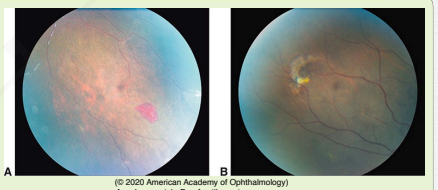
Sickle Cell Retinopathy

Introduction
- hemoglobin
  - two alpha-globin subunits
  - two beta-globin subunits
- *HBB* gene mutation — 11p15.4
  - abnormal versions of beta-globin → causes sickle cell hemoglobinopathy
  - unusually low levels of beta-globin → causes beta thalassemia
- mutant hemoglobins S, C, or both are of greatest ocular importance
- most prevalent in the black population
  - sickle cell trait (Hb AS) 8%
  - sickle cell disease (Hb SS) 0.4%
  - hemoglobin SC disease 0.2%

Nonproliferative Sickle Cell Retinopathy
- pathogenesis
  - arteriolar and capillary occlusion
  - anastomosis and remodeling occur in the periphery
- salmon-patch hemorrhage
  - area of intraretinal hemorrhage after a peripheral retinal arteriolar occlusion



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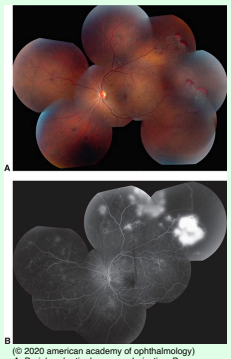
A, salmon patch. B, refractile spots.

- refractile spots — old, resorbed hemorrhages with hemosiderin deposition within the inner retina just beneath ILM
- black "sunburst" lesions — localized areas of retinal pigment epithelial hypertrophy, hyperplasia, and pigment migration in the peripheral retina
  - caused by hemorrhage
- occlusion of parafoveal capillaries and arterioles
- spontaneous occlusion of the central retinal artery

proliferative sickle cell retinopathy (PSR)
- Introcuction
  - retinal ischemia — secondary to infarction of the retinal tissue by means of arteriolar, precapillary arteriolar, capillary, or venular occlusions
  - sickle cell hemoglobin C (SC) and sickle cell thalassemia (SThal) — more serious ocular complications
  - SS disease — more systemic complications
- clinical findings
  - retinal neovascularization
    - located more peripherally
    - neovascularization in PDR generally begins postequatorially



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A, Peripheral retinal neovascularization. B, fluorescein angiogram shows leakage from the sea fan lesions in the periphery and peripheral nonperfusion

  - white sea fan neovascularization — results from autoinfarction of the peripheral neovascularization — auto infarction not common in PDR
  - preretinal or vitreous hemorrhage
  - tractional retinal detachment — secondary to extraretinal fibrovascular proliferation

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other clinical findings
- conjunctiva
  - segmentation of blood in the conjunctival blood vessels
  - comma sign
    - comma-shaped thrombi dilate and occlude capillaries
    - inferior bulbar conjunctiva and fornix
- disc sign of sickling
  - dark red spots
  - small vessels on the surface of the optic disc can exhibit intravascular occlusions
- angioid streaks
  - 6% of cases of SS disease
  - AS trait

5 stages
1. peripheral arteriolar occlusions
2. peripheral arteriovenular anastomoses — dilated, preexisting capillary channels
3. preretinal sea fan neovascularization — at the posterior border of areas of nonperfusion
4. vitreous hemorrhage
5. tractional retinal detachment

Lab
- sickling and solubility tests (sickle cell preparations) — for screening
  - do not distinguish between heterozygous and homozygous states
- Hemoglobin electrophoresis — should be performed for patients testing positive on sickle cell preparations

Management
- hyphema
  - all black patients with traumatic hyphema should be screened for sickling hemoglobinopathy (including trait)
  - control of IOP may be difficult
  - ischemic optic neuropathy may result from short intervals of a modest increase in IOP → increased risk of complications
  - consider early anterior chamber washout in presence of hyphema with increased IOP
  - may worsen sickling through the production of systemic acidosis
  - be cautious in the use of carbonic anhydrase inhibitors
- ***Photocoagulation***
  - more commonly after such treatment in PSR than in proliferative diabetic retinopathy
  - lower-intensity light burns to the ischemic peripheral retina
  - peripheral scatter photocoagulation
  - retinal tears and subsequent rhegmatogenous retinal detachment can occur — caution
- ***Vitreoretinal surgery in PSR***
  - indications
    - nonclearing vitreous hemorrhage
    - rhegmatogenous, tractional, schisis, or combined retinal detachment
    - retinal detachment
      - usually begins in the ischemic peripheral retina
      - tears typically occur at the base of sea fans
      - often precipitated by photocoagulation
  - precautions
    - adequate patient hydration
    - supplemental nasal oxygenation
    - judicious use of expansile gases to minimize IOP elevations
    - exchange transfusion before vitreoretinal surgery is no longer favored
    - particularly when combined with extensive diathermy or cryopexy — 360° scleral buckling can cause anterior segment ischemia or necrosis