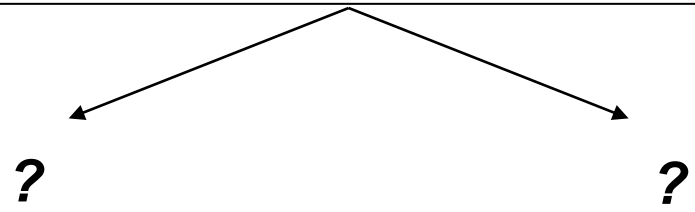
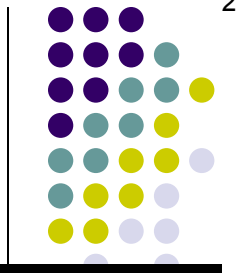
Congenital/Stationary Retinal Disease



Two very basic categories

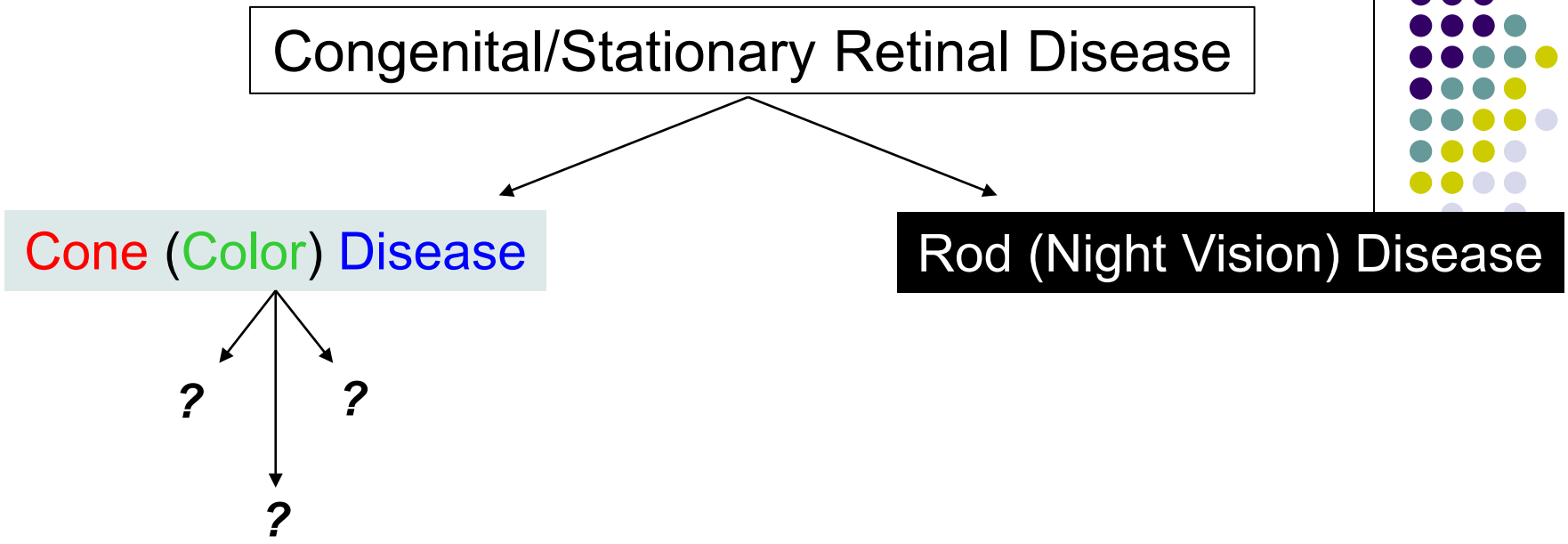


Congenital/Stationary Retinal Disease

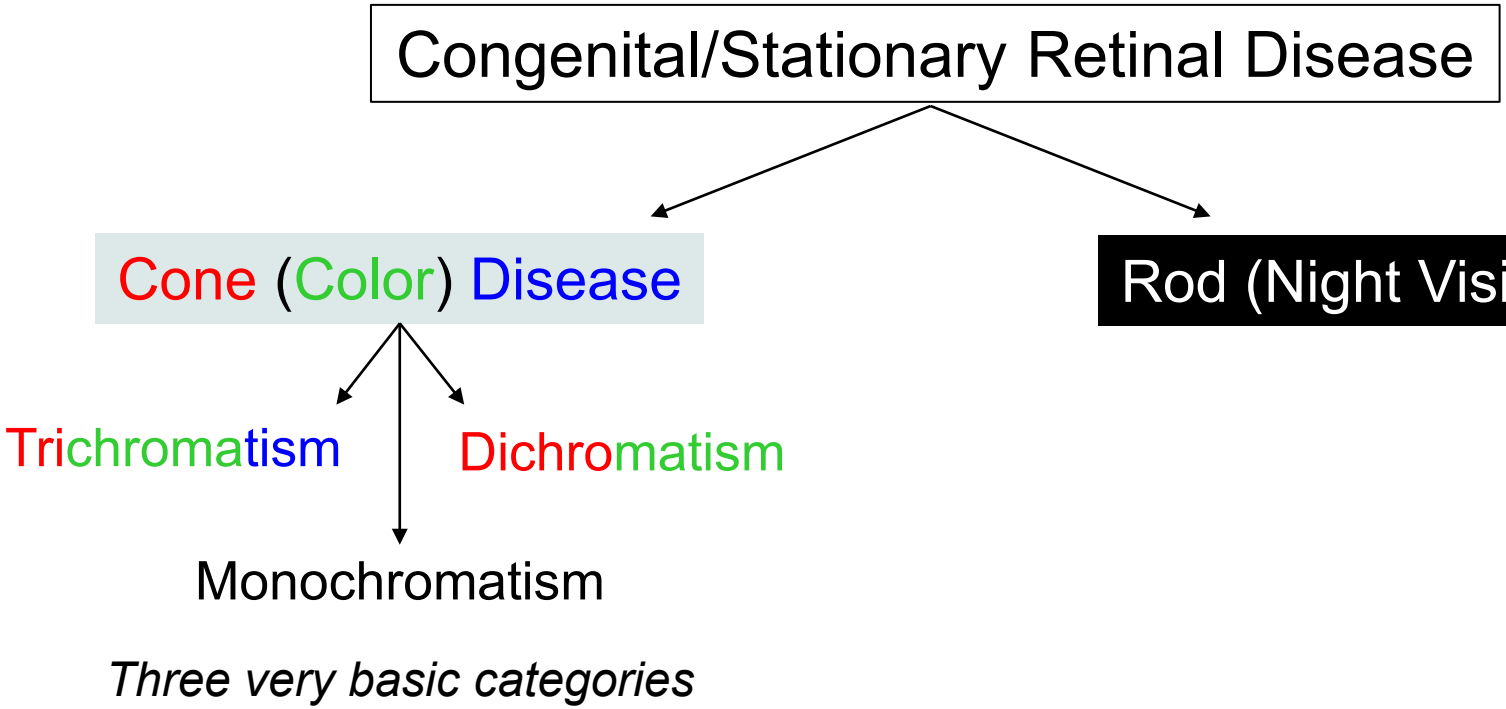
Cone (Color) Disease

Rod (Night Vision) Disease

Two very basic categories



Three very basic categories



Three very basic categories

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

What does it mean to say someone is a 'trichromat'?

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

What does it mean to say someone is a 'trichromat'?
It concerns performance on a *color-matching test*. In this test, the participant is asked to match a test color by mixing primary-color lights (note--**not** mixing paints!).

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What does it mean to say someone is an 'anomalous' trichromat?

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

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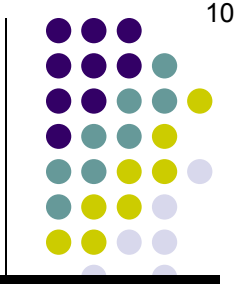
What does it mean to say someone is an 'anomalous' trichromat?
It means he needs all three colored lights to do the matching, but that the relative intensities among the lights differs significantly from that employed by people with normal color vision (which color is abnormally intense is a function of what sort of anomalous trichromacy he has)

Congenital/Stationary Retinal Disease



Cone (Color) Disease

Rod (Night Vision) Disease

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Dude, wussup with the gendered language?

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

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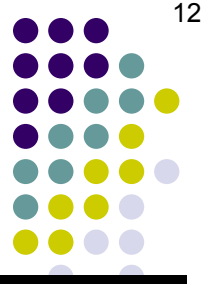
Dude, wussup with the gendered language?
The genetics relevant to anomalous color vision are predominantly X-linked recessive, so the **vast** majority of individuals with color deficiencies are males (including yours truly)

Congenital/Stationary Retinal Disease



Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

*What does it mean to say someone is a **dichromat**?*

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

What does it mean to say someone is a **dichromat***?*
It means that, on the color-matching test, he can match any test color using only *two* lights. (Which two depends upon the form of dichromacy, but the missing one is almost always vs never blue.)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

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Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

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The fact that a dichromat can match any color with only two primaries indicates what about his cones?

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
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  - **Dichromatism**
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The fact that a dichromat can match any color with only two primaries indicates what about his cones?
It indicates his cones possess only *two* photopigments, not three as do the cones in trichromats

Congenital/Stationary Retinal Disease

Cone (Color) Disease

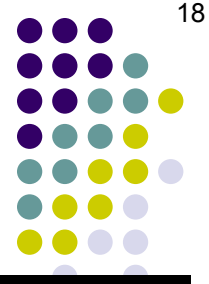
Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

By what other name is monochromatism known?



Congenital/Stationary Retinal Disease

Cone (Color) Disease

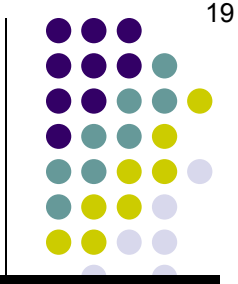
Rod (Night Vision) Disease

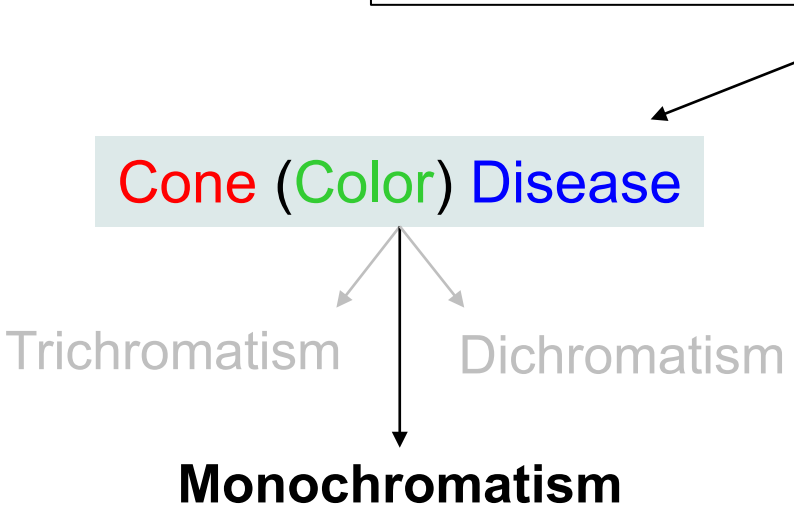
Trichromatism

Dichromatism

Monochromatism

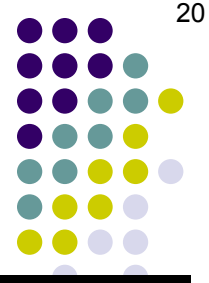
By what other name is monochromatism known?
Achromatopsia





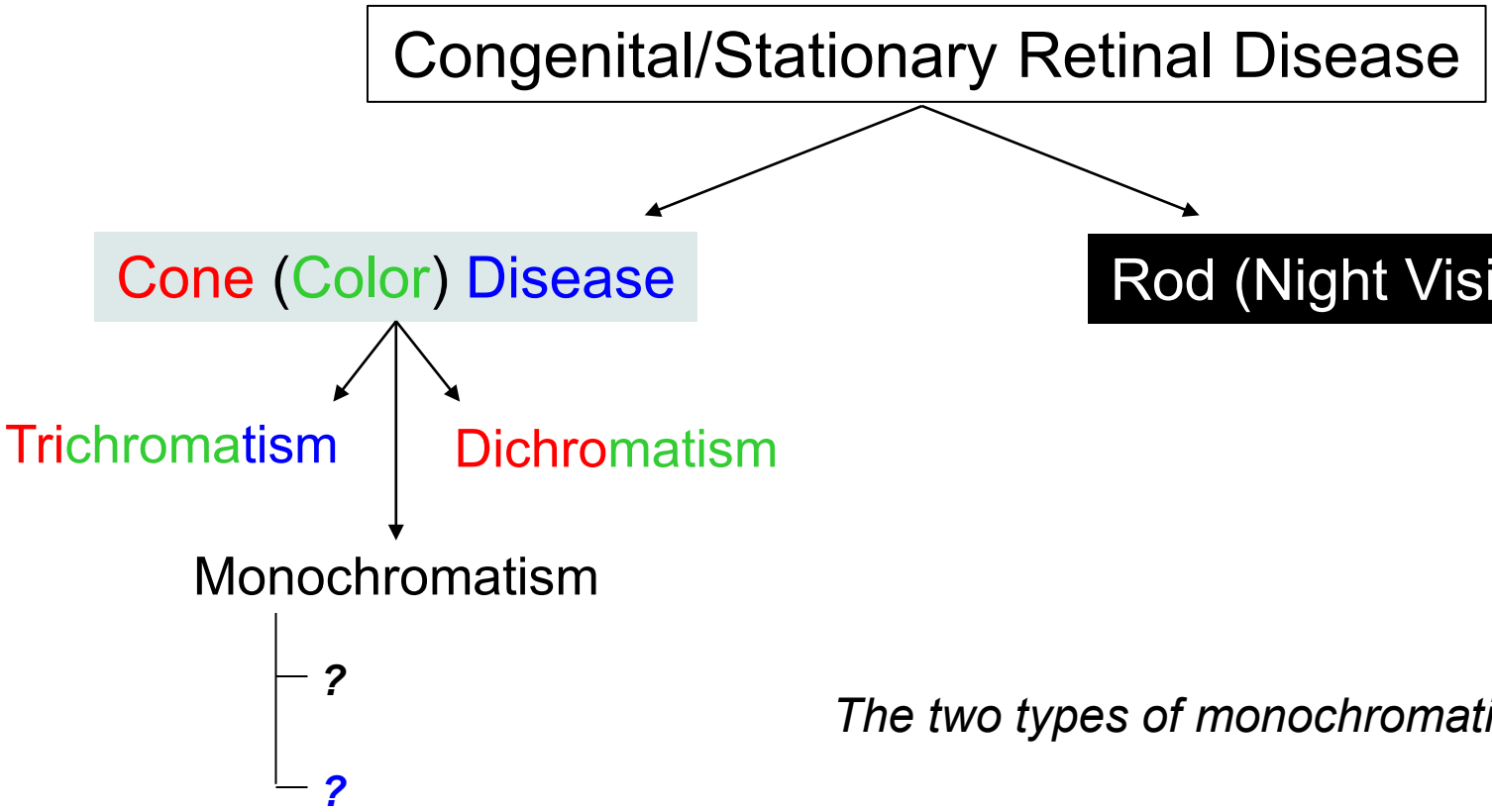
By what other name is monochromatism known?
Achromatopsia

Does monochromatism/achromatopsia mean what I think it does?

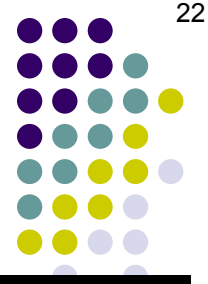


By what other name is monochromatism known?
Achromatopsia

Does monochromatism/achromatopsia mean what I think it does?
Yes--it is the state in which an individual can match any test color using just **one** color of light



The two types of monochromatism are…



Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
    - Rod monochromatism
    - Blue-cone monochromatism
- Rod (Night Vision) Disease

The two types of monochromatism are…

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Rod monochromatism

Blue-cone monochromatism

Rod monochromatism

--Inheritance…

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
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  - **Rod monochromatism**
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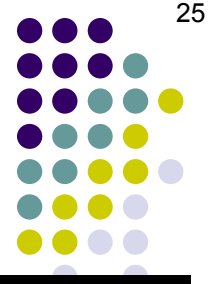
Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - **Rod monochromatism**
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

-- EOM issue always present

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

-- **Nystagmus** always present

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

-- **Nystagmus** always present

--VA range:

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- **Rod monochromatism**
- Blue-cone monochromatism

Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

-- **Nystagmus** always present

--VA range: **20/80–20/200**

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- **Rod monochromatism**
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

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--**VA range: 20/80–20/200**

Why the broad range in VA?

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- **Rod monochromatism**
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

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--**VA range: 20/80–20/200**

Why the broad range in VA?
Because the dz manifests partial expression in some cases (ie, some pts will have a few functioning cones)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
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Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

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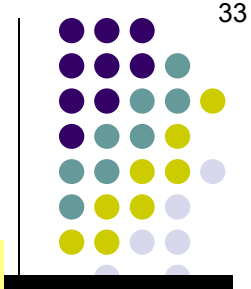
-- **Nystagmus** always present

--VA range: **20/80–20/200**

--ERG: ***?***

Congenital/Stationary Retinal Disease



Disease

Before we get into the weeds on this…What does ERG *stand for?*

ness

└ Blue-cone monochromatis

--VA range: **20/80–20/200**

--**ERG**: *?*

Congenital/Stationary Retinal Disease

Before we get into the weeds on this…What does ERG *stand for?*
Electroretinogram (or electroretinography)

Disease

ness

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Congenital/Stationary Retinal Disease

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In one sentence, what is it?

Disease

ness

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In one sentence, what is it?
An [redacted] test that measures how [redacted] cells respond to a [redacted] stimulus

Disease

ness

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Disease

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

Disease

ness

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Disease

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Disease

ness

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Disease

ness

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What are the three main types of ERG?
Full-field (ffERG, aka German word ERG), **multifocal** (mfERG), and **pattern** (pERG)

Disease

ness

└ Blue-cone monochromatis

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--VA range: **20/80–20/200**

ERG: *?*

ffERG: Demonstrates the response of the central vs entire retina to flash stimuli
mfERG
pERG

Congenital/Stationary Retinal Disease

Disease

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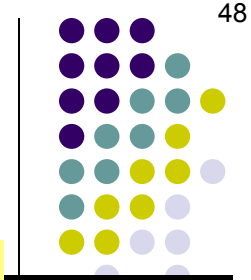
--**ERG**: *?*

ffERG: Demonstrates the response of the entire retina to flash stimuli
mfERG: Produces a [ ] map of central [cone vs rod] function
pERG

Congenital/Stationary Retinal Disease



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--VA range: **20/80–20/200**

--**ERG**: *?*

ffERG: Demonstrates the response of the entire retina to flash stimuli
mfERG: Produces a topographic map of central cone function
pERG

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Disease

ness

└ Blue-cone monochromatis

--VA range: **20/80–20/200**

ERG: *?*

ffERG: Demonstrates the response of the entire retina to flash stimuli
mfERG: Produces a topographic map of central cone function
pERG: Flashes a [redacted] pattern of rapidly alternating light-and-dark areas

Congenital/Stationary Retinal Disease

Disease

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Electroretinogram (or electroretinography)

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An electrophysiologic test that measures how retinal cells respond to a light stimulus

How is it performed?
The pt is dilated , and usually dark- adapted. Electrodes are attached to the pt's cornea and/or periocular skin, and a series of standardized visual stimuli (usually brief flashes) are presented.

What are the three main types of ERG?
Full-field (ffERG, aka *Ganzfeld* ERG), **multifocal** (mfERG), and **pattern** (pERG)

ness

└ Blue-cone monochromatis

--VA range: **20/80–20/200**

--**ERG**: *?*

ffERG: Demonstrates the response of the entire retina to flash stimuli
mfERG: Produces a topographic map of central cone function
pERG: Flashes a checkerboard pattern of rapidly alternating light-and-dark areas

Congenital/Stationary Retinal Disease

Disease

Before we get into the weeds on this…What does ERG *stand for?*
Electroretinogram (or electroretinography)

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

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How does a mfERG accomplish this?

Congenital/Stationary Retinal Disease

Disease

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***mfERG*: Produces a topographic map of central cone function**
pERG: Flashes a checkerboard pattern of rapidly alternating light and dark areas

How does a mfERG accomplish this?
Instead of flashing the entire retina, mfERG flashes are limited to small, hexagon-shaped areas of the macula. By divvying the macula up into hexagons and systematically testing each, mfERG can map out the functional status of the macula.

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- **Rod monochromatism**
- Blue-cone monochromatism

Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**

--No cones present—true color blindness

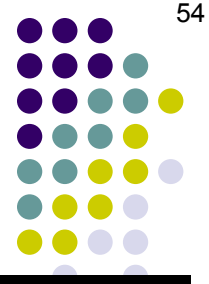
-- **Nystagmus** always present

--VA range: **20/80–20/200**

--ERG:

- --Cone response: ***?***
- --Rod response: ***?***

Next questions



Congenital/Stationary Retinal Disease

Cone (Color) Disease

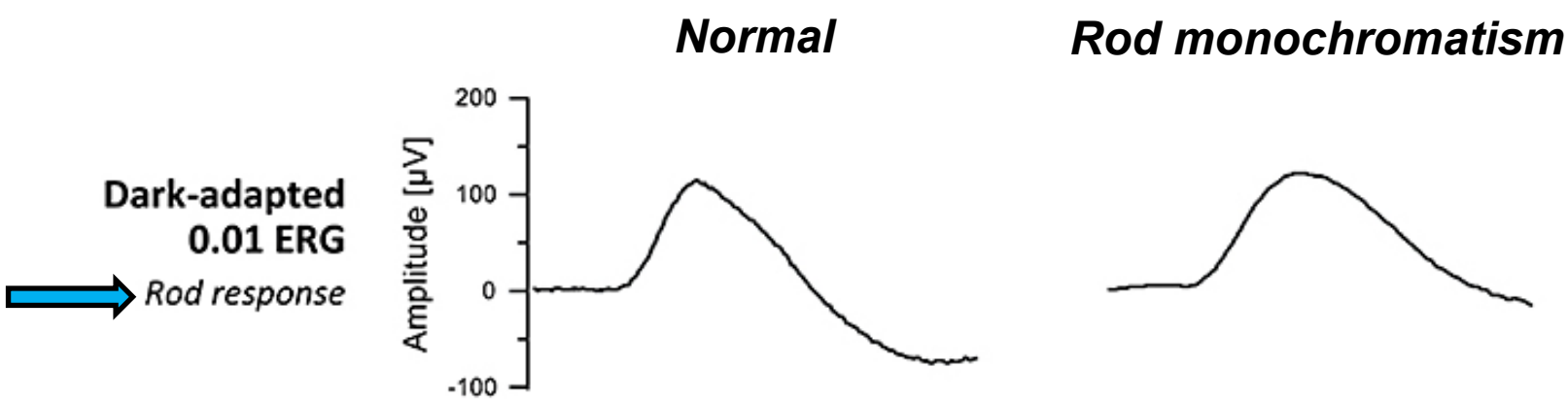
- Trichromatism
- Dichromatism
- Monochromatism
  - **Rod monochromatism**
  - Blue-cone monochromatism

Rod (Night Vision) Disease

Rod monochromatism

--Inheritance…**AR**
--No cones present—true color blindness
-- **Nystagmus** always present
--VA range: **20/80–20/200**
--ERG:
  --Cone response: **Absent**
  --Rod response: **Normal**

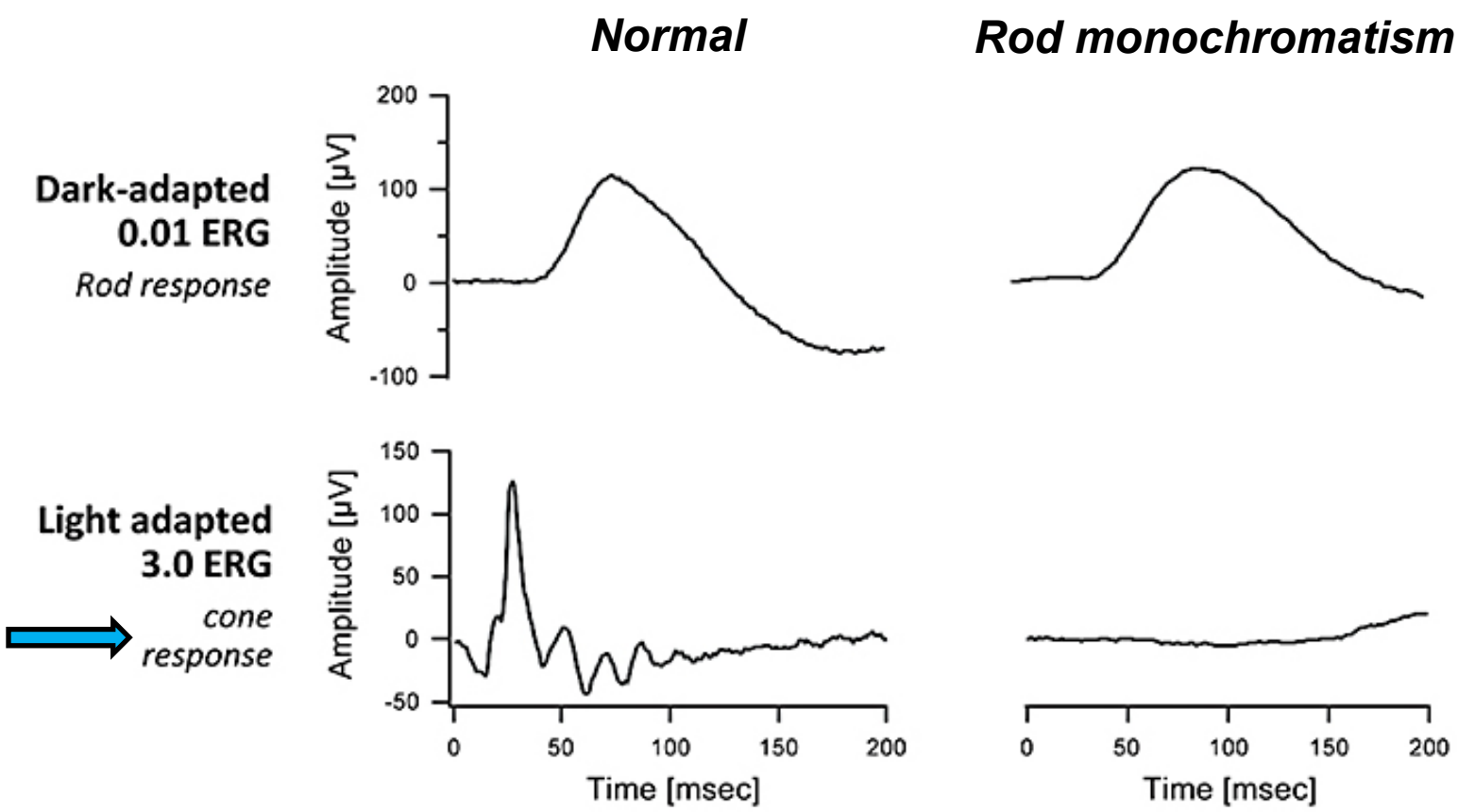
Congenital/Stationary Retinal Disease



In rod monochromatism, the rod response is (relatively) normal

Rod monochromatism: ERG

Congenital/Stationary Retinal Disease



However, the cone response is essentially nonexistent, as expected

Rod monochromatism: ERG

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

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Rod (Night Vision) Disease

Rod monochromatism

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-- **Nystagmus** always present

--VA range: **20/80–20/200**

--ERG:

--Cone response: **Absent**

--Rod response: **Normal**

Classic presentation of rod monochromatism:

-- two words *and*

-- *and*

--

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

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Rod (Night Vision) Disease

Rod monochromatism

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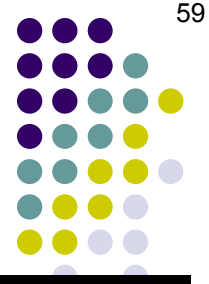
Classic presentation of rod monochromatism:

-- Poor acuity *and*

-- Nystagmus *and*

-- Photophobia



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: ***?***

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism
--Inheritance: **X-linked**

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
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  - Rod monochromatism
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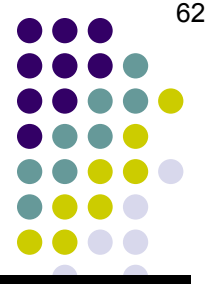
Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only duh cones present



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
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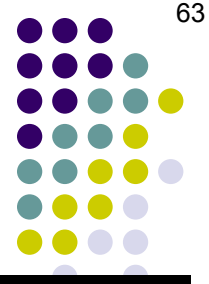
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present



Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Rod monochromatism

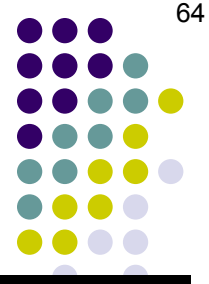
Blue-cone monochromatism

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…***?***



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

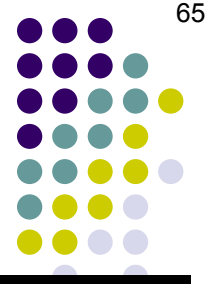
Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…**20/80**



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…20/80

Why is VA better than in many rod monochromats?

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- Rod monochromatism
- **Blue-cone monochromatism**

Rod (Night Vision) Disease

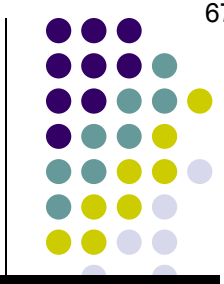
Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…20/80

Why is VA better than in many rod monochromats?
Because all blue-cone monochromats have a set of functioning cones (specifically, the blue ones)



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

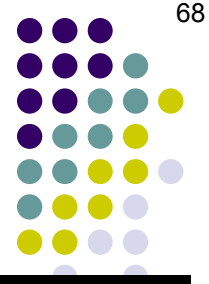
Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…**20/80**

--Diagnose via specialized…***?***



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…**20/80**

--Diagnose via specialized…**color ERG**

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- Rod monochromatism
- **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

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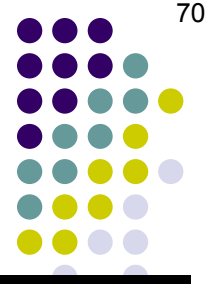
0

d...**color ERG**

What are the findings of color-ERG testing in blue-cone monochromatism?



Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

0

d…**color ERG**

What are the findings of color-ERG testing in blue-cone monochromatism?

Perhaps unsurprisingly, findings include a normal blue-cone response along with absent or greatly attenuated green- and red-cone responses

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

- Rod monochromatism
- **Blue-cone monochromatism**

Rod (Night Vision) Disease

Blue-cone monochromatism

--Inheritance: **X-linked**

--Only **blue** cones present

--VA usually about…**20/80**

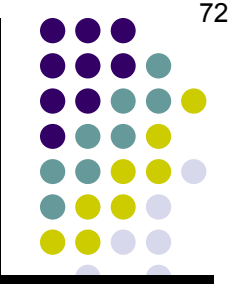
--Diagnose via specialized…**color ERG**

Classic presentation of blue-cone monochromatism:

-- two words *and*

-- *and*

--



Congenital/Stationary Retinal Disease

Cone (Color) Disease

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- Dichromatism
- Monochromatism
  - Rod monochromatism
  - **Blue-cone monochromatism**

Rod (Night Vision) Disease

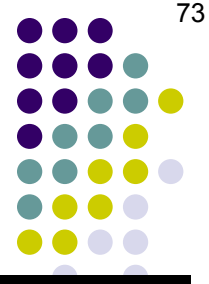
Blue-cone monochromatism

--Inheritance: **X-linked**
--Only **blue** cones present
--VA usually about…**20/80**
--Diagnose via specialized…**color ERG**

Classic presentation of blue-cone monochromatism:

-- Poor acuity *and*
-- Nystagmus *and*
-- Photophobia

(Yes, just like rod monochromatism)



Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Speaking of conditions that present very early in life with poor VA, nystagmus and photophobia…While there are many, the others that should come first to mind are what?

--Rod monochromatism

--Blue-cone monochromatism

--***?***

--***?***

--***?***

ochromatism

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s present

…**20/80**

cialized…**color ERG**

Classic presentation of blue-cone monochromatism:

-- **Poor acuity** ***and***

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Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Speaking of conditions that present very early in life with poor VA, nystagmus and photophobia…While there are many, the others that should come first to mind are what?

--Rod monochromatism
--Blue-cone monochromatism
--**Albinism**
--**Aniridia**
--**Leber's congenital amaurosis**

ochromatism
nked
s present
…**20/80**
ecialized…**color ERG**

Classic presentation of blue-cone monochromatism:

-- **Poor acuity** ***and***
-- **Nystagmus** ***and***
-- **Photophobia**

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
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  - Monochromatism
    - Rod monochromatism
    - Blue-cone monochromatism
- Rod (Night Vision) Disease

Blue-cone monochromatism
--Inheritance: **X-linked**
--Only **blue** cones present
--VA usually about…**20/80**
--Diagnose via specialized…**color ERG**

Classic presentation of
-- ~~Poor~~ Good acuity *and*
-- Nystagmus *and*
-- ~~Photophobia~~

If a pt has nystagmus plus **good** *vision, what condition does s/he most likely have?*

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
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Classic presentation of
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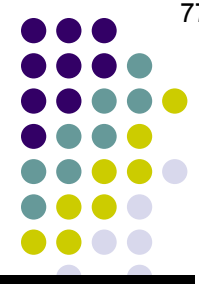
If a pt has nystagmus plus ***good*** *vision, what condition does s/he most likely have?*
Congenital motor nystagmus

Congenital/Stationary Retinal Disease



Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

- Rod monochromatism
- Blue-cone monochrom

Briefly, what is congenital motor nystagmus*?*

ERG

Classic presenta

-- ~~Poor~~ Good acuity *and*

-- Nystagmus *and*

-- ~~Photophobia~~

what condition does s/he most likely have?

Congenital motor nystagmus

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
    - Rod monochromatism
    - Blue-cone monochrom
- Rod (Night Vision) Disease

Briefly, what is congenital motor nystagmus*?*
A nystagmus arising in the first few months of life that is not secondary to either sensory or CNS pathology

ERG

Classic presenta
-- ~~Poor~~ Good acuity *and*
-- Nystagmus *and*
-- ~~Photophobia~~

what condition does s/he most likely have?
Congenital motor nystagmus

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - Blue-cone monochrom

Rod (Night Vision) Disease

Briefly, what is congenital motor nystagmus*?*
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Is the nystagmus vertical, horizontal or both/either?

ERG

Classic presenta

Good
-- ~~Poor~~ acuity *and*
-- Nystagmus *and*
-- ~~Photophobia~~

what condition does s/he most likely have?

Congenital motor nystagmus

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
    - Rod monochromatism
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Is the nystagmus vertical, horizontal or both/either?
It is virtually always

ERG

Classic presenta
-- ~~Poor~~ Good acuity *and*
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what condition does s/he most likely have?
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Congenital/Stationary Retinal Disease

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Congenital/Stationary Retinal Disease

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  - Trichromatism
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A nystagmus arising in the first few months of life that is not secondary to either sensory or CNS pathology

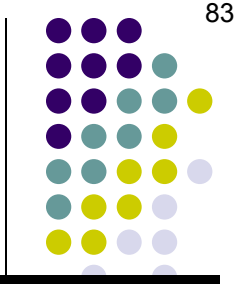
Is the nystagmus vertical, horizontal or both/either?
It is virtually always horizontal

Rule of thumb: If a pt has nystagmus + good VA, it's congenital motor nystagmus

ERG

Classic presenta
-- ~~Poor~~ Good acuity *and*
-- Nystagmus *and*
-- ~~Photophobia~~

what condition does s/he most likely have?
Congenital motor nystagmus



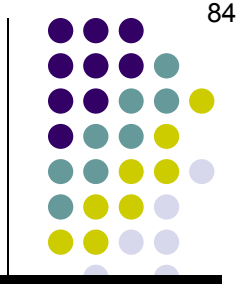
Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - aka... ~~Blue-cone~~ **monochromatism**

Rod (Night Vision) Disease

Finally: Note that blue-cone monochromatism is also known as monochromatism



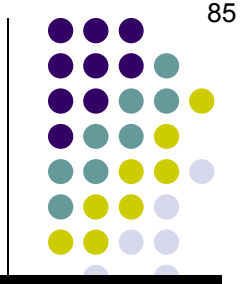
Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism
  - Rod monochromatism
  - ~~Blue-cone~~ S-cone **monochromatism**

Rod (Night Vision) Disease

Finally: Note that blue-cone monochromatism is also known as S-cone monochromatism



Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

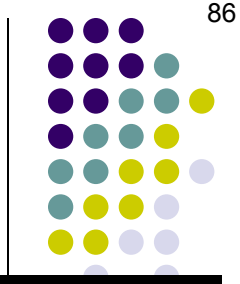
Dichromatism

Monochromatism

- Rod monochromatism
- ~~Blue-cone~~ S-cone **monochromatism**

Finally: Note that blue-cone monochromatism is also known as
S-cone monochromatism

Why is it aka S-cone monochromatism? What's the 'S' stand for?



Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Rod monochromatism

S-cone ~~Blue-cone~~ **monochromatism**

Finally: Note that blue-cone monochromatism is also known as
S-cone monochromatism

Why is it aka S-cone monochromatism? What's the 'S' *stand for?*
As noted earlier in the slide-set, blue light is of short wavelength, so blue cones are aka *short-wavelength cones*--S-cones for short (see what I did there?)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

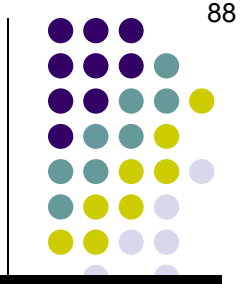
Monochromatism

Rod monochromatism

S-cone ~~Blue-cone~~ **monochromatism**

Finally: **Note that** blue-cone monochromatism is also known as **S-cone monochromatism**

Other than knowing that the condition goes by two names, is there another reason that an awareness of the name S-cone monochromatism *is noteworthy?*



Congenital/Stationary Retinal Disease

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Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Rod monochromatism

S-cone ~~Blue-cone~~ **monochromatism**

Finally: **Note that** blue-cone monochromatism is also known as **S-cone monochromatism**

Other than knowing that the condition goes by two names, is there another reason that an awareness of the name S-cone monochromatism *is noteworthy?*
Indeed there is, and it's this: To make certain not to confuse S-cone monochromatism with the similarly-named but completely different condition *enhanced S-cone syndrome*

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Speaking of conditions with two names…'Enhanced S-cone syndrome' is also known as what?

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In what ways are photoreceptors affected?

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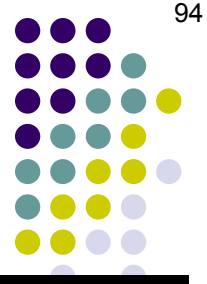

Cone (Color) Disease

Rod (Night Vision) Disease

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Dichromatism

In what ways are photoreceptors affected?
--Rods: ***?***

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--Blue cones: **Increased** in number

What are the ERG findings?
--Rod response: Undetectable
--Red/green cone response: Attenuated

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Rod (Night Vision) Disease

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--Rods: Non-functioning
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--Blue cones: *Increased* in number

What are the ERG findings?
--Rod response: Undetectable
--Red/green cone response: Attenuated
--Blue cones: **Enhanced** (hence the name of the syndrome)

wn as

monochromatism with the similarly-named but completely different
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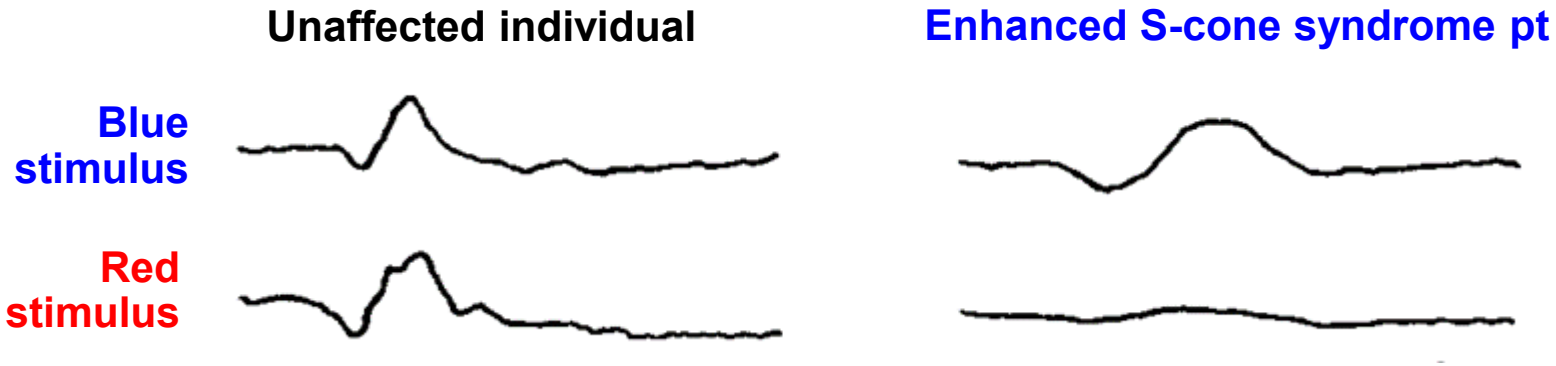
Congenital/Stationary Retinal Disease



Full-field ERG in response to color stimuli for an unaffected individual and a patient with **enhanced S-cone syndrome**. Note that in the patient, responses to blue stimuli are **larger** than that of the unaffected individual. Note further that the pt's response to the red stimulus is essentially nonexistent.

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

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Dichromatism

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How does it present?

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How does it present?
With decreased acuity as well as night blindness

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What is the appearance of enhanced S-cone syndrome on DFE?

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How does it present?
With decreased acuity as well as night blindness

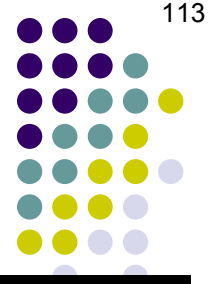
What is the appearance of enhanced S-cone syndrome on DFE?
Unlike the relatively normal appearance of the posterior pole in S-cone monochromatism, the posterior pole in enhanced S-cone syndrome is decidedly abnormal—retinoschisis as well as RP-like changes are the rule

nown as

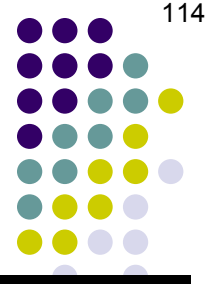
monochromatism with the similarly-named but completely different condition ***enhanced S-cone syndrome***

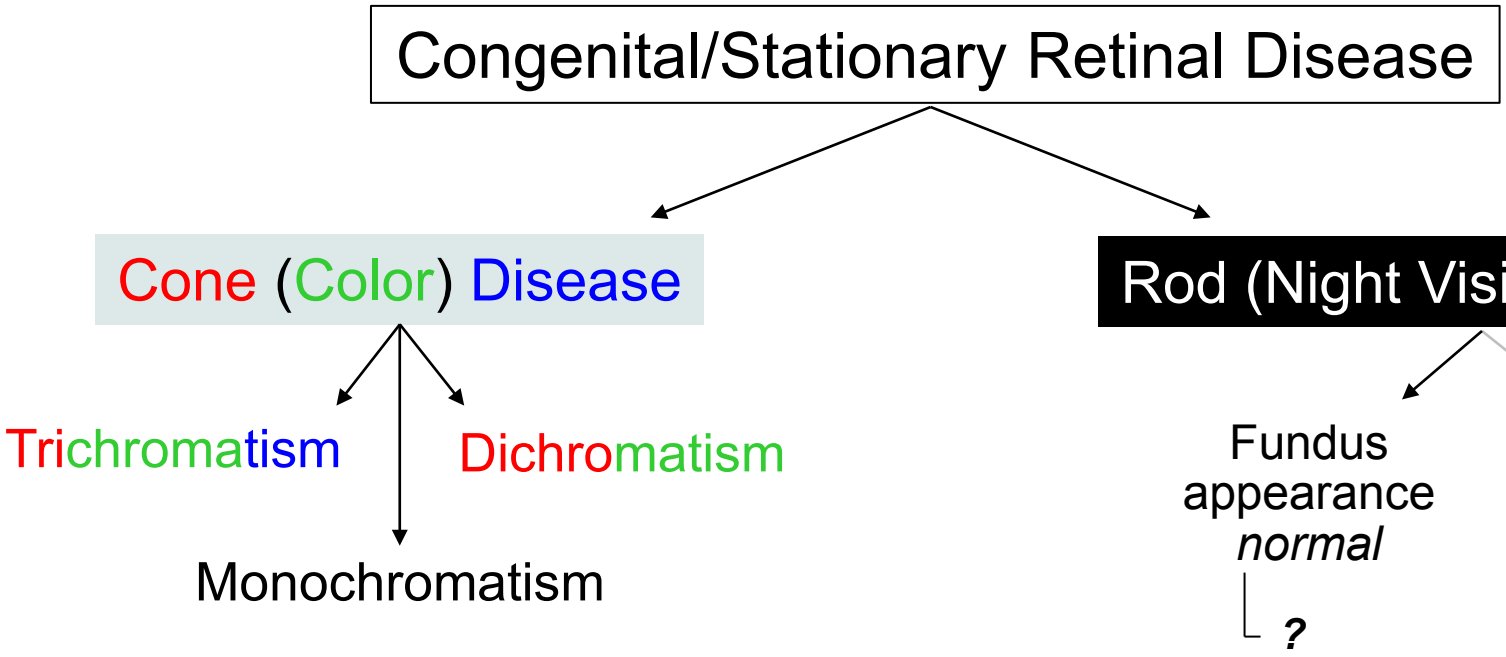
Congenital/Stationary Retinal Disease

Enhanced S-cone syndrome



Two general categories, not specific conditions




Congenital/Stationary Retinal Disease
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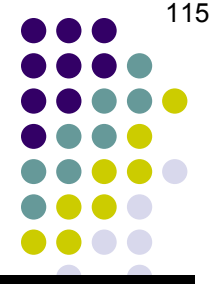
Trichromatism
Dichromatism
Monochromatism
Fundus appearance normal
Fundus appearance abnormal
?



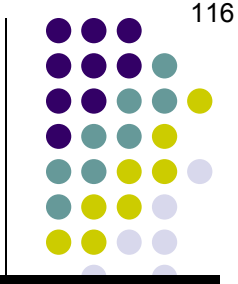
Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism

Rod (Night Vision) Disease

- Fundus appearance *normal*
  - CSNB
- Fundus appearance *abnormal*



Congenital/Stationary Retinal Disease

Cone (Color) Disease

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Rod (Night Vision) Disease

- Fundus appearance *normal*
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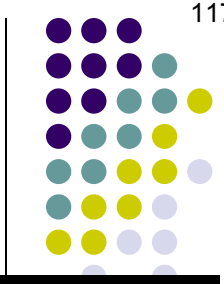
What does CNSB *stand for in this context?*



Congenital/Stationary Retinal Disease

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  - Monochromatism
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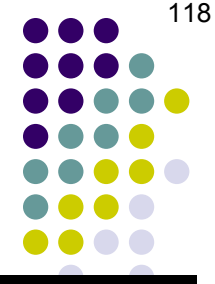
What does CNSB *stand for in this context?*
Congenital stationary night blindness



Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- Rod (Night Vision) Disease
  - Fundus appearance ***normal***
    - **CSNB**
  - Fundus appearance *abnormal*

Foreshadowing alert: We will soon see that while, strictly speaking, the *fundus* appears normal in CSNB, the *posterior pole* may not!

Congenital/Stationary Retinal Disease

Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism

Rod (Night Vision) Disease

- Fundus appearance *normal*
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Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common =

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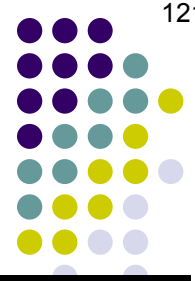
Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked



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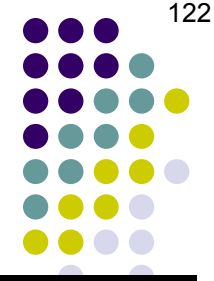
--Pathology: Communication failure between…

[two cell types]



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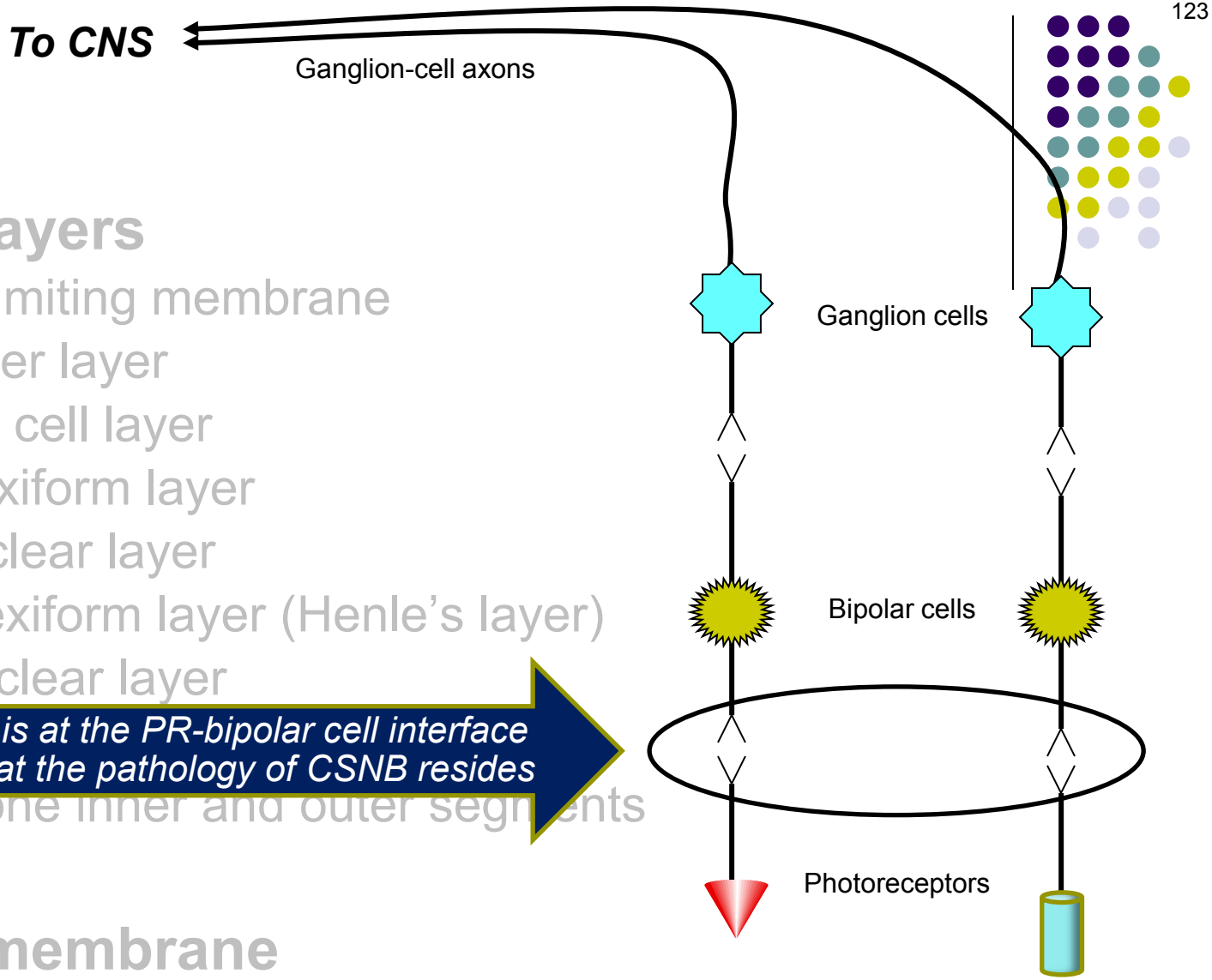
Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

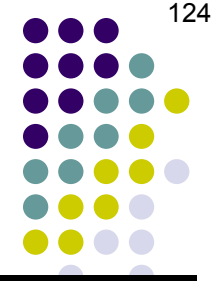
--Pathology: Communication failure between…
photoreceptors & bipolar cells

- **Retinal Layers**
  - Internal limiting membrane
  - Nerve fiber layer
  - Ganglion cell layer
  - Inner plexiform layer
  - Inner nuclear layer
  - Outer plexiform layer (Henle's layer)
  - Outer nuclear layer
  - Exter
  - Rod & cone inner and outer segments
- **RPE**
- **Bruch's membrane**

It is at the PR-bipolar cell interface that the pathology of CSNB resides



The ***photoreceptors***, ***bipolar*** and ***ganglion cells*** comprise the vertical retinal pathway—*vertical* in the sense that it is the direct path from photic stimulation to the CNS processing centers.



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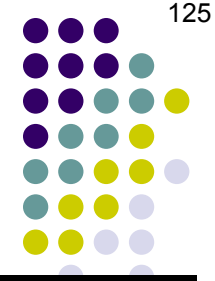
Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

--Pathology: Communication failure between…
photoreceptors & bipolar cells

--VA range: ***?***



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Cone (Color) Disease

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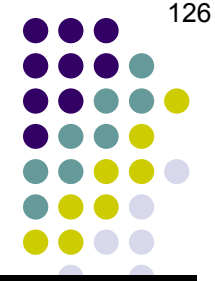
Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

--Pathology: Communication failure between…
photoreceptors & bipolar cells

--VA range: 20/20 - 20/200



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Congenital Stationary Night Blindness (CSNB)

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--VA range: 20/20 - 20/200

--Refractive error: Usually…***?***

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Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

--Pathology: Communication failure between…
photoreceptors & bipolar cells

--VA range: 20/20 - 20/200

--Refractive error: Usually…myopia

Congenital/Stationary Retinal Disease

Cone (Color) Disease

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Rod (Night Vision) Disease

- Fundus appearance *normal*
  - **CSNB**
- Fundus appearance *abnormal*

Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

--Pathology: Communication failure between… photoreceptors & bipolar cells

--VA range: 20/20 - **20/200**

--Refractive error: Usually…**myopia**

When VA is poor in CSNB, it's usually due to the (high) myopia, not the photoreceptors

Congenital/Stationary Retinal Disease

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- Rod (Night Vision) Disease
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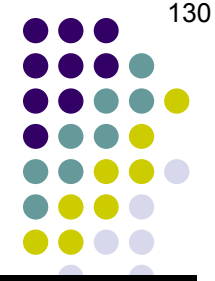
Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked
--Pathology: Communication failure between…
photoreceptors & bipolar cells
--VA range: 20/20 - 20/200
--Refractive error: Usually…myopia
--Presents in childhood with:
--***?***
--***?***
--***?***



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    - **CSNB**
  - Fundus appearance *abnormal*

Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked
--Pathology: Communication failure between…
photoreceptors & bipolar cells
--VA range: 20/20 - 20/200
--Refractive error: Usually…myopia
--Presents in childhood with:
--Nystagmus
--Decreased vision
--Nyctalopia

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

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--**Nyctalopia**

What is nyctalopia*?*

Congenital/Stationary Retinal Disease

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*Many CSNB children do **not** complain of nyctalopia. Why not?*

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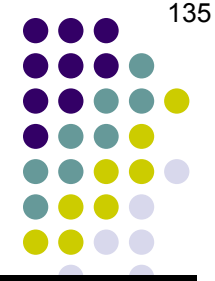
--**Nyctalopia**

What is nyctalopia*?*
Night blindness

*Many CSNB children do **not** complain of nyctalopia. Why not?*
As they have had extremely poor night vision their entire lives, it seems normal to them—they don't know any different



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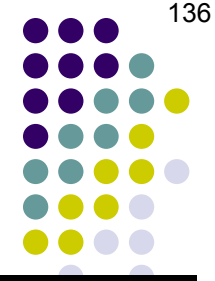
--Classified according to…*[Psychophysical test]*



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In other words, the *dark-adapted* ERG. (Remember, it's in the dark that CSNB pts have their difficulty.) Abnormalities of the photopic or light-adapted ERG also occur in CSNB, but are much more subtle.

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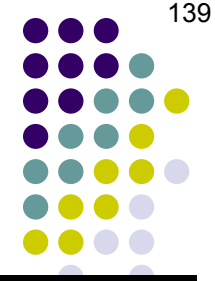

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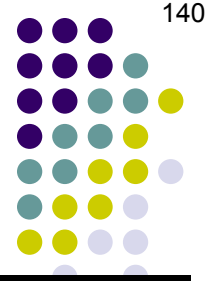
--Most common pattern: *Negative* ERG



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--*Negative* ERG =

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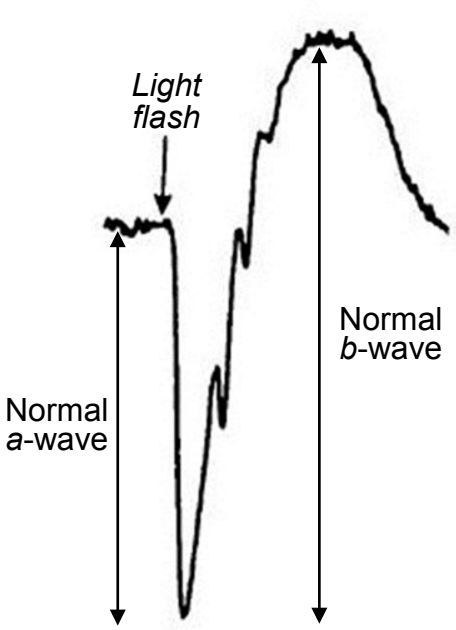
--Classified according to…Scotopic ERG pattern
--Most common pattern: *Negative* ERG
--*Negative* ERG = Large *a*-wave, smaller *b*-wave

Congenital/Stationary Retinal Disease

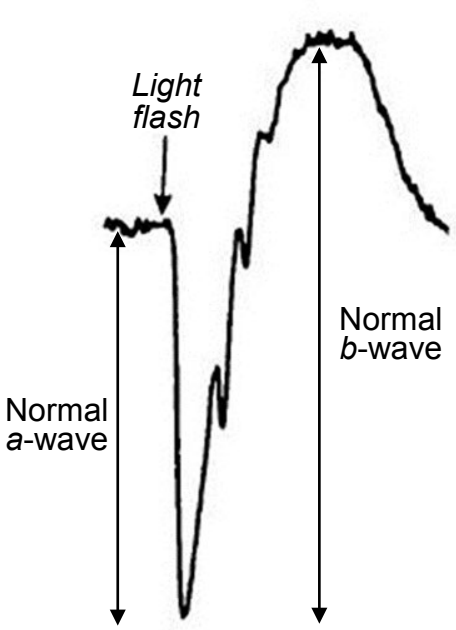
Normal



In a normal ERG, the *b*-wave is much larger than the *a*-wave

Congenital/Stationary Retinal Disease

Normal



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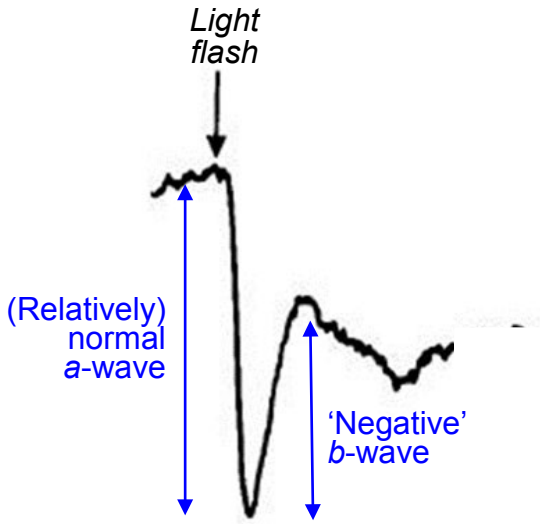
CSNB



The *b*-wave is said to be 'negative' when it is smaller than the *a*-wave, as is the case in CSNB

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

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Fundus appearance *abnormal*

Congenital Stationary Night Blindness (CSNB)

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Another condition—not common, but moreso than CSNB—also presents with a negative ERG. What is it?

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--Classified according to…Scotopic ERG pattern

--Most common pattern: *Negative* ERG

--*Negative* ERG = Large *a*-wave, smaller *b*-wave

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What does XLR *stand for in this context?*

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What does XLR *stand for in this context?*

X-linked retinoschisis

--Classified according to…Scotopic ERG pattern

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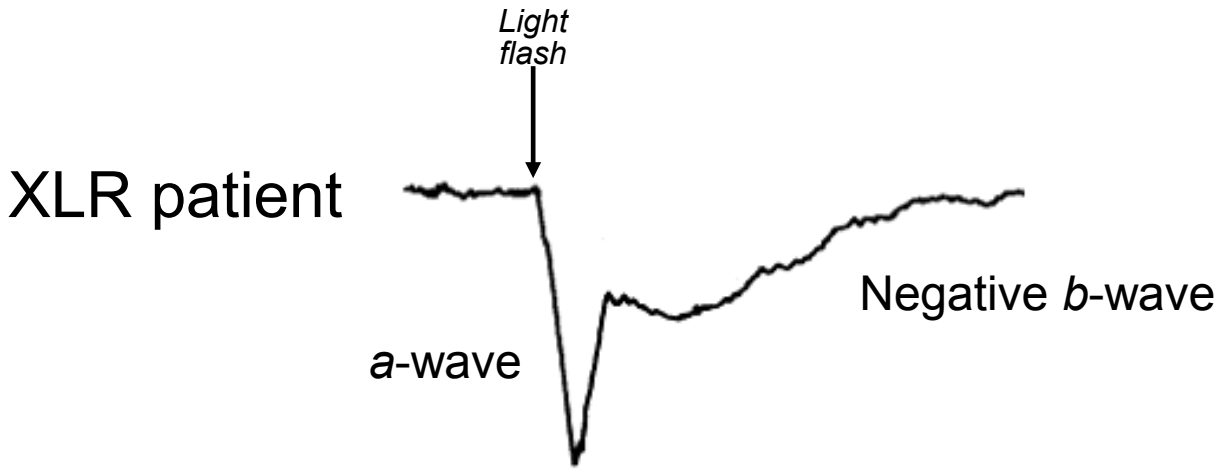
Congenital/Stationary Retinal Disease



X-linked retinoschisis: ERG

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

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XL

W

X-linked **retinoschisis**

Before we get any deeper—what does retinoschisis *refer to in this context?*

--Classified according to…Scotopic ERG pattern

--Most common pattern: *Negative* ERG

--*Negative* ERG = Large *a*-wave, smaller *b*-wave

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X-linked **retinoschisis**

Before we get any deeper—what does retinoschisis *refer to in this context?*

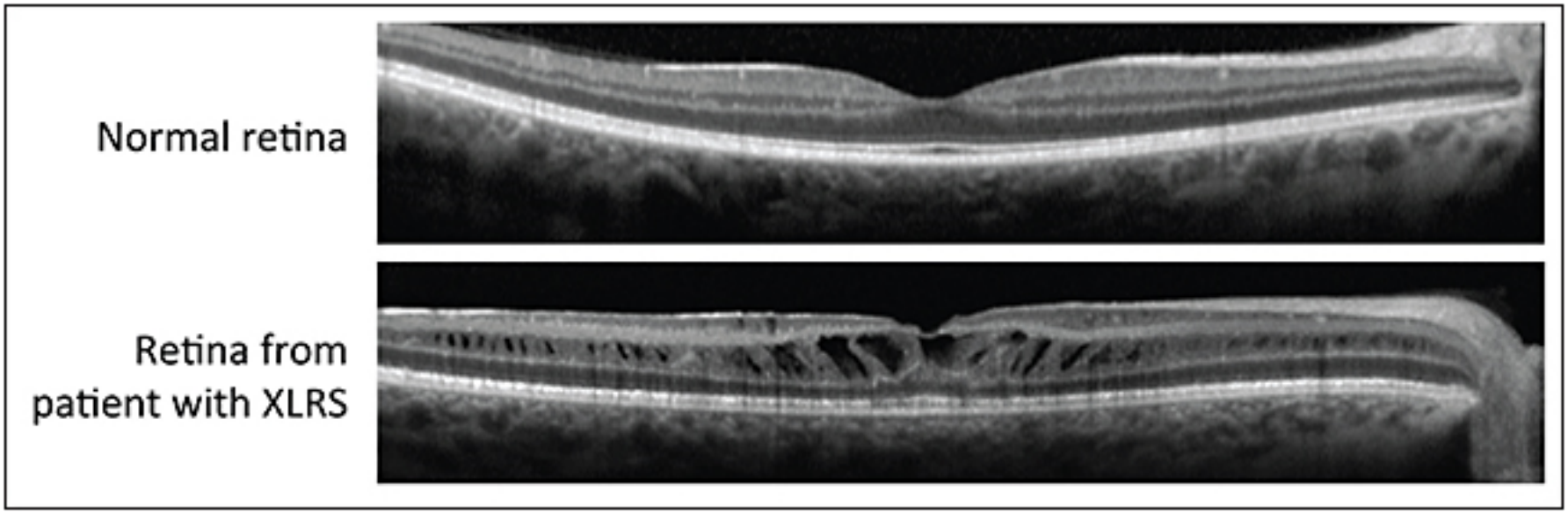
Splitting within the layers of the neurosensory retina

--Classified according to…Scotopic ERG pattern

--Most common pattern: *Negative* ERG

--*Negative* ERG = Large *a*-wave, smaller *b*-wave

Congenital/Stationary Retinal Disease



X-linked retinoschisis

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

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Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

By what other (very similar) name is X-linked retinoschisis (XLR) known?

What does XLR stand for in this context?

X-linked retinoschisis

--Nyctalopia

--Classified according to…Scotopic ERG pattern

--Most common pattern: *Negative* ERG

--*Negative* ERG = Large *a*-wave, smaller *b*-wave

Congenital/Stationary Retinal Disease

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Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

By what other (very similar) name is X-linked retinoschisis (XLR) known?
X-linked juvenile retinoschisis (XLJR)

What does XLR stand for in this context?
X-linked retinoschisis

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--Classified according to…Scotopic ERG pattern

--Most common pattern: *Negative* ERG

--*Negative* ERG = Large *a*-wave, smaller *b*-wave

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Fundus appearance *normal*

Fundus appearance *abnormal*

By what other (very similar) name is X-linked retinoschisis (XLR) known?

X-linked **juvenile** retinoschisis (XL**J**R)

What is implied—correctly—by the word juvenile *above?*

What does XLR stand for in this context?

X-linked retinoschisis

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What is implied—correctly—by the word juvenile *above?*

That the condition manifests early in life (in fact, it is [redacted])

What does XLR stand for in this context?

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X-linked juvenile retinoschisis (XLJR)

How does it present on DFE?

What does XLR stand for in this context?
X-linked retinoschisis

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By what other (very similar) name is X-linked retinoschisis (XLR) known?
X-linked juvenile retinoschisis (XLJR)

How does it present on DFE?
With macular schisis in a [redacted] pattern, +/- peripheral schisis

What does XLR stand for in this context?
X-linked retinoschisis

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How does it present on DFE?
With macular schisis in a radial pattern, +/- peripheral schisis

What does XLR stand for in this context?
X-linked retinoschisis

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What layer(s) of the retina are involved in the schisis?

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Mainly the [abb.], but the [abb.] can be involved as well

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What does XLR stand for i

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All of them, essentially

What does XLR stand for i

X-linked retinoschisis

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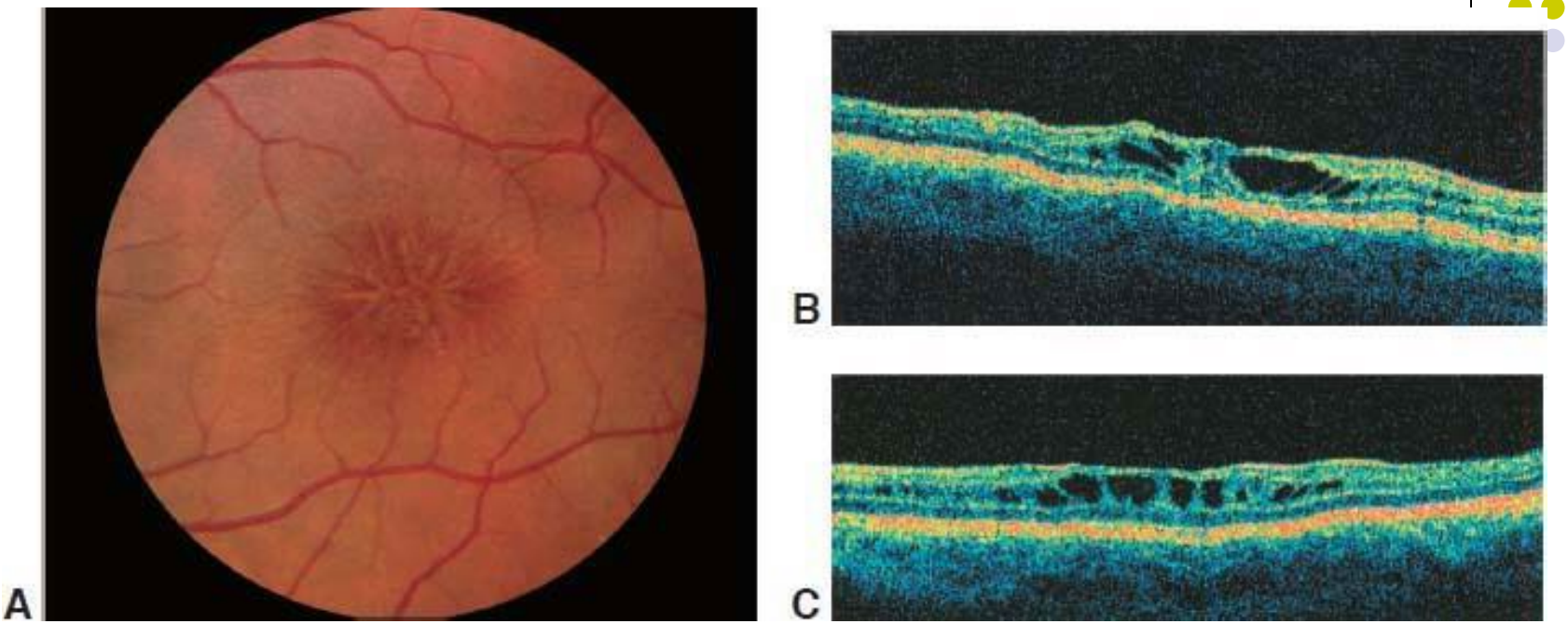
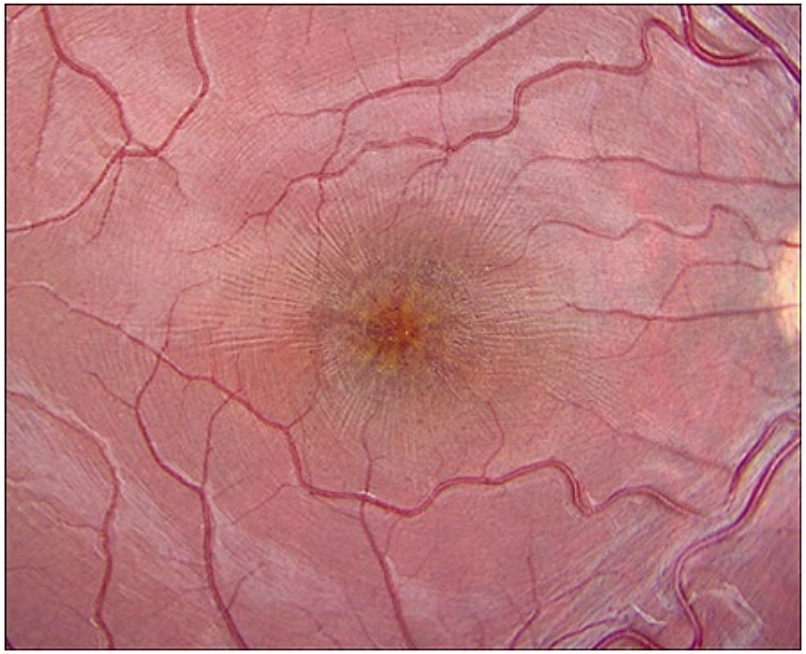
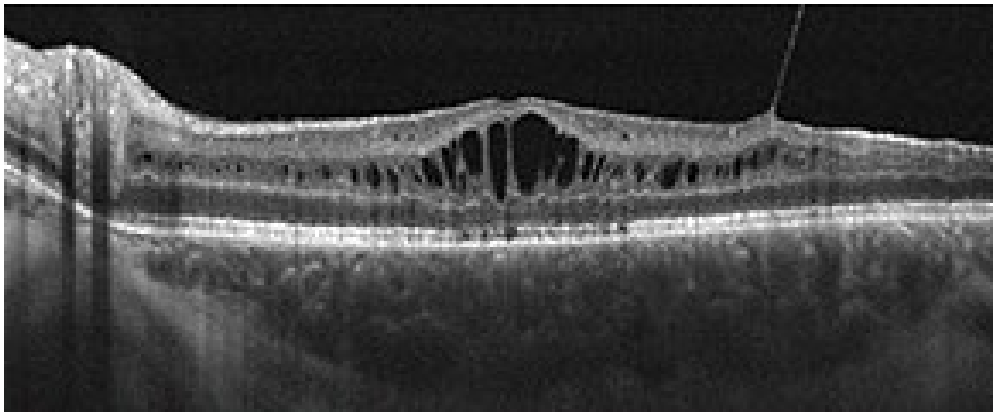


A, Color fundus photograph shows the characteristic pattern of macular schisis, a more consistent finding than peripheral changes. Vertical (B) and horizontal (C) OCT scans demonstrate schisis spaces in the middle layers of the macula.

X-linked retinoschisis

Congenital/Stationary Retinal Disease





X-linked juvenile retinoschisis: Foveal cysts

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Monochromatism

Fundus appearance *normal*

Fundus appearance *abnormal*

By what other (very similar) name is X-linked retinoschisis (XLR) known?
X-linked juvenile retinoschisis (XLJR)

How does it present on DFE?
With macular schisis in a radial pattern, +/- peripheral schisis

How does it present clinically?

What does XLR stand for in this context?
X-linked retinoschisis

--Nyctalopia
--Classified according to…Scotopic ERG pattern
--Most common pattern: *Negative* ERG
--*Negative* ERG = Large *a*-wave, smaller *b*-wave

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With macular schisis in a radial pattern, +/- peripheral schisis

How does it present clinically?
With modestly v severely decreased VA in life stage.

What does XLR stand for in this context?
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With macular schisis in a radial pattern, +/- peripheral schisis

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With modestly decreased VA in childhood. Over time, VA will drop to Snellen or so.

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X-linked retinoschisis

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How does it present clinically?
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Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

Another condition presents with c/o night blindness, and ERG reveals patterns identical to those of CSNB. Thus, this condition is high on the DDx for CSNB. What is it?

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Melanoma-associated retinopathy

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What is melanoma-associated retinopathy?

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What is melanoma-associated retinopathy?

A paraneoplastic process in which retinal cells display antigens that are identical to, or cross-react with, melanoma cells within the body

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In what very important way does the night blindness of MAR differ from that of CSNB?

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In addition to night blindness, there is another complaint that is classic for MAR. What is it?

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Photopsias, often described as 'shimmering'

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What other psychophysical test is always abnormal in CSNB?

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What other psychophysical test is always abnormal in CSNB?

Dark adaptometry

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-- *What other psychophysical test is always abnormal in CSNB?*
Dark adaptometry

-- *What does dark adaptometry assess?*

--

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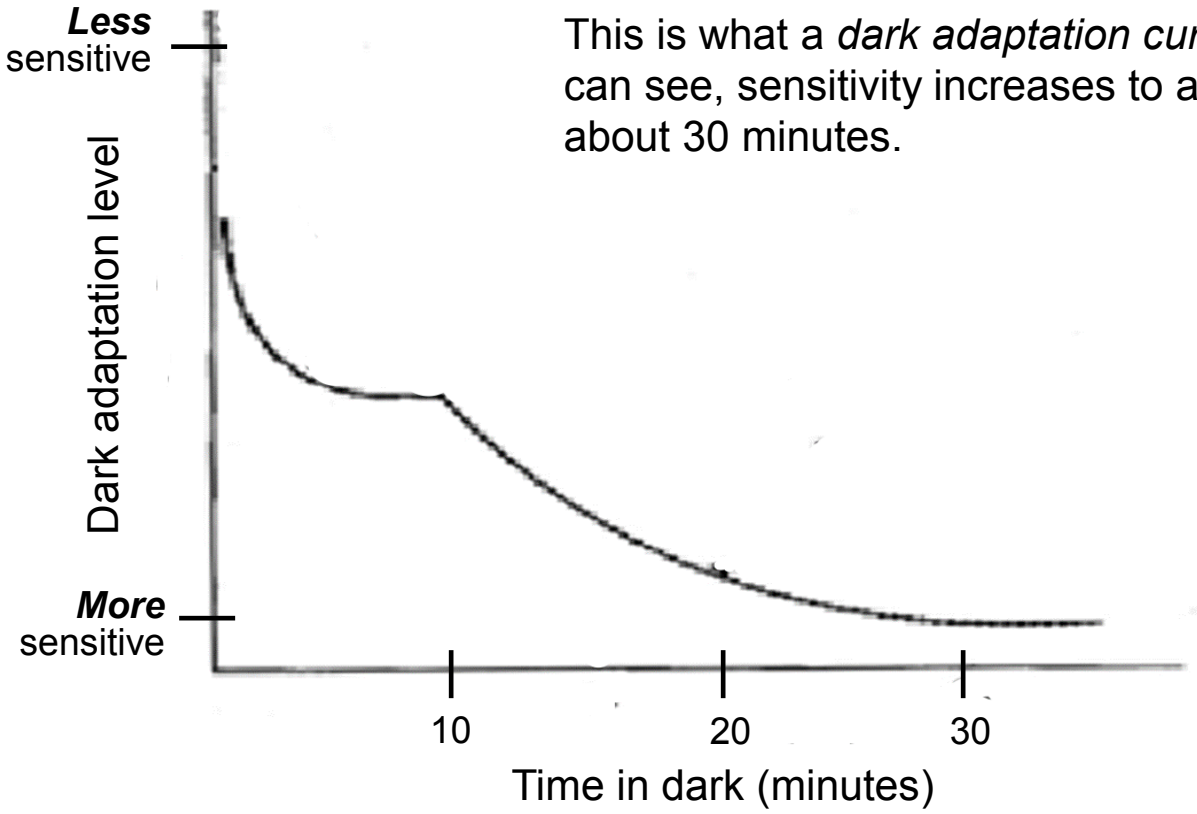
Dark adaptometry

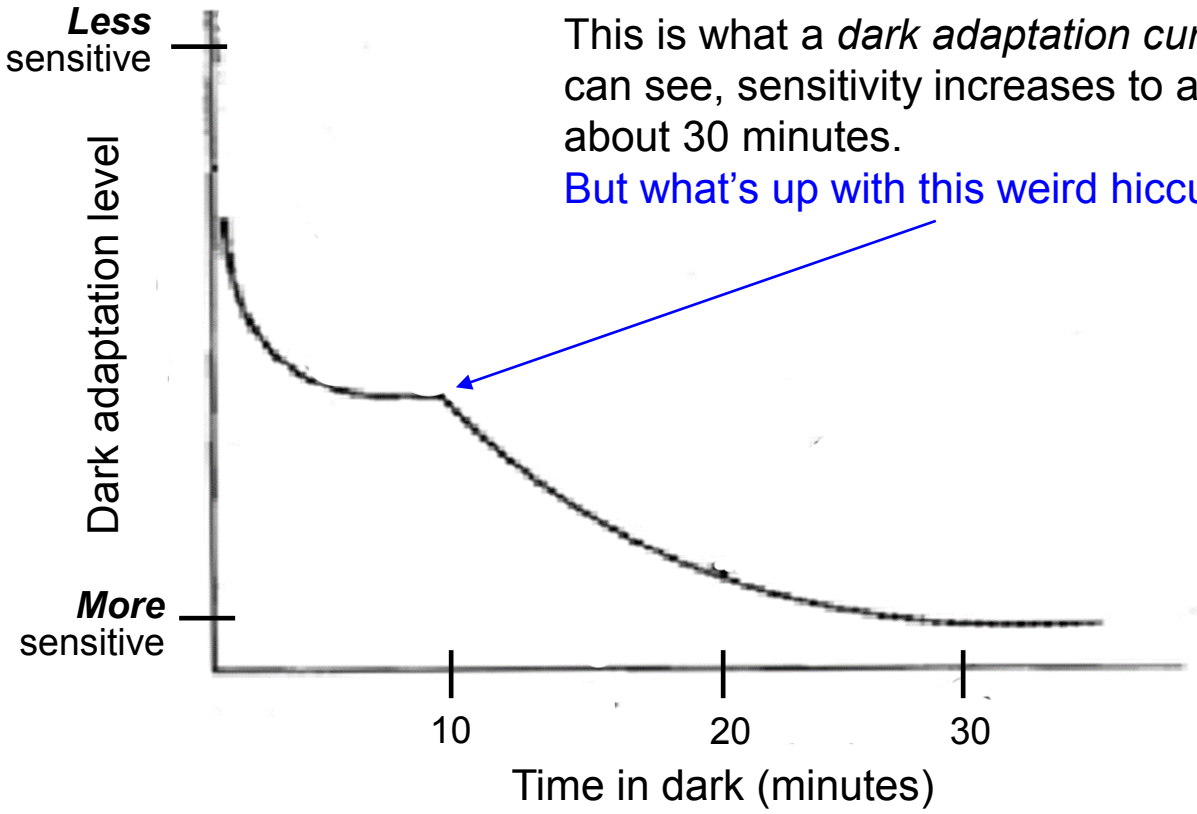
What does dark adaptometry assess?
The increase in sensitivity that occurs when the background illumination is low. That is to say, the longer an eye is in the dark, the dimmer the light it can perceive (up to a point).

Congenital/Stationary Retinal Disease



This is what a *dark adaptation curve* looks like. As you can see, sensitivity increases to a maximum after about 30 minutes.

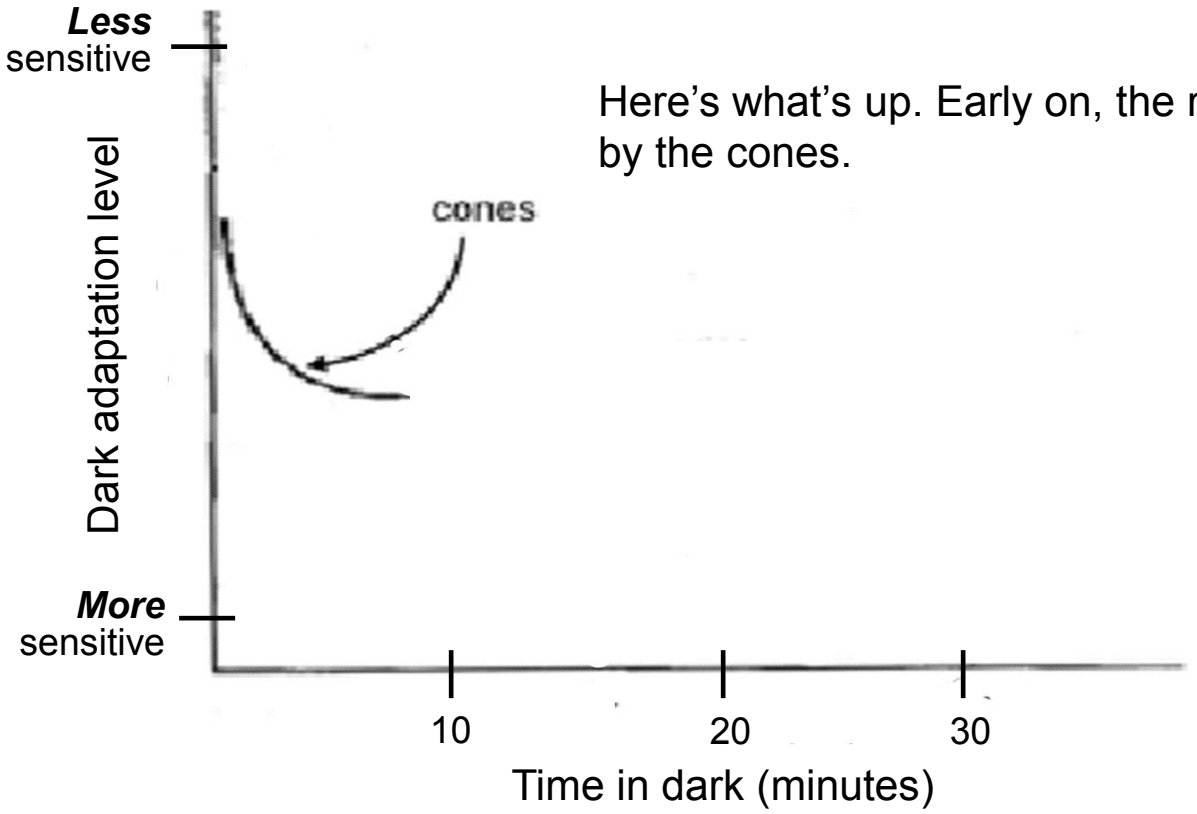
Congenital/Stationary Retinal Disease



This is what a *dark adaptation curve* looks like. As you can see, sensitivity increases to a maximum after about 30 minutes.

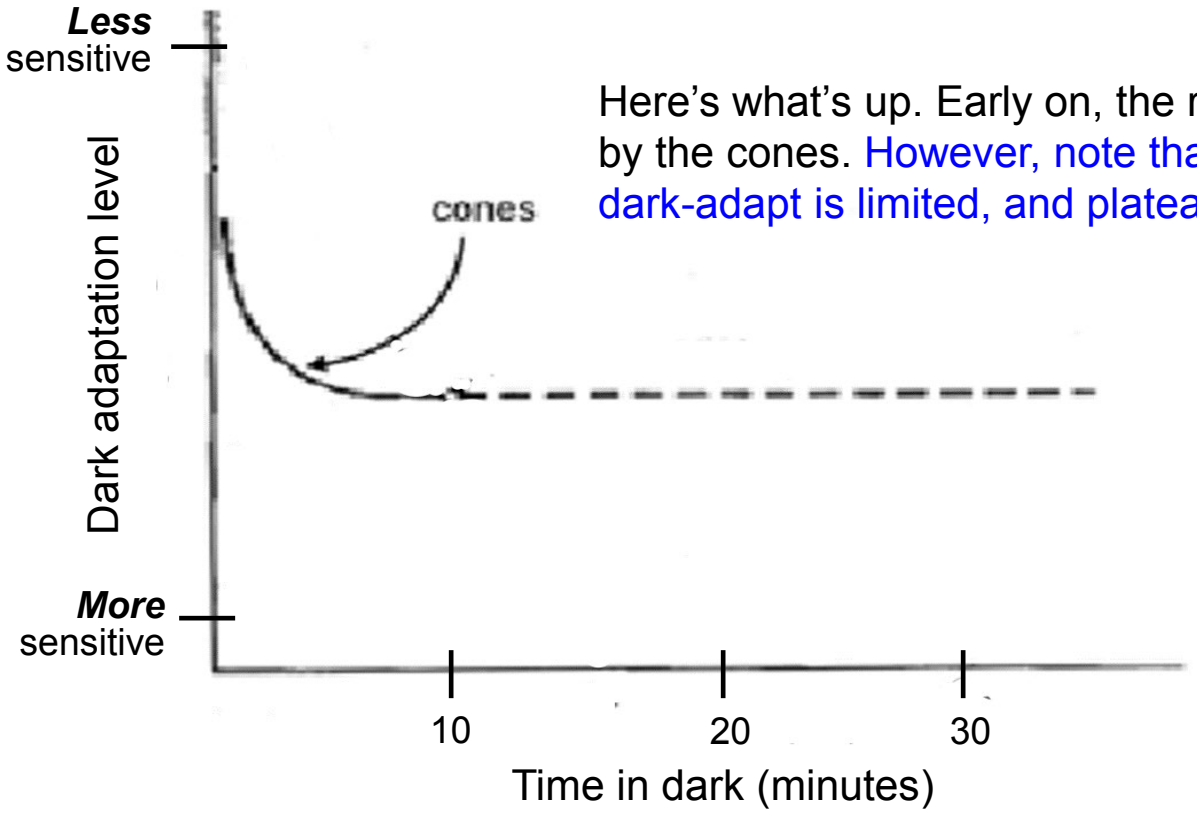
But what's up with this weird hiccup in the curve?

Congenital/Stationary Retinal Disease



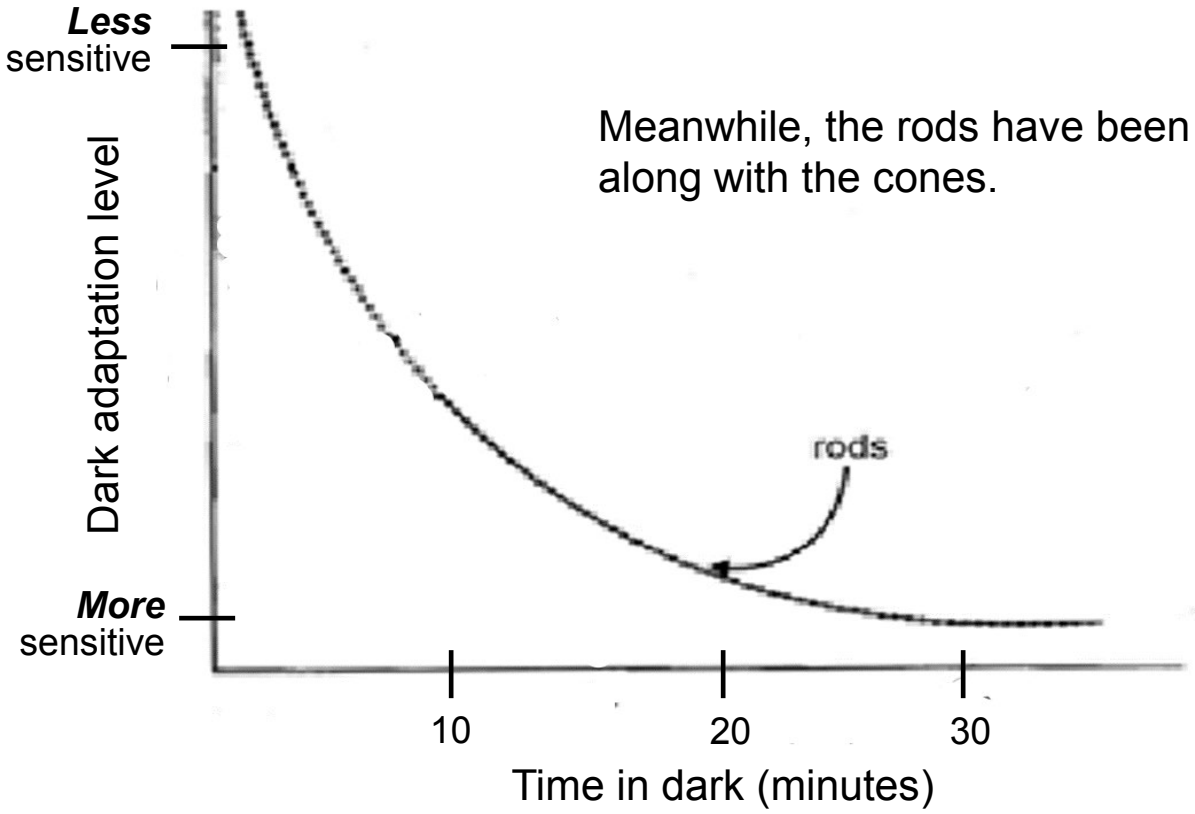
Here's what's up. Early on, the response is dominated by the cones.

Congenital/Stationary Retinal Disease



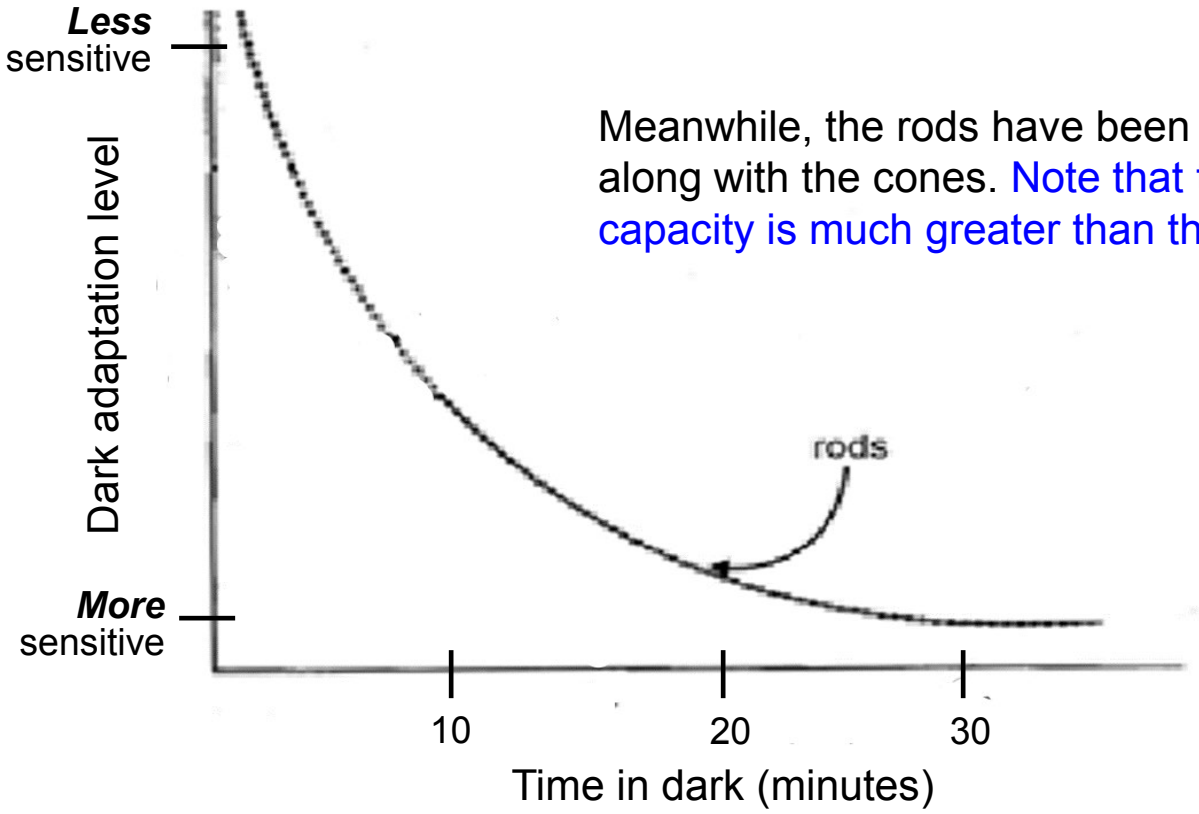
Here's what's up. Early on, the response is dominated by the cones. However, note that the cones' ability to dark-adapt is limited, and plateaus after ~10 minutes.

Congenital/Stationary Retinal Disease



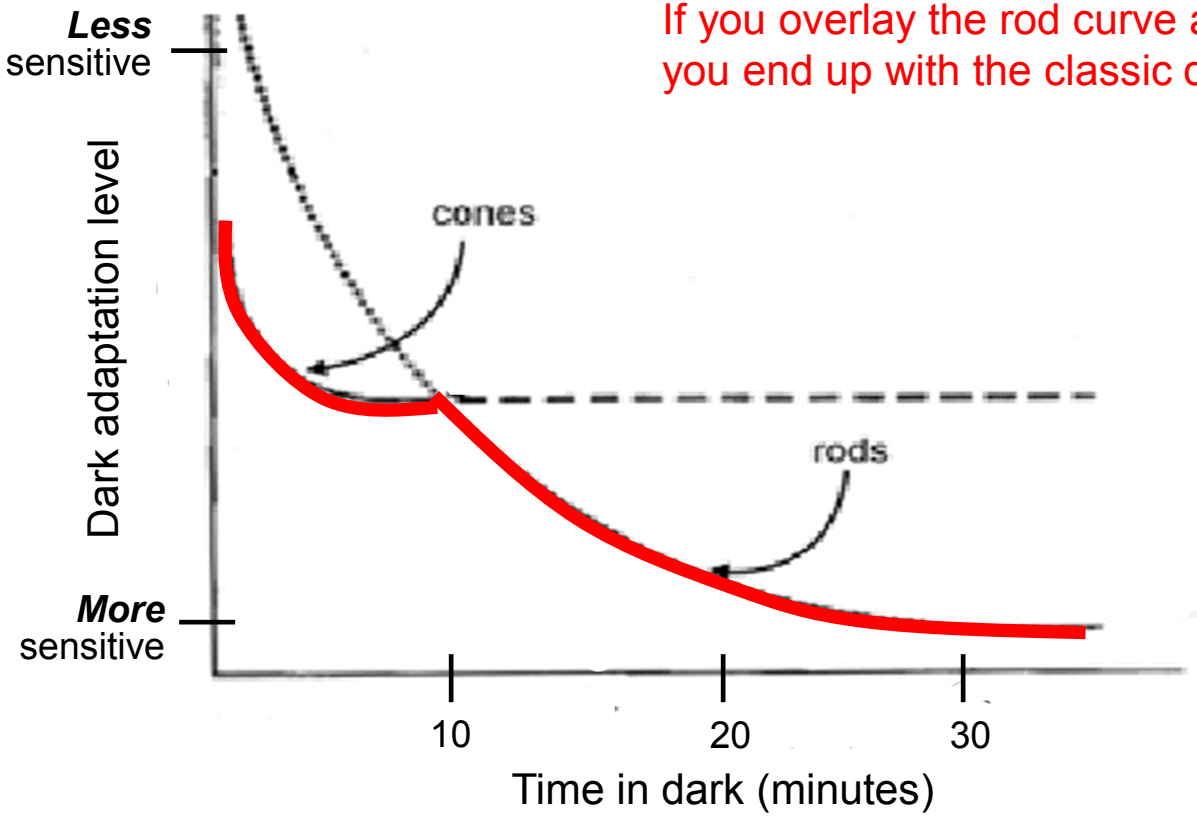
Meanwhile, the rods have been quietly dark-adapting along with the cones.

Congenital/Stationary Retinal Disease



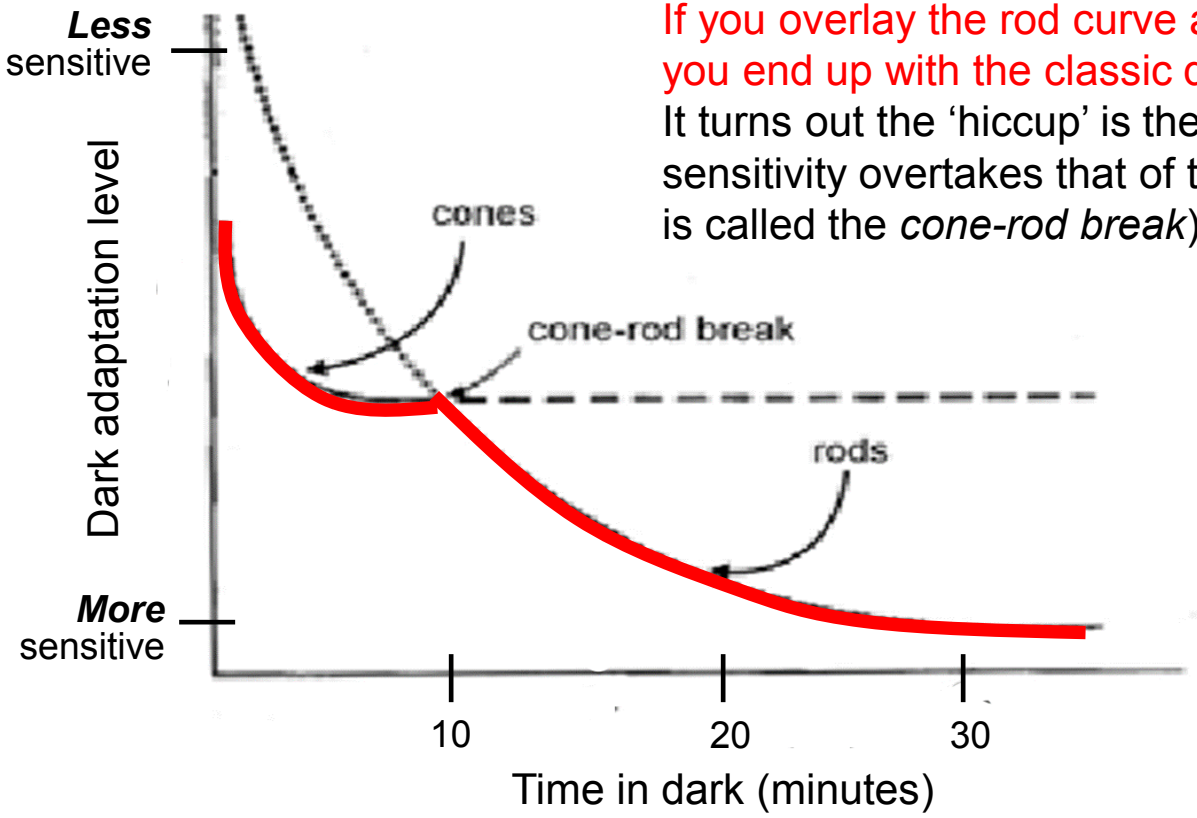
Meanwhile, the rods have been quietly dark-adapting along with the cones. Note that their dark-adaptation capacity is much greater than that of the cones.

Congenital/Stationary Retinal Disease



If you overlay the rod curve atop the cone curve, you end up with the classic dark-adaptation curve.

Congenital/Stationary Retinal Disease



If you overlay the rod curve atop the cone curve, you end up with the classic dark-adaptation curve. It turns out the 'hiccup' is the point where rod sensitivity overtakes that of the cones (and hence is called the *cone-rod break*).

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

CSNB

Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

-- *What other psychophysical test is always abnormal in CSNB?*
Dark adaptometry

-- *What does dark adaptometry assess?*
The increase in sensitivity that occurs when the background illumination is low. That is to say, the longer an eye is in the dark, the dimmer the light it can perceive (up to a point).

-- *In what way is dark adaptometry abnormal in CSNB?*

Next question

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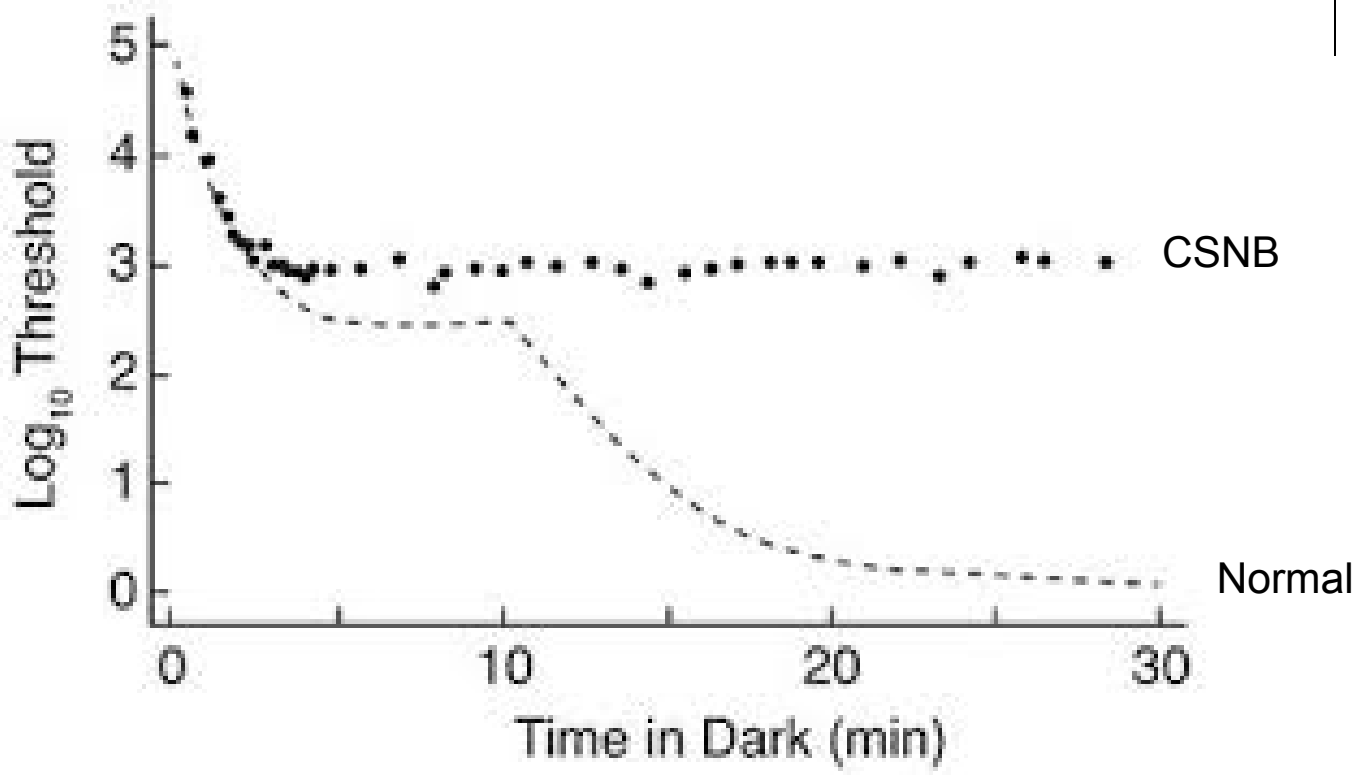
-- *What does dark adaptometry assess?*

-- The increase in sensitivity that occurs when the background illumination is low. That is to say, the longer an eye is in the dark, the dimmer the light it can perceive (up to a point).

-- *In what way is dark adaptometry abnormal in CSNB?*
Due to the lack of functioning rods, the cone-rod break never kicks in—adaptation remains at the cone maximum, with the result being poor vision under very dim conditions

Congenital/Stationary Retinal Disease



Dark-adaptometry curve in CSNB (filled circles). Note the lack of rod adaptation (ie, it looks just like the *cones only* graph a few slides back).

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--Several inheritance patterns; most common = X-linked

--Pathology: Communication failure between…
photoreceptors & bipolar cells

--VA range: 20/20 - 20/200

--Refractive error: Usually…myopia

Recall we said earlier that the posterior pole exam may not be normal.

(No question yet—keep going)

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What structure in particular may be abnormal in appearance?

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What structure in particular may be abnormal in appearance?

The optic nerve head

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Recall we said earlier that the posterior pole exam may not be normal.
What structure in particular may be abnormal in appearance?
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In what manner is the ONH likely to be abnormal in CSNB?

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It may manifest a [two words]

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Recall we said earlier that the posterior pole exam may not be normal.
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In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic tilt

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Recall we said earlier that the posterior pole exam may not be normal.

What structure in particular may be abnormal in appearance?

The optic nerve head

If you can remember one of these facts, it should help you remember the other

In what manner is the ONH likely to be abnormal in CSNB?

It may manifest a **myopic tilt**

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- Rod (Night Vision) Disease
  - Fundus appearance *normal*
    - **CSNB**
  - Fundus appearance *abnormal*

Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked
--Pathology: Communication failure between…
photoreceptors & bipolar cells
--VA range: 20/20 - 20/200
--Refractive error: Usually…myopia

Recall we said earlier that the posterior pole exam may not be normal.
What structure in particular may be abnormal in appearance?
The optic nerve head

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic tilt , and its nasal vs temporal aspect may be pallorous

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

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In what manner is the ONH likely to be abnormal in CSNB?

It may manifest a myopic tilt , and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
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In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic tilt , and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

CSNB

Congenital Stationary Night Blindness (CSNB)

--Several inheritance patterns; most common = X-linked

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It may manifest a myopic tilt , and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?

Fuchs coloboma (Dr Fuchs strikes again—I stan an eye king!)

Ernst Fuchs
1851-1930

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
    - **CSNB**
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its inferior vs superior pole is elevated and its inferior vs superior pole is posteriorly displaced (ie, staphyloma-like)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Tilted disc

Congenital/Stationary Retinal Disease

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  - Trichromatism
  - Dichromatism
  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

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  - Dichromatism
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Congenital Stationary Night Blindness (CSNB)
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What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Huh? I thought situs inversus meant all the organs were on the wrong side of the body, or something. What does it mean in this context?

It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Huh? I thought situs inversus meant all the organs were on the wrong side of the body, or something. What does it mean in this context?
It means the nasal v temp vessels run direction for a short interval before heading off in the right direction

It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

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  - Monochromatism
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Huh? I thought situs inversus meant all the organs were on the wrong side of the body, or something. What does it mean in this context?
It means the temporal vessels run nasally for a short interval before heading off in the right direction

It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Tilted disc: Situs inversus

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
    - **CSNB**
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Does any of this impact vision?

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

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--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a three words

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
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--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
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  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a description defect).

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
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  - Dichromatism
  - Monochromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
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Congenital Stationary Night Blindness (CSNB)
--Several inheritance patterns; most common = X-linked

What does it mean to say the ONH is 'tilted'?
It means its superior pole is elevated and its inferior pole is posteriorly displaced (ie, staphyloma-like)

The vessels on a tilted disc may run in an unusual pattern. What is the two-word name for this pattern?
'Situs inversus'

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a bitemporal defect)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
  - Fundus appearance *abnormal*

How on earth does a tilted disc produce a bitemporal VF defect?

Does any of this impact vision?

It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?

It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?

Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

How on earth does a tilted disc produce a bitemporal VF defect?
It's actually pretty straightforward.
Recall that the area including and adjacent to the inferior pole of the ONH is staphylomatous. This means the 'axial length' of the photoreceptors within this region is greater than that of the rest of the posterior pole. Because of this extra axial length, the correction used during VF testing (which is based on the refraction of the non-staphylomatous fovea) is not myopic enough for the inferior peripapillary region.

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

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Dichromatism

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Recall that the area including and adjacent to the inferior pole of the ONH is staphylomatous. This means the 'axial length' of the photoreceptors within this region is greater than that of the rest of the posterior pole. Because of this extra axial length, the correction used during VF testing (which is based on the refraction of the non-staphylomatous fovea) is not myopic enough for the inferior peripapillary region. Because this region is out of focus, it will manifest a *refractive scotoma* on the test. And because the retina involved in this scotoma is **inferonasal** to the fovea, it follows that the resulting VF defect will be **superotemporal** to fixation.

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
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Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

Trichromatism

Dichromatism

Fundus appearance *normal*

Fundus appearance *abnormal*

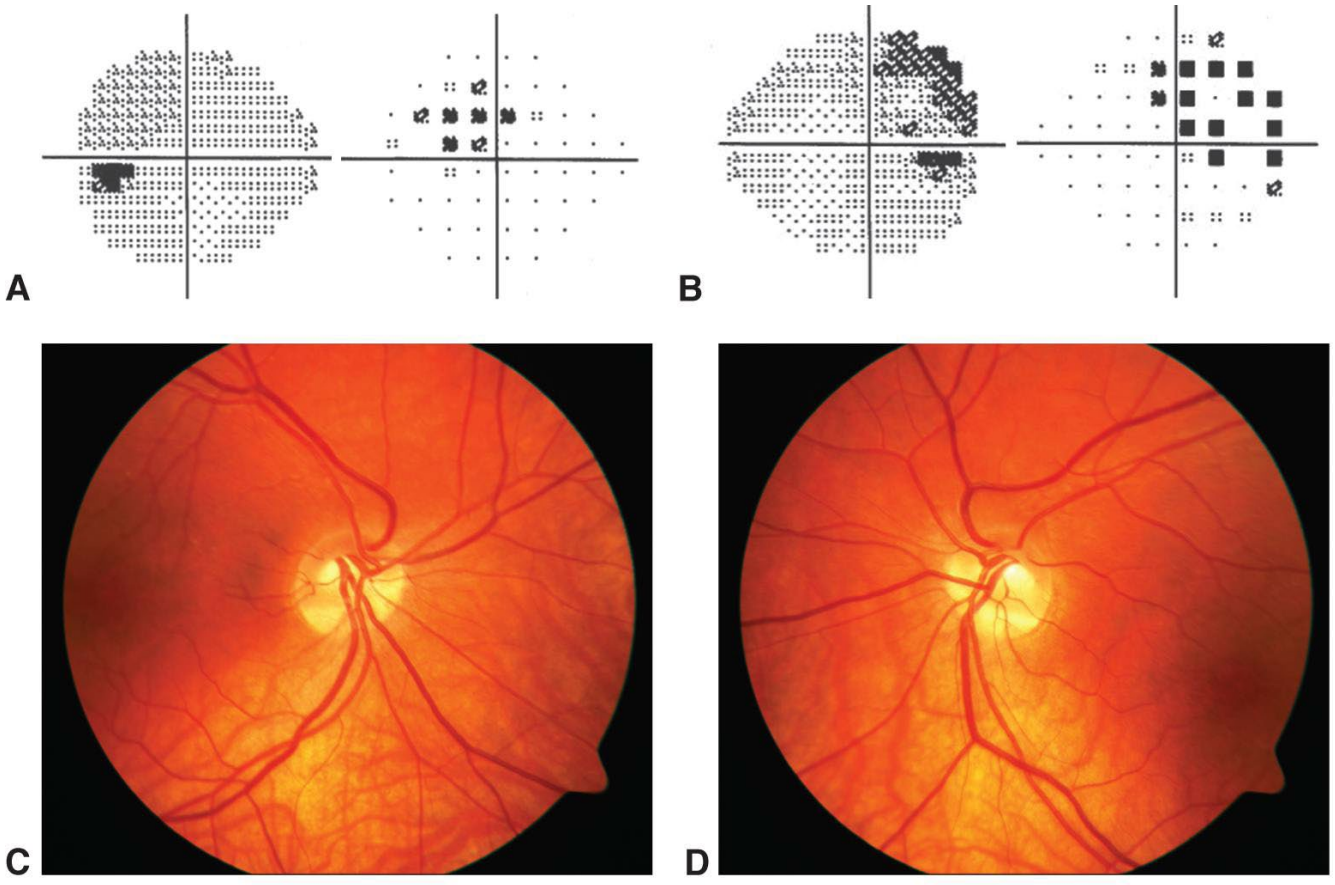
How on earth does a tilted disc produce a bitemporal VF defect?
It's actually pretty straightforward.
Recall that the area including and adjacent to the inferior pole of the ONH is staphylomatous. This means the 'axial length' of the photoreceptors within this region is greater than that of the rest of the posterior pole. Because of this extra axial length, the correction used during VF testing (which is based on the refraction of the non-staphylomatous fovea) is not myopic enough for the inferior peripapillary region. Because this region is out of focus, it will manifest a *refractive scotoma* on the test. And because the retina involved in this scotoma is **inferonasal** to the fovea, it follows that the resulting VF defect will be **superotemporal** to fixation. And, as Fuchs coloboma is virtually always bilateral, these superotemporal VF defects are present *bilaterally*.

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease



Tilted disc: Superior bitemporal VF defects

Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
- **Rod (Night Vision) Disease**
  - Fundus appearance *normal*
  - Fundus appearance *abnormal*

How on earth does a tilted disc produce a bitemporal VF defect?
It's actually pretty straightforward.
Recall that the area including and adjacent to the inferior pole of the ONH is staphylomatous. This means the 'axial length' of the photoreceptors within this region is greater than that of the rest of the posterior pole. Because of this extra axial length, the correction used during VF testing (which is based on the refraction of the non-staphylomatous fovea) is not myopic enough for the inferior peripapillary region. Be… *refractive scotoma* on the test. And becau… al to the fovea, it follows that the resulting VF d… uchs coloboma is virtually always bilateral, thes… y.

This implies the VF defects resolve if the proper refractive correction is employed. Do they?

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

Fundus appearance *abnormal*

How on earth does a tilted disc produce a bitemporal VF defect?
It's actually pretty straightforward.
Recall that the area including and adjacent to the inferior pole of the ONH is staphylomatous. This means the 'axial length' of the photoreceptors within this region is greater than that of the rest of the posterior pole. Because of this extra axial length, the correction used during VF testing (which is based on the refraction of the non-staphylomatous fovea) is not myopic enough for the inferior peripapillary region. Be… *refractive scotoma* on the test. And becau… al to the fovea, it follows that the resulting VF d… uchs coloboma is virtually always bilateral, thes… .

This implies the VF defects resolve if the proper refractive correction is employed. Do they?
Indeed they do, and this cinches the diagnosis

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

*When I hear 'superior bitemporal VF defect' two words come to mind, and they **ain't** Fuchs coloboma—they are* [redacted].

Trichr… undus …earance …normal

How o…
It's act…
Recall…
This m…
rest of… g
(which… -like)
inferio…
scoton… …ern?
it follo… …na
is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

*When I hear 'superior bitemporal VF defect' two words come to mind, and they **ain't** Fuchs coloboma—they are **pituitary tumor** .*

Trichr… undus …earance …normal

How o…
It's act…
Recall…
This m…
rest of… g
(which… -like)
inferio…
scoton… …ern?
it follo… …a
is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

*When I hear 'superior bitemporal VF defect' two words come to mind, and they **ain't** Fuchs coloboma—they are **pituitary tumor** . How am I supposed to know whether a bitemporal VF cut results from a pituitary tumor as opposed to Fuchs coloboma?*

Trichr… undus
…earance
…normal

How o…
It's act…
Recall…
This m…
rest of… g
(which… -like)
inferio…
scoto… …a, …ern?
it follo… …na
is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt** , and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

*When I hear 'superior bitemporal VF defect' two words come to mind, and they **ain't** Fuchs coloboma—they are **pituitary tumor** . How am I supposed to know whether a bitemporal VF cut results from a pituitary tumor as opposed to Fuchs coloboma?*
You mean, other than the fact that the ONHs in a Fuchs pt will be highly tilted and manifest situs inversus, whereas the ONHs of a pituitary-tumor pt will be edematous? (Remember: *When all else fails, examine the pt*.)

Trichr … undus … earance … normal

How o…
It's act…
Recall…
This m…
rest of… g
(which… -like)
inferio…
scoto… …a, …ern?
it follo… …na
is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

*When I hear 'superior bitemporal VF defect' two words come to mind, and they **ain't** Fuchs coloboma—they are **pituitary tumor** . How am I supposed to know whether a bitemporal VF cut results from a pituitary tumor as opposed to Fuchs coloboma?*
You mean, other than the fact that the ONHs in a Fuchs pt will be highly tilted and manifest situs inversus, whereas the ONHs of a pituitary-tumor pt will be edematous? (Remember: *When all else fails, examine the pt*.)

OK, fair. (Unnecessarily rude, but fair.) But is there a way to tell from the VF itself?

Trichr … undus … earance … normal

How o
It's act
Recall
This m
rest of … g
(which … like)
inferio
scoto … a, … ern?
it follo … na
is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

Does any of this impact vision?
It does indeed—the tilt of the ONH may produce a visual field defect (classically, a **bitemporal defect**)

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
Fuchs coloboma (Dr Fuchs strikes again—I stan for an eye king!)

Congenital/Stationary Retinal Disease

Cone (Color) Disease

Rod (Night Vision) Disease

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OK, fair. (Unnecessarily rude, but fair.) But is there a way to tell from the VF itself?
Indeed there is—a bitemporal VF defect 2ndry to a pituitary tumor will always respect the vertical vs horizontal midline, whereas one 2ndry to Fuchs coloboma will not.

is virtually always bilateral, **these superotemporal VF defects are present *bilaterally*.**

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Congenital/Stationary Retinal Disease

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You mean, other than the fact that the ONHs in a Fuchs pt will be highly tilted and manifest situs inversus, whereas the ONHs of a pituitary-tumor pt will be edematous? (Remember: *When all else fails, examine the pt*.)

OK, fair. (Unnecessarily rude, but fair.) But is there a way to tell from the VF itself?
Indeed there is—a bitemporal VF defect 2ndry to a pituitary tumor will always respect the vertical midline, whereas one 2ndry to Fuchs coloboma will not. Put another way: A pituitary tumor, but not a Fuchs coloboma, is expected to produce bitemporal **hemianopic** VF loss.

these superotemporal VF defects are present *bilaterally*.

bitemporal defect

In what manner is the ONH likely to be abnormal in CSNB?
It may manifest a myopic **tilt**, and its temporal aspect may be pallorous

What is the eponymous name for a tilted disc of this sort?
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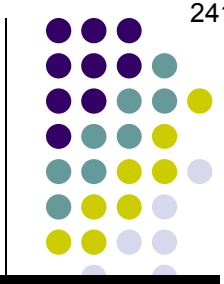


Congenital/Stationary Retinal Disease

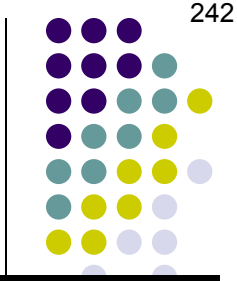
Cone (Color) Disease

- Trichromatism
- Dichromatism
- Monochromatism

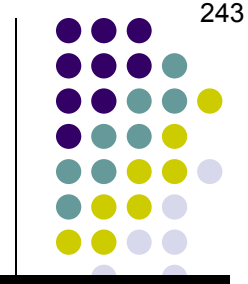
Rod (Night Vision) Disease

- Fundus appearance *normal*
  - CSNB
- Fundus appearance *abnormal*
  - ?
  - ?

Next question



Congenital/Stationary Retinal Disease
Cone (Color) Disease
Rod (Night Vision) Disease
Trichromatism
Dichromatism
Monochromatism
Fundus appearance normal
Fundus appearance abnormal
CSNB
Fundus albipunctatus
Oguchi disease



Congenital/Stationary Retinal Disease

Cone (Color) Disease

Trichromatism

Dichromatism

Monochromatism

Rod (Night Vision) Disease

Fundus appearance *normal*

CSNB

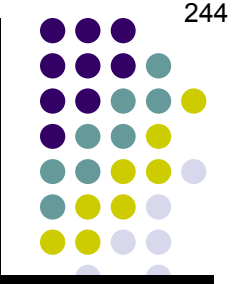
Fundus appearance *abnormal*

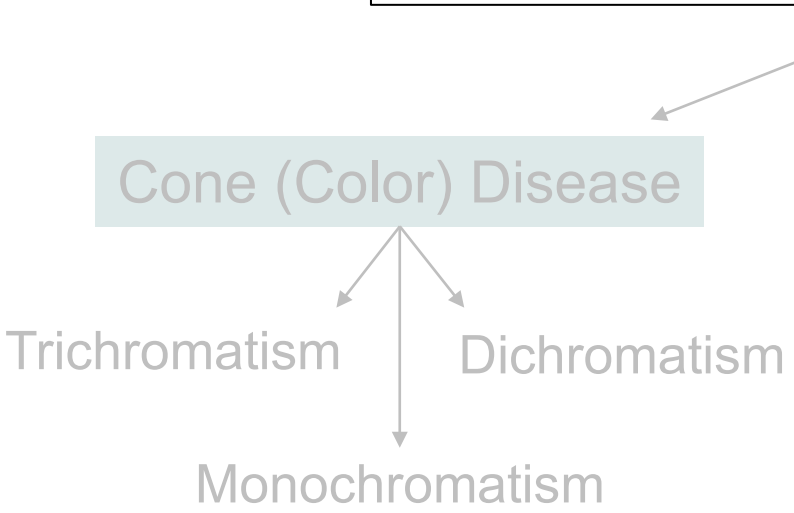
Fundus albipunctatus

Oguchi disease

Fundus Albipunctatus

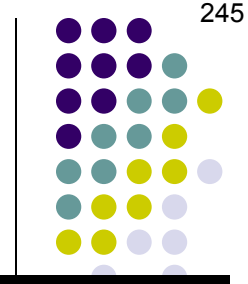
--Pathology: Delayed regeneration of the photopigment…

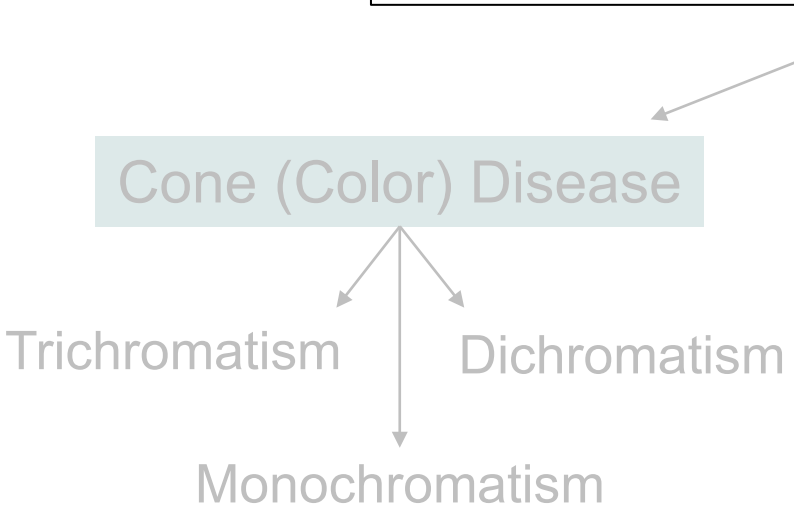




Fundus Albipunctatus

--Pathology: Delayed regeneration of the photopigment…rhodopsin



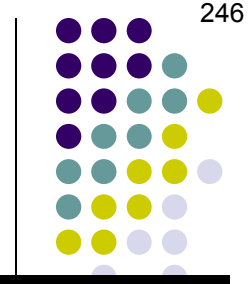


Fundus Albipunctatus

--Pathology: Delayed regeneration of the photopigment…rhodopsin

--Dark adaptation is abnormal:

--Initially, patients are…*[condition]*, with abnormal… *[test]*

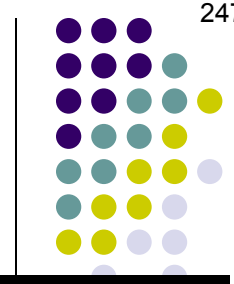


Fundus Albipunctatus

--Pathology: Delayed regeneration of the photopigment…rhodopsin

--Dark adaptation is abnormal:

--Initially, patients are…night-blind, with abnormal…rod ERG



Fundus Albipunctatus

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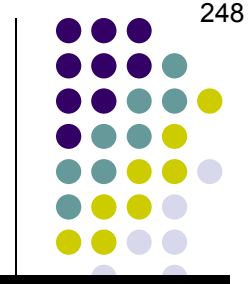
--With enough time, will dark-adapt, and ERG normalizes



Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- Rod (Night Vision) Disease
  - Fundus appearance *normal*
    - CSNB
  - Fundus appearance *abnormal*
    - **Fundus albipunctatus**
    - Oguchi disease

Fundus Albipunctatus

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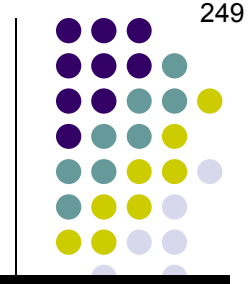
How much time are we talking about?



Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- Rod (Night Vision) Disease
  - Fundus appearance *normal*
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  - Fundus appearance *abnormal*
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Fundus Albipunctatus

--Pathology: Delayed regeneration of the photopigment…rhodopsin

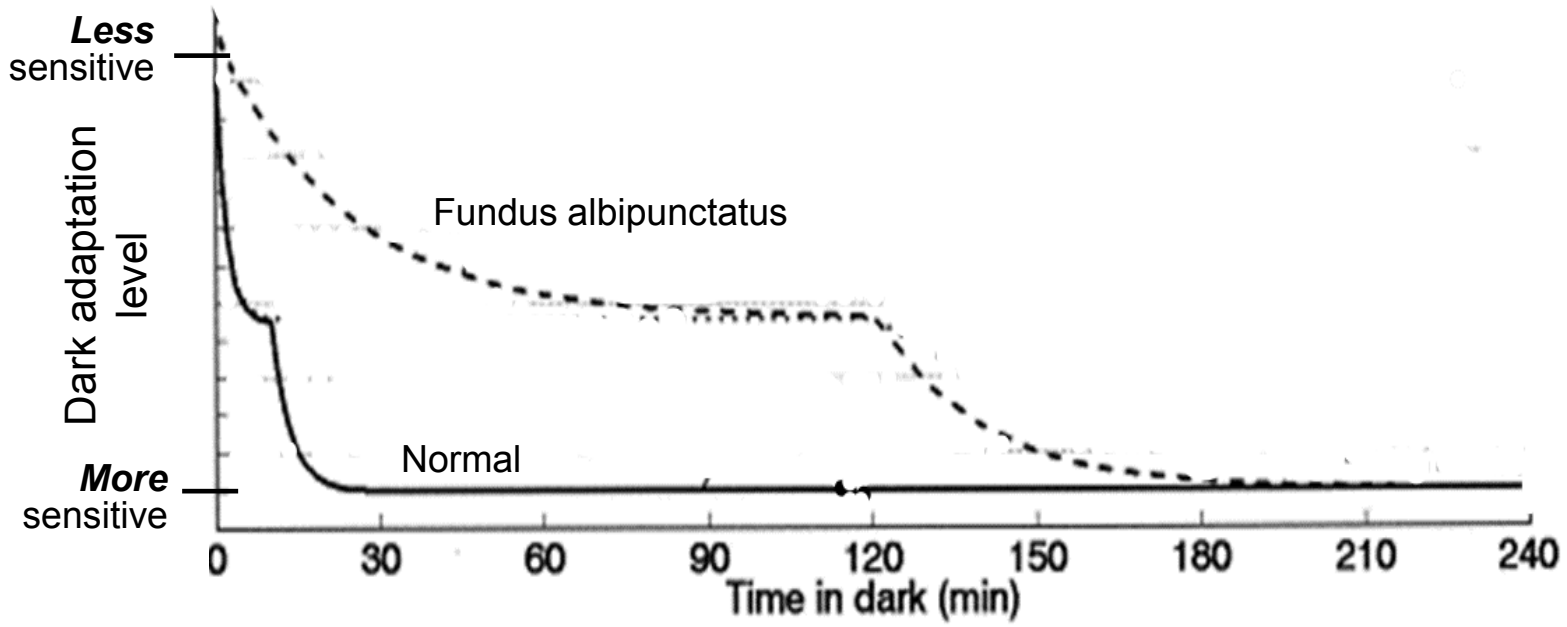
--Dark adaptation is abnormal:

--Initially, patients are…night blind, with abnormal…rod ERG

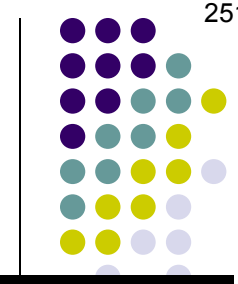
--**With enough time**, wil

How much time are we talking about?
Several hours at least

Congenital/Stationary Retinal Disease



Delayed dark adaptation in fundus albipunctatus



Fundus Albipunctatus

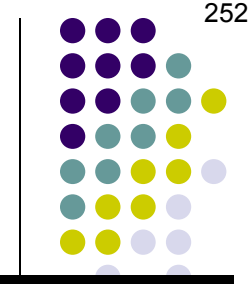
--Pathology: Delayed regeneration of the photopigment…rhodopsin

--Dark adaptation is abnormal:

--Initially, patients are…night-blind, with abnormal…rod ERG

--With enough time, will dark-adapt, and ERG normalizes

--DFE: Striking array of…***?***



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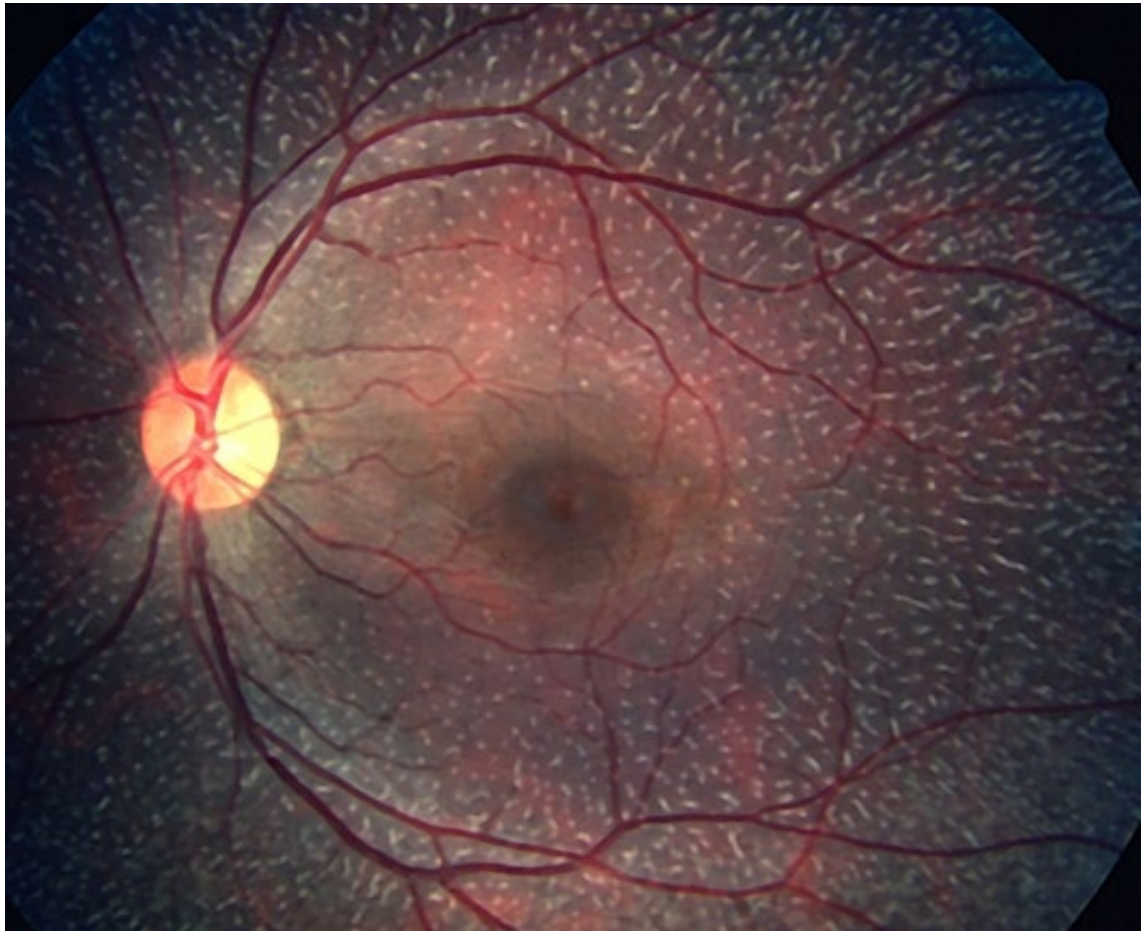
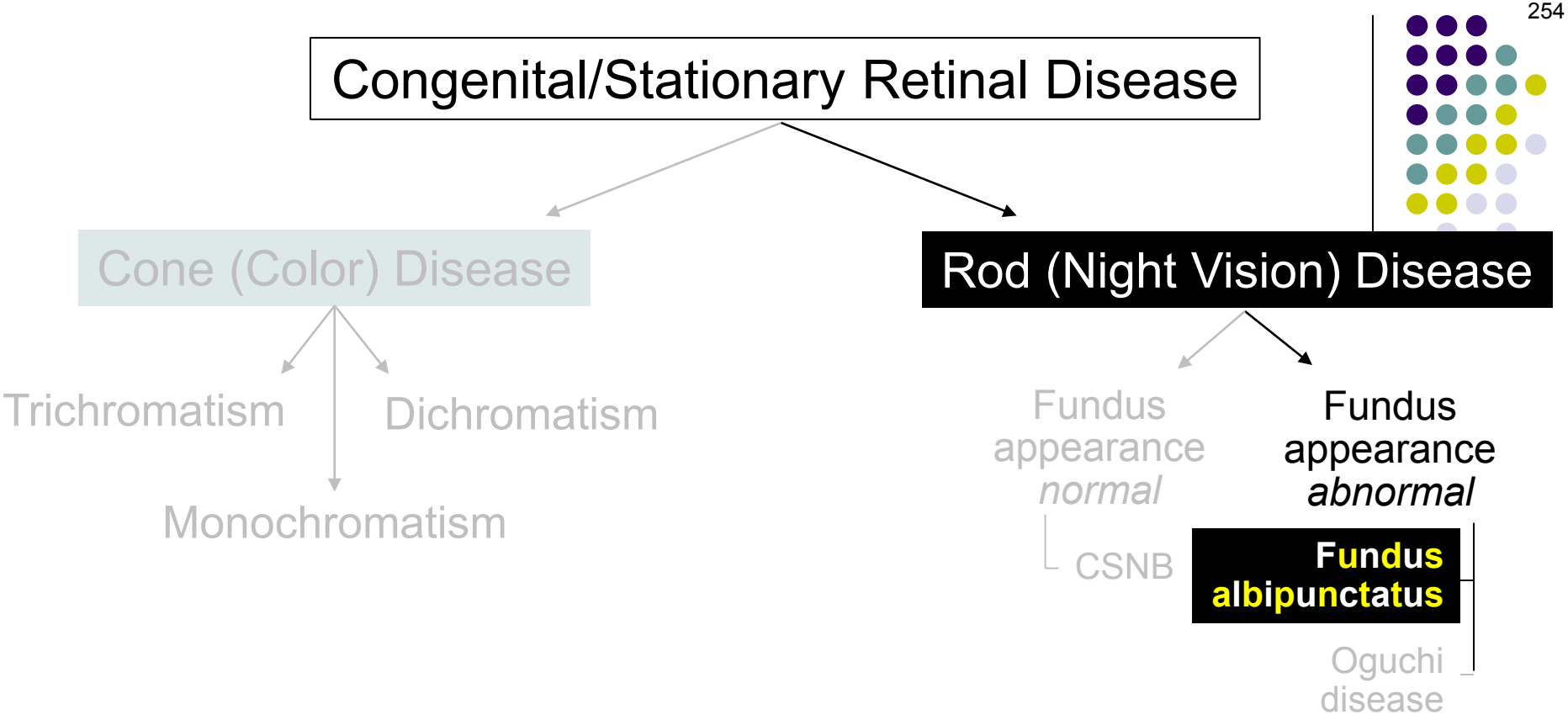
--DFE: Striking array of…**yellow - white dots**

Congenital/Stationary Retinal Disease



Fundus albipunctatus



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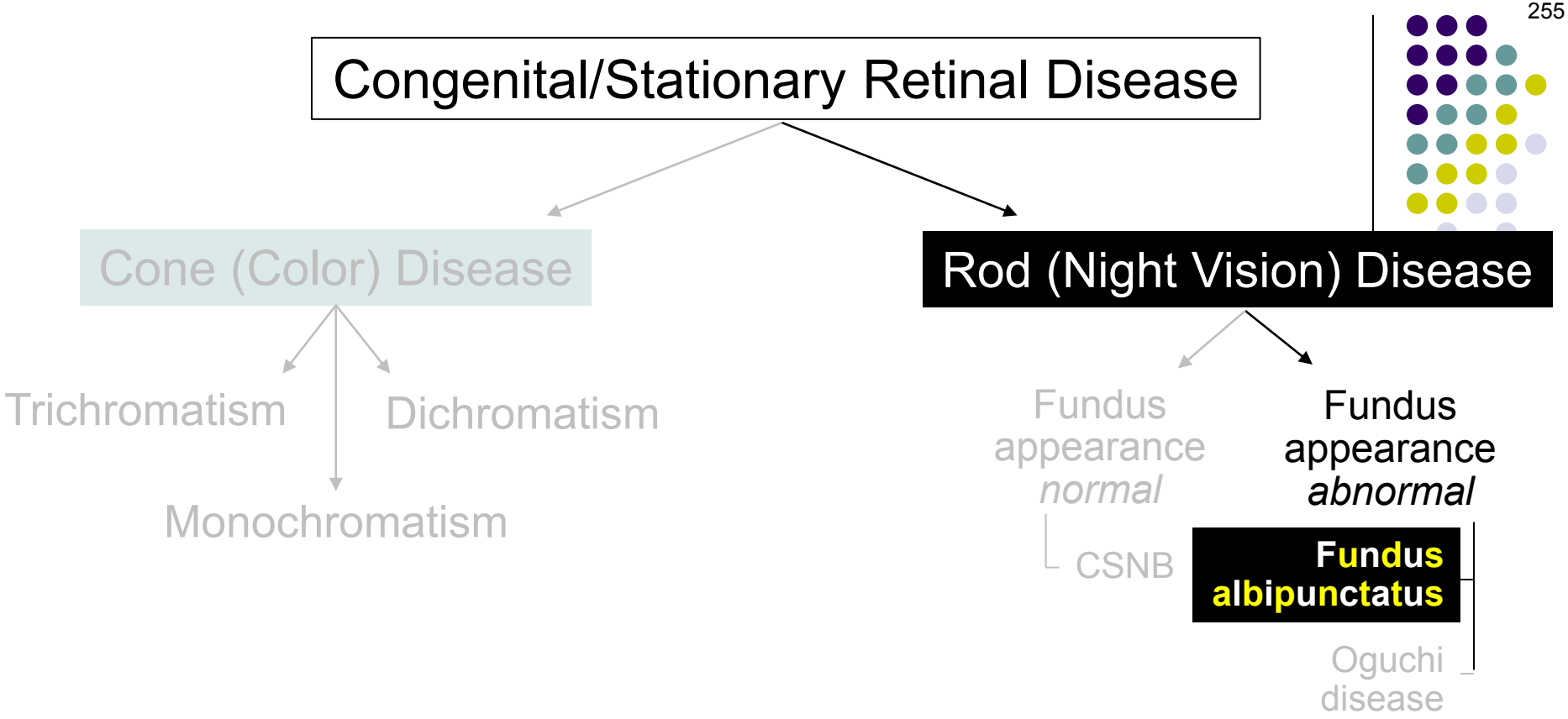
--DFE: Striking array of…**yellow - white dots**

--Dots found in entire posterior pole *except…**?***



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--DFE: Striking array of…**yellow - white dots**

--Dots found in entire posterior pole *except*…fovea

Congenital/Stationary Retinal Disease

What is the main disease that must be differentiated from fundus albipunctatus?

Disease

Tri

undus
earance
normal

undus
ctatus

guchi
isease

Fundus Albipunctatus

--Pathology: Delayed regeneration of the photopigment…rhodopsin
--Dark adaptation is abnormal:
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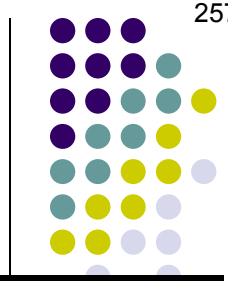
Congenital/Stationary Retinal Disease



Disease

Tri

undus
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Oguchi
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What is the main disease that must be differentiated from fundus albipunctatus?
Retinitis punctata albescens

Fundus Albipunctatus

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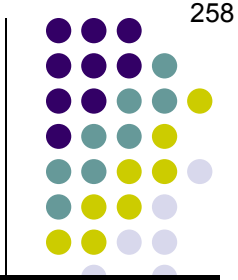
Congenital/Stationary Retinal Disease



Disease

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What is retinitis punctata albescens?

undus
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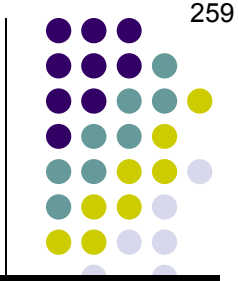
Congenital/Stationary Retinal Disease



Disease

What is the main disease that must be differentiated from fundus albipunctatus?
Retinitis punctata albescens

What is retinitis punctata albescens?
An abb. variant characterized by white - yellow dots similar to those of albipunctatus

undus
earance
normal

undus
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Fundus Albipunctatus

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Congenital/Stationary Retinal Disease

What is the main disease that must be differentiated from fundus albipunctatus?
Retinitis punctata albescens

What is retinitis punctata albescens?
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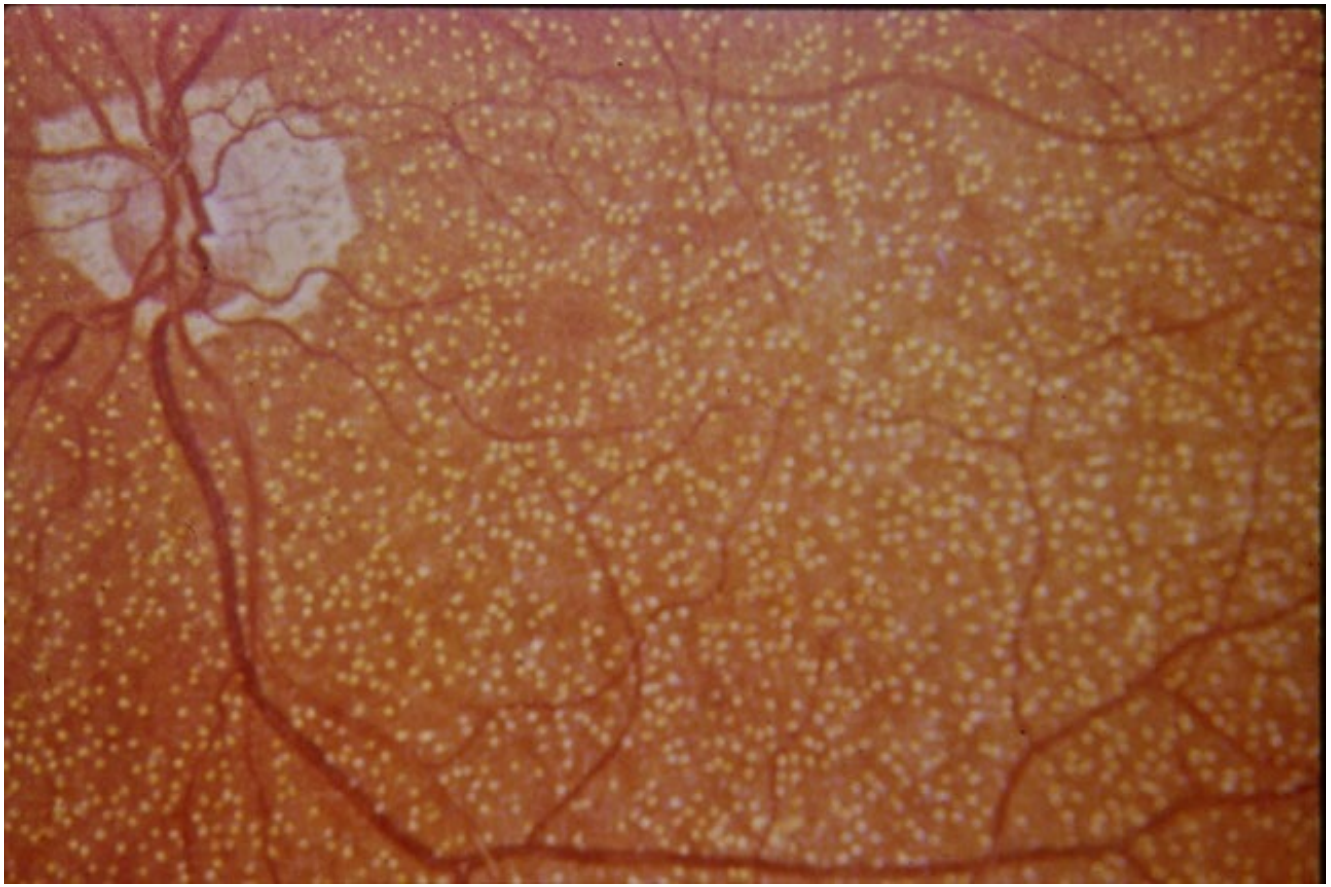
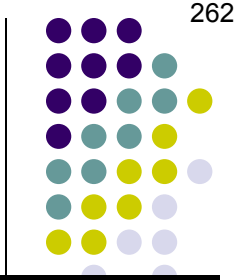
Congenital/Stationary Retinal Disease



Retinitis punctata albescens

Congenital/Stationary Retinal Disease



Disease

What is the main disease that must be differentiated from fundus albipunctatus?
Retinitis punctata albescens

What is retinitis punctata albescens?
An RP variant characterized by white - yellow dots similar to those of albipunctatus

How do fundus albipunctatus and retinitis punctata albescens differ?

Tri

undus
earance
normal

undus
ctatus

guchi
sease

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--On DFE: ***?***

--On ERG

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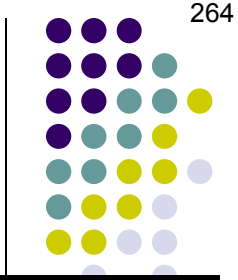
Congenital/Stationary Retinal Disease



Disease

What is the main disease that must be differentiated from fundus albipunctatus?
Retinitis punctata albescens

What is retinitis punctata albescens?
An RP variant characterized by white - yellow dots similar to those of albipunctatus

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

Fundus Albipunctatus
--Pathology: Delayed regeneration of the photopigment…rhodopsin
--Dark adaptation is abnormal:
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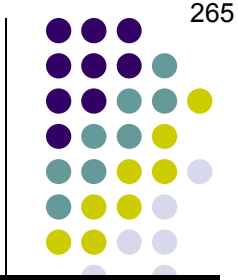
Congenital/Stationary Retinal Disease



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Fundus Albipunctatus
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Congenital/Stationary Retinal Disease

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How do fundus albipunctatus and retinitis punctata albescens differ?
--On DFE: Like other forms of RP, retinitis punctata albescens demonstrates **arteriolar narrowing**, whereas albipunctatus does not
--On ERG: Fundus albipunctatus is a disease of abnormal rhodopsin regeneration, which manifests as slow but ultimately successful dark adaptation.

Fundus Albipunctatus
--Pathology: Delayed regeneration of the photopigment…rhodopsin
--Dark adaptation is abnormal:
--Initially, patients are…night-blind, with abnormal…rod ERG
--With enough time, will dark-adapt, and ERG normalizes
--DFE: Striking array of…**yellow - white dots**
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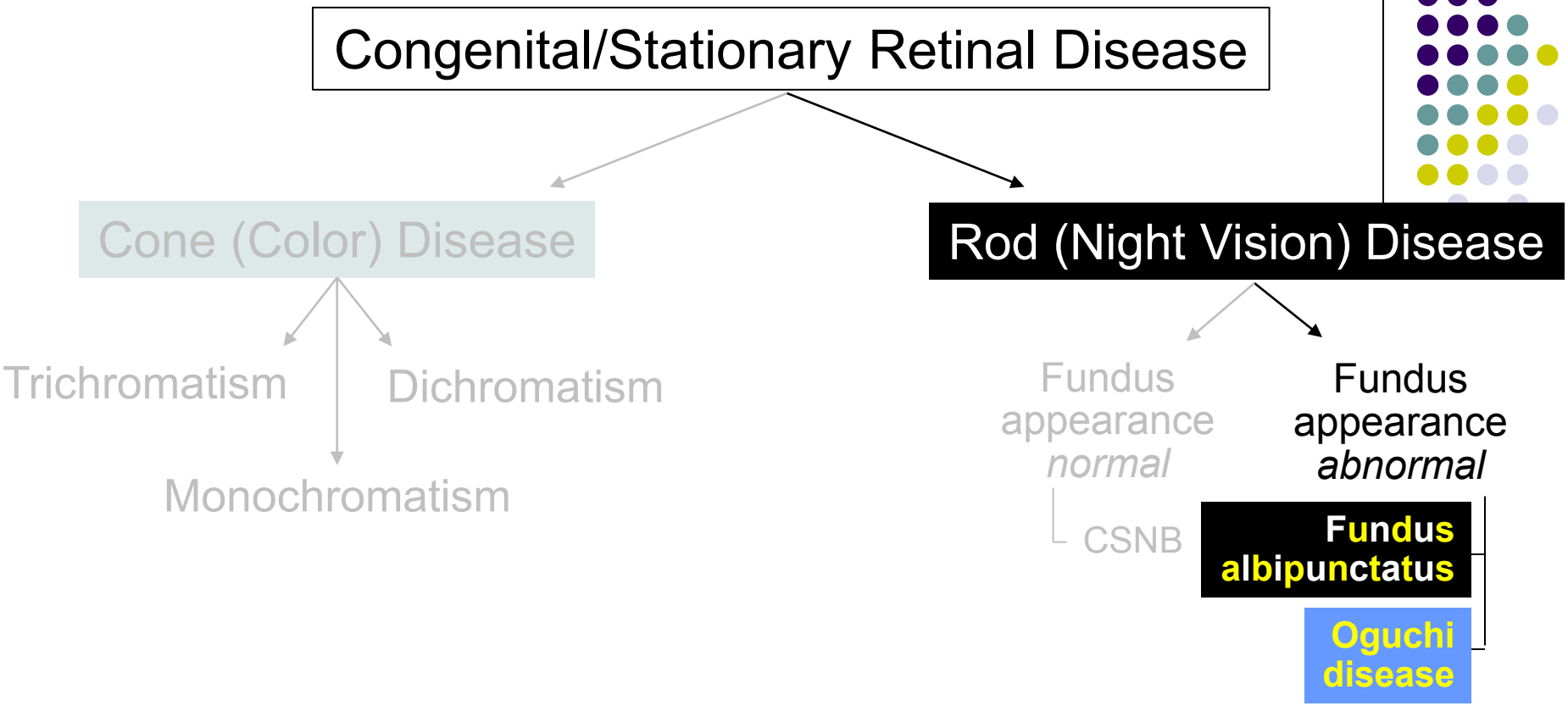
What is retinitis punctata albescens?
An RP variant characterized by white - yellow dots similar to those of albipunctatus

How do fundus albipunctatus and retinitis punctata albescens differ?
--On DFE: Like other forms of RP, retinitis punctata albescens demonstrates **arteriolar narrowing**, whereas 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.

Fundus Albipunctatus

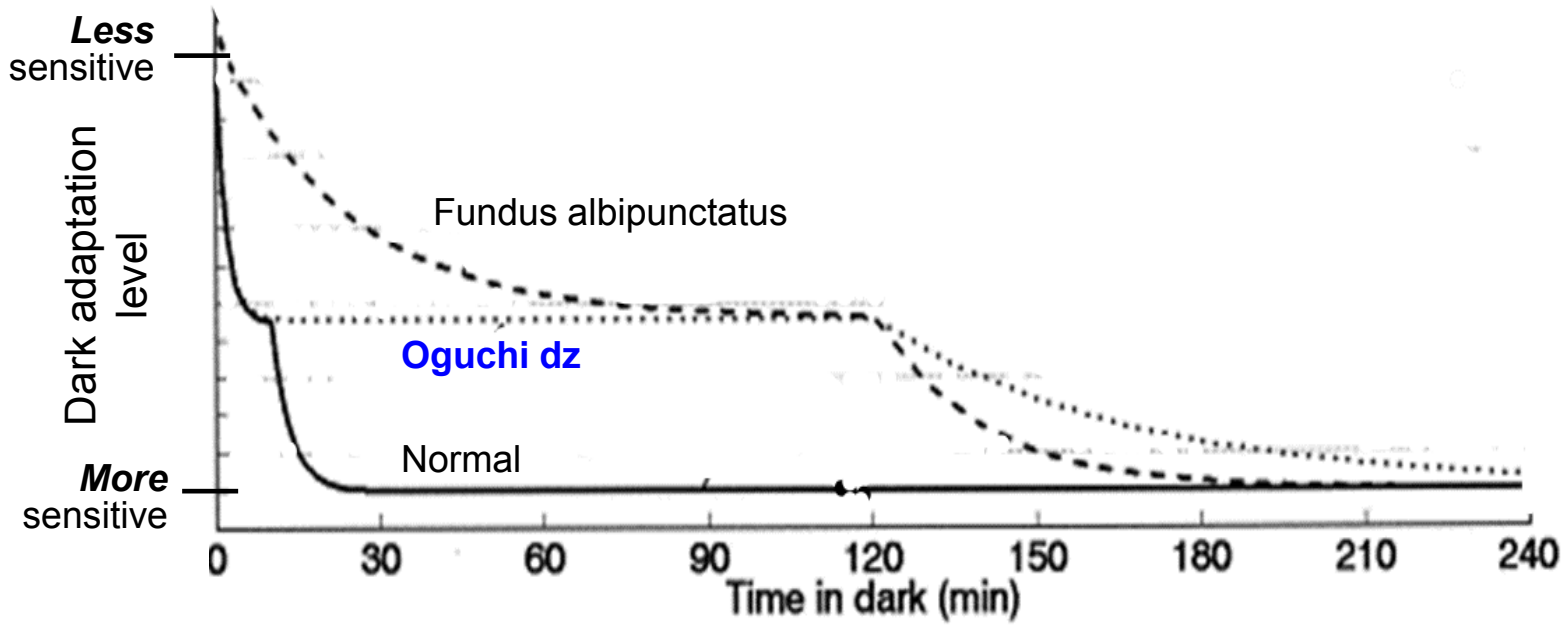
--Pathology: Delayed regeneration of the photopigment…rhodopsin
--Dark adaptation is abnormal:
  --Initially, patients are…night-blind, with abnormal…rod ERG
  --With enough time, will dark-adapt, and ERG normalizes
--DFE: Striking array of…**yellow - white dots**
  --Dots found in entire posterior pole *except*…fovea



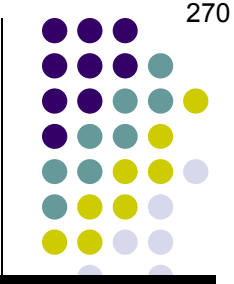
Oguchi Disease

--Also have slow dark adaptation (*not* a pigment regeneration issue, though)

Congenital/Stationary Retinal Disease



Delayed dark adaptation in Oguchi dz



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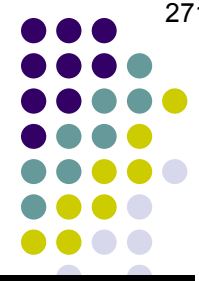
--Once dark-adapted, dark sensitivity lost with a single…[*event*]



Congenital/Stationary Retinal Disease

- Cone (Color) Disease
  - Trichromatism
  - Dichromatism
  - Monochromatism
- Rod (Night Vision) Disease
  - Fundus appearance *normal*
    - CSNB
  - Fundus appearance *abnormal*
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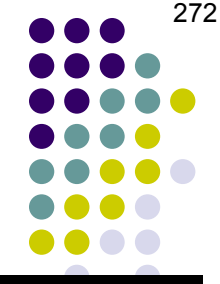

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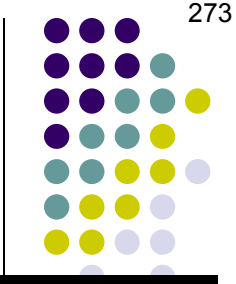
--DFE:

--Normal appearance when…*[state of adaptation]*



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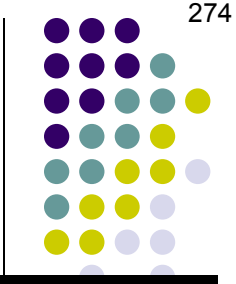

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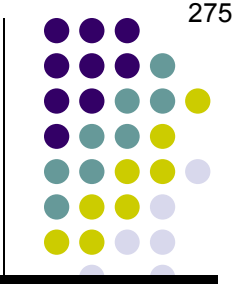
  --After light exposure, posterior pole takes on a…*[appearance]*



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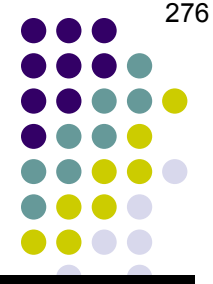

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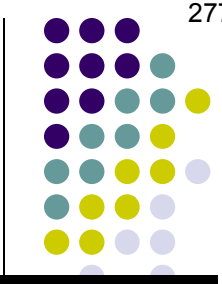
    --This color change is known as the…[*eponym-eponym*]



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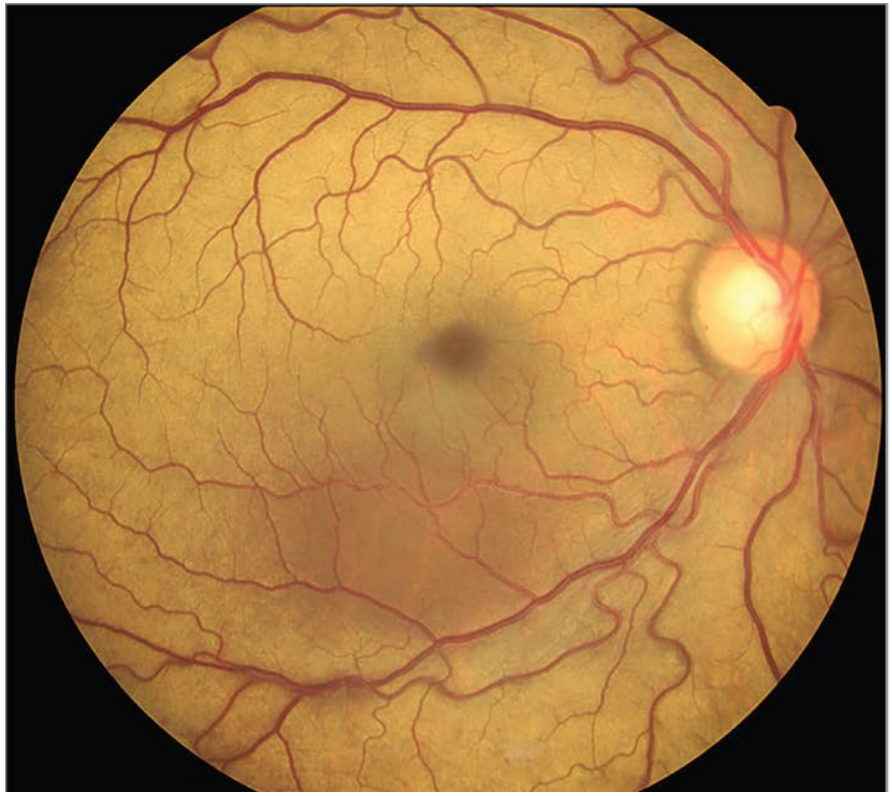
Congenital/Stationary Retinal Disease



Appearance after exposure to light

Appearance in dark-adapted state

Mizuo-Nakamura phenomenon in Oguchi dz

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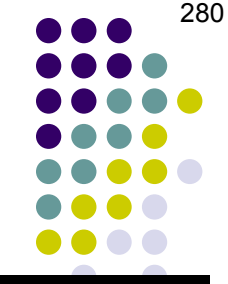
--DFE:

--Normal app…

--After light e…**ow iridescent sheen**

--This color …**ura phenomenon**

Is Oguchi dz common, or rare?



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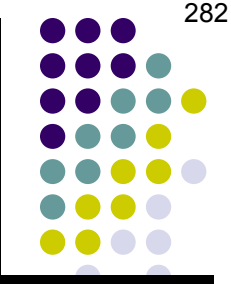
With what ethnicity is it closely associated?



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Japanese