Top 5 Most Common Ocular Manifestations from Systemic Conditions

Mark Schaeffer, OD FAAO

Intrepid

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  Our Main Stage sessions feature free, promotional content for all attendees.
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- Exhibit Hall Hours Conferee Cafe – Exhibit Hall – Booth P19087

  Thursday, Sept 18 9:30am – 6:00pm
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  Saturday, Sept 20 9:30am – 3:00pm Conferee Happy Hour Thur, Sept 18 4:30 - 5:30pm

vision expo

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Financial Disclosures – Mark Schaeffer, OD FAAO

I Have Received Honoraria From:

- AesculaTech - Consultant
- Alcon – Consultant, Speaker
- Allergan – Consultant, Speaker
- Bausch + Lomb – Consultant, Speaker
- CooperVision – Consultant
- Harrow - Consultant, Speaker
- Johnson & Johnson Vision Care – Consultant
- LENZ Therapeutics - Consultant
- Optase – Consultant
- Science Based Health - Consultant
- Sight Sciences – Consultant
- Tarsus - Consultant
- Zeiss - Consultant
  - Founder, Dr. MES Consulting
  - Founding Member, Intrepid Eye Society

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How do you manage systemic disease patients?

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Case #1

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Comprehensive Eye Exam

39 year old female

- Comprehensive Eye Exam, updating glasses and contact lenses
  - During case history, lenses have been bothering her off and on
  - Getting more dryness as the end of the day
  - Has been to multiple ODs, has been in daily disposables
  - Here today for updated Rx
- Medical history unremarkable
- SPEED score- 8

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Eye Exam and findings

	OD	OS
Refraction	Plano-2.25x015 20/15	-0.50-1.00x176 20/15
Slit lamp exam	Decreased tear break up time (+) NaFL staining inferior cornea	Decreased tear break up time (+) NaFL staining inferior cornea
Cup-to-disc ratio	0.3/0.3	0.3/0.3
Fundus evaluation	(-) H, T, D 360 OU	(-) H, T, D 360

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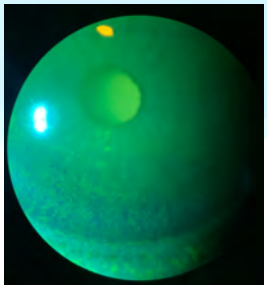
Let's look at the cornea…



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Plan of Action

-Sidebar discussion: Do you discontinue contact lenses?

- Refit into premium Silicone Hydrogel daily disposable contact lens
- Started on Lifitegrast bid OU
- RTC 2-3 months

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Patient returns to clinic

12 months later

- Patient felt better on medication
- Contact lenses felt better at the end of the day
  - Patient commented, "This is what usually happens for a little bit"
- Visual acuity 20/15 OD, OS, OU
- Patient felt better after 2-3 months, discontinued the treatment and is now feeling worse

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Do we want to ask some more questions?

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Systemic questions

- Do you suffer from dry mouth?
- Any other areas of dryness?
- Do you frequently have to drink liquids when swallowing food?

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Patient responses

- Points to water bottle
  - "I can't go anywhere without it. Constantly consuming water"
- Was planning on discussing with OB/GYN regarding vaginal dryness

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Patient encouraged to get testing from PCP

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Current plan

- Patient resumed Lifitegrast
- On shorter return schedule for compliance
- Returns in a couple months
  - "Did you schedule with your primary care?"
    - (No response)
  - "Do you want us to schedule with your primary care?"
    - (No response)

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COVID happens

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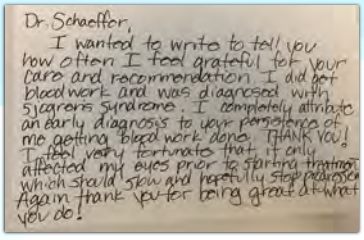
Then I get this in the mail to my office…



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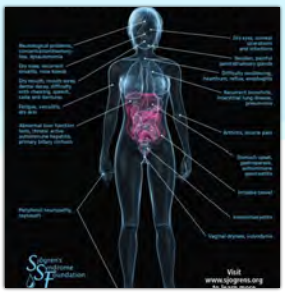
Sjogren's Syndrome

- Auto-immune deficiency of body's salivary glands to produce fluid
- Of individuals with significant aqueous deficient dry eye, 10% are likely to have Sjogren's syndrome
- 26% of patients with either aqueous tear deficiency or evaporative dry eye have an underlying rheumatic condition, including Sjogren's syndrome
- OF 1208 participants in international Sjogren's syndrome registry, 85% reported symptoms of dry eye



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New questions in Sjogren's

1. Is your mouth dry while eating a meal?
2. Can you eat a cracker without drinking any fluids?
3. How often do you have excessive tearing?
4. Are you able to produce tears?

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One more on the "surface"

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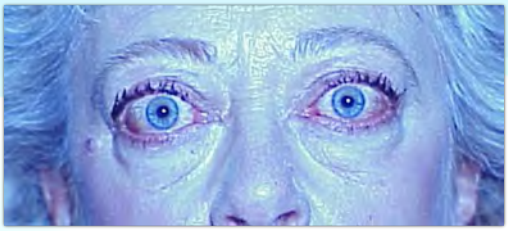
When It's Not Just Ocular Surface Disease



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Thyroid Eye Disease

- Auto-immune disease of the eye and orbital muscles
- Characterized by acute, progressive inflammatory stage then chronic, fibrotic stage 1-3 years later
- Typically found in patients with Grave's disease (40% have TED)
  - Can be found in patients with hypothyroid and euthyroid as well

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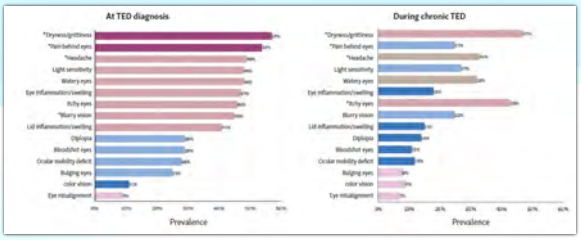
Signs of TED



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Signs of TED

- Orbital congestion (not to be mistaken for conjunctivitis)
- Allergic conjunctivitis without any papillary reaction that doesn't improve with allergy drops
- Unexplained changes in vision that are inconsistent with corneal changes from dryness or other pathologies, which can actually be caused by low-grade chronic compressive optic neuropathy
  - Resistance to retropulsion, an unsatisfactory response to a careful motility check and lid lag on down-gaze can help with this diagnosis
  - Optic nerve imaging with OCT and visual field testing can be helpful in these cases

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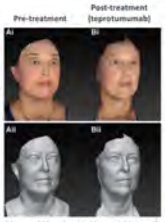
TED is a Condition of the Eye, But Also the Face

In addition to orbital soft tissue expansion, TED is associated with expansion of soft tissue in other regions of the face, including the brow, temples, and mid and lower face

Teprotumumab reduced mean soft tissue volume across facial regions

(n=23 patients with mean TED duration of 29 months)



59-year-old female with 10-month history of TED and expansion of facial soft tissue

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Clinical Experience with Teprotumumab for Chronic TED

- Patient selection
  - 68-year-old Black male with a 9-year history of TED
  - Previous treatment with steroids, bilateral orbital radiation, bilateral orbital decompression, and strabismus surgery
  - Progressively worsening proptosis
    - Exophthalmometry: 32 mm OD, 32.5 mm OS
  - CAS of 3
- After 8 infusions of teprotumumab, the patient saw clinically significant reductions in proptosis and reached a CAS of 0
  - Exophthalmometry: 26 mm OD, 28 mm OS
  - Reduction of 6 mm and 4.5 mm, respectively

Before teprotumumab treatment

After teprotumumab treatment

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Cell-ebrate Good Times!

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27 year old female reports to the clinic

Red Eye / Urgent care appointment

- Complains of pain, redness, and light sensitivity in the right eye
- Started yesterday, but has gotten worse
- Used Visine but didn't help
- Medical history unremarkable*
- Denied prescription medication use, but uses NSAID for headaches, muscle pain

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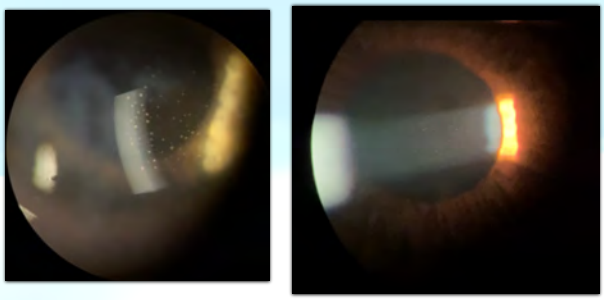

Representative Photos

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Uveitis

Anterior Granulomatous Non-infectious Uveitis OD

- Patient was started on difluprednate qid OD
- Dosed with Cyclopentolate 1% in office
- RTC 24 hours
- Referral to PCP for lab work

- Resolved with steroid treatment
- Quick aside on tapering with a steroid

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Patient (+) RF, ANA
Working diagnosis: Systemic Lupus Erythematosus

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LAB WORKUPS

WHEN DO YOU ORDER LABS?

WHAT DO YOU TELL THE PATIENT?

HOW DO YOU CO-MANAGE WITH SUB-SPECIALTIES?

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WHEN SHOULD LAB TESTS BE ORDERED?

- Bilateral cases
- Atypical age group
- Recurrent uveitis
- Recalcitrant cases
- Hyperacute cases
- Worsens with tapering
- VA worsening
- Immunosuppressed patient

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WHAT LABS TO ORDER?

- *Complete Blood Count with differential (CBC with diff)
- *Erythrocyte Sedimentation Rate (ESR)
- *C-Reactive Protein (CRP)
- *Anti-Nuclear Antibody (ANA)
- *Angiotensin Converting Enzyme (ACE)
- Chest X-ray (CXR)
- Anti-neutrophil Cytoplasmic Antibody (ANCA)
- *Venereal Disease Research Labratory (VDRL) / Rapid Plasma Reagin (RPR)
- *Fluorescent Treponema Antibody Absorption Test (FTA-Abs)
- *Purified Protein Derivative (PPD) or Mantaux Test
- Enzyme Linked Immunosorbent Assay (ELISA) or Immunofluorescence Assay (IFA)
- Western blot
- *Rheumatoid Factor (RF)
- *Human Leukocyte Antigen (HLA)
- CD4 count

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SYSTEMIC LUPUS ERYTHEMATOSUS

Chronic, microvascular inflammatory condition
- Generation of autoantibodies
- Defect in cell apoptosis signal
- Increased cell death

Epidemiology
- 5.1 per 10,000 (number on the rise)
- Average ~6 years from first symptoms to diagnosis

Patient Profile
- F>>>M (7:1 and 11:1 during child-bearing years)
- Affects minorities more

Prognosis
- Had 50% mortality rate previous, has decreased to 10%

Diagnostic Testing
- Anti-Nuclear Antibody (ANA)

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Case

- Dry Eye Consult
- 35yo F
- CC; Blurred vision

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Case

- CC; Blurred vision
- Lupus, Sjogrens
- "Sparkly" Vision
- Kidney issues

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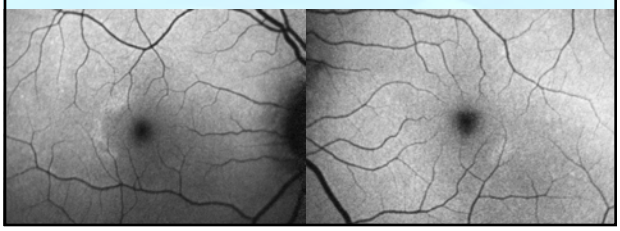
FAF



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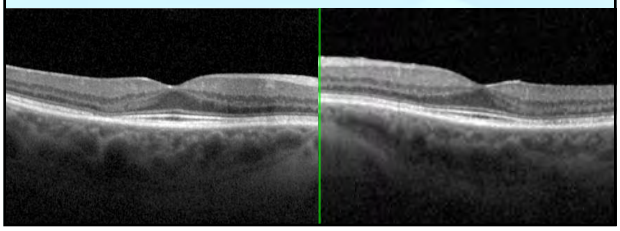
OCT



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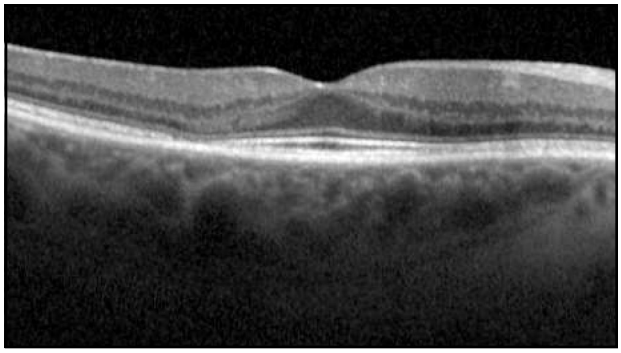

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Chloroquine and Hydroxychloroquine Retinopathy

Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy (2016 Revision)

Michael F Marmor [1], Ulrich Kellner [2], Timothy Y Y Lai [3], Ronald B Melles [4], William F Mieler [5]; American Academy of Ophthalmology

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Chloroquine and Hydroxychloroquine Retinopathy

- Asian patients often show an extramacular pattern of damage.
- DOSE: We recommend a maximum daily HCQ use of ≤5.0 mg/kg real weight, which correlates better with risk than ideal weight.
- There are no similar demographic data for CQ, but dose comparisons in older literature suggest using ≤2.3 mg/kg real weight.

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Chloroquine and Hydroxychloroquine Retinopathy

- The risk of toxicity is dependent on daily dose and duration of use. At recommended doses,
- The risk of toxicity up to 5 years is under 1%
- And up to 10 years is under 2%,
- But it rises to almost 20% after 20 years.
- However, even after 20 years, a patient without toxicity has only a 4% risk of converting in the subsequent year.

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Chloroquine and Hydroxychloroquine Retinopathy

- A baseline fundus examination should be performed to rule out pre-existing maculopathy.
- Begin annual screening after 5 years for patients on acceptable doses and without major risk factors.

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Chloroquine and Hydroxychloroquine Retinopathy

- High dose and long duration of use are the most significant risks.
- Other major factors are concomitant;
  - Renal disease
  - Tamoxifen.

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Chloroquine and Hydroxychloroquine Retinopathy

- The primary screening tests are
  - Automated visual fields (10-2 STD)
  - Spectral-domain optical coherence tomography (SD OCT).
    - The multifocal electroretinogram (mfERG) can provide objective corroboration for visual fields
    - Fundus autofluorescence (FAF) can show damage topographically

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Chloroquine and Hydroxychloroquine Retinopathy

- Retinopathy is not reversible, and there is no present therapy.
- Recognition at an early stage (before any RPE loss) is important to prevent central visual loss.
- However, questionable test results should be repeated or validated with additional procedures to avoid unnecessary cessation of valuable medication.

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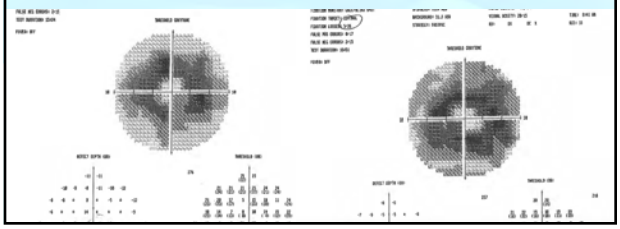
VF



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53 year old male reports for eye exam

- Complains of blurred vision, worse in the evening in the right eye
- History of Type 2 diabetes, "controlled" on medications
  - A1c: 8.2
  - Taking Metformin, Glipizide
- Previous fundus examinations unremarkable for diabetic retinopathy with similar A1c

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Eye Exam and findings

	OD	OS
Refraction	-4.25-0.50x084 20/25-	-3.00-0.25x096 20/20+
Slit lamp exam Pre-dilation	Healthy, unremarkable	Healthy, unremarkable
Cup-to-disc ratio	0.25/0.25	0.25/0.25
Fundus evaluation	(-) hemes, exudates, cotton wool spots, NVD, NVE	(-) hemes, exudates, cotton wool spots, NVD, NVE

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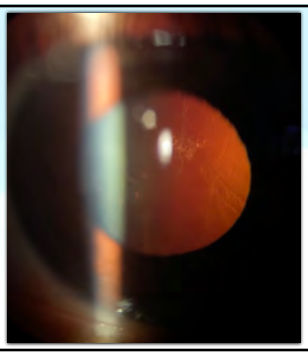

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Diabetic Posterior Subcapsular cataract

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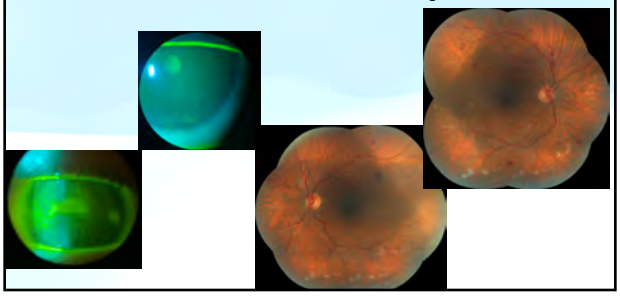
Where diabetes can affect the eye



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Why diabetes can affect the lens

- Protein build-up in the lens
- Can change the index of refraction leading to fluctuating prescriptions
  - Bag of glasses

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Do you test A1c in your office?

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Routine Care that's anything but…

68 year old female reports to the clinic for routine eye exam → Haven't had an eye exam since pandemic began → Wants to update glasses → Hx of hypertension, hyperlipidemia, anxiety

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Patient findings

	OD	OS
BCVA	20/15	20/15
IOP	18mmHg	17mmHg
Cornea	Clear	Clear
Conjunctiva	White and quiet	White and quiet
Iris / Ant Chamber	Deep and quiet	Deep and quiet
Lens	Clear	Clear

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Patient findings continued

	OD	OS
Discs	.2/.2, large area of PPA	.2/.2, large area of PPA
Vitreous	Clear	Clear
Macula	Normal	Normal
Retina	See photo	See photo

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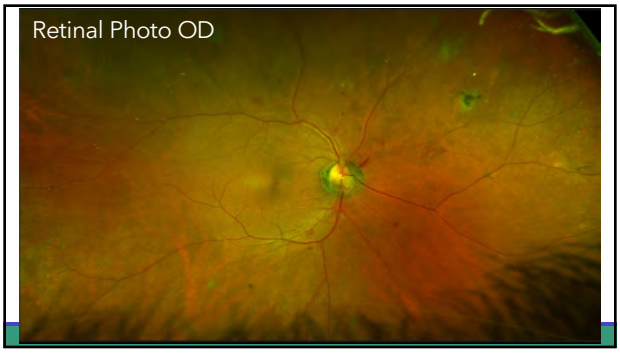
Retinal Photo OD



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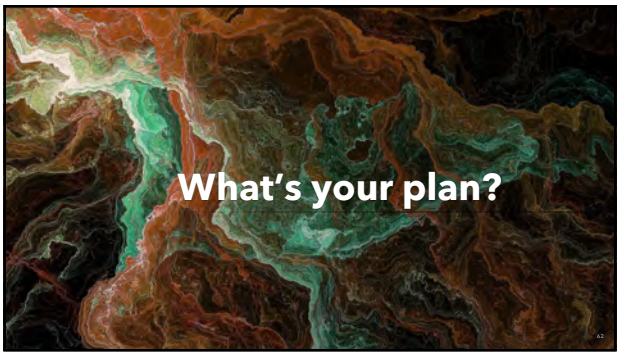

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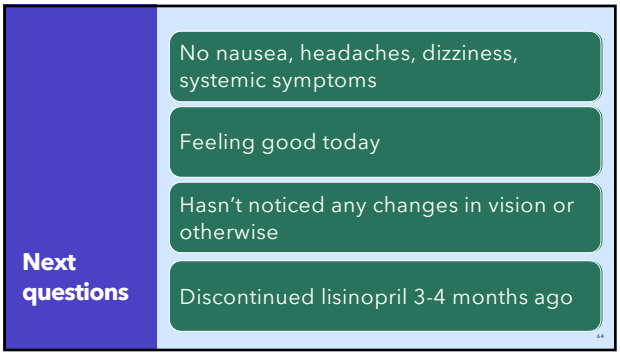

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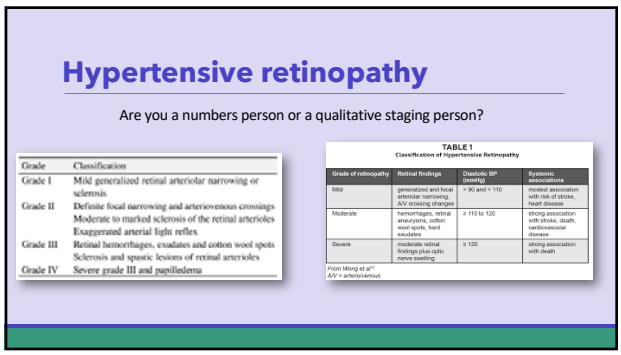

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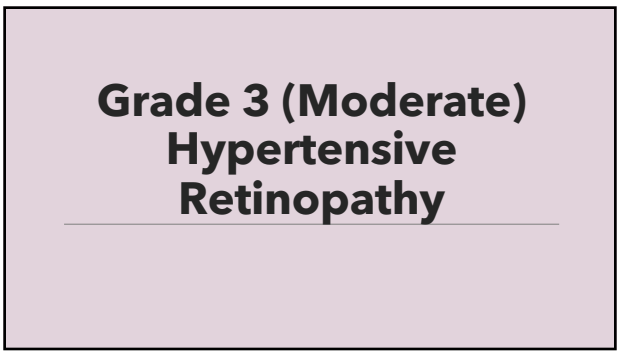

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Next steps

- Go directly to the ER!
- Patient lost to follow-up

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9 months later…

28 year old new patient reports to the office

"Doc, you don't know me but I know who you are. I want to say thank you"

"Because of you, my grandmother got to hold her great granddaughter who was born 4 months ago"

Grandmother went to the ICU for 7 days for impeding stroke

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THANK YOU!

Common Ocular Manifestations From Systemic Associations: Front to Back

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