# ***Retinal Urgencies and Emergencies***

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I do not have any personal relevant financial or commercial interest to disclose. My husband has a salaried position at Heidelberg Engineering as a Clinical Development Manager. He works with researchers and did not have any direct or indirect influence on this lecture.

**Course Description/Summary:**
The primary care eye physician must have a broad range of knowledge and skills to diagnose and manage retinal disorders and to be able to distinguish which ones require immediate treatment/referral versus observation. Through this course, the primary eye care physician will be able to identify various retinal disorders, distinguish which ones require immediate management, and how to provide the appropriate follow-up and testing needed for retinal urgent/emergent cases.

**Learning Objectives:**
1. To learn how to identify retinal urgencies or emergencies through a patient's presenting signs and symptoms and how to confirm these with the appropriate clinical examination.
2. To understand how rapidly an urgent or emergent patient should be treated or referred.
3. To understand and recognize the possible complications of retinal urgencies/emergencies and how to appropriately follow-up with these patients.
4. To learn the systemic complications of retinal urgencies/emergencies and the appropriate systemic work-up recommended.

**Course Outline**

I. **What constitutes a retinal urgency or emergency?**
- A. Loss of vision or risk of loss of vision
- B. Changes in vision
- C. Pain
- D. Patient perceived anxiety or stress
- E. Doctor's comfort zone, level of expertise, or experience

II. **Posterior Vitreous Detachment**
- A. Symptoms: flashes, floaters
- B. Signs: Weiss ring, anterior vitreous pigment ("tobacco dust")-assume retinal tear, retinal hemorrhage (torn blood vessel)
- C. Examination: DFEX w/scleral depression, wide field imaging, biomicroscopy to assess for tobacco dust and Weiss ring
- D. Management:
  1. Retinal tear detected: Immediate referral to retinal specialist for laser or cryopexy (depending on location)
  2. Hemorrhage, no tear: Follow in 1 week; signs/symptoms of retinal detachment (R.D.) given to patient
  3. PVD, no hemorrhage, no tear: Follow in 1 month; signs and symptoms of R.D.

III. **Retinal Tear:** PVD, high myopia, ocular trauma, previous ocular surgery, increased age
- A. Symptoms: photopsias, new floaters, shadows or veils in vision
- B. Signs: Visible tear; rule out surrounding retinal detachment
- C. Examination: DFEX w/scleral depression, widefield imaging
- D. Management:
  1. Refer immediately to retinal specialist for laser or cryopexy especially if tear is superior
  2. R.D. signs and symptoms for possible future occurrences, as patient is at risk. Continue to monitor after repair.
  3. Counsel patient to avoid sudden head movements or jarring, as adequate scarring takes 1 week after tear repair.
- E. Follow-up after retinal tear repair:
  1. Encourage protective eyewear especially when playing sports, working in the yard, DYI projects, etc.
  2. If phakic, monitor for rapid cataract formation and myopic shift.

IV. **Retinal Detachment:**
- A. Symptoms: photopsias, new floaters, veils, shadows, curtain sensation, no pain
- B. Signs: Retinal partially or fully detached, macula on or off, location of R.D. matters
- C. Examination: DFEX w/scleral depression, widefield imaging, macular OCT
- D. Management:
  1. Macula-on R.D.: Risk of R.D. converting from macula-on to macula-off R.D. within 24 hrs is low (0.5-0.11%)
     a. Superior R.D.: Refer immediately for surgery. **TRUE EMERGENCY!**
     b. Inferior, nasal, or temporal R.D.: Refer immediately or within 1 day.
     c. Bedrest/Positioning: superior tear-supine; temp.-lay on temp. side of affected eye; nasal-nasal side; inf.-sit upright
  2. Macula-off R.D.: immediate repair vs. waiting

E. Follow-up:
1. Repeat DFEX, OCT-macula
2. If phakic patient, cataract will quickly form causing myopic shift. If SBP, axial length will change causing myopic shift.
3. Caution patient on the signs and symptoms of retinal detachment
4. Recommend protective eyewear.
5. Assess for muscle imbalance if SBP performed

V. **Macular Degeneration (Exudative or Wet ARMD)**
A. Symptoms: new metamorphopsia, scotoma, or decreased central vision (usually moderate VA loss) occurring over days/wks
B. Signs: CNVM; serous detachment; macular blood or fluid; elevated drusen, RPE, or PED
C. Examination: DFEX, OCT, OCT-A, FA, fundus photos
D. Management:
1. If suspect CNVM, refer immediately to retinal specialist within 1-3 days for OCT, OCT-A, FA, anti-VEGF therapy
2. Home Amsler's grid
3. ForseeHome AMD Monitoring System (Notal Vision): to monitor fellow eye for conversion to wet ARMD
4. Dark Adaptation Testing: to help stage fellow eye
5. Nutritional supplementation
6. Lifestyle counseling
7. High powered adds or low vision devices

VI. **Submacular Hemorrhage:**
A. Symptoms: sudden decreased vision (20/200 to light perception), metamorphopsia
B. Signs: blood collection between neurosensory retina & RPE. Important to distinguish: subretinal blood vs. sub-RPE blood
1. Sub-retinal blood: bright red, more harmful to photoreceptors (early photoreceptor damage wi/24 hrs)
a. iron-related toxicity to photoreceptors
b. oxygen and nutrient diffusion impairment
c. clot contraction causes mechanical damage to photoreceptors
2. Sub-RPE blood: darker
C. Cause: Valsalva, trauma, pathological myopia, ARMD, ocular histopl., polypoidal choroidal vasculopathy, macroaneurysm
D. Examination: DFEX, OCT, OCT-A, FA, fundus photos, widefield imaging
E. Management: intravitreal or subretinal injection of tPA (recombinant plasminogen activator) w/pneumatic displacement (gas bubble); anti-VEGF intravitreal injections w/ or wo/ CNVM

VII. **Vascular Occlusions:**
A. **Branch retinal vein occlusion:**
1. Symptoms: sudden, painless VA loss (macular edema); asymptomatic (no mac. edema); floaters (vit. heme-neovasc.)
2. Signs: localized flame-shaped retinal hemorrhage, CWS's; mac. edema (5-15% wi/1 yr., < 50% spontaneously resolve); Possible mac. ischemia; thickened/tortuous B.V.'s; most BRVO occur at AV crossing (common adventitial sheath)
3. Risk Factors:
a. Older Patients-uncontrolled hypertension, obesity, cardiovascular disease, glaucoma
b. Younger patients-inherited abnormal tendency to develop blood clotting
4. Diagnostic Testing: DFEX, OCT, OCT-A (mac/extramac. fluid), photos, FA (venous stasis, edema, ischemia, neovasc.)
5. Treatment:
a. Systemic: Identify/treat underlying risk factors w/PCP-monitor BP, cholesterol/lipid levels, abnormal blood clotting
b. Eye:
1) No macular edema/neovasc.: follow w/ DFEX's, OCT
2) Macular edema present:
a) Anti-VEGF intravitreal injections (lessens leakage, stops neovascularization); used alone or w/laser
b) Laser Treatment (hard to treat cases: light grid laser, 2/3 of pts. show improvement after 3 yrs of f/u
c) Intraocular Steroids (eyes non-responsive to anti-VEGF therapy): Ozurdex (dexamethasone implant), triamcinolone (intraocular injection)-increased risk of cataract, elevated IOP
6. Prognosis: Good; > 60% (treated/untreated) maintain >20/40 after 1 year
a. Presenting VA influences prognosis
b. Macular/foveal extent correlates w/prognosis
7. Complications: Neovascularization (not common)-treat w/scatter laser, vit. heme, R.D., glaucoma (2% iris neovasc.)

B. **Central Retinal Vein Occlusion:**
1. Symptoms: Blurred VA; metamorphopsia (M.E.); unilateral, TVO's, floaters (vit. heme), pain/redness (neovasc. glauc.)
2. Signs: Flame-shaped hemorrhages in all quadrants; CWS's; macular edema (common); vitreous hemorrhage, neovasc.
3. Risk Factors: Diabetes, hypertension, hypercoagulable states (bilateral CRVO or younger patient)
4. Diagnostic Testing: DFEX, OCT, OCT-A, photos, FA; PCP-r/o DM, HTN; hematologist-r/o hypercoagulable state
5. Treatment: anti-VEGF for edema/neovascularization, intraocular steroids (triamcinolone-injection, Ozurdex-implant)
6. Prognosis: Better-younger person; Older person wo/treatment: 1/3 improve wo/treatment, 1/3 no change, 1/3 worse
a. Non-ischemic CRVO: milder type of CRVO. Leaky retinal vessels w/macular edema
b. Ischemic: more severe type of CRVO. Poorer VA and prognosis. Higher risk of neovascularization, NVG, vit. heme. Treatment: anti-VEGF (temporary), laser (more permanent solution), combination (anti-VEGF & laser)
7. Follow-up: Usually monthly (non-ischemic can become ischemic). Look for neovascularization of iris or angle. Earlier neovascularization and/or macular edema detection-better VA prognosis.

C. **Retinal Artery Occlusion:** embolus or thrombus
1. Types: Central Retinal Artery Occlusion (CRAO) and Branch Retinal Artery Occlusion (BRAO)
2. Symptoms: CRAO-sudden VA loss (25% have cilioretinal artery); BRAO-sectoral VF loss or no symptoms
3. Signs: CRAO-"cherry red spot" in center of macula; BRAO-sectoral retinal whitening along blocked vessel

4. Risk Factors: Carotid artery disease, atherosclerosis, valvular heart disease, cardiac tumor, arrhythmia, diabetes, Hypertension, IV drug use, GCA, blood clotting diseases (sickle cell), oral contraceptives, homocystinuria, pregnancy, Blood platelet abnormalities. Males>females. Only 1-2% bilateral.
5. Diagnostic testing: DFEX, OCT, OCT-A, FA
6. Treatment: No proven treatment for CRAO-hyperventilation, paracentesis, medically lowering IOP, ocular massage. Artery occlusions-theoretical emergency. If treatment is going to be effective, needs to be within 4-6 hrs of onset.
7. Work-up for acute CRAO/BRAO: Refer immediately to ER or PCP to evaluate for CVA risk (risk factors for Retinal Artery Occlusion are the same for CVA/cardiac disease: Carotid Doppler, Cardiac Echo, GCA w/u.
8. Prognosis:
   a. CRAO: severe, permanent VA loss (if cilioretinal artery present, >80% recover $\geq$ 20/50).
   b. BRAO: Permanent VF loss in area of occlusion; 80% recover to $\geq$ 20/40.
9. Complications: Neovascularization w/2ndary vitreous hemorrhage (rare), neovascular glaucoma (rare). Treatment: Laser, anti-VEGF therapy.

**VIII. Anterior Uveitis/Posterior Uveitis**

A. **Signs and Symptoms:**
   1. **Anterior Uveitis** (accounts for 50-60% of uveitis**):** pain, redness, photophobia, decreased VA
   2. **Posterior Uveitis**: Decreased VA, Floaters, Scotoma

B. **Uveitic Emergencies:** Risk of optic nerve/macular damage and permanent VA loss without immediate treatment
   1. Acute Retinal Necrosis (ARN): optic neuropathy, vascular occlusion, retinal detachment
   2. Progressive Outer Retinal Necrosis (PORN): vascular occlusion, retinal detachment
   3. Cytomegaolvirus (CMV) Retinitis: maculopathy, optic neuropathy, retinal detachment
   4. Ocular Toxoplasmosis: macular retinochoroidal scarring, macular edema, ERM, CNVM, RD, glaucoma
   5. Behcet's Disease (BD): maculopathy, optic nerve atrophy, cataract, glaucoma, retinal ischemia, RD, phthisis bulbi
   6. Vogt-Koyanani-Harada (VKH) Disease: Exudative RD, maculopathy, subretinal neovascular membrane, fibrosis
   7. Sympathetic Ophthalmia: Exudative retinal detachment, cataract, glaucoma, maculopathy

C. **Uveitis Evaluation:**
   1. **Patient History:** age, sex, general health (immunocompetent vs. immunocompromised), PMHx, surgical hx, trauma
      a. Immunocompetent: young male (ARN, Bechet's, Toxoplasmosis, VKH); young female (ARN, VKH, toxoplasmosis)
      b. Immunocompromised (HIV or organ transplant patient): CMV retinitis, PORN
   2. **Diagnosis:** usually made clinically (SLEX, DFEX, OCT, OCT-A, FA), confirmed through laboratory evaluation
      a. Vitreous opacity present: peripheral retinal infiltrate-ARN; central retinal infiltrate-Bechet's; exud. R.D.-VKH, S.O.
      b. Vitreous opacity absent: central retinal infiltrate-CMV retinitis, PORN, toxoplas.; periph. Ret. infiltrate-toxoplas.
      c. Retinitis (location), exudative retinal detachment, retinal scarring.
   3. **Basic Uveitis Work-up:** CBC w/diff., liver function, renal profile, RPR, tuberculin skin test, chest x-ray, urinalysis
   4. **Treatment:** topical, sub-tenon's, oral, IV
   5. **Prognosis:** disease dependent, treatment expediency, patient's immune system

**IX. Retained Lens Fragments:** Occurs following cataract surgery

A. Signs and symptoms: inflammation, decreased VA, high IOP, pain
B. Management (see immediately): control IOP (no prostaglandins), Diamox, topicals, keep corneal clear, post-up meds, surgery if IOP/cornea not controlled.

**X. Endophthalmitis:** True Medical Emergency

A. Exogenous: intraocular surgery (cataract, trabeculectomy, glaucoma tube, intraocular injections, etc.), usually bacterial
B. Endogenous: rare, severely immune-compromised patient
C. Signs/symptoms: decreased vision, pain, redness, hypopyon
D. Management: Refer ***immediately*** to retinal specialist (not ER). Hours counts. Blindness within 24 hours

**XII. Ocular Trauma-Globe Rupture:** 3 per 100,000 in US; ***SURGICAL EMERGENCY***

A. Symptoms: PAIN, greatly decreased vision, diplopia, acute rise in IOP
B. Signs: teardrop pupil, prolapsed iris, hyphema, severe subconjunctival. hemorrhage, shallow or very deep AC, limited EOM's, extrusion of globe contents. Rupture is in location where eye is weakest:
   1) No h/o prior ocular surgery: rupture most commonly posterior to extraocular muscles where sclera is weakest
   2) H/o intraocular surgery: rupture is often at prior incision site
   3) Also commonly ruptures at limbus w/blunt trauma
C. Diagnosis: history, clinical exam (VA, pupils, slit lamp-R/o penetrating FB, scleral/corneal lacerations, uveal prolapse, iris abnormalities, Seidel sign (contraindicated w/obvious globe rupture), DFEX, maxillofacial CT scan (no MRI), ultrasound (?)
D. Epidemiology/Demographics: Males > females
   1) Most commonly in males <40 yrs of age
   2) Males >40 yrs of age causes include assault, workplace injuries
   3) >75 yrs of age: ground level falls
   4) Globe rupture: can occur at any location
   5) Indirect trauma: rupture more common at nasal-superior quadrant close to limbus
   6) Substance abuse: associated with higher rate of ocular trauma
E. Causes
   1. Penetrating trauma/perforating injury (entrance/exit wound): glass, metal, shotgun pellets, BB pellets, wood shavings (grinding)
   2. Blunt trauma: mechanical falls, syncope, seizures, MVA w/airbag, assault w/blunt object
   3. Children: sharp objects (scissors): injury usually occurs at home
   4. Adults: workplace injuries, assaults, MVA

5. Elderly: ground-level falls
6. Other causes: gunshot, sporting injuries, stab wounds, blast wounds

F. Management: do not patch, place a shield, keep patient upright, send to ER immediately for CT to R/O intraocular foreign body, tetanus shot
1. Patient should remain NPO
2. Place a shield (do not patch)
3. Keep patient upright, bedrest, pain control, antiemetics
4. Initially, no tonometry, no lid retraction, no ultrasound
5. Pre-op sterile antibiotics to decrease risk of endophthalmitis
6. ***MANAGEMENT OF GLOBE RUPTURE IS A SURGICAL EMERGENY UNTIL PROVEN OTHERWISE***: Decreased risk for endophthalmitis if surgery within 24 hours

G. Differential Diagnosis: subconjunctival. hemorrhage, orbital/wall fracture, corneal abrasion, orbital hemorrhage, corneal ulcer, glaucoma, traumatic iritis

H. Prognosis: The most important factor is presenting VA

I. Complications: permanent blindness, endophthalmitis, chronic pain. Risk for Sympathetic Ophthalmia.

J. Deterrence and Patient Education: proper eye protection, avoid harmful lifestyle choices, PKP risk