From Retina to Neuro

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Kelly Malloy: Nothing to disclose
Carlo Pelino: Has received honorarium from Carl Zeiss and Thrombogenics
The Eye is an extension of the Brain
Axons of ganglion cells become optic nerve, etc, etc.
• **DURA MATER**
  1. Periosteal layer (next to bone)
     - forms periorbitum
  2. Investing layer (inner)
     - forms optic nerve sheath

• **ARACHNOID MATER**

• **PIA MATER**

Then b v’s penetrate deep into the brain; $O_2$, gluc, etc

- Cerebrospinal Fluid
- Dura
- Arachnoid
- Subdural Space
- Subarachnoid Space
- Pia
- Brain
Dural Venous Sinuses

http://epomedicine.com
Anterior and Posterior Blood supply
Similarities between the eye and the brain

web.stanford.edu
Choroid plexus

Blood brain barrier (one part)

Blood retinal barrier (one part) same concept

Blood aqueous barrier (one part) same concept

Fenestrated Capillaries

Choroid Epithelium

Tight junctions

CSF in ventricle of brain

Brain/ neurons
CAUSES OF UNEXPLAINED VISION LOSS

• RETINA
  – Foveal ischemia
  – Macular Edema
    » CME
    » CSME
  – Macular Hole
  – Epiretinal membranes
  – Central serous retinopathy
  – Degenerative Myopia
  – Macular Degeneration

• NEURO
  – Functional Vision Loss
  – Ocular Ischemic Syndrome
  – Optic Neuropathy
    » Optic Neuritis
    » Ischemic Optic Neuropathy
  – Visual Pathway Damage
    » Stroke
    » Tumor
CAUSES OF OPTIC DISC EDEMA

• RETINA
  – Diabetic Papillopathy
  – Malignant hypertension
  – Neuro-retinitis
    » Toxo
    » Lyme
    » Sarcoid

• NEURO
  – Optic Neuritis (Papillitis)
  – Peri-Optic Neuritis
  – Papilledema
CAUSES OF VASCULAR SHEATHING

• RETINA
  – Vascular Occlusions
  – Sarcoid
  – Other systemic diseases

• NEURO
  – Multiple Sclerosis
  – Sarcoid
  – Other systemic diseases
CAUSES OF HEMES/CWS

• RETINA
  – Diabetic Retinopathy
  – Hypertensive Retinopathy
  – Retinal Vein Occlusions

• NEURO
  – Papilledema
  – Ischemic Optic Neuropathy
EXAM FINDINGS THAT DO NOT MATCH WITH PRESUMED RETINAL DX

– Large relative afferent pupillary defect
– Field that does not match retinal and/or optic disc appearance
– Visual acuity that does not match macular appearance
– Macular vs Optic Disc OCT findings
EXAM FINDINGS THAT DO NOT MATCH WITH PRESUMED NEURO DX

- Abnormal macular OCT
- Lack of relative afferent pupillary defect (if unilateral)
- Abnormal ERG
CASE 1
65 year old woman

- C/o red, painful eye x 3 days
- Pain is a “10”
- 3 days ago, episode of bilateral vision loss
  - Complete blackness
  - Lasted a few seconds
  - Associated dizziness

- Shortly after, OS became red
- Headache and left eye pain
- VA OS decreasing since then
65 year-old woman
• VA OD 20/40 and OS 20/60
• + red desat OS
• + decreased brightness sense OS
• + APD OS
• CF: inferior temporal constriction OS
• TA: OD 12  OS 10
• BP: 129/72
CAUSES OF SEGMENTAL DISC EDEMA

- Papilledema
- Optic Neuritis
- Disc Drusen
- Emboli/Ischemia
- NA-AION
- Neuro-Retinitis
- A-AION
• Systemic Hx:
  • HTN x 15 yrs – on Accupril
  • Hypercholesterolemia – no tx / allergies
  • + SOB / fatigue, had carotid US
    • Stenosis of left carotid artery
    • Saw vascular surgeon
    Ordered CTA of neck and cardiology consult
  Will F/U in one month
• CT- angio neck
• Report showed
  – Severe stenosis (90%) of left subclavian artery
  – Severe stenosis of mid-portion of left common carotid
    • Extends for 2.5 cm
Assessment / Plan

• Acute Ocular Ischemic Syndrome
• STAT hospital admission
• Carotid endarterectomy vs stent
• **DIAGNOSIS: Acute Ocular Ischemic Syndrome**
  
  • STAT hospital admission
  • High-risk for stroke – pt not allowed to move
  • Carotid endarterectomy was determined to be too risky
  • Patient underwent endovascular stenting
  • Patient did suffer a stroke during surgery
  • resulting in R hemiparesis and memory loss

• **FOLLOW-UP:**
  
  • Underwent motor and speech therapy
  • Recovered well
Statistics Regarding Ocular Ischemic Syndrome

- A 40% mortality rate has been reported in patients with OIS
- The most common symptom is slowly progressive vision loss, but 10% report sudden vision loss
- 40% present with pain
- 67-87% present with iris neovascularization
- 10-20% are asymptomatic at time of diagnosis
- 86% of patients are smokers
- Risk factors include diabetes, ischemic heart disease, cerebro-vascular disease, trauma, and vasculitis (need to R/O GCA)
Ocular/Visual Manifestations of Carotid Occlusion

- Transient vision loss (amarosis fugax)
- Red eye (episcleral injection)
- Mid-peripheral retinal hemorrhages
- Iris neovascularization
- Emboli (Hollenhorst plaques)
- Optic Disc Edema (Ischemic Optic Neuropathy)
- Retinal artery occlusion (CRAO / BRAO)
- Ophthalmic artery occlusion
- Retinal or disc neovascularization
- Venous stasis retinopathy
- Homonymous hemianopia
- Ocular motility problems (cranial nerve palsies / brainstem motility disorders)
- Supranuclear gaze abnormalities
Neurologic manifestations of Carotid Occlusion

- Carotid bruit
- Hemiplegia
- Hemianesthesia
- Homonymous Hemianopia
- Aphasia
- Headache
- Neglect
- Speech disturbances
NASCET


• NORTH AMERICAN SYMPTOMATIC CAROTID ENDARTERECTOMY TRIAL
SURGICAL TREATMENT (NASCET)

SYMPTOMATIC with STENOSIS > 70%
CAROTID ENDARTERECTOMY IS RECOMMENDED

SYMPTOMATIC with STENOSIS 50-70%
CAROTID ENDARTERECTOMY IS INDICATED
2.1% PERIOPERATIVE RISK OF STROKE AND DEATH

SURGICAL TX of ASYMPTOMATIC CAROTID STENOSIS IS CONTROVERSIAL
Fig. 4. Progression of carotid stenosis incurs an increasing risk of stroke until the point of occlusion, when the stroke risk drops precipitously.
SURGICAL TREATMENT

• PERICUTANEOUS TRANSLUMINAL ANGIOPLASTY
• INTERNAL CAROTID ARTERY STENT
• FOR PATIENTS UNABLE TO UNDERGO ENDARTERECTOMY

SALUS UNIVERSITY
Any TIA or Retinal Ischemia/Emboli Treated the Same

¼ of patients with acute retinal ischemia (even if transient) had an acute brain infarction on brain DWI-MRI

10-15% of patients will have a disabling stroke within 3 months after a TIA, with half occurring within 48 hours after resolution of TIA.
What needs to be done?

• DWI-MRI within 24-48 hours of vision loss
• Imaging (CTA) of cervical and intracranial vessels.
• EKG and echocardiogram
• Laboratory testing
  – CBC with platelets
  – Coagulation studies
  – Fasting lipid profile
How does this get done?

Do NOT send these patients to their PCP, cardiologist, neurologist, neuro-ophthalmologist, or retinal specialist.

Do NOT try to obtain the work-up yourself.

Send to an ED with an Acute Stroke Care Center!
ADDITIONAL CASES
Although CRAO is the ocular equivalent of an ischemic stroke, patients don’t consider it as urgent as they would a cerebral stroke; thus, they rarely seek emergency medical help.

Identify the etiology: The cause (emboli, thrombus, vasospasm, giant cell arteritis, etc.) of the occlusion may affect workup and management.

Treatment: Perform what is considered standard therapy (e.g., IOP drops, anterior chamber paracentesis, ocular massage) and to promptly refer the patient to an acute stroke center for a vascular evaluation because of their knowledge about the type of vascular workup needed.

Take home of CRAO: minimizing the risk of hemispheric stroke.

Evaluate for carotid artery and cardiac diseases, and should have a brain MRI.

Do not have a lack of urgency and wait for weeks to complete a workup.

The highest window of stroke risk is within the first week. The incident rate ratio for ischemic stroke peaks 1 to 7 days after CRAO (44.51; 95% CI, 27.07-73.20) and remains elevated for the first 30 days (14.0; 95% CI, 8.90-22.00).

CRAO: Harbinger of Ischemic Stroke
Written By: Annie Stuart, Contributing Writer, interviewing Andrew G. Lee, MD, Marc H. Levin, MD, PhD, and Neil R. Miller, MD
Include **noninvasive imaging of cervical vessels**, such as carotid ultrasound, to look for stenosis or compromise of the internal carotid artery

Undergo **electrocardiography** to check for an arrhythmia as soon as possible, and they should have an **echocardiogram to evaluate for cardiac ischemia**

CRAO if the occlusion is detected early, requires a brain **MRI with diffusion-weighted imaging within 24 hours**

CRAO and other types of sudden monocular vision loss—such as **branch retinal arterial occlusion** or **retinal transient ischemic attack**—increase the risk of silent brain infarctions.

One study found that approximately 90% of strokes following monocular visual loss were silent. Since these 3 conditions significantly increase the risk for subsequent clinical strokes, the researchers advised urgent and thorough evaluation on a stroke unit.
Diffusion-weighted magnetic resonance (MR) imaging provides image contrast that is different from that provided by conventional MR techniques. It is particularly sensitive for detection of acute ischemic stroke and differentiation of acute stroke from other processes that manifest with sudden neurologic deficits.

Diffusion-weighted MR imaging also provides adjunctive information for other cerebral diseases including neoplasms, intracranial infections, traumatic brain injury, and demyelinating processes.

Because stroke is common and in the differential diagnosis of most acute neurologic events, diffusion-weighted MR imaging should be considered an essential sequence, and its use in most brain MR studies is recommended.
CASE 2
37 year-old woman

- Complaint of blurry vision
- Episodes of darkening vision x few seconds
- Occasional flashes
- Feels like looking through a glaze
Systemic History

- DM x 14 years (known diabetic retinopathy)
- Hypercholesterolemia
- Iron deficiency anemia
- Proteinuria
- s/p MI
Medications

- Atenolol
- Plavix
- Diovan
- Bystolic
- 325 mg aspirin
- Insulin
- Metformin
- iron
• Social History
  – unremarkable
• Family History
  – DM, HTN
  – Glaucoma
  – Blindness (from DR)
Exam Findings

- BCVA: OD 20/40  OS 20/80
- Color: OD 13/14  OS 14/14
- PERRLA (0.6 log) RAPD OS
- CF: inferior nasal constriction OU, red desat OS
- No ptosis or proptosis
- Normal ocular motility
Exam Findings

• SLE: unremarkable, no NVI
• TA: OD 14 mm Hg  OS 16 mm Hg
• BP: 110/80
Other symptoms over the past year

- Nausea, vomiting, headaches, pulsatile tinnitus
- Saw a neurologist, had a CT – apparently normal
- She thinks her symptoms began after she gained a significant amount of weight, which she attributes to starting to use insulin
- Several months later, her headaches improved somewhat; then they worsened again
• Went to the ER due to dizziness; told of vertigo
• She said they wanted to do a LP, but instead checked her eye pressure, which was normal
• Later, she developed pain in her left arm and back
• Went to ER again, and told of an MI; she subsequently had 2 stents placed
• At that time, both her BS and cholesterol were very elevated
• As far as she knows, her BP has been okay
After released from the hospital, she had an episode where both eyes went black and she lost feeling in her legs.
• What does this patient have? What would you do with this patient?
CAUSES OF INDISTINCT OPTIC DISC MARGINS

- Papilledema
- Optic Neuritis
- Anomalous Disc
- Disc Drusen
- NA-AION
- A-AION
- Neuro-Retinitis
CAUSES OF MACULAR STAR

- Hypertensive retinopathy
- Neuro-retinitis
- Diabetic Retinopathy
- NA-AION
- Retinal Vein Occlusion
- Von Hippel Lindau
- Radiation Retinopathy
- Papilledema
Bilateral Diabetic Papillopathy AKA NA - AION
Hypertensive Retinopathy – grade 4
Modified Frisén Papilledema Scale

Grade 0

Grade 1

Grade 2

Grade 3

Grade 4

Grade 5
Papilledema With Hemorrhages

• Need to think of:
  – Acute increase in intracranial pressure
  – Severe chronic papilledema
• Does this all have to be explained by just one diagnosis?
20 year-old man

• Pain OD on upgaze x few days
• Today, vision OD is “off”
• Denies diplopia, transient vision loss
• Denies headache
• A few days ago, he felt feverish, but did not check his temperature
• Fam HX: Father dx with Lupus in 20s
• BCVA: OD 20/50 and OS 20/20
• Color 14/14 OD and 14/14 OS
• PERRLA (-) APD
• CF: full OU
• HVF: essentially normal OU
• SLE and IOP normal OU
• BP: 104/70
• Temp: 98.8 degrees
Initial Presentation

Right Eye

Left Eye
CAUSES OF SEGMENTAL DISC EDEMA

- Papilledema
- Optic Neuritis
- Emboli/Ischemia
- NA-AION
- A-AION
- Neuro-Retinitis
- Disc Drusen
- Pt denies any rashes (only when asked)
- He does admit to a scratch by a cat (kitten) several weeks ago (only when asked)
- A few weeks ago, his right eyelid was swollen
- Pt has several scars on his forehead, above right eye, and on his nose
Labs Ordered - told to have done today!

- CBC
- C-reactive protein
- ESR
- Platelet count
- Lyme titer (if + get Western Blot Lyme IgG and IgM)
- ANA with reflex titer
- ACE
- RPR
- FTA-ABS
- Bartonella Quintana titer
- Bartonella Henselae titer
Follow-up 5 days later

- Pt notes a spot in right vision, that is getting larger
- Reduced central vision
- Since last visit, has had chills and fever
- Has also had headache
- Decreased appetite
- Unable to work – doesn’t feel right
- Labs not done until 2 days ago – not complete
Lab results (so far)

- ANA ( + ) titer and pattern not yet known
- Lyme titer is ( + )  WB IgG (-), IgM (+)
- ACE slightly elevated at 70
- ESR: 44
- CRP: pending
- Bartonella titers: pending
• BCVA: OD 20/200 and OS 20/20
• Color 1/14 OD and 14/14 OS
• Pupils – trace RAPD OD
• CF: very large blindspot OD
• HVF: large blindspot OD – extending past fixation and superiorly out to 10 degrees
• SLE and IOP normal OU (-) cells
Humphrey Visual Field: 6 days after initial presentation. Note significant increase in blind spot in the right eye. The left field was unreliable due to patient fatigue.
Follow-Up: 6 days after initial presentation
Follow-Up: 6 days after initial presentation
• Need to r/o Lyme, sarcoid, auto-immune disease
• Ds DNA (-)
• Repeat ACE (-), CXR (-)
• LP (-) for Lyme, sarcoid
• Bartonella titers
• Bartonella Quintana (-)
• Bartonella Hensalea (+)
• (+) IgG > 1:2560
• (+) IgM > 1:800

• DX: Cat-scratch Disease
Follow-Up: 15 days after initial presentation
CAUSES OF MACULAR STAR

- HTN retinopathy
- Neuro-retinitis
- NA-AION
- DM Retinopathy
- RVO
- Papilledema
- Radiation Retinopathy
- Von Hippel Lindau
Follow-Up: 34 days after initial presentation
Optical Coherence Tomography (OCT) - 2 different sections through the right macular region. Note the OCT appearance of the retinal exudates (arrows).

Follow-Up: 34 days after initial presentation
Humphrey Visual Field: 34 days after initial presentation. Note reduction in blindspot size in the right eye corresponding with a reduction in optic disc edema.
Follow-Up: 34 days after initial presentation
• **Treatment:**
  – Antibiotics
    » Doxycycline (pt vomited every time he took this medication)
    » Rifampin
    » Bactrim prescribed in place of Doxycycline
    » Pt then switched to Azithromycin by Infectious Disease
Cat Scratch Disease

- **Treatment / Response:**
  - Excellent prognosis - Most cases are self-limiting and fully resolve, even when involving the CNS
  
  - Drugs of choice – Bactrim, Gentamicin, Ciprofloxacin, Rifampin, Azithromycin
Cat Scratch Disease

Typically transmitted by a kitten (by a scratch or a lick)

Only a minority of the exposures to B. Henselae result in cat-scratch disease. The ability of the cat to transmit the disease is transient

Most cases occur in fall / early summer - related to kitten births and flea infestations

80% of cases occur in patients under age 21

Starts with local infection, then lymphadenopathy, and rarely progresses – eg. Neuroretinitis, etc.
62 year-old woman

- History of glaucoma
  - Using Travatan and Azopt OU

- No eyecare x 1.5 months due to lack of insurance
  - PCP did refill drops during that time
• Chief complaint
  – Blur with prolonged reading
  – Rare headaches from lack of sleep

• Systemic History:
  – Diabetes
  – Hypertension
  – Hypercholesterolemia
  – Rheumatoid Arthritis
• BCVA: OD 20/20  OS 20/20
• Color: OD 12/14  OS 14/14
• PERRLA (-) RAPD
• Normal ocular motility
• SLE: only mild lens changes
• TA: OD 17 mm Hg  OS 18 mm Hg
• BP: 142/80
CAUSES OF COLLATERAL / OPTOCILIARY SHUNT VESSELS

End Stage Glaucoma

Congenital

Retinal Vein Occlusion

Sphenoid Wing Meningioma

Chronic Atrophic Papilledema

Optic Nerve Sheath Meningioma
• Pt is of Haitian descent
  – Came to US 6 years ago

• She recalls that in Haiti about 20 years ago
  – Decrease in vision in OS
  – Unsure of diagnosis, but remembers being told about some bleeding in the eye
OPTOCHOROIDAL (optociliary) SHUNTS

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

- Pre-existing channels
- Dilate in response to chronic obstruction of CRV
- Shunt venous flow to choroidal circulation

- Can be congenital (single) – RARE – only 5% (work-up)
- Acquired (usually multiple) in 95% - to edge of disc
ACQUIRED OPTOCHOROIDAL SHUNTS

• If the patient has no history or clinical findings of CHRONIC PAPILLEDEMA, CRVO, or GLAUCOMA, then we must rule-out a:

  – MENINGIOMA of the optic nerve or sphenoid wing
67 year-old woman

- Presents emergently due to a spot in her vision for 6 months
- She initially thought it was a smudge on her glasses, but then realized it didn’t go away when she removed her glasses
- The spot is stable and stationary
• Systemic history
  – Hypertension
  – Hypercholesterolemia
  – “borderline” (untreated) diabetes
  – Osteoarthritis
  – Denies trauma
• BCVA: OD 20/30  OS 20/25+
• Color: OD 14/14  OS 14/14
• PERRLA (+ 1.5-1.8 log) RAPD OD
• CF: Constricted inferiorly OD, Full OS
• Exophthalmometry: OD 22 mm, OS 21 mm
• Palpebral apertures: OD 5 mm , OS 5 mm
• SLE: mild upper lid edema OU
• TA: OD 16 mm Hg  OS 16 mm Hg
• BP: 147/85
Ocular Motility

OD

75

95

100

OS

100

100

90

6 eso

8 LH

1 eso

6 eso

2 RH

4 eso

6 eso
Right Optic Nerve Sheath Meningioma
• Optochoroidal shunt vessels are not necessarily always present with optic nerve sheath meningiomas.

• They need to be in the differential for any optic neuropathy, and certainly with the presence of any optic disc shunt vessels.
CASE 5
76 year-old woman

- c/o blurry vision OU x 3 years
- Having difficulty reading
- Occasional vertical diplopia when watching tv
- Eyes teary and look bulgy
• Diabetes x 7 years
• Hypertension x 7 years
• Arthritis
• Hypercholesterolemia
• s/p Congestive heart failure
• + pacemaker
• Currently being tested for kidney and lung issues
• Lupus x 40 years
• Lupus – diagnosed at about age 30
• Was medicated on (hydroxy)chloroquine 250 mg daily for years
• About 3-4 years ago, diagnosed with bulls-eye maculopathy, and d/c (hydroxy)chloroquine
• Put on a different med for Lupus
• Followed by Rheumatology
- s/p appendectomy
- s/p cataract surgery
- s/p fractured back due to fall 1.5 years ago

- MEDS:
  - Furosemide, gabapentin, lisinopril, vitamin D3, tramadol HCl, aspirin, alendronate sodium
- BCVA: OD 20/50- and OS 20/50-
- Color 0/14 OD and 0/14 OS
- PERRL (-)RAPD, (+)LND OU
- CF: superior central defects OU, superior arcuate OS
• Apertures: 11mm OD, 12 mm OS
• Lid crease 7 mm OD, 7 mm OS
• Levator function: 19 mm OD, 21 mm OS
• Exophthalmometry: 22 mm OD, 24 mm OS
• Ductions: 95% normal abduction and supraduction OU
• SLE: pseudophakic OU
• GAT: 10 mm Hg OD 11 mm Hg OS
• BP: 180/68
• DFE:
CAUSES OF BULL’S EYE MACULOPATHY

- Chloroquine / Hydroxychloroquine
- Tapetoretinal degenerations
- Cone / Rod dystrophy
- Batten's disease
- Progressive Cone Dystrophy
- Stargardt's disease
- Benign concentric macular dystrophy

BULL’S EYE MACULOPATHY
• What would you expect the visual field to look like?
• Is any of this vision loss or visual field loss due to vitreomacular traction (VMT)?

• May there also be an optic neuropathy? Do we need to do a work-up for optic neuropathy?
• Patient was lost to follow-up, and returned a few years later.
• If this is Plaquinil maculopathy alone, would you expect there to be progression?
• By this time, fundus examination showed retinal pigment changes suggestive of tapetoretinal degeneration (Plaquenil toxicity)
HYDROXYCHLOROQUINE TOXICITY
Hydroxychloroquine Toxicity

IR 30° ART + OCT 30° (8.9 mm) ART (23) Q: 28 [HS]

http://webeye.ophth.uiowa.edu/
PLAQUENIL

Hydroxychloroquine (Plaquinil) - Anti-malarial

• Ophthalmic side effects (infrequent with current dosing ranges):
  – Irreversible retinal damage has been observed ("chloroquine retinopathy").
  – If there are any indications of abnormality in the color vision, visual acuity, visual field, or retinal macular areas, or any visual symptoms (eg, light flashes or streaks), confer with prescribing practitioner (likely d/c drug stat)
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.
PATTERN OF RETINOPATHY

• Although the locus of toxic damage is parafoveal in many eyes, Asian patients often show an extramacular pattern of damage.
DOSE

• 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
  – concomitant renal disease
  – 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.
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