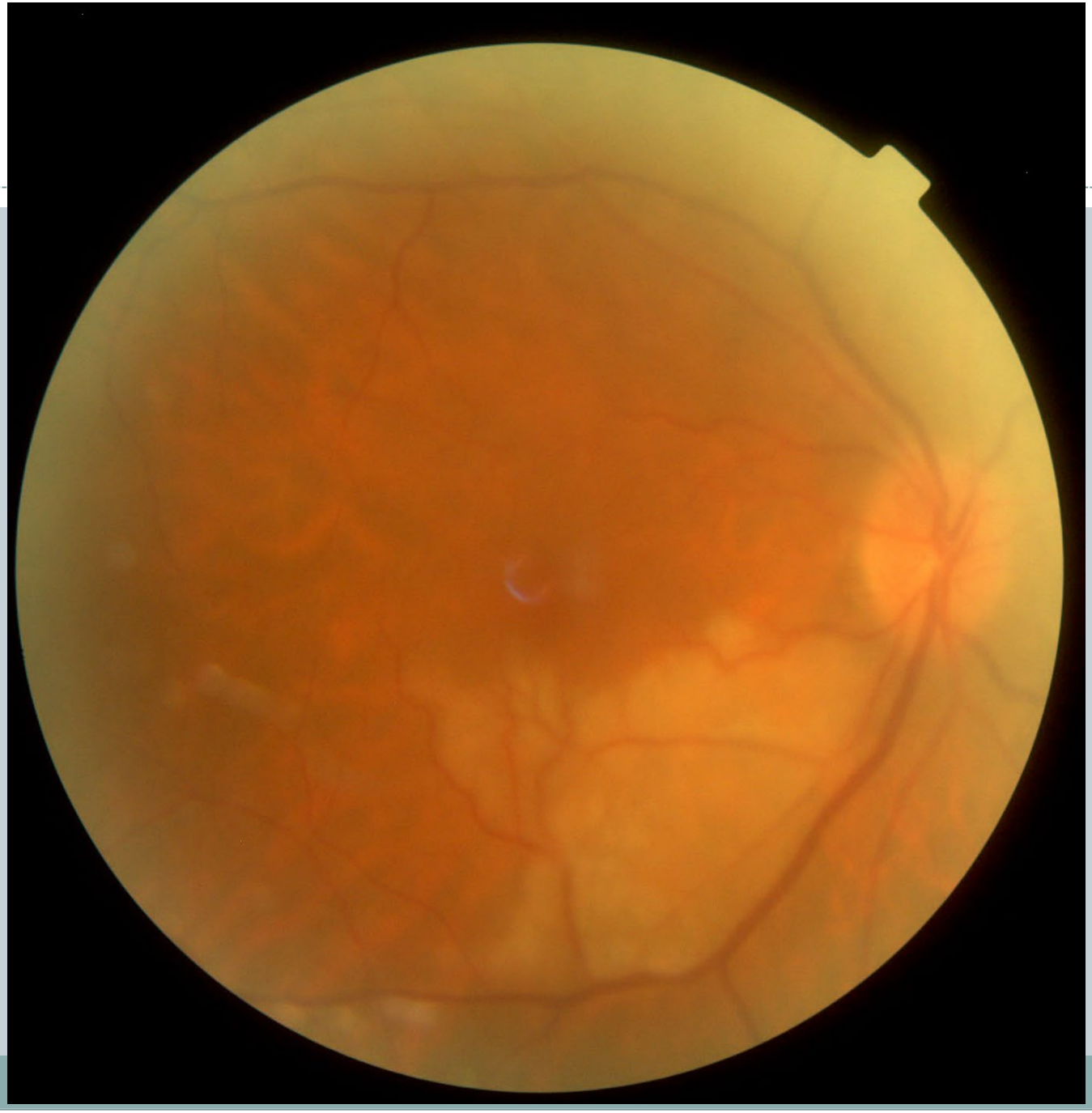
↓

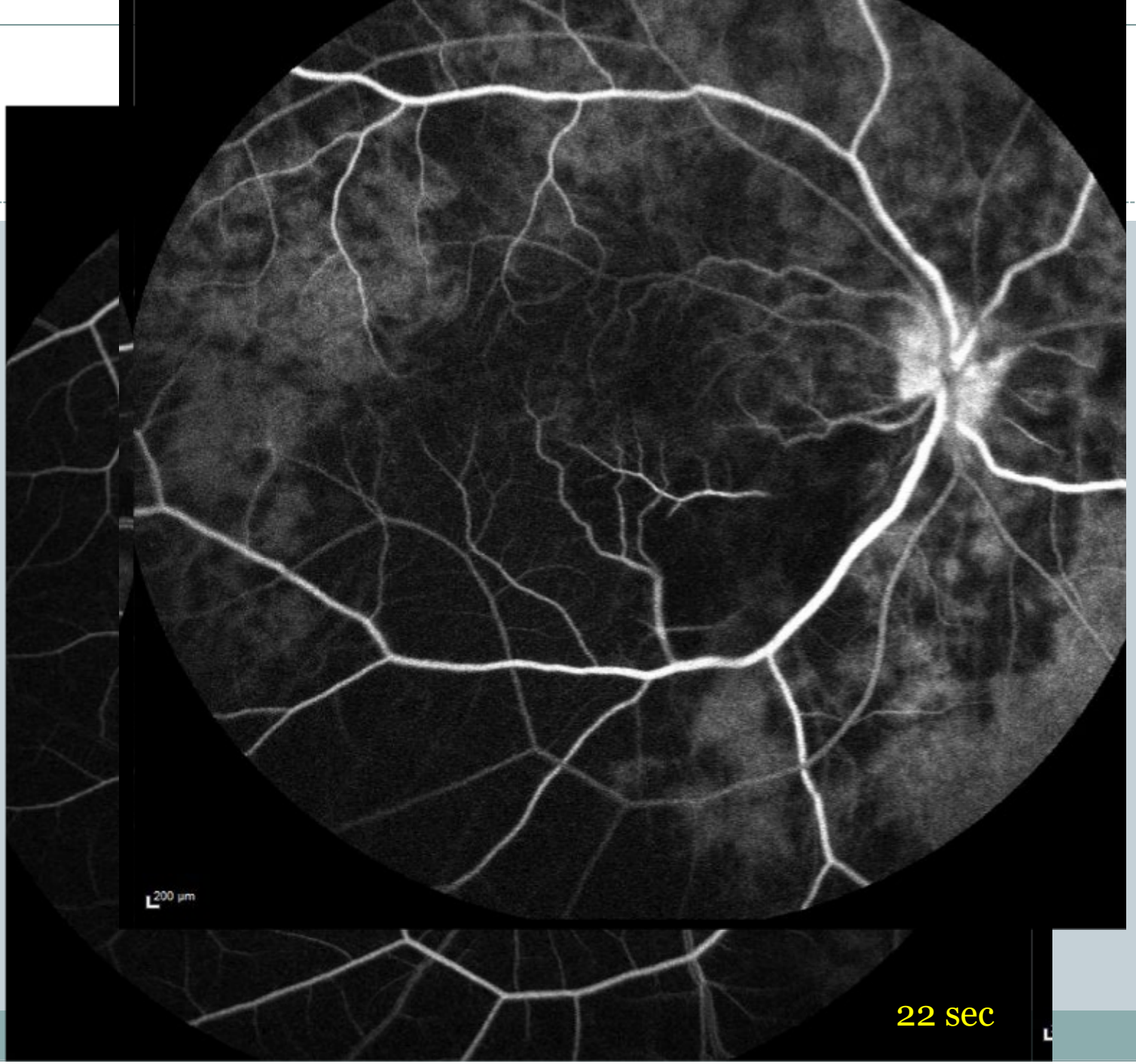
optic nerve head & choroid



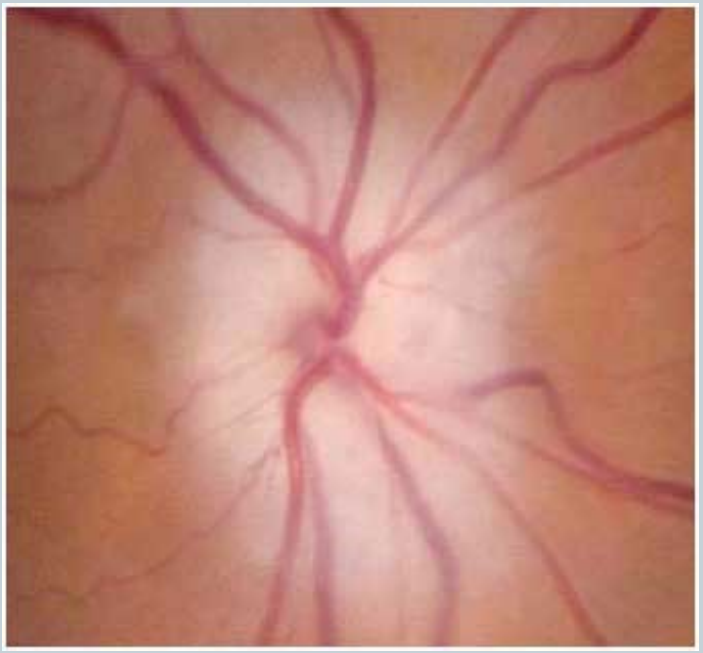



CRP 59
ESR 90
21 sec : absence of choroidal flush
44 Sec : large areas with persisting temporal choroidal non perfusion



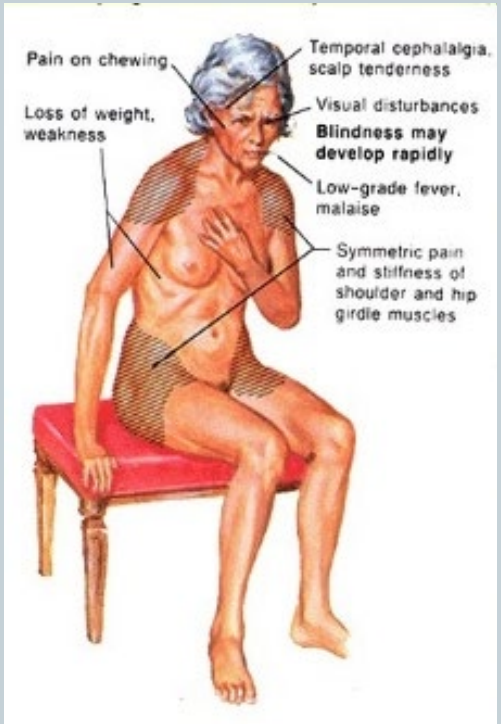

200 µm
22 sec

ARTERITIC
“pallid oedema”



In a susceptible subject…

→ consider likelihood of GCA URGENTLY when :
→ recent **acquired diplopia** or
→ **acute arterial ischemic signs or symptoms**
→ (CRAO, BRAO, amaurosis fugax, non- explained reduction of vision)



- Consequences of missing potential GCA weighted against potential harm from steroids.
- Involve 4th oncall/ consultant
- Unless diagnosis is unequivocal MUST be supported by a temporal artery biopsy→ book and arrange

- The second eye may follow quickly→ no DELAYS in treatment!

- A. Optic neuropathies
- **B. Transient vision loss/ acute arterial ischemia**
- **C. Papilloedema**
- D. Double vision
  - Orthoptics assessment
  - Complete cranial nerve examination
  - Intermittent in the elderly
- E. Non-explained visual loss
  - In the elderly = always consider GCA
- F. Pupils

Acute arterial ocular ischemia transient visual loss

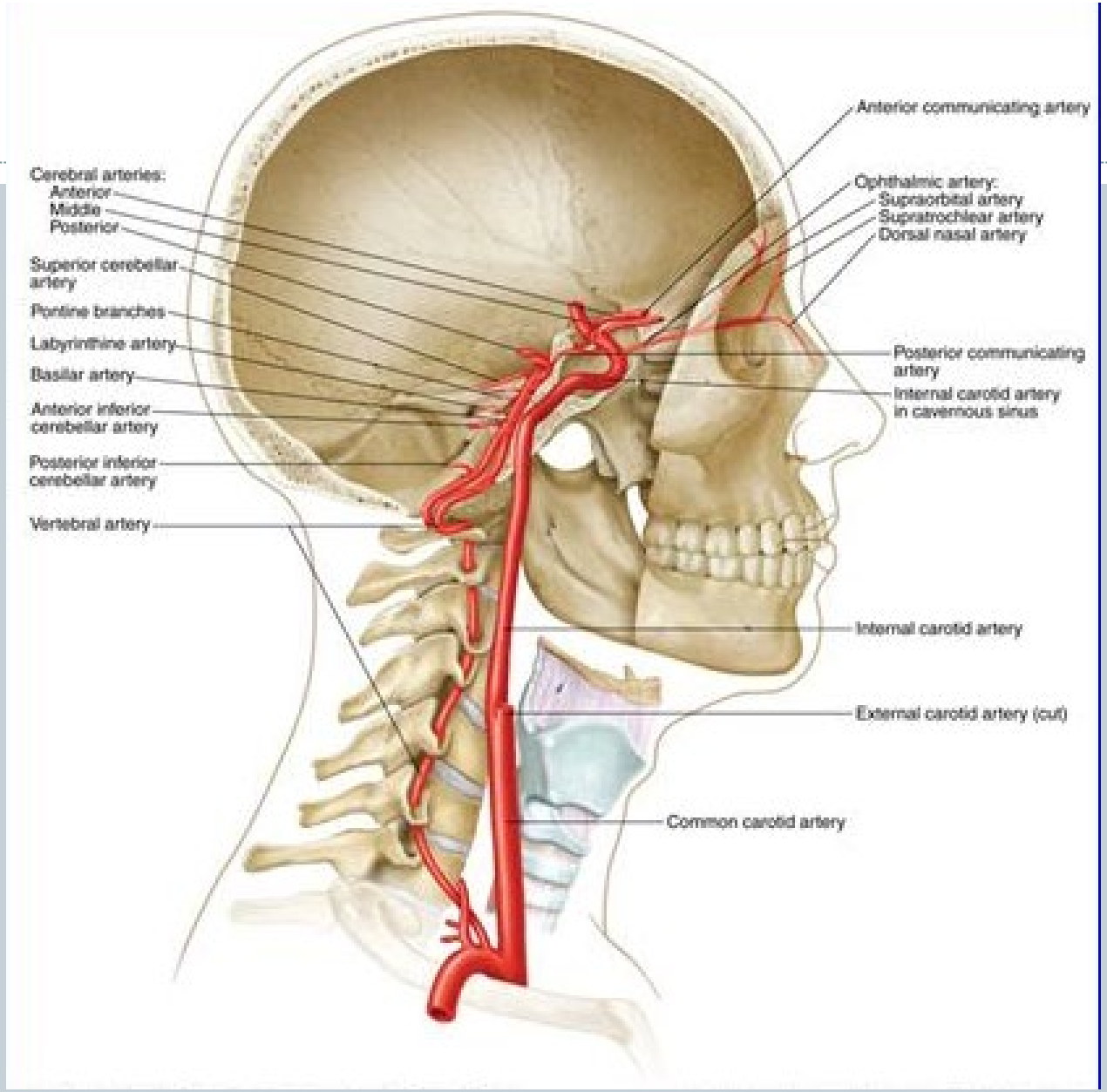
1- TRANSIENT VISUAL LOSS " amaurosis fugax"

→ Black out of vision in one eye starting suddenly lasting between 1 minute to 5-10 minutes- 1 h

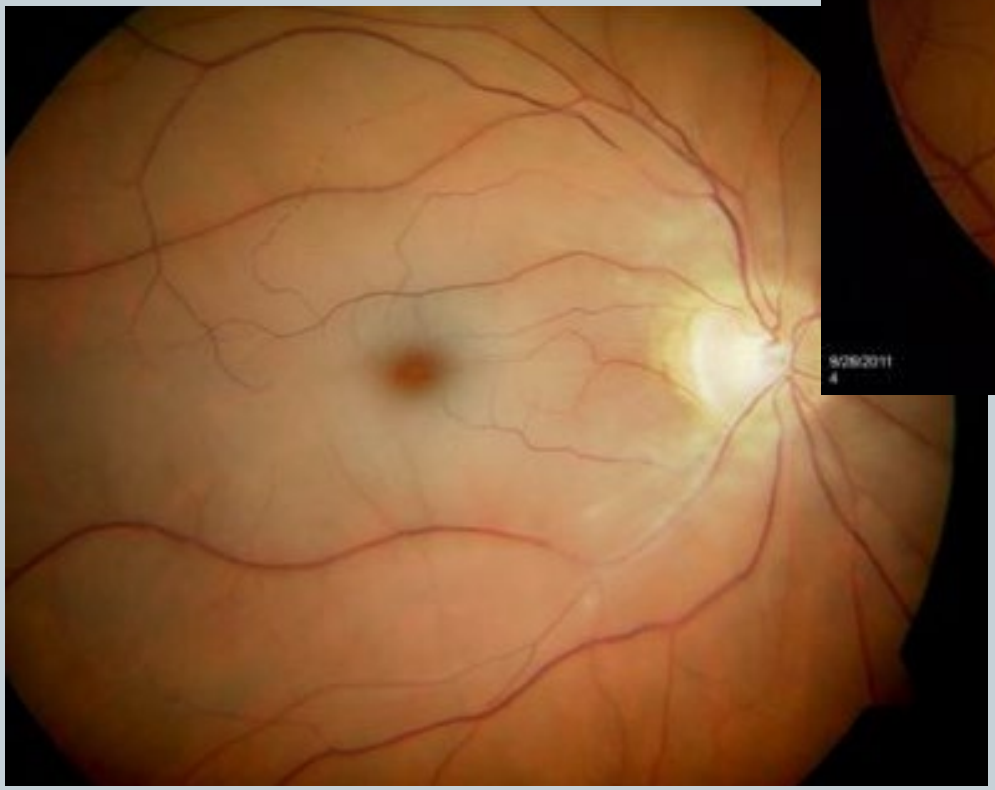
In > 50 -> IS THIS GCA? - ESR/CRP

ABNORMAL→ consider a diagnosis of GCA

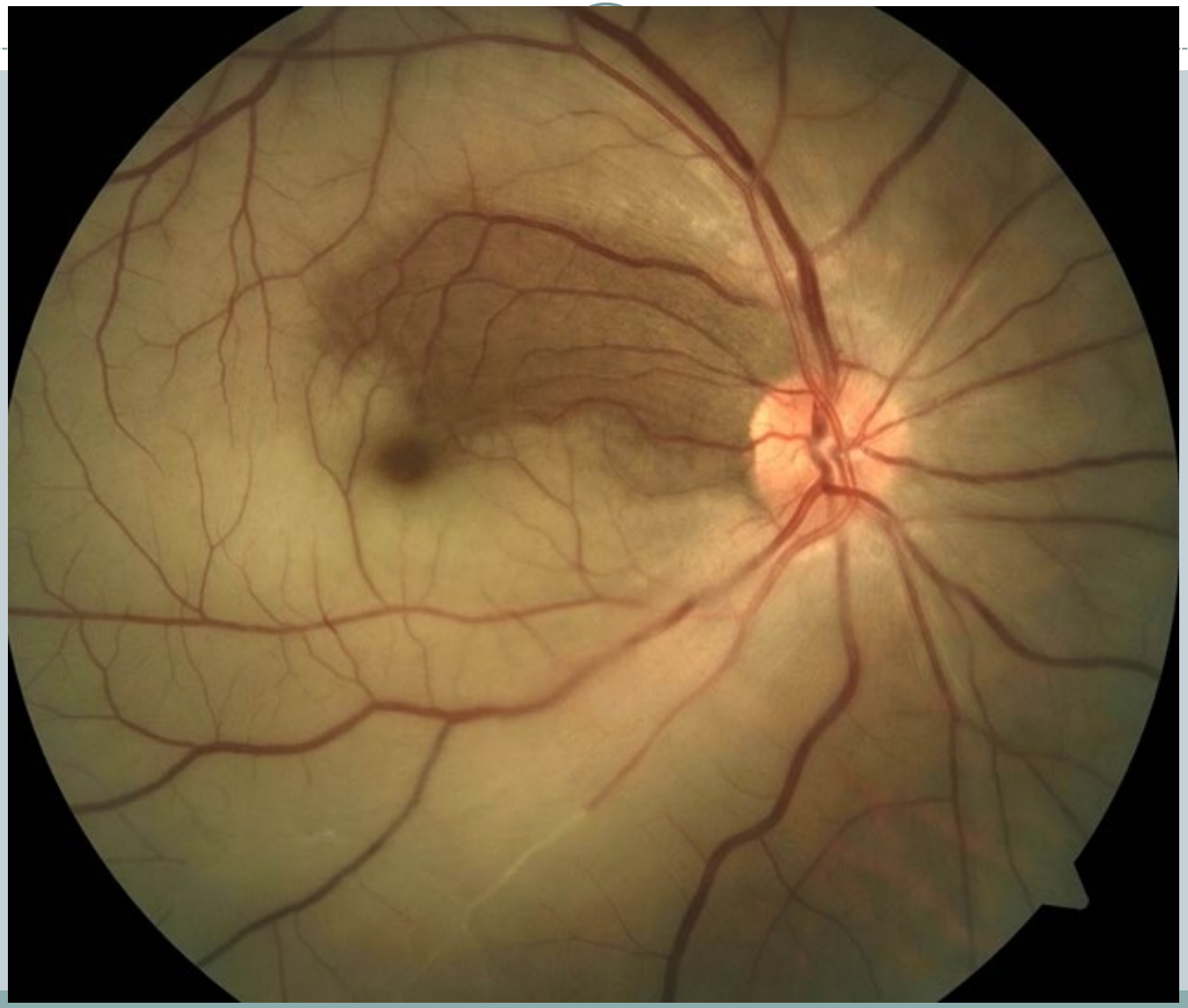
NORMAL→ refer STROKE UNIT (TIA)


Anterior communicating artery
Cerebral arteries:
Anterior
Middle
Posterior
Ophthalmic artery:
Supraorbital artery
Supratrochlear artery
Dorsal nasal artery
Superior cerebellar artery
Pontine branches
Labyrinthine artery
Basilar artery
Anterior inferior cerebellar artery
Posterior communicating artery
Internal carotid artery in cavernous sinus
Posterior inferior cerebellar artery
Vertebral artery
Internal carotid artery
External carotid artery (cut)
Common carotid artery

CENTRAL RETINAL ARTERY OCCLUSION



BRANCH RETINAL ARTERY OCCLUSION

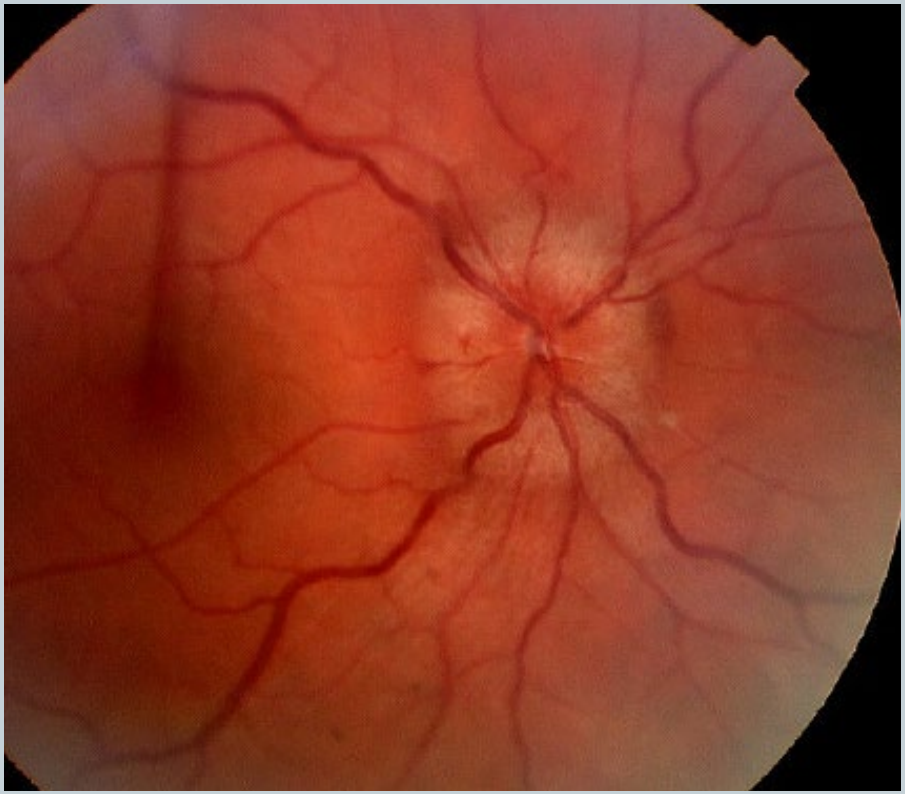


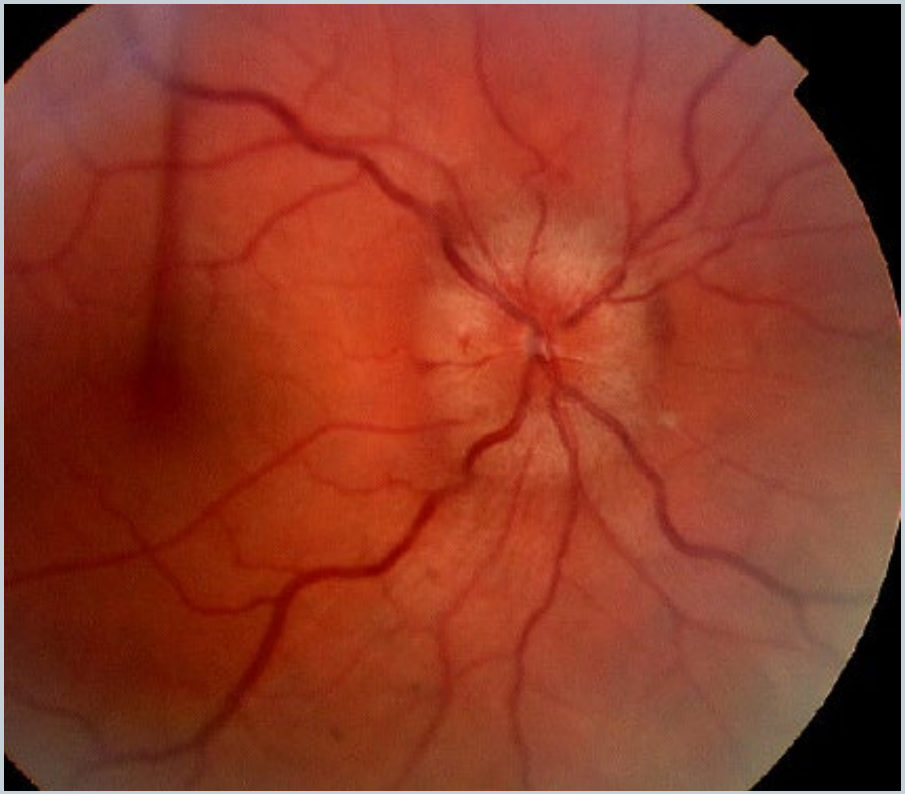
- High ++ risk of stroke (15% 1st 3 months)
- Maximal in the first 2 weeks (80-90% events)

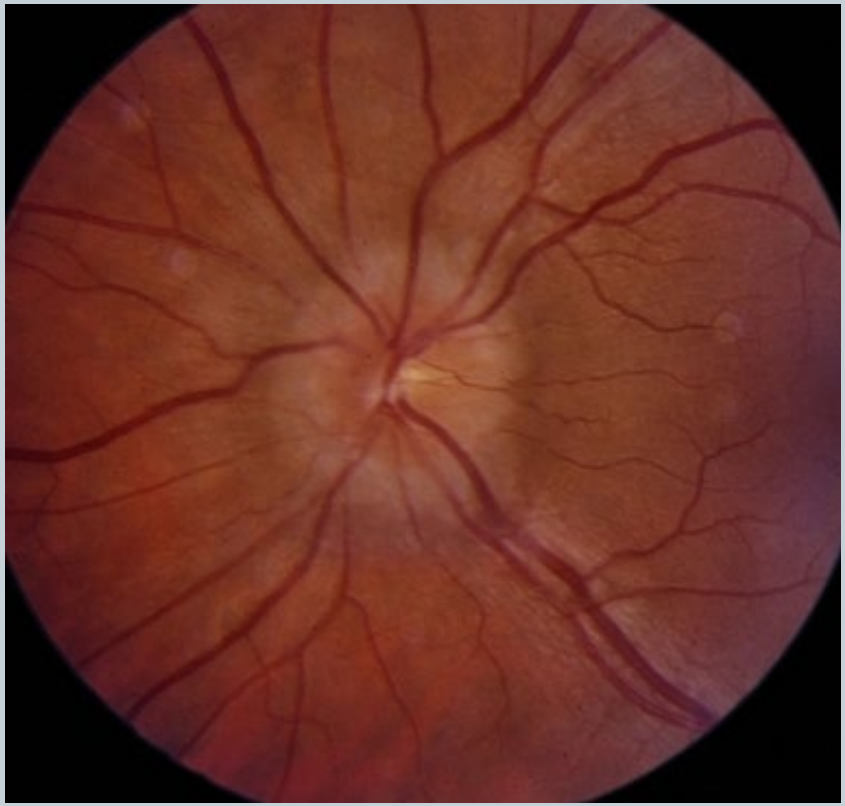
- A. Optic neuropathies
- B. Transient vision loss/ acute arterial ischemia
- **C. Papilloedema**
- D. Double vision
- E. Non-explained visual loss
- F. Pupils – urgent causes of anisocoria

Papilloedema

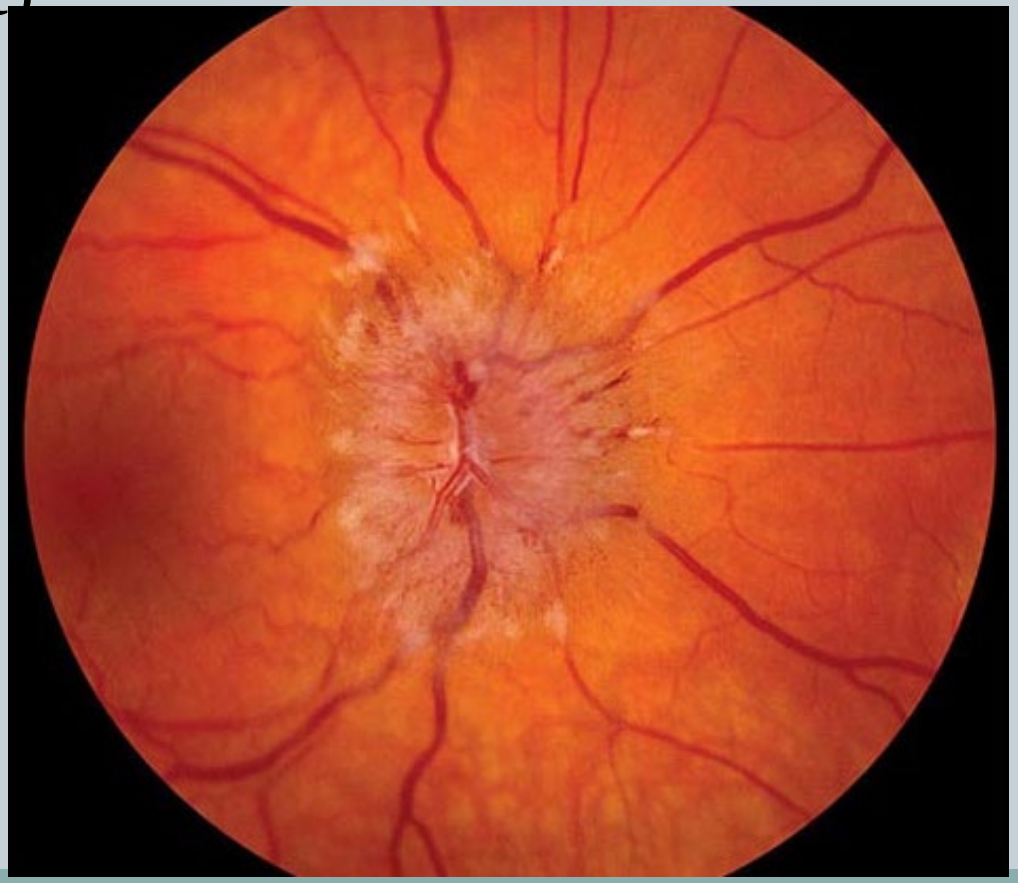
Interruption of normal axoplasmic flow 2ary to raised ICP→ swelling of axons → optic nerve head congestion



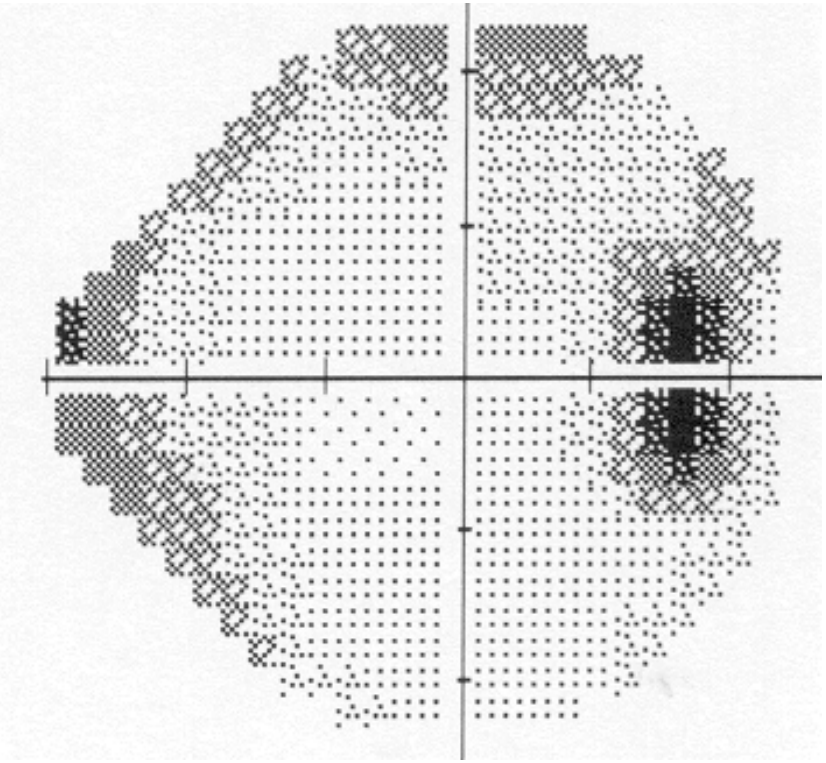




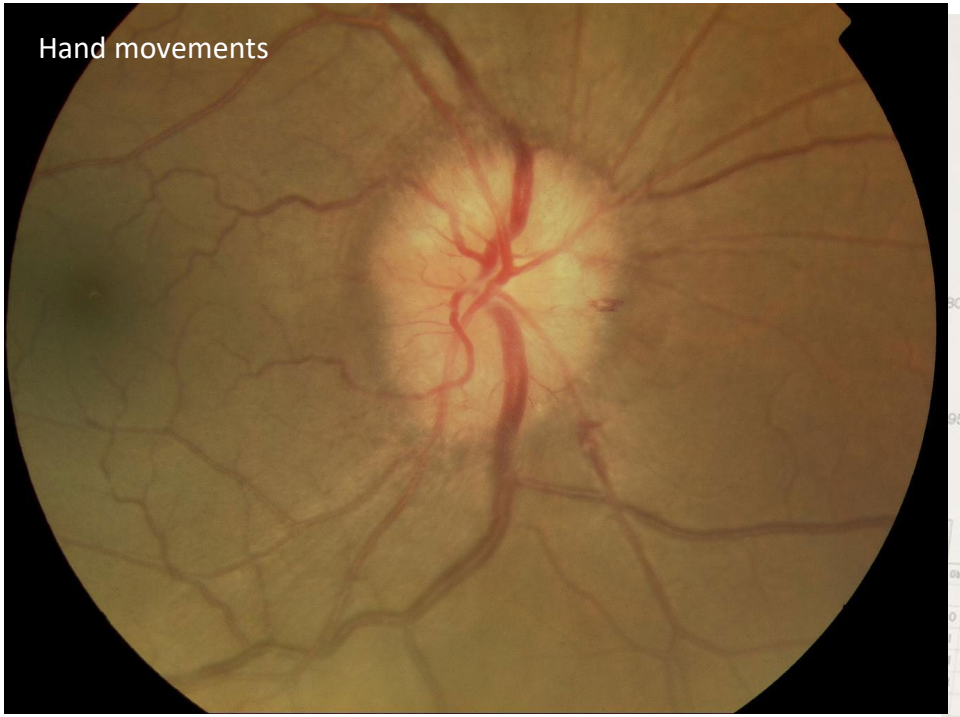
- Preserved vision unless very severe,/florid (and this would be very urgent)

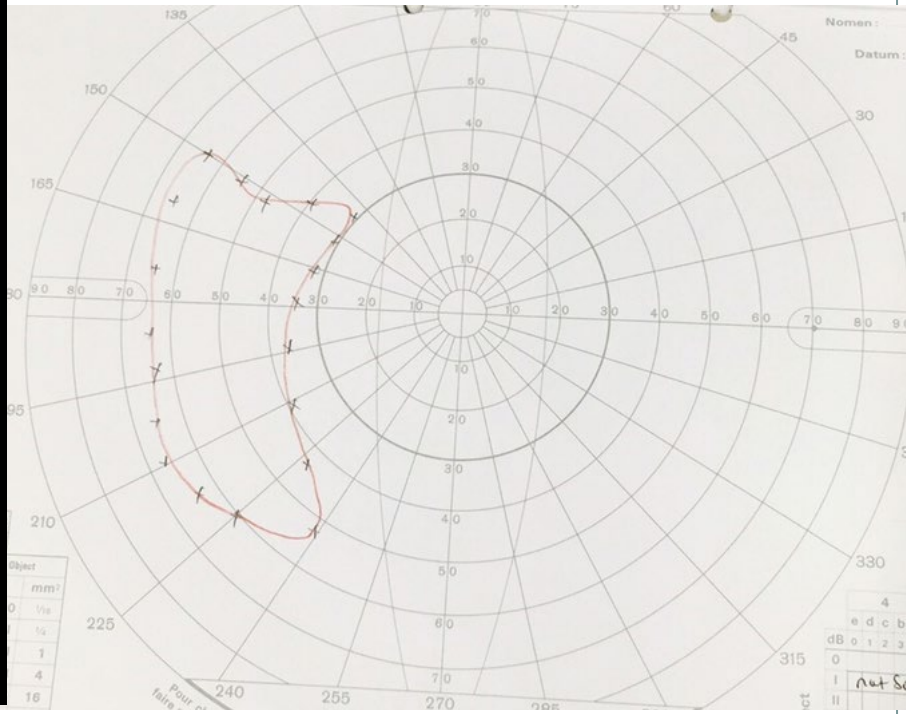


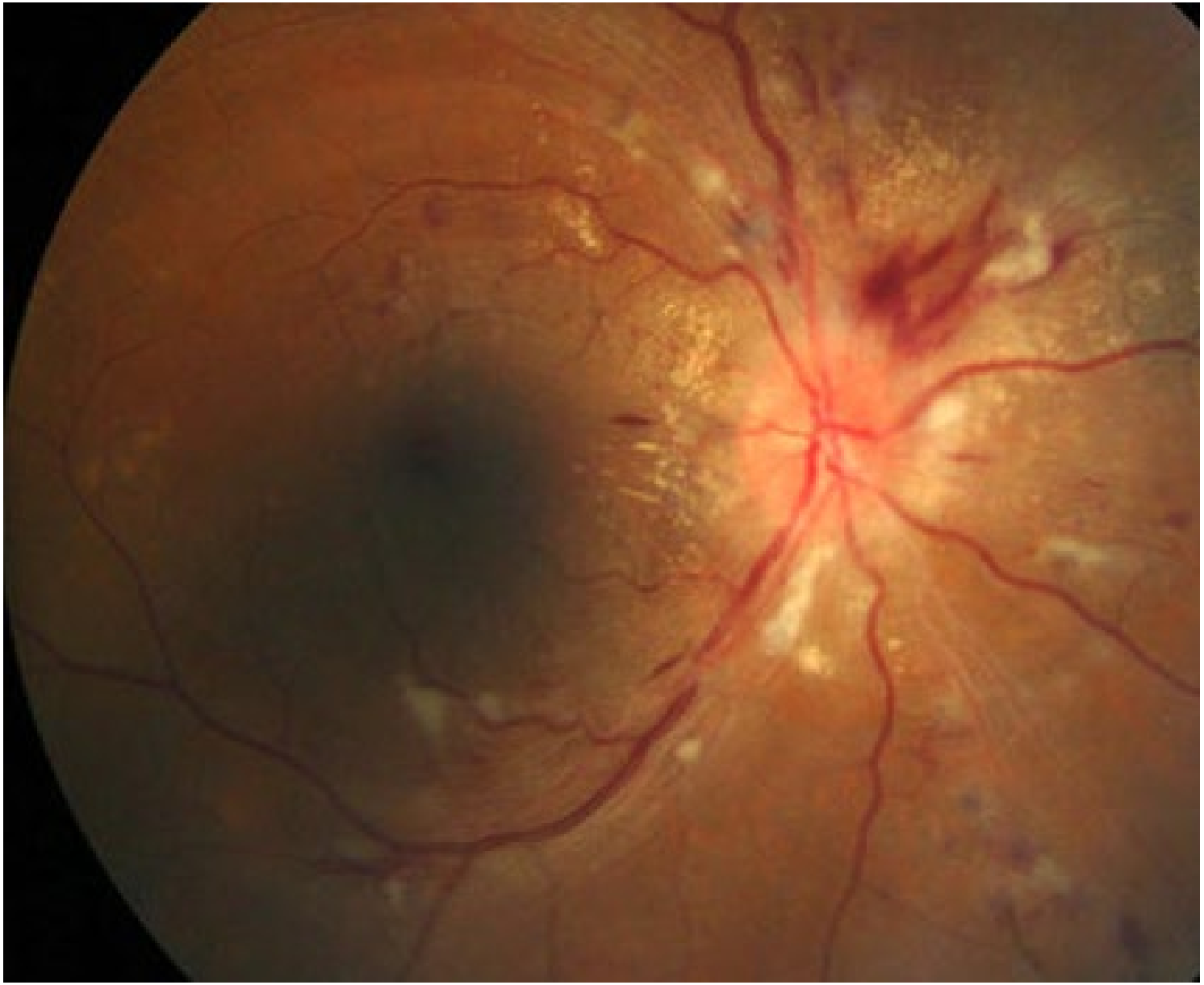
Papilloedema



NOT papilloedema

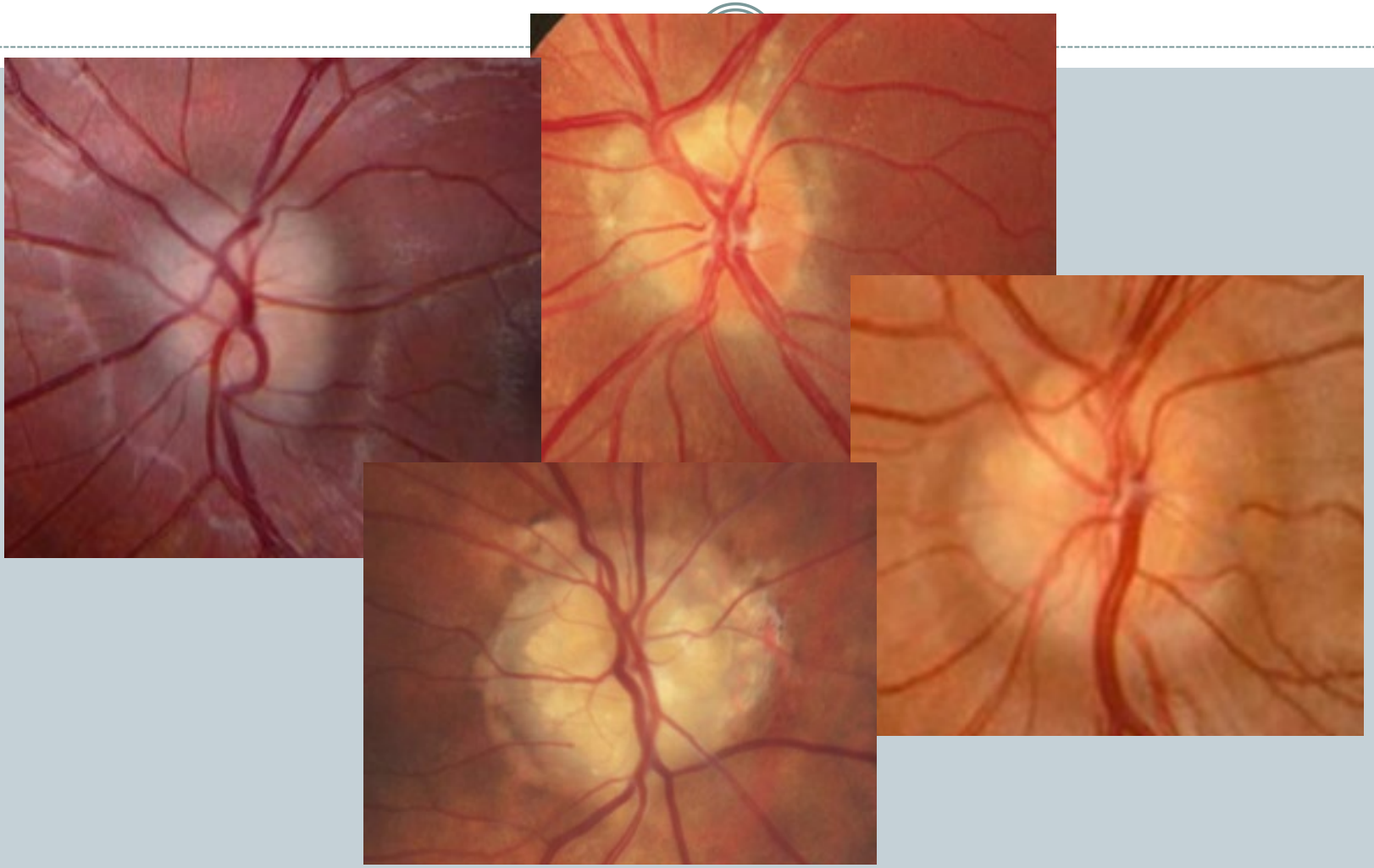






Malignant hypertension >180/120

Pseudopapilloedema



- Not easy to differentiate sometimes
- Requires experience→ most senior doctor to examine!
- Implications of true papilloedema
  - Intracranial mass, venous thrombosis…

Blood pressure → (exclude malignant hypertension)

Disc photos or OCT optic nerve fiver layer: Document appearance at presentation as accurately as possible

Asymptomatic with normal vision
No headaches
Normal acuity and fields

Symptomatic (visual symptoms or headaches) **with normal vision**
Normal best corrected acuity and fields

Symptomatic with significantly *reduced vision or* **RAPD (VA <6/12)**

Occasional headaches and no change in pattern, floaters, visual auras

Significant headaches with high pressure features * +/- visual symptoms (transient visual obscurations, scotomas..)

Is it really papilledema ?

Yes

No

AMU admission for investigations and **neuro-ophthalmology referral**

AMU admission for investigation + URGENT Neuro-ophthalmology opinion <24 H

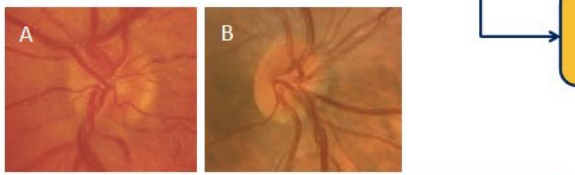
A Small crowded discs?
B Tilted discs?
C Drusenoid appearance?
D Other pseudo-papilledema?

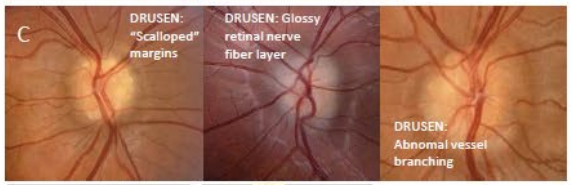
Discharge +/- senior opinion & referral if concerns

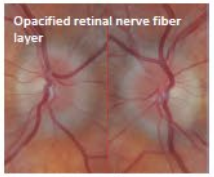
E Unilateral disc swelling only?

See unilateral disc swelling guideline

* High pressure headaches:: increase with valsalva & lying down (night-time, first on waking up) + whooshing tinnitus







- A. Optic neuropathies
- B. Transient vision loss/ acute arterial ischemia
- C. Papilloedema
- **D. Double vision (diplopia)**
- E. Non-explained visual loss
- F. Pupils

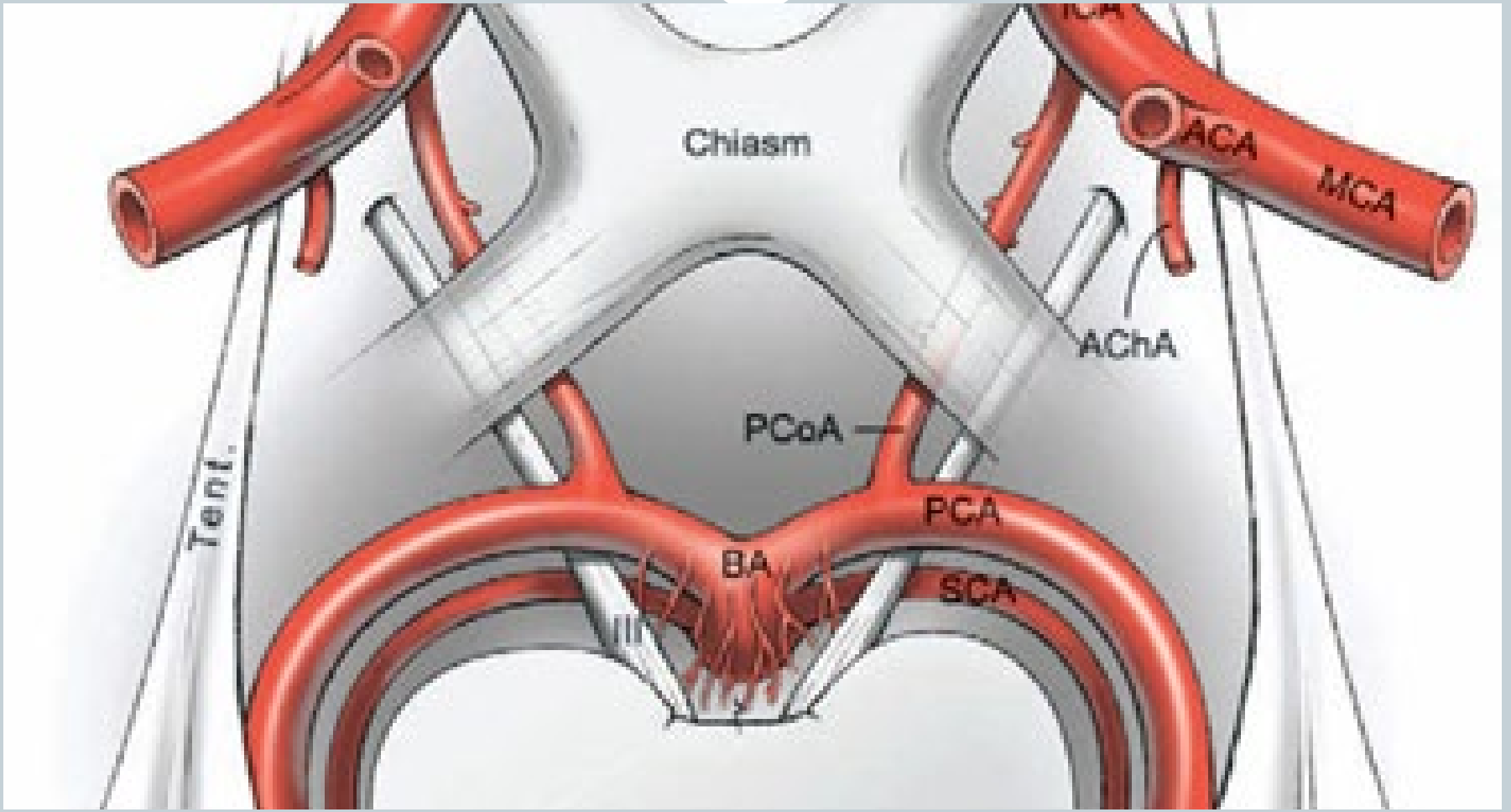
Diplopia

- Orthoptics
- Neurogenic (paretic)/ restrictive/fatigable
- Longstanding/ acute
- Young / > 50-60

Diplopia

- Is it fitting a pattern of **isolated cranial nerve palsy**? (explore cranial nerves!)
- Multiple causes
- Fourth- congenital/ longstanding?
- Sixth
- Third→ s admit for CT ANGIOGRAM- rule out aneurysm

Acute third nerve palsy



CT angiogram

- Isolated cranial nerve palsies (VI or IV)
  - **VI or IV sudden onset over a few hours/ couple of days in a context of HT + elderly, diabetic ++→ often microvascular. Case by case.**
  - **Always have early F/U with orthoptics (liaise with neuro-opthalmology if needed)**

Attention to any Red flags

- More than one cranial nerve (multiple)
- Young patient
- Orbital signs
- Excessive pain
- Other neurological/new systemic symptoms
- History of cancer
- Presence of papilloedema…

Decision to admit + neuro-ophthalmology referral during admission

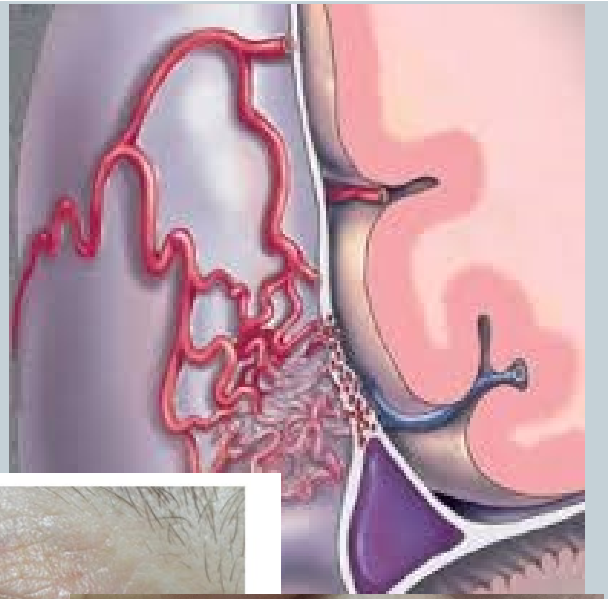
CC fistula

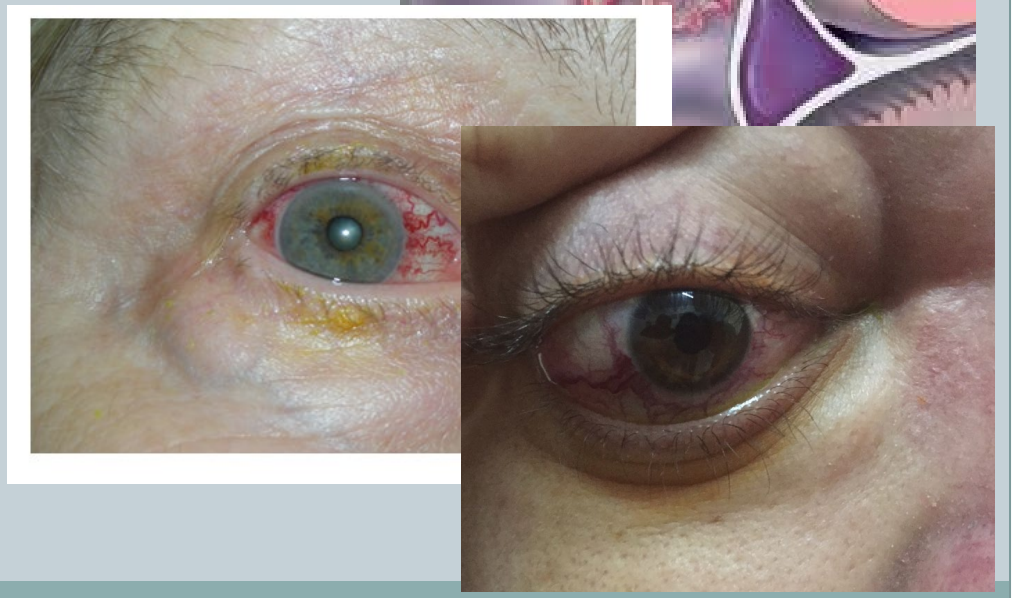
Arterialisation of the cavernous sinus

Often secondary venous thrombosis

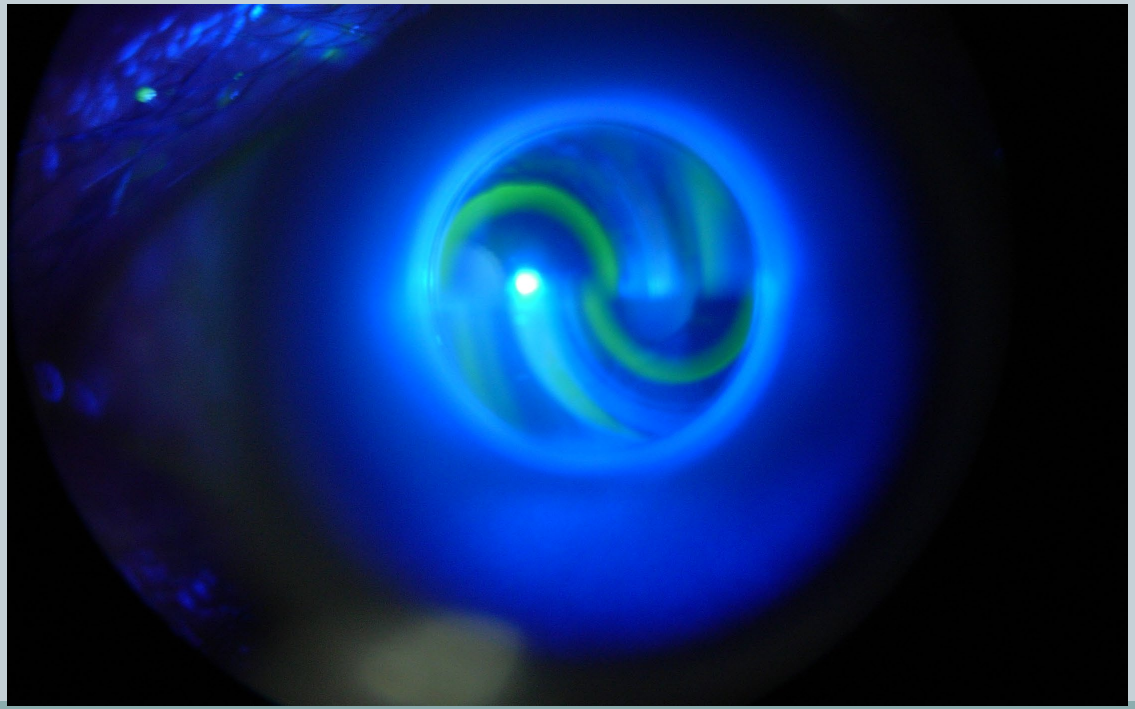
Whooshing/ audible " machinery" noise

Focal venous ischemia/ infarction→ focal symptoms





- Uveoscleral flow- high IOP 10 mmHg
- Oscillating aplanation tonometre
- Urgent neuro-surgical input



- Myasthenia
- Supra-nuclear gaze disorders (occ stroke)
- Orthoptics→ liaise with neuro-ophthalmology

- A. Optic neuropathies
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- C. Papilloedema
- D. Double vision (diplopia)
- **E. Non-explained visual loss**
- F. Pupils

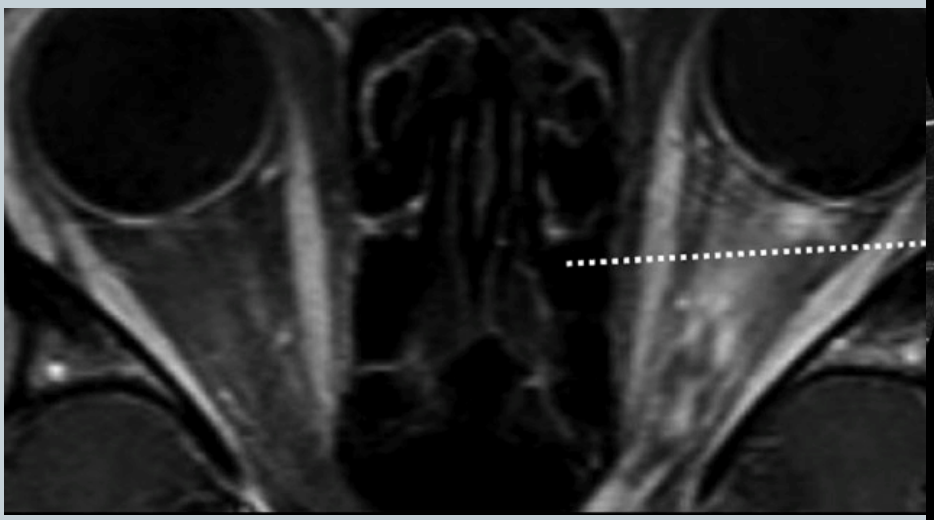
‘Unexplained’ loss of vision

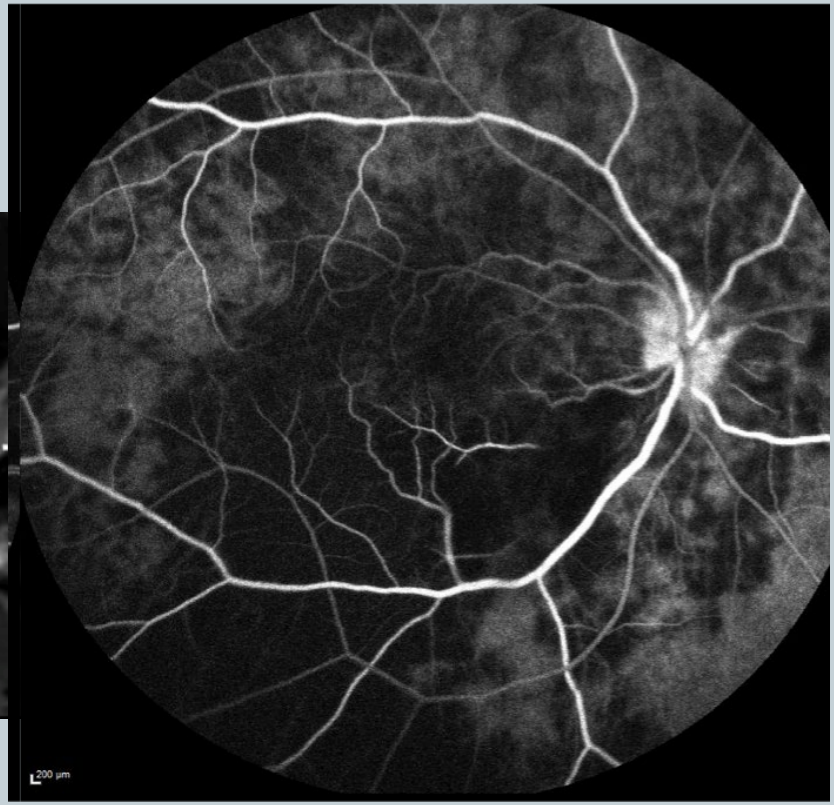
Retrobulbar, cortical… occ functional (exclusion!)

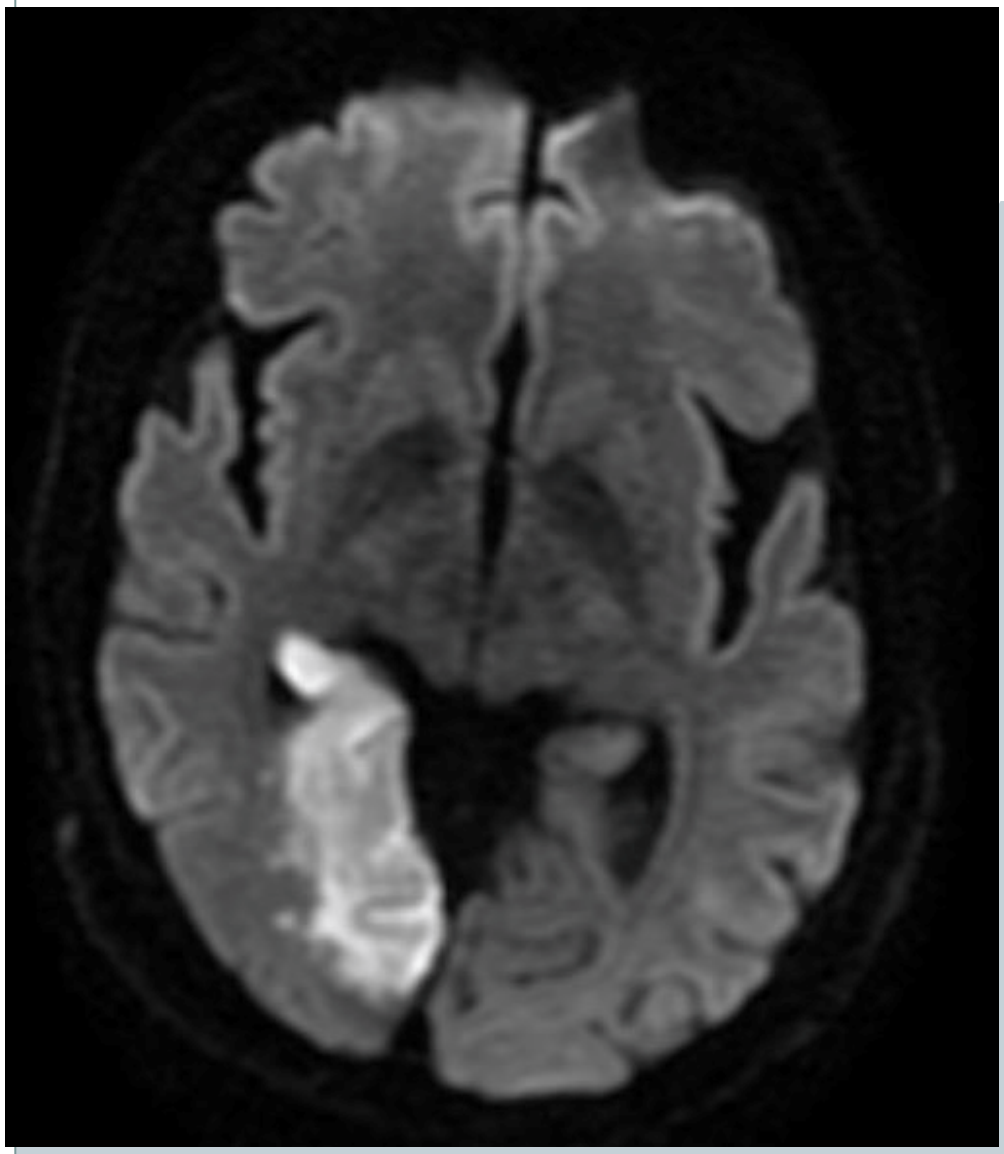
Unilateral : RAPD???

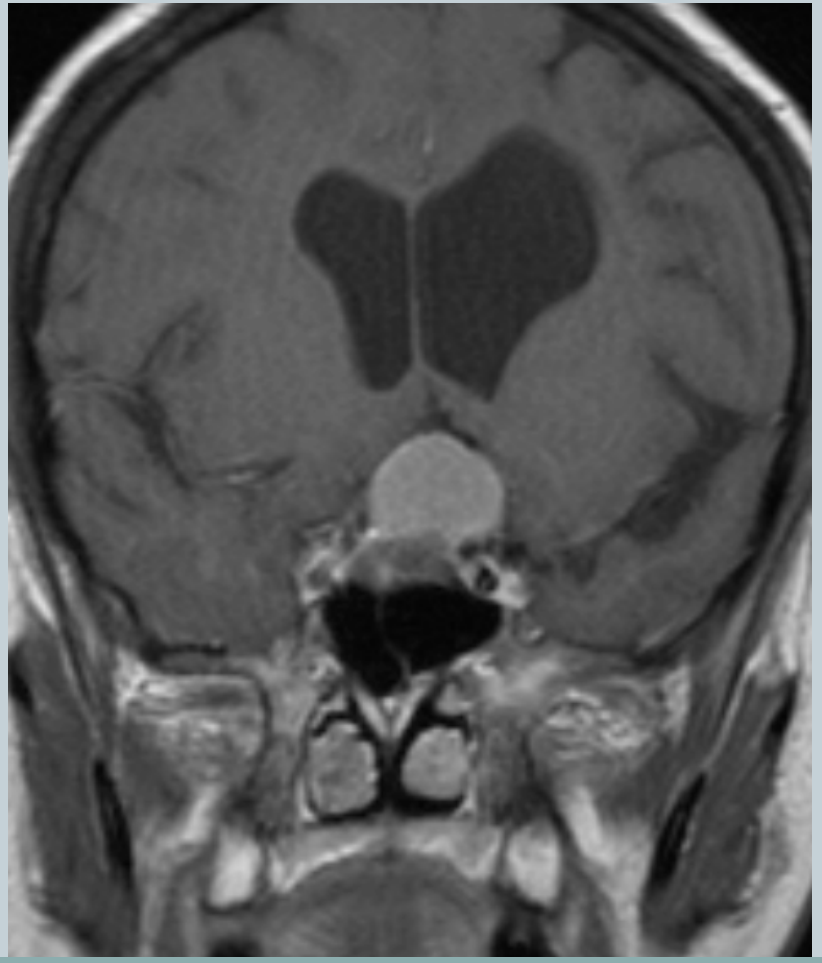
Confrontation visual fields ++

Goldmann visual fields ++




200 µm





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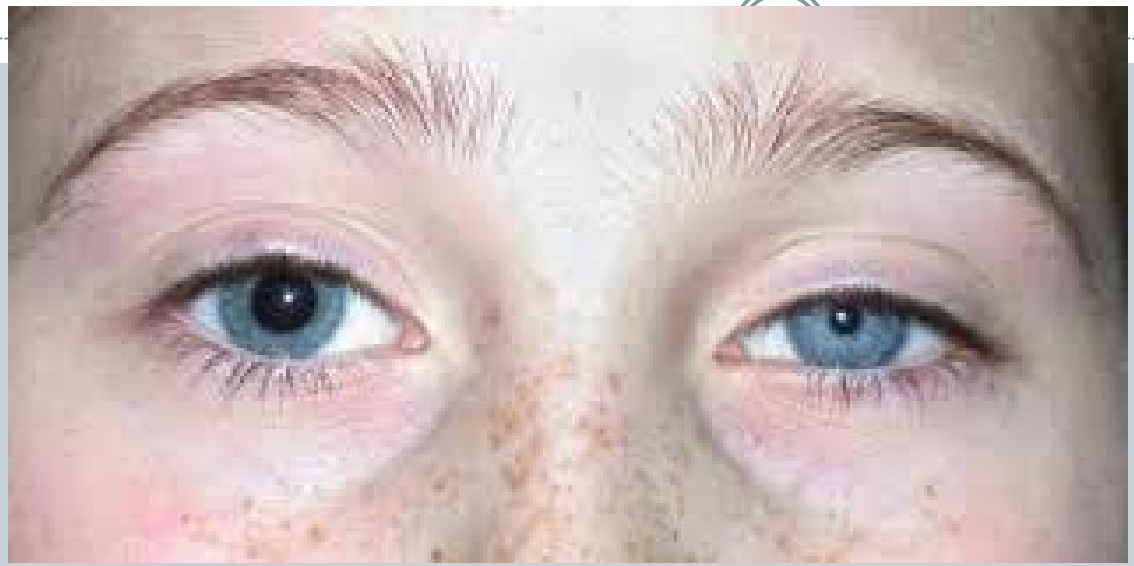
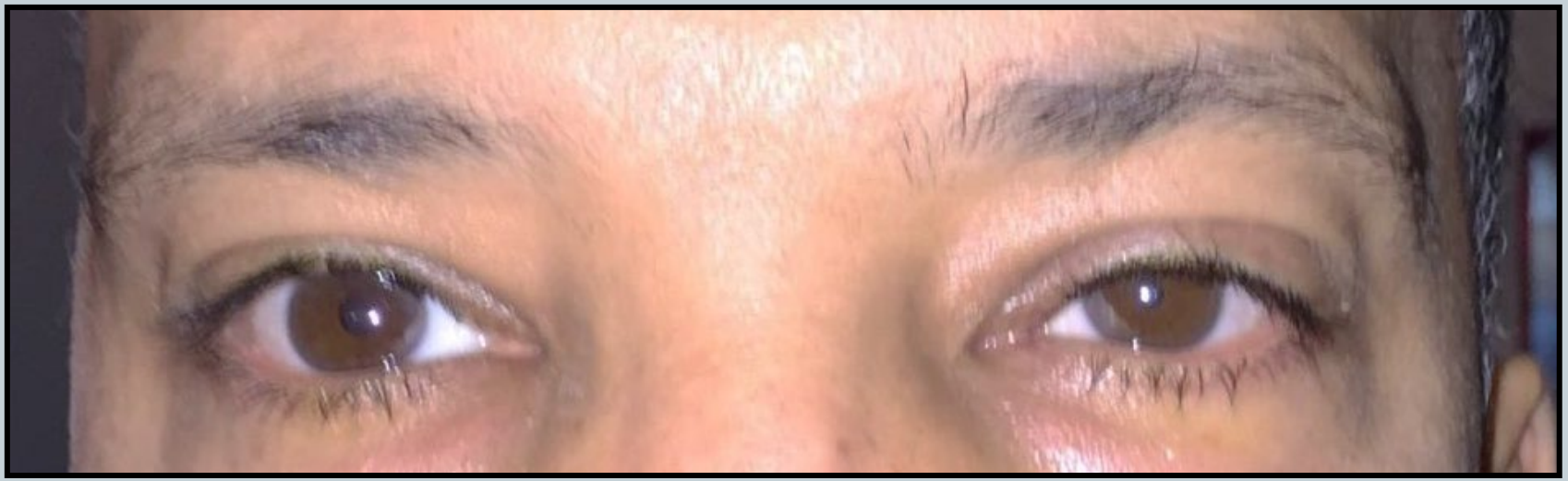
- **F. Pupils**

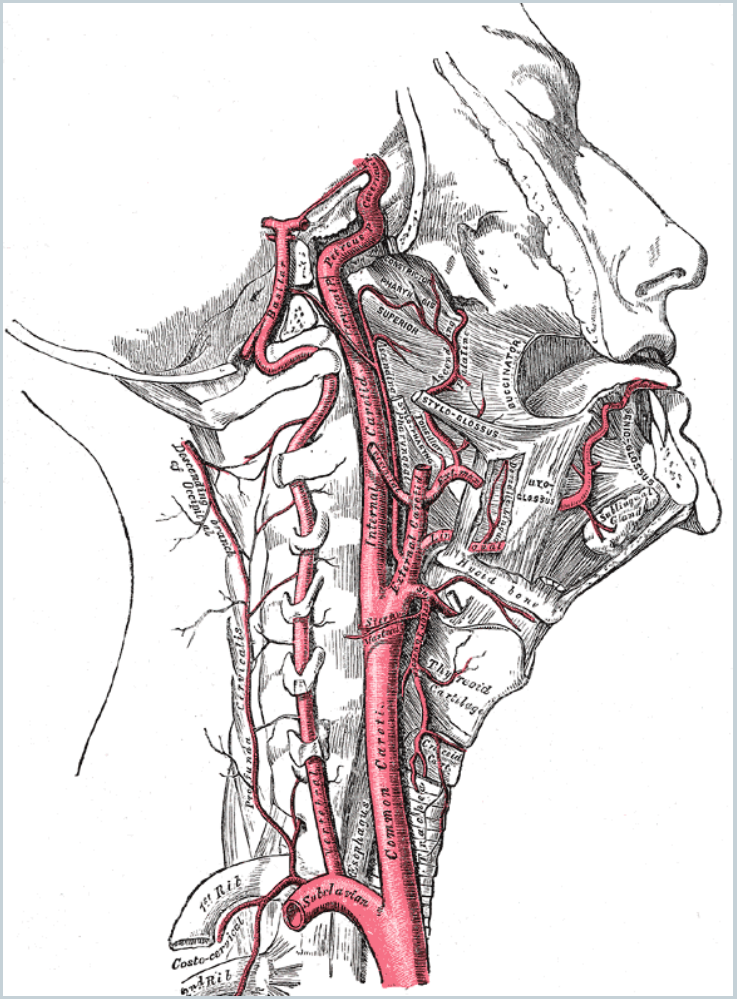
Efferent defect

- Urgent causes of anisocoria
  - **Horner syndrome (painful)**
  - **Dilated pupil from partial third nerve palsy (aneurysmal)**
  - Local iris causes (eg pupil block)





Painful Horner syndrome



Carotid artery dissection until proven otherwise

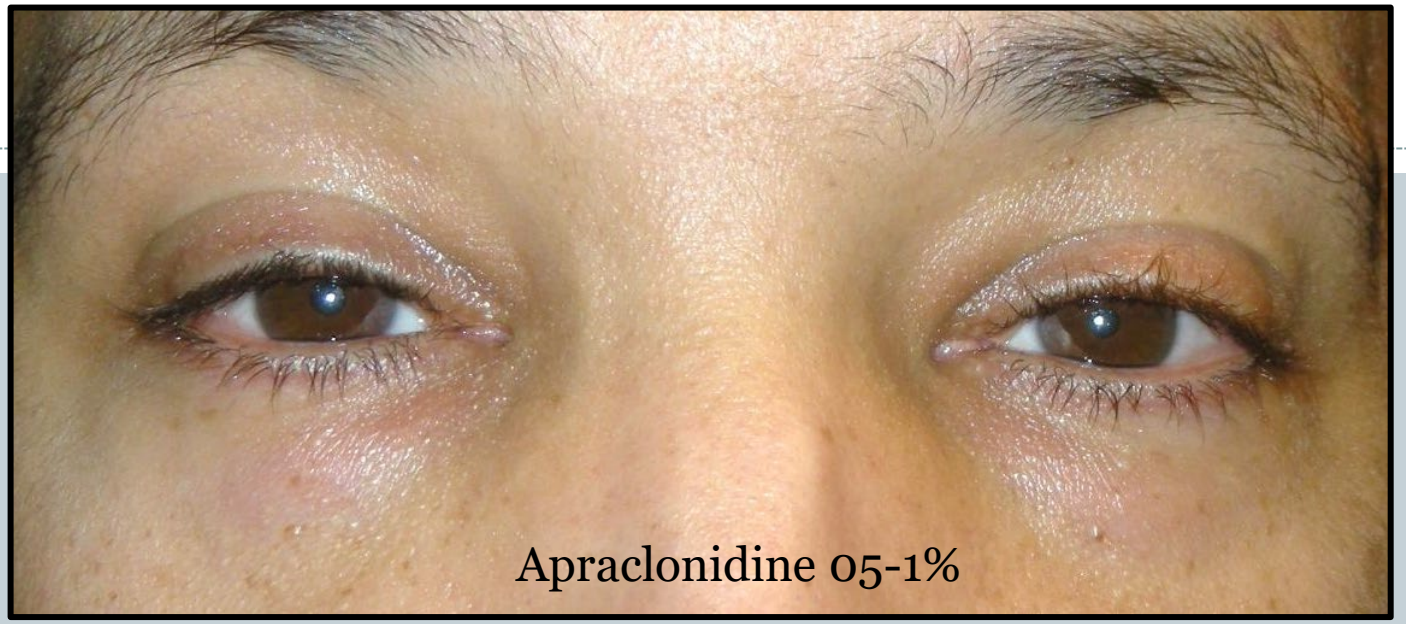
Amaurosis fugax

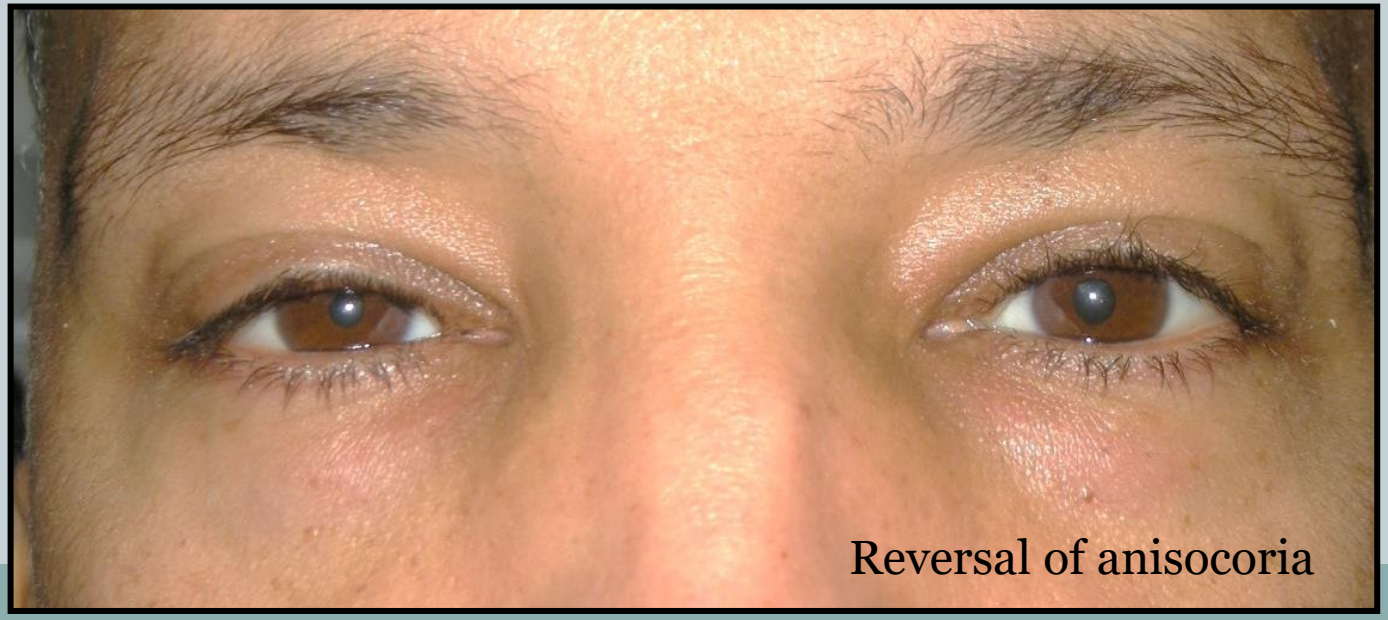
VI nerve palsy

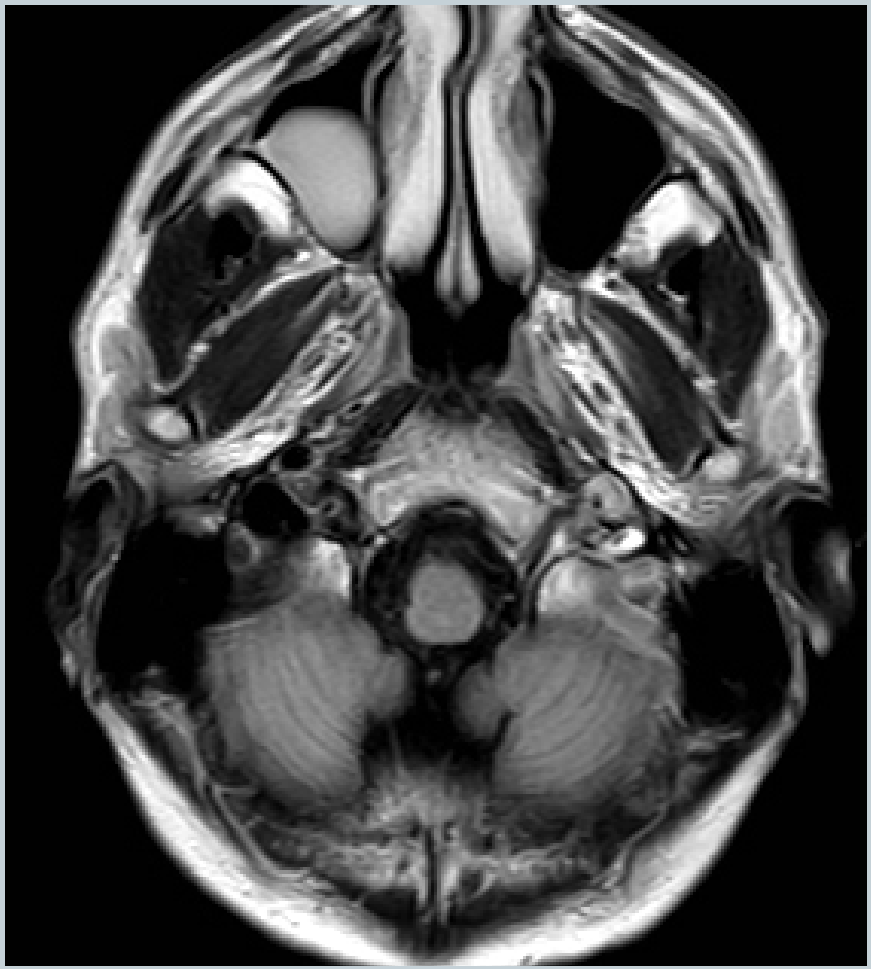
Medical admission for angiogram (CTA/MRA)

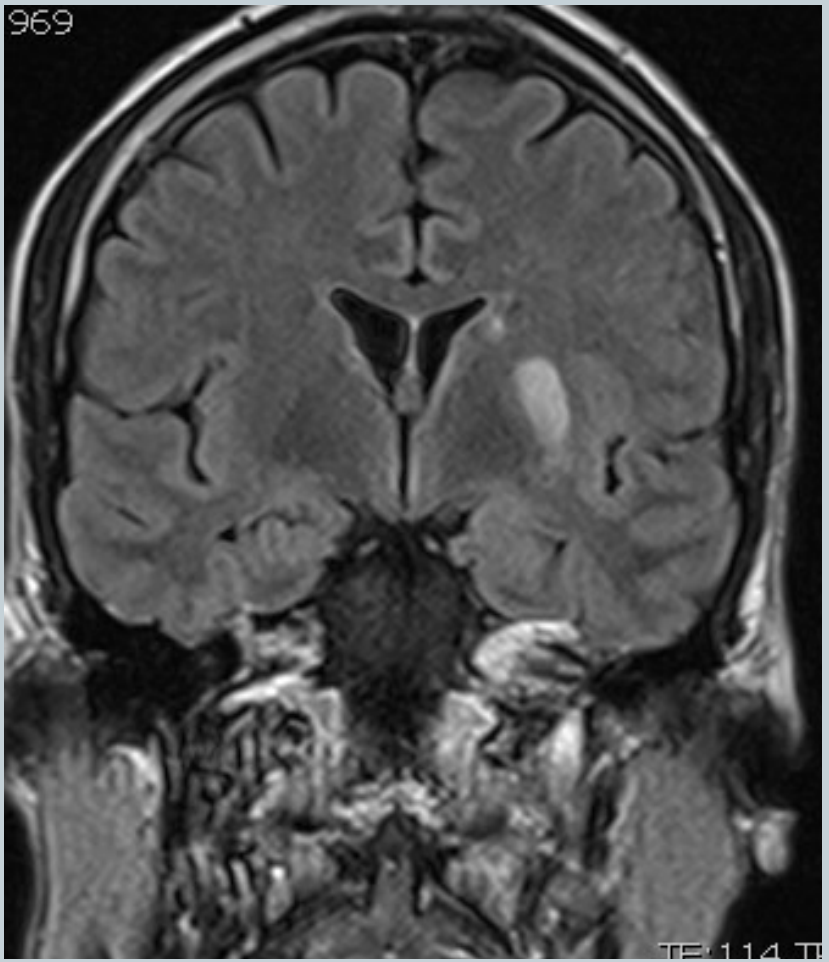
Aspirin/ anticoagulation

High risk of stroke ++


Apraclonidine 05-1%


Reversal of anisocoria

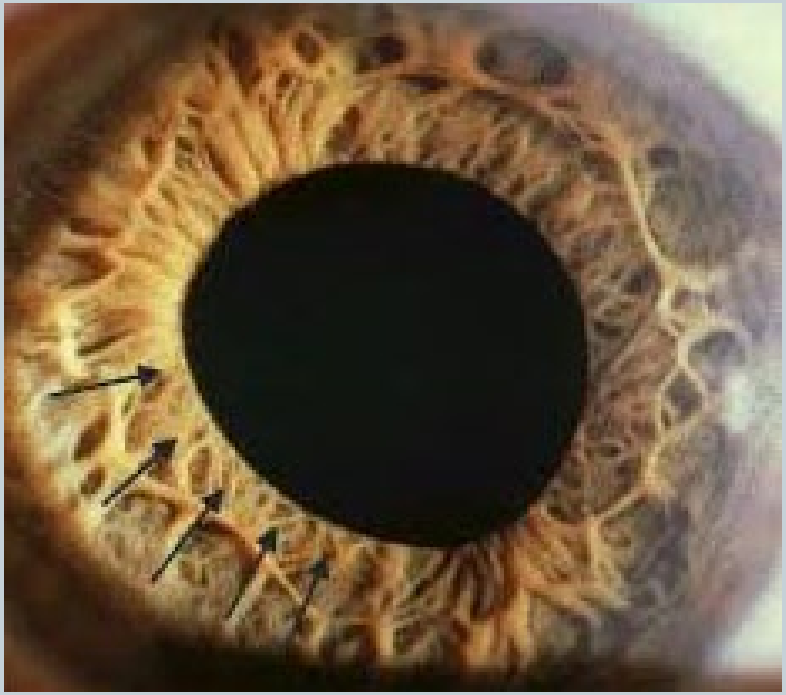


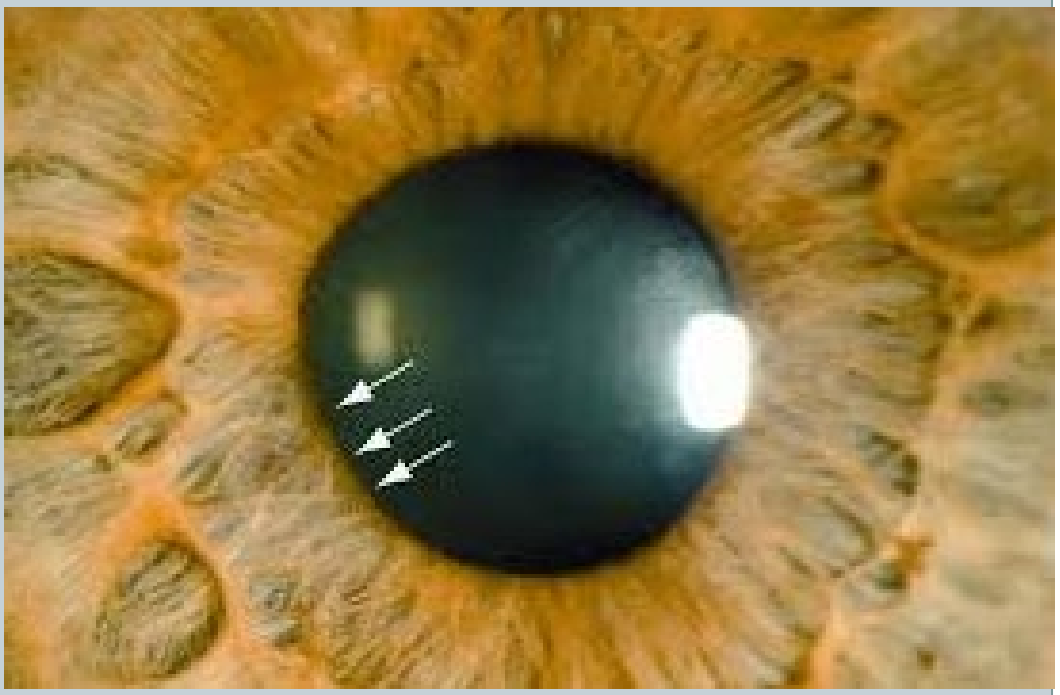


Anticoagulant

Other causes of anisocoria

- Irregular pupils with light near dissociation
  - Post ganglionic denervation of the PS fibers to iris sphincter→ tonic/ Adies pupil
  - Central → midbrain syndrome (gaze palsy, convergence retraction nystagmus, eyelid retraction)
  - Infection (syphilis)
- Pharmacological dilation





- How to refer
  - **Neuro-ophth team BMEC/ QE fellow**
  - **Direct hand over (phone, +/- email)**
  - “Notes to”

Thank you