Broadly speaking, what sort of disease is sickle-cell dz?
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An amino-acid substitution in the hemoglobin beta-chain leads to its malfolding under certain metabolic conditions (eg, low $O_2$ tension). This results in the characteristic ‘sickling’ of affected RBCs.
Sickle cell: RBC sickling
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What are the four common genotypes of sickle-cell disease?
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--SC
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What is the key difference between SS, SC and S-Thal vs SA disease?
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<tr>
<th>Genotype</th>
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<tr>
<td>SS</td>
<td>Clinically apparent disease</td>
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<tr>
<td>SC</td>
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<tr>
<td>S-Thal</td>
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<td>SA</td>
<td>Asymptomatic carrier state</td>
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What is the key difference between SS, SC and S-Thal vs SA disease?
The first three manifest as clinically apparent dz, whereas SA is an asymptomatic (under most conditions) carrier state--aka ‘sickle trait’
Broadly speaking, what sort of disease is sickle-cell dz?
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In America, people of which two ethnic identities are at greatest risk?
--African-American: 1 in 500
--Hispanic-American: 1 in 36,000

What percent of African-Americans test positive for sickle trait?
8% (1 in 12)
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(People of Mediterranean and Southeast Asian ancestry are also at some risk)
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<thead>
<tr>
<th>Location</th>
<th>DBR</th>
<th>SR</th>
</tr>
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<td>Location</td>
<td><em>Posterior</em> to the equator (usually in the posterior pole)</td>
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*This is an important difference to bear in mind!*
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and/or occlusion
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Capillary and/or arteriolar occlusion
Sickle cell: Vascular occlusion
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What is a salmon patch?
A retinal hemorrhage trapped under the internal limiting membrane

It represents the rupture of an arteriole occluded by sickled RBCs

What accounts for its salmon color?
It is in the process of hemolyzing
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NPSR manifests as three lesions. What are they?
- Salmon patches
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What is the DFE appearance of refractile spots?
- Retinal areas of focal iridescence
- Hemosiderin deposit just beneath the ILM
- Refractile spots are the hemosiderin left when the hemorrhage is resorbed

Refactile spots represent the final stage in the evolution of another retinal lesion—which one?
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Sickle Cell Disease and the Eye

Acute preretinal hemorrhage. The hemorrhage is bright red. Anterior to the hemorrhage, a black sunburst lesion is seen.

Same lesion, 4 weeks later. The hemorrhage is pink (salmon patch) with a surrounding schisis cavity.

Same lesion, 6 weeks later. A schisis cavity is seen with multiple iridescent spots.

Sickle cell: Refractile spots
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**What does a sickle-cell sunburst lesion look like?**
Flat areas of hyperpigmentation, usually round and somewhat stellate (hence the name)

RPE hypertrophy and hyperplasia, along with an accumulation of pigment
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**What is the incidence of PSR in:**
- --SS dz?
- --SC dz?
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What is the incidence of PSR in:

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- **SC dz? 33%**
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What is the incidence of PSR in:
- SS dz? 3%
- SC dz? 33%
- SThal dz?
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
- PSR is more common in SS than SC disease **False—it is more common in SC**

What is the incidence of PSR in:
- **SS dz? 3%**
- **SC dz? 33%**
- **SThal dz? 13%**

What about SThal—what the incidence of PSR for it?
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) True dat
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole False—they are peripheral
- NPSR is more common in SS than SC disease True
- PSR is more common in SS than SC disease False—it is more common in SC

What is the incidence of PSR in:
--SS dz? 3%
--SC dz? 33%
--SThal dz? 13%

So the incidence of PSR is SC > SThal > SS

No question—continue when ready
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: *Nonproliferative* (NPSR), and *proliferative* (PSR) **True**
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- NPSR is more common in SS than SC disease **True**
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*Relatedly: Is PSR a disease of young people, or the elderly?*
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR)  
  
  - True dat

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole  
  
  - False—they are peripheral

- NPSR is more common in SS than SC disease  
  
  - True

- PSR is more common in SS than SC disease  
  
  - False—it is more common in SC

Relatedly: Is PSR a disease of young people, or the elderly? 
Young. It can occur during the teens, and is uncommon after the 30s.
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: *Nonproliferative* (NPSR), and *proliferative* (PSR) \textit{True dat}
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole \textit{False—they are peripheral}
- NPSR is more common in SS than SC disease \textit{True}
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\textit{In a very few words, what is the pathogenesis of PSR?}
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- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: **Nonproliferative** (NPSR), and **proliferative** (PSR) True dat

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*In a very few words, what is the pathogenesis of PSR?*
As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia
Sickle cell: Retinal ischemia
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: *Nonproliferative* (NPSR), and *proliferative* (PSR) True dat
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole False—they are peripheral
- NPSR is more common in SS than SC disease True
- PSR is more common in SS than SC disease False—it is more common in SC

*In a very few words, what is the pathogenesis of PSR?*
As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

*By what appearance-based name are sickle-cell neovascular lesions known?*
Concerning sickle-cell, get your true/false on:

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As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

*By what appearance-based name are sickle-cell neovascular lesions known?*
‘Sea fans’
Sickle-Cell Disease and the Eye

Sickle cell: ‘Sea fans’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
- PSR is more common in SC than SS disease **False—it is more common in SS**

In a very few words, what is the pathogenesis of PSR?

- The pathogenesis underlying PSR is divided into five stages. What are they?

**Stage I:**
- Stage II:
- Stage III:
- Stage IV:
- Stage V:

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’
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- NPSR is more common in SS than SC disease. **True**
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In a very few words, what is the pathogenesis of PSR?

As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia.

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’
Stage I PSR. Peripheral arteriolar occlusions are seen as 'silver-wire' vessels.

Stage I PSR. FA shows the occluded peripheral vessels with the adjacent avascular retina.

Sickle cell: PSR: Stage I
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
- PSR is more common in SC than SS disease **False—it is more common in SS**

In a very few words, what is the pathogenesis of PSR? 
- As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known? 
- ‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?
- **Stage I:** Peripheral arteriolar occlusions
- **Stage II:**
- **Stage III:**
- **Stage IV:**
- **Stage V:**
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- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole. False—they are peripheral

- NPSR is more common in SS than SC disease. True

- PSR is more common in SS than SC disease. False—it is more common in SC.

In a very few words, what is the pathogenesis of PSR?
The pathogenesis underlying PSR is divided into five stages. What are they?

Stage I: Peripheral arteriolar occlusions
Stage II: Anastomosis formation
Stage III:
Stage IV:
Stage V:

By what appearance-based name are sickle-cell neovascular lesions known?
‘Sea fans’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: *Nonproliferative* (NPSR), and *proliferative* (PSR) **True dat**

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In a very few words, what is the pathogenesis of PSR? As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?

- Stage I: Peripheral arteriolar occlusions
- **Stage II: Anastomosis formation**
- Stage III:
- Stage IV:
- Stage V:

Which vessels become anastomosed to one another? The occluded arterioles anastomose to nearby terminal venules by way of pre-existing capillaries

Do the anastomoses leak on FA? No (that’s how you know they are not neo vessels, which are notoriously leaky)
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: **Nonproliferative** (NPSR), and **proliferative** (PSR) **True dat**
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In a very few words, what is the pathogenesis of PSR?

- As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known?

- ‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?

Stage I: Peripheral arteriolar occlusions

Stage II: Anastomosis formation

Stage III: Stage IV:

Stage V:

Which vessels become anastomosed to one another?

The occluded arterioles anastomose to nearby terminal venules by way of pre-existing capillaries
Stage II PSR. FA shows the arteriolar-venular anastomoses with the adjacent avascular retina.
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) True dat

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole False—they are peripheral

- NPSR is more common in SS than SC disease True

- PSR is more common in SC False—it is more common in SS disease

In a very few words, what is the pathogenesis of PSR?

As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?

- Stage I: Peripheral arteriolar occlusions
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Which vessels become anastomosed to one another?

The occluded arterioles anastomose to nearby terminal venules by way of pre-existing capillaries

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No (that’s how you know they are not neo vessels, which are notoriously leaky)
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR). True dat.

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole. False—they are peripheral.

- NPSR is more common in SS than SC disease. True.

- PSR is more common in SS than SC disease. False—it is more common in SC.

In a very few words, what is the pathogenesis of PSR? As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia.

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’.

The pathogenesis underlying PSR is divided into five stages. What are they?

1. **Stage I:** Peripheral arteriolar occlusions
2. **Stage II:** Anastomosis formation
3. **Stage III:** The occluded arterioles anastomose to nearby terminal venules by way of pre-existing capillaries
4. **Stage IV:**
5. **Stage V:**

**Do the anastomoses leak on FA?** No (that’s how you know they are not neo vessels, which are notoriously leaky).
Stage II PSR. FA shows the arteriolar-venular anastomoses with the adjacent avascular retina.

*Note the absence of leakage*

Sickle cell: PSR: Stage II
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True**

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**

- NPSR is more common in SS than SC disease **True**

- PSR is more common in SS than SC disease **False—it is more common in SC**

In a very few words, what is the pathogenesis of PSR?

- The pathogenesis underlying PSR is divided into five stages. **What are they?**
  - Stage I: Peripheral arteriolar occlusions
  - Stage II: Anastomosis formation
  - **Stage III:**
  - Stage IV:
  - Stage V:

By what appearance-based name are sickle-cell neovascular lesions known?

- ‘Sea fans’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is **more common in SS than SC disease** **True**
- PSR is more common in SC **False—it is more common in SC**

**In a very few words, what is the pathogenesis of PSR?**

*The pathogenesis underlying PSR is divided into five stages. What are they?*

- **Stage I:** Peripheral arteriolar occlusions
- **Stage II:** Anastomosis formation
- **Stage III:** Neovascularization (ie, sea-fan formation)
- **Stage IV:**
- **Stage V:**

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) True dat
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole False—they are peripheral
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- PSR is more common in SS than SC disease False—it is more common in SC

The pathogenesis underlying PSR is divided into five stages. What are they?
- Stage I: Peripheral arteriolar occlusions
- Stage II: Anastomosis formation
- Stage III: Neovascularization (ie, sea-fan formation)
- Stage IV: 
- Stage V: 

In which direction do the sea fans ‘grow’: Anteriorly (ie, toward the ora), or posteriorly (toward the macula)?

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: *Nonproliferative* (NPSR), and *proliferative* (PSR)  
  True dat

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole  
  False—they are peripheral

- NPSR is more common in SS than SC disease  
  True

- PSR is more common in SC than SS disease  
  False—it is more common in SC

In a very few words, what is the pathogenesis of PSR?  
As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known?  
'Sea fans'

The pathogenesis underlying PSR is divided into five stages. What are they?

**Stage I:** Peripheral arteriolar occlusions

**Stage II:** Anastomosis formation

**Stage III:** Neovascularization (ie, sea-fan formation)

**Stage IV:**

**Stage V:**

In which direction do the sea fans ‘grow’: Anteriorly (ie, toward the ora), or posteriorly (toward the macula)?  
Anteriorly
(a) Fluorescein angiography of characteristic sea fan neovascularization. (b) The sea fan neovascularization shows evidence of leakage of dye. Inferior to the neovascularization, the arteriolar-venular anastomosis is seen with early neovascularization.

Sickle cell: Stage III
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) True dat
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole False—they are peripheral
- NPSR is more common in SS than SC disease True
- PSR is more common in SC than SS disease False—it is more common in SC

In a very few words, what is the pathogenesis of PSR?

As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

What are these vessels called?

The ‘feeding’ and ‘draining’ vessels, respectively

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?

- Stage I: Peripheral arteriolar occlusions
- Stage II: Anastomosis formation
- Stage III: Neovascularization (ie, sea-fan formation)
- Stage IV:
- Stage V:

In which direction do the sea fans ‘grow’: Anteriorly (ie, toward the ora), or posteriorly (toward the macula)?

Anteriorly

Do sea fans leak on FA?

Yes (as neo lesions usually do)
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR). **True**

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole. **False**—they are peripheral.

- NPSR is more common in SS than SC disease. **True**

- PSR is more common in SC than SS disease. **False**—it is more common in SS.

In a very few words, what is the pathogenesis of PSR? As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia.

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’

The pathogenesis underlying PSR is divided into five stages. What are they?

- **Stage I:** Peripheral arteriolar occlusions
- **Stage II:** Anastomosis formation
- **Stage III:** Neovascularization (ie, sea-fan formation)
- **Stage IV:**
- **Stage V:**

In which direction do the sea fans ‘grow’: Anteriorly (ie, toward the ora), or posteriorly (toward the macula)? **Anteriorly**

Do sea fans leak on FA? **Yes** (as neo lesions usually do)
(a) Fluorescein angiography of characteristic sea fan neovascularization. (b) The sea fan neovascularization shows evidence of leakage of dye. Inferior to the neovascularization, the arteriolar-venular anastomosis is seen with early neovascularization.

*Note the leakage*

Sickle cell: Stage III
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: **Nonproliferative** (NPSR), and **proliferative** (PSR).
  - True dat

- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole.
  - False—they are peripheral.

- NPSR is more common in SS than SC disease.
  - True

- PSR is more common in SS than SC disease.
  - False—it is more common in SC.

In a very few words, what is the pathogenesis of PSR?

- As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia.

By what appearance-based name are sickle-cell neovascular lesions known?

- 'Sea fans'

The pathogenesis underlying PSR is divided into five stages. What are they?

- **Stage I:** Peripheral arteriolar occlusions
- **Stage II:** Anastomosis formation
- **Stage III:** Neovascularization (ie, **sea-fan formation**)
- **Stage IV:** [Irrelevant]
- **Stage V:** [Irrelevant]

In which direction do the sea fans ‘grow’: Anteriorly (ie, toward the ora), or posteriorly (toward the macula)?

- Anteriorly

Do sea fans leak on FA?

- Yes (as neo lesions usually do)

Sea fans are associated with a prominent arteriole and a prominent venule. What are these vessels called?

- The ‘feeding’ and ‘draining’ vessels, respectively.

---

*Sickle-Cell Disease and the Eye*
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR) and proliferative (PSR) True dat
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- PSR is more common in SC than SS disease False—it is more common in SC

The pathogenesis underlying PSR is divided into five stages. What are they?

- Stage I: Peripheral arteriolar occlusions
- Stage II: Anastomosis formation
- Stage III: Neovascularization (i.e., sea-fan formation)
- Stage IV:
- Stage V:

In which direction do the sea fans ‘grow’: Anteriorly (i.e., toward the ora), or posteriorly (toward the macula)?
Anteriorly

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Yes (as neo lesions usually do)

Sea fans are associated with a prominent arteriole and a prominent venule. What are these vessels called?
The ‘feeding’ and ‘draining’ vessels, respectively
(a) Sea fan neovascularization with a single feeder vessel and two draining venules.  
(b) Sea fan neovascularization with multiple feeder arterioles and draining venules.

Sickle cell: Stage III
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**

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- NPSR is more common in SS than SC disease **True**

- PSR is more common in SS than SC disease **False—it is more common in SC**

In a very few words, what is the pathogenesis of PSR?

As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’
Concerning sickle-cell, get your true/false on:

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  - **Stage III:** Neovascularization (ie, sea-fan formation)
  - **Stage IV:** Vitreous hemorrhage
  - **Stage V:** Sickle-Cell Disease and the Eye

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’
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In a very few words, what is the pathogenesis of PSR? As with NPSR, vascular occlusion is the culprit, only it's severe enough to result in significant ischemia

By what appearance-based name are sickle-cell neovascular lesions known? ‘Sea fans’
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As with NPSR, vascular occlusion is the culprit, only it’s severe enough to result in significant ischemia.

By what appearance-based name are sickle-cell neovascular lesions known?

‘Sea fans’
Sickle Cell Disease and the Eye

Sickle cell: PSR: TRD
Sickle-Cell Disease and the Eye

Sickle cell: PSR: Stages
Sickle-Cell Disease and the Eye

Stages of Proliferative Sickle Cell Retinopathy - Fundus Views

Sickle cell: PSR: Stages
Concerning sickle-cell, get your true/false on:

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- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
- PSR is more common in SS than SC disease **False—it is more common in SC**
- Sea-fan lesions frequently regress spontaneously
Concerning sickle-cell, get your true/false on:

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- PSR is more common in SS than SC disease **False—it is more common in SC**
- Sea-fan lesions frequently regress spontaneously **True**
Sickle-Cell Disease and the Eye

Autoinfarcted sea fan neovascularization. The white appearance is classic.

Sickle cell: Autoinfarction
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR) and Proliferative (PSR). True
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole. False—They are peripheral.
- NPSR is more common in SS than SC disease. True
- PSR is more common in SC than SS disease. False—It is more common in SC.
- Sea-fan lesions frequently regress spontaneously. True

<table>
<thead>
<tr>
<th>Location</th>
<th>DBR</th>
<th>SR</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Proliferative lesions regress spontaneously?</strong></td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Another important difference
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
- PSR is more common in SS than SC disease **False—it is more common in SC**
- Sea-fan lesions frequently regress spontaneously **True**
- Laser photocoagulation of sea-fan feeder vessels is indicated
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: **Nonproliferative** (NPSR), and **proliferative** (PSR) **True dat**
- As in DBR, lesions in sickle-cell retinopathy are typically located in the posterior pole **False—they are peripheral**
- NPSR is more common in SS than SC disease **True**
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The final stage of PSR is tractional RD, not rhegmatogenous. Why the concern over RRD?
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**Rhegmatogenous** RD is a significant concern in PSR **True**

The final stage of PSR is tractional RD, not rhegmatogenous. Why the concern over RRD? Because the sickle-cell retina is prone to developing tears when it is lasered. For this reason, the decision to treat must be made judiciously.
Concerning sickle-cell, get your true/false on:

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The final stage of PSR is tractional RD, not rhegmatogenous. Why the concern over RRD? Because **the sickle-cell retina is prone to developing tears when it is lasered**. For this reason, the decision to treat must be made judiciously.

<table>
<thead>
<tr>
<th></th>
<th>DBR</th>
<th>SR</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Location</strong></td>
<td><strong>Posterior</strong> to the equator (usually in the posterior pole)</td>
<td><strong>Anterior</strong> to the equator (ie, peripherally)</td>
</tr>
<tr>
<td><strong>Proliferative lesions regress spontaneously?</strong></td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Prone to developing retinal tears when lasered?</strong></td>
<td>No</td>
<td>Yes</td>
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</tbody>
</table>

Another important difference

"Sickle-Cell Disease and the Eye"
Concerning sickle-cell, get your true/false on:

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**Bonus**: What are the three classic nonretinal ocular stigmata of sickle-cell disease?

---

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- Comma sign
- Disc sign
- Angioid streaks

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--Comma sign? What is comma sign?
--Disc sign
--Angioid streaks

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**Bonus:** *What are the three classic nonretinal ocular stigmata of sickle-cell disease?*

--- **Comma sign:** Segmented heme in occluded conj vessels, esp in the inferior fornix
--- **Disc sign**
--- **Angioid streaks**

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- Rhegmatogenous RD is a significant concern in PSR **True**
The *comma sign* of sickle-cell dz. Blocked small conj vessels are seen as comma-shaped lines.

Sickle cell: ‘Comma sign’
Concerning sickle-cell, get your true/false on:

- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
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- Rhegmatogenous RD is a significant concern in PSR **True**
The *disc sign* of sickling. Blocked small vessels are seen as dark spots or lines.

**Sickle cell: ‘Disc sign’**
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- Like diabetic retinopathy (DBR), sickle-cell retinopathy comes in two basic forms: Nonproliferative (NPSR), and proliferative (PSR) **True dat**
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Sickle-Cell Disease and the Eye

Angioid streaks (arrowheads). Note that only a few of the many present have been marked.
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What is the well-known mnemonic for the DDx for angioid streaks?

- ?
- ?
- ?
- ?
- ?
- ?
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- ?
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What is the well-known mnemonic for the DDx for angioid streaks?

- P
- E
- P
- S
- I

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What is the well-known mnemonic for the DDx for angioid streaks?
- P
- E
- P
- Sickle-cell anemia
- I

What does each letter stand for (other than the ‘S’, duh)?
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- P
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- **Angioid streaks:** Reddish linear abnormalities in peripapillary Bruch’s

What is the well-known mnemonic for the DDx for angioid streaks?
- **P**seudoxanthoma elasticum
- **E**hlers-Danlos dz
- **P**aget’s dz of bone
- **S**ickle-cell anemia
- **I**diopathic

Rhegmatogenous RD is a significant concern in PSR _True_
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For more on angioid streaks, see slide-set R61

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