TO REFER OR NOT TO REFER?… THAT IS THE QUESTION

Presented by Kelly 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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47 Year-Old Asymptomatic Man

- Has been under care of an eye care provider x years
  - Called a glaucoma suspect
    - Large cupping OU
    - Visual field defects OS
  - Previously recommended that he start Xalatan
    - Opted not to due to possibility of iris pigmentation changes
  - Now recommended that he have an SLT
    - He wants to know if he should proceed with this

Ocular	Systemic	Medications	Family	Social
• Large cupping • Normal IOP	• Unremarkable	• None	• Mother • Narrow Angle Glaucoma	• unremarkable

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The SLT has been recommended, so…

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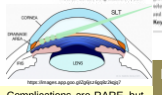
Clinical Ophthalmology — Dovepress

REVIEW

Complications of selective laser trabeculoplasty: a review

Julia Song



Complications are RARE, but we must always assess the risk vs benefit of any treatment.

- Elevated IOP
- Iritis
- Hyphema
- Macular Edema
- Foveal Burn
- Corneal Haze

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We know that the risks of SLT are low, but the benefits must outweigh them to proceed.

We need to do his exam…

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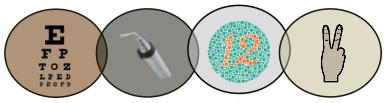
AFFERENT FINDINGS



BCVA
OD 20/15
OS 20/15

Pupils
PERRLA
(-) RAPD

Color
OD 14/14
OS 14/14

CF
OD
full-to-finger count;
No red desaturation

OS
full-to-finger count;
No red desaturation

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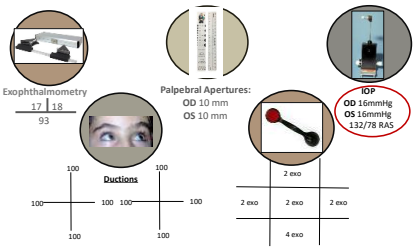
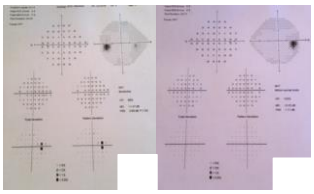
Efferent Findings



Obtained Past Records

- Highest documented IOP
  - 18 mm Hg OD
  - 18 mm Hg OS

We obtained several VFs over several years, and all looked exactly the same… repeatable defect.



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Pressure Never Was Elevated

- What does this mean?
- If this is thought to be an optic neuropathy, and IOP was never elevated, does that mean a work-up is warranted to look for non-glaucomatous causes (imaging, labs, etc)?

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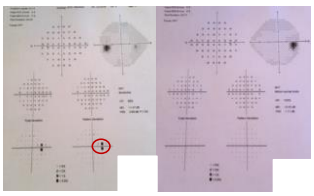
Obtained Past Records

- Highest documented IOP
  - 18 mm Hg OD
  - 18 mm Hg OS



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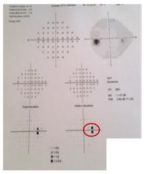
Glaucoma is a Disease of the Arcuate RNFL Bundles



- Nasal Step ?
- Bjerrum Scotoma
- Arcuate Defect

- Glaucomatous defects are mainly nasal
- Be suspicious of a "glaucomatous defect" which is mainly temporal!

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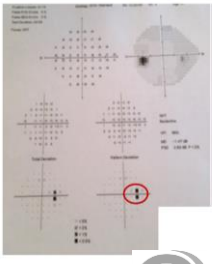

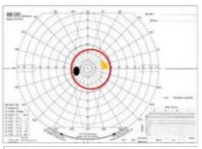

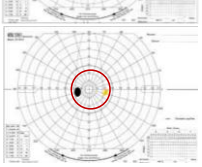

Remember the Horizontal Raphe!

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Let's Do Our Own VF - GVF

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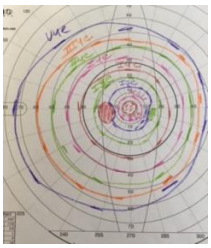

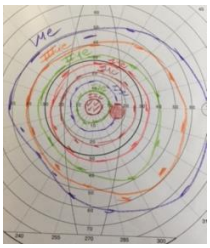

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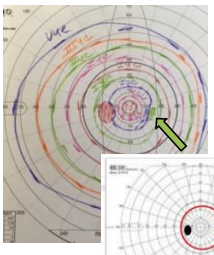

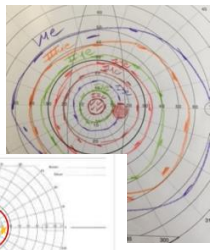

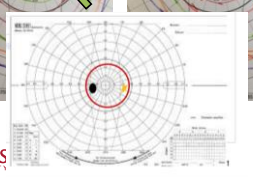

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Maybe a 10-2 will pick up more defect of the proximal arcuate bundles?

Published in final edited form as:
Ophthalmol Glaucoma. 2019 ; 2(2): 95–102. doi:10.1016/j.ogla.2019.01.003.

Comparing 10-2 and 24-2 Visual Fields for Detecting Progressive Central Visual Loss in Glaucoma Eyes with Early Central Abnormalities

Zhichao Wu, BAppSc(Optom), PhD[1,2], Felipe A. Medeiros, MD, PhD[3], Robert N. Weinreb, MD[4], Christopher A. Girkin, MD, MSPH[5], Linda M. Zangwill, PhD[1]

[1]Hamilton Glaucoma Center, Department of Ophthalmology, University of California, San Diego, La Jolla, California.

[2]Centre for Eye Research Australia, Royal Victorian Eye and Ear Hospital, East Melbourne, Australia

[3]Ophthalmology, Department of Surgery, The University of Melbourne, Melbourne, Australia

[4]Duke Eye Center and Department of Ophthalmology, Duke University School of Medicine, Durham, North Carolina

[5]Department of Ophthalmology, University of Alabama at Birmingham, Birmingham, Alabama.

Conclusions: Trend-based analyses using 10-2 MD resulted in a mild reduction (7-9%) in the time to detect central visual field progression compared to C24-2 MD in glaucoma eyes with early central visual field abnormalities. Further studies are needed to determine whether other progression analyses can better exploit the increased sampling of 10-2 tests. These findings provide evidence-based guidance on the potential value-add of 10-2 testing in the clinical management of glaucoma patients.

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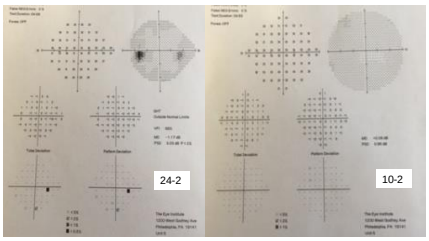
Maybe a 10-2 will pick up more defect of the proximal arcuate bundles?



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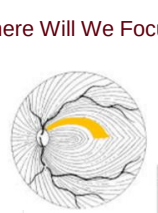
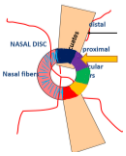
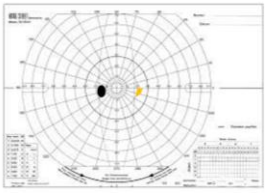
Where Will We Focus on Optic Disc?





- Where proximal superior arcuate RNFL bundle inserts onto disc



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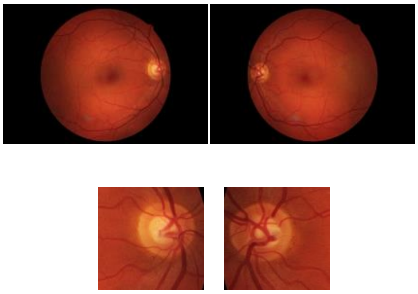

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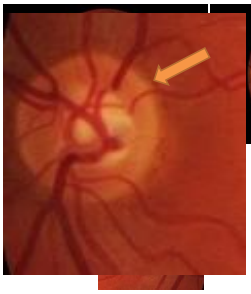

No pallor
No notching

LET'S LOOK AT THE OCT!

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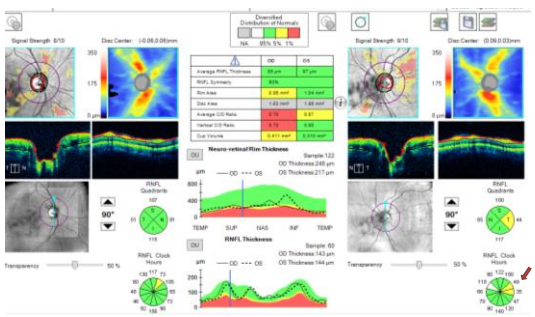

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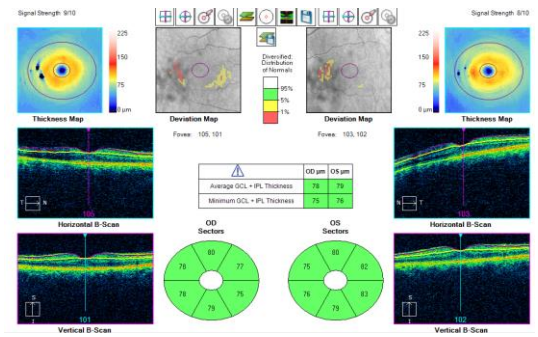

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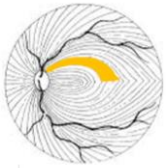
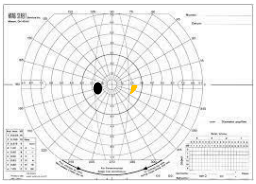
What about the corresponding retina?





- In the region of the superior proximal arcuate bundle

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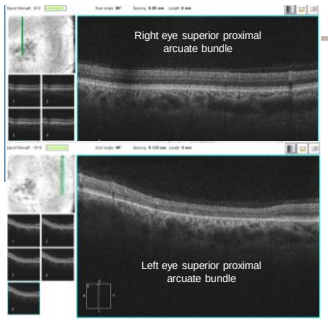
Corresponding retinal areas in each eye.

Do you notice a difference?



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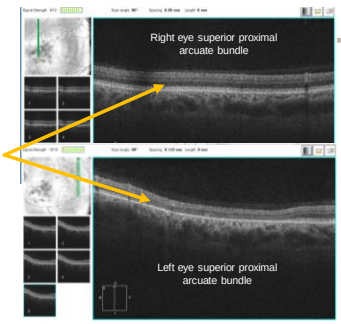
In a similar region in each eye, note the difference in the ONL and IS/OS



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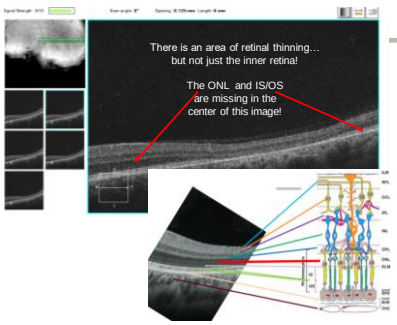

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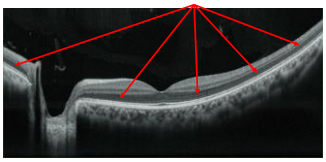
The ONL should not be missing anywhere in the retina!



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BMJ Journals

British Journal of Ophthalmology

Home / Archive / Volume 103, Issue 9

Clinical science

Outer retinal layer thickness in patients with glaucoma with horizontal hemifield visual field defects

Jayme R Vianna [1], Ziad Butty [1, 2], Lucas A Torres [1], Glen P Sharpe [1], Donna M Hutchison [1], Lesya M Shuba [1], Marcelo T Nicolela [1], Balwantray C Chauhan [1]

Correspondence to Dr Jayme R Vianna, Department of Ophthalmology and Visual Sciences, Dalhousie University, Halifax, Nova Scotia, Canada.

Conclusion Our study showed no evidence that glaucoma has an effect on the outer retinal layer thickness. In contrast, a large impact was observed in inner layer thickness.

http://dx.doi.org/10.1136/bjophthalmol-2018-312753

There have been some reports of damage to photoreceptors in chronic, end-stage glaucoma, or with elevated IOPS.

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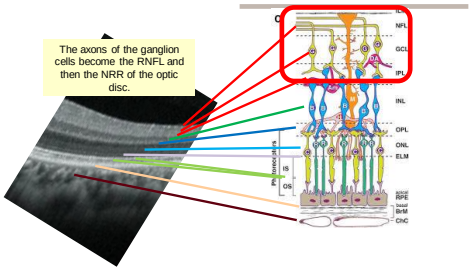
Glaucoma is a disease of the INNER retina



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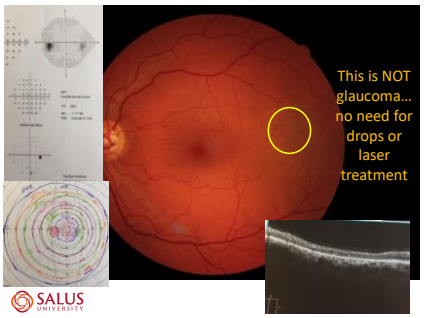

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Sometimes it is NOT Glaucoma, OR another Optic Neuropathy

- It is important to make that distinction
  - There can be side effects to glaucoma drops and laser treatment
  - This treatment can be costly
- How did we know where to look with a 5-line raster on the OCT?
  - " Back to the Basics…. ANATOMY"

It is just as important to know when it is NOT glaucoma as to know when it IS glaucoma!

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Case 2

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79 year-old woman

- **Vertical diplopia x several mos, pressure around left eye**
- Denies: difficulty swallowing, breathing, arm/leg weakness
- No other visual, ocular, or neurologic symptoms

Ocular History	Systemic History	Medications
• Diabetic retinopathy • s/p blepharoplasty OU • s/p cataract surgery OU	• DM • HTN • Hypercholesterolemia	• Lisinopril • Glyburide • Januvia • Atorvastatin

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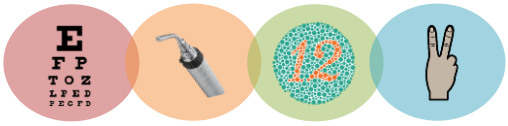
AFFERENT FINDINGS



BCVA
OD 20/20
OS 20/20

Pupils
PERRLA
(-) APD

Color
OD 14/14
OS 14/14

CF

OD
full-to-finger count
no red desat

OS
full-to-finger count
no red desat

Normal

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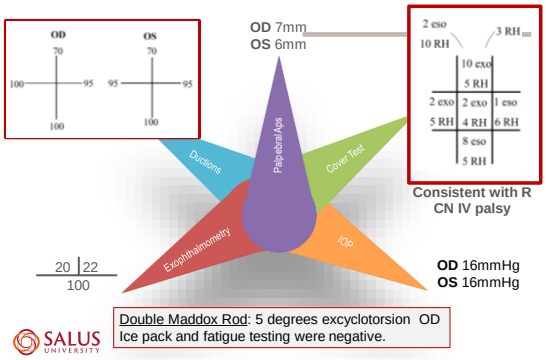
Efferent Findings



Consistent with R CN IV palsy

Double Maddox Rod: 5 degrees excyclotorsion OD
Ice pack and fatigue testing were negative.

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- No Referral Needed
- Refer to PCP for further testing
- Refer to Neuro-Op for first available appt
- Refer to Neuro-Op Expedited
- Send to the Emergency Room STAT

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REMEMBER…

- Her complaint was pressure around the LEFT eye
- We find a pattern consistent with a RIGHT CN IV palsy

- Does this make sense?
- What else should we be thinking / considering?

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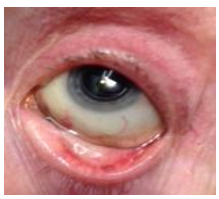
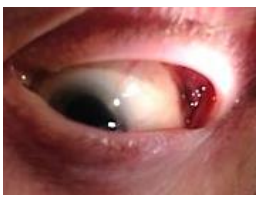
Remember, she complained about pressure around the LEFT eye!





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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

No Referral Needed	Refer to PCP for further testing
Refer to Neuro-Op for first available appt	Refer to Neuro-Op Expedited
Send to the Emergency Room STAT	Refer to Ocular Oncology

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The Pivotal Test In This Case

- **Slit Lamp Examination (with careful assessment of deep fornix)!**
  - Just because the patient reports diplopia, do not ignore the rest of the ocular health examination
  - An ocular or orbital process could be the etiology of the diplopia
  - **Be suspicious of unilateral conjunctival pigment, especially in Caucasians**
- **History**
  - The CT was consistent with a R CN IV palsy, but she was complaining of pressure around the left eye

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What We Did

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- Referral to ocular oncology
- Extensive facial surgery to remove mucus membrane from the fornix 360 degrees with cryotherapy, biopsy, and reconstruction
- Dx: **CONJUNCTIVAL MELANOMA**

- On follow-up
- Improvement in diplopia, less vertical deviation (less right hyper)

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Case 3

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67 year-old woman

- Presents emergently due to **a spot in her vision x 6 months**
- She initially thought it was a smudge on her glasses, but then realized it didn't go away when she removed her glasses
- The spot is stable and stationary

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HISTORY

Ocular	Systemic	Social
• Unremarkable	• "borderline" DM • HTN • Hypercholesterolemia • Osteoarthritis	• Unremarkable

She denies any trauma to her eyes or head.
She denies any thyroid disorders.

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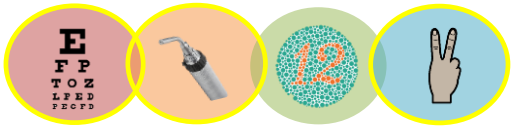
AFFERENT FINDINGS



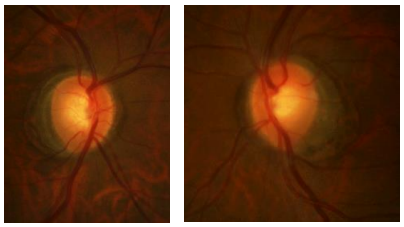
BCVA
OD 20/30
OS 20/25+

Pupils
PERRLA
(+) 1.5-1.8 log RAPD OD

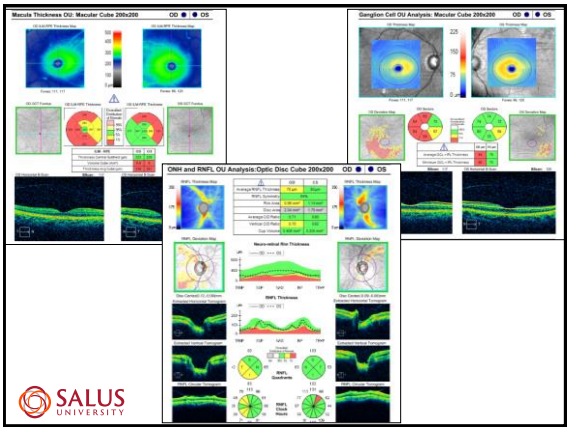
Color
OD 14/14
OS 14/14

CF
OD
Inferior constriction

OS
full-to-finger count



? Trace temporal pallor OD



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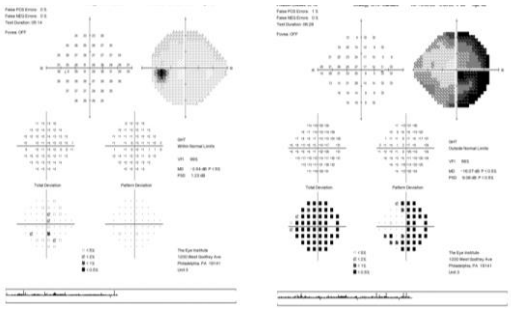

MD: -2.54dB

MD: -16.07dB

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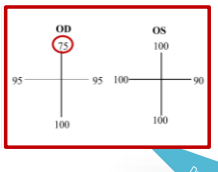
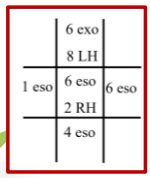
Efferent Findings



OD 5 mm
OS 5 mm



22 | 21
98

OD 16mmHg
OS 16mmHg
147/85 RAS

Positive forced duction test OD

TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- No Referral Needed
- Refer to PCP for further testing
- Refer to Neuro-Op for first available appt
- Refer to Neuro-Op Expedited
- Send to the Emergency Room STAT

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The Pivotal Test In This Case

- **Ductions, cover testing in different positions of gaze, and forced ductions**
  - to see that this is affecting BOTH the afferent and the efferent visual system, which localizes to an **orbital** space-occupying lesion.

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What We Did

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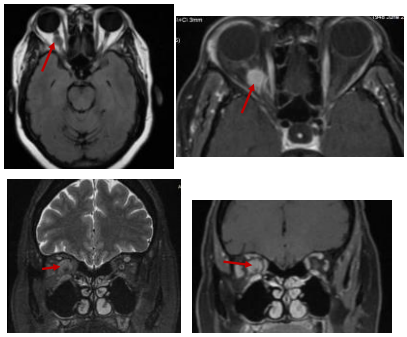

Right Optic Nerve Sheath Meningioma
(contrast enhancement)

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- What would you do at this point?
  - A. Follow the patient yearly
  - B. Follow the patient every 4 months
  - C. Refer to Ophthalmology
  - D. Refer to Neurology
  - E. Refer to Neurosurgery

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- What would you do at this point?
  - A. Follow the patient yearly
  - B. Follow the patient every 4 months
  - C. Refer to Ophthalmology
  - D. Refer to Neurology
  - E. Refer to Neurosurgery

We co-managed the patient with neurosurgery.

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1.5 Years Later

- **Patient called** reporting **worsening of vision, mainly diplopia. She is now noticing horizontal diplopia in left gaze.**
- She told this to her neurosurgeon, so she was scheduled to get fit with a mask in preparation for radiation due to presumed worsening of right ONSM.

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- What would you do at this point?
  - A. Tell patient to proceed with radiation
  - B. Tell patient to see you for pre-treatment baseline measurements first
  - C. Tell patient to see you to determine if radiation is necessary

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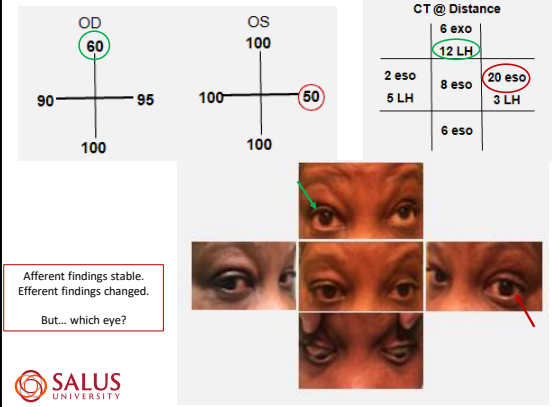

Afferent findings stable.
Efferent findings changed.

But… which eye?

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The Pivotal Test In This Case

- Ductions and cover testing in different positions of gaze to see that change is in the fellow eye!

Radiation is only warranted for ONSM if it is progressively worsening. It is not worsening; the change is in the fellow eye.

Radiation is NOT indicated in this case.

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What We Did

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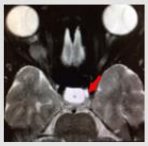
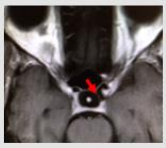
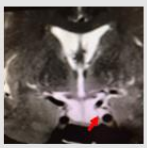
F/U NEURO-IMAGING

Stable right ONSM.

Left medial cavernous sinus arachnoid cyst (arrow) appears bright on T2 images and dark on T1 images.

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Case 4

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12 year-old boy

- Presents for a routine eye exam
- Last eye exam was 1 ½ years ago
- He was diagnosed with refractive amblyopia and was instructed to patch OS for 2 hours per day

Refractive Error:	
OD: +1.50 – 4.50 x 10	20/25-
OS: -0.25 sphere	20/20

- Was supposed to return for further amblyopia evaluation, but he was unable to do so
- He lost his glasses a year ago and has not been patching; feels his vision is OK

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- Asthma
  - Uses an inhaler as needed
- Seasonal Allergies
- Headaches
  - < 1 x / week
  - Usually occur in the evening
  - Both eyes can hurt during the headache
  - No medication is needed
  - Goes away with rest
  - His grandfather never heard him complain of headaches
- He denies any other visual, ocular, or neurologic symptoms

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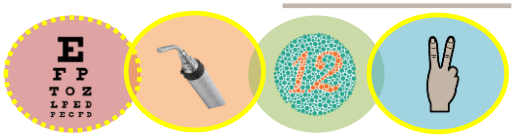
AFFERENT FINDINGS



BCVA
OD 20/25-1
OS 20/20

Pupils
PERRLA
(+) 0.6 APD OD

Color
OD 14/14
OS 14/14

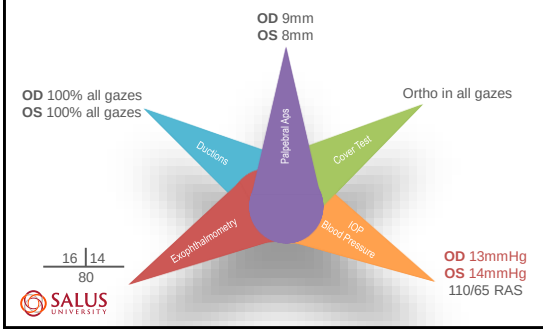
CF
OD
Inferior constriction, Red desat IN > IT

OS
full-to-finger count
IN red desat

- Pupils
  - 1 second test:
    - no change in pupil size when swinging from eye to eye
  - 3 second test
    - Escape begins at 1 second OD
    - Escape begins at 3 seconds OS

Red Desaturation Between the Eyes: 20% desat OD
Brightness Sense: 20% reduced OD

Efferent Findings



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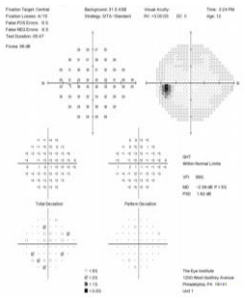
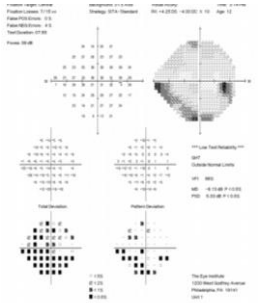
HVF



MD: -2.39dB



MD: -8.13dB

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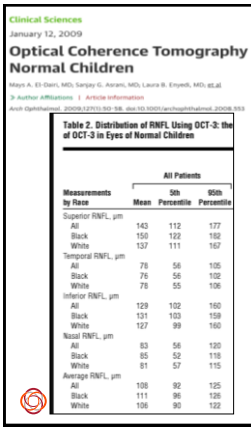
OCT



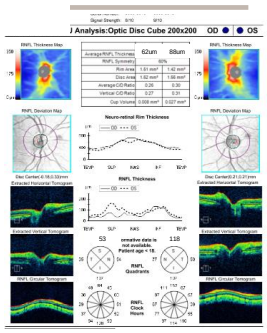
Clinical Sciences

January 12, 2009

Optical Coherence Tomography in the Eyes of Normal Children

Table 2. Distribution of RNFL Using OCT-3: the of OCT-3 in Eyes of Normal Children

Measurements by Race	All Patients Mean	5th Percentile	95th Percentile
Superior RNFL, μm			
All	143	112	177
Black	150	122	182
White	137	111	167
Temporal RNFL, μm			
All	78	56	105
Black	76	56	102
White	78	55	106
Inferior RNFL, μm			
All	129	102	160
Black	131	103	159
White	127	99	160
Nasal RNFL, μm			
All	83	56	120
Black	85	52	118
White	81	57	115
Average RNFL, μm			
All	108	92	125
Black	111	96	128
White	106	90	122



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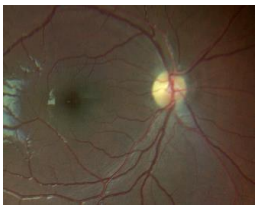

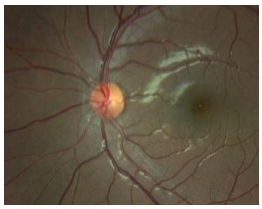

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- No Referral Needed
- Refer to PCP for further testing
- Refer to Neuro-Op for first available appt
- Refer to Neuro-Op Expedited
- Send to the Emergency Room STAT

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The Pivotal Test In This Case

- Swinging Flashlight Test!
  - Even if you have an explanation for possible reduced VA, careful pupil testing is essential to rule out an overlying pathology
  - If not for the RAPD, the VF and OCT would not have been done
  - Do pupil testing YOURSELF
  - Do BOTH the 1 second and the 3 second test

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What We Did

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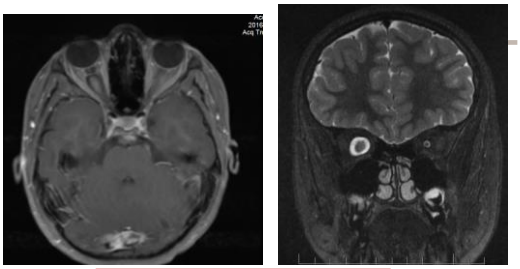
MRI Results



OPTIC PATHWAY GLIOMA

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- Referral to Pediatric Neuro-Ophthalmology for management of optic pathway glioma.
  - **Rule-out association with Neurofibromatosis type 1**
  - Refer to oncology

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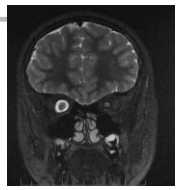
Optic Pathway / Hypothalamic Gliomas (OPHG)

- Optic nerve gliomas account for 1.7-7% of gliomas
- Approximately 75% of patients with these tumors present during the first decade of life and 90% become symptomatic before 20 years of age.
- Usually low grade tumors with variable clinical course



- Children age 2 and under:
  - failure to thrive
  - Macrocephaly
  - vision loss
- Children age 2 to 5:
  - endocrine dysfunction
- Children 6+ and adults:
  - vision complaints

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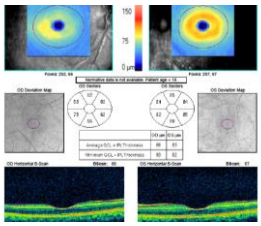
OPHG TREATMENT

- Observation if stable
- Chemotherapy **ONLY** if progressive visual acuity or visual field loss
  - Can be difficult to assess in children
- OCT can be helpful!!
  - Need to look at ganglion cell analysis (GCL-IPL measurements)



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82

OPHG

- Our patient has been observed for several years, and has been stable
- Overall, optic pathway gliomas tend to be low grade, slow-growing, and associated with long patient survival.
- However, they must be identified and differentiated from other childhood ocular diseases, including amblyopia, as they require different management and treatment strategies.

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83

Case 5

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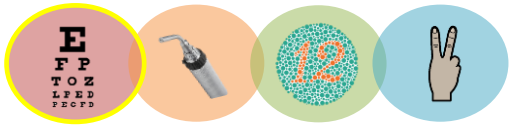
14 Year-Old Girl

- Reduced VA OD
- OD has been blurry for a long time
- No new complaints / feels vision is stable
- Previously told of a "lazy eye"
- The patient reports that her vision is stable
- She currently wears contact lenses
- She does not recall ever being told to patch an eye

- Medical history - unremarkable
- Social history - unremarkable
- Family history
  - Grandmother had eye muscle surgery due to "lazy eye"
  - Father has bad eyesight (high myope)

AFFERENT FINDINGS



BCVA	Pupils	Color	CF
OD 20/80 (single letter*) **OS** 20/20	PERRLA (-) APD	**OD** 14/14 **OS** 14/14	**OD** full-to-finger count no red desat **OS** full-to-finger count no red desat

- 5 Years Ago BCVA: OD 20/80 OS 20/20
- 1 Year Ago BCVA: OD 20/100 OS 20/20
- 1 Month Ago BCVA: OD 20/125 OS 20/20

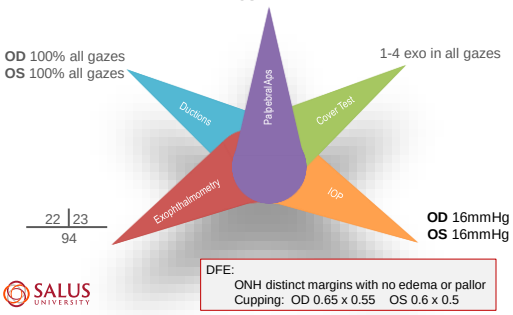
There was a concern for progressive vision loss OD.

Efferent Findings

OD 12mm
OS 12mm

OD 100% all gazes
OS 100% all gazes

1-4 exo in all gazes



22 | 23
94

OD 16mmHg
OS 16mmHg

DFE:
ONH distinct margins with no edema or pallor
Cupping: OD 0.65 x 0.55 OS 0.6 x 0.5

Does the Refractive Error Explain the Reduced VA?

- BCVA:
  OD -4.25 – 2.00 x 180 20/80
  OS: -5.75 – 1.50 x 10 20/20

If yes, then we are finished…
If not, then what do we do next…

TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- No Referral Needed
- Refer to PCP for further testing
- Refer to Neuro-Op for first available appt
- Refer to Neuro-Op Expedited
- Send to the Emergency Room STAT

Does This Patient Need Neuro-Imaging?

- A. YES
- B. NO
- C. I want to do more testing before I decide that

Does This Patient Need Neuro-Imaging?

- A. YES
- B. NO
- C. I want to do more testing before I decide that

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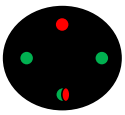
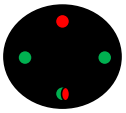
Worth 4-Dot Test

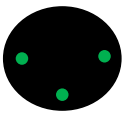
Near



Intermediate



Distance



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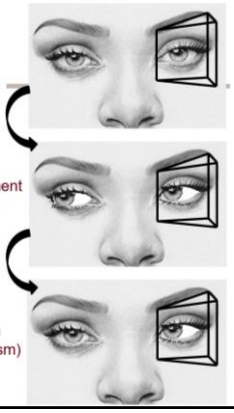
4 Base-Out Test

Expected Results
(if no suppression scotoma)



Vergence Movement
(both eyes)

Same result should occur with prism placed over each eye.

Fusional Duction
(eye without the prism)

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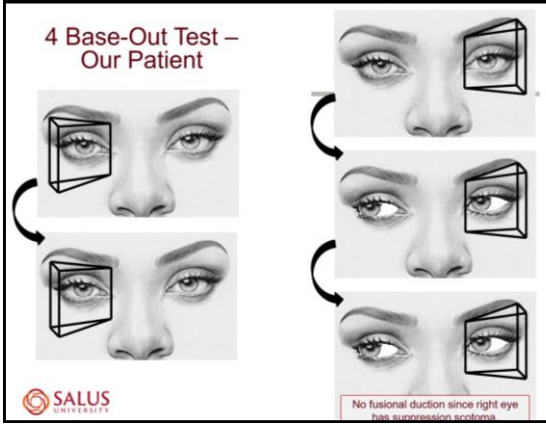
4 Base-Out Test – Our Patient



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No fusional duction since right eye has suppression scotoma

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The Pivotal Test In This Case

- Visual Acuity!
  - Be sure to test single letter VA through best correction, even with pinhole as necessary
- 4 Base-Out Prism Test!
  - Even if you have an explanation for possible reduced VA, is the degree of refractive error and the symmetry between the eyes consistent with the measured VA?
  - If you cannot explain amblyopia by either refractive error or strabismus, you need to
    - Look for another amblyogenic factor
      - **REMEMBER MICROTROPIA**
    - Rule out neuro-ophthalmic disease (ALWAYS be sure you completed the first before you move on to the second)

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What We Did

- Nothing… (well, nothing neuro-related anyway)
- Reassured the patient and her grandmother
- Suggested polycarbonate lenses

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Case 6

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62 year-old woman

- **Longstanding history of glaucoma**
  - **Using Travatan and Azopt OU**
- No eyecare x 1.5 months due to lack of insurance
  - PCP did refill drops during that time

- **Chief complaint**
  - Blur with prolonged reading
  - Rare headaches from lack of sleep
- **Systemic History:**
  - Diabetes
  - Hypertension
  - Hypercholesterolemia
  - Rheumatoid Arthritis

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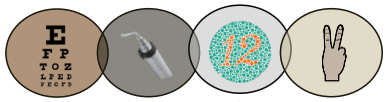
AFFERENT FINDINGS

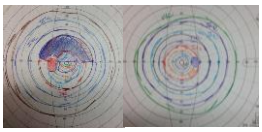


BCVA
OD 20/20
OS 20/20

Pupils
PERRLA
(+) trace
RAPD OS

Color
OD 12/14
OS 14/14

VF
Unilateral superior arcuate defect



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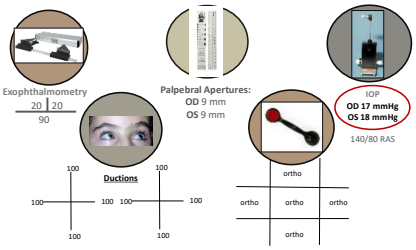
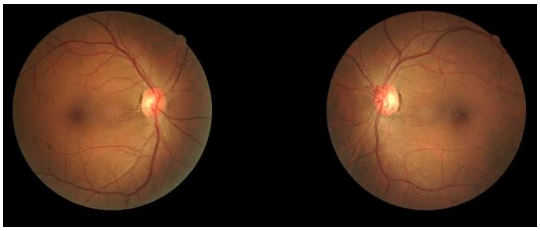
Efferent Findings





We see asymmetric cupping, OS > OD.
But, what else do we see?

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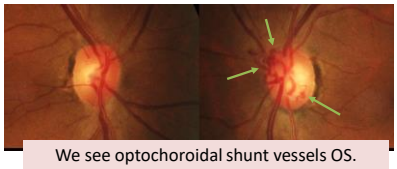
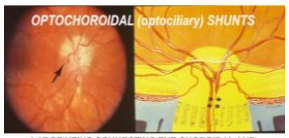


We see optochoroidal shunt vessels OS.



LARGE VEINS CONNECTING THE CHOROIDAL AND RETINAL CIRCULATION AT THE OPTIC NERVE HEAD

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CAUSES OF OPTOCHOROIDAL / OPTOCILIARY SHUNT VESSELS

- If the patient has no history or clinical findings of CHRONIC PAPILLEDEMA, CRVO, or ADVANCED GLAUCOMA, then we must rule-out a:
  - MENINGIOMA of the optic nerve or sphenoid wing
    - (with neuroimaging)

Let's look again, to see if there is any evidence of a cause…

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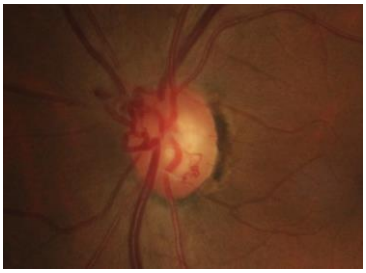

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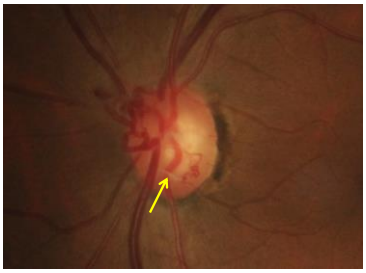

This is a past retinal vein occlusion!

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•Ask more questions!

- She recalls that about 20 years ago
  - Sudden decrease in vision in OS
  - Unsure of diagnosis, but remembers being told about some bleeding in the eye

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Case 7

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58 Year-Old Woman

- **Concerned about cosmetic appearance of her eyelids**
- **Noticed eyelid asymmetry x 15 years, after trauma OS**
- Punched OS 15 years ago; no treatment sought
- Vision is good in each eye
- Denies diplopia, eye pain, or headaches

SYSTEMIC HISTORY

- Remarkable only for hypertension

MEDICATIONS: hydrochlorothiazide

SOCIAL HISTORY: smokes 1 pack of cigarettes per week

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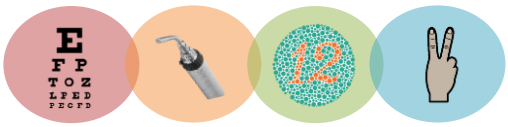
AFFERENT FINDINGS



BCVA
OD 20/20
OS 20/20

Pupils
PERRLA
(-) APD

Color
OD 14/14
OS 14/14

CF
OD
full-to-finger count
no red desat
OS
full-to-finger count
no red desat

Normal

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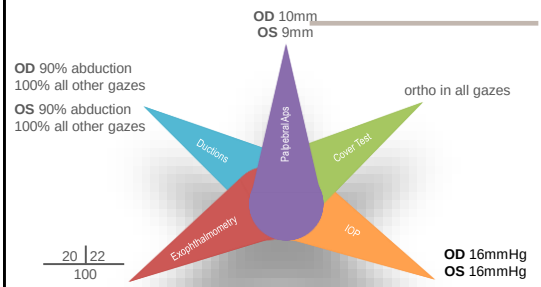
Efferent Findings



Normal

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This is why it is good to do the testing yourself!

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TO REFER OR NOT TO REFER …THAT IS THE QUESTION

- No Referral Needed
- Refer to PCP for further testing
- Refer to Neuro-Op for first available appt
- Refer to Neuro-Op Expedited
- Send to the Emergency Room STAT

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The Pivotal Test In This Case

- **Exophthalmometry (and patient observation)!**
  - Whenever there is eyelid asymmetry, exophthalmometry should also be done
  - Could also appreciate the pulsations due to pulsating mires on tonometry
  - Be sure to ask about trauma. Even if trauma is very remote (15 years ago), work-up is still necessary
    - To r/o other cause such as CC fistula
    - To identify and direct neurosurgical consultation for meningoencephalocele in attempt to prevent intracranial infection

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What We Did

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DIFFERENTIAL DIAGNOSES CAUSES OF PULSATILE EXOPHTAHALMOS

- **Aortic Regurgitation**
- **Carotid-Cavernous Fistula**
- **Arachnoid Cyst**
- **Aplasia of Sphenoid Wing in Neurofibromatosis 1**
- **Congenital or Acquired Bony Defects of Orbit**
  - **(usually the orbital roof)**

This list indicates that neuro-imaging is needed.

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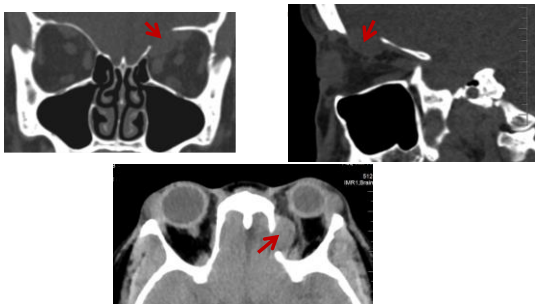
Meningoencephalocele



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Meningoencephalocele

- A meningoencephalocele is a protrusion of the meninges and the brain through a defect in the cranium.
- Most patients who develop intraorbital encephalocele after trauma develop pulsatile exophthalmos, usually within 1 year.
- Surgical approaches require multimodal strategies involving neurosurgeons, plastic surgeons, and ophthalmologists.

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Case 8

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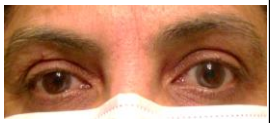
51 Year-Old Woman



- Presents due to new onset ptosis x 3 months
  - Noticed this upon awaking from open heart surgery
    - Prosthetic mitral valve secondary to stenosis
      - ON-X mechanical mitral valve
    - Sutured PFO
    - Pacemaker inserted due to persistent atrial fibrillation
- She denies any changes with vision, headaches, or eye pain
- She denies any other visual or neurologic symptoms

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51 year-old woman

- Noticed right sided ptosis since open heart surgery 3 months prior
  - Followed by PCP and cardiologist (no known cause for ptosis identified)

Ocular	Systemic	Medications	Family	Social

• Unremarkable	• Persistent atrial fibrillation
	• Congenital patent foramen ovale
	• Mitral valve stenosis
	• s/p surgery
	  • Mechanical aortic valve
	  • Pacemaker
	  • Sutured PFO
	• Mild anemia
		• 81 mg aspirin
		• Metoprolol succinate 50mg
		• Warfarin 5mg
			• unremarkable	• unremarkable

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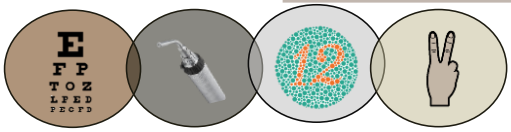
AFFERENT FINDINGS



BCVA
OD 20/20
OS 20/20

Pupils
PERRLA
(-) RAPD

EFFERENT PUPILS
Bright
OD 2.5, OS 3

Dim
OD 4.25, OS 5

Color
OD 14/14
OS 14/14

CF
OD
full-to-finger count;
No red desaturation

OS
full-to-finger count;
No red desaturation

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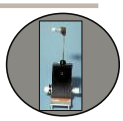
Efferent Findings

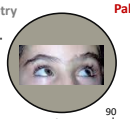
Normal SLE and DFE

Exophthalmometry
16 | 16
94

Palpebral Apertures:
OD 8 mm
OS 10 mm



IOP
OD 16mmHg
OS 15mmHg
132/70 RAS
Pulse: 70bpm
REGULAR



Ductions

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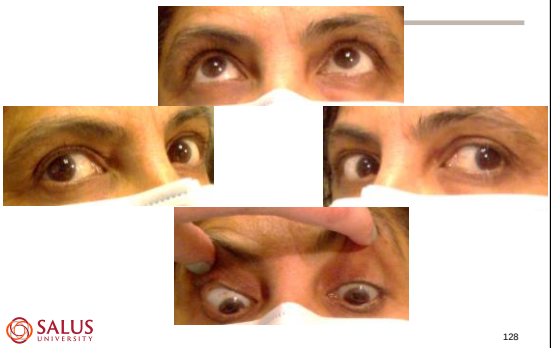

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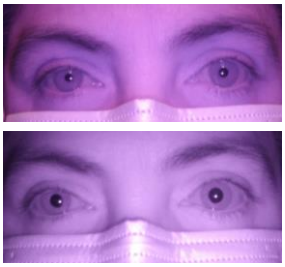
Concern for Right Horner Syndrome

EFFERENT PUPILS

Bright
OD 2.5, OS 3

Dim
OD 4.25, OS 5

Palpebral Apertures:
OD 8 mm
OS 10 mm



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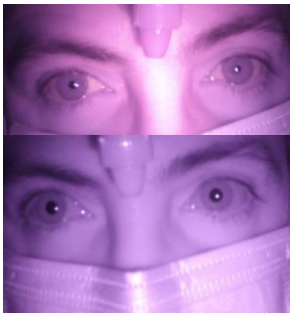
Diagnostic Apraclonidine Testing

Diagnostic Pupil Testing
Type: 0.5% Apraclonidine

30 Min:	Bright	Dim
OD	3.00	5.00
OS	3.25	5.25

Still negative test after 30 minutes

Now need to wait another 30 minutes



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Diagnostic Apraclonidine Testing

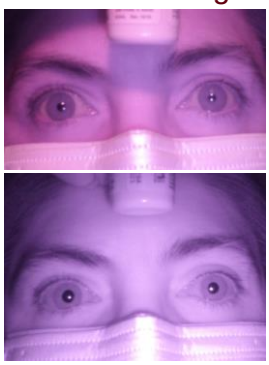
Diagnostic Pupil Testing
Type: 0.5% Apraclonidine

30 Min:	Bright	Dim
OD	3.00	5.00
OS	3.25	5.25
1 hr:	Bright	Dim
OD	2.75	5.25
OS	2.50	5.00

Findings: +
Horner's Syndrome
Eye: OD

Positive Test after 1 hour

Confirms right Horner syndrome



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Assessment & Plan

- What needs to be done?
- How urgent is the work-up?
  - She noticed this 3 months ago, upon awaking from surgery
- What is the cause?
  - She noticed it after heart surgery, but her cardiologist said it is not related to the surgery

Is This An Emergency?

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Assessment & Plan

- If we cannot connect the right Horner syndrome with the heart surgery, she is in need of imaging to look for a causative etiology
  - MRA or CTA of head and neck
    - R/O carotid dissection
  - MRI with contrast (brain, C-spine, soft tissue neck- including lung apex, orbits)
    - R/O mass, structural abnormality, abnormal enhancement
- Think about sympathetic pathway
  - Would open heart surgery affect this?
  - On the right side?

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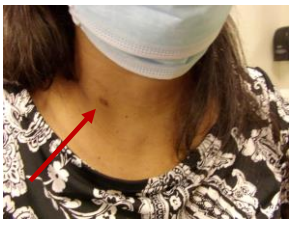
Be Observant & Get Records!

- She had a central venous line placed in the right jugular vein in the lower neck.
- Placement or removal of such likely nicked the right sympathetic chain.



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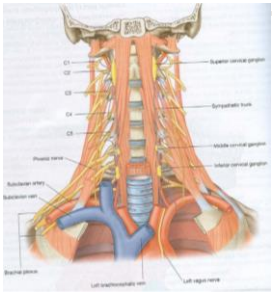
Timing & Location vs. Coincidence

- She had a central venous line placed in the right jugular vein in the lower neck.
- Placement or removal of such likely nicked the right sympathetic chain.
- Knowing the anatomy can be very helpful in determining your management plan!



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Case 9

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52 Year-Old Woman

- Presents emergently due to sudden ptosis OS
  - Noticed x few days
- Vision also seems to be getting blurry
- She denies diplopia
- She has had headaches
  - But they have recently improved

MEDICATIONS:
Aubagio
Baclofen
Tirosint

SYSTEMIC HISTORY:
- Multiple sclerosis
- Hypothyroidism

- Followed by neurology
- Last saw neurology 2 weeks ago

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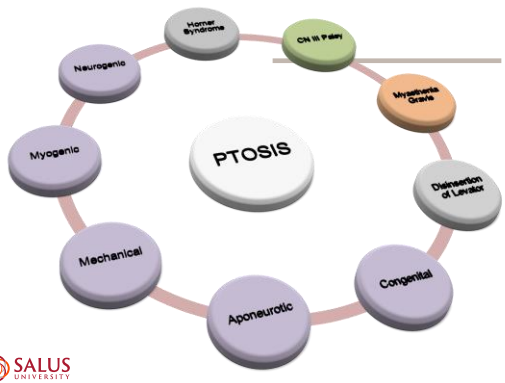

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Eye Exam Results

- Afferent exam normal
- Pupils isocoric
- Exophthalmometry: 11 OD and 11 OS
- EOMs: full OU
- Comitant small exo deviation
- No nystagmus
- Lids:
  - Apertures: 10 mm OD, 9 mm OS
  - Pre-fatigue: 9mm OD, 8mm OS
  - Post-fatigue 8mm OD 7mm OS
  - Pre-ice: 8mm OD, 7mm OS
  - Post-ice 8mm OD, 7mm OS
  - Lid crease 5mm OD, 6mm OS
  - Levator function: 15mm OD, 15mm OS

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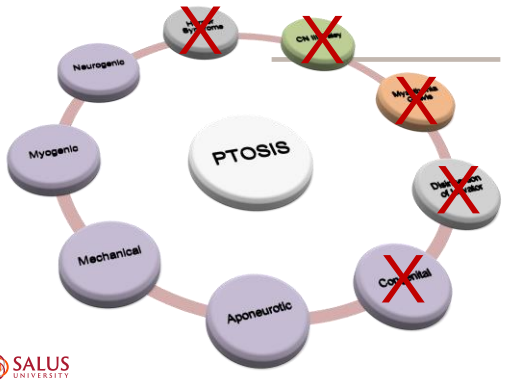

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Neurologic Exam Results

- Motor weakness of bilateral lower extremities
  - History of MS
- Unable to wrinkle frontalis muscle on either side
  - Possibly more weak on left side
  - Could smile normally on both sides
    - Rules out CN VII palsy…

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- Ask more questions!

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- Ultimately determined that pt had 1st Botox injection 2 wks ago at neurology appointment because of chronic headache
- She admits to eye rubbing
  - Likely that Botox has migrated to left eyelid causing ptosis
- Educated patient that effects of Botox will gradually wear off over the next month.

When patients get a droopy eyelid from Botox, it usually begins about 1-3 weeks after the injection and lasts about 2-3 weeks on average.

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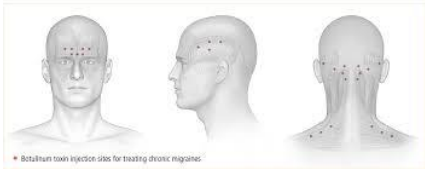
BOTOX treatment for chronic migraine

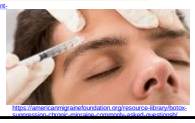
Findings from two 24-week multicenter randomized controlled trials (PREEMPT 1 AND PREEMPT 2) suggest that botulinum toxin type A is effective for the treatment of **chronic** migraine.



* Botulinum toxin injection sites for treating chronic migraines

https://www.bridgemilneurological.com/migraine-management-center/botox-injections-for-migraine-botulinum-toxin/

The use of botulinum toxin therapy is **not** recommended for the preventive treatment of **episodic** migraine.



https://americanmigrainefoundation.org/resource-library/botox-suppression-chronic-migraine-commonly-asked-questions/

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BOTOX SIDE EFFECTS

- dry mouth
- discomfort or pain at the injection site
- Tiredness
- **Headache**
- neck pain
- eye problems:
  - **double vision**
  - **blurred vision**
  - **drooping eyelids**
  - **swelling of eyelids**
  - **dry eyes**
  - **drooping eyebrows**

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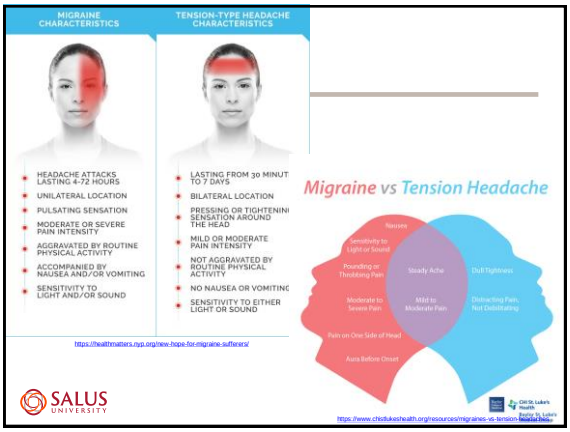

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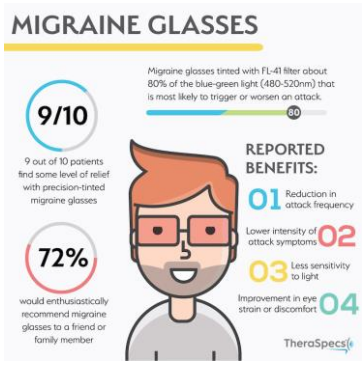
FL-41 Tinted Lenses

These are the same lenses recommended for blepharospasm, and other causes of photophobia.



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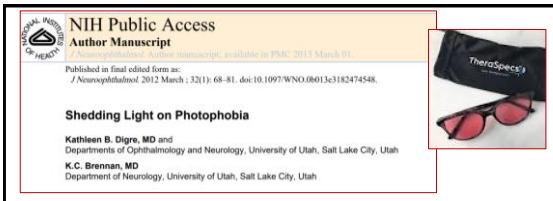

headaches and those with light sensitivity. Some tints have been successful in migraine. Good et al. (29) found that FL-41 tint, a rose-colored tint, reduced migraine frequency in children by over one-half. Subjects reported a decrease in photophobia and glare in between attacks, but no change in the light sensitivity associated with the migraine attack. FL-41 tint filters 80% of short wavelength 50 or 60 Hz flicker that is seen with fluorescent lights. As flicker stimuli can be particularly noxious to patients with migraine (125), the authors reasoned that flicker reduction contributed to the reduction in headaches.

We studied FL-41 tinted lenses and found that they increased the threshold to discomfort in all subjects (controls, migraineurs, and patients with blepharospasm), but they did not differ from gray tinted lenses in reducing light sensitivity (44). To test whether patients preferred

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Case 10

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29 Year old woman

- Presents emergently due to transient blurring of vision OD in the setting of headaches
  - Awoke with a headache (did not wake her up)
  - 10/10 pain in back of head **(unilateral)**
  - **Pounding / throbbing headache**
  - She has been getting 5-6 x/month
    - Not always as severe
    - Sometimes pain is more anterior in head
- No nausea or vomiting
- Does experience photophobia and phonophobia
- Family history of migraine

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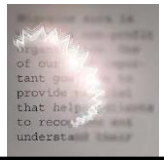
- Vision changes described as:
  - Right side of vision was blurry
    - Thinks is was both eyes
  - Parts of vision were missing
  - Distorted and shimmering
  - Lasted about ½ **hour**
  - **Gradually** moved from center to the right side of her VF

This is her 3rd such episode in the past year.

The last 2 times, the vision change occurred prior to the HA. Today, the HA and vision change occurred at the same time.



MIGRAINE AURA

Migraine visual aura may occur independently of headache (acephalgic migraine) and may be confused with ischemia, especially in older patients

- Most common cause of binocular TVL in young
- But migrainous visual aura is not rare in older adults
  - Onset after age 50 can even occur
- Presents with positive visual phenomena (scintillations)
- Can also be negative visual phenomena (scotoma)
- Classic migraine aura lasts 20-30- minutes, rarely up to an hour
- has a characteristic build-up, or evolution (unlike other TBVL)
- MECHANISM: neuronal depression after a period of cortical excitation (cortical spreading depression)

https://en.wikipedia.org/wiki/Photopsia

MIGRAINE AURA

The best known visual aura is called a fortification spectrum because its pattern resembles the walls of a medieval fort.

- May start as a small hole of light, sometimes bright geometrical lines and shapes in visual field
- May expand into a sickle- or C-shaped object, with zigzag lines on the leading edge.
- As it moves, it may appear to grow.
- Sometimes accompanied by a partial loss of vision / scotoma.
- Commonly last 15 to 30 minutes.

Fortification spectra

Vauban 17th century

http://www.neurovascularmedicine.com/migraine.php

Auras are not the same for all people. Some might experience bright spots or flashes.

MIGRAINE AURA

Characteristics of the Visual Aura

Positive phenomena, negative phenomena, or both	Either may occur alone; positive phenomena often occur first and are followed by negative phenomena
Visual field	Scotoma often start centrally and migrate peripherally
Shape	Fortification spectra often "C"-shaped; scotoma bean shaped
Motion	Objects may rotate, oscillate, or boil
Flicker	Rate 10 cycles per second; may change during the course of the aura
Color	Gray, red, green, gold, yellow, blue, or purple; often have no specific color except excessively bright white
Clarity	May be blurry or fuzzy
Brightness	Often very bright
Expansion	Buildup occurs in both fortification spectra and scotoma
Migration	Spectra may "march" from the central area to periphery or sometimes vice versa

MIGRAINE AURA

Patients having a migraine with aura have transient episodes of focal neurologic dysfunction appearing 1 to 2 hours before the onset of a migraine headache and resolving within 60 minutes.

Aura symptoms

Symptom	Prevalence (%)
Nausea	87
Photophobia	82
Lightheadedness	72
Scalp tenderness	65
Vomiting	56
Visual disturbances	36
Photopsia	26
Fortification spectra	10
Paresthesias	33
Vertigo	33
Alteration of consciousness	18
Syncope	10
Seizure	4
Confusional state	4
Diarrhea	16

After Raskin NH. Headache, 2nd ed. Churchill Livingston: New York (1998).

ICHD-3 beta

Cephalalgia

International Headache Society

Cephalalgia 33(9) 629-808
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DOI: 10.1177/0333102413485658
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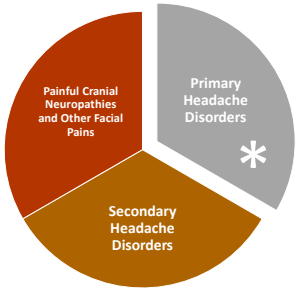
Headache Classification Committee of the International Headache Society (IHS)

The International Classification of Headache Disorders, 3rd edition (beta version)

Jes Olesen, Denmark (Chairman)

Lars Bendtsen, Denmark
David Dodick, USA
Anne Ducros, France
Stefan Evers, Germany
Michael First, USA
Peter J Goadsby, USA
Andrew Hershey, USA
Zaza Katsarava, Germany
Morris Levin, USA
Julio Pascual, Spain
Michael B Russell, Norway
Todd Schwedt, USA
Timothy J Steiner, UK (Secretary)
Cristina Tassorelli, Italy
Gisela M Terwindt, The Netherlands
Maurice Vincent, Brazil
Shuu-Jiun Wang, Taiwan

HEADACHE CLASSIFICATION – 3 main categories



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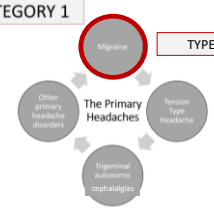
CATEGORY 1

TYPE 1

Migraine is a common disorder that affects approximately 17 percent of women and 6 percent of men.

Migraine is most common in those aged 30-39.



1.1 Migraine without aura
1.2 Migraine with aura (including 1.2.4 Retinal migraine)
1.3 Chronic migraine
1.4 Complications of migraine
1.5 Probable migraine
1.6 Episodic syndromes that may be associated with migraine

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MIGRAINE WITHOUT AURA

A. At least **5 attacks** fulfilling criteria B-D

B. Headache attacks **lasting 4-72 h** (untreated or unsuccessfully treated)

C. Headache has ≥**2** of the following characteristics:
1. **unilateral** location
2. **pulsating** quality
3. moderate or severe pain intensity
4. aggravation by or causing avoidance of routine physical activity (*eg*, walking, climbing stairs)

In those age 18 or younger, the headache may last 2-72 hours.

D. During headache ≥**1** of the following:
1. **nausea and/or vomiting**
2. **photophobia and phonophobia**

Migraine without aura is the most common migraine type, accounting for approximately **75 percent of cases**.

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MIGRAINE WITH AURA

1.2.1 Migraine with typical aura
1.2.2 Migraine with brainstem aura
1.2.3 Hemiplegic migraine
1.2.4 Retinal migraine

A. At least **2 attacks** fulfilling criteria B and C

B. ≥**1** of the following fully reversible aura symptoms:
1. **visual**
2. **sensory**
3. **speech and/or language**
4. **motor**
5. **brainstem**
6. **retinal**

Migraine without aura is the most common migraine type, accounting for approximately **75 percent of cases**.

C. ≥**2** of the following 4 characteristics:
1. ≥1 aura symptom **spreads gradually over ≥5 min, and/or ≥2 symptoms occur in succession**
2. each individual aura symptom **lasts 5-60 min**
3. ≥1 aura symptom is **unilateral**
4. aura accompanied or **followed in <60 min by headache**

D. TIA excluded

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This is her 3rd such episode in the past year.

Getting headaches 5-6x /month, not always with aura.

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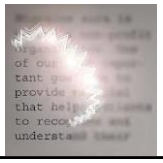
- Vision changes described as:
  - Right side of vision was blurry
    - Thinks is was both eyes
  - Parts of vision were missing
  - Distorted and shimmering
  - Lasted about **½ hour**
  - **Gradually** moved from center to the right side of her VF



Referred her to neurology, where she was diagnosed with and treated for migraine with aura.

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ABORTIVE MIGRAINE TREATMENT

Symptomatic therapy. More effective when used early in the course of the headache.

- **Simple Analgesics**
  - Aspirin, NSAIDS, Acetaminophen

Combined use of triptan and NSAID is more effective than either used alone.

- **Triptans (serotonin 1b/1d agonists)**
  - Sumatriptan, zolmitriptan, naratriptan, rizatriptan, almotriptan, eletriptan, frovatriptan
- **Antiemetics (dopamine receptor agonists)**
  - Metoclopramide, chlorpromazine, prochlorperazine
- **Selective Serotonin 1F Receptor Agonist**
  - Lasmiditan
- **CGRP Antagonists (calcitonin gene-related peptide antagonists)**
  - Rimegepant (Nurtec), Ubrogepant (Ubrelvy) - NOT 1st LINE THERAPY
- **Ergots (5-HT 1b/1d receptor agonist)**
  - Dihydroergotamine (DHE 45), Ergotamine

Abortive therapy plus parenteral dexamethasone reduces the rate of early headache recurrence.

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PREVENTIVE TREATMENT FOR MIGRAINE

- **Beta-blockers**
  - Metoprolol
  - Propranolol
  - Timolol

Beta blockers are **NOT** used in smokers or pts over age 60 due to higher rate of stroke / cardiovascular events.

Sometimes, calcium channel blockers and ACE inhibitors are used, but don't have much supportive data.

- **Antidepressants**
  - Amitriptyline (tricyclic antidepressant)
  - Venlafaxine (serotonin-norepinephrine reuptake inhibitor)
- **Anticonvulsants**
  - Topiramate
  - Valproate
- **Calcitonin gene-related peptide (CGRP) antagonists**
  - Erenumab
  - Fremanezumab
  - Galcanezumab
  - Eptinezumab

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MIGRAINE – precipitating / exacerbating factors

- Emotional stress (80 %)
- Hormones in women (65 %)
- Not eating (57 %)
- Weather (53 %)
- Sleep disturbances (50 %)
- Odors (44 %)
- Neck pain (38 %)
- Lights (38 %
- Alcohol (38 %)
- Smoke (36 %)
- Sleeping late (32 %)
- Heat (30 %)
- Food (27 %)
- Exercise (22 %)
- Sexual activity (5 %)

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1.2.2 Migraine with brainstem aura

A. At least 2 attacks

B. Aura of **fully reversible** visual, sensory and/or speech/language symptoms, but **not motor or retinal**

C. **≥2 of the following brainstem symptoms:**
1. dysarthria
2. vertigo
3. tinnitus
4. hypacusis
5. diplopia
6. ataxia
7. decreased level of consciousness

1.2.3 Hemiplegic migraine

A. At least **2 attacks**

B. Aura consisting of **both** of the following:
1. **fully reversible motor weakness**
2. fully reversible visual, sensory and/or speech/language symptoms

C. ≥2 of the following 4 characteristics:
1. ≥1 aura symptom spreads gradually over ≥5 min, and/or ≥2 symptoms occur in succession
2. each individual non-motor aura symptom lasts 5-60 min, and **motor symptoms last <72 h**
3. ≥1 aura symptom is unilateral
4. aura accompanied or followed in <60 min by headache

These are all types of migraine with aura.

Many of these would be a diagnosis of exclusion, and would need a work-up to rule out other, more sinister causes.

1.2.4 Retinal migraine

A. At least **2 attacks**

B. Aura of **fully reversible monocular positive and/or negative visual phenomena** confirmed during an attack by either or both of the following:
1. **clinical visual field examination**
2. **patient's drawing of a monocular field defect**

C. **≥2** of the following 3 characteristics:
1. aura spreads gradually over ≥5 min
2. aura symptoms last 5-60 min
3. aura accompanied or followed in <60 min by headache

D. Other causes of amaurosis fugax excluded

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- Differentiating neuro-ophthalmic disease from other non-neuro etiologies can be difficult.
- You have many tools available to help you make the distinction.
- **If you cannot rule out a neuro-ophthalmic disease process, ALWAYS err on the side of caution and REFER!**

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Thank You

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