

Q

Sickle-Cell Disease and the Eye

Broadly speaking, what sort of disease is sickle-cell dz?



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A hemoglobinopathy



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What is the underlying problem?

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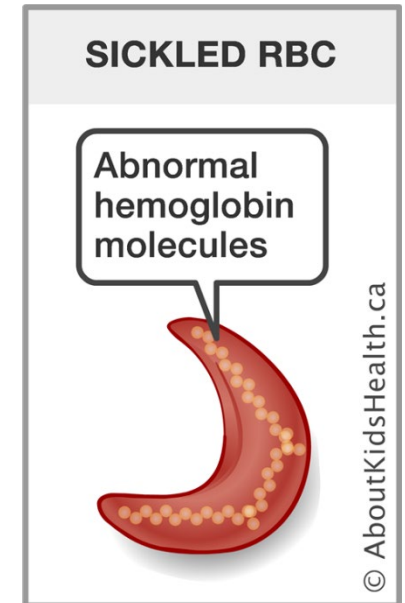
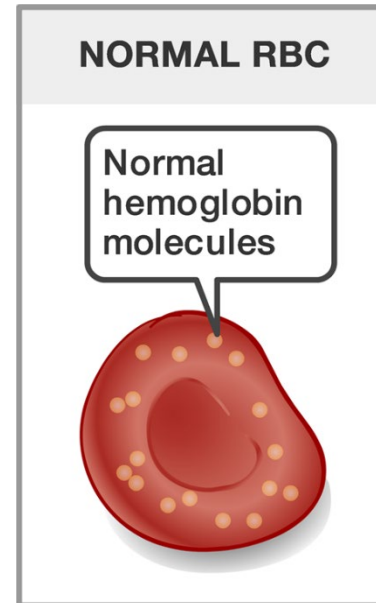
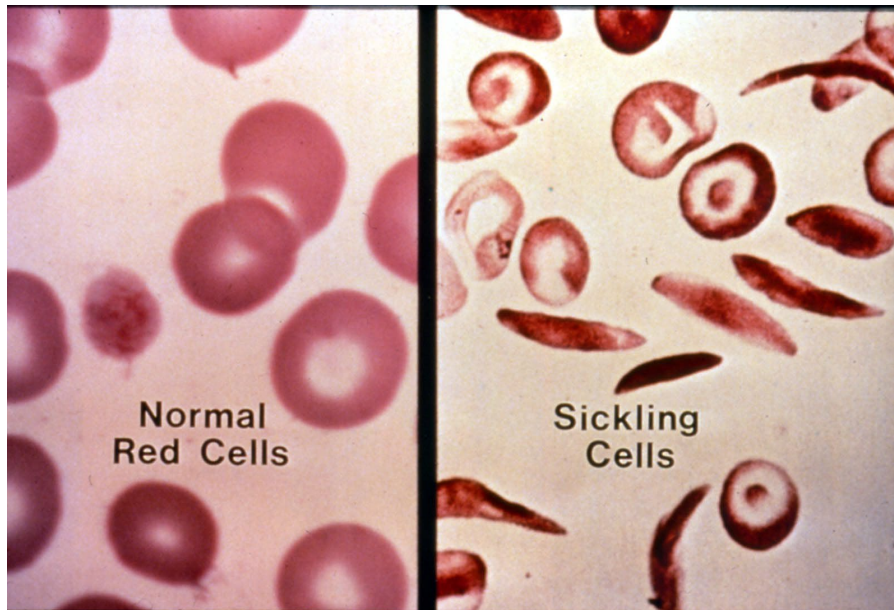
A hemoglobinopathy

What is the underlying problem?

An amino-acid substitution in the hemoglobin beta-chain leads to its misfolding under certain metabolic conditions (eg, low O₂ tension). This results in the characteristic 'sickling' of affected RBCs.



Sickle-Cell Disease and the Eye



Sickle cell: RBC sickling

Q

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6

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--SC

--S-Thal

--SA

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8



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



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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'

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10

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In America, people of which two ethnic identities are at greatest risk?

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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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--African-American: 1 in ?

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



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What percent of African-Americans test positive for sickle trait?



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8% (1 in 12)

Sickle-Cell Disease and the Eye

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



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Sickle-Cell Disease and the Eye

17



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20



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	DBR	SR
<i>Location</i>	Posterior to the equator (usually in the posterior pole)	Anterior to the equator (ie, peripherally)

This is an important difference to bear in mind!

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22



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



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and/or occlusion



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Sickle-Cell Disease and the Eye

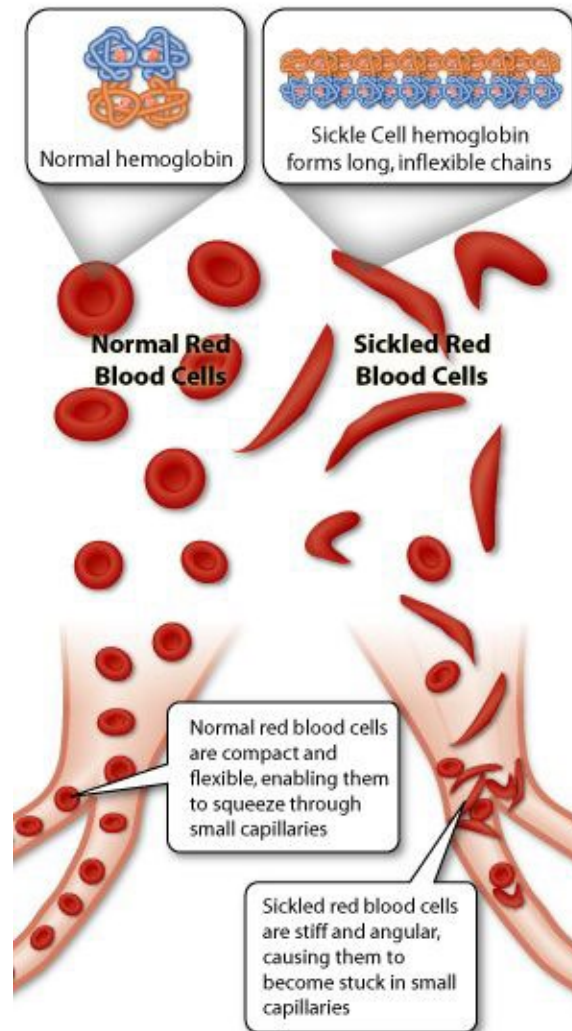
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Capillary and/or arteriolar occlusion



Sickle-Cell Disease and the Eye



Sickle cell: Vascular occlusion



Q

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NPSR manifests as three lesions. What are they?

- Salmon patches
- Refractile spots
- Sunburst lesions

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Sickle-Cell Disease and the Eye

30



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In a very few words
Capillary and/or a

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Sickle-Cell Disease and the Eye

31



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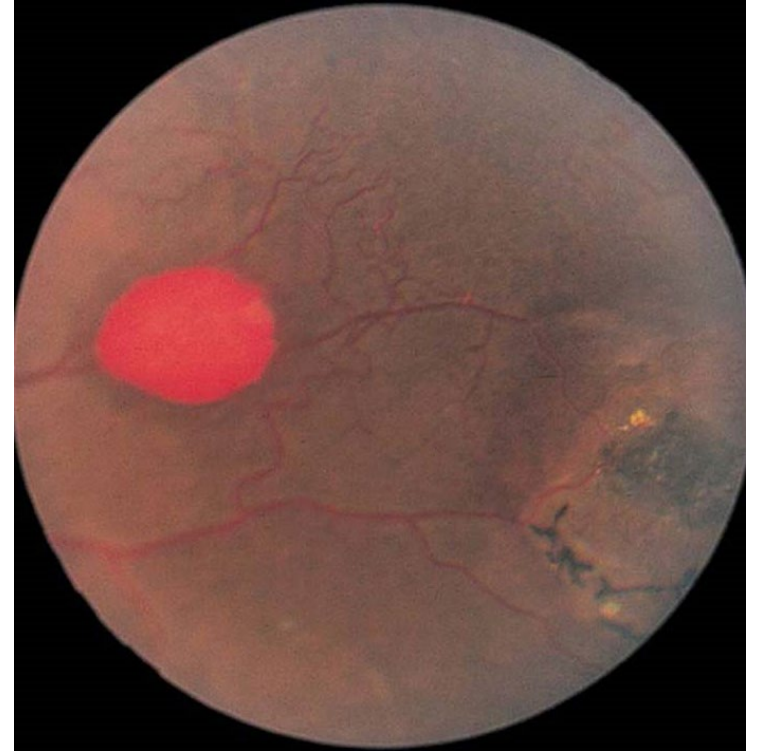
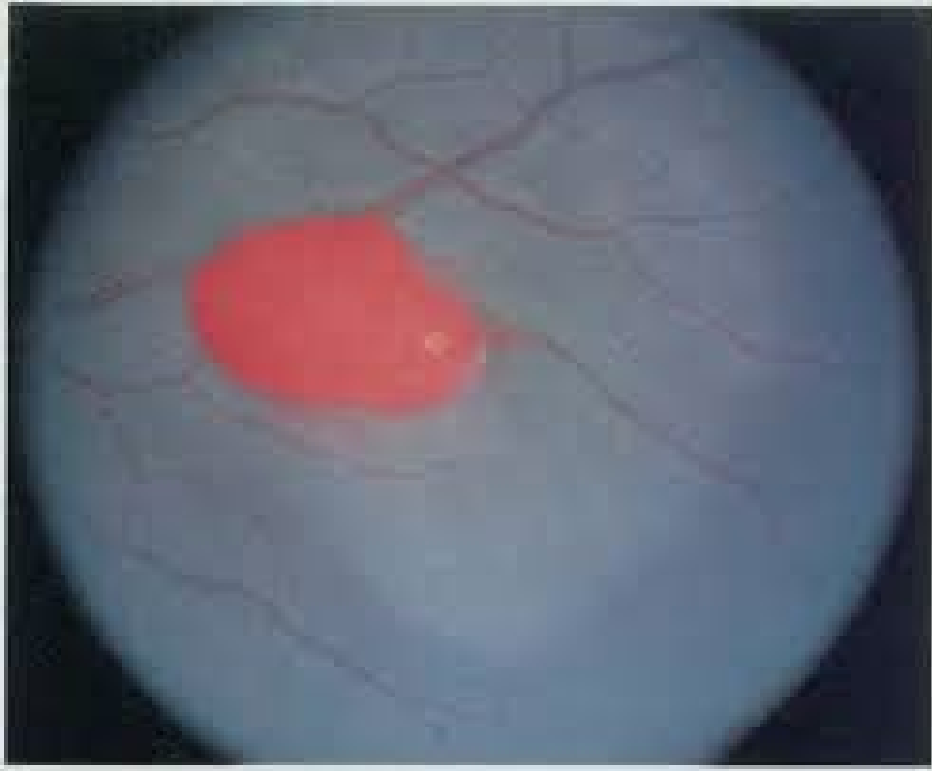
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What is a salmon patch?

A retinal hemorrhage trapped under the internal limiting membrane

Sickle-Cell Disease and the Eye



Sickle cell: Salmon patch



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34



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35



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It is in the process of hemolyzing

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37



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41



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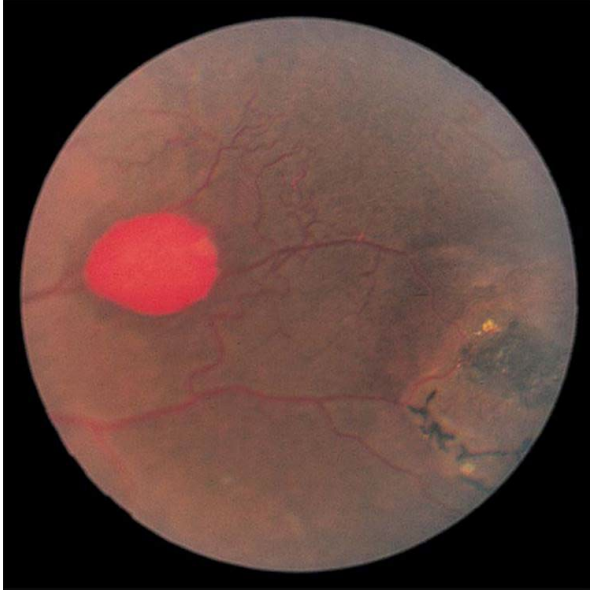
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Refractile spots
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Refractile spots represent the final stage in the evolution of another retinal lesion—which one?
Salmon patches. Refractile spots are the hemosiderin left when the hemorrhage is resorbed

Sickle-Cell Disease and the Eye



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

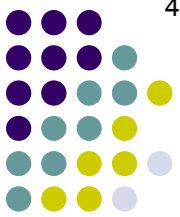


Same lesion, 4 weeks later. The hemorrhage is pink (salmon patch) with a surrounding schisis cavity.



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

Sickle cell: Refractile spots



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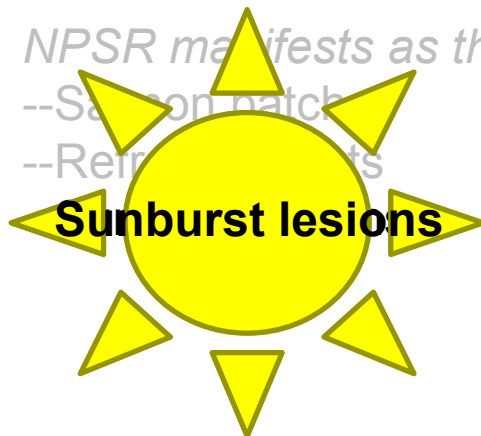
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NPSR manifests as three lesions. What are they?

--Sawson patches
--Rendu's



What does a sickle-cell sunburst lesion look like?

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Sickle-Cell Disease and the Eye



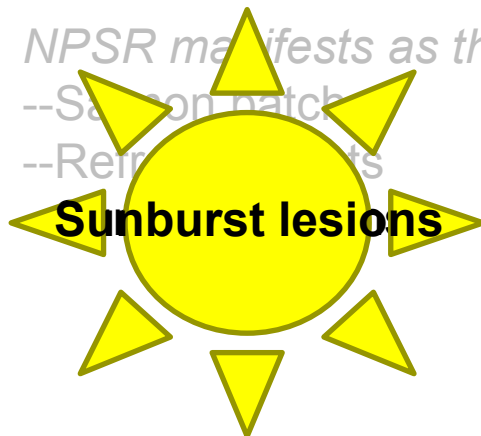
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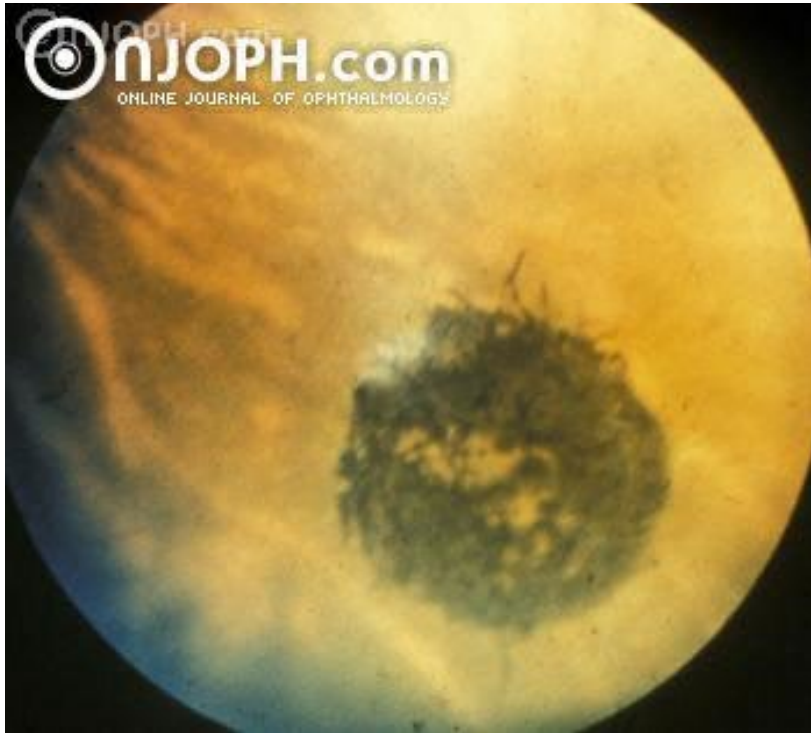


What does a sickle-cell sunburst lesion look like?

Flat areas of hyperpigmentation, usually round and somewhat stellate (hence the name)



Sickle-Cell Disease and the Eye



Salmon patches too

Sickle cell: Sunburst lesions

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Sickle-Cell Disease and the Eye



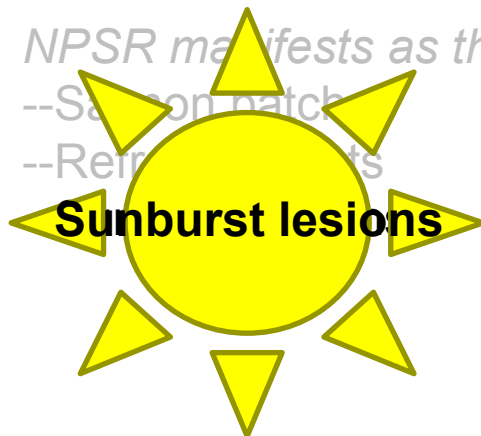
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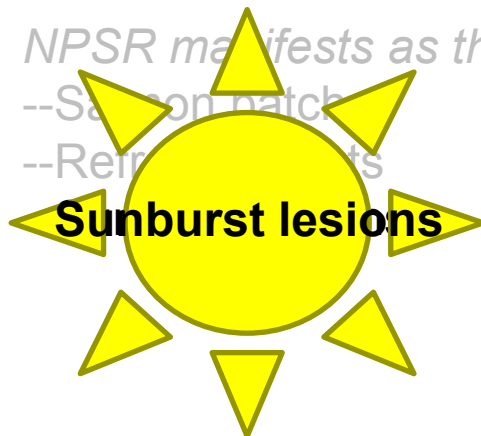
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--Retrolental fibroplasia



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What causes them?

RPE hypertrophy and hyperplasia, along with an accumulation of pigment



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Sickle-Cell Disease and the Eye

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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

A

Sickle-Cell Disease and the Eye

50



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Q

Sickle-Cell Disease and the Eye

51



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What is the incidence of PSR in:
--SS dz?
--SC dz?



A

Sickle-Cell Disease and the Eye

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



Q

Sickle-Cell Disease and the Eye

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What is the incidence of PSR in:
--SS dz? 3%
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A

Sickle-Cell Disease and the Eye

54



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What is the incidence of PSR in:

--SS dz? 3%

--SC dz? 33%



Q

Sickle-Cell Disease and the Eye

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What about SThal—what the incidence of PSR for it?

What is the incidence of PSR in:

--SS dz? **3%**

--SC dz? **33%**

--SThal dz?



A

Sickle-Cell Disease and the Eye

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What about SThal—what the incidence of PSR for it?

What is the incidence of PSR in:

--SS dz? **3%**

--SC dz? **33%**

--SThal dz? **13%**



Sickle-Cell Disease and the Eye

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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

Q

Sickle-Cell Disease and the Eye

58



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Relatedly: Is PSR a disease of young people, or the elderly?

A

Sickle-Cell Disease and the Eye

59



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Relatedly: Is PSR a disease of young people, or the elderly?

Young. It can occur during the teens, and is uncommon after the 30s.



Q

Sickle-Cell Disease and the Eye

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



A

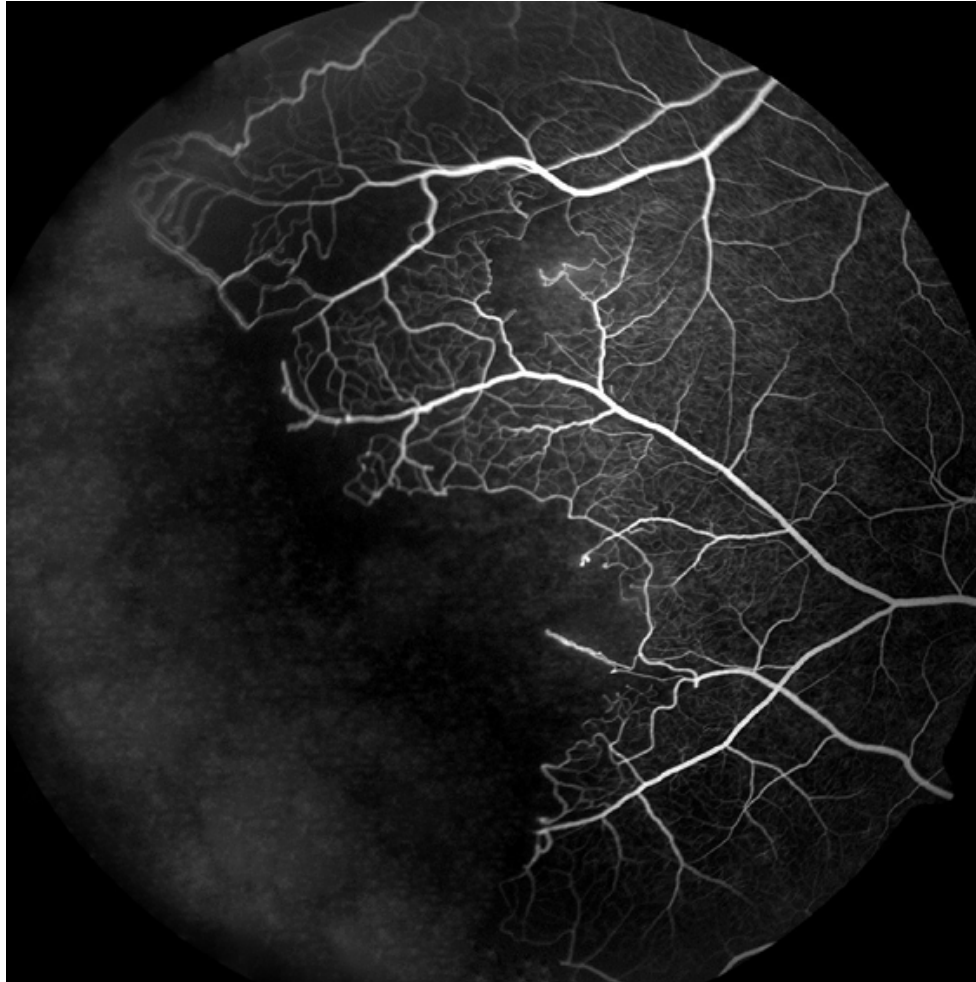
Sickle-Cell Disease and the Eye

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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 Disease and the Eye



Sickle cell: Retinal ischemia





Q

Sickle-Cell Disease and the Eye

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Sickle-Cell Disease and the Eye

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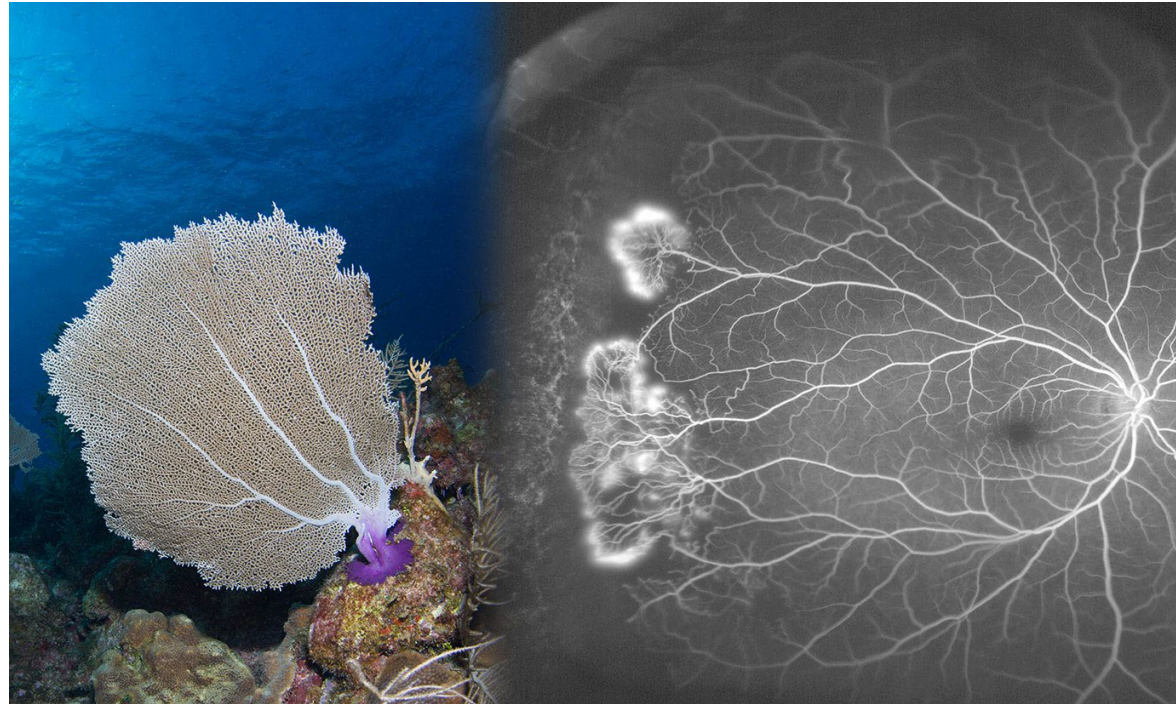
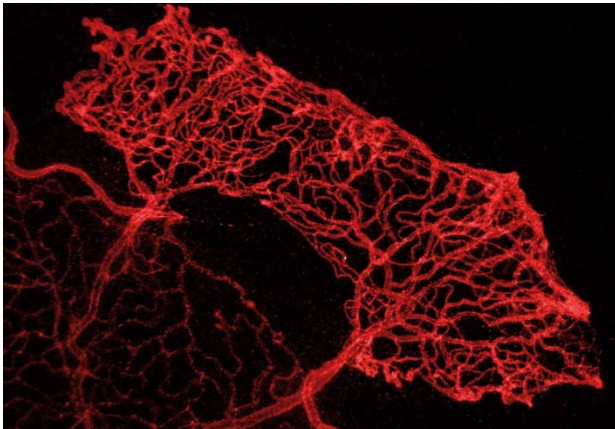
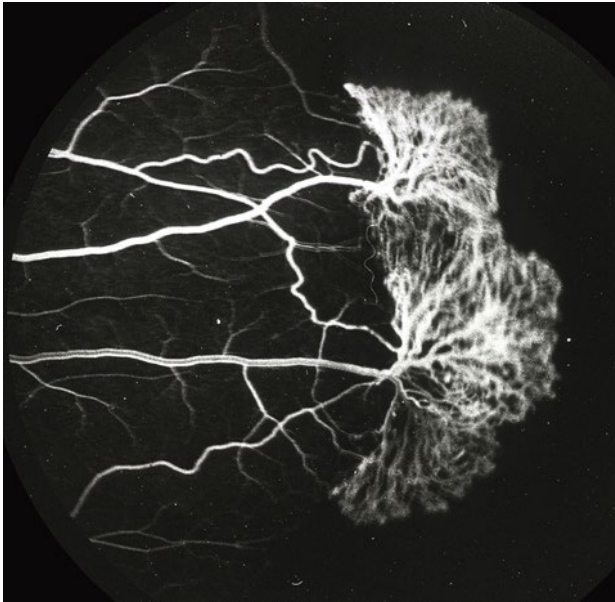
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'Sea fans'

Sickle-Cell Disease and the Eye



Sickle cell: 'Sea fans'



Q

Sickle-Cell Disease and the Eye

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

common

In a very few

As with NPSR

it's severe enough to result in significant ischemia

is more

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Sickle-Cell Disease and the Eye

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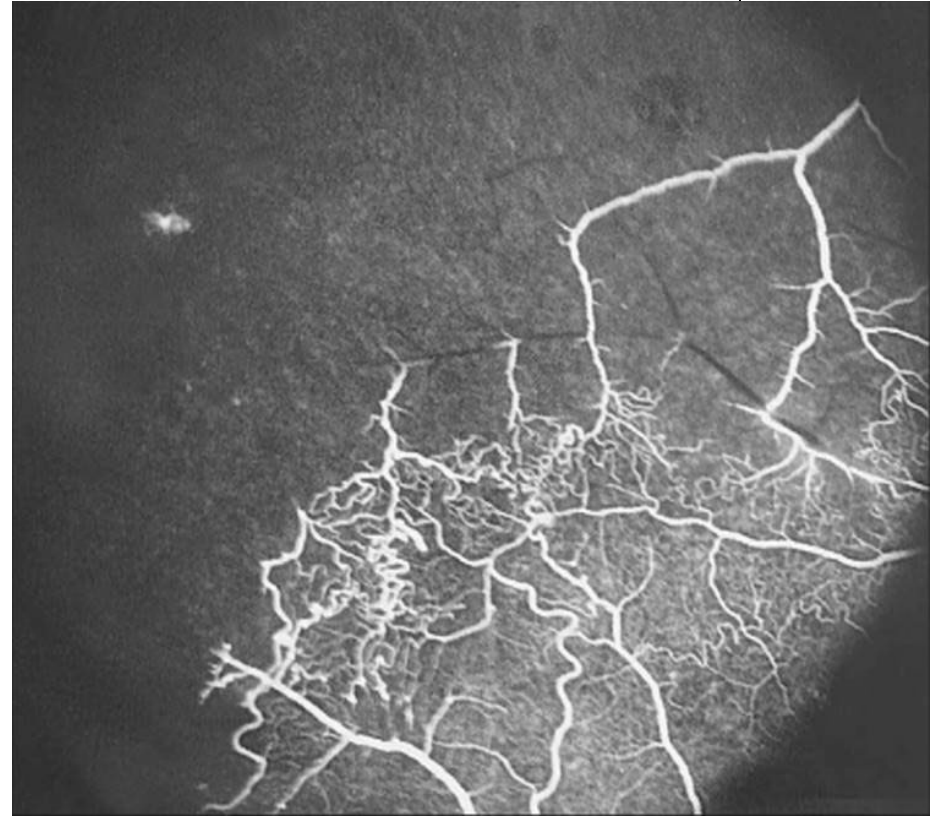
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Sickle-Cell Disease and the Eye



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



Q

Sickle-Cell Disease and the Eye

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- In a very few*
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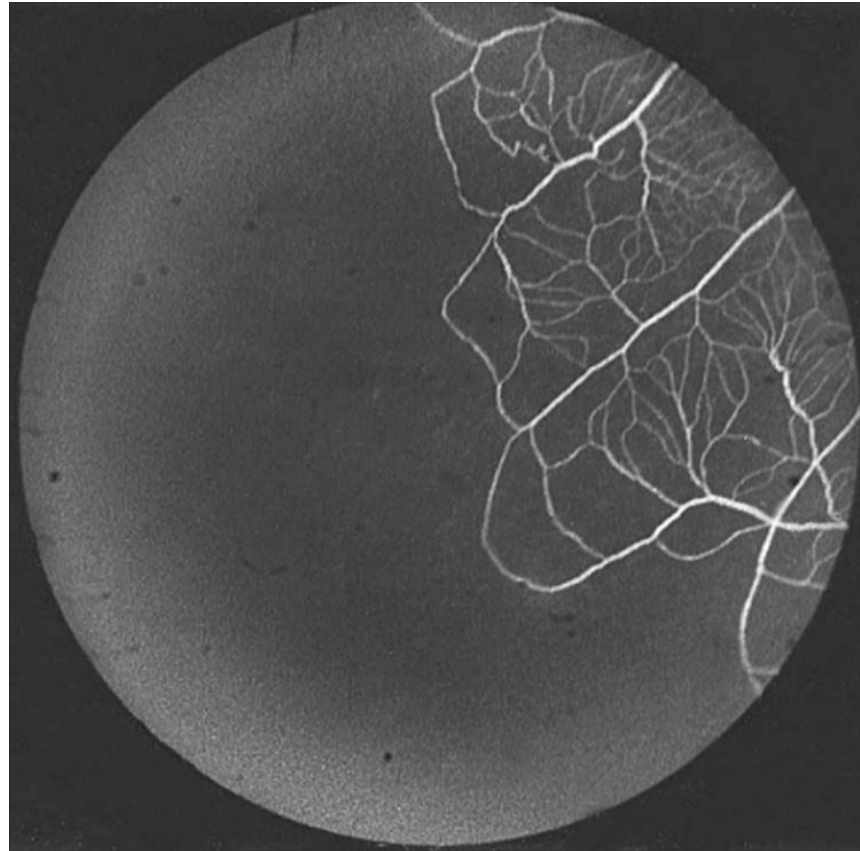


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Sickle-Cell Disease and the Eye

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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
- In a very few*
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Sickle-Cell Disease and the Eye



Stage II PSR. FA shows the arteriolar-venular anastomoses with the adjacent avascular retina.

Sickle cell: PSR: Stage II



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Stage II: Anastomosis formation

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?

In a very few

As with NPSR

it's severe end

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'Sea fans'

is more

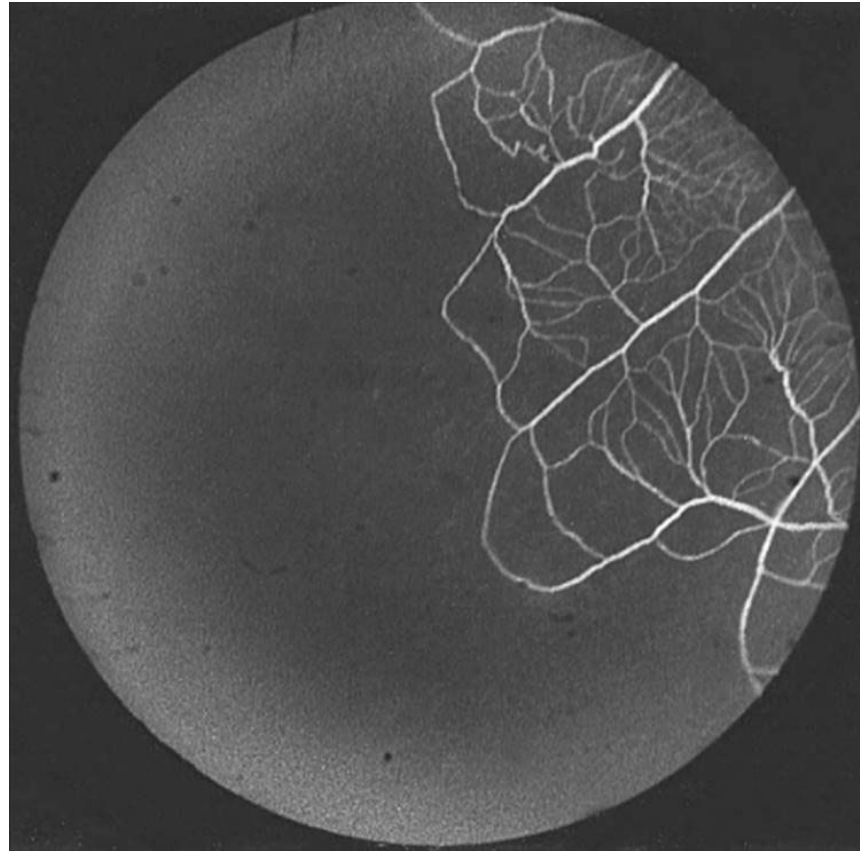


A

Sickle-Cell Disease and the Eye

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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
 - Do the anastomoses leak on FA?*
 - No (that's how you know they are not neo vessels, which are notoriously leaky)
- In a very few*
- As with NPSR*
- it's severe end*
- By what appe*
- neovascular lesions known:*
- 'Sea fans'*

Sickle-Cell Disease and the Eye



Stage II PSR. FA shows the arteriolar-venular anastomoses with the adjacent avascular retina.

Note the absence of leakage

Sickle cell: PSR: Stage II

Q

Sickle-Cell Disease and the Eye



77

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Stage IV:
Stage V:
is more common
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As with NPSR
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 'Sea fans'



A

Sickle-Cell Disease and the Eye

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

Stage IV:

Stage V:

In a very few

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'Sea fans'

is more



Q

Sickle-Cell Disease and the Eye

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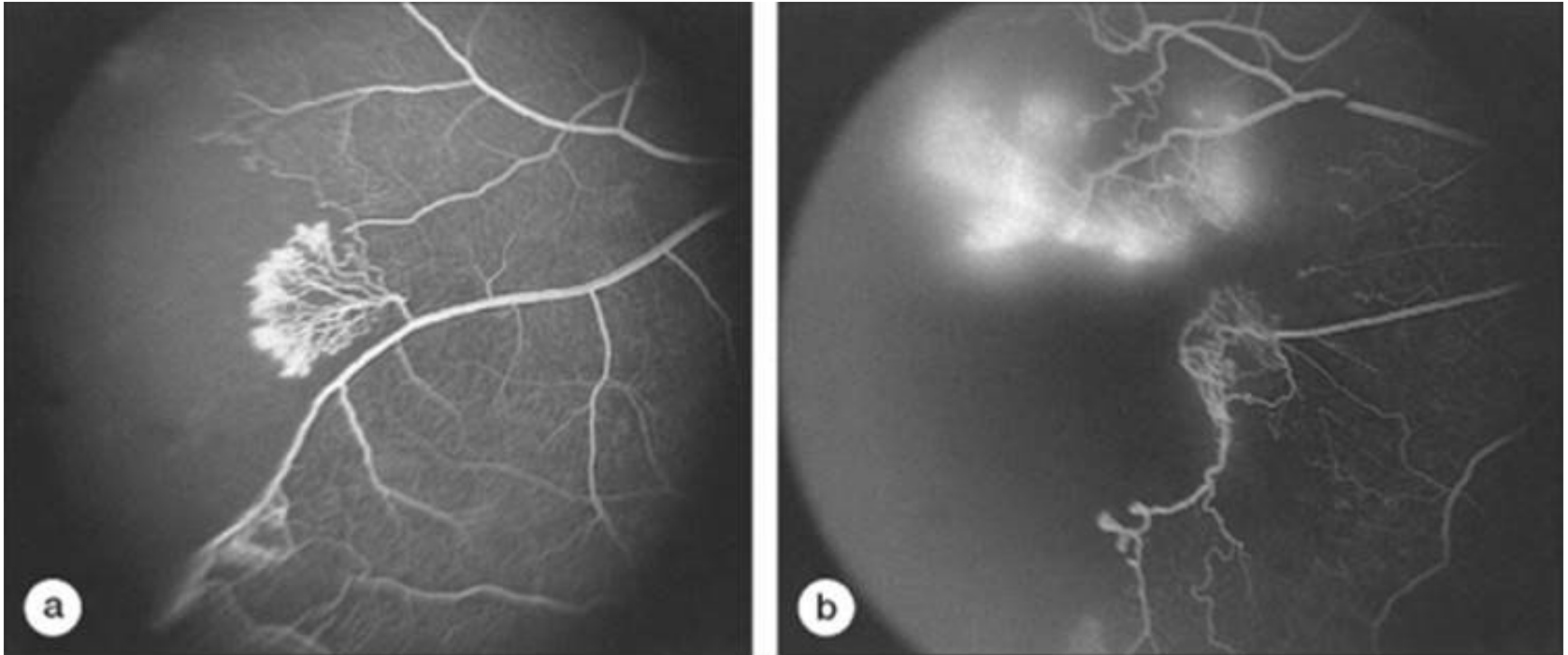
In which direction do the sea fans 'grow': Anteriorly (ie, toward the ora), or posteriorly (toward the macula)?



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Anteriorly



Sickle-Cell Disease and the Eye



(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



Q

Sickle-Cell Disease and the Eye

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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?



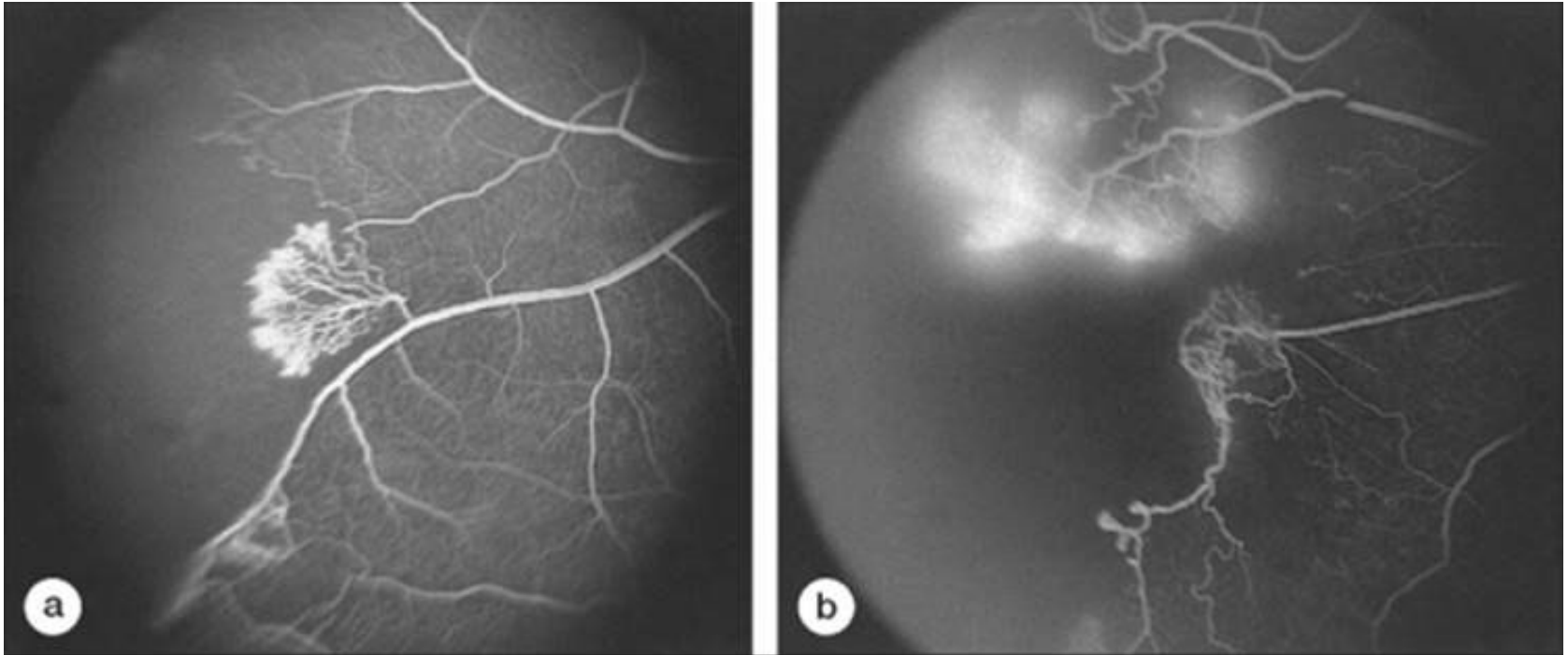
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Sickle-Cell Disease and the Eye



(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



Q

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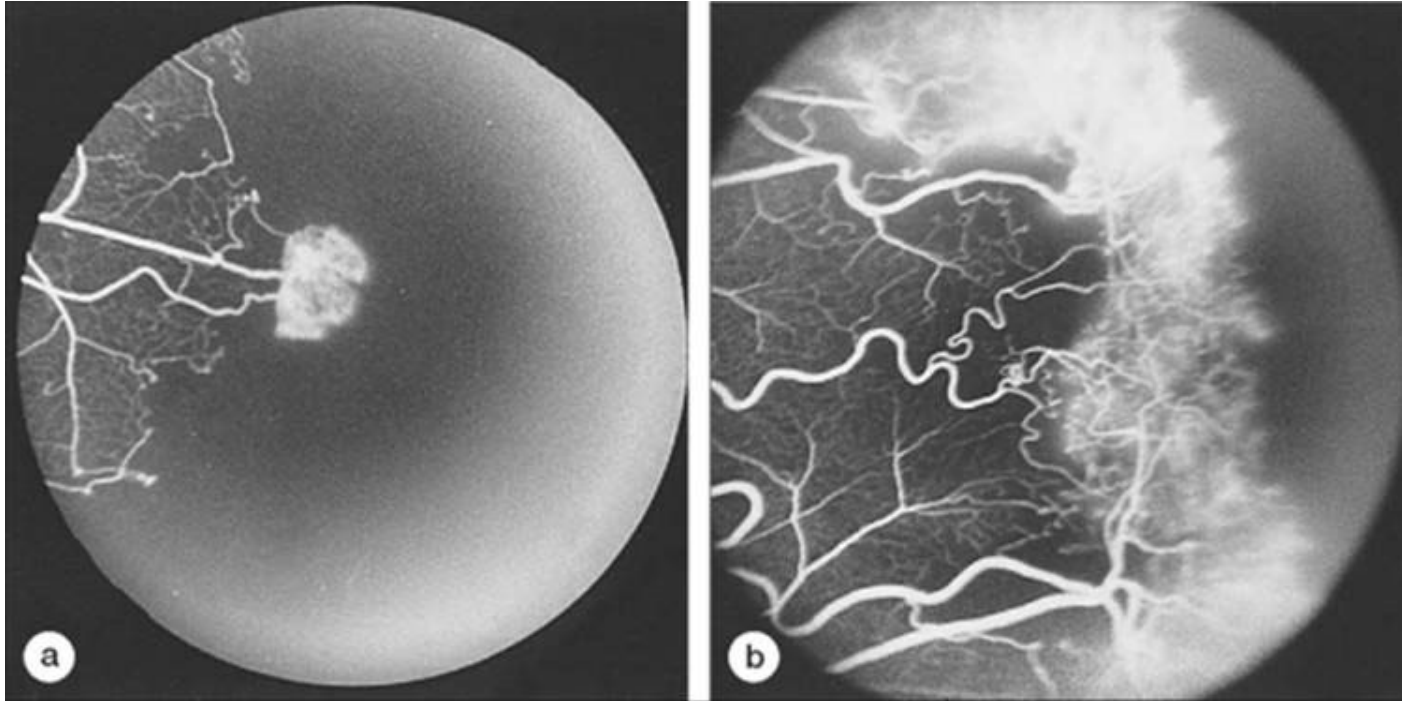
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 - By what appearance* *Do sea fans leak on FA?* *Yes (as neo lesions usually do)* *neovascular lesions* *Sea fans are associated with a prominent arteriole and a prominent venule. What are these vessels called?* *The 'feeding' and 'draining' vessels, respectively* *'Sea fans'*



Sickle-Cell Disease and the Eye



- (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



Q

Sickle-Cell Disease and the Eye

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

Stage IV:

Stage V:

In a very few

As with NPSR

it's severe enough to result in significant ischemia

is more

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

'Sea fans'



A

Sickle-Cell Disease and the Eye

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In a very few

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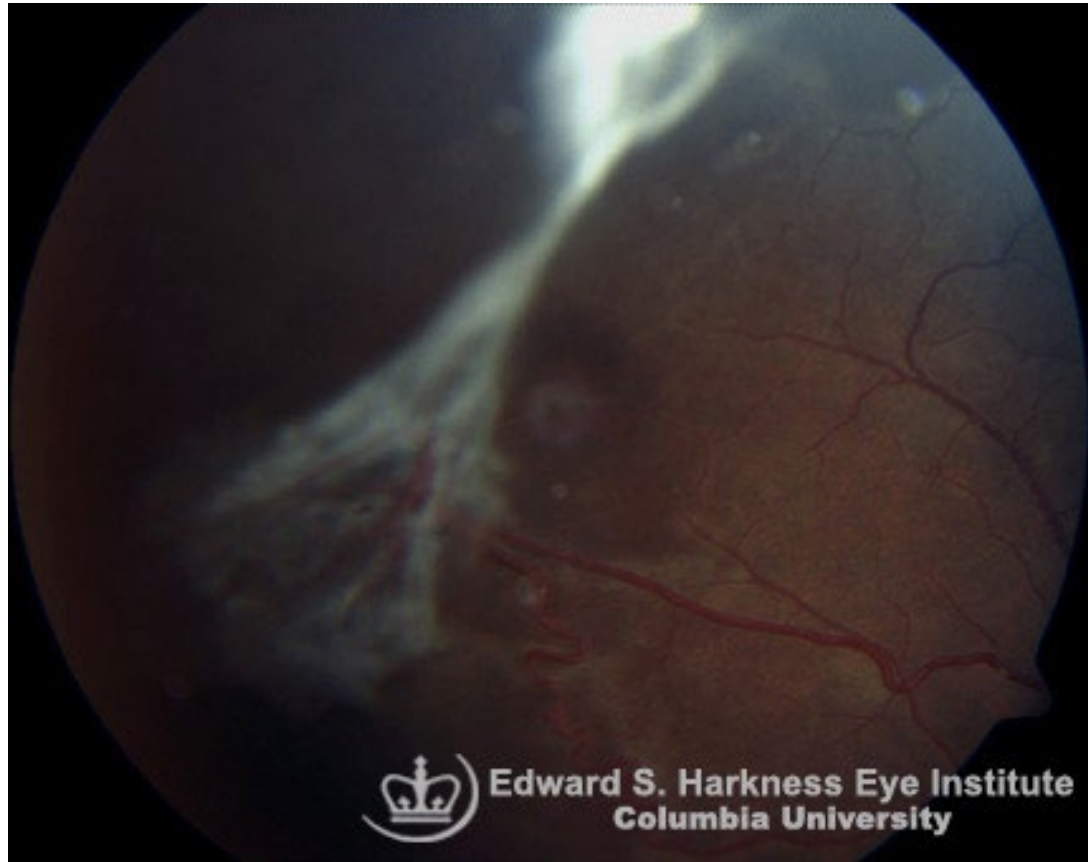
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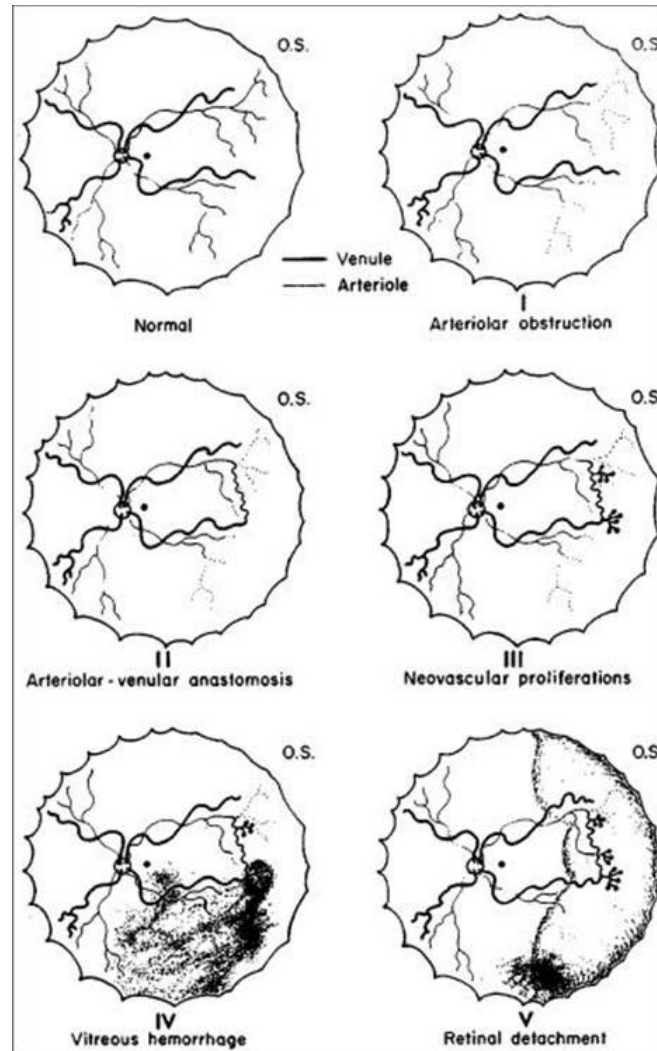


Sickle-Cell Disease and the Eye



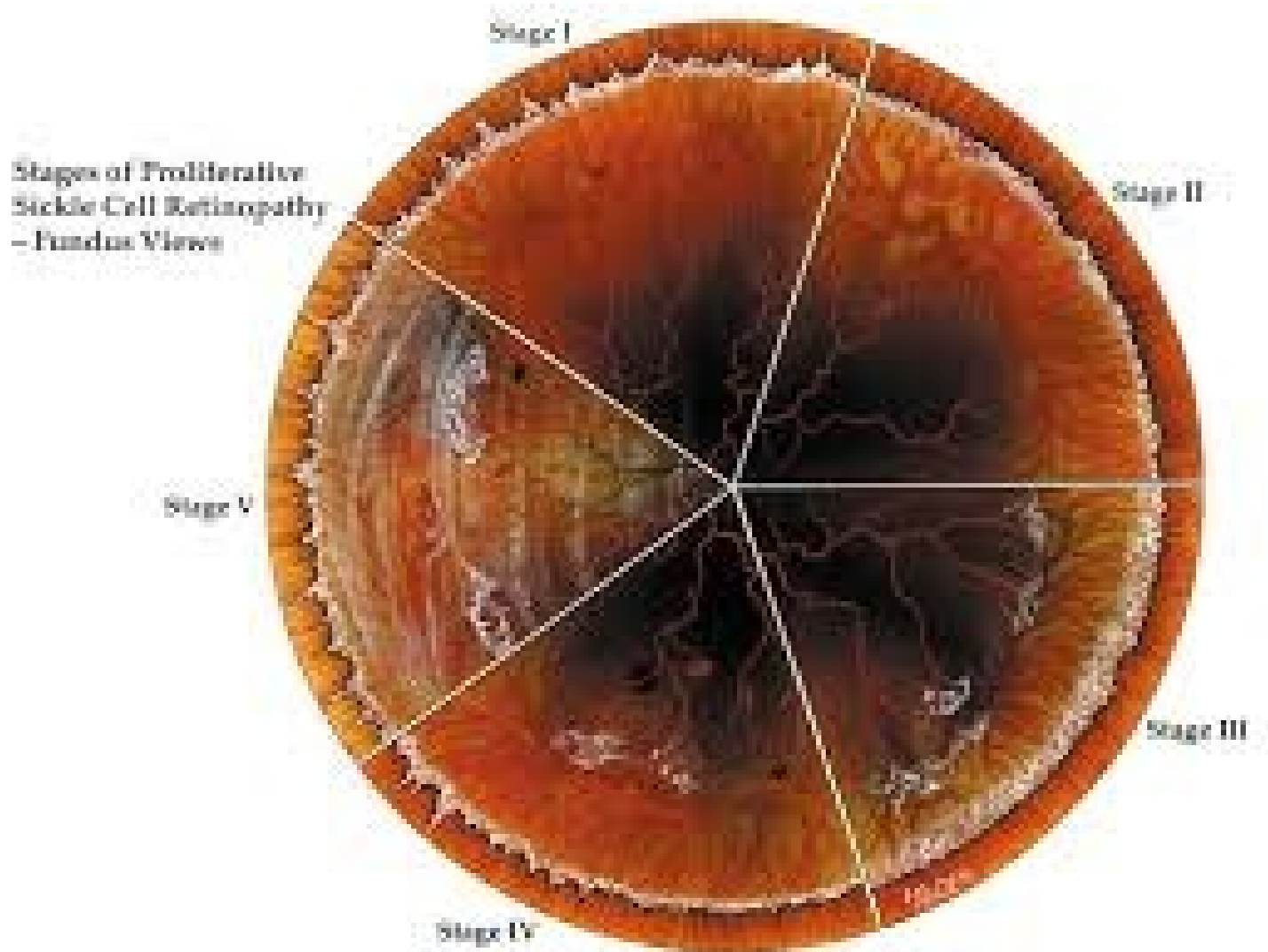
Sickle cell: PSR: TRD

Sickle-Cell Disease and the Eye



Sickle cell: PSR: Stages

Sickle-Cell Disease and the Eye



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Q

Sickle-Cell Disease and the Eye

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A

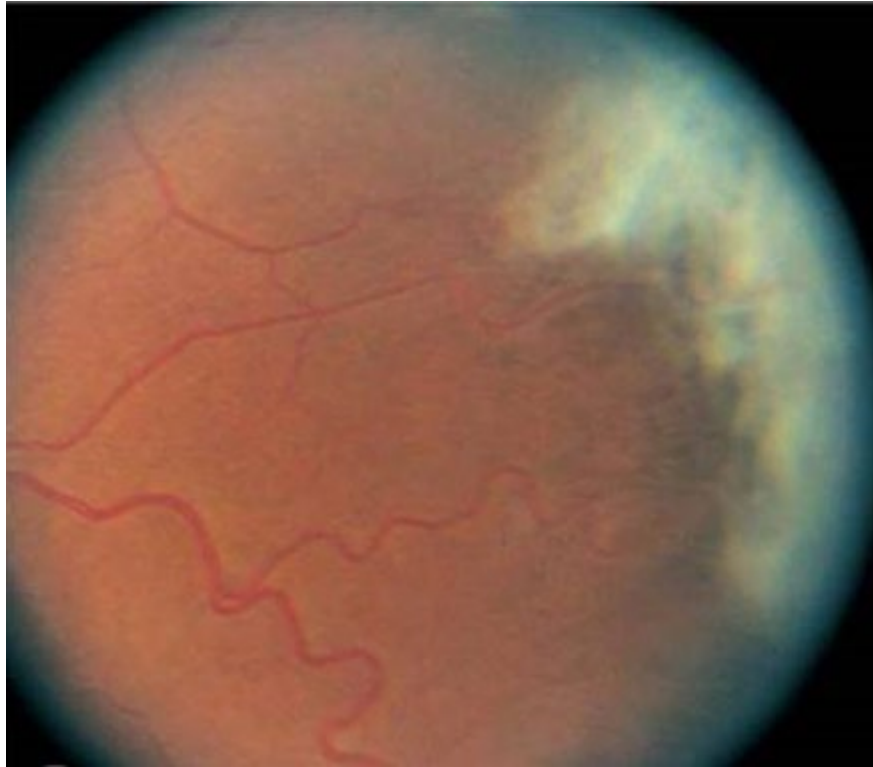
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Sickle-Cell Disease and the Eye



Autofluorescent sea fan neovascularization. The white appearance is classic.

Sickle cell: Autofluorescence



Sickle-Cell Disease and the Eye

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

- As in DBR, NPSR is located in the posterior pole.

- NPSR is more common in SC.
- PSR is more common in SC.

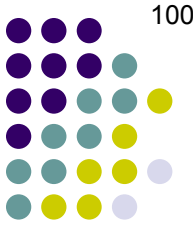
	DBR	SR
<i>Location</i>	Posterior to the equator (usually in the posterior pole)	Anterior to the equator (ie, peripherally)
<i>Proliferative lesions regress spontaneously?</i>	No	Yes

Another important difference

- Sea-fan lesions frequently **regress spontaneously** *True*

Q

Sickle-Cell Disease and the Eye



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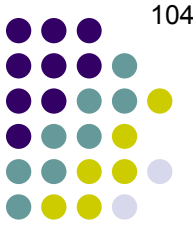
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The final stage of PSR is tractional RD, not rhegmatogenous. Why the concern over RRD?

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 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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False—treats

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

Another important difference

- 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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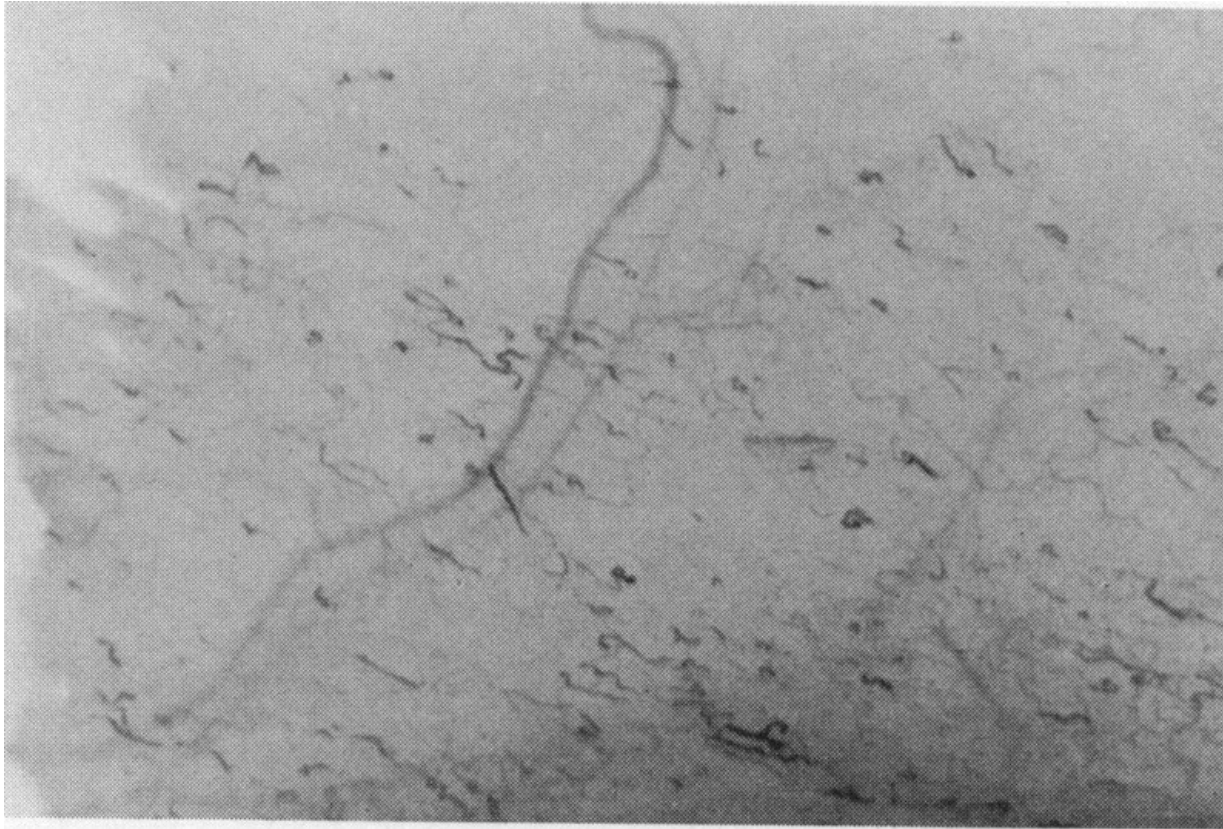
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Sickle-Cell Disease and the Eye



The *comma sign* of sickle-cell dz. Blocked small conj vessels are seen as comma-shaped lines.

Sickle cell: 'Comma sign'



Q

Sickle-Cell Disease and the Eye

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--**Comma sign:** Se *In what infectious condition is comma sign a well-known finding?* fornix

--**Disc sign**

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Sickle-Cell Disease and the Eye



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

Sickle cell: 'Disc sign'

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Sickle-Cell Disease and the Eye



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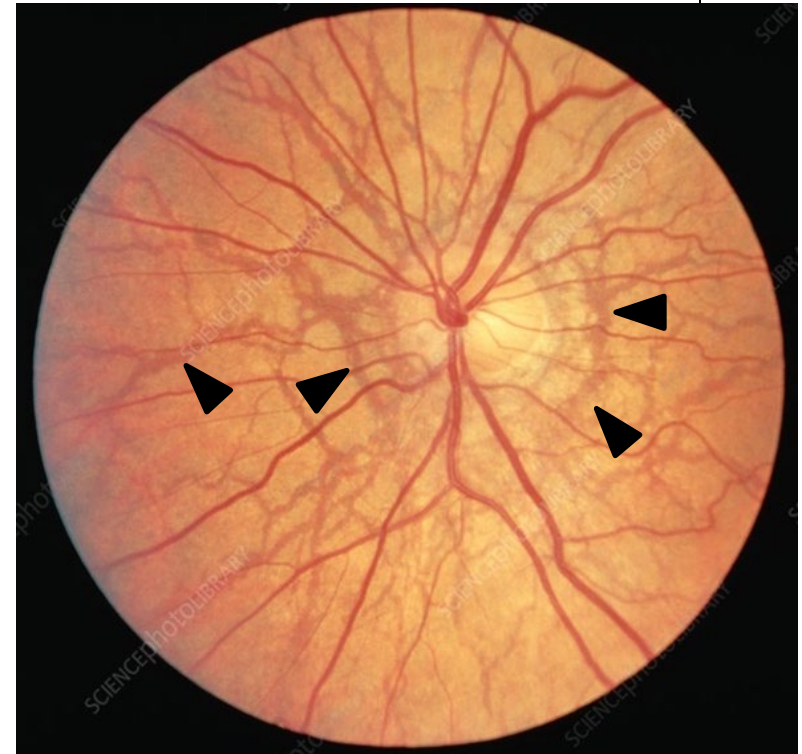
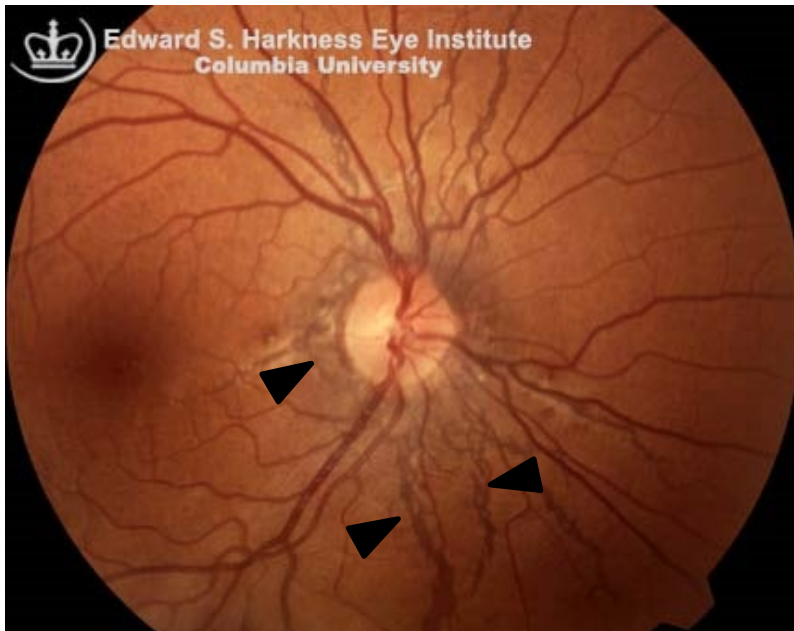
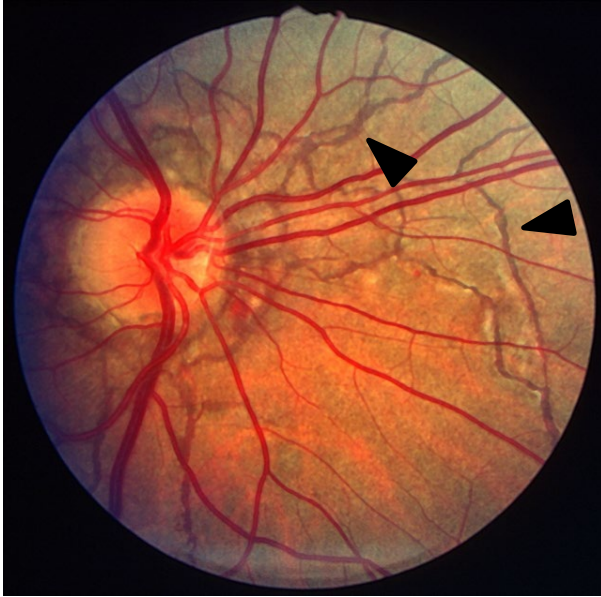
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Sickle-Cell Disease and the Eye



Angioid streaks (arrowheads).
Note that only a few of the many
present have been marked.

Q

Sickle-Cell Disease and the Eye



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--Comma sign: Seg

--Disc sign: Dark re

--Angioid streaks:

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

- Sea-fan les

- Laser phot

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--Comma sign: Segmental infarction

--Disc sign: Dark retinal disc

--**Angioid streaks:**

- Sea-fan lesions

- Laser photocoagulation

False—treat

not the feeder vessels

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

--P

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--Pseudoxanthoma elasticum

--Ehlers-Danlos dz

--Paget's dz of bone

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--Idiopathic

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Sickle-Cell Disease and the Eye

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For more on angioid streaks, see slide-set R61

- Comma sign: Segmental retinal depigmentation *What is the well-known mnemonic for the DDX for angioid streaks?*
- Disc sign: Dark retinal spots *What does each letter stand for (other than the 'S', duh)?*
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 - Laser photocoagulation *--Ehlers-Danlos dz*
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