Sickle cell disease:

Pathophysiology:

normal Hg molecule is Hg A: 2 alpha + 2 beta globin chains

Hg S: 2 alpha + substitution of valine for glutamic acid
Hg C: 2 alpha + substitution of lysine for glutamic acid

insoluble under deoxygenated conditions
leads to distortion of shape, membrane changes, cellular dehydration and
decreased deformability
microinfarction of vital organs, painful bony crisis, splenic autoinfarction and
renal damage

Sickle cell disease:
SS disease
SC disease
Sickle cell trait: AS disease

SC has worst ocular effects because
hematocrit is higher than SS and thus in situation that promotes sickling there
is higher viscosity of blood and more vaso occlusive disease in retina
lower hct and viscosity of SS, protects the retina

Epidemiology

8% of Blacks have sickle cell trait
0.4% SS disease
0.2% SC disease

Nonproliferative Sickle Cell Retinopathy

salmon patch
refractile spots
black sunburst
Proliferative sickle retinopathy

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Peripheral arteriolar occlusions

**HOW?**
- sickled erythrocyte can act as microemboli
- increased viscosity to the point where cessation of flow or thrombus formation

**WHY?**
- peripheral portions of retina more involved
- not dependent only on critical lumen size
- different critical closing pressures
- decreased O2 tensions in peripheral retina

Peripheral arteriolar-venular anastomoses

- anastomoses shunt blood from the occluded arterioles to the nearest venules
- arise at interface of post perfused retina and ant non perfused retina
- enlargement of existing capillaries
- resemble telangetasia and microaneurysm on exam

Peripheral neovascularization
neovascular capillary buds sprout at previous art-venous anastomoses
growth is toward the ora serrata
?vasoformative growth factor in ischemic retina
fan-shaped configuration lie flat in the retina: resemble *Gorgonia flabellum*
= sea fan sign with one feeder arteriole and one draining venule
with time complex arborizing NV patch
protrude from the retina toward center of globe---- lead to VH or
spontaneously involute to white fibrous tissue
SC 59-72% with ocular symptom
supero temporal> inferotemporal> superonasal> inferonasal

FA: profuse leakage of dye into vitreous within 2-3 minutes

**vitreous hemorrhage**
spontaneous vitreous hemorrhage in Black think Sickler
23% SC
3% SS
rare AS
arise from seafans due to vitreous collapse or minor trauma
small or large: near sea fan or center of vitreous cavity
localized peripheral hemorrhages become organized into fibrous tissue
usually peripheral therefore only symptomatic when more severe

**retinal detachment**
usually rhegmatogenous
tears are adjacent or subadjacent to to fibrovascular tissue
shape: ovoid or horseshoe
most occurs with SC disease

**Other findings:**
comma sign inferior fornix
disc sign of sickling
Black pt with traumatic hyphema
CRAO, CRVO, macular arteriole occlusions
angiod streaks

TREATMENT

Neovascularization
- laser: xenon or argon (less VF loss but higher hemorrhage)
- cryotherapy, diathermy
- capillary bed treatment
- feeder vessel treatment: need more energy vs PRP in ischemic retina
  best results in conjunction with FA and cobalt blue filter

Retinal detachment
- Sx difficult:
  1) Retinal breaks are obscured by preretinal fibrovascular tissue
  2) vitreous traction is often pronounced need extensive buckle
  3) high rate of post ant segment necrosis (71% vs 3%)
- local anesthesia without epi
- no removal of EOM
- limit cryopexy
- adequate hydration
- nasal O2

Retinal vascular occlusion
Arteriovenous anastomosis  Hemorrhage

Neovascularization  preretinal  intraretinal  subretinal

Autoinfarction  vitreous hemorrhage  vit fibrosis  refractile  sunburst

Retinal break

RD