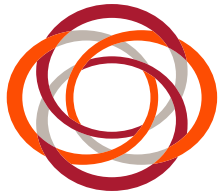
NEURO-OPHTHALMIC PRESENTATIONS IN OLDER ADULTS

Presented by Kelly Malloy, OD



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There are several conditions that are EMERGENT that we always have to consider in the older population!

Let's go through some cases. You can determine what you would do and if you think each is a medical emergency!

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CASE 1

93 year-old woman

- Sudden onset **vertical diplopia**
  - X 1 week
  - At distance and near
  - Relief with closing either eye
- Associated blur
- **Left upper eyelid droopy**
  - x 1 week
- **Eye pain OU** x 1 week
- **Generalized weakness** x 1 week

- Systemic history
  - Hypertension x 30 years
  - Hypercholesterolemia
  - Arthritis
  - Ovarian cancer 30 years ago

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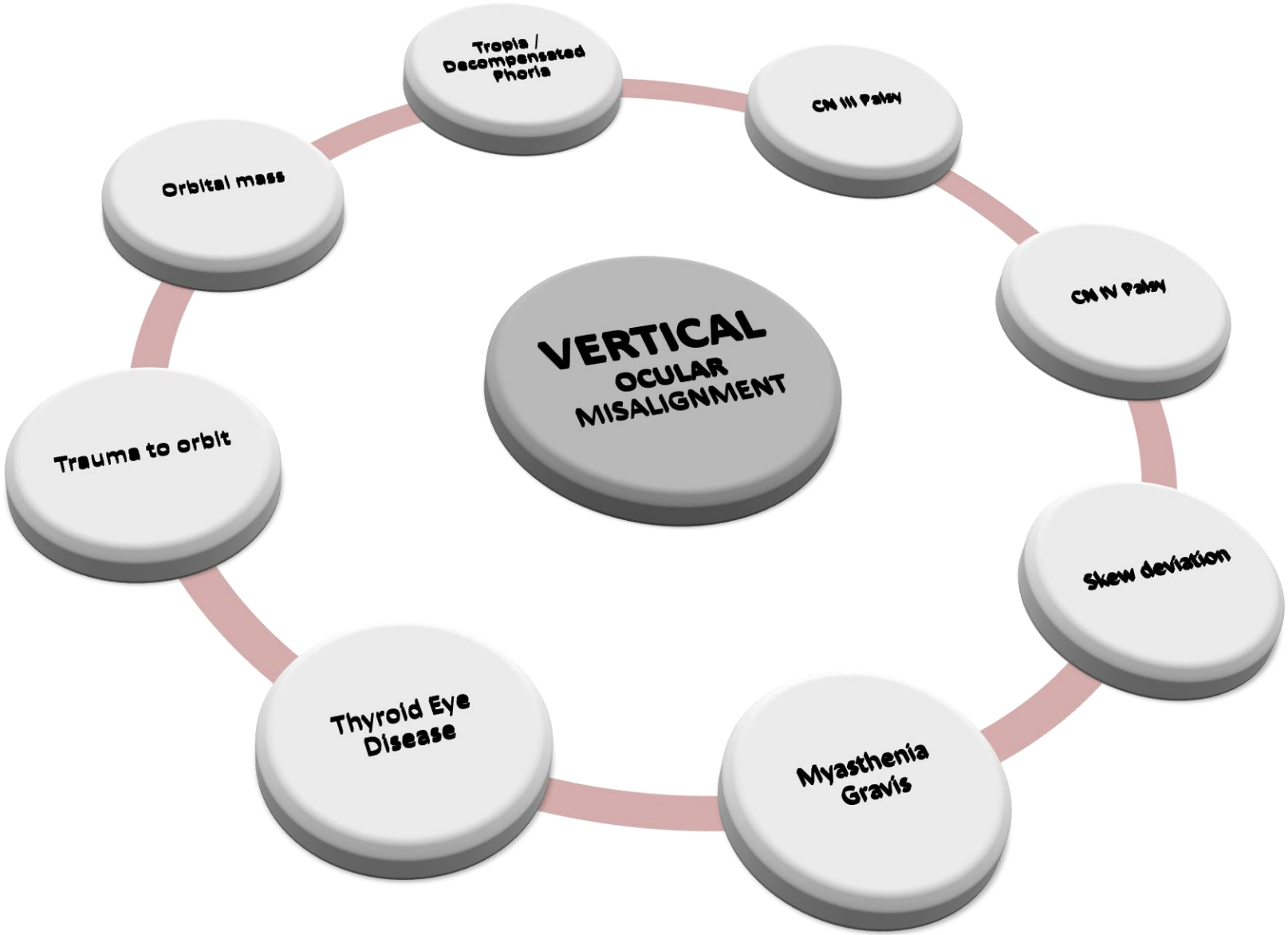
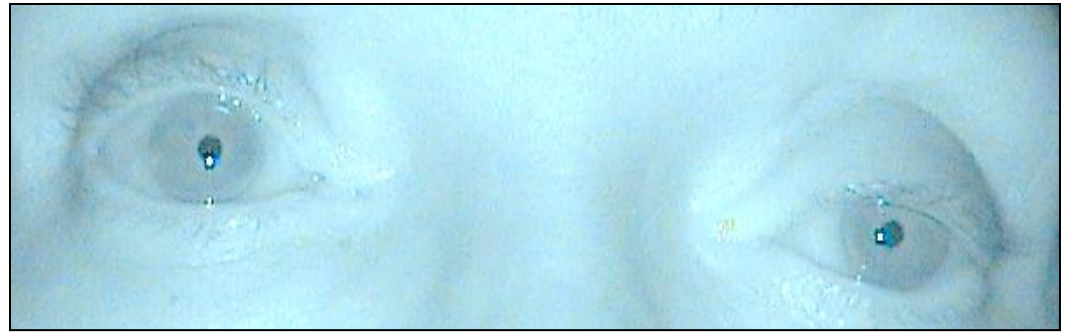

VERTICAL
OCULAR
MISALIGNMENT
Tropia / Decompensated Phoria
CN III Palsy
CN IV Palsy
Skew deviation
Myasthenia Gravis
Thyroid Eye Disease
Trauma to orbit
Orbital mass

Exam Findings

- VA: 20/25- OD 20/20- OS
- Color 13/14 OD 13/14 OS
- (-) RAPD
- CF: full OU
- IOP: 10 mm Hg OU
- DFE: Healthy optic discs, small cupping OU
- (-) edema OU , (-) pallor OU
- **Neurologic examination**
  - **Mild left upper extremity weakness**



Palpebral apertures: 11 mm OD and 7 mm OS

Exophthalmometry: 17 mm OD and 18 mm OS

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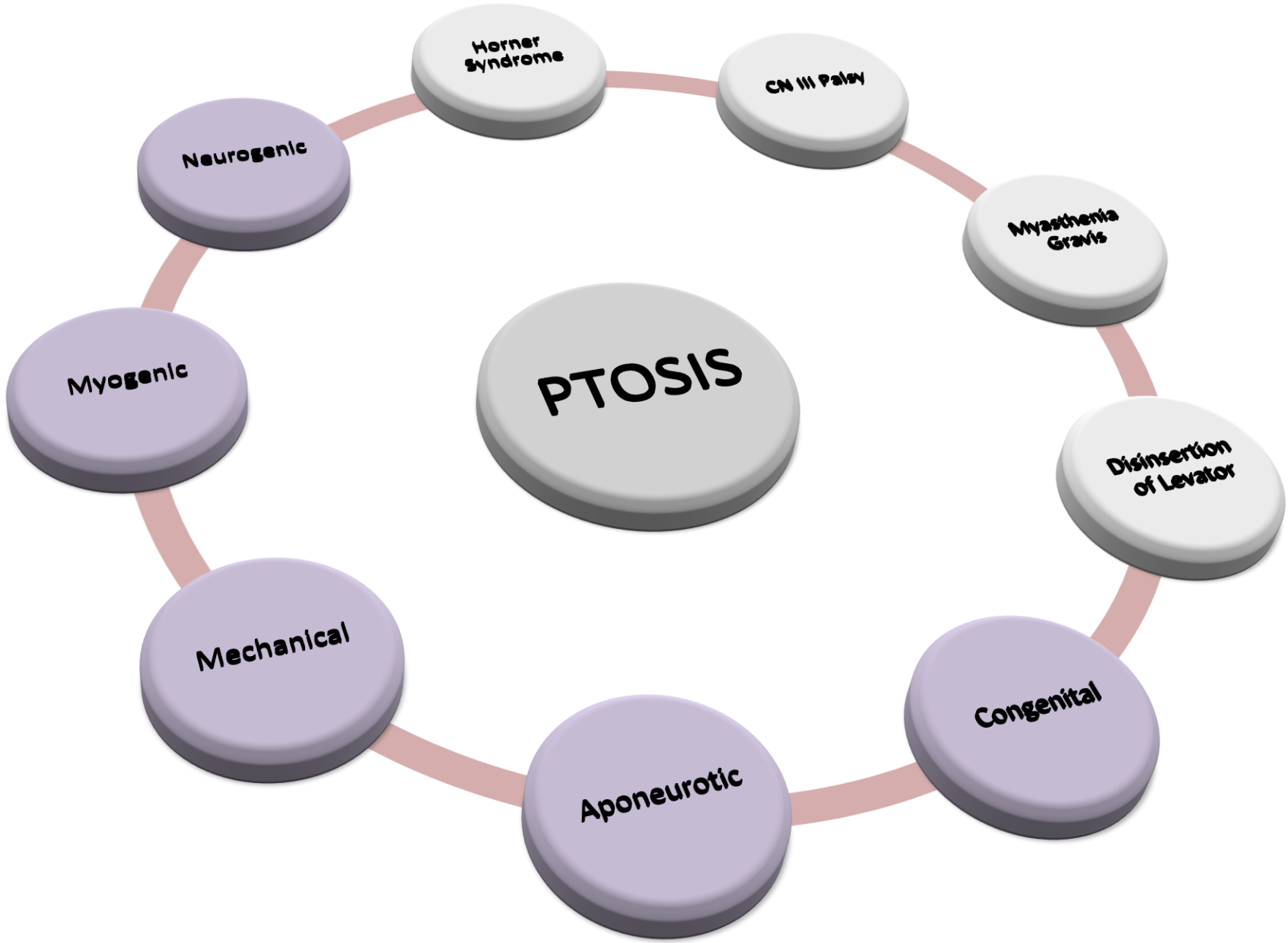
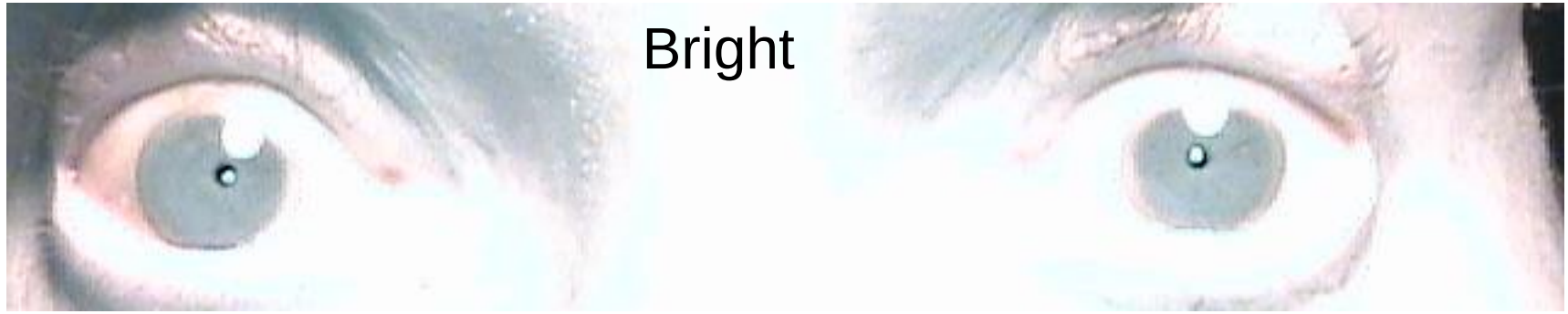

PTOSIS
Neurogenic
Horner Syndrome
CN III Palsy
Myasthenia Gravis
Disinsertion of Levator
Congenital
Aponeurotic
Mechanical
Myogenic



Pupils Isocoric (not suggestive of autonomic NS problem)

When there is asymmetric palpebral apertures, we must measure pupil sizes in bright and dim to determine if there may be a problem with the autonomic nervous system.

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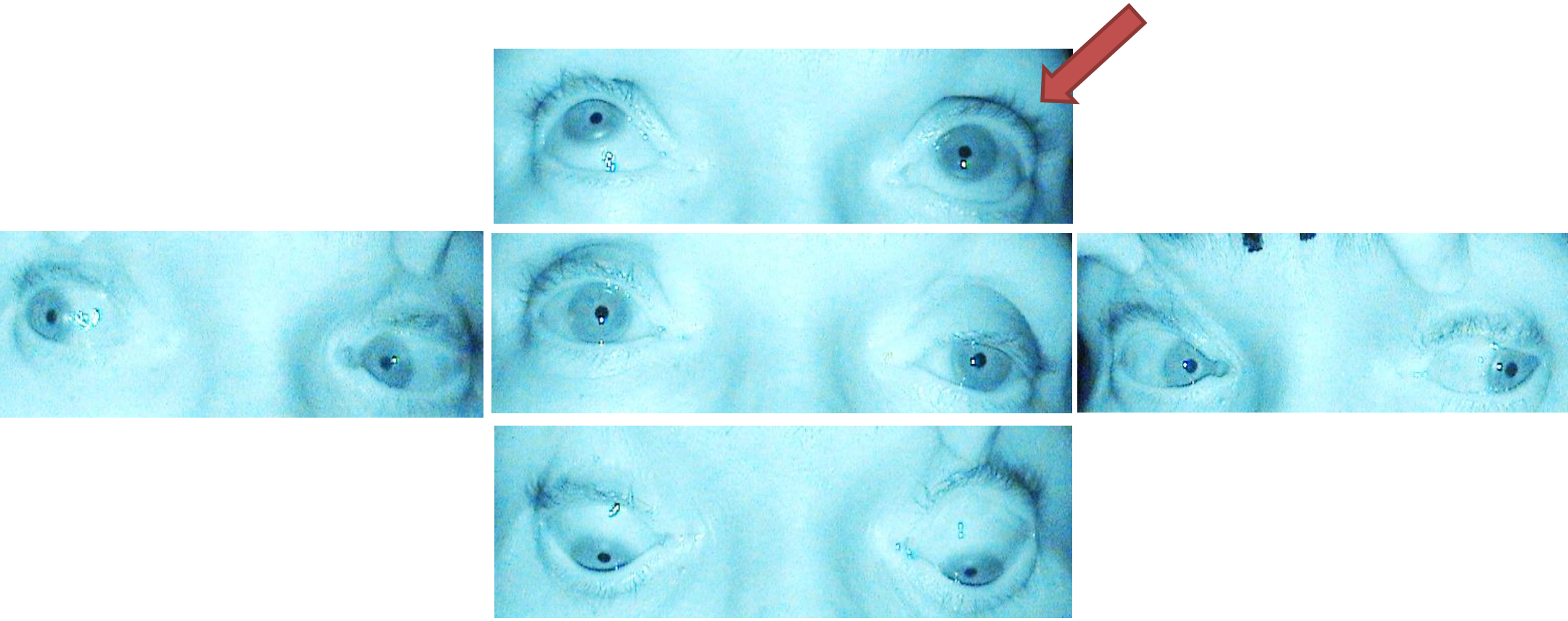
Ocular Motility



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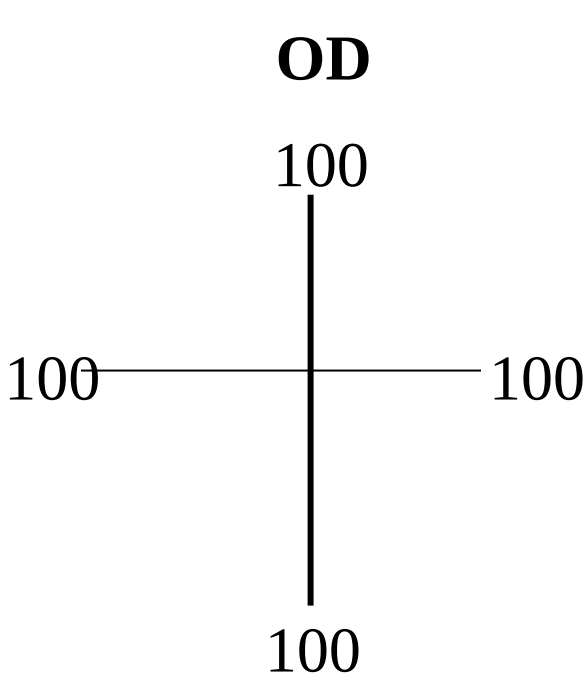
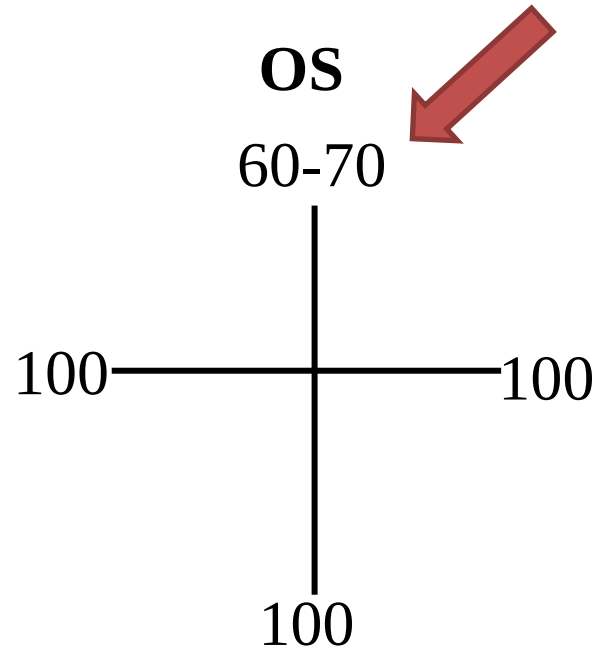
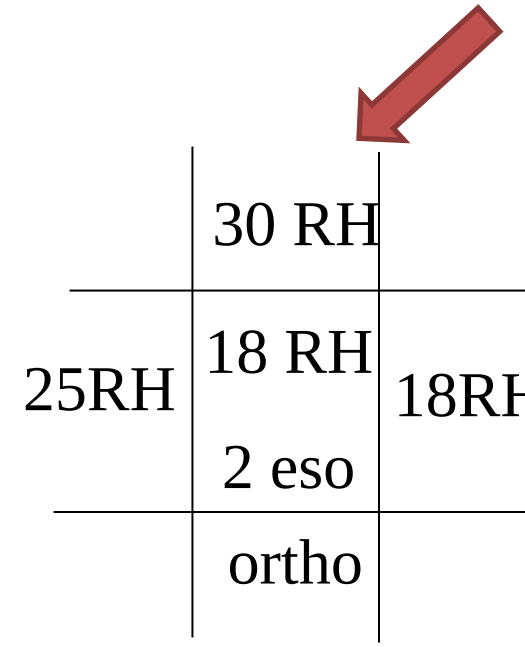
Ocular Motility







Head tilt: 20 right hyper on right head tilt, and 18 right hyper on left head tilt

What is the cause of the supraduction deficit?

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What is the cause of the vertical misalignment / supraduction deficit?

- CN III Palsy – superior division
- Myasthenia Gravis
- Thyroid Orbitopathy
- Skew Deviation
- CN IV Palsy

Is the asymmetric palpebral aperture related to the ocular misalignment? Or is that an unrelated aponeurotic ptosis?

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What is the cause of the vertical misalignment / supraduction deficit / ptosis?

- **CN III Palsy –** partial ? superior division?
  - Forced Duction: negative (no restriction)
- **Myasthenia Gravis**
  - Forced Duction: negative (no restriction)
- **Orbital Mass**
  - Forced Duction: positive (restriction)
- **Thyroid Orbitopathy**
  - Forced Duction: positive (restriction)
- **Skew Deviation**
  - Torsion: higher eye intorted
- **CN IV Palsy**
  - Torsion: higher eye extorted

CN III and skew deviation would be emergent. We would need to r/o an aneurysm and stroke, respectively.

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The eye moved up somewhat, but not fully.
We called it an equivocal forced duction test.

There was no torsion on double Maddox rod testing.

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WHICH IS IT?

Work-Up

- **Could be a partial CN III palsy**
  - Get MRI / MRA to r/o aneurysm

- Get labs to r/o other causes
  - CBC with differential, platelet count, ESR (Westergren), C-reactive protein
  - Lyme titer
  - RPR, FTA-ABS
  - ACE
  - Acetylcholine receptor antibody testing (binding, blocking and modulating),
  - TSH, t4 and anti-thyroid antibodies (anti-thyroperoxidase, anti-thyroglobulin, TSI – thyroid stimulating immunoglobulin, TSH receptor antibody)

CN III palsy could be caused by GCA!

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Results

- MRI report
  - mild bilateral proptosis
  - prominent orbital fat
  - mild fatty atrophy of inferior, lateral, and medial rectus muscles OU

- Lab testing
  - elevated TSH at 9.55
  - T3 at 37
  - thyroglobulin antibodies at 1790
  - thyroid peroxidase antibodies at > 1000

Elevated TSH indicates hypothyroidism.
Low TSH indicates hyperthyroidism.

Confirms thyroid dysfunction and likely thyroid eye disease (Hashimoto's thyroiditis)!

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Diagnosis / Treatment

- DX: autoimmune thyroid disease with subclinical hypothyroidism and mild thyroid orbitopathy
- Endocrine consult
  - started on levothyroxine 25mcg daily

But, since it could have been a partial CN III palsy, we still needed to r/o aneurysm!

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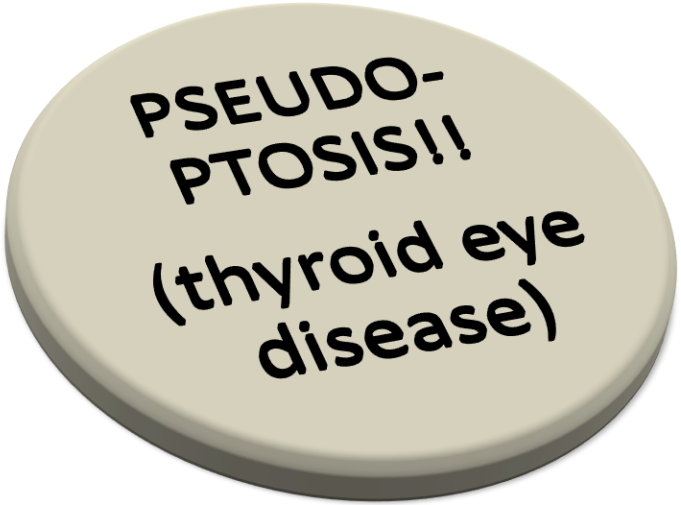
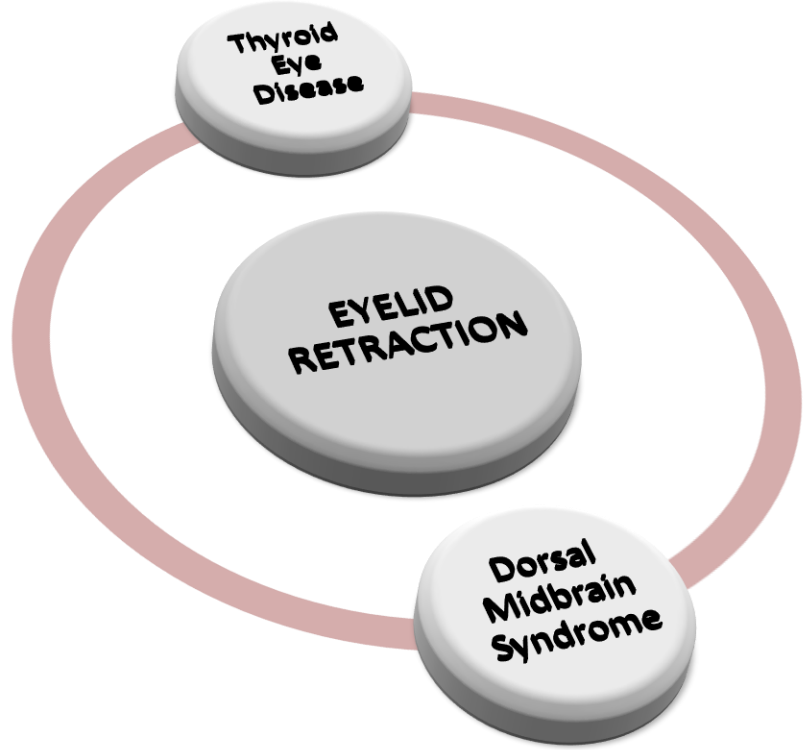
What about the ptosis?



It was lid retraction in the fellow eye!



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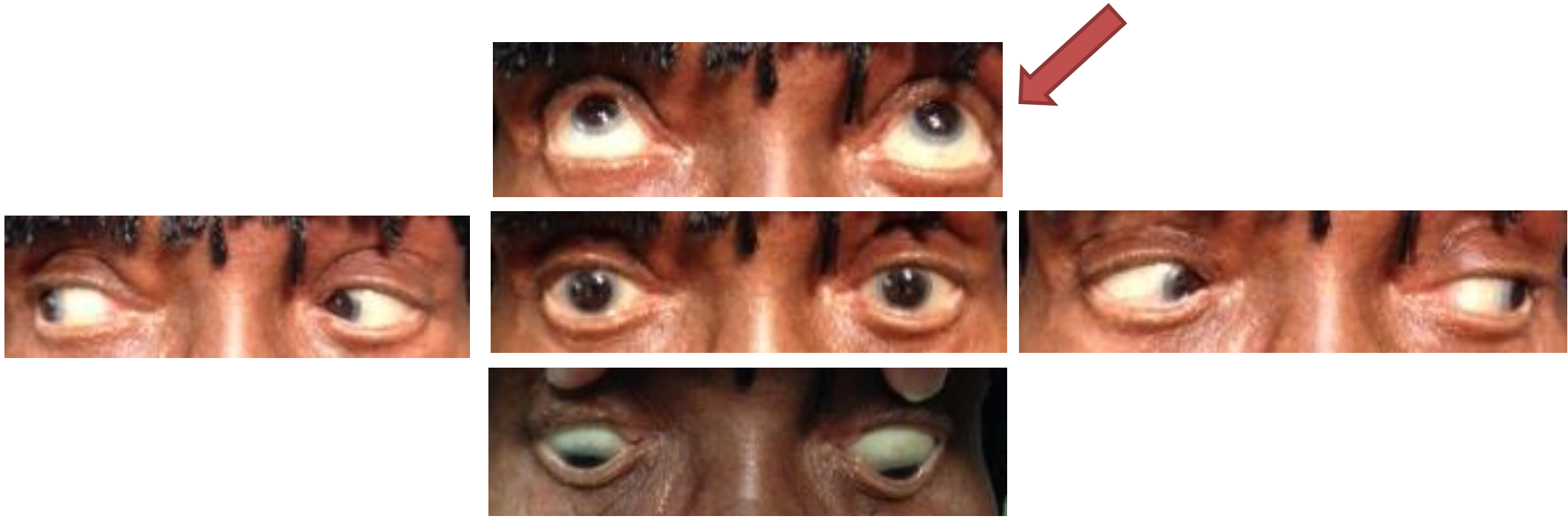
6 week follow-up

Pt reports no diplopia x 2 weeks, and lids now symmetric.



MILD TED

58% regress with achieving a euthryoid state

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THYROID ANTIBODIES

- In many patients with hypothyroidism or hyperthyroidism, lymphocytes react against the thyroid (thyroid autoimmunity) and make antibodies against thyroid cell proteins.

- THYROID ANTIBODIES
  - **Thyroid peroxidase antibody**
  - **Thyroglobulin antibody**
    - **Ex: elevated thyroid peroxidase antibody or thyroglobulin antibody in a patient with hypothyroidism indicate HASHIMOTO's. In this case, the antibodies are NOT used to monitor response to treatment. We use the TSH and FT4 for that.**
  - **Thyroid Stimulating Immunoglobulin (TSI)**
  - **Thyrotropin receptor antibody test (TSHR or TRAb)**
    - **Ex: elevated TSI and TRAb in a patient with hyperthyroidism can be associated with Graves' Disease. In this case, we CAN use the antibodies to assess response to treatment.**

Elevated TSH indicates hypothyroidism.
Low TSH indicates hyperthyroidism.

TSH Receptor Antibody (TRAB) is the primary autoantigen in Thyroid Eye Disease or Graves Disease

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COMMON THYROID CONDITIONS

- **Grave's Disease**
  - Autoimmune disease in which the immune system attacks the thyroid and causes it to make more thyroid hormone than the body needs
  - Common cause of **HYPERTHYROIDISM**
  - Affects more women than men
  - About ¼ - ½ people with Graves' develop TED

TED is more common with Graves, but can also be seen in Hashimoto's.

- **Hashimoto's Thyroiditis**
  - Autoimmune disease that damages the thyroid gland, decreasing production of T3 and T4
  - Most common cause of **HYPOTHYROIDISM**
  - Affects more women than men

Hypothyroidism is among the most frequent chronic diseases in the elderly

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THYROID EYE DISEASE

- INFILTRATIVE ORBITOPATHY
  - Red Eye
  - Ocular Inflammation
  - Eyelid Edema
  - Proptosis
  - Motility Restriction
  - Optic Nerve Compression (OPTIC NEUROPATHY)

TED can affect efferent visual system and the afferent visual system!

It is important to diagnose TED early and treat as needed in attempt to prevent TED associated optic neuropathy.

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MOTILITY RESTRICTION

- INFERIOR RECTUS 60-70%
- MEDIAL RECTUS 25%
- SUPERIOR RECTUS 10%
- DIURNAL VARIATION
- GAZE INDUCED IOP RISE (upgaze > lateral gaze)

INFERIOR Rectus is the most commonly involved muscle in TED. Which would cause a problem with UPGAZE (muscle BELLY is involved not letting back of globe move down)

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TOBACCO USE AND TED

Need to Educate Patients on Worse Prognosis of Thyroid Eye Disease (TED) with Smoking.

Pts with TED, or even thyroid dysfunction, need to be educated to stop smoking.

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HOW TO CLASSIFY AND STAGE TED TO DETERMINE THE PROPER MANAGEMENT PLAN (newer classifications)

Hindawi Publishing Corporation
Journal of Ophthalmology
Volume 2015, Article ID 249125, 16 pages
http://dx.doi.org/10.1155/2015/249125

- **VISA** (vision, inflammation, strabismus, appearance)

  EUGOGO (European Group of Graves' Orbitopathy)

Review Article

Graves' Ophthalmopathy: VISA versus EUGOGO Classification, Assessment, and Management

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Both Systems Consider Both Severity and Activity

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Review Article

Clinical Activity Score

Graves' Ophthalmopathy: VISA versus EUGOGO Classification, Assessment, and Management

Hindawi Publishing Corporation
Journal of Ophthalmology
Volume 2015, Article ID 249125, 16 pages
http://dx.doi.org/10.1155/2015/249125

EUGOGO

Table 2: Clinical Activity Score (CAS) (amended by EUGOGO after Mourits et al.). One point is given for the presence of each of the parameters assessed. The sum of all points defines clinical activity: active ophthalmopathy if the score is above 3/7 at the first examination or above 4/10 in successive examinations.

	For initial CAS, only score items 1–7
1	Spontaneous orbital pain
2	Gaze evoked orbital pain
3	Eyelid swelling that is considered to be due to active GO
4	Eyelid erythema
5	Conjunctival redness that is considered to be due to active GO
6	Chemosis
7	Inflammation of caruncle OR plica
	Patients assessed after follow-up (1–3 months) can be scored out of 10 by including items 8–10
8	Increase of >2 mm in proptosis
9	Decrease in uniocular ocular excursion in any one direction of >8°
10	Decrease of acuity equivalent to 1 Snellen line

VISA

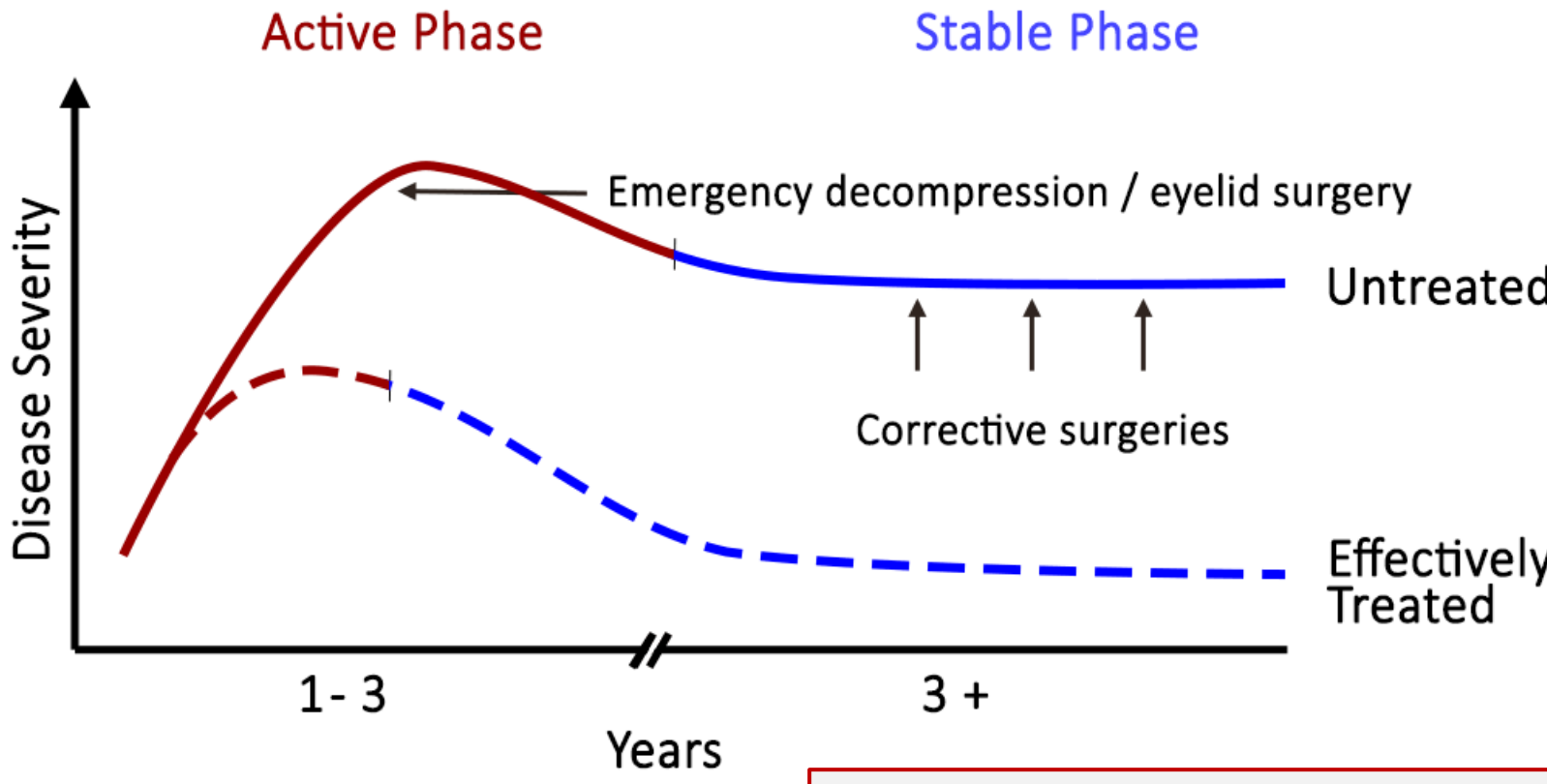
Table 3: VISA Inflammatory Index (I) (Dolman and Rootman 2006 [25], ITEDS modified). Patients with moderate inflammatory index (less than 4 of 10) are managed conservatively. Patients with high scores (above 5 of 10) or with evidence of progression in the inflammation are offered a more aggressive therapy.

Sign or symptom	Score
Caruncular edema	0: absent 1: present
Chemosis	0: absent 1: conjunctiva lies behind the grey line of the lid 2: conjunctiva extends anterior to the grey line of the lid
Conjunctival redness	0: absent 1: present
Lid redness	0: absent 1: present
Lid edema	0: absent 1: present but without redundant tissues 2: present and causing bulging in the palpebral skin, including lower lid festoon
Retrobulbar ache At rest With Gaze	 0: absent; 1: present 0: absent; 1: present
Diurnal variation	0: absent; 1: present

Rundel's Curve

Without early treatment, will go on to need more surgeries.

Disease Time Course and Intervention Strategy



So, we need to be able to identify ACTIVE TED!

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SIGHT THREATENING TED
(Compressive optic neuropathy)

- Occurs in 3-5% of patients with Graves' Disease

Patient needs to be sent to the ED of a hospital for immediate IV steroid therapy (dexamethasone 4mg) and for potential urgent orbital decompression surgery, if needed.

Orbital decompression can stop progression of optic neuropathy if performed without delay and by an experienced surgeon.

Sight threatening TED is a MEDICAL EMERGENCY!!

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TREATMENT OF TED

(Depends on Severity)

- PRISMS & PATCHES (for diplopia)
- DROPS & OINTMENTS (for dry eye)
- ELEVATED SLEEPING HEAD POSITION (for edema)
- STOP SMOKING !!! (for prognosis)

- WHAT ELSE CAN BE DONE…
  - Prednisone
  - Steroid Sparing Agents
  - Orbital Radiation
  - Orbital Decompression

We now have a specific treatment for Thyroid Eye Disease!

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Teprotumumab (Tepezza)

- Human monoclonal antibody
- Administered by **IV infusion**
- **Every 3 weeks for 8 infusions**
- Acts as an insulin-like growth factor-1 receptor inhibitor to block secretion of inflammatory cytokines and reduce proptosis
- After 24 weeks, the patients who used the drug were more likely to achieve a proptosis reduction of at least 2 mm from baseline

First FDA approved medication for TED (January 2020).

> J Neuroophthalmol. 2021 Jan 4;Publish Ahead of Print. doi: 10.1097/WNO.0000000000001134. Online ahead of print.

Teprotumumab in Clinical Practice: Recommendations and Considerations From the OPTIC Trial Investigators

Raymond S Douglas [1], Yao Wang, Roger A Dailey, Gerald J Harris, Sara T Wester, Jade S Schiffman, Rosa A Tang, Brian Fowler, James Fleming, Terry J Smith

Affiliations + expand

PMID: 33417417 DOI: 10.1097/WNO.0000000000001134

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Teprotumumab (Tepezza)

- **To date, studies have shown that it is effective in:**
  - Decreasing proptosis
  - Eliminating diplopia
  - Reducing orbital pain, redness, and swelling
  - Improving functional vision
  - Improving appearance

Currently, only approved by insurance for those with clinically active TED (need to measure CAS)

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Teprotumumab (Tepezza)

- Warnings and Precautions
  - **Pre-existing IBD**
    - May cause exacerbation of preexisting IBD
    - Monitor for flare of IBD
    - May need to d/c med
  - **Hyperglycemia (IGF-1)**
    - 10% of patients (not all in preexisting diabetics)
    - BS should be checked before and during treatment
- Potential adverse reactions:
  - muscle spasms
  - hearing impairment (hypoacusis/tinnitus)
  - Nausea
  - Vomiting
  - Diarrhea
  - Alopecia
  - Headache

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CASE 2

74 YEAR-OLD WOMAN

- Presents emergently with her husband and sister for vision loss OS x 8 days
- Systemic Hx:
  - Alzheimer's disease
  - Hypertension
  - Hypercholesterolemia
  - Type II NIDDM
  - Anxiety
- Ocular Hx:
  - Glaucoma
- Medications:
  - Systemic: Metformin, unknown meds for conditions above
    - Ocular: Lumigan QHS OU, Combigan BID OU

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- **Prior to 8 days ago, she was still independent, despite her Alzheimer's; she was able to dress, feed herself and use her glaucoma medications on her own**

- **Her husband and sister both noticed a change in her mental status around the time of her vision changes. Now, she can no longer dress herself and she has been eating with her hands. They also notice that she has been walking into walls. Her husband has been putting in her eye drops.**

- **She also has been very tired and she, herself notes that something is not right, she just can't explain how she feels**

EYE EXAM FINDINGS

- Visual acuity:
  - OD: 20/30 OS: NLP
- Confrontation Fields:
  - OD: dense superior and inferior nasal defect
  - OS: unable to see fingers
- Ductions:
  - Limited supraduction bilaterally: 50%
  - No nystagmus
- Pupils:
  - 1.5 APD OS

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EYE EXAM FINDINGS

- Anterior Segment
  - Unremarkable
- Intraocular pressure:
  - OD: 18 OS: 18
- Posterior Segment: (undilated)
  - Lens: OD: 2+ NS, OS: 2-3+ NS
  - Optic Nerve: OD: unable to view, OS: .99/1.0

WHAT WOULD YOU DO WITH THIS PATIENT?

- Is she just noticing her vision loss from glaucoma?
- Is there anything concerning or urgent?
- What is the best management plan?

ASSESSMENT & PLAN

- Visual field defect
  - Dense nasal loss OD and no light perception vision OS.
  - Visual field loss, may be glaucomatous…
  - But, **cannot rule out a homonymous hemianopia** based on the pattern of loss, especially in the setting of the recent change in mental status.

  - Sent patient to the ED for **emergent CT scan of the brain without contrast to rule out acute intracranial pathology**.
  - **Alzheimer's dementia would not progress so quickly.**
  - **Alzheimer's pts are prone to falls…**

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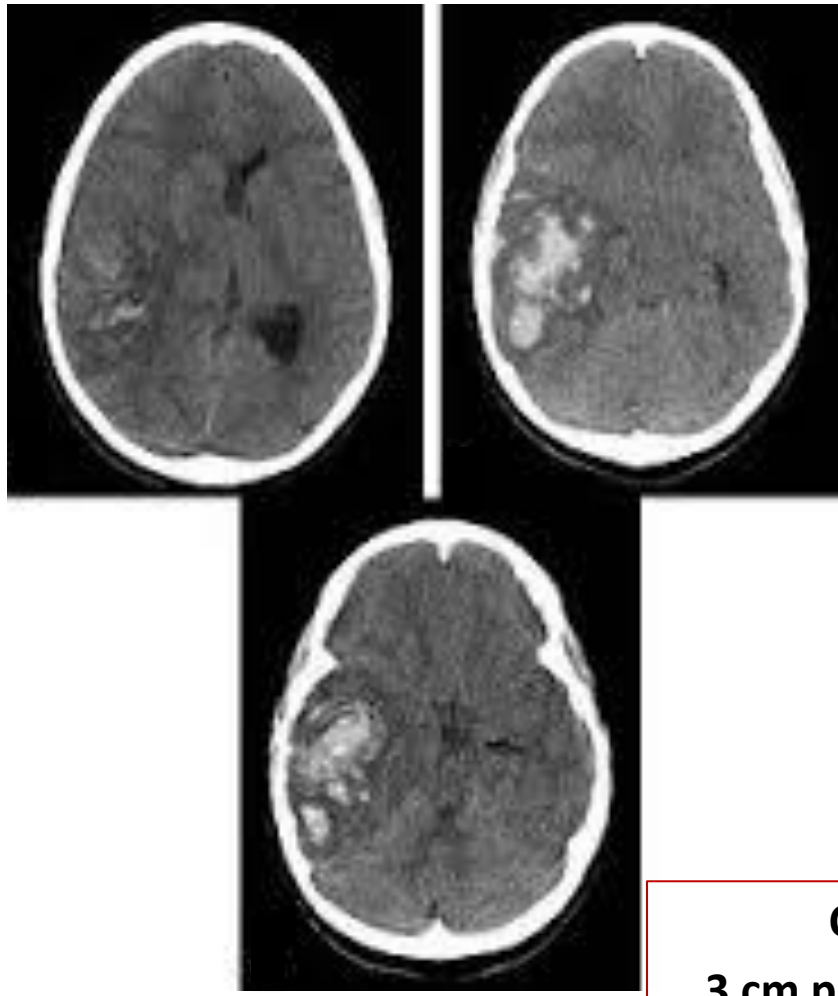
RESULTS OF CT



We need to be sure NOT to attribute ALL new findings to known underlying neuro-degenerative disease.

We need to be aware of clinical findings that suggest acute intracranial pathology.

Alzheimer's dementia does NOT rapidly progress in 1 week!

CT SCAN FINDING:

3 cm parenchymal bleed in the parietal and temporal lobes, with mass effect

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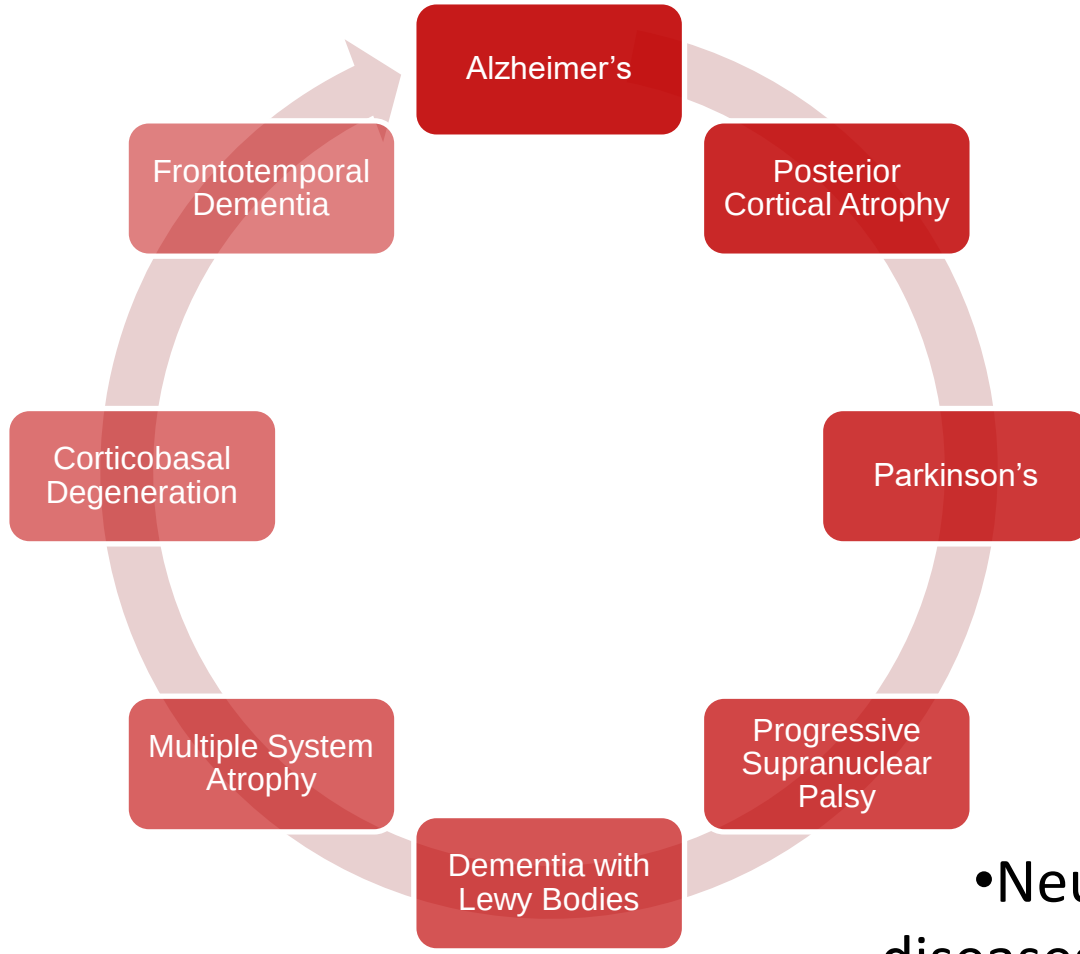
Neurodegenerative Diseases Causing Dementia



- Neurodegenerative diseases are both incurable and debilitating

Alzheimer's Disease

- Progressive neurodegenerative disease
- **Predominantly a disorder affecting memory**
- As the disease progresses, can affect
  - Orientation
  - Attention
  - Language
  - Executive function
  - Visuospatial

Memory impairment, specifically **short-term memory loss** is the most frequent feature of AD and is

Executive dysfunction and impaired visuospatial skills tend to be affected relatively early, while deficits in **language function and behavioral symptoms often manifest later** in the disease course.

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ALZHEIMER'S DISEASE (AD)

- patients with AD have **problems reading, driving, and performing tasks that depend on visual cognition**
- We need to determine ways to assess and manage visual cognition in AD
- We need to determine if patients can benefit from other treatments / rehab as well

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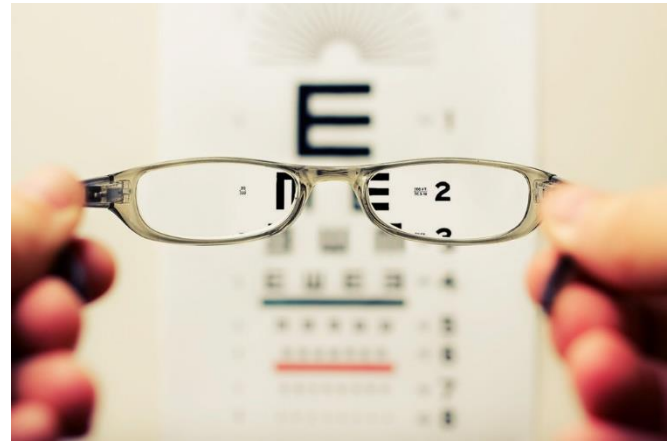
AD and Afferent Function Symptoms

- Blur
- Difficulty seeing in twilight or in rain
- Colors are washed out
- Vision just is not right



- Reduced Visual Acuity
- Reduced Color Discrimination
- Reduced Contrast Sensitivity
- Visual Field Defects

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SAGE-Hindawi Access to Research
Parkinson's Disease
Volume 2011, Article ID 908306, 9 pages
doi:10.4061/2011/908306

Research Article

Visual Symptoms in Parkinson's Disease

R. A. Armstrong

Visual symptoms are very common in PD!

Table 1: Visual signs and symptoms of Parkinson's disease (PD).

Ocular aspect	Change in PD
Visual acuity	Poor, especially at low contrast
Colour vision	Vision blurred for coloured stimuli Shortened colour fusion time Progressive deterioration
Visual fields	Increase in glaucomatous visual field defects Side effects of surgery
Saccadic eye movement	Reaction time and max. velocity of horizontal gaze slower Hypometria Amplitude increased after cued saccades
Smooth pursuit movement	Affected early in disease process Superimposed saccades Reduction in response magnitude
Optokinetic Nystagmus	Abnormal in some patients
Convergence	Impaired, associated with large exophoria, diplopia
Blink frequency	Reduced, causing abnormal tear film, dry eye and reduced vision
Blink reflex	Habituation not observed
Pupil diameter	Larger after light adaptation with anisocoria
Light reflex	Longer latency
Constriction time	Increased
Contraction amplitude	Reduced
Contrast sensitivity (CS)	Abnormal in some cases, intermediate to high frequencies
Temporal processing	Impaired ability to track rapid fluctuations Duration perception affected
Flash ERG	Reduced amplitude of "b" wave
PERG	Reduced amplitudes. Specific defect at medium SF Delayed P50
Cortical VEP	Delayed P100
Chromatic VEP	Increased latency and reduced Amplitude (esp. blue-yellow)
ERP	Abnormal. Delayed reaction times
Visuo-spatial	Difficulty in judging verticals, position of body parts, and in route-walking tasks
Orientation and motion discrimination	Impaired
Facial perception	Impaired ability to perceive and imagine emotional faces
Visual hallucinations	Chronic in 30–60% of treated cases

Abbreviations: ERG: Electroretinogram, ERP: event-related potentials, PERG: Pattern electroretinogram, SF: Spatial frequency, VEP: Visual evoke

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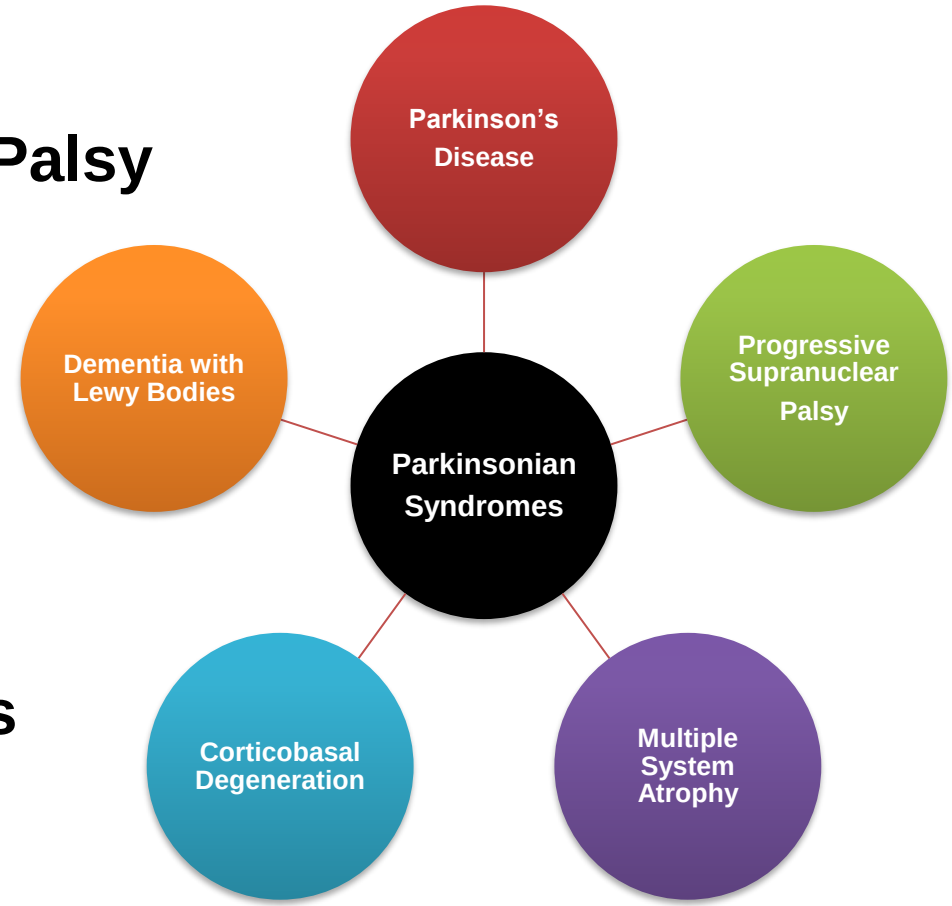
Parkinson Disease (PD)

- The most common cause of **parkinsonism**, a syndrome manifested by:
  - Rest tremor
  - Rigidity
  - Bradykinesia
  - Postural instability
  - Autonomic dysfunction

Remember:
Bradykinesia means slowness of movement

Parkinsonian Syndromes

- **Parkinson's Disease**
- **Progressive Supranuclear Palsy**
- **Multiple System Atrophy**
- **Corticobasal Degeneration**
- **Dementia with Lewy Bodies**



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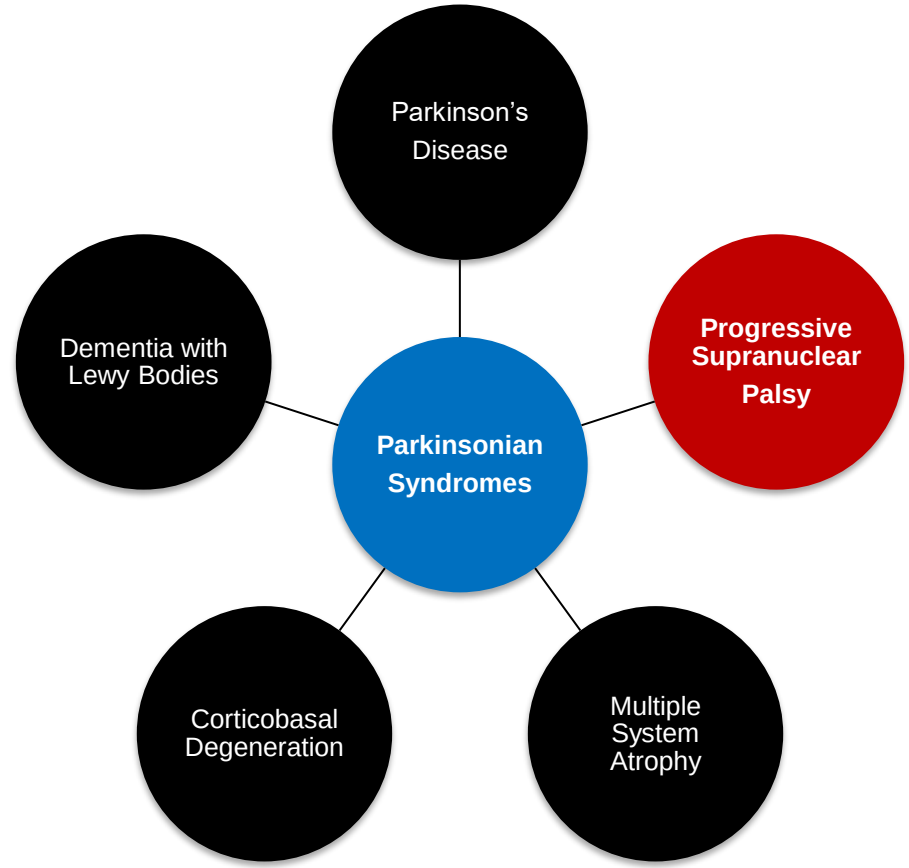
How To Differentiate From Parkinson's Disease

- **PD should not demonstrate limited vertical ductions**
  - **There is jerkiness and cogwheeling, but not significant ductional limitations**
- If **limited vertical eye movements** (particularly downgaze), need to consider **Progressive Supranuclear Palsy (PSP)**



CASE 3

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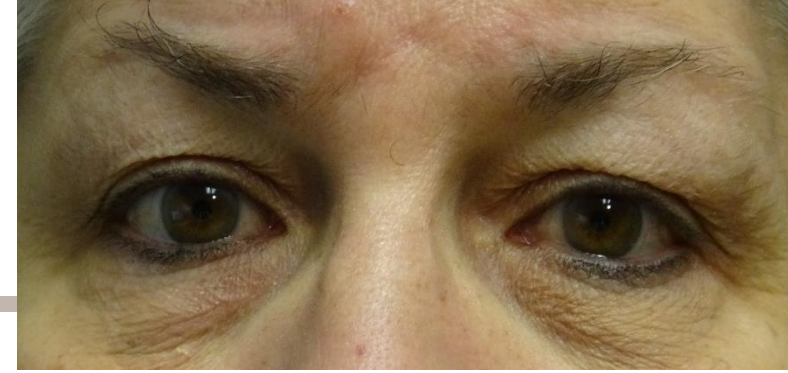
66 Year-Old Woman

- Referred by neurology due to abnormal EOMs
- Has been having trouble walking, but she attributes it to arthritis of her knees
- When she gets up, she has to wait for a while before moving
- She holds onto her husband when she walks



- Currently wearing progressive lenses
- Having some difficulty with prolonged reading
- She would like to spend more time reading
- Denies diplopia

- Not currently driving; her husband is not comfortable with letting her drive until they figure out what is going on
- She wants to get back to driving

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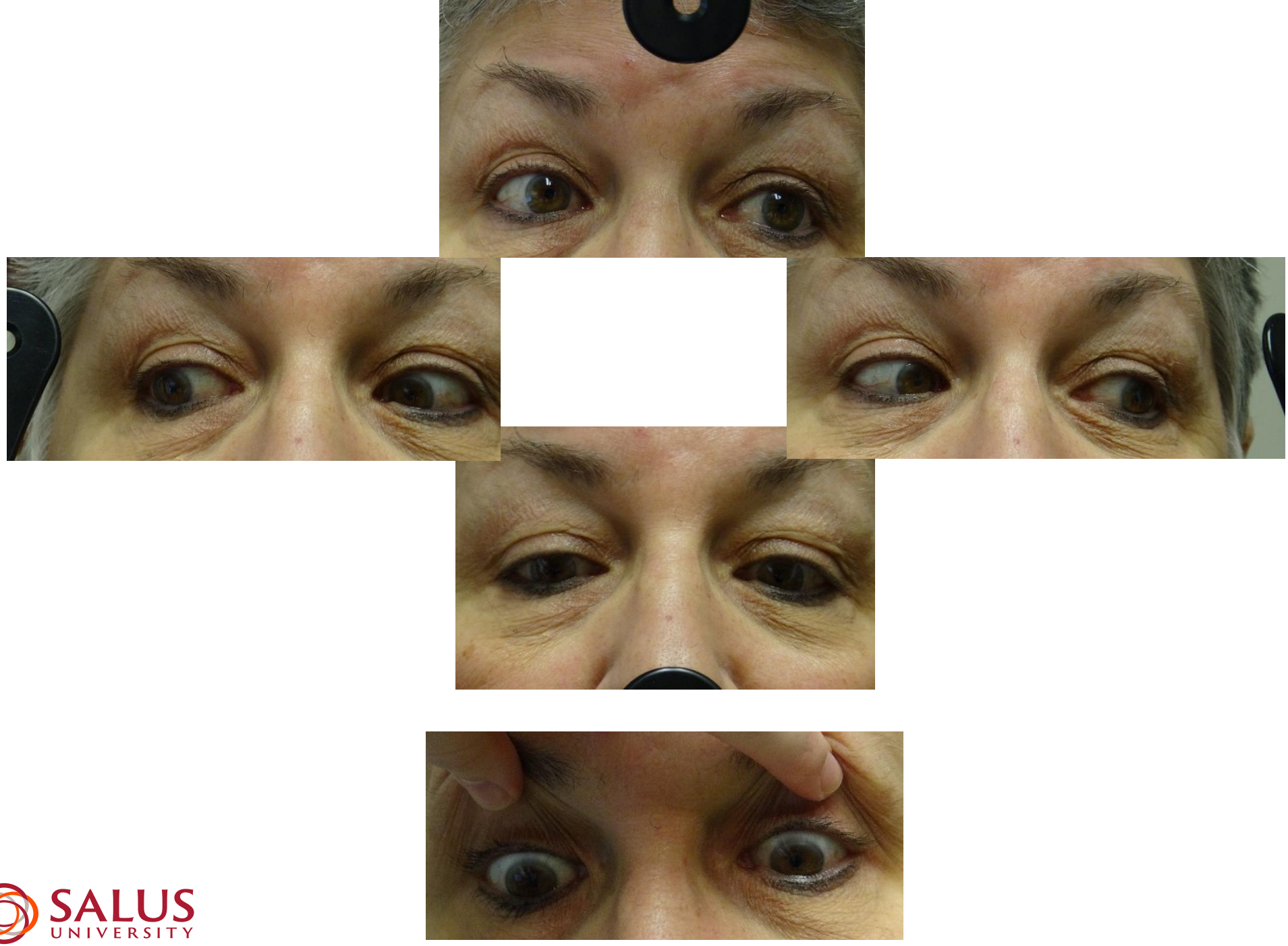

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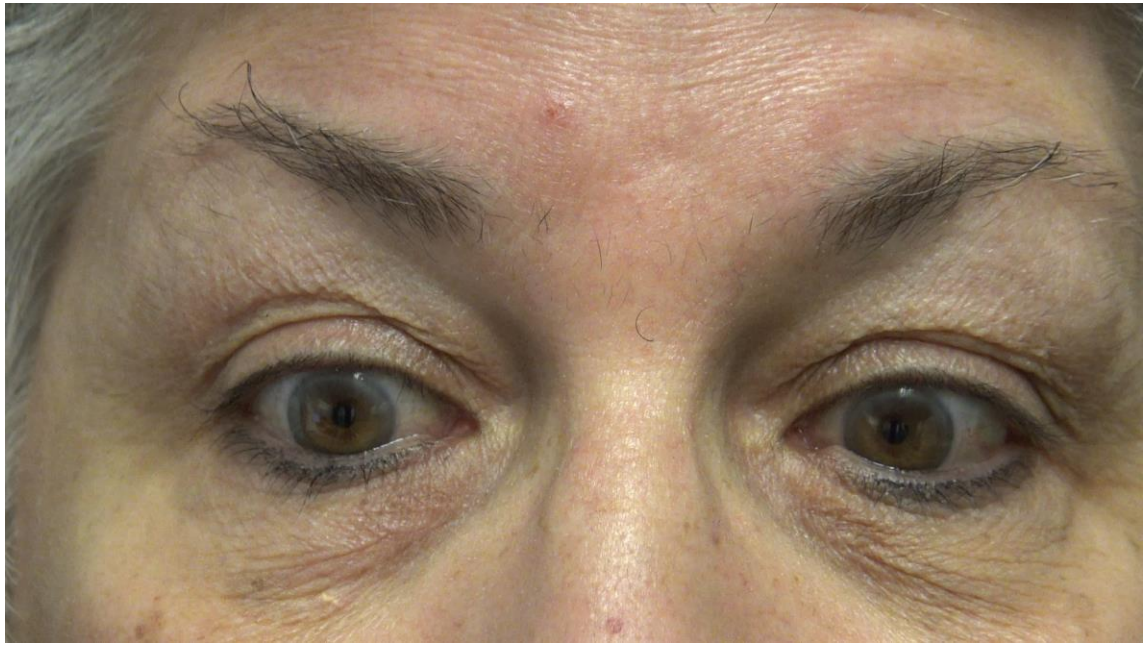

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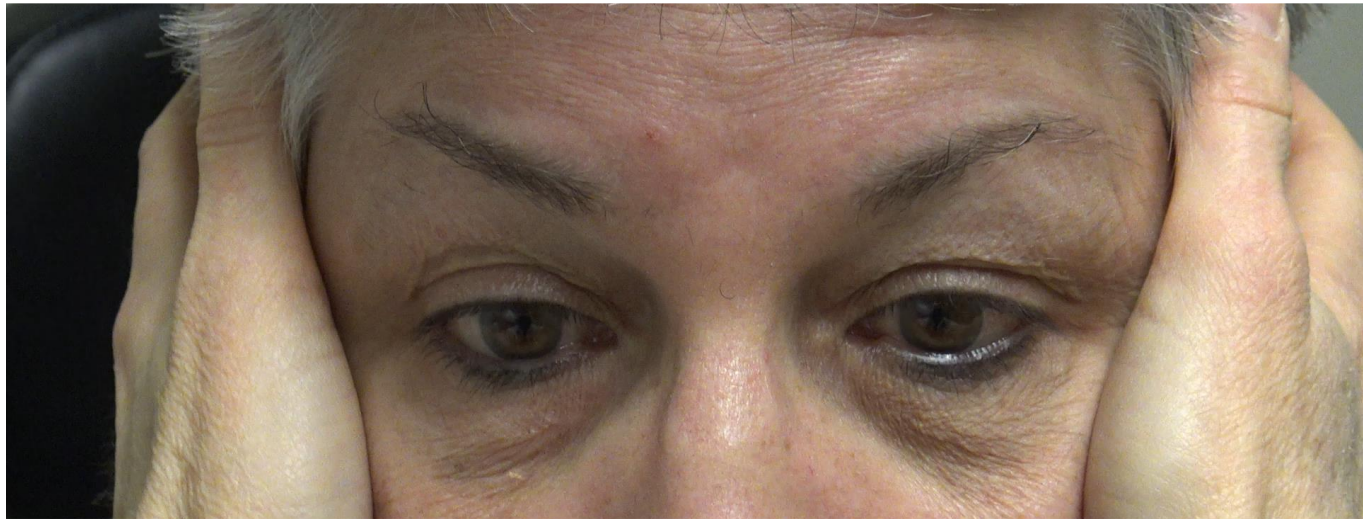
Doll's Head (Oculocephalic) Reflex



A dissociation between impaired voluntary eye movements (pursuits and saccades) and preserved involuntary eye movements (VOR) is called a supranuclear gaze palsy. This is most commonly seen in PSP.

The Doll's head maneuver assesses the vestibulo-ocular reflex (VOR). We discussed this as part of the HINTS testing for nystagmus, but it is also helpful to distinguish limited ductions as either supranuclear or infranuclear in origin.

The fact that the ability to move the eyes up and down is greater with the Doll's Head maneuver than just with ductions confirms that it is a **supranuclear problem**. Therefore, we know it is not a problem with the nerves or, NMJ, or muscles.

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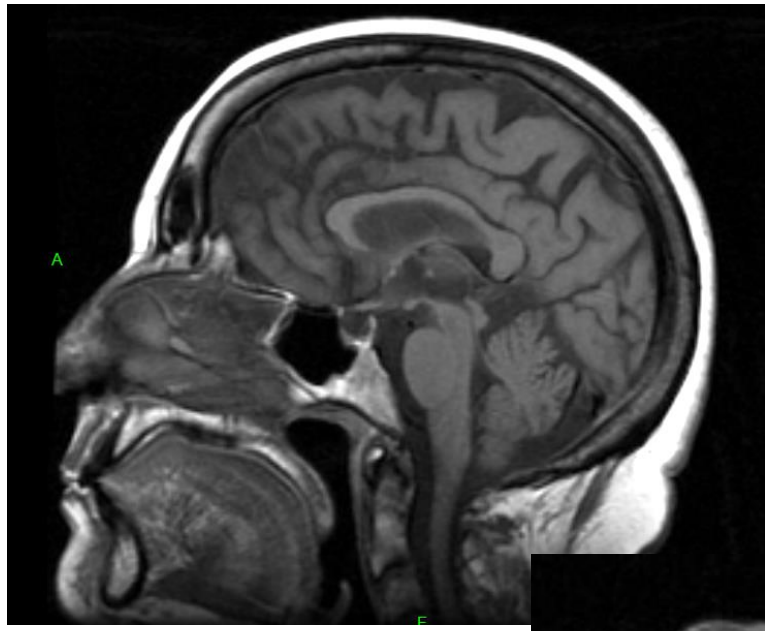

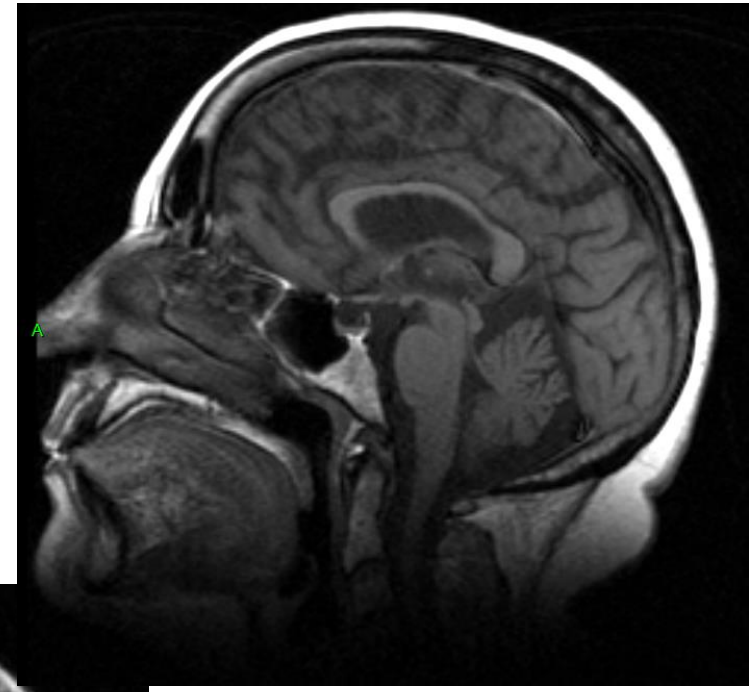

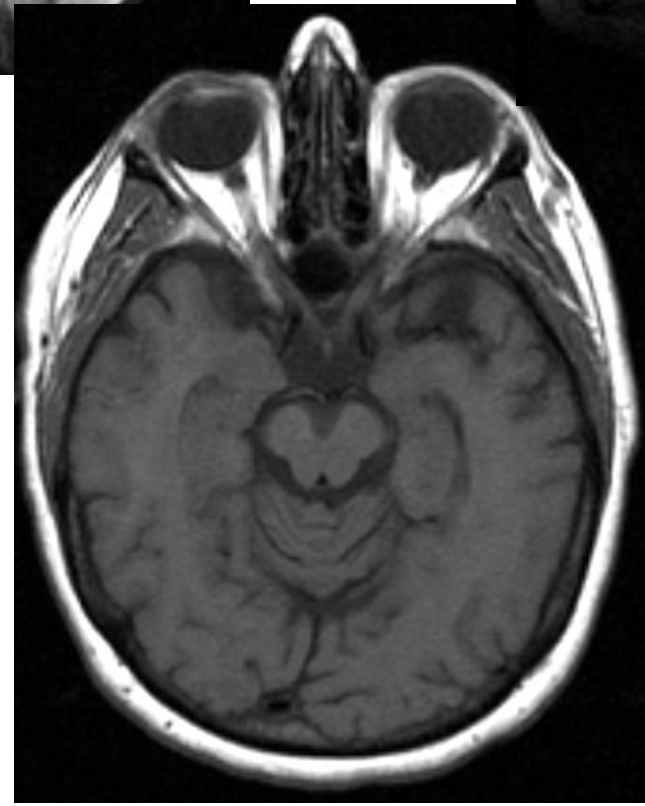

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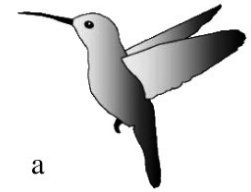
PSP - diagnosis

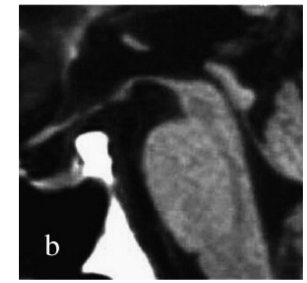
Characteristic eye movements help confirm the diagnosis of PSP

- **Initially as slowing of vertical saccades**
- **Gradually evolve into hypometric saccades, square-wave jerks (fixation instability)**
- **Eventually supranuclear vertical gaze palsy**

The radiologic **hummingbird sign** (also called the penguin silhouette sign) results from the prominent midbrain atrophy in PSP with a relatively preserved pons, resembling a hummingbird or penguin in silhouette on midsagittal MRI of the brain.



Remember that the midbrain is the vertical gaze center.



Classic Hummingbird sign of MRI

(a) Hummingbird, (b) mid-sagittal plain MRI in PSP. The region including the most rostral midbrain, the midbrain tegmentum, the pontine base, and the cerebellum appears to correspond to the bill, crown, body, and wing, respectively, of a hummingbird (i.e., the "hummingbird sign"). Adapted from "Kato N, Arai K, Hattori T. Study of the rostral midbrain atrophy in progressive supranuclear palsy. J Neurol Sci. 2003 Jun 15;210(1–2):57–60. "

From: Progressive Supranuclear Palsy

StatPearls [Internet].
Treasure Island (FL): StatPearls Publishing; 2018 Jan-.

Neuropathologic examination remains the gold standard for its definitive diagnosis

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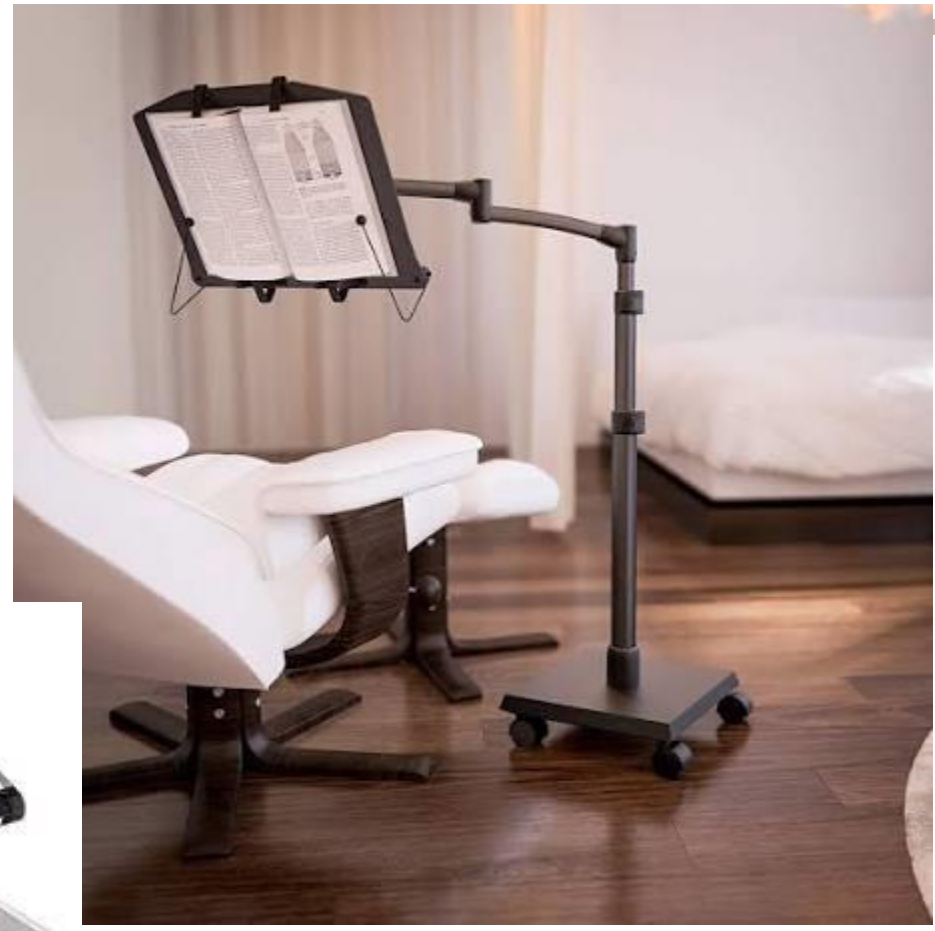
What Can We Do To Help?

- Since her vertical motilities are limited (superior > inferior) we need to get her out of progressive lenses
  - They are constantly making her try to make vertical eye movements
- She will do better with separate DVO and NVO lenses
- Incorporated yoked prism since she cannot move her eyes up at all.
  - Want to move image down slightly so it is move comfortable for her
  - 3 BU prism OU (3 BU in each eye)

- Also, recommended a reading stand so that her reading material can be held at eye level



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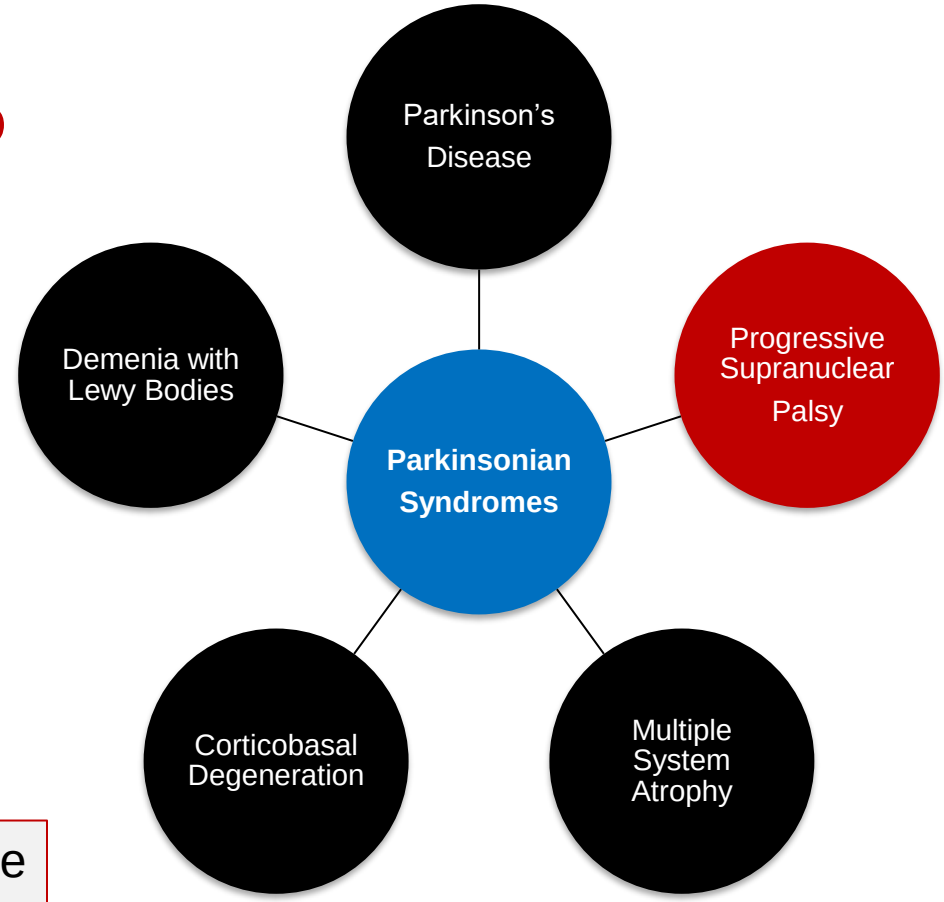
Progressive Supranuclear Palsy (PSP)

- **More severe and faster progression compared with PD**
- Responds poorly to L-dopa
- Early falls (often backwards)
- Mean age of diagnosis 65
- Average time to death is 7 years

PSP has never been diagnosed before the age of 40.



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CASE 4

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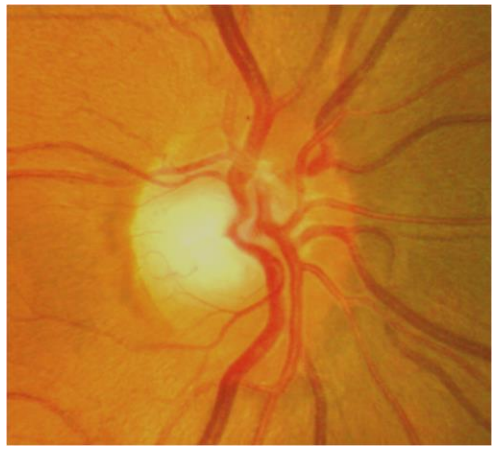
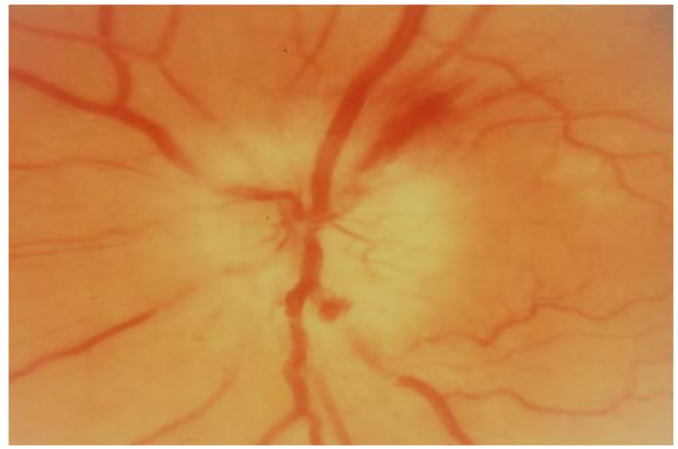
84 Year-old woman

- **HISTORY:**
- Ocular History:
  - Bilateral OAG, bilateral Fuch's endo dystrophy
  - S/P PKP OS
  - S/P BRVO c focal laser tx OS
- Systemic History:
  - DM, HTN, MI, CABG, arthritis, anemia (currently tx c iron supp)
- Symptoms: Reduced VA OS x 3 wks, L temple pain, scalp tenderness, jaw claudication, malaise, recent flare-up of asthma

Exam Findings

- **VA:** OD 20/200 **BCVA**: OD 20/200 (+) RAPD OS
  OS CF OS 20/400 (-6 D cyl)
  VF: significant constriction OS

DFE : OD - distinct margins / NRR pink

OS – diffusely pale / residual edema S and I / hemes

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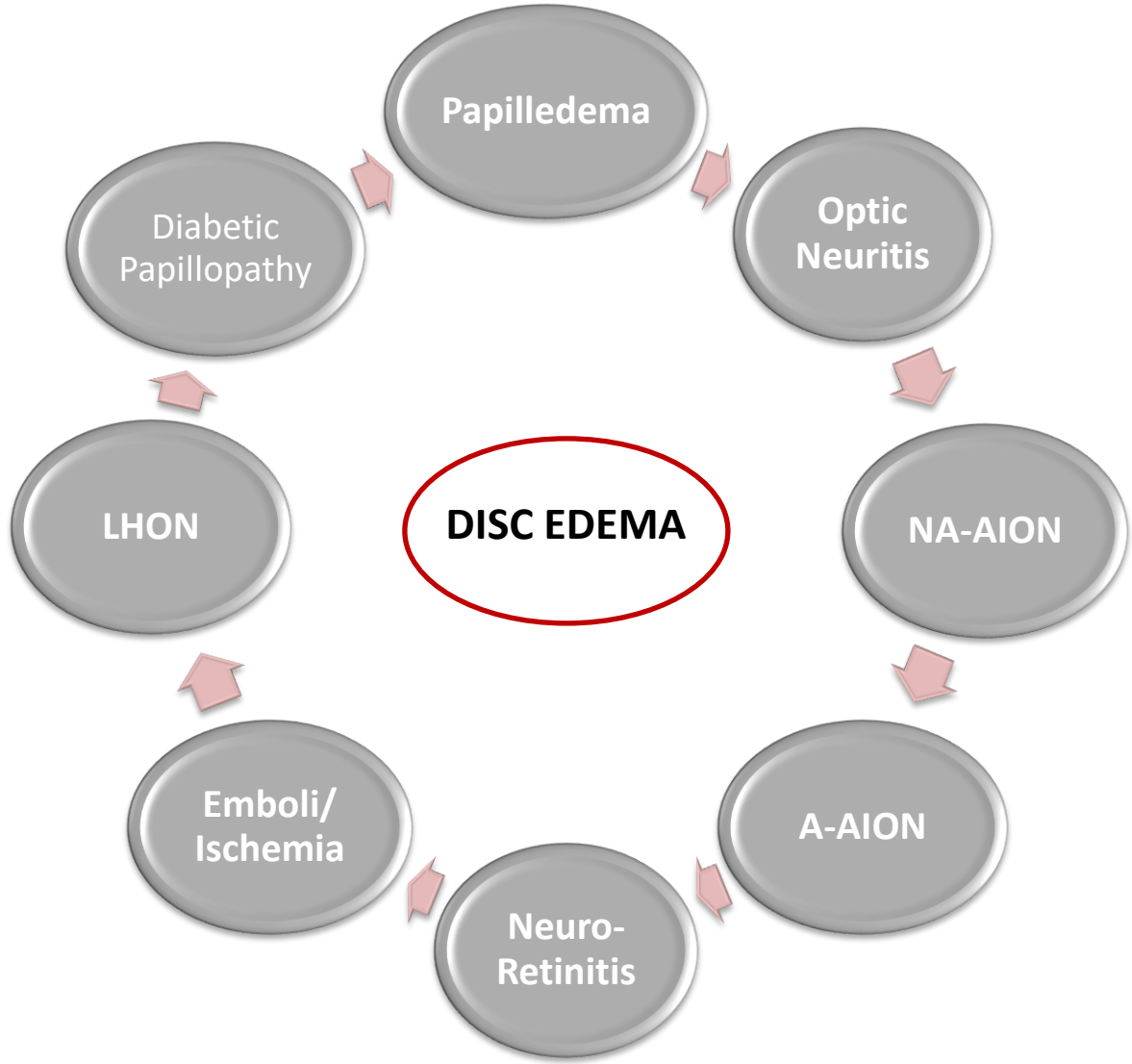
CAUSES OF DISC EDEMA



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WHICH IS IT?

GCA SYMPTOMS

- **HEADACHE**
- **TENDER TEMPORAL ARTERY**
- **MYALGIA/ ARTHRALGIA**
- **FEVER**
- **WEIGHT LOSS**
- **JAW CLAUDICATION**
- **ANOREXIA (poor appetite)**
- **MALAISE**

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Laboratory Results

- STAT ESR: **125 mm /sec**
- **CRP : 12.7 mg/L**
- **Platelets: 267**
- **TAB: POSITIVE**

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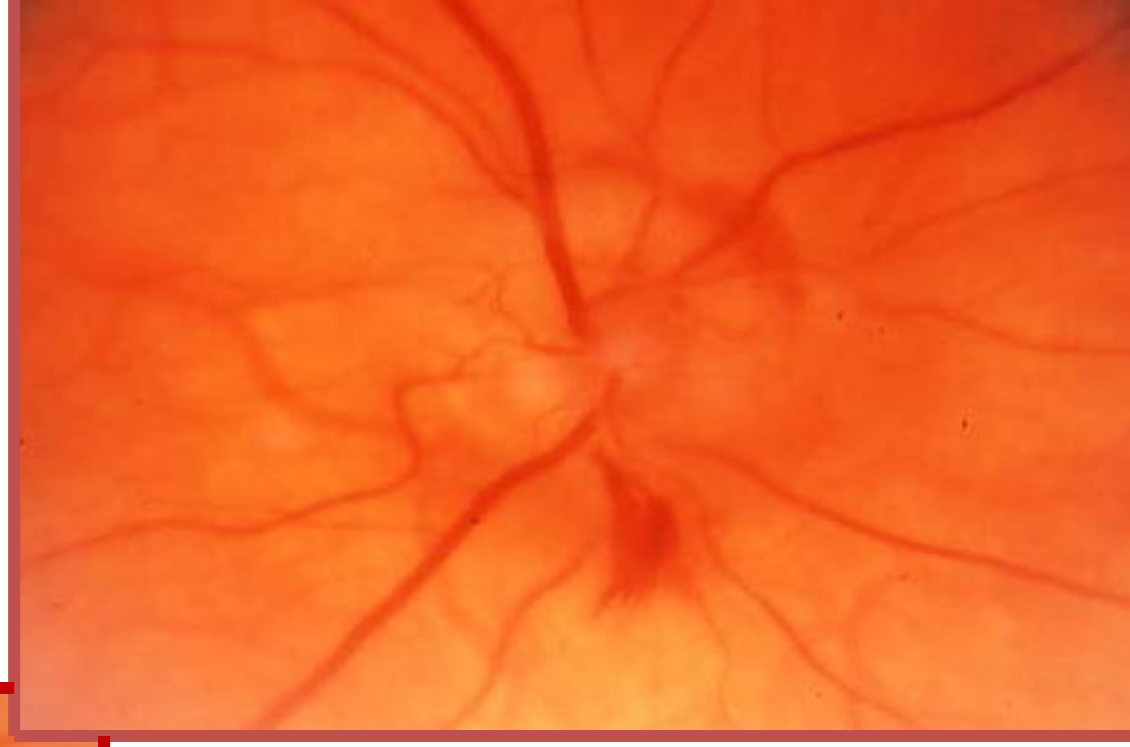
NAAION

- Segmental edema
- Hemorrhage
- Luxury perfusion
- NRR pink
- No pain
- Nadir: 3.4 days
- Fellow eye: disc at risk

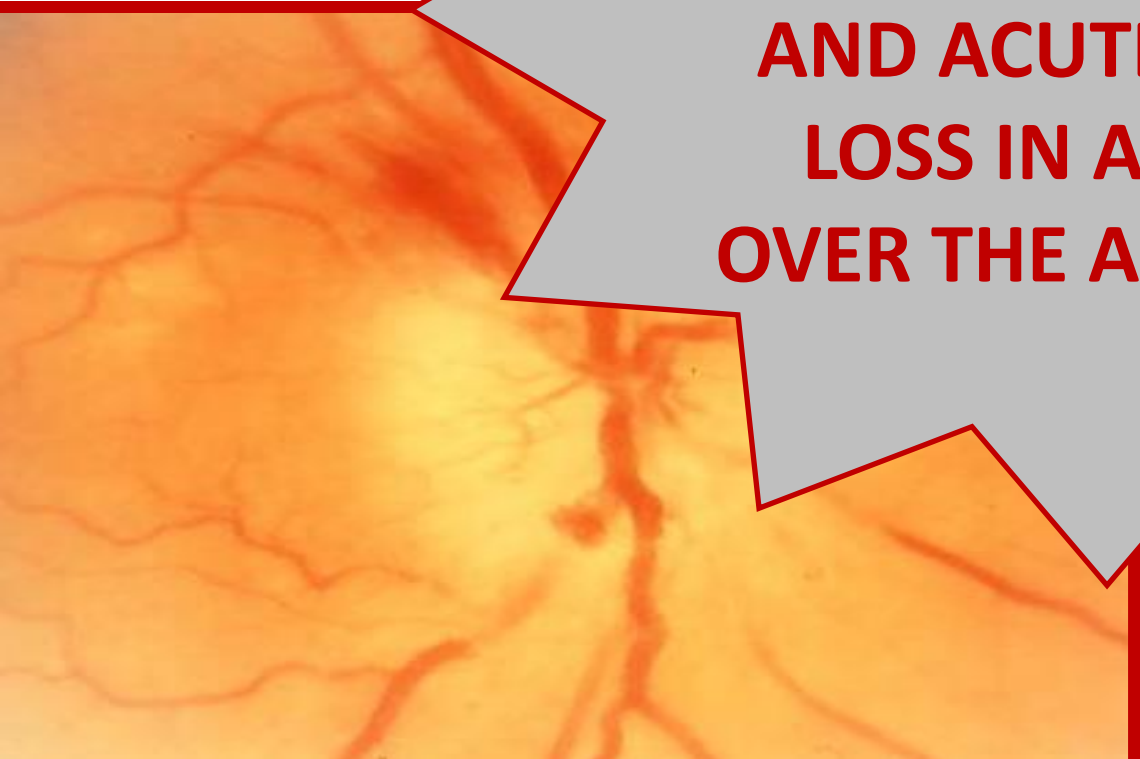


AAION

- Diffuse edema
- Pallid
- Hemorrhage
- 79% painful
- Premonitory TMB
- Nadir: minutes
- Fellow disc: anything

- Segmental edema
- Hemorrhage
- Luxury perfusion
- NRR pink
- No pain
- Nadir: 3.4 days
- Fellow eye: disc

NEED TO R/O GIANT CELL ARTERITIS WITH ANY DISC EDEMA AND ACUTE VISION LOSS IN ANYONE OVER THE AGE OF 50!



dema

morrhage

% painful

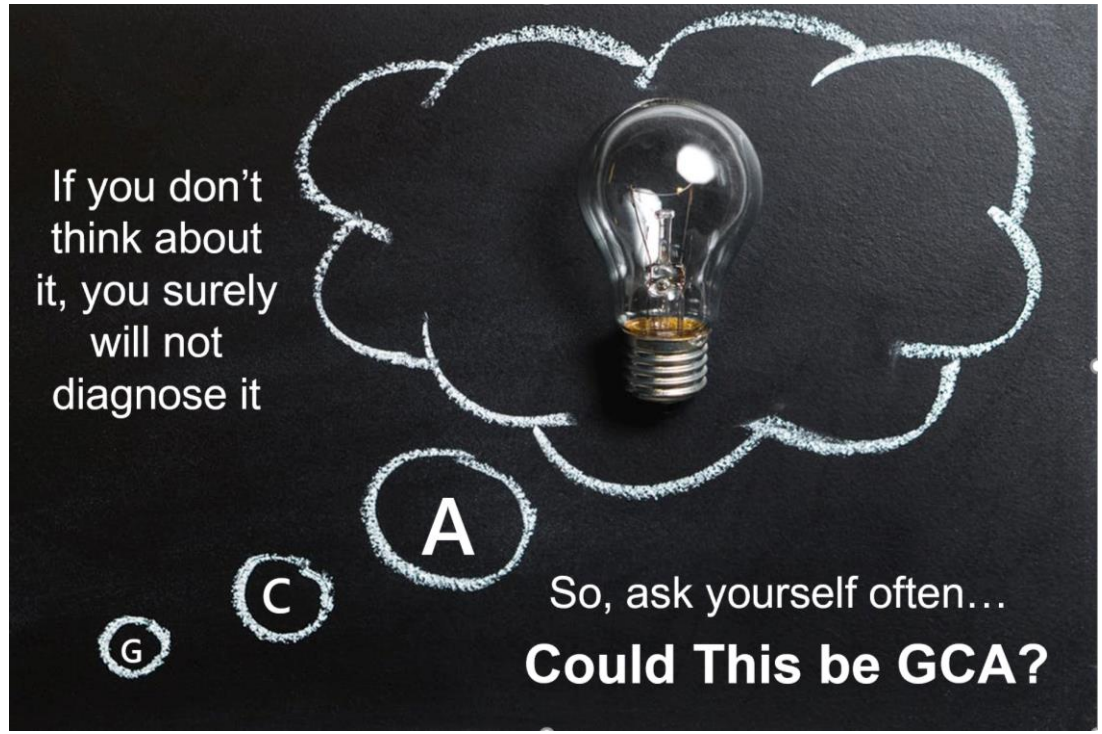
- Premonitory TMB
- Nadir: minutes
- Fellow disc: anything

GIANT CELL ARTERITIS

- **We need to keep up with the literature**
- Can cause devastating vision loss, stroke, death!
- DO NOT MISS THIS DIAGNOSIS!!!!
  - SAVE LIVES!
  - SAVE VISION!
  - STAY OUT OF COURT!

- GCA accounts for 14% of all Patients with permanent blindness (71% AAION)

- 94% of blindness occurs <u>prior to</u> treatment



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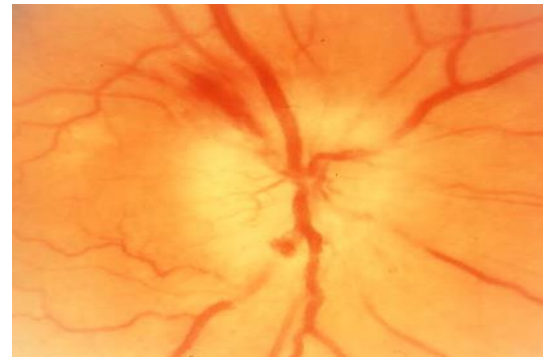
GCA ≠ AAION



- NOT every person with GCA gets AAION, or has visual involvement
- AAION is NOT the only ocular / visual manifestation of GCA
  - Retinal artery occlusion, Homonymous hemianopia, **transient vision loss**
- But… every AAION IS caused by GCA.
- GCA can also effect the efferent visual system (diplopia)

- **GCA NEEDS to be considered and ruled out in EVERY new onset neurologic deficit in anyone over the age of 50!**
  - (any abnormality with Neurologic Exam, including CN testing (II, III, IV, VI).

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GCA – What's New?

- 1. What's new in DIAGNOSIS?
- 2. What's new in TREATMENT?
- 3. What's new in TIMING?

Eye (2020) 34:1013–1026
https://doi.org/10.1038/s41433-019-0608-7

The ROYAL COLLEGE of OPHTHALMOLOGISTS

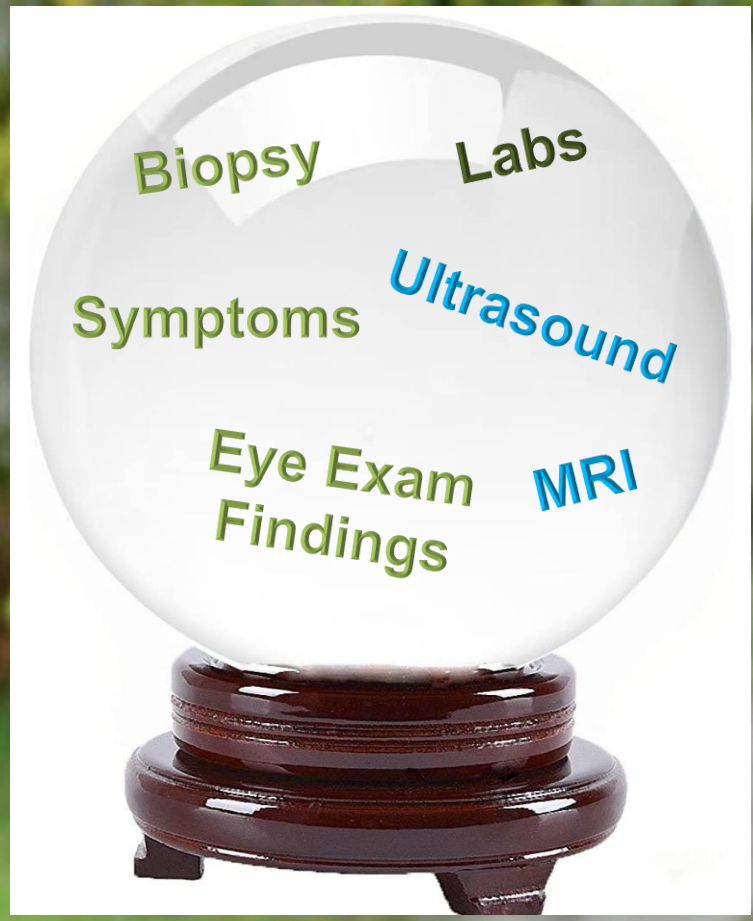
REVIEW ARTICLE

A new era for giant cell arteritis

H. S. Lyons[1] · V. Quick[2] · A. J. Sinclair[3,4] · S. Nagaraju[5] · S. P. Mollan[3]

Received: 17 September 2018 / Revised: 8 April 2019 / Accepted: 10 April 2019 / Published online: 3 October 2019

Who has GCA?



… anyone over the age of 50

GCA INITIAL WORK-UP

OLD

- Labs
  - CBC
  - ESR
  - CRP
- Temporal Artery Evaluation
  - Biopsy

NEW

- Labs
  - CBC
  - ESR
  - CRP
  - Comprehensive metabolic panel
    - Liver function, glucose
- Temporal Artery Evaluation
  - **Color Doppler US** or
  - Biopsy

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CBC, ESR & CRP

- Still important first line tests!

Laboratory data aid in the evaluation of GCA and its differential diagnosis, but they are not specific and cannot be relied on as definitive evidence for or against a possible diagnosis of GCA.

Why Comprehensive Metabolic Panel ?

- Liver Function
  - Up to 20% of GCA patients can have elevated liver enzymes
- Glucose
  - Because steroid treatment can elevate glucose level

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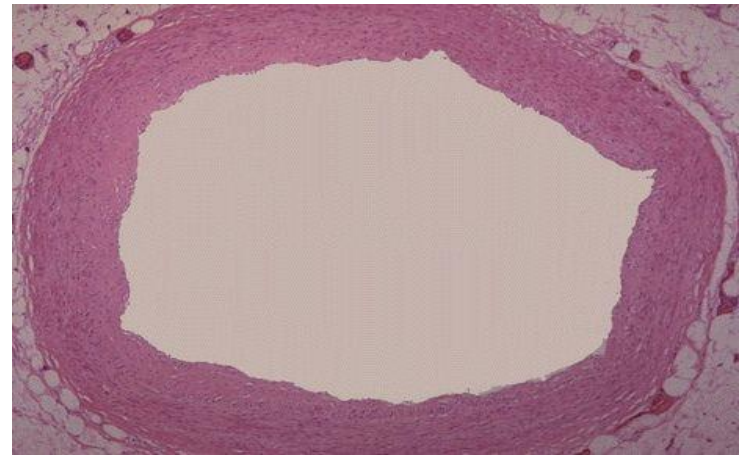
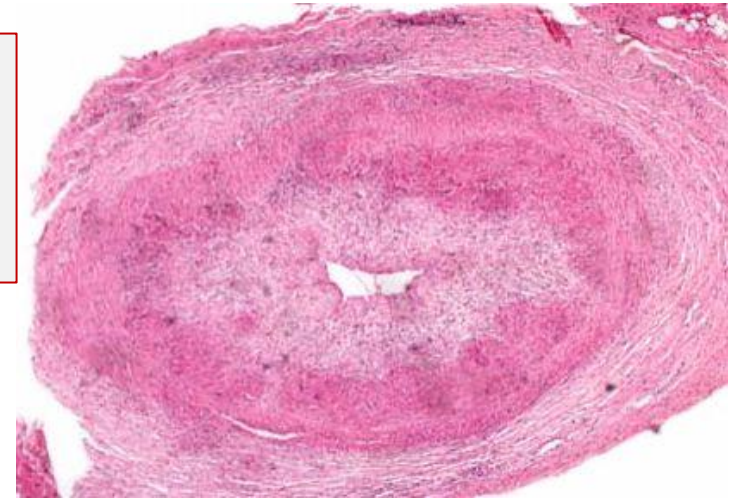
GCA is a VASCULITIS

- **Attacks medium and large sized arteries**
- **Specifically targets arteries with ILM**
  - Elastin likely is inciting antigen (auto-immune)
  - Cytokine production
    - IL-1, **IL-6**, TNF-a
  - Destroys/fragments the elastic lamina
  - Infiltrates the vessel wall with giant cells
  - Causes intimal thickening
- Thrombosis and Occlusion

Any vasculitis can cause stroke and vision loss



GCA does not only affect the temporal artery (that is just easy to biopsy).



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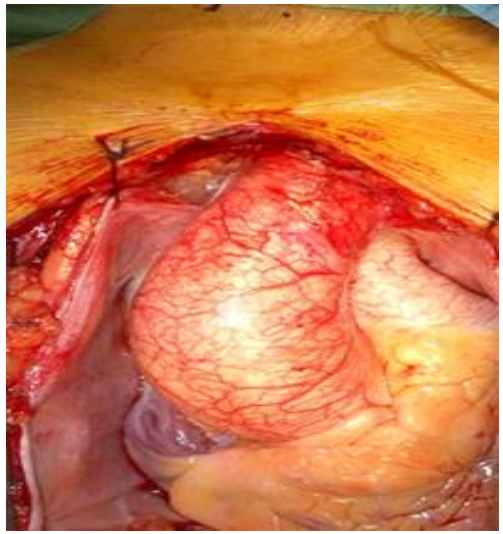
GCA and aortic involvement

Large vessel involvement in GCA



- The initial evaluation of GCA should include baseline and annual abdominal ultrasound, chest radiography (or more sensitive CT / MRI) and transthoracic echocardiography.
- **Patients with GCA need long term cardiology follow-up** since catastrophic aortic complications can occur years after the typical GCA symptoms resolve, even in the setting of a normalized ESR and C-reactive protein.
- **GCA patients have increased risk of aortic aneurysm**, being 17.3 and 2.4 times more likely to develop a thoracic (TAA) or abdominal aneurysm (AAA), respectively.
- GCA patients often have no acute symptoms of the aneurysm; therefore the mean time to detection is > 1 year after GCA diagnosis.

Unexplained chest pain in a patient with giant cell arteritis? Think aortic dissection

Joshua Abishek[1], Richard Akintayo[2] and Chris Isles[3]

Journal of the Royal College of Physicians of Edinburgh
2022, Vol. 52(2) 124–127

DOI: 10.1177/14782715221103685
journals.sagepub.com/home/rcp
SAGE

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Curr Rheumatol Rep
https://doi.org/10.1007/s11926-022-01086-2

VASCULITIS (C DEJACO AND C DUFTNER, SECTION EDITORS)

Spectrum of Large and Medium Vessel Vasculitis in Adults: Primary Vasculitides, Arthritides, Connective Tissue, and Fibroinflammatory Diseases

Luca Seitz[1,2] · Pascal Seitz[1] · Roxana Pop[3] · Fabian Lötscher[1]

Accepted: 26 July 2022

We can look at many of these vessels with color Doppler ultrasound.

Mainly affected large and medium vessels or vessel beds (i)

Large	Medium
‡ Aorta and all direct large branches, supraaortal (subclavian, axillary, brachial) > lower extremities (iliac, femoral, popliteal) [8]	‡ Head/neck (any artery, esp. TA, facial, occipital, ophthalmic) +/- Mesenteric, renal, lateral/internal thoracic, infrabrachial, infrapopliteal, coronary, cerebral, ovarian, uterine, breast, skin, liver, testes [9–13••]

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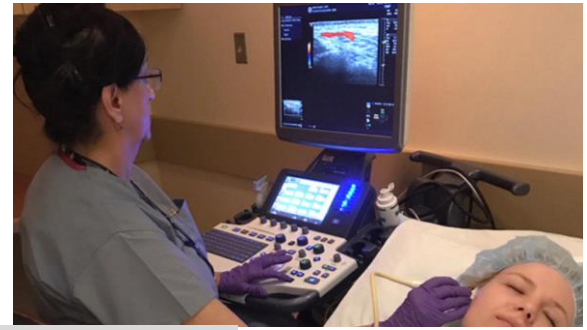
COLOR DOPPLER ULTRASOUND (CDUS)

- CDUS of the head, neck, and upper extremities can serve as a diagnostic surrogate for temporal artery biopsy when performed by **clinicians skilled in this technique.**

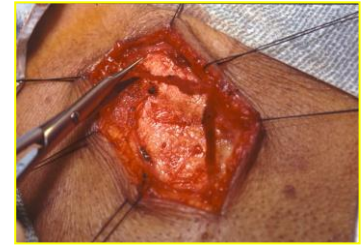
Dependent on equipment, technique, and operator. (as with all ultrasounds!)



Has a resolution of 0.1 mm

Along with lab testing, this is **a newer FIRST-LINE test for GCA**.

- In the absence of extensive experience with this technology in routine clinical practice, however, **temporal artery biopsy remains an essential diagnostic measure** for the evaluation of suspected GCA.



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RHEUMATOLOGY

Rheumatology 2018;57:ii22–ii31
doi:10.1093/rheumatology/kex461

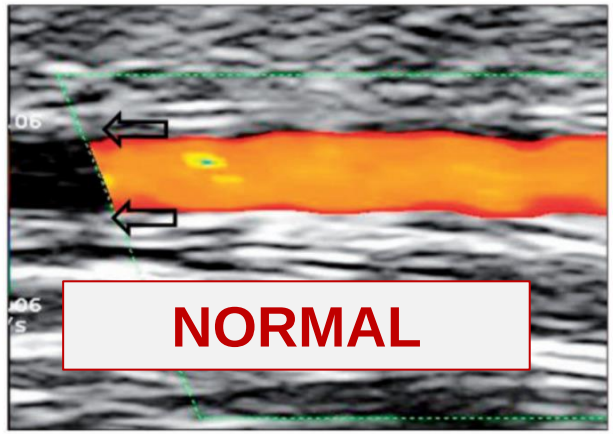
Should be performed ASAP
Before or early after initiation of steroids
- Ideally within 1 week

Ultrasound in the diagnosis and management of giant cell arteritis

Wolfgang A. Schmidt[1]

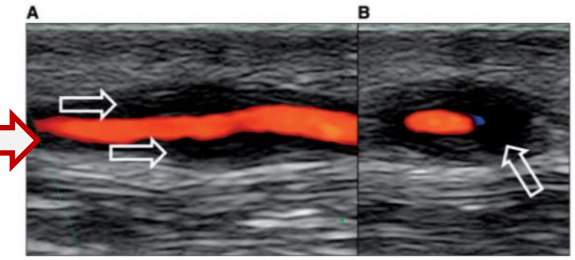
Treatment with glucocorticoids rapidly reduces the sensitivity of US for GCA diagnosis

FIG. 1 Normal intima–media complex of a temporal artery parietal branch (22-MHz probe)



HALO SIGN:
hypoechoic wall thickening

FIG. 2 Halo sign of temporal artery frontal branch

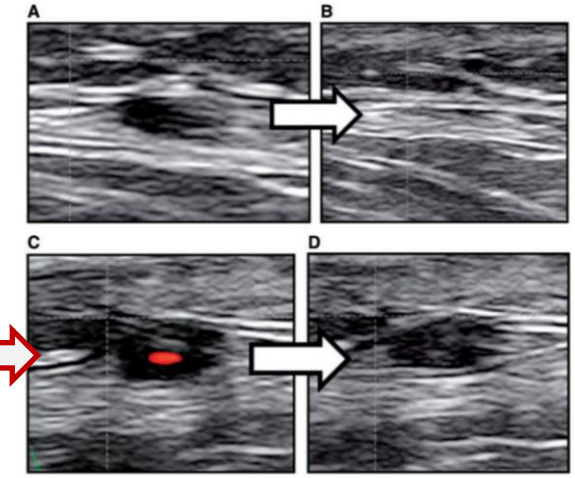


(A) Longitudinal; (B) transverse.

The presence of bilateral halo signs of the temporal arteries is highly specific for GCA

COMPRESSION SIGN:
visibility of the TA upon transducer-imposed compression of the artery.

FIG. 3 Compression sign of temporal artery branches



(A and B), normal; (C and D) abnormal.

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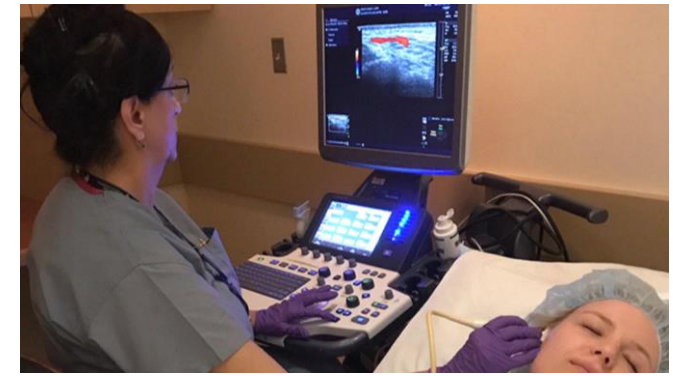
Color Doppler Ultrasound

Advantages

- Non-invasive
- Immediate interpretation
- No radiation exposure
- Can be used on various arteries
  - Medium vessel vasculitis
    - Temporal, cranial
  - Large vessel vasculitis
    - Axiliary, subclavian

Disadvantages

- Experience
- Interpretation
- What constitutes positive result? (cut-off)



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RHEUMATOLOGY ADVANCES IN PRACTICE

Rheumatology Advances in Practice 2021;00:1–9
https://doi.org/10.1093/rap/rkab083
Advance Access Publication 10 November 2021

Original article

Colour Doppler ultrasound and the giant cell arteritis probability score for the diagnosis of giant cell arteritis: a Canadian single-centre experience

Farah Zarka [1], Maxime Rhéaume[1], Meriem Belhocine[1], Michelle Goulet[1], Guillaume Febrer[2], Anne-Marie Mansour[1], Yves Troyanov[3], Tara Starnino[3], Rosalie-Sélène Meunier[1], Isabelle Chagnon[1], Nathalie Routhier[1], Valérie Bénard[1], Stéphanie Ducharme-Bénard[1], Carolyn Ross [1] and Jean-Paul Makhzoum [1]

Abstract

Objectives. The aim was to compare the accuracy of colour Doppler ultrasonography (CDUS) and temporal artery biopsy (TAB) to establish the final diagnosis of GCA and to determine how the GCA probability score (GCAPS) performs as a risk stratification tool.

Methods. Descriptive statistics were performed on a retrospective cohort of patients referred to our vasculitis referral centre between 1 July 2017 and 1 October 2020 for suspected GCA. CDUS, TAB, centre-specific TAB (vasculitis centre *vs* referring hospitals) and GCAPS were compared against the final diagnosis of GCA as determined by a GCA expert; CDUS was also compared with TAB results.

Results. Data from 198 patients were included: 60 patients with GCA and 138 patients without GCA. Sixty-two patients had a TAB. Using the final diagnosis by a GCA expert as a reference, the sensitivity, specificity, positive predictive value and negative predictive value were 93.3%, 98.5%, 96.6% and 97.1% for CDUS and 69.2%, 100%, 100% and 81.8% for TAB, respectively. The false-negative rate was 6.7% for CDUS and 30.8% for TAB. False-negative TAB mostly occurred when performed in referring hospitals (57.1%) as opposed to our vasculitis centre (21.1%). With a cut-off at 9.5 points, sensitivity for GCAPS was 98.3% and specificity 74.3%.

Conclusion. CDUS of the temporal and axillary arteries showed a high sensitivity and specificity and helped to diagnose GCA in patients with negative TAB. We validated that GCAPS is a useful clinical tool, with a score of <9.5 making the diagnosis of GCA improbable.

Patients with cranial manifestations
typically have a positive temporal artery biopsy OR positive CDUS

Key messages

- Colour Doppler ultrasonography is a highly effective tool when performed by a skilled ultrasonographer.
- Colour Doppler ultrasonography has a better sensitivity than temporal artery biopsy for the diagnosis of GCA.

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TREATMENT OF GCA

OLD

- Visual Involvement: **PROTECT OTHER EYE**
  - STAT IV Methylprednisolone
  - (Solumedrol 250mg IV q 6 hrs x 3 days)
  - Followed by Oral Prednisone
- No Visual Involvement: **PROTECT VISION**
  - Oral Prednisone (high dose 60-80 mg daily)

Steroids are still first line treatment, especially in cases of vision loss.

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Treatment Approved Specifically for GCA

NEW

- Tocilizumab
  - A recombinant monoclonal antibody directed against interleukin-6 receptors (IL-6R).
  - Previously used to treat moderate to severe rheumatoid arthritis (IV)
  - **It was initially FDA approved as a sub-cutaneous treatment for GCA**

First FDA approved therapy specific to GCA (May 2017)

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RHEUMATOLOGY

Rheumatology 2022;61:2915–2922
https://doi.org/10.1093/rheumatology/keab780
Advance Access publication 29 October 2021

Original article

New-onset versus relapsing giant cell arteritis treated with tocilizumab: 3-year results from a randomized controlled trial and extension

John H. Stone [1], Helen Spotswood[2], Sebastian H. Unizony[1], Martin Aringer [3], Daniel Blockmans[4], Elisabeth Brouwer[5], Maria C. Cid[6], Bhaskar Dasgupta[7], Juergen Rech[8], Carlo Salvarani[9], Robert Spiera[10] and Min Bao[11]

Remission:
- absence of symptoms
- normalization of labs

Patients on tocilizumab
- used much lower doses of prednisone

GiACTA Trial

July 27, 2017
N Engl J Med 2017; 377:317-328
DOI: 10.1056/NEJMoa1613849

Tocilizumab, received weekly or every other week, **combined with a 26-week prednisone taper** was superior to either 26-week or 52-week prednisone tapering plus placebo with regard to **sustained glucocorticoid-free remission** in patients with giant-cell arteritis.

Rheumatology key messages

- Tocilizumab delayed time to first flare and reduced glucocorticoid exposure in new-onset and relapsing GCA.
- Tocilizumab dosed weekly delayed time to first flare to a greater degree than every-other-week dosing.

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Contents lists available at ScienceDirect

Journal of Autoimmunity

journal homepage: www.elsevier.com/locate/jautimm

Divergent regulatory T cell responses to high-dose methylprednisolone and tocilizumab in giant cell arteritis

Godehard A. Scholz [a], Michaela Fux [b], Lisa Christ [a], Joseena Iype [c], Yara Banz [d], Peter M. Villiger [e,*]

[a] University Clinic of Rheumatology and Immunology, Inselspital, University Hospital Bern, Switzerland
[b] Institute of Social and Preventive Medicine, University of Bern, Switzerland
[c] University Institute of Clinical Chemistry, Inselspital, University Hospital Bern, Switzerland
[d] Institute of Pathology, University of Bern, Switzerland
[e] University of Bern and Medical Center Monbijou, Switzerland

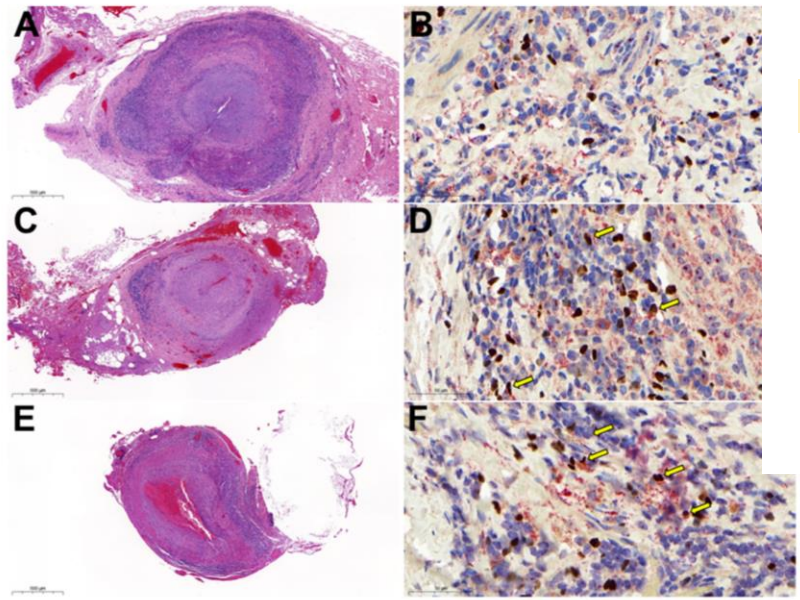

Fig. 2. Hematoxylin and Eosin staining images (left panels, 5x) of TABs taken at baseline (A and B), post methylprednisolone treatment (C and D) and post tocilizumab treatment (E and F). Immunohistochemical staining of formalin-fixed, paraffin-embedded samples (right panels, 63x). FOXP3 staining is denoted by a brown nuclear positivity. CD15s stain is shown in red. Double positive cells are highlighted with the yellow arrow.

Tocilizumab is not used as a monotherapy, but always in combination with steroids.

In a longitudinal approach, we present mechanistic insights into the respective roles of GCs and TCZ on Treg cells in GCA. We identified two complementary responses: 1) MP leads to a rapid systemic augmentation of CD15s+ activated Treg cells and thereby promotes their entry into the inflamed vessel wall. This finding is supported by the presence of CD15s+ Treg cells at sites of active inflammation in TABs. Thus, CD15s+ Treg cells appear to be important players when active inflammation needs to be controlled. 2) TCZ does not interfere with CD15s+ Treg cells, but, as recently shown, restores the suppressive function of Treg cells by augmenting the expression of FOXP3 EXON2 [3]. The prevention of Treg cells to polarize into IL-17 producing effector T cells will likely result in a delayed clinical response. Indeed, time kinetics of clinical response to MP and to TCZ, respectively, were different, they perfectly match cellular findings [2]. Collectively, our data might provide the basis for next-generation therapies. A future real life scenario could be the adoptive transfer of previously *ex vivo* sorted or genetically manufactured Treg cells highly expressing CD15s and/or FOXP3 EXON2, thereby mimicking MP and TCZ effects.

Methylprednisolone and tocilizumab cause different T-cell responses

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Tocilizumab is recommended in all cases of GCA with visual involvement.

THERAPEUTIC ADVANCES in
Musculoskeletal Disease

Original Research

Tocilizumab in visual involvement of giant cell arteritis: a multicenter study of 471 patients

Ther Adv Musculoskelet Dis
2022, Vol. 14: 1–16
DOI: 10.1177/1759720X221113747

Javier Loricera*, Santos Castañeda*, Clara Moriano*, Javier Narváez,

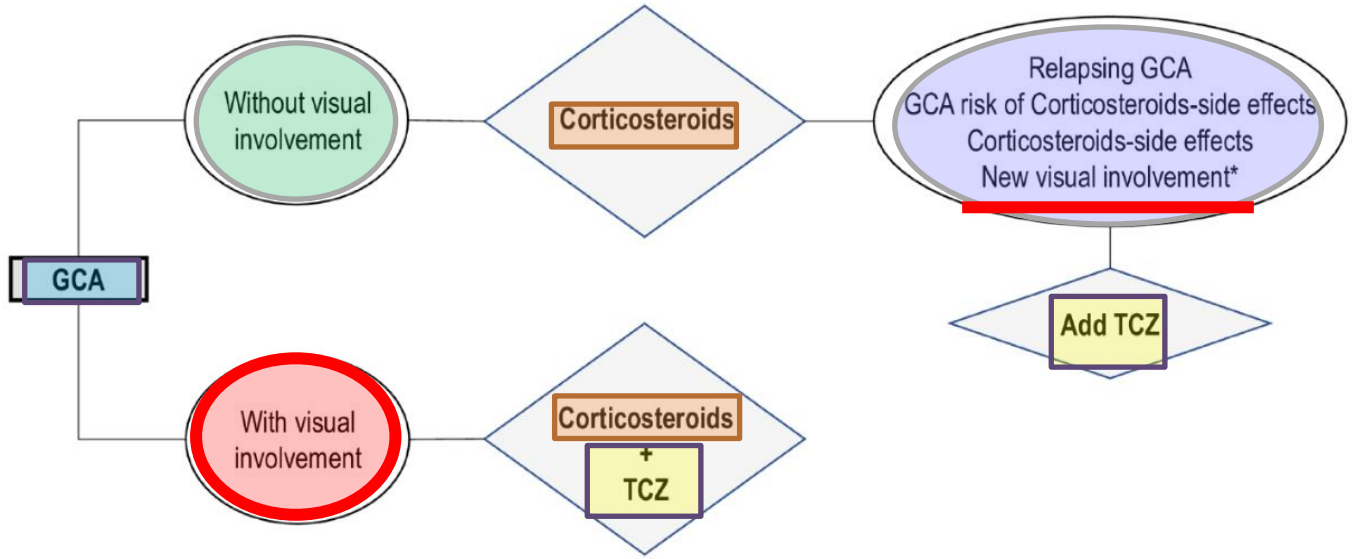

Figure 3. Proposed algorithm for the use of tocilizumab in giant cell arteritis with and without visual involvement.
GCA, giant cell arteritis; TCZ, tocilizumab.
*If new visual involvement, in addition to additional tocilizumab we recommend increased glucocorticoids dose.

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J Neuroophthalmol. 2022 Jun 1;42(2):173-179. doi: 10.1097/WNO.0000000000001514.
Epub 2022 Apr 27.

Does Tocilizumab Influence Ophthalmic Outcomes in Giant Cell Arteritis?

Marc A Bouffard [1], Sashank Prasad, Sebastian Unizony, Fiona Costello

Affiliations + expand

PMID: 35482901 DOI:

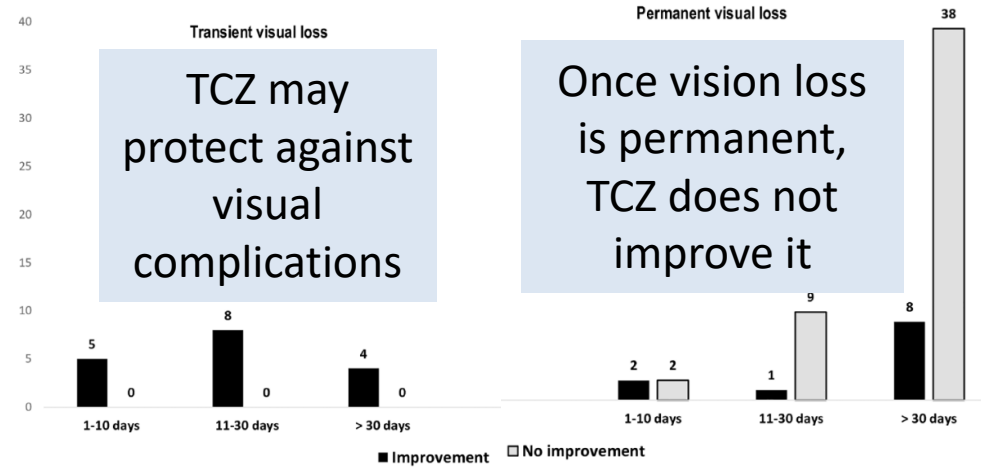
Conclusions: Compared with conventional corticosteroid monotherapy, treatment of GCA with both corticosteroids and tocilizumab may decrease the likelihood of permanent vision loss. Further prospective, collaborative investigation between rheumatologists and neuro-ophthalmologists is required to clarify the ophthalmic and socioeconomic impact of tocilizumab on the treatment of GCA.

THERAPEUTIC ADVANCES in *Musculoskeletal Disease* — *Original Research*

Tocilizumab in visual involvement of giant cell arteritis: a multicenter study of 471 patients

Javier Loricera*, Santos Castañeda*, Clara Moriano*, Javier Narváez, Vicente Aldasoro, Olga Maiz, Rafael Melero, Ignacio Villa, Paloma Vela, Susana Romero-Yuste, José L. Callejas, Eugenio de Miguel, Eva Galíndez-Agirregoikoa, Francisca Sivera, Jesús C. Fernández-López, Carles Galisteo, Iván Ferraz-Amaro, Julio Sánchez-Martín, Lara Sánchez-Bilbao, Mónica Calderón-Goercke, Alfonso Casado, José L. Hernández†, Miguel A. González-Gay† and Ricardo Blanco†; on behalf of the Tocilizumab in Giant Cell Arteritis Spanish Collaborative Group‡

Ther Adv Musculoskelet Dis
2022, Vol. 14: 1–16



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WHO FOLLOWS GCA PTS?

- Eye Doctor
- Rheumatologist
- Neurologist
- General Practitioner / PCP (BS)
- Cardiologist (REFER!!)

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SUMMARY – What's New in GCA?

- **If acute AAION or TVL, send patient to ED immediately for dx/tx**
- Otherwise, if GCA is in DDX for non-acute process, need urgent evaluation with labs and imaging
  - CBC, CRP, ESR, CMP (liver function)
  - **CDUS is now considered first line testing** (operator dependent!)
  - Biopsy is still used if CDUS is negative
  - Biopsy of other side can increase yield by 5-7%
- Steroids are still the first line treatment
- **Tocilizumab helps to reduce steroid use/complications and achieve remission**
  - Tocilizumab recommended in all cases with vision involvement
  - Once on tocilizumab, cannot use labs to monitor progress

AMAUROSIS FUGAX

- Transient monocular blindness
- Most often caused by ON ischemia, can be light-induced
- Present in 31-46% of GCA pts.
- Precedes permanent visual loss 50-63% of the time
- Prodrome to AAION

Even if vision is back to normal, still need to R/O GCA and stroke!

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CASE 5

55 YEAR OLD WOMAN

- **Complete vision loss of the right eye that she noticed upon waking** this morning around 1:00 am.
- She reported that the vision began to improve around 6:00 am at which point she could see shadows and lights.
- She denies any ocular pain or GCA symptoms.

- SYSTEMIC HISTORY
  - hypercholesterolemia for 2 years (not treated)
  - iron deficiency anemia
- OCULAR HISTORY
  - mild cataracts and primary open angle glaucoma bilaterally
- MEDICATIONS
  - iron, fish oil, vitamin B-12, multivitamin.
  - given medication in the past for hypercholesterolemia, did not use it because of side effects.
  - Her PCP apparently recently prescribed cholesterol mediation again, but the patient has not yet had it filled.

We need to consider GCA in any case of vision loss, transient or not.

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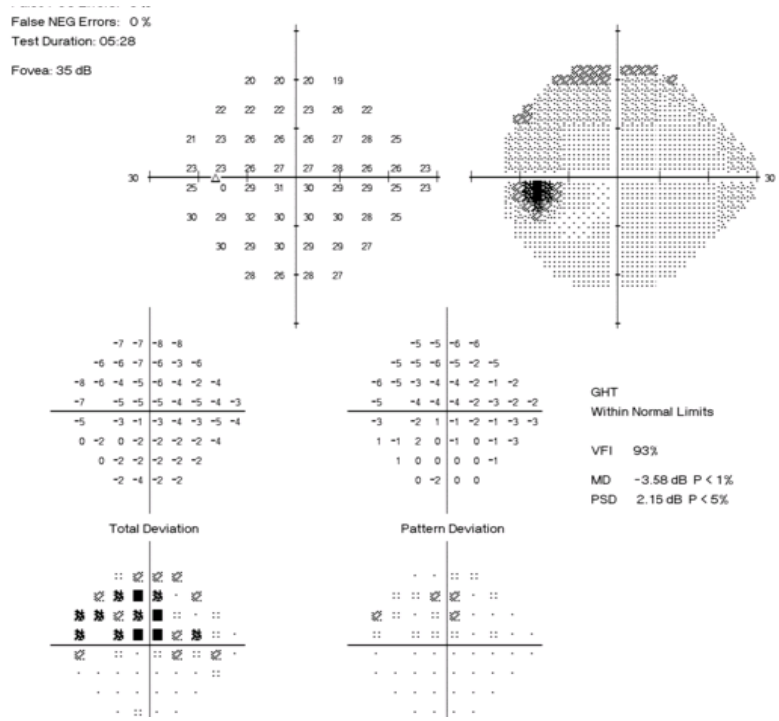
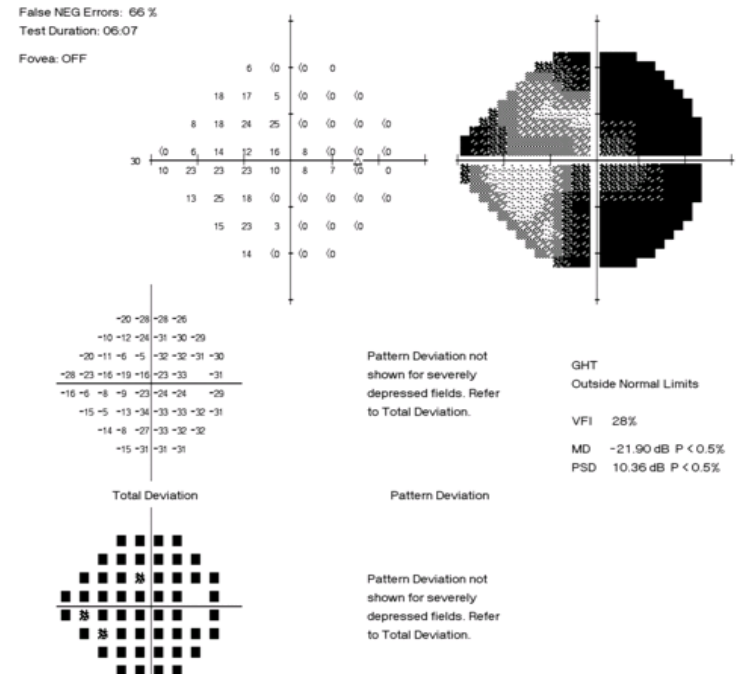
- BCVA: HM at 3 feet OD and 20/20- OS
- (+) RAPD OD >1.8 log
- CF: severe constriction OD, full OS
- Ocular motility testing: normal
- SLE: mild lens changes OU
- GAT: 14 mmHg OD and 13 mmHg OS

- BP: 150/70 RAS
- Pulse: 70 bpm (regular)





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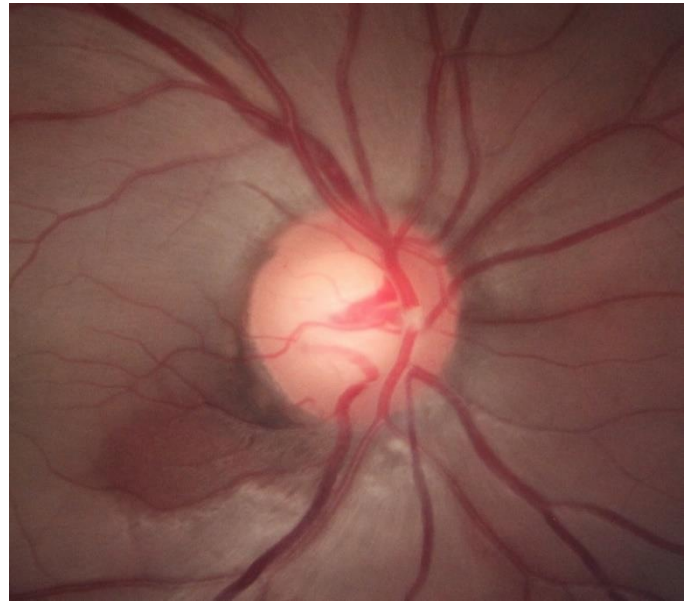

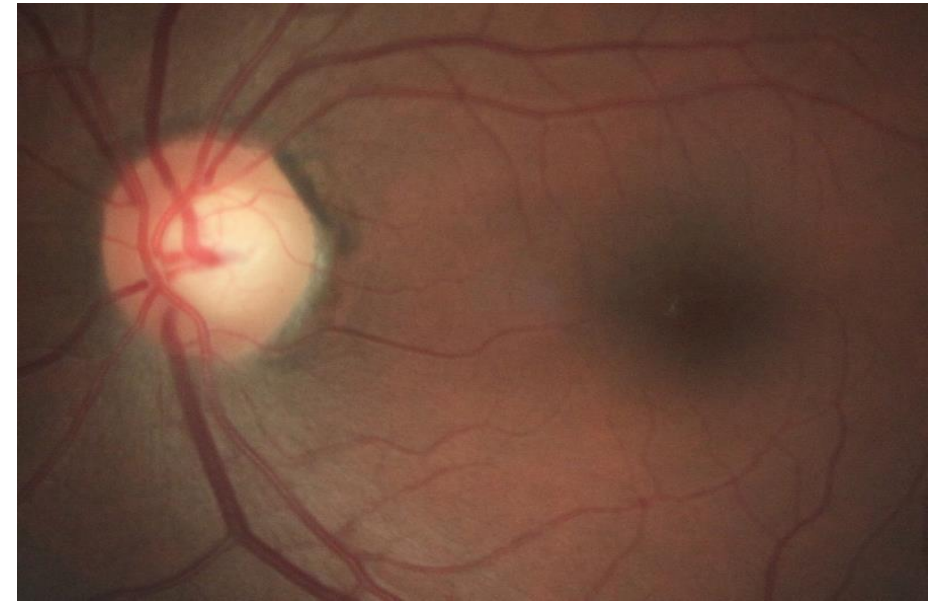

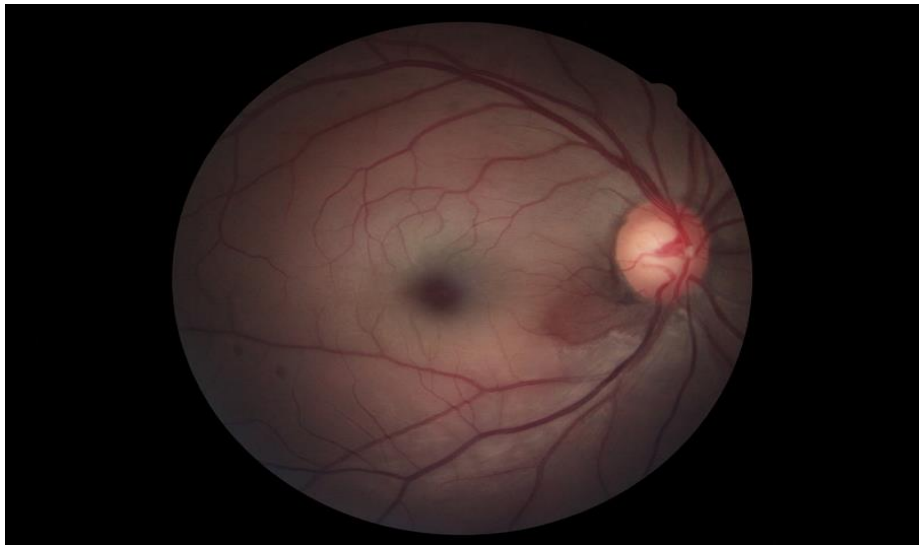

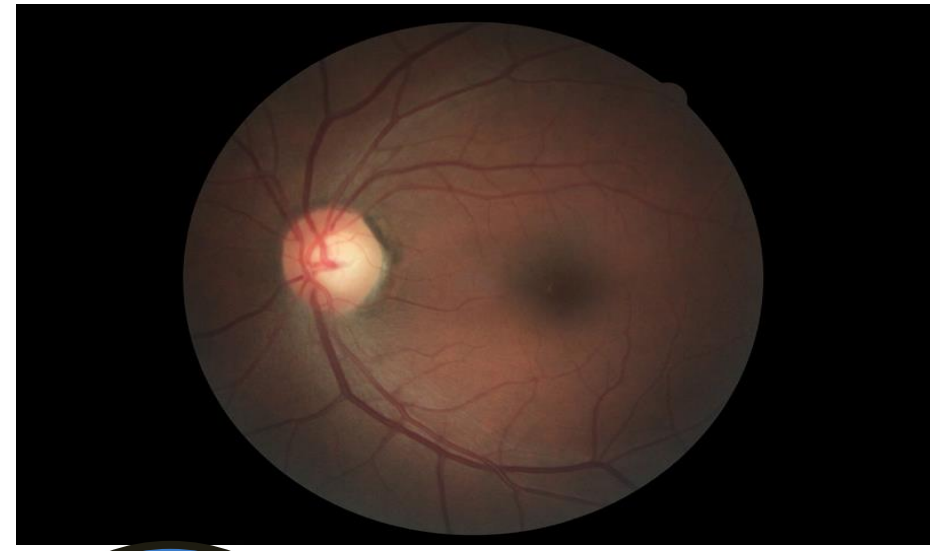

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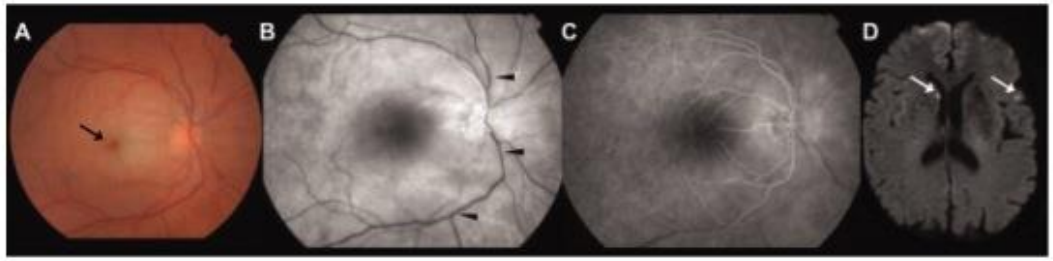
Retinal ischemia vs cerebral ischemia

Concurrent Acute Brain Infarcts in Patients with Monocular Visual Loss

Johanna Helenius, MD,[1] E. Murat Arsava, MD,[1] Joshua N. Goldstein, MD, PhD,[2]
Dean M. Cestari, MD,[3] Ferdinando S. Buonanno, MD,[4]
Bruce R. Rosen, MD, PhD,[1] and Hakan Ay, MD[1,4]

ANN NEUROL 2012;72:286–293



American Journal of Ophthalmology
June 2014; 157:1119-1121

EDITORIAL

Acute Retinal Arterial Ischemia: An Emergency Often Ignored

VALÉRIE BIOUSSE

- SAME MECHANISMS!
- SAME Guidelines for
  - CRAO
  - BRAO
  - Transient vision loss
  - & STROKE!!

AHA/ASA Guideline

Guidelines for the Prevention of Stroke in Patients With Stroke and Transient Ischemic Attack

A Guideline for Healthcare Professionals From the American Heart Association/American Stroke Association

Any patient with suspected TIA or those with acute retinal ischemia (including transient vision loss) should be evaluated urgently in order to identify those at high risk of immediate cerebral infarction and cardiac ischemia.

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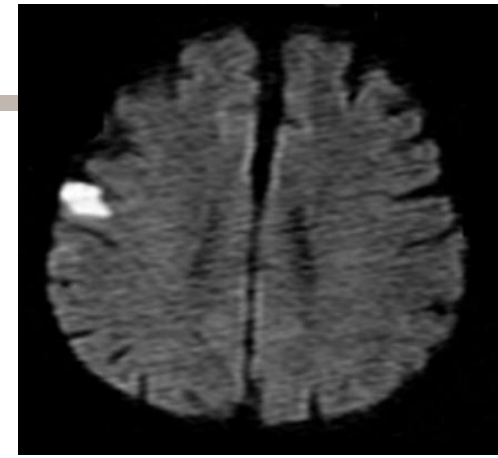
Any TIA or Retinal Ischemia/Emboli Treated the Same

Co-occurrence of Acute Retinal Artery Occlusion and Acute Ischemic Stroke: Diffusion-Weighted Magnetic Resonance Imaging Study

JUNWON LEE*, SEUNG WOO KIM*, SUNG CHUL LEE, OH WOONG KWON, YOUNG DAE KIM, AND SUK HO BYEON

Am J Ophthalmol 2014; 157: 1231-1238



¼ of patients with acute retinal ischemia (even if transient) had an acute brain infarction on brain DWI-MRI

10-15% of patients will have a disabling stroke within 3 months after a TIA, with half occurring within 48 hours after resolution of TIA.

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What needs to be done?

- DWI-MRI within 24-48 hours of vision loss
- Imaging (CTA) of cervical and intracranial vessels.
- EKG and echocardiogram
- Laboratory testing
  - CBC with platelets
  - Coagulation studies
  - Fasting lipid profile

How does this get done?

Do NOT send these patients to their PCP, cardiologist, neurologist, neuro-ophthalmologist, or retinal specialist.

Do NOT try to obtain the work-up yourself.

Send to an ED with an Acute Stroke Care Center!

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OUR PT NEEDS IMMEDIATE HOSPITALIZATION FOR STROKE WORK-UP

- High total cholesterol and high LDL
- Anemia
- mild stenosis of the proximal right internal carotid artery (no stroke on MRI DWI or ADC)
- TREATMENT: Crestor and aspirin 325 mg, as well as a multivitamin and iron.

Even if the patient said she lost her vision and it returned to normal, we would still need to send her to the hospital to rule out stroke.

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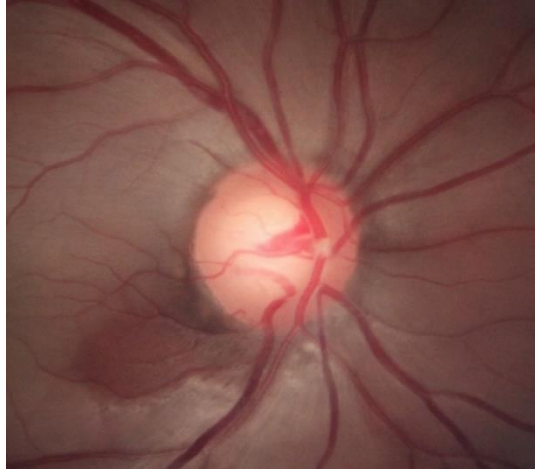
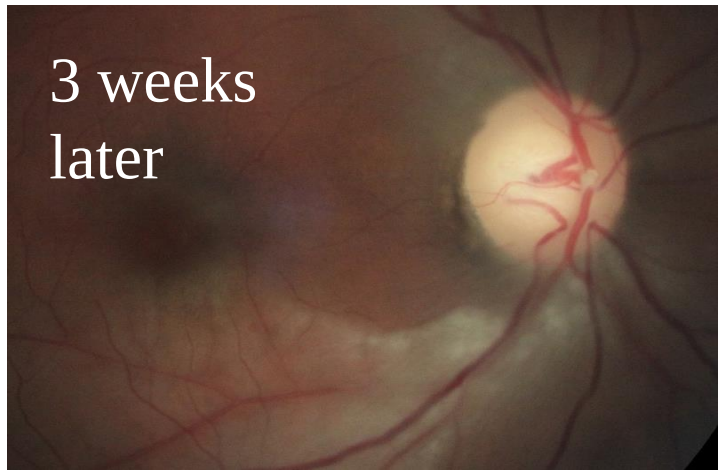
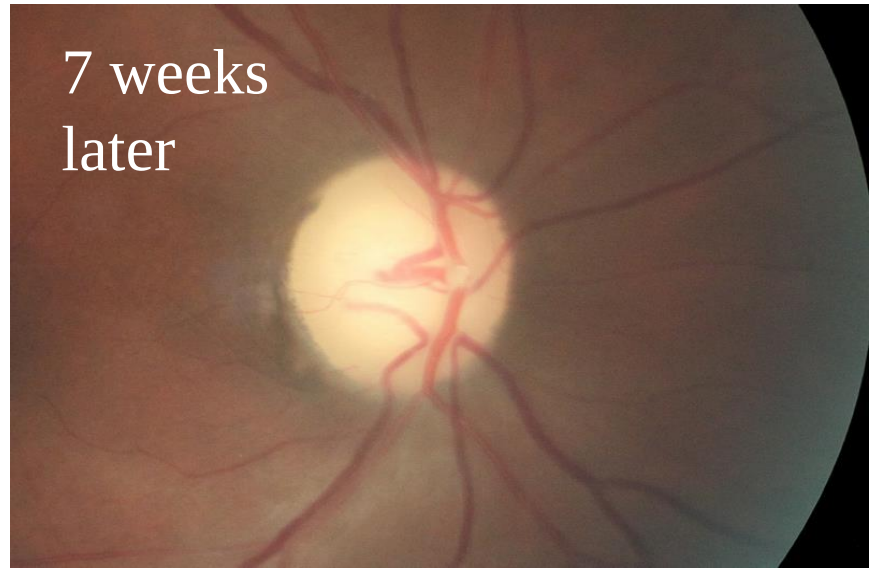
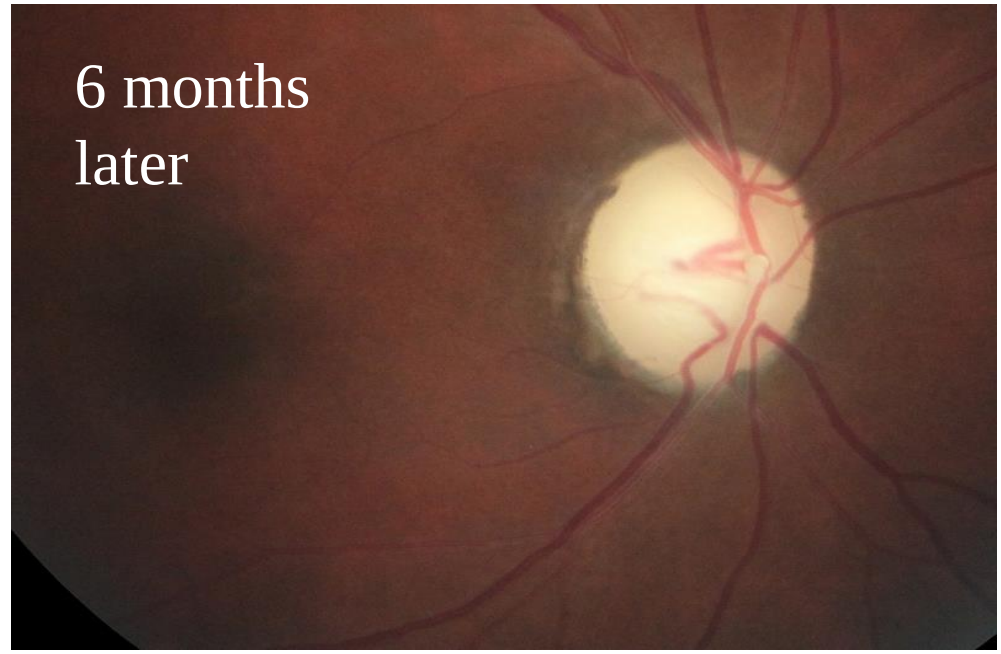



10 days
later


3 weeks
later


7 weeks
later


6 months
later

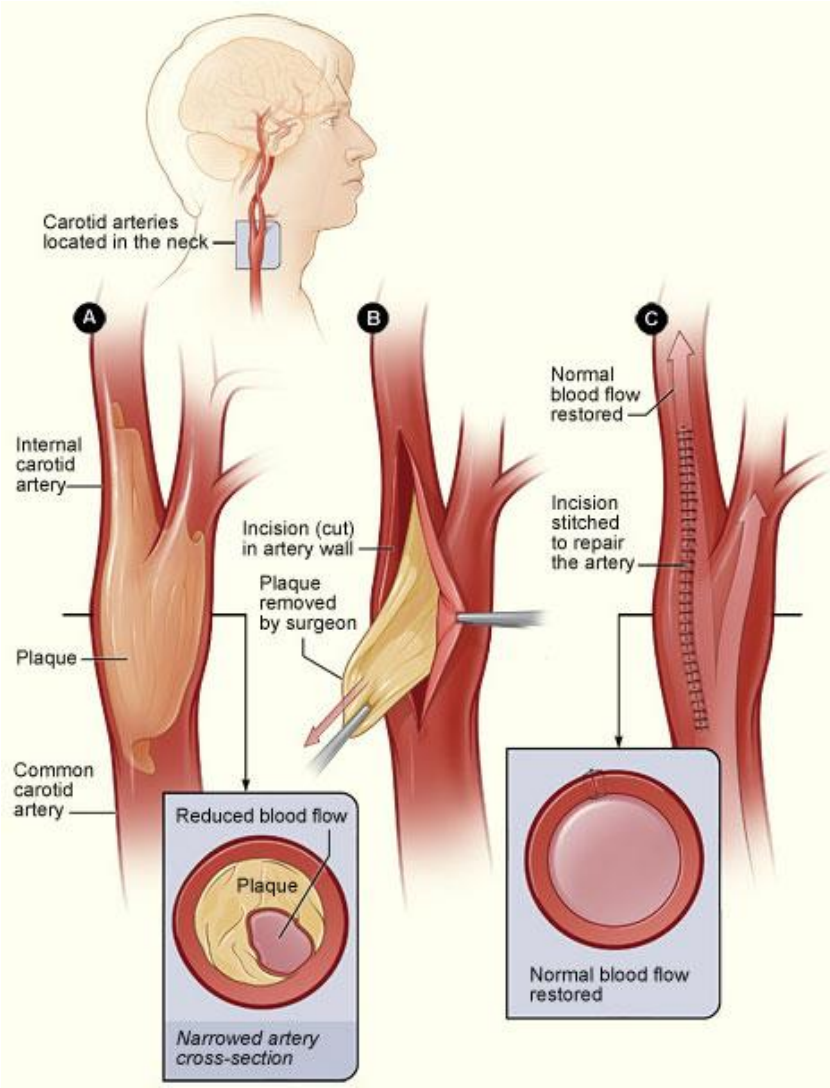
Most health care professionals consider retinal TIAs benign with a low risk of stroke.

The care of patients with retinal TIAs is often delayed.

Carotid stenosis surgery was delayed by 1-2 months when the symptom was a retinal TIA.

Carotid endarterectomy for symptomatic patients with carotid stenosis should be performed within 2 weeks (ideally within 2 days of the TIA).

CAROTID ENDARTERECTOMY VS STENT



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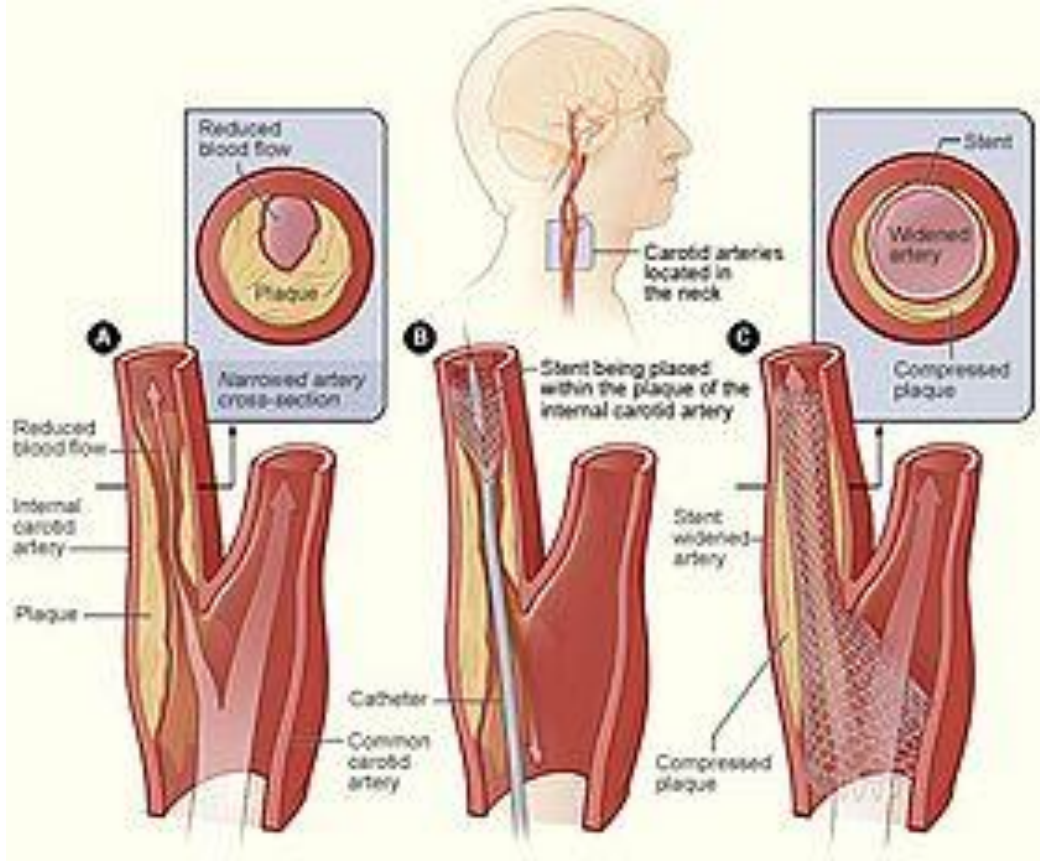

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NORTH AMERICAN SYMPTOMATIC CAROTID ENDARTERECTOMY TRIAL (NASCET)

N Eng J Med. 1991; 325: 445-453.

SYMPTOMATIC with STENOSIS > 70%

CAROTID ENDARTERECTOMY IS RECOMMENDED

SYMPTOMATIC with STENOSIS 50-70%

CAROTID ENDARTERECTOMY IS INDICATED

2.1% PERIOPERATIVE RISK OF *STROKE* AND *DEATH*

SURGICAL TX of ASYMPTOMATIC CAROTID STENOSIS IS CONTROVERSIAL

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Low stroke risk if 100% occluded

CASE 6

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68 year-old woman

- Shadow on left side of vision x 2 days
- Headache and flashes of light for 2 days
- 3 weeks ago, noted stiffness in left hand and foot

DO THESE SYMPTOMS LOCALIZE ANYWHERE?

- Diabetes x 20 years
- Hypertension x 20 years
- Hypercholesterolemia
- Kidney cancer 5 years ago
  - Left kidney removed
- Breast cancer 5 years ago
  - Right breast removed
  - Chemotherapy
- Defibrillator
- Glaucoma
  - Travatan

If we decide to get neuroimaging, does this patient have any contraindications to either imaging or use of contrast?

- The patient does drive
  - drove to office today

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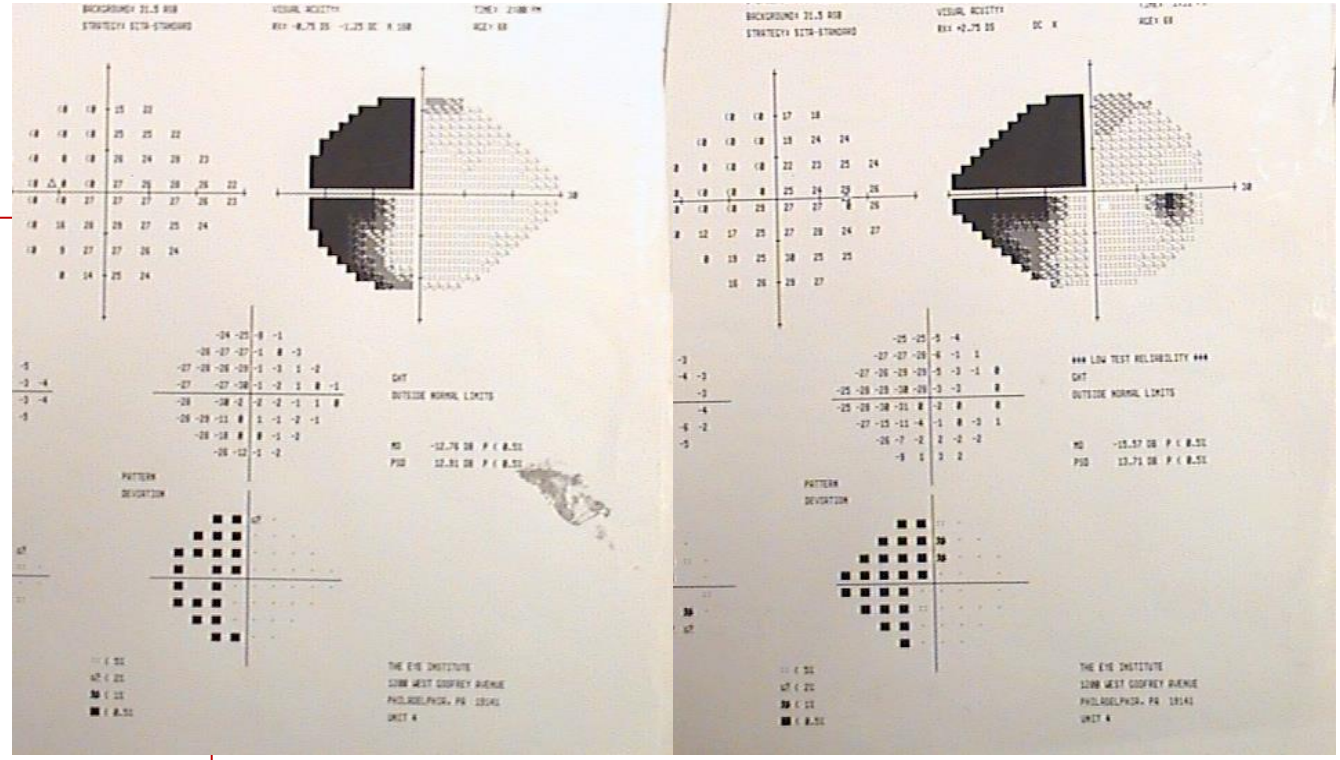

- VA: OD 20/25, OS 20/30
- Color: 14/14 OD 14/14 OS
- PERRLA (-) APD

- No ptosis or proptosis
- Normal ocular motility

- Slit lamp: cataracts OU
- IOP: OD 24 mm Hg , OS 21 mm Hg
- DFE: asymmetric cupping OD > OS, otherwise unremarkable OU

- BP: 136/86
- Pulse: normal/regular

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- Neurologic Exam:
  - ? Left-sided weakness
  - Difficulty with tandem gait
  - Difficulty with rapid alternating movements of the left hand

- **Admit to Hospital**
  - Rule out acute stroke vs. metastatic lesion
    - Right side of brain
    - Behind the optic chiasm
    - Appears congruous – so occipital lobe (due to neuro exam findings)
    - Also findings suggestive of cerebellum involvement
  - Will need to have CT, likely without contrast
  - MRI contraindicated

STROKE WAS CONFIRMED

- Visual Field defect persists
- Patient will no longer be able to drive
- In the aging population, we have to deal with assessing driving ability

KNOW THE DRIVING VISUAL REQUIREMENTS IN YOUR STATE

- Homonymous Hemianopia can improve as edema resolves
  - 84% have some amount of recovery at 6 months
  - **Allow 1 year to get to maximum recovery**
    - » Usually not enough recovery to regain driving ability in those not meeting visual driving requirements right after stroke

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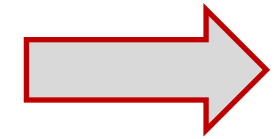
WHAT'S THE CAUSE OF THE OCCIPITAL LOBE LESION?

(TROBE et al 1983)

➢ <u>Age > 40 Years</u>:

- **INFARCT 89%**
- TUMOR 3%

Let's talk more about stroke



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Incidence of First-time Stroke

- > 800,000 per year
- Steadily increasing
- 1/5 die within a year of 1st stroke

80% of strokes are preventable (NSA, 2009)

Strokes can occur at any age, but are more common over the age of 65!

More women than men have strokes

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Role of Optometrists in Regard to Stroke

- Identify Acute Stroke / TIA
  - Get patient immediate (emergent) work-up/treatment as necessary
- Determine if Deficits are from old strokes or if they indicate a new stroke/other process
- Preventative Measures
  - Identify stroke risk factors
  - Educate patients
  - Refer as necessary
- Assess visual function after stroke
  - Improve visual function where possible
  - Comment on driving ability

https://www.stroke.org/en/help-and-support/resource-library/fast-materials/stroke-hero-poster

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ACUTE STROKE (EMERGENCY)

- Must assume that every acute stroke is hemorrhagic
- CT is done immediately to r/o bleeding
- Blood is seen better on CT than on MRI
- If no blood is noted, and stroke is not seen, MRI is done with diffusion weighted imaging (DWI) to identify acute ischemic infarct

If you suspect an acute stroke

Time is Brain!

(for BOTH ischemic and hemorrhagic stroke)

- Do not complete eye exam
- Only do what you need to confirm your stroke suspicion
- Do not let the patient drive to the hospital
- **Call 911 immediately**
- Tell the dispatcher the patient has had an acute stroke

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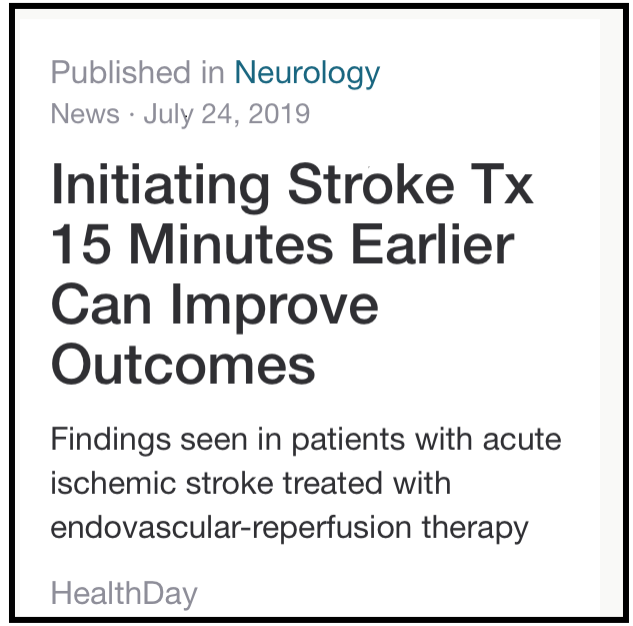
Time to brain death based on % normal blood flow

% NORMAL BLOOD FLOW	TIME TO BRAIN DEATH
NO flow	10 minutes
<30% normal flow	1 hour
30-40% normal flow	Hour to several hours
With collateral and residual flow	Up to 6 hours



Published in Neurology

News · July 24, 2019

Initiating Stroke Tx 15 Minutes Earlier Can Improve Outcomes

Findings seen in patients with acute ischemic stroke treated with endovascular-reperfusion therapy

HealthDay

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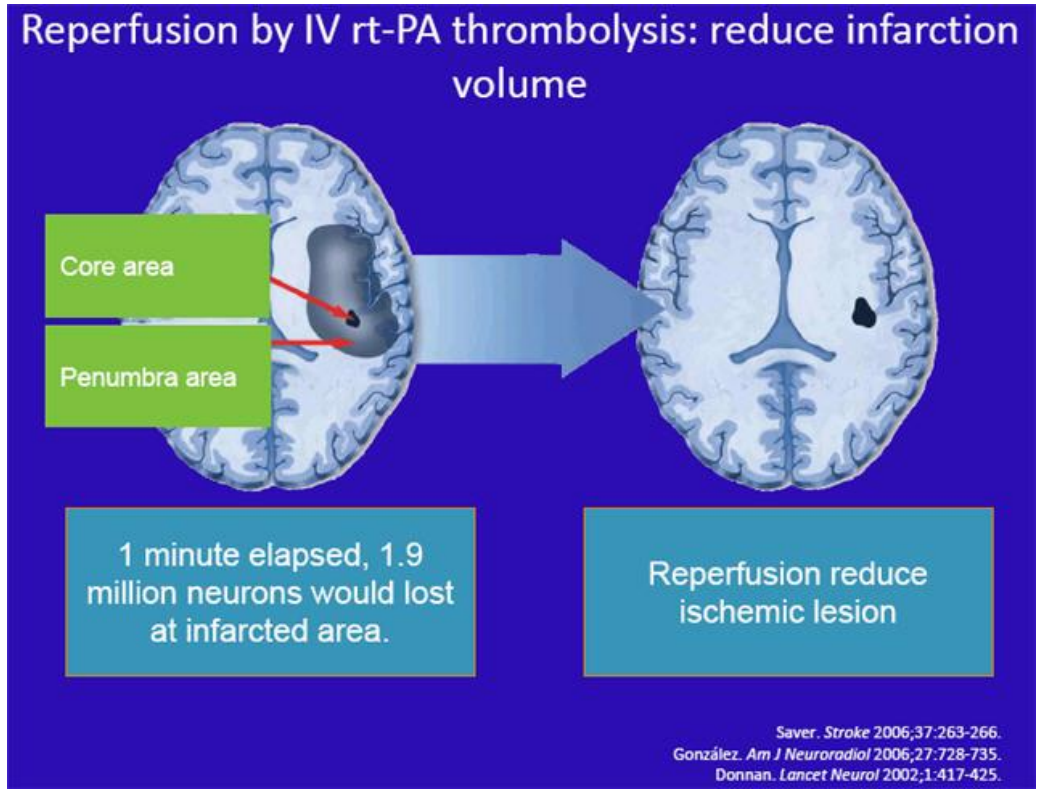
Tissue-Plasminogen Activator

t-PA (for acute ischemic stroke)

Effective in ¼ to 1/3 of patients

If patients are not candidates, the time frame has passed, or the treatment was ineffective, there is now another alternative treatment option…



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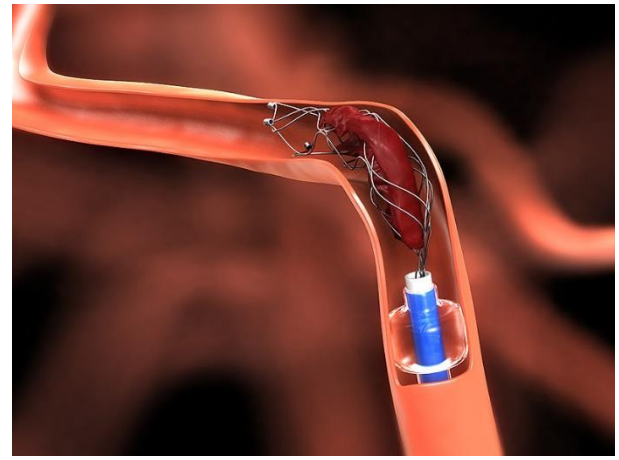
The Stent Retriever

Used primarily at advanced comprehensive stroke centers



- Tiny wire cage
- Threaded through a catheter into a blood vessel in the groin
- Guided up to the blocked artery (only for large vessel strokes) in the brain
- Cage opens up and captures the clot
- The stent, along with the clot, is removed
- Immediately blood begins flowing again to the brain

NEURO-OPHTHALMIC MANIFESTATIONS OF STROKE

- **ALMOST ALL AREAS OF THE BRAIN, WHEN DAMAGED, RESULT IN VISUAL MANIFESTATIONS**
  - Retina – CRAO, BRAO
  - Optic Nerve – Optic neuropathy
  - Post-chiasm– homonymous hemianopia
  - Cerebellum – ataxia, nystagmus
  - Brainstem – diplopia, skew deviation, INO, nystagmus
  - Cerebrum- supranuclear gaze palsy

REGARDLESS OF THE PRESENTATION, ANY ACUTE STROKE IS A MEDICAL EMERGENCY, AND PT SHOULD BE SENT TO THE HOSPITAL IMMEDIATELY!

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CASE 7

CASE: 63 year-old woman

- Sudden onset diplopia x 5 days
  - At distance and near
  - Horizontal and diagonal
  - Worse in right gaze
  - Resolves with covering either eye
- Headache 2 days ago
  - Above right eye, frontal

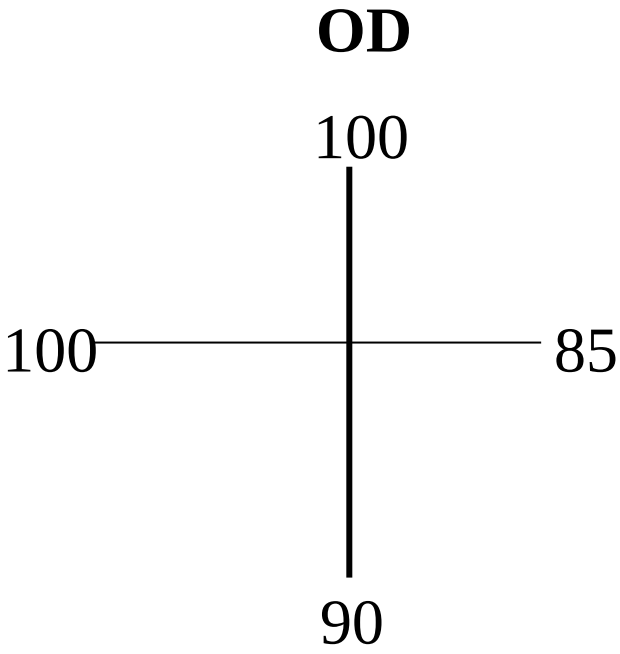
- **SYSTEMIC HEALTH**
  - Diabetes x 15 years
  - Hypertension x 15 years
  - Hypercholesterolemia
  - Arthritis
  - s/p stroke x 3 (last 5 years ago)
    - Residual weakness
  - Medications
    - Naprosen, Detrol, Minocycline, Enalapril, Nefedipine, Aggrenox, Alendronate, Metformin, and Pravastatin.

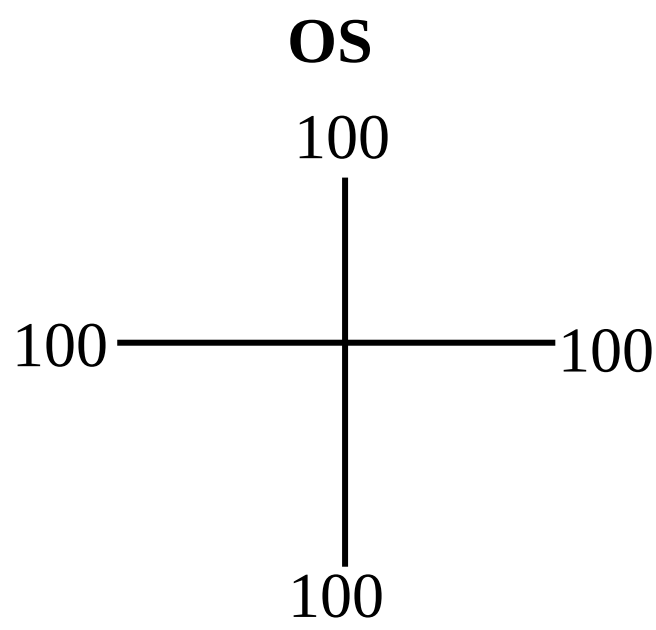
- **OCULAR HISTORY**
  - Cataracts
  - Glaucoma (longstanding)
    - s/p PI OU
    - Supposed to be on Cosopt and latanaprost
      - Ran out of meds yesterday

- **SOCIAL HISTORY**
  - Smokes 3-4 cigarettes / weekend x years
  - Few beers per weekend

- VA: 20/25 OD 20/30 OS
- Color 14/14 OD 14/14 OS
- (-) RAPD, anisocoric
- Bright: 3 OD, 2.75 OS dim: 4 OD, 3.75 OS
- CF: full OU
- Palpebral apertures: 7 mm OD 7 mm OS
- IOP: 20 mm Hg OD, 21 mm Hg OS
- DFE: Large cupping OU
- (-) edema OU , (-) pallor OU
- Neurologic examination
- BP: 178/94, pulse 50bpm

Ocular Motility





	4 RH 8 eso	
20 RH 10 eso	12 RH 10 eso	6 RH 10 exo
	6 RH 6 eso	

Head tilt testing demonstrated 16 right hyper on right head tilt, and 12 right hyper and 14 eso on left head tilt

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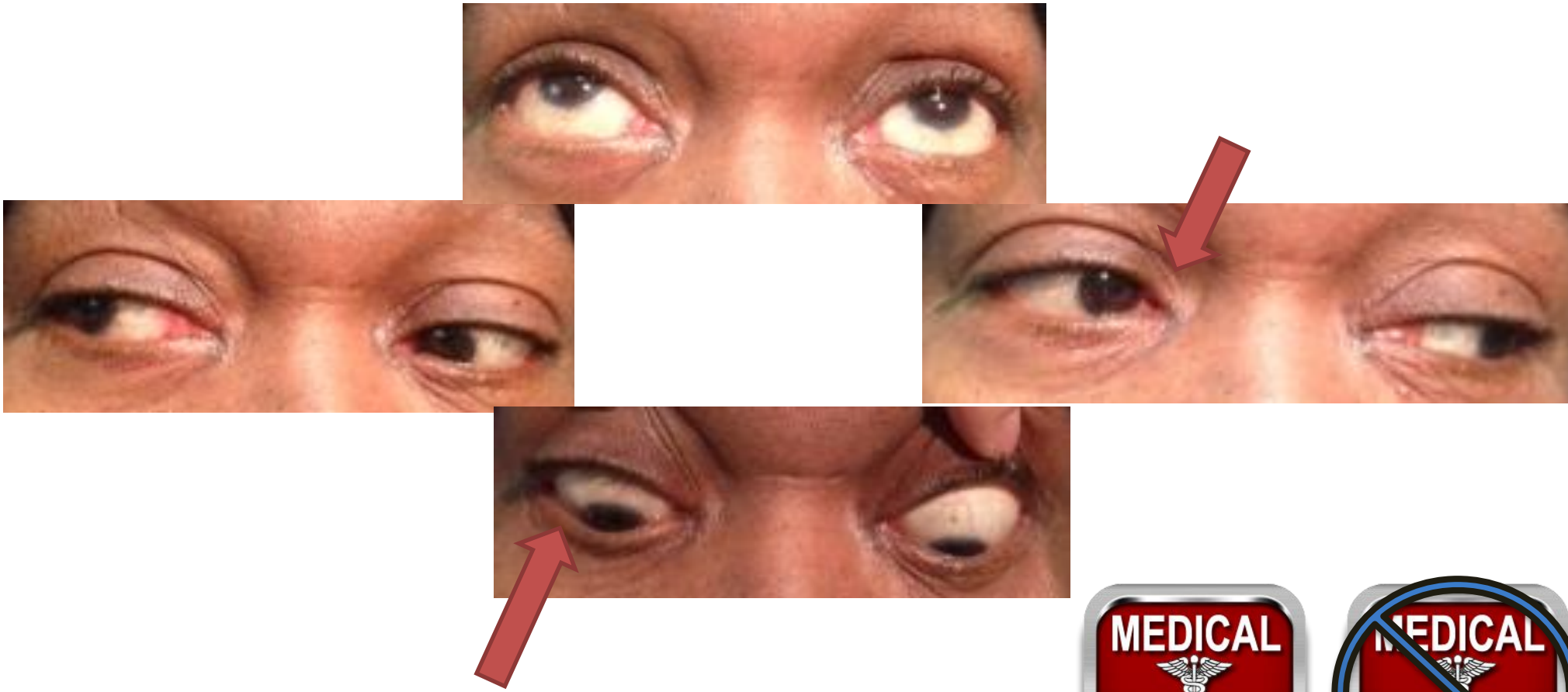
What is the cause of the adduction deficit and infraduction deficit?



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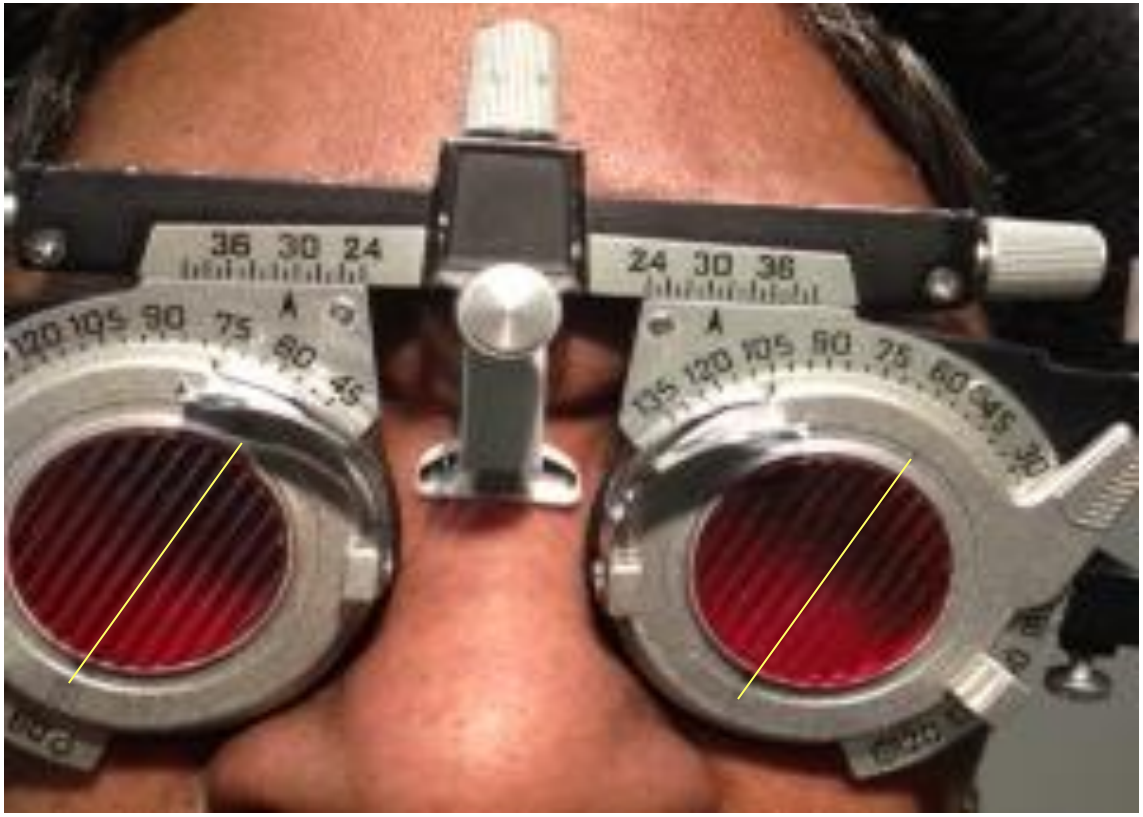
What is the cause of the adduction deficit and infraduction deficit?

- CN III Palsy
- Thyroid Orbitopathy
- Myasthenia Gravis
- INO and Skew Deviation

What is the cause of the adduction deficit and infraduction deficit?

- CN III Palsy
  - Negative forced duction test
- Thyroid Orbitopathy
  - Positive forced duction test
- Myasthenia Gravis
  - Negative forced duction test
  - Fatigue
- INO and Skew Deviation
  - Abducting nystagmus
  - Higher eye intorted, Lower eye extorted
  - Negative forced duction test



Double Maddox rod testing :
- 15-20 degrees of incyclotorsion OD
- 15-20 degrees of excyclotorsion OS

CONFIRMS SKEW DEVIATION!

- **INO / Skew**
  - Likely secondary to stroke
    - In setting of elevated BP and low pulse
- Admit to hospital for emergent work-up
  - MRI
    - **Acute brainstem lesion noted**

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INTERNUCLEAR OPHTHALMOPLEGIA

MOST COMMON ETIOLOGY

OLDER PATIENTS

- VASCULAR / STROKE

YOUNGER PATIENTS

- DEMYELINATING

ANY BRAINSTEM MOTILITY PROBLEM IN OLDER PATIENT, ALWAYS NEED TO R/O STROKE.

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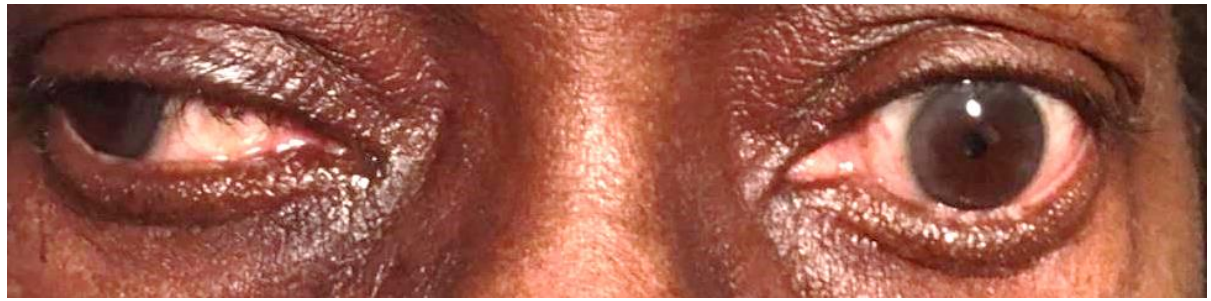
CASE 8

76 Year-Old Woman



- Complaint of diplopia and ptosis

- Arthritis
- GERD
- Osteoporosis
- Denies Diabetes, Hypertension, Hypercholesterolemia

- Meds: Alendronate, Allegra, Caltrate + D3, Centrum Silver, Colase, Omeprazole

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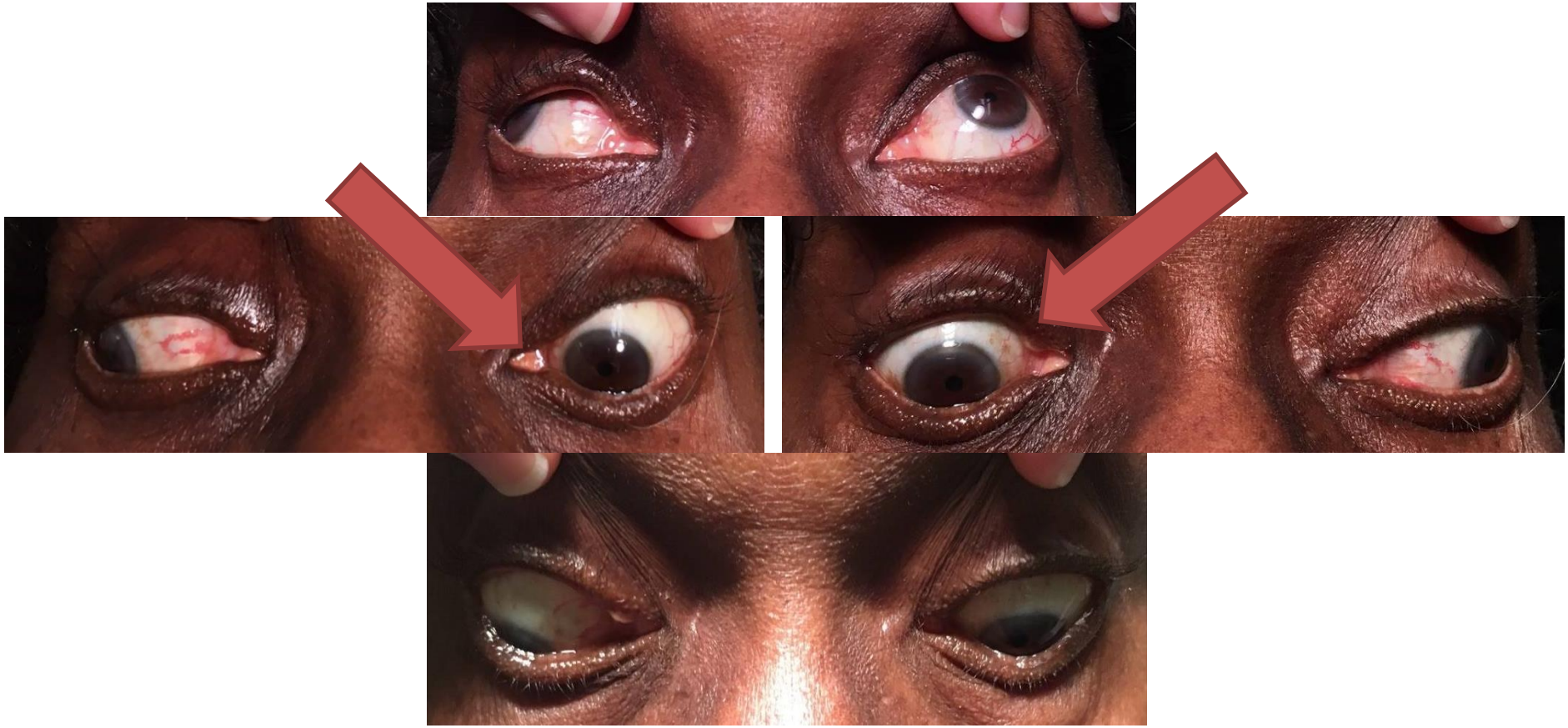
EXAM RESULTS

- VA 20/20 OD 20/25 OS
- Color (Ishihara): 14/14 OD, 14/14 OS
- PERRLA (-) RAPD
- CF: full OU
- Exophthalmometry: 20 OD 20 OS
- BP: 118/64
- Normal SLE, IOP, and DFE



Bilateral Adduction Deficit

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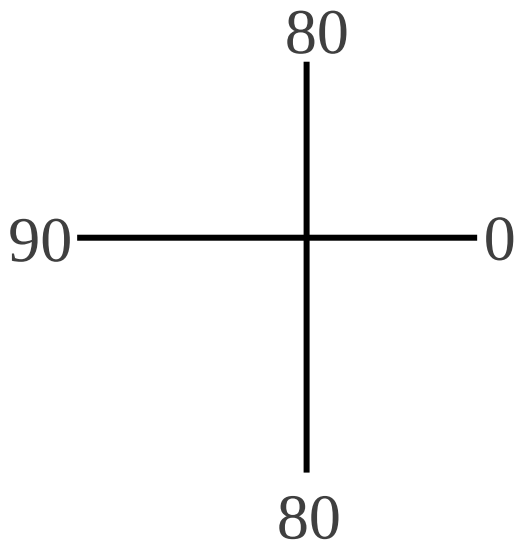
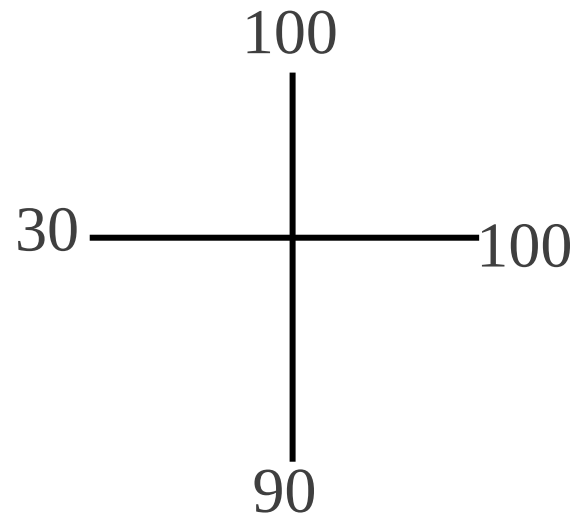
Ocular Motility





	90 exo 5 RH	
70 exo 3 RH	85 exo 8 RH	75 exo 4 LH
	65 exo 5 RH	

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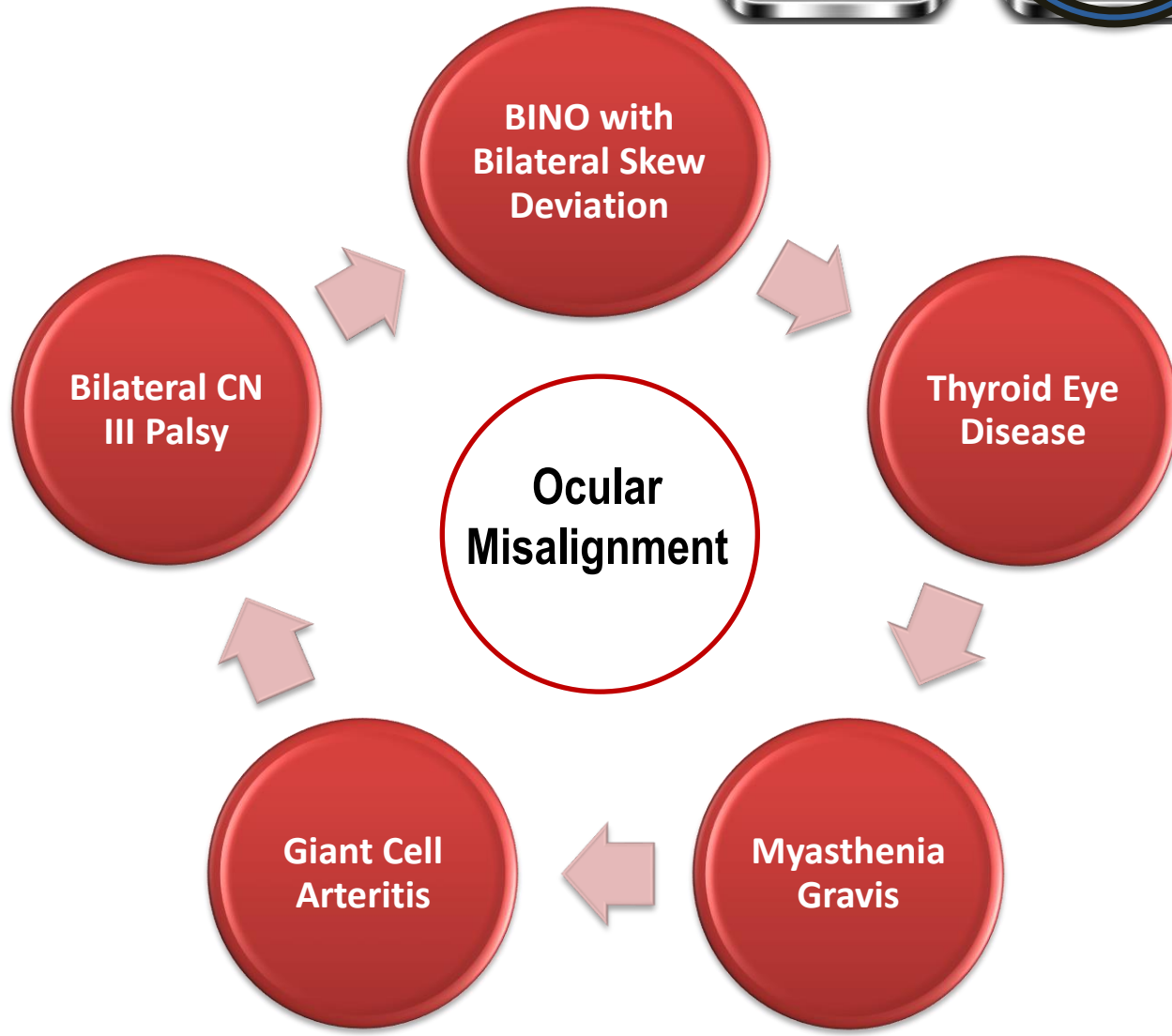
DDX FOR OCULAR MISALIGNMENT

- Looks like a Bilateral Internuclear Ophthalmoplegia (BINO) with Bilateral Skew Deviation
- What would be the management plan?
  - Send to the ER to rule out acute brainstem stroke

BUT…



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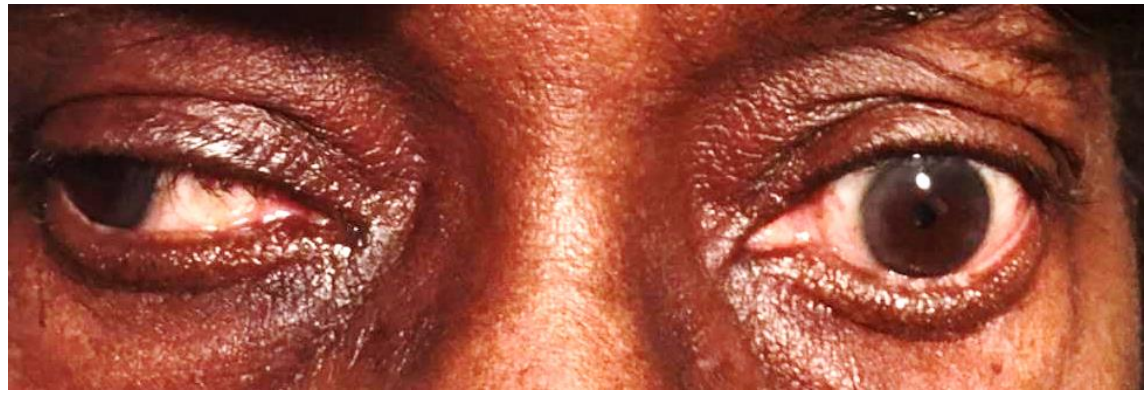
- **How do we explain the ptosis if this is a BINO and skew deviation?**



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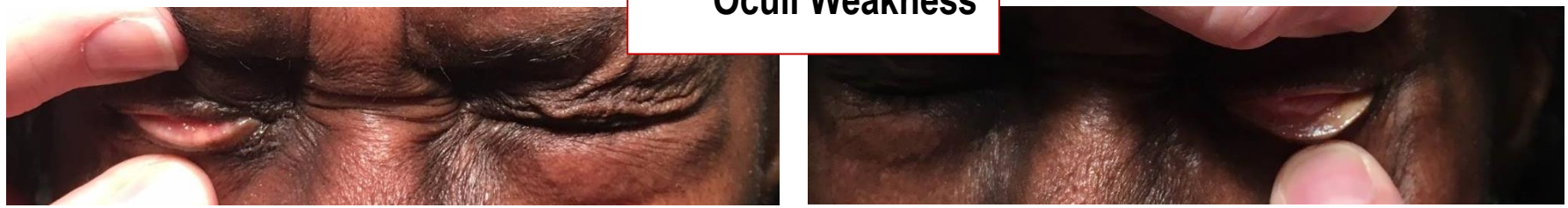
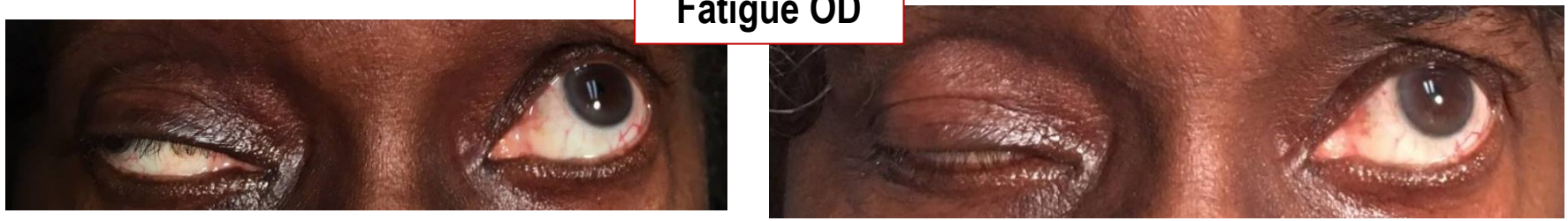
Additional In-Office Testing

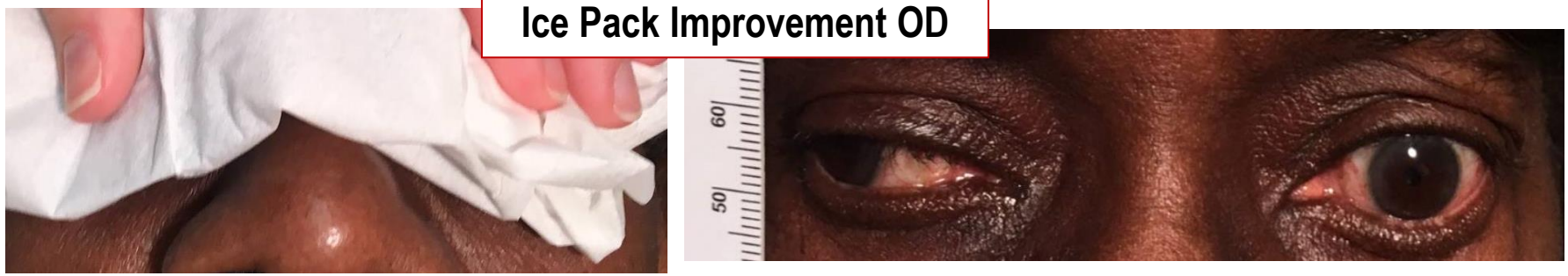
Bilateral Orbicularis Oculi Weakness



Fatigue OD



Ice Pack Improvement OD



Need to consider myasthenia gravis in ALL cases of diplopia! … and in ALL cases of ptosis!

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- Patient later admits to other symptoms:
  - Swallowing difficulty
    - X 6 months
    - She scheduled an ENT appt
    - Has had difficulty swallowing liquids
    - Has choked on food
  - Denies any breathing difficulty or weakness

Work-Up for MG

- Acetylcholine Receptor Antibodies 
- Single Fiber EMG of orbicularis oculi 
- Chest CT scan 

Treatment

- Was treated with Mestinon 60 mgTID
  - did not provide sufficient relief of symptoms
  - Mestinon was discontinued
- She was put on Prednisone with a gradual increase in dosage over several weeks, up to a maximum of 60mg of Prednisone QD
  - Once her symptoms improved, her dosage was tapered

MG TREATMENTS

SYMPTOMATIC TREATMENT

- Acetylcholinesterase Inhibitors

CHRONIC IMMUNOSUPPRESSIVES

- Glucocorticoids
- NSAIDS

RAPID, SHORT-ACTING IMMUNOMODULATORS

- PLEX
  IVIG

SURGICAL TREATMENT

- Thymectomy

WHO GETS MG? …. ANYONE!

- PEAK INCIDENCE (generalization)
  - –YOUNGER WOMEN (15-20)
    - But, occurs in older women as well!
  - –OLDER MEN (50-60)
    - But, occurs in younger men as well!
- OVERALL F:M 2:1
- UNDER 3O: F:M 4.5:1
- Can occur at any age!

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MYASTHENIA GRAVIS

- 60-70% present initially with ocular signs
- 90% of all myasthenics have ocular signs
- 15% will ONLY have eye signs (OCULAR MYASTHENIA)

Causes painless, variable diplopia and ptosis

- Ocular muscles include levator, orbicularis oculi and EOMs
- Does not affect pupils (no voluntary muscle)

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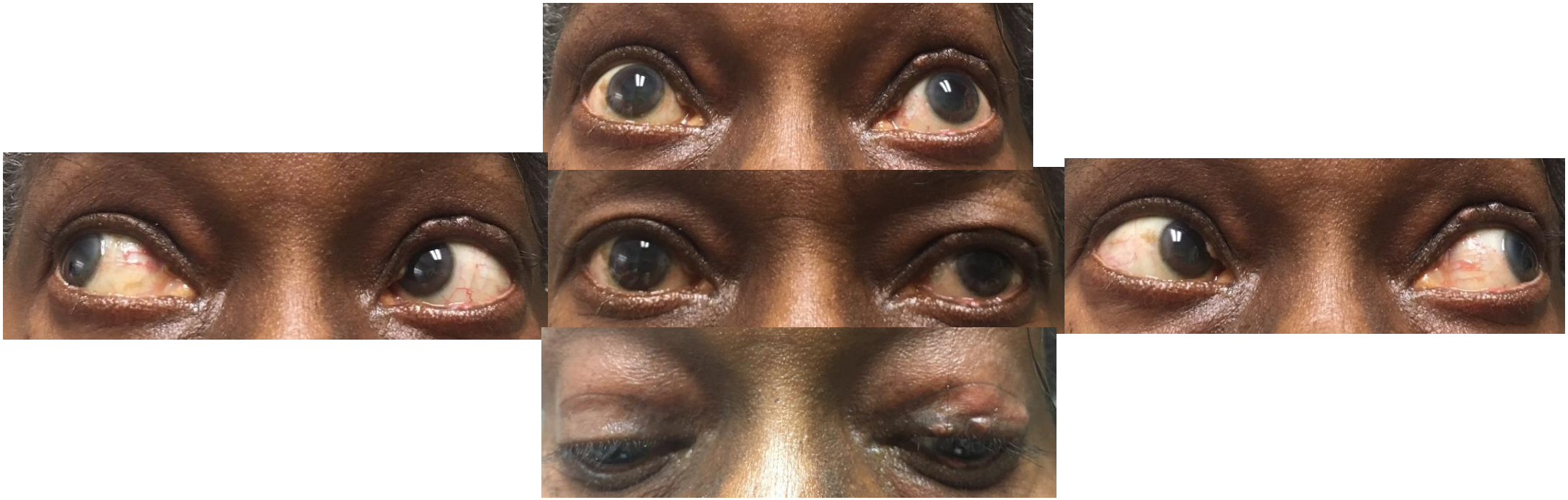
While ocular MG is not a medical emergency, some cases of MG (bulbar symptoms, myasthenic crisis) can be emergent!

IMPROVEMENT ON FOLLOW-UP



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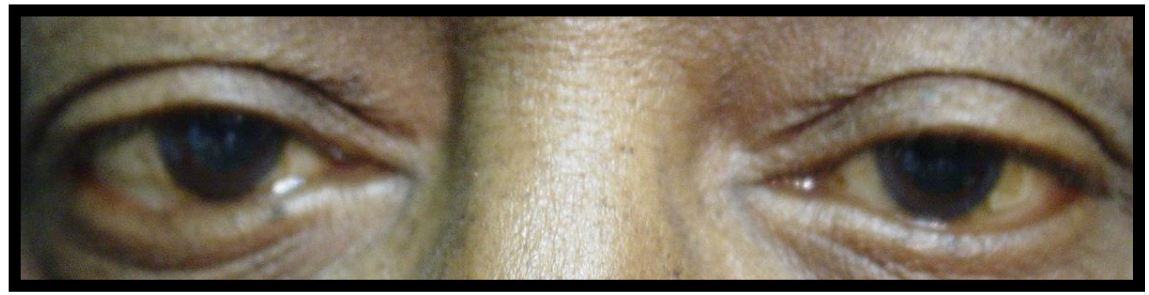
CASE 9

66 Year-old man



- Sudden onset blurry vision x 4 days
- "glare" in left gaze, no diplopia
- Wife notes - OS sometimes turns in
- Examined at ER, told BP (190/90) cause of blur
- HTN x10 yrs, prostate CA-chemo q 3 m – no surg/rad
- HCTZ, Nifedipine, unspecified chemo agent
- Denies eye / head pain, neuro or GCA symptoms

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VA: OD 20/30

OS 20/25

Pupils isocoric, (-)RAPD

CF: OD full OS full

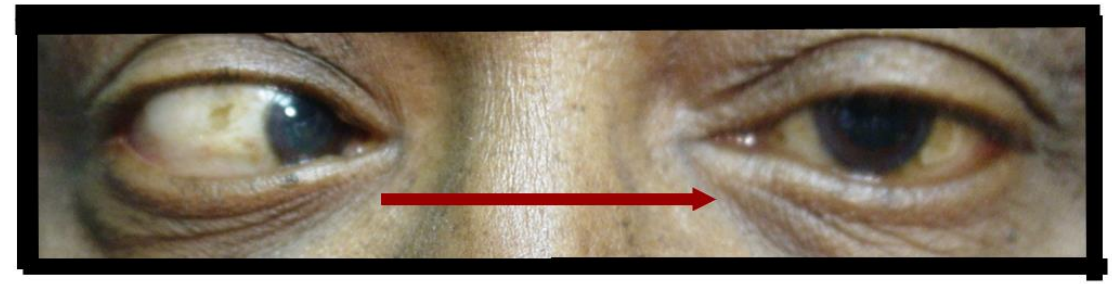
Color: OD 14/14 OS 14/14

Exophthalmometry:

- OD 23mm OS 22mm

SLE: mild cataracts

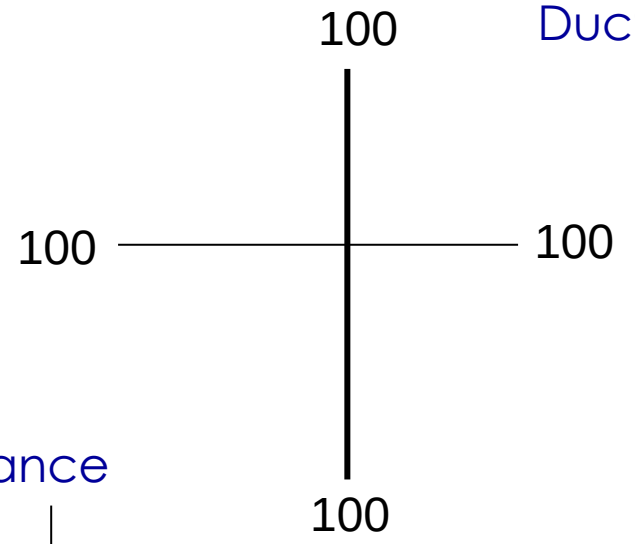
DFE: unremarkable

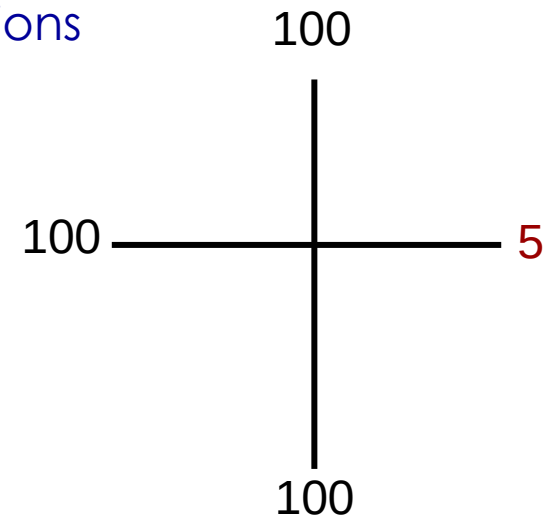


OD

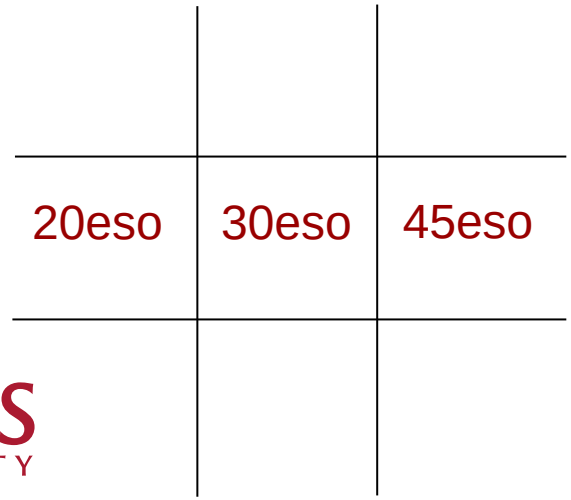
OS

Ductions





CT @ Distance



- **Slowed saccades to left OS**
- **Negative forced duction test OS**

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NEUROLOGIC EXAMINATION

CN V, VII – XII intact

Motor, sensory, coordination testing unremarkable

Left Abduction deficit +

Slowed Abducting saccades +

Negative Forced Duction Test

= Neurogenic CN VI Palsy

Could be vasculopathic, **BUT** need to R/O other etiology, especially mets due to prostate CA, pontine stroke, and GCA!

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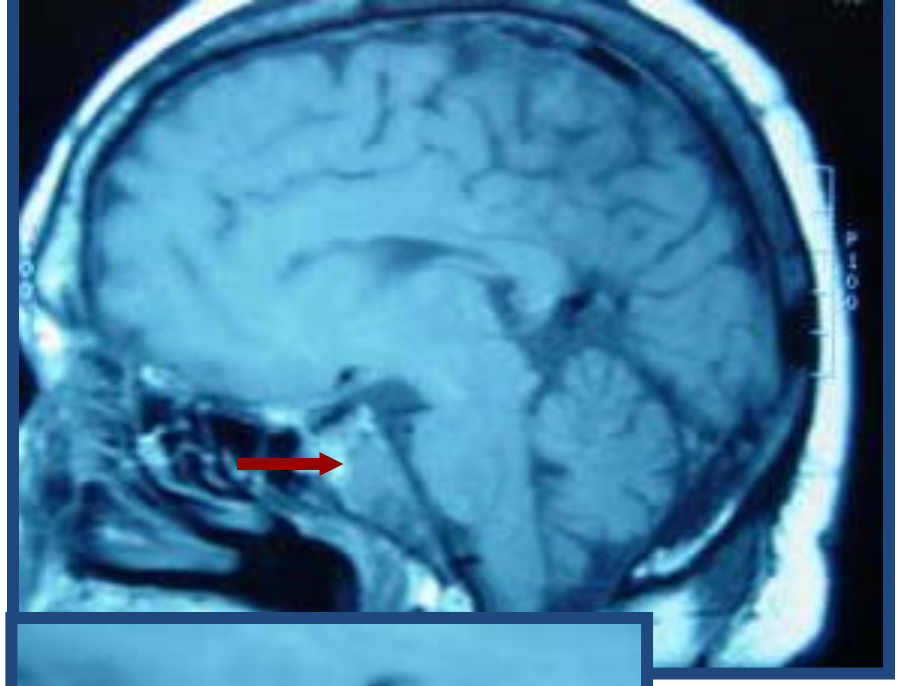
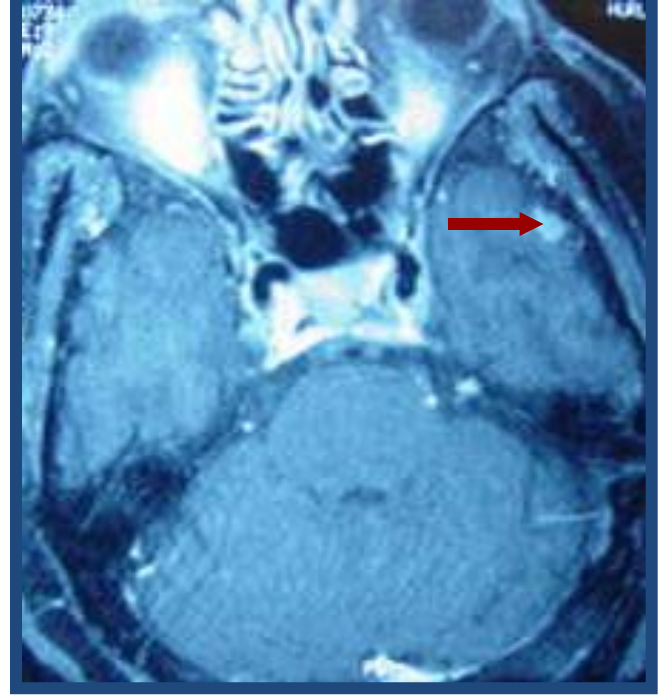
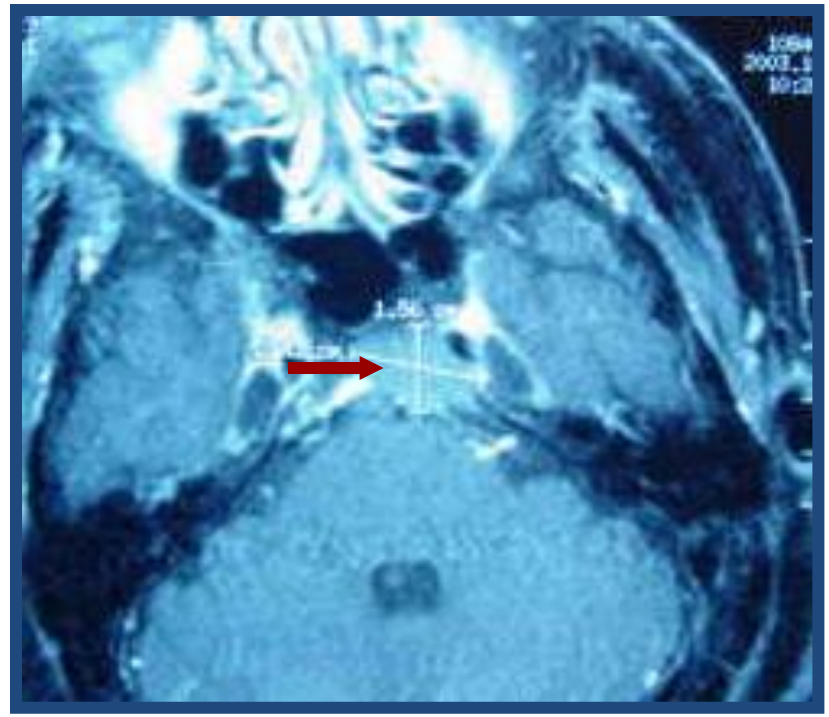




Prostate CA metastasis to clivus



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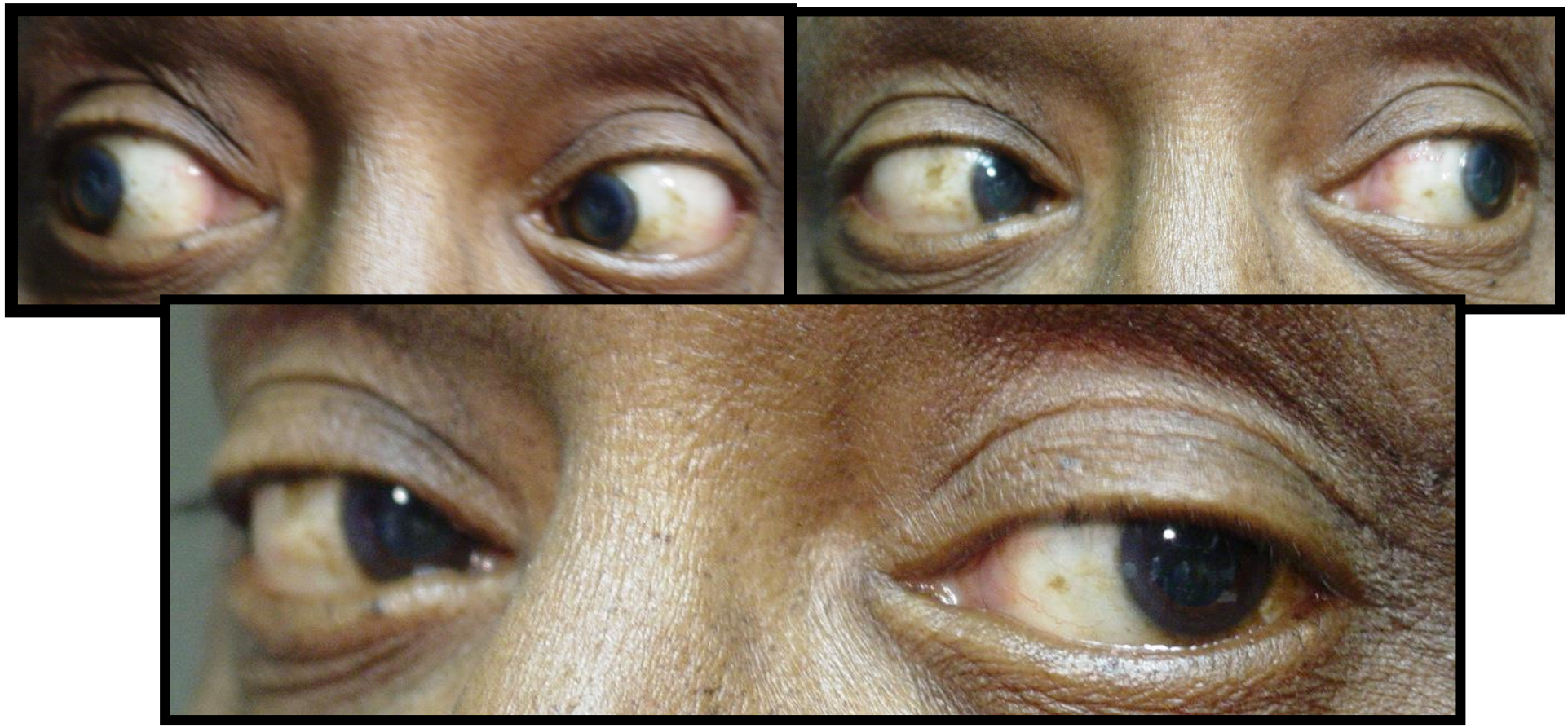

- S/P radiation treatment to involved areas
- Now 90 % normal abducting capacity OS

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BRAIN METASTASES

- **Location of Brain metastases**
  - Based on **blood supply**, since the metastases occurs through the bloodstream
    - **Cerebrum** (80-85%)
    - **Cerebellum** (10-15%)
      - Renal cancer
      - GI cancer
      - Pelvic cancer
    - **Brainstem** (3-5%)
- **Number of Brain metastases**
  - **Isolated**
    - Thyroid cancer
    - Colon cancer
    - Renal cancer
  - **Multiple**
    - Melanoma
    - Lung cancer
    - Breast cancer

If there appears to be an isolated brain metastasis on CT, there are often found to be multiple mets on MRI

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BONE METASTASES

- Primary bone cancers (to cause neuro-ophthalmic manifestations) are rare
- Bone mets are associated with lytic lesions and pain
- **Sites of Origin**
  - **Prostate**
  - **Breast**
  - **Lung**
- **Sites of metastases** (to cause neuro-ophthalmic disorder)
  - **Clivus**
  - **Other regions of skull base**

Lytic lesions involve destruction of bone.

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Advancing age is the most important risk factor for cancer overall, and for many individual cancer types.

The median age of a cancer diagnosis is 66 yrs.

Always consider cancer (or metastasis) as an etiology of a clinical presentation, especially in older patients, even if there is no known history of cancer.

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CASE 10

70 year-old man

- c/o a funny sensation on his right lower eyelid and under his right eye, and involving his right nose and upper lip. V2
- He suddenly noticed this about 1 month ago, and it seems constant since then, with nothing making it better or worse.
- He doesn't feel the right side above his lip when he shaves, or the upper right teeth/gums when brushing his teeth
- He denies any other visual, ocular, or neurologic symptoms.

He mentioned this to his PCP and cardiologist

They did not feel any additional work-up was necessary.

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- Hypertension x 12 years
- Hypercholesterolemia
- Osteoarthritis
- Coronary artery disease
- Status post 2 stent placements in 2000
- Status post spinal surgery because of herniated discs.
- Medications
  - Lipitor, Metoprolol, and 81 mg aspirin.
- Ocular history is unremarkable.
- Social history is remarkable for a past history of tobacco use, which was discontinued 15 years ago.

- VA: 20/20 OD, 20/20 OS
- Color: 5/6 OD, 5/6 OS
- PERRLA (-) RAPD
- CF: full OU
- EOMs: no restrictions
- No ptosis or proptosis

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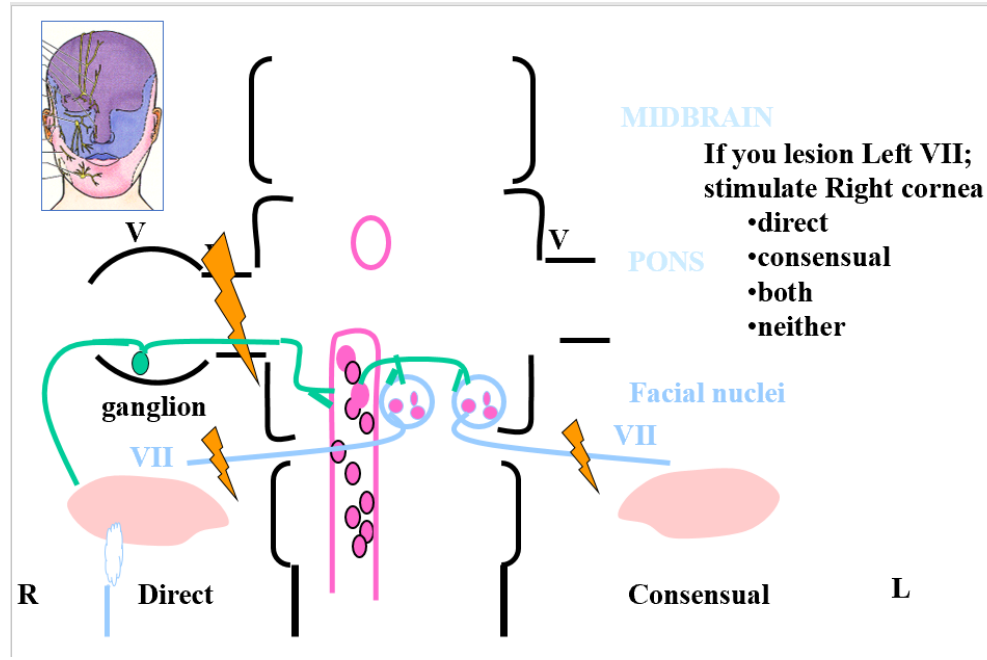
- Normal slit lamp exam
- Decreased corneal sensation OD as compared with OS (V1)
- IOP: 16 OD, 16 OS
- BP: 135/80
- DFE: 0.3 x 0.3 cupping OU
- (-) edema (-) pallor

NEUROLOGIC EXAMINATION:
Reduced sensation in the distribution of V2 on the right side as compared with left side

WORK-UP: MRI brain and orbits w/w/o



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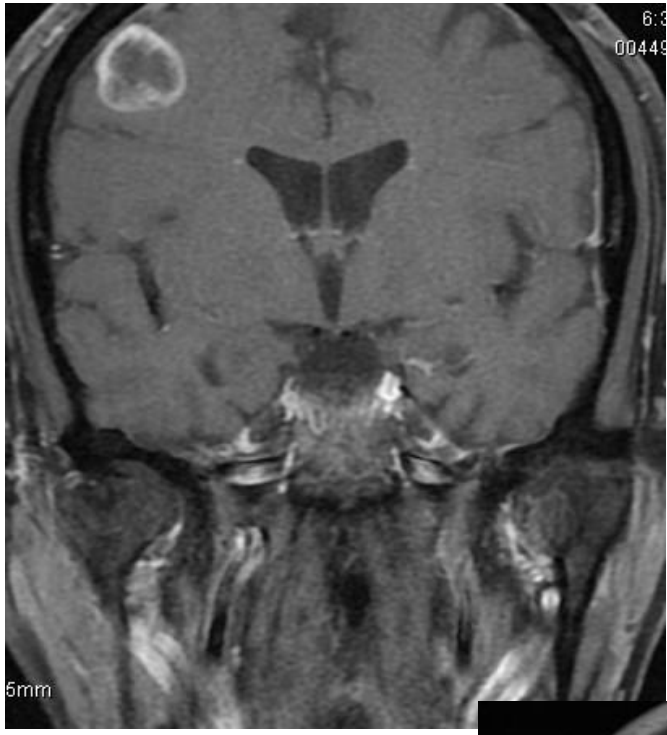

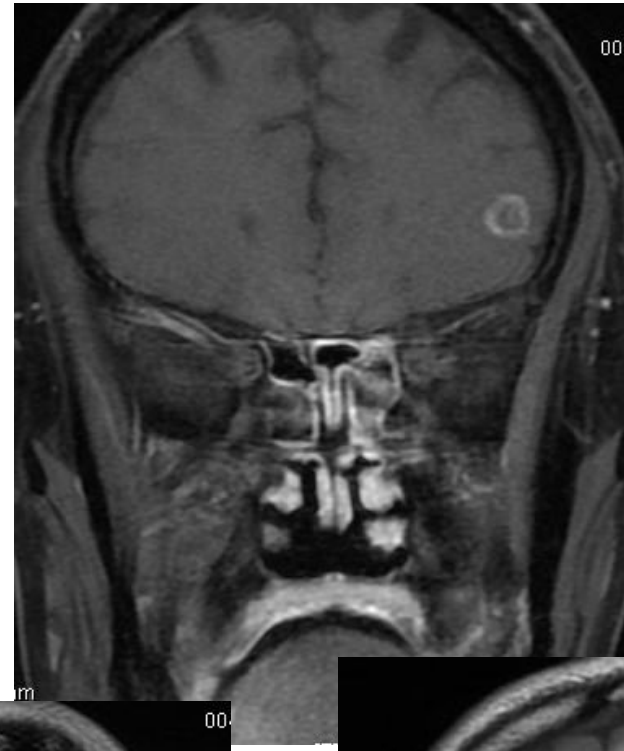

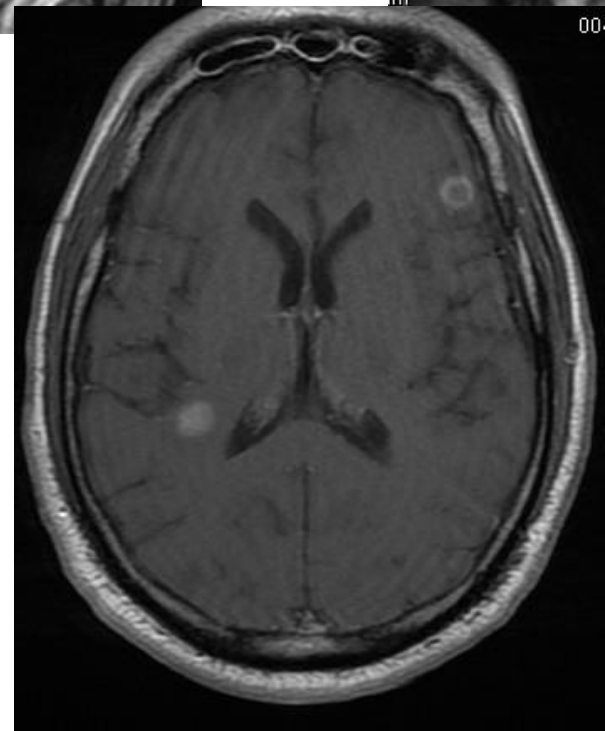

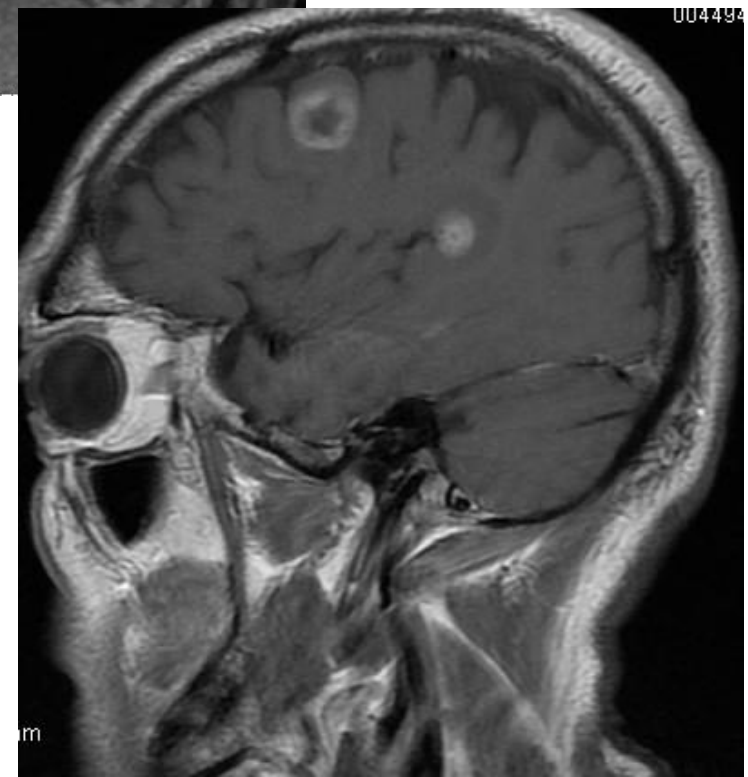

Multiple ring-enhancing lesions

Ring-enhancing lesions are concerning for metastases and abscess.

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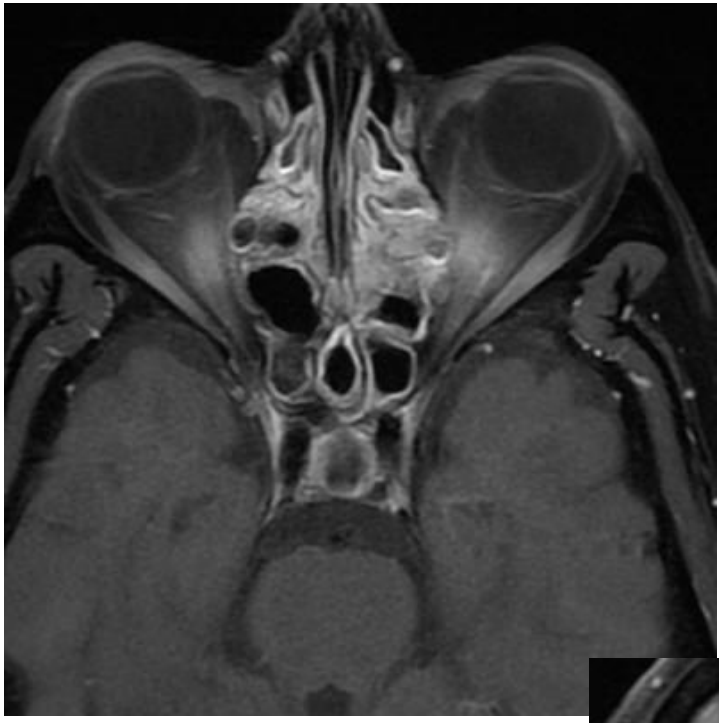

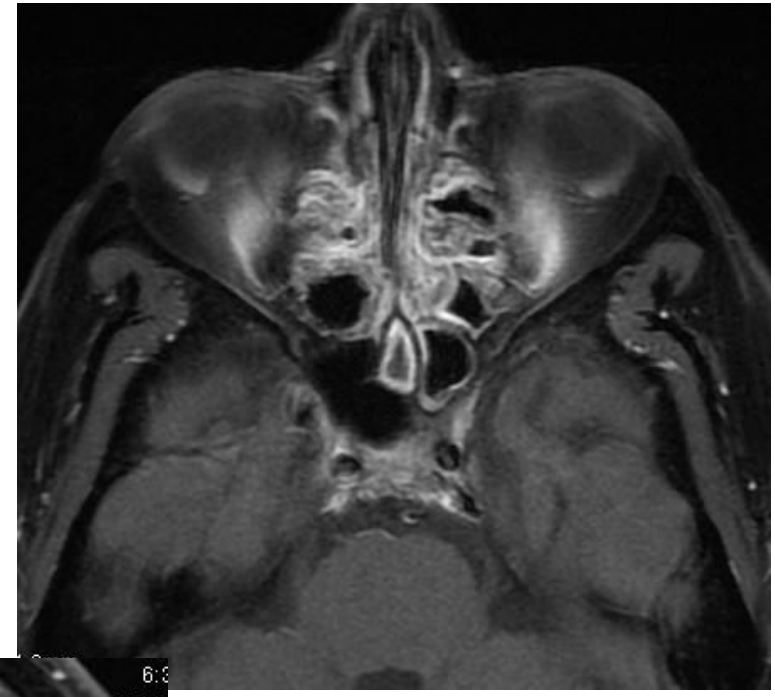

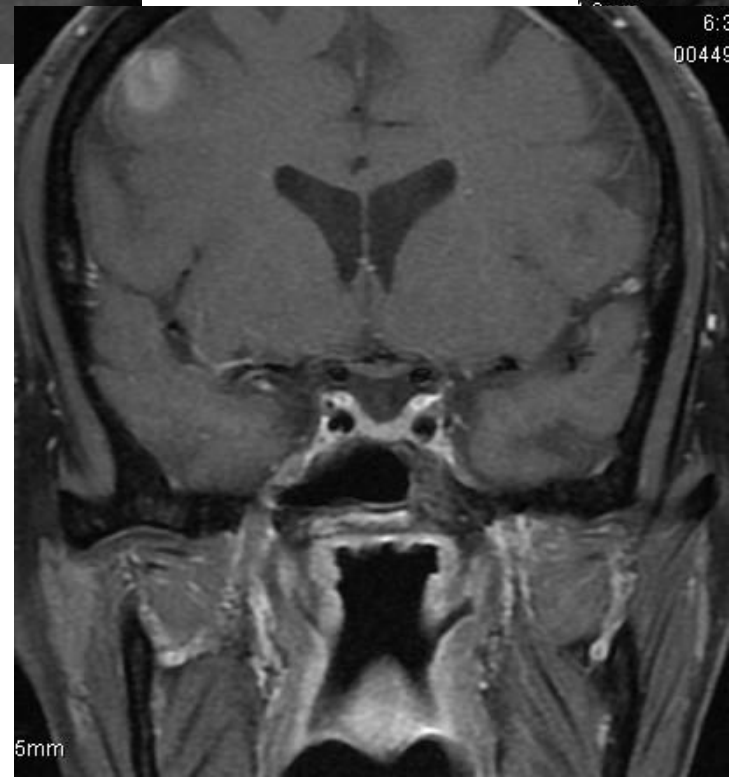

Highly concerning for metastatic disease

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- The patient has no known history of cancer.
- A work-up is needed to find the primary site
- He reports he had a normal colonoscopy a few months ago

Suspect these are brain mets.
(He has no fever, or other indication of infectious process)
Need to find primary cancer.

- He now admits to some frontal headaches
  - X 1-2 months
  - May be getting more frequent
  - Relief with ibuprofen
- No changes in weight or appetite

BRAIN METASTASES

- Result from systemic cancers involving:
- Lung ****
- Breast ***
- Melanoma ****
- GI tract (colon, pancreatic)
- Lymphoma
- Genitourinary (kidney, testes, ovary, cervix)
- Thyroid

**** = most common

Isolated brain met: thyroid, renal, colon

Multiple brain mets: (breast),lung and melanoma

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- WORK-UP:
  Chest x-ray: nodular density in the right lower lobe (shadow vs. nodule)
- CT of chest / abdomen / pelvis with contrast:
  - Consistent with lung cancer (carcinoma)

Tx: radiation to brain, chemotherapy

SITES OF CANCER TO CAUSE NEURO-OPHTHALMIC MANIFESTATIONS

- Brain
- Bone (skull)
- Meninges
- Orbit
- Lymph nodes
- Others (less applicable to us)

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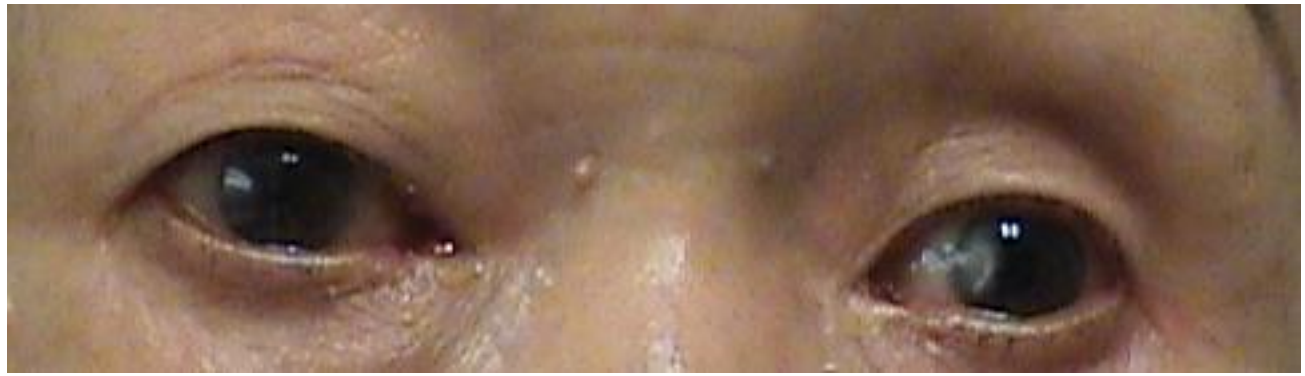
CASE 11

88 Year Old Woman



PAST HISTORY: B-cell lymphoma of groin
15 years ago
s/p chemo; in remission

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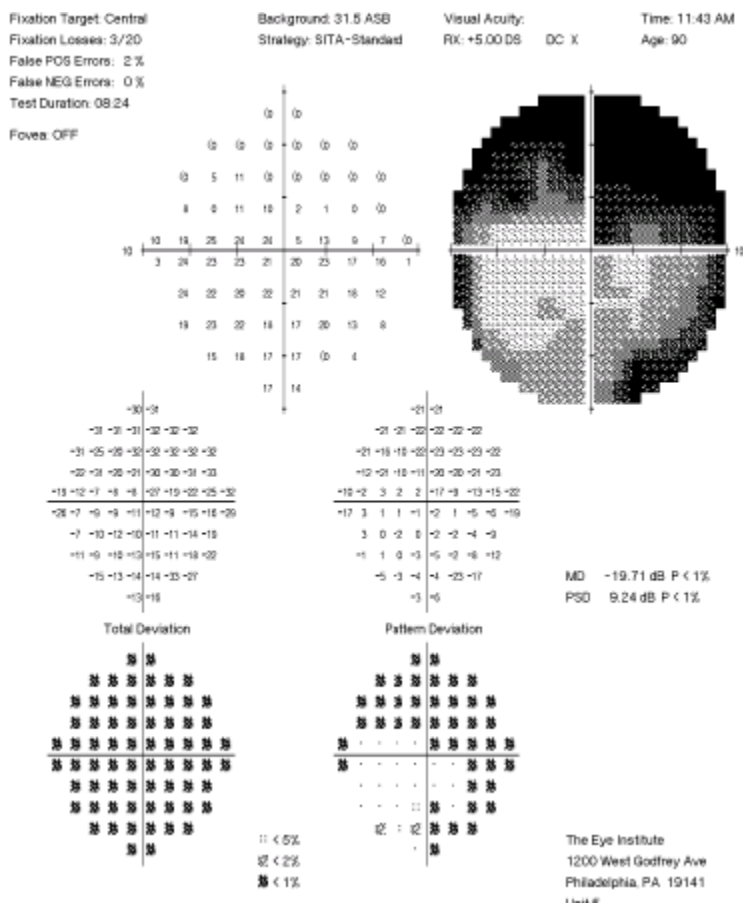

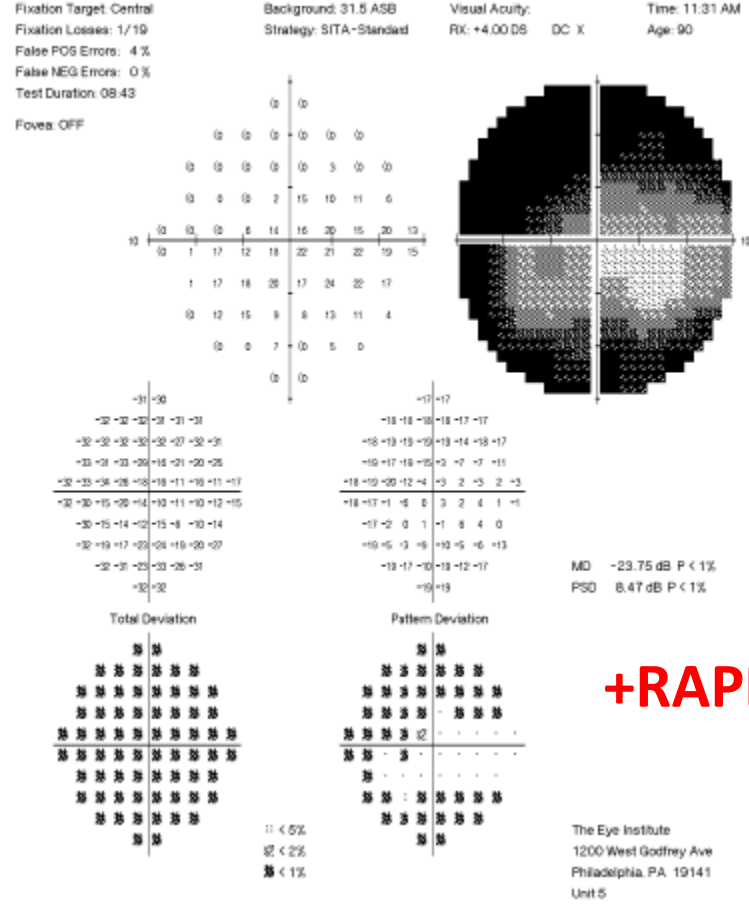

+RAPD OD

IOP: OD 17mm Hg, OS 11 mm Hg on treatment

DFE: OD 0.95/0.95, OS 0.95/0.95 (-)edema OU

Longstanding history of severe glaucoma OU

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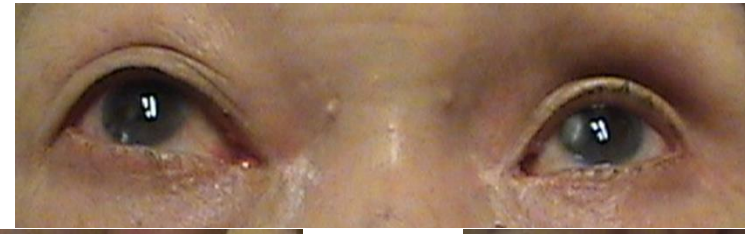

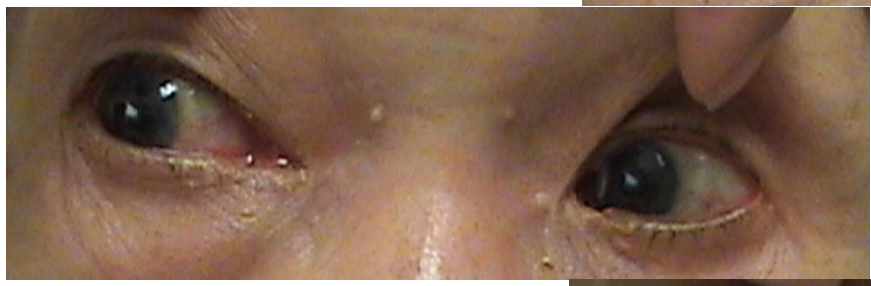

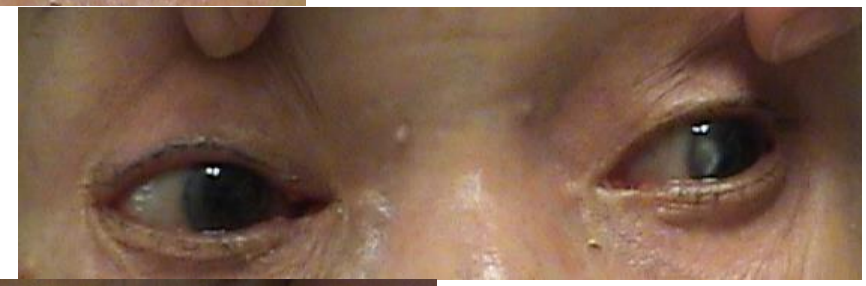

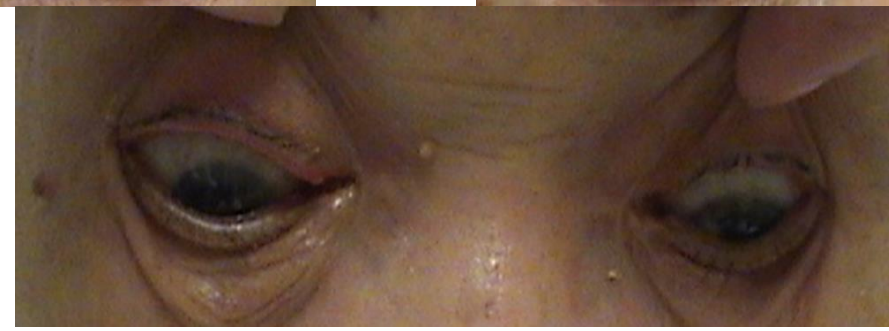

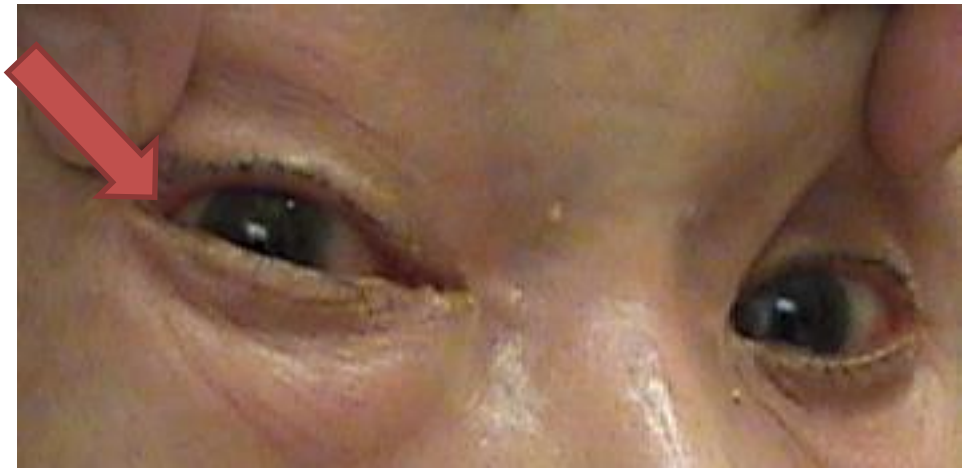

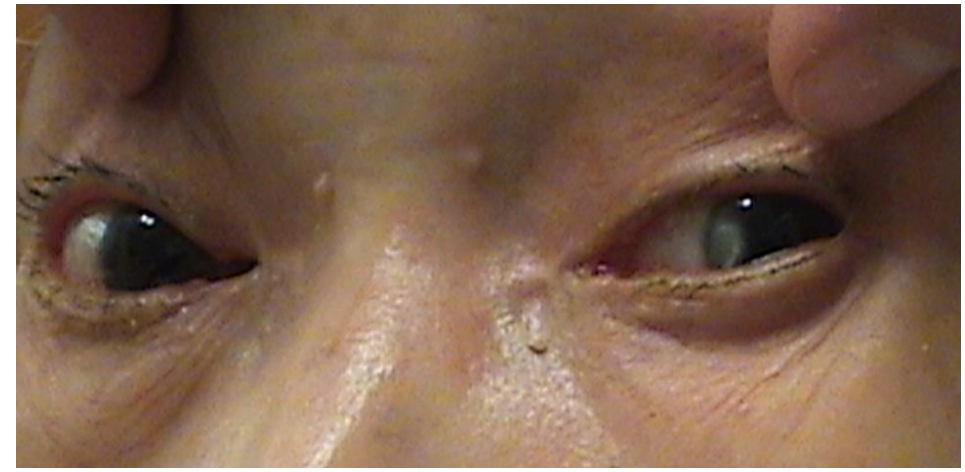

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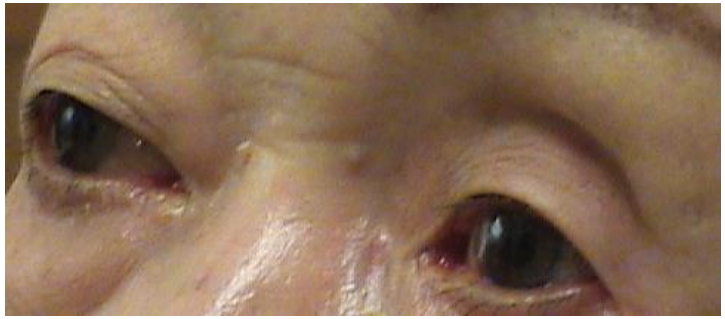

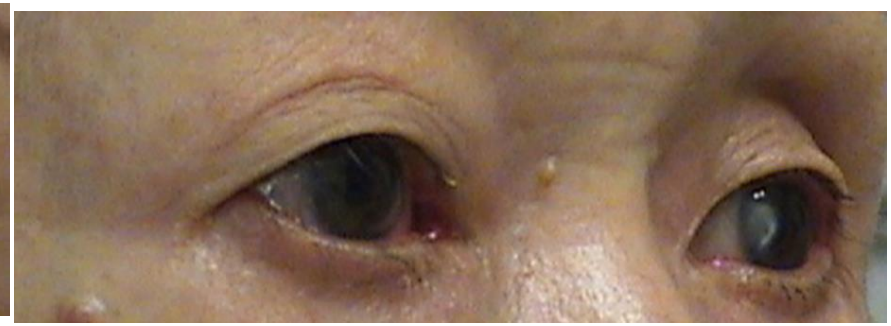

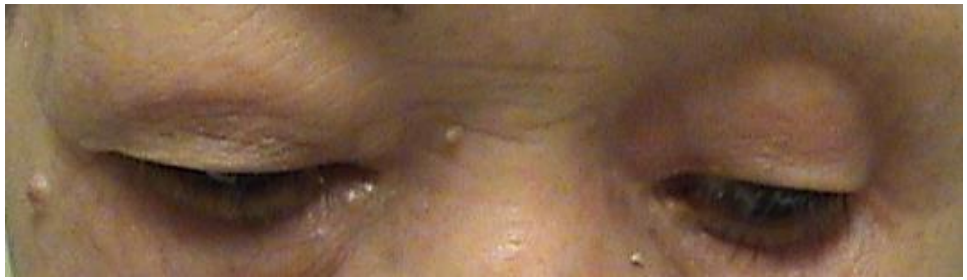

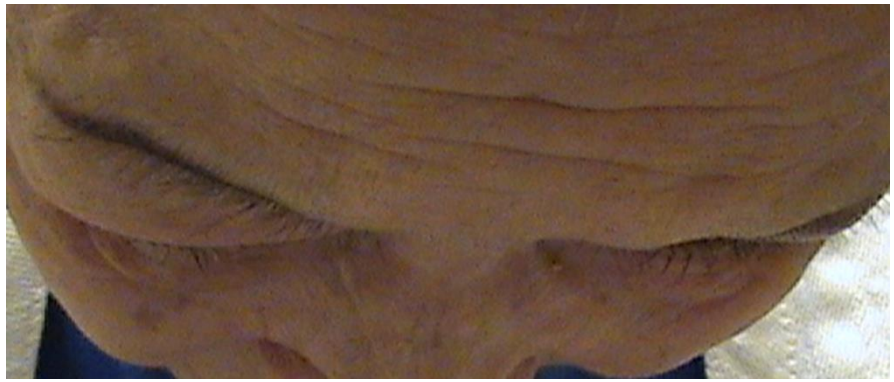

DDX of Painless Proptosis

- Lymphoma **
- Sarcoidosis
- Tuberculosis
- Granulomatous Polyangiitis (Wegener's)
- Other Orbital Mass (cystic, neoplastic, neural, vascular, fibrosing, lymphoid)
- IOIP (Pseudotumor)

Exophthalmometry:
OD 13mm OS 9mm

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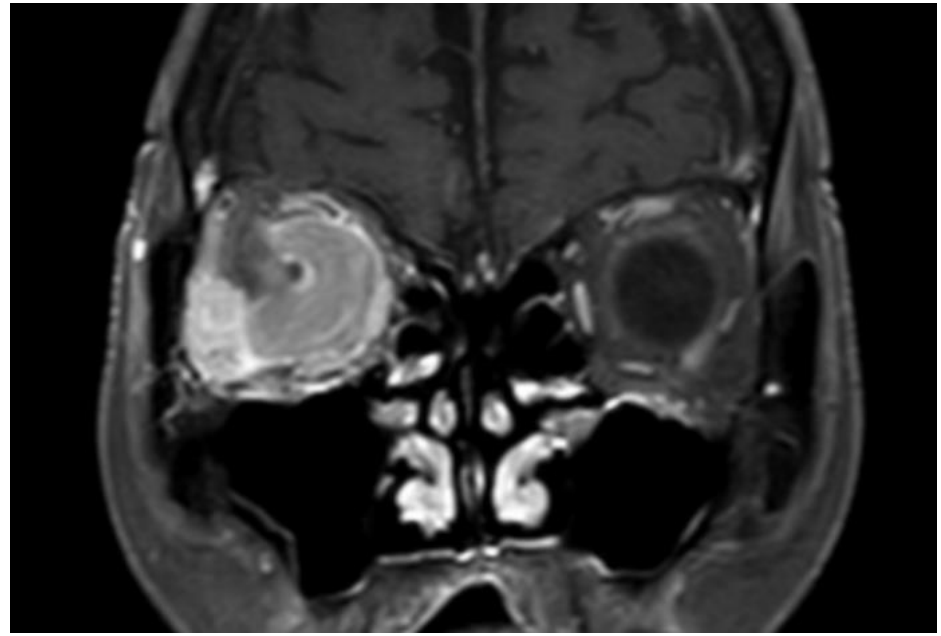
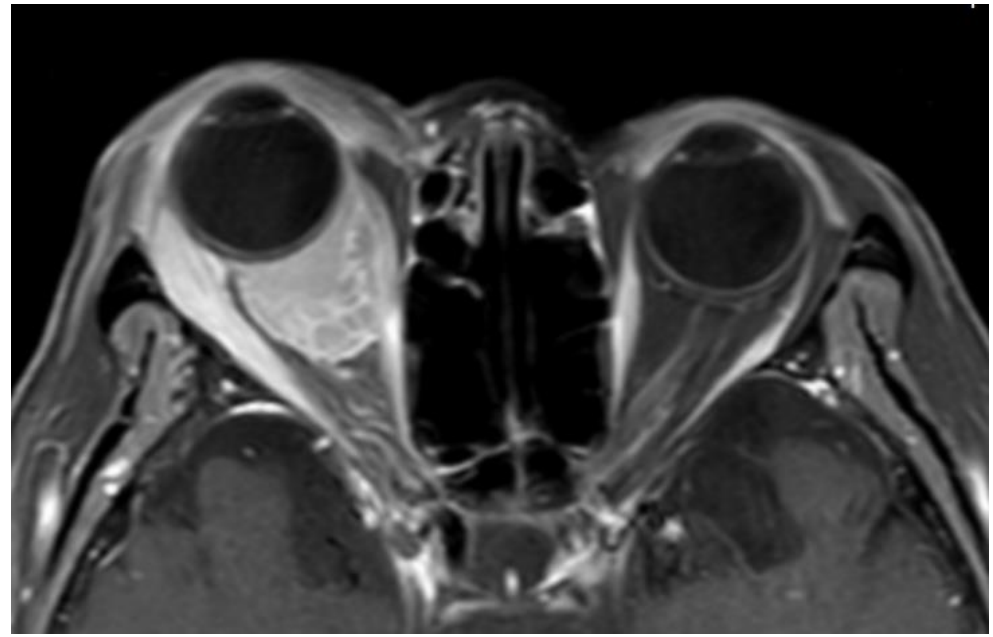
We ordered MRI

Orbital Lymphoma





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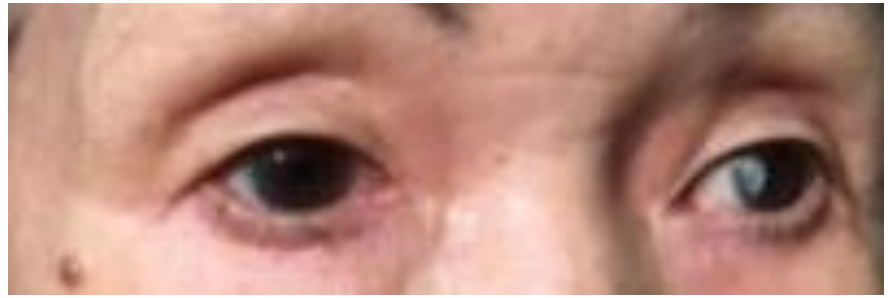
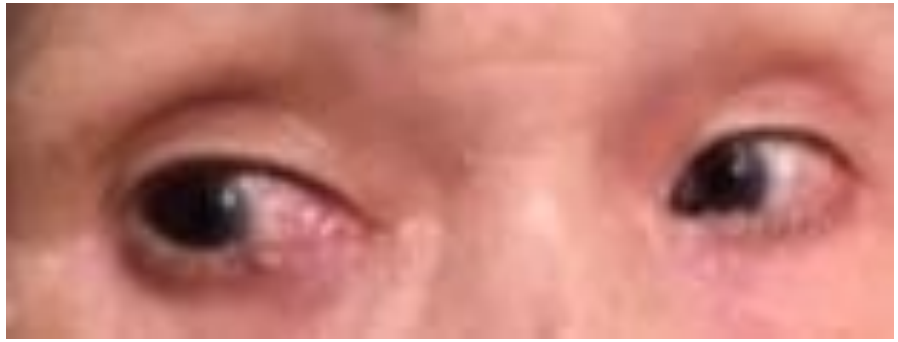
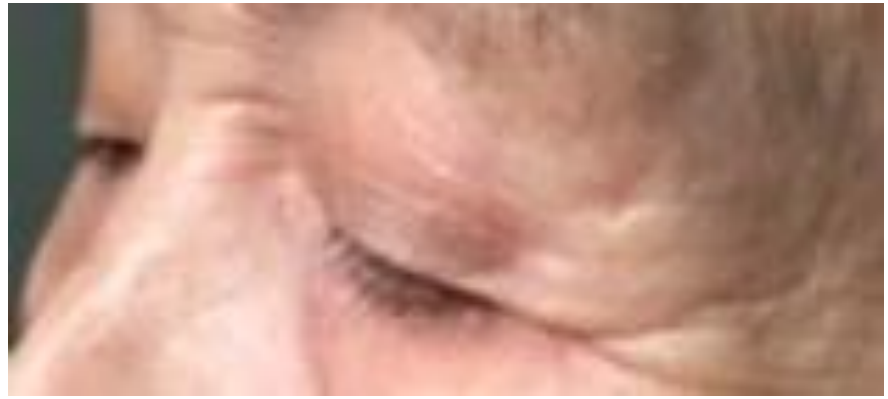
S/P biopsy & radiation







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- Always consider cancer as an etiology for any afferent and efferent neuro-ophthalmic presentation!

- It could be a primary or secondary cancer…

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CASE 12

77 year old man

- Reports 3 week history of blurred vision OD
  - Notices especially when reading
  - Right-sided weakness
- Visual acuities 20/20 OD 20/20 OS
- PERRL (trace +) RAPD OD
- Confrontation fields: right homonymous hemianopia denser superiorly
- Medical history
  - Hypertension

How Does The RAPD Help With Localization?

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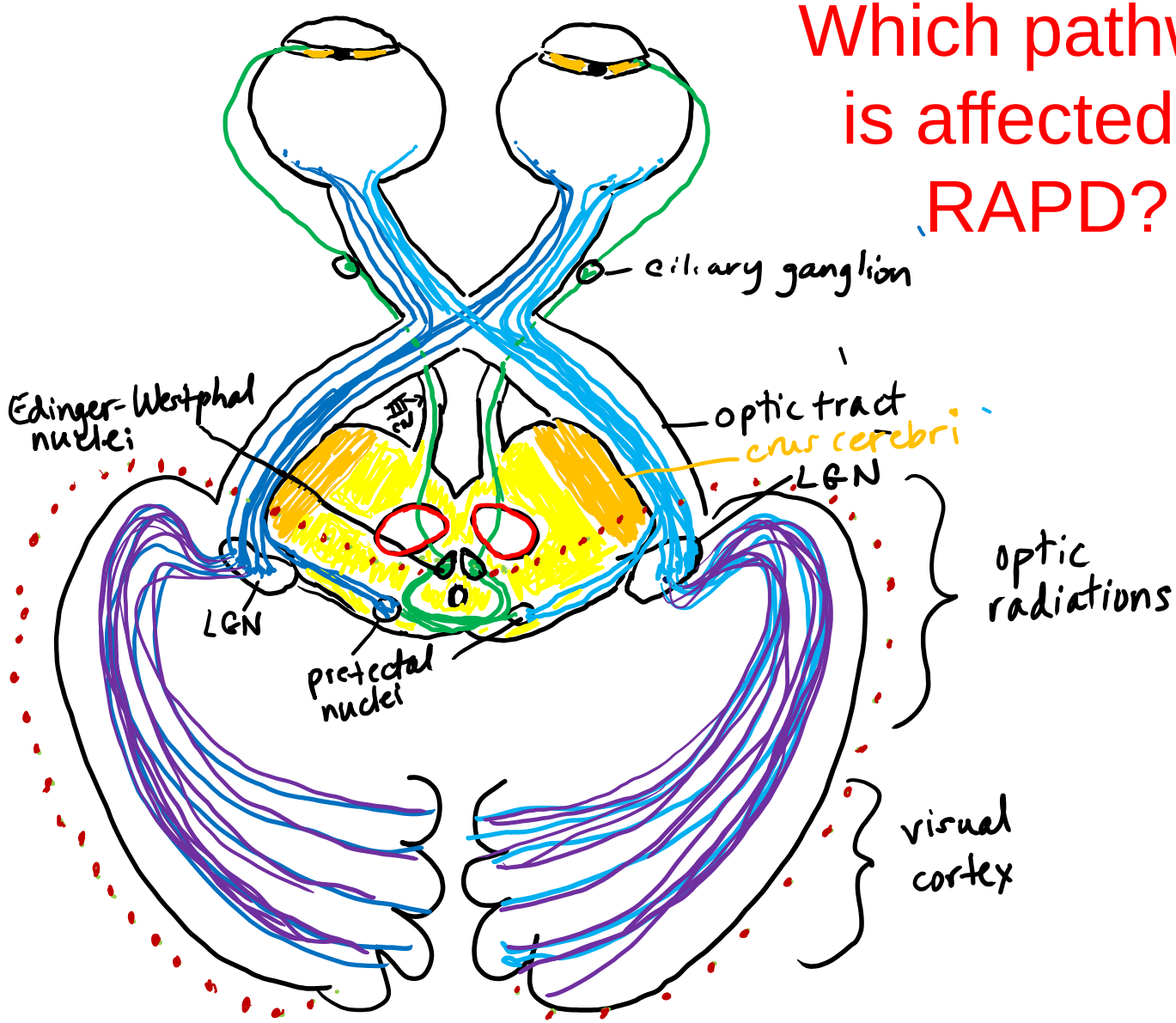
Super-imposed Light, Near, and Visual Pathways



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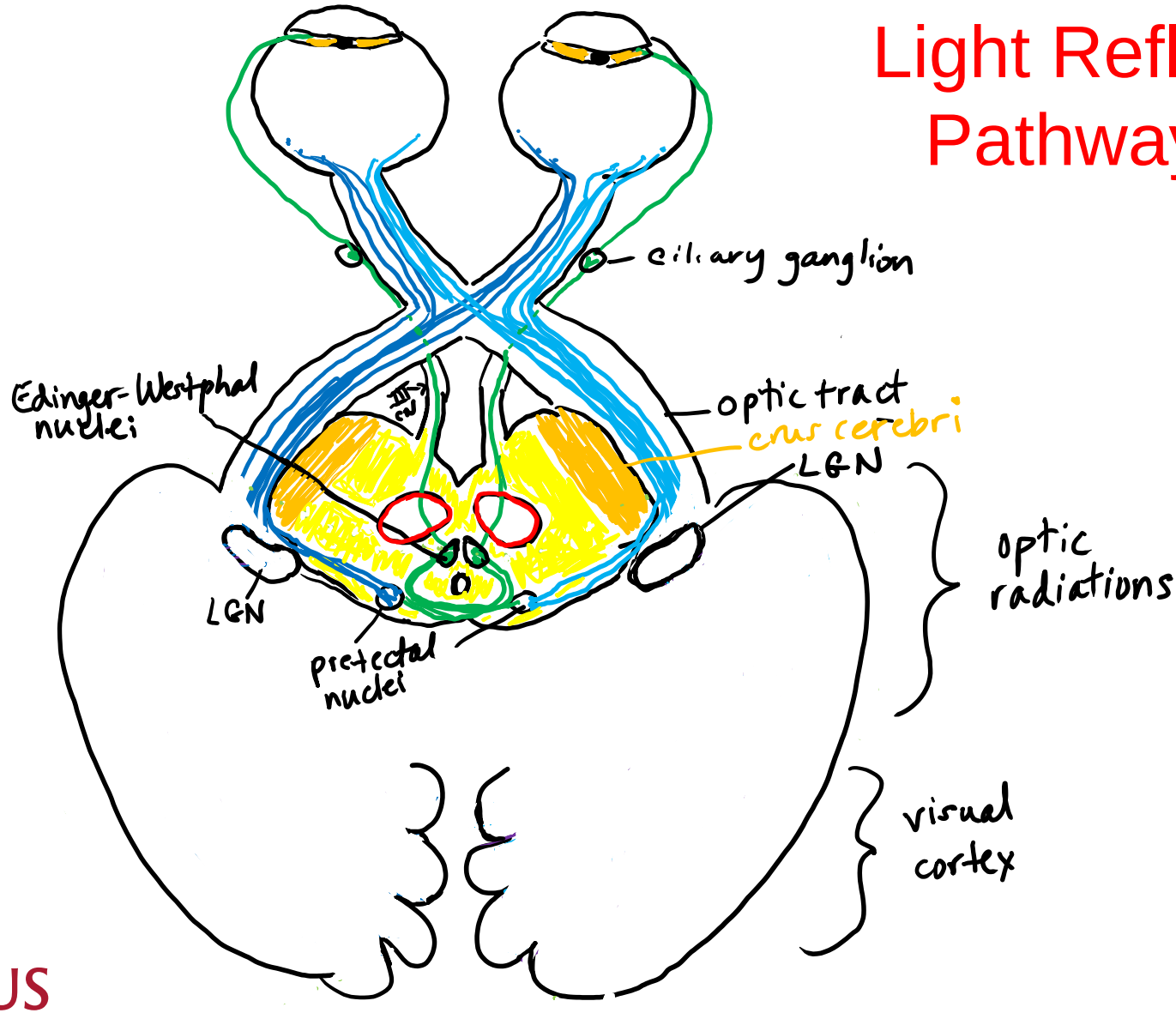
Pupillary Light Reflex Pathway

Afferent Limb of Light Reflex Pathway



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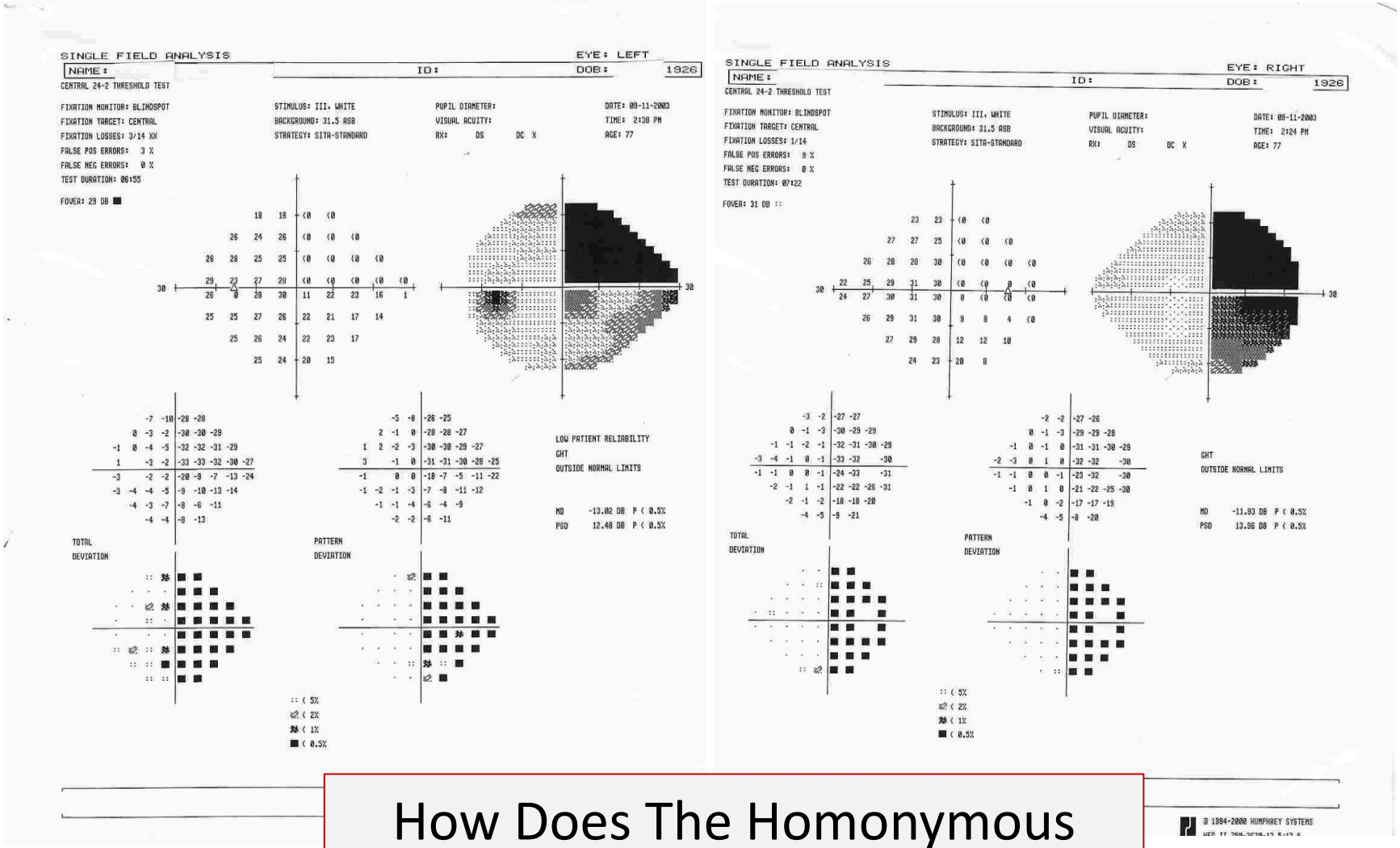
Visual Field Results



How Does The Homonymous Hemianopia Help With Localization?

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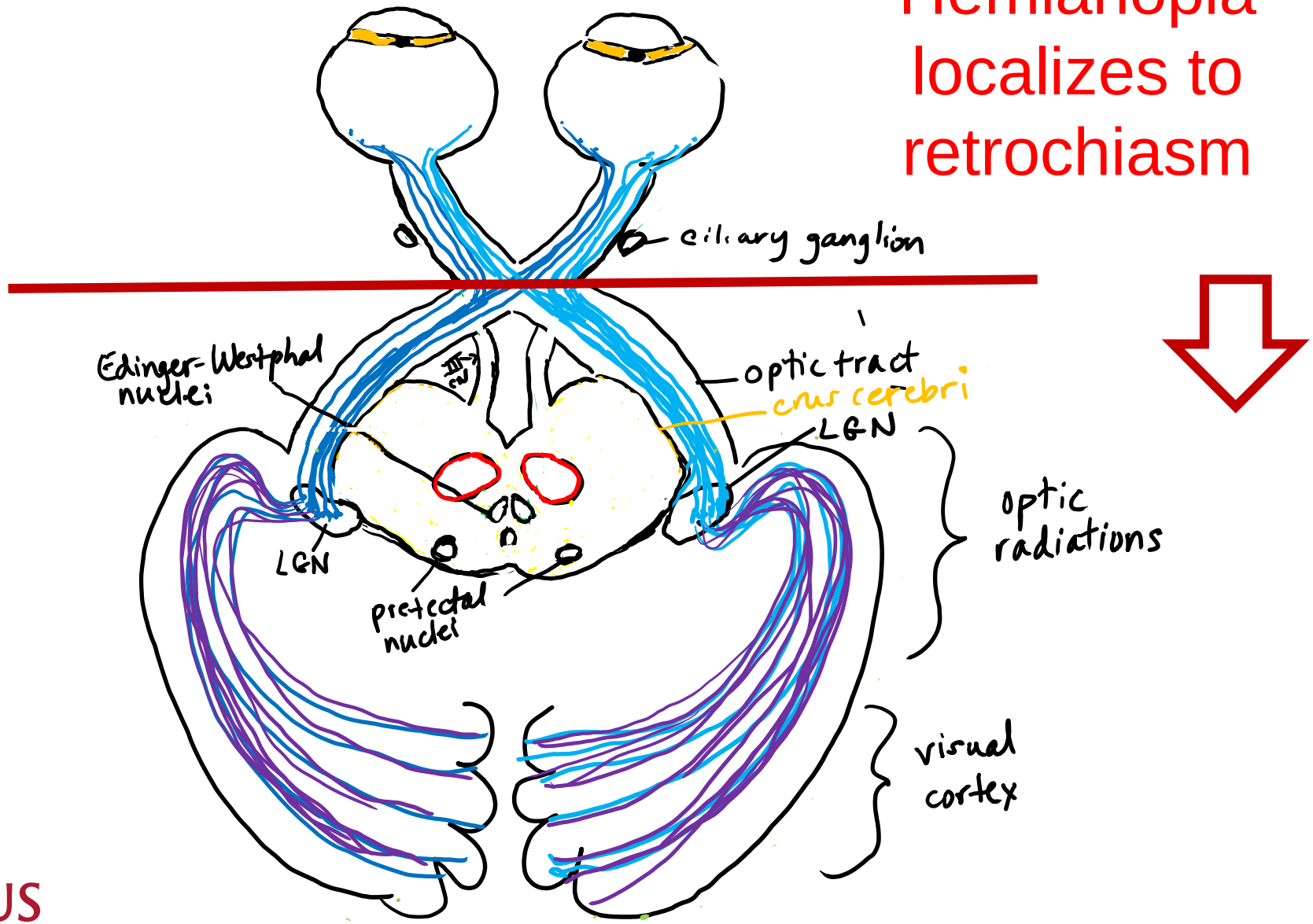
Visual Pathway

Homonymous Hemianopia localizes to retrochiasm



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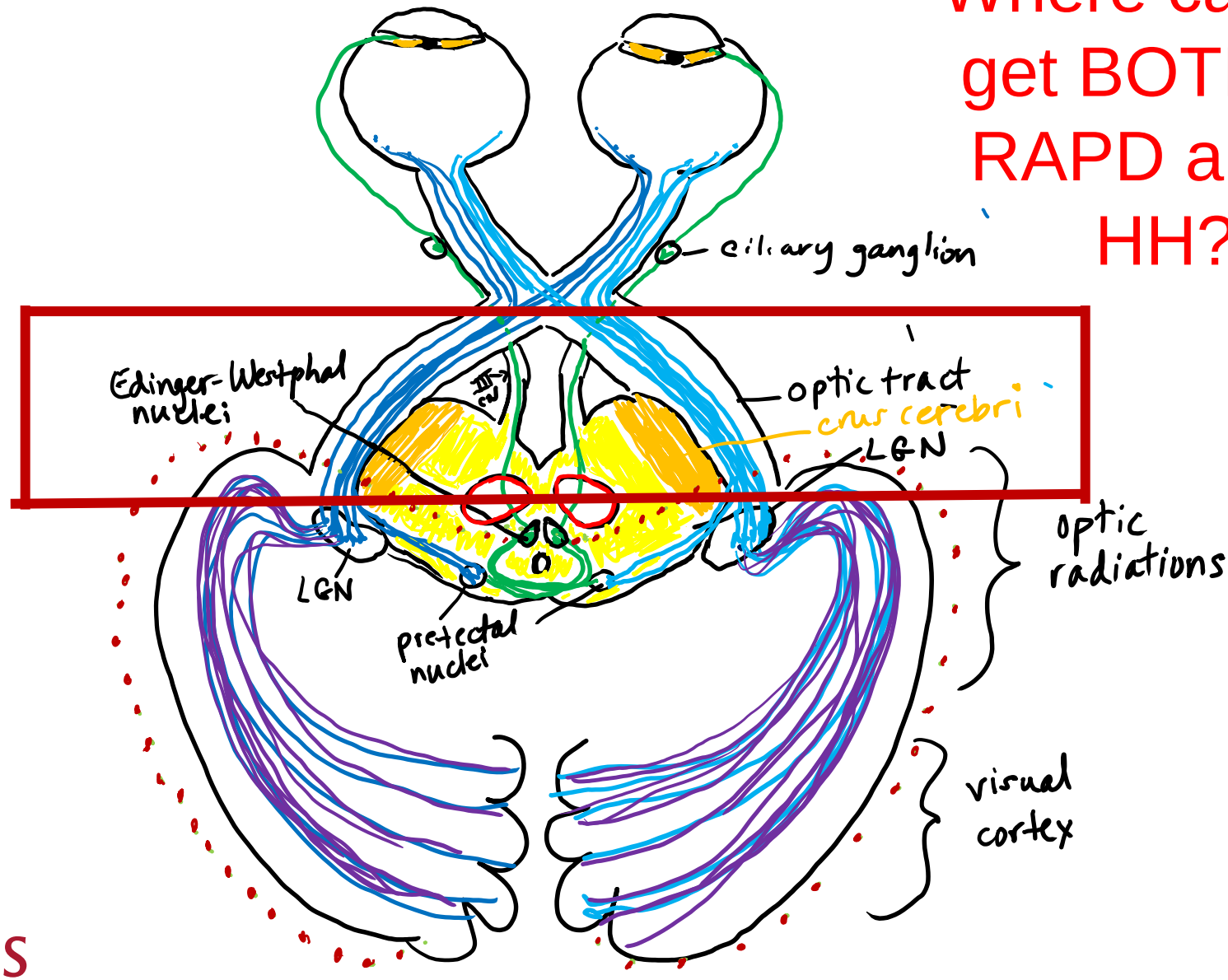
Super-imposed Light, Near, and Visual Pathways

Where can we get BOTH an RAPD and a HH?



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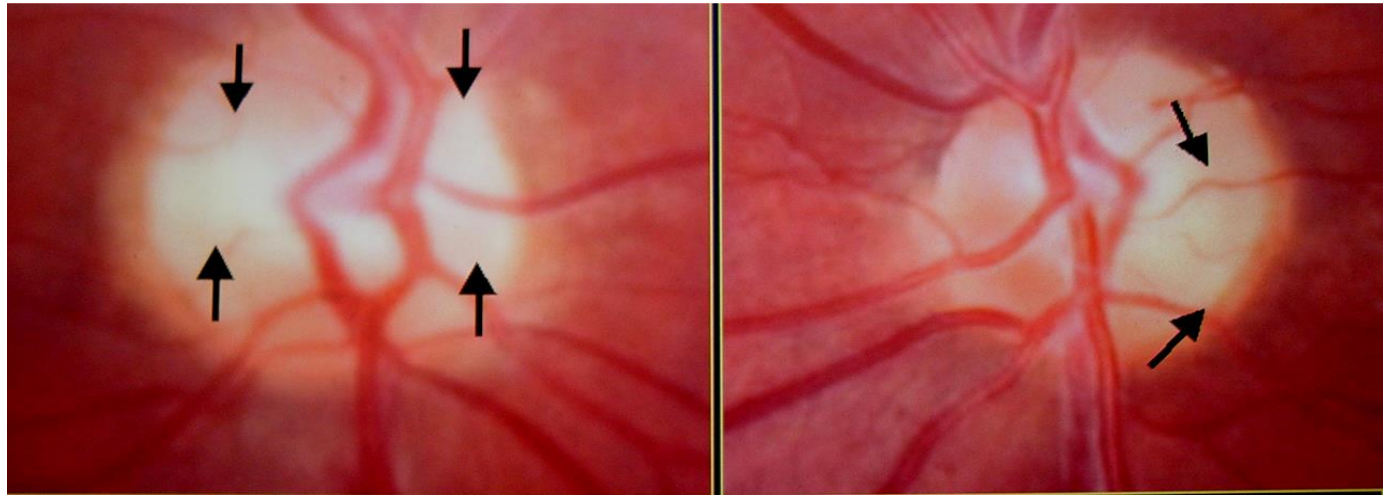
Optic Tract

- **Comprised of the axons of retinal ganglion cells**
  - Most synapse in LGN
  - Some synapse in pretectal nucleus
- **Lesions here can result in:**
  - Homonymous hemianopia
  - RAPD
  - NRR pallor



Band or "bow-tie" optic atrophy

Temporal pallor or atrophy

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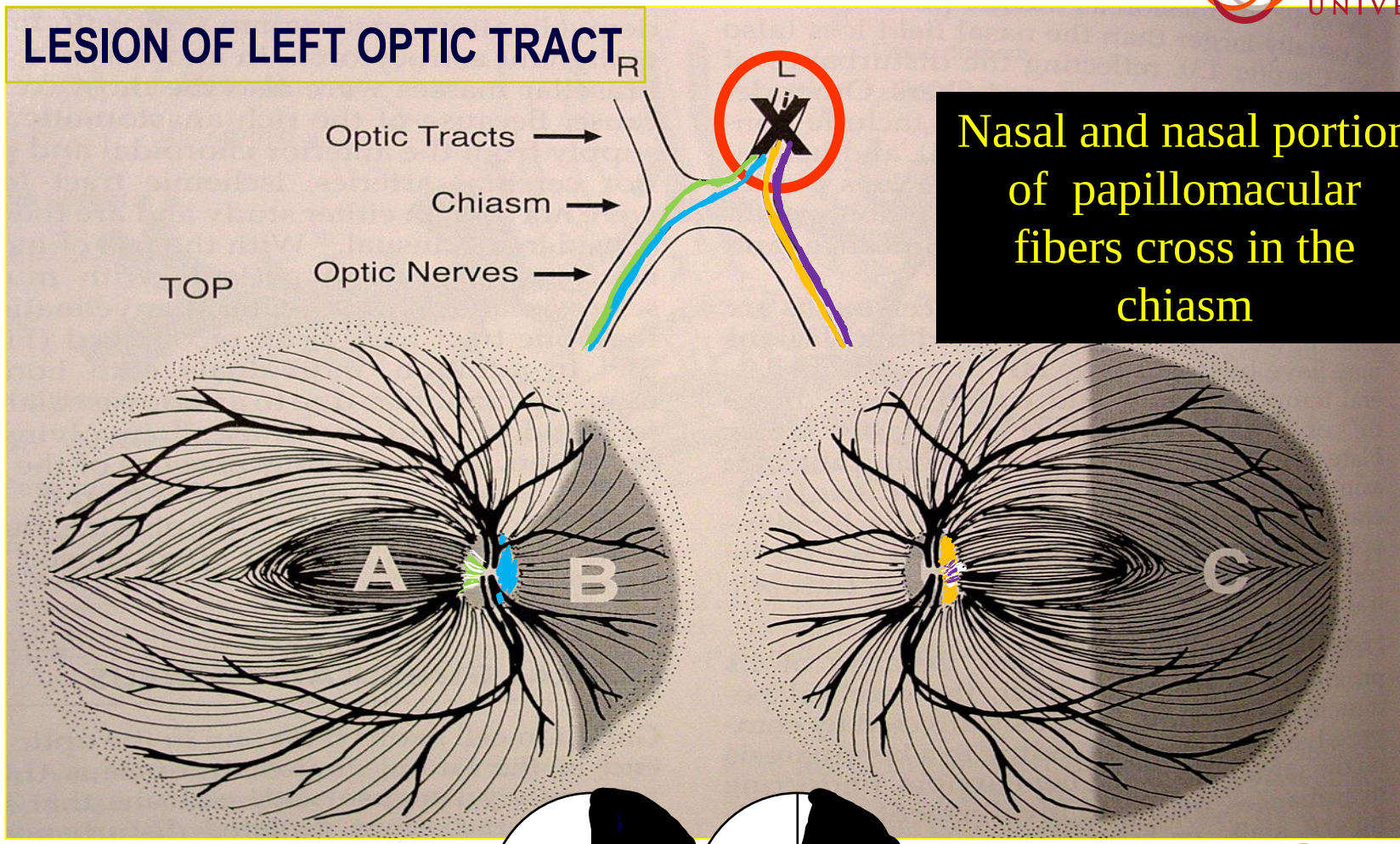
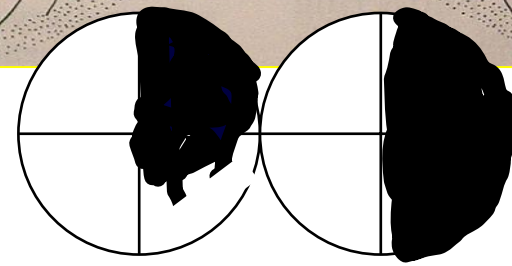


Small RAPD OD



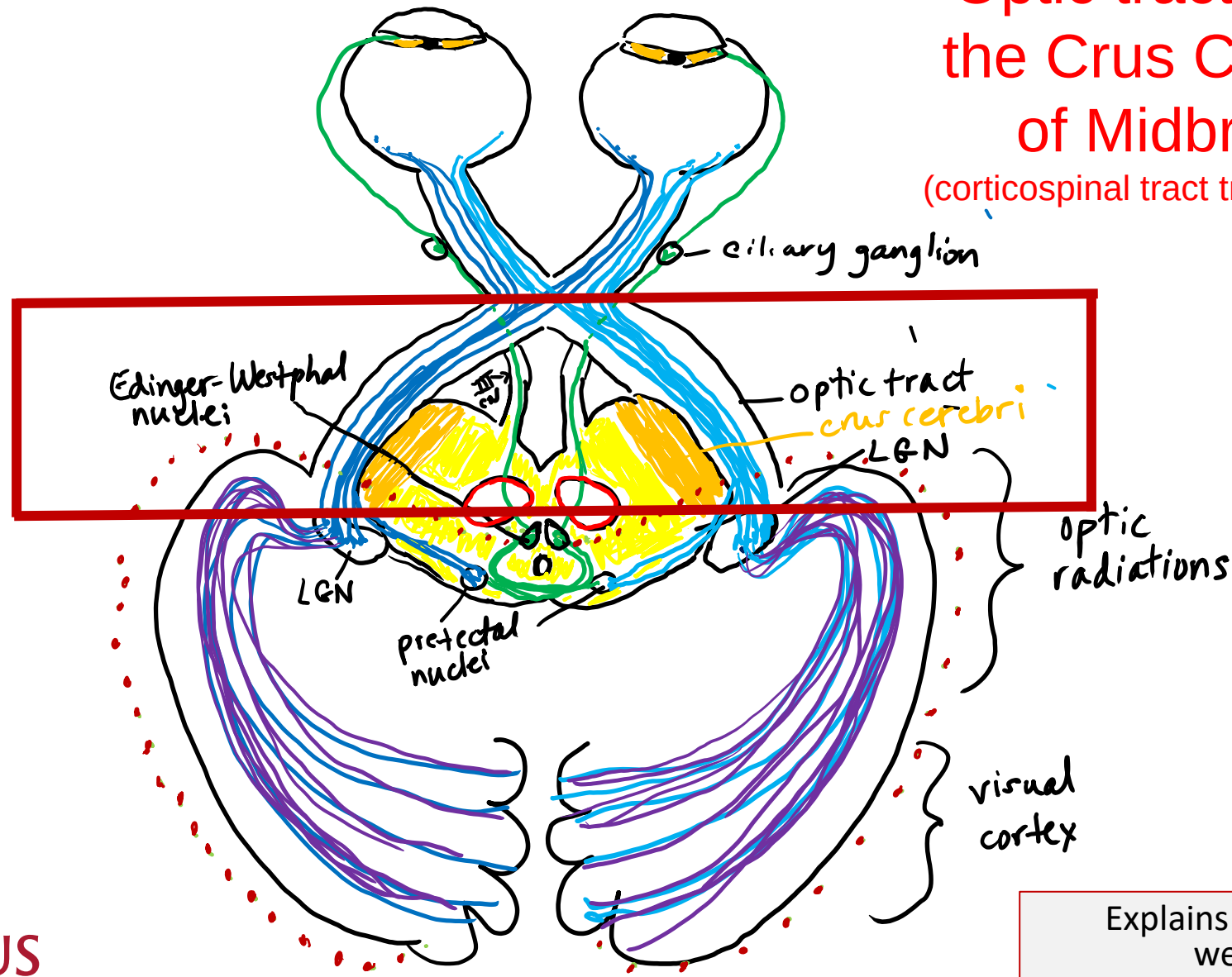
● = temporal fibers
● = nasal fibers
● = nasal macular fibers
● = temporal macular fibers

INCONGRUOUS RIGHT HOMONYMOUS HEMIANOPIA

Super-imposed Light, Near, and Visual Pathways

Optic tract hugs the Crus Cerebri of Midbrain

(corticospinal tract travels here)



Explains contralateral weakness

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What Is Our Work-Up?

- We need to look at this anatomical region
- Concern of damage to left optic tract
    - Neuro-imaging
    - MRI is preferable to CT
    - Contrast should be used if possible

CONCERN for MASS, STROKE

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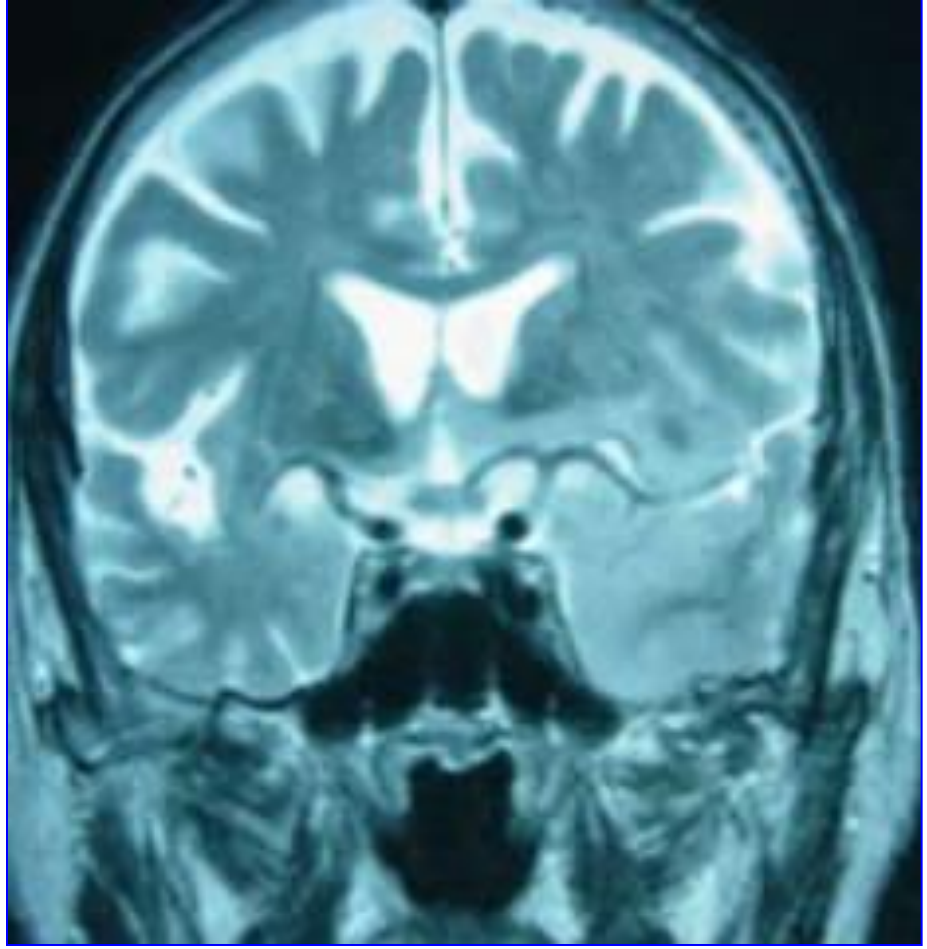
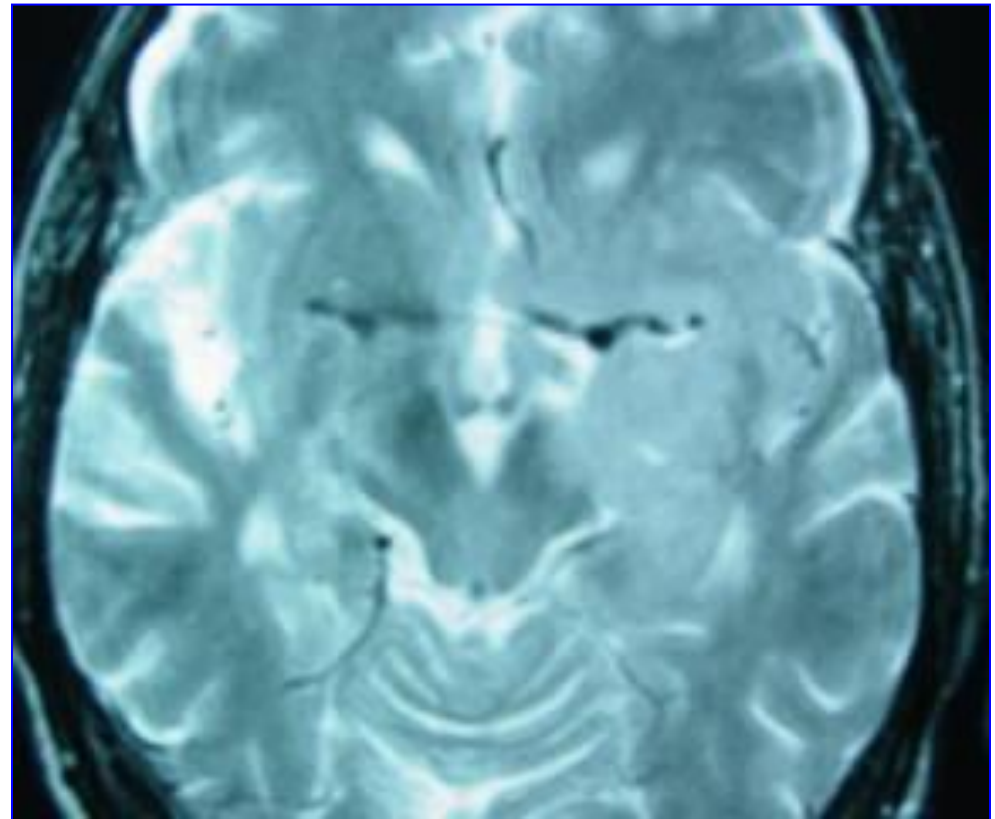


MRI “NORMAL”



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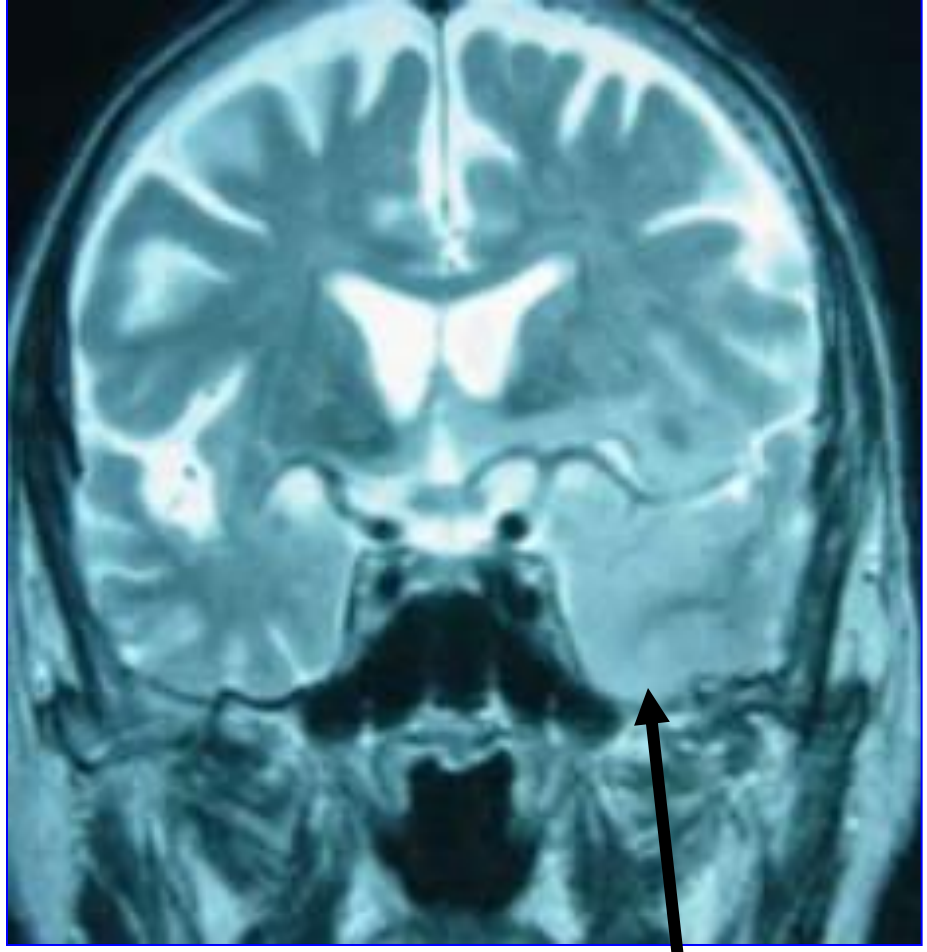
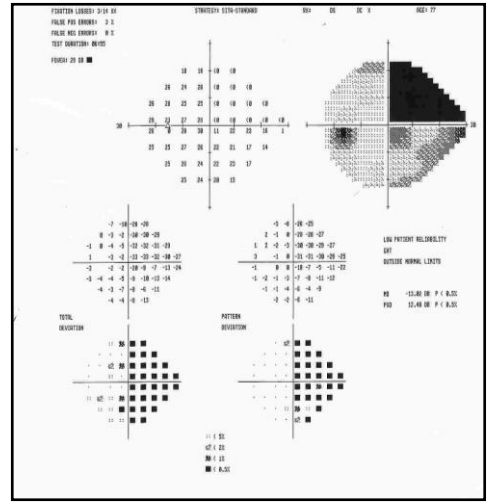
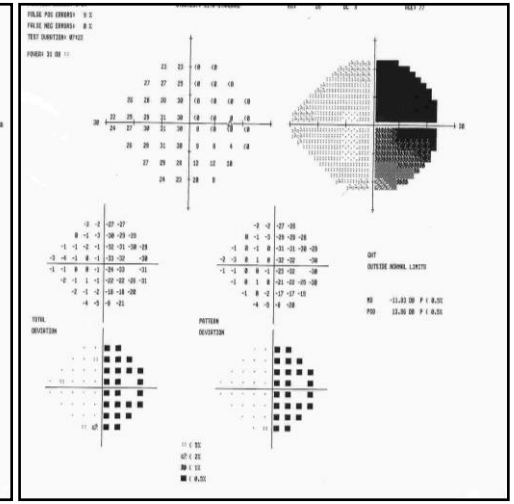
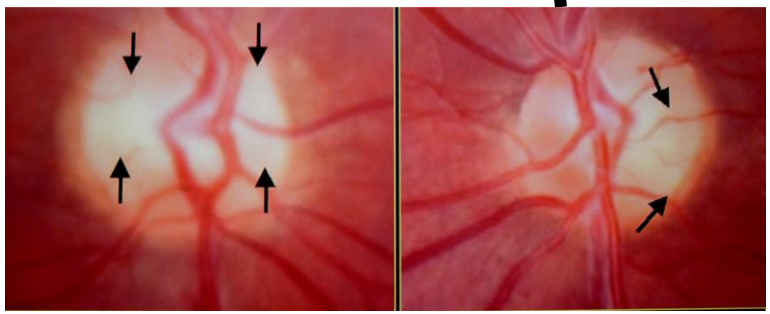
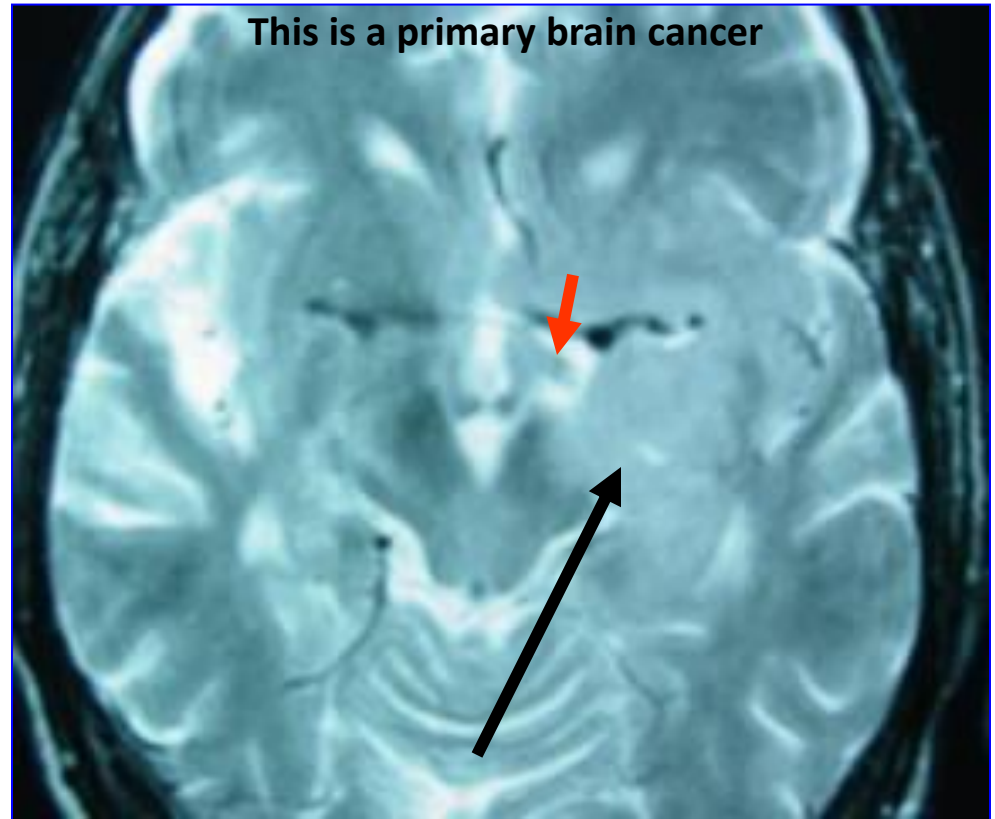






Grade 4 Astrocytoma





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You Will See These Conditions More Frequently!

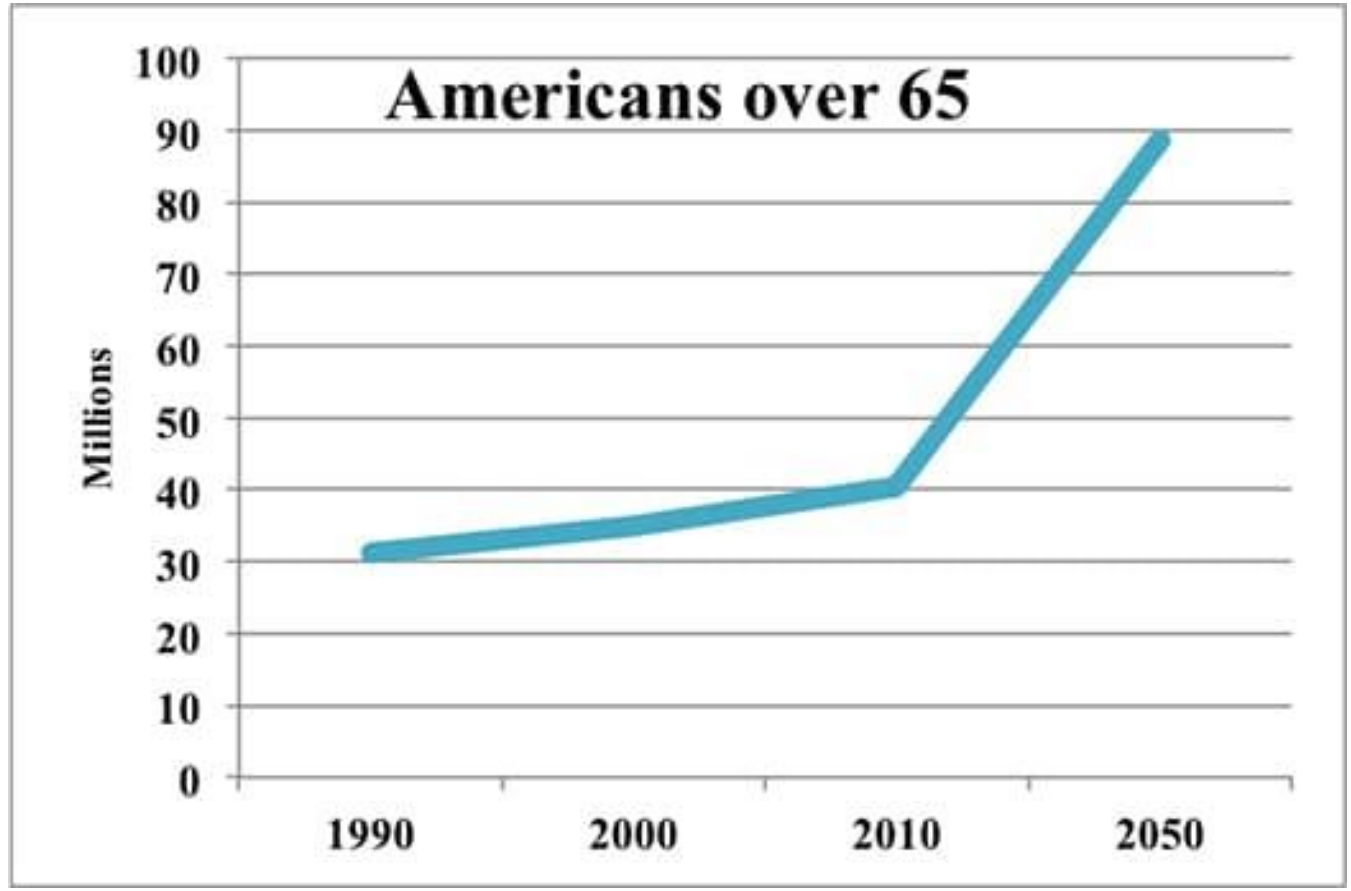
GCA

STROKE

CANCER

THYROID DISEASE

NEURO-DEGENERATIVE DISEASE



https://images.app.goo.gl/r8bhhxbv3q2ZxpZc8

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THANK YOU.

ANY QUESTIONS?

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