CNVM DDx:
- ARMD
- OHS
- Pathologic myopia
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- Sorsby macular dystrophy (SMD)
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- **ARMD**
  
  ARMD is addressed extensively in a series of slide-sets—see the Table of Contents
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What does OHS stand for in this context?

Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

Is there a gender predilection in OHS?
No

Is there a racial predilection?
Yes, OHS occurs almost exclusively among whites of Northern European heritage.

Is there a geographic predilection?
Yes, the majority of cases are found in people who reside in the Mississippi/Ohio River valleys of the US.

Does OHS manifest unilaterally, or bilaterally?
Bilaterally (although it can be somewhat asymmetric).

Is OHS associated with vitritis?
Never. If vitritis is present, it’s not OHS.

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In a nutshell, what are histo spots?
Discrete, focal, atrophic scars.

What are you looking for on DFE again?
Histo spots.

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- Histospots

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Where are they typically located?
They can be anywhere in the posterior pole, ie, macula or near-to-mid periphery.

Are they usually larger, or smaller than the ONH?
Smaller.

What two-word phrase is used to describe them?
‘Punched out’

Do they evolve over time?
Generally no.

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</tr>
</thead>
<tbody>
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</tr>
<tr>
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What about Iatrogenic?
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Cystoid macular edema, aka DME

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In another nutshell, what is PPA?
Like histo spots, PPA represent atrophic scars.

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What about AC cell? Never. If AC cell is present, it’s not OHS.
CNVM DDx

Active lesion

Disciform scar

OHS
What does OHS stand for in this context?
Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

How is the diagnosis of OHS made?
It is a clinical diagnosis based on DFE findings.

What are you looking for on DFE?
The so-called ‘classic triad’ of OHS.

What is the classic triad of OHS?
--Histo spots?
--Peripapillary atrophy?
--Disciform macular lesion(s)?

In the last nutshell, what are disciform lesions?
--Active lesions represent either the presence of CNVM under the retina, or a hemorrhagic retinal detachment.
--Inactive lesions (aka disciform scars) are fibrovascular remnants of previous CNVM and/or subretinal hemorrhage.

Which lesion(s) require treatment?
Only active disciform lesions.

What treatment modalities are used to treat active disciform lesions?
--Thermal laser
--Photodynamic therapy (PDT)
--Anti-VEGF therapy
--Submacular surgery
--Intravitreal corticosteroids
--Combination therapy (of some of the above modalities)
What does OHS stand for in this context?
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How is the diagnosis of OHS made?
It is a clinical diagnosis based on DFE findings.

What are you looking for on DFE?
The so-called ‘classic triad’ of OHS
--Histospots
--Peripapillary atrophy
--Disciform macular lesion(s)

What is the classic triad of OHS?

Which lesion(s) require treatment?
Only active disciform lesions

What about AC cell?
Never.

In the last nutshell, a summary:
Active lesions:
--Histospots
--Peripapillary atrophy
--Disciform macular lesion(s)

Inactive lesions:
--Histospots
--Peripapillary atrophy

Which lesion(s) require treatment?
Only active disciform lesions

What about AC cell?
Never.

In the last nutshell, a summary:
Active lesions:
--Histospots
--Peripapillary atrophy
--Disciform macular lesion(s)

Inactive lesions:
--Histospots
--Peripapillary atrophy
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The so-called ‘classic triad’ of OHS.

**What is the classic triad of OHS?**
--Histospots
--Peripapillary atrophy
--Disciform macular lesion(s)

**Which lesion(s) require treatment?**
Only active disciform lesions.

**What treatment modalities are used to treat active disciform lesions?**
Never.
What does OHS stand for in this context? Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

How is the diagnosis of OHS made? It is a clinical diagnosis based on DFE findings.

What are you looking for on DFE? The so-called ‘classic triad’ of OHS.

What is the classic triad of OHS? Histospots, peripapillary atrophy, disciform macular lesion(s).

In the last nutshell, active lesions are those of hemorrhage, retinal detachment, or a homogenous retinal scar.

Inactive lesions (aka disciform scars) are fibrovascular remnants of previous CNVM and/or subretinal hemorrhage.

Which lesion(s) require treatment? Only active disciform lesions.

What treatment modalities are used to treat active disciform lesions? Thermal laser, photodynamic therapy (PDT), anti-VEGF therapy, submacular surgery, intravitreal corticosteroids, combination therapy (of some of the above modalities).
**CNVM DDx**

**OHS**

*What does OHS stand for in this context?*
Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

*How is the diagnosis of OHS made?*
It is a clinical diagnosis based on DFE findings.

*What are you looking for on DFE?*
The so-called 'classic triad' of OHS:

- Histospots
- Peripapillary atrophy
- Disciform macular lesion(s)

*What is the classic triad of OHS?*

- Histospots
- Peripapillary atrophy
- **Disciform macular lesion(s)**

*In the last nutshell, which lesion(s) require treatment?*
Only active disciform lesions.

*Which lesion(s) require treatment?*

- Active lesions represent either the presence of CNVM under the retina, or a hemorrhagic retinal detachment.
- Inactive lesions (aka disciform scars) are fibrovascular remnants of previous CNVM and/or subretinal hemorrhage.

*What treatment modalities are used to treat active disciform lesions?*
- Thermal laser
- Photodynamic therapy (PDT)
- Anti-VEGF therapy
- Submacular surgery
- Intravitreal corticosteroids
- Combination therapy (or some of the above modalities)

*Do antifungals play a role in the treatment of OHS?*
**No**
What does OHS stand for in this context?
Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

How is the diagnosis of OHS made?
It is a clinical diagnosis based on DFE findings.

What are you looking for on DFE?
The so-called ‘classic triad’ of OHS:

- Histospots
- Peripapillary atrophy
- Disciform macular lesion(s)

What is the classic triad of OHS?

- Inactive lesions (aka disciform scars) are...fibrovascular remnants of previous CNVM and/or subretinal hemorrhage

Which lesion(s) require treatment?
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What treatment modalities are used to treat active disciform lesions?

- Thermal laser
- Photodynamic therapy (PDT)
- Anti-VEGF therapy
- Submacular surgery
- Intravitreal corticosteroids
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Do antifungals play a role in the treatment of OHS?
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What are you looking for on DFE?
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In the last nutshell, which lesion(s) require treatment?
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Which treatment modalities are used to treat active disciform lesions?
--Thermal laser
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Do antifungals play a role in the treatment of OHS?
No.

For a closer look at OHS, see slide-set U21.

In the last nutshell, which lesion(s) require treatment?
Only active disciform lesions.

What treatment modalities are used to treat active disciform lesions?
--Thermal laser
--Photodynamic therapy (PDT)
--Anti-VEGF therapy
--Submacular surgery
--Intravitreal corticosteroids
--Combination therapy (of some of the above modalities)

Do antifungals play a role in the treatment of OHS?
No.
What is the classic DFE appearance of angioid streaks?

Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch's membrane.
Q/A

- **CNVM DDx:**
  - ARMD
  - POHS
  - **Angioid streaks**

---

**What is the classic DFE appearance of angioid streaks?**

Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch's membrane.

**What proportion of angioid streaks are associated with systemic abnormalities?**

About half.

**What is the well-known mnemonic for angioid streak's associations? What are these associations?**

- PXE (Pseudoxanthoma elasticum)
- Ehlers-Danlos syndrome
- Paget's disease of bone
- Sickle-cell disease
- Idiopathic
What is the classic DFE appearance of angioid streaks?

**Reddish-brown** lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.
Angioid streaks (arrowheads). Note that only a few of the many present have been marked.
What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane

What proportion of angioid streaks are associated with systemic abnormalities?
What is the classic DFE appearance of angioid streaks?

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About half

What is the well-known mnemonic for angioid streak’s associations?
• CNVM DDx:
  • ARMD
  • POHS
  • Angioid streaks

What is the classic DFE appearance of angioid streaks?
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What is the well-known mnemonic for angioid streak’s associations?
P E P S I
What is the classic DFE appearance of angioid streaks?
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What proportion of angioid streaks are associated with systemic abnormalities?
- About half

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
- **P**
- **E**
- **P**
- **E**
- **S**
- **I**
What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

What proportion of angioid streaks are associated with systemic abnormalities?
About half.

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
Pseudoxanthoma elasticum (PXE)
Ehlers-Danlos syndrome
Paget’s disease of bone
Sickle-cell disease
Idiopathic (ie, no association)
CNVM DDx:

- ARMD
- POHS
- Angioid streaks

What is the classic DFE appearance of angioid streaks? Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

What proportion of angioid streaks are associated with systemic abnormalities? About half.

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
- Pseudoxanthoma elasticum (PXE)
- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)

~50% of cases are associated with one of these

~50% of cases have no known systemic association
What is the classic DFE appearance of angioid streaks?
**Reddish-brown** lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

What proportion of angioid streaks are associated with systemic abnormalities?
About half.

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
- **P**seudoaxanthoma elasticum (PXE)?
- **E**hlers-Danlos syndrome?
- **P**aget’s disease of bone?
- **S**ickle-cell disease?
- Idiopathic (ie, no association)

**Which condition has the strongest association with angioid streaks?**
What is the classic DFE appearance of angioid streaks? Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

What proportion of angioid streaks are associated with systemic abnormalities? About half.

What is the well-known mnemonic for angioid streak’s associations? What are these associations? Pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget’s disease of bone, Sickle-cell disease, Idiopathic (i.e., no association).

Which condition has the strongest association with angioid streaks? PXE, by a mile.
What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

What proportion of angioid streaks are associated with systemic abnormalities?
About half.

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
Pseudoxanthoma elasticum (PXE)
Ehlers-Danlos syndrome
Paget’s disease of bone
Sickle-cell disease
Idiopathic (ie, no association)

Which condition has the strongest association with angioid streaks? PXE, by a mile.

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?
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
Briefly, what sort of disorder is PXE?

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

What is the well-known mnemonic for angioid streak’s associations? What are these associations?

Pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget’s disease of bone, Sickle-cell disease, Idiopathic (ie, no association).

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Which condition has the strongest association with angioid streaks? PXE, by a mile.

Pseudoxanthoma elasticum (PXE)

Ehlers-Danlos syndrome
Page's disease of bone
Sickle-cell disease
Idiopathic (ie, no association)

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**CNVM DDx**

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Rare

*Which condition has the strongest association with angioid streaks?*
PXE, by a mile

**Pseudoxanthoma elasticum (PXE)**
- Ehlers-Danlos syndrome
- Paget's disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)
Briefly, what sort of disorder is PXE?
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Rare

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Yes, \(\text{♀} \) are twice as likely to be affected

What is the well-known mnemonic for angioid streak’s associations? What are these associations?
Pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget’s disease of bone, Sickle-cell disease, Idiopathic (ie, no association)

Which condition has the strongest association with angioid streaks?
PXE, by a mile
**Q/A**

**CNVM DDx**

*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?*
Rare

*Is there a gender predilection?*
Yes, \( \square \) \( \text{MVF} \) are twice as likely to be affected

*What is the well-known mnemonic for angioid streak’s associations? What are these associations?*

\textbf{Pseudoxanthoma elasticum (PXE)}

- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)

*Which condition has the strongest association with angioid streaks?*
PXE, by a mile
### CNVM DDx

- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Choroidal rupture after trauma
- Iatrogenic

---

#### What is the classic DFE appearance of angioid streaks?

Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

#### What proportion of angioid streaks are associated with systemic abnormalities?

About half.

#### What is the well-known mnemonic for angioid streak’s associations? What are these associations?

**Pseudoxanthoma elasticum (PXE)**
- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)

---

#### Briefly, what sort of disorder is PXE?

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Rare.

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Yes, ♀ are twice as likely to be affected.

#### Which condition has the strongest association with angioid streaks?

PXE, by a mile.
**CNVM DDx**

**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?**
Rare

**Is there a gender predilection?**
Yes, ♂️ are twice as likely to be affected

**Other than the eye, what organ-systems are affected?**
- Bruch’s membrane
- Other

**What is the well-known mnemonic for angioid streak’s associations? What are these associations?**
- **P**seudoxanthoma **E**lasticum (PXE)
- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)

**Which condition has the strongest association with angioid streaks?**
PXE, by a mile
**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?**

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

**Which condition has the strongest association with angioid streaks?**

PXE, by a mile

**What is the well-known mnemonic for angioid streak’s associations? What are these associations?**

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- Ehlers-Danlos syndrome
- Paget’s disease of bone
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Which condition has the strongest association with angioid streaks? PXE, by a mile.

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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
- GI tract
- Eye

What is the appearance of affected skin?
An area of waxy-yellow, papule-like lesions.

What is the classic informal descriptor for this appearance?
‘Chicken skin’.

In what two locations is ‘chicken skin’ most often found?
- The neck
- The axillae
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?
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Which condition has the strongest association with angioid streaks?
PXE, by a mile
PXE skin
**CNVM DDx**

**Briefly, what sort of disorder is PXE?**
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PXE, by a mile.

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- Ehlers-Danlos syndrome
- Paget’s disease of bone
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- Idiopathic (ie, no association)
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**What is the appearance of affected skin?**
An area of waxy-yellow, papule-like lesions

**What is the classic informal descriptor for this appearance?**
‘Chicken skin’

**What are the well-known associations of angioid streaks?**
Which condition has the strongest association with angioid streaks? PXE, by a mile

**What is the classic DFE appearance of angioid streaks?**
Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane

**What proportion of angioid streaks are associated with systemic abnormalities?**
About half

**What is the well-known mnemonic for angioid streak’s associations?**
Pseudoxanthoma elasticum (PXE)
**CNVM DDx**

**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?**
Rare

**Is there a gender predilection?**
Yes, ♀ are twice as likely to be affected

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**What is the classic informal descriptor for this appearance?**
‘Chicken skin’

**In what two locations is ‘chicken skin’ most often found?**
-- Skin
-- Eye

**Other than the eye, what organ-systems are affected?**
-- Vascular system
-- GI tract
-- Skin

**What is the well-known mnemonic for angioid streak’s associations?**
What are these associations?

**Pseudoxanthoma elasticum (PXE)**

**Which condition has the strongest association with angioid streaks?**
PXE, by a mile
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What is the classic informal descriptor for this appearance?
‘Chicken skin’

In what two locations is ‘chicken skin’ most often found?
--The neck
--The axillae

Which condition has the strongest association with angioid streaks?
PXE, by a mile
PXE: ‘Chicken skin’
Q

Briefly, what sort of disorder is PXE?
An elastorrhexis, i.e., a condition characterized by progressive calcification and fragmentation of elastic tissues.

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
--GI tract
--Eye

What is the well-known mnemonic for angioid streak's associations?
What are these associations?

Pseudoxanthoma elasticum (PXE)
Ehlers-Danlos syndrome
Paget’s disease of bone
Sickle-cell disease
Idiopathic (i.e., no association)

Which condition has the strongest association with angioid streaks?
PXE, by a mile.

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

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**Which condition has the strongest association with angioid streaks?**
PXE, by a mile

**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
**CNVM DDx**

**Q**

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An elastorrhexis, i.e., a condition characterized by progressive calcification and fragmentation of elastic tissues.

*Is it common, or rare?*

Rare.

*Is there a gender predilection?*

Yes, ♀ are twice as likely to be affected.

**A**

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

--Skin
--Vascular system
--GI tract
--Eye

*What is the well-known mnemonic for angioid streak’s associations? What are these associations?*

Pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget’s disease of bone, Sickle-cell disease, Idiopathic (i.e., no association).

*Which condition has the strongest association with angioid streaks?*

PXE, by a mile.

*What mellifluous name is used to describe the RPE mottling?*

Peau d’orange.

*What is the classic DFE appearance of angioid streaks?*

Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch’s membrane.

*What proportion of angioid streaks are associated with systemic abnormalities?*

About half.

*What is the well-known mnemonic for angioid streak’s associations? What are these associations?*

Pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget’s disease of bone, Sickle-cell disease, Idiopathic (i.e., no association).
CNVM DDx

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

- Skin
- Vascular system
- GI tract
- Eye

**There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?**
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**What mellifluous name is used to describe the RPE mottling?**
Peau d’orange.

**Which condition has the strongest association with angioid streaks?**
PXE, by a mile.

**What is the well-known mnemonic for angioid streak’s associations?**

Pseudoxanthoma elasticum (PXE)

- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (i.e., no association)
PXE: *Peau d’orange* fundus
Briefly, what sort of disorder is PXE?

An elastorrhexis, i.e., a condition characterized by progressive calcification and fragmentation of elastic tissues

Is it common, or rare?

Rare

Is there a gender predilection?

Yes, ♀ are twice as likely to be affected

For more on angioid streaks, see slide-set R61

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

What mellifluous name is used to describe the RPE mottling?

Peau d’orange

Which condition has the strongest association with angioid streaks?

PXE, by a mile

What is the well-known mnemonic for angioid streak’s associations? What are these associations?

Pseudoxanthoma elasticum (PXE)

Ehlers-Danlos syndrome

Page’s disease of bone

Sickle-cell disease

Idiopathic (i.e., no association)
CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- **Pathologic myopia**
- Idiopathic
- Sorsby macular dystrophy
- Choroidal rupture after trauma
- Iatrogenic

*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*
- CNVM DDx:
  - ARMD
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  - Iatrogenic

*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Q

- CNVM DDx:
  - ARMD
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  - Angioid streaks
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  - Iatrogenic

**What proportion of eyes longer than 26.5 mm will develop CNV?** About 10%

**What axial length serves as a useful cutoff for defining pathologic myopia?**

26.5 mm
**A**

- **CNVM DDx:**
  - ARMD
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**What proportion of eyes longer than 26.5 mm will develop CNV?**

*About 10%*

**What axial length serves as a useful cutoff for defining pathologic myopia?**

*26.5 mm*
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*

26.5 mm

*What is the classic finding on DFE that puts high myopes at risk for CNVM?*
CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- **Pathologic myopia**
- Idiopathic
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*
26.5 mm

*What is the classic finding on DFE that puts high myopes at risk for CNVM?*
**Lacquer cracks**
**Q**

- **CNVM DDx:**
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia

Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm

**Angioid streaks vs lacquer cracks: Compare and contrast**

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Q/A

- **CNVM DDx:**
  - ARMD
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  - **Angioid streaks**
  - Pathologic myopia

**Angioid streaks vs lacquer cracks: Compare and contrast**

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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Angioid streaks
‘Reddish-brown’

Lacquer cracks
‘Yellowish’
Q/A

- **CNVM DDx:**
  - ARMD
  - POHS
  - **Angioid streaks**
  - Pathologic myopia

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**Angioid streaks vs lacquer cracks: Compare and contrast**

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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Q/A

- CNVM DDx:
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Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm
Angioid streaks
‘Reddish-brown’
‘Peripapillary’

Lacquer cracks
‘Yellowish’
‘Macular’
**Q/A**

- **CNVM DDx:**
  - ARMD
  - POHS
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**CNVM DDx:**
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**Q/A**

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- POHS
- **Angioid streaks**
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Q

- CNVM DDx:
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  - POHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic
  - **Sorsby macular dystrophy**
  - Choroidal rupture
  - Iatrogenic

**In two words, what sort of condition is Sorsby?**
(Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)
A: **dystrophy**
A CNVM DDx:

- ARMD
- POHS
- Angioid streaks
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- Iatrogenic

*In two words, what sort of condition is Sorsby? (Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)*

A macular dystrophy
Q

● CNVM DDx:
  ● ARMD
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  ● **Sorsby macular dystrophy**
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A macular dystrophy

*What is the inheritance pattern?*
CNVM DDx:
- ARMD
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- **Sorsby macular dystrophy**
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**In two words, what sort of condition is Sorsby?**
(Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)

A macular dystrophy

What is the inheritance pattern?
AD

A young adult with large numbers of “drusenlike deposits” in the maculae

What is the classic presentation of a pt who has become symptomatic?
Bilateral subfoveal CNVMs at age 40
Q

- CNVM DDx:
  - ARMD
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**CNVM DDx**

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What is the inheritance pattern?
AD

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*What is the inheritance pattern?*

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A young adult with large numbers of “drusenlike deposits” in the macula bilaterally
Sorsby: Drusenlike deposits
Q

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*What is the classic presentation of a pt who has become symptomatic?*

CNVMs by age # or so
A

- **CNVM DDx:**
  - ARMD
  - POHS
  - Angioid streaks
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Bilateral subfoveal CNVMs by age 40 or so

The story you’re looking for (on a test) is of an adult who had good vision bilaterally until their early 40s, when they noted sudden VA decrease in one eye, then the other.

(No question, proceed when ready)
CNVM DDx:
- ARMD
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What is the classic presentation of a pt who has become symptomatic?
Bilateral subfoveal CNVMs by age 40 or so

The story you’re looking for (on a test) is of an adult who had good vision bilaterally until their early 40s, when they noted sudden VA decrease in one eye, then the other. DFE and imaging will reveal CNVM in the affected eye(s) if vision loss is recent, extensive scarring if remote.

*(No question, proceed when ready)*
Sorsby: Late extensive fibrosis/scarring
Q

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What is the classic presentation of a pt who has become symptomatic?
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What is the subheader name?
**'Early-onset "drusenoid" macular dystrophies'**
A CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
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- Choroidal rupture
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**What is the inheritance pattern?**
AD

**What is the subheader name?**
‘Early-onset “drusenoid” macular dystrophies’

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A macular dystrophy

What is the inheritance pattern?
AD

What is the subheader name?
‘Early-onset “drusenoid” macular dystrophies’

For context and completeness’ sake: What are the other early-onset “drusenoid” macular dystrophies with which Sorsby’s is grouped?
--?
--?
--?
--?
--?

(Hints forthcoming)
Q

● **CNVM DDx:**
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic
  - **Sorsby macular dystrophy**
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What is the inheritance pattern?
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‘Early-onset “drusenoid” macular dystrophies’

For context and completeness’ sake: What are the other early-onset “drusenoid” macular dystrophies with which Sorsby’s is grouped?
--? The most common inherited macular dystrophy
--?
--?
--?

Bilateral subfoveal CNVMs by age 40 or so
CNVM DDx:
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--Stargardt
--?
--?
--?

Bilateral subfoveal CNVMs by age 40 or so
In two words, what sort of condition is Sorsby? (Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)
A macular dystrophy

What is the inheritance pattern?
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‘Early-onset “drusenoid” macular dystrophies’

For context and completeness’ sake: What are the other early-onset “drusenoid” macular dystrophies with which Sorsby’s is grouped?
--Stargardt
--? The best macular dystrophy?
--?
CNVM DDx:

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- POHS
- Angioid streaks
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--Stargardt
--Best dz
--?

Bilateral subfoveal CNVMs by age 40 or so
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**CNVM DDx**

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- A macular dystrophy

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For context and completeness’ sake: What are the other early-onset “drusenoid” macular dystrophies with which Sorsby’s is grouped?

- Stargardt
- Best dz
- Best dz’s older sibling

Bilateral subfoveal CNVMs by age 40 or so
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  --Stargardt
  --Best dz
  --Adult-onset foveomacular vitelliform dystrophy

Bilateral subfoveal CNVMs by age 40 or so
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A macular dystrophy

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--Stargardt
--Best dz
--Adult-onset foveomacular vitelliform dystrophy

**Sorsby macular dystrophy**

--? *Eg, butterfly; reticular; pulverulentus*

--?

Bilateral subfoveal CNVMs by age 40 or so
A

- **CNVM DDx:**
  - ARMD
  - POHS
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  - Pathologic myopia
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**CNVM DDx**

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--Best dz
--Adult-onset foveomacular vitelliform dystrophy

**Sorsby macular dystrophy**

--The pattern dystrophies
--?
--?

Bilateral subfoveal CNVMs by age 40 or so
Q

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic
  - **Sorsby macular dystrophy**
  - Choroidal rupture
  - Iatrogenic

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**CNVM DDx**

In two words, what sort of condition is Sorsby? (Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)

A macular dystrophy

What is the inheritance pattern?

AD

What is the subheader name?

‘Early-onset “drusenoid” macular dystrophies’

For context and completeness’ sake: What are the other early-onset “drusenoid” macular dystrophies with which Sorsby’s is grouped?

--Stargardt
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Not breast-related, despite its name

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