

# Eye-conic Gazes

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## Case History

- ❖ 18-year-old Caucasian male reports for comprehensive eye exam and complains of blurry vision after prolonged near work
- ❖ Full-time high school senior student using primarily textbooks for homework
- ❖ 4 hours daily screen time
- ❖ PMHx: Unremarkable
- ❖ FMHx: Unremarkable
- ❖ Medications: None
- ❖ Allergies: None

## Pertinent Findings

- ❖ BCVA 20/20 OD and 20/20 OS
- ❖ Pupils round and reactive to light with no APD
- ❖ Confrontation Fields: unremarkable
- ❖ EOMS: -4 under action in lateral and medial gazes with retraction OD, OS
- ❖ Constant slight right head tilt
- ❖ Cover Test: ortho at distance and near with corrected head posture
- ❖ Maddox Rod:
  - Horizontal - fluctuating crossed and uncrossed diplopia at distance, uncrossed at intermediate and near
  - Vertical - 5 Lt Hyper
- \*fluctuating results due to patient assuming compensatory head posture and eyes trying to maintain binocularity
- ❖ Stereo: Global – nil, Local – 200'

## References

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## Differential Diagnoses

- ❖ Duane Syndrome Type I, II, III
- ❖ Moebius Syndrome
- ❖ Okiihiro Syndrome
- ❖ Wildervanck Syndrome
- ❖ Goldenhar Syndrome
- ❖ Sixth Nerve Palsy
- ❖ Brown Syndrome
- ❖ Fourth Nerve Palsy
- ❖ One and a half syndrome
- ❖ Internuclear ophthalmoplegia
- ❖ Congenital/infantile esotropia

## Associated Findings

- ❖ Amblyopia
- ❖ Strabismus
- ❖ Marcus Gunn jaw-winking phenomenon
- ❖ Crocodile tears
- ❖ Head turn or tilt
- ❖ Convergence insufficiency
- ❖ Globe retraction
- ❖ Upshoot/Downshoot

## Diagnosis Discussion

Duane Retraction Syndrome (DRS) presents as a unilateral or bilateral horizontal eye movement deficit with globe retraction and narrowing of palpebral fissure upon attempted adduction. DRS is congenital and thought to be an absence of abducens nerve and aberrant misinnervation of the lateral rectus by axons of oculomotor nerve. It can be caused by a CHN1 mutation, seen in ~5% of patients who have a parent with DRS. The etiology remains unclear and ~90% of patients have no family history. There are 3 types of DRS with variable clinical presentations, but all present with globe retraction and narrowing of palpebral fissure upon attempted adduction. Type I is the most common and is characterized by an abduction deficit, Type II is the rarest and presents with an adduction deficit, and Type III presents with both an abduction and adduction deficit. This case represents Type III DRS, which comprises about 15% of Duane patients. About 30% of DRS patients also have other congenital anomalies.

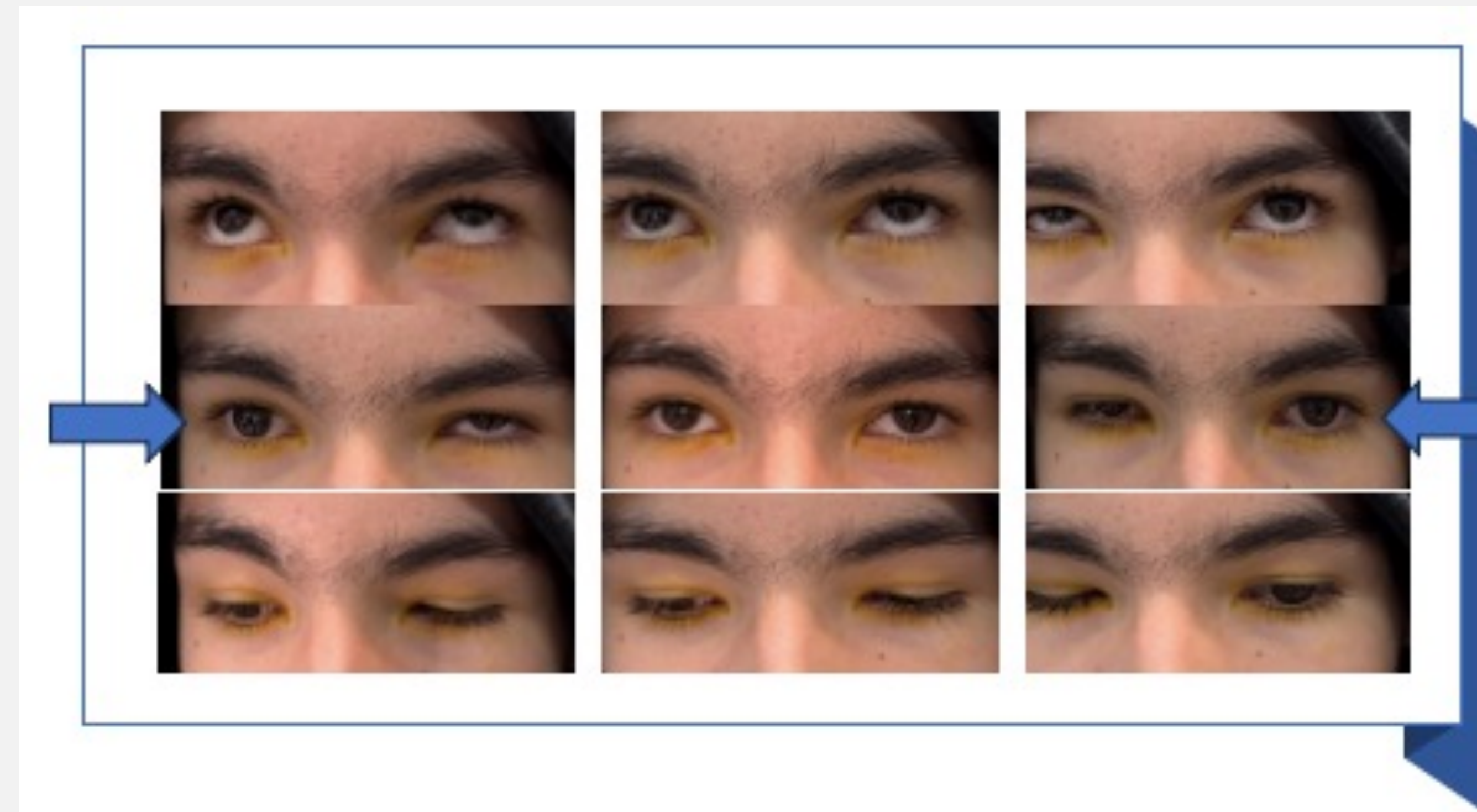


Figure 1. Duane Syndrome Type III in 9 gazes

## Treatment/Management

Due to the congenital nature of Duane Syndrome, many patients have developed compensatory head turns to eliminate diplopia or other binocular vision symptoms. It is appropriate to educate and monitor patients who have no complaints. For symptomatic patients, accommodations may be recommended. For example, a Left Type I patient may need to sit on the left side of the classroom and place reading material on the right side. For a patient with accompanied convergence insufficiency due to adduction deficits, a slant board and bar magnifier may help for near work. Prism may be used to eliminate diplopia. Yoked prism can be used to improve a compensatory head turn or strictly move the image into the patient's field of vision. Vision therapy can improve accommodative and vergence skills thereby relieving symptoms and improving binocularity. Surgery is recommended in severe cases to eliminate or improve compensatory head posture, misalignment, retraction, and upshoots/downshoots. In this case, a slant board and bar magnifier were re-introduced to the patient to utilize during near work as needed. If symptoms persist, vision therapy and/or base in prism would be recommended.

## Conclusion

Differentiating Duane Syndrome from more threatening diagnoses can be challenging but is essential. Associated congenital anomalies should be recognized and a systemic evaluation and genetic analysis recommended. Addressing symptoms is important in these patients and can be done via accommodations, prism, vision therapy, or surgery.



## Background

Granulomatous uveitis is characterized as a chronic condition leading to inflammation of the uveal tract. It is poorly understood which particular systemic condition will lead to certain presentation of chronic uveitis. The treatment of uveitis can be convoluted due to the high association of systemic etiology. Determining infectious or inflammatory and determining the underlying cause is crucial to long term management. Prognosis is dependent on proper treatment of causative systemic conditions and co-management with appropriate providers to manage the systemic condition with ocular sequelae.

## Case History

A 36 year old African American female presented for an annual comprehensive exam. The chief complaint was mild constant blurry vision of the left eye more than her right through her current habitual spectacle for around six months. She also reported longstanding mild light sensitivity in both eyes. Patient reports a history of chronic granulomatous uveitis, optic nerve head edema, epiretinal membrane and cystoid macular edema of both eyes. She reports receiving kenalog injections in both eyes with mild improvement in vision around one month prior to visit with an outside specialist.

Her corrected distance visual acuity (VA) was 20/20 in the right eye (OD) and 20/30 in the left eye (OS) with pinhole no improvement. Her pupils were irregular and minimally reactive to light with no relative afferent pupillary defect. Upon slit lamp examination, her lids and lashes were normal. The right eye (OD) presented white and quiet conjunctiva, 2+ inferior superficial punctate keratitis, clear stroma, large mutton fat keratic precipitates (KP) on the endothelium, deep chamber with 1+ cell, koepple nodules, irregular pupil with posterior synechiae from three to eight clock hours. The left eye (OS) presented with 1-2+ circumlimbal injection, 2+ inferior superficial punctate keratitis, clear stroma, large mutton fat KP on the endothelium, shallow chamber with 2+ cell, 1+ flare, koepple nodules, irregular pupil with convex iris approach and iris bombe secondary to posterior synechiae from one to 12 clock hours. The intraocular pressure was 37 mm Hg OD and 50 mm Hg OS at 2:36 PM measured by applanation after installation of fluorescein sodium and benoxinate hydrochloride 0.25%/0.40% to each eye. After consulting with an internal ophthalmologist on site, the patient was given one round Simbrinza 1%/0.25% and 250 mg Diamox PO. The patient consented to dilation with 1% Tropicamide/ 2.5% Phenylephrine and 1% Atropine in both eyes in an attempt to break the posterior synechiae. Her crystalline lens reveals trace nuclear sclerosis and diffuse pigment on anterior lens capsule. The vitreous showed 2+ cells in the posterior chamber. There was a limited view of the fundus due to poor dilation secondary to iridolenticular adhesion.

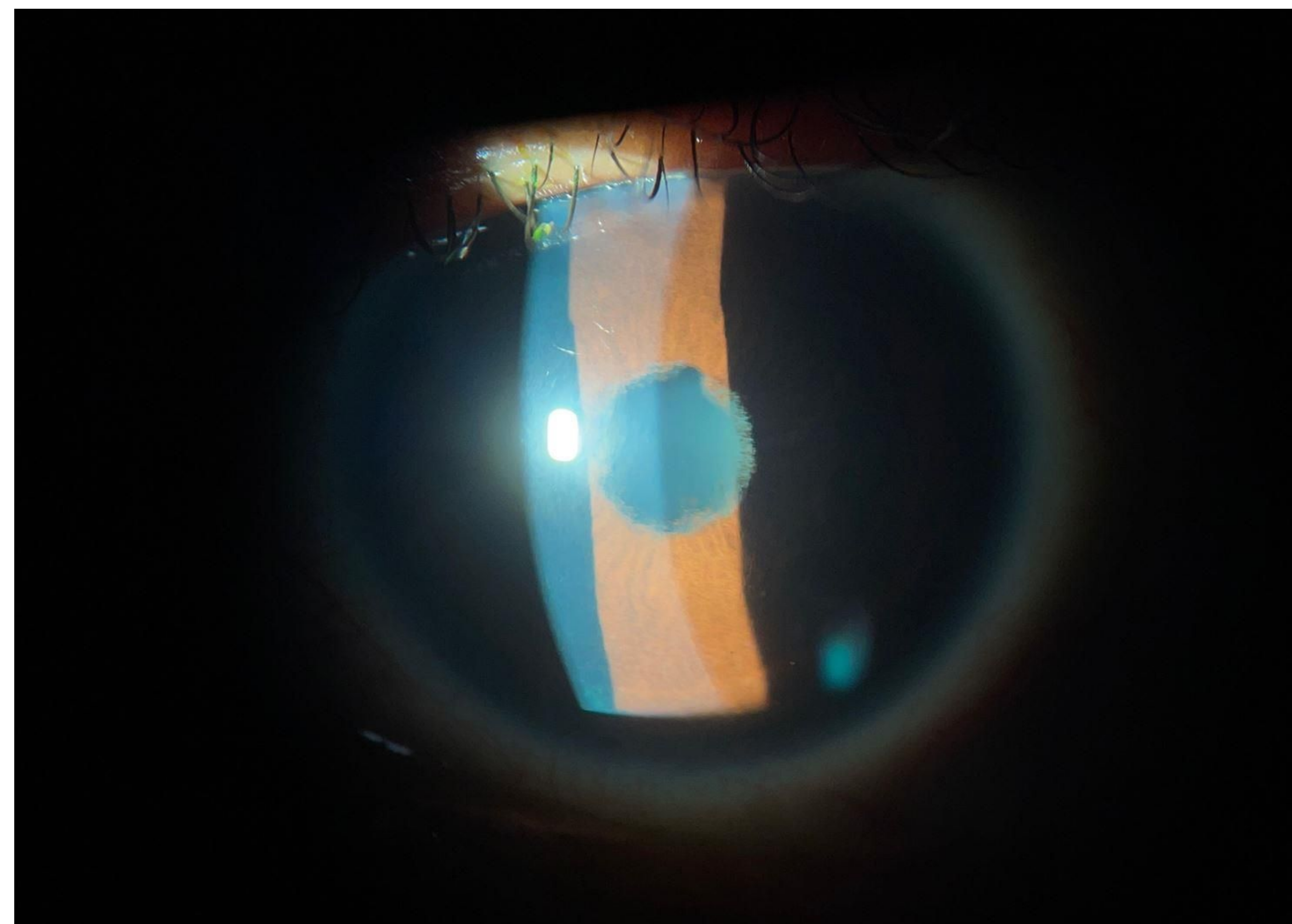


Figure 1: The left eye presented in clinic with posterior synechiae 360 degrees upon initial examination.

## Treatment and Management

The patient had an emergent LPI (Laser Peripheral Iridotomy) performed in the office on the left eye the same day to create another area for outflow through the iris. Considering the urgency and nature of the condition, the patient was to continue on Diamox 250 mg twice daily along with max topical medications to reduce intraocular pressure. The patient was scheduled for next available cataract surgery. During this time, the patient was to return to rheumatology for evaluation, obtain past medical records and begin a more aggressive systemic intervention for underlying conditions. Upon reviewing medical records, it was discovered that the patient had a history of active tuberculosis in the past and was treated with a course of TB medications. She was later on diagnosed with scleroderma and rheumatoid arthritis (RA). Co-management with rheumatology became crucial to maintain systemic control and prevent ocular flare ups in the future, requiring acute communication between each managing physician. After cataract surgery and iridotomy was performed in both eyes, the pressure began to improve significantly. The patient was to continue on Simbrinza twice daily along with Pred Forte once daily. At this point, the patient began a new course of systemic medications and was able to control ocular and systemic symptoms.

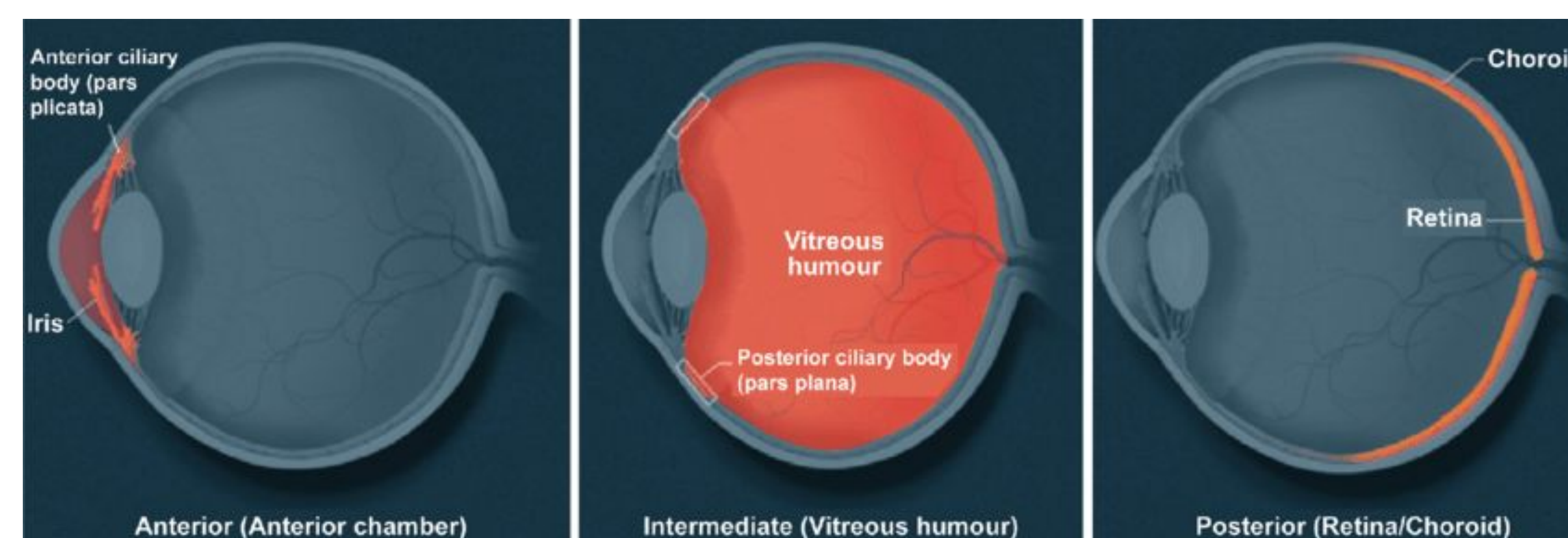


Figure 2: Uveitis may affect different parts of the eye. In this particular case, both the anterior and intermediate segments of the eye were affected.

## Discussion

Uveitis is inflammation of the uveal tract, affecting different segments of the eye. Presentation may be acute or chronic, and it will present in different manners. Understanding the different signs within the eye can point towards whether presentation is due to an infectious, traumatic or inflammatory cause. In this particular case, the patient had a longstanding history of both infectious and inflammatory systemic conditions leading to the presentation seen on initial examination. The patient has presented with chronic findings in the anterior and intermediate segment, causing chronic adhesions of the iris to the lens, leading to the formation of iris bombe. It is also of note that the long term inflammation present on examination may not correlate with patient symptoms as it would in an acute attack of uveitis. The importance of careful slit lamp examination plays a role in understanding and differentiation of type of uveitis present. As our understanding and management of systemic etiology grows, our understanding of uveitis and proper management will grow as well. Most importantly, the co-management between rheumatology and primary care providers becomes increasingly prevalent in the proper management of these cases.

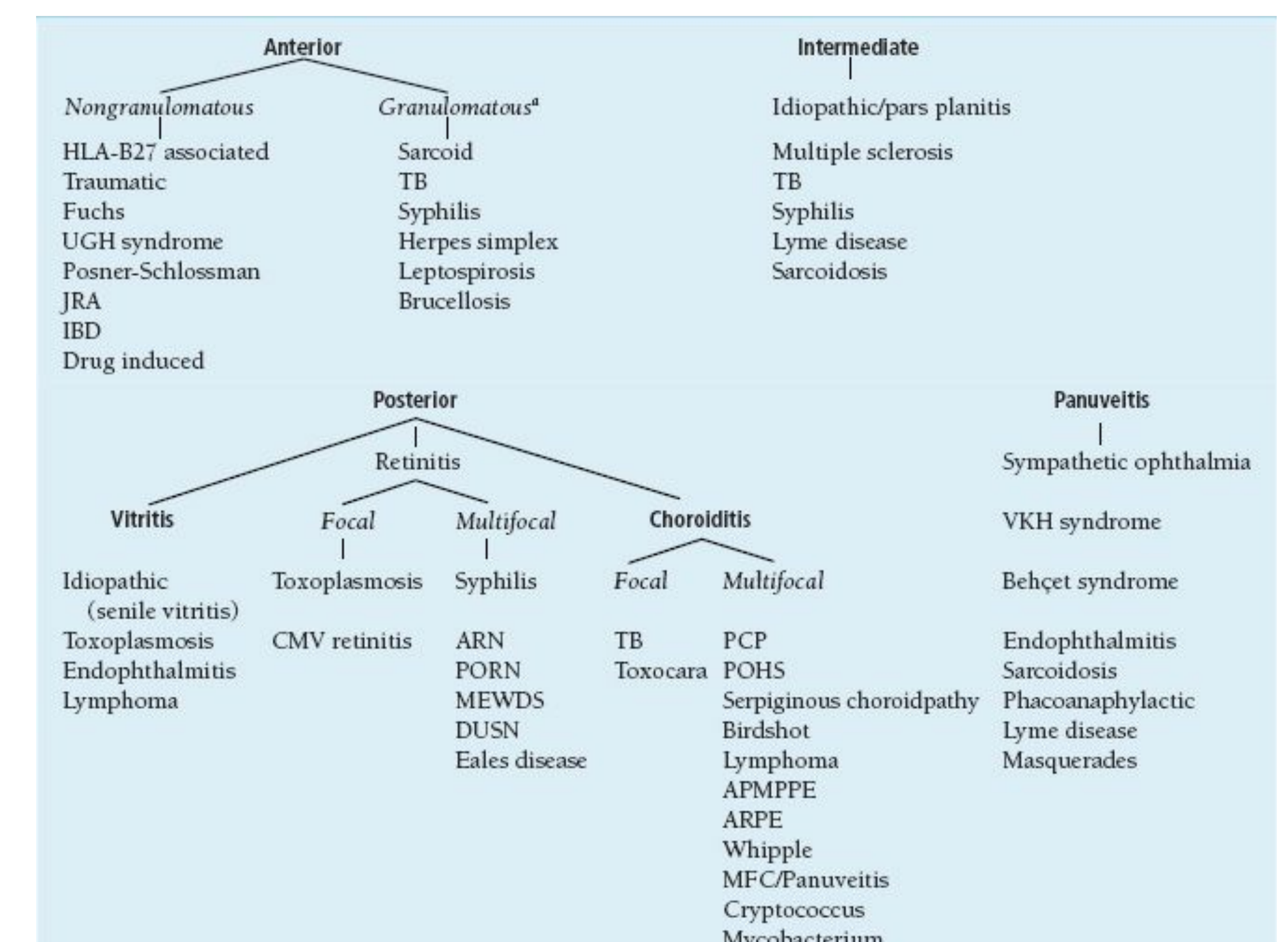


Figure 3: This diagram indicates the many variations of uveitis and which particular presentation is most commonly associated with each systemic condition.

## Acknowledgments and Conflicts of Interest

Eric Fazio O.D.




## References

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- <https://entokey.com/4-intraocular-inflammation-and-uveitis/>



## Atypical Corneal Lesion Masquerading as Herpetic Keratitis




Alberta Pengo, OD

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## Financial Disclosures




- None

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## Case History




- 54-year-old female presents for painless, blurred vision in the right eye for 2 months
- Past ocular history: Unremarkable
- Past medical history: Unremarkable
- Previous smoker, quit 10 years ago
- No known medical allergies

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## Base Eye Exam

- BCVA: 20/30 OD, 20/20 OS
- Pupils: PERRL, (-) APD OD, OS
- EOMS: Full
- CVF: FTFC 4Q
- IOP: 19 mmHg OD, 19 mmHg OS




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## Anterior Segment Exam

Slit Lamp Exam	Right	Left
External	Demodex, Meibomian gland dysfunction	Demodex, Meibomian gland dysfunction
Lids/Lashes	Diffuse bulbar injection, Temporal pterygium	Normal
Conjunctiva/Sclera	Dendritic-like lesion involving the visual axis	White and quiet
Cornea	Clear	Clear
Anterior Chamber	Deep and quiet	Deep and quiet
Iris	Round and flat	Round and flat
Lens	Trace nuclear sclerosis	Trace nuclear sclerosis

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### Posterior Segment Exam

Slit Lamp Exam	Right	Left
Vitreous	Normal	Normal
Disc	Pink and Healthy	Normal
C/D	0.45	0.45
Macula	Normal, Flat	Normal, Flat
Vessels	Normal	Normal
Periphery	Normal	Normal

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### Diagnosis: Herpes Simplex Keratitis

- Start oral Acyclovir 500mg 3 times a day for 10 days
- Return to clinic in 5 days

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### 5 day Follow Up

- VA OD: 20/30, PH 20/25
- No improvement of the corneal lesion
- Acyclovir treatment was extended to a two-week course

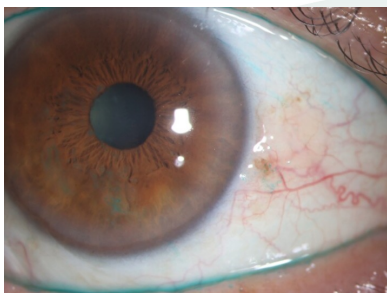
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### 2 Week Follow Up

- VA OD: 20/30, PH 20/25
- No improvement
- Slit lamp exam: grey-white opalescent lesions with finger-like projections
- (+) Staining with Lissamine Green



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### Differential Diagnosis

- Herpetic Keratitis
- Pannus
- Corneal Scar
- Map Dot Dystrophy
- Salzmann Nodular Degeneration
- Ocular Surface Squamous Neoplasia

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Next Step?

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Anterior Segment Ocular Coherence Tomography

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AS-OCT: Conjunctiva

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Diagnosis: Corneal Ocular Surface Squamous Neoplasia

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Learning Objectives

- Review Ocular Surface Squamous Neoplasia (OSSN)
- Ocular presentation of OSSN
- Interpret High-Resolution Anterior Segment OCT (AS-OCT)
- Discuss the management options

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Ocular Surface Squamous Neoplasia (OSSN)

- The most common non-pigmented tumor of the ocular surface
- Abnormal growth of dysplastic squamous epithelial cells of the conjunctiva and/or cornea
- Slow-growing tumors that rarely metastasize but can cause local tissue destruction
- Spectrum of tumors:
  - Conjunctival/Corneal Intraepithelial Neoplasia (CIN)
  - Carcinoma in Situ (CIS)
  - Squamous Cell Carcinoma (SCC)

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### OSSN "Umbrella" Term

Normal	CIN1	CIN2	CIN3	Invasive Cancer

**Squamous cells**  
**Basal cells**

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### Pathophysiology

- OSSN arises from basal epithelial cells
  - limbal epithelial crypts located in the basal layer of limbal stem cells
- Corneal involvement begins at limbus and spreads centrally

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### Etiology

- UV Exposure
  - Populations within 30-degree latitude from the equator
- Age
- Cigarette smoking
- Xeroderma pigmentosum (Vitamin A deficiency)
- Human Papilloma Virus (Strains 16 and 18)
- HIV/Immunosuppression
  - Karp et al: half of patients under the age of 50 diagnosed with OSSN were seropositive for HIV

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### Ocular Presentation

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### OSSN: Conjunctival Presentation

- Limbal location: Nasal
  - Highest exposure to sunlight
- Irregular Borders
- Abnormal Vessels:
  - Branching
  - Hairpin loops
- Three presentations

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### OSSN: Conjunctival Presentation

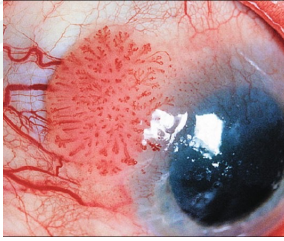
- Gelatinous lesions (Most Common)
  - Nodular
    - Defined margins
    - Rapid growth
  - Diffuse
    - Resembles chronic conjunctivitis

Basu, S.S., Chakraborty, S., Basu, C., Chakraborty, S., Chakraborty, S. (2022). Review of the molecular and clinical epidemiology, staging systems and treatment of Ocular Squamous Neoplasia. *Journal of Ocular Inflammation and Immunology*, 15(1), 1-10. doi:10.1186/s12944-022-00100-0

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### OSSN: Conjunctival Presentation

- Papillomatous lesions
  - Pedunculated
  - Stippled red appearance
  - Fibrovascular core

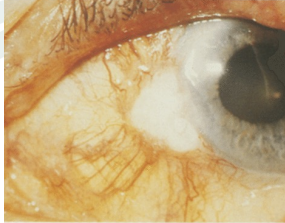


Smith, C.L. & Shields, J.A. (2018). Conjunctival Tumors of the eye and orbit. In: Atlas of ophthalmology. Elsevier, pp. 100-110. https://doi.org/10.1016/j.yes.2018.05.001

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### OSSN: Conjunctival Presentation

- Leukoplakic
  - 10% of lesions
  - Thickened, white conjunctiva
  - Hyperkeratinization

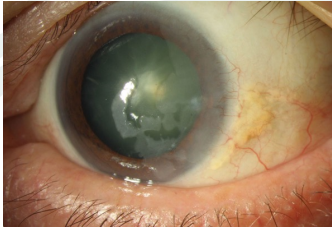


Lee, G.S. & Wink, L.W. (2005). Ocular surface squamous neoplasia. Survey of Ophthalmology, 50(3), 439-450. https://doi.org/10.1016/j.surv.2005.05.001

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### OSSN: Corneal Presentation


- Opalescent
- Avascular superficial corneal opacity
  - Feathery/irregular borders
- Feeder vessel: suspicious for SCC



Scott, (2002, August 27). Prevalence of ocular surface squamous neoplasia increases with age. EyeWorld. Retrieved from https://www.eyeworld.com/2002/08/27/prevalence-of-ocular-surface-squamous-neoplasia-increases-with-age.aspx

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### Diagnosis



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### OSSN: Diagnosis

- Gold Standard: Biopsy
- Impression Cytology: non-invasive, superficial tissue
- Fine needle aspiration: non-invasive, nonspecific
- Diagnostic Dyes
  - Lissamine Green
  - Rose Bengal
  - Trypan Blue
- High Resolution AS-OCT

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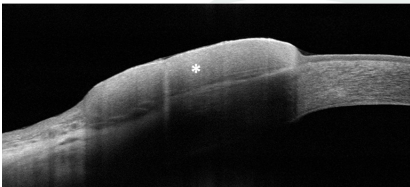
### High-Resolution Anterior Segment OCT



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### AS-OCT: Conjunctiva

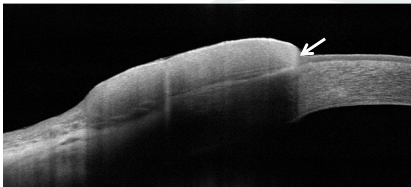


Thickened hyper-reflective epithelium with an abrupt transition line and a distinct plane between the lesion and the underlying tissue

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### AS-OCT: Conjunctiva

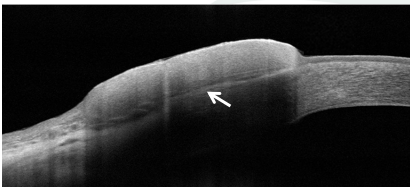


Thickened hyper-reflective epithelium with an abrupt transition line and a distinct plane between the lesion and the underlying tissue

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### AS-OCT: Conjunctiva



Thickened hyper-reflective epithelium with an abrupt transition line and a distinct plane between the lesion and the underlying tissue

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### High-Resolution Optical Coherence Tomography as an Adjunctive Tool in the Diagnosis of Corneal and Conjunctival Pathology

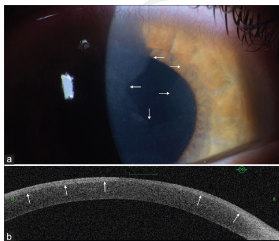
Afshan A. Nanji MD, MPH<sup>1</sup>, Fouad E. Savvyad MD<sup>1</sup>, Anat Galor MD, MSPH<sup>1,2</sup>, Sander Dubovy MD<sup>1,3</sup>, Carol L. Karp MD<sup>1</sup>

Using **120 μm** as a cutoff, the sensitivity of HR-OCT for differentiating between OSSN and pterygia with **100% sensitivity and 100% specificity**.

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### AS-OCT: Cornea



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### Treatment Options

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## Surgery

Wide excision with cryotherapy and closure with amniotic membrane

**Pros**

- Gold standard
- Diagnostic and therapeutic
- Small or localized lesions

**Cons**

- Residual lesion
- Scarring
- Symblepharon
- Limbal stem cell deficiency

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## Topical Therapy

**Pro**

- Treats the entire lesion
  - Diffuse lesions
  - Recurrent
- Fewer side effects
- Well-tolerated

**Con**

- Longer treatment time
- Off-label (Not FDA approved)
- Side effects:
  - Limbal stem cell deficiency
  - Secondary infections
  - Punctal stenosis

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## Topical Therapy

	MOA	Cost	Side Effects	Considerations
Mitomycin-C	Alkylating agent	\$400/bottle Compounding pharmacy	Painful, Significant ocular toxicity	Punctal Plug Topical Steroid PF AT
5-Fluorouracil 1%	Cytostatic drug	\$50	Less painful and less ocular toxicity than MMC	Topical Steroid PF AT
Interferon alpha-2-b	Immune modulator	\$600/month Compounding pharmacy	Minimal Flu-like symptoms	PF AT Acetaminophen

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## Topical Chemotherapy Regimen

	Interferon alpha-2b	5-Fluorouracil	Mitomycin C
<b>Regimens</b>	<b>Drops:</b>	<b>Drops:</b>	<b>Drops:</b>
	<ul style="list-style-type: none"> <li>• 1 million IU/mL QID until resolution (mean, 4 months)(also 1-3MIU/ml used) [2,3,58-61]</li> </ul>	<ul style="list-style-type: none"> <li>• 1% QID for 1 week followed by 3 weeks off medication [22](mean, 4 cycles)</li> <li>• 1% QID for 2-4 days followed by 30 days off medication [10,60]</li> <li>• 1% QID for 4 weeks continuously with 3 months between courses. [68]</li> </ul>	<ul style="list-style-type: none"> <li>• 0.04% QID for 1 week followed by 3 weeks off medication(mean, 4 cycles) [30].</li> <li>• 0.04% QID 1 week on 1 week off [38,39][11]reported range 2 to 6 cycles)</li> <li>• 0.02% QID for 2-4 weeks [23]</li> <li>• 0.002% 4 times daily until resolution [22]</li> </ul>
	<b>Injections:</b>		
	<ul style="list-style-type: none"> <li>• 3 million IU/5cc injection/1-3*week (mean, 4-5 weeks) or 10 million IU/month [55] [56]</li> </ul>		

40

## Surgical versus Topical Therapy

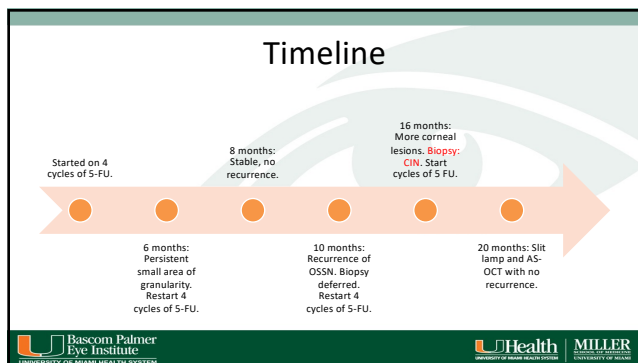
- Topical chemotherapy has become favored among corneal specialists
  - Topical therapy treats the entire ocular surface with fewer associated side effects
- Recurrence rates are similar between surgical and medication management

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## Back to our Patient

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### Conclusion

- OSSN is the most common non-pigmented tumor of the ocular surface
- Variable presentation and can masquerade as other benign and malignant pathologies of the cornea or conjunctiva
- Biopsy is the gold standard for diagnosis, but AS-OCT is a non-invasive tool to confirm the diagnosis of suspicious corneal and conjunctival lesions
- AS-OCT: three classic features and epithelial thickness of greater than 120 microns is pathognomonic for OSSN
- Topical chemotherapy is the preferred treatment option for corneal specialist

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### Thank You

- Dr. Carol Karp
- Dr. Lily Zhang
- Dr. Andrew Rouse
- Fellow Co-Residents

¿Preguntas?

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An aerial photograph of a city, likely New York City, showing a dense urban landscape with many buildings and streets. The image is framed by a circular vignette effect, making the center of the city more prominent and the edges fade out. A black banner with white text is overlaid on the top half of the image.

**“I only see half of the vision chart”**

**Yuwei Wang, O.D., M.S.  
Ocular Disease Resident  
Braverman Eye Center**



**No financial disclosures**

# Clinical Presentation

- 40 year old hispanic female
- CC: suddenly “lost” vision and seeing “white cloud” OU for the last 2 weeks
- Past Ocular History: unremarkable
- Family History: unremarkable
- Past Medical History: Hypertension; High cholesterol; Migraine; Fibromyalgia; Sinusitis; Arthritis
- Systemic Medications: Losartan; Amlodipine; Lisinopril; Gabapentin
- Allergies: NKDA
- Ocular Medications: none



**Visual Acuity - Patient reports seeing only half of the chart**

	OD	OS
Dist cc	20/80+1	20/70+1
Dist ph cc	20/70+1	NI
Near sc	J3 -2	J5

**Refraction**

	OD	OS
Current Rx (DV)	-2.75 sph	-2.50 -0.50 x 144
Manifest	-3.00 sph	-2.75 -0.50 x 145
BCVA	20/70+1	20/70+1

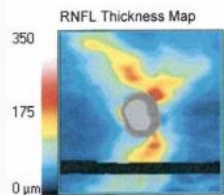
<b>Entrance tests</b>		
	OD	OS
Pupil	ERRL (-)APD	
EOMs	FULL; no diplopia	
CF	FTFC	
Cover Test	Ortho @ D & N	
Pressure(GAT@3:08pm)	18	20

<b>Blood Pressure</b>		
	BP	Repeated BP
Automatic	178/126	186/122

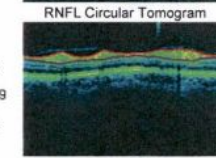
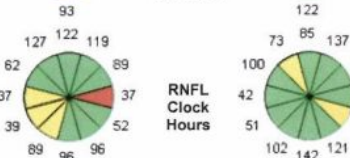
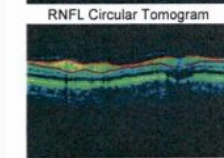
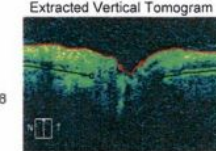
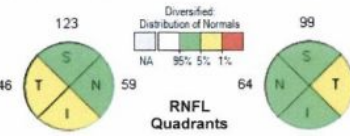
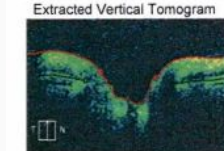
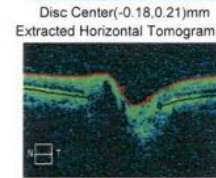
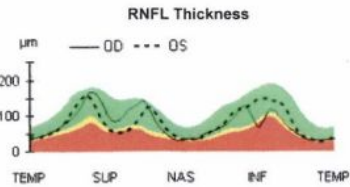
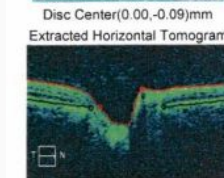
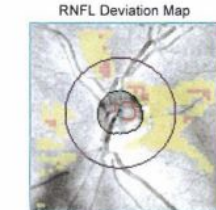
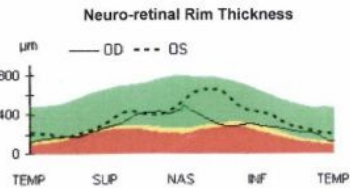
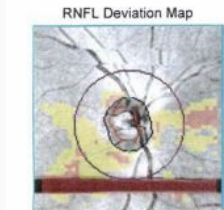
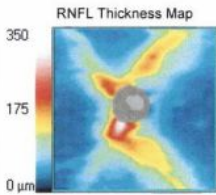


<b>Slit Lamp Exam</b>		
	OD	OS
Lids/Lashes	Decreased TM	Decreased TM
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior Chamber	Deep and quiet	Deep and quiet
Iris	Round and flat	Round and flat
Lens	Trace NS/ALCS	Trace NS/ALCS
<b>DFE - Unremarkable for vitreous/macula/vessels/peripheral</b>		
	OD	OS
Optic Nerve Head	C/D 0.20/0.20	C/D 0.15/0.15

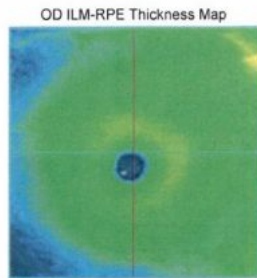
# ONH and RNFL OU Analysis: Optic Disc Cube 200x200 OD ● ● OS



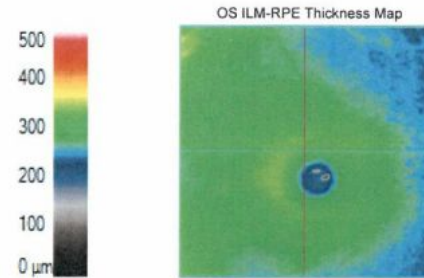
	OD	OS
Average RNFL Thickness	80 $\mu\text{m}$	93 $\mu\text{m}$
RNFL Symmetry	77%	
Rim Area	1.15 mm <sup>2</sup>	1.23 mm <sup>2</sup>
Disc Area	1.96 mm <sup>2</sup>	1.53 mm <sup>2</sup>
Average C/D Ratio	0.63	0.44
Vertical C/D Ratio	0.64	0.40
Cup Volume	0.251 mm <sup>3</sup>	0.049 mm <sup>3</sup>



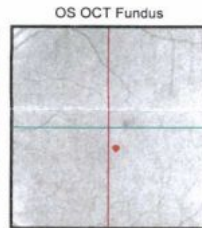
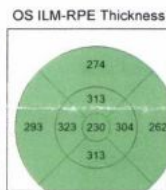
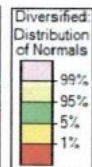
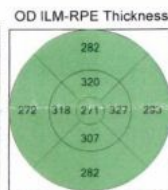
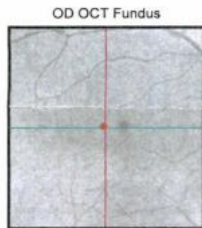
# Macula Thickness OU: Macular Cube 200x200 OD ● ● OS



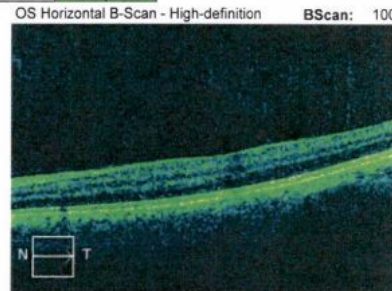
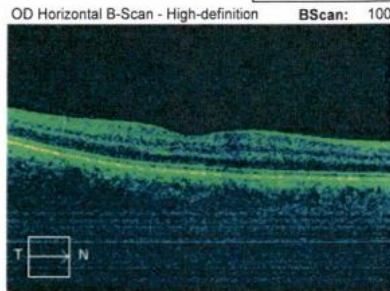
Fovea: Not found



Fovea: 111, 122



ILM - RPE	OD	OS
Thickness Central Subfield ( $\mu\text{m}$ )	271	230
Volume Cube (mm <sup>3</sup> )	10.3	10.1
Thickness Avg Cube ( $\mu\text{m}$ )	286	281





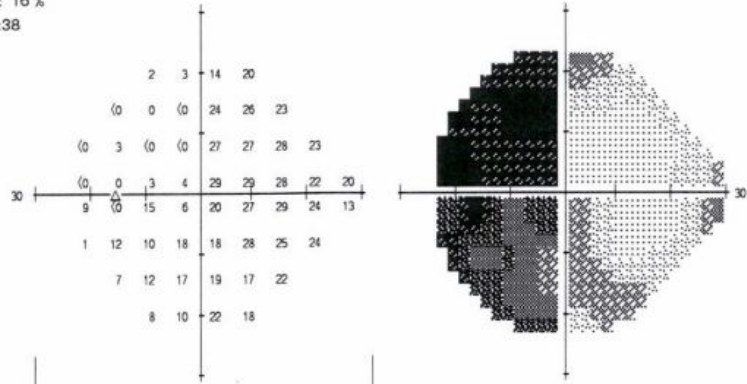
Fixation Losses: 0/12  
 False POS Errors: 0 %  
 False NEG Errors: 16 %  
 Test Duration: 04:38

Strategy: SITA-Fast

RX: DS DC X

Age: 40

Fovea: OFF

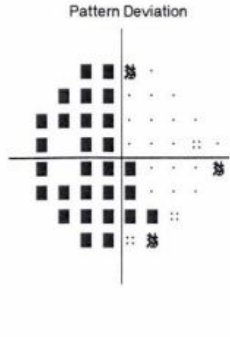


-26	-25	-15	-8				
-32	-30	-33	-7	-4	-7		
-32	-28	-34	-5	-6	-4	-8	
-33	-30	-30	-4	-5	-4	-9	-8
-23	-18	-28	-14	-6	-4	-7	-16
-30	-20	-22	-15	-16	-5	-7	-6
-25	-19	-15	-14	-14	-9		
-22	-21	-9	-12				

-21	-21	-10	-4				
-27	-25	-28	-2	0	-2		
-28	-23	-29	-30	0	1	1	-3
-28	-25	-25	0	0	0	-4	-4
-18	-14	-23	-9	-2	1	-2	-11
-25	-15	-18	-10	-11	0	-2	-2
-20	-15	-11	-9	-10	-4		
-18	-17	-4	-7				

GHT  
 Outside Normal Limits

VFI 54%  
 MD -15.21 dB P < 0.5%  
 PSD 10.66 dB P < 0.5%



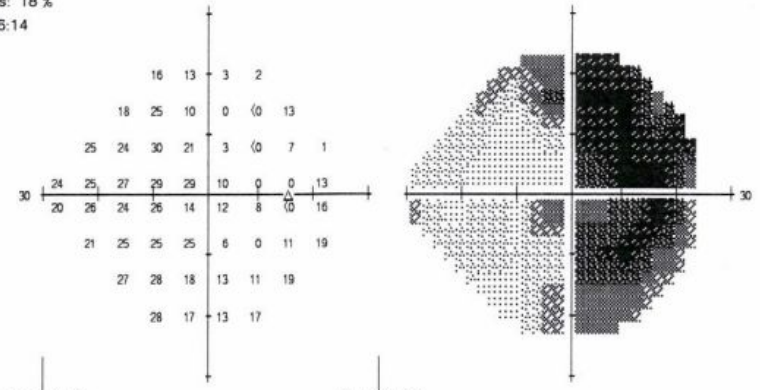
Fixation Losses: 1/14  
 False POS Errors: 2 %  
 False NEG Errors: 18 %  
 Test Duration: 06:14

Strategy: SITA-Fast

RX: DS DC X

Age: 40

Fovea: OFF

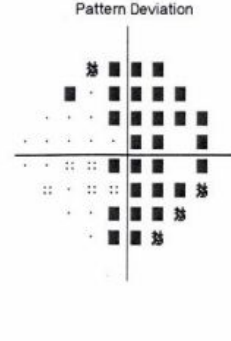
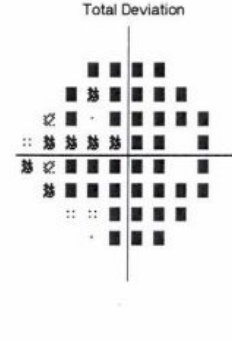


-12	-16	-25	-26				
-12	-6	-21	-31	-32	-17		
-5	-8	-2	-11	-29	-34	-24	-30
-5	-5	-5	-4	-23	-33	-18	
-9	-5	-8	-8	-20	-22	-25	-15
-9	-7	-8	-8	-27	-33	-21	-12
-4	-3	-14	-19	-21	-13		
-2	-13	-17	-13				

-8	-11	-21	-22				
-8	-1	-16	-26	-27	-12		
-1	-3	2	-7	-25	-29	-20	-25
0	-1	-1	0	0	-19	-28	-14
-5	0	-3	-3	-17	-21	-11	
-5	-2	-3	-3	-22	-28	-16	-7
1	1	-10	-14	-16	-8		
2	-9	-13	-9				

GHT  
 Outside Normal Limits

VFI 54%  
 MD -14.51 dB P < 0.5%  
 PSD 9.93 dB P < 0.5%



# Differential Diagnosis: Bitemporal Hemianopsia

## Lesions of the chiasm

- Pituitary Adenoma (most common)
- Hypothalamic Glioma (more common)
- Meningioma (less common)
- Craniopharyngioma (rare)
- Aneurysm
- Pituitary Apoplexy
- Traumatic Chiasmal Syndrome

## Others

- Tilted Disc Syndrome
- Bilateral Nasal Staphyloma
- Nasal Retinitis Pigmentosa

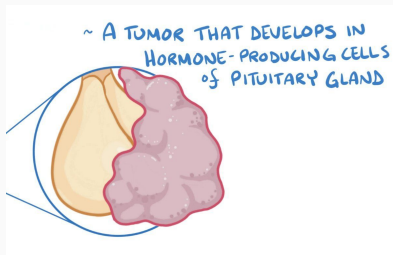


# Differential Diagnosis: Bitemporal Hemianopsia

Most common

## Pituitary Adenoma

- Benign
- 15% of all intracranial neoplasms



## Meningioma

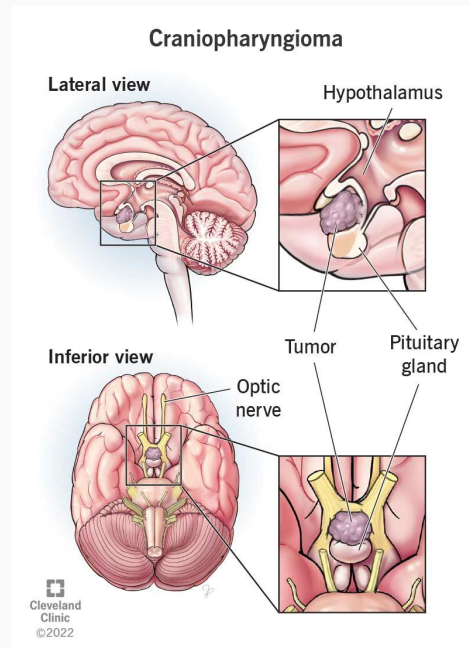
- Benign
- Most common primary brain tumor

## Hypothalamic Glioma

- Mostly Benign
- Young children
- 2% of all CNS tumors

Rare

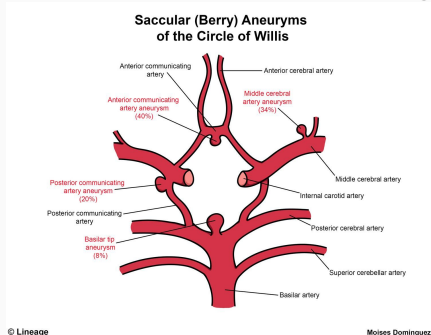
## Craniopharyngioma



# Differential Diagnosis: Bitemporal Hemianopsia

## Aneurysm

- Rare
- Anterior cerebral artery, internal carotid artery, or anterior communicating artery

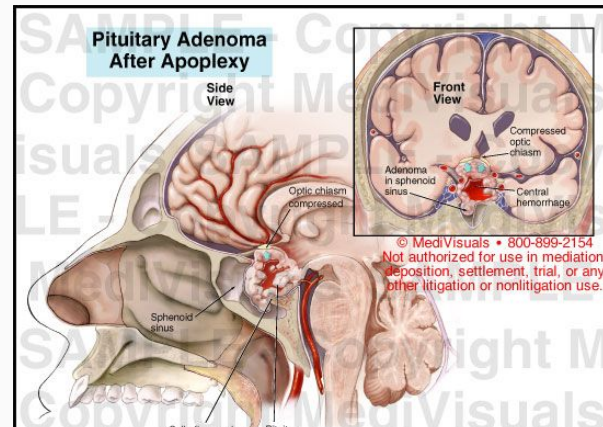


## Pituitary Apoplexy

- Rare
- Acute hemorrhagic infarction and acute swelling

## Traumatic Chiasmal Syndrome

- Rare
- High-velocity head trauma





## **Next Step**

**Refer to neuro  
ophthalmologist**

# Neuro-Ophthalmology Visit The Day After

CC/HPI/Review of system		
Headache x 2 months	Nausea x 1 week	Balance difficulty
Neck stiffness/pain	Retrobulbar pain OD>OS	Fatigue/ Muscle weakness

	OD	OS
BCVA	20/60	<b>20/400</b>
Color Vision	0/8	0/8
Confrontational Fields	Normal	Abnormal(Sup/temp restriction)
Optic Nerve Head	C/D 0.20/0.20	C/D 0.15/0.15
	1+ temporal pallor	1-2+ temporal pallor

# Diagnostic Impression

## Bitemporal Hemianopsia

- likely secondary to pituitary mass
- Loss of vision OS>OD
- Dramatic decrease in vision OS over two days
- Color vision loss OU

# Management Plan

- Sent to ER for same day MRI (pituitary) and surgical evaluation
- Follow up 1 week for repeat HVF and vision/pupil check



## The Same Day ER Story...

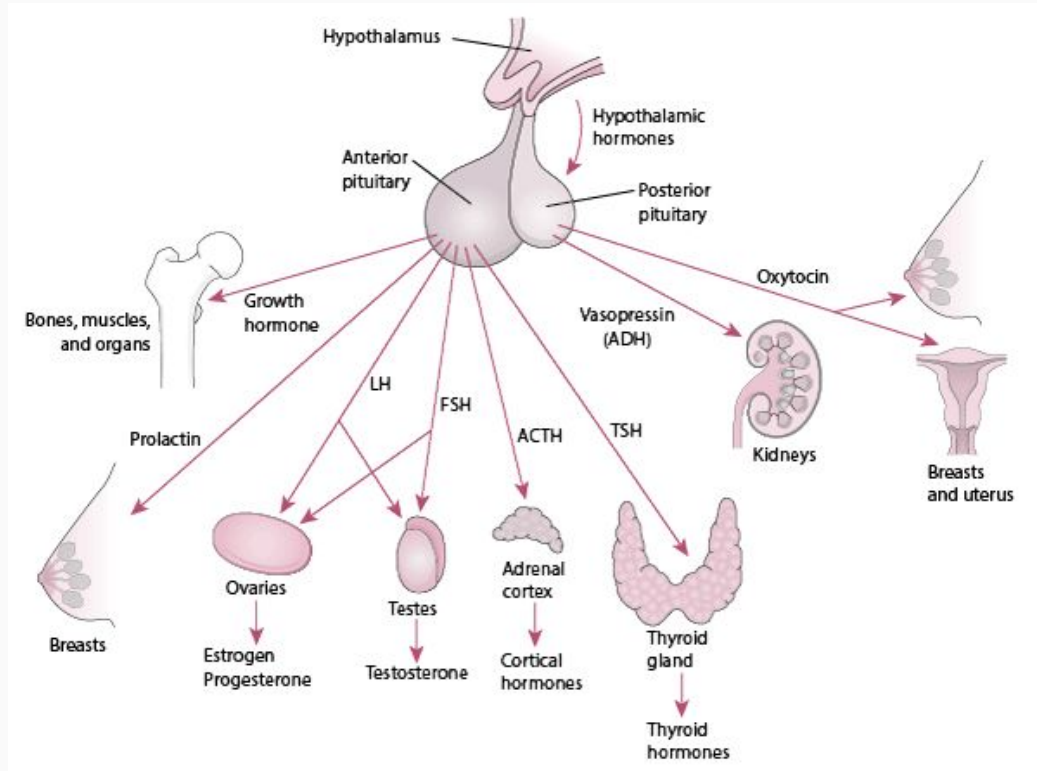
- Upon admission and after labs were sent, emergency MRI demonstrated a 2x1.5 cm pituitary mass with cystic severe compression of the optic chiasm: **at least 15 cc of necrotic thick yellow fluid under pressure with additional hemorrhagic tissue lining the internal aspect of this large pituitary apoplexy.**
- Notified the OR that the patient needed to proceed as soon as possible with emergency transsphenoidal surgery

# Diagnoses and Procedure Performed

- Preoperative diagnoses
  1. Pituitary apoplexy with rapid progressive visual loss bilaterally with dense bitemporal hemianopsia and visual acuity loss
  2. Rapid progressive visual loss with severe headache and pituitary apoplexy
- Assessments
  1. HTN 2. Pituitary apoplexy 3. Pituitary macroadenoma 4. Bitemporal hemianopia
- Procedure performed
  1. IV Decadron (dexamethasone) to cover adrenal function
  2. Emergency transsphenoidal microscopic removal of pituitary macroadenoma, status post pituitary apoplexy.

# Pituitary Labs

- **Cortisol**
- TSH
- T4/T3
- FSH
- Luteinizing Hormone
- **Prolactin**
- Human growth hormone
- ACTH
- Electrolytes
- Glucose



<https://www.merckmanuals.com/home/hormonal-and-metabolic-disorders/pituitary-gland-disorders/overview-of-the-pituitary-gland>



# 4 week Follow up

## Visual acuity cc

OD 20/25      OS 20/30

## Color vision

OD 7/8      OS 8/8

## Confrontational fields

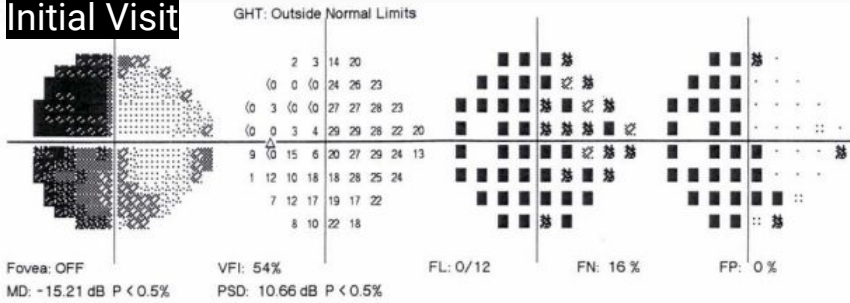
OD Full      OS Full

## ONH

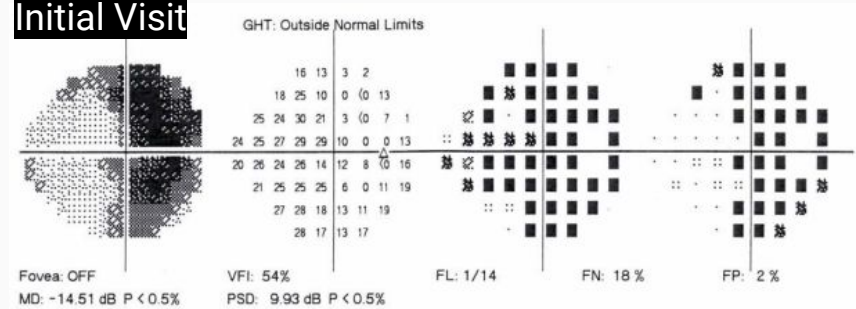
OU 1-2+ bitemporal pallor



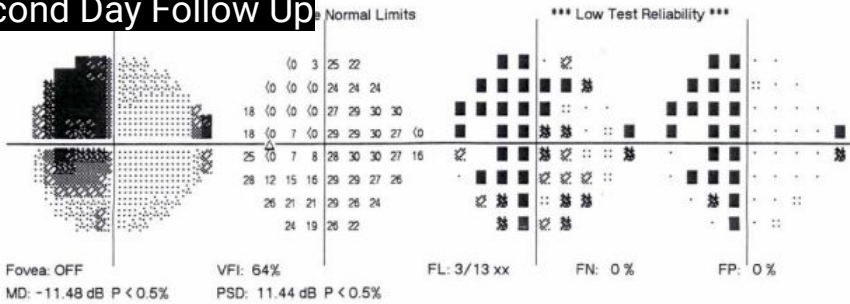
## Initial Visit



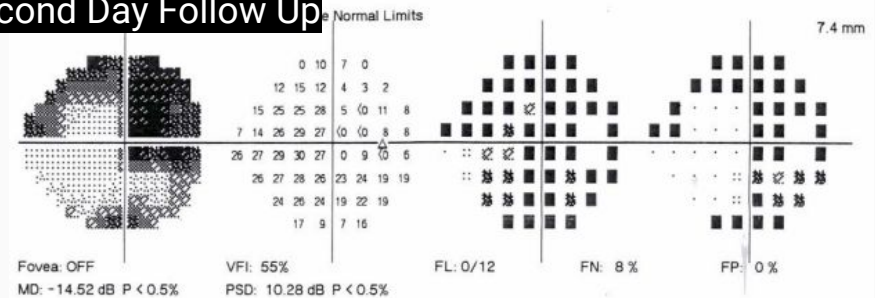
## Initial Visit



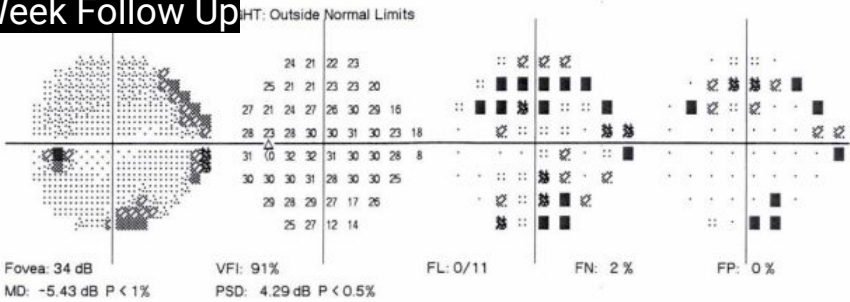
## Second Day Follow Up



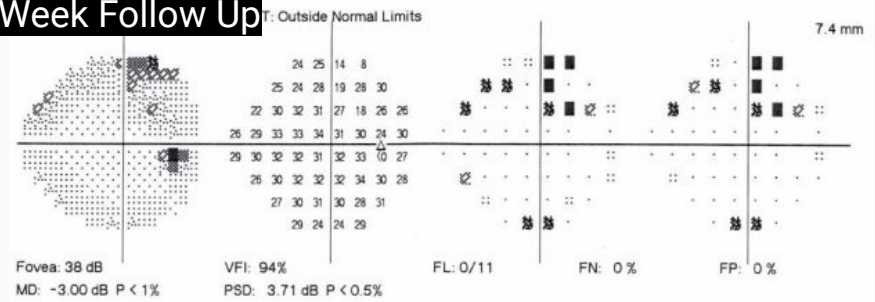
## Second Day Follow Up



## 4 Week Follow Up



## 4 Week Follow Up

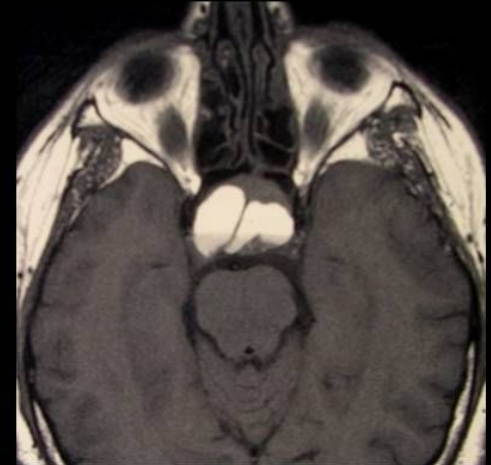


# Pituitary Apoplexy

- Apoplexy means bleeding into an organ or loss of blood flow to an organ. Pituitary apoplexy is commonly caused by bleeding inside a noncancerous (benign) tumor of the pituitary.

## Neuro-ophthalmic Emergency !

### Pituitary apoplexy





# What hormones does the pituitary gland make?

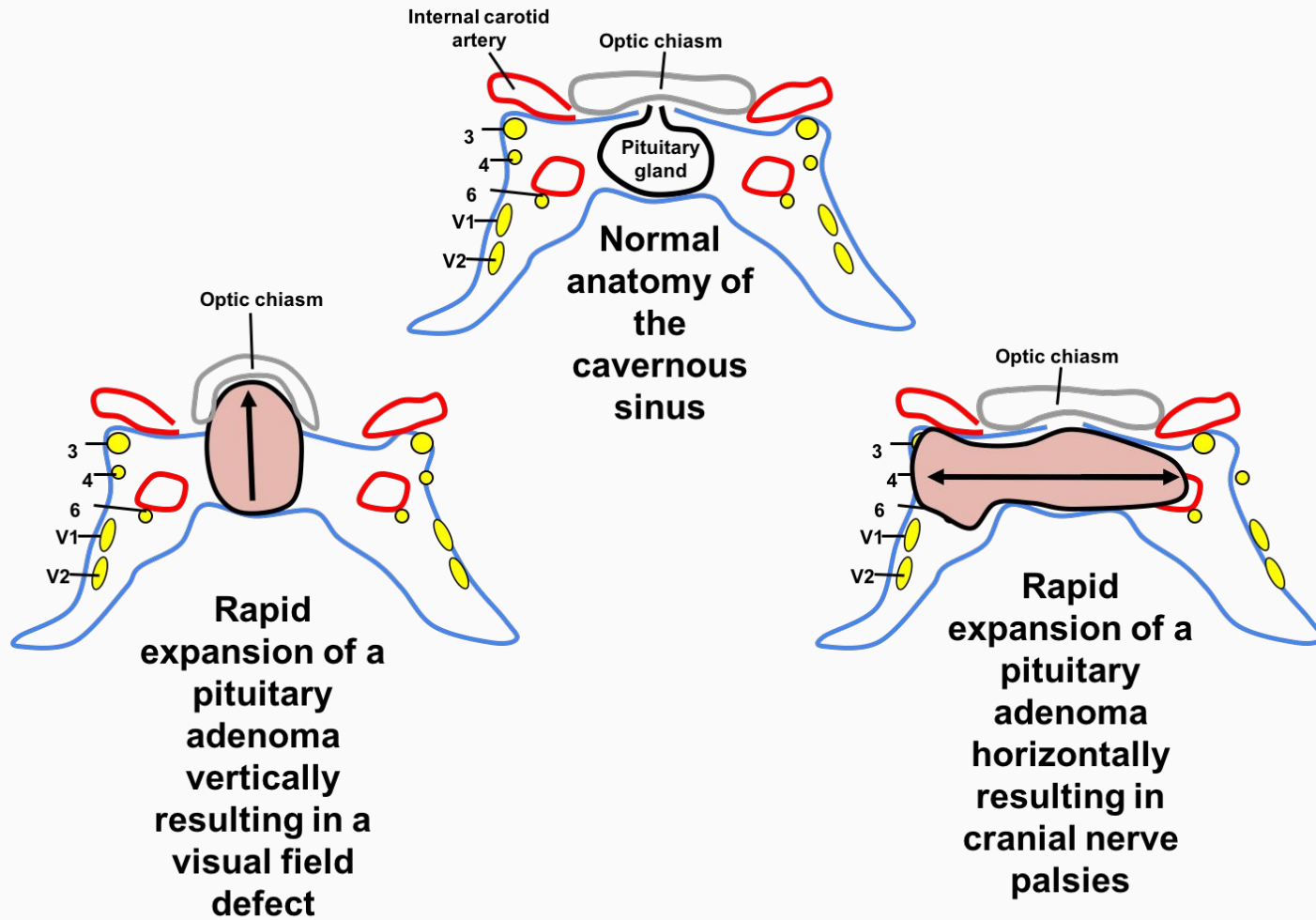
- Adrenocorticotrophic hormone (ACTH or corticotropin)
- Follicle-stimulating hormone (FSH)
- Growth hormone (GH)
- Luteinizing hormone (LH)
- Prolactin
- Thyroid-stimulating hormone (TSH)

# Pituitary Apoplexy

- Sudden onset of severe **headache** (>90%); **nausea/vomiting** (70%)
- **Bitemporal defect** (50-70%): bitemporal superior quadrantic defect
- **Diplopia**: cranial nerves III, IV, and VI palsies vulnerable to compression
- Altered mental status/consciousness (30%)
- Hypopituitarism: hormonal dysfunction; life-threatening hypotension, hypoglycemia, shock from low ACTH/cortisol
- May be hemorrhagic or nonhemorrhagic
- May involve a pre-existing pituitary adenoma or a nonadenomatous gland

# Associated Triggers

- Head trauma
- Hypertension
- Pregnancy/Postpartum - Sheehan syndrome
- Iatrogenic: major surgery, angiography, radiotherapy of head, dynamic pituitary function tests
- Medications: anticoagulants, estrogens, bromocriptine

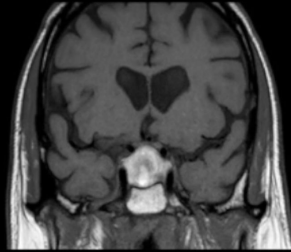




# MRI Signs of Pituitary Apoplexy

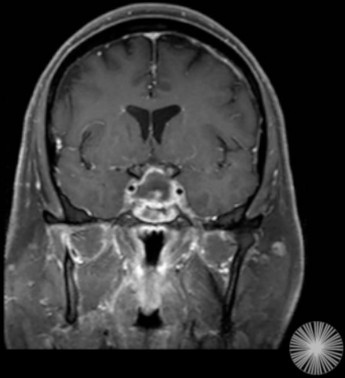
**Hemorrhagic**

Without contrast

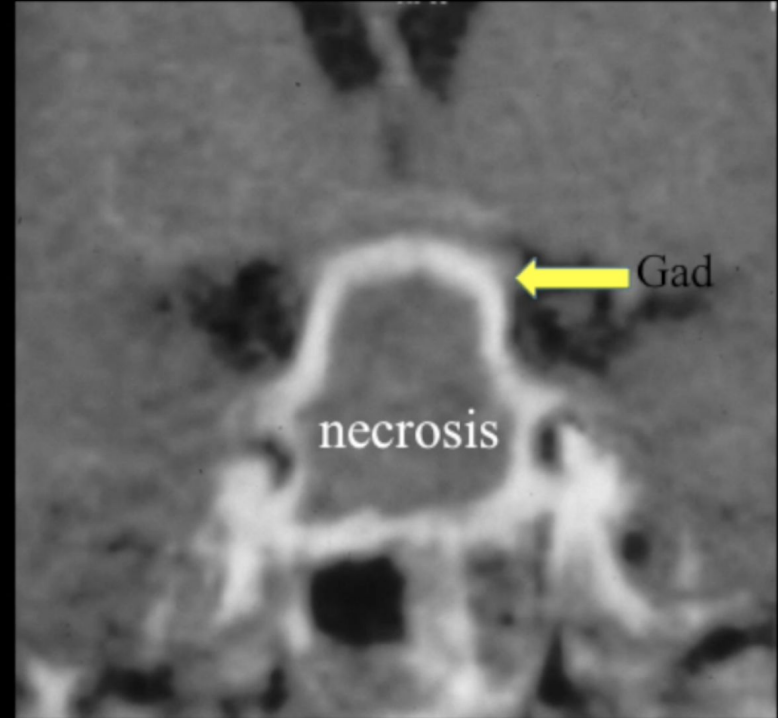


**Ischemic**

Contrast

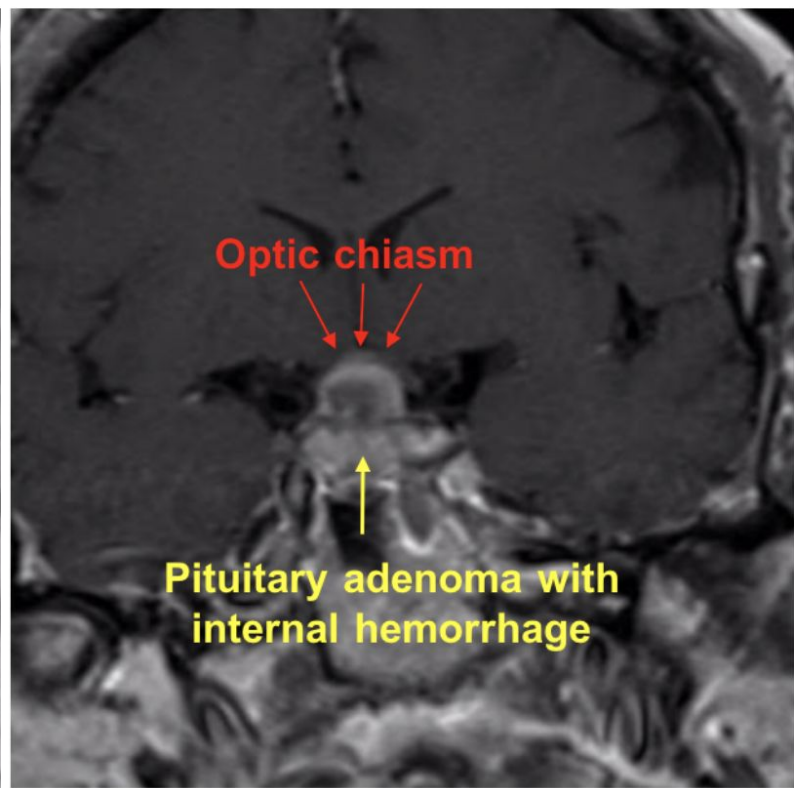
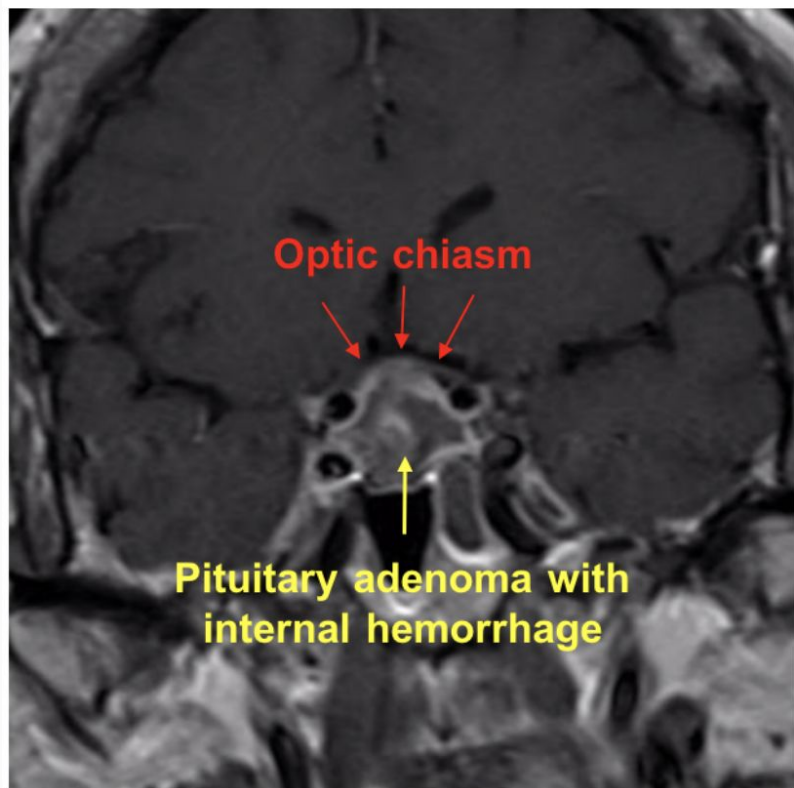


## Ischemic Apoplexy



## Pituitary Ring Sign

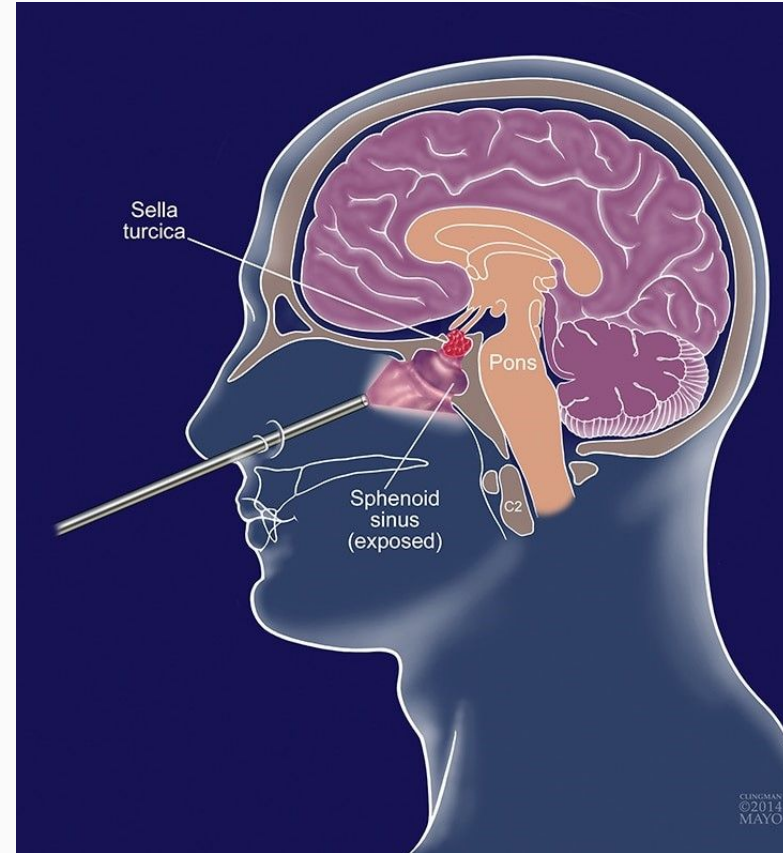
<https://www.aaopt.org/education/clinical-video/mri-signs-of-pituitary-apoplexy>



**Coronal T1 MRI of the brain with gadolinium showing a pituitary adenoma (yellow arrow) with internal hemorrhage and mass effect and superior displacement of the optic chiasm (red arrows).**

# Treatment

- Mild conditions
  - Closely monitor daily over weeks to months
  - Check visual acuity; color vision; EOMs
- Severe conditions
  - Surgery immediately (transsphenoidal)
  - IV Steroids/hormone therapy
  - Blood tests for endocrine function



# What happens if not treated emergently?

- Acute panhypopituitarism
- Adrenal crisis (acute cortisol insufficiency)
- Permanent visual acuity loss/blindness
- Permanent visual field loss
- Death



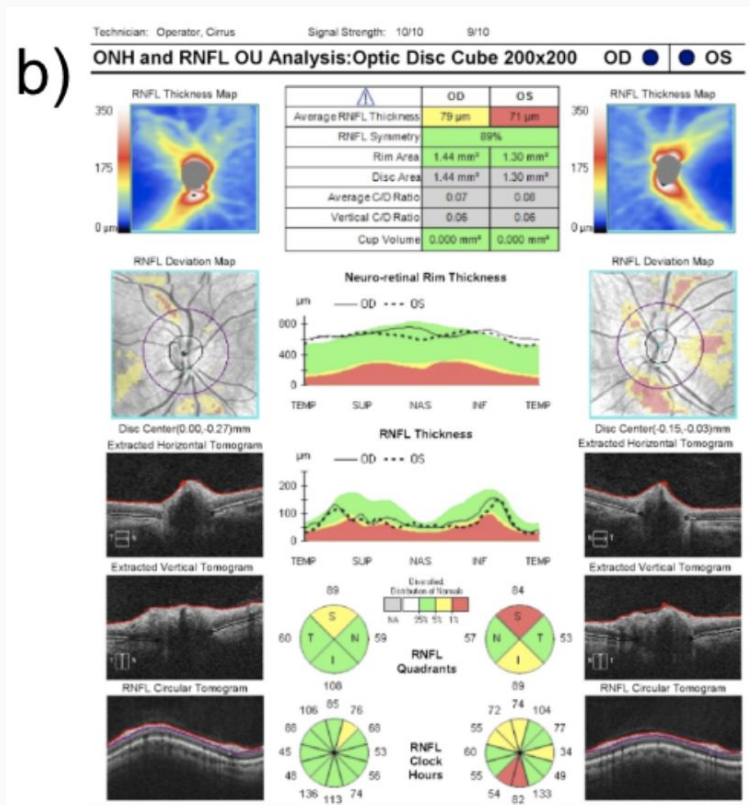
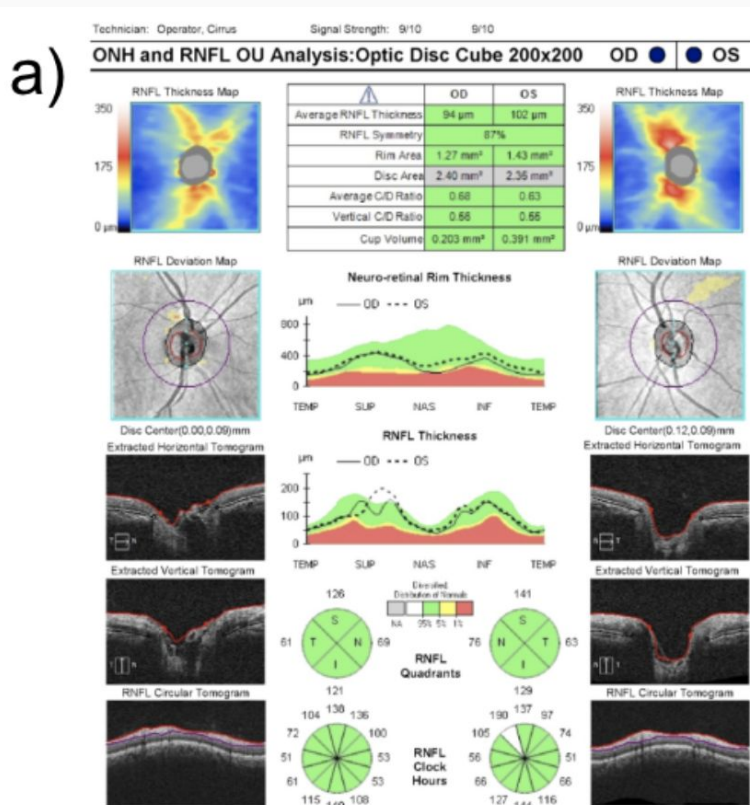


# Variables of visual recovery

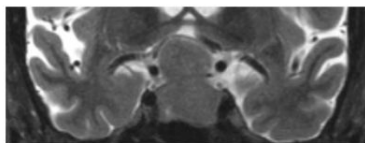
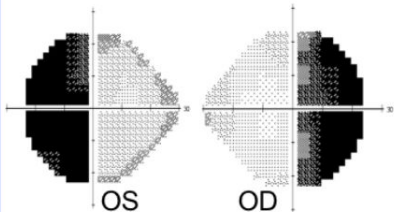
- Duration of symptoms
- Size of the tumor
- Preoperative visual acuity and visual field
- Optic nerve appearance
- OCT: thickness of the RNFL and ganglion cell complex

**Prognosis for vision recovery is usually good with timely intervention**

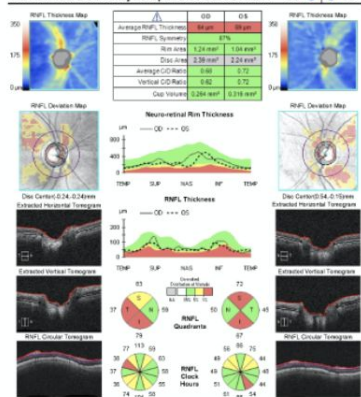
Which of the following OCTs of the retinal nerve fiber layer (RNFL) most likely represents a patient with a new bitemporal hemianopia from pituitary apoplexy?



# Patient A

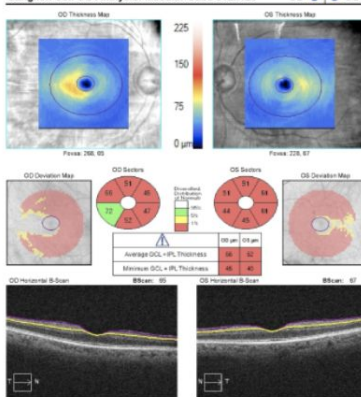


Technician: Operator, Cirrus Signal Strength: 7/10 8/10  
**ONH and RNFL OU Analysis: Optic Disc Cube 200x200** OD ● ● OS



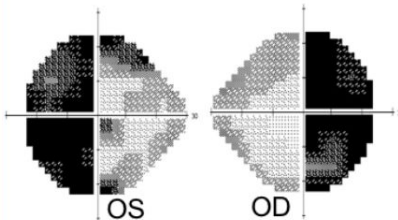
	OD	OS
Average RNFL Thickness	64 μm	59 μm
RNFL Symmetry	87%	
Rim Area	1.24 mm <sup>2</sup>	1.04 mm <sup>2</sup>
Disc Area	2.39 mm <sup>2</sup>	2.24 mm <sup>2</sup>
Average C/D Ratio	0.68	0.72
Vertical C/D Ratio	0.62	0.72
Cup Volume	0.264 mm <sup>3</sup>	0.316 mm <sup>3</sup>

Technician: Operator, Cirrus Signal Strength: 9/10 10/10  
**Ganglion Cell OU Analysis: Macular Cube 512x128** OD ● ● OS

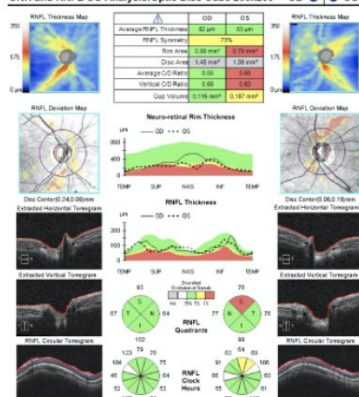


	OD μm	OS μm
Average GCL + IPL Thickness	56	52
Minimum GCL + IPL Thickness	45	40

# Patient B

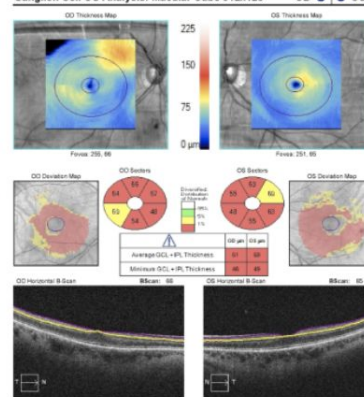


Technician: Operator, Cirrus Signal Strength: 9/10 9/10  
**ONH and RNFL OU Analysis: Optic Disc Cube 200x200** OD ● ● OS



	OD	OS
Average RNFL Thickness	82 μm	83 μm
RNFL Symmetry	73%	
Rim Area	0.99 mm <sup>2</sup>	0.79 mm <sup>2</sup>
Disc Area	1.46 mm <sup>2</sup>	1.39 mm <sup>2</sup>
Average C/D Ratio	0.55	0.65
Vertical C/D Ratio	0.55	0.63
Cup Volume	0.116 mm <sup>3</sup>	0.187 mm <sup>3</sup>

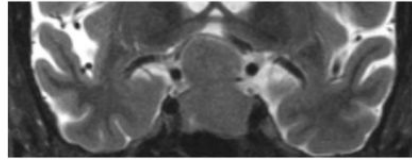
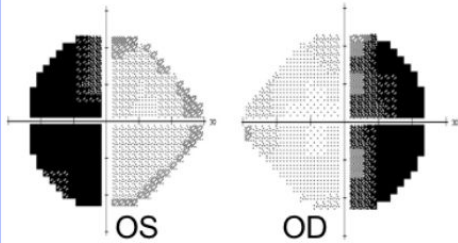
Technician: Operator, Cirrus Signal Strength: 9/10 9/10  
**Ganglion Cell OU Analysis: Macular Cube 512x128** OD ● ● OS



	OD μm	OS μm
Average GCL + IPL Thickness	61	59
Minimum GCL + IPL Thickness	46	49

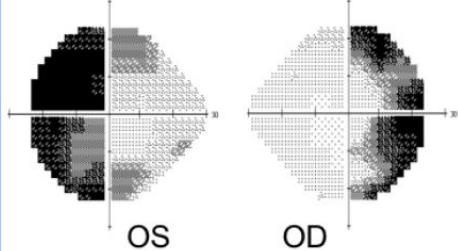
# Patient A

Pre-operative visual field



	OD	OS
Average RNFL Thickness	64 $\mu\text{m}$	59 $\mu\text{m}$
RNFL Symmetry	87%	
Rim Area	1.24 mm <sup>2</sup>	1.04 mm <sup>2</sup>
Disc Area	2.39 mm <sup>2</sup>	2.24 mm <sup>2</sup>
Average C/D Ratio	0.68	0.72
Vertical C/D Ratio	0.62	0.72
Cup Volume	0.264 mm <sup>3</sup>	0.316 mm <sup>3</sup>

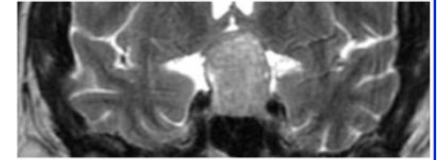
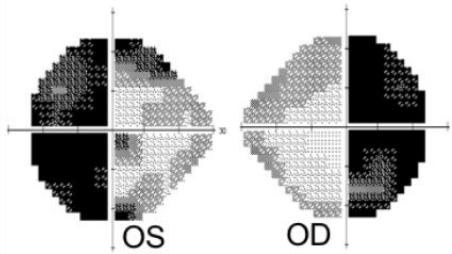
6 months after surgery



	OD $\mu\text{m}$	OS $\mu\text{m}$
Average GCL + IPL Thickness	56	52
Minimum GCL + IPL Thickness	45	40

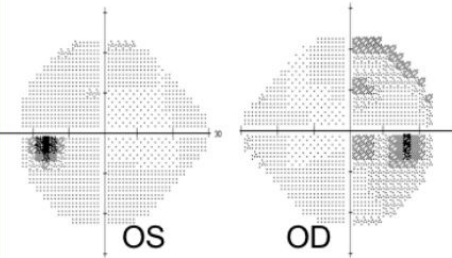
# Patient B

Pre-operative visual field



	OD	OS
Average RNFL Thickness	82 $\mu\text{m}$	83 $\mu\text{m}$
RNFL Symmetry	73%	
Rim Area	0.99 mm <sup>2</sup>	0.79 mm <sup>2</sup>
Disc Area	1.46 mm <sup>2</sup>	1.39 mm <sup>2</sup>
Average C/D Ratio	0.55	0.65
Vertical C/D Ratio	0.55	0.63
Cup Volume	0.115 mm <sup>3</sup>	0.187 mm <sup>3</sup>

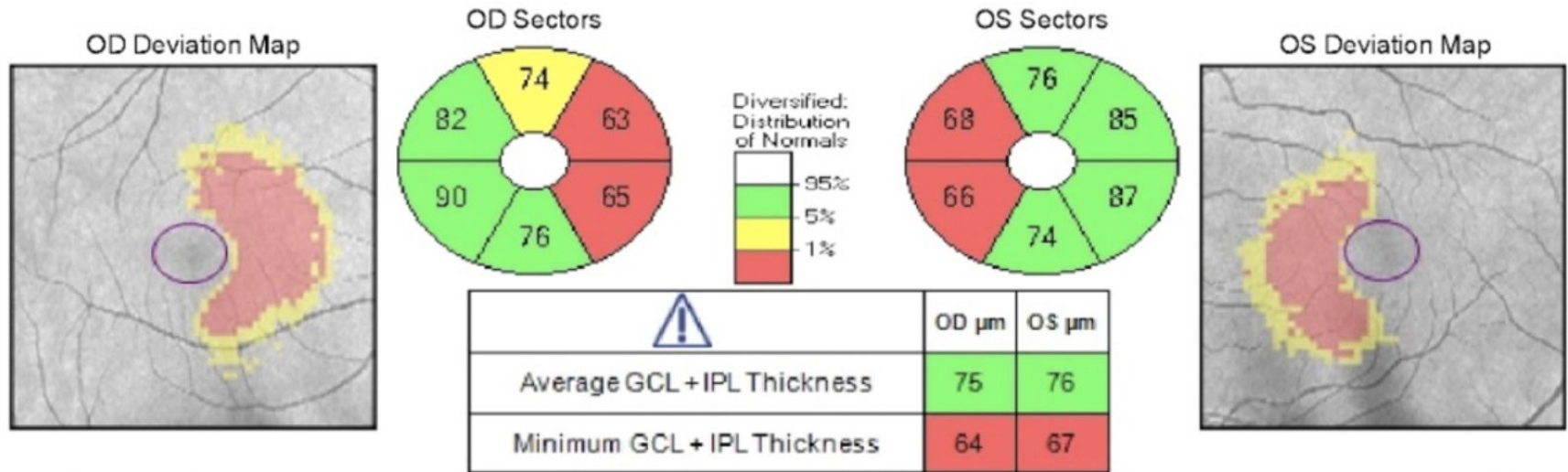
6 months after surgery



	OD $\mu\text{m}$	OS $\mu\text{m}$
Average GCL + IPL Thickness	61	59
Minimum GCL + IPL Thickness	46	49



# Macular ganglion cell complex often shows changes earlier than the RNFL



OCT of the macular ganglion cell complex shows binasal thinning in both eyes, indicating that the sellar mass is having an effect on the anterior visual pathways.

# Optometrist Role

- Recognize the risk of pre-existing macroadenoma
- Monitor vision, color vision and EOM status closely
- 24-2 HVF and OCT RNFL/GCC
- Referral to Neuro-OMD/Neurosurgeon/ER/Hospital

# Reference

1. <https://pressbooks.pub/casebasedneuroophthalmology/chapter/pituitary-apoplexy/>
2. <https://www.aao.org/education/current-insight/pituitary-apoplexy-syndrome-of-acute-visual-dysfun>
3. <https://www.sciencedirect.com/topics/neuroscience/bitemporal-hemianopsia#:~:text=Lesions%20of%20the%20chiasm%20lead,occipital%20cortex%20or%20optic%20tract.>
4. <https://www.aao.org/education/clinical-video/mri-signs-of-pituitary-apoplexy>
5. <https://my.clevelandclinic.org/health/diseases/23935-panhypopituitarism>