Be Sure Not To Miss These Diagnoses

March 1, 2020 online section of 10 hour course

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JOHNS HOPKINS
MEDICINE

A WIT-D

approach to avoiding mistakes

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Financial Disclosure

- I have no financial disclosures

A Witty (WIT-D) Approach to Avoiding Mistakes

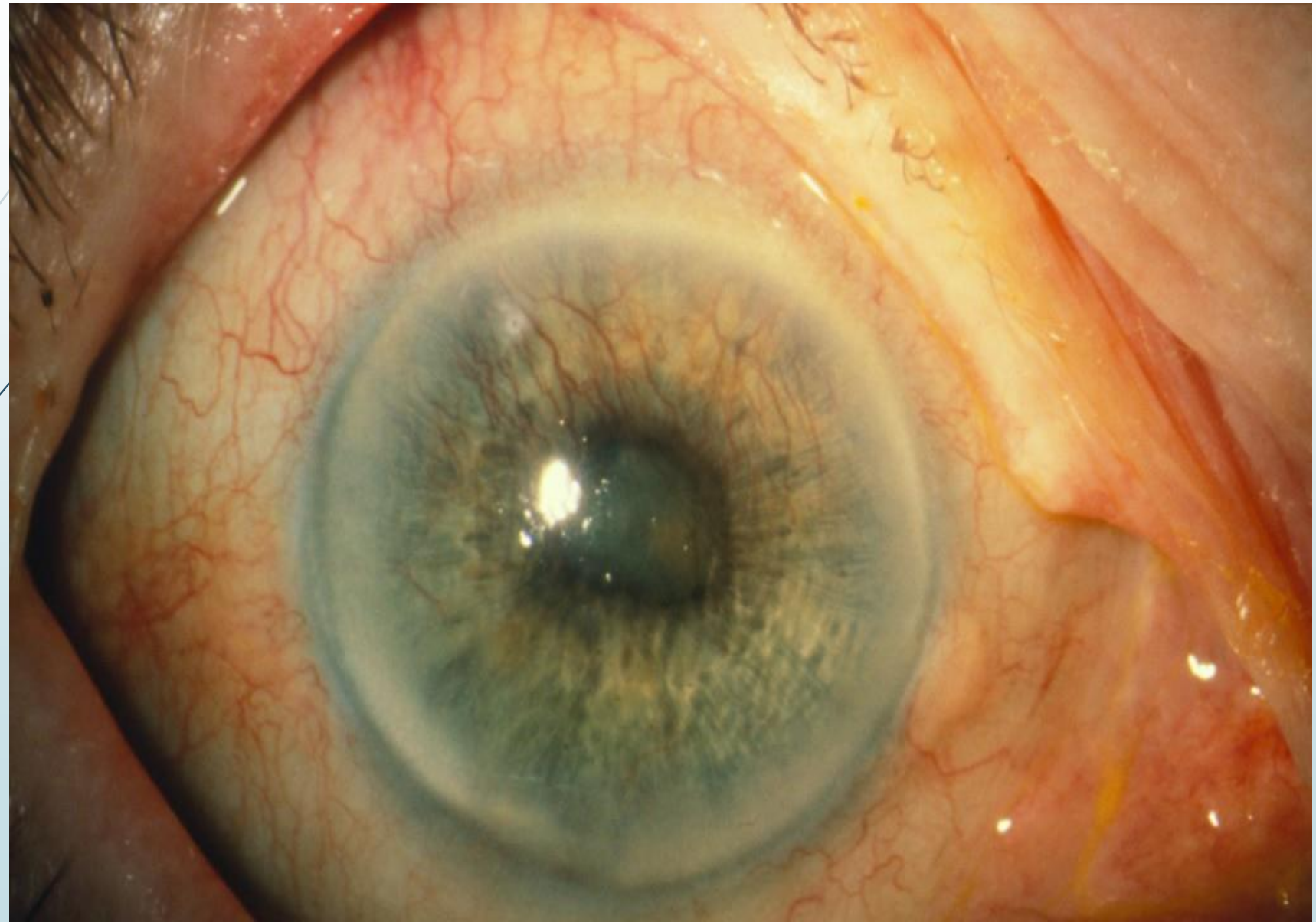
- W= what is worst thing (identify it, rule out)
- I= Information (test as you can for worst thing, in or out)
- T= tell someone (tell patient and others)
- D= document (if not in chart = did not do it?)

Reference; Carolyn Buppert OMIC, gold sheet 4(6) 2002

61 y/o M, moderate cataracts

- Visually significant with reduced ADL
- Diagnosed with DM II , 1 year ago
- few scattered dot hemorrhage
- Macula appears normal

What does this image depict? Actual online quiz



What does this image depict?
Ammonia burn - 7% (1030 votes)

Arcus senilis - 48% (6697 votes)

Diabetic cataract - 14% (1985 votes)

Rubeosis iridis - 31% (4348 votes)

Answer: Rubeosis iridis. Total votes: 14,060

A more prominent example of a serous finding not to be missed

Iris Neovascularization

Wilmer Eye Institute

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Four Neuro-ophthalmic Emergencies You'd Prefer Not To Miss

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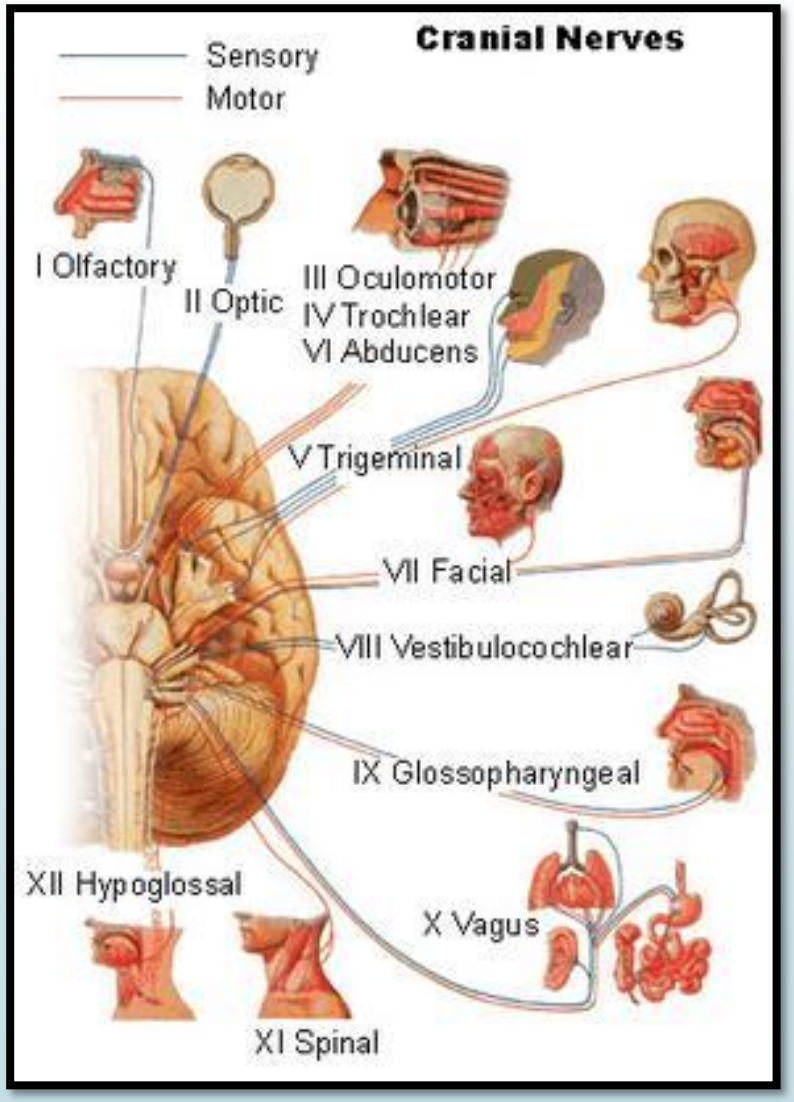
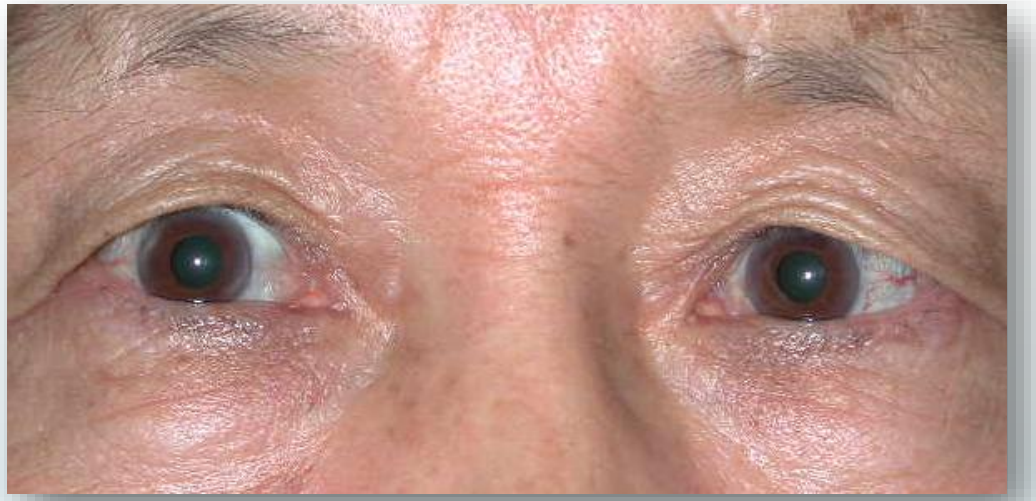
Overview

1. Cranial Nerve Palsy
2. Myasthenia Gravis
3. Papilledema
4. Giant Cell Arteritis

Cranial Nerve Palsy

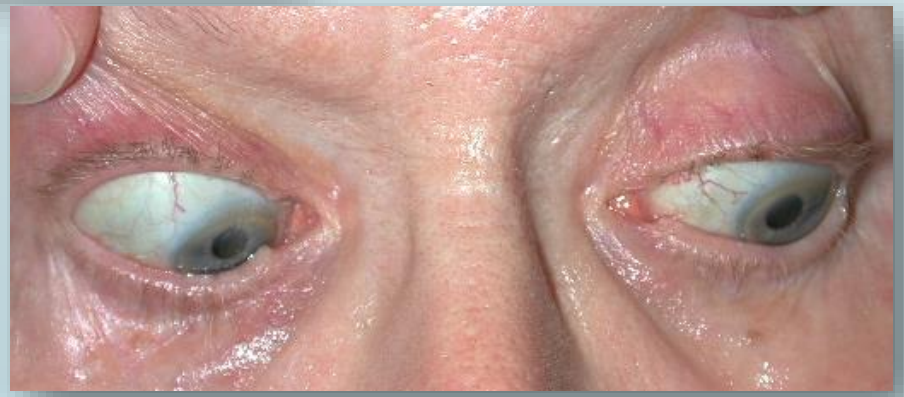


CN VI



Esotropia which worsens when looking in the direction of the involved eye

CN IV



CN III

Bonus Points

3rd Nerve Palsy

- Etiology
  - Aneurysm
  - Trauma
  - Ischemic
    - Diabetes and hypertension
- Decreased levator function
- **Associated findings**
    - **Ocular motility abnormality**
    - **Dilated pupil**

How concerned should you be?

Complete, pupil not involved, and vasculopath

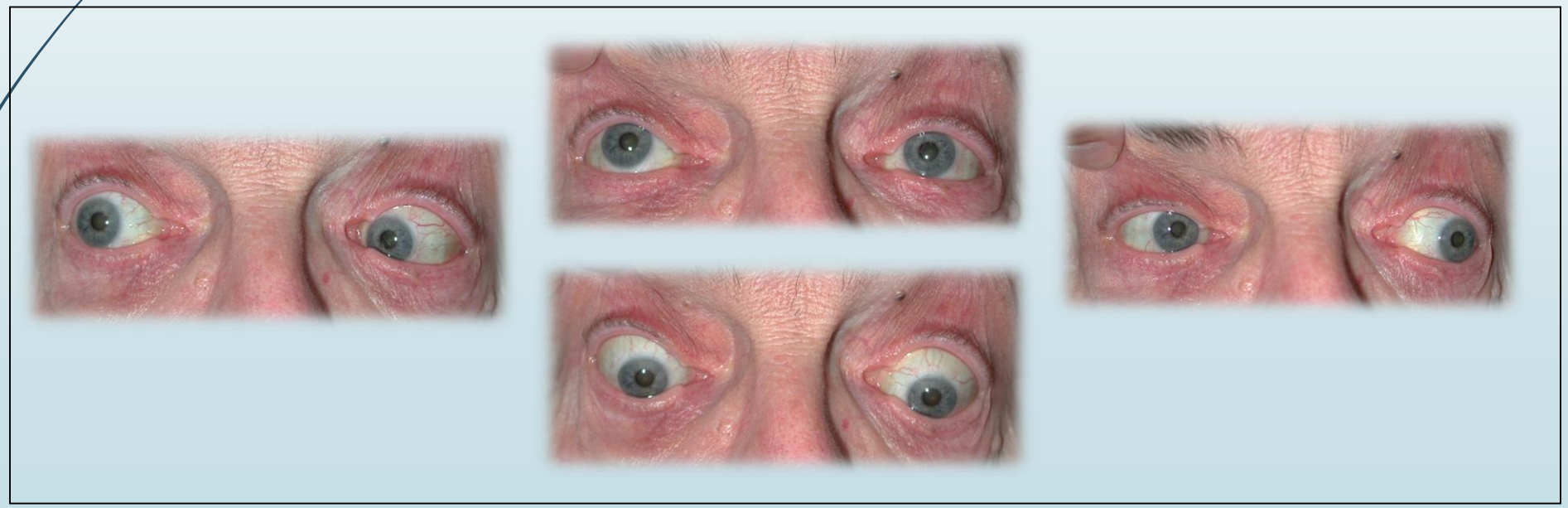
Observation?

Partial, pupil involved, and non-vasculopath

MRI with MRA or CTA

Consider Conventional angiogram if above normal

Abnormal Extraocular Motility



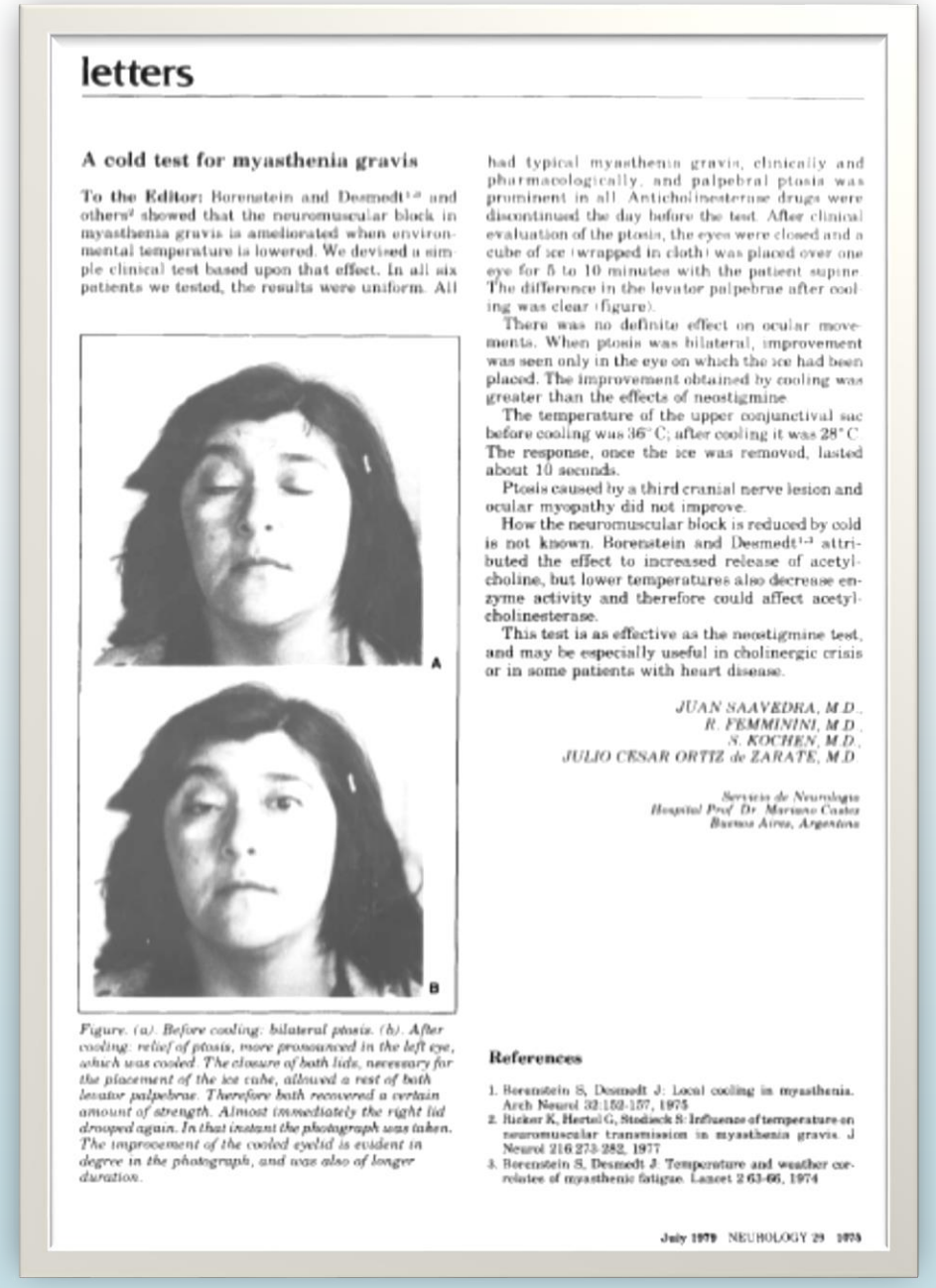
Fatigue

Time: 0

Time: 30 seconds

Time: 1 minute

Saavedra J, Femminini R, Kochen S, deZarate JC. A cold test for myasthenia gravis. Neurology 1979;29:1075.



letters

A cold test for myasthenia gravis

To the Editor: Borenstein and Desmedt[1,2] and others[3] showed that the neuromuscular block in myasthenia gravis is ameliorated when environmental temperature is lowered. We devised a simple clinical test based upon that effect. In all six patients we tested, the results were uniform. All had typical myasthenia gravis, clinically and pharmacologically, and palpebral ptosis was prominent in all. Anticholinesterase drugs were discontinued the day before the test. After clinical evaluation of the ptosis, the eyes were closed and a cube of ice (wrapped in cloth) was placed over one eye for 5 to 10 minutes with the patient supine. The difference in the levator palpebrae after cooling was clear (figure).

There was no definite effect on ocular movements. When ptosis was bilateral, improvement was seen only in the eye on which the ice had been placed. The improvement obtained by cooling was greater than the effects of neostigmine.

The temperature of the upper conjunctival sac before cooling was 36° C; after cooling it was 28° C. The response, once the ice was removed, lasted about 10 seconds.

Ptosis caused by a third cranial nerve lesion and ocular myopathy did not improve.

How the neuromuscular block is reduced by cold is not known. Borenstein and Desmedt[1,2] attributed the effect to increased release of acetylcholine, but lower temperatures also decrease enzyme activity and therefore could affect acetylcholinesterase.

This test is as effective as the neostigmine test, and may be especially useful in cholinergic crisis or in some patients with heart disease.

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Figure. (a): Before cooling: bilateral ptosis. (b): After cooling: relief of ptosis, more pronounced in the left eye, which was cooled. The closure of both lids, necessary for the placement of the ice cube, allowed a rest of both levator palpebrae. Therefore both recovered a certain amount of strength. Almost immediately the right lid drooped again. In that instant the photograph was taken. The improvement of the cooled eyelid is evident in degree in the photograph, and was also of longer duration.

References

1. Borenstein S, Desmedt J: Local cooling in myasthenia. Arch Neurol 32:152-157, 1975
2. Rücker K, Hertel G, Stodieck S: Influence of temperature on neuromuscular transmission in myasthenia gravis. J Neurol 216:273-282, 1977
3. Borenstein S, Desmedt J: Temperature and weather correlates of myasthenic fatigue. Lancet 2:63-66, 1974

Ice test

BEFORE

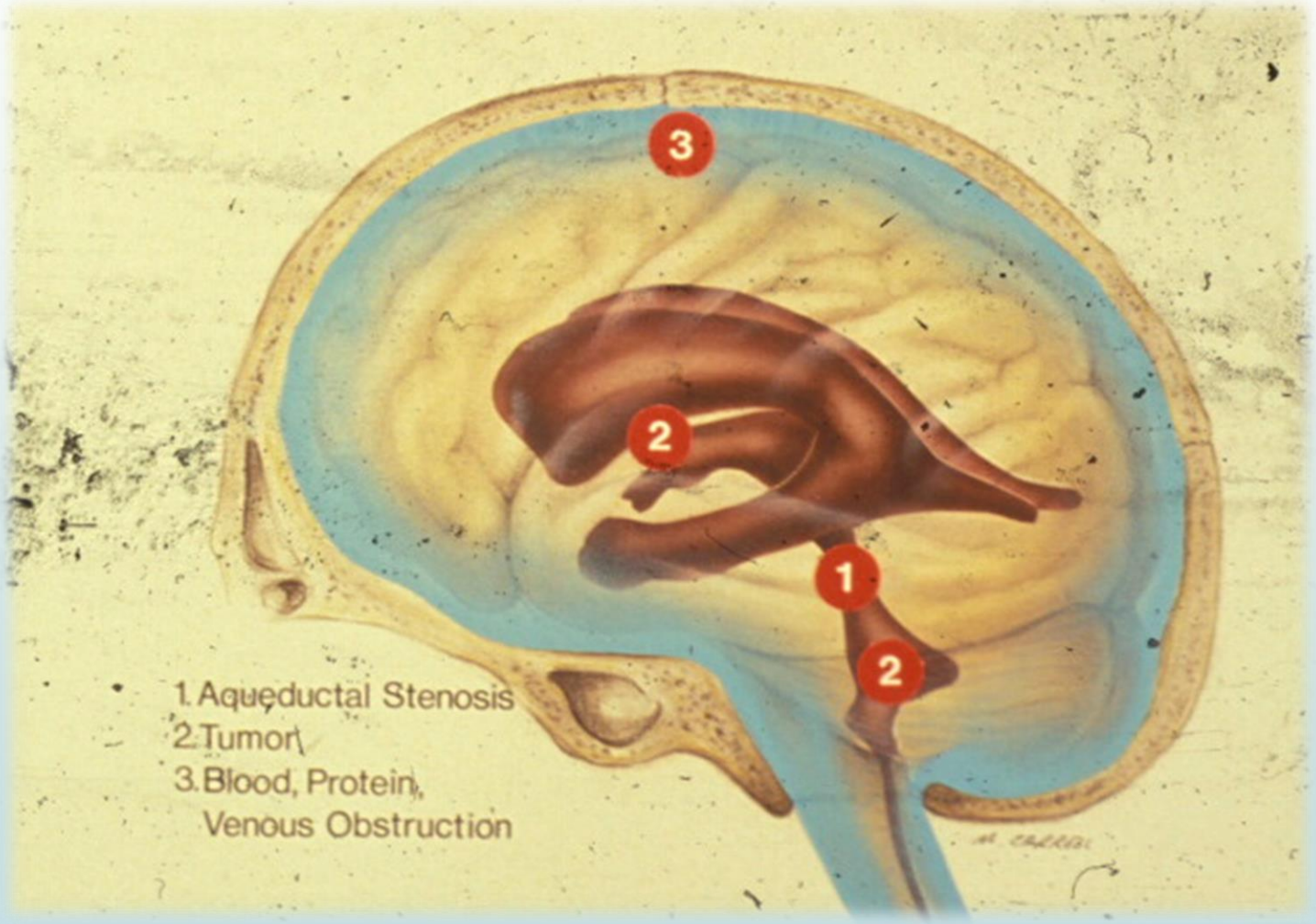
AFTER

Myasthenia Gravis

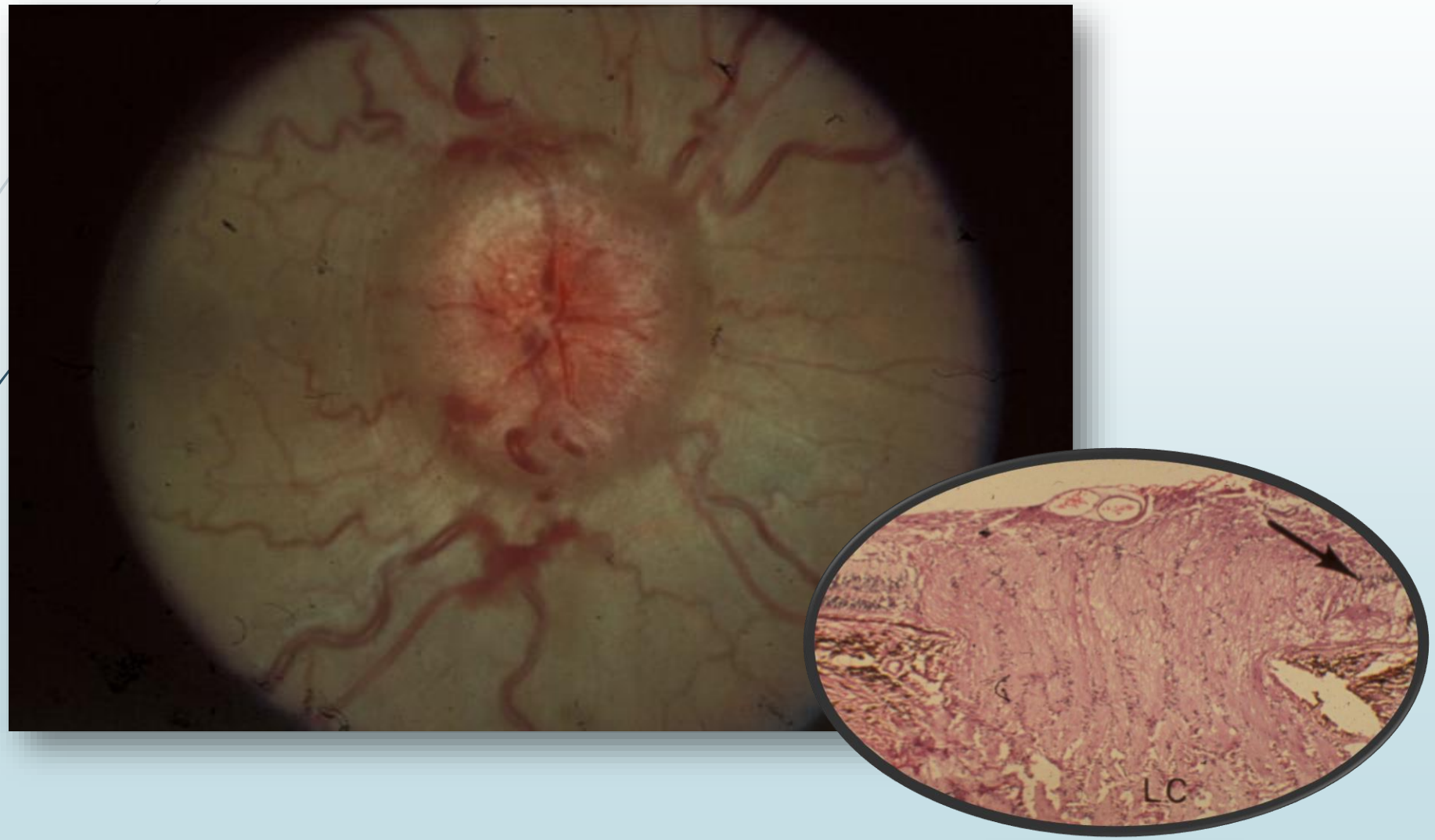
- Autoimmune disease
  - Antibodies block neuromuscular junction
- History
  - Variability: daily and diurnal
  - Usually bilateral
  - Diplopia
  - Weakness
    - *Breathing*
    - *Swallowing*
    - Upper extremity: drying hair, reaching top shelf
    - Lower extremity: stairs

- Examination
  - Levator function decreased
  - Fatigue
  - Ice test
  - Antibodies
  - Tensilon test: inhibits acetylcholine esterase

Papilledema



Papilledema ?definition?



Edematous Nerve

Papilledema

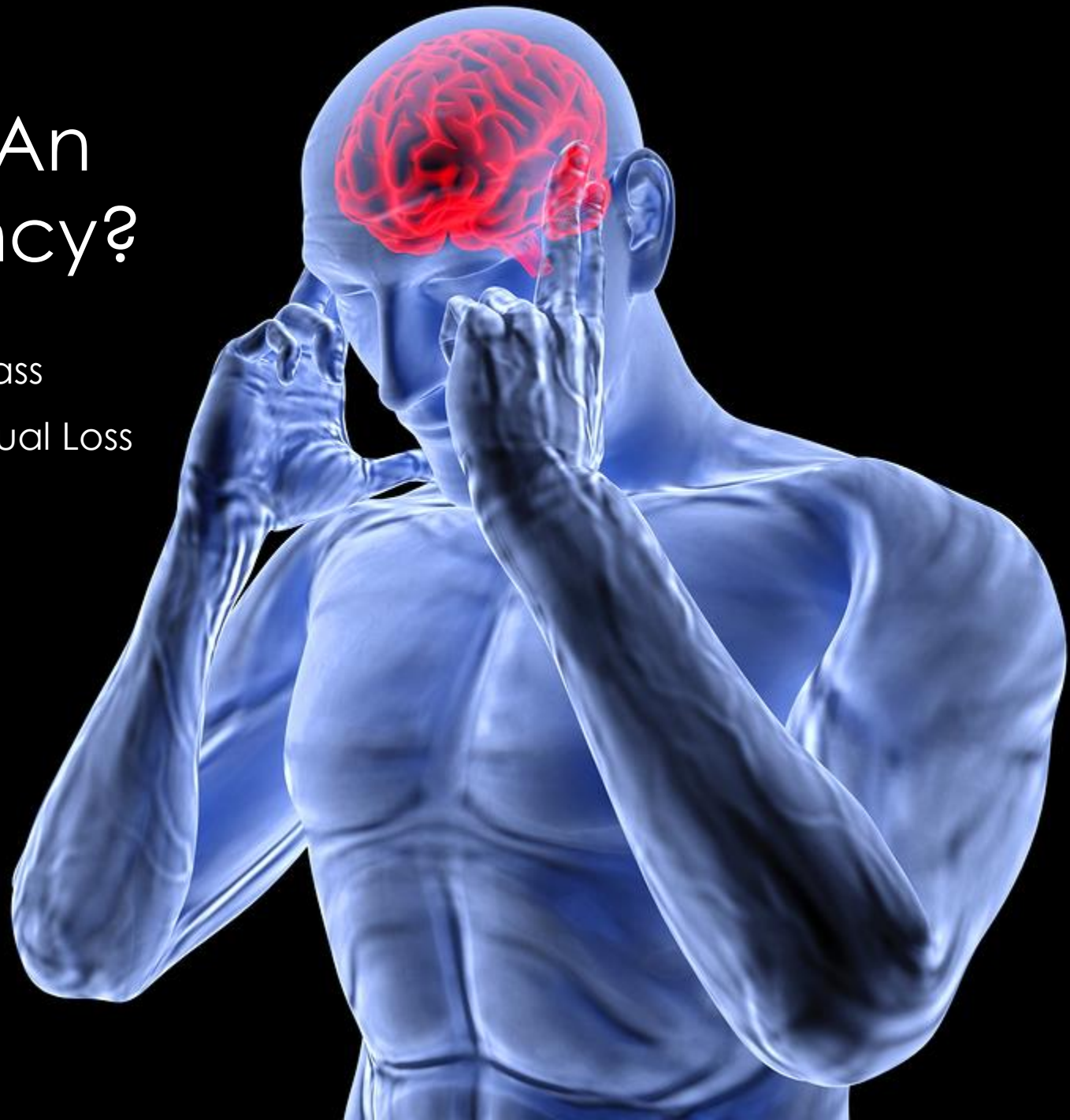
- Visual Acuity
  - Normal
  - Decreased
    - Chronic
    - Macular heme/exudate
- Field
  - Enlarged blindspot
- SVP
  - Absent
- Bilateral (usually)

Inflammation/Infarct

- Visual Acuity
  - Decreased
  - Highly Variable
- Field
  - Abnormal (any pattern)
- SVP
  - May be present
- Unilateral or Bilateral

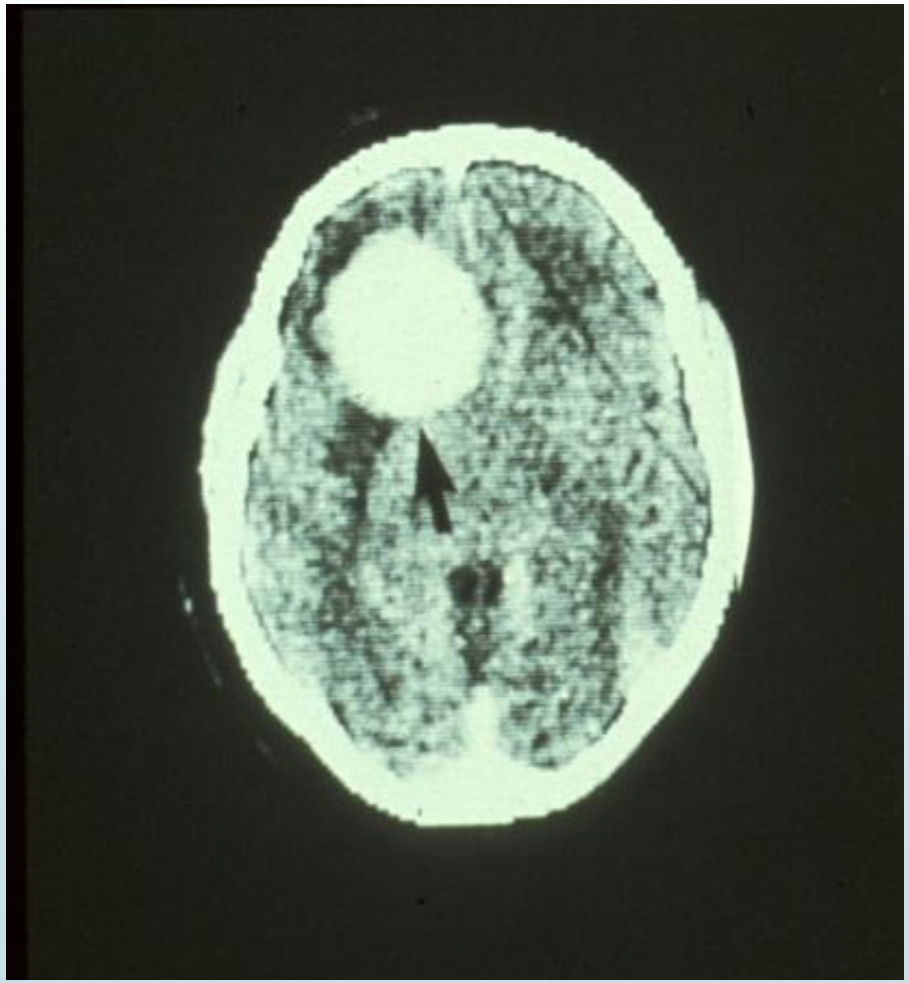
Why Is It An Emergency?

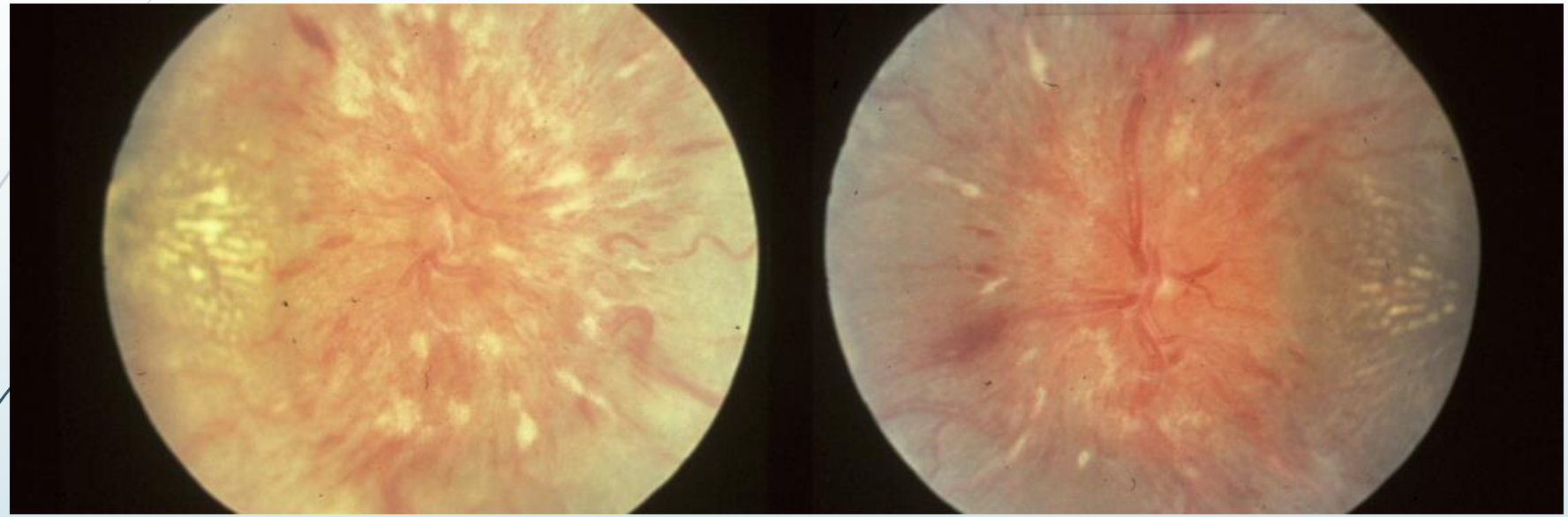
- Intracranial Mass
- Impending Visual Loss



Unknown: 47 yo with headache

24 hours later





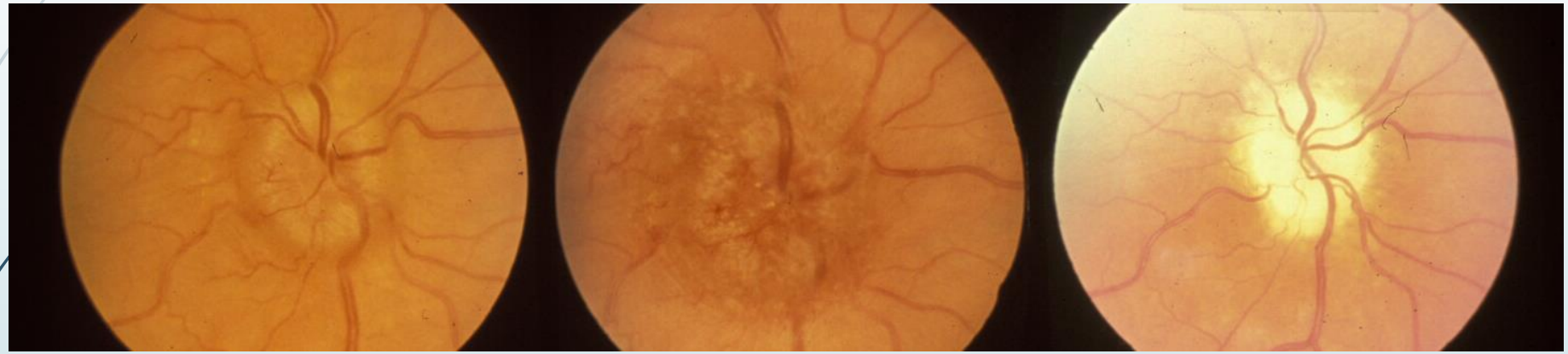
Idiopathic Intracranial Hypertension (IIH)

- Diagnostic Criteria
  - Increased ICP
  - Normal neuroimaging (?small ventricles)
  - Normal CSF composition
- Symptoms
  - Headache, nausea/vomiting
  - Transient visual obscurations (TVO)
  - Diplopia (due to 6th nerve palsy)
  - Pulsatile tinnitus

IIH: Visual Loss

- Peripheral field
  - Progressive constriction
- Enlarged blind spot
  - Distortion of adjacent retina
- Central visual loss
  - Advanced constriction
  - Edema/hemorrhage/exudate

25% will have visual loss >20/200



IIH: Management

- Observation
  - Visual field
  - Color vision
  - Disk photo comparison
- Weight loss
  - Minimal (6%) may help
- Medication
  - Acetazolamide
  - Furosemide

- Surgery
  - Nerve sheath decompression
  - Shunting
- Gastric bypass surgery
- Temporary management
  - Repeated LP
  - Corticosteroids

Case

- An 82 year old female
- Vision loss in the right eye two weeks ago and one day ago noticed decreasing vision in the left eye as well.
- She complains of temporal headache, scalp tenderness, and jaw pain after chewing.
- Normal CBC, an ESR of 99 mm/hr, and a CRP of 2.7 mg/dl (normal <0.5).
- Her visual acuity is 20/200 in the right eye and CF in the left.
- No relative afferent pupillary defect.
- Slit lamp exam is unremarkable.


4


12

Non-arteritic Vs. Arteritic

- Segmental
- More hemorrhages
- Usually crowded

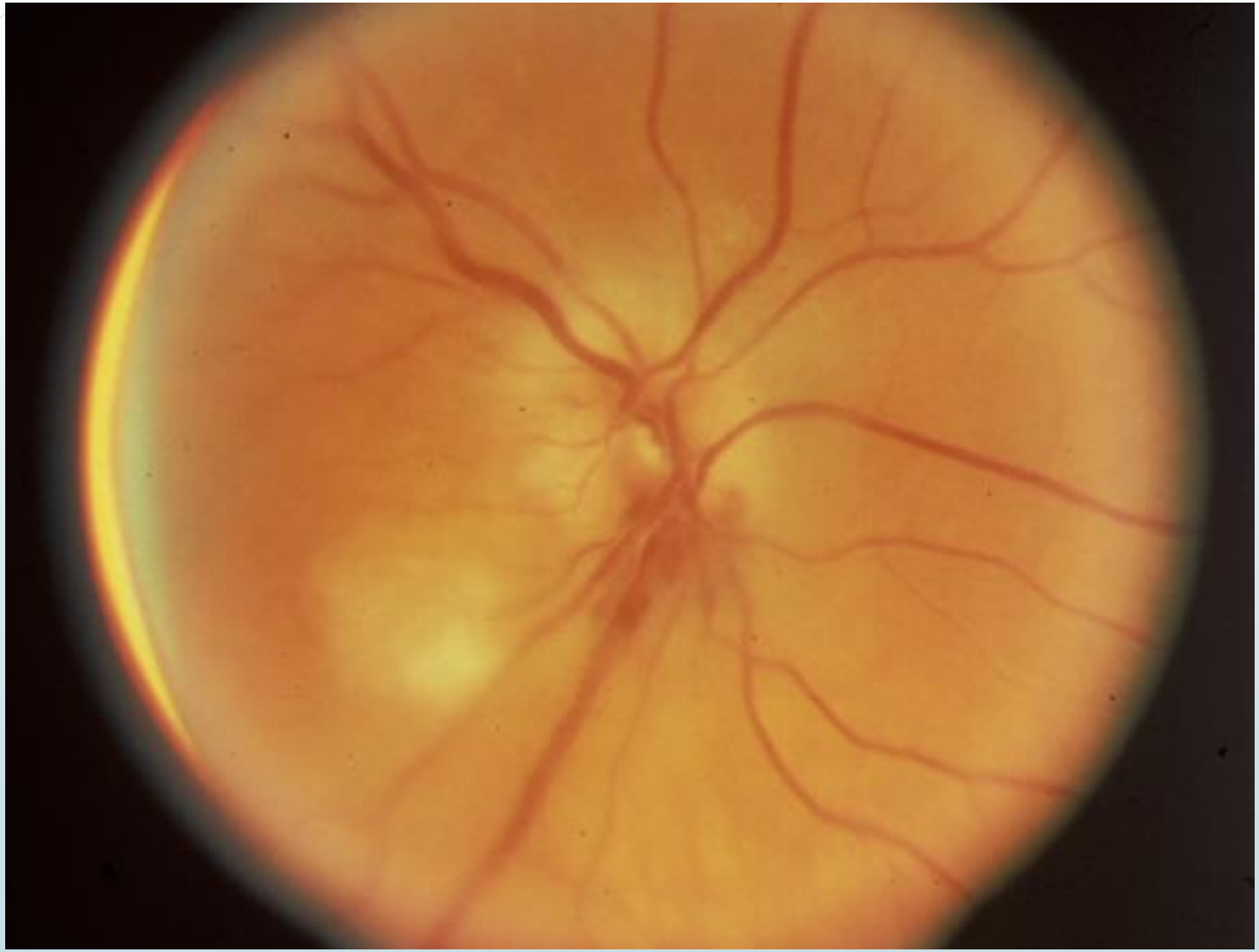
- Diffuse
- More pallor
- Not-necessarily crowded

Giant Cell Arteritis (GCA)

- 5 to 10% of AION cases
- Mean age: 70
- Gender M:F = 1:2

- Systemic symptoms
  - Scalp tenderness/headache
  - Jaw/tongue claudication
  - Malaise/fatigue
  - Joint/muscle pain
  - Weight loss
  - "Occult temporal arteritis"
    - without systemic symptoms
    - 20%
  - TVO may precede event

Unknown

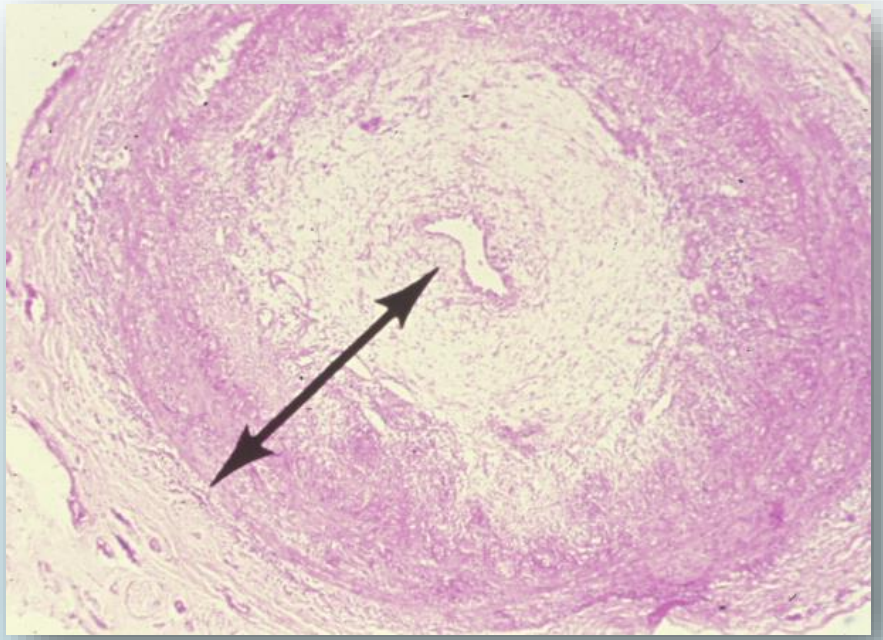


Laboratory

- ESR
  - Mean 70 mm/hr
  - Normal in 15%
  - Rises with anemia (Hct/nl x ESR)
  - Rises with age
    - Women: (age+10)/2 and Men: age/2
- CRP (elevated ESR and CRP 97% specific)

Biopsy

- Within one week of starting steroids
- 3 to 9% false negative
- Unilateral vs. bilateral: controversial



Facial Nerve Injury: A Complication of Superficial Temporal Artery Biopsy

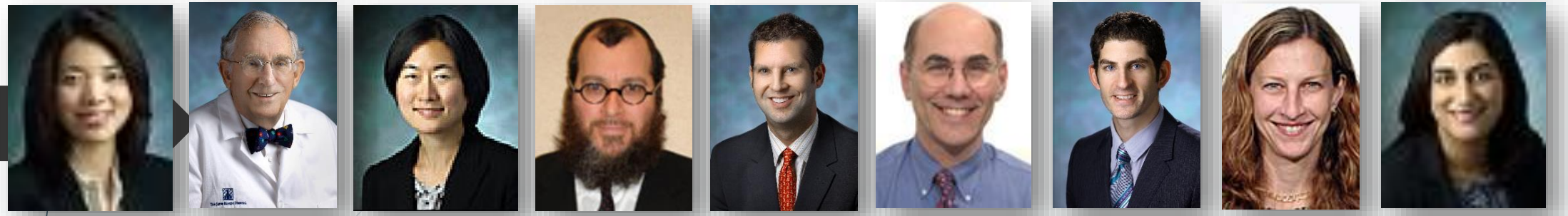
MICHAEL K. YOON, JONATHAN C. HORTON, AND TIMOTHY J. MCCULLEY

Disc edema with visual loss.

- When to be concerned
  - Age >65
  - Any age with characteristic symptoms/findings
- What to do
  - Immediate referral to neuro-ophthalmologist
  - Send to emergency room
  - Document

Closing

1. Cranial Nerve Palsy
   - Could be an aneurysm
2. Myasthenia Gravis
   - Systemic involvement can be fatal
3. Papilledema
   - Could be a tumor, can be blinding
4. Giant Cell Arteritis
   - Blindness is imminent



Divisions of Neuro-ophthalmology and Oculoplastic Surgery