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

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

Retinitis Pigmentosa
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
Retinitis Pigmentosa

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

Retinitis Pigmentosa
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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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

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

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

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

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

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

Classic fundus appearance:
- two words
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 appearance:
--Bone spicules

What are bone spicules?

Bone spicules:
Focal accumulations of pigment released when dying RPE cells disintegrate.
Retinitis Pigmentosa

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

Classic fundus appearance:
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What are bone spicules?
Focal accumulations of pigment released when dying RPE cells disintegrate.
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
--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 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
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

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

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

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?

Syphilis
Retinitis Pigmentosa

RP-like fundus in syphilis
Retinitis Pigmentosa

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

Group of inherited retinal diseases characterized by:
-- Poor scotopic vision
-- Constricted VF
-- Abnormal ERG
-- Characteristic fundus appearance

Two well-recognized non-classic phenotypes:
-- Three words
-- One word

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

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

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

Retinitis Pigmentosa
Group of inherited retinal diseases characterized by:
-- Poor scotopic vision
-- Constricted VF
-- Abnormal ERG
-- Characteristic fundus appearance

Retinitis Pigmentosa

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

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

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

Appearance-wise, what is the hallmark of choroideremia?

Appearance-wise, what is the hallmark of choroideremia?

Appearance-wise, what is the hallmark of choroideremia?
Retinitis Pigmentosa

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

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

Retinitis Pigmentosa
Retinitis Pigmentosa

Retinitis punctata albescens
Retinitis Pigmentosa

- Bone spicules
- Waxy disc pallor
- Arteriolar narrowing

Retinitis Pigmentosa is a group of inherited retinal diseases characterized by:
- Poor scotopic vision
- Constricted VF
- Abnormal ERG
- Characteristic fundus appearance

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

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

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

Retinitis Pigmentosa

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

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

Choroideremia
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.

**Retinitis Pigmentosa**

- Bone spicules
- Waxy disc pallor
- Arteriolar narrowing

Retinitis Pigmentosa

**Two well-recognized non-classic phenotypes:**

- Retinitis punctata albescens
- Choroideremia

**Retinitis punctata albescens**

- Myriad white dots/flecks in the deep retina

**Choroideremia**

- Pronounced atrophic changes of the RPE, choriocapillaris and choroid
Retinitis Pigmentosa

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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?

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

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

Group of inherited retinal diseases characterized by:
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--constricted VF
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Retinitis Pigmentosa

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

Group of inherited retinal diseases characterized by:
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Two well-recognized non-classic phenotypes:
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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?

Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of CSNB

CSNB

Congenital stationary night blindness
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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Two well-recognized non-classic phenotypes:
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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
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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Retinitis Pigmentosa

- Bone spicules
- Waxy disc pallor
- Arteriolar narrowing

Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

Retinitis Pigmentosa

- OK, what’s the difference?
Retinitis Pigmentosa

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Retinitis Pigmentosa
Group of inherited retinal diseases characterized by:
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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

**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.

**Abb.**

Congenital stationary night blindness
Retinitis Pigmentosa

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

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

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

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

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

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

OK, what’s the difference?
Whereas retinitis punctata albscens 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

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--Retinitis punctata albescens
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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

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 it mean to say the night blindness is stationary?

Congenital **stationary** night blindness
Retinitis Pigmentosa

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Whereas retinitis punctata albescens is a non-classic phenotype of RP, fundus albipunctatus is a non-classic phenotype of CSNB

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)

Congenital stationary night blindness
Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

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

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

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

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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 albscessens

How else do fundus albipunctatus and retinitis punctata albscessens differ?

Retinitis Pigmentosa

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

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

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

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Retinitis Pigmentosa

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--poor scotopic vision
--constricted VF
--abnormal ERG
--characteristic fundus appearance

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

---Retinitis Pigmentosa

Retinitis Pigmentosa

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

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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)

How else do fundus albipunctatus and retinitis punctata albescens differ?
--On DFE:
Like other forms of RP, retinitis punctata albescens demonstrates arteriolar narrowing, whereas fundus albipunctatus does not

--On ERG:
Fundus albipunctatus is a disease of abnormal rhodopsin regeneration, which manifests as slow but ultimately successful dark adaptation. In contrast, retinitis punctata albescens is a photoreceptor disease; therefore, dark adaptation does not occur and the ERG never normalizes, no matter how much time is allowed to elapse.
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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Two well-recognized non-classic phenotypes:
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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

In terms of both its name and appearance, with what disease is fundus albipunctatus often confused?

Fundus flavimaculatus

By what other name is fundus flavimaculatus/Stargardt disease known?
Juvenile macular dystrophy

Pts with fundus albipunctatus c/o night blindness. What do pts with fundus flavimaculatus complain of?
Decreased visual acuity
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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Retinitis Pigmentosa

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

In terms of both its name and appearance, with what disease is fundus albipunctatus often confused?
Fundus flavimaculatus

Two well-recognized non-classic phenotypes:
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Retinitis Pigmentosa

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Fundus flavimaculatus

CSNB

Congenital stationary night blindness

In terms of both its name and appearance, with what disease is fundus albipunctatus often confused?
Fundus flavimaculatus

Is fundus albipunctatus the same thing as fundus flavimaculatus?
No, and it's very important to know 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

Fundus albipunctatus

Fundus flavimaculatus
Retinitis Pigmentosa

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Fundus flavimaculatus

Is fundus albipunctatus the same thing as fundus flavimaculatus?

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**Fundus alipunctatus**

In terms of both its name and appearance, with what disease is fundus alipunctatus often confused?
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Is fundus alipunctatus the same thing as fundus flavimaculatus?
No, and it's very important to know the difference

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

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Is fundus albipunctatus the same thing as fundus flavimaculatus?
No, and it’s very important to know the difference

OK, what’s the difference?

Fundus flavimaculatus

By what other name is fundus flavimaculatus/Stargardt disease known?
Juvenile macular dystrophy

Pts with fundus albipunctatus c/o night blindness. What do pts with fundus flavimaculatus complain of?
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Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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CSNB
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--Bone spicules
--Waxy disc pallor
--Arteriolar narrowing

Other common signs:
--ONH: not waxy pallor

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Cystic foveal changes in RP

Courtesy of Dr. Stephen Tsang
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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?
Steroids

What about CM? How is it treated?
It’s not treatable
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Topical, or PO?

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Periocular or intravitreal

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Vitreous cell? Does this mean RP is an inflammatory condition?

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Vitreous cell? Does this mean RP is an inflammatory condition? No. The vitreous cells seen in RP are mainly RPE cells liberated from the degeneration of that structure.

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Classic fundus appearance:
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Can the PSC in RP become visually significant, thereby rendering surgical extraction warranted?

Retinitis Pigmentosa

Absolutely, yes

Is CE in RP associated with an increased risk of intra-op complications?
Yes

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--CME
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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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Per the newest (at the time of this writing) edition of the Retina book, the term retinitis pigmentosa is “no longer preferred.” Per the book, what term is preferred instead?
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4 most common inheritance patterns:
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Of the 4 most common inheritance patterns, which is… …most common?

Of the 4 most common inheritance patterns, which is… …carries the worst visual prognosis?
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Typical pattern of VF loss: [specific pattern of VF loss found in early RP]

(Start here)
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Typical pattern of VF loss: Mid-peripheral scotomata → [how the VF evolves next]
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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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[how the VF evolves next]
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Group of inherited retinal diseases characterized by:
--poor scotopic vision
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Classic fundus appearance:
--Bone spicules
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Other common signs:
--ONH: Drusen
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--Early:
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Normal ERG
Early RP

a-wave
b-wave

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Late RP

a-wave

b-wave

'b-wave'

'a-wave'

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

- **Group of inherited retinal diseases characterized by:**
  - Poor scotopic vision
  - Constricted VF
  - Abnormal ERG
  - Characteristic fundus appearance

- **Characteristic ERG changes in RP:**
  - Early: Reduced *a* and *b* waves
  - Late: Undetectable

- **3 classic ‘variant’ forms of RP:**
  - Sectorial
  - Sine pigmento
  - Central

- **Typical pattern of VF loss:**
  - Mid-peripheral scotomata → coalesce into partial ring → coalesce into complete ring → expand rapidly outward → expand slowly inward

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

- **4 most common inheritance patterns:**
  - Sporadic
  - AD
  - AR
  - X-linked

- **Other common signs:**
  - ONH: Drusen
  - Fovea: CME/CMD
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- **Fundamentally, RP is a disease of photoreceptor dysfunction; pigment changes are secondary**

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- **Leber's congenital amaurosis** is an age-related variant of RP.
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Whether LCA is a form of RP is another issue currently in flux!

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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
--Waxy disc pallor
--Arteriolar narrowing

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

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

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Two well-recognized non-classic phenotypes:
--Retinitis punctata albescens
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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? How do they manifest?
--Type I manifests...in the first decade with profound hearing loss, RP and vestibular dysfunction
--Type II manifests...in the second decade with moderate hearing loss, RP; vestibular function is intact
--Type III has... hearing loss; the RP...in severity; vestibular function is...

4 classic 'variant' forms of RP:
--Sectorial
--sin pigmento
--Pigmented paravenous atrophy
--centro-peripheral

4 most common inheritance patterns:
--Sporadic
--AD
--AR
--X-linked

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Characteristic ERG changes in RP:
--Early: Reduced a and b waves
--Late: Undetectable

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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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