Q

- CNVM DDx:
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- ARMD
- OHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy
- Traumatic choroidal rupture
- Iatrogenic
**CNVM DDx:**

- **ARMD**
  
  *ARMD is addressed extensively in a series of slide-sets—see the Table of Contents*

- OHS
- Angioid streaks
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- Iatrogenic
What does OHS stand for in this context?

OHS stands for Ocular Histoplasmosis Syndrome. The P in OHS stands for 'presumed.'

Is there a gender predilection in OHS?
No.

Is there a racial predilection?
Yes, OHS occurs almost exclusively among white people 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.

What about AC cell?
Never. If AC cell is present, it's not OHS.
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In a nutshell, what are histo spots?

Discrete, focal, atrophic scars.

Where are they typically located?

They can be anywhere in the posterior pole, i.e., 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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In another nutshell, what is PPA? Like histo spots, PPA represent atrophic scars.
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--Disciform macular lesion(s)

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

What about vitritis?
Never. If vitritis is present, it’s not OHS.

What about AC cell?
Never. If AC cell is present, it’s not 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, are --Active lesions representing a CNVM 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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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, active lesions (which are expected to be hemorrhage-rich) represent: (I) active CNVM, (II) hemorrhagic retinal detachment, or an exudative retinal detachment.

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

What about AC cell?
Never. If AC cell is present, it's not OHS.

What is the classic triad of OHS?
--Histo spots
--Peripapillary atrophy
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The so-called ‘classic triad’ of OHS.

**What is the classic triad of OHS?**
--Histo spots
--Peripapillary atrophy
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**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?**
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**What treatment modalities are used to treat active disciform lesions?**
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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, we see a hemorrhage-retinal detachment or a homogenous-retinal detachment.

--Active lesions (aka disciform active lesions) -- either previous CNVM and/or hemorrhage

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

What treatment modalities are used to treat active disciform lesions? --Thermal laser
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The so-called ‘classic triad’ of OHS
-- Histo spots
-- Peripapillary atrophy
-- **Disciform macular lesion(s)**

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

In the last nutkhalk, an **Active lesions**
-- Active lesions (aka: hemorrhagic retinopathy)
-- Inactive lesions (aka: 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)

Do antifungals play a role in the treatment of OHS?
-- **Antifungals?**
A

• CNVM DDx:
  - ARMD
  - OHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic Sorsby macular dystrophy (SMD)
  - Traumatic choroidal rupture
  - Iatrogenic

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

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What 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
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The so-called ‘classic triad’ of OHS:
--Histo spots
--Peripapillary atrophy
--Disciform macular lesion(s)

What is the classic triad of OHS?

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

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)
--Antifungals? No!

Do antifungals play a role in the treatment of OHS?
No.
CNVM DDx:

- ARMD
- POHS

**Angioid streaks**

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

- CNVM DDx:
  - ARMD
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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
Q

• 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?
A

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks

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?
P
P
E
P
S
I
**Question:**

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

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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?

**PEPSI**

- P = POHS
- E = Eales disease
- P = Pathologic myopia
- S = Sorsby macular dystrophy (SMD)
- I = Iatrogenic
**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)
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?**
- **P**seudoxanthoma elasticum (PXE)
- **E**hlers-Danlos syndrome
- **P**aget’s disease of bone
- **S**ickle-cell disease
- **I**diopathic (ie, **no association**)

~50% of cases are associated with one of these
~50% of cases have no known systemic association.

*(No question, proceed when ready)*
CNVM DDx:
- ARMD
- POHS
- **Angioid streaks**

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

Which condition has the strongest association with angioid streaks?
**CNVM DDx:**

- ARMD
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- **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**seudoxanthoma **E**lasticum (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?*

PXE, by a mile.
CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Choroidal rupture after trauma
- Iatrogenic

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- Paget’s disease of bone
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- 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)
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About half.

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- 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? What are these associations?
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
**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)

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

PXE, by a mile.

---

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

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Yes, ♀ are twice as likely to be affected.

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- Skin
- Vascular system
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- Eye
**CNVM DDx:**
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Choroidal rupture after trauma
- Iatrogenic

---

**What is the classic DME 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)

---

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Rare.

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

Is there a gender predilection? Yes, females are twice as likely to be affected.

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

Is it common, or rare?
- Rare

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

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
**CNVM DDx:**
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Choroidal rupture after trauma
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- Sickle-cell disease
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Rare.

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

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Rare.

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Yes, ♀ are twice as likely to be affected.

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

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- Vascular system
- GI tract
- Eye

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

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About half.

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**P**seudoxanthoma elasticum (PXE), **E**hlers-Danlos syndrome, **P**aget’s disease of bone, **S**ickle-cell disease, Idiopathic (i.e., no association)

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

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 system, 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.
CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
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What other organ-systems are affected?
- Skin
- Vascular system
- GI tract
- Eye

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**In what two locations is 'chicken skin' most often found?**
- The neck
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Paget’s disease of bone
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Idiopathic (ie, no association)

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

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'Chicken skin'.

In what two locations is 'chicken skin' most often found?
Skin, Eye.

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 association? What are these associations?
Pseudeoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget's disease of bone, Sickle-cell disease, Idiopathic (ie, no association).
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?
- Pseudoxanthoma elasticum (PXE)
- Ehlers-Danlos syndrome
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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.

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

Is there a gender predilection?
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Other than the eye, what organ-systems are affected?
--Skin
--Vascular
--GI tract
About 1 in 10 patients present breaks in Bruch's membrane

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

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**For more on angioid streaks, see slide-set R61**

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

26.5 mm
Q

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  - Angioid streaks
  - **Pathologic myopia**
  - Idiopathic
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What proportion of eyes longer than 26.5 mm will develop CNV?

About 10%
CNVM DDx:
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*
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*What is the classic finding on DFE that puts high myopes at risk for CNVM?*
CNVM DDx:
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*What is the classic finding on DFE that puts high myopes at risk for CNVM?*

**Lacquer cracks**
Q

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
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Angioid streaks vs lacquer cracks: Compare and contrast

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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 CNVM DDx:

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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
**Q**

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A macular dystrophy

**For context and completeness’ sake: What are the other macular dystrophies with which Sorsby’s is addressed?**

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- Stargardt
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- Adult-onset vitelliform
- The ‘pattern’ dystrophies
- Central areolar choroidal dystrophy
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Q

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

Bilateral subfoveal CNVMs at age 40
Q

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What is the classic DFE finding in a pt who has the condition, but has yet to become symptomatic?
(Hint: It’s indicated by the subheader of the section in the Retina book in which it is presented.)
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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’

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