1. Retinopathy of Prematurity (ROP)
   A. Stages of ROP
      Stage 1        Demarcation line
      Stage 2        Ridge
      Stage 3        Ridge with extraretinal fibrovascular proliferation (IVF)
      Stage 4        Subtotal retinal detachment
      Stage 5        Total retinal detachment
      Plus disease   Added when vascular shunting is so marked that the veins are enlarged and the arteries are tortuous in the posterior pole

   B. Type 1 ROP (New Threshold) Administer Peripheral Ablation Treatment
      Zone II
      Plus disease with stage 2 or 3

      Zone I
      Plus disease with stage 1, 2, or 3

      Stage 3 without plus disease
         - IVF
      Treatment - laser, Avastin

   C. Incontinentia Pigmenti
      1. X-linked dominant inheritance
      2. Fundus findings
         a. Peripheral avascular retina
         b. Absence of normal foveal avascular zone
         c. Retinal folds
         d. Retinal detachment
         e. Retinal dysplasia
         f. Cortical blindness due to occipital lobe infarction
      3. Systemic Findings
         a. Incomplete dentition
         b. Vesicular skin eruptions (infancy)
         c. Alopecia at the vertex of the skull
      4. Candidate gene for inherited IP: NEMO-NF kappaB essential modulator
         a. IVF for asymptomatic family members
D. X-Linked Retinoschisis-Gene localized to distal short arm of X-Chromosome (Xp.22.1-p.22.3)
1. Nerve fiber layer dehiscence
2. Stellate maculopathy No stain on IVF, OCT shows cysts
3. Vitreous hemorrhage **B scan**
4. Retinal detachment **B scan**

E. . FEVR
1. Clinical features
   a. Retinal fold with peripheral fibrovascular mass
   b. Non-rhegmatogenous retinal detachment simulating Stage 5 retinopathy of prematurity (ROP)
   c. Termination of peripheral vasculature with fimbriated border (IVF)
   d. Retinal exudation
   e. Dragging of the retina usually temporally
   f. Rhegmatogenous retinal detachment (RD) (rare) (**OCT & B scan**)
2. Inheritance
   a. Autosomal dominant
   b. Gene locus – 11 q 13-23; no candidate gene yet identified (Frizzled-4-gene)
   c. X-linked – allelic with Norrie’s disease
   d. Autosomal recessive – 2 families

F. Coats Disease
1. Clinical features
   a. Most common in males (8-10% in females)
   b. Age of onset: 8-10 years most common, but may present in infancy
   c. Unilateral in 80-90% of patients
   d. When bilateral, it is often asymmetric with one eye only mildly affected
   e. Not hereditary, no familial or associated systemic disease
   f. No known etiology
2. Ocular findings
   a. Retinal telangiectatic “lightbulb” lesions usually in the fundus periphery with capillary dropout demonstrable with fluorescein angiography
   b. Exudation with a predilection for the posterior pole. Cholesterol clefts are noted histologically
   c. Non-rhegmatogenous retinal detachment
   d. Cataract
   e. Neovascular glaucoma
   f. Phthisis bulbi
3. Diagnostic tests
   a. Examination under general anesthesia, if necessary
   b. Fluorescein angiography
   c. Ultrasound
   d. CAT scan (CT scan) – important to delineate calcium to aid in ruling out retinoblastoma
REFERENCES


