

EYE CATCHING CASES

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Financial Disclosures

☐ I do not have any financial interest in any of the parts of the lecture today.

Objectives

☐ Describe the purpose of ancillary testing such as visual field, corneal topography, fundus photography and OCT

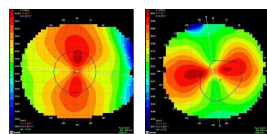
☐ Discuss normal vs abnormal findings in these tests

☐ Discuss how these tests apply to the cases presented

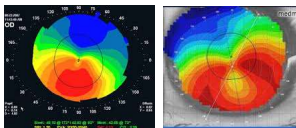
Corneal Topography

- Placido Disc
 - Rings projected onto cornea and then curvature is measured over 1000's of points
- Slit Scanning
 - Light based analysis based on elevation of the front and back surface of cornea
- Corneal ectasia (KCN, PMD)
 - Diagnosis
 - Progression
- Irregular Astigmatism
 - Scar
 - Pterygium
- Contact Lens fitting

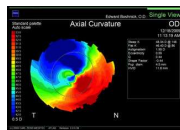




- Normal
 - Bow tie
 - Hour glass
 - Figure 8



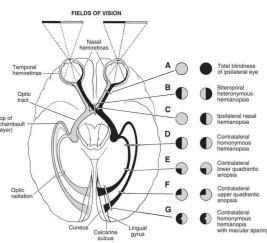
- Corneal Ectasia
 - Keratoconus
 - Pellucid Marginal Degeneration (PMD)



- Irregular Astigmatism

Visual Field

- Subjective test analyzing the function of the visual pathway
- Retina
 - Plaquenil toxicity
- Optic Nerve
 - Glaucoma
 - Optic Disc Edema
 - Papilledema
- Optic Pathway
 - Stroke
 - Tumor

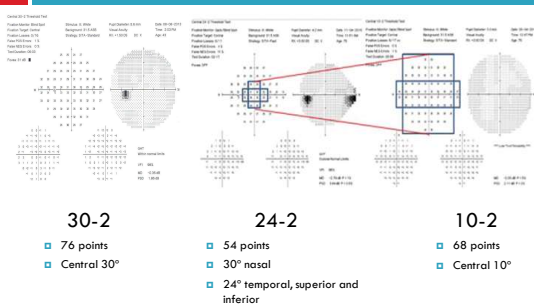


Visual Field



- 10-2
 - Central 10° of vision
 - Retinal toxicity
 - Advanced Glaucoma
 - AMD
- 24-2
 - 24° temporal, superior and inferior
 - 30° nasal
 - Glaucoma
- 30-2
 - Central 30°
 - Optic nerve
 - Neurological

Visual Field



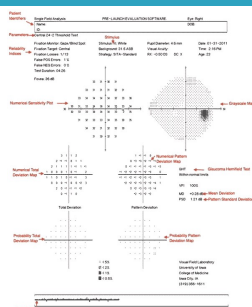
Visual Field

Reliability Measurements

- Fixation Monitoring
 - <20%
- False Positives
 - <15%
- False Negatives


Performance Measurements

- Mean Deviation
 - Difference from normal
- Pattern Deviation
 - Localized loss for specific patient
- Visual Field Index
 - Overall Performance
- Glaucoma Hemifield Test
 - Glaucoma analysis



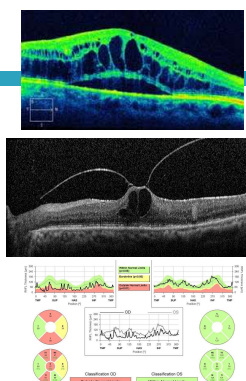
OCT

- Ocular Coherence Tomography
- Uses light waves to create cross-section images
 - ▣ Retina
 - ▣ Optic Disc
 - ▣ Anterior Segment



OCT

- Structural Assessment
 - ▣ Optic Nerve
 - Glaucoma
 - Disc edema
 - Atrophy
 - ▣ Retina
 - Diabetic Retinopathy
 - AMD
 - Vitreomacular disease
 - Dystrophies and Degenerations



Handout: Retinal Layers

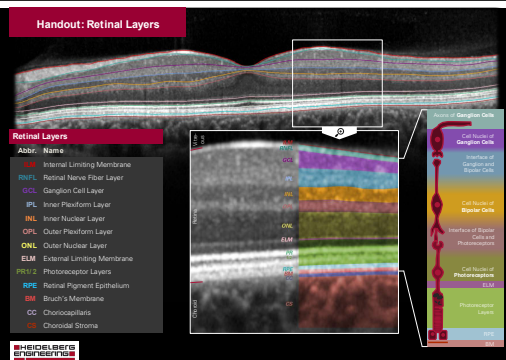
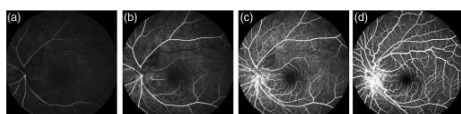


Abb.	Name
ILM	Internal Limiting Membrane
RNFL	Retinal Nerve Fiber Layer
GCL	Ganglion Cell Layer
IPL	Inner Plexiform Layer
INL	Inner Nuclear Layer
OPL	Outer Plexiform Layer
ONL	Outer Nuclear Layer
ELM	External Limiting Membrane
PRIS	Photoreceptor Layers
RPE	Retinal Pigment Epithelium
BM	Bruch's Membrane
CC	Choriocapillaris
CS	Chorioid Stroma

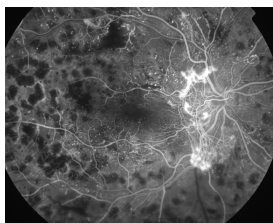
Fluorescein Angiography

- ▣ Fluorescein dye injected into arm
- ▣ Choroidal flush after 8-12 seconds
- ▣ Retinal circulation 11-18 seconds
 - Early arteriovenous
 - Late arteriovenous phase
- ▣ Peaks around 30 seconds then recirculation



Fluorescein Angiography

- ▣ Hypofluorescence
 - ▣ Blocking defect
 - ▣ Filling defect
- ▣ Hyperfluorescence
 - ▣ Leakage
 - ▣ Staining



A COMPLICATED
CORNEA...AND MORE

COMPLICATED CORNEA

HISTORY

- 33 year old Hispanic Female
- CC: Here for cataract evaluation
 - LEE: 6 months ago – no mention of cataract
 - Progressively decreasing vision over last year
 - Light sensitivity and glare with sunlight and night driving
 - Itching and irritation after spraying insecticide in eyes 10 days ago
 - Went to ER – eyes were irrigated extensively, no drops prescribed.
 - Improving symptoms with AT's and cool compresses
- Past Medical Hx
 - High cholesterol
- Past Ocular Hx
 - None
- Family Hx
 - No blindness, glaucoma or AMD

COMPLICATED CORNEA

EXAM

- VA
 - OD: 20/100
 - OS: 20/80
- Current Glasses
 - OD: -6.50 -2.50x043
 - OS: -3.50 -4.75x097
- Keratometry
 - OD: 47.25 / 50.50 @ 146
 - OS: 48.50 / 55.00 @ 30
- Manifest Refraction
 - OD: -7.75 -3.50x065 20/70
 - OS: -5.50 -3.75x089 20/80

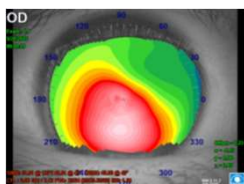
COMPLICATED CORNEA

EXAM

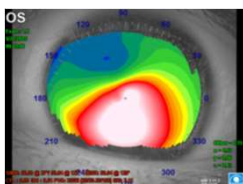
- | | |
|--------------------------------|---------------------------|
| □ Adnexa | □ Optic Disc |
| □ WNL OD/OS | □ C/D: 0.30 OD/OS |
| □ Conjunctiva | □ No edema or pallor |
| □ 1+ papillae | |
| □ Trace injection | □ Macula |
| □ Cornea | □ Normal OD/OS |
| □ Munson's Sign OD/OS | □ Vessels |
| □ No striae, no scarring OD/OS | □ Normal OD/OS |
| □ A/C | □ Periphery |
| □ Deep quiet OD/OS | □ No holes/tears RD OD/OS |
| □ Lens | |
| □ Clear OD/OS | |

COMPLICATED CORNEA

TOPOGRAPHY AND SCLERAL



Scleral Contact Lens:
BC: 49.00
Dia: 15.6
Power: -10.25
VA: 20/25



Scleral Contact Lens:
BC: 50.00
Dia: 15.6
Power: -13.00
VA: 20/25

COMPLICATED CORNEA

SCLERAL FITTING #2

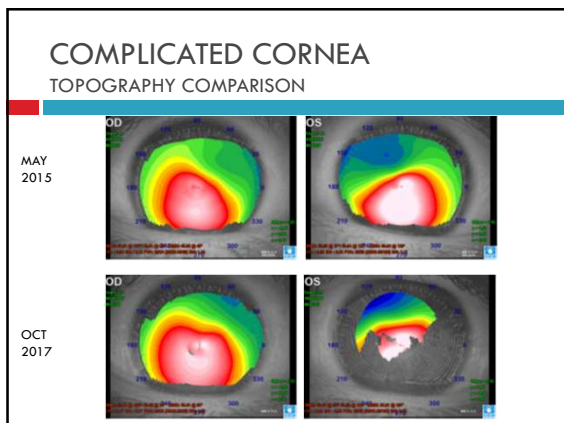


- OCT 2017 (2 ½ Years later)
- Blurry vision, film over contacts
 - ▣ Filling scleral lens with multipurpose solution
- VA (w/scleral)
 - ▣ 20/300
 - ▣ 20/70

COMPLICATED CORNEA

SCLERAL FITTING #2

- | | |
|--------------------------------|----------------------|
| □ Adnexa | □ Optic Disc |
| ▣ WNL OD/OS | ▣ C/D: 0.30 OD/OS |
| □ Conjunctiva | ▣ No edema or pallor |
| ▣ 1+ papillae | □ Macula |
| □ Cornea | ▣ Normal OD/OS |
| ▣ Munson's Sign OD/OS | □ Vessels |
| ▣ No striae, no scarring OD/OS | ▣ Normal OD/OS |
| □ A/C | |
| ▣ Deep quiet OD/OS | |
| □ Lens | |
| ▣ Clear OD/OS | |



COMPLICATED CORNEA

SCLERAL FITTING #2

2015	2017
OD:	OD:
BC: 49.00	BC: 52.00
Dia: 15.6	Dia: 15.6
Power: -10.25	Power: -13.50
VA: 20/25	VA 20/20
OS:	OS:
BC: 50.00	BC: 54.00
Dia: 15.6	Dia: 15.6
Power: -13.00	Power: -16.50
VA: 20/25	VA: 20/20

COMPLICATED CORNEA

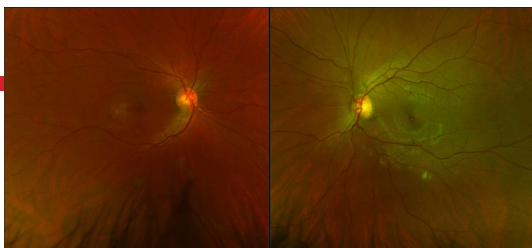
SCLERAL DISPENSE #2

	□ Scleral Dispense (5 months later)
	□ 20/40 OD
	■ PH 20/25
	■ Plano Over-refraction
	■ Good fit and comfort
	□ 20/20 OS
	■ Good fit and comfort
	□ F/U 1 month

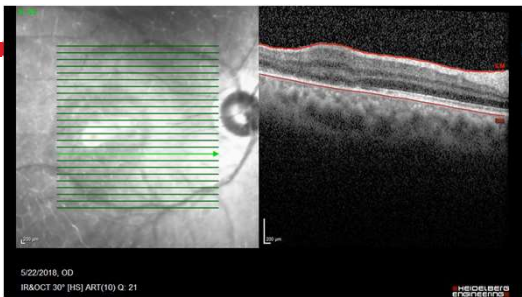
COMPLICATED CORNEA

...AND MORE

- Scleral F/U –2 months
 - 20/80 OD
 - distortion to left side of fixation
 - OR: Plano
 - Fitting: good clearance, limbal and edge fit
 - 20/20 OS



- OD: flat, round, grey subretinal lesion under macula
- OS: WNL



5/22/2018, OD
IRAOCT 30° (HS) ART(10) Q. 21
HEIDELBERG ENGINEERING

COMPLICATED CORNEA

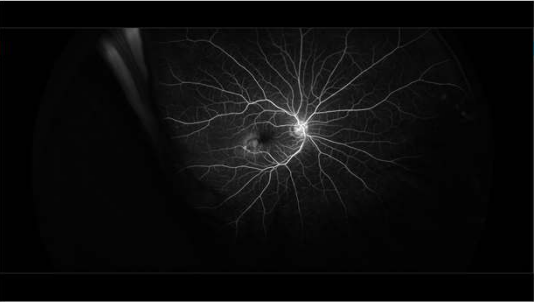
...AND MORE

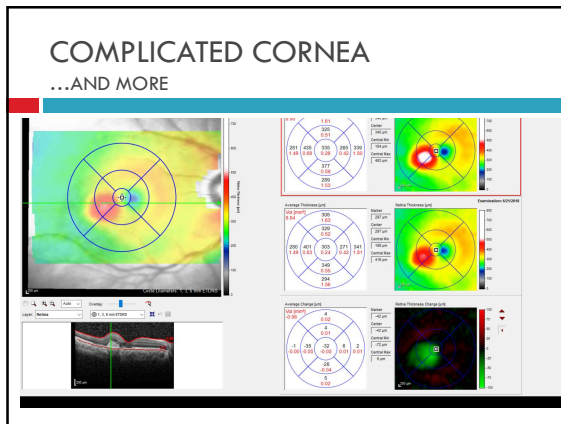
- Differential Diagnosis
 - ▣ Myopic CNVM
 - ▣ AMD CNVM
 - ▣ Ocular Histoplasmosis
 - ▣ Multifocal Choroiditis
 - ▣ Idiopathic CNVM

COMPLICATED CORNEA

...AND MORE

- Suspected Myopic CNVM
 - ▣ Referred to retina clinic
 - ▣ IVA
 - ▣ F/U 1 month
- Retina F/U (1 Month)
 - ▣ 20/60 with scleral lens
 - ▣ Improved vision and distortion
 - ▣ IVA #2
 - ▣ F/U 1 month

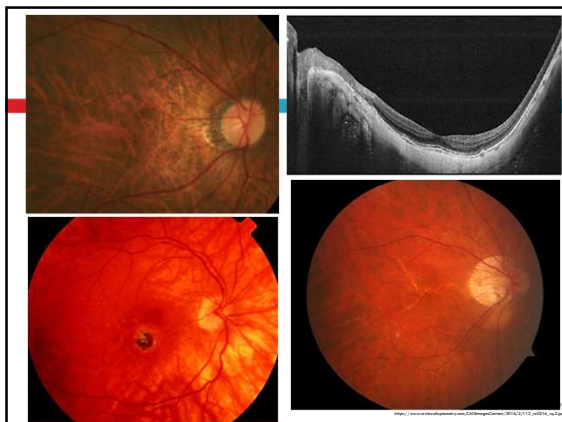




MYOPIC CNVM

RISK FACTORS

- ☐ Pathological myopia
 - ☐ Spherical equivalent $> -6D$
 - ☐ Axial length $> 26.5mm$
 - ☐ Tessellated Fundus
 - ☐ Posterior Staphyloma
 - ☐ Fuch's Spot
 - ☐ RPE Atrophy (Patchy $>$ Diffuse)
 - ☐ Lacquer Cracks



MYOPIC CNVM

PATHOLOGY

- Lacquer cracks induce intracellular changes in RPE that release VEGF
- Mechanical stretching of RPE cells up-regulates pro-angiogenic factors, including VEGF
- New vessels grow thru breaks in Bruch's membrane (Lacquer cracks) or result from an atrophic area within the RPE

MYOPIC CNVM

EXAM

- Highly myopic patient with sudden decrease in VA (20/40 to 20/100) or onset of metamorphopsia
- Small (<1DD) flat, greyish subretinal membrane



MYOPIC CNVM

TREATMENT

- Anti-VEGF
 - ▢ Lucentis = Eylea = Avastin
- Average improvement in BCVA was 13.8 letters after 12 months

MYOPIC CNVM

PEARLS

- ~30% will develop mCNVM in the other eye
- Early detection and treatment is key
- Don't forget about pathological myopia and CNVM in high myopes
 - ▣ Educate on risk factors
 - ▣ Amsler Grid

BLESSED BY BACKUP

BACKUP


HISTORY

- 67 year old, white male
- CC: cloudy circle in vision OD
 - ▣ Referred by endocrinologist for cloudy vision
 - ▣ Woke up 10 days ago and OD had a cloudy circle in vision; "looks like a donut" central vision is good
 - ▣ No new neurological symptoms
- Medical Hx
 - ▣ Type 2 Diabetes
 - ▣ HTN
 - ▣ HLD

BACKUP

EXAM

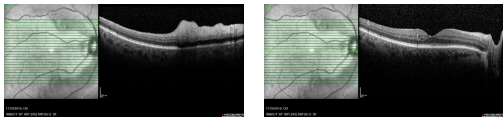
OD:	OS:
<ul style="list-style-type: none"> VA: 20/25 Pupils: PERRL, no APD EOM and Alignment: <ul style="list-style-type: none"> Smooth and full Ortho Confrontations: FTCF SLE: 1+ NS DFE: 	<ul style="list-style-type: none"> VA: 20/25 Pupils: PERRL, no APD EOM and Alignment: <ul style="list-style-type: none"> Smooth and full Ortho Confrontations: FTCF SLE: 1+ NS DFE:



<ul style="list-style-type: none"> Optic Disc: <ul style="list-style-type: none"> 0.30 No edema or Pallor Macula: appears flat Vessels: CWS along temporal arteries; sluggish perfusion Periphery: WNL 	<ul style="list-style-type: none"> Optic Disc: <ul style="list-style-type: none"> 0.30 No edema or pallor Macula: WNL, flat Vessels: <ul style="list-style-type: none"> Few scattered DBH Periphery: WNL
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BACKUP

OCT

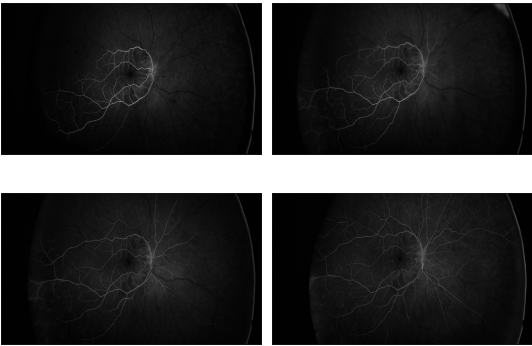


OD: Inner retinal thickening with trace CME
OS: flat

BACKUP

DDX

- DDx:
 - BRAO
 - CRAO
 - Ocular Ischemic Syndrome
- Assessment and Plan:
 - Central retinal artery occlusion
 - With patent cilioretinal artery
 - Next day referral to retina clinic
 - Patient education about diagnosis and systemic complications including risk of stroke



FA REPORT OD: delayed arterial filling with hyperfluorescent changes in veins

BACKUP

WORKUP

- Systemic Workup (within 1 week)
 - Holter
 - Echocardiogram
 - Carotid Ultrasound
 - ESR/CRP
- 80% blockage of right carotid artery
 - Now s/p angioplasty
- Maintains 20/25 VA OD

CRAO

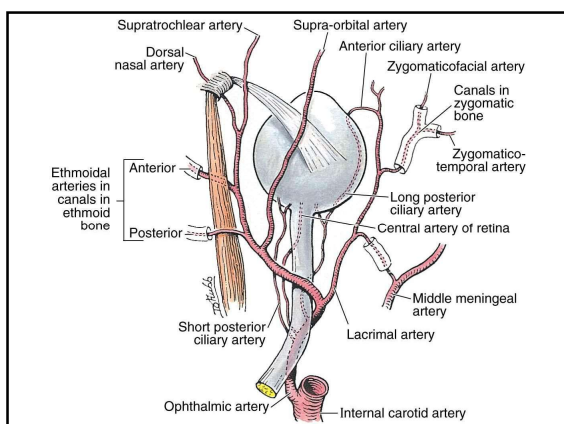
RISK FACTORS

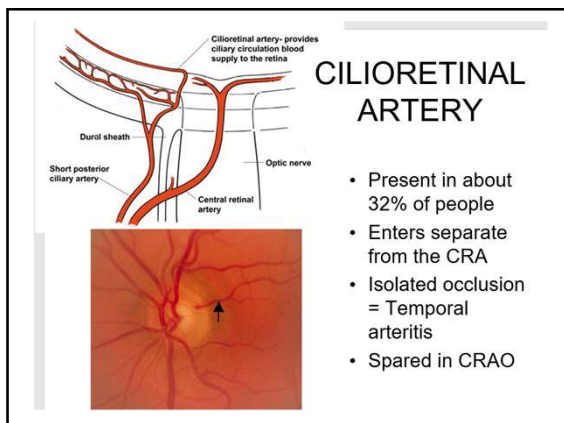
- Age
- Male > Female
- Smoking
- HTN
- Obesity
- Diabetes
- Hyperlipidemia
- Cardiovascular disease
- Coagulopathy

CRAO

PATHOLOGY

- Obstruction of the retinal vessel by embolus, thrombus, trauma, inflammation or spasm





CRAO

EXAM

- ☐ Vision loss
 - ☐ Sudden
 - ☐ Painless
 - ☐ Worse than 20/400 (75%)
- ☐ APD
- ☐ Cherry Red Spot
 - ☐ Develops within a few hours
 - ☐ Resolves in 4-6 weeks

CRAO

TREATMENT

- ☐ First 3-4 hours
 - ☐ Lower IOP
 - ☐ Topical drops
 - ☐ A/C Paracentesis
 - ☐ Ocular Massage
 - ☐ Contact gonio lens
- ☐ Emergency Room?

CRAO

PEARLS

- Ocular findings are a warning sign for an ischemic stroke
 - ▢ High correlation with ischemic finding on brain MRI
- Systemic workup is key
- Visual prognosis is poor

ARE YOU SER(I)OUS!

ARE YOU SER(I)OUS!

HISTORY

- 49 year old, white male
- CC: spot in OS that blocks central vision
 - ▢ Onset 5-6 weeks ago
 - ▢ Seems to be intermittent; present more when he pays attention to it
 - ▢ Looks like a dim disc floating around

ARE YOU SER(I)OUS!

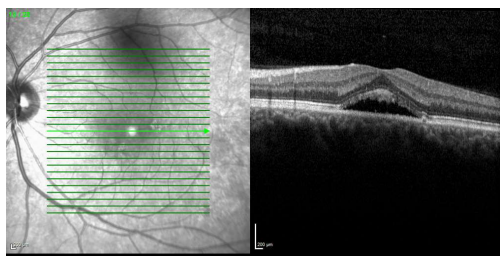
EXAM

OD:

- ☐ VA: 20/20 cc
- ☐ Pupils: PERRL, no APD
- ☐ EOM: full
- ☐ Confrontations: full
- ☐ SLE: unremarkable
- ☐ DFE:
 - ☐ C/D: 0.20
 - ☐ Macula: WNL
 - ☐ Vessels: WNL
 - ☐ Periphery: WNL

OS

- ☐ 20/20 cc
- ☐ Pupils: PERRL, no APD
- ☐ EOM: full
- ☐ Confrontations: full
- ☐ SLE: unremarkable
- ☐ DFE:
 - ☐ C/D: 0.20
 - ☐ Macula: round serous elevation
 - ☐ Vessels: WNL
 - ☐ Periphery: WNL



OCT:

- ☐ Neurosensory Detachment OS
- ☐ Small PED temporal to fovea

ARE YOU SER(I)OUS!

DIAGNOSIS

DDx:

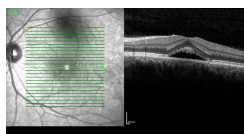
- ☐ Central Serous Chorioretinopathy
- ☐ CNVM

Diagnosis: Central Serous Chorioretinopathy

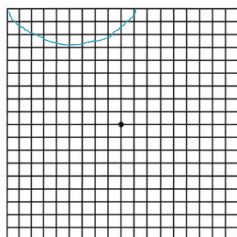
- ☐ Amsler grid provided
- ☐ F/U 1 month

ARE YOU SER(I)OUS!

6 WEEKS

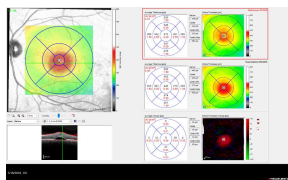


CC: Stable, no changes
 VA: 20/20 OS
 Macula: unchanged
 OCT: unchanged
 F/U: 1 Month



ARE YOU SER(I)OUS!

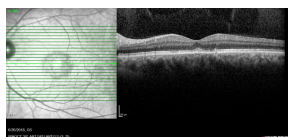
4 MONTHS



- CC: Left eye seems to have improved
- VA: OS 20/25
- SLE: unremarkable
- DFE: stable serous elevation OS
- OCT: slightly worsened NSD
- F/U 1 month in retina clinic

ARE YOU SER(I)OUS!

5 MONTHS



- CC: Distortions dissipating OS
- VA: 20/20- OS
- DFE:
 - Flat
 - RPE changes with few drusen like opacities temporal macula
- OCT:
 - Small PED, no NSD
- F/U
 - 6 Months

CSCR

□ RISK FACTORS

- ▣ Stress
- ▣ Corticosteroids
- ▣ H. Pylori
- ▣ Autoimmune diseases
- ▣ Sleep Disturbances
- ▣ Hypertension

□ PATHOLOGY

- ▣ Increased vascular permeability of choroid

CSCR

EXAM

- Blurry vision
- Visual distortion
- Dark area in central vision
- VA: 20/20 – 20/200
- Hyperopic shift (+1.00)
- Round serous elevation of macula
- OCT
 - ▣ Neurosensory detachment
 - ▣ Pigment epithelial detachment

CSCR

EXAM

- Treatment
 - ▣ Self Limiting (weeks to months)
 - ▣ Stop steroids
 - ▣ Anti-corticosteroid therapy
 - ▣ Anti-VEGF
 - ▣ Laser photocoagulation
 - ▣ PDT

CSCR

PEARLS

- Treatment = Observation
- Associated PED is not uncommon
- Referral and FA for un-resolving or atypical presentation

NUMB AND NUMBER

NUMB AND NUMBER

HISTORY

- | | |
|--|---|
| <ul style="list-style-type: none"> □ 50 year old, white female □ CC: DM eye exam <ul style="list-style-type: none"> ■ history of abrasion OD ■ Long history of things to "fix her cornea" ■ Denies pain, redness, irritation □ Ocular Hx: <ul style="list-style-type: none"> ■ Diabetic retinopathy 2017 ■ PCIOL ~ 2015 ■ Lateral tarsorrhaphy OD <ul style="list-style-type: none"> ■ Poorly healing abrasion OD ■ Presumed herpetic in nature ■ No pain thru this episode | <ul style="list-style-type: none"> □ Medical Hx: <ul style="list-style-type: none"> ■ DM, HTN, HLD, Kidney disease □ Ocular Medications: <ul style="list-style-type: none"> ■ Prednisolone acetate QD OD ■ Restasis BID OU ■ Valacyclovir 1 g QD ■ Refresh TID-QID □ Systemic medications <ul style="list-style-type: none"> ■ Atorvastatin ■ Levothyroxine ■ Gabapentin ■ NovoLog ■ Warfarin |
|--|---|

NUMB AND NUMBER

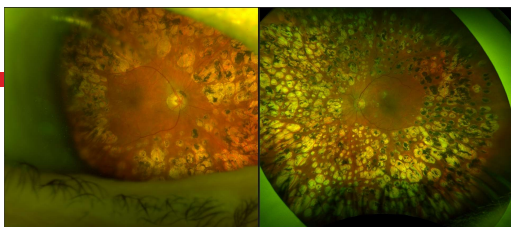
EXAM

OD:

- ☐ EOM: Full
- ☐ Confrontations: constricted
- ☐ Adnexa: lateral tarsorrhaphy
- ☐ Conjunctiva: WNL
- ☐ Cornea: 2-3+ SPK, worst centrally; faint stromal scar?
- ☐ A/C: deep and quiet
- ☐ Lens: PCIOL in good position

OS:

- ☐ EOM: Full
- ☐ Confrontations: Full
- ☐ Adnexa: WNL
- ☐ Conjunctiva: WNL
- ☐ Cornea: 1-2+ inferior SPK
- ☐ A/C: deep and quiet
- ☐ Lens: PCIOL

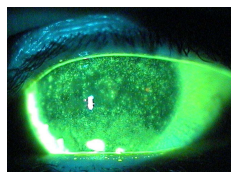


- | | |
|--|--|
| <input type="checkbox"/> Vitreous: VH inferiorly | <input type="checkbox"/> Vitreous: no heme |
| <input type="checkbox"/> C/D: 0.40 | <input type="checkbox"/> C/D: 0.40 |
| <input type="checkbox"/> Macula: flat, no fluid | <input type="checkbox"/> Macula: flat, no fluid |
| <input type="checkbox"/> Vessels: arteriolar narrowing | <input type="checkbox"/> Vessels: arteriolar narrowing |
| <input type="checkbox"/> Periphery: s/p full PRP | <input type="checkbox"/> Periphery: s/p full PRP |

NUMB AND NUMBER

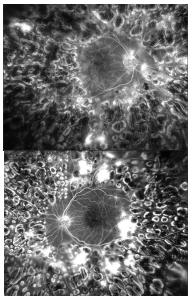
DDX

- ☐ DDx for cornea
 - ☐ Dry eye
 - ☐ Neurotrophic keratitis
 - ☐ Topical drug toxicity
 - ☐ Chemical injury
 - ☐ Limbal stem cell deficiency



NUMB AND NUMBER


PLAN



- Neurotrophic keratitis
 - CPM
 - Prednisolone acetate QD OD
 - Restasis BID OU
 - Valacyclovir 1g QD
 - NPAT's Q1-2 hr OU
 - F/U 1 month
 - PDR
 - Active VH OD
 - Inactive OS
 - Retina follow up 2 weeks

NEUROTROPHIC KERATITIS


RISK FACTORS



- HZV, HSV
- Topical Anesthetic
- Diabetes
- Ocular Surgery
 - LASIK/PRK
 - Retinal laser
- Any damage to V
 - Surgical
 - Traumatic

NEUROTROPHIC KERATITIS

PATHOLOGY



- Degenerative disease of cornea resulting from impaired corneal innervation
 - Lack of neuromodulation leads to degeneration

NEUROTROPHIC KERATITIS

STAGES

- Stage 1
 - Irregular epithelium; SPK
 - Corneal edema
 - Corneal neovascularization
 - Corneal scarring
- Stage 2
 - Recurrent or persistent epithelial defects
- Stage 3
 - Stromal involvement
 - Ulcer
 - Melting
 - Perforation

NEUROTROPHIC KERATITIS

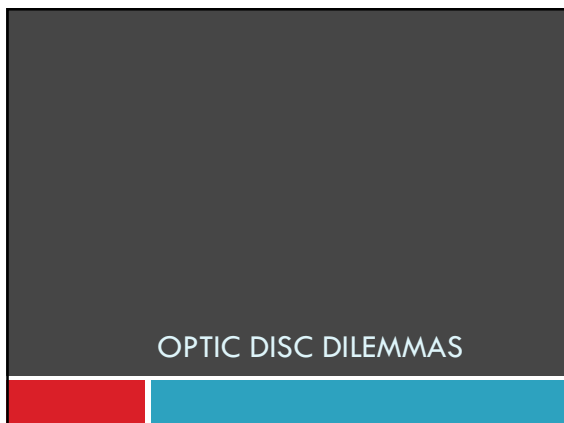
TREATMENT

- Stage 1
 - Artificial tears/ointment
 - Autologous serum tears
 - Bandage contact lens
- Stage 2
 - Bandage contact lens
 - Amniotic membrane graft
 - Antibiotic drops
 - Steroid?
 - Tarsorrhaphy
- Stage 3
 - Ulcer treatment and healing
 - Prevention of perforation
 - Oxervate
 - Nerve growth factor
 - Surgical

NEUROTROPHIC KERATITIS

PEARLS

- Can be easily confused with moderate/severe dry eye
- Consider NK in patients with a history of HSV or HZV
- Educate patient on potential vision threatening complications and importance of follow up



OPTIC DISC DILEMMAS

Case #1

- 42 year old, white male
- CC: blurry distance vision; headaches with extended periods of computer work and reading
- Ocular Hx: unremarkable
- Medical Hx: unremarkable
- Medications: None

OPTIC DISC DILEMMAS

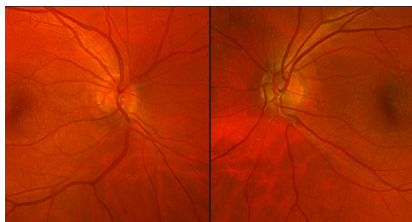
Case #1

- MR: +0.25-0.50x135
- 20/20
- SLE: unremarkable
- DFE:
 - <0.10
 - Elevation 360
 - Sharp disc margins
 - No pallor

- MR: +0.75-1.00x060
- 20/20
- SLE: unremarkable
- DFE:
 - <0.10
 - Elevation 360
 - Sharp disc margins
 - No pallor

OPTIC DISC DILEMMAS

Case #1



- | | |
|---|---|
| <ul style="list-style-type: none"> □ OD □ <0.10 □ Elevation 360 □ Sharp disc margins □ No pallor | <ul style="list-style-type: none"> □ OS □ <0.10 □ Elevation 360 □ Sharp disc margins □ No pallor |
|---|---|

OPTIC DISC DILEMMAS

Case #2



- 11 year old, white female
- CC: referred for exam due to result of convergence insufficiency survey at school
- Headaches when she reads
 - 2-3x per week
 - Forehead and brow area
 - Relieved with ibuprofen
 - Do not disrupt daily activities
- Medical Hx: Unremarkable
- Medications: None

OPTIC DISC DILEMMAS

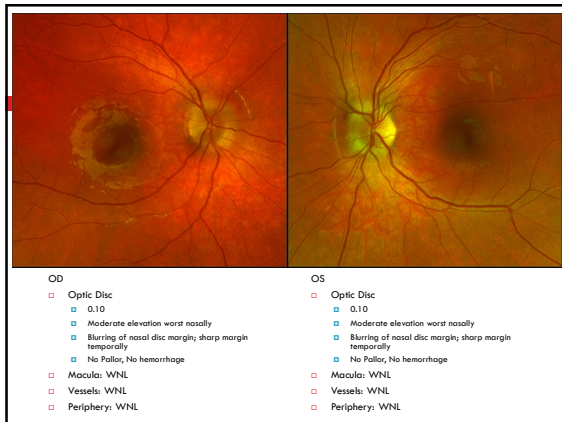
Case #2

OD:

- VA: 20/20 sc
- Pupils: PERRL, no APD
- EOM: smooth and full
- Binocular Vision
 - CT: 4 XP
 - NPC: nose
 - Amps: 13
 - BO: 20/25/20
- Cyclo MRx: Plano -0.75x010
- SLE: WNL
- DFE:

OS:

- VA: 20/20 sc
- Pupils: PERRL, no APD
- EOM: smooth and full
- Binocular Vision
 - CT: 4 XP
 - NPC: nose
 - Amps: 14
 - BO: 20/25/20
- Cyclo MRx: Plano -0.25x175
- SLE: WNL
- DFE:



OPTIC DISC DILEMMAS

Case #3

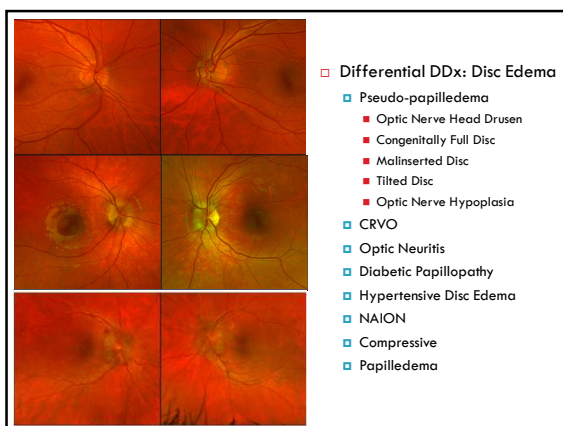
- 36 year old white female
- CC: vision will turn all white sometimes
 - Patient passed out and fell onto face 6/1/18 (3 months ago)
 - Headache starting at the base of the neck and extending thru right temple and behind right eye
 - Exacerbated by stress and activity
 - Variable pain from 4-8
 - Present daily
 - Improved with use of Excedrin
 - Episodes of vision turning white, this will last a few seconds and then return to normal
 - Occurs multiple time per day
- Medical Hx
 - Unremarkable
- Radiology
 - C/T done at ER was normal per patient; no MRI performed
- Medications:
 - Tramadol
 - Metaxalone
- Vital Signs
 - BP: 148/85
 - BMI: 35.2 (Obesity)

OPTIC DISC DILEMMAS

Case #3

OD:	OS:
VA: 20/40cc	VA: 20/40
MRx: -1.00 -1.00x095	MRx: -0.75-1.00x093
20/25	20/25
EOM and Alignment:	EOM and Alignment:
Full, Ortho	Full, Ortho
Confrontations: Full	Confrontations: Full
SLE: WNL	SLE: WNL
DFE:	DFE:






OPTIC DISC DILEMMAS

IMPORTANT TERMINOLOGY

- Optic disc edema \neq Papilledema
 - Papilledema is optic disc edema due to elevated intracranial pressure
- Papilledema \neq Pseudotumor Cerebri or Idiopathic Intracranial Hypertension

DDx by Dr. Freedman

Causes	Disc Edema
<p>High ICP - Papilledema</p> <p>Ischemia - AION</p> <p>Inflammation / Optic Neuritis</p> <ul style="list-style-type: none"> - Classic Demyelinating Optic Neuritis associated with or without MS - Post Viral, Meningitis, Vasculitis, Neuroretinitis, Autoimmune, Infectious (e.g. Toxoplasmosis) <p>Infiltrative - e.g. Leukemia, Sarcoidosis, Lymphoma, Germopathy</p> <p>Compression - Tumors (e.g. orbital or sellar / parasellar), Large Mucocele (e.g. Graves) or Vessel (e.g. carotid a.)</p> <p>Hereditary: e.g. LHON</p> <p>Toxic - e.g. Methanol, Ethylene Glycol, Chemotherapy, Ethambutol, Anti-TNFα</p> <p>Other - orbital venous obstruction, POEMS[®] syndrome</p> <p>Ocular / False Localizing Sign[®]: e.g. not directly optic nerve disease, but coexisting eye problem</p> <ul style="list-style-type: none"> Venous stasis (CRVO, BRVO), Hypotony Posterior Scleritis, Uveitis Acute Multifocal Placoid Pigment Epitheliopathy (AMPPE) Multiple Evanescent White Dot Syndrome (MEWDS) 	
<p>OR</p> <p>MAYBE NOT EDEMA, BUT SOMETHING THAT LOOKS LIKE IT</p> <p>e.g. Anomalous Congenital Disc Elevation or</p> <p>Abnormal Disc Vessels or Growths on Disc (see next 2 lists)</p>	

OPTIC DISC DILEMMAS

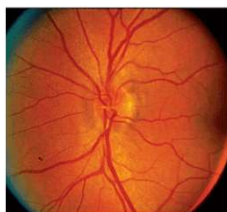
Pseudopapilledema

- ☐ Optic Nerve Head Drusen
- ☐ Congenitally Full Disc
- ☐ Malinserted Disc
- ☐ Tilted Disc
- ☐ Optic Nerve Hypoplasia

Papilledema

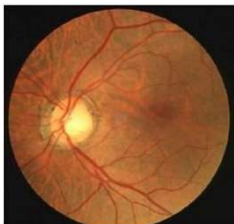
- ☐ Intracranial Tumor
- ☐ Malignant HTN
- ☐ Venous sinus thrombosis
- ☐ Chiari malformation
- ☐ AV Malformation
- ☐ Meningitis/Encephalitis
- ☐ Idiopathic Intracranial Hypertension

Crowded Disc



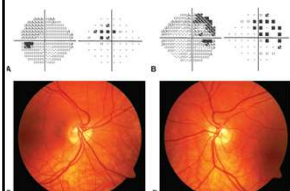
- ☐ Asymptomatic
- ☐ Smaller than average disc
 - ☐ 1.2 – 2.5 mm
 - ☐ Average 1.88mm
 - ☐ Correction Factor
 - ☐ 78D x 1.11
 - ☐ 90D x 1.33
- ☐ Hyperopic eyes
- ☐ Slightly hyperemic

Malinserted Disc



- Oblique insertion of optic nerve to the globe
 - ▢ Tilted along vertical axis
- Nasal elevation, temporal depression
 - ▢ Scleral crescent
- Myopes

Tilted Disc Syndrome



Triad of findings:

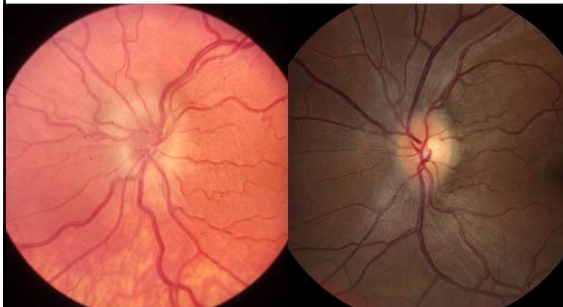
- ▢ Tilted Disc
 - Vertical axis itself is rotated downward
 - Superior elevation, inferior depression
- ▢ Decreased acuity
- ▢ Bi-temporal visual field defects

Optic Nerve Head Drusen



- Buried or Visible
- Elevation of disc
 - ▢ With or without blurred margins
- Anomalous vasculature
 - ▢ Early branching at disc
 - ▢ Tortuosity
- Move to surface as patient ages
 - ▢ Scalloped borders
- Usually Asymptomatic
 - ▢ Can have VF defects

The Real Diagnostic Dilemma...



OPTIC DISC DILEMMA

EXAM

- ☐ Comprehensive History
 - ☐ Headache
 - ☐ Transient visual obscurations
 - ☐ Tinnitus, Vertigo
 - ☐ Diplopia
- ☐ Medical Hx
 - ☐ Medications
 - ☐ Systemic risk factors

DDx by Dr. Freedman

Papilledema and Causes of Increased Intracranial Pressure



1. Hypertension – Malignant*
2. Intracranial Tumor (by mere mass effect or by causing obstructive hydrocephalus), Carcinomatous Meningitis
3. Medications – Vitamin A, Accutane, Tetracyclines, Contraceptives, Corticosteroid withdrawal, Thyroid Replacement, Growth Hormone Supplement
4. Toxic: Ethylene Glycol, Lead (Pb), Hyper-ammonemia
5. CNS Inflammation: Meningitis** Encephalitis (e.g. Lyme, HIV, post-Varicella, Malaria, Abscess) other inflammatory - e.g. CNS Vasculitis, Lupus, HeNOL Syndrome***
6. Trauma, Hematomas, Sub - Arachnoid hemorrhage
7. Vascular abnormalities: AVM, AV fistula (e.g. dural AV fistulae after longstanding venous sinus thrombosis)
8. Obstruction to Venous Drainage – Jugular Vein (e.g. paraganglioma-glossus tumor), Vena Cava, Venous Sinus Thrombosis – hypercoagulable states, middle ear or mastoid infections, Erythropoietin abuse
9. Hydrocephalus, Significant Chiari Malformation or Dandy Walker Syndrome, Craniosynostosis
10. Endocrine: Addison's, Hypoparathyroidism, Weight Gain
11. Other: Sleep Apnea, Anemia, Thyroid dysfunction, elevated protein levels
12. Idiopathic: Pseudotumor Cerebri or Idiopathic Intracranial Hypertension (IIH)****
 - seen most frequently in young women of child bearing age – including with onset of puberty
 - first need to rule out above conditions as well as possible, and if you make the diagnosis of IIH, then you need to stay open to other causes (like Thrombosis) if pt does respond well to treatment


OPTIC DISC DILEMMA

EXAM

- Acuity
 - Refractive error
- EOM and Cover test
 - VI palsy most common with PE (38%)
 - Esotropia
 - Abduction deficit

OPTIC DISC DILEMMA

EXAM

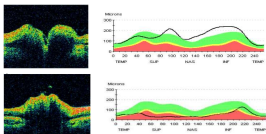
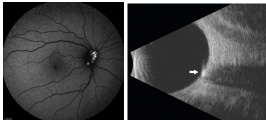


Evaluation of the Optic Disc

- Size
- Cup
- Margins
- Vasculature
- SVP

OPTIC DISC DILEMMA

ANCILLARY TESTING

Visual Field

- Papilledema
 - Enlarged blind spot
 - Peripheral Constriction
 - Can also be present in ONHD

OCT

- Papilledema: thickened RNFL
- ONHD: thinning of RNFL
 - Can also show nodular shadows (drusen)

FAF

- ONHD can show auto-fluorescence

B-Scan

- ONHD show as highly reflective

FA

- Papilledema: ONH will show hyperfluorescence and peripapillary leakage

DDx by Dr. Freedman

Differentiating between Congenital and Acquired Disc Elevation		
Feature	Congenital	Acquired
Nerve Fiber Layer	Clear	Opacified
Large Disc Vessels	Anomalous	Normal
Small Disc Vessels	Normal	Telangiectatic
NFL Hemorrhage	Rare	Frequent
Physiologic Cup	Small or absent	Normal (But may be obscured by edema)
Drusen	Sometimes present	Absent



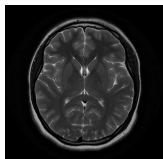
OPTIC DISC DILEMMA

HISTORY AND EXAM

Key Features	ONHD	Papilledema
Visual Symptoms	TVL, VFD	TVL, VFD, Diplopia
Headaches	None	If Present are worse upon awakening and/or postural changes
Neurological Symptoms	None	Tinnitus, vertigo, nausea/vomiting, peripheral neuralgias
Optic Nerve Appearance	Elevation confined to disc, SVP	Elevated swollen nerve, hyperemia, peripapillary vessel obscuration, +/- FSH, +/- CWS, +/- Paton's lines, SVP absent
Vasculature	Anomalous branching pattern	Microvascular dilation

OPTIC DISC DILEMMA


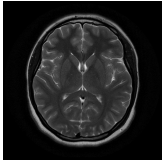
WORKUP



- Brain MRI with and without contrast
 - Urgent/Emergent
 - Consider symptoms and exam
- Lumbar Puncture
 - After imaging
 - Opening pressure
 - CSF Study
- MRV
 - Venous sinus thrombosis

OPTIC DISC DILEMMA

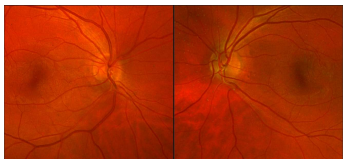
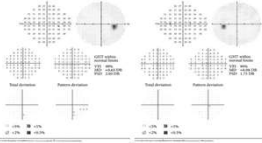
WORKUP

- Follow Up
 - Weeks to month(s)
 - Monitor Symptoms
 - Fundus Photos
 - Visual Field
- Treatment – IIH
 - Stop any causative medications
 - Weight Loss
 - Acetazolamide or Lasix

OPTIC DISC DILEMMAS

CASE #1

Optic Disc

- Size: Normal
- Cup: Crowded
- Margins: Sharp
- Vasculature
 - No obscuration
- SVP: Absent

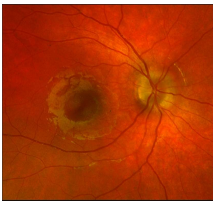
■ HVF: Full

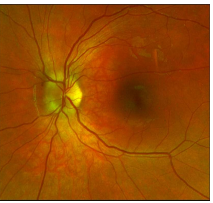
■ Plan:

- F/U 3-4 months
- Optic Disc Eval

OPTIC DISC DILEMMAS

CASE #2



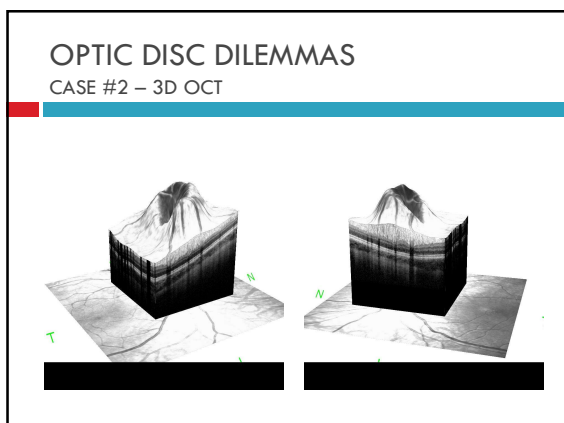


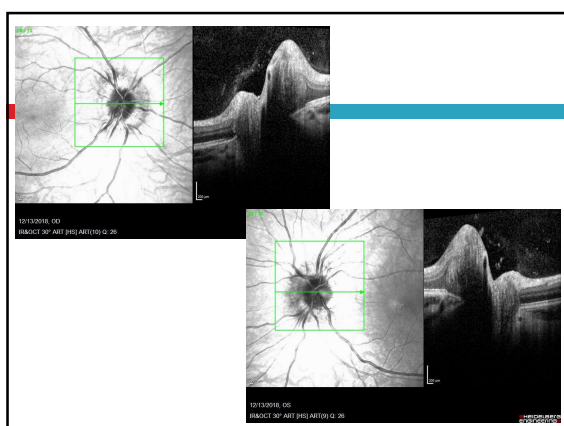
OD

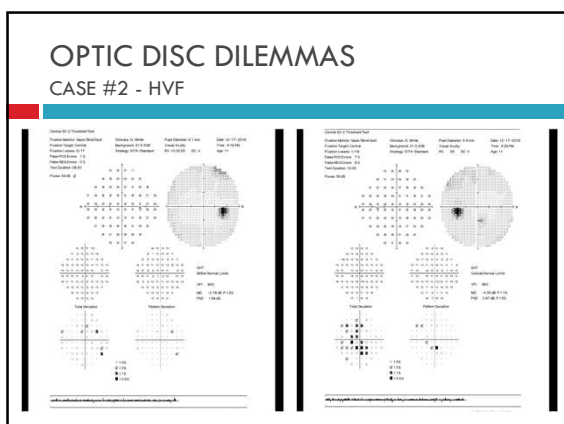
- Optic Disc
- 0.10
- Moderate elevation worst nasally
- Blurring of nasal disc margin; sharp margin temporally
- No Pallor, No hemorrhage

OS

- Optic Disc
- 0.10
- Moderate elevation worst nasally
- Blurring of nasal disc margin; sharp margin temporally
- No Pallor, No hemorrhage





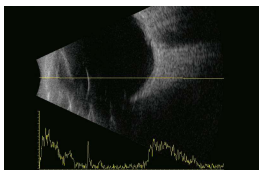
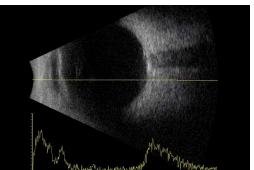


OPTIC DISC DILEMMAS

CASE #2 – B-Scan



OD

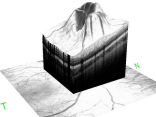

OS

OPTIC DISC DILEMMAS

CASE #2

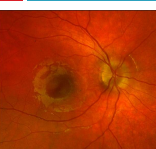




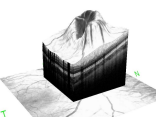
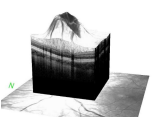



- Optic Disc
 - Size: Normal
 - Cup: Crowded
 - Margins: Blurred nasally
 - Vasculature
 - No obscuration
 - Early bifurcations OS
 - SVP: Absent
- HVF: Full
- OCT: Suspected ONHD
- B-Scan: Suspected ONHD

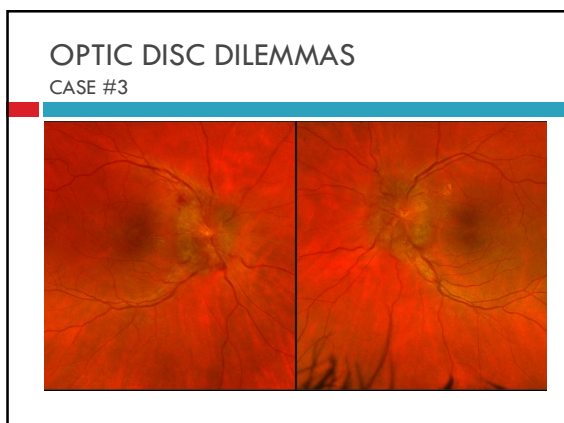
OPTIC DISC DILEMMAS

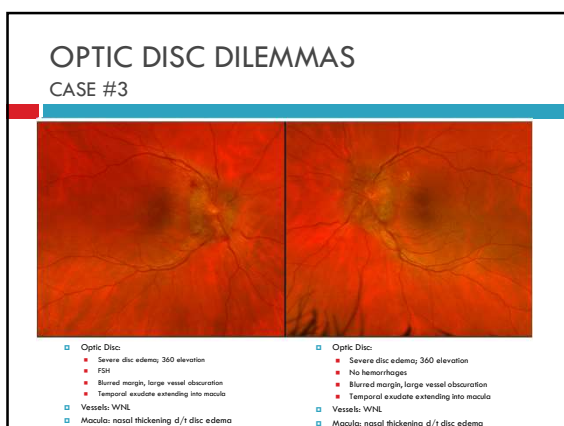
CASE #2

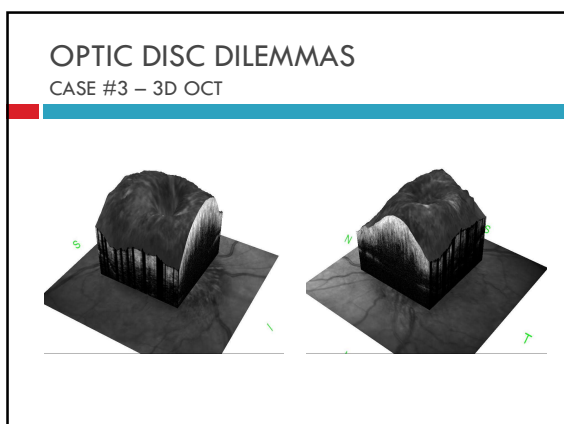



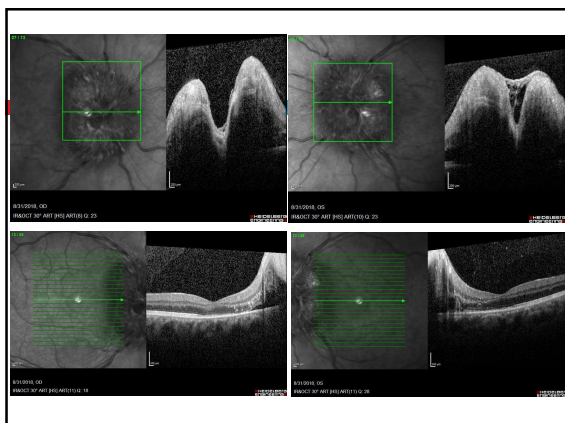



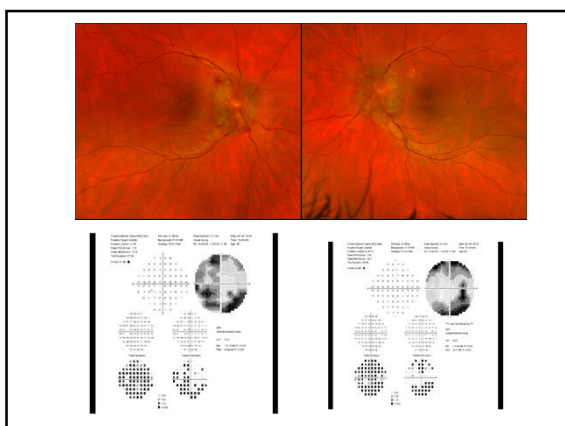
- Diagnosis
 - Suspected Optic Nerve Head Drusen OU
- Plan:
 - F/U 2 months
 - Optic Disc Eval
 - HVF





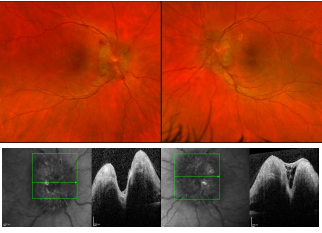






OPTIC DISC DILEMMAS

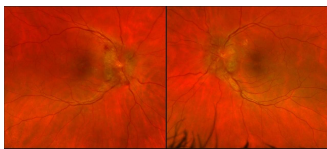
CASE #3



- Optic Disc
 - Size: ?
 - Cup: obscured
 - Margins: blurred 360
- Vessels:
 - large vessel obscuration
 - FSH
 - SVP: Absent
- HVF:
 - Enlarged blind spot
 - Constriction
- OCT
 - Severe 360 elevation

OPTIC DISC DILEMMAS

CASE #3

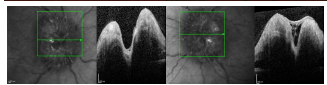


□ Diagnosis:

- ▣ Bilateral optic disc edema
- Suspicious for papilledema


□ Plan

- ▣ MRI head within 1 week
- ▣ F/U 1 week



OPTIC DISC DILEMMAS

CASE #3 – FOLLOW UP


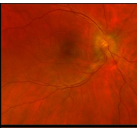
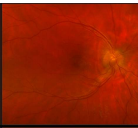




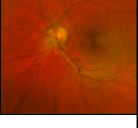


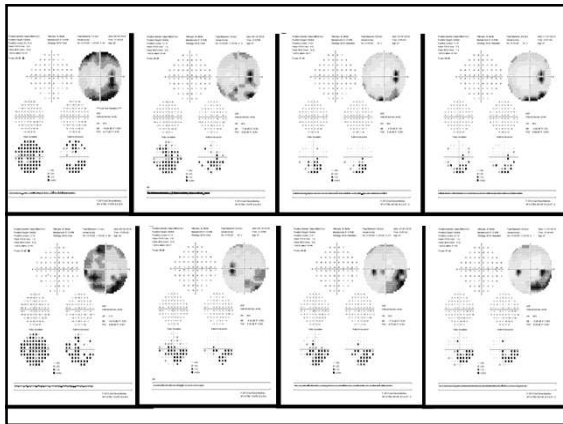
□ 1 Week

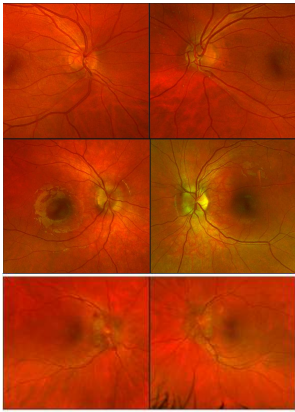
- ▣ MRI normal
- ▣ Vision stable
- ▣ Symptoms stable
- ▣ LP within 1 week

□ 2 week

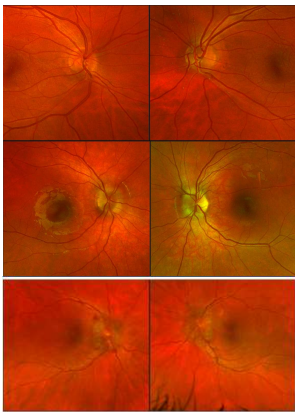
- ▣ LP - opening pressure 25
- ▣ CSF study normal
- ▣ Start acetazolamide 500mg BID
- ▣ Follow up 1 month

Initial	1 month	2 month	3 month
			
			





- Case 1
 - Congenital disc anomaly
 - F/U 3 months
- Case 2
 - Suspected buried optic nerve head drusen
 - F/U 2 months
- Case 3
 - Papilledema
 - Pseudotumor cerebri
 - Normal MRI
 - High Opening Pressure
 - Resolving on acetazolamide



PEARLS

- DON'T PANIC!
 - Comprehensive History
 - Optic Disc Evaluation
 - HVF
 - OCT
- Consider all exam findings
 - Workup, Refer, Follow

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QUESTIONS?

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