Ocular Manifestations of Systemic Disease

COPE#45545-SD

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Disclosures

Walter O. Whitley, OD, MBA, FAAO has received consulting fees, honorarium or research funding from:

- Alcon
- Allergan
- Bausch and Lomb
- Biotissue
- Beaver-Visitec
- Ocusoft
- Science Based Health
- Shire
- Sun Pharmaceuticals
- TearLab Corporation
- Tearscience

- Publications
  - Advanced Ocular Care – Co-Chief Medical Editor
  - Review of Optometry – Contributing Editor
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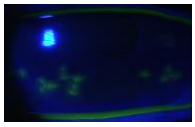
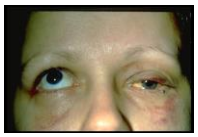
Ocular ER: Big 5 Do Not Miss

- Herpes simplex keratitis
- Intraocular foreign bodies
- Orbital blow-out fracture
- Endophthalmitis
- Temporal arteritis





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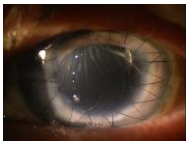
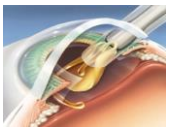
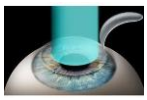
The Herpes Virus Family

- Herpes Simplex
  - HSV-1: Orofacial and ocular infections
  - HSV-2: Genital infections
- Herpes Zoster
- Epstein Barr
- Cytomegalovirus



What are the Triggers?





Herpes Simplex Virus

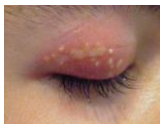
- Primary vs. recurrent infections
- More common as a recurrent HSV
- Remain dormant in the sensory ganglia
- More than 90% carry the latent virus
- Active phase can lead to destructive inflammatory phase

HSV Ocular Signs and Symptoms

- Symptoms
  - Pain
  - Photophobia
  - Blurred VA
  - Tearing
  - Redness
  - FB sensation
- Signs
  - Recurrent
  - Unilateral
  - Eyelid vesicles
  - Epithelial dendrites
  - Decreased K sensitivity
  - K edema
  - KPs
  - Iris stroma / sphincter
  - High IOP
  - Vitritis
  - Retinitis
  - Papillitis

Primary Ocular HSV Infection

- Unilateral blepharoconjunctivitis
  - Follicular conjunctivitis
  - Palpable preauricular lymphadenopathy
- Skin or eyelid vesicles



- Epithelial keratitis
- Stromal keratitis / uveitis are rare

Photo accessed from http://zizzur.com/viewarticle.php?id=173.

Recurrent Ocular Infection

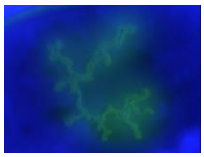
- Reactivation of virus in latently infected sensory ganglion
- Can occur in almost any ocular tissue
  - Blepharoconjunctivitis
  - Epithelial keratitis – lowest risk
  - Stromal keratitis – highest risk
  - Iridocyclitis

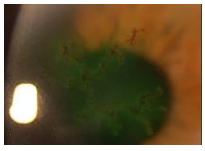
Case Example

- 71YOWF
- OS has a film over it, red, blurry
  - Started 3 days ago
  - Onset was sudden with constant irritation
  - May have gotten eye cream in the eye
- Used Pred Forte and Acuvail last night
- Was out in the garden working on her bushes

Initial Presentation

- BCVA
  - OD: 20/20
  - OS: 20/40
- OS Findings:
  - 2+ Injection
  - Peripheral scars but doesn't recall previous episodes
- IOP:
  - OD 14
  - OS Not taken





Considerations

- Dendrites vs. Pseudodendrites
- Can present as marginal keratitis
- Decreased corneal sensitivity
- Neurotrophic keratopathy

Treatment for HSV Epithelial Keratitis

- Dendritic keratitis usually resolves within 3 weeks
- Goal to minimize stromal damage and scarring
- Consider epithelial debridement
- Topical / Oral antivirals
- Topical steroids??

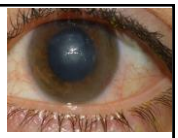
Diagnosis

- HSV Dendritic Keratitis OS
- Treatment:
  - Zirgan 5X daily
- Zirgan 0.15% ganciclovir ophthalmic gel
  - Approved for treatment of acute herpetic keratitis
  - Dosage – One drop 5 times a day until healed, then one drop 3 times a day for 7 days
  - Supplied in 5 gm tube

Ganciclovir Mechanism of Action

- Penetrates cell infected with the virus
- Phosphorylated within the cell to ganciclovir monophosphate by a viral thymidine-kinase
- Activation continues due to several cell kinases leading to formulation of ganciclovir triphosphate
  - Inhibits viral DNA polymerase
  - Incorporates into viral DNA
  - Prevents replication by chain termination

HSV Stromal Keratitis



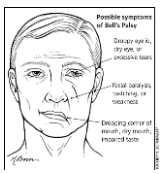
- Interstitial – Non-necrotizing
  - Type III Hypersensitivity
  - Unifocal or multifocal stromal haze
    - With or without neovascularization
  - Disciform
    - Endotheliitis
    - Stromal and epithelial edema
    - Iritis w/ keratic precipitates
- Necrotizing
  - Dense area of stromal inflammation with epithelial defect
  - Difficult to distinguish from bacterial and fungal infections
  - Consider cultures and stains

Treatment for HSV Stromal Keratitis

- Topical corticosteroids
  - Prednisolone acetate1% q2h with taper over 1-2 weeks
  - Difluprednate qid
- Topical / oral antiviral
  - Trifluridine QID
    OR
  - Acyclovir 400 mg BID
    OR
  - Valacyclovir 500 mg QD
  - Use concurrently until patient off steroids

Herpes and Bell's Palsy

- HSV or HZV has been shown to cause Bell's Facial Nerve Palsy
- Main concern is dry eye secondary to poor lid function



Complications of Herpetic Eye Disease

- Epitheliopathy
- Neurotrophic keratopathy
- Severe / chronic recurrent disease
  - Bullous keratopathy
  - Corneal scarring / vascularization
  - Irregular astigmatism
- Penetrating keratoplasty

Oral Antivirals

- Inhibit viral DNA polymerase without inhibiting normal cellular activity
- Works best if treatment initiated within 72 hours
- Pregnancy category B
- Caution in patients with renal disease

Antiviral Drug	HSV	HZO
Acyclovir	400 mg 5x/day for 1 week	800 mg 5x/day for 1 week
Valacyclovir	500 mg TID for 1 week	1000 mg TID for 1 week
Famciclovir	250 mg TID for 1 week	500 mg TID for 1 week

Herpetic Eye Disease Study I

- Herpes Stromal Keratitis, Not on Steroid Trial
  - Pred Phosphate faster resolution and fewer treatment failures
  - Delaying treatment did not affect outcome
- Herpes Stromal Keratitis, on Steroid Treatment
  - No apparent benefit in the addition of oral acyclovir to the treatment of topical corticosteroid and topical antiviral
- ***HSV Iridocyclitis, Receiving Topical Steroids***
  - ***Trend in the results suggests benefit in adding oral acyclovir***

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Herpetic Eye Disease Study II

- HSV Epithelial Keratitis Trial
  - No benefit from oral ACV with topical trifluridine in preventing the development of stromal keratitis / iritis
- ***Acyclovir Prevention Trial***
  - ***Reduced by 41% the probability of recurrence***
  - ***50% reduction in the rate of return of the more severe form***
- Ocular HSV Recurrence Factor Study
  - No results available

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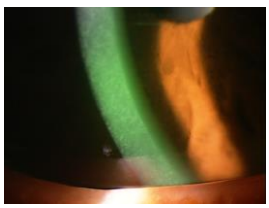
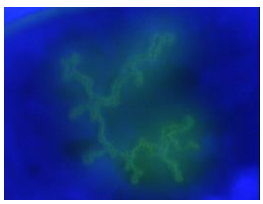
Orals for Simplex???





Case Example - BL

- 63YOWM Referred by PCP for sudden decrease VA OD and swelling of eyelids OD>OS for 1 week
  - Pressure from forehead to cheek
  - Worse in evenings
  - Mild seasonal allergies
  - Some tearing and redness OD

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Examination

- Non-healing scab on R forehead
- Conjunctiva: 2+ injection OD
- K: 2+SPK, 2+ MCE, 1+ KPs, No dendrites OD
- AC: 2+ Cells OD
- Lens: 2+ NS OD / 1+NS OS
- IOP: 31/13

Diagnosis???

- Considerations:
  - PCP told him he had an infection not shingles
  - Episode started 3 weeks prior
- Treatment
  - Valacyclovir 1000mg TID po
  - Difluprednate QID OD
  - Timolol 0.5% QAM OD
  - F/u 1 week

Herpes Varicella-Zoster Virus

- Primary infections: Chicken pox
  - Remains latent in dorsal root or other sensory ganglia after primary infection
  - May lie dormant for years to decades
- Later infections: Shingles
  - Virus specific cell-mediated immune responses decline
  - Localized cutaneous rash erupting in a single dermatome
  - HZO accounts for 10-25% of all cases of shingles

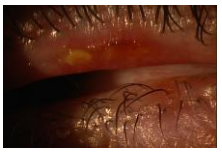
Herpes Zoster Ophthalmicus

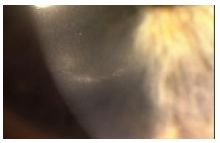
- 90% of U.S. population infected with VZV by adolescence
- 100% of U.S. population by 60 years of age
- 1.5-3.4 cases per 1,000 individuals

http://emedicine.medscape.com/article/783223-overview#aw2aab6b4

Herpes Zoster Ophthalmicus

- Conjunctivitis
- Scleritis
- Pseudodendrites
- Keratic precipitates
- Iritis
- Synechiae
- Neurotrophic keratitis
- Elevated IOP
- Potential vascular occlusion
- Nerve palsies
- Glaucoma (longer-term)





HZO: Signs and Symptoms

- Prodromal phase: fatigue, malaise, low-grade fever
- Unilateral rash over the forehead, upper eyelid, and nose
  - 60% of patient have dermatomal pain prior to rash
  - Erythematous macules to papules to vesicles to pustules to crusts
  - Other symptoms: eye pain, conjunctivitis, tearing, decrease VA, eyelid rash
  - Hutchinson's sign
- Post-herpetic neuralgia: >12 months for 50%

HZO: Treatment

- Local wound care
- Analgesia
- Antivirals
  - Valtrex 1g TID
- Antibiotics??
- Oral corticosteroids
- Post-herpetic neuralgia
  - Tricyclic antidepressants
  - Topical capsaicin ung
  - Gabapentin

Vaccines for HZO - Zostavax

- Zostavax is live attenuated herpes zoster (HZ) virus
  - >50% reduction in the incidence of HZ
  - >60% reduction in symptom severity in patients who developed HZ
  - 66.5% reduction in postherpetic neuralgia.
- Must have chicken pox as a child
- May help patients who've had HZO already

1. Oxman MN, Levin MJ, Johnson GR, et al. A vaccine to prevent herpes zoster and postherpetic neuralgia in older adults. N Engl J Med. 2005 Jun 2;352(22):2271-84.

"The Common Eyeritis"

- 32YOWM, Red, Painful Eye OD, Photophobic, No discharge
- No previous episodes
- Ocular/Medical Hx: unremarkable
- No other associated symptoms
- SLE: 2+ injection / 2+ cells

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Case Example - AM

- 44yo Asian American c/o blurred VA, redness, tearing, periorbital edema starting 2-3 days prior
- Med Hx: Uncontrolled DM (Dx in 1998)
- Vasc: OD 20/60 PH 20/30
  OS 20/80 PH 20/40
- IOP: 21 / 18

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Uveitis

- Classic Symptoms
  - Acute onset
  - Decreased vision
  - Redness
  - Photophobia
  - Pain
  - Excessive tearing

Retrieved from http://eyecaremanual.com/eye-conditions/photophobia/photophobia.html on 3/22/11

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Clinical Signs

- VA
- Conjunctiva
- Cornea
- Anterior chamber
- Iris
- Pupil
- IOP
- Lens
- Vitreous
- Disc edema
- Macular edema
- Periphlebitis

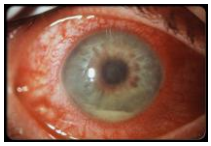

Photo accessed from http://www.medicinenet.com/image-collection/uveitis_picture/picture.htm

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What is Your Treatment?

- Prednisolone acetate 1% vs. difluprednate 0.05% vs. loteprednol etabonate .5%
- Homatropine 5% vs. Scopolamine 0.25% vs. Atropine 1%
- Would you prescribe an oral medication?
- Would you consider lab testing?

Case Example

- Acute, non-granulomatous, anterior uveitis OS
- Cause???
- Treatment
  - Ordered labs – CBC w/diff, ESR, SMA-12, HLA-B27, Urinalysis, FTA-ABS, HgA1c, FBS, RPR, Lyme Western Blot
  - Difluprednate q2h OS
  - Homatropine 0.5% TID OS
  - Doxycyline 100 mg BID po

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Pulse Therapy

- QID to Q 1 Hour for 7 to 10 Days
- Zero Tolerance for AC Cells
- Avoids Surface Toxicity
- Quick & Dirty
- Hit It Hard and Fast: Aggressive

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Lab Testing

- Minimum lab testing
  - CBC with differential
  - Erythrocyte sedimentation rate (ESR)
  - Angiotensin converting enzyme (ACE)
  - Venereal disease research laboratory (VDRL)
  - Fluorescent treponemal antibody absorption (FTA-ABS)
  - Lyme titers in endemic areas***
  - HLA-B27
  - Antinuclear antibody (ANA)
  - Urinalysis
  - Chest X-ray
  - PPD***

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Considerations

- Joint pain???
- Breathing problems???
- Retrobulbar eye pain???
- Skin lesions???
- Retinal scars???

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Condition	Clinical Features	Test Indicated
Ankylosing spondylitis	Young male, low back pain, chest pain	HLA-B27, sacroiliac X-ray
Reiter's syndrome	Young male, arthritis, urethritis, conjunctivitis	HLA-B27, ESR, CRP
Juvenile idiopathic arthritis	Slight male predilection, sacroiliitis common	ANA, RF, knee radiograph
Inflammatory bowel disease	Ulcerative colitis, diarrhea, abdominal cramps	HLA-B27, GI referral for endoscopy
Sarcoidosis	African Americans, females, vasculitis, vitritis	ACE, chest X-ray or CT scan
Tuberculosis	Prolonged cough, fever, chills, night sweats, weight loss	PPD, chest X-ray
Syphilis	Hx of sexual contact with infected person, rash, fever, malaise, headache, joint pain	FTA-ABS, VDRL, RPR
Toxoplasmosis	Immunocompromised status, exposure to cats, hx of eating raw meat, punched-out retinal lesions	Toxoplasma IgG or IgM for acute acquired cases
Lyme disease	Recent tick bite	Lyme Western Blot

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Name:
Address:

RX:
- ✓ CBC /Diff -**86361**
- ___ ESR
- ___ CRP
- ✓ SMA-12
- ___ HLA B27
- ✓ ACE
- ___ Urinalysis
- ___ RHEUMATOID FACTOR
- ___ CH SO
- ✓ ANA -**86039**
- ___ Lysozyme Total Serum 1g E
- ___ RAST Zone II (Southeast)
- ___ Cardiolipin Antibody, profile- **86147x3**
- ___ Bartonella Antibodies- **86611 x 4**
- ___ Brucella Antibodies - **86622 x 2**
- ___ Sjogren's Antibodies -**86237 x 2**
- ✓ RPR- **86593**
- ___ FTA- Ab-**86781**
- ___ Lyme Western Blot IgG & IgM-**86617 x 2**
- ___ Toxoplasmosis IgG

DX: ✓
- ___ Uveitis
- ___ Pan
- ___ Arthristis
- ___ Diffise
- ___ Severe Atropic Disease
- ___ Sarcoidosis
- ___ Scleritis

O.D.

Uveitis: Common Systemic Associations

- Most common cause
  - Idiopathic : 38-70%
- Other systemic causes
  - HLA-B27 related disease
    - Ankylosing spondylitis
    - Reactive arthritis
    - Psoriatic arthritis
    - Inflammatory bowel disease
  - Sarcoidosis
  - Systemic Lupus Erythematosus
  - Rheumatoid Arthritis
  - Behcets Disease

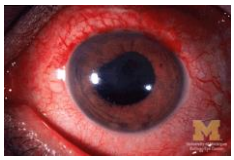

Photo accessed from http://www.aao.org/theeyeshaveit/red-eye/images/anterior-uveitis.jpg

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Lyme Titer

- Ordered based on suspicion
- Erythema migrans is the only manifestation of Lyme disease in the United States for which clinical diagnosis should be made in the absence of laboratory confirmation
- A patient with a significantly characteristic symptom with the appropriate history of possible exposure should be started on antibiotics after appropriate laboratory studies have been drawn

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Tx for Lyme Disease

- Early infection or nonspecific symptoms with positive Lyme titers in the adult may be treated with oral doxycycline (100 mg twice daily for 4 to 6 weeks) or tetracycline (500 mg four times a day for 4 to 6 weeks).
- Severe infection in adults with definitive ocular, neuroophthalmic, neurological, or cardiac involvement may be treated with penicillin G (24 million units, intravenous, daily in four divided doses for 21 days) or intravenous ceftriaxone (2 g/day in two divided doses for 21days.

Case Example - You've Got to be Kidding Me!

- 27yowm presents with red, painful, blurry VA OS. Started 10 days ago after returning from a trip to Italy. Taking 500mg Naprosyn for HA.
- Health – Unremarkable
- Allergy to PCN
- Vasx: OD 20/20-3 OS 20/25-3 with NI
- IOP: 9 / 10
- SLE:
  - OD Mild limbal flush / 1+ Cells
  - OS 2+ Inj / 2+ Cells

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What is Your Treatment?

- Prednisolone acetate 1% vs. difluprednate 0.05% vs. loteprednol etabonate .5%
- Homatropine 5% vs. Scopolamine 0.25% vs. Atropine 1%
- Would you consider lab testing?
- Would you prescribe an oral medication?

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Case Example

- Acute, bilateral non-granulomatous, anterior uveitis OU
- Cause???
- Treatment
  - Difluprednate qid OD, q2h OS
  - Cyclopentolate 2% TID OU
- Labs???

Screening Tests for Syphilis

- Venereal Disease Research Lab (VDRL)
  - VDRL may become non-reactive in latent syphilis or after successful treatment
  - False positives may occur in:
    - Pregnancy
    - Infectious mononucleosis
    - Systemic lupus erythematosis
- Rapid Plasma Reagin (RPR)
  - Alternative to VDRL

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Fluorescent Treponemal Antibody Absorption (FTA-ABS)

- Detects specific antibodies against T pallidum
- Confirms diagnosis of syphilis
  - More specific than VDRL
  - More sensitive in primary syphilis
- Test may remain positive for life
- Reactive:
  - Primary syphilis 95%
  - Secondary 100%
  - Late latent 100%
  - Tertiary 96%
  - False positives may occur in pregnancy and SLE

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Syphilis

- STD caused by T pallidum / great imitator / any tissue and organ
- Sexually active / multiple partners
- Systemic Sx – Depends on stage – primary painless ulcer / secondary skin rash palms, soles, trunk / tertiary neurosyphilis
- All types of ocular inflammation
- Labs
  - VDRL / RPR
  - FTA – ABS
  - ESR elevated
- Tx – penicillin therapy
- Good prognosis if treated early

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So He Has an Allergy to PCN?

- Augenbraun M, Workowski K. Ceftriaxone therapy for syphilis: report from the emerging infections network. Clin Infect Dis. 1999 Nov. 29(5):1337-8
  - Tetracycline, erythromycin, and ceftriaxone have shown antitreponemal activity in clinical trial
- Slow taper of steroid
- Lost to follow up

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Uveitis - Take Home Pearls

- Be a detective and find the cause
- Be aggressive with treatment
- Don't taper too soon
- Treat and follow

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Case Example - SD

- 38 year old, African American, Female presents with red, painful, and photophobic OS
- Started 3 weeks ago / similar episode 10 years ago
- Tried dexamethasone 0.1% but no relief
- BCVA OD 20/25 OS 20/20
- IOP: 17 mmHg

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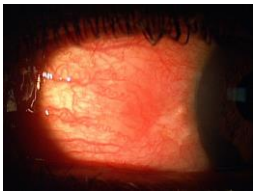
Differentials

- Conjunctivitis
- Episcleritis
- Scleritis
- Uveitis



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When Should Lab Tests Be Ordered?

- Bilateral cases
- Atypical age group
- Recurrent uveitis
- Scleritis
- Recalcitrant cases
- Hyperacute cases
- Worsens with tapering
- VA worsens
- Immunosuppressed

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Treatment for Scleritis

- NSAIDS
- Systemic steroids
- Immunosuppressive therapy
- Topical steroids???

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Rheumatoid Arthritis

- Middle aged women
- Arthritis affecting both sides equally
- Morning stiffness
- Inflammation of joints and tissue
- Diagnostic Testing
  - Positive rheumatoid factor
  - Anti-CCP present
  - Elevated CBC
  - Joint X-ray

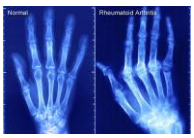

Photo accessed from http://img.webmd.com/dtmcms/live/webmd/consumer_assets/site_images/articles/health_tools/rheumatoid_arthritis_overview_slideshow/PRinc_rm_x-ray_of_rheumatoid_arthritis.jpg

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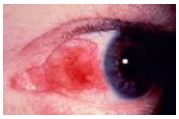
Rheumatoid Arthritis

- 25% RA patients have ocular manifestations
  - Keratoconjunctivitis sicca - 15-25% patients
    - Sjogrens frequently accompanies RA
  - Episcleritis
  - Most common systemic condition associated with scleritis
  - Uveitis



Rheumatoid Factor (RF)

- Differentiates RA from other chronic arthritides
- Positive values (titers > 1:80) occur in approximately 70% of patients with rheumatoid arthritis
- Positive in only 5% of patients with JRA
- Can be positive in the following
  - Sjogren's
  - SLE
  - Syphilis
  - Chronic infections
  - Sarcoidosis
  - Liver disease

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Rheumatoid Arthritis Treatment

- NSAIDs
- Steroids
- Disease Modifying Anti-rheumatic Drugs
  - Methotrexate
  - Sulfasalazine
  - Hydroxychloroquine

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Plaquenil (hydroxychloroquine sulfate)

- Indicated for the treatment of discoid and systemic lupus erythematosus, rheumatoid arthritis, and malaria
- Dosage: 200mg to 400mg per day
- Primary risk factors
  - Duration > 5 years
  - Cumulative dose >1000g
  - Age
  - Systemic – High BMI, liver, kidney dysfunction
  - Ocular – retina or macular changes

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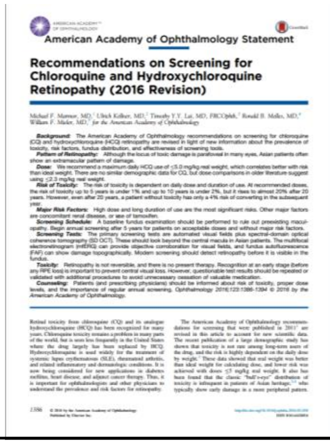

American Academy of Ophthalmology Statement

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

Michael F. Marmor, MD, Ulrich Kellner, MD, Timothy Y.Y. Lai, MD, FRCOphth, Ronald B. Melles, MD, William F. Mieler, MD, for the American Academy of Ophthalmology

Background: The American Academy of Ophthalmology recommendations on screening for chloroquine (CQ) and hydroxychloroquine (HCQ) retinopathy are revised in light of new information about the prevalence of toxicity, risk factors, fundus distribution, and effectiveness of screening tools.
Pattern of Retinopathy: Although the locus of toxic damage is parafoveal in many eyes, 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.
Risk of Toxicity: 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.
Major Risk Factors: High dose and long duration of use are the most significant risks. Other major factors are concomitant renal disease, or use of tamoxifen.
Screening Schedule: A baseline fundus examination should be performed to rule out preexisting maculopathy. Begin annual screening after 5 years for patients on acceptable doses and without major risk factors.
Screening Tests: The primary screening tests are automated visual fields plus spectral-domain optical coherence tomography (SD OCT). These should look beyond the central macula in Asian patients. The multifocal electroretinogram (mfERG) can provide objective corroboration for visual fields, and fundus autofluorescence (FAF) can show damage topographically. Modern screening should detect retinopathy before it is visible in the fundus.
Toxicity: 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.
Counseling: Patients (and prescribing physicians) should be informed about risk of toxicity, proper dose levels, and the importance of regular annual screening. Ophthalmology 2016;123:1386-1394 © 2016 by the American Academy of Ophthalmology.

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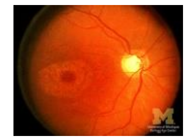
Plaquenil Examinations

- Complete dilated examination
- Color vision / Amsler??
- Central visual field testing 10-2
- Fundus photography for co-existing retinal disease
- Spectral domain OCT, FAF, mfERG (if available)



Accessed from http://www.kellogg.umich.edu/theeyeshaveit/side-effects/chloroquine.html

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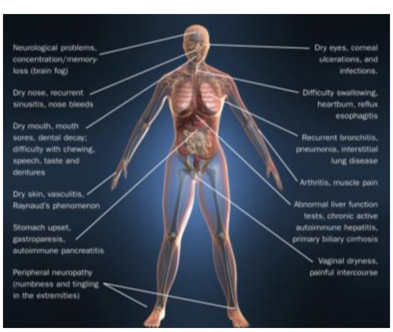
Sjögren's Is More than Dry Eye[1]


Neurological problems, concentration/memory loss (brain fog)
Dry nose, recurrent sinusitis, nose bleeds
Dry mouth, mouth sores, dental decay; difficulty with chewing, speech, taste and dentures
Dry skin, vasculitis, Raynaud's phenomenon
Stomach upset, gastroparesis, autoimmune pancreatitis
Peripheral neuropathy (numbness and tingling in the extremities)
Dry eyes, corneal ulcerations, and infections.
Difficulty swallowing, heartburn, reflux esophagitis
Recurrent bronchitis, pneumonia, interstitial lung disease
Arthritis, muscle pain
Abnormal liver function tests, chronic active autoimmune hepatitis, primary biliary cirrhosis
Vaginal dryness, painful intercourse

1. http://www.sjogrens.org/home/about-sjogrens-syndrome/symptoms.

Recent Clinical Findings for Sjögren's Diagnostics[1-4]

Current Screening	New SS Panel
• Combined serology sensitivity & specificity is around 40-60%	• Combined serology sensitivity & specificity is 87% and 82.5% respectively • Cumulative specificity of 92.2% for CA6, SP-1, and PSP
• None of the serology test diagnose SS early • Misses approximately 25-35 % cases	• Approximately 50% of the **early & new** cases are identified (Ro and La Negative) • Picks up additional cases
• All serology tests identifies are non-organ specific auto-antibodies and could occur in other autoimmune diseases	• Comprises of both organ/non-organ specific auto-antibodies

1. Tincani A, et al. Novel aspects of Sjögren's Syndrome in 2012. BMC Med Apr 4 2013;11:93. doi: 10.1186/1741-7015-11-93. 2. Shen L, et al. Novel autoantibodies in Sjogren's Syndrome. Clin Immunol 2012;145:251-255. 3. Huang Y, et al. The immune factors involved in the pathogenesis, diagnosis, and treatment of Sjogren's Syndrome. Clin Dev Immunol 2013; Article ID 160491. doi:10.1155/2013/160491.
4. Ramos-Casals M, Brito-Zeron P, Siso-Almirall A, Bosch X. Primary Sjogren's Syndrome. BMJ 2012;344:e3821

Sarcoidosis

- Systemic inflammatory disease forming granulomas in organs (lungs, lymph nodes, skin, eyes)
- Often young, African American females
- Enlarged lymph nodes
- Shortness of breath
- Fatigue
- Diagnostic Testing
  - Chest X-ray
  - Elevated ACE
  - PPD: TB vs. Sarcoid
  - Biopsy of nodule

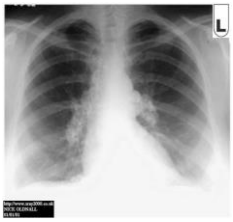


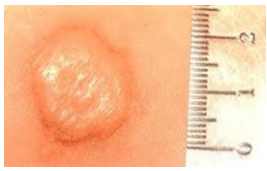
Photo accessed from http://www.e-radiography.net/radpath/s/sarcoidosis.jpg

Sarcoidosis

- Ocular manifestations
  - Redness, pain welling of lids or lacrimal gland
  - Painless subcutaneous nodular mass of eyelids
  - Ptosis
  - Diplopia
  - Cicatrizing conjunctival inflammation
  - Conjunctival nodules
  - Keratoconjunctivitis sicca
  - Band keratopathy
  - Granulomatous anterior or posterior uveitis
  - Cataract
  - Chorioretinitis
  - Retinal periphlebitis or neovascularization
  - Optic nerve disease or glaucoma

Purified Protein Derivative (PPD)

- Skin test to screen for tuberculosis
- Intradermal injection of 0.1ml of soluble antigen from a given TB organism in forearm
  - Positive test – 5 – 15 induration in 2-3 days
- Specificity increased with chest x-ray
- False positives include prior exposure to TB



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QuantiFeron TB Gold (QTF-G)

- An alternative to skin testing of cell-mediated immune response to antigens simulating the mycobacterial proteins ESAT-6, CFP-10, and TB7.7
- < 12 hours
- A positive result indicates that Mycobacterium tuberculosis infection is likely
  - Positive tests should be followed by further medical and diagnostic evaluation for tuberculosis disease (eg, acid-fast bacilli smear and culture, chest x-ray).
- QuantiFERON-TB Gold is usually negative in individuals vaccinated with Mycobacterium bovis bacille Calmett-Guerin

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Tuberculosis

- Poverty is the primary risk factor
- Lungs most commonly affected
- Uveitis is the most common eye complication
- Immune suppression
- Fever, Night Sweats, Fatigue
- Posterior and Pan-uveitis most common

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Angiotensin Converting Enzyme (ACE)

- Produced by a variety of cells including granulomatous cells
- Serum ACE levels reflect the total amount of granulomatous tissue in the body
- Screen for sarcoidosis
  - 75% sensitive
  - 95% specific
- False positives include:
  - TB
  - Lymphomas
  - Leprosy
- Consider serum lysozyme / calcium assay

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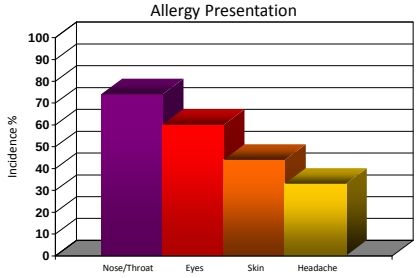
Sarcoidosis

- Treatment
  - Aforementioned blood work
  - Uveitis topical and possible oral steroids
  - Dacryoadentitis treated with systemic steroids
  - Consult with PCP

Incidence of Allergic Symptoms

Eye Symptoms Are the Second Most Common Allergy Presentation



6 out of 10 allergy patients suffer ocular allergy symptoms

The 1999 Gallup Study of Allergies and Allergic Symptoms Affecting the Nose, Throat, Eyes, and Skin

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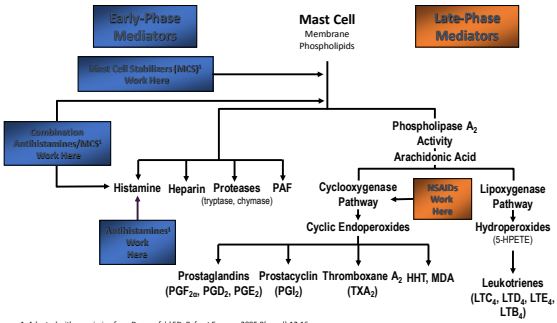
Most Prescription Treatment Options Have a Limited Effect on the Inflammatory Cascade



1. Adapted with permission from Donnenfeld ED. *Refract Eyecare*. 2005;9(suppl):12-16.
2. Slonim CB. *Rev Ophthalmol*. 2000:101-112.

Graded Pharmacotherapy

Stepwise Treatment Strategies for Allergic Conjunctivitis	
Mild	Avoidance, cold compresses, tears, over-the-counter medications
	Topical antihistamines/mast cell stabilizers
	Oral antiallergics (allergists may already have patients on orals; may exacerbate the ocular condition while improving the nasal condition) Montelukast
Moderate	+ Mast cell stabilizers (treats allergy before mediator is released)
	+ Combination antihistamine/mast cell stabilizers
	+ Topical corticosteroids (most beneficial for severe outbreaks)
Severe	Topical corticosteroids (short course; fluorometholone/dexamethasone/loteprednol/prednisolone)
	Topical immunomodulating agents (tacrolimus, cyclosporine)
	Oral steroids

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Specific Allergy Therapy

- Preventive
- Palliative
- Alternative
- Immunotherapy
- Pharmacologic
  - Topical, Nasal, Inhaled
  - Dermatologic
  - Systemic

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Singulair (montelukast sodium)

- Leukotriene receptor antagonist
- Indications:
  - Prophylaxis and chronic treatment for asthma
  - Acute prevention of exercise-induced brochoconstriction
  - Relief of symptoms of allergic rhinitis
- 10 mg tablet qd
- Side effects
  - Behavior or mood changes, URI, fever, headache, sore throat, cough, stomach pain, diarrhea, ear ache or ear infection, flu, runny nose, and sinus infection

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12 Patient Allergy Tips

- Never rub your eyes
- Wash your hands
- Use allergy free pillows
- Stay indoors
- Use drops for eyes, sprays for nose
- Avoid "get the red" out vasoconstrictors
- Chill your drops
- Use cool compresses
- Apply allergy drops proactively
- Pets out of the house or bedroom
- Know and avoid your personal antigens

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Cataract / Refractive Surgery Complications

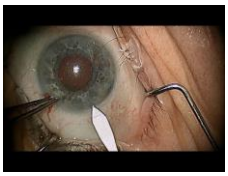
- Operative Complications
  - Surgeon makes the call
- Post-operative Complications
  - Co-managing doctor makes the call
- ***Successful cataract surgery is the result of continuous communication!!***

Benign Prostatic Hyperplasia

- Histologic diagnosis characterized by proliferation of the cellular elements of the prostate
- An estimated 50% of men demonstrate histopathologic BPH by age 60 years. This number increases to 90% by age 85 years
- Symptoms: Urinary frequency and urgency, Hesitancy, Incomplete bladder emptying, Straining, Decreased force of stream

Flomax (tamsulosin)

- Indication for the treatment of benign prostatic hyperplasia
- Intraoperative floppy iris syndrome
- Importance to communicate prior to cataract surgery



Omidria™

(phenylephrine and ketorolac injection) 1% / 0.3%

- OMIDRIA™ is an α1-adrenergic receptor agonist and nonselective cyclooxygenase inhibitor indicated for:
  - Maintaining pupil size by preventing intraoperative miosis
  - Reducing postoperative ocular pain
  - OMIDRIA is added to an irrigation solution used during cataract surgery or intraocular lens replacement.

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http://sagespoonliving.com/wp-content/uploads/2012/11/diabesity.jpg accessed on 10/15/15

Neuropathy is end stage organ damage

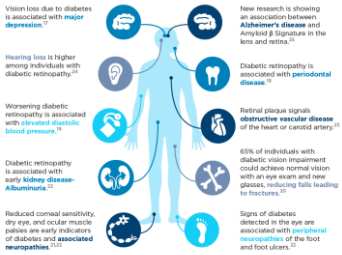
- Diabetics know this first hand
- All diabetics get dry eye, few complain about it.

Epidemiology

- Systemic, microvascular disease affecting (not limited to) the liver, kidneys, and eyes.
  - Type I caused by destruction of the Islets of Langerhans in the Pancreas.
  - Type II caused by the body's developed resistance to insulin.
- It is the most common cause of blindness in the 20-70 year old population.
  - Diabetic retinopathy is prevalent in 30% of the diabetic population.

Eye Care Disease Prevalence

What You Might Not Know



Eye Disease Management

A Key Component of Diabetes Care

People with diabetes do have a higher risk of prominent eye conditions and blindness than people without diabetes. Prevalent diseases include:

People with diabetes are **40% more likely** to suffer from glaucoma than people without diabetes. The longer someone has had diabetes, the more common glaucoma is. Risk also increases with age. [5]

Many people without diabetes get cataracts, but people with diabetes are 60% more likely to develop this eye condition. People with diabetes also tend to get cataracts at a younger age and have them progress faster. [6]

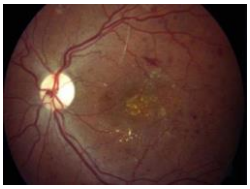
Diabetic retinopathy accounts for approximately 12% of all new cases of blindness each year. It can cause vision loss in two ways: Macular Edema and Proliferative Retinopathy and Vitreous Hemorrhage [7]

Diabetic Retinopathy

- Occurs due to a breakdown in the retina's ability to auto regulate its blood supply properly.
  - Hyperglycemia increases retinal blood flow and therefore causes "capillary hypertension."
- This hypoxic environment causes an up-regulation of the angiogenic factor VEGF.
  - VEGF stimulates the growth of new blood vessels to meet the needs of the starving retina.

Risk factors for developing DR

- Duration of DM
- Control of DM will not prevent but delays
  - Fasting BS <126 and A1C <7%
- Hypertension/Hyperlipidaemia
- Renal Disease
- Pregnancy
- Sleep apnea
- Obesity
- Smoking
- Anaemia



ETDRS Classification of DR

DR Level	Retinal Findings
Mild NPDR	At least one MA and 1 or more of following • Retinal hemorrhages • Hard exudates • Soft exudates
Moderate NPDR	Hemorrhages and MA or soft exudates, VB, and IRMA present
Severe NPDR	Any of the following and no signs of PDR (4-2-1 rule) • >20 intraretinal hemorrhages in each of 4 quadrants • Definite venous beading in 2 or more quadrants • Prominent IRMA in 1 or more quadrants
Very Severe NPDR	• 2 or more of lesions of Severe NPDR
PDR	One of either • Neovascularization • Vitreous/preretinal hemorrhage

ETDRS PDR

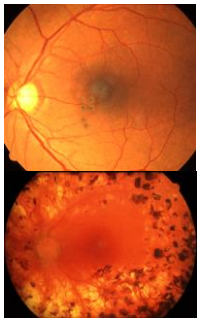
- Presence of Neovascularization with/without pre-retinal hemes.
- Early/Low-risk PDR:
  - Any size NVE without vitreous heme.
  - NVD <1/4 in size without vitreous heme.
- High-Risk PDR - 1 or more of the following
  - NVD approximately 1/4DD- 1/3DD or more in size
  - NVD less than ¼ DD in size when fresh VH or PRH is present
  - NVE greater than or equal to 1/2DD in size when fresh VH or PRH is present
    - If a patient is at this stage, severe vision loss is likely if no treatment initiated.

New Treatments Diabetic Retinopathy

- Proliferative Disease:
  - Pan Retinal Photocoagulation or Focal Laser
  - Ranivizumab (lucentis) and Aflibercept (Eylea)-anti-VEGF
- Diabetic Macular Edema:
  - Dexamethasone (Ozurdex),Fluocinolone acetonide (Iluvien)
  - Anti-VEGF (above)
- RISE/RIDE studies showed a 3 line VA improvement in diabetic eyes treated with anti VEGF

PDR Treatment

- LASER: Light Amplification by the Stimulated Emission of Radiation
  - Focal
  - Grid
  - Panretinal photocoagulation
- Anti-VEGF ocular injections



Treatment for PDR: Vitrectomy

- Indicated after weeks to months of blood not clearing from vitreous heme
- Best results if done within 6 months of heme (DRVS)
- Usually done at 6 weeks
- Alleviate retinal traction, ERM
- When PRP is not enough
- Cataracts!

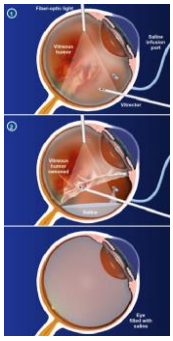


Photo accessed from http://www.eyecentersofsetexas.com/college_location/treatments.php?qi=78

Diabetic Macular Edema

- DME incidence based on duration and type of diabetes
  - IDDM
    - <8 years rare
    - 10 years 7-10% incidence DME
    - 20 years 25-30% incidence DME
  - NIDDM
    - 10 years 5% incidence DME
    - 20 years 15% incidence DME
    - NIDDM w/ insulin use
      - 10 years 10% incidence DME
      - 20 years 30-35% incidence DME

DME

- DME is also closely associated with degree of DR present
  - Mild NPDR ~ 3 % incidence
  - Moderate- Severe NPDR ~40% incidence
  - PDR ~71% incidence

DME

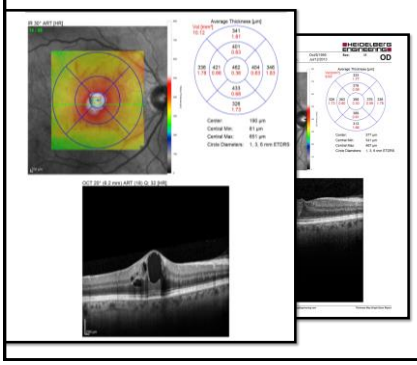
- Signs: circinate ring of exudates, retinal thickening, retinal elevation.
- Clinically Significant Macular Edema:
  - Thickening of retina at or within 500 microns of the center of the macula
  - Hard exudates at or within 500 microns of the center of the macula
  - A zone or zones of retinal thickening 1 disc area or larger in size that is within 1 disc diameter of the center of the macula.

Current Treatment

- New mainstay treatment: Intravitreal Injections
  - Lucentis (FDA)
  - Eyelea (recent FDA)
  - Ozurdex (recent FDA)
  - Avastin
  - Triamcinolone
- Focal laser with intravitreal injection

Before Injections OD

After Injections OD



Case Study

- 2/13 ROV: 52 YO Asian Female / Follow up 4 month dry eye check. Intermittent foreign body sensation and fogged vision over 1 year
- Ocular Hx: DES, LASIK 12.08.11
  - Ocular Medications: Restasis BID OU
- Medical Hx: Allergies, Borderline Diabetes, Acid Reflux
  - Systemic Medications: Multivitamin, Iron

Slit Lamp Examination

- BCVA
  - OD 20/25-
  - OS 20/20-
- MR
  - OD pl – 0.75 x 005
  - OS -0.50 DS
- External: normal OU
- Conjunctiva: 2+ injection
- Cornea: 1+ Diffuse SPK OU
- Tear Eval:
  - 4 sec NIBUT
  - Schirmer 8/9
- Iris: flat OU
- A/C: deep & quiet OU
- Lens: clear OU

Diagnostic Testing

- Screening questionnaire
- Blink rate
- Tear meniscus
- Tear film osmolarity
- Tear film break up time
- Ocular surface staining
- Schirmer / Red Thread Test
- Lid Evaluation
  - Lid and MG morphology
  - MG Expression
- Tear interferometry
- Presence of MMP-9

Superior Limbic Keratitis

- Definition
  - Uncommon chronic disease
  - Superior bulbar and tarsal conjunctiva and limbus
  - Bilateral
  - Middle aged women
  - Abnormal thyroid function
  - Symptoms worse than signs
  - Remission occurs spontaneously
- Pathogenesis
  - Blink-related trauma
  - Tear film insufficiency
  - Excess of lax conjunctival tissue
  - Inflammatory process
  - Self-perpetuating cycle

SLK and Treatment

- Lubrication
- Acetylcysteine
- Mast cell stabilizers
- Steroids
- Cyclosporin A
- Soft contact lens
- Silver nitrate
- Autologous serum
- Botulinum toxin
- Supratarsal steroid injection
- Resection
- Conjunctival ablation
- Consider thyroid evaluation

Thyroid Disease Causes

- Hypothyroidism
  - Hashimotos Disease
  - Thyroid removal
  - Pituitary gland malfunctions (TSH)
  - Low iodine intake*
  - Lithium exposure
- Hyperthyroidism
  - Autoimmune (Grave's Disease)
  - Toxic adenomas
  - Subacute thyroiditis
  - Pituitary gland malfunctions (TSH)
  - Cancerous growths in thyroid

Symptoms: Hypo vs. Hyper

- Hypothyroidism
  - Fatigue, sleepiness
  - Weight gain (decreased appetite)
  - Cold intolerance
  - Depression
  - Menstrual disturbances
  - Hair loss
  - Dry skin
- Hyperthyroidism
  - Nervousness
  - Anxiety
  - Increased perspiration
  - Heat intolerance
  - Hyperactivity
  - Palpitations
  - Weight loss

Ocular Manifestations

- Anterior segment
  - Evaporative DES
  - SLK (65% have thyroid dysfunction)*
  - Lid retraction (Dalrymple's sign)
  - Lid lag
  - Exophthalmos
- Posterior Segment
  - Optic nerve hypoplasia
  - Optic nerve swelling/compression
  - Chorioretinal striae
- Intra-Orbital
  - EOM restriction (IM SLO)
  - EOM enlargement
  - Optic nerve compression

Thyroid Eye Disease

- **N**o signs or symptoms
- **O**nly signs (limited to upper lid retraction and stare, with or without lid lag)
- **S**oft tissue involvement (conjunctiva, lids, etc.)
- **P**roptosis
- **E**xtraocular muscle involvement (diplopia)
- **C**orneal involvement (lagophthalmos)
- **S**ight loss (due to optic nerve involvement)

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Thyroid ED

- Autoimmune Disease
- Women > Men (3-10 times)
- Occurs in 4th – 5th decade of life
- When men are affected, symptoms are worse
- Ocular manifestation generally appear 2.5 years after onset of disease
- 25-50% Grave's dz patients develop ocular manifestations.
- Most common in Hyperthyroid, but can occur with hypothyroidism

*http://www.eyecalcs.com/DWAN/pages/v5/v5c021.html
** Yanoff & Duker Ophthlmology 3rd edition.
^Kendler DL, Lippa J, Bootman J. The initial clinical characteristics of Graves' orbitopathy vary with age and sex. Arch Ophthalmol. 1993;111:197–201.

Thyroid ED

- Symptoms are easy to expect, when process is understood.
  - Antibodies affect extraocular muscles, orbital fat, and the levator, causing swelling and immobility.
  - Swelling of EOM can cause optic nerve compression, and result in exophthalmos
  - Levator involvement results in lid retraction and lag
  - Antibodies can also affect lacrimal gland

Diagnosis/Testing

- Tonometry (primary gaze and up gaze)
- Exophthalmeter
- Appearance
- Thyroid panel/ Autoimmune markers
- Imaging (CT and MRI)
- Forced duction/motility

Thyroid Eye Disease

- Optic Nerve Compression
  - Visual field defects
  - Contrast abnormalities
  - Color vision defects
  - RAPD
  - Decreased Visual Acuity
  - Pale atrophic optic disc
  - Rare: Occurs in 10% *
  - 40-50% of patients with compression have normal appearing fundus^*.

Treatment options

- Self Limiting: Graves' disease usually runs a progressive course for 3–5years and then stabilizes.*
- Concern is patient comfort and treatment of Dry eye concerns.
- Lid weights/taping/tarsorraphy may be required to decrease exposure

Transient Vision Loss

- 78 yo F
- Noted 5-8 seconds of fluctuating vision
- Qualify vision loss

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Similar Case

78 yo Male with headache (of any shape or form)

- What three questions should come to mind?

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82 yowf Sudden Loss of VA

- Ocular history:
  - Primary open angle glaucoma OU
  - Epithelial basement membrane dystrophy OU
  - Pseudophakia OU
  - Early Dry ARMD OU
- Medical history:
  - Arthritis
  - Hypertension
  - High Cholesterol
  - Peripheral Neuropathy
  - Restless leg Syndrome
  - GERD

- Ocular Medication
  - Combigan BID OS
  - Travatan Z QHS OU

- Systemic Medication
  - Crestor 5mg
  - Amlodipine-Benazepril 5/10mg
  - Pramipexole 0.125mg
  - Tramadol HCL
  - Nexium 40mg
  - Lidoderm patch
  - Gabapentin 300mg
  - Celebrex 200mg
  - Iron supplement
  - Krill oil supplement

Case Example

- VAcc:
  - OD: LP
  - OS: 20/50 +2
- Pupils
  - OD: 1+ APD
  - OS: round and reactive
- EOM
  - Full OU
- CVF
  - OD: constricted inferior 180
  - OS: Full to finger counting
- IOP: 18mmHg/18mmHg by Goldmann

- Assessment
  - Ischemic Optic Neuropathy OD
    - Pt denied any jaw pain, headaches, shoulder or hip pain, change in weight and malaise
- Plan
  - Labs ordered: ESR, CRP, CBC w/diff
  - Medication: Prednisone 20mg 3 PO QD and Ranitidine 150mg BID PO
  - Meds are not to be started before having blood drawn
  - Follow up in 1 week pending lab results

- Lab Results:
  - ESR: 95 (High)
  - CRP: 7.09 (High)
  - Platelet: 465 (High)
- Temporal artery biopsy scheduled in 2 weeks

Giant Cell Arteritis

- Temporal Artery Biopsy Result
  - Active arteritis with rare giant cells, consistent with temporal arteritis
  - Mild arteriosclerosis
  - Disruption and focal loss of internal elastic lamina
- Informed the patient that her PCP will monitor her labs from now on and adjust her oral prednisone dose accordingly. She is to continue on the 60mg/day dosing for right now until he instructs her otherwise
- Follow up in 1 month

Giant Cell Arteritis

- The most common vasculitis in >50 years old
- Incidence increases with age, peaking in the 8th decade
- More common in females and in those of northern European descent/Scandinavians
- Large autopsy studies strongly suggest that the true prevalence of GCA may be ~1% of the population

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Giant Cell Arteritis

- Any elderly pt with headache
- Any elderly pt with diplopia
- Any elderly pt with transient vision loss

- Ask
  - Temporal tenderness
  - Pain with combing hair, wearing glasses, hats
  - Pain with chewing, swallowing

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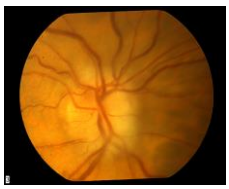
Giant Cell Arteritis

- Ischemic symptoms
  - Headache
  - Scalp pain
  - Jaw Claudication



- Systemic inflammation symptoms
  - Polymyalgia rheumatica
  - Fatigue
  - Malaise
  - Fever
  - Anorexia/weight loss
  - Night sweats

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GCA – Ocular Manifestations

- Anterior Ischemic Optic Neuropathy
  - Rapid, profound vision loss
  - (+) APD
  - A swollen optic nerve with a chalky white appearance
  - VF defect varies
- Central Retinal Artery Occlusion
  - ~ 10% of CRAO's are caused by GCA
- Cranial Nerve Palsy
  - Rare and usually CN 6

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Now what?

- Differentials for unilateral swollen nerves
  - Anterior Ischemic Optic Neuropathy
    - NAION
    - AAION
  - Optic Neuritis
  - CRVO
  - Compression (Mass/lesion, Thyroid)
  - Inflammation
  - Infection

GCA - Testing

- Blood Work
  - ESR, CRP, CBC w/diff
- TA Biopsy
- Simultaneous color Doppler and duplex ultrasonography

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AAION Treatment

- Standard of care: **Corticosteroids**
  - In acute cases of AAION (same day) patients should be started on IV methylprednisolone 250-1000mh x 3 days
    - Possible vision recovery, most importantly save the other eye (?)
  - Oral Prednisone 80-100mg/day if vision loss is not acute
- Referral to internist / rheumatology
  - Will taper steroids to the lowest required dose
  - Usually GCA patients require treatment for 2 or more years
  - Recurrences can happen

Oral Corticosteroid Considerations

- Accurate diagnosis is essential
- Indicated for acute inflammatory eye, orbital and eyelid conditions
- Pregnancy category C
- Dosepaks available
  - 24 mg, 30 mg, 60 mg with taper
- Best taken with meals
- Short term rarely has ocular side effects

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Ranitidine

- Histamine-2 blockers
- Works by reducing the amount of acid your stomach produces
- 150 mg BID po
- Generally well tolerated
- HA

- Side effects of long term steroids use:
  - Bone fractures
  - Infections
  - GI Bleeding/perfusion
  - Hypertension
  - Diabetes mellitus
- New Alternatives
  - Methotrexate
    - can be useful as steroid-sparing agents in patients who require prolonged treatment with high doses of steroids (more than 5-10 mg/d) and those who experience significant steroid-related complications

Viagra (sildenafil citrate)

- Selective inhibitor of phosphodiesterase type 5
- Impairment of color discrimination (B/G)
- Non-arteritic ischemic optic neuropathy

NAION

- Vascular insufficiency from small vessel disease
- There is insufficient perfusion of the optic nerve head which causes infarction of the prelaminar region
- Occurs usually between ages 40-60
- Loss of vision occurs upon wakening in majority of cases
- More common in Caucasians

- Risk factors:
  - Diabetes (#1)
  - Hypertension
  - Ischemic heart disease
  - Nocturnal hypotension
  - Hypotensive agents taken at night
  - Sleep Apnea
  - Smoking
  - Viagra or other ED medication
  - Recent surgery (cardiac or neck sx, any ocular surgery)

Can Anything be Done?

- Hayreh SS, Zimmerman MB. Non-arteritic anterior ischemic optic neuropathy: Role of systemic corticosteroid therapy. Graefes Archives of Clinical Exp Ophthalmology 2008; 246:1029-1046.
- Initial VA 20/70 or worse, treated within 2 weeks of onset of symptoms
  - Visual outcome at 6 mo
    - Treated eyes 70% improved
    - Untreated eyes 41%.
  - Visual Fields
    - Treated – 40.1% improvement
    - Untreated – 24.5% improvement

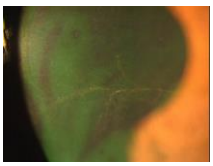
Remember!

- AAION
  - >60 years
  - VA loss is profound, often 20/200 or worse
  - Prodromal symptoms
  - Chalky white swollen disc
  - ONH c/d can be any size
  - Cup enlarges when edema resolves
  - FA shows patchy choroidal perfusion and late disc leakage
  - Risk of fellow eye involvement is high and can occur in days

- NAION
  - 40-60 years
  - VA can be as good or better than 20/60
  - Hyperemic swollen disc
  - Disc at risk
  - Pallor'd cup when edema resolves
  - FA will show delayed filling and late disc leakage
  - Fellow eye may occur years later

Cordarone (amiodorone)

- Indicated for the treatment of life-threatening recurrent ventricular arrhythmia
- Side Effects
  - Halos
  - Photosensitivity
  - Optic neuropathy
  - Optic neuritis
  - Disc swelling



Differentials for Vortex Keratopathy

- Drug induced
  - Amiodorone
  - Chloroquine
  - Tamoxifen
  - Ibuprofen
  - Indomethacin
- Stem cell deficiency
- Fabry's disease

Fabry Disease

- X-linked disorder due to a deficiency of alpha-galactosidase resulting in the buildup of globotriaosylceramide
- Signs and symptoms include:
  - Severe pain in the extremities
  - Exercise intolerance
  - Renal involvement
  - Skin lesions – angiokeratoma corporis discusum consists of clusters of superficial cutaneous dark-red angiokeratomas
  - Tortuosity of conjunctival and retinal vessels

THANK YOU

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