Course Description:
Using case presentations, this course will provide the most up-to-date information on retinal conditions such as polypoidal choroidal vasculopathy (PCV), angioid streaks, macular telangiectasia, optic disc pit maculopathy, retinal arterial microaneurysm, and plaquenil maculopathy.

Goal:
Provide attendees with recent developments in the early diagnostic strategies and therapeutic advances each retinal disorder and discuss integration of these innovations into clinical practice.

Learning Objectives:
At the conclusion of this presentation the participant should be able to:

1. Know the variable clinical findings and presentations of the entities presented.
2. Appreciate the latest technologies in early diagnosis of each condition, including multi-modal imaging with SD-OCT, OCT-A, FA, IGCA, and fundus autofluorescence (FAF), and ultrawide field imaging.
3. Understand the current therapeutic strategies for each retinal disorder.
4. Discuss the latest in the management of each clinical entity.

Abstract:
Retinal diseases are on the rise. In some cases, clinical findings can lead be a diagnostic dilemma for many practitioners. Nonetheless, proper identification is important. The clinical findings of each entity and current treatment options will be reviewed in this presentation.
Retina Grand Rounds  
Sherrol A. Reynolds OD, FAAO  
Beata Lewandowska, OD, DABO  

NSU College of Optometry

Course Outline

I. PCV: Polypoidal choroidal vasculopathy (PCV)
   - Multiple recurrent serosanguineous bilateral retinal pigment epithelial detachments (PEDs). Known as posterior uveal bleeding syndrome
   - Clinical presentation(s)
     - #1 misdiagnosis is AMD: Variant of Type I CNVM
       - Predilection for pigmented races. Highest in Black women and Asian
       - Average age is 50’s-70’s
       - Predilection for juxtapapillary or macula
       - Commonly bilateral but can be UNILATERAL
   - Diagnostic tests
     - FA appearance may look like an occult CNV
     - ICG- Abnormal branching choroidal vascular channels with associated dilated aneurysmal polyps.
     - SD-OCT findings
       - Bolus sing: small elevation associated with the polyp
       - Double layer sign-dual reflective layers
       - Serosanguineous neurosensory and/or RPE detachment with high slopes
     - OCTA- visualization of branching vascular network (BVN)
       - Sea-fan or medusa
   - Management
     - Unpredictable natural history:
       - Spontaneous regression or recurrent with vision loss
     - Combination PDT/ Avastin therapy (EVEREST study)

II. Angioid streaks
   - Crack-like dehiscences of Bruch membrane
     - with deposition of calcium, magnesium, or iron salts due to disturbed metabolism
   - Case presentation (s)
   - Differential diagnoses
     - Lacquer cracks
     - Choroidal rupture
   - Choroidal neovascular membrane (CNVM)
   - Diagnostic tests
     - SD-OCT- discontinuity in Bruch's membrane
     - OCTA- CNVM
o Fundus autofluorescence (FAF)
o Fluorescein angiography (FA)

• Management
  o Determine Systemic causes
    ▪ Pseudoxanthoma elasticum (PXE)
    ▪ Ehler-Danlos syndrome (EDS)
    ▪ Paget's disease of bone
    ▪ Sickle cell disease and other hemoglobinopathies
    ▪ Idiopathic
  o AVT for CNMV

III. Macular telangiectasia
• Macular telangiectasia (MacTel) type 1 or 2
• Case Presentation(s)
  o MacTel 1: Aneurysmal telangiectasia
    ▪ Unilateral
    ▪ 90% male
    ▪ Lipid exudate
  o MacTel 2: Bilateral - MOST COMMON
  o Non- proliferative: Perifoveal telangiectasia
  o Proliferative: CNMV

• Diagnostic tests
  o SD-OCT: ILM draping
  o OCTA: CNVM
  o Fluorescein angiography (FA)

• Management:
  o Observation: Typically for type II non-proliferative
  o AVT
  o Consider testing for HTN and DM in patients with parafoveal hemorrhaging. If these diseases are not present, then telangiectasia is the likely cause

IV. Optic disc pit maculopathy (ODP-M)
• Optic disk pit (ODP) is a rare congenital abnormality
• Case presentation (s)
  o Symptoms and clinical signs
  o Serous macular detachment
  o Differential diagnoses
    ▪ Central serous retinopathy
• Diagnostic tests
• Management
  o oral corticosteroids
  o laser photocoagulation at the temporal disc margin
  o intravitreal gas injection
  o macular buckling surgery
  o pars plana vitrectomy (PPV) and a creation of inner retinal fenestrations
V. Retinal arterial macroaneurysm (RAM)
- Typically associated with Hypertension or atherosclerosis
  - Quiescent
  - Hemorrhagic
  - Exudative
- Case Presentation
- FA/ICG appearance
  - FA shows weaken of the vasculature at site of macroaneurysms = associated with hyperfluorescence
- OCT (A)
  - Monitor associated retinal edema from RAM and to determine the presence and extent of subretinal fluid, macular edema and hemorrhage.

Management
- Observation
  - Unpredictable natural history: Spontaneous regression or recurrent with vision loss
- Argon laser photocoagulation
- Anti-VEGF

VI. Plaquenil Toxicity
- Case presentations
- Latest Plaquenil Macular Toxicity Update- AAO 2016
  High risk factors:
  - Duration of use > 5 years
  - Dosage >5.0 mg/kg (real weight)
  - Renal disease
  - Retinal or Macular disease
  - Tamoxifen use
Lesser Risk Factors
  - Age
  - Liver disease.
  - Genetic factors.
- Screening tests:
  - Visual field- White 10-2
    - 24-2 or 30-2 VF for Asian patients because HCQ toxicity often manifests changes beyond the macula
  - SD-OCT
    - Perifoveal thinning
    - Loss of Inner-/Outer segment line
    - “Flying Saucer Sign”
  - Fundus Autofluorescence (FAF)
    - Increased (Hyper) autofluorescence- early damage
    - May precede thinning on SD-OCT
    - Reduced (Hypo) autofluorescence-Late RPE loss appears as a dark area Plaquenil related toxicity
  - Management:
- Baseline: before or during the 1st year of medication initiation
- Monitoring: Non-high-risk patients: exam perform at 5yrs and annually thereafter
- High risk patients: annually after baseline

References