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

Group of inherited retinal diseases characterized by:

-- poor scotopic vision

VF

-- ?

-- ?
Retinitis Pigmentosa

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

(We will drill down on this shortly)
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 \textit{abb.}
--?
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

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

(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 appearance:
--Bone spicules
--?
--?

What are 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?
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

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

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

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
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

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:
-- three words
-- one word
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
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?

Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

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

Retinitis Pigmentosa
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

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

Retinitis Pigmentosa

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

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

**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

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

**Retinitis Pigmentosa**

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

**Retinitis Pigmentosa**

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

What is the hallmark of choroideremia?

Retinitis Pigmentosa

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

Retinitis Pigmentosa

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

Choroideremia
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 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?
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

- 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:
- Retinitis punctata albescens
- Choroideremia

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.

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
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 albescens
--Choroideremia

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

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

Retinitis punctata albescens

Fundus albipunctatus
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 albescens
- Choroideremia

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?

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
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 albescens
--Choroideremia

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
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

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?

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

Congenital stationary night blindness
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 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.

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

**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?**
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

**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

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

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

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 CSNB stand for?

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

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 CSNB stand for?
Congenital stationary night blindness
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

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

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:
--Retinitis punctata albescens
--Choroideremia

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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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?
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

CSNB

Congenital stationary night blindness
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

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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

How else do fundus albipunctatus and retinitis punctata albescens differ on DFE?

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

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

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

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

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

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

OK, what’s the difference?
Whereas retinitis punctata albsens 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 albsens (which, like all RP, is relentlessly progressive)

How else do fundus albipunctatus and retinitis punctata albsens differ on DFE?
Like other forms of RP, retinitis punctata albsens demonstrates arteriolar narrowing, whereas fundus albipunctatus does not
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

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

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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)

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

Retinitis Pigmentosa

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

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

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

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

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

Retinitis Pigmentosa

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

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

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:
--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

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

Retinitis Pigmentosa

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

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
Retinitis Pigmentosa

Fundus albipunctatus

Fundus flavimaculatus
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 albescens
-- Choroideremia

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

Is fundus albipunctatus the same thing as fundus flavimaculatus?

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

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?

Whereas fundus albipunctatus is a non-classic phenotype of CSNB, fundus flavimaculatus is a classic variant of Stargardt disease.

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

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

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

Fundus flavimaculatus

Is congenital stationary night blindness the same as Stargardt disease?
Yes, they are synonymous.

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

Fundus albipunctatus

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

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

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?
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OK, what’s the difference?
Whereas fundus albipunctatus is a non-classic phenotype of CSNB, fundus flavimaculatus is a classic variant of Stargardt disease.

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What does CSNB stand for?
Congenital stationary night blindness

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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:
--poor scotopic vision
--constricted VF
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--characteristic fundus appearance

- Bone spicules
- Waxy disc pallor
- Arteriolar narrowing

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Retinitis Pigmentosa

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Juvenile macular dystrophy

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

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--poor scotopic vision
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--characteristic fundus appearance

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

Other common signs:
--ONH: not waxy pallor
--?
--?
--?

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

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---?
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Other common signs:
--ONH: Drusen
--Fovea: CME/CMD
--?
Cystic foveal changes in RP
Retinitis Pigmentosa

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What do these stand for?
CME:
CMD:

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Retinitis Pigmentosa

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CME is…‘wet’ (ie, leaks on FA)
CMD is…‘dry’ (no leakage on FA)
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What is the difference between the two?
CME is…‘wet’ (ie, leaks on FA)
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What proportion of RP pts will develop CME?

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

Retinitis Pigmentosa

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

What is second-line?
Steroids

CM:
It's not treatable
Retinitis Pigmentosa

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CME: Cystoid macular edema
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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?
Steroids

What about CMD? How is it treated?
It’s not treatable
Retinitis Pigmentosa

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What about CM?
CME is not treatable
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- What about CM? How is it treated?
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What proportion of RP pts will develop CME?
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What is the first-line tx for CME in RP?
Acetazolamide

Topical, or PO?

What about CM?
Topical can be effective, but most clinicians probably go PO
Retinitis Pigmentosa

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Topical can be effective, but most clinicians probably go PO

CM: Cystoid macular degeneration

What about CM: Cystoid macular degeneration? How is it treated?

It's not treatable

Topical, or PO?

Topical can be effective, but most clinicians probably go PO
Retinitis Pigmentosa

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**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

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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

- Retinitis Pigmentosa
Retinitis Pigmentosa

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10-20% or so

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Acetazolamide

What is second-line?
Steroids

What route(s)?
Periocular, or intravitreal

Retinitis Pigmentosa

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Steroids

Retinitis Pigmentosa
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What about CMD? How is it treated?
Retinitis Pigmentosa

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What is second-line?
Steroids

What about CMD? How is it treated?
It’s not treatable
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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-- 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:

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

Retinitis Pigmentosa
Retinitis Pigmentosa

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

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

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

A group of inherited retinal diseases characterized by:
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Classic fundus appearance:
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- ONH: Drusen
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Vitreous cell? Does this mean RP is an inflammatory condition?

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Retinitis Pigmentosa
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--ONH: Drusen
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Retinitis Pigmentosa

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.
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
--poor scotopic vision
--constricted VF
--abnormalVF
--characteristic fundus appearance:
--Bone spicules
--Waxy disc pallor

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

Retinitis Pigmentosa

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.

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

Retinitis Pigmentosa

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Classic fundus appearance:
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Retinitis Pigmentosa

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--Lens: 

Retinitis Pigmentosa

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Can the PSC in RP become visually significant, thereby rendering surgical extraction warranted?

Retinitis Pigmentosa

Yes

Yes
Retinitis Pigmentosa

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Retinitis pigmentosa

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

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**Can the PSC in RP become visually significant, thereby rendering surgical extraction warranted?**

Absolutely, yes

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

Yes

**Is CE in RP associated with an increased risk of post-op complications?**

Yes

**What is it about RP eyes that puts them at risk for intra-op complications?**

*Intra-op complications*
Retinitis Pigmentosa

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

Is CE in RP associated with an increased risk of post-op complications?
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What is it about RP eyes that puts them at risk for intra-op complications?
They tend to have zonular instability
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Is CE in RP associated with an increased risk of intra-op complications?
Yes

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

What two post-op complications do RP pts get at a higher rate than non-RP pts?
--?
--?
Retinitis Pigmentosa

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Yes

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

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

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

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Retinitis Pigmentosa

Poorest scotopic vision
Constricted VF
Abnormal ERG
Characteristic fundus appearance

Retinitis Pigmentosa

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

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

Is RP a rod-cone dystrophy, or a cone-rod dystrophy?

It is a rod-cone dystrophy.
Retinitis Pigmentosa

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

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Rod-cone dystrophy
Retinitis Pigmentosa

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4 most common inheritance patterns:
--?
--?
--?
--?
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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4 most common inheritance patterns:
--Sporadic
--AD
--AR
--X-linked

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Retinitis Pigmentosa

Of the 4 most common inheritance patterns, which is...
...most common?

Retinitis Pigmentosa

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Retinitis Pigmentosa

Of the 4 most common inheritance patterns, which is…
…most common? Sporadic

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Of the 4 most common inheritance patterns, which is...
...most common? **Sporadic**
...least common?

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Retinitis Pigmentosa is a disease of photoreceptor dysfunction; pigment changes are secondary.
Retinitis Pigmentosa

Group of inherited retinal diseases characterized by:
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Of the 4 most common inheritance patterns, which is...
...most common? **Sporadic**
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...carries the worst visual prognosis?

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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
Retinitis Pigmentosa

Classic fundus appearance:
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--ONH: Drusen
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Typical pattern of VF loss: Mid-peripheral scotomata → [how the VF evolves next]
Two well-recognized non-classic phenotypes:
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--Choroideremia

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

4 most common inheritance patterns:
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Typical pattern of VF loss: Mid-peripheral scotomata → coalesce into partial ring → coalesce into complete ring → [how the VF evolves next]
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In one sentence, what is it?
An [ ] test that measures how [ ] cells respond to a [ ] stimulus
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In one sentence, what is it?
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The pt is dilated vs undilated, and usually light- vs dark-adapted
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Normal ffERG

The normal ffERG. Note:
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--The first deflection—the name of it—is sharply downward.
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The b-wave represents function of the inner retina

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.
Retinitis Pigmentosa

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

Now we’re ready to tackle these Qs

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

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

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

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

Early RP

a-wave

b-wave

3 most common inheritance patterns:
--Sporadic
--AD
--AR
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Fundamentally, RP is a disease of photoreceptor dysfunction; pigment changes are secondary

Other common signs:

Normal ERG Early RP

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**Normal ERG**

- *a*-wave
- *b*-wave

**Early RP**

- *b*-wave
- *a*-wave

**Late RP**

- ؟

**Removal**

-- AR
-- X-linked
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Whether LCA is a form of RP is another issue currently in flux!

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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

Retinitis Pigmentosa
Leber's congenital amaurosis is an age-related variant of RP.

Characteristic ERG changes in RP:
- Early: Reduced a and b waves
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4 classic 'variant' forms of RP:
- Sectorial
- Sin pigmento
- Pigmented paravenous atrophy
- Centro-peripheral

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

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

Other common signs:
- ONH: Drusen
- Fovea: CME/CMD
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Classic fundus appearance:
- Bone spicules
- Waxy disc pallor
- Arteriolar narrowing

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...progressive hearing loss; the RP varies in severity; vestibular function is sporadic.

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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Usher syndrome tl;dr:
= RP + hearing loss +/- vestibular dysfunction
-- Type I: Early, severe
-- Type II: Later, less severe
-- Type III: Variable

Retinitis Pigmentosa