

# Q

## CNVM DDx



- CNVM DDx:

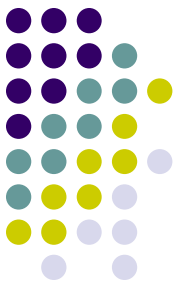
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# A

## CNVM DDx

- CNVM DDx:
  - 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*
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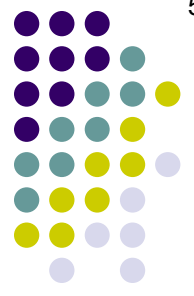
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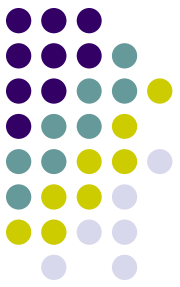


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Ocular histoplasmosis syndrome, aka **POHS** (the P is for **presumed**)



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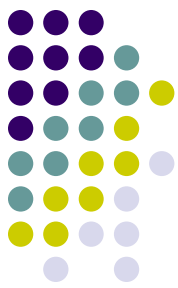
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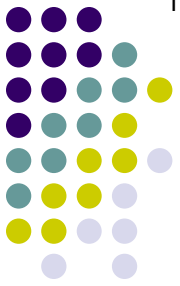
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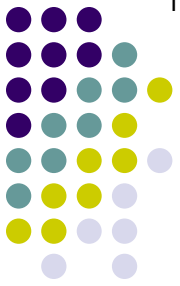
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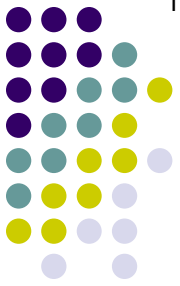
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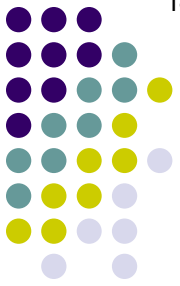
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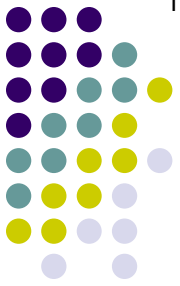
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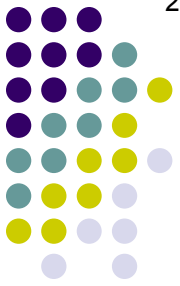
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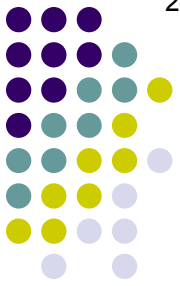
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--Histo spots

--Peripapillary atrophy

--Disciform macular lesion(s)

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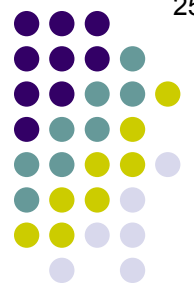
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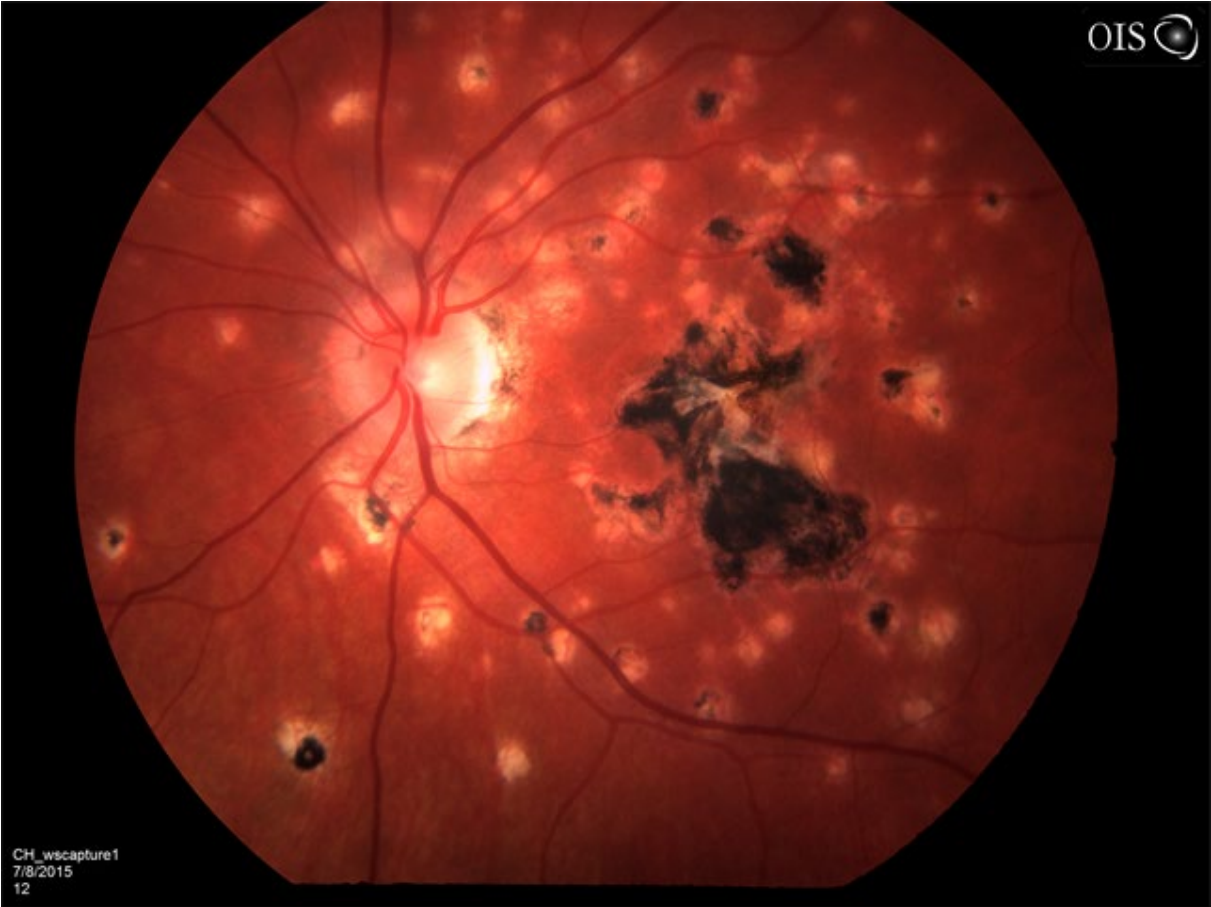
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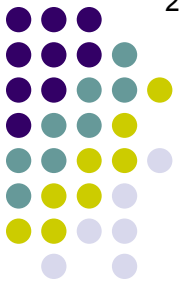




# CNVM DDX



OHS: The classic triad



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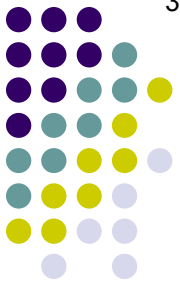
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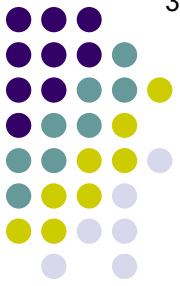
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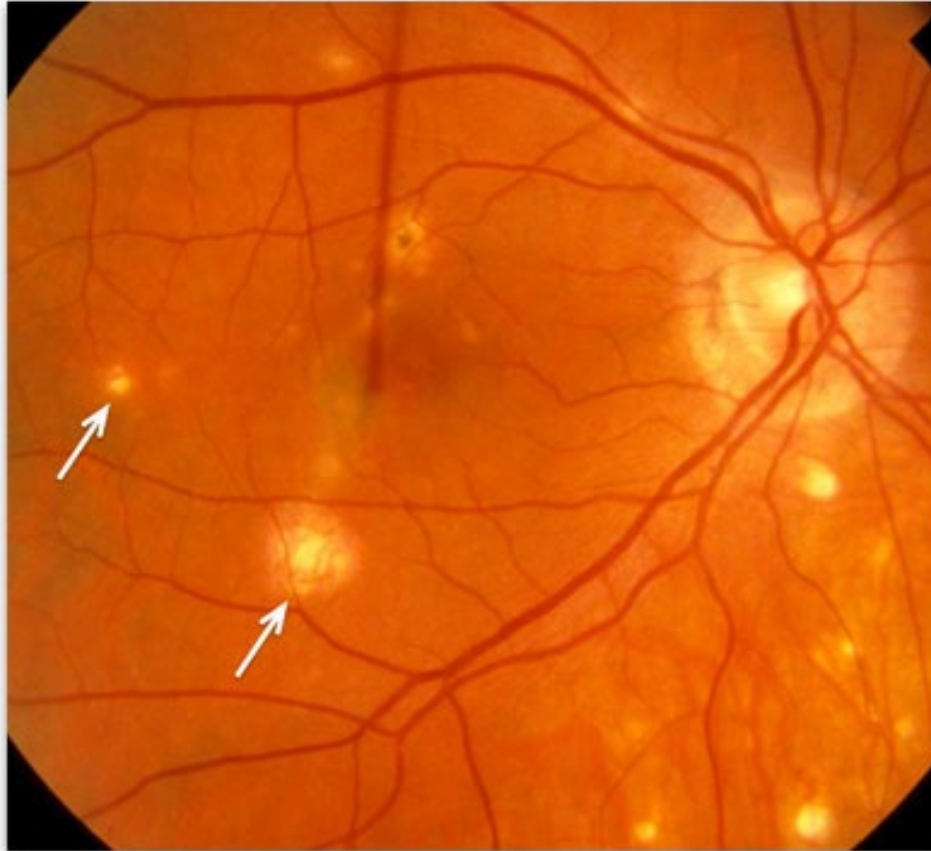
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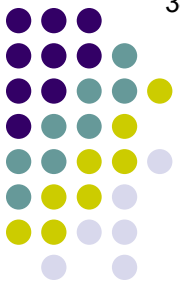
*What about AC cells?*

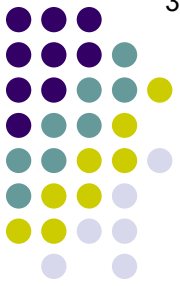
**Never.** If AC cell is present, it's not OHS.

# CNVM DDx



OHS: Histo spots





## CNVM DDx

Q

### ● CNVM DDx

● ARMD

● **OHS**

● Angioid

● Pathologic

● Idiopathic

● Sorsby

● Trauma

● 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'

*In a nutshell, what are histo spots?*

Discrete, focal, atrophic scars

*What is the classic class*

-**Histo spots**

--Peripapillary atrophy

--Disciform maculopathy

*Where are they typically located?*

They can be anywhere in the posterior pole, ie, macula or near-to-mid periphery

reside in the Misen

*Are they usually larger, or smaller than the ONH?*

Smaller

*Does OHS manifest*

Bilaterally (although

*What two-word phrase is used to describe them?*

'Punched out'

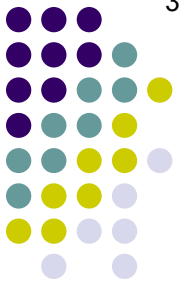
*Is OHS associated*

**Never.** If vitritis is

*Do they evolve over time?*

*What about AC cells?*

**Never.** If AC cell is present, it's not OHS.



## CNVM DDx

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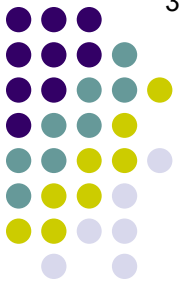
**Never.** If vitritis is

*Do they evolve over time?*

Generally no

*What about AC cell?*

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## CNVM DDx

Q

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● ARMD

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*What is the classic triad of OHS?*

--Histo spots

--**Peripapillary atrophy**

--Disciform macular lesion

*In another nutshell, what is PPA?*

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?*

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## CNVM DDx

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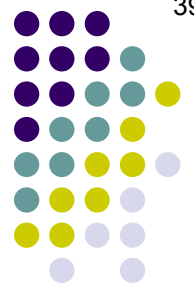
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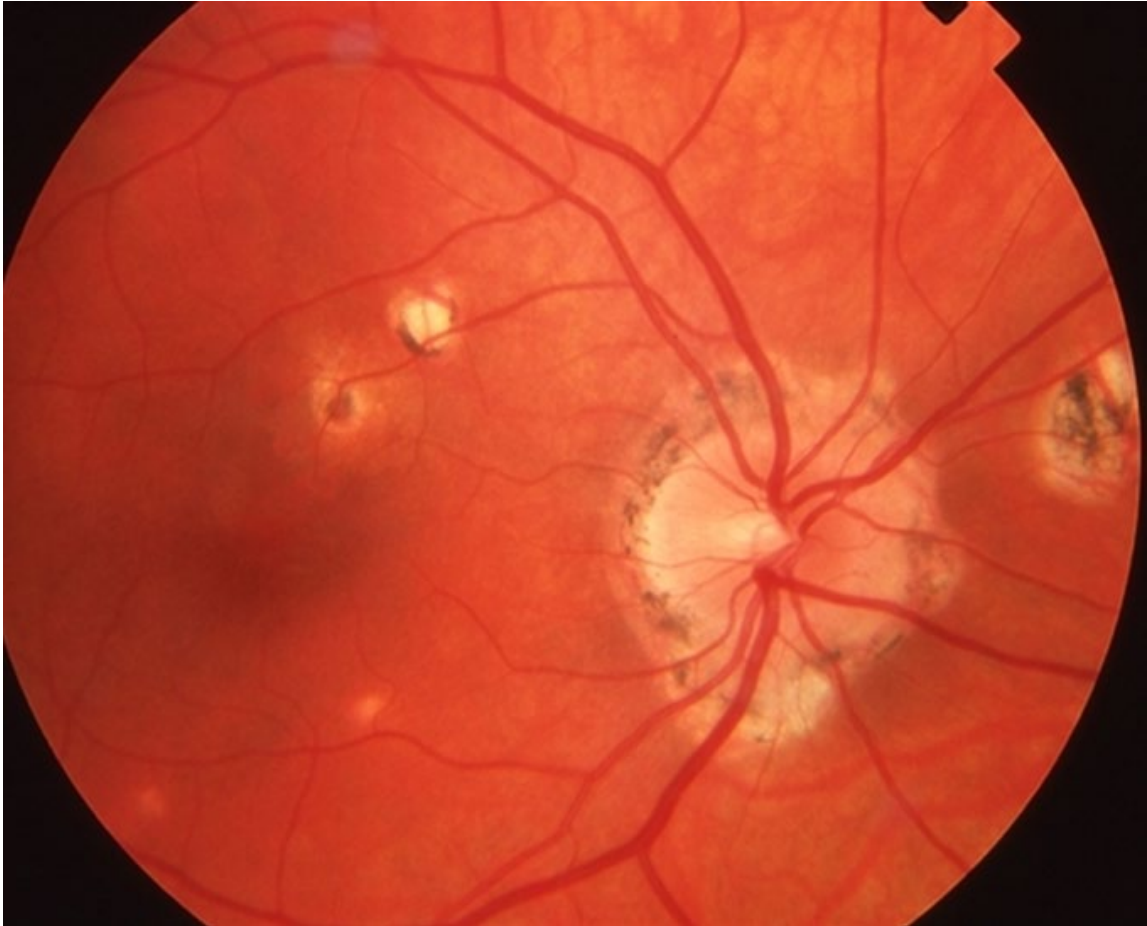
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**CNVM DDx**



OHS: PPA



# Q

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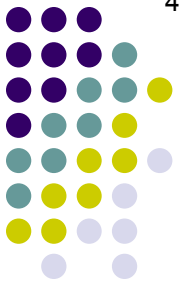
*In the last nutshell, what are disciform lesions?*

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# Q/A

## CNVM DDx

- CNVM DDx

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*What is the classic triad of OHS?*

--Histo spots

--Peripapillary atrophy

- Idiopathic

--**Disciform macular lesion(s)**

who reside in the Mississippi/Ohio River valleys of the US

- *In the last nutshell, what are disciform lesions?*

--**Active** lesions represent...

- --

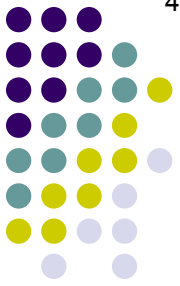
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--**Active** lesions represent...either the presence of CNVM under the retina, or a hemorrhagic retinal detachment; whereas

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CNVM DDx



Active lesion

OHS



# Q/A

## CNVM DDx

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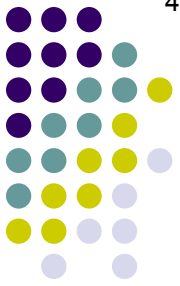
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# Q/A

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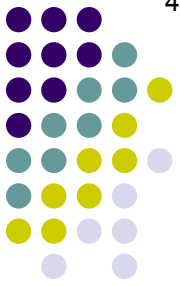
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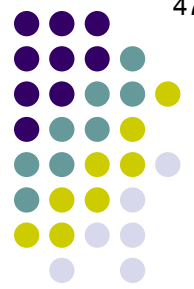
*In the last nutshell, what are disciform lesions?*

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

*What about AC cell?*

**Never.** If AC cell is present, it's not OHS.



**CNVM DDx**



Active lesion



Disciform scar

OHS



## CNVM DDx

Q

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**--Histo spots?**

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reside in the Mississippi/Ohio River valleys of the US

*In the last nutshell, w/*

**--Active** lesions repre

or a hemorrhagic retin

**--Inactive** lesions (aka

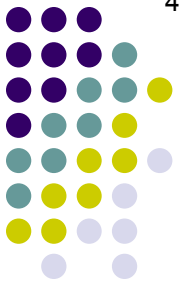
previous CNVM and/or

*What a*

**Never.**

*Which lesion(s) require treatment?*





# A

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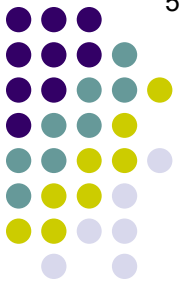
previous CNVM and/or

*What a*

*Never.*

*Which lesion(s) require treatment?*

Only active disciform lesions



# CNVM DDx

# Q

## ● CNVM DDx

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

● ARMD

*How is the diagnosis of OHS made?*  
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● **OHS**

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

● Angioid

*What is the classic triad of OHS?*

● Pathologic

- Histo spots
- Peripapillary atrophy

● Idiopathic

- Disciform macular lesion(s)**
- who reside in the Mississippi/Ohio River valleys of the US

● *In the last nutshell, w/*

*Which lesion(s) require treatment?*

● **-Active lesions**

Only active disciform lesions

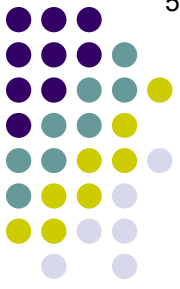
● or a hemorrhagic retina

● **-Inactive lesions** (aka previous CNVM and/or

*What treatment modalities are used to treat active disciform lesions?*

- 
- 
- 
- 
- 
- 

*What a*  
*Never.*



# A

## CNVM DDx

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#### ● ARMD

#### ● OHS

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--**Disciform macular lesion(s)**

reside in the Mississippi/Ohio River valleys of the US

*In the last nutshell, what are the key features of a hemorrhagic retina?*

--**Active lesions**

--Inactive lesions (aka)

--previous CNVM and/or

--Never.

*What are the key features of a hemorrhagic retina?*

Never.

*Which lesion(s) require treatment?*

Only active disciform lesions

*What treatment modalities are used to treat active disciform lesions?*

--Thermal laser

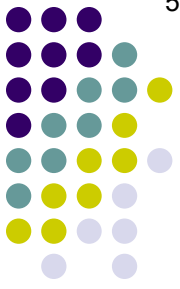
--Photodynamic therapy (PDT)

--Anti-VEGF therapy

--Submacular surgery

--Intravitreal corticosteroids

--Combination therapy (of some of the above modalities)



## CNVM DDx

Q

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--**Disciform macular lesion(s)**

who reside in the Mississippi/Ohio River valleys of the US

*In the last nutshell, what are the key features of OHS?*

--**Active lesions**

or a hemorrhagic retina

--**Inactive lesions** (aka

previous CNVM and/or

atrophic CNVM)

*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

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

--Combination therapy (or some of the above modalities)

--**Antifungals?**



# A

## CNVM DDx

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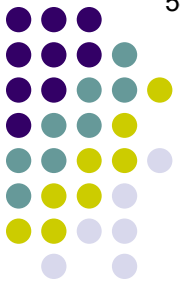
--Anti-VEGF therapy

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

No

--Combination therapy (or some of the above modalities)

--**Antifungals? No!**



## CNVM DDx

- CNVM DDx

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The so-called 'classic triad' of OHS

- Angioid

*What is the classic triad of OHS?*

ites

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

- Idiopathic

reside in the Mississippi/Ohio River valleys of the US

- *In the last nutshell, w/*

*Which lesion(s) require treatment?*

*--Active lesions (repre*

Only active disciform lesions

- *of a hemorrhagic retina*

*--Inactive lesions (aka*

*What treatment modalities are used to treat active disciform lesions?*

- *previous CNVM and/or*

--Thermal laser

*never*

--Photodynamic therapy (PDT)

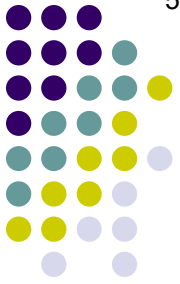
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*Do antifungals play a role in the treatment of OHS?*

**No**

--Combination therapy (or some of the above modalities)

--*Antifungals? No!*



## CNVM DDx

# Q

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

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



# Q/A

## CNVM DDx

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

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

color-color

retinal structure

lines radiating out from the peripapillary region; these lines represent breaks in





## CNVM DDx

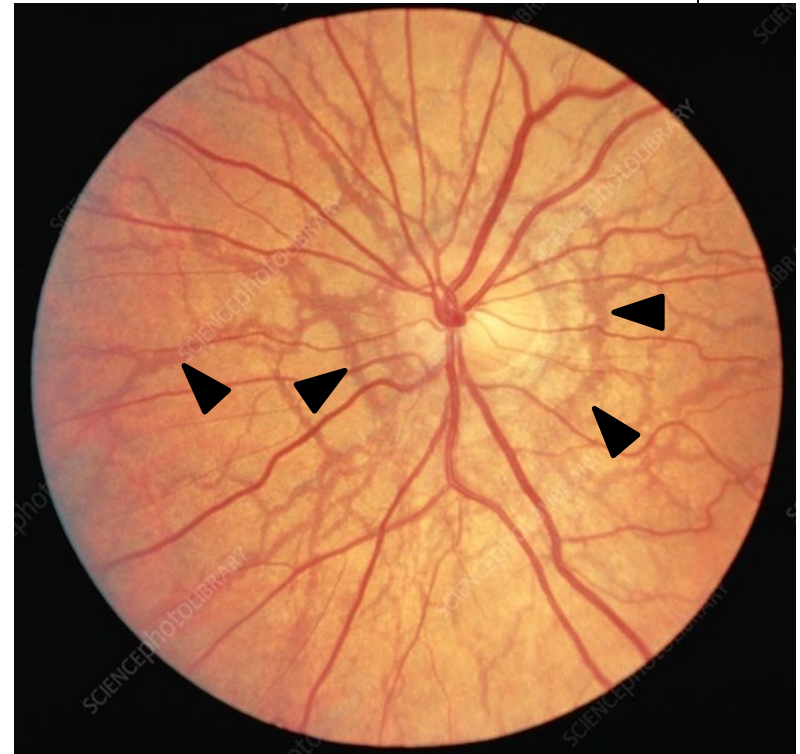
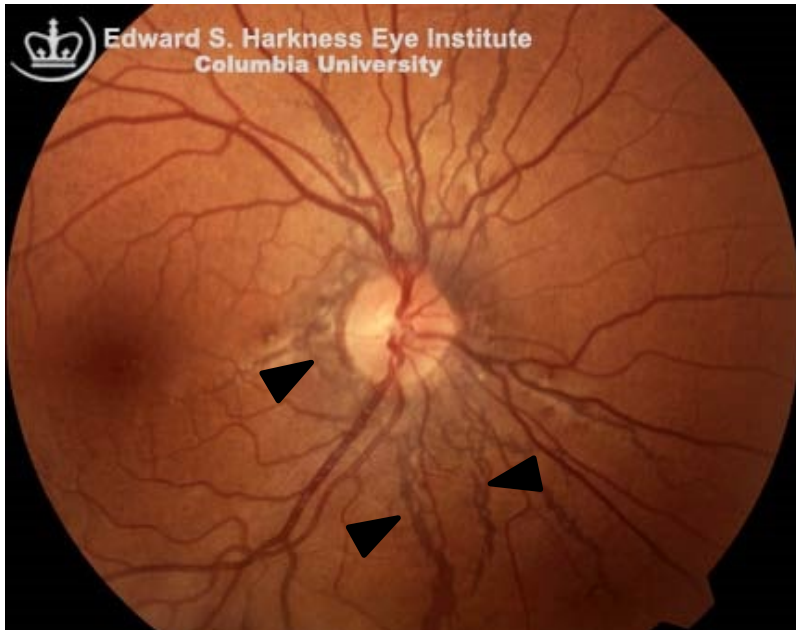
# A

- 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



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



## CNVM DDx

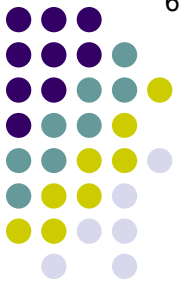
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- CNVM DDx:
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*What proportion of angioid streaks are associated with systemic abnormalities?*



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  - ARMD
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## CNVM DDx

# Q

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  - ARMD
  - POHS
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*What is the well-known mnemonic for angioid streak's associations?*



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**P  
E  
P  
S  
I**



## CNVM DDx

# Q

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

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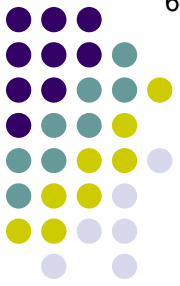
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**P  
E  
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*What proportion of angioid streaks are associated with systemic abnormalities?*

About half

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

**P**seudoxanthoma elasticum (PXE)

**E**hlers-Danlos syndrome

**P**aget's disease of bone

**S**ickle-cell disease

**I**diopathic (ie, no association)



## CNVM DDx



- 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

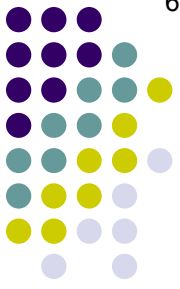
*What proportion of angioid streaks are associated with systemic abnormalities?*

About half

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

<b>P</b> seudoxanthoma elasticum (PXE)	}	~50% of cases are associated with one of these
<b>E</b> hlers-Danlos syndrome		
<b>P</b> aget's disease of bone		
<b>S</b> ickle-cell disease		
<b>I</b> diopathic (ie, <b>no association</b> )	}	~50% of cases have no known systemic association

*(No question, proceed when ready)*



## CNVM DDx

# Q

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

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

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**P**seudoxanthoma 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?*



## CNVM DDx

# A

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

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

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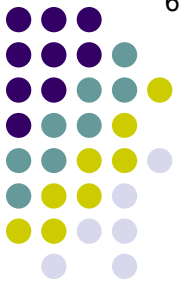
**P**aget's disease of bone

**S**ickle-cell disease

**I**diopathic (ie, no association)

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

PXE, by a mile



## CNVM DDx

Q

Briefly, what sort of disorder is PXE?

- CNM
- A
- F
- A

What is

Reddis

Bruch's

What p

About

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

**P**seudoxanthoma elasticum (PXE)

Ehlers-Danlos syndrome

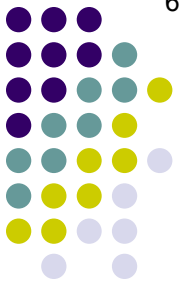
Paget's disease of bone

Sickle-cell disease

Idiopathic (ie, no association)

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

sent breaks in



## CNVM DDx

# A

- CNVM DDx
  - A
  - P
  - A

*Briefly, what sort of disorder is PXE?*

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

*What is*

**Reddis**

Bruch's

sent breaks in

*What p*

About

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

**P**seudoxanthoma elasticum (PXE)

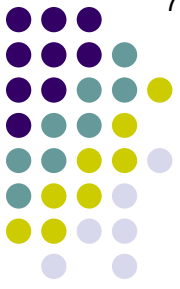
Ehlers-Danlos syndrome

Paget's disease of bone

Sickle-cell disease

~~I~~diopathic (ie, no association)

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



## CNVM DDx

Q

*Briefly, what sort of disorder is PXE?*

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

*Is it common, or rare?*

What is

Reddis

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What p

About

*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

sent breaks in



## CNVM DDx

# A

- CNVM DDx
  - A
  - P
  - A

*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

*What is*

**Reddis**

Bruch's

sent breaks in

*What p*

About

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

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PXE, by a mile



## CNVM DDx

Q

- 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?*

What is

Reddis

Bruch's

What p

About

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

**P**seudoxanthoma 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*

sent breaks in





# Q/A

## CNVM DDx

- **CN**
  - **A** Briefly, what sort of disorder is PXE?  
An elastorrhesis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues
  - **F** Is it common, or rare?  
Rare
  - **A** Is there a gender predilection?  
Yes, **MVF** are twice as likely to be affected

What is

Reddis

Bruch's

What p

About

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)

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

sent breaks in



## CNVM DDx

# A

- CNVM DDx
  - *Briefly, what sort of disorder is PXE?*  
An elastorrhesis, 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

Reddis

Bruch's

What p

About

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

**P**seudoxanthoma 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

sent breaks in



## CNVM DDx

Q

● CN

*Briefly, what sort of disorder is PXE?*

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

● A

*Is it common, or rare?*

● F

Rare

● A

*Is there a gender predilection?*

Yes, ♀ are twice as likely to be affected

*What is*

**Reddis**

Bruch's

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

--

--

*What p*

About

--

--

*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)

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



## CNVM DDx

# A

- CNVM
  - A: *Briefly, what sort of disorder is PXE?*  
An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues
  - F: *Is it common, or rare?*  
Rare
  - A: *Is there a gender predilection?*  
Yes, ♀ are twice as likely to be affected

What is

Reddis  
Bruch's

What p  
About

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

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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

sent breaks in



## CNVM DDx

Q

● CN

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

● A

*Is it common, or rare?*

● F

Rare

● A

*Is there a gender predilection?*

Yes, ♀

*What is the appearance of affected skin?*

What is

Reddis

Bruch's

Other th

---Skin

---vascu

---GI trac

---Eye

What p

About

sent breaks in

*What is the well-known mnemonic for angiod streaks' associations? What are these associations?*

**P**seudoxanthoma elasticum (PXE)

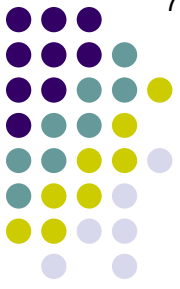
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*Which condition has the strongest association with angiod streaks?*  
PXE, by a mile



## CNVM DDx

# A

- CNVM DDx
  - *Briefly, what sort of disorder is PXE?*  
An elastorrhesis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

- *Is it common, or rare?*

- Rare

- *Is there a gender predilection?*

Yes, ♀

*What is the appearance of affected skin?*

An area of waxy-yellow, papule-like lesions

**Skin**

*What is the appearance of affected skin?*

Reddish

Bruch's

*Other than skin*

--Vasculature

--GI tract

--Eye

*What part of the body is affected?*

About 1/2500

*What is the well-known mnemonic for angiod streaks associations? What are these associations?*

**P**seudoxanthoma elasticum (PXE)

**E**hlers-Danlos syndrome

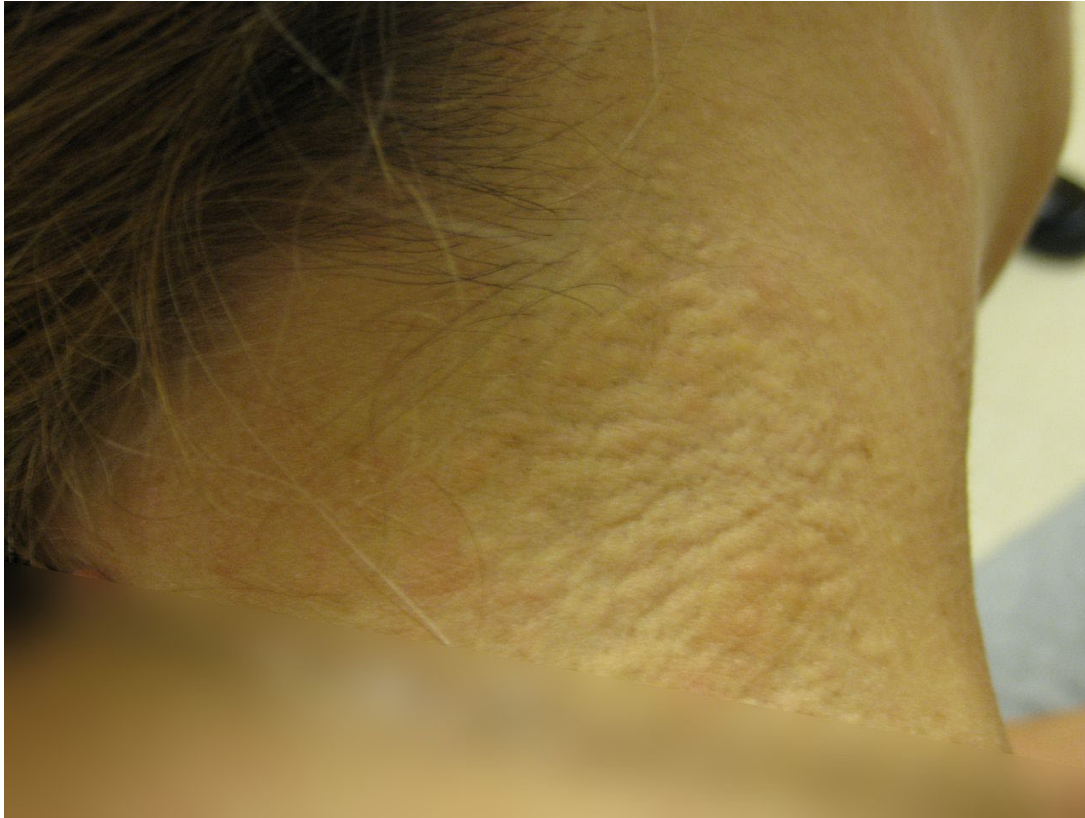
**P**aget's disease of bone

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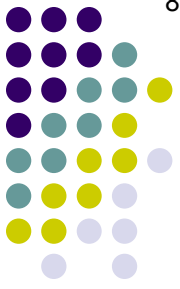
**I**diopathic (ie, no association)

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

# CNVM DDx



PXE skin



## CNVM DDx

Q

● CN

*Briefly, what sort of disorder is PXE?*

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

● A

*Is it common, or rare?*

● F

Rare

● A

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Yes, ♀

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Reddis

Bruch's

Other th

—Skin

*What is the classic informal descriptor for this appearance?*

--vascu

--GI trac

--Eye

What p

About

sent breaks in

*What is the well-known mnemonic for angiod streaks' associations? What are these associations?*

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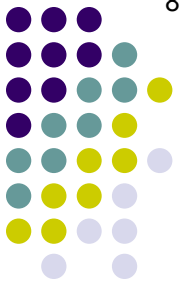
**S**ickle-cell disease

**I**diopathic (ie, no association)

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

PXE, by a mile





## CNVM DDx

# A

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Bruch's

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*What is the classic informal descriptor for this appearance?*

'Chicken skin'

What p

--vascu

About

--GI trac

--Eye

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PXE, by a mile



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Bruch's

---Skin

*What is the classic informal descriptor for this appearance?*

'Chicken skin'

What p

---vascu

About

---GI trac

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

---Eye

--

What is

---

*What is the well-known mnemonic for angiod streaks' associations? What are these associations?*

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

PXE, by a mile



## CNVM DDx

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- CN

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

- A

*Is it common, or rare?*

- F

Rare

- A

*Is there a gender predilection?*

Yes, ♀

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Reddis

Other th

Bruch's

---Skin

*What is the classic informal descriptor for this appearance?*

'Chicken skin'

What p

---vascu

About

---GI trac

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

---Eye

--The neck

--The axillae

What is the well-known mnemonic for angiod streaks' associations? What are these associations?

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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 angiod streaks?*

PXE, by a mile

## CNVM DDx



PXE: 'Chicken skin'



## CNVM DDx

Q

● CN

*Briefly, what sort of disorder is PXE?*

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

● A

*Is it common, or rare?*

● F

Rare

● A

*Is there a gender predilection?*

Yes, ♀ are twice as likely to be affected

What is

Reddis

Bruch's

What p

About

What is the well-

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

--Skin

--Vascul

--GI trac

--Eye

--

--

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

--Angioid streaks

--

--

sent breaks in

What are these associations?

**P**seudoxanthoma 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

# A

- CN

*Briefly, what sort of disorder is PXE?*

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

- A

*Is it common, or rare?*

- R

Rare

- A

*Is there a gender predilection?*

Yes, ♀ are twice as likely to be affected

*What is*

**Reddis**

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

Bruch's

--Skin

--Vascul

*What p*

--GI tract

About

--**Eye**

*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 is the well-*

*What are these associations?*

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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?  
PXE, by a mile*



## CNVM DDx

Q

● CN

*Briefly, what sort of disorder is PXE?*

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

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Rare

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What is

Reddis

Bruch's

What p

About

What is the well-

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

--Skin

--Vascul

--GI trac

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

Eye

RPE mottling

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

--Optic disc drusen

**Pseudoxanthoma elasticum (PXE)**

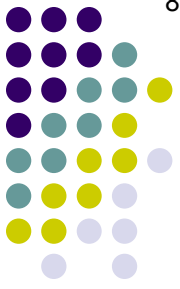
Ehlers-Danlos syndrome

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Sickle-cell disease

Idiopathic (ie, no association)

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



## CNVM DDx

# A

- CNVM
  - A: *Briefly, what sort of disorder is PXE?*  
An elastorrhesis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues
  - F: *Is it common, or rare?*  
Rare
  - A: *Is there a gender predilection?*  
Yes, ♀ are twice as likely to be affected

What is

Reddish

Bruch's

What p

About

What is the well-

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

--Skin

--Vascul

--GI trac

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

Eye

RPE mottling

*What mellifluous name is used to describe the RPE mottling?*  
**Peau d'orange**

**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

sent breaks in

What are these associations?



# CNVM DDx



PXE: *Peau d'orange* fundus

## CNVM DDx



- CNVM
  - *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

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

--SKIN

--Vascul

--GI trac

--Eye

*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**

*What is the well-*

*known association with PXE?*

*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)

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

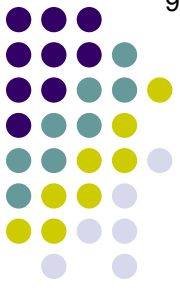


## CNVM DDx

# Q

- 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?*



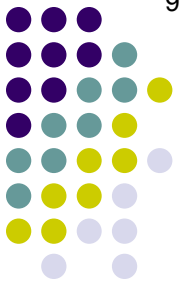
## CNVM DDx

# A

- 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?*

**26.5 mm**



## CNVM DDx

Q

- CNVM DDx:

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

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

*ok, what axial length cutoff for defining*

*a?*

**26.5 mm**



## CNVM DDx

# A

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

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

ok, what axial length cutoff for defining

26.5 mm



## CNVM DDx

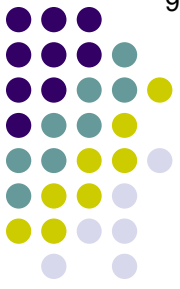
# Q

- 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?*

26.5 mm

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



## CNVM DDx

# A

- 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?*

26.5 mm

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

**Lacquer cracks**





## CNVM DDx

# 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*

*...focusing on DFE that ask for CNVM?*

	Angioid streaks	Lacquer cracks
Color	?	?



# Q/A

## CNVM DDx

- 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*

*...ing on DFE that ask for CNVM?*

	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish

## CNVM DDx



**Angioid streaks**  
'Reddish-brown'



**Lacquer cracks**  
'Yellowish'



# Q/A

## CNVM DDx

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

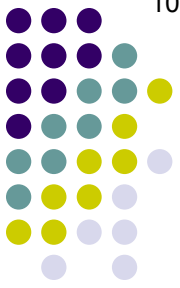
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*...ing on DFE that ask for CNVM?*

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



# Q/A

## CNVM DDx

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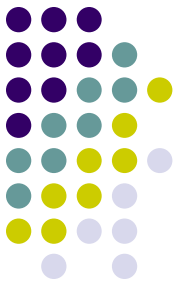
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*Angioid streaks vs lacquer cracks: Compare and contrast*

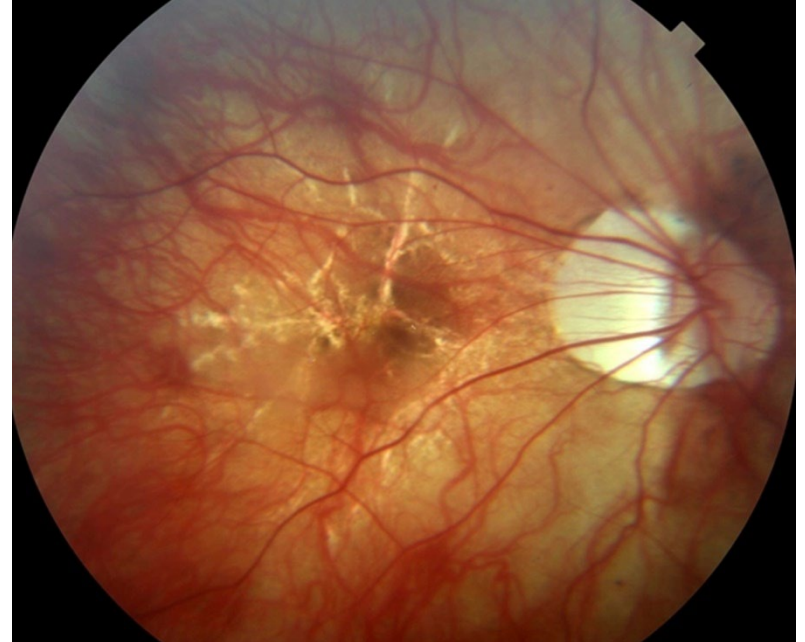
*...ing on DFE that ask for CNVM?*

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

## CNVM DDx



**Angioid streaks**  
'Reddish-brown'  
'Peripapillary'



**Lacquer cracks**  
'Yellowish'  
'Macular'



# Q/A

## CNVM DDx

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

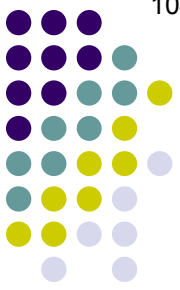
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	Angioid streaks	Lacquer cracks
Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	?	?



# Q/A

## CNVM DDx

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Color	Reddish-brown	Yellowish
Location	Peripapillary	Macula
Subretinal heme?	Yes	Yes
Locus for CNV?	?	?





# Q/A

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Subretinal heme?	Yes	Yes
Locus for CNV?	Yes	Yes
Associated with...	?	?



## CNVM DDx

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*...ing on DFE that ask for CNVM?*

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



# Q

- CNVM DDx:

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

## 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

**Sorsby macular dystrophy**



# A

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
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A macular dystrophy

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*What is the inheritance pattern?*

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AD

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- CNVM DDx:
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A macular dystrophy

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*What is the classic DFE finding in a pt who has the condition, but has yet to become symptomatic?  
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**Sorsby macular dystrophy**

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A macular dystrophy

*What is the inheritance pattern?*

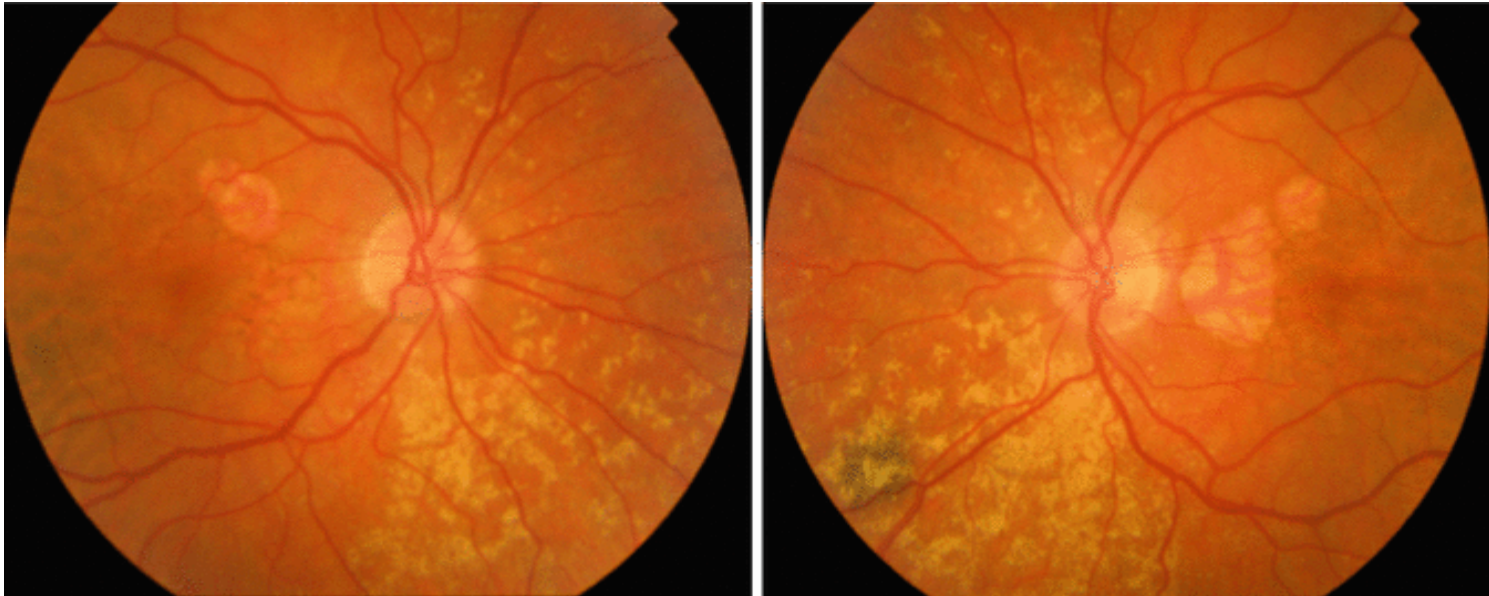
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*What is the classic DFE finding in a pt who has the condition, but has yet to become symptomatic?  
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A young adult with large numbers of "drusenlike deposits" in the macula bilaterally



# CNVM DDx



Sorsby: Drusenlike deposits

# Q

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

laterality, and location

CNVMs by age # or so

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

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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

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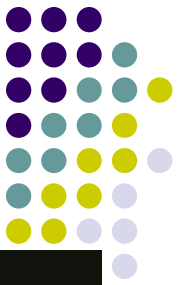
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Bilateral subfoveal CNVMs by age 40 or so

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. 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)*

## CNVM DDx



Sorsby: Late extensive fibrosis/scarring

Q

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
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*What is the inheritance pattern?*

AD

*What is the subheader name?*

'Early-onset "drusenoid" macular dystrophies'

*What is the classic presentation of a young adult with large numbers of "drusenlike deposits" in the macula bilaterally?  
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*For context and completeness' sake: What are the other early-onset "drusenoid" macular dystrophies with which Sorsby's is grouped?*

--?

--?

*(Hints forthcoming)*

--?

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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--? *The most common inherited macular dystrophy*

--?

--?

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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  - ARMD
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--Stargardt

--?

--?

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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--? *The best macular dystrophy?*

--?

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

# A

- CNVM DDx:
  - ARMD
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--Best dz

--?

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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--Stargardt

--Best dz

--? *Best dz's older sibling*

--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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--Best dz

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--?

--?

--?

Bilateral subfoveal CNVMs by age 40 or so



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

--?

--?

Bilateral subfoveal CNVMs by age 40 or so

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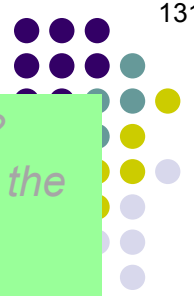
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--?

--?

Bilateral subfoveal CNVMs by age 40 or so



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- Stargardt
- Best dz
- Adult-onset foveomacular vitelliform dystrophy
- The pattern dystrophies
- ? *Not breast-related, despite its name*
- ?

Bilateral subfoveal CNVMs by age 40 or so

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--Best dz

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--The pattern dystrophies

--Central areolar choroidal dystrophy

--?

Bilateral subfoveal CNVMs by age 40 or so

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**Sorsby macular dystrophy**

--The pattern dystrophies

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--? *The only one named for a US state*

Bilateral subfoveal CNVMs by age 40 or so

# A

- CNVM DDx:
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  - Choroidal rupture
  - Iatrogenic

## 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

--Best dz

--Adult-onset foveomacular vitelliform dystrophy

--The pattern dystrophies

--Central areolar choroidal dystrophy

--North Carolina macular dystrophy

Bilateral subfoveal CNVMs by age 40 or so