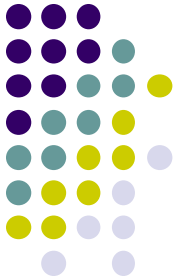
# Retinitis Pigmentosa



*A note before we begin:* The Academy seems to be phasing out the term *retinitis pigmentosa*. Further, the scope of conditions covered by the term is shrinking. The point being, the facts concerning RP are in flux at the moment, and may have changed by the time you read this. *Caveat emptor!*

Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor brightness vision
--?
--?
--?

Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--***?***
--*?*
--*?*

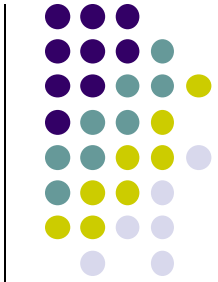
Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
- [redacted] VF
--?
--?

Retinitis Pigmentosa

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--?
--?

*(We will drill down on this shortly)*

Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor **scotopic** vision
--**constricted** VF
--*?*
--*?*

**Night blindness** and **progressive peripheral VF loss** are the two **hallmark symptoms** of RP

Retinitis Pigmentosa

# Retinitis Pigmentosa

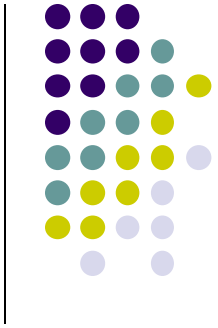
Group of inherited retinal diseases characterized by:
--poor **scotopic** vision
--constricted VF
--*?*
--*?*

**Night blindness** and **progressive peripheral VF loss** are the two **hallmark symptoms** of RP

*Some RP pts do not c/o poor night vision. Why not?*

Retinitis Pigmentosa

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--?
--?

**Night blindness** and progressive peripheral VF loss are the two hallmark symptoms of RP

*Some RP pts do not c/o poor night vision. Why not*?
Such pts have had poor scotopic vision their entire lives, and thus are unable to recognize it; ie, their poor night vision seems normal to them

Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal abb.
--?

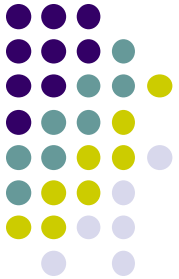
Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG *(This too)*
*--?*

Retinitis Pigmentosa

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--*?*

**Night blindness, VF loss** and **abnormal ERG** are the **defining features of RP**. If it ain't got all three, it very likely ain't RP!

Retinitis Pigmentosa

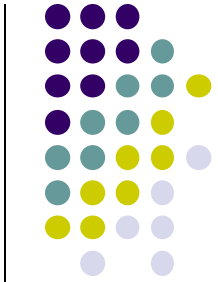
# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Retinitis Pigmentosa

*(No question yet—proceed when ready)*

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

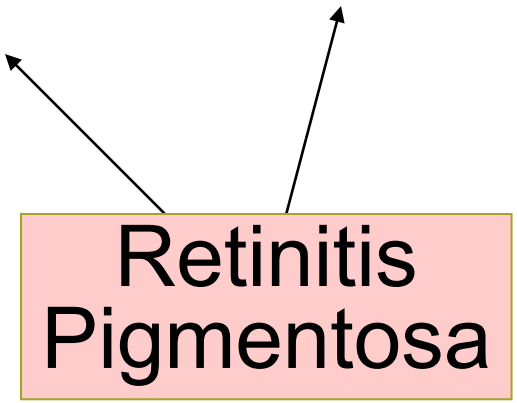
Classic fundus appearance:
-- two words
--?
--?

Retinitis Pigmentosa

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--*?*
--*?*



# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appe
**--Bone spicules**
--*?*
--*?*

*What are* bone spicules*?*

Retinitis Pigmentosa

# Retinitis Pigmentosa

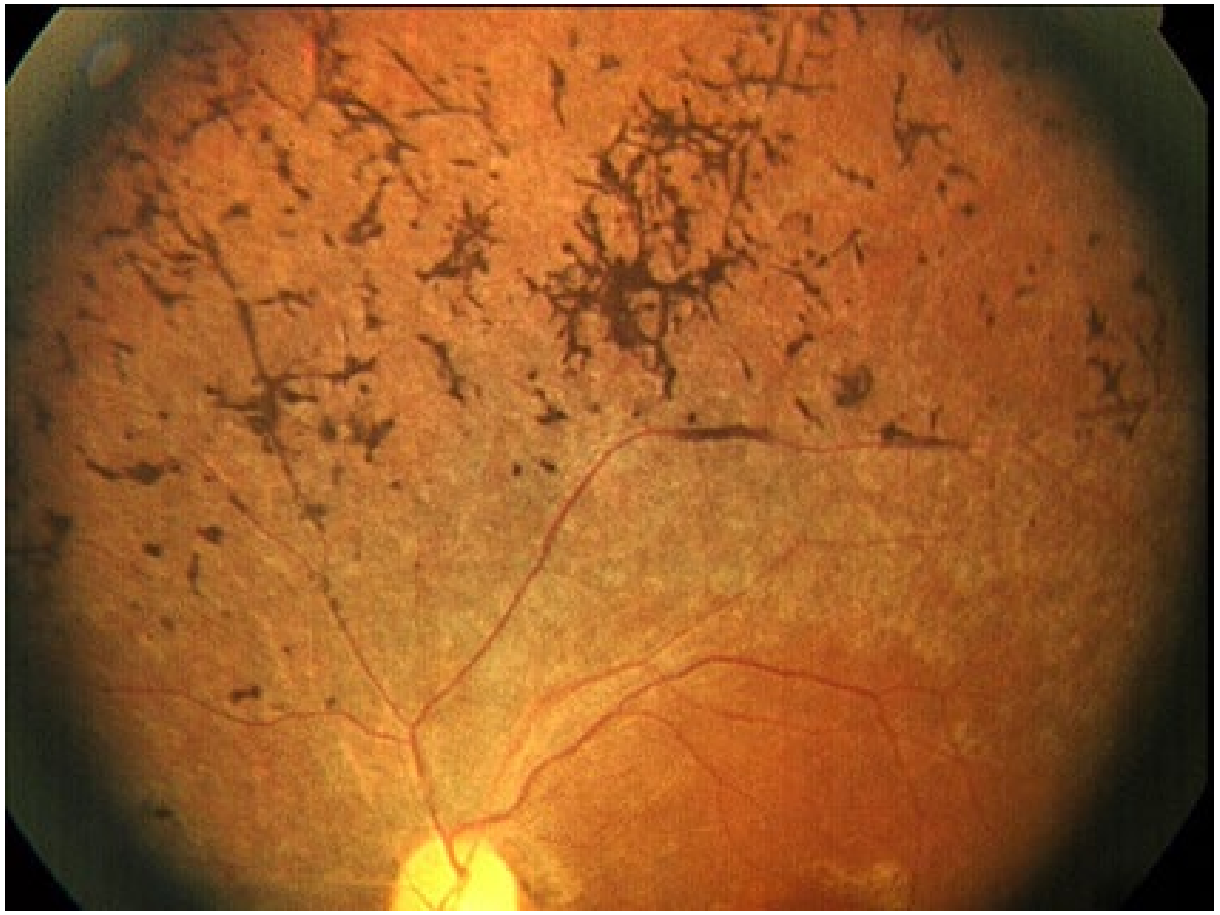
Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appe
**--Bone spicules**
--*?*
--*?*

*What are* bone spicules*?*
Focal accumulations of pigment released when dying RPE cells disintegrate

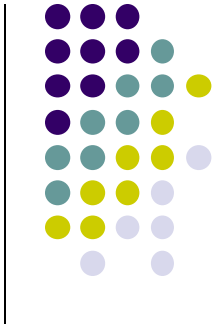
Retinitis Pigmentosa

# Retinitis Pigmentosa



RP: Bone spicules

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

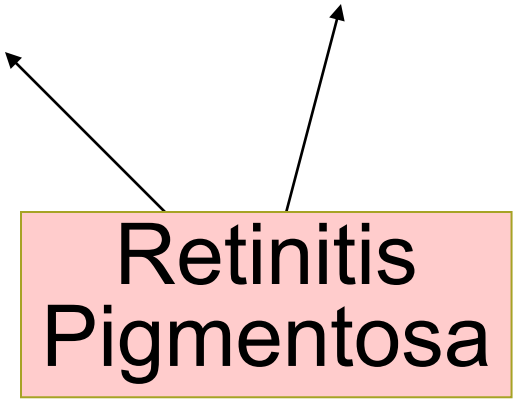
Classic fundus appearance:
--Bone spicules
--classic description disc pallor
--?

Retinitis Pigmentosa

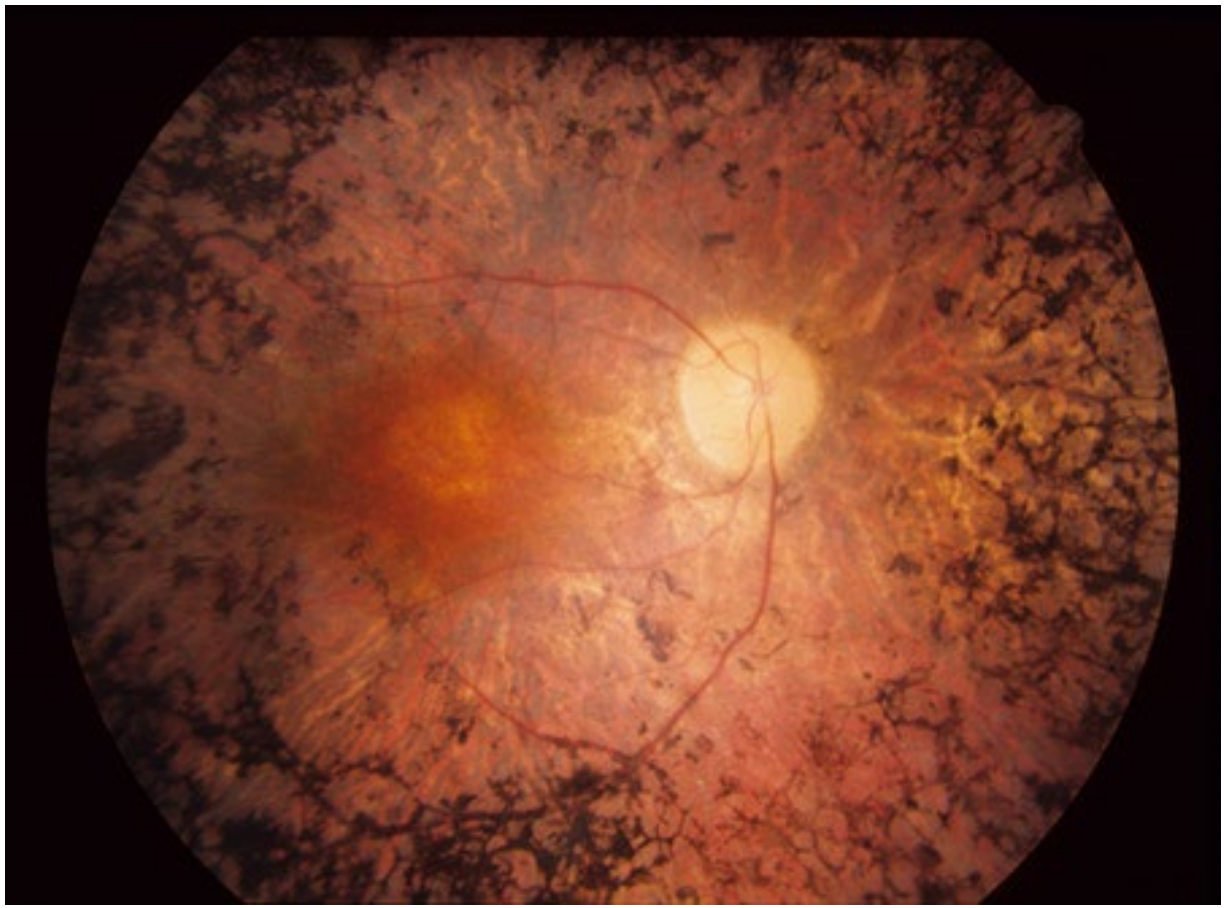
# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--?



# Retinitis Pigmentosa

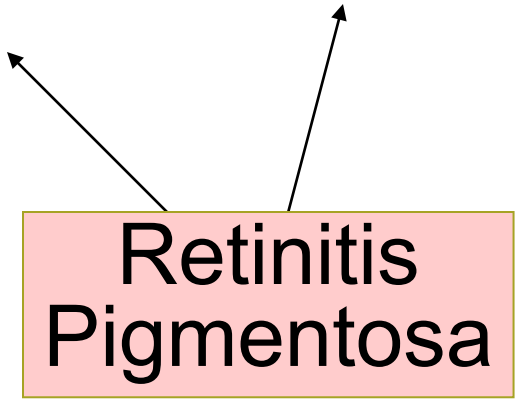


RP: Waxy disc pallor

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

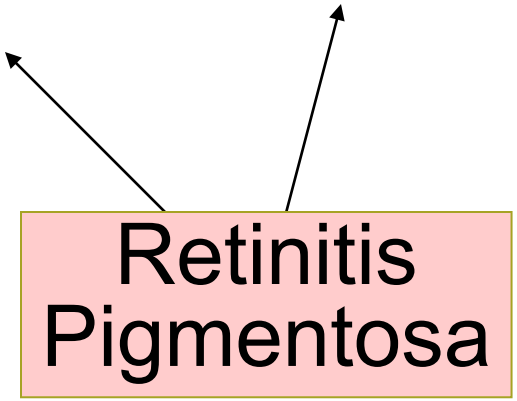
Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--Arteriolar [redacted]



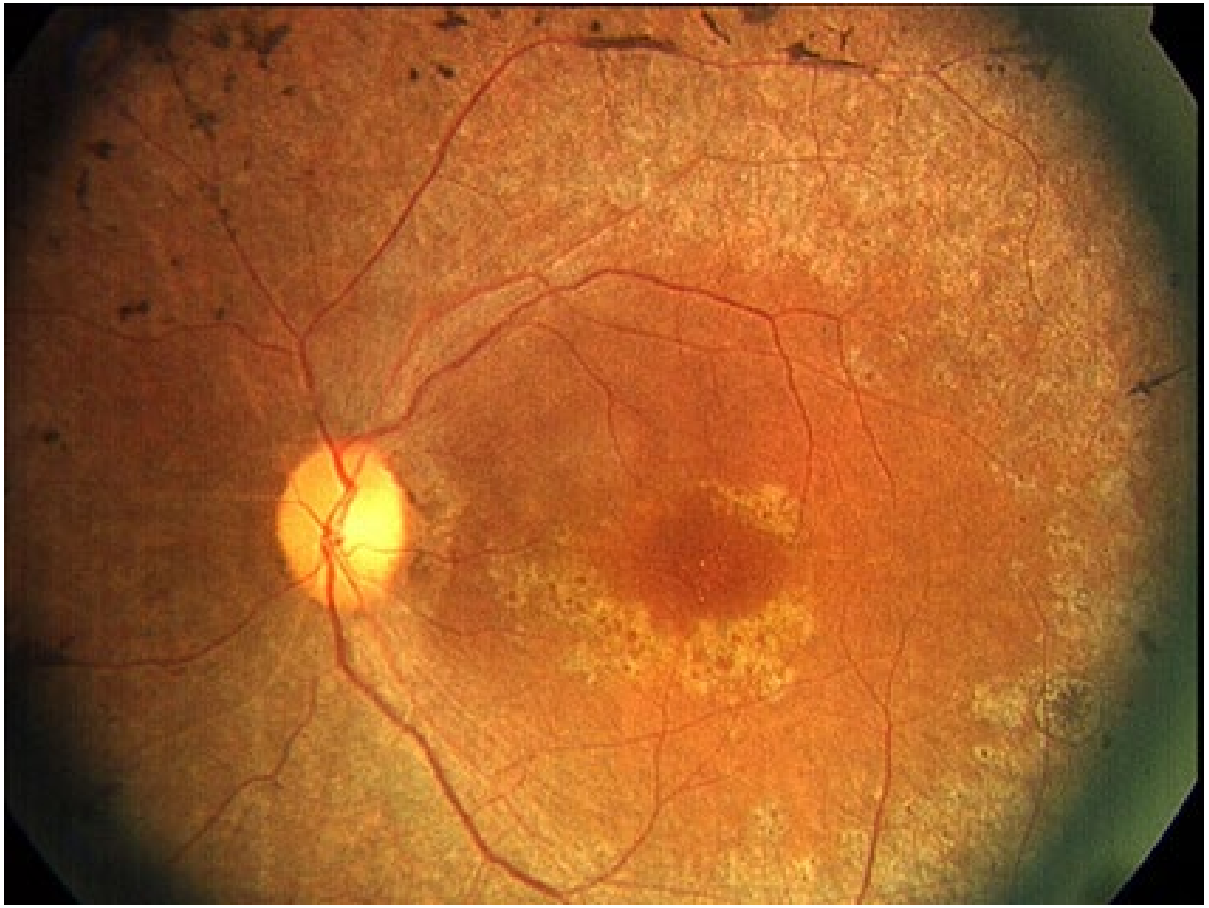
# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--Arteriolar narrowing

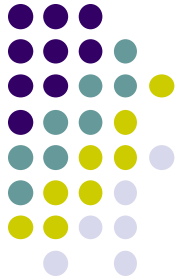


# Retinitis Pigmentosa



RP: Arteriolar narrowing

# Retinitis Pigmentosa



Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--Arteriolar narrowing

*Which of the three appears first?*

Retinitis Pigmentosa

# Retinitis Pigmentosa

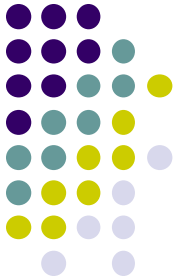
Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--**Arteriolar narrowing**

*Which of the three appears first?*
Arteriolar narrowing

Retinitis Pigmentosa

# Retinitis Pigmentosa



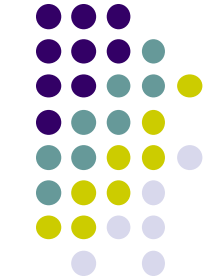
Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Possible* **fundus appearance:**
**--Bone spicules**
**--Waxy disc pallor**
**--Arteriolar narrowing**

**?**

*What infectious (and therefore treatable) disease can produce a similar fundus appearance, and must always, **always** be at least considered in a patient with an RP-like fundus?*

# Retinitis Pigmentosa



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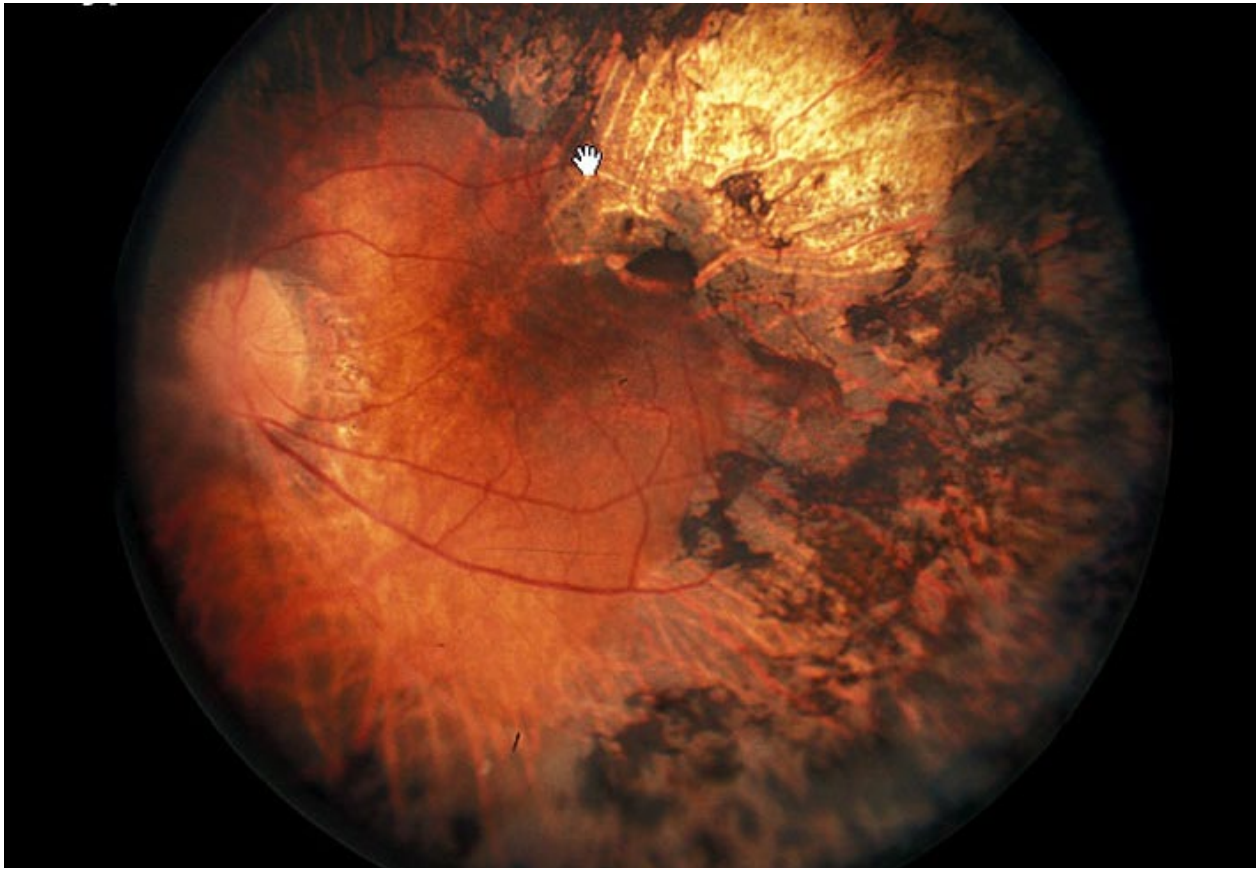
--abnormal ERG
--characteristic fundus appearance

*Possible* **fundus appearance:**
**--Bone spicules**
**--Waxy disc pallor**
**--Arteriolar narrowing**

Syphilis

*What infectious (and therefore treatable) disease can produce a similar fundus appearance, and must always, **always** be at least considered in a patient with an RP-like fundus?*
Syphilis

# Retinitis Pigmentosa



RP-like fundus in syphilis

# Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

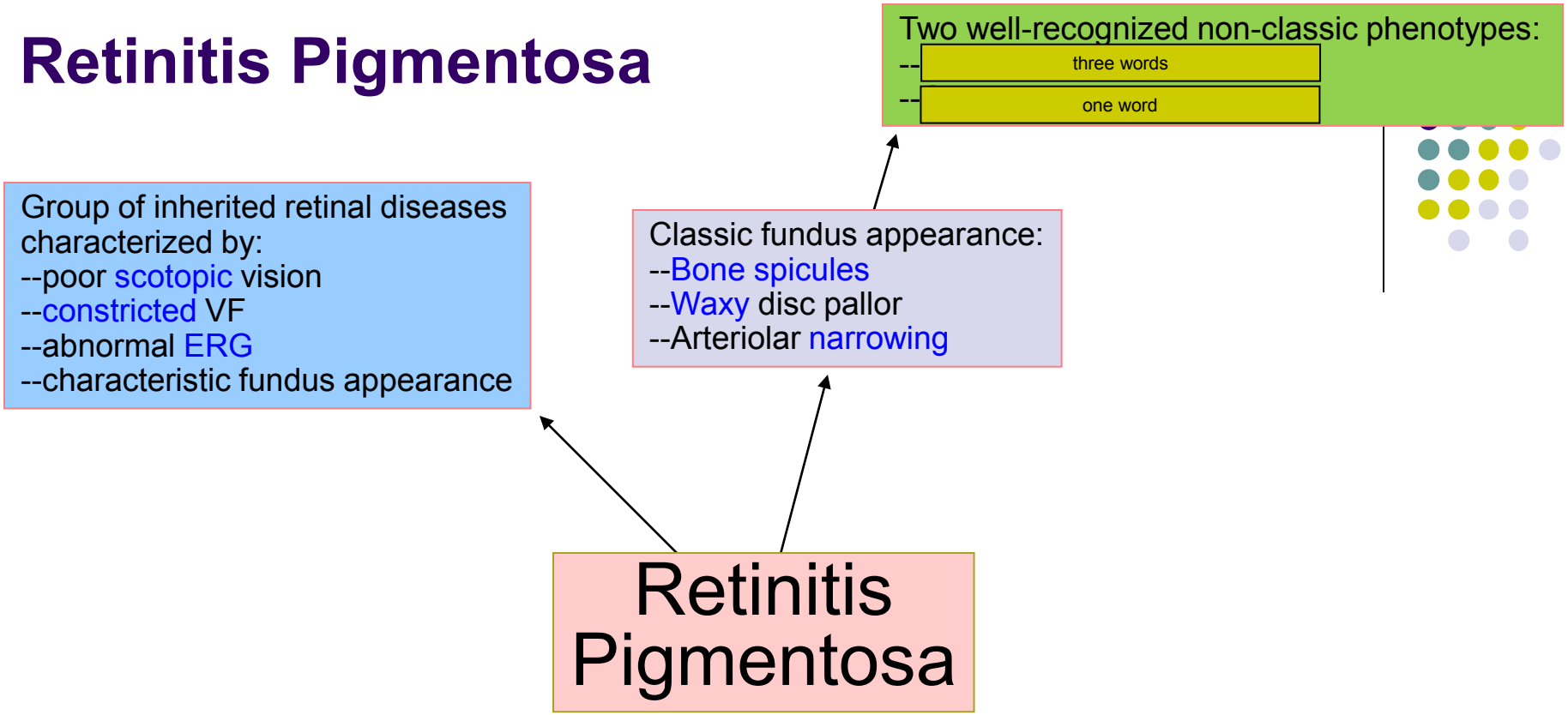
*Possible* **fundus appearance:**
**--Bone spicules**
**--Waxy disc pallor**
**--Arteriolar narrowing**

*Syphilis*

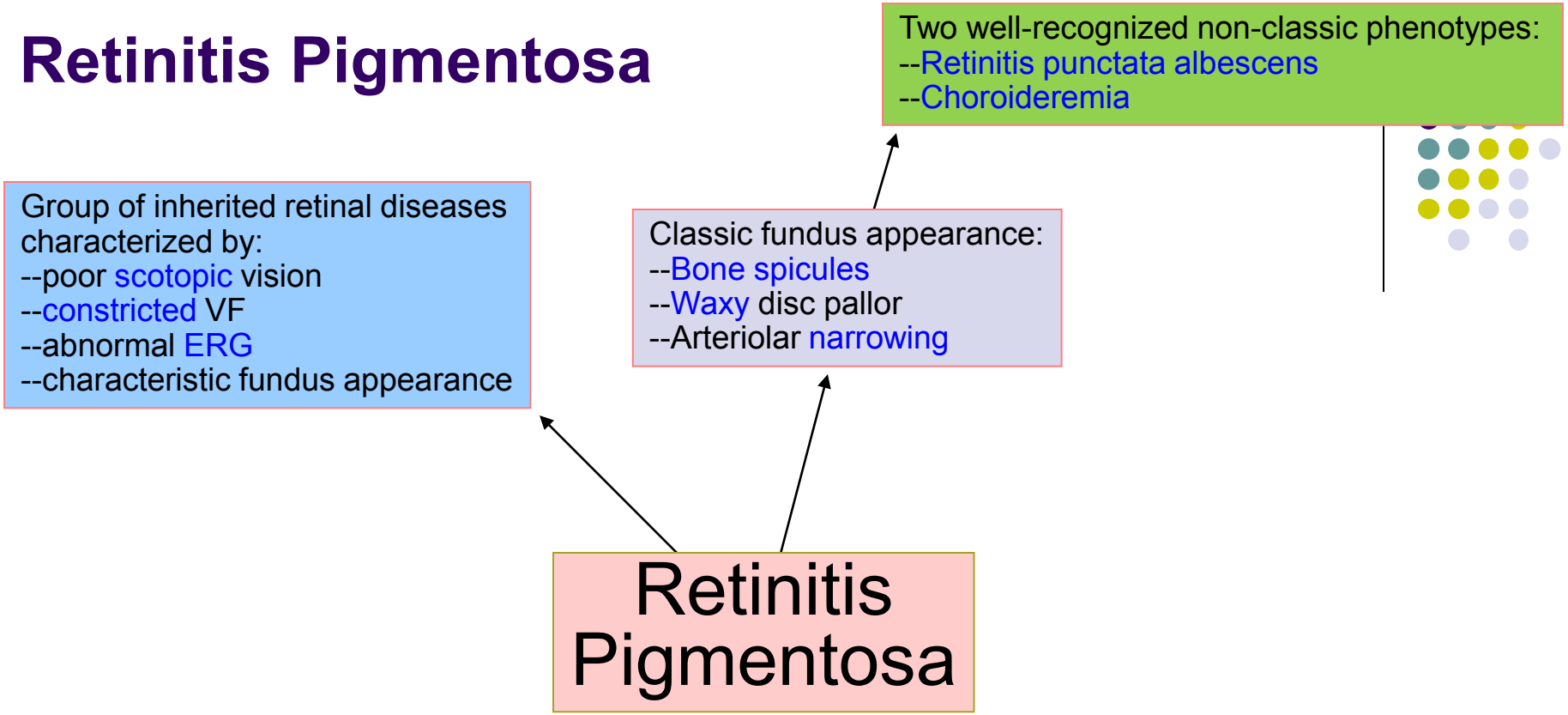
There are a number of other conditions that can mimic RP. Once you've got this set on lock, consider reviewing *R5, Differential for a Retinitis Pigmentosa-like Fundus*.

*similar fundus appearance, and must always, **always** be at least considered in a patient with an RP-like fundus?*
Syphilis

# Retinitis Pigmentosa



# Retinitis Pigmentosa



# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--**Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*

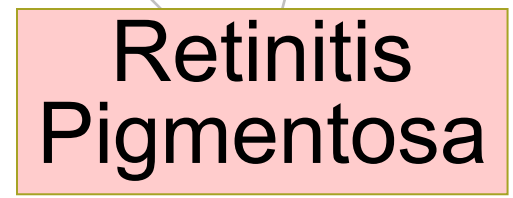
Retinitis Pigmentosa

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--**Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad color **dots/flecks** in the deep retina



# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
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Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
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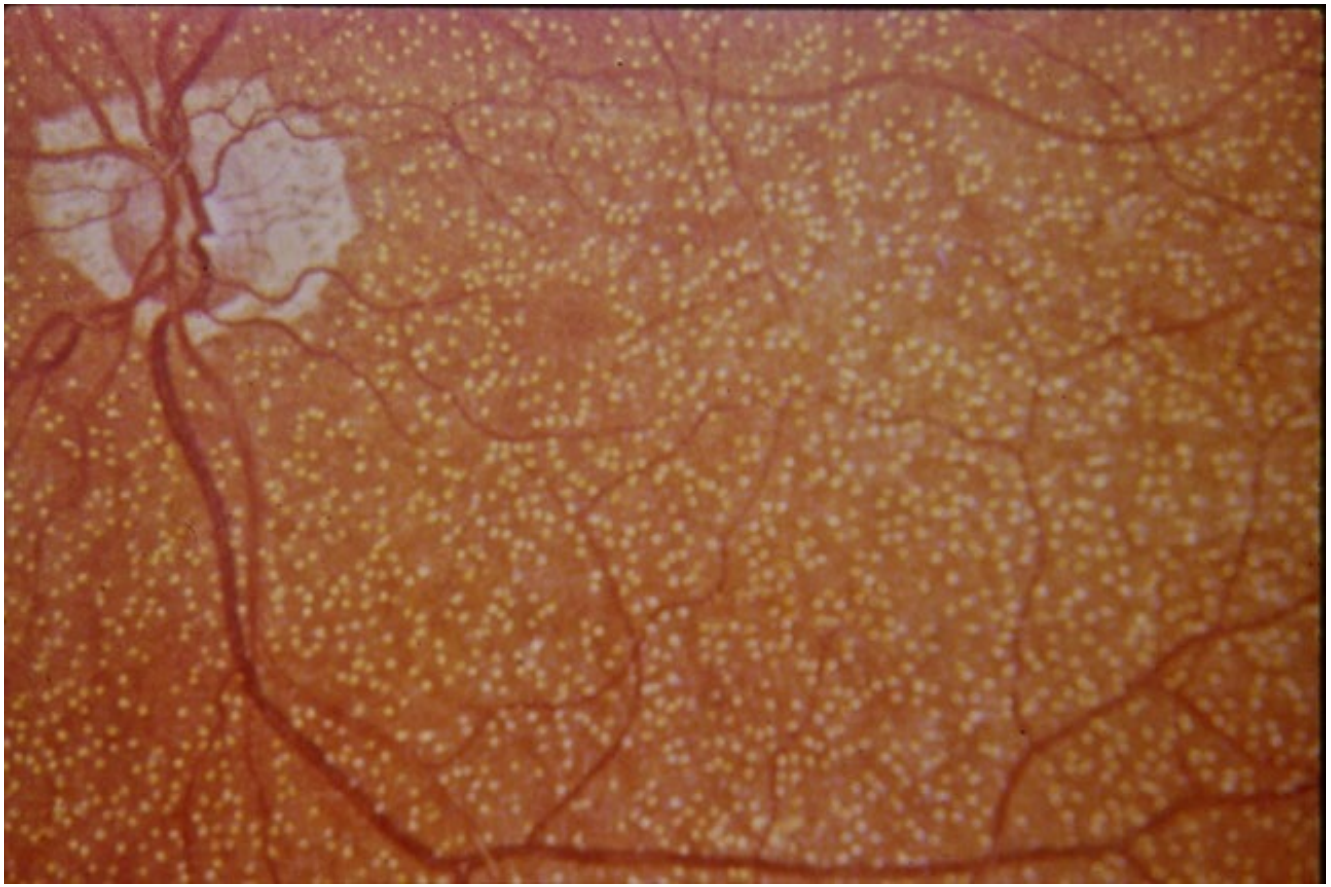
*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

Retinitis Pigmentosa

# Retinitis Pigmentosa



Retinitis punctata albescens

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--**Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

I remember the dots are white because *albescens* sounds like *albino*

Retinitis Pigmentosa

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
--**Choroideremia**

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*What is the hallmark of choroideremia?*

Retinitis Pigmentosa

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
--**Choroideremia**

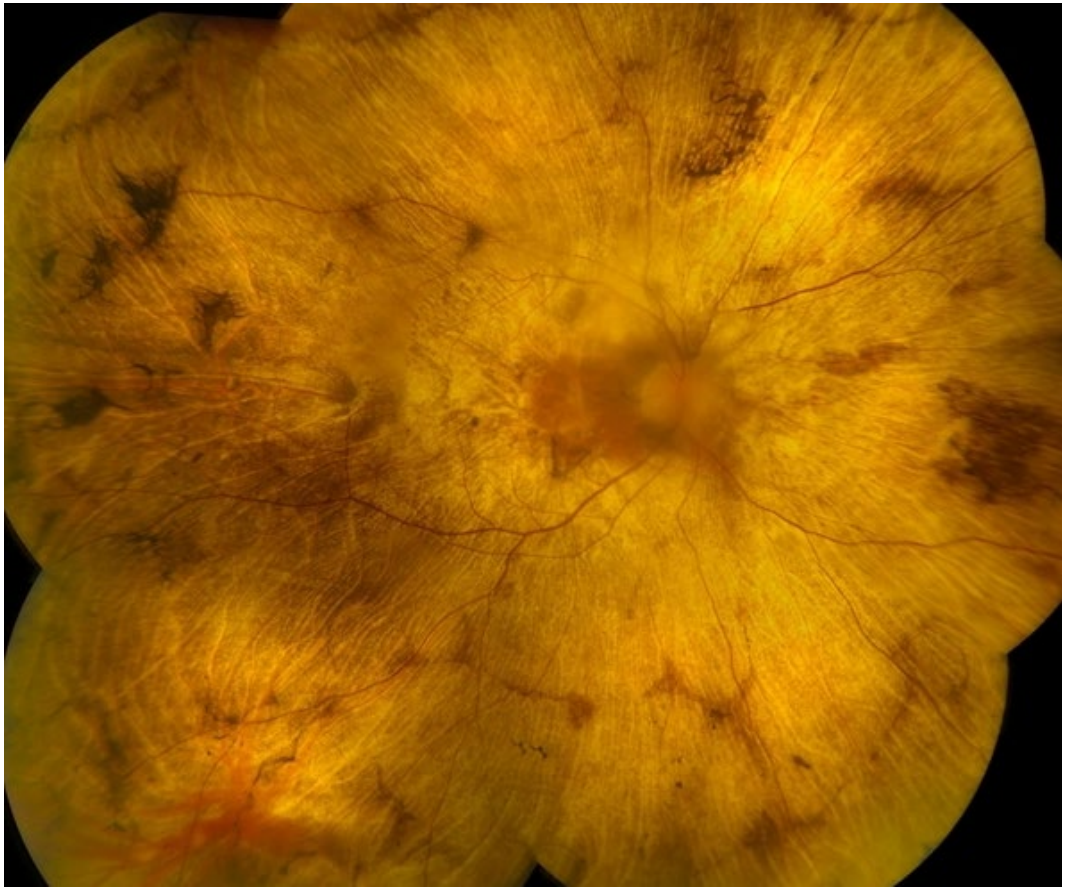
Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*What is the hallmark of choroideremia?*
Pronounced atrophic changes of the RPE, choriocapillaris and choroid

Retinitis Pigmentosa

# Retinitis Pigmentosa



Choroideremia

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
--**Choroideremia**

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*What is the hallmark of* ***choroideremia****?*
Pronounced atrophic changes of the RPE, choriocapillaris and choroid

Note: Whether choroideremia is a form of RP is one of the points in flux. The most recent *Retina* revision states it is not, whereas the immediately preceding version—which, at the time of this writing, is still being used by residents-in-training—maintains that it is. Caveat emptor.

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*W*
Pr
oroid

*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
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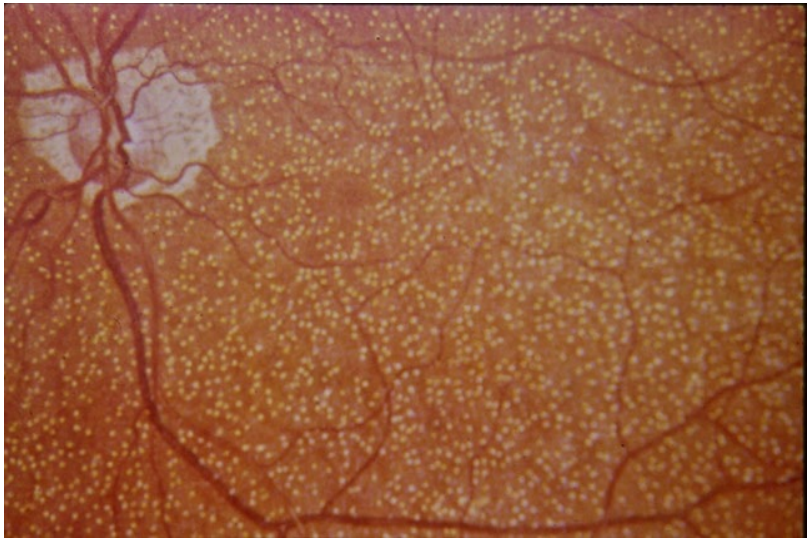
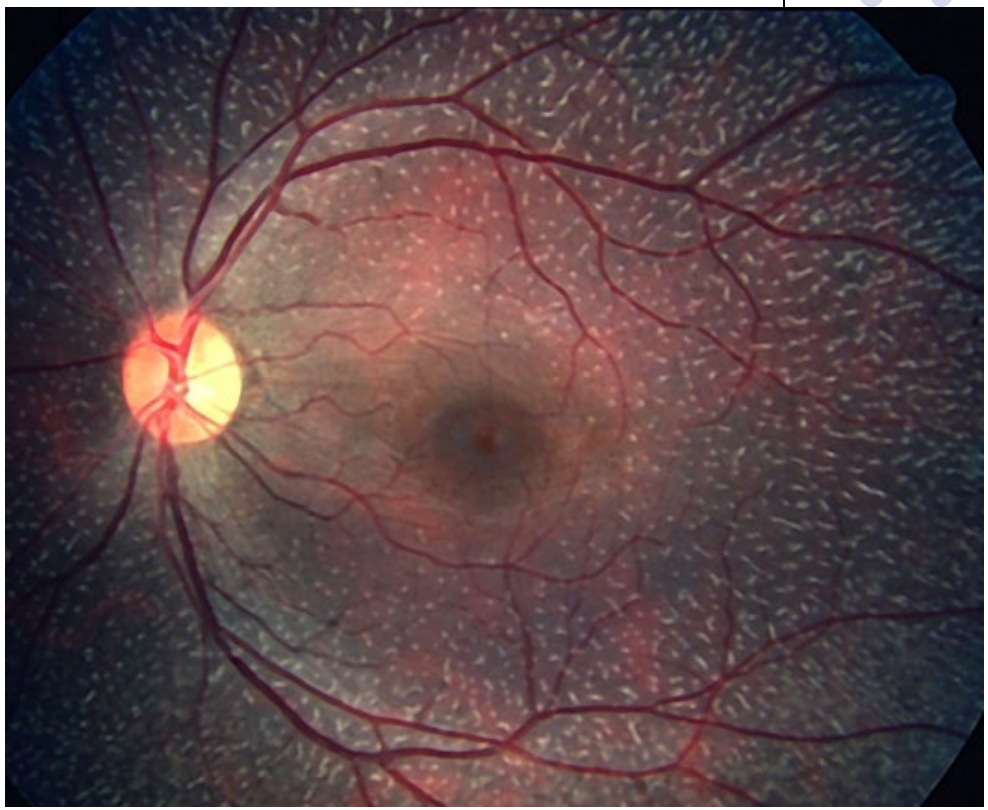
*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*
Fundus albipunctatus

# Retinitis Pigmentosa



Retinitis punctata albescens



Fundus albipunctatus

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
Myriad **white dots/flecks** in the deep retina

*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*
Fundus albipunctatus

*Is retinitis punctata albescens the same thing as fundus albipunctatus?*

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

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--poor scotopic vision
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*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*
Fundus albipunctatus

*Is retinitis punctata albescens the same thing as fundus albipunctatus?*
No, and it's very important to know the difference

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
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*Is retinitis punctata albescens the same thing as fundus albipunctatus?*
No, and it's very important to know the difference

*OK, what's the difference?*

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
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--poor scotopic vision
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Myriad **white dots/flecks** in the deep retina

*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*
Fundus albipunctatus

*Is retinitis punctata albescens the same thing as fundus albipunctatus?*
No, and it's very important to know the difference

*OK, what's the difference?*
Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of abb.

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
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*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
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*In terms of both its name and appearance, with what disease is retinitis punctata albescens often confused?*
Fundus albipunctatus

*Is retinitis punctata albescens the same thing as fundus albipunctatus?*
No, and it's very important to know the difference

*OK, what's the difference?*
Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of CSNB

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
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*Appearance-wise, what is the hallmark of retinitis punctata albescens?*
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Fundus albipunctatus

*Is retinitis punctata albescens the same thing as fundus albipunctatus?*
No, and it's very important to know the difference

*OK, what's the difference?*
Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of CSNB

*What does* CSNB *stand for?*

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
--Choroideremia

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
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No, and it's very important to know the difference

*OK, what's the difference?*
Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of CSNB

*What does* CSNB *stand for?*
Congenital stationary night blindness

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
**--Retinitis punctata albescens**
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Group of inherited retinal diseases characterized by:
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*W*
Pr
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*What does it mean to say the night blindness is* stationary*?*
It means it is nonprogressive, an important way in which fundus albipunctatus differs from retinitis punctata albescens (which, like all RP, is relentlessly progressive)

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Like other forms of RP, retinitis punctata albescens demonstrates [two words], whereas fundus albipunctatus does not

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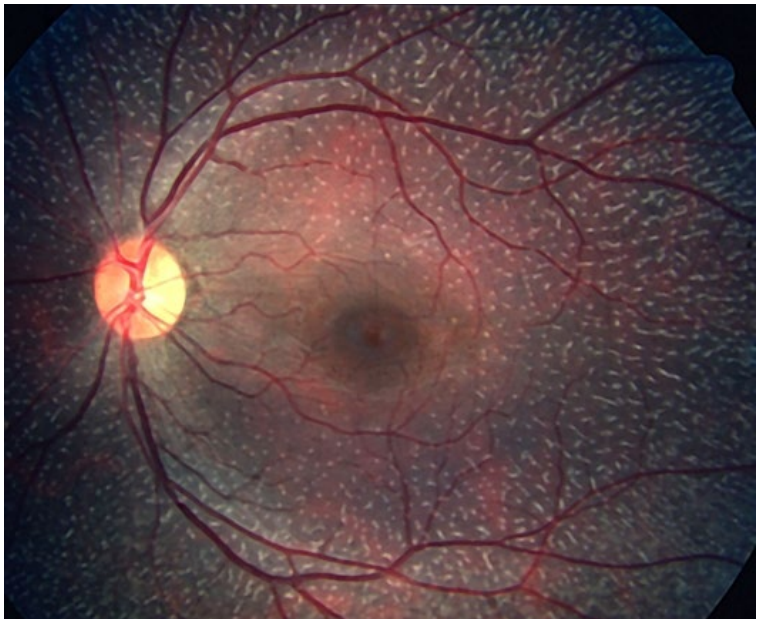
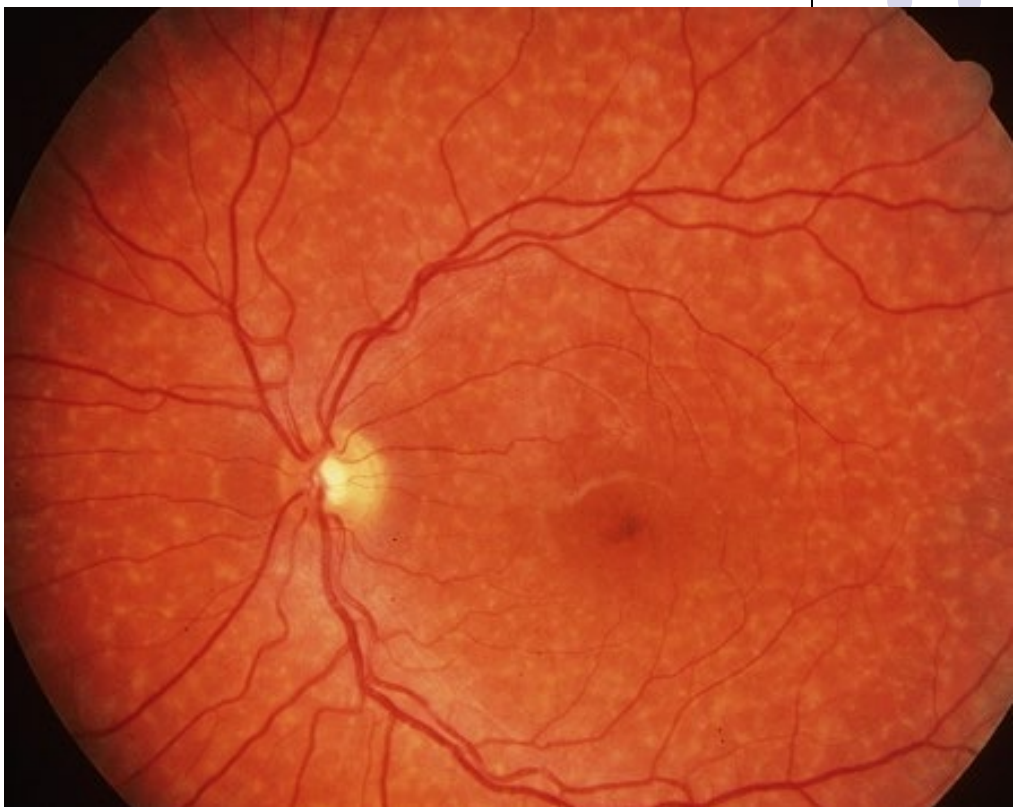


Fundus albipunctatus



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Juvenile macular dystrophy

*(Of historical interest only; this name is now considered obsolete)*

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Decreased visual acuity

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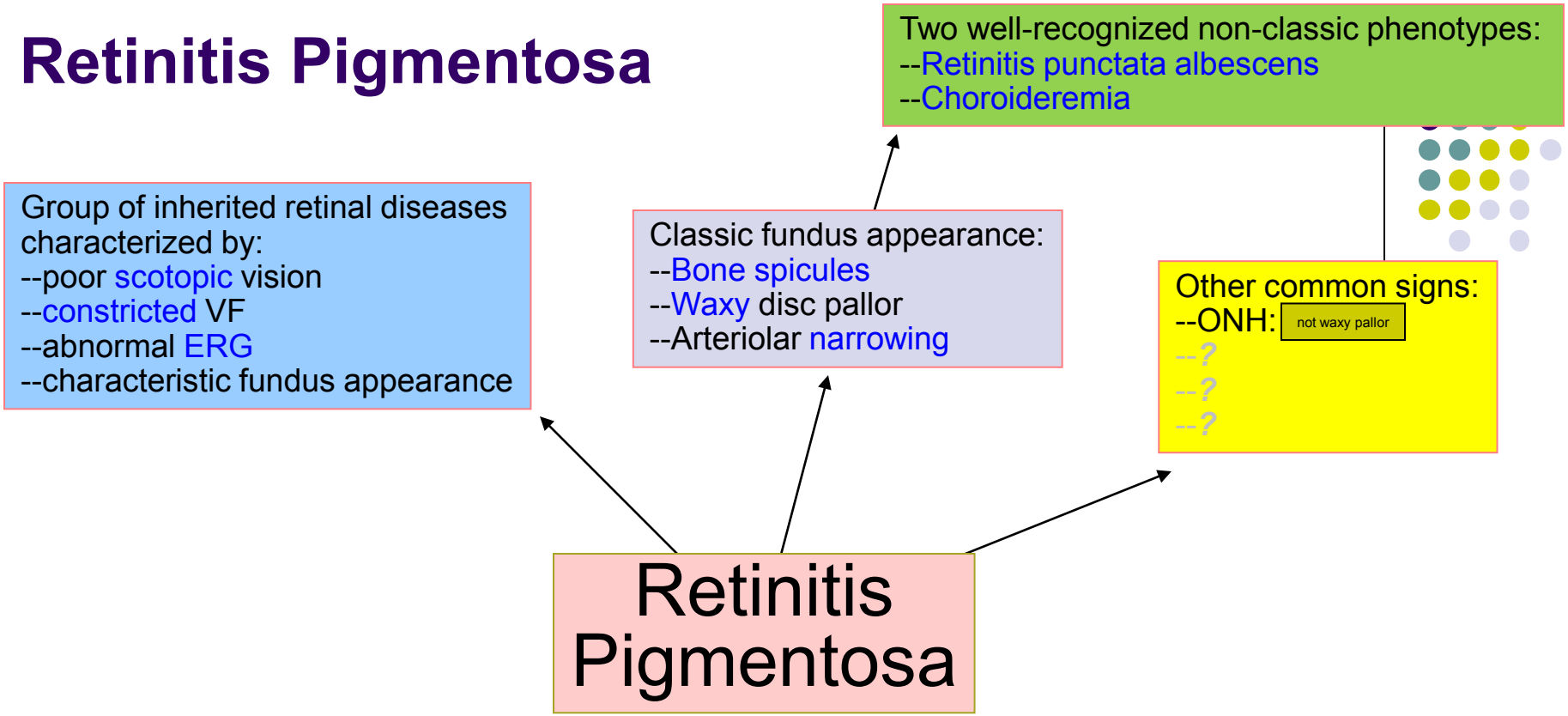

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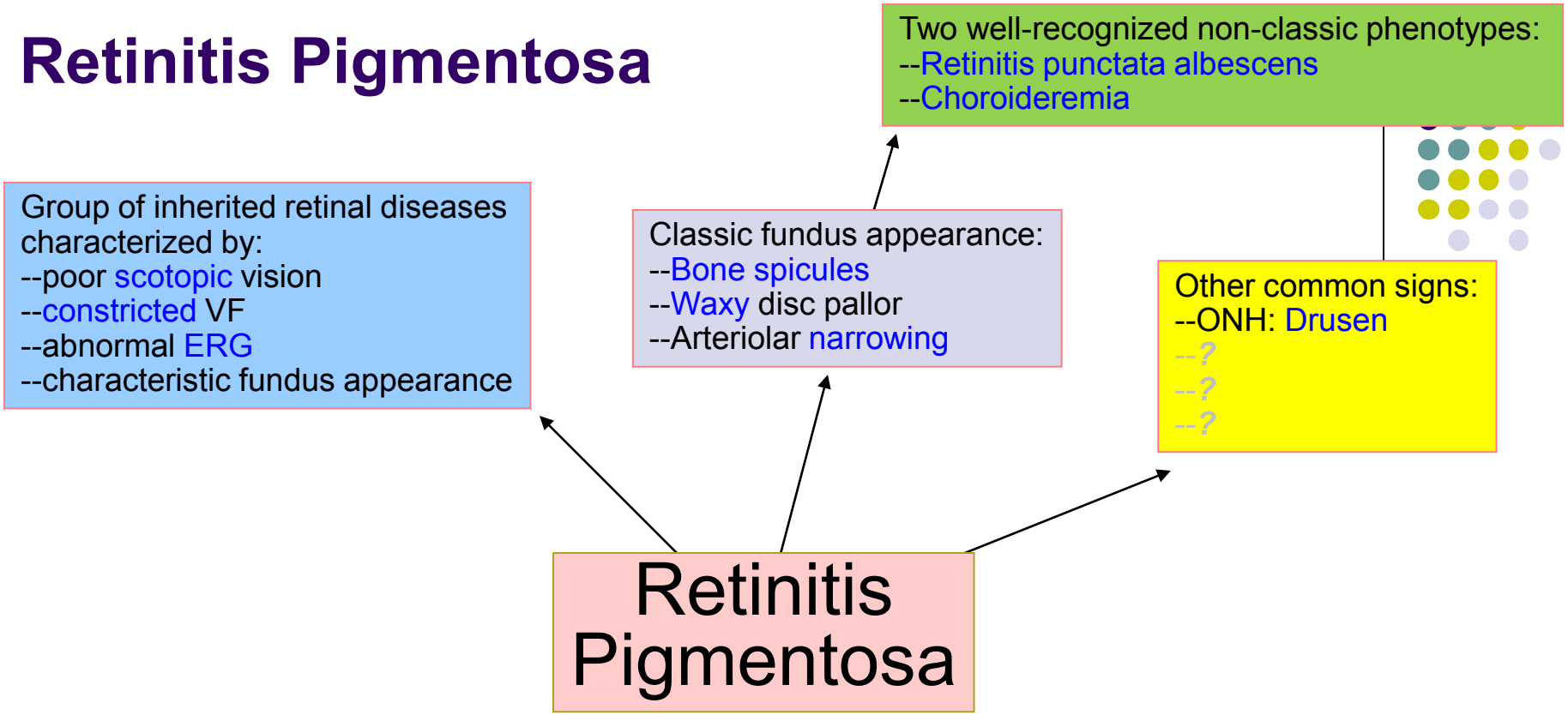

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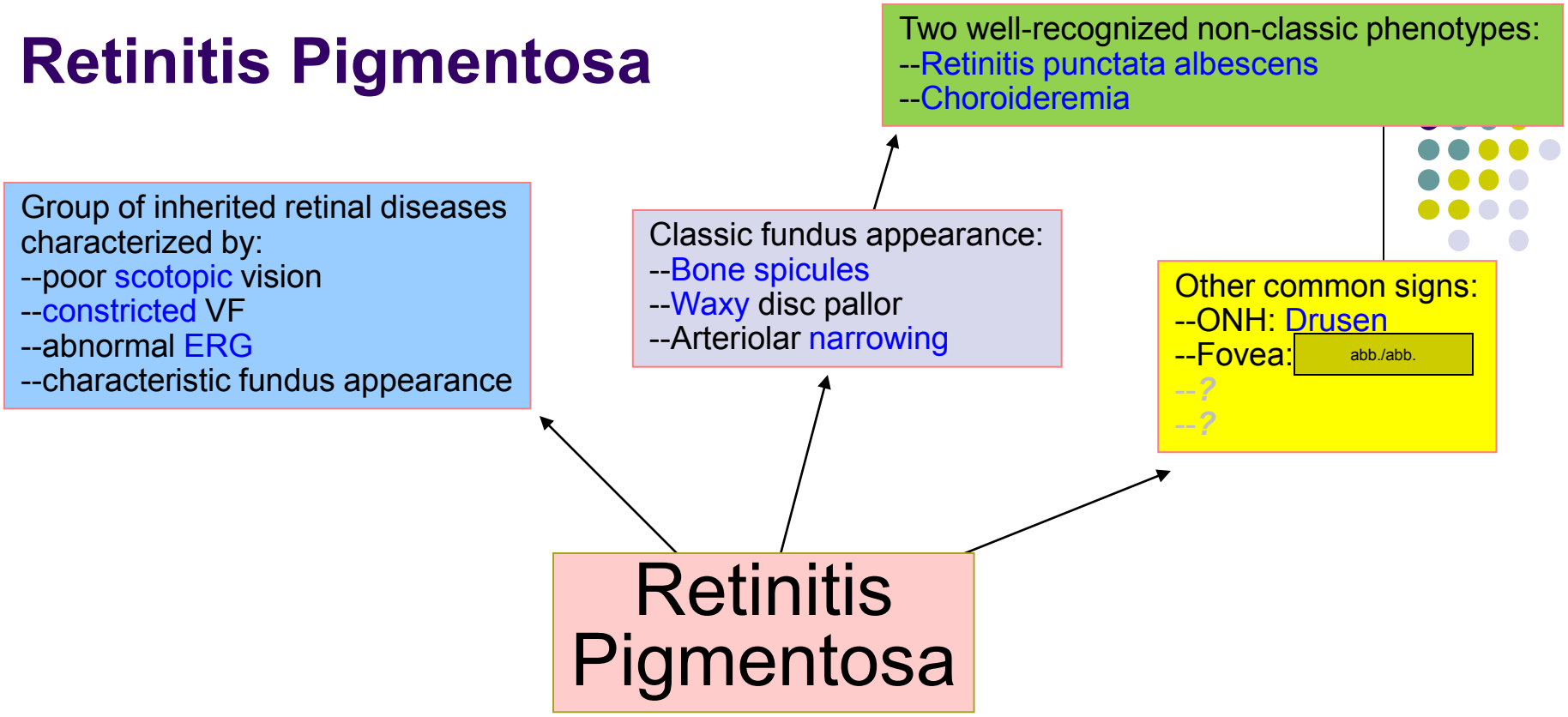

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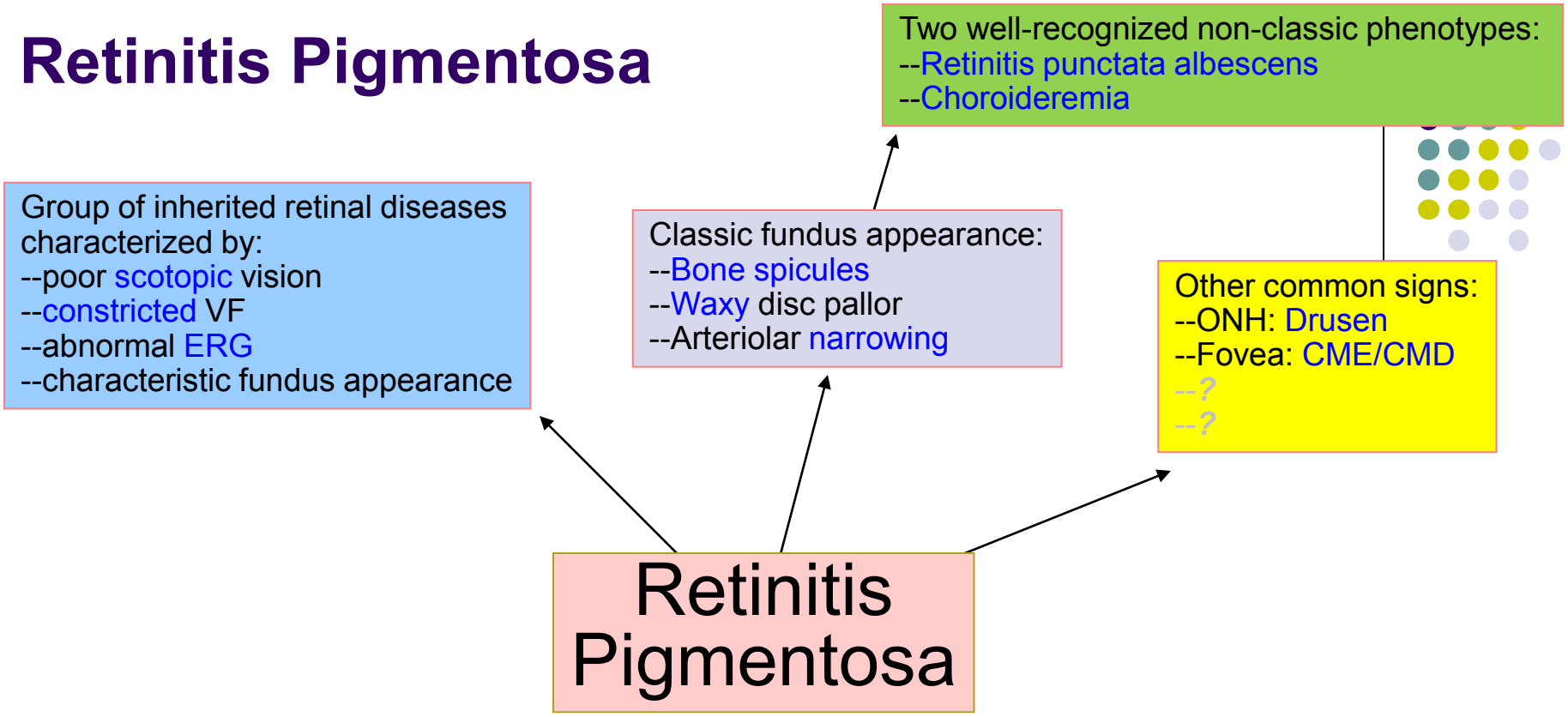

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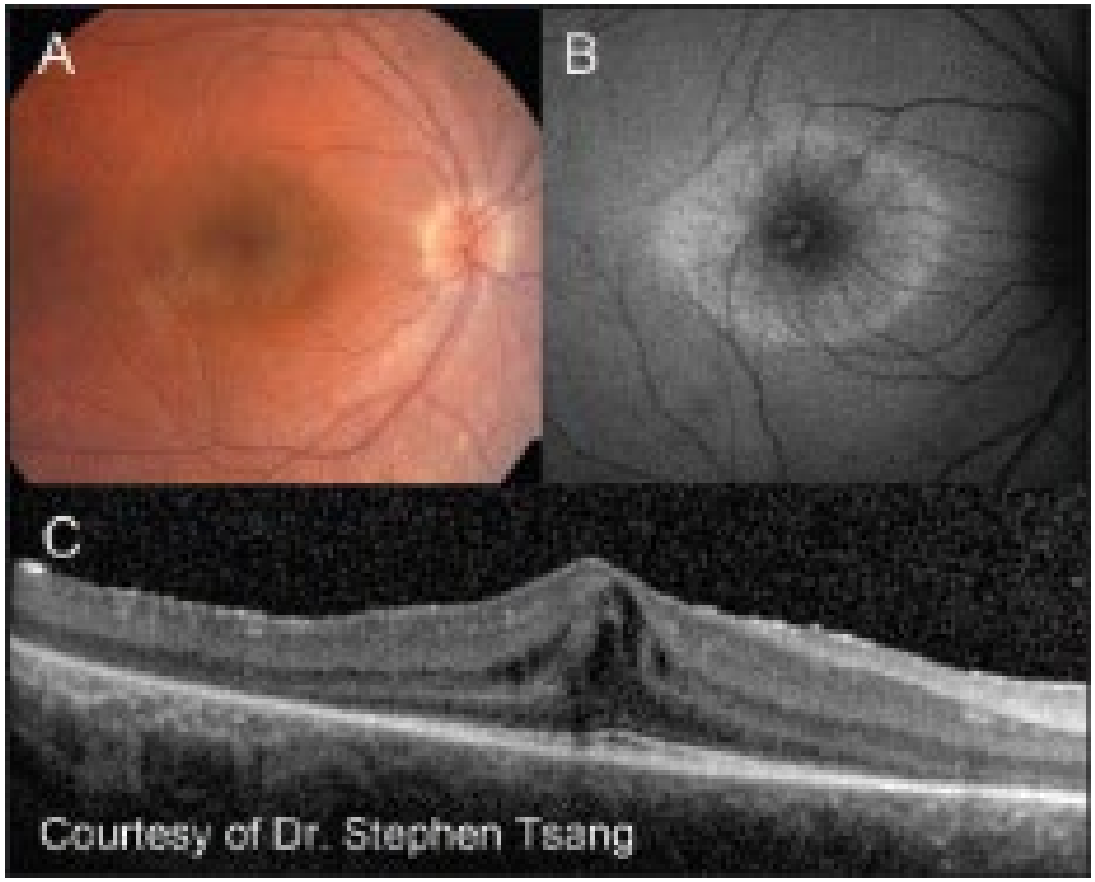


Cystic foveal changes in RP

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--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--Arteriolar narrowing

Other common signs:
--ONH: Drusen
**--Fovea: CME/CMD**
--?
--?

*What do these stand for?*
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*CMD:*

Retinitis Pigmentosa

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CME is…
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*What proportion of RP pts will develop CME?*

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Topical can be effective, but most clinicians probably go PO

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Retinitis Pigmento

*What do these stand for?*
***CME:* Cystoid macular edema**
*CMD:* Cystoid macular degeneration

*What is the difference between the two?*
CME is…'wet' (ie, leaks on FA)
CMD is…'dry' (no leakage on FA)

*What proportion of RP pts will develop CME?*
10-20% or so

*What is the first-line tx for CME in RP?*
Acetazolamide

*What is second-line?*

# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
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Other common signs:
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*What route(s)?*

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*What route(s)?*
Periocular or intravitreal

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*What about CMD? How is it treated?*

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*What about* CMD*? How is it treated?*
It's not treatable

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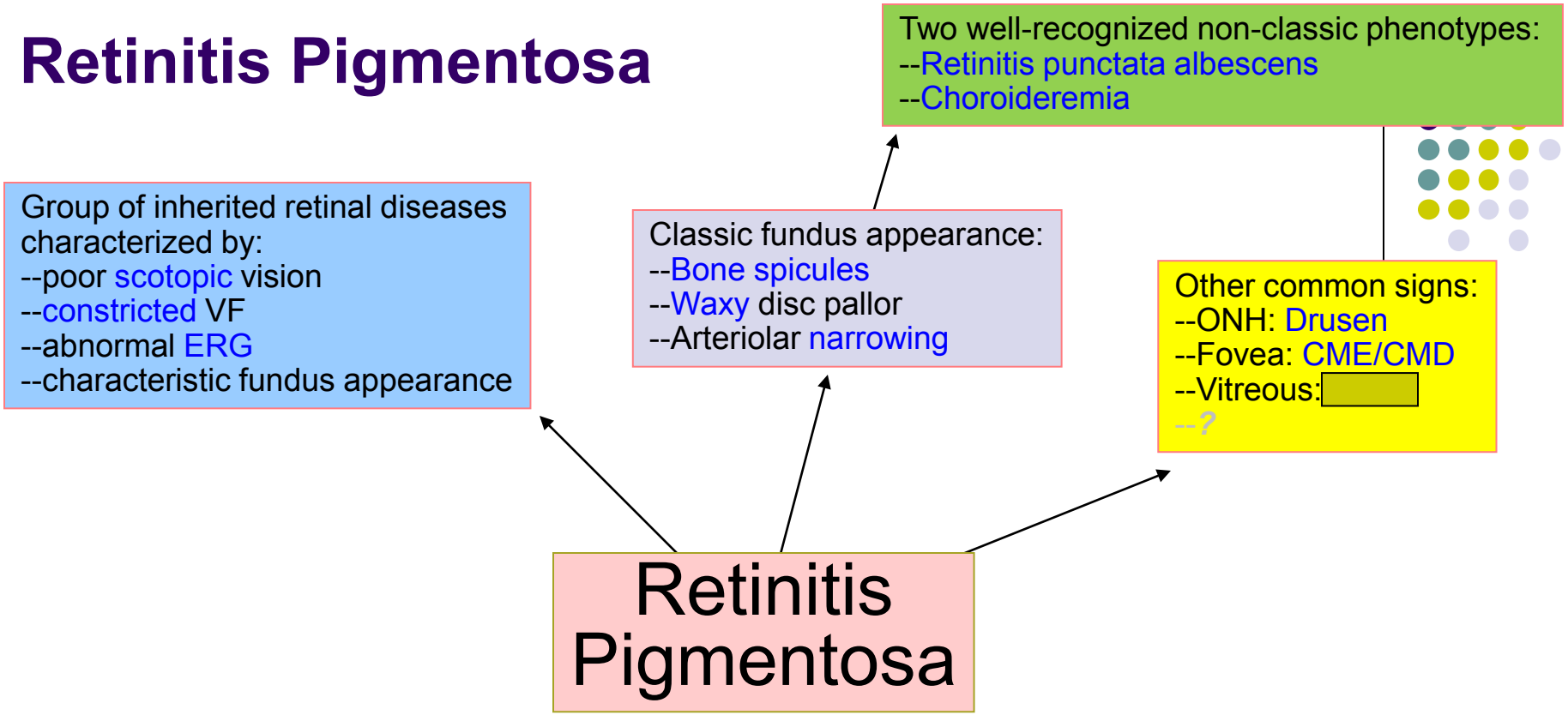

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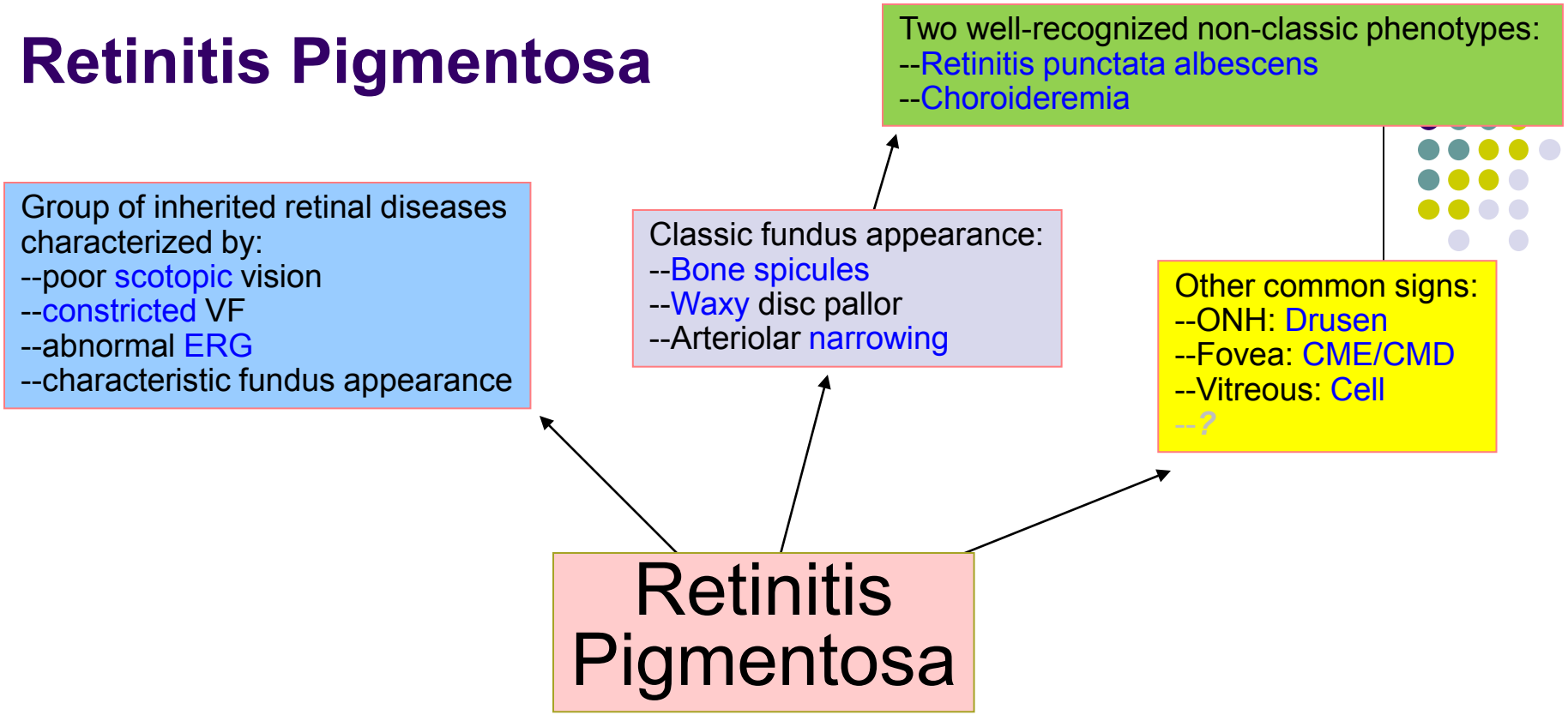

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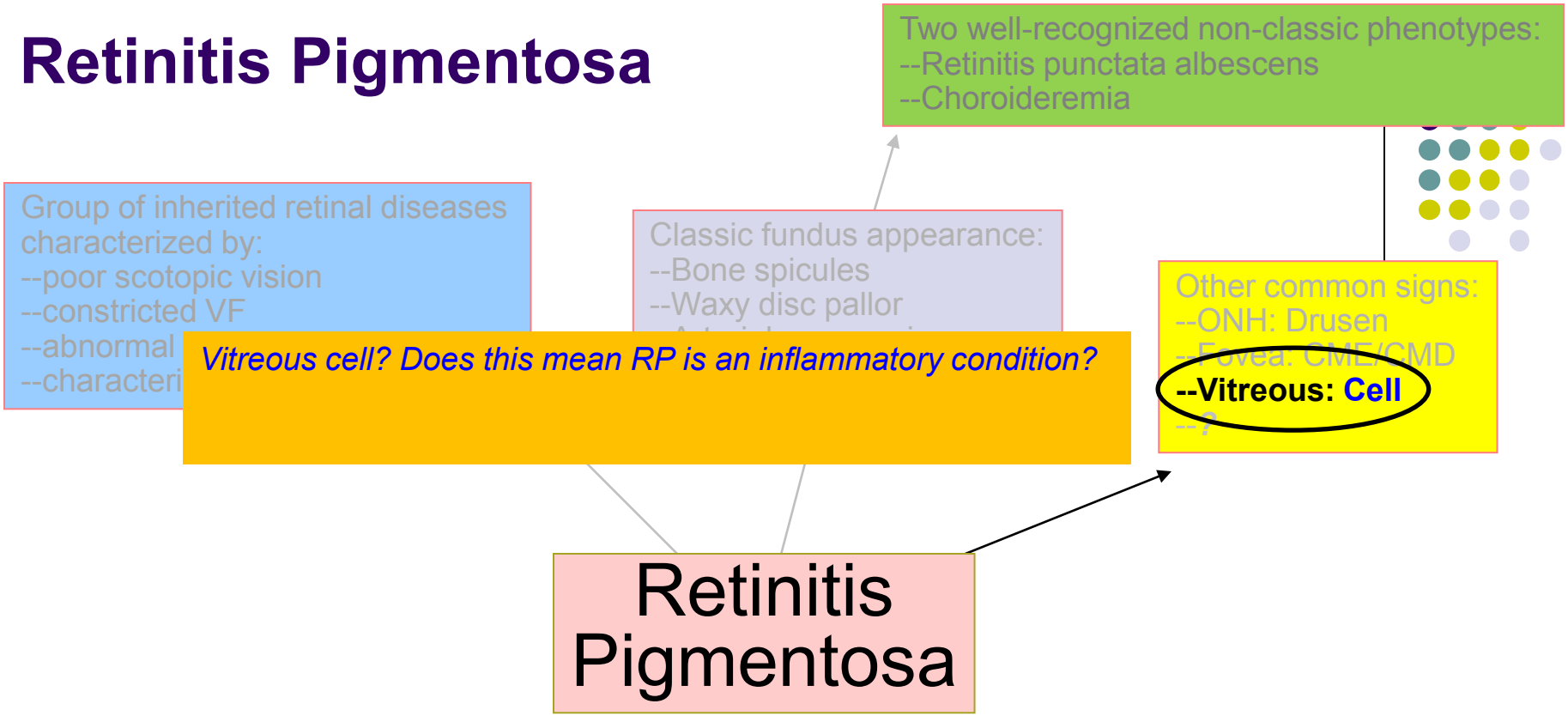

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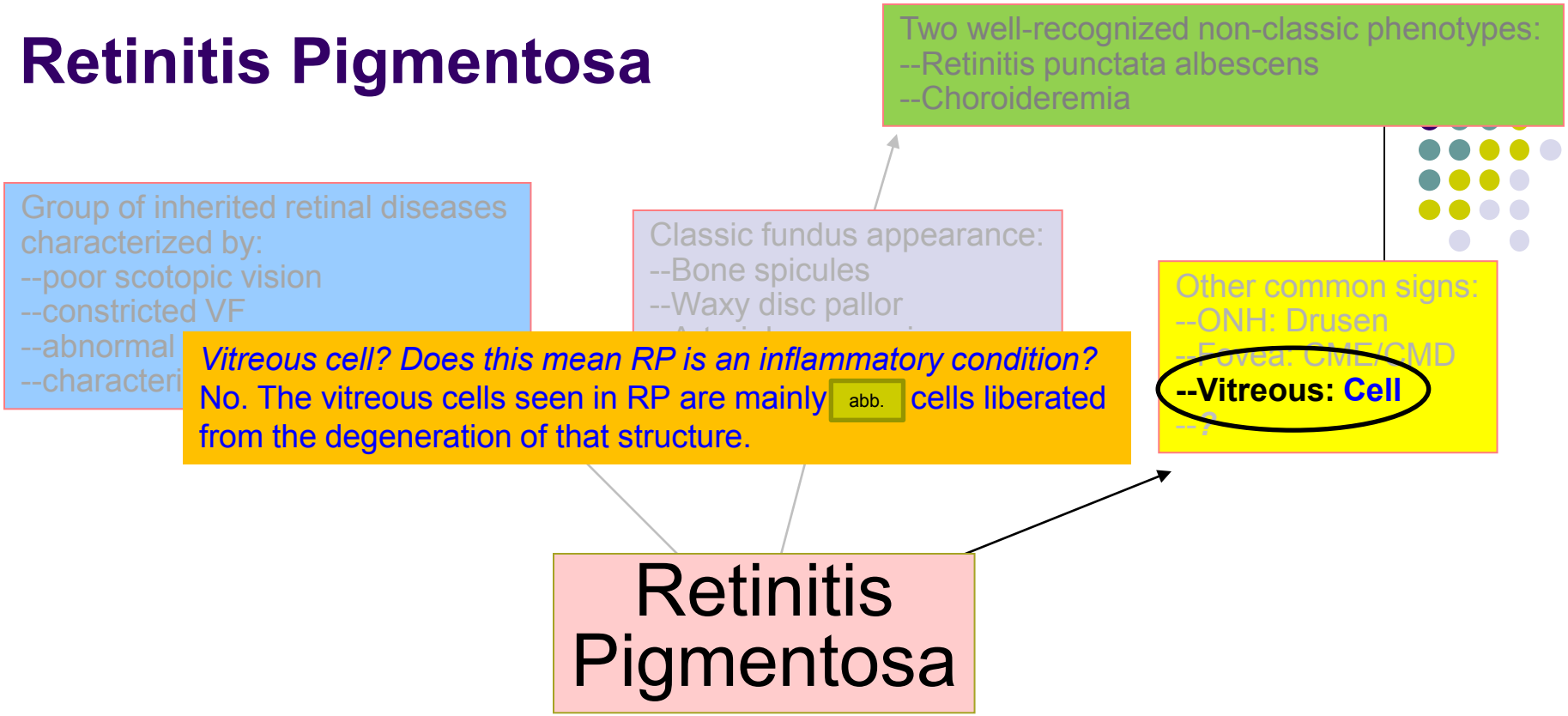

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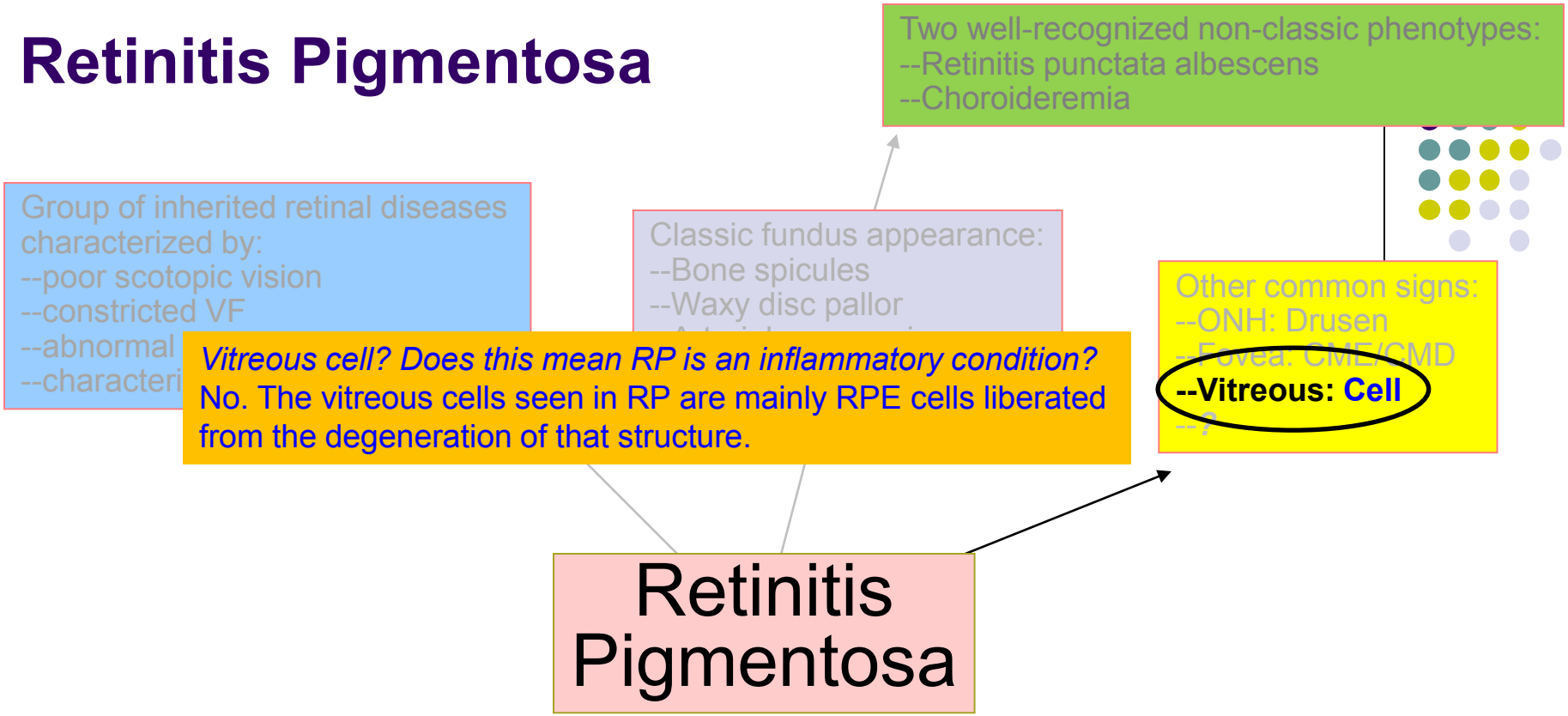

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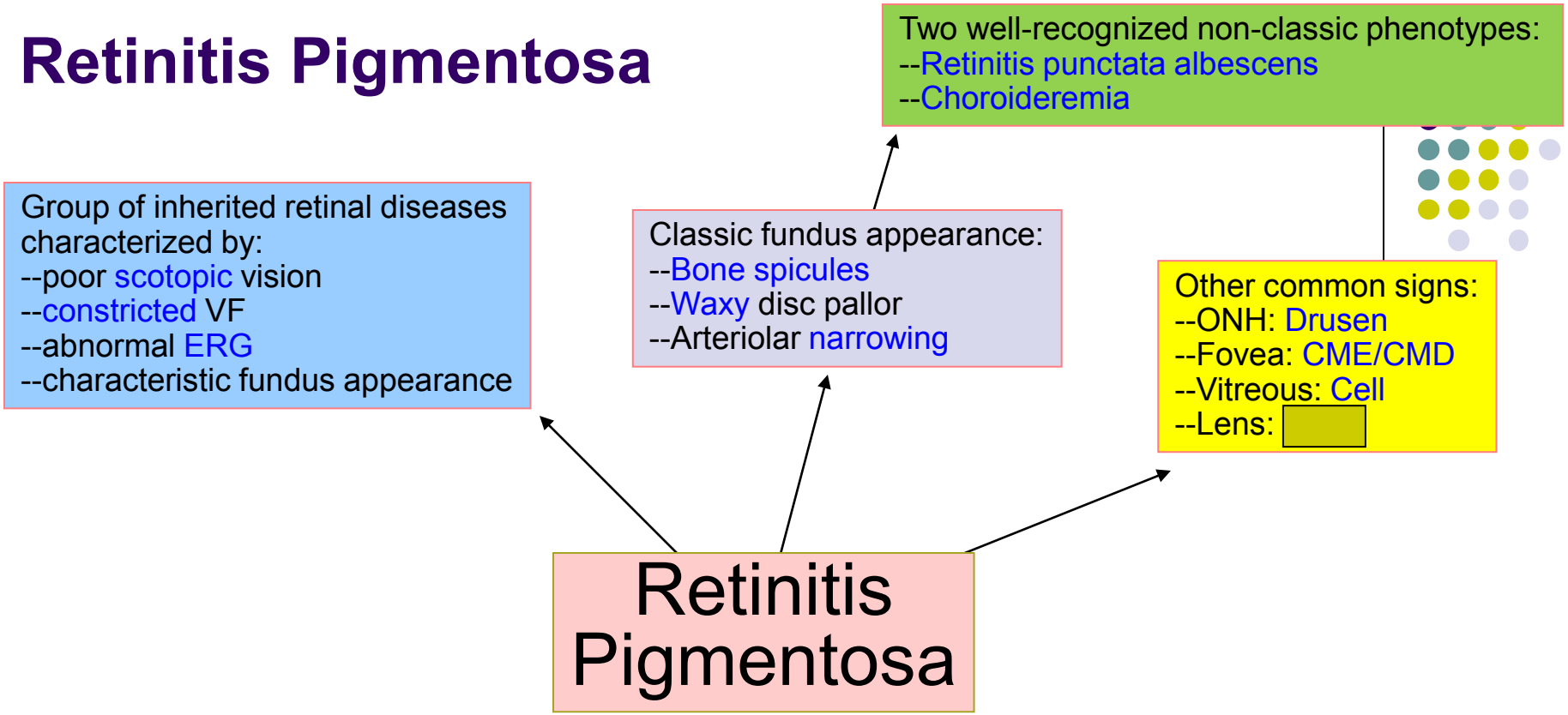

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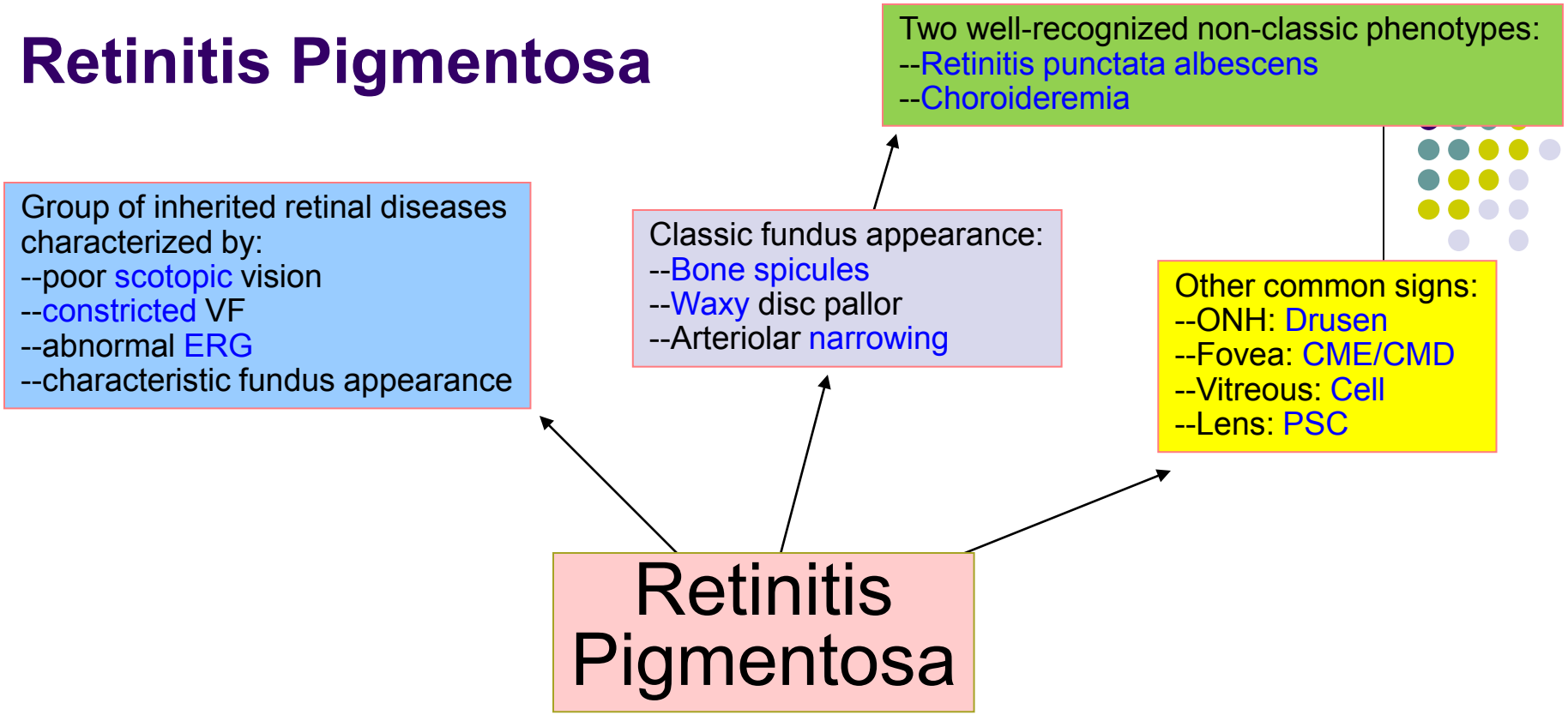

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*What two post-op complications do RP pts get at a higher rate than non-RP pts?*
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--PCO formation
--CME

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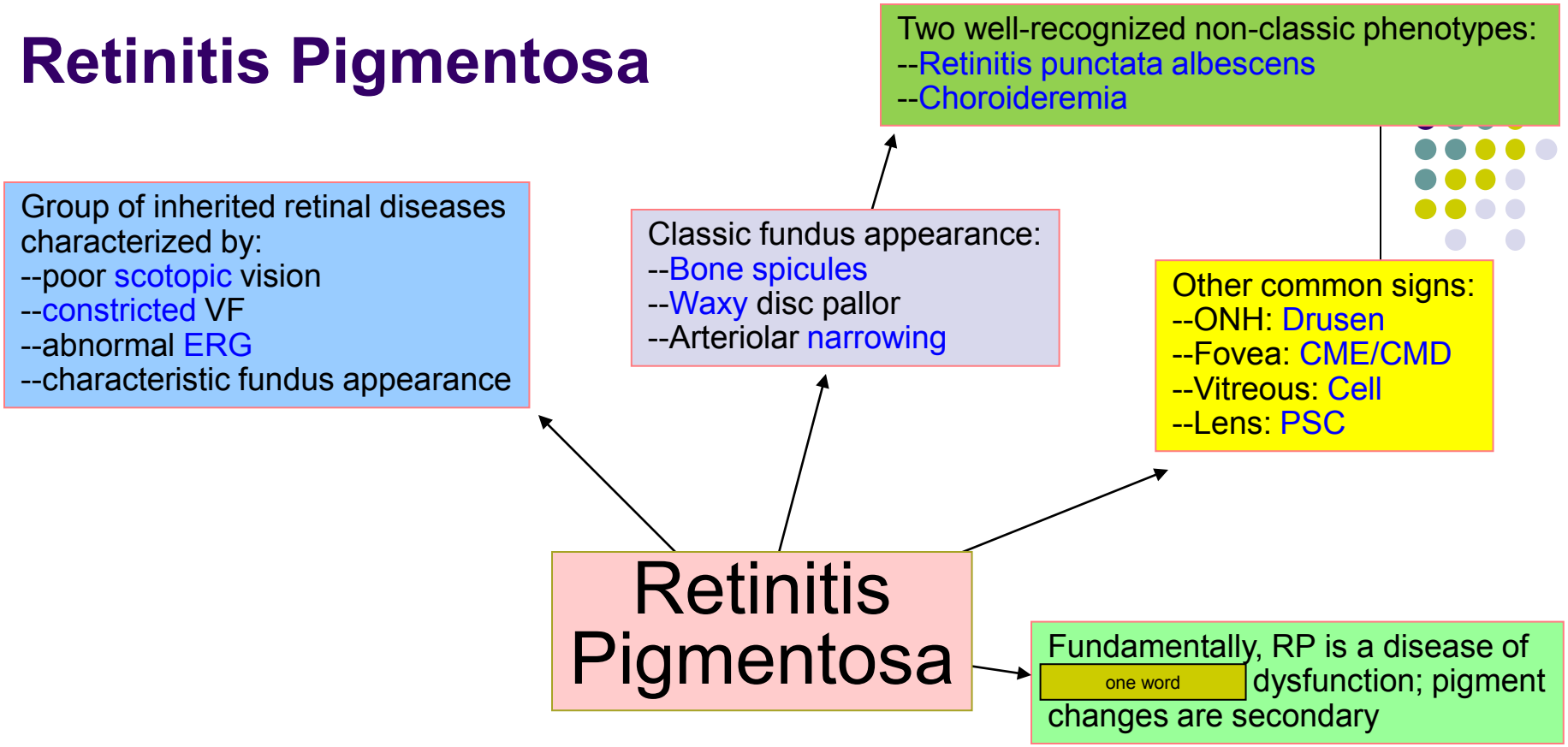

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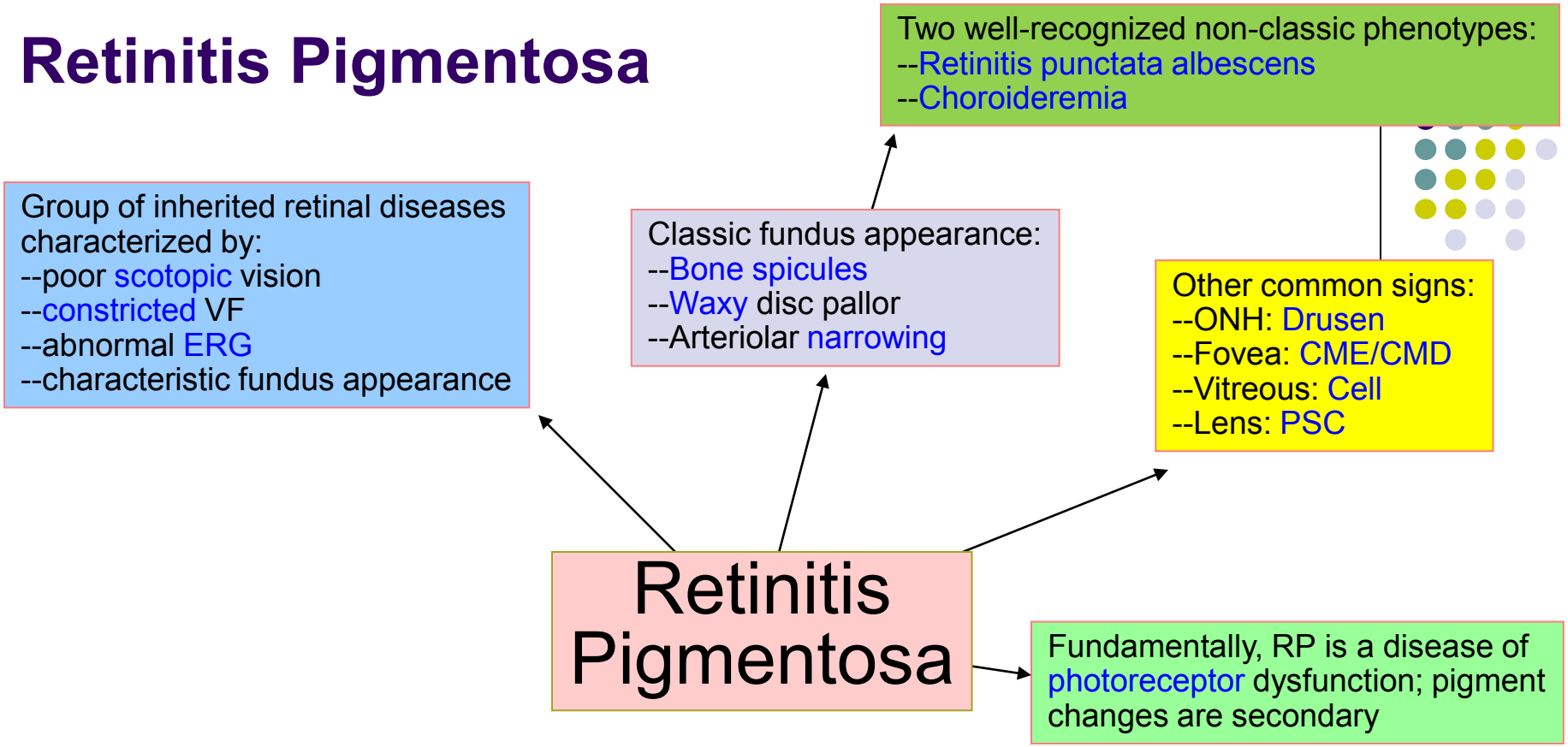

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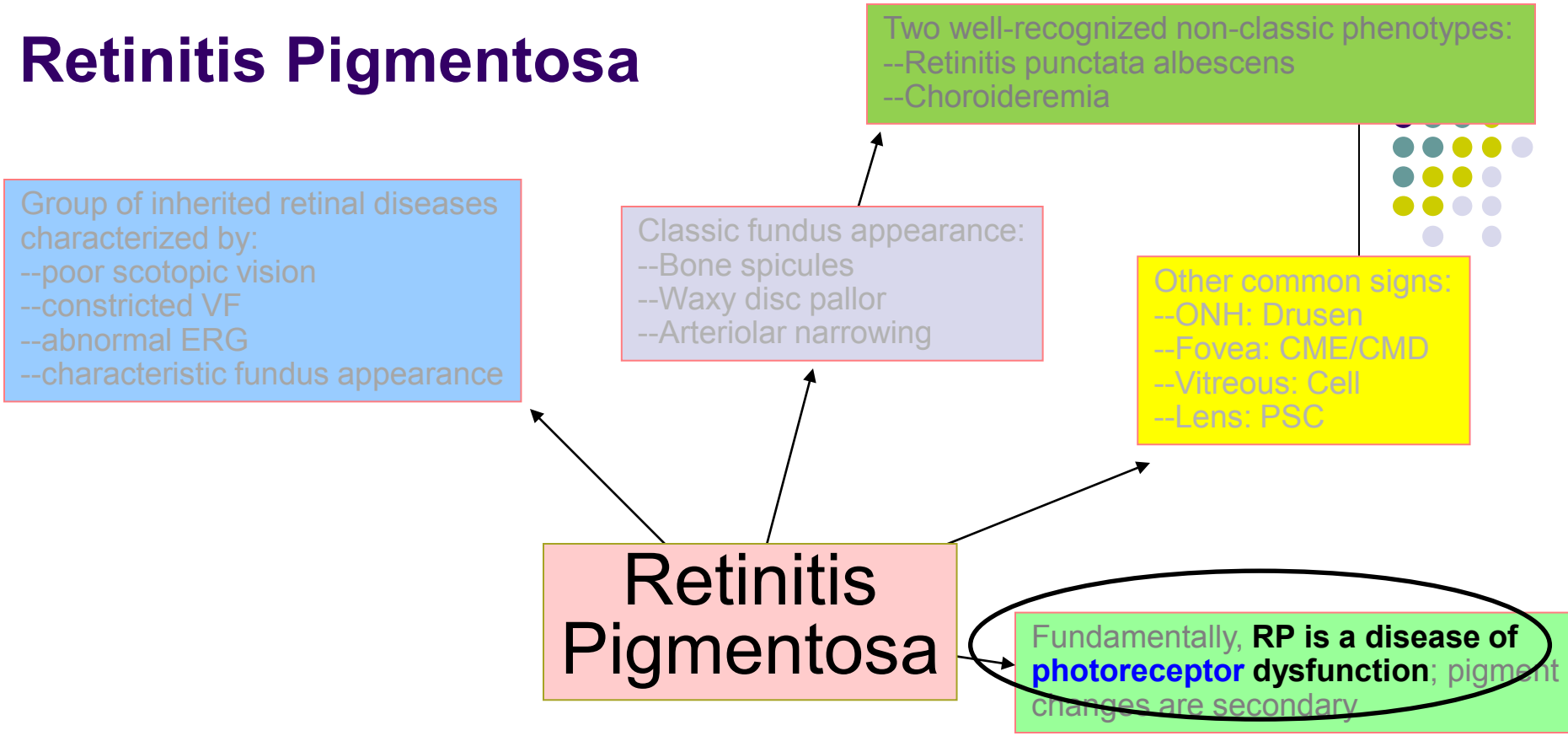


*What is the difference between a rod-cone dystrophy and a cone-rod dystrophy?*

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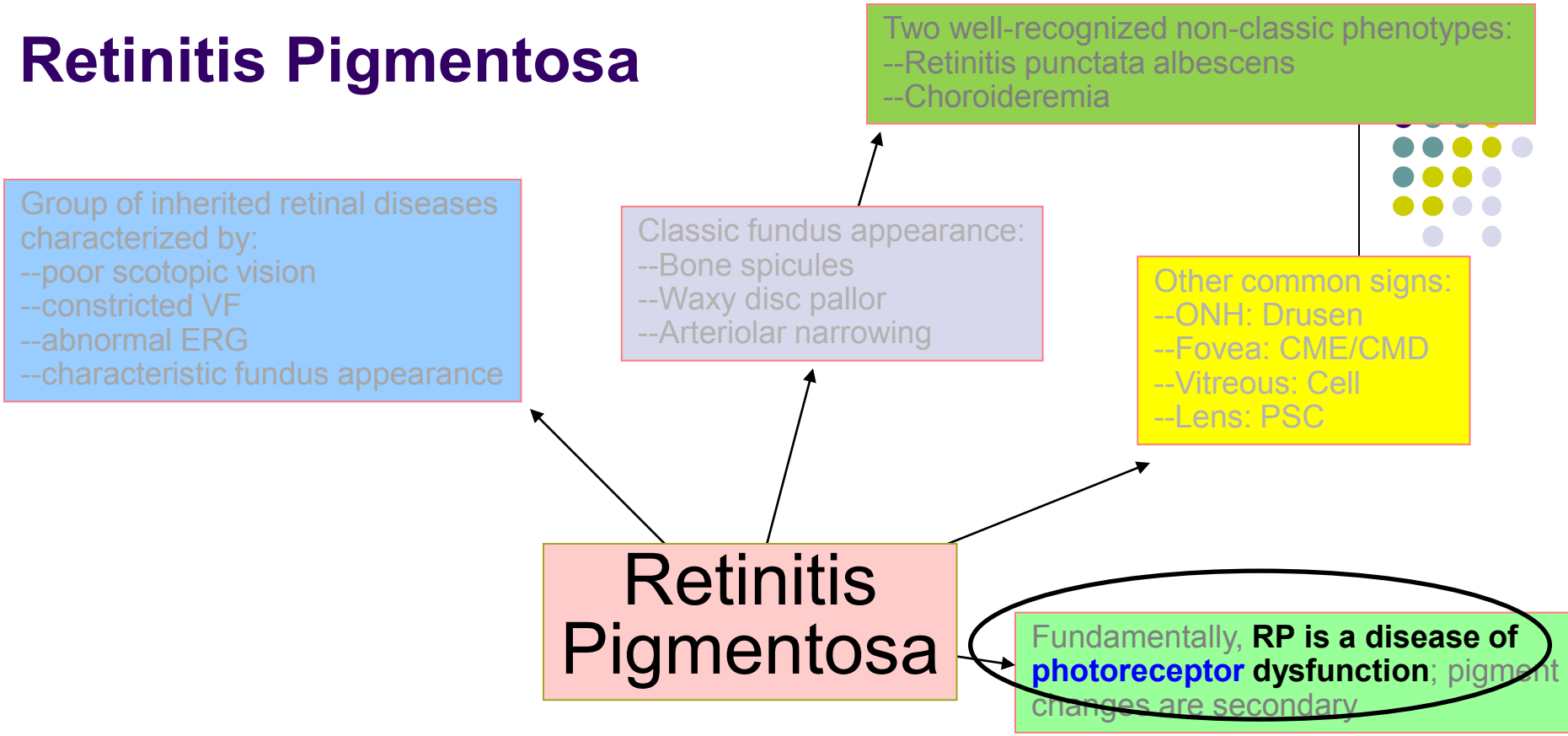


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The difference is the order in which those two populations of photoreceptors are affected by the dystrophy

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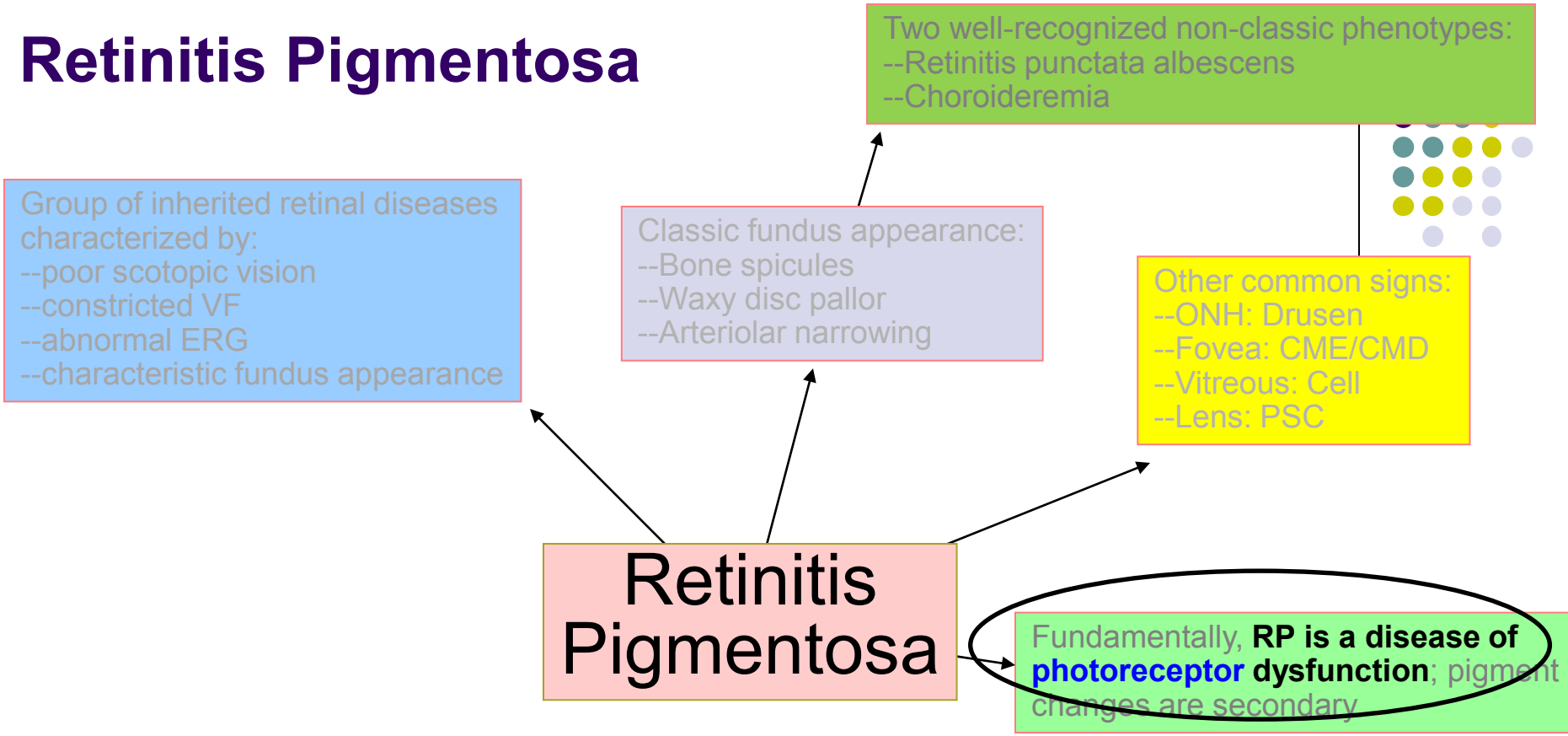


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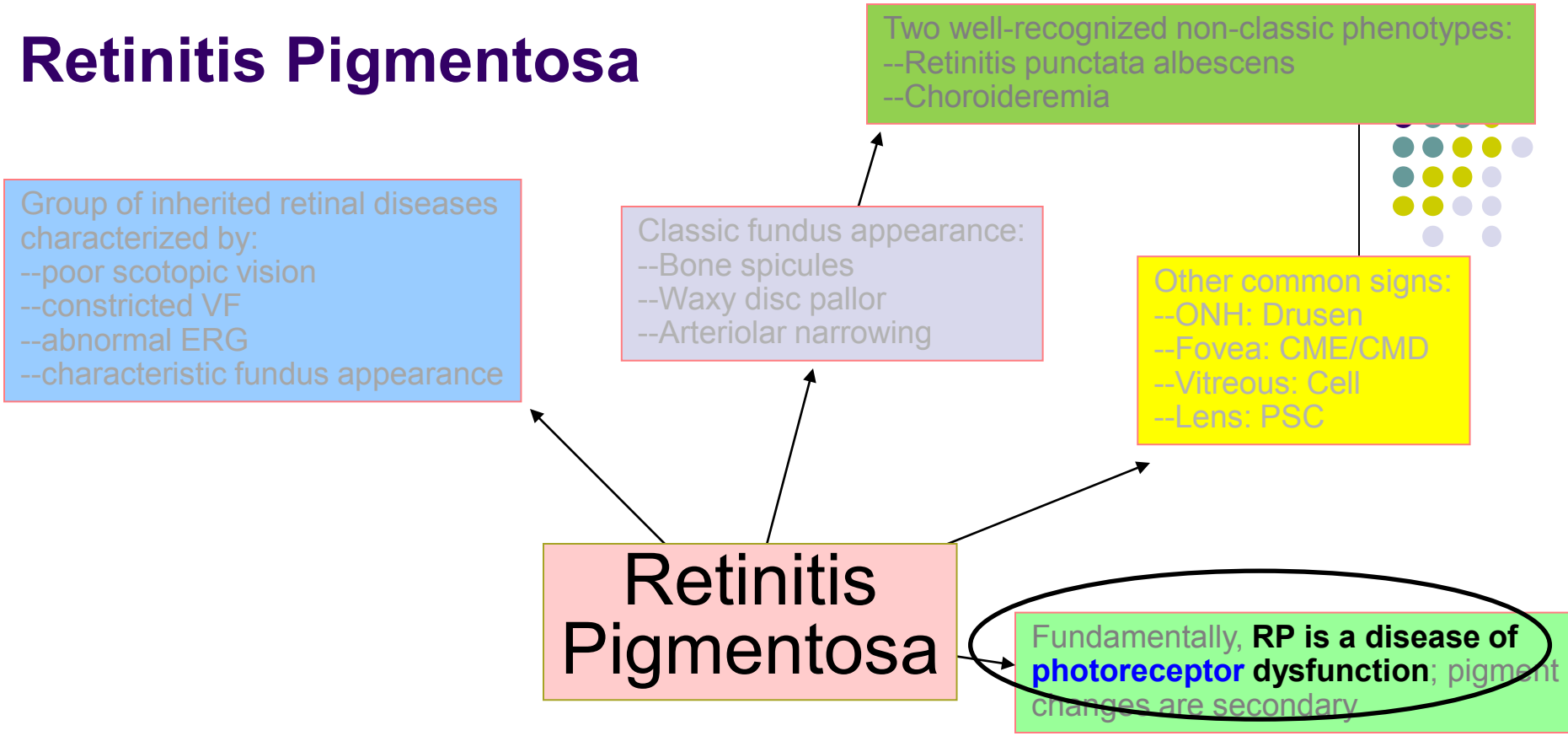


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Retinitis Pigmentosa

Fundamentally, **RP is a disease of photoreceptor dysfunction**; pigment changes are secondary

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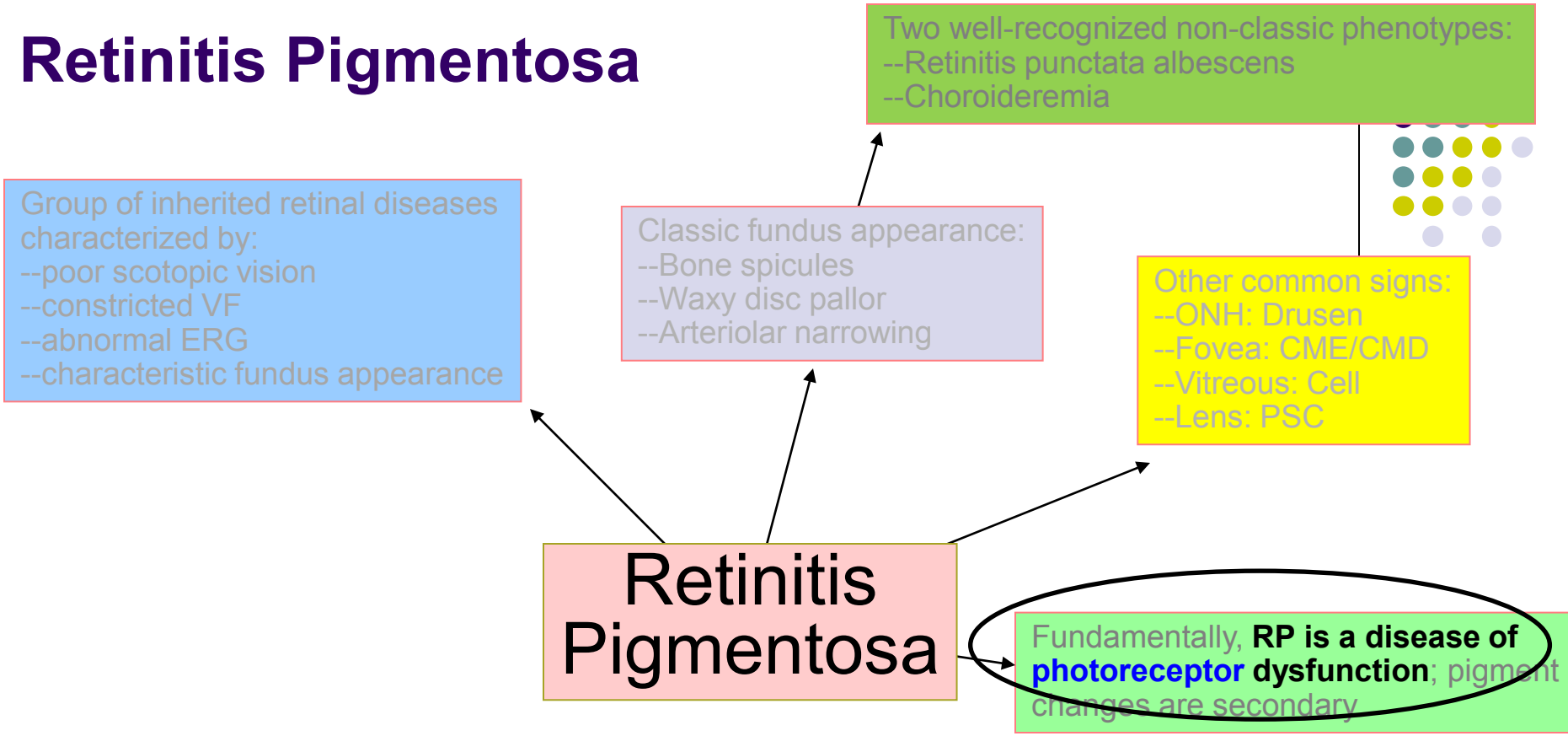


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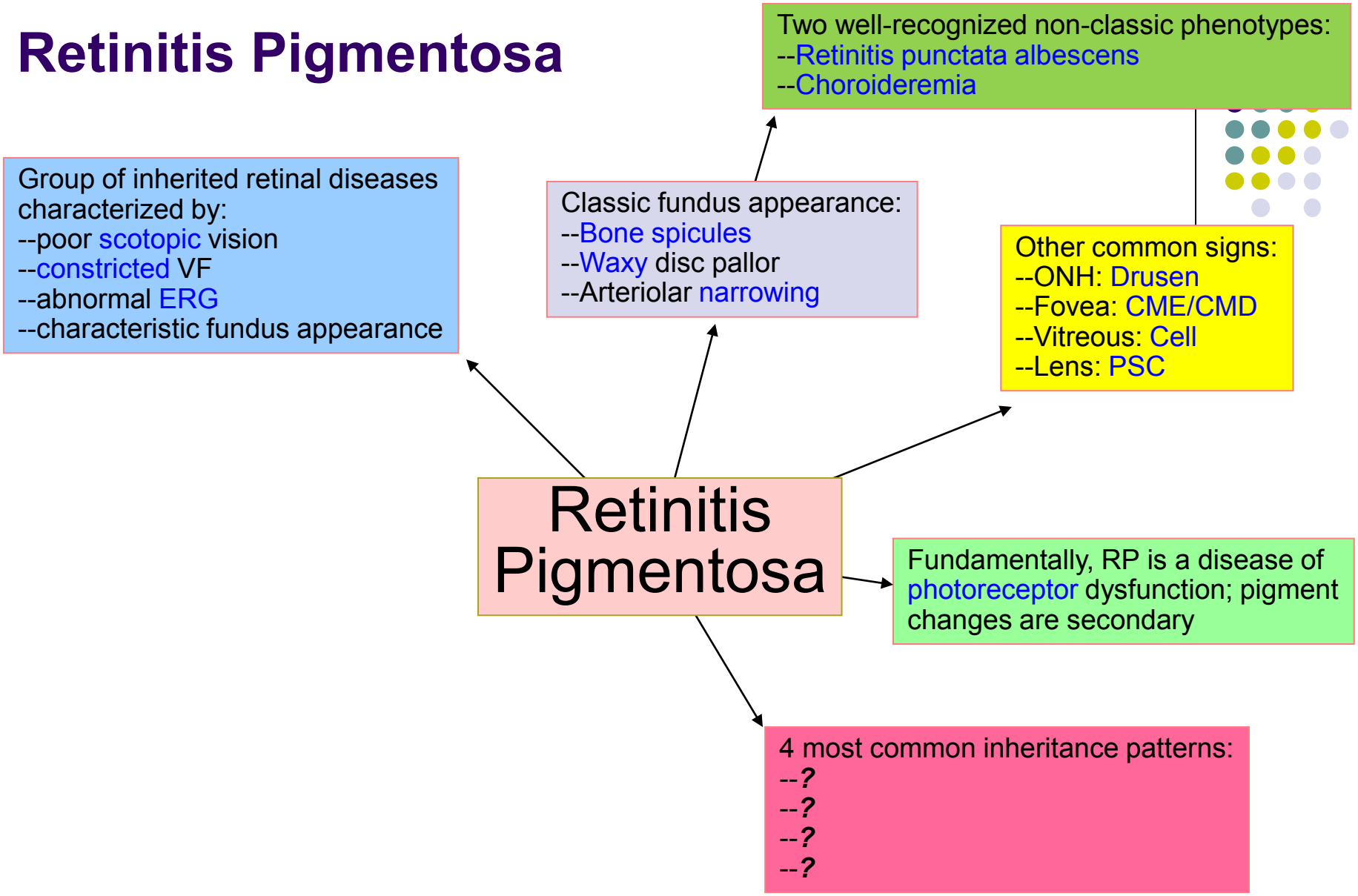

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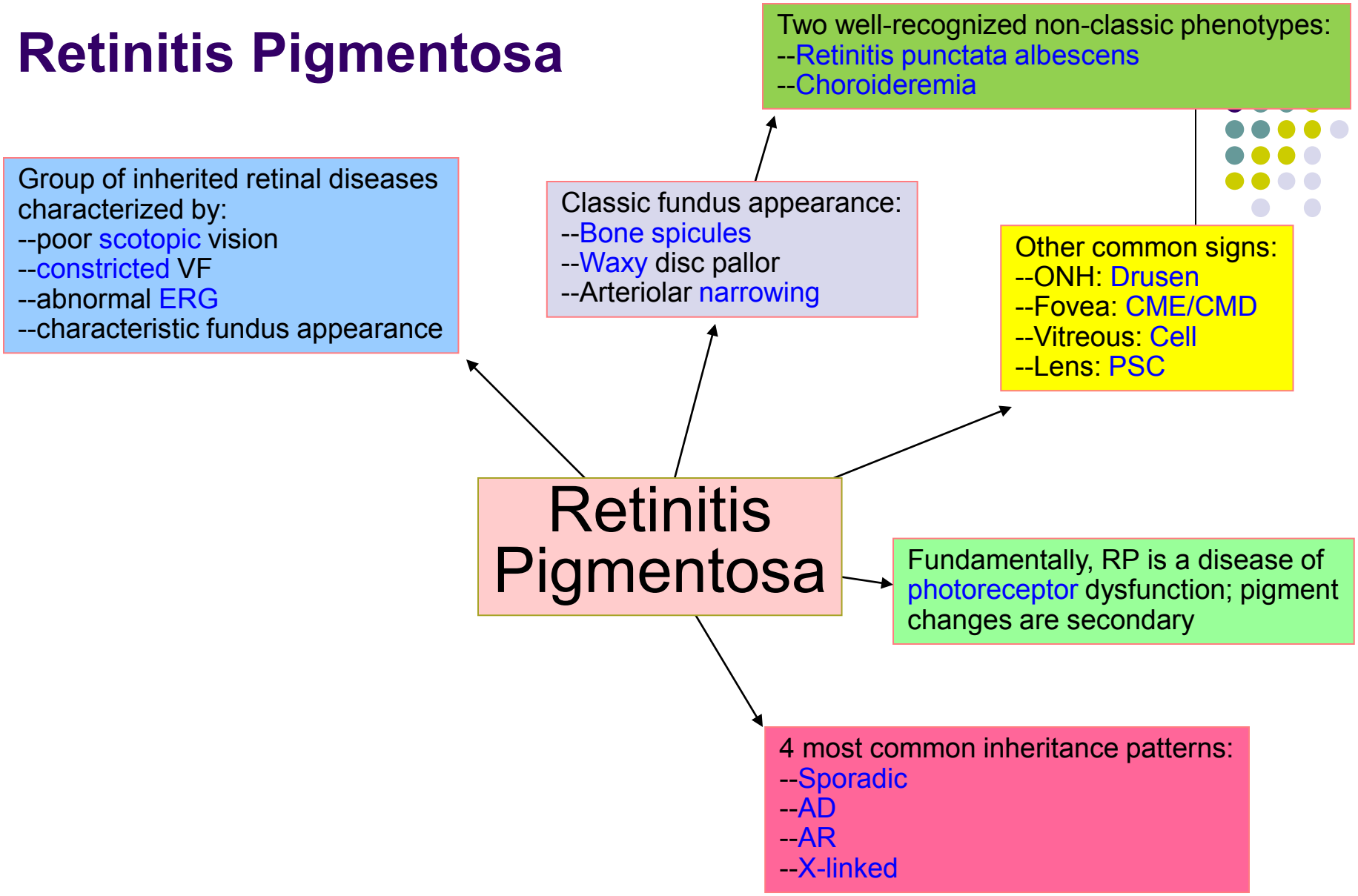

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Retinitis P

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*Of the 4 most common inheritance patterns, which is…*
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**4 most common inheritance patterns:**
**--Sporadic**
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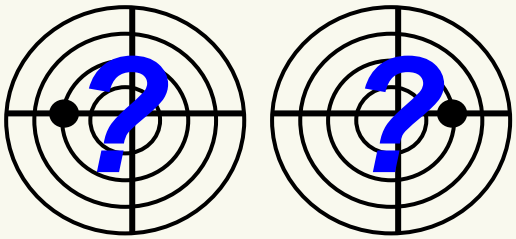


*(Start here)*

*Typical pattern of VF loss:* *[specific pattern of VF loss found in **early** RP]*

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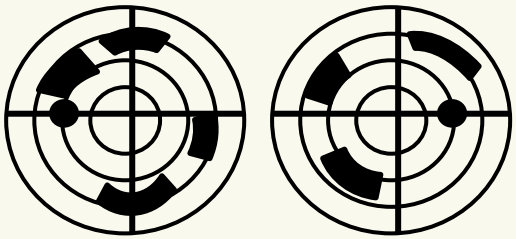

*Typical pattern of VF loss:* Mid-peripheral scotomata

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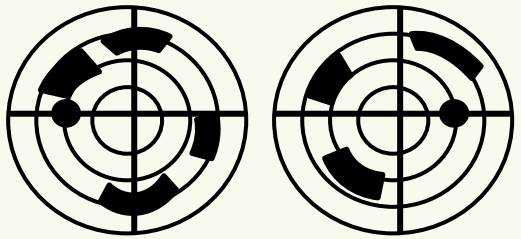

? ?

*Typical pattern of VF loss:* Mid-peripheral scotomata → *[how the VF evolves next]*

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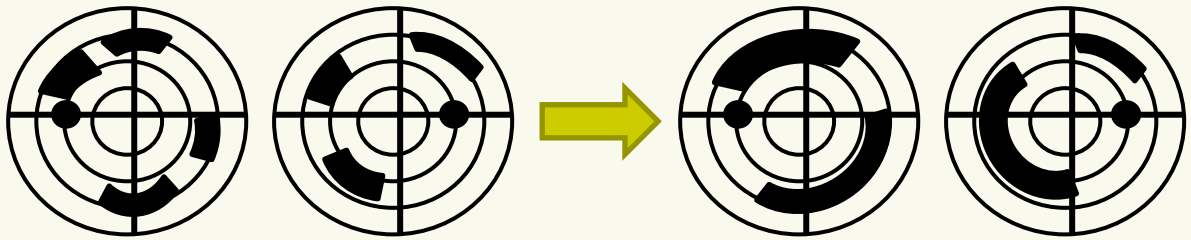

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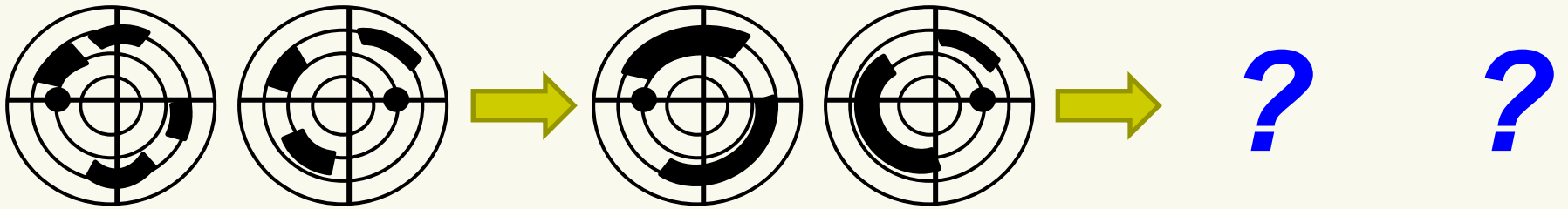


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*[how the VF evolves next]*

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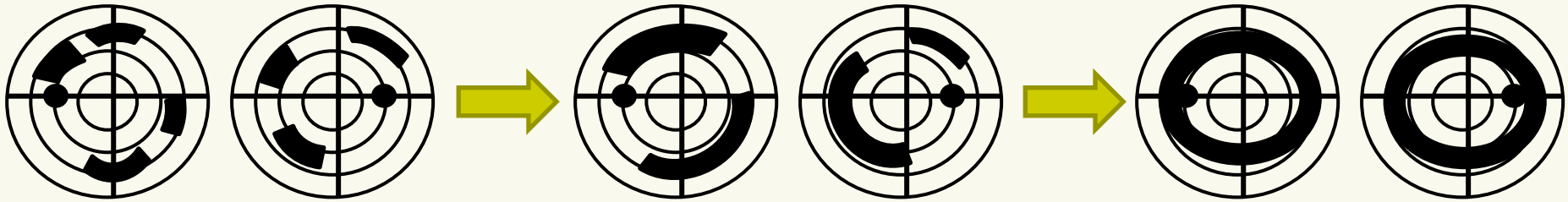

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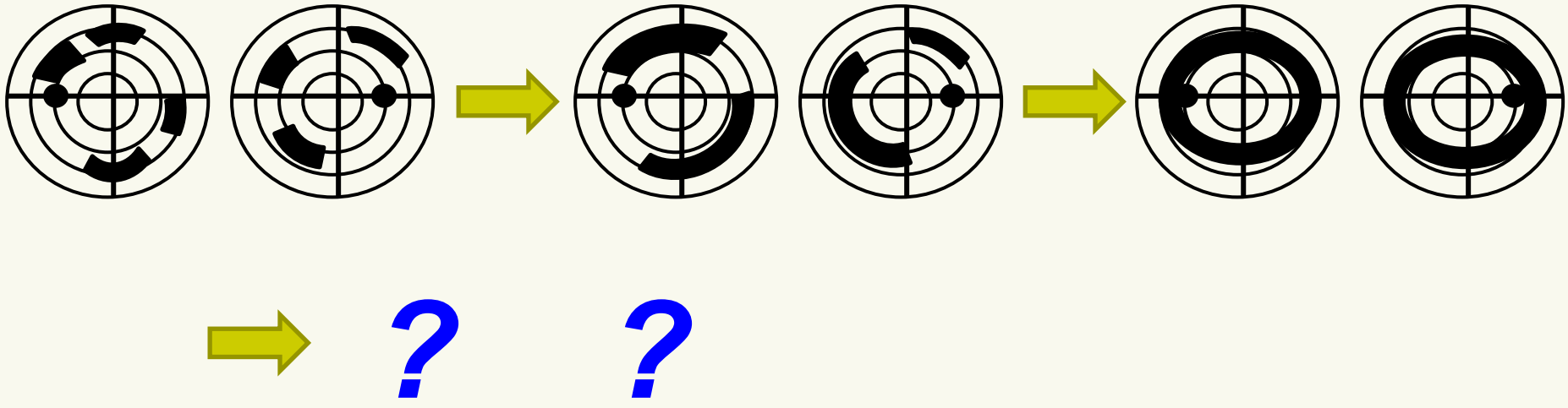


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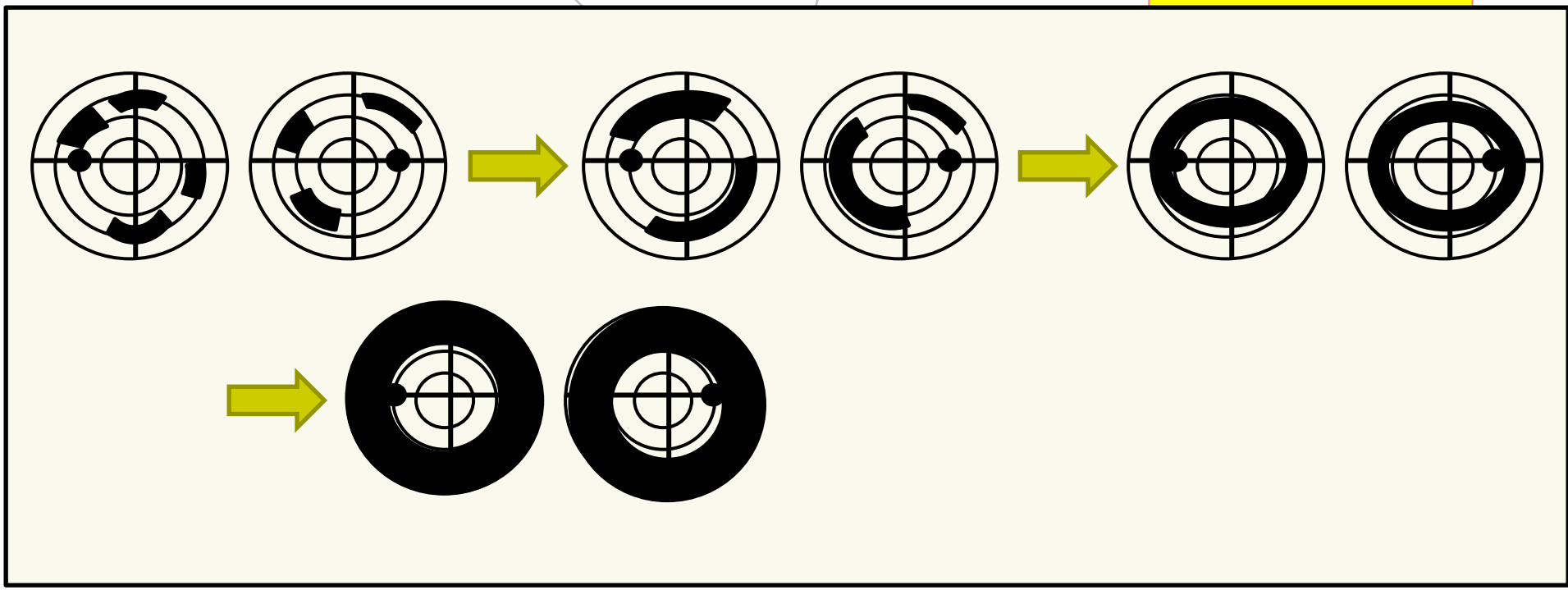

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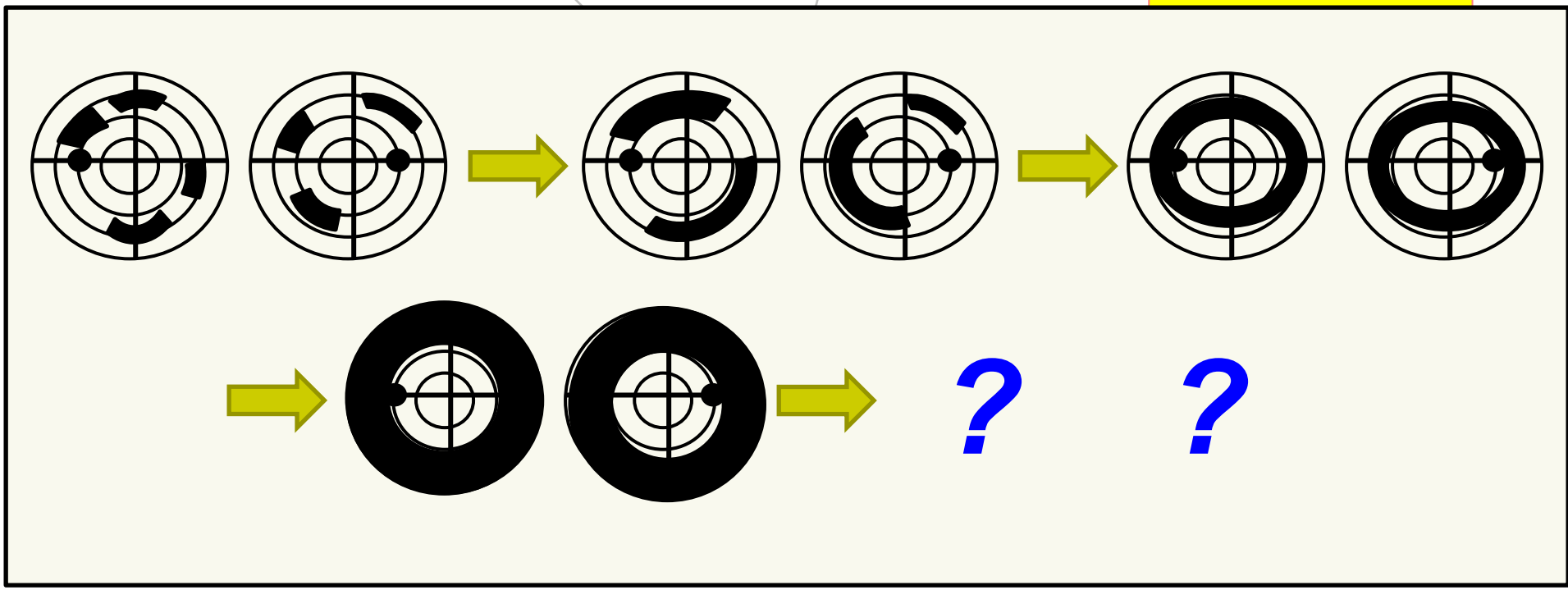


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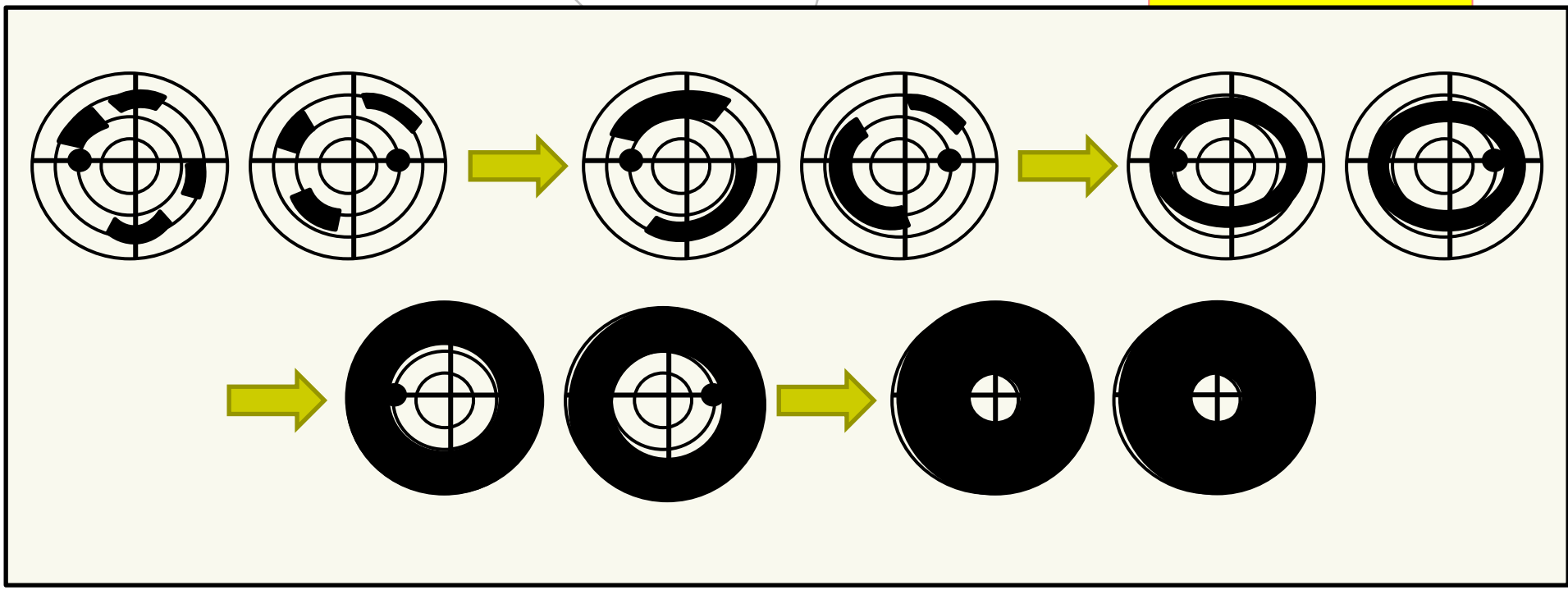

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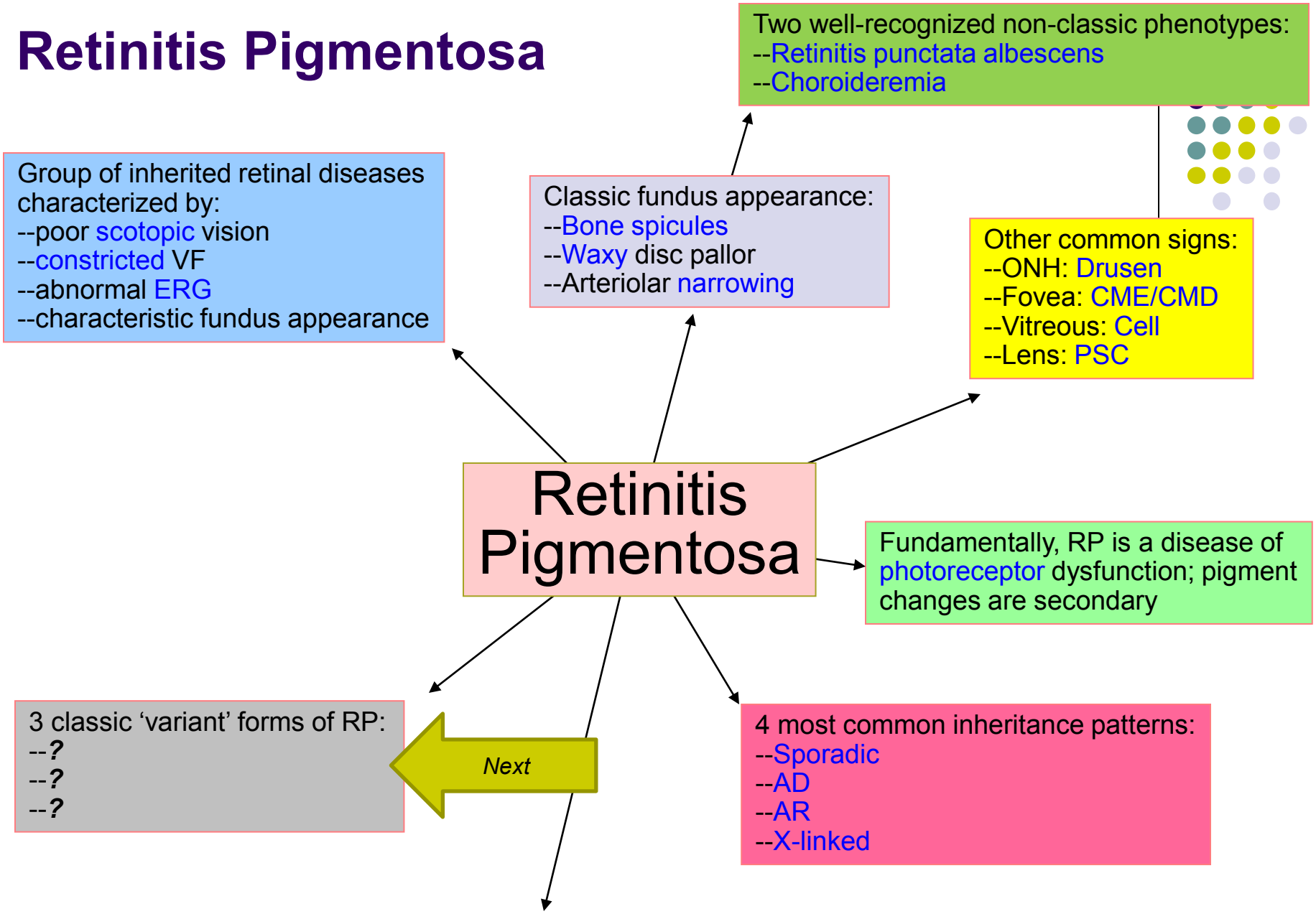

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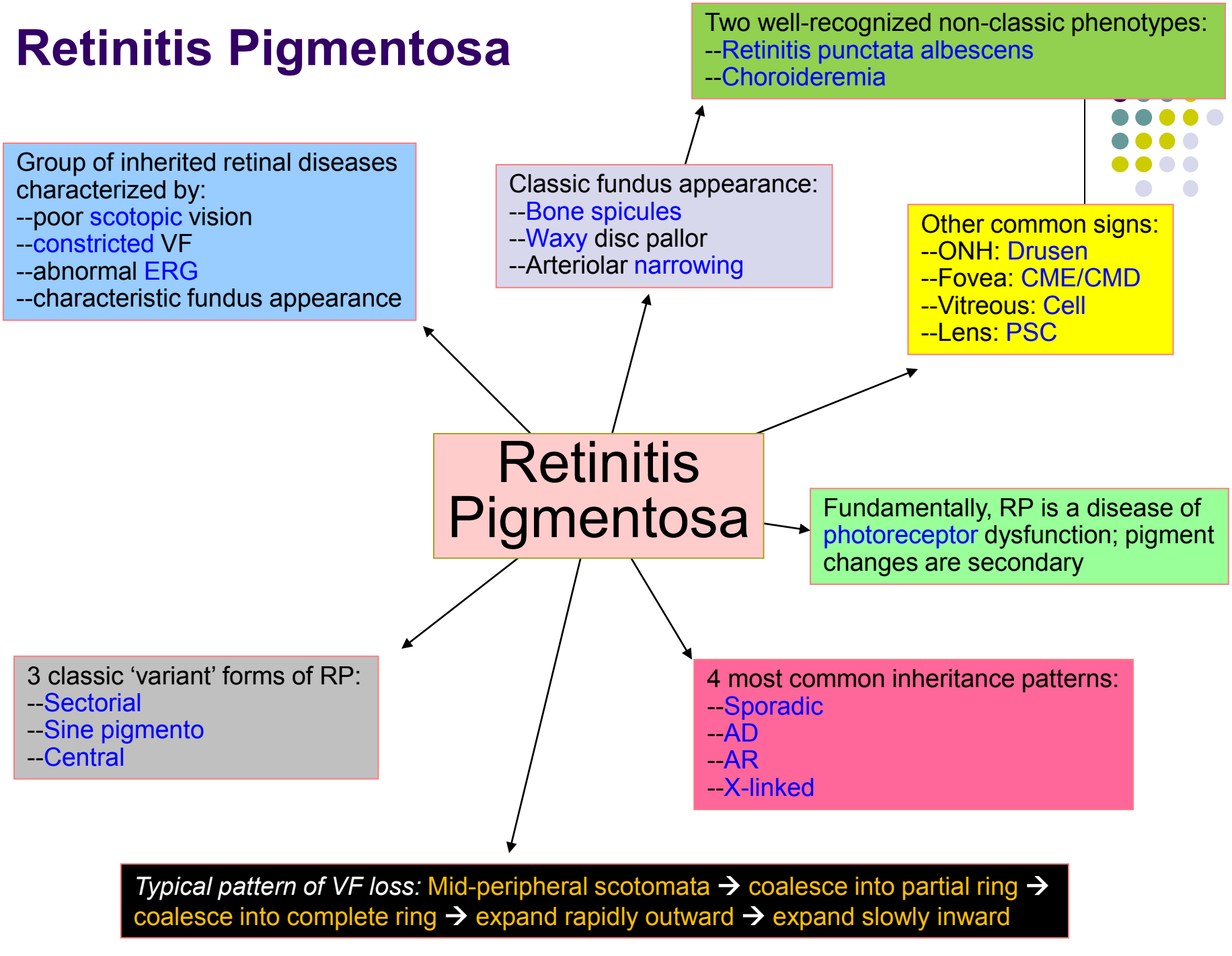

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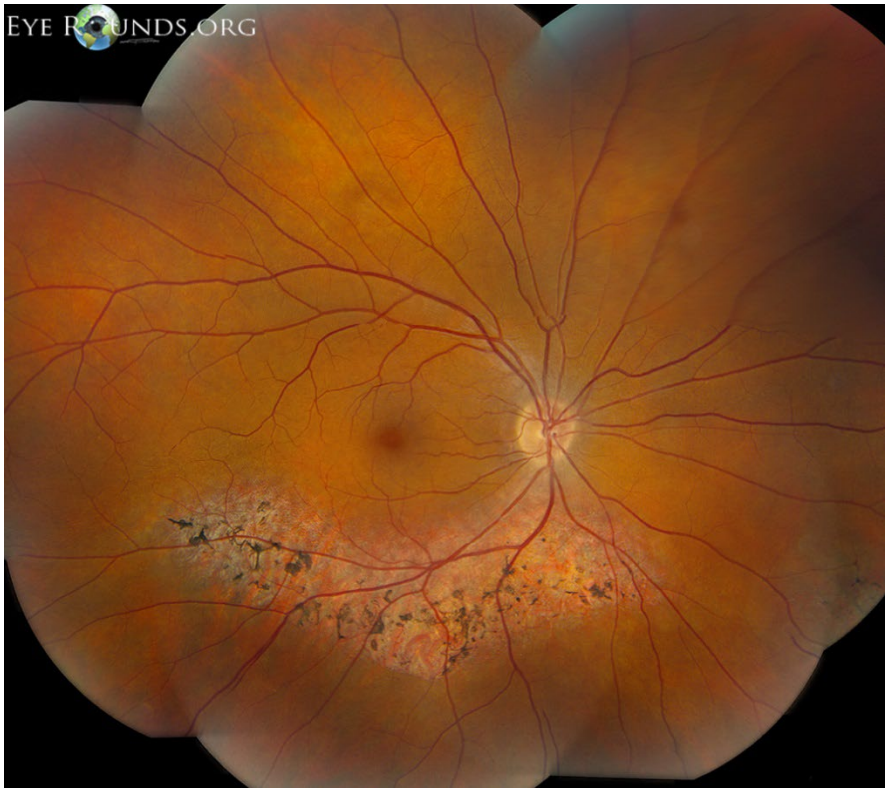


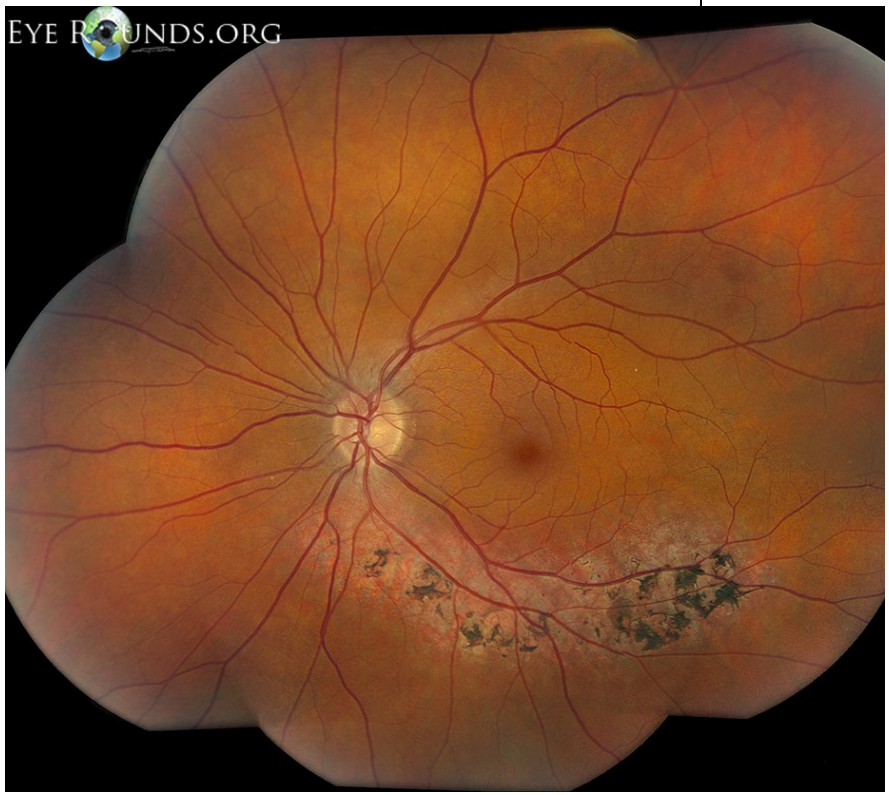


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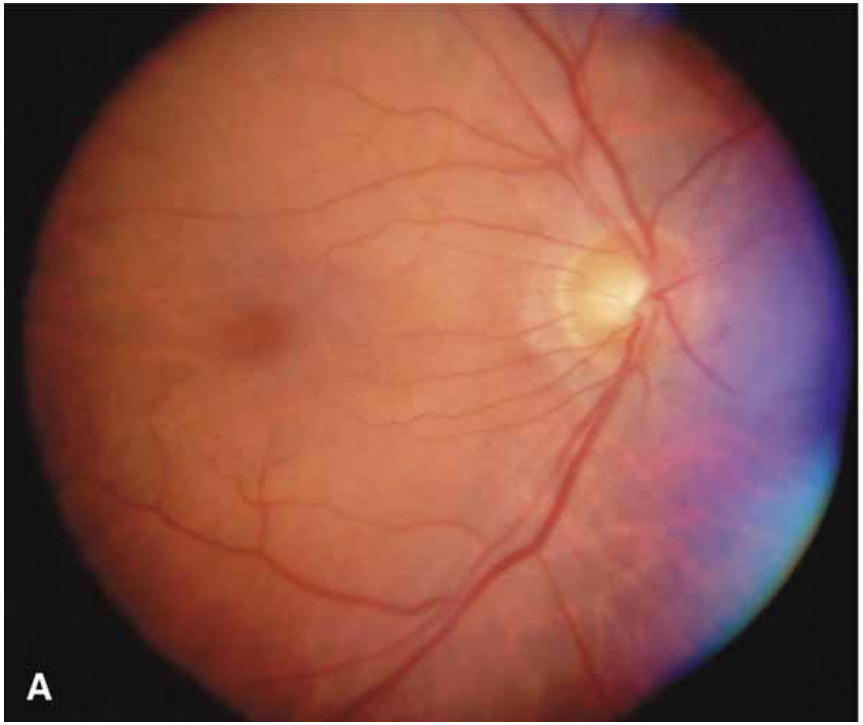


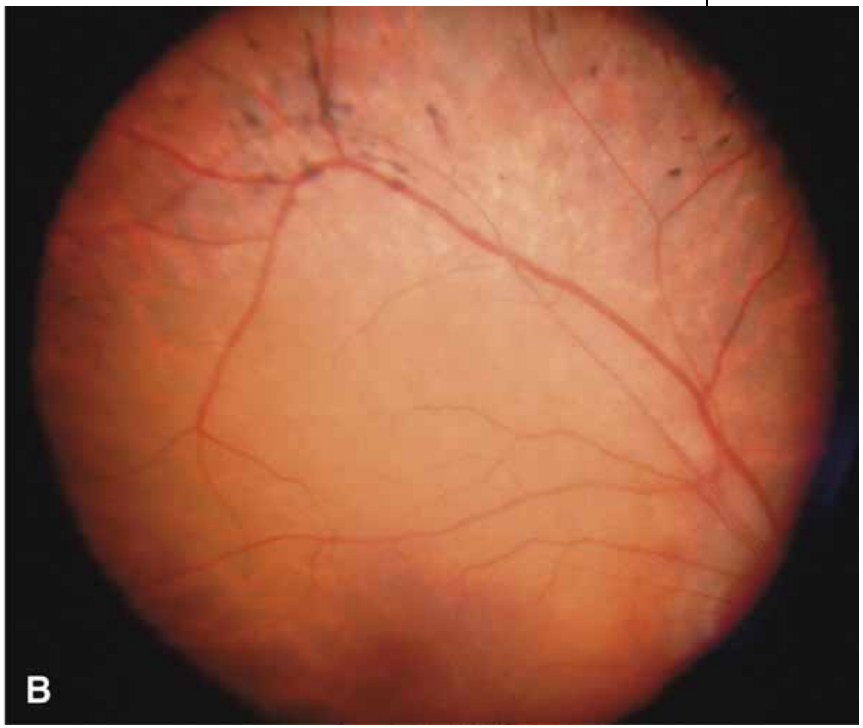


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*What is* central RP*?*
In essence, a reversed form of RP. Whereas in 'normal' RP visual acuity and the central visual field are spared until late in the disease process, in central RP acuity is profoundly affected early on, and VF loss progresses outward from fixation.

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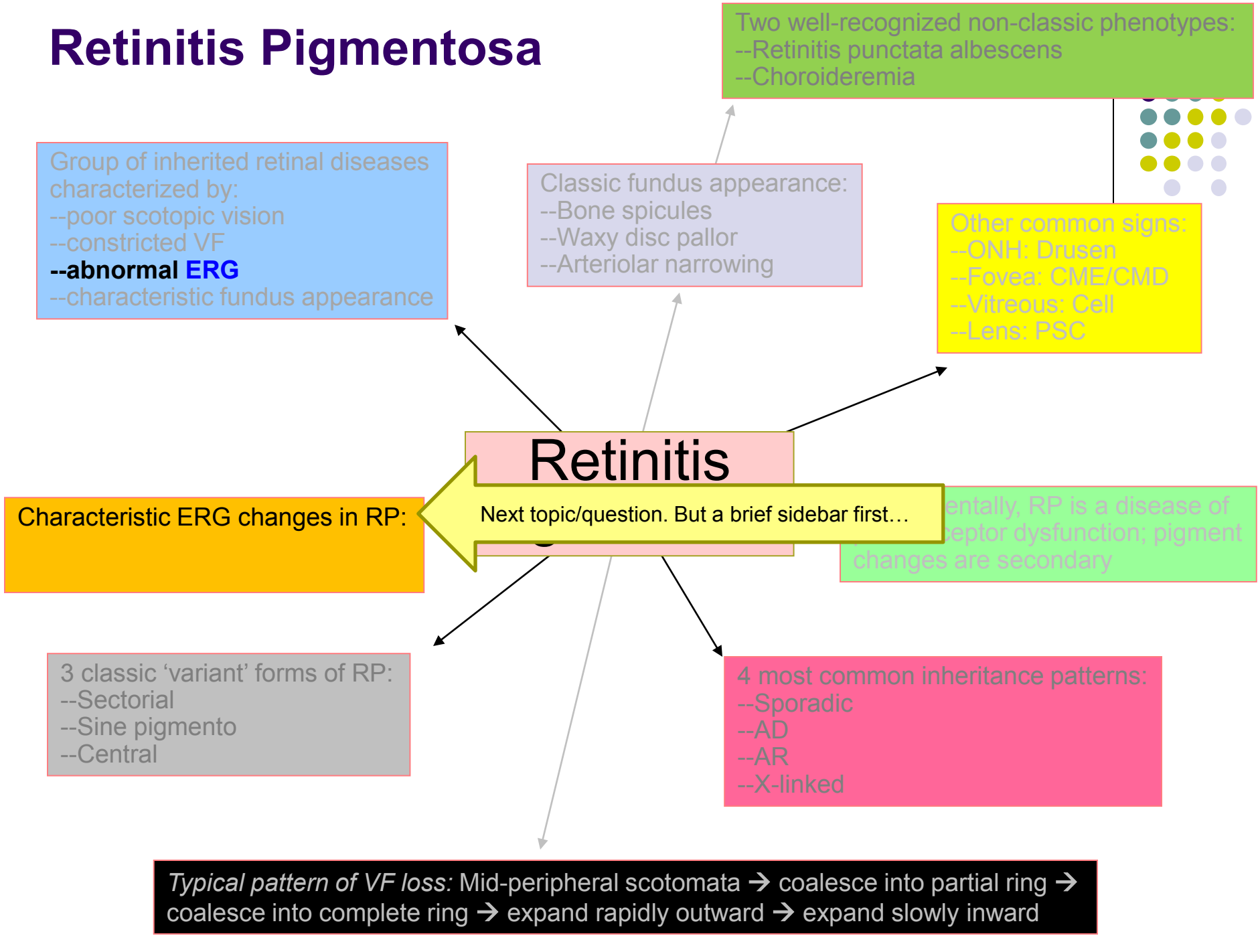

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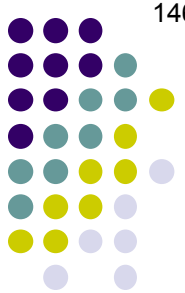

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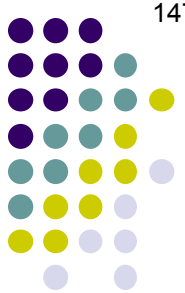

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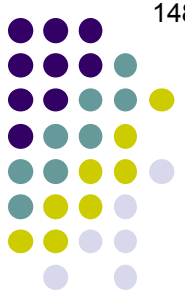

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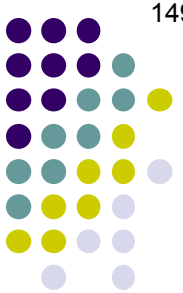

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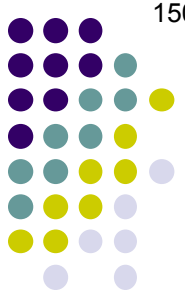

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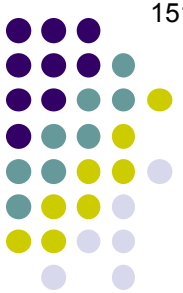

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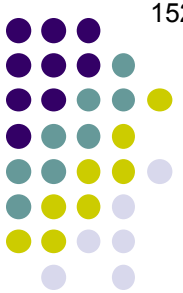
*How is it performed?*

# Retinitis Pigmentosa



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Electroretinogram (or electroretinography)

*In one sentence, what is it?*
An electrophysiologic test that measures how retinal cells respond to a light stimulus

*How is it performed?*
The pt is dilated vs undilated, and usually light- vs dark-adapted

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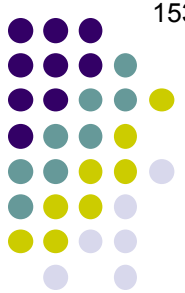

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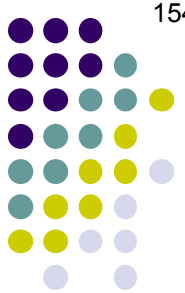

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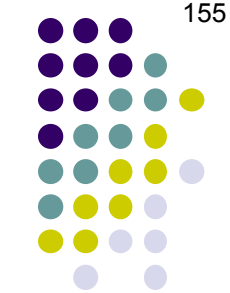

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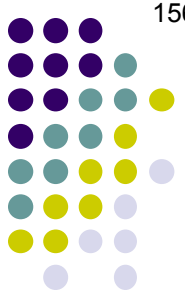

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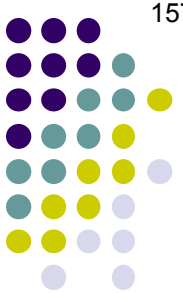

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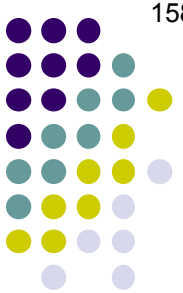

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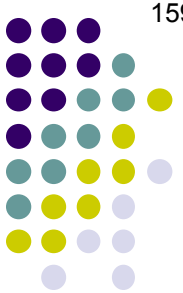

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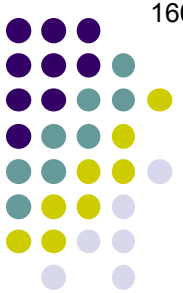

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Full-field

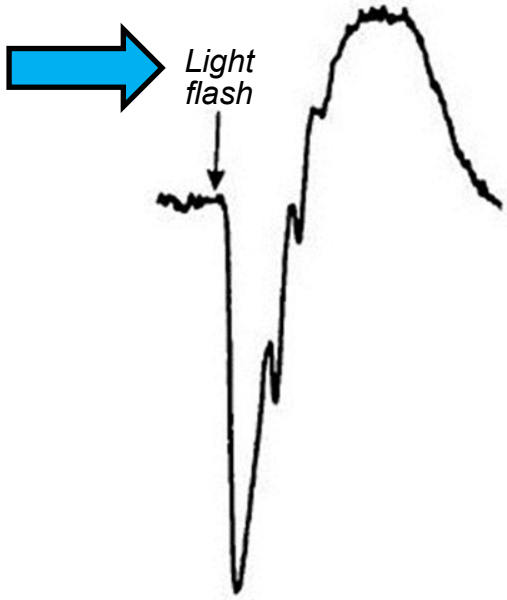
# Retinitis Pigmentosa

***Normal ffERG***

**The normal ffERG**. Note:

# Retinitis Pigmentosa

***Normal ffERG***




**The normal ffERG**. Note:
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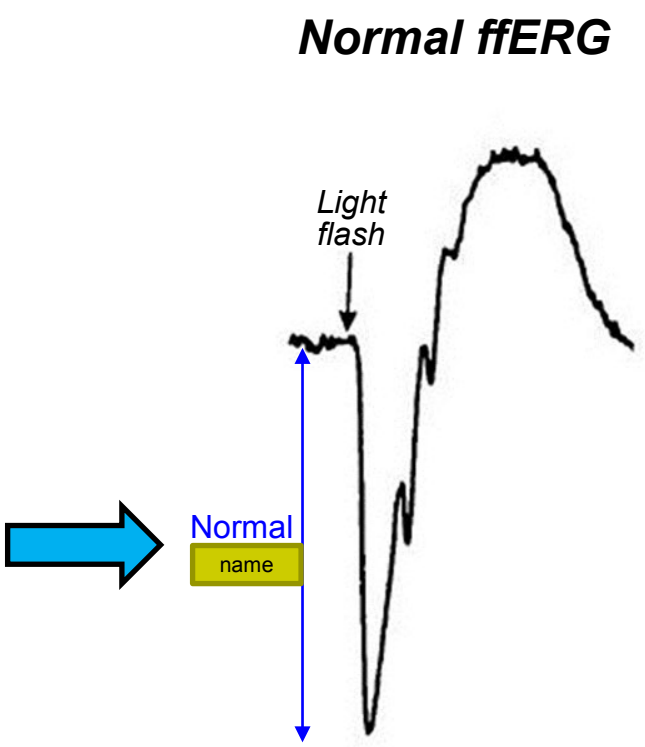
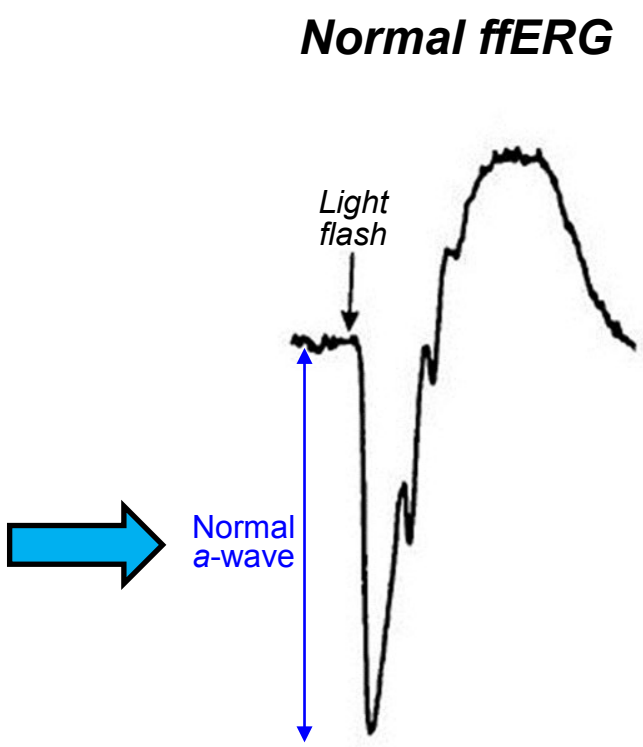



**The normal ffERG**. Note:
--The response commences with the stimulus flash
--The first deflection—the name of it—is sharply downward.

# Retinitis Pigmentosa

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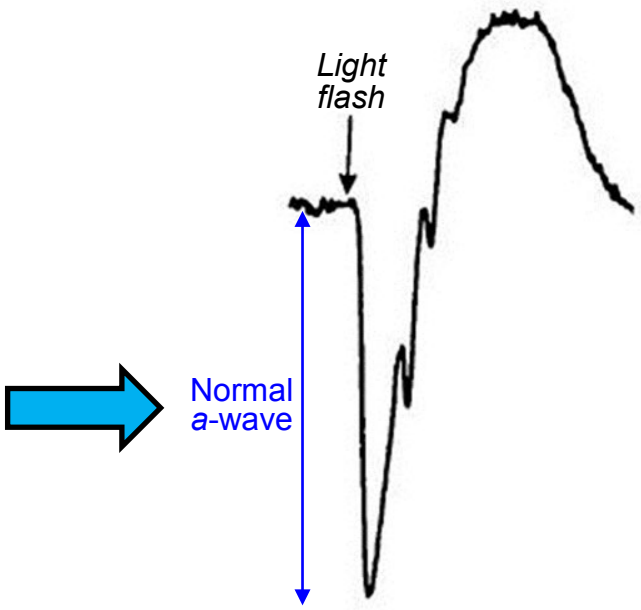


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The *a*-wave represents [redacted] function

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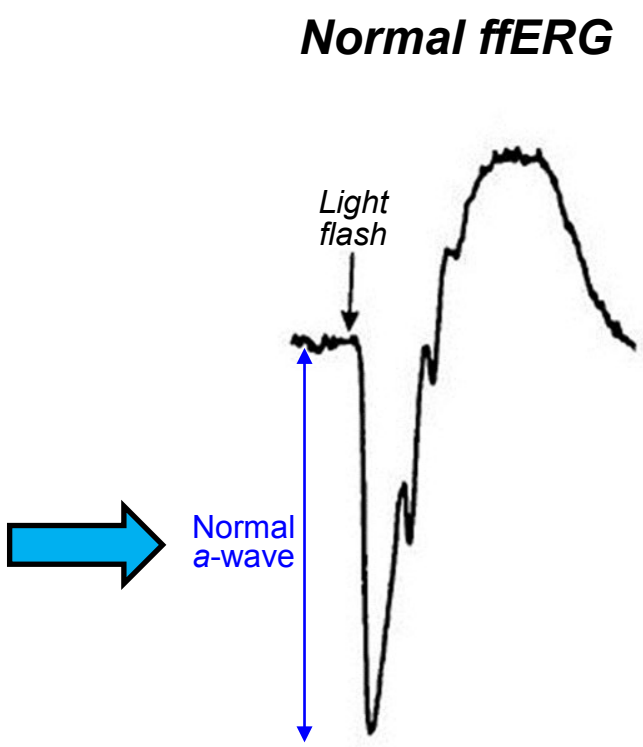


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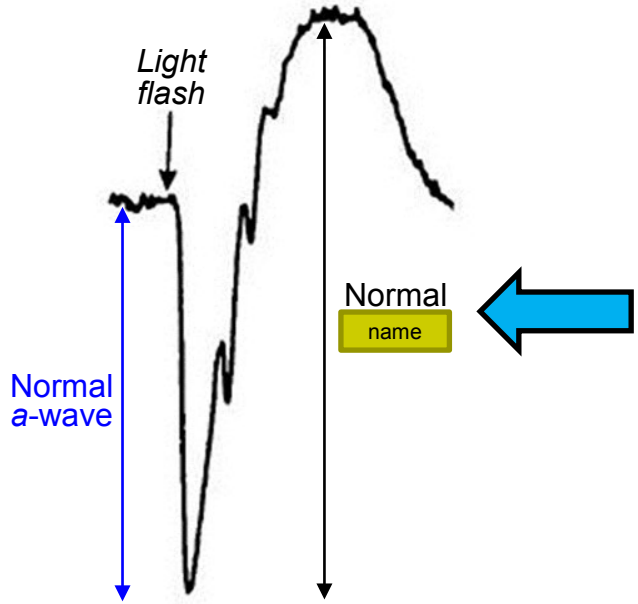


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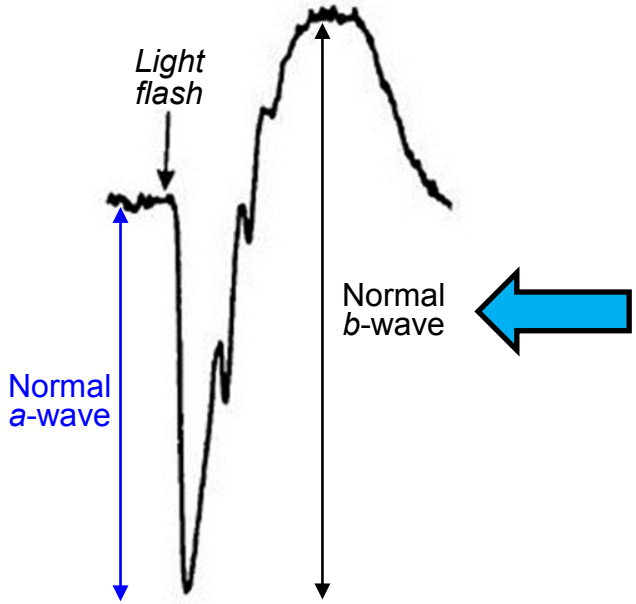


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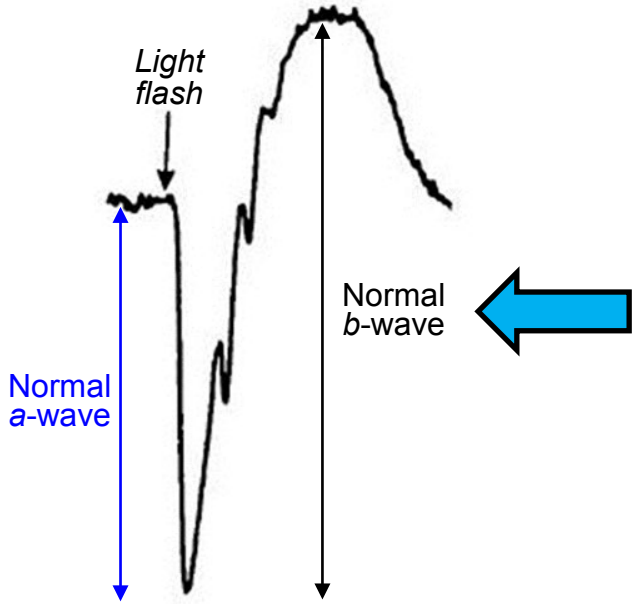


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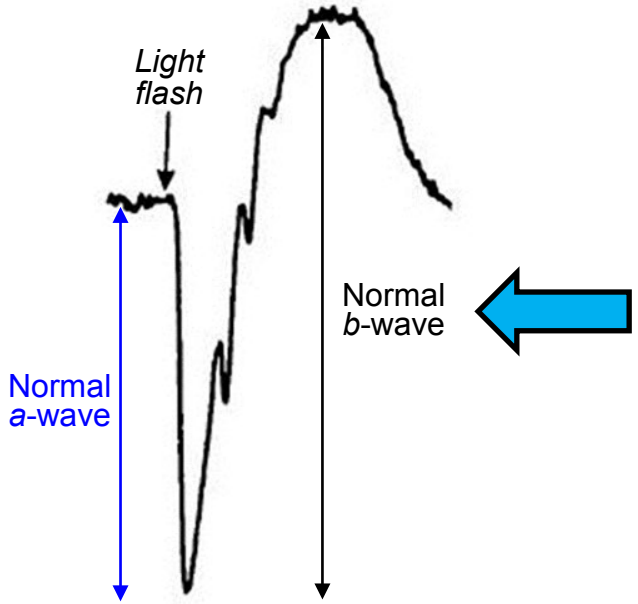


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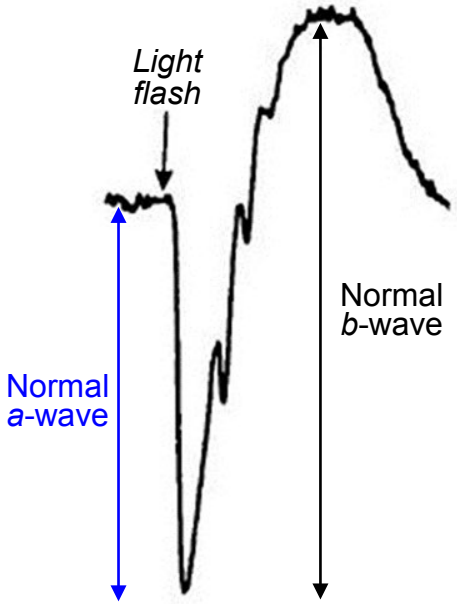


Note: This is a significant oversimplification of ERG interpretation, That said, I *think* it's enough to get you through the OKAP and Boards. But caveat emptor, bro.

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# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
--Choroideremia

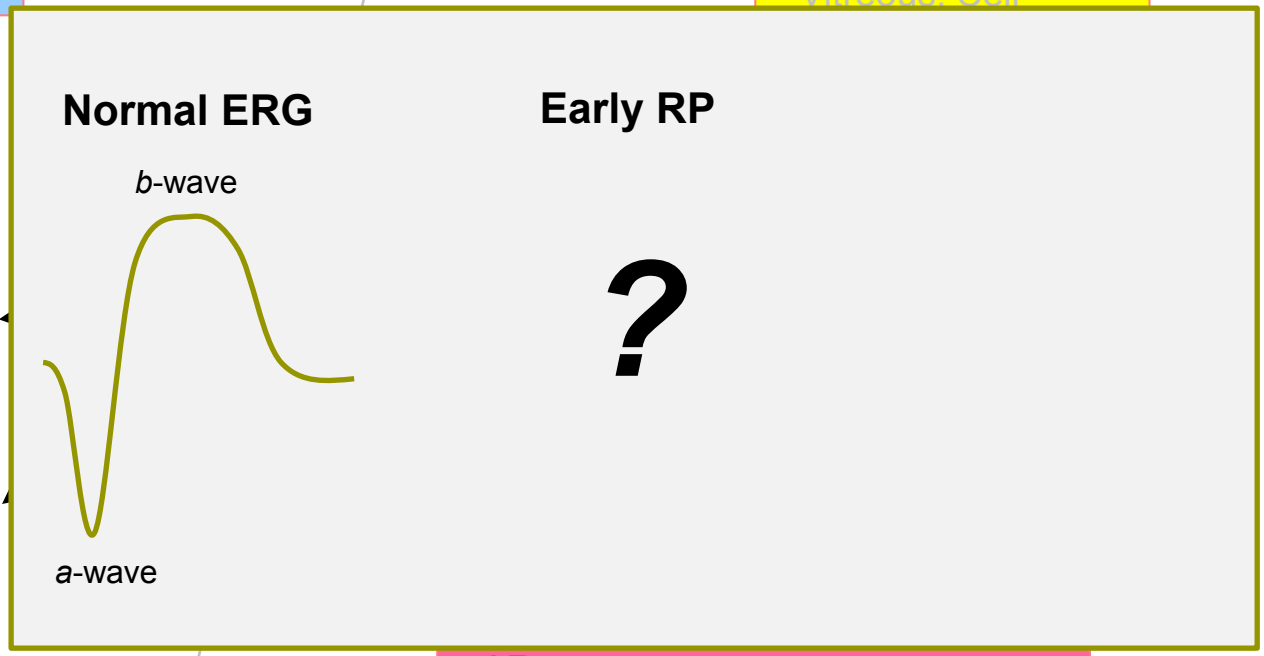
Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
**--abnormal ERG**
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
--Waxy disc pallor
--Arteriolar narrowing

Other common signs:
--ONH: Drusen
--Fovea: CME/CMD
--Vitreous: Cell

*Now we're ready to tackle these Qs*

Characteristic ERG changes in RP:
--Early:
--Late:




3 classic 'variant' forms of RP:
--Sectorial
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--AR
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*Typical pattern of VF loss:* Mid-peripheral scotomata → coalesce into partial ring → coalesce into complete ring → expand rapidly outward → expand slowly inward

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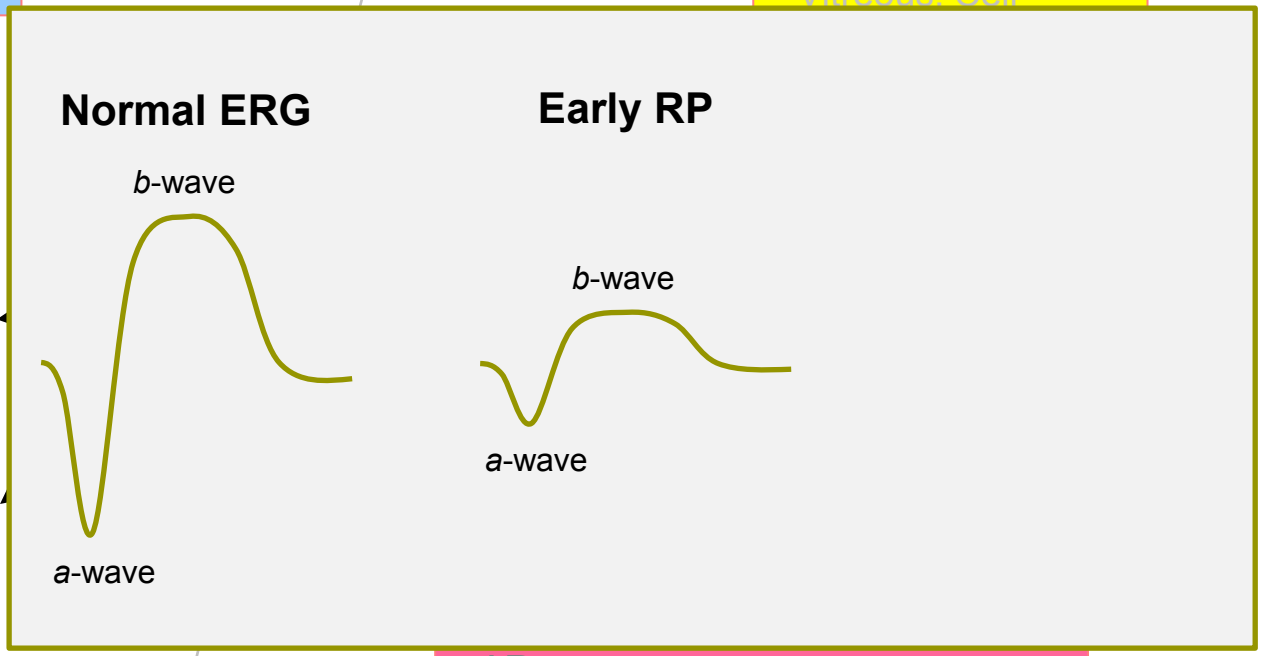


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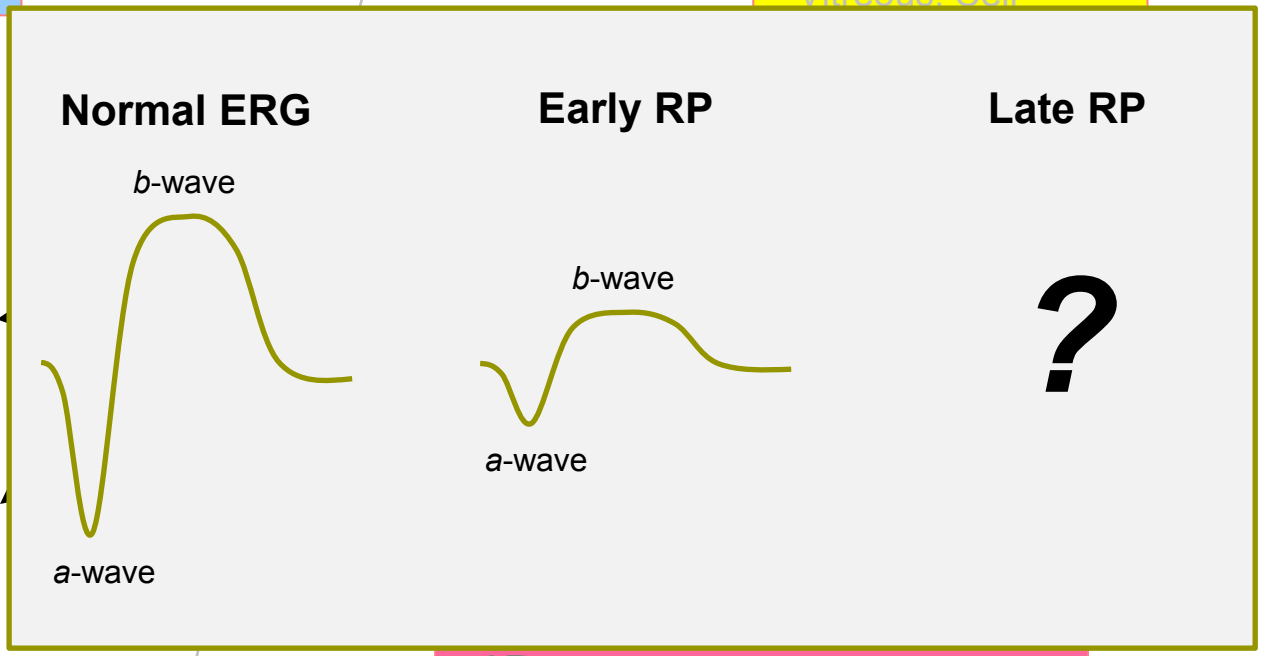


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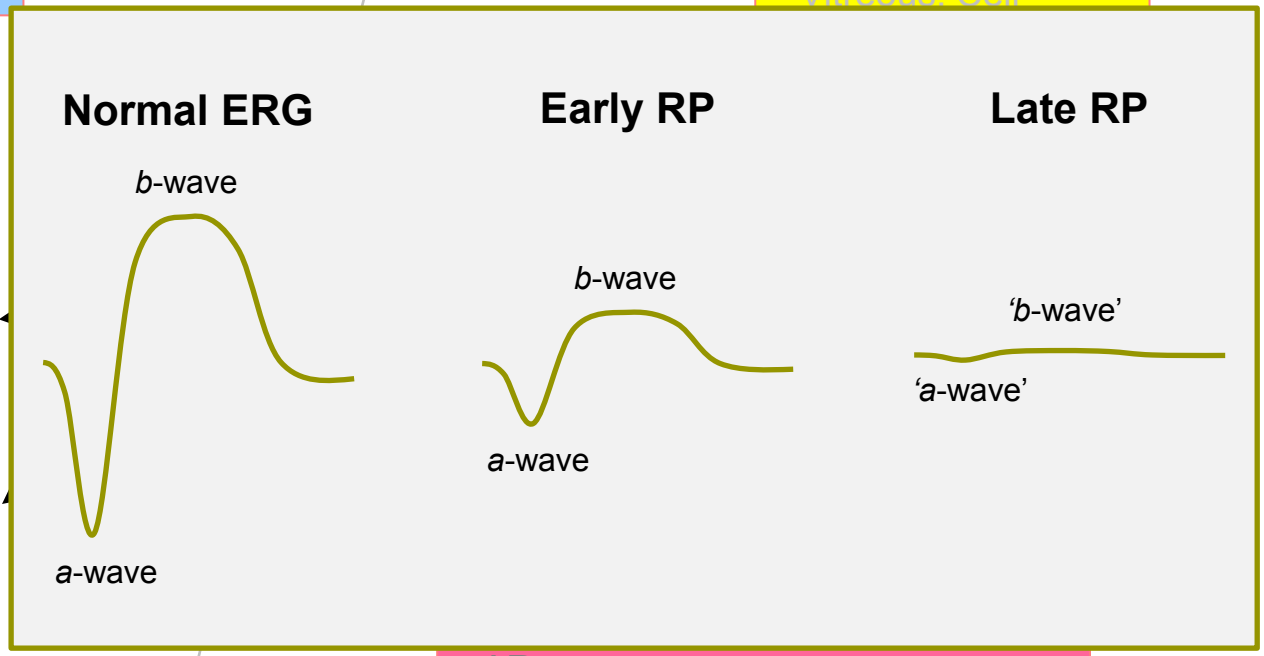


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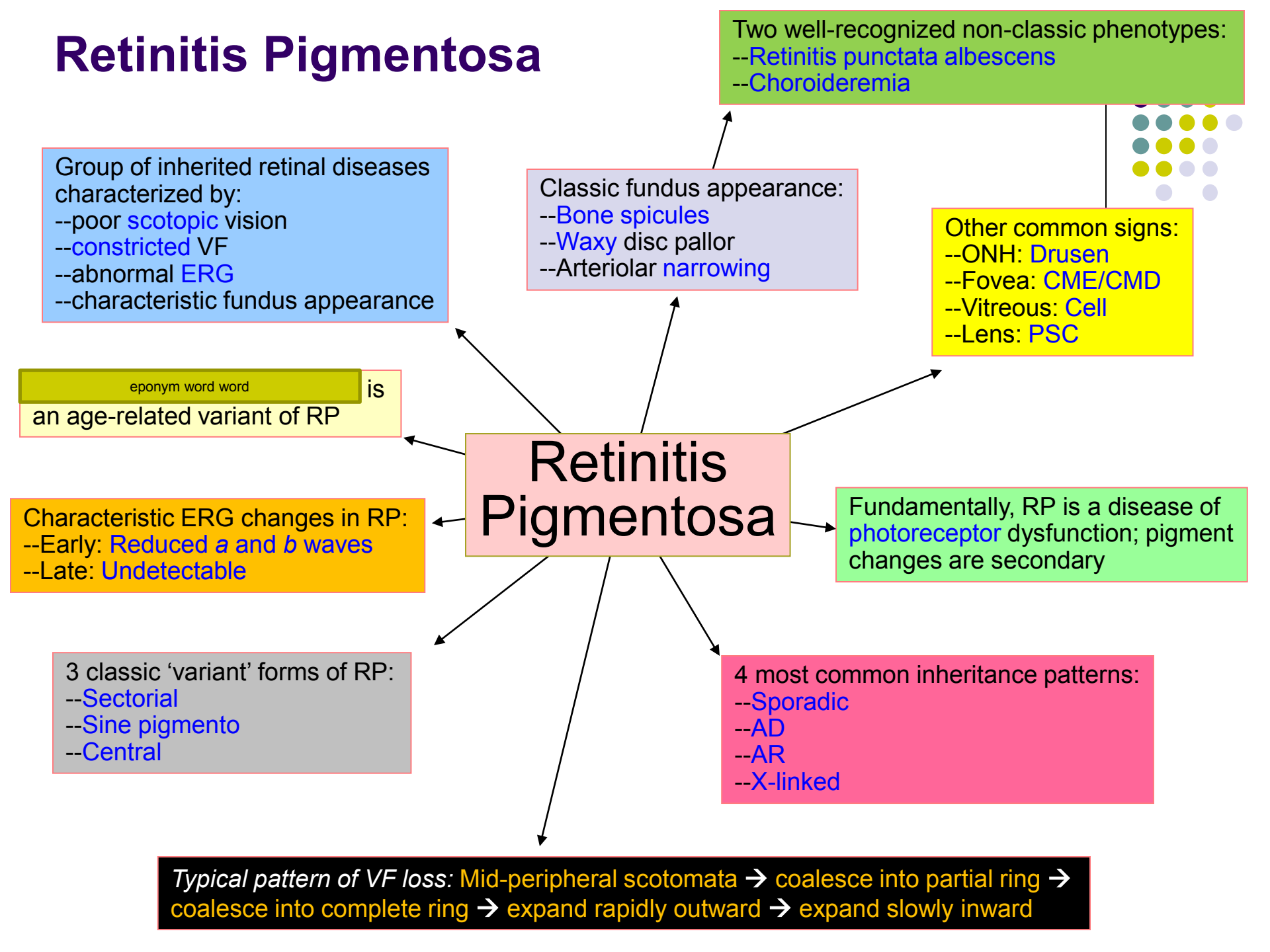

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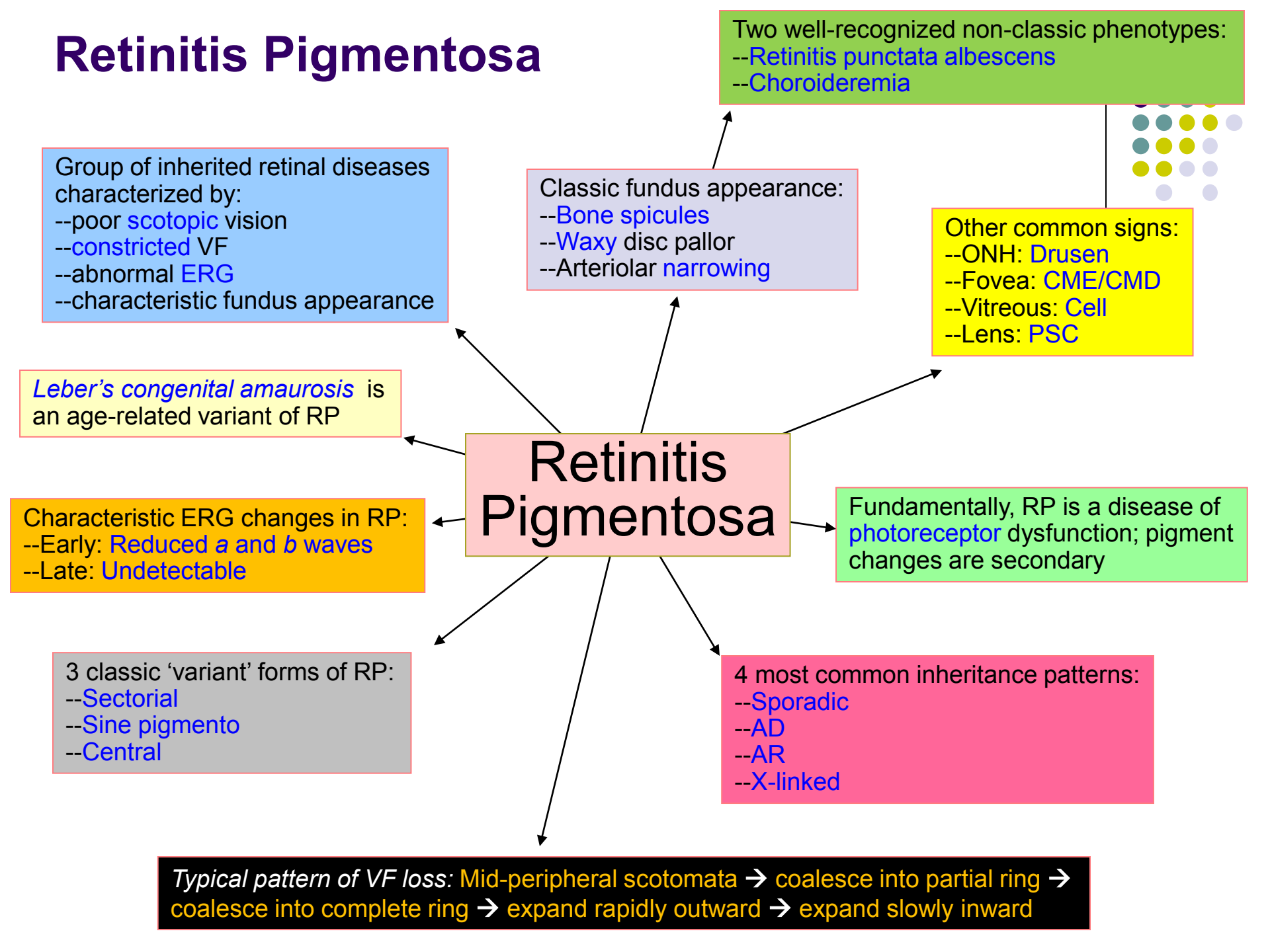

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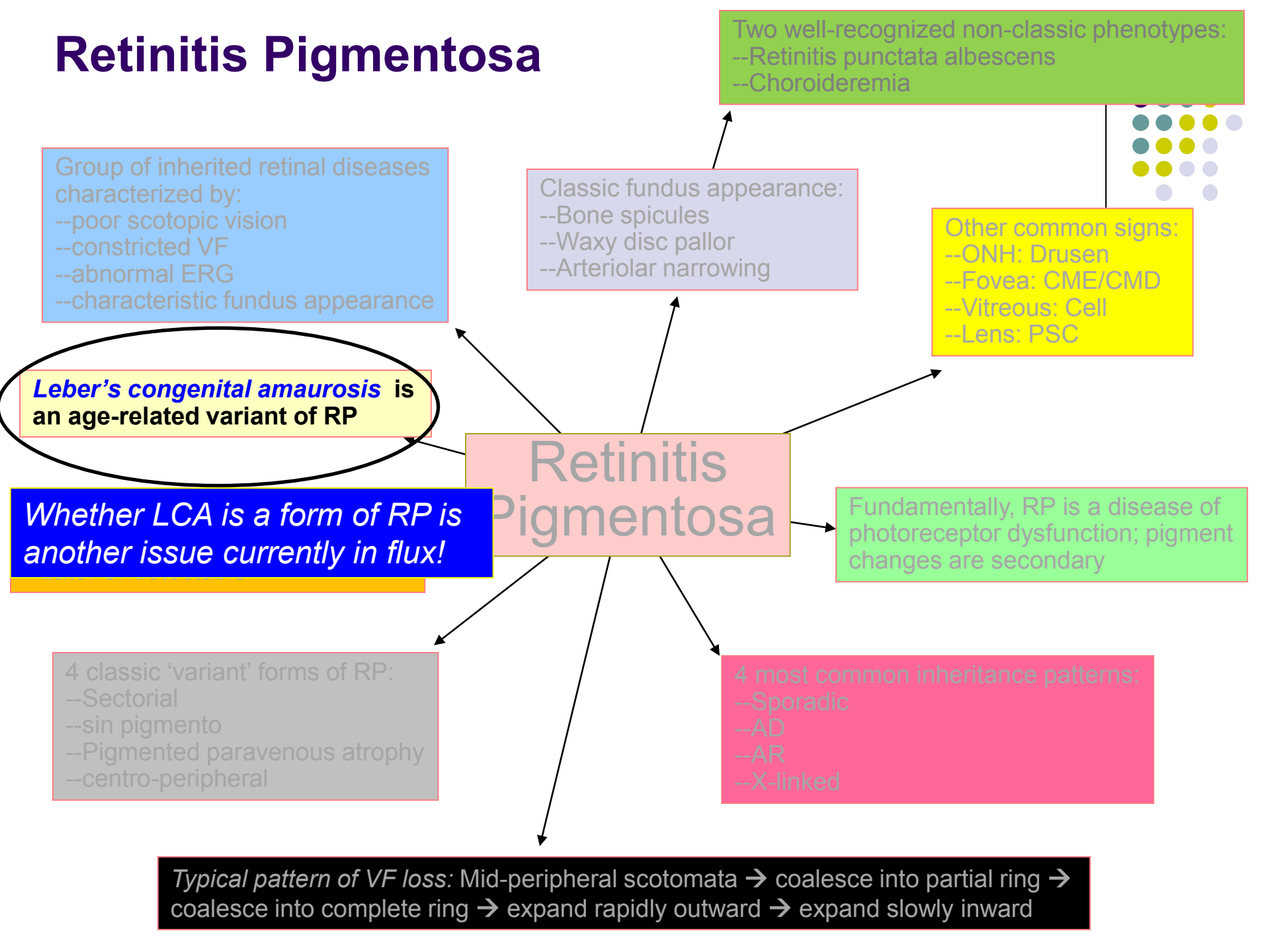

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**Usher syndrome =** *Retinitis pigmentosa* + two words

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# Retinitis Pigmentosa

Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
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Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

Classic fundus appearance:
--Bone spicules
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Other common signs:
--ONH: Drusen
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**Usher syndrome =** *Retinitis pigmentosa* + sensorineural deafness

*Where does Usher syndrome rank as a cause of deaf-blindness in the US?*
It is the most common cause thereof

*There are three types of Usher syndrome--what are they called?*
--**Type I**
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4 classic 'variant' forms of RP:
--Sectorial
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**Usher syndrome tl;dr**
= RP + hearing loss +/- vestibular dysfunction
--Type I: Early, severe
--Type II: Later, less severe
--Type III: Variable

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