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A hemoglobinopathy
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What is the underlying problem?
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An amino-acid substitution in the hemoglobin beta-chain leads to its malfolding under certain metabolic conditions (e.g., low O₂ 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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Sickle-Cell Disease and the Eye

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--SC
--S-Thal
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- SS
- SC
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What is the key difference between SS, SC and S-Thal vs SA disease?
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--SS
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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
--Hispanic-American

(People of Mediterranean and Southeast Asian ancestry are also at some risk)
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--SC
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--SA

In America, people of which two ethnic identities are at greatest risk?
--African-American: 1 in ?
--Hispanic-American: 1 in ?

What is the sickle-cell dz birthrate for these groups?
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

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--African-American: 1 in 500
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What percent of African-Americans test positive for 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)
Concerning sickle-cell, get your true/false on:

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

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<table>
<thead>
<tr>
<th>Location</th>
<th>DBR</th>
<th>SR</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Posterior</em> to the equator (usually in the posterior pole)</td>
<td><em>Anterior</em> to the equator (ie, peripherally)</td>
<td></td>
</tr>
</tbody>
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*This is an important difference to bear in mind!*
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*In a very few words, what is the pathogenesis of NPSR?*
Concerning sickle-cell, get your true/false on:

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

**Capillary and/or arteriolar occlusion**
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In a very few words, what is the pathogenesis of NPSR? Capillary and/or arteriolar occlusion
Sickle cell: Vascular occlusion
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 manifests as three lesions. What are they?
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*In a very few words, what is the pathogenesis of NPSR?*
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What is a salmon patch?

A retinal hemorrhage trapped under the internal limiting membrane

What is its origin?

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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What is the DFE appearance of refractile spots? Retinal areas of focal iridescence

What substance accounts for their iridescence? Hemosiderin deposit just beneath the ILM

Refractile spots represent the final stage in the evolution of another retinal lesion—which one? Salmon patches. Refractile spots are the hemosiderin left when the hemorrhage is resorbed
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- **NPSR** is more common in SS than SC disease. **True**

**In a very few words, what is the pathogenesis of NPSR?**
Capillary and/or arteriolar occlusion

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**NPSR**

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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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Capillary and/or arteriolar occlusion

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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)

What causes them?
RPE hypertrophy and hyperplasia, along with an accumulation of pigment
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Sickle-Cell Disease and the Eye

Salmon patches too

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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:*
  --*SS dz? 3%*
  --*SC dz?*
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- NPSR is more common in SS than SC disease **True**
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What is the incidence of PSR in:
- --SS dz? **3%**
- --SC dz?
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- NPSR is more common in SS than SC disease True
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What is the incidence of PSR in:
- SS dz? 3%
- SC dz? 33%
Concerning sickle-cell, get your true/false on:

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Relatedly: Is PSR a disease of young people, or the elderly?
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*In a very few words, what is the pathogenesis of PSR?*
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?*
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:

- 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’
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 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:**

**Stage II:**

**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 disease. False—it is more common in SS disease.

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:**

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

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**

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:**

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

As with NPSR, 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:

- 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

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 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:
- Stage IV:
- Stage V:

Which vessels become anastomosed to one another?

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 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

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

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.

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.

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 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 disease 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
- Stage II: Anastomosis formation
- Stage III:

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
- 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

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)

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

- 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 76

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 (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)?
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**

**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**) (Corrected)
- **Stage IV:**
- **Stage V:**

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

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

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.**
(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 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)?
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
- 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

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

Do sea fans leak on FA?

Yes (as neo lesions usually do)

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

‘Sea fans’
(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: \textit{Nonproliferative} (NPSR), and \textit{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}

- PSR is more common in SS than SC disease \textit{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'

What are these vessels called?

The 'feeding' and 'draining' vessels, respectively

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

\begin{itemize}
  \item \textbf{Stage I}: Peripheral arteriolar occlusions
  \item \textbf{Stage II}: Anastomosis formation
  \item \textbf{Stage III}: Neovascularization (\ie, \textit{sea-fan formation})
  \item \textbf{Stage IV}:
  \item \textbf{Stage V}:
\end{itemize}

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

\textit{Anteriorly}

Do sea fans leak on FA?

\textit{Yes (as neo lesions usually do)}

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

\textit{The 'feeding' and 'draining' vessels, respectively}
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

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)

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
(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
- 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

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

- NPSR is more common in SS than SC disease True

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

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: Vitreous hemorrhage
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 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’
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

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:** Vitreous hemorrhage
- **Stage V:** Retinal detachment

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 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: Neovascularization (i.e., sea-fan formation)
Stage IV: Vitreous hemorrhage
Stage V: Tractional retinal detachment

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: PSR: Stages

Sickle-Cell Disease and the Eye
Sickle-Cell Disease and the Eye

Sickle cell: PSR: Stages
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
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**
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 disease. True
- PSR is more common in SC disease. False—it is more common in SC
- Sea-fan lesions frequently regress spontaneously. True

<table>
<thead>
<tr>
<th></th>
<th>DBR</th>
<th>SR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Location</td>
<td>Posterior to the equator (usually in the posterior pole)</td>
<td>Anterior to the equator (ie, peripherally)</td>
</tr>
<tr>
<td>Proliferative lesions regress spontaneously?</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**
- 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 **False—treatment is directed at the areas of nonperfusion, not the feeder vessels**
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**
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- Rhegmatogenous RD is a significant concern in PSR
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
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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 **True**
- Laser photocoagulation of sea-fan feeder vessels is indicated **False—treatment is directed at the areas of nonperfusion, not the feeder vessels**
- **Rhegmatogenous** RD is a significant concern in PSR **True**

*The final stage of PSR is tractional RD, not rhegmatogenous. Why the concern over RRD?*
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 **False—treatment is directed at the areas of nonperfusion, not the feeder vessels**

**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.
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**Sickle-Cell Disease and the Eye**

<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>Proliferative lesions regress spontaneously?</td>
<td><strong>No</strong></td>
<td><strong>Yes</strong></td>
</tr>
<tr>
<td>Prone to developing retinal tears when lasered?</td>
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</tr>
</tbody>
</table>

Another important difference

---

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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: Segmented heme in occluded conj vessels, esp in the inferior fornix
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The *comma sign* of sickle-cell dz. Blocked small conj vessels are seen as comma-shaped lines.

**Sickle cell: ‘Comma sign’**
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In what infectious condition is comma sign a well-known finding? HIV

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--Comma sign: Segmented heme in occluded conj vessels, esp in the inferior fornix

--Disc sign? What is disc sign?

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Sickle cell: ‘Disc sign’

The disc sign of sickling. Blocked small vessels are seen as dark spots or lines.

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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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- --?
- --?
- --?
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- **P**
- **E**
- **P**
- **S**
- **I**

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What does each letter stand for (other than the ‘S’, duh)?

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- **P**seudoxanthoma elasticum
- **E**hlers-Danlos dz
- **P**aget’s dz of bone
- **S**ickle-cell anemia
- **I**diopathic

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