

Q

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?

A

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic)
peripapillary region

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Angioid Streaks



- What is the classic DFE appearance of **angioid streaks**?
Reddish-brown lines radiating out from the (atrophic) peripapillary region

Why are they called 'angioid' streaks?

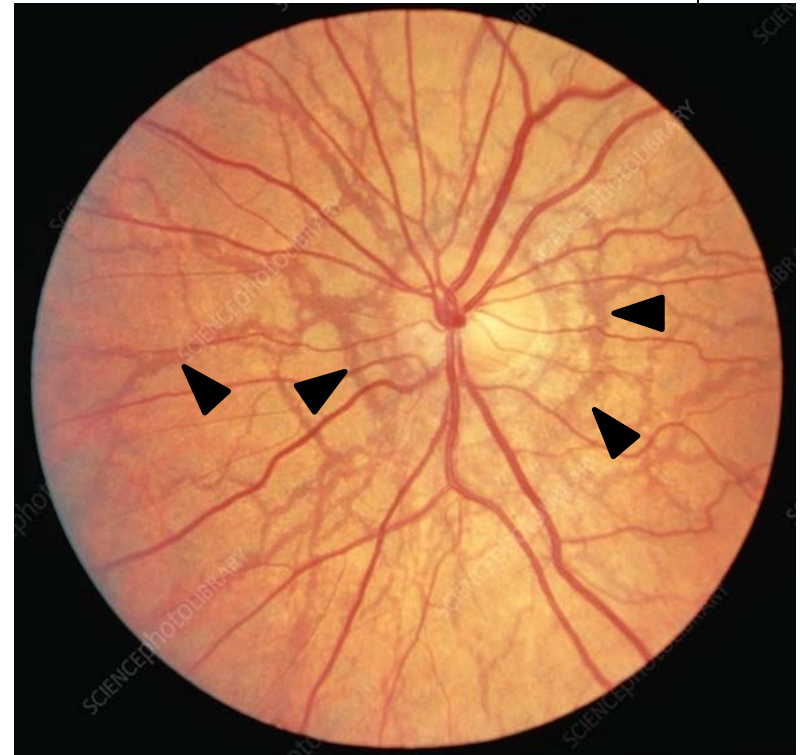
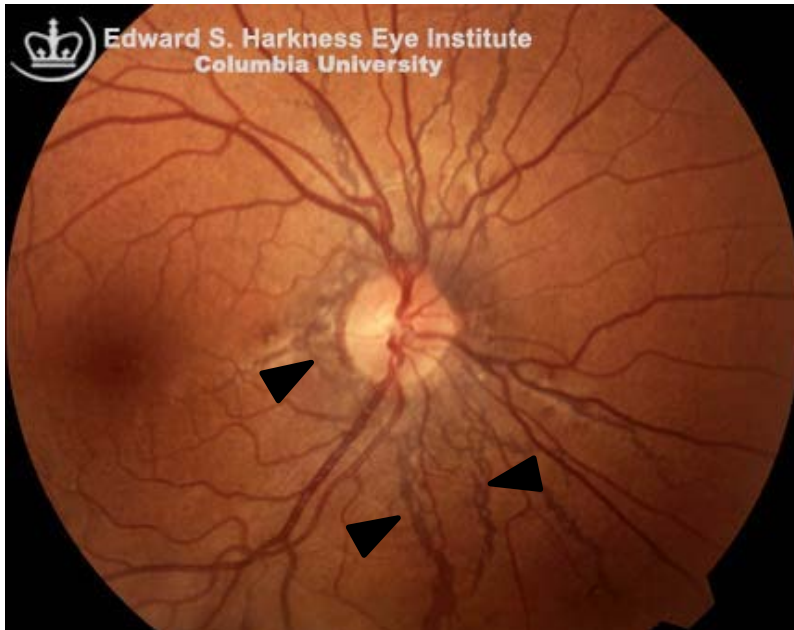
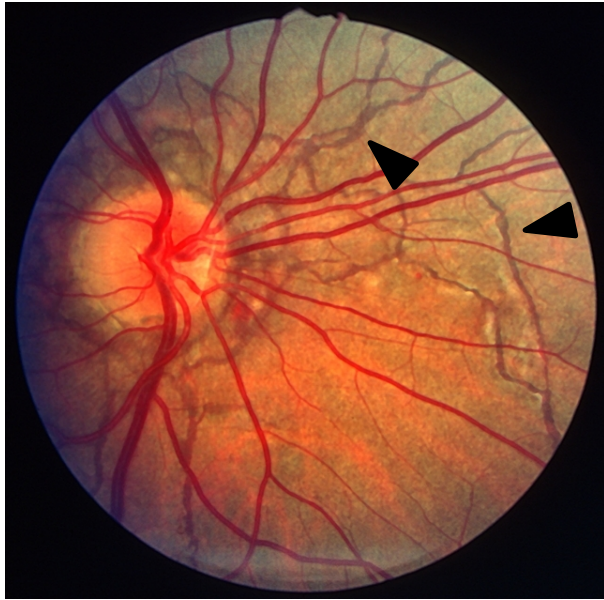


- What is the classic DFE appearance of **angioid streaks**?
Reddish-brown lines radiating out from the (atrophic) peripapillary region

Why are they called 'angioid' streaks?

Because the streaks resemble **blood vessels**

Angioid Streaks



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

Q

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic)
peripapillary region *bilaterally? unilaterally?*

Does angioid streaks present unilaterally, or bilaterally?

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Bilaterally

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With what symptom(s) does/do angioid streaks present?

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With what symptom(s) does/do angioid streaks present?

Nothing—it is asymptomatic (until/unless the fovea is involved)

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Nothing—it is asymptomatic (until/unless the fovea is involved)

Can angioid streaks occur congenitally?



- What is the classic DFE appearance of angioid streaks?
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Bilaterally

With what symptom(s) does/do angioid streaks present?

Nothing—it is asymptomatic (until/unless the fovea is involved)

Can angioid streaks occur congenitally?

Apparently not—there has never been a reported case of their presence in the fundus of an infant

Q

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal?

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Angioid Streaks



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- Which layer of the retina is abnormal? Bruch's membrane

Angioid Streaks

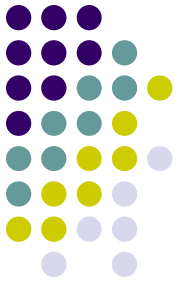


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- Which layer of the retina is abnormal? **Bruch's membrane**

Let's briefly review the anatomy of Bruch's membrane...

Q

Angioid Streaks



What are the five layers of Bruch's membrane?

Bruch's membrane

- 1) (Start here)
- 2)
- 3)
- 4)
- 5)

Innermost



Outermost



What are the five layers of Bruch's membrane?

Bruch's membrane

- 1) two words of RPE
- 2)
- 3)
- 4)
- 5)

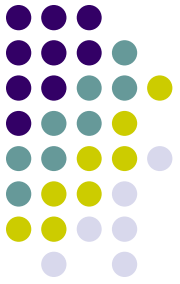
Innermost



Outmost

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Angioid Streaks



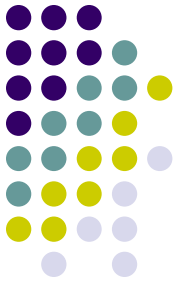
What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) (Next)
 - 3)
 - 4)
 - 5)

Innermost



Outermost



What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner one word layer
 - 3)
 - 4)
 - 5)

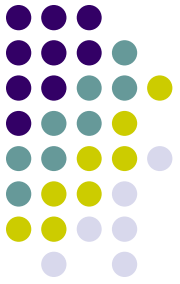
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Outmost

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Angioid Streaks



What are the five layers of Bruch's membrane?

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 - 2) Inner collagenous layer
 - 3) (Next)
 - 4)
 - 5)

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Outermost



What are the five layers of Bruch's membrane?

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 - 4)
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Innermost



Outmost

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Angioid Streaks



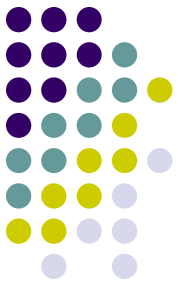
What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) (Next)
 - 5)

Innermost



Outermost



What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer one familiar word layer
 - 5)

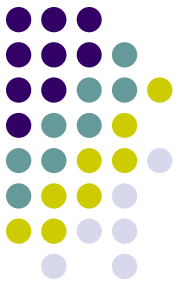
Innermost



Outermost

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Angioid Streaks



What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) (Next)

Innermost



Outermost



What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) two familiar words of choriocapillaris

Innermost



Outermost

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Angioid Streaks



What are the five layers of Bruch's membrane?

- Bruch's membrane
- 1) Basement membrane of RPE
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 - 5) Basement membrane of choriocapillaris

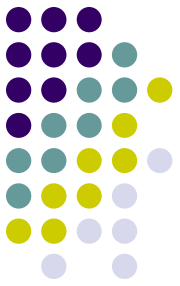
Innermost



Outermost

Q

Angioid Streaks



What are the five layers of Bruch's membrane?

O) ?

What (non-Bruch's) structure goes here?

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) Basement membrane of choriocapillaris

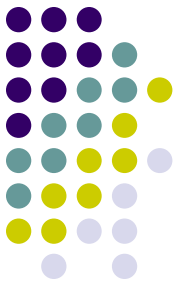
Innermost



Outermost

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Angioid Streaks



What are the five layers of Bruch's membrane?

0) *RPE cells*

What (non-Bruch's) structure goes here? **The RPE cells themselves**

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) Basement membrane of choriocapillaris

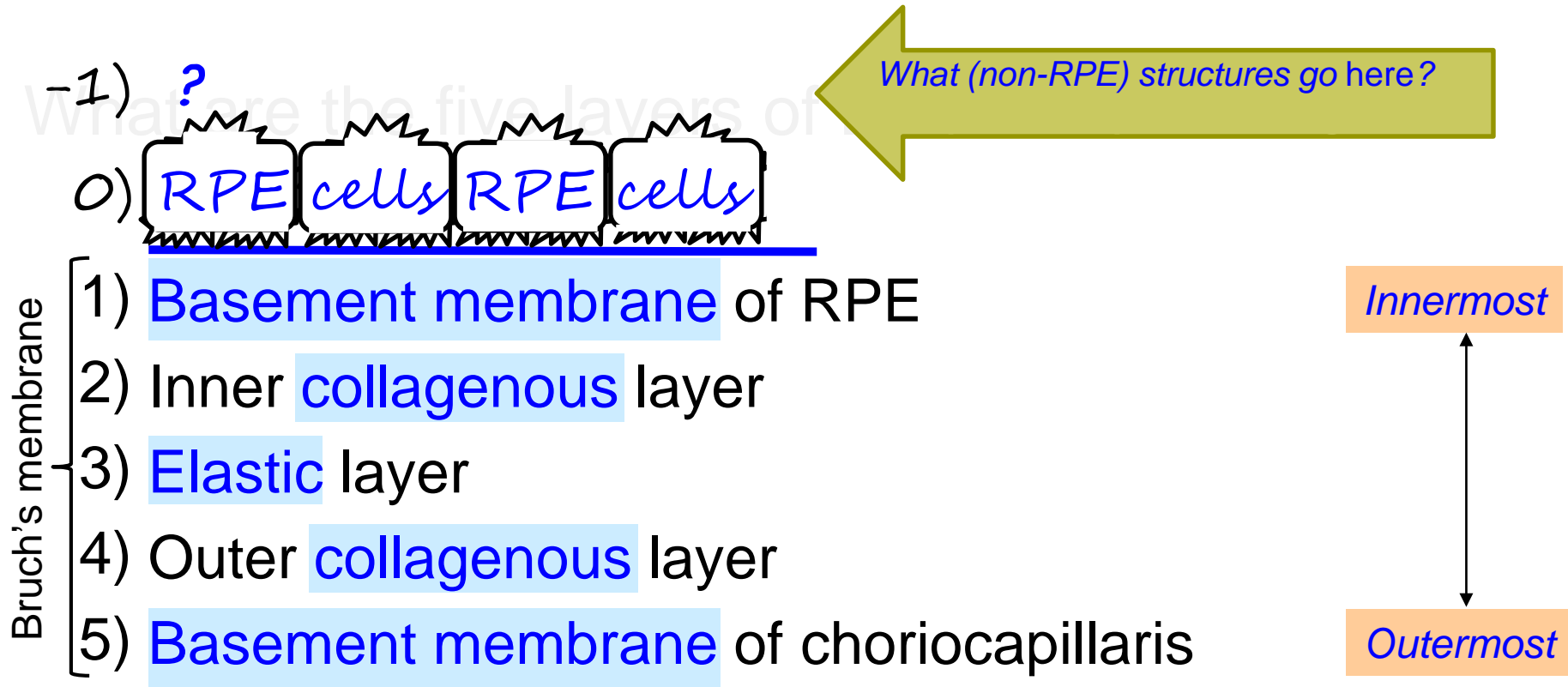
Innermost



Outermost

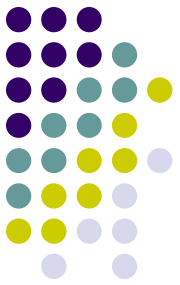
Q

Angioid Streaks



A

Angioid Streaks



-1) PR outer segs

0) RPE cells RPE cells

What (non-RPE) structures go here?
The photoreceptor outer segments

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) Basement membrane of choriocapillaris

Innermost

Outermost

Q

-2) ?

Angioid Streaks

What cell type is this?

-1) PR outer segs

0) RPE cells RPE cells

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) Basement membrane of choriocapillaris

Innermost

Outermost



A

Angioid Streaks

-2) Bipolar cells

What cell type is this?
Bipolar cells

-1) PR outer segs

0) RPE cells RPE cells

- Bruch's membrane
- 1) Basement membrane of RPE
 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
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Innermost

Outermost




Q

Angioid Streaks

-2) Bipolar cells

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- 1) Basement membrane of RPE
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 - 6)  ?

Innermost

Outermost

What structure is this?

A

Angioid Streaks

-2) Bipolar cells

-1) PR outer segs

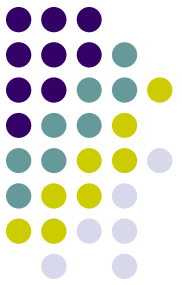
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 - 2) Inner collagenous layer
 - 3) Elastic layer
 - 4) Outer collagenous layer
 - 5) Basement membrane of choriocapillaris
 - 6) ~~Choriocapillaris~~ Choriocapillaris

Innermost

Outermost

What structure is this?
The choriocapillaris



Q

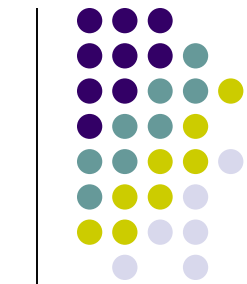
Angioid Streaks

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 - 6) ~~Choriocapillaris~~
 - 7) ~~?~~



Innermost

Outermost

What structure is this?

A

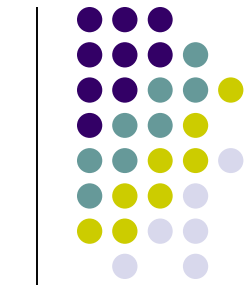
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 - 5) Basement membrane of choriocapillaris
 - 6) ~~Choriocapillaris~~
 - 7) ~~Choroid~~



Innermost

Outermost

What structure is this?
The choroid

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Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? **Bruch's membrane**

What's abnormal about Bruch's in angioid streaks?

--
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What's abnormal about Bruch's in angioid streaks?
--It is...[thickened vs thinned]
--It is...[common pathologic process]

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Angioid streaks represent breaks in the brittle Bruch's



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What serious complications can result from the breaks in Bruch's?

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What serious complications can result from the breaks in Bruch's?
Choroidal neovascular membrane and subretinal hemorrhage



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*To be clear: Does this refer to hemorrhage **not** related to a CNVM?*



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Yes



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Indeed it is—estimates of CNVM prevalence in angioid streaks run as high as %

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Does CNVM in angioid streaks carry a good, or poor prognosis?

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Poor

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Does the brittleness of **Bruch's**...
predispose these eyes to traumatic
choroidal rupture with subsequent **CNVM**
development?

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Are there steps the managing
ophthalmologist should take in this regard?

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predispose these eyes to traumatic
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development?*
Indeed it does

*Are there steps the managing
ophthalmologist should take in this regard?*
Encourage the use of safety glasses, and
the avoidance of risky activities

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- Which layer of the retina is abnormal? **Bruch's membrane**

What is the typical appearance of angioid streaks on **FA**?

What about Bruch's in angioid streaks?

(and thus brittle)

What is related to Bruch's?

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Poor



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? **Bruch's membrane**

What is the typical appearance of angioid streaks on **FA**?

The streaks **hypo- v hyperfluoresce** early due to a window defect, and late due to **mechanism**

about Bruch's in angioid streaks?

and thus **brittle**)

related to Bruch's?

Angioid streaks represent **breaks in the brittle Bruch's**

What serious complications can result from the breaks in Bruch's?

Choroidal neovascular membrane and subretinal hemorrhage

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Angioid Streaks



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What is the typical appearance of angioid streaks on **FA**?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

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and thus **brittle**)

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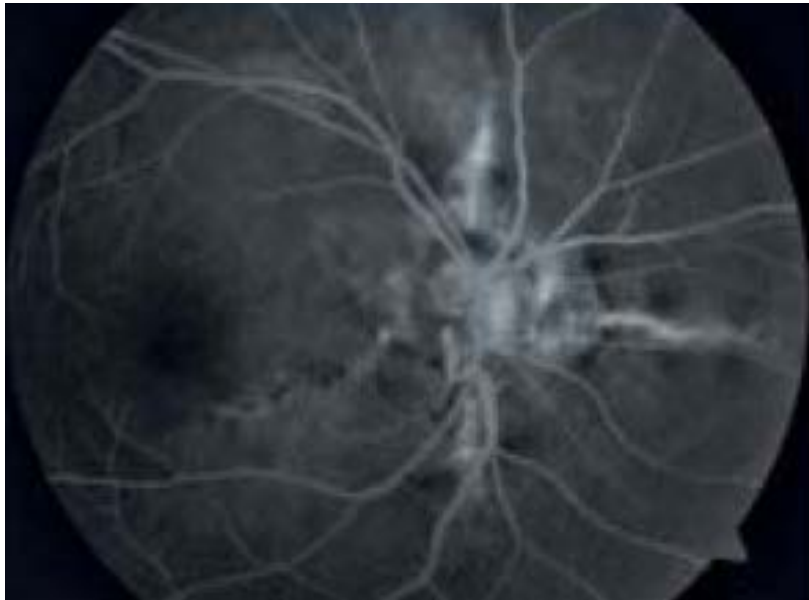
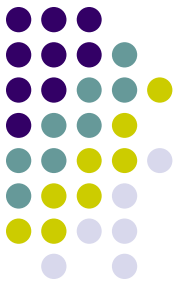
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Angioid Streaks



Early



Late

FA



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Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? **Bruch's membrane**

What is the typical appearance of angioid streaks on **FA**?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hypo- v hyperfluoresce** during the **late** phase

What about Bruch's in angioid streaks?

(and thus **brittle**)

What is related to Bruch's?

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Poor

Q

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

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- What is the typical appearance of angioid streaks on FA?
The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**
- Which layer of the retina is abnormal? **Bruch's membrane**

Reddish-brown, linear, branching, peripapillary region	Angioid streaks	Lacquer cracks
--	-----------------	----------------

What is the typical appearance of angioid streaks on FA?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase

What is the typical appearance of angioid streaks on FA?

and thus **brittle**)

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Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the typical appearance of angioid streaks on FA?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	?

- Which layer of the retina is abnormal? Bruch's membrane

What is the typical appearance of angioid streaks on FA?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase

What is the typical appearance of angioid streaks on DFE?

What is the typical appearance of angioid streaks on OCT?

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Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the typical appearance of angioid streaks on FA?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish

- Which layer of the retina is abnormal? **Bruch's membrane**

What is the typical appearance of angioid streaks on FA?
 The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**.
 Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase.

What is the typical appearance of angioid streaks on DFE?
 (about Bruch's in angioid streaks?
 and thus **brittle**)

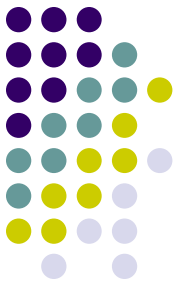
What is the typical appearance of angioid streaks on OCT?
 (related to Bruch's?
 Angioid streaks represent **breaks in the brittle Bruch's**)

What serious complications can result from the breaks in Bruch's?
Choroidal neovascular membrane and subretinal hemorrhage

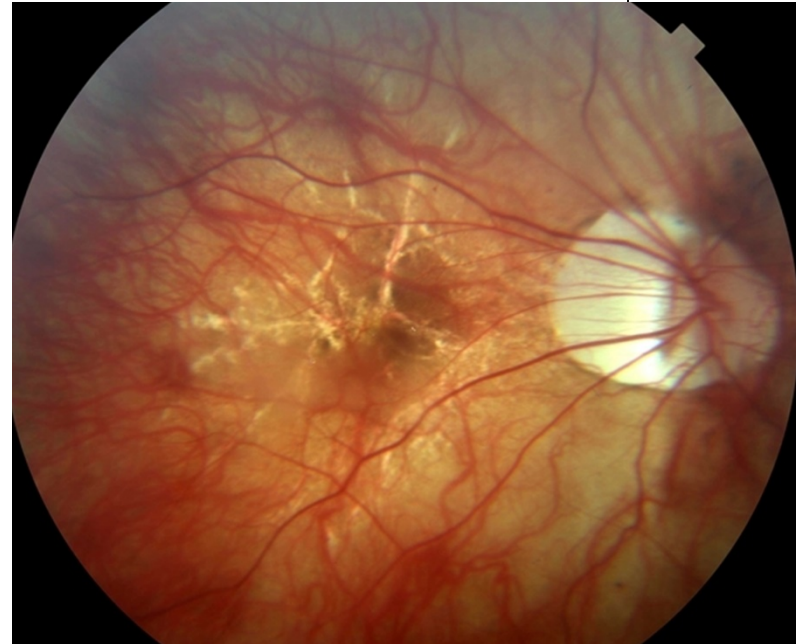
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Angioid Streaks



Angioid streaks:
'Reddish-brown'



Lacquer cracks:
'Yellowish'

Q

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What

Reddish
peripap

- Which

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	?

membrane

What is the typical appearance of angioid streaks on FA?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase

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Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What

Reddish
peripapillary

- Which

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula

retina

What is the typical appearance of angioid streaks on FA?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

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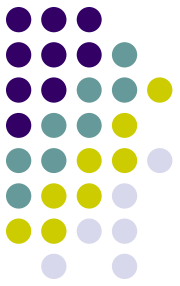
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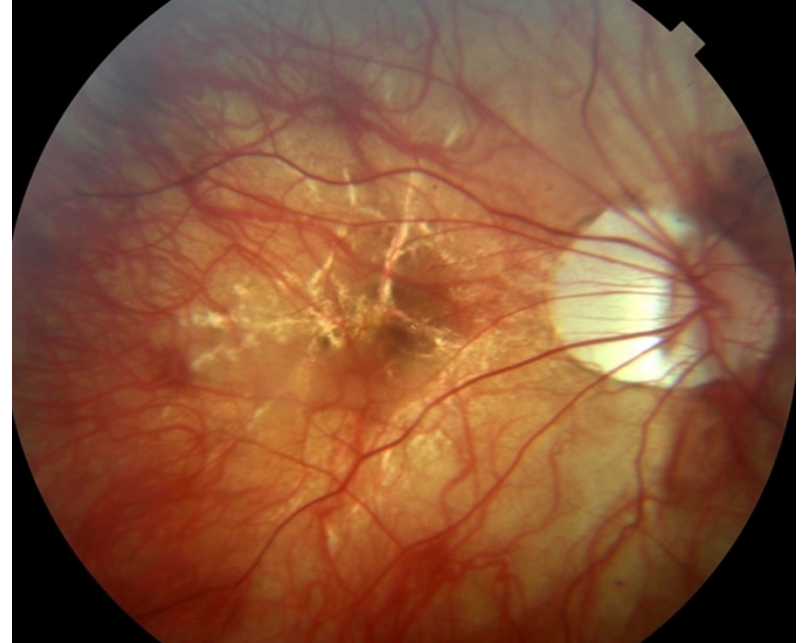
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Poor

Angioid Streaks



Angioid streaks:
'Reddish-brown'
'Peripapillary'



Lacquer cracks:
'Yellowish'
'Macular'

Q

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the typical appearance of angioid streaks on FA?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	?

Reddish-brown
peripapillary

- Which is more brittle?

What is the typical appearance of angioid streaks on FA?

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase

What is the typical appearance of angioid streaks on FA?

and thus **brittle**)

What is the typical appearance of angioid streaks on FA?

Angioid streaks represent **breaks in the brittle Bruch's**

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Does CNVM in angioid streaks carry a good, or poor prognosis?
Poor

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Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the typical appearance of angioid streaks on IFA?
- Which

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes

The streaks **hyperfluoresce** early due to a window defect, and late due to **staining**

Of course, if a CNVM develops, it will **hyperfluoresce** during the choroidal phase

about Bruch's in angioid streaks? (and thus brittle)

Angioid streaks represent **breaks in the brittle Bruch's**

What serious complications can result from the breaks in Bruch's? **Choroidal neovascular membrane** and subretinal hemorrhage

Is CNVM a common occurrence in angioid streaks? Indeed it is—estimates of CNVM prevalence in angioid streaks run as high as 80%

Does CNVM in angioid streaks carry a good, or poor prognosis? Poor

Q

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the type of finding on DFE?
- Which Bruch's-based condition is it?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	?

What is the type of finding on DFE?
The streaks hyperfluoresce and late due to staining

Of course, if a CNVM develops, it will hyperfluoresce during the choroidal phase

What is the type of finding on DFE?
The streaks hyperfluoresce and late due to staining

Angioid streaks represent breaks in the brittle Bruch's

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Poor

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Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the type of finding on DFE?

Reddish-brown peripapillary

- Which

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	Yes

membrane

What is the type of finding on DFE?

The streaks hyperfluoresce and late due to staining

Of course, if a CNVM develops, it will hyperfluoresce during the choroidal phase

What is the type of finding on DFE?

and thus brittle)

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Does CNVM in angioid streaks carry a good, or poor prognosis?
Poor

Q

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the type of finding on DFE?
- Which Bruch's-based condition is associated with angioid streaks?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	Yes
Associated with...	We'll see shortly	?

What is the type of finding on DFE? The streaks hyperfluoresce early and late due to

Of course, if a CNVM develops, it will hyperfluoresce during the choroidal phase

Angioid streaks represent breaks in the brittle Bruch's

What serious complications can result from the breaks in Bruch's? Choroidal neovascular membrane and subretinal hemorrhage

Is CNVM a common occurrence in angioid streaks? Indeed it is—estimates of CNVM prevalence in angioid streaks run as high as 80%

Does CNVM in angioid streaks carry a good, or poor prognosis? Poor

A

Angioid Streaks



There is another Bruch's-based condition that, similar to angioid streaks, presents with a linear finding on DFE. What is that condition?

- What is the typical color of angioid streaks?
- Which location is most common for angioid streaks?
- Which condition is associated with angioid streaks?

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	Yes
Associated with...	We'll see shortly	High/pathologic myopia

What is the typical color of angioid streaks? The streaks are typically reddish-brown and late due to

Of course, if a CNVM develops, it will hyperfluoresce during the choroidal phase

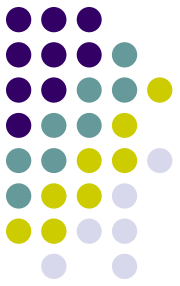
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Angioid Streaks



Angioid streaks:
'Reddish-brown'
'Peripapillary'



Lacquer cracks:
'Yellowish'
'Macular'
'High myopia' (note the optic disc)

Angioid Streaks

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	Yes
Associated with...	We'll see shortly	High/pathologic myopia

Review slide—no question

Angioid streaks represent breaks in the brittle Bruch's

What serious complications can result from the breaks in Bruch's?
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- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? **Bruch's membrane**
- What proportion of angioid streaks are associated with systemic abnormalities?

A

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? Bruch's membrane
- What proportion of angioid streaks are associated with systemic abnormalities? About half



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- *What is the well-known mnemonic for angioid streaks' associations?*
 -
 -
 -
 -
 -



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 - P
 - E
 - P
 - S
 - I



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 - P
 - E
 - P
 - S
 - I

A

Angioid Streaks



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- Which layer of the retina is abnormal? **Bruch's membrane**
- What proportion of angioid streaks are associated with systemic abnormalities? **About half**
- *What is the well-known mnemonic for angioid streaks' associations? What are the associations?*
 - **P**seudoxanthoma elasticum (PXE)
 - **E**hlers-Danlos syndrome
 - **P**aget's disease of bone
 - **S**ickle-cell disease
 - **I**diopathic (I know, it's not really an association)

Angioid Streaks



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 - **Sickle-cell disease**
 - **I**diopathic
- Mnemonic caveat #1:**
A number of hemoglobinopathies (eg, β -thal) besides sickle-cell dz are associated with angioid streaks

Angioid Streaks



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- **S**ickle-cell disease
- **I**diopathic

Mnemonic caveat #2:

The list of conditions that have been associated with angioid streaks is **much** longer than the PEPSI mnemonic suggests.
Caveat emptor.



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Of these, which has the strongest association with angioid streaks?



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 - Ehlers-Danlos syndrome
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 - Sickle-cell disease
 - **Idiopathic!**

Of these, which has the strongest association with angioid streaks?
Idiopathic



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 - **I**diopathic!

Of these, which has the strongest association with angioid streaks?

Idiopathic

What proportion of cases are idiopathic?



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Of these, which has the strongest association with angioid streaks?

Idiopathic

What proportion of cases are idiopathic?

About half (as implied above)



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 - **Idiopathic!**

Of these, which has the strongest association with angioid streaks?

Idiopathic

What should you do if you see an angioid-streak pt who does not carry the dx of a known associated systemic condition?



- What is the classic DFE appearance of angioid streaks?
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 - **Idiopathic!**
- Of these, which has the strongest association with angioid streaks?*
- Idiopathic**
- What should you do if you see an angioid-streak pt who does not carry the dx of a known associated systemic condition? Refer her for a workup, just to make certain*



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? Bruch's membrane
- What proportion of angioid streaks are associated with systemic abnormalities? About half
- *What is the well-known mnemonic for angioid streaks' associations? What are the associations?*
 - **P***pseudoxanthoma elasticum (PXE)?*
 - **E***hlers-Danlos syndrome?*
 - **P***aget's disease of bone?*
 - **S***ickle-cell disease?*
 - ~~I~~*diopathic*

Of the cases associated with a condition, which condition is most likely?



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*Of the cases associated with a condition, which condition is most likely?
PXE, by a mile*

Angioid Streaks



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 - Sickle-cell disease
 - ~~Idiopathic~~

Of the cases associated with a condition, which condition is most likely?

Paget's and sickle-cell are a distant second and third



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which condition is most likely?
PXE, by a mile



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Well over half
 - **P**seudoxanthoma
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which condition is most likely?
PXE, by a mile



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Of those angioid-streaks pts with a systemic association, what proportion have PXE?

Well over half

Of pts with PXE, what proportion have angioid streaks?

which condition is most likely?

PXE, by a mile

A

Angioid Streaks



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Of those angioid-streaks pts with a systemic association, what proportion have PXE?

Well over half

Of pts with PXE, what proportion have angioid streaks?
Essentially all of them, eventually

which condition is most likely?

PXE, by a mile

Q



Briefly, what sort of disorder is PXE?

- What are the clinical features of PXE? (e.g. skin, eyes, cardiovascular, renal, etc.)
- Which organ systems are affected by PXE?
- What is the inheritance pattern of PXE?
- What are the associated conditions of PXE?

Pseudoxanthoma elasticum (PXE)!

- Ehlers-Danlos syndrome
- Paget's disease of bone
- Sickle-cell disease
- ~~Idiopathic~~

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile



A

Briefly, what sort of disorder is PXE?

An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

- What are the clinical features of PXE? (e.g. skin, eyes, heart, kidneys, etc.)
- Which organs are most affected by PXE?
- What are the genetic causes of PXE?
- What are the systemic manifestations of PXE?
- What are the differential diagnoses for PXE?

Pseudoxanthoma elasticum (PXE)!

- Ehlers-Danlos syndrome
- Paget's disease of bone
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Of the cases associated with a condition, which condition is most likely?

PXE, by a mile



Q

Briefly, what sort of disorder is PXE?

An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

Is it common, or rare?

- What
- Redd
- peripa
- Which
- What
- syste
- What
- assoc

breaks?
(nic)

membrane
d with

breaks'

Pseudoxanthoma elasticum (PXE)!

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PXE, by a mile



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Is it common, or rare?

Rare

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PXE, by a mile



Q

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Is it common, or rare?

Rare

Is there a gender predilection?

Pseudoxanthoma elasticum (PXE)!

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Of the cases associated with a condition, which condition is most likely?

PXE, by a mile



Q/A

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Is it common, or rare?

Rare

Is there a gender predilection?

Yes, **MvF** are twice as likely to be affected

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- Ehlers-Danlos syndrome
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PXE, by a mile



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PXE, by a mile



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Is there a gender predilection?

Yes, ♀ are twice as likely to be affected

Other than the eye, what organ-systems are affected?

--

--

--

--Eye

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

Paget's disease of bone

Sickle-cell disease

~~Idiopathic~~

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile



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Other than the eye, what organ-systems are affected?

--Skin

--Vascular system

--GI tract

--Eye

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Rare

Is there a gender predilection?

Yes, ♀ > ♂

What is the appearance of affected skin?

Other than

Skin

--Vascula

--GI tract

--Eye

Pseudoxanthoma elasticum (PXE)!

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Is there a gender predilection?

Yes, ♀ > ♂

What is the appearance of affected skin?

An area of waxy-yellow, papule-like lesions

Other than

--Skin

--Vascula

--GI tract

--Eye

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

Paget's disease of bone

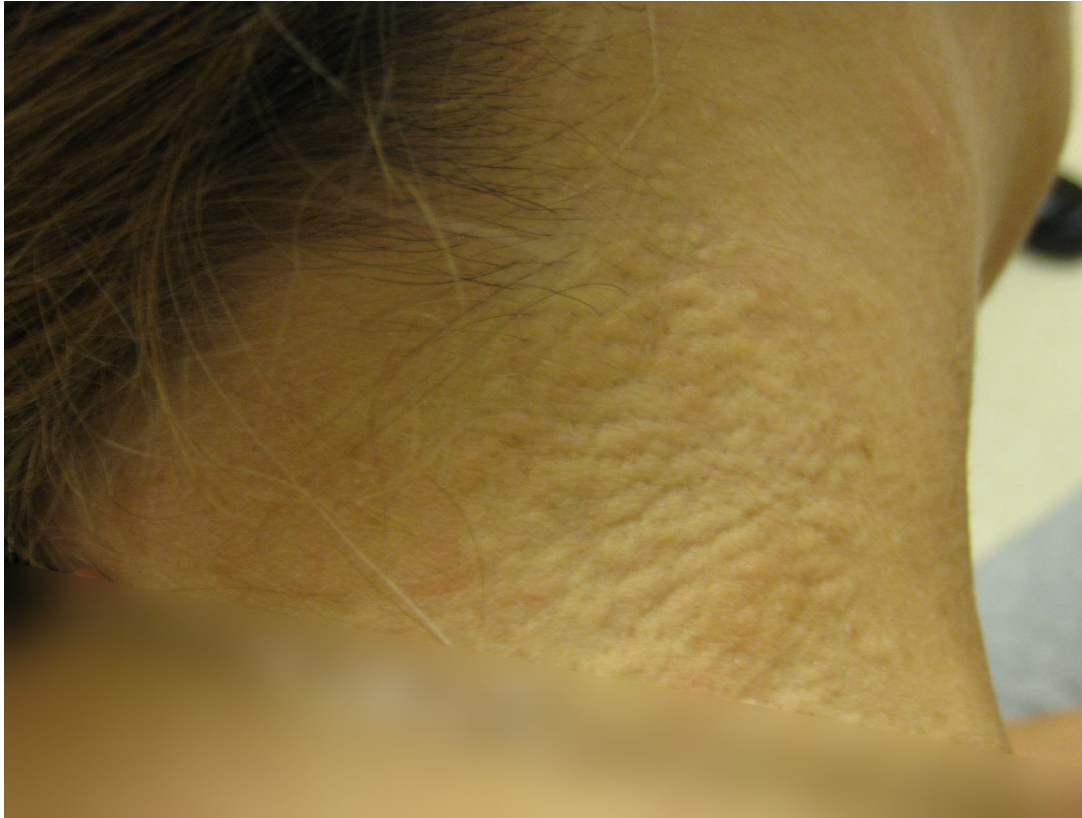
Sickle-cell disease

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Of the cases associated with a condition, which condition is most likely?

PXE, by a mile

Angioid Streaks



PXE skin



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Rare

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Yes, ♀ > ♂

What is the appearance of affected skin?

An area of waxy-yellow, papule-like lesions

Other than skin

--Skin

--Vascula

--GI tract

--Eye

What is the classic informal descriptor for this appearance?

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

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What is the appearance of affected skin?

An area of waxy-yellow, papule-like lesions

Other than

Skin

What is the classic informal descriptor for this appearance?

'Chicken skin'

--Vascula

--GI tract

--Eye

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--Eye

In what two locations is 'chicken skin' most often found?

--

--

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Other than

Skin

What is the classic informal descriptor for this appearance?

'Chicken skin'

--Vascula

--GI tract

--Eye

In what two locations is 'chicken skin' most often found?

--The neck

--The axillae

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

Paget's disease of bone

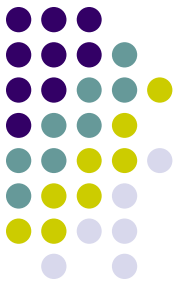
Sickle-cell disease

~~Idiopathic~~

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile

Angioid Streaks



PXE: 'Chicken skin'



Q

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Is it common, or rare?

Rare

Is there a gender predilection?

Yes, ♀ are twice as likely to be affected

Other than the eye, what organ-systems are affected?

--Skin

--**Vascular system**

--GI tract

--Eye

How does PXE affect the vascular system?

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

Paget's disease of bone

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How does PXE affect the vascular system?

In a number of ways; the two mentioned in the *Retina* book are calcific coronary artery dz, and an increased risk of CVA

Pseudoxanthoma elasticum (PXE)!

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--Eye

What is the classic GI manifestation of PXE?

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

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Sickle-cell disease

~~Idiopathic~~

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Other than the eye, what organ-systems are affected?

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--GI tract

--Eye

What is the classic GI manifestation of PXE?

Hemorrhage, which is common, and life-threatening in severity

Pseudoxanthoma elasticum (PXE)!

Ehlers-Danlos syndrome

Paget's disease of bone

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--Skin

--Vascular

--GI tract

Eye

There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?

--Angioid streaks

--

--

Pseudoxanthoma

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PXE, by a mile



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--Skin

--Vasculature

--GI tract

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There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?

--Angioid streaks

--RPE mottling

--Optic disc drusen

Pseudoxanthoma

Ehlers-Danlos syndrome

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PXE, by a mile



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--RPE mottling

--Optic disc drusen

What mellifluous name is used to describe the RPE mottling?

Pseudoxanthoma

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile

● Ehlers-Danlos syndrome

● Paget's disease of bone

● Sickle-cell disease

● ~~Idiopathic~~



A

Briefly, what sort of disorder is PXE?

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Is it common, or rare?

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--Skin

--Vasculature

--GI tract

--**Eye**

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--Angioid streaks

--**RPE mottling**

--Optic disc drusen

What mellifluous name is used to describe the RPE mottling?

Peau d'orange

Pseudoxanthoma

Ehlers-Danlos syndrome

Paget's disease of bone

Sickle-cell disease

~~Idiopathic~~

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile



An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

Is there a gender predilection?

Other than the eye, what organ-systems are affected?

--Vascula

--GI tract

There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?

--Android strokes

RPE mottling

--Optic disc druse

What mellifluous name is used to describe the RPE motting?

Peau d'orange

What does peau d'orange mean in English?

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile

Pseudoxan

Ehlers-Danlos syndrome

Paget's disease of bone

Sickle-cell disease

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QA

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Is there a gender predilection?

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Other than the eye, what organ-systems are affected?

--Skin

--Vascular

--GI tract

--Eye

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--RPE mottling

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Peau d'orange

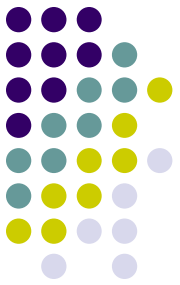
What does peau d'orange mean in English? 'Orange peel'--refers to a fine stippling like that found on the skin of an orange

Of the cases associated with a condition, which condition is most likely?

PXE, by a mile

- What Redd peripa
- Which
- What syste
- What assoc
- Pseudoxan
- Ehlers-Danlos syndrome
- Paget's disease of bone
- Sickle-cell disease
- Idiopathic

Angioid Streaks



PXE: *Peau d'orange* fundus

Q

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? Bruch's membrane
- What proportion of angioid streaks are associated with systemic abnormalities? About half

- *What is the well-known m associations? What are th*
In two, hyphenated words, what sort of disorder is Ehlers-Danlos?

It is a disorder

- Pseudoxanthoma elasticum (P)

- **Ehlers-Danlos syndrome**

- Paget's disease of bone

- Sickle-cell disease

- ~~Idiopathic~~

A

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It is a connective-tissue disorder

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Briefly, what are its main nonocular findings?

--
--
--

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Briefly, what are its main nonocular findings?

--Fragile

-- joints

--Easy

two related words

• Pseudoxanthoma elasticum (P)

• **Ehlers-Danlos syndrome**

• Paget's disease of bone

• Sickle-cell disease

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A

Angioid Streaks



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 - **Ehlers-Danlos syndrome**
 - Paget's disease of bone
 - Sickle-cell disease
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In two, hyphenated words, what sort of disorder is Ehlers-Danlos?

It is a connective-tissue disorder

Briefly, what are its main nonocular findings?

--Fragile skin

-- Hyperextensible joints

--Easy bruising/bleeding

Angioid Streaks



Ehlers-Danlos syndrome

Q

Angioid Streaks



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--Fragile skin
-- Hyperextensible joints
-- Easy **bruising/bleeding**

What colorful eye finding is associated with Ehlers-Danlos?

- Pseudoxanthoma elasticum (P)

- **Ehlers-Danlos syndrome**

- Paget's disease of bone

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In two, hyphenated words, what sort of disorder is Ehlers-Danlos?

It is a connective-tissue disorder

Briefly, what are its main nonocular findings?

--Fragile skin
-- Hyperextensible joints
-- Easy **bruising/bleeding**

What colorful eye finding is associated with Ehlers-Danlos?

Blue



A

Angioid Streaks



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is abnormal? Bruch's membrane
- What proportion of angioid streaks are associated with systemic abnormalities? About half

- *What is the well-known m... associations? What are th...*

In two, hyphenated words, what sort of disorder is Ehlers-Danlos?

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Blue sclera

- Pseudoxanthoma elasticum (P)

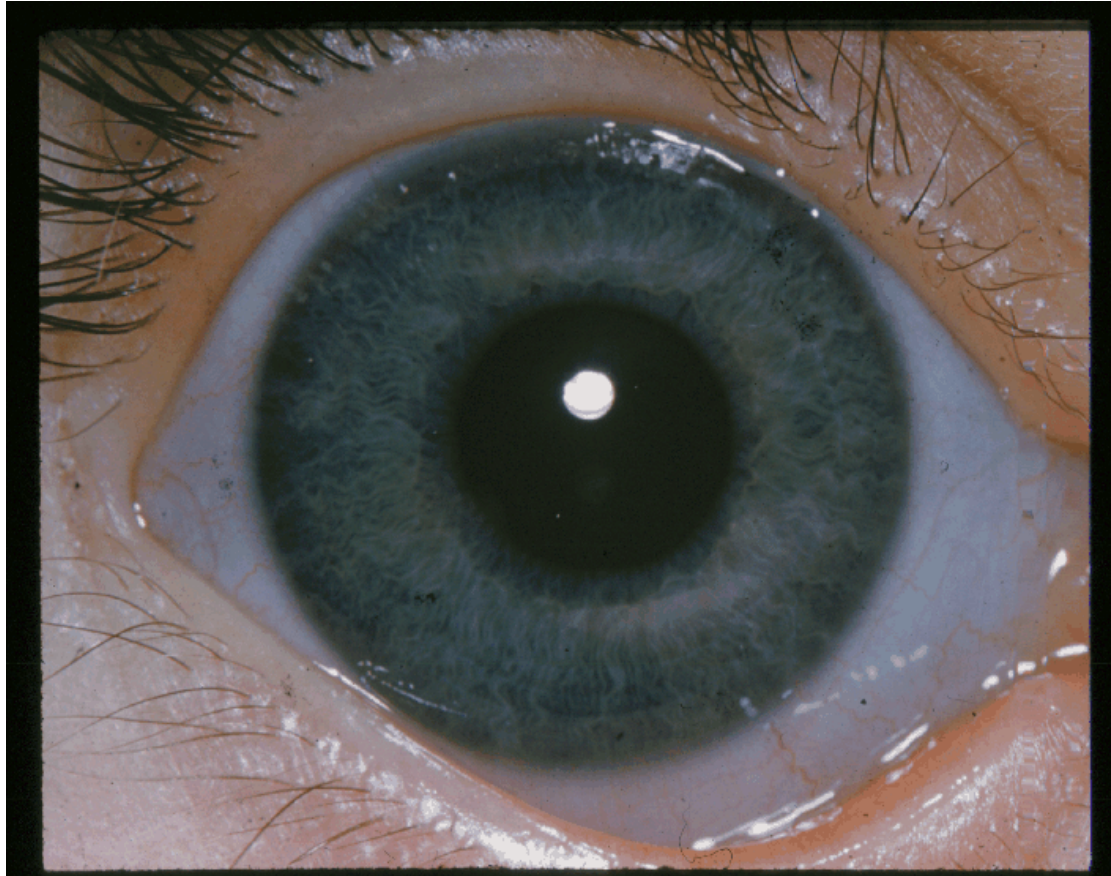
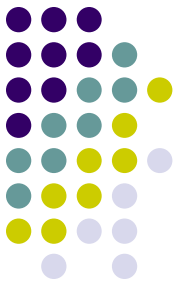
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- Paget's disease of bone

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Angioid Streaks



Blue sclera in Ehlers-Danlos



- What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the (atrophic) peripapillary region
- Which layer of the retina is involved?
- What proportion of angioid streaks are associated with systemic abnormalities? About 50%
- What is the well-known mnemonic for the systemic associations? What are the associations?
 - Pseudoxanthoma elasticum (PXE)
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Is Paget's a common, or uncommon condition?



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Is Paget's a common, or uncommon condition?
Quite common; it is the most common bone disease after [redacted]



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Increased osteoclastic and osteoblastic activity lead to focal abnormal bone structure



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It is usually asymptomatic, but can present with skeletal deformities pain



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As noted previously, essentially all PXE pts develop angioid streaks. Is the same true of Paget's pts?

No--only ~10% will develop angioid streaks



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Broadly speaking, what sort of disease is sickle-cell anemia?
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- What is the association with Ehler-Danlos syndrome?
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--SS

--SC

--S-Thal

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What are the forms of sickle-cell anemia?

--SS
--SC

What is the incidence of significant retinopathy in:

--SS dz? 3%

--SC dz? 33%

S-Thal vs SA disease?

whereas SA is an
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--S-Thal
--SA

What percent of African-Americans test positive for sickle trait?
8% (1 in 12)

In America, people of which two ethnic heritages are at greatest risk?
African-American; Hispanic-American



Sickle-cell retinopathy comes in two basic forms—what are they?

- What is sickle cell retinopathy? How does it come in two basic forms - what are they?
- Recurrent retinopathy
- What is the incidence of sickle cell retinopathy?
- What is the incidence of sickle cell retinopathy in the S-Thal vs SA disease?
 - What is the incidence of significant retinopathy in:
 - --SS dz? 3%
 - --SC dz? 33%
 - --S-Thal vs SA disease?
 - whereas SA is an 'inherited trait'
- What is the incidence of sickle cell retinopathy in African-Americans?
 - What percent of African-Americans test positive for sickle trait?
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- In America, people of which two ethnic heritages are at greatest risk?
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- Sickle-cell retinopathy comes in two basic forms—what are they?*
Nonproliferative (NPSR), and proliferative (PSR)

What are



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Angioid Streaks



Sickle-cell retinopathy comes in two basic forms—what are they?
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Nonproliferative and proliferative—just like diabetic retinopathy?
Indeed

What are the frequencies of S-Thal vs SA disease?
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- What are the two basic forms of sickle-cell retinopathy? Nonproliferative (NPSR), and proliferative (PSR)
- Recurrence of sickle-cell retinopathy? Nonproliferative and proliferative—just like diabetic retinopathy? Indeed
- What is the relationship between sickle-cell retinopathy and DBR? Like DBR, does sickle retinopathy tend to occur in the posterior pole?
- What is the relationship between sickle-cell retinopathy and the retinal vascular system?

- What is the incidence of significant retinopathy in:
 - SS
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What is the incidence of significant retinopathy in: S-Thal vs SA disease?
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- Pseudoexfoliation syndrome
- Ehlers-Danlos syndrome
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Angioid Streaks



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Recall: Nonproliferative and proliferative—just like diabetic retinopathy? Indeed
- What is the distribution of sickle retinopathy? Like DBR, does sickle retinopathy tend to occur in the posterior pole? No—unlike DBR, sickle retinopathy tends to occur well into the periphery (usually anterior to the equator)
- What is the incidence of significant retinopathy in: S-Thal vs SA disease? --SS dz? 3% --SC dz? 33% whereas SA is an 'trait'
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- What are the two basic forms of sickle-cell retinopathy? Nonproliferative (NPSR) and proliferative (PSR)

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

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- --Salmon patches
- --Refractile spots
- --Sunburst lesions

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- NPSR manifests as three lesions. What are they? What is their appearance?
- --**Salmon patches**: Retinal hemorrhage trapped under the internal limiting membrane
- --**Refractile spots**: Iridescent retinal areas
- --**Sunburst lesions**: Hyperpigmented patches, usually somewhat stellate

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What is the pathogenesis of PSR?

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What is the pathogenesis of PSR?

As with NPSR, it's vascular occlusion, but severe enough to produce significant ischemia

Like

No-

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Sickle-cell disease

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Nonproliferative (**NPSR**), and proliferative (**PSR**)

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

--SS
--SC

--SS dz? 3%

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whereas SA is an
trait'

--S-Thal

--SA

What percent of African-Americans test positive for sickle trait?
8% (1 in 12)

In America, people of which two ethnic heritages are at greatest risk?
African-American; Hispanic-American

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~~Idiopathic~~



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No, they may spontaneously regress, and laser may precipitate a rhegmatogenous RD

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Angioid Streaks



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

Sickle-cell disease

Review slide—no questions