NEURO-OPHTHALMIC DISEASE CASES

Presented by Kelly A. Malloy, OD

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Dr. Malloy is a consultant and speaker for Osmotica Pharmaceuticals and RVL Pharmaceuticals, which has no association with anything in this lecture.

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CASE 1

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5 year-old girl

- Had routine eye exam 3 months ago, but was unable to stay for dilation.
- She is just here for the DFE
  - She has been getting occasional frontal headaches
  - They have not affected her daily activities
  - Already mentioned it to her pediatrician
  - Normally complains of headaches when at school (1x/month)
  - Only complained of headache once on a weekend, but she was in the mall and hungry

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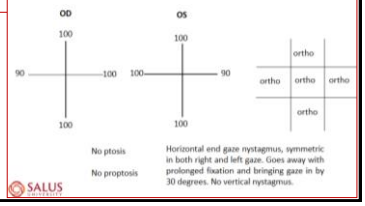
Afferent Exam Findings

- VA: OD 20/20 -
  OS 20/20-
- Pupils isocoric, (-)RAPD
- CF: full to finger count and red targets OU
- Color: OD 13/14 OS 13/14
- Unable to do HVF

Efferent Exam Findings



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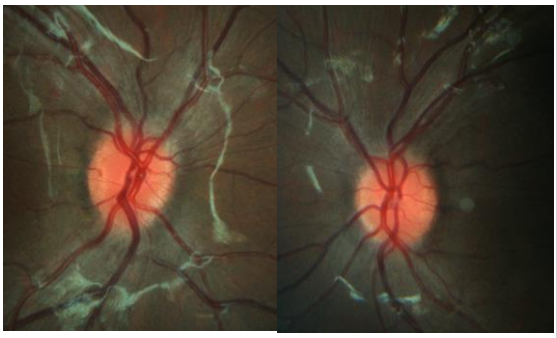
ONH Appearance (no SVP)



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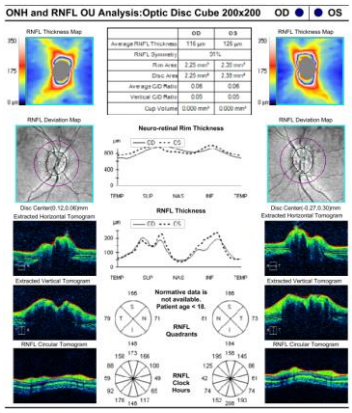
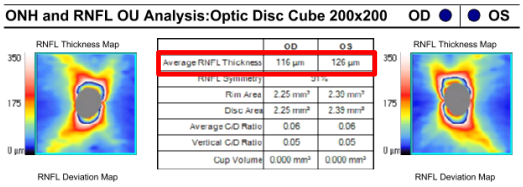
ONH OCT

There are no age-matched norms for children.

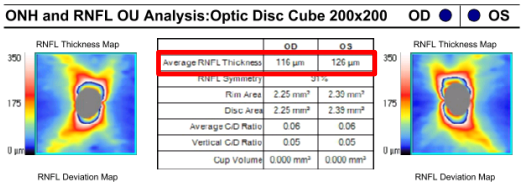




Average RNFL Thickness
OD 116um, OS 126um

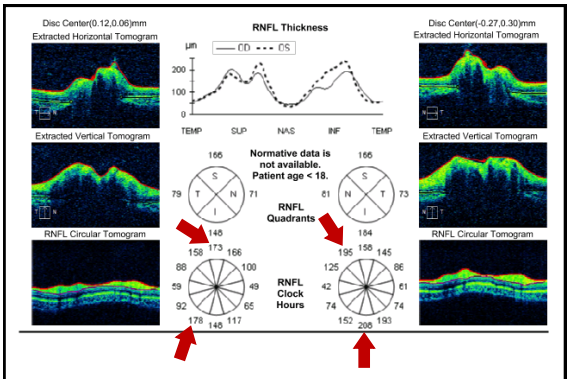
Yanni et al Am J Ophthalmol. 2013 February ; 155(2): 354–360

- 83 healthy North American **children aged 5-15**
  - attempt to determine normative data using the Spectralis SDOCT.
- Taking the 5th to 95th percentile data from these children as representing normal values
  - **mean peripapillary RNFL thickness 107.6um.**
- this is significantly higher than normative data for adults
- This difference is explained by the fact that even healthy individuals experience RNFL thinning with age.



Average RNFL Thickness
OD 116um, OS 126um

Normal mean peripapillary RNFL thickness 107.6um



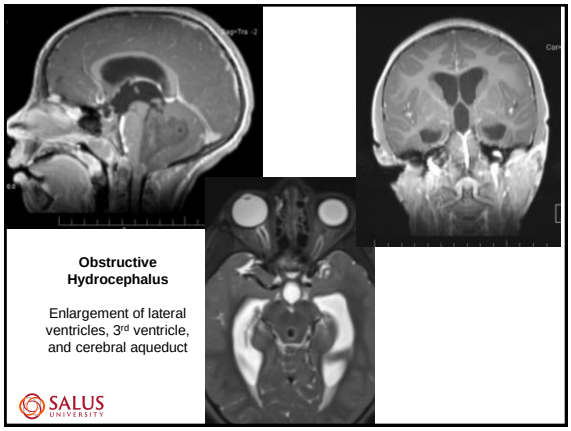
Inferior & superior nerve fibers swell first

Medulloblastoma with Drop Mets

PAPILLEDEMA IS PAPILLEDEMA

THE DEGREE OF SWELLING GIVES NO INDICATION OF THE ETIOLOGY

SO, EVEN MILD PAPILLEDEMA IS A MEDICAL EMERGENCY



Obstructive Hydrocephalus

Enlargement of lateral ventricles, 3rd ventricle, and cerebral aqueduct

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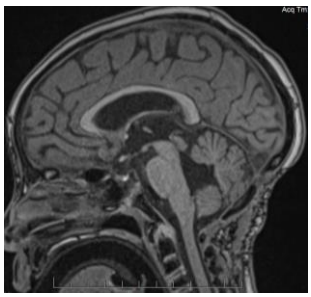
5 year old: One year post-surgery, radiation and chemo



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CASE 2

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A 40 year-old male emergently presents with progressive vision loss in his left eye.

- Onset 3 weeks ago, blur and darkening of vision
- No pain, headache, abnormal gait, dizziness, galactorrhea, acromegaly, skin or hair growth, weight gain, or weakness
- Reports recent occupational stress, decreased libido, and hypertension controlled by diet and exercise
- No medications reported
- No previous ocular diagnoses reported

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Pertinent Examination Findings:

- DVA sc: OD: 20/25-2, PH NI OS: 20/40-2, PH NI

- Pupils: 0.6 Log APD OS

- CVF:
OD: Mild peripheral temporal constriction with red desaturation ST>IT
OS: Severe temporal constriction with red desaturation

- Red Cap Desaturation: 90% desaturation OS

- Brightness Desaturation: 50% desaturation OS

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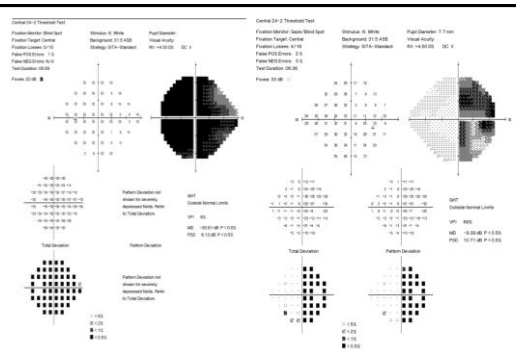

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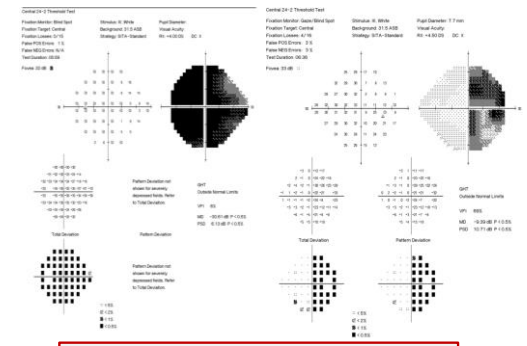

LEFT JUNCTIONAL SCOTOMA

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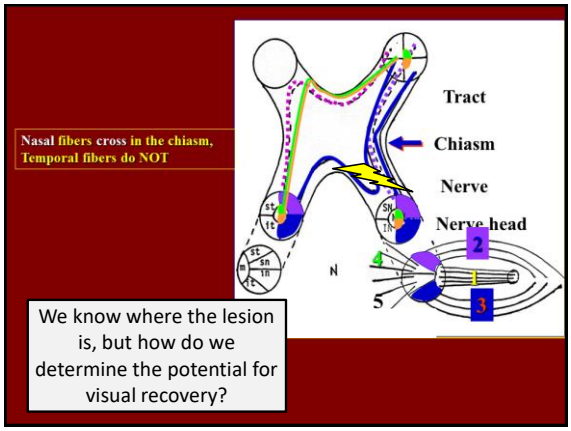
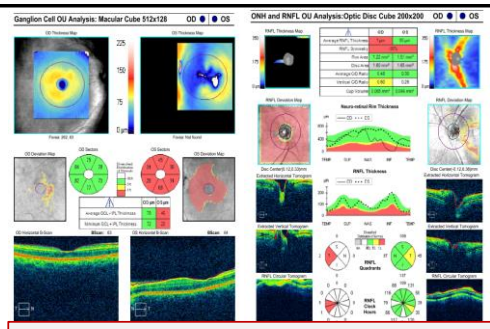


We know where the lesion is, but how do we determine the potential for visual recovery?



OCT can be helpful in determining the potential for visual recovery!

Is this a reliable OCT? Can it help us in predicting visual recovery in this case?

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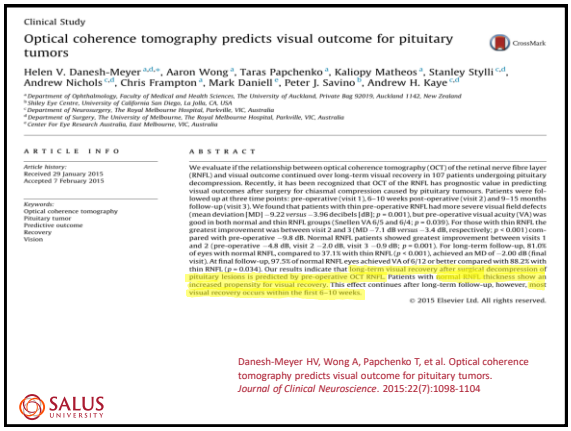

Clinical Study

Optical coherence tomography predicts visual outcome for pituitary tumors

Helen V. Danesh-Meyer, Aaron Wong, Taras Papchenko, Kaliopy Matheos, Stanley Stylli, Andrew Nichols, Chris Frampton, Mark Daniell, Peter J. Savino, Andrew H. Kaye

ARTICLE INFO

ABSTRACT

We evaluate if the relationship between optical coherence tomography (OCT) of the retinal nerve fibre layer (RNFL) and visual outcome continued over long-term visual recovery in 107 patients undergoing pituitary decompression. Recently, it has been recognized that OCT of the RNFL has prognostic value in predicting visual outcomes after surgery for chiasmal compression caused by pituitary tumours. Patients were followed up at three time points: pre-operative (visit 1), 6–10 weeks post-operative (visit 2) and 9–15 months follow-up (visit 3). We found that patients with thin pre-operative RNFL had more severe visual field defects (mean deviation [MD] −9.22 versus −3.96 decibels [dB]; $p = 0.001$), but pre-operative visual acuity (VA) was good in both normal and thin RNFL groups (Snellen VA 6/5 and 6/4; $p = 0.039$). For those with thin RNFL the greatest improvement was between visit 2 and 3 (MD −7.1 dB versus −3.4 dB, respectively; $p = 0.001$) compared with pre-operative −9.8 dB. Normal RNFL patients showed greatest improvement between visits 1 and 2 (pre-operative −4.8 dB, visit 2 −2.0 dB, visit 3 −0.9 dB; $p = 0.001$). For long-term follow-up, 81.0% of eyes with normal RNFL, compared to 37.1% with thin RNFL ($p < 0.001$), achieved an MD of −2.00 dB (final visit). At final follow-up, 97.5% of normal RNFL eyes achieved VA of 6/12 or better compared with 88.2% with thin RNFL ($p = 0.034$). Our results indicate that long-term visual recovery after surgical decompression of pituitary lesion is predicted by pre-operative OCT RNFL. Patients with normal RNFL thickness show an increased propensity for visual recovery. This effect continues after long-term follow-up, however, most visual recovery occurs within the first 6–10 weeks.

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Danesh-Meyer HV, Wong A, Papchenko T, et al. Optical coherence tomography predicts visual outcome for pituitary tumors. *Journal of Clinical Neuroscience*. 2015:22(7):1098-1104

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Optic disc appearance can also be helpful in determining the potential for visual recovery!

What do you think about potential visual recovery based on this optic disc appearance?
Does that, in turn, tell us anything about the urgency of surgical intervention?

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WHICH IS IT?

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Based on the VF defect, we localize this to the sellar region.

The VF loss is dense and both superiorly and inferiorly, so we know the lesion is large.

The OCT tells us that although there is compression, it is very likely that the visual function can improve significantly with surgical intervention.

Surgery needs to be done before the vision loss becomes irreversible.

So, this becomes more urgent…

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When should neurosurgery be contacted urgently or emergently?

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When should neurosurgery be contacted urgently or emergently in regard to sellar lesions?

Answers:

1. Acute visual loss
2. Apoplexy (Sudden onset severe HA, vision decline, lethargy)
3. Significant endocrinopathy with fatigue, hypotension (hypocortisolism, hyponatremia, hypothyroid)

How should that be done?

Get acquainted with your local neurosurgeon.

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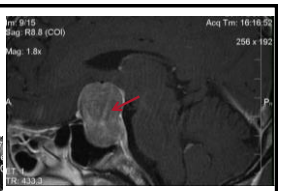
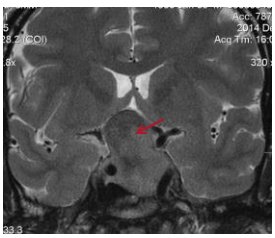
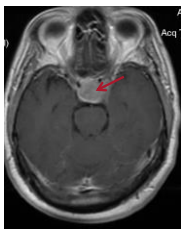
Large Pituitary Macroadenoma







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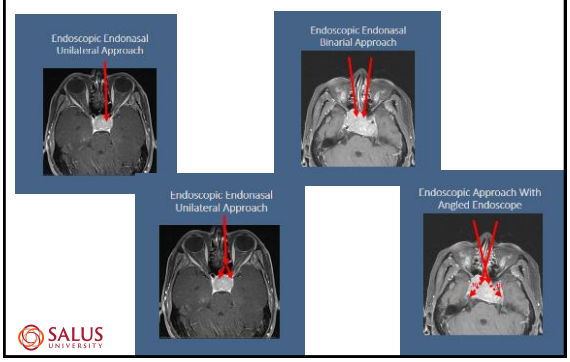
Surgical Treatment



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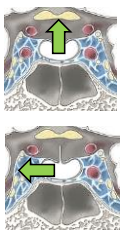
Pituitary Macroadenoma

Post-operative Management

- Vision → Afferent
- Fluid balance (DI) → Efferent
- Hypocortisolism
- Hyponatremia / SIADH
- Sinonasal issues



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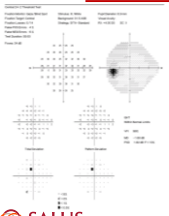
Underwent endoscopic transnasal resection within 24 hours of presenting to The Eye Institute Emergency Service.

Post-surgical testing showed somewhat low am cortisol levels
Started on Decadron 0.5mg.
Scheduled with endocrinology.

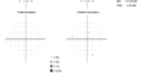
One Month Follow-Up Eye Exam:



VA OD 20/20 OS 20/20

Color: OD 14/14 OS 14/14

CF: Normal OU



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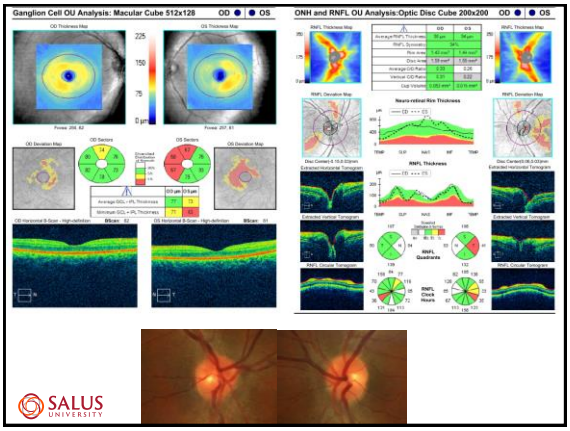

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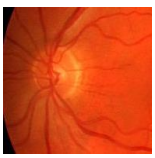
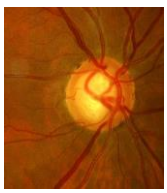
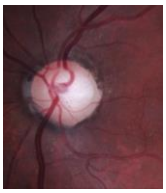
CHIASMAL SYNDROME

- PAINLESS, ASYMMETRIC VISUAL LOSS
- OPTIC NERVE DYSFUNCTION (tests of afferent function)
- VF RESPECT FOR THE VERTICAL
- ACQUIRED CUPPING & NEURORETINAL RIM PALLOR ****(50% HAVE NORMAL ONH)*







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If Pituitary Adenoma is Suspected but Without Acute Vision Loss

Both imaging and pituitary function lab testing should be obtained prior to the neurosurgical consult (except in emergent cases).

- Prolactin
- AM Cortisol
- ACTH (adrenocorticotropic hormone)
- IGF-1 (insulin-like growth factor 1)
- GH (growth hormone)
- TSH (thyroid stimulating hormone), fT4
- LH, FSH (luteinizing hormone, follicle stimulating hormone)
- Testosterone / Estrogen

* Random cortisol and ACTH if possible

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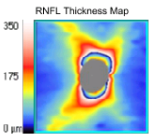
IMPORTANCE OF OCT TESTING

- The last 2 cases demonstrated how the OCT can be critical in both diagnosis an determination of treatment / timeframe of treatment
- Be aware of OCT norms and artifacts
- Use your OCT to its full potential!



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CASE 3

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54 Year-Old Woman

- Referred by OD for evaluation /management of presumed optic neuritis
- Patient awoke this am with reduced vision OS
  - Vision loss persists into afternoon
- Denies eye pain (even with movements) or headache, or any other symptoms

- Systemic history is unremarkable.
- No DM, HTN, hypercholesterolemia
- Does not know her family history
- No tobacco, alcohol or drug use

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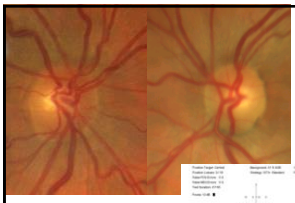
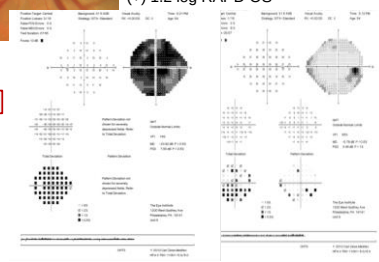


Exam Findings

VA: OD 20/20 OS 20/50
Color: OD 14/14 OS 2/14
Pupils:
Equally anisocoric in bright & dim
(+) 1.2 log RAPD OS



So, is this optic neuritis?

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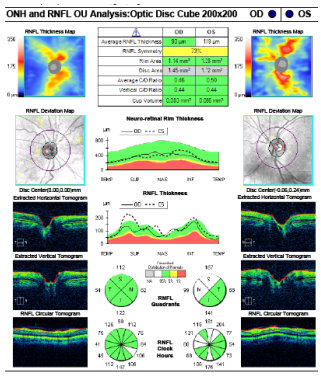
- There is some RNFL elevation
- Does this mean optic neuritis?



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Let's look a little more closely…

So, is this optic neuritis?

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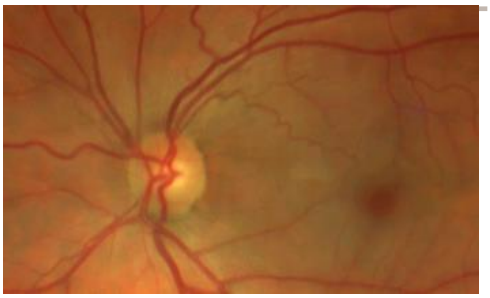
What about now?



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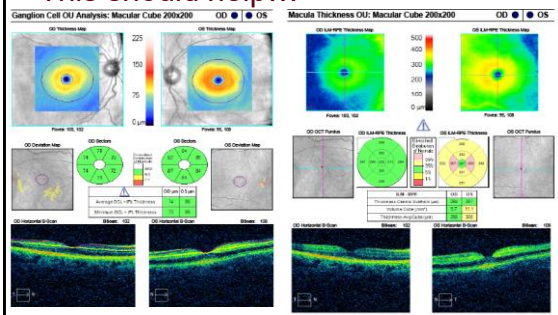
This should help…



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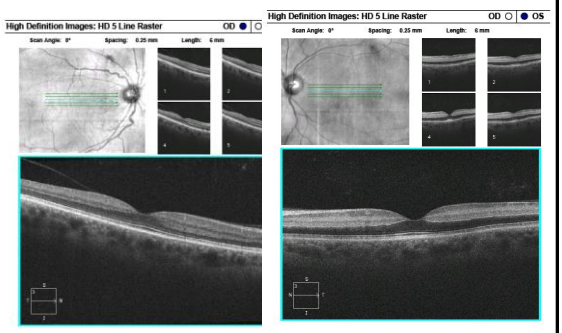

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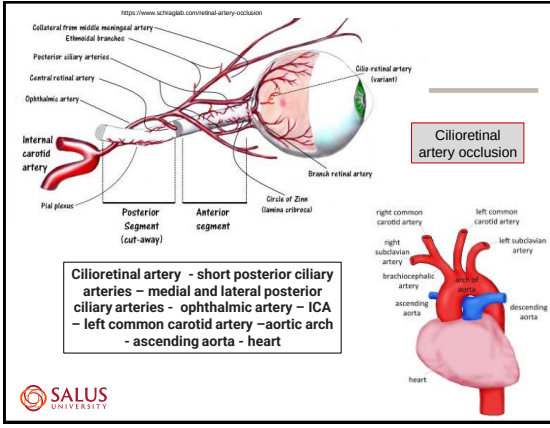

Cilioretinal artery occlusion

Cilioretinal artery - short posterior ciliary arteries – medial and lateral posterior ciliary arteries - ophthalmic artery – ICA – left common carotid artery –aortic arch - ascending aorta - heart

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Assessment & Plan

- What needs to be done?
- How urgent is the work-up?

Is This An Emergency?

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Assessment & Plan

- What needs to be done?
  - TREAT THIS LIKE AN ACUTE STROKE
    - Neuroimaging to rule our stroke
    - Cardiac eval to rule out MI / other heart / vascular issues
    - Labs
- How urgent is the work-up?
  - Emergent – to nearest ED with a stroke center

MEDICAL EMERGENCY

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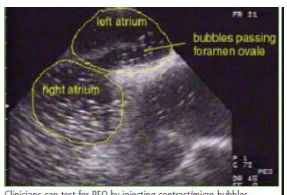
- **Acute retinal artery occlusion IS an acute stroke.**
- **Send to ER to r/o other infarcts to brain (MRI)**
- Work-up to otherwise include:
  - Carotid circulation evaluation
  - Cardiac evaluation
  - Lab testing
  - **Bubble Study**
  - Others

MEDICAL EMERGENCY



Clinicians can test for PFO by injecting contrast/micro bubbles (agitated saline) intravenously and monitoring their passage through the atria.

DX: Retinal artery occlusion secondary to patent foramen ovale (PFO).

TX: Blood thinners vs PFO closure

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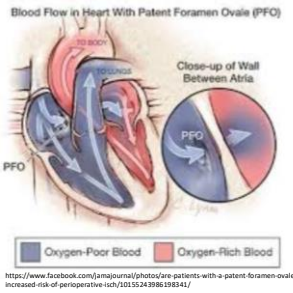

https://www.facebook.com/jamajournal/photos/are-patients-with-a-patent-foramen-ovale-at-increased-risk-of-perioperative-isch/10155243986198341/

This hole allows small venous blood clots, which would normally get filtered by the lungs, to get into the arterial blood stream, causing a cerebral stroke or a retinal artery occlusion.

Patent foramen ovale (PFO) [or atrialseptal defect] needs to be considered in all cases of cryptogenic, or unexplained, stroke.

Treated with anticoagulant therapy or surgical device closure.

AMPLATZER™ PFO occluder device (Abbott) is the only device approved by the US Food and Drug Administration for transcatheter closure of a PFO.

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CASE 4

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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

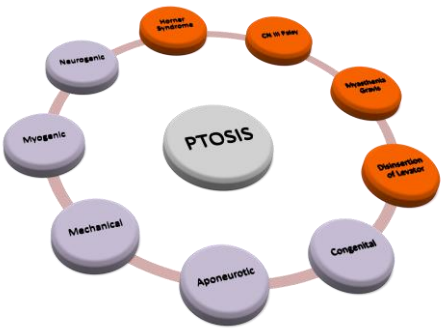
- Systemic history
  - Hypertension x 30 years
  - Hypercholesterolemia
  - Arthritis
  - Ovarian cancer 30 years ago

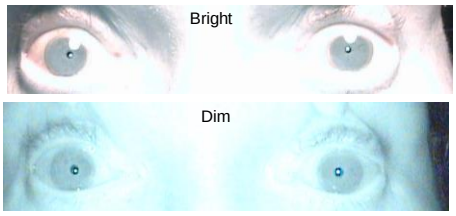
Exam Findings

- 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

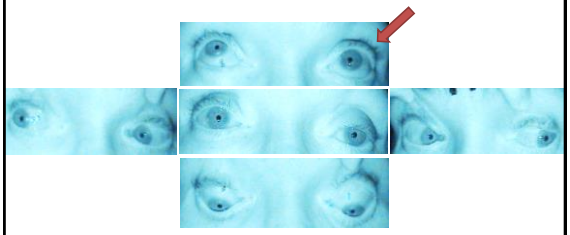




Pupils Isocoric (not suggestive of autonomic NS problem)

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.

Ocular Motility



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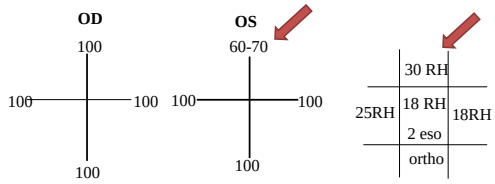
Ocular Motility



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

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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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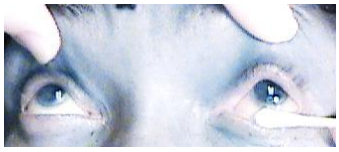

The eye moved up somewhat, but not fully. We called it an equivocal forced duction test.

There was no torsion on double Maddox rod testing.

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WHICH IS IT?

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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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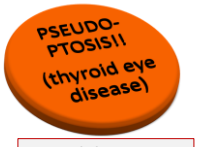
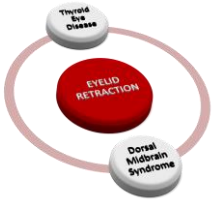
What about the ptosis?



It was lid retraction in the fellow eye!



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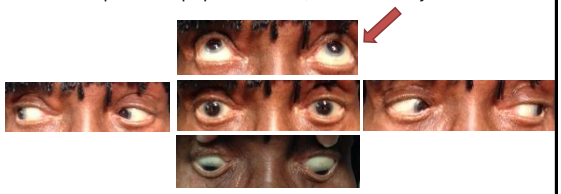
6 week follow-up

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



MILD TED
58% regress with achieving a euthyroid 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
- **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

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

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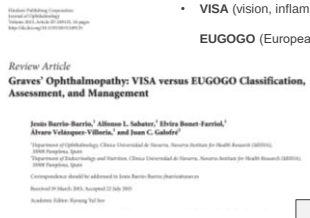
HOW TO CLASSIFY AND STAGE TED TO DETERMINE THE PROPER MANAGEMENT PLAN (newer classifications)

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

EUGOGO (European Group of Graves' Orbitopathy)



Review Article

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

Jesús Barrio-Barrio,[1] Alfonso L. Sabater,[2] Elvira Bonet-Farriol,[3] Álvaro Velázquez-Villoria,[4] and Juan C. Galofré[5]

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

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

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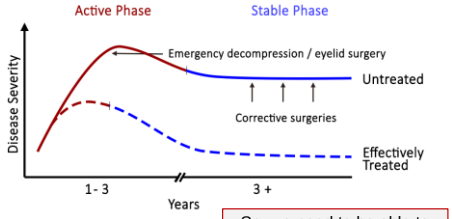
Rundel's Curve

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, 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: 33477417 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 5

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30 yr-old woman with diplopia

- Diplopia noticed 1 month ago
- Constant horizontal diplopia
- Intermittent headache and eye pain
- Had eye exam 2 weeks ago – given glasses to correct astigmatism
  - Diplopia remains
- Other symptoms: fatigue, coldness (anemia)

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- Medical Hx:
  - 2 full-term pregnancies and births
  - Severe anemia after 2nd child, requiring a transfusion
    - Has not been using iron regularly
  - On Depa-Provera (medroxyprogesterone injection q 3 months to inhibit ovulation)
  - Notes occasional numbness in L hand x 1 month
  - Fam HX: +Lupus

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- BCVA: OD 20/20 OS 20/20
- Color: 7/7 OD, 7/7 OS
- PERRL (-) RAPD OS
- CF: full OU
- Palpebral apertures: 9 mm OD and 9 mm OS
- Exophthalmometry: 21 mm OD and 20 mm OS
- Normal anterior segment health OU
- Normal GAT, BP
- Normal DFE
- Neurologic exam: ? Difficulty with tandem gait

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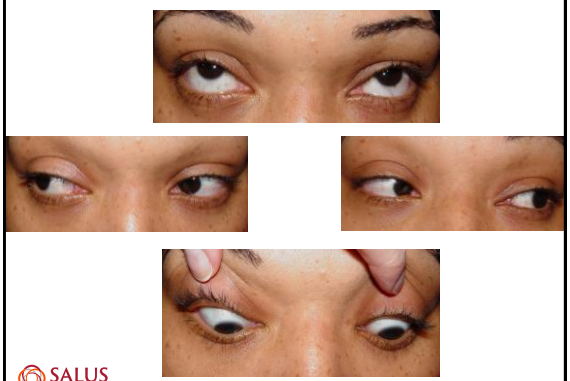

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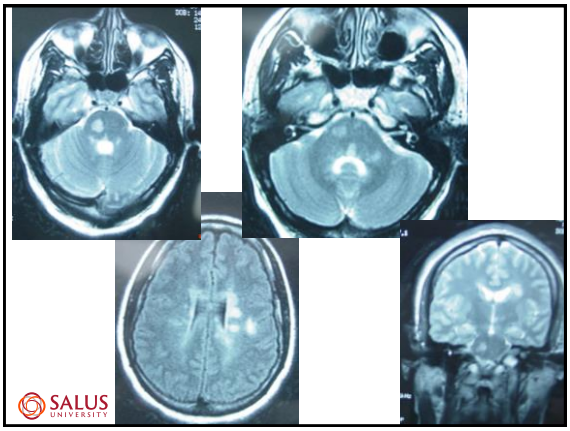
CT @ Distance

16 eso	20 eso	30 eso

Negative Forced Duction Test

Potential Labs for Diplopia

- CBC, platelet
- C-reactive protein, ESR
- Lyme titer (if + get Western blot IgG and IgM)
- ANA with reflex titer
- ACE
- RPR
- FTA-ABS
- Acetylcholine Receptor Antibodies (binding, blocking, modulating)
- Thyroid studies (TSH, T3, T4, thyroid stimulating immunoglobulin, thyroperoxidase antibodies, thyroglobulin antibodies)



WHAT ARE THE MRI DIAGNOSTIC CRITERIA FOR MS? MCDONALD CRITERIA (REVISED IN 2017)

NEED TO SHOW DISSEMINATION IN SPACE AND TIME!

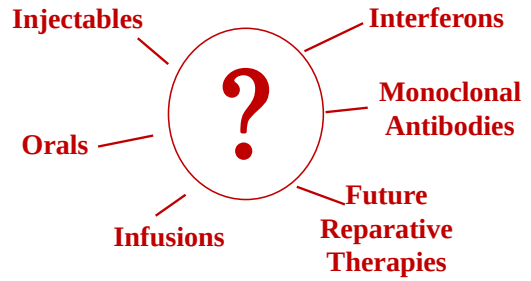
Summary of 2017 McDonald Criteria for the Diagnosis of MS

✓ Requires elimination of more likely diagnoses
✓ Requires demonstration of dissemination of lesions in the central nervous system in space and time

CLINICAL PRESENTATION	ADDITIONAL CRITERIA TO MAKE MS DIAGNOSIS
...in a person who has experienced a typical attack/CIS at onset	
• 2 or more attacks and clinical evidence of 2 or more lesions, OR • 2 or more attacks and clinical evidence of 1 lesion with clear historical evidence of prior attack involving lesion in different location	None. DIS and DIT have been met.
• 2 or more attacks and clinical evidence of 1 lesion	DIS shown by one of these criteria: - additional clinical attack implicating different CNS site - 1 or more MS-typical T2 lesions in 2 or more areas of CNS: periventricular, cortical, juxtacortical, infratentorial or spinal cord
• 1 attack and clinical evidence of 2 or more lesions	DIT shown by one of these criteria: - Additional clinical attack - Simultaneous presence of both enhancing and non-enhancing MS-typical MRI lesions, or new T2 or enhancing MRI lesion compared to baseline scan (without regard to timing of baseline scan) - CSF oligoclonal bands
• 1 attack and clinical evidence of 1 lesion	DIS shown by one of these criteria: - Additional attack implicating different CNS site - 1 or more MS-typical T2 lesions in 2 or more areas of CNS: periventricular, cortical, juxtacortical, infratentorial or spinal cord AND DIT shown by one of these criteria: - additional clinical attack - Simultaneous presence of both enhancing and non-enhancing MS-typical MRI lesions, or new T2 or enhancing MRI lesion compared to baseline scan (without regard to timing of baseline scan) - CSF oligoclonal bands
...in a person who has steady progression of disease since onset	
1 year of disease progression (retrospective or prospective)	DIS shown by at least two of these criteria: - 1 or more MS-typical T2 lesions (periventricular, cortical, juxtacortical or infratentorial) - 2 or more T2 spinal cord lesions - CSF oligoclonal bands

DIT = Dissemination in time
DIS = Dissemination in space
CNS = central nervous system
T2 lesion = hyperintense lesion on T2-weighted MRI
CSF = cerebrospinal fluid

MS MEDICATIONS



SOME MS MEDS HAVE BEEN ASSOCIATED WITH MACULAR EDEMA

- SPHINGOSINE 1-PHOSPHATE RECEPTOR MODULATORS
  - Fingolimod
  - Siponimod
  - Ozanimod
  - Ponesimod

THESE ARE ALL ORAL MEDICATIONS

- The American Academy of Ophthalmology has recommended a complete ophthalmologic exam **(ophthalmoscopy with evaluation for macular edema / OCT)**
  - at baseline
  - 3-4 months after medicine initiation
  - repeat evaluation 6 months later
  - then annually thereafter

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This case reminds us…

We tend to think of MS affecting the AFFERENT visual system.

MS effects the EFFERENT visual system as well!

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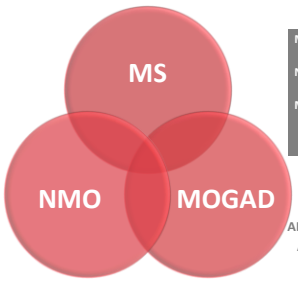
DEMYELINATING DISEASE DILEMMA



MS= MULTIPLE SCLEROSIS

NMO= NEUROMYELITIS OPTICA

MOGAD= MYELIN OLIGODENDROCYTE GLYCOPROTEIN ASSOCIATED DISEASE

ALL HAVE AUTOIMMUNE COMPONENT
ALL HAVE DEMYELINATING LESIONS
ALL AFFECT THE CNS
… SO, ALL CAN AFFECT THE OPTIC NERVE
BUT – TREATMENTS DIFFER! 93

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Work-up for Neurodegenerative Demyelinating Diseases

Multiple Sclerosis (Labs only to R/O other conditions)	NMOSD	MOGAD
(There is NO specific serum test for Multiple Sclerosis)	AQP4-IgG Cell based assay recommended	Anti-MOG antibody Cell based assay with full length human MOG target antigen
ACE -Angiotensin Converting Enzyme (sarcoid) FTA-ABS Fluorescent Treponemal Antibody test Absorption test (syphilis) RPR – Rapid Plasmin Reagin (syphilis) ANA - Antinuclear Antibody (autoimmune disease) Lyme titer or Western Blot (Lyme disease)	These should still be ruled out	These should still be ruled out
If any are positive, consider alternate etiology instead of MS or any other demyelinating disease.	**If positive**, then only 1 core clinical feature is required (optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, acute diencephalic clinical syndrome, symptomatic cerebral syndrome)	If positive, then one clinical event typically associated with MOG antibody is required (optic neurits, myelitis, ADEM, cerebral monofocal or polyfocal deficits, brainstem or cerebellar deficits, cortical encephalitis often with seizures)
If all negative (and no indication of NMOSD or MOGAD), then consider MS based on neuroimaging and revised McDonald Criteria demonstrating dissemination of CNS lesions in both space and time.	**If negative**, then 2 core clinical characteristics and their specific MRI qualifications (fulfilling dissemination in space) are required, one of which must be either optic neuritis, acute longitudinally extensive transverse myelitis, or area postrema syndrome.	**If negative**, and MOGAD still suspected, repeat the test (during an acute attack, a time of treatment-free intervals, or 1-3 months after plasma exchange, IVIg, or steroid treatment.

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New MOGAD Diagnostic Criteria - 2023

- Published in The Lancet Neurology on January 24th, 20231.
- Patients with low positive serum MOG-Ab titers can be diagnosed with MOGAD if they possess at least one supporting clinical or MRI feature.
  - bilateral simultaneous optic neuritis
  - longitudinally extensive spinal cord
  - optic nerve involvement
  - a conus lesion
- Supporting features can also be applied to patients with positive MOG-Ab results without reported titers and patients with negative serum but positive cerebrospinal fluid (CSF) MOG-Ab.

Overview of Neurodegenerative Demyelinating Diseases

	Multiple Sclerosis	NMOSD	MOGAD
Pathophysiology	Microglia activation and cascade of oxidative and mitochondrial injury	Antibodies targeting AQP4 water channels	Antibodies targeting MOG proteins on oligodendrocytes
Frequent MRI Optic Neuritis Features	Unilateral, short-segmented	Bilateral, long-segmented	Bilateral, long-segmented
Typical Visual Prognosis	Good (varies by subtype)	Poor	Good
Optic Neuritis Treatment	IVMP- followed by short oral taper	IVMP-followed by short oral taper Adjunct Therapy- IVIG and PLEX	IVMP- followed by long oral taper Adjunct Therapy- IVIG and PLEX
Demyelinating Disease Treatment	Immunomodulator	Immunosuppressant	Immunosuppressant

Overview of Neurodegenerative Demyelinating Diseases

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CASE 6

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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

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- **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

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- 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

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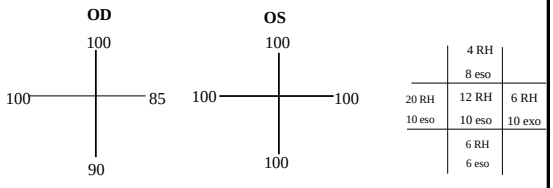
Ocular Motility



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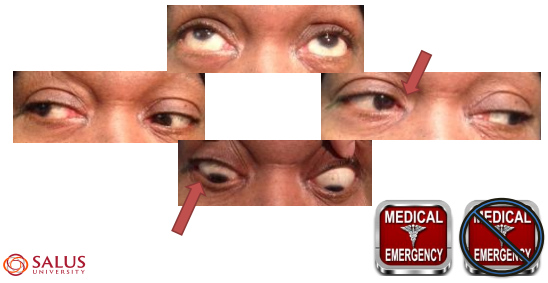
What is the cause of the adduction deficit and infraduction deficit?



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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

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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

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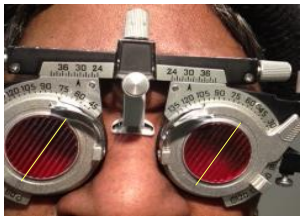

Double Maddox rod testing :
- 15-20 degrees of incyclotorsion OD
- 15-20 degrees of excyclotorsion OS

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	YOUNGER PATIENTS
• VASCULAR / STROKE	• DEMYELINATING

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

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CASE 7

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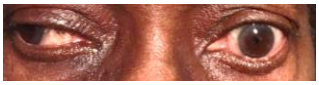
76 Year-Old Woman



- 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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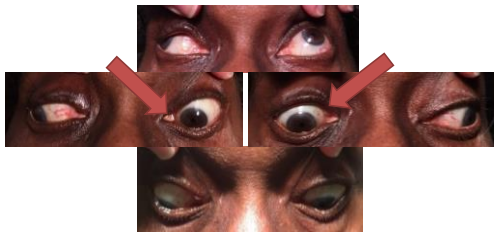
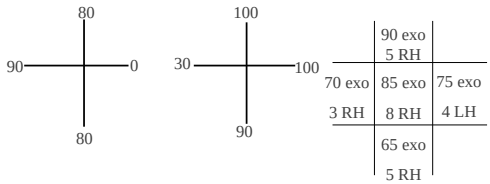
EXAM RESULTS

- 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

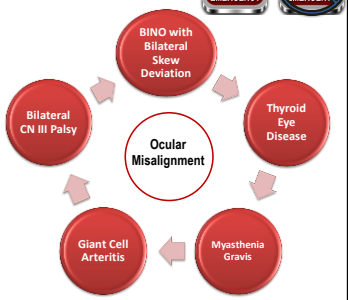
Ocular Motility



DDX FOR OCULAR MISALIGNMENT

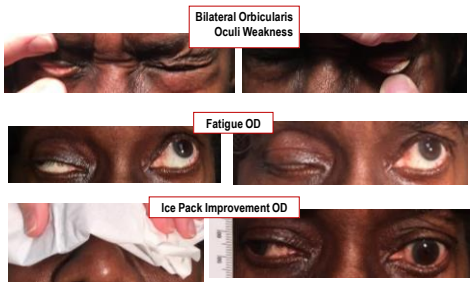
- Looks like a Bilateral Internuclear Ophthalmoplegia (BINO) with Bilateral Skew Deviation
  - Seen in MS or stroke
- What would be the management plan?
  - Send to the ER to rule out acute brainstem stroke

BUT…



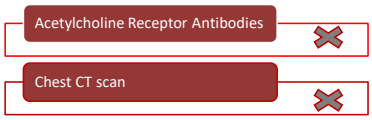
- **How do we explain the ptosis if this is a BINO and skew deviation?**

Additional In-Office Testing



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

- 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



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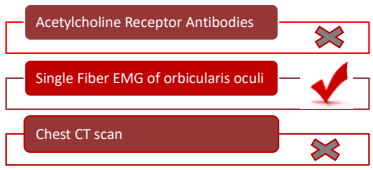
Work-Up for MG



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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

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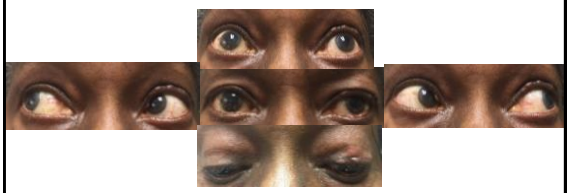
IMPROVEMENT ON FOLLOW-UP



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MG TREATMENTS

SYMPTOMATIC TREATMENT
- Acetylcholinesterase Inhibitors

CHRONIC IMMUNOSUPPRESSIVES
- Glucocorticoids
- NSAIDS

RAPID, SHORT-ACTING IMMUNOMODULATORS
- PLEX
  IVIG

SURGICAL TREATMENT
- Thymectomy

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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)

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

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CASE 8

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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

100 100 100 100 100 5 100

CT @ Distance

20eso 30eso 45eso

- 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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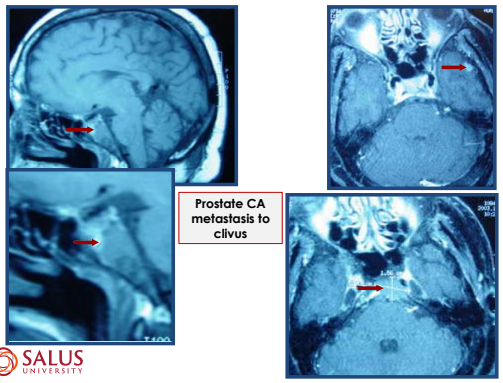

Prostate CA metastasis to clivus

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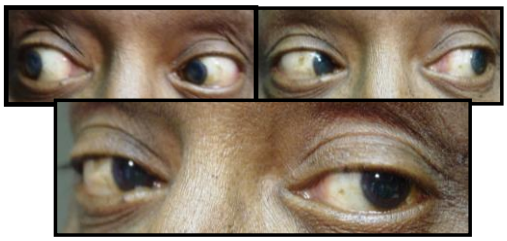


- S/P radiation treatment to involved areas
- Now 90 % normal abducting capacity OS

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

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.

CASE 9

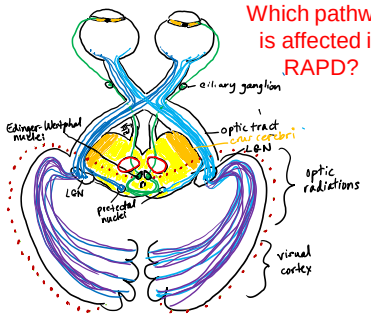
77 year old man

- 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?

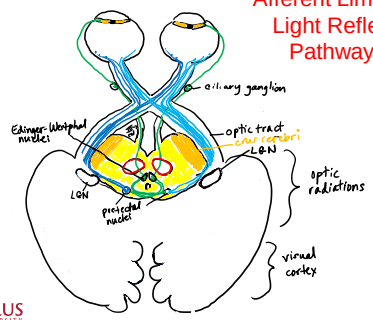
Super-imposed Light, Near, and Visual Pathways

Which pathway is affected in RAPD?



Pupillary Light Reflex Pathway

Afferent Limb of Light Reflex Pathway



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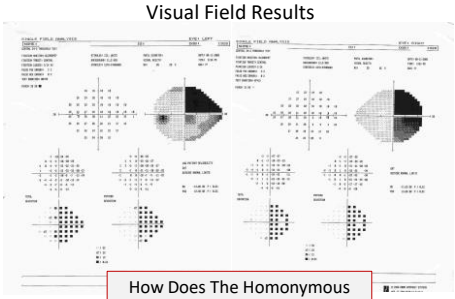
Visual Field Results



How Does The Homonymous Hemianopia Help With Localization?

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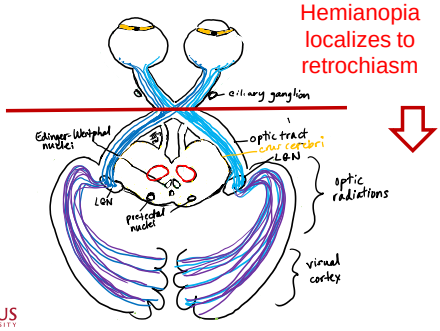
Visual Pathway

Homonymous Hemianopia localizes to retrochiasm



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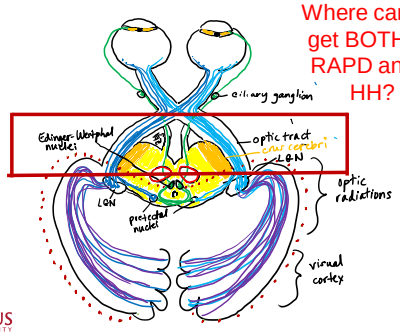
Super-imposed Light, Near, and Visual Pathways

Where can we get BOTH an RAPD and a HH?



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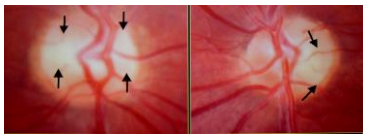
Optic Tract

- **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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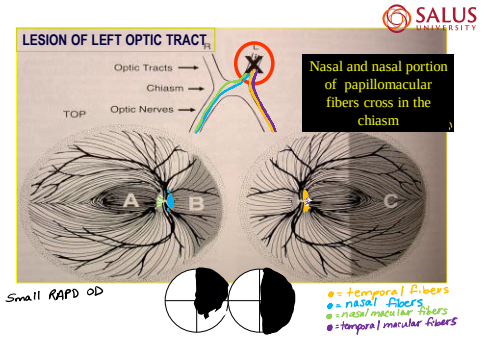
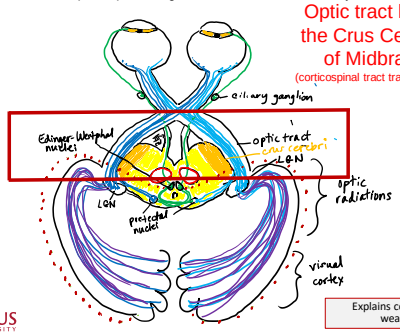


INCONGRUOUS RIGHT HOMONYMOUS HEMIANOPIA

Super-imposed Light, Near, and Visual Pathways



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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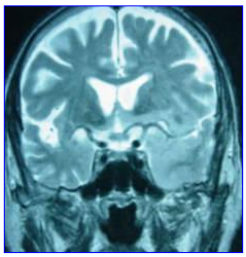
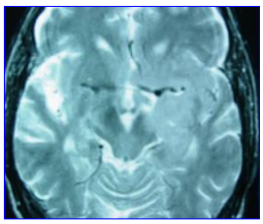


MRI "NORMAL"



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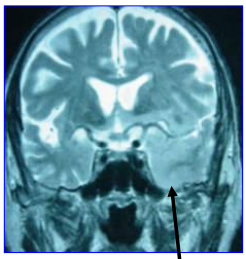
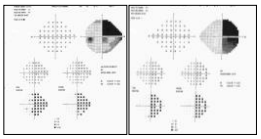
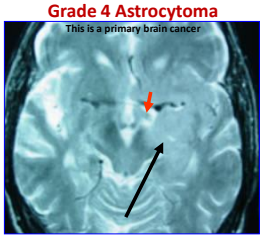




Grade 4 Astrocytoma

This is a primary brain cancer



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CASE 10

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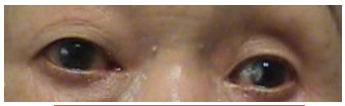
88 Year Old Woman



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

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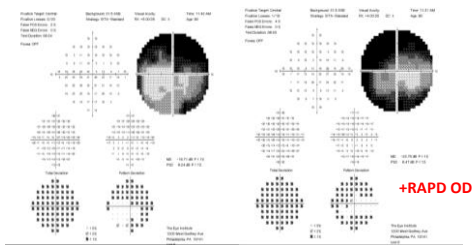

IOP: OD 17mm Hg, OS 11 mm Hg on treatment
DFE: OD 0.95/0.95, OS 0.95/0.95 (-)edema OU
Longstanding history of severe glaucoma OU

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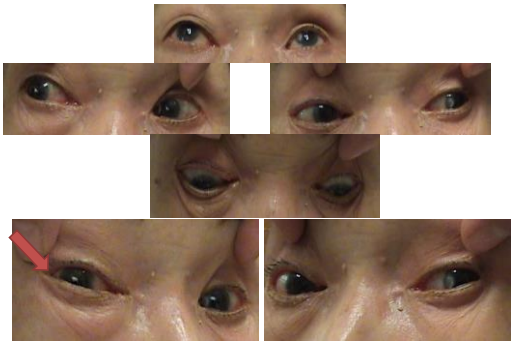

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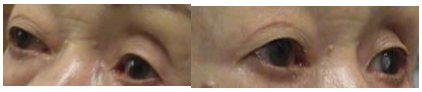

DDX of Painless Proptosis
- Lymphoma **
- Sarcoidosis
- Tuberculosis
- Granulomatous Polyangiitis (Wegener's)
- Other Orbital Mass (cystic, neoplastic, neural, vascular, fibrosing, lymphoid)
- IOIP (Pseudotumor)

Exophthalmometry: OD 13mm OS 9mm

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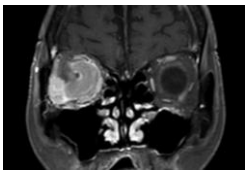
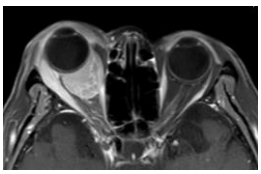
We ordered MRI

Orbital Lymphoma





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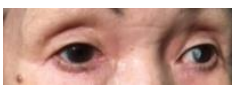
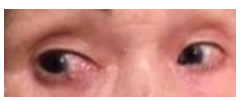
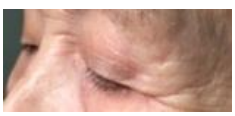
S/P biopsy & radiation







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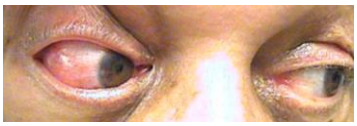
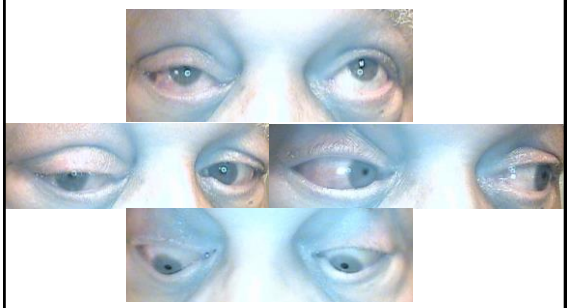
- Always consider cancer as an etiology for any afferent and efferent neuro-ophthalmic presentation!
- It could be a primary or secondary cancer…

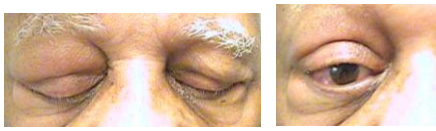
CASE 11

- 69 year-old man
- c/o right eye redness x 10 days
- No pain
- Horizontal diplopia in right gaze x 3 days





- VA: 20/20 OD, 20/20 OS
- Color: 14/14 OD, 14/14 OS
- PERRLA (-) RAPD
- CF: full OU
- Palpebral apertures: 5mm OD, 8mm OS
- Levator function: 15mm OD, 20mm OS
- Exophthalmometry: 25mm OD, 21mm OS



- Slit lamp exam
  - Grade 2 conjunctival injection OD
  - R upper lid edema
  - R lacrimal gland enlargement
- IOP: 20 OD, 18 OS
- BP: 135/80
- DFE: 0.2 x 0.2 cupping OU
- (-) edema (-) pallor

Remember the DDX for lacrimal gland enlargement from NOD2!

Knowing that helps to understand what questions you need to ask.

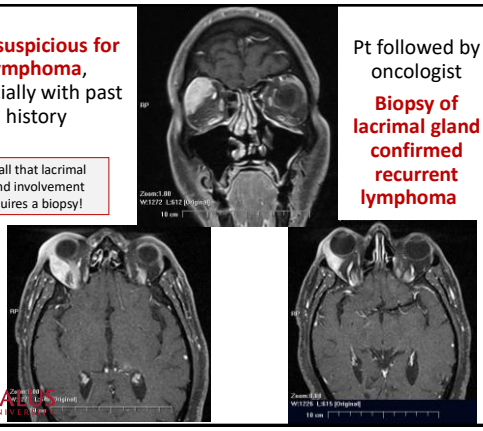
- History of Non-hodgkin's lymphoma 8 years prior
- affecting right leg/groin region(s/p surgery)

Once we know this information, lymphoma becomes our leading differential diagnosis.

So, what is our A/P?

MRI suspicious for lymphoma, especially with past history

Recall that lacrimal gland involvement requires a biopsy!



Pt followed by oncologist

Biopsy of lacrimal gland confirmed recurrent lymphoma

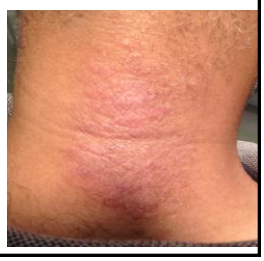
- Even when patients present due to a red eye, we need to look for early signs of proptosis or motility issues to suggest an orbital process.

CASE 12

48 YEAR-OLD MAN

- Swollen eyelid OD x 2 weeks
- Worse in the am
- Ocular irritation OD x 2 weeks prior
- Feels hard nodule on upper lid
- Right upper lid getting droopy
- (-) eye pain
- (-) headache
- (-) diplopia

- Rash around both eyelids, on and off x few years
- Now similar lesions on back of neck
- Denies any recent infections
- Had a cat with fleas (house fumigated)
- Was caring for a family member with HIV



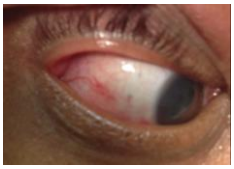
- Medical history unremarkable
- Social alcohol use
- Denies tobacco or drug use

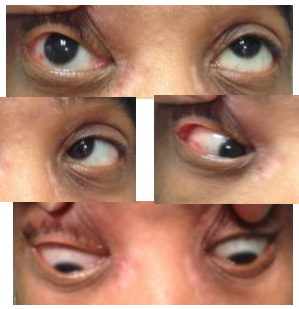
- Saw eye doctor last week
- Rx'd Keflex, Claritin and cold compresses
- No significant improvement

EXAMINATION RESULTS

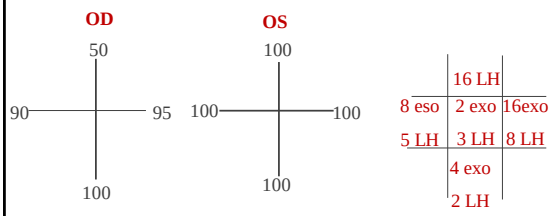
- VA: OD 20/25 OS 20/20
- Color (Ishihara): OD 0/14 OS 0/14
- Pupils: PERRL (-) APD OS
- CF: Full OU
- IOP: OD 23 mm Hg OS 19 mm Hg

Exophthalmometry: OD 24 mm, OS 20 mm
Palpebral apertures: OD 3 mm, OS 10 mm
Levator Function: OD 9 mm, OS 19 mm





OCULAR MOTILITY



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WORK-UP

- Recommend lab tests and imaging
- Pt was in the process of getting insurance
- Wants to wait until end of month to do testing; should have insurance then
- Refuses any work-up or additional referral
- Will do tests and return early next month

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FOLLOW-UP 2-3 WEEKS LATER

- Didn't do any tests
- Still no insurance; may take another month

- Skin lesions worsened
- Went to a free clinic; given ointment

- Still unable to fully open OD
- Vision remains good

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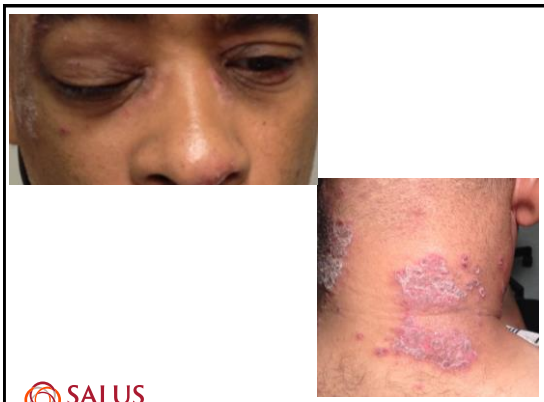

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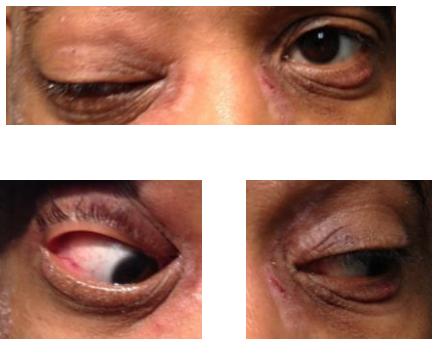

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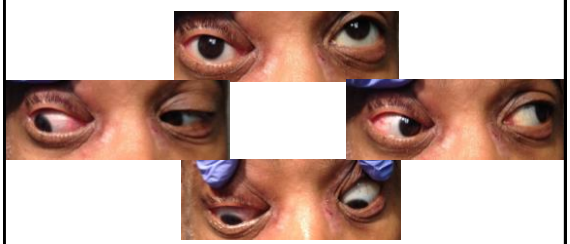

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Differential Diagnoses

NON-SPECIFIC ENHANCING ORBITAL MASS:

- Idiopathic Orbital Inflammatory Pseudotumor
- Orbital Lymphoma
- **Orbital Sarcoid**
- Orbital Tuberculosis
- Granulomatous Polyangiitis (a vasculitis)
- Eosinophilic Granulomatous Polyangiitis
- IgG4 and IgG4 related disease

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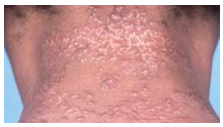
SARCOIDOSIS

- Inflammatory disease characterized by the growth of tiny collections of inflammatory cells (granulomas) in any part of the body
  - lungs
  - lymph nodes
  - eyes
  - skin
  - heart
  - brain
  - other organs



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- Needs work-up ASAP
- Need to R/O lymphoma, sarcoid, etc
- Since no insurance, patient went to ER

- Biopsy ultimately confirmed sarcoidosis

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Importance of Understanding Orbital Anatomy - Enlarged Lacrimal Gland and Skin Lesions

- Skin lesions give a clue as to the pathology; also easy access for biopsy
- Commonly see associated skin lesions in both sarcoid and Lupus
- Cutaneous involvement occurs in 20-35% of patients with systemic sarcoidosis

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CASE 13

39 year-old woman

- c/o facial asymmetry (superior nasal bump OD x 1 yr)
- Diplopia – is getting worse
- Told past eye doctor (a year ago) & PCP of symptoms
  - no work-up done (was told nothing to worry about!)
- S/p right endoscopic sinus surgery 10 years prior
- No history of thyroid dysfunction; health otherwise unremarkable

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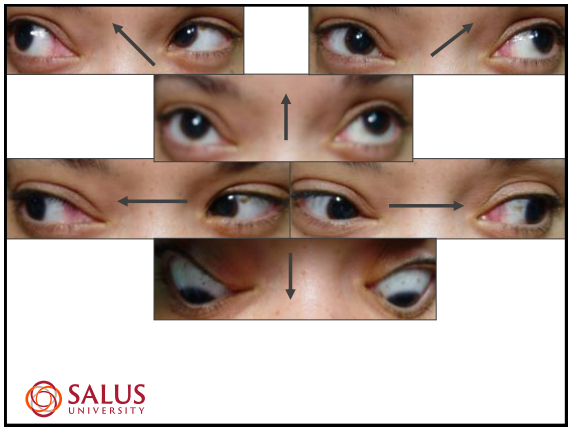
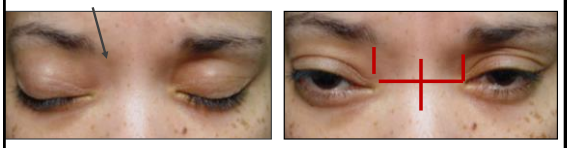
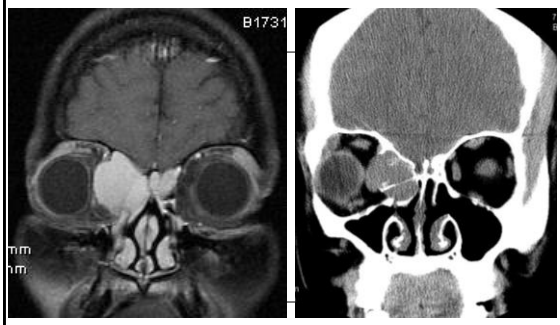
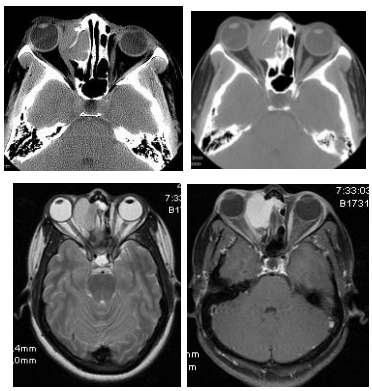


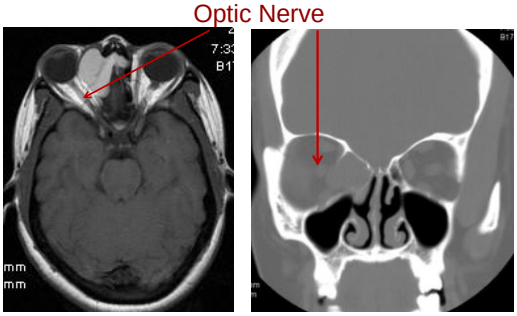


What is the necessary work-up?
Should we get an MRI or CT?



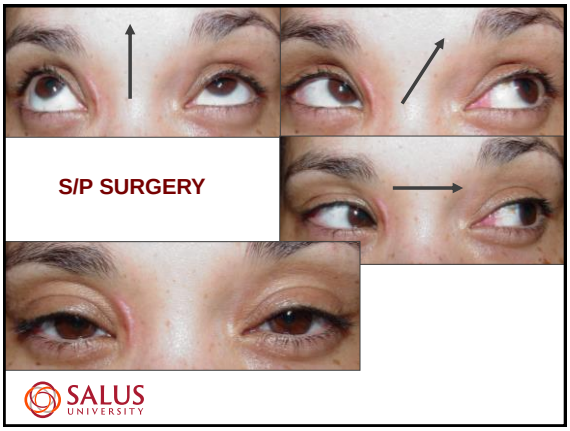


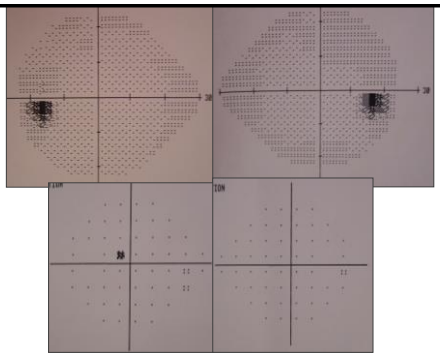
Optic Nerve



Importance of Understanding Orbital Anatomy - Mucocele

- Knowing this was not a more typical cause of diplopia due to the facial disfigurement
- Realize the localization of the source to the ethmoid and frontal sinuses (the bump present for the past year)
- Understand the increased likelihood of mucoceles with a history of sinus surgery
- Recognize the need for urgent surgery to prevent vision loss from optic nerve involvement, as well as rupture and spread of infection





Meningoencephalocele

- A meningoencephalocele is a protrusion of the meninges and the brain through a defect in the cranium.
- The most common causes of intraorbital encephaloceles are trauma, tumors, and congenital malformations.
- Most patients who develop intraorbital encephalocele after trauma develop pulsatile exophthalmos, usually within 1 year.

THANK YOU.

ANY QUESTIONS?