Sickle Cell and the Eye

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Disclosures

- No financial interest in this subject matter
Sickle Cell and the Eye- Take Home Points

• Sickle cell disease can affect any part of the eye
• Retinopathy risk variable, depends on genotype
• Male sex, older age, visual symptoms and floaters associated with retinopathy
• Dilated fundus exam required at least annually beginning at age 10, younger if possible
• Treatment can prevent vision loss
Orbital Involvement

- Retro-orbital and Orbital Involvement
  - Orbital bone infarction
  - Orbital compression syndrome

Schundeln et al., Journal of Pediatrics, 2014
Anterior Segment
Sickle Cell Retinopathy

- Goldberg staging (AJO 1971)
  - Stage I: peripheral arteriolar occlusions
  - Stage II: peripheral arteriolar-venular anastomoses
  - Stage III: neovascular and fibrous proliferation
  - Stage IV: vitreous hemorrhage
  - Stage V: retinal detachment

*Top photo by Andrew R. Marks, M.D.*
Non-Proliferative Sickle Cell Retinopathy
Stage 1: Peripheral arteriolar occlusions
Stage 2: Arteriolar-venular anastomoses
Stage 3: Neovascularization
Stage IV: vitreous hemorrhage
Stage IV: vitreous hemorrhage
Seafans visible despite vitreous heme
Stage V: vitreous hemorrhage
Combined TRD/RRD
Sickle Cell Maculopathy

- Numerous macular abnormalities possible
  - Hairpin venular loops
  - Microaneurysmal dots
  - Foveal avascular zone irregularities
- Macular ischemia does not develop neovascularization
- Many patients with abnormalities but no visual consequence
Fig 2.—Right macular region of patient 1. Microaneurysm-like dots (m) are in temporal macula. Pathologic avascular zone (PAZ) is just discernable temporally. Dashed lines enclose area of Fig 4.

Fig 3.—Left macular region of patient 1. Microaneurysm-like dots (m) surround center of macula. Dashed lines enclose area of Fig 5.
Multimodal Imaging in SCR

Temporal parafovea  Fovea  Montage

Fluorescein angiography

Superficial plexus

Deep plexus
Multimodal Imaging in SCR
Treatments for Sickle Cell Retinopathy

• Mostly observation, unless....
Treatments

• Laser photocoagulation
• Intravitreal injection of anti-vascular endothelial growth factor agents (anti-VEGF)
Practical Questions

• What about glasses?
  – Refractive error separate

• Is all this retinal imaging testing necessary?
  – Not sure, a thorough dilated exam will pick up serious retinal pathology

• So do all with SCD have retinopathy?
  – Probably, and will increase as they age

• Is vision loss in SCD certain?
  – Probably not, risk is low, but subtle vision loss may be underdiagnosed

• When are sickle cell retinopathy screening exams necessary?
  – At least annually, age 5 and older, NHLBI consensus suggests age 10
  – Floaters, or vision loss, HbSC/Beta thal
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