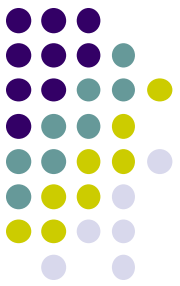
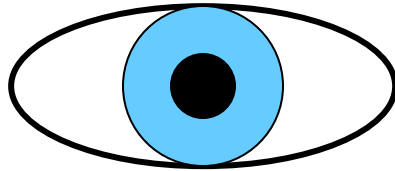


Q

1

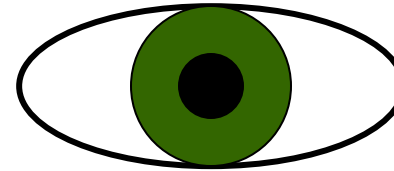


Heterochromia iridis:  
Divide these...



**LIGHTER** iris abnormal

...into their respective  
categories



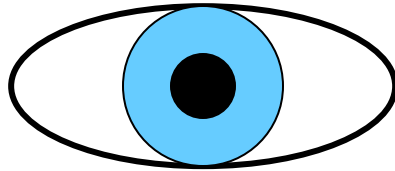
**DARKER** iris abnormal

*Start at the top and  
work down the list* →

Siderosis  
Rb  
Congenital Horner's  
Melanoma  
JXG  
Waardenburg syndrome  
Unilateral Xalatan use  
Oculodermal melanocytosis  
Incontinentia pigmenti  
Fuchs heterochromic iridocyclitis

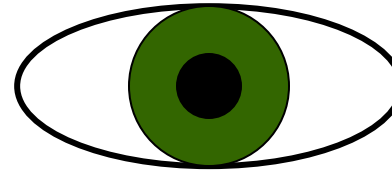
A

Heterochromia iridis:  
Divide these...



**LIGHTER** iris abnormal

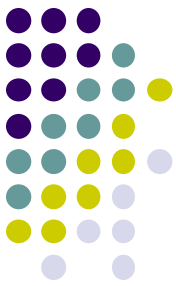
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**DARKER** iris abnormal

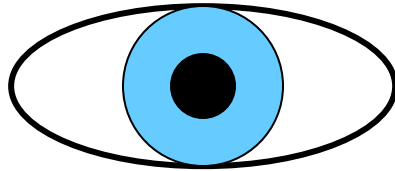
Siderosis

Rb  
Congenital Horner's  
Melanoma  
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Waardenburg syndrome  
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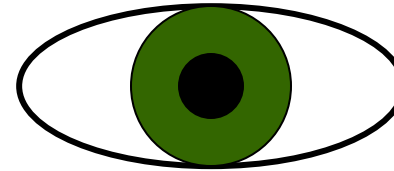
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Heterochromia iridis:  
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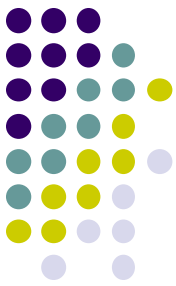
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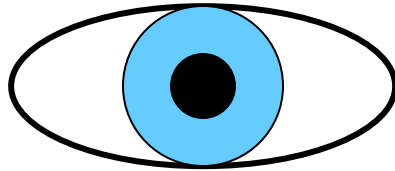
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Congenital Horner's  
Melanoma  
JXG  
Waardenburg syndrome  
Unilateral Xalatan use  
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Fuchs heterochromic iridocyclitis



A

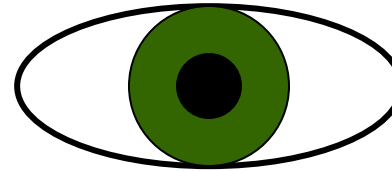
Heterochromia iridis:  
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**LIGHTER** iris abnormal

Congenital Horner's

...into their respective  
categories



**DARKER** iris abnormal

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Rb

Melanoma  
JXG

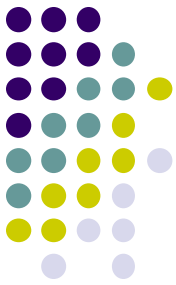
Waardenburg syndrome

Unilateral Xalatan use

Oculodermal melanocytosis

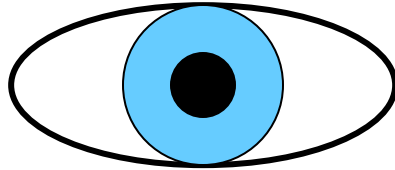
Incontinentia pigmenti

Fuchs heterochromic iridocyclitis



A

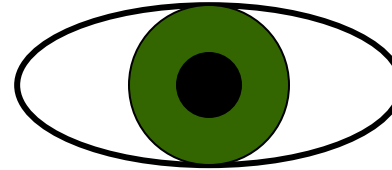
Heterochromia iridis:  
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Congenital Horner's

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**DARKER iris abnormal**

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Rb

Melanoma

JXG

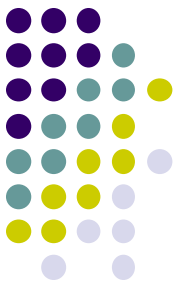
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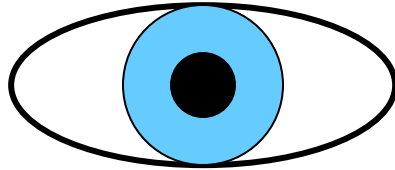
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Fuchs heterochromic iridocyclitis



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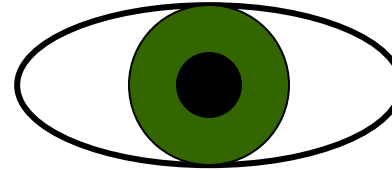
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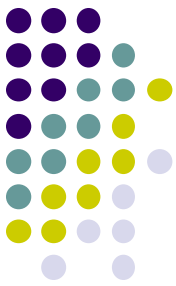
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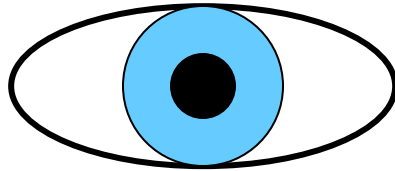
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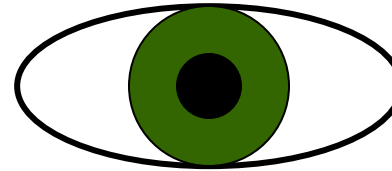
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Fuchs heterochromic iridocyclitis

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categories

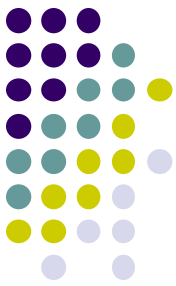


**DARKER** iris abnormal

Siderosis

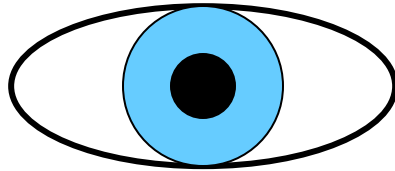
Rb

Melanoma



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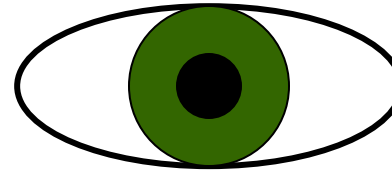
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**DARKER** iris abnormal

Siderosis

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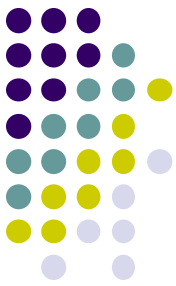
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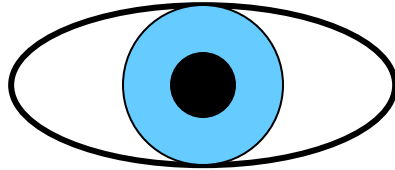
Fuchs heterochromic iridocyclitis





A

Heterochromia iridis:  
Divide these...



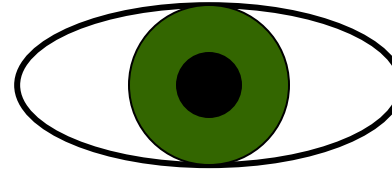
**LIGHTER iris abnormal**

Congenital Horner's

JXG

Waardenburg syndrome

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**DARKER iris abnormal**

Siderosis

Rb

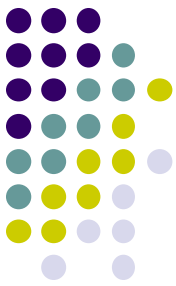
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Unilateral Xalatan use

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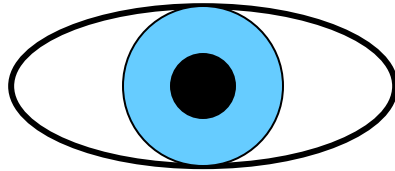
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Congenital Horner's

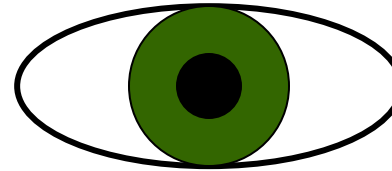
JXG

Waardenburg syndrome

Incontinentia pigmenti

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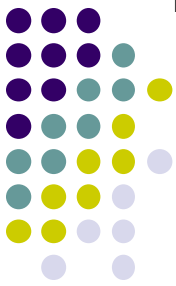
**DARKER** iris abnormal

Siderosis

Rb

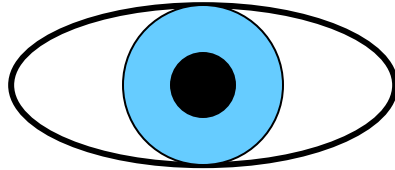
Melanoma

Unilateral Xalatan use  
Oculodermal melanocytosis



A

Heterochromia iridis:  
Divide these...



## LIGHTER iris abnormal

Congenital Horner's

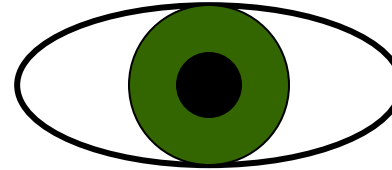
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Waardenburg syndrome

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Fuchs heterochromic iridocyclitis

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## DARKER iris abnormal

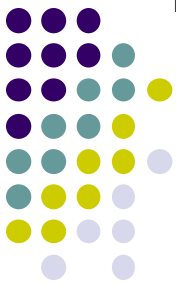
Siderosis

Rb

Melanoma

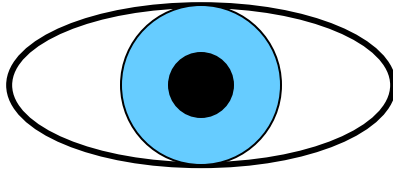
Unilateral Xalatan use

Oculodermal melanocytosis



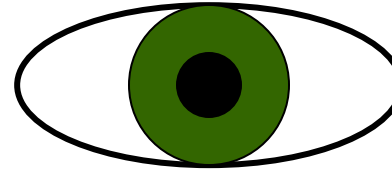
Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

...into their respective  
categories



DARKER iris abnormal

**Congenital Horner's**

JXG

Waardenburg syndrome

Incontinentia pigmenti

Fuchs heterochromic iridocyclitis

*What is the Horner syndrome triad?*

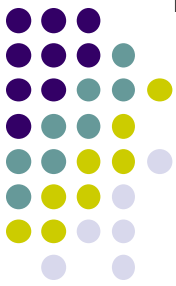
--?

--?

--?

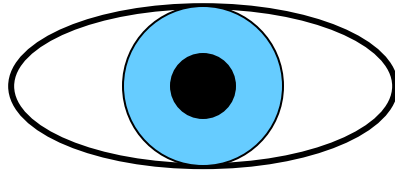
Siderosis

Unilateral Xalatan use  
Oculodermal melanocytosis



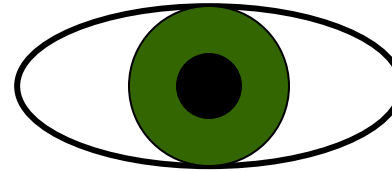
A

Heterochromia iridis:  
Divide these...



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DARKER iris abnormal

**Congenital Horner's**

JXG

Waardenburg syndrome

Incontinentia pigmenti

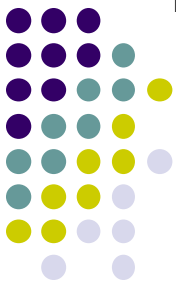
Fuchs heterochromic iridocyclitis

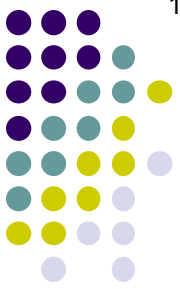
*What is the Horner syndrome triad?*

- Ptosis
- Miosis
- Anhydrosis

Siderosis

Unilateral Xalatan use  
Oculodermal melanocytosis

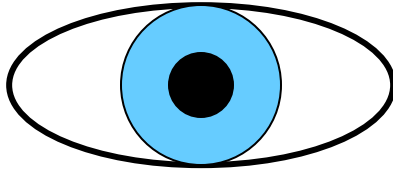




Congenital Horner's (note the ptosis and miosis in the lighter-colored eye)

Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

**Congenital Horner's**

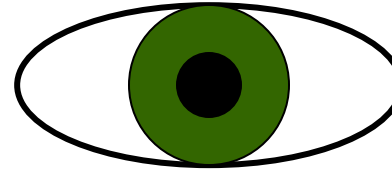
JXG

Waardenburg syndrome

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Fuchs heterochromic iridocyclitis

...into their respective  
categories



DARKER iris abnormal

Siderosis

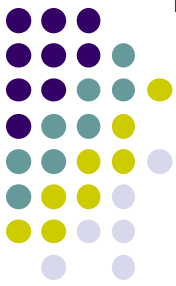
*What **nonocular** findings would suggest the presence of a congenital Horner's?*

--

--

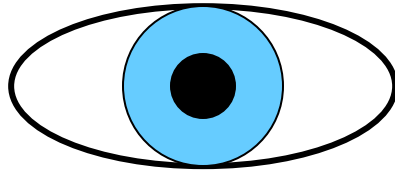
Unilateral Xalatan use

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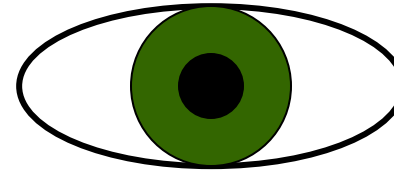
# A/Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

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DARKER iris abnormal

**Congenital Horner's**

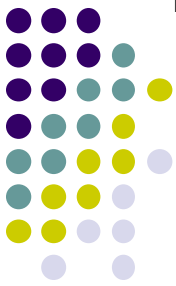
(The term is invented, but the finding is not!)

What **nonocular** findings would suggest the presence of a congenital Horner's?

--'Heterochromia nipplea' (ipsilateral areola lighter)  
--Fairer hair on the ipsi- vs  
contralateral side of the head

Unilateral Xalatan use  
Oculodermal melanocytosis

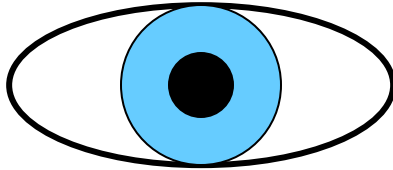
Incontinentia pigmenti  
Fuchs heterochromic iridocyclitis





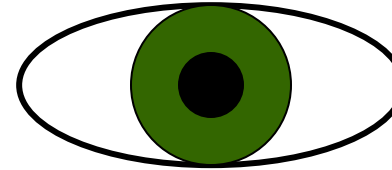
A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

...into their respective  
categories



DARKER iris abnormal

**Congenital Horner's**

(The term is invented, but the finding is not!)

What **nonocular** findings would suggest the presence of a congenital Horner's?

- 'Heterochromia nipplea' (ipsilateral areola lighter)
- Fairer hair on the ipsilateral side of the head

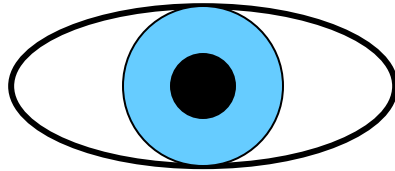
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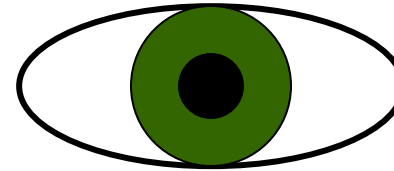
Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

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categories



DARKER iris abnormal

## Congenital Horner's

Siderosis  
*What nonocular findings would suggest the presence of a congenital Horner's?*

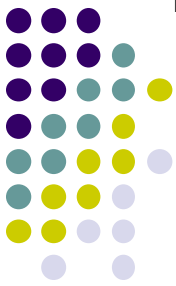
--'Heterochromia nipplea' (ipsilateral areola lighter)

--**Fairer hair on the ipsilateral side of the head**

In addition to being fairer, if the child has naturally curly hair, the hair on that side will be noticeably more vs less curly

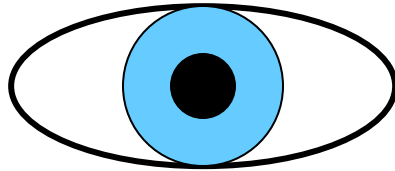
Unilateral Xalatan use  
Oculodermal melanocytosis

Incontinentia pigmenti  
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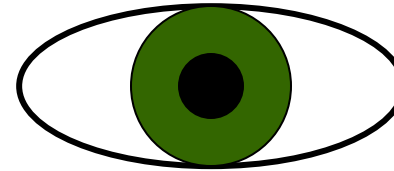
A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

...into their respective  
categories



DARKER iris abnormal

## Congenital Horner's

Siderosis

*What nonocular findings would suggest the presence of a congenital Horner's?*

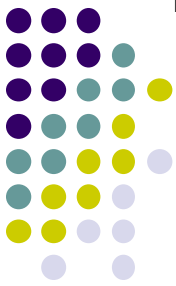
--'Heterochromia nipplea' (ipsilateral areola lighter)

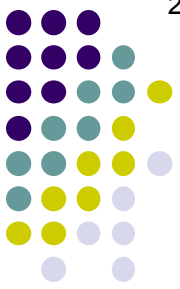
**--Fairer hair on the ipsilateral side of the head**

In addition to being fairer, if the child has naturally curly hair, the hair on that side will be noticeably **less** curly

Incontinentia pigmenti  
Fuchs heterochromic iridocyclitis

Unilateral Xalatan use  
Oculodermal melanocytosis

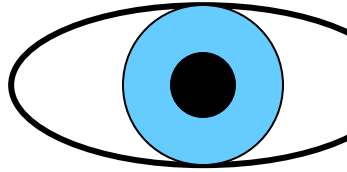




Right congenital Horner's (note the straighter hair on the right side)

Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

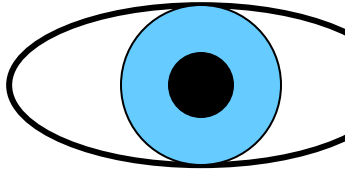
Incontinentia pigmenti

Fuchs heterochromic iridocyc

*What does JXG stand for?*

A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

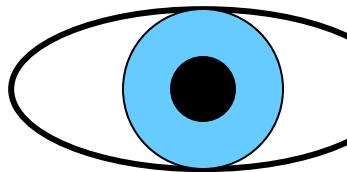
Incontinentia pigmenti

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What does JXG stand for?  
Juvenile xanthogranuloma

Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

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Waardenburg syndrome

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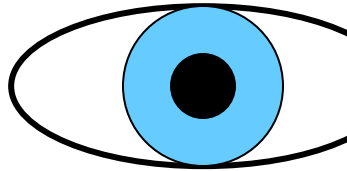
What does JXG stand for?

**Juvenile** xanthogranuloma

*The modifier juvenile implies that adult-onset xanthogranuloma is a thing—is it?*

A

Heterochromia iridis:  
Divide these...



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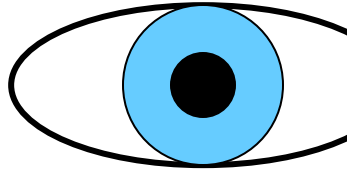
**Juvenile** xanthogranuloma

*The modifier juvenile implies that adult-onset xanthogranuloma is a thing—is it?*  
Indeed it is



Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

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What does JXG stand for?

**Juvenile** xanthogranuloma

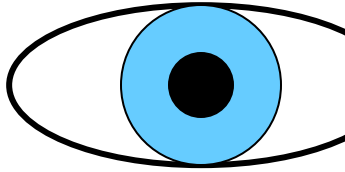
*The modifier juvenile implies that adult-onset xanthogranuloma is a thing—is it?*

Indeed it is

*What is adult-onset xanthogranuloma known as?*

A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

Incontinentia pigmenti

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What does JXG stand for?

**Juvenile** xanthogranuloma

*The modifier juvenile implies that adult-onset xanthogranuloma is a thing—is it?*

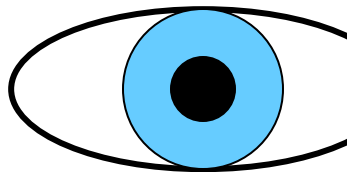
Indeed it is

*What is adult-onset xanthogranuloma known as?*

It's known as 'adult-onset xanthogranuloma' (AOX)

Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

Incontinentia pigmenti

Fuchs heterochromic iridocyc

What does JXG stand for?

**Juvenile** xanthogranuloma

*The modifier juvenile implies that adult-onset xanthogranuloma is a thing—is it?*

Indeed it is

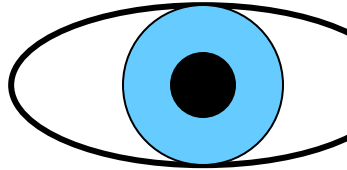
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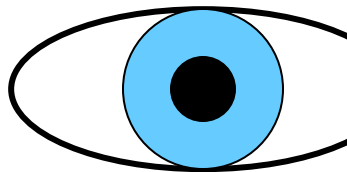
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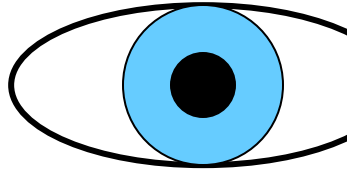
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*(Hints forthcoming)*

# Q/A

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--? *This one's name is rather on-the-nose*

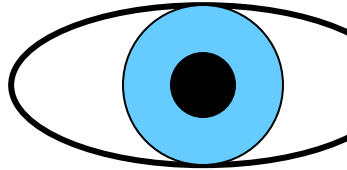
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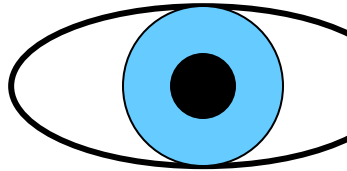
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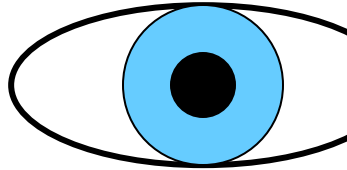
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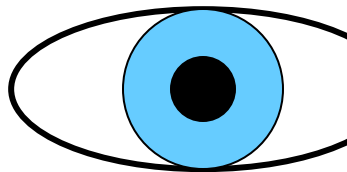
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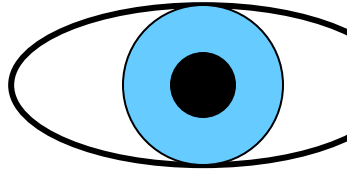
--Necrobiotic xanthogranuloma

--Adult-onset [wheeze] with periorcular xanthogranuloma

--?

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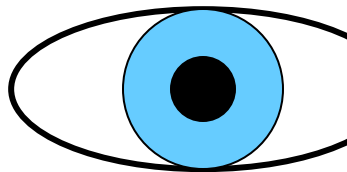
--Necrobiotic xanthogranuloma

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--?

Q

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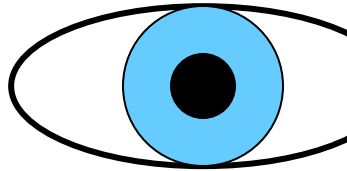
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- eponym-eponym** disease

A

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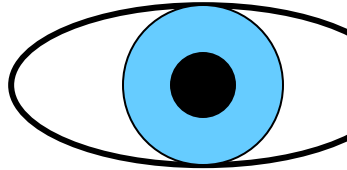
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***For more on the AOXs, see slide-set K20***

JXG

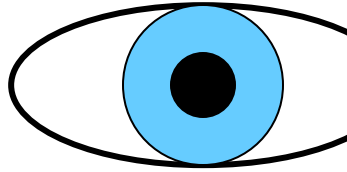
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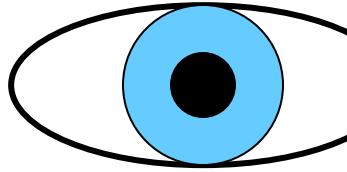
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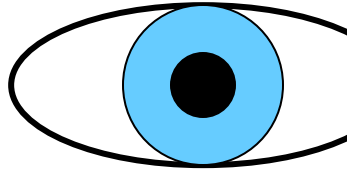
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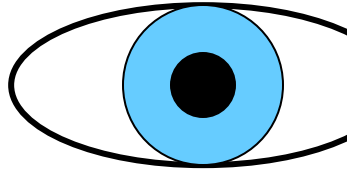
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The presence of...?  
The presence of...?

Q/A

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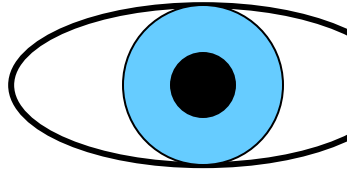
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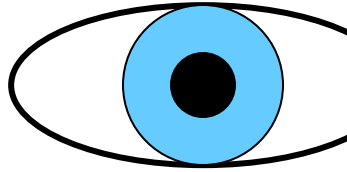
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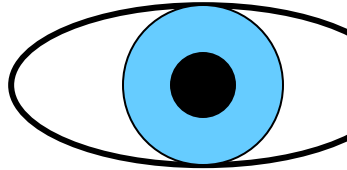
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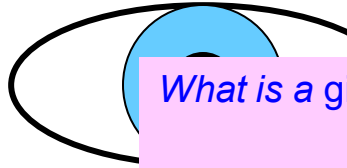
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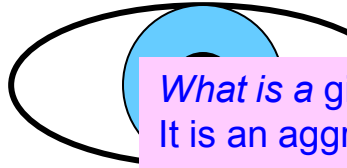
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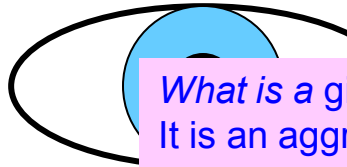
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It is an aggregate of

cell type

A

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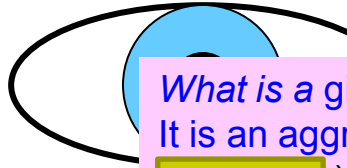
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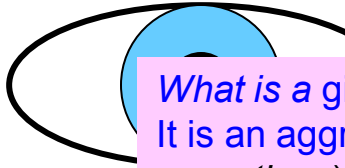
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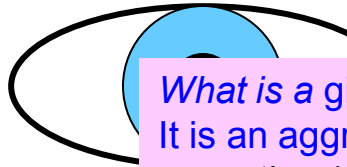
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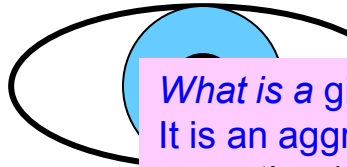
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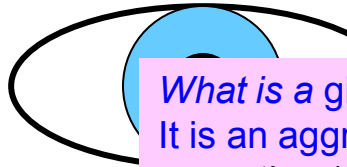
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What is the histologic hallmark of a giant cell?

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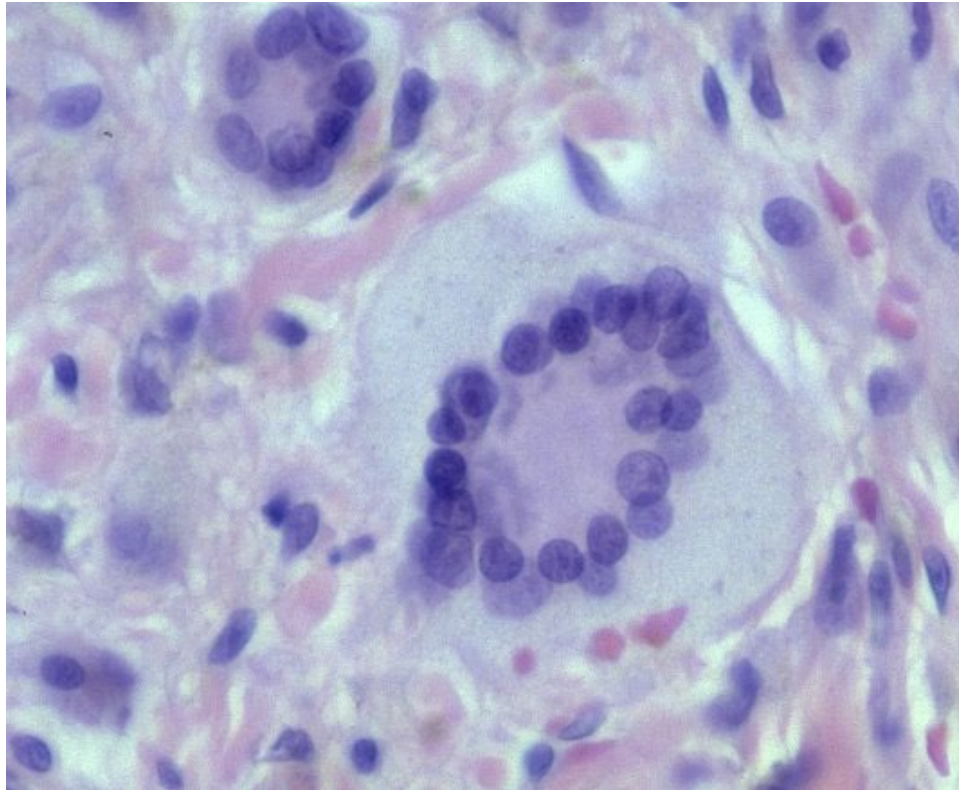
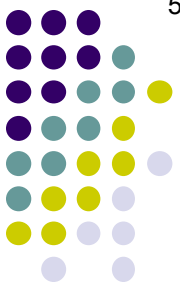
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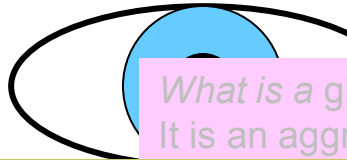
It is *multinucleated*—the myriad nuclei of the involved histiocytes are all visible within it



JXG: Touton giant cells

Q

Heterochromia iridis:  
Divide these...



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Histiocytes are also known as...?

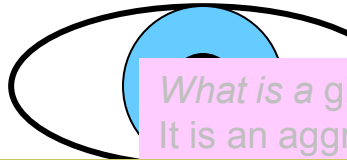
cell?

The involved histiocytes are all visible

Incontinentia pigmenti  
Fuchs heterochromic iridocyclitis

A

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Macrophages

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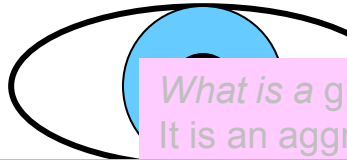
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Histiocytes are also known as...?

Macrophages

Histiocytes derive from what type of cell?

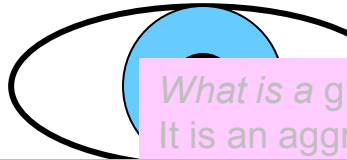
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What is a giant cell?

It is an aggregate of **histiocytes**. (The formal term for the aggregate is histiocytes that have glommed together.

Histiocytes are also known as...?

Macrophages

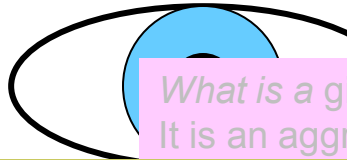
Histiocytes derive from what type of cell?

Monocytes

Incontinentia pigmenti  
Fuchs heterochromic iridocycl

Q

Heterochromia iridis:  
Divide these...



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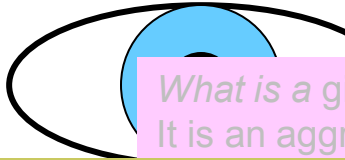
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What does derive mean here? How does a monocyte become a histiocyte?

Incontinentia pigmenti  
Fuchs heterochromic iridocycl

# Q/A

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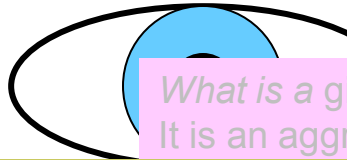
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Monocytes are found within the

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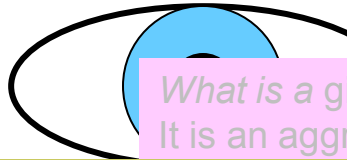
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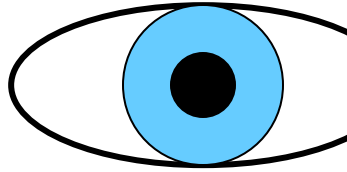
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Monocytes are found within the vasculature. When a monocyte leaves the circulation and settles down within tissue, it becomes (ie, is re-classified) as a histiocyte.

Incontinentia pigmenti  
Fuchs heterochromic iridocyclitis

Q

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

Incontinentia pigmenti

Fuchs heterochromic iridocycl

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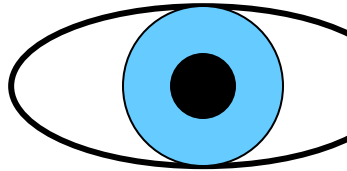
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'Foamy macrophages'? I thought they were 'lipid-laden histiocytes.'  
What's the deal?

A

Heterochromia iridis:  
Divide these...



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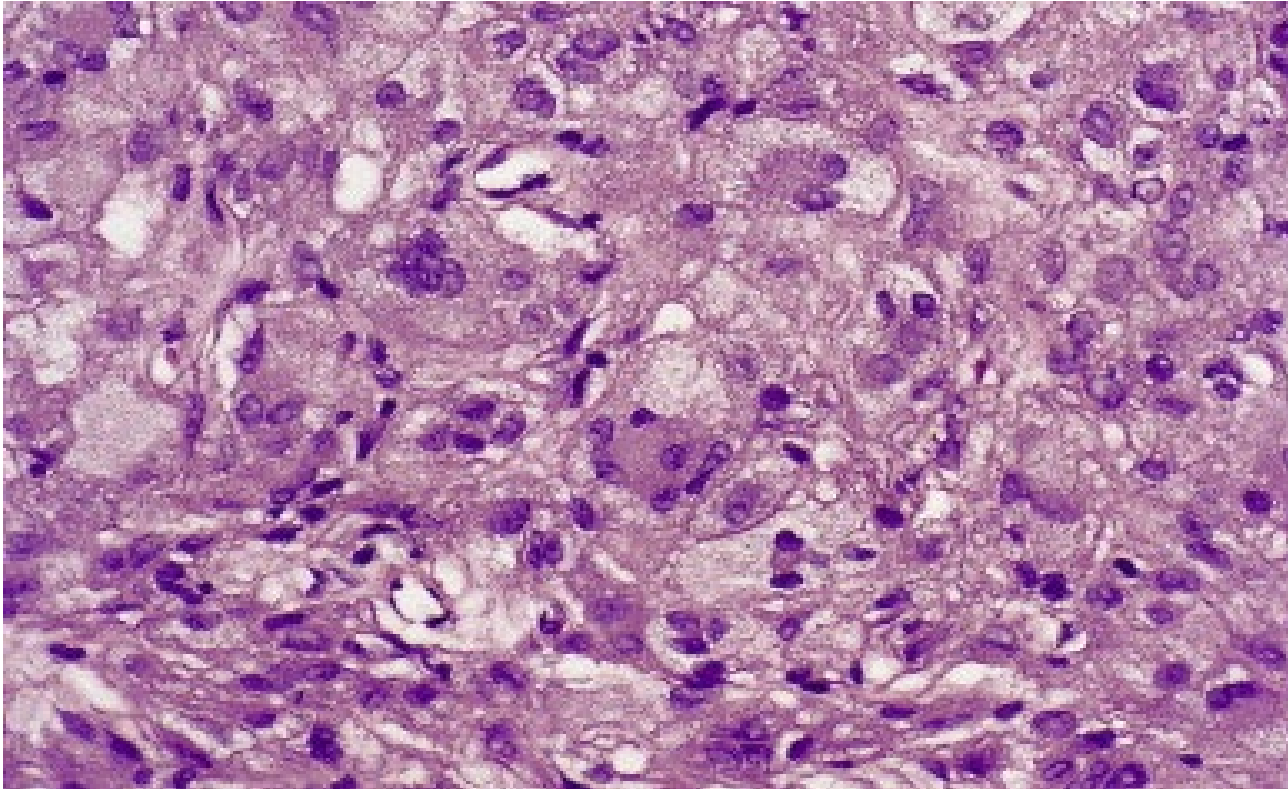
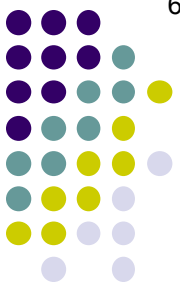
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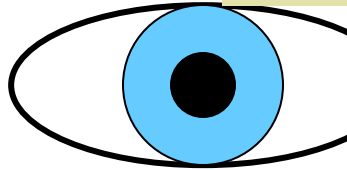




JXG: Foamy macrophages

Q

Heterochromia i  
Divide these



Are Touton giant cells and foamy macrophages present in the AOXs as well?

Touton giant cells  
'foamy' macrophages

LIGHTER iris abnormal

Congenital Horner's

**JXG**

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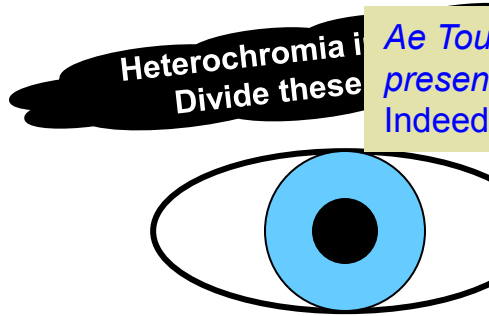
What is the histological

What is the histology?

The presence of...

The presence of...

A



*Are Touton giant cells and foamy macrophages present in the AOXs as well?*  
Indeed they are

**Touton giant cells**  
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LIGHTER iris abnormal

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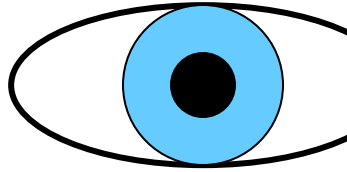
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The presence of...  
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Q

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Divide these...



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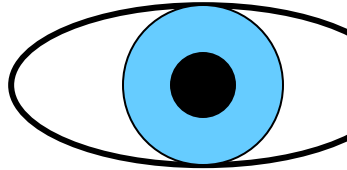
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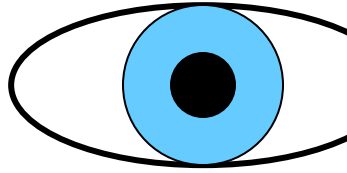
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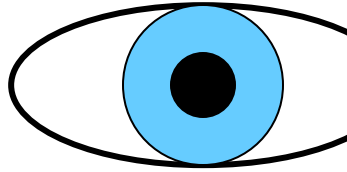
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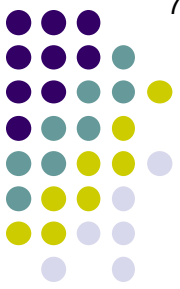
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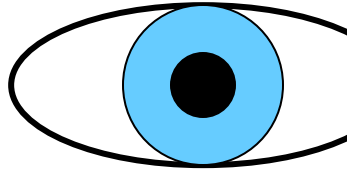


JXG: Skin papules. The orangish color is classic



Q

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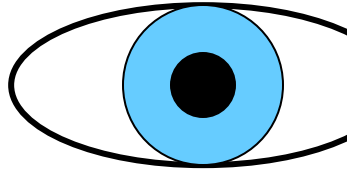
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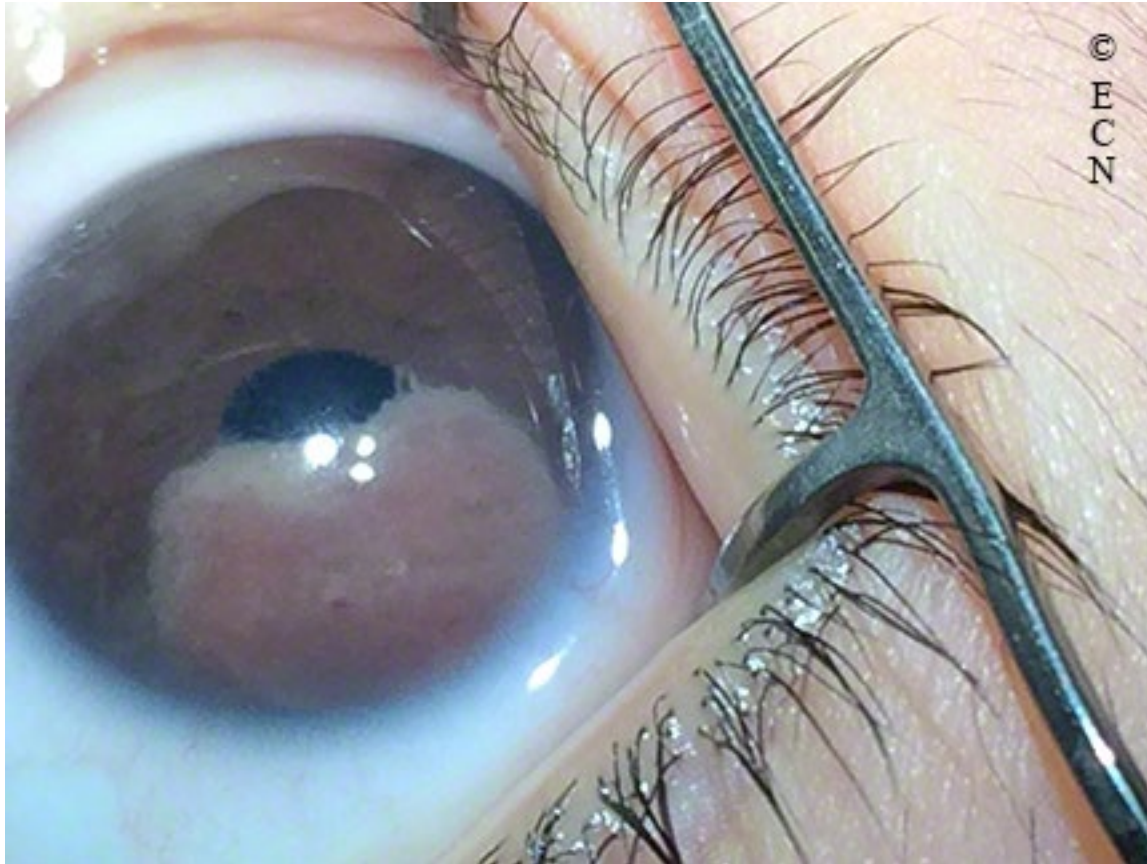
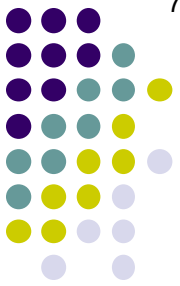
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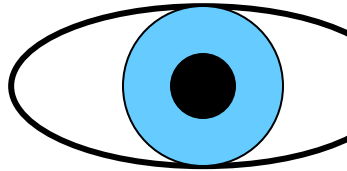
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Unilaterally



JXG: Iris lesion

Q

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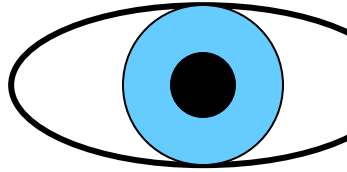
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# Q/A

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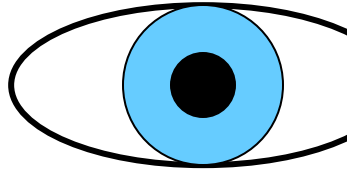
--?

--If enough are present, they will cause

duh (two words)

A

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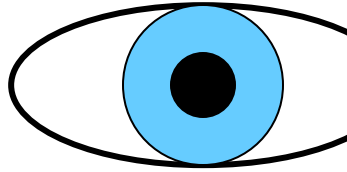
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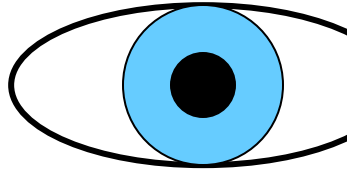
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--?

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--They are prone to spontaneous bleeding

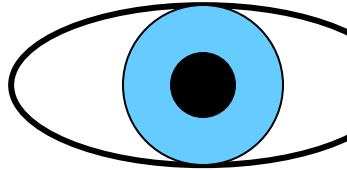
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When iris JXG nodules are present, is it uni-, or bilaterally?  
Unilaterally

In what three ways are the iris nodules clinically significant?  
--They are prone to spontaneous bleeding, with subsequent

bad thing #1

and

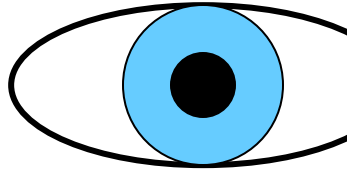
bad thing #2 (two words)

--?

--If enough are present, they will cause heterochromia iridis

A

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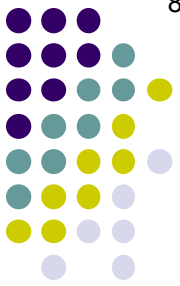
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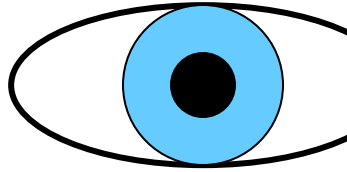
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--?  
--If enough are present, they will cause heterochromia iridis



JXG: Spontaneous hyphema

Q

Heterochromia iridis:  
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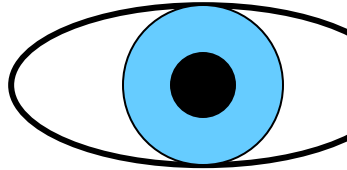
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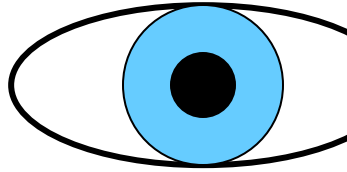
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*How does JXG usually present? (Hint: It's not ophthalmic)*  
As skin papules

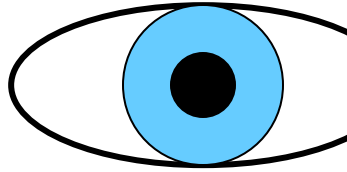
*When iris JXG nodules are present, is it uni-, or bilaterally?*  
Unilaterally

*In what three ways are the iris nodules clinically significant?*  
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma  
--They are in the peds uveitis DDx as a 'masquerade syndrome'  
--If enough are present, they will cause heterochromia iridis

*Should they be removed surgically?*

# Q/A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

**JXG**

Waardenburg syndrome

Incontinentia pigmenti

Fuchs heterochromic iridocycl

*What does JXG stand for?*  
Juvenile xanthogranuloma

*In three words, what sort of condition is it?*  
It is a nonneoplastic histiocytic proliferation

*What are the two hallmarks of JXG histology?*  
The presence of... Touton giant cells  
The presence of... 'foamy' macrophages

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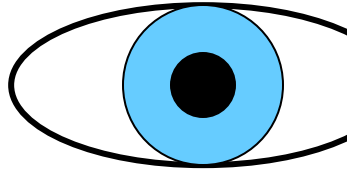
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*Should they be removed surgically?*  
Only if the [redacted] is uncontrollable

A

Heterochromia iridis:  
Divide these...



LIGHTER iris abnormal

Congenital Horner's

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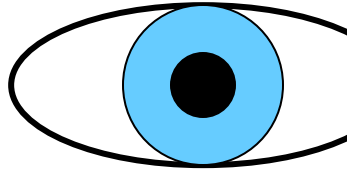
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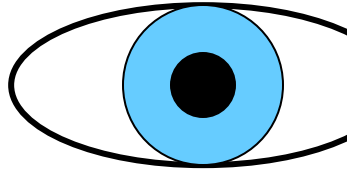
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*Should they be removed surgically?*  
Only if the glaucoma is uncontrollable

*What is the natural history of the disease?*

Q/A

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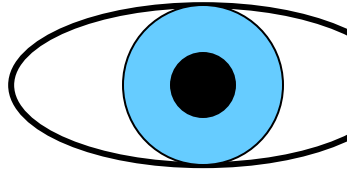
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*Should they be removed surgically?*  
Only if the glaucoma is uncontrollable

*What is the natural history of the disease?*  
JXG is self-limited, usually resolving by age

A

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Divide these...



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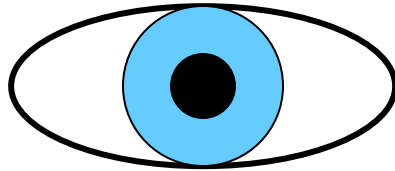
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*Should they be removed surgically?*  
Only if the glaucoma is uncontrollable

*What is the natural history of the disease?*  
JXG is self-limited, usually resolving by age 5 years

Q

Heterochromia iridis:  
Divide these...



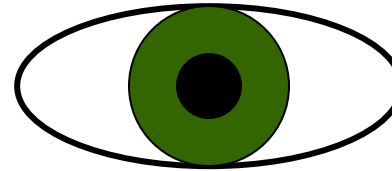
LIGHTER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

...into their respective  
categories



DARKER iris abnormal

Siderosis

Rb

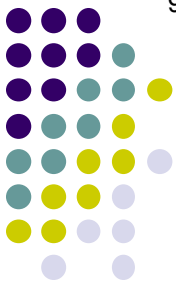
Melanoma

*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

--Heterochromia iridis (duh)

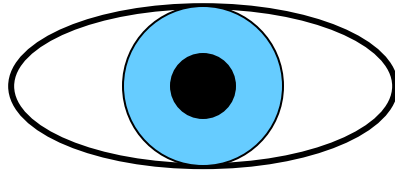
--?

--?



A

Heterochromia iridis:  
Divide these...



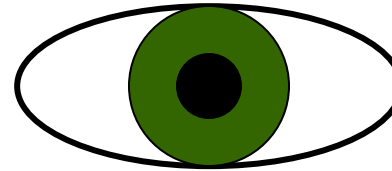
LIGHTER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

...into their respective  
categories



DARKER iris abnormal

Siderosis

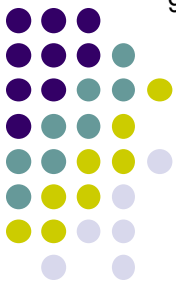
Rb

Melanoma

*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

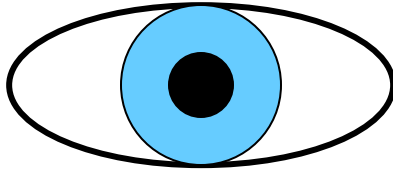
- Heterochromia iridis (duh)
- Synophrys
- Dystopia canthorum

*(Note: Some pts have sectoral fundus pigmentation changes, so that would be a good answer too)*



Q

Heterochromia iridis:  
Divide these...



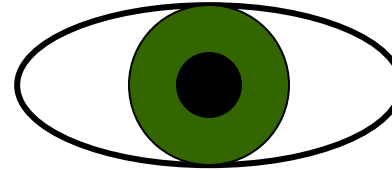
LIGHTER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

...into their respective  
categories



DARKER iris abnormal

Siderosis

Rb

Melanoma

What 3 ophthalmic findings are classic for Waardenburg syndrome?

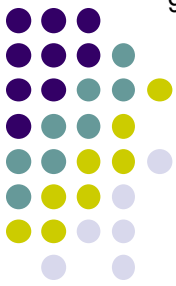
--Heterochromia iridis

--**Synophrys**

--Dystopia canthorum

What is synophrys?

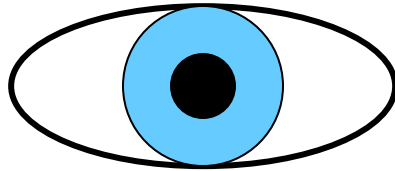
Unilateral Xalatan use  
normal melanocytosis





# A/Q

Heterochromia iridis:  
Divide these...



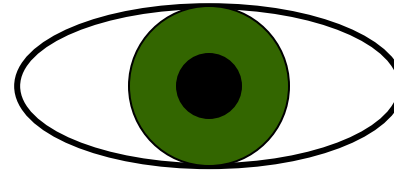
LIGHTER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

...into their respective  
categories



DARKER iris abnormal

Siderosis

Rb

Melanoma

What 3 ophthalmic findings are classic for Waardenburg syndrome?

--Heterochromia iridis

--**Synophrys**

--Dystopia canthorum

What is synophrys?

The formal medical term for a

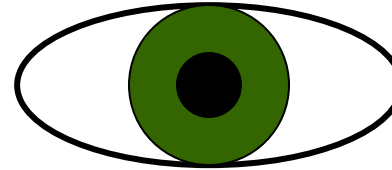
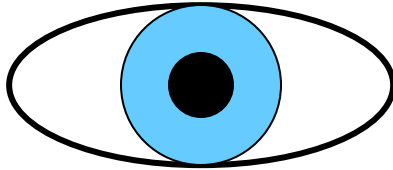


Unilateral Xalatan use  
normal melanocytosis

A

Heterochromia iridis:  
Divide these...

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categories



LIGHTER iris abnormal

DARKER iris abnormal

Congenital Horner's

Siderosis  
Rb

JXG

Melanoma

**Waardenburg syndrome**

What 3 ophthalmic findings are classic for Waardenburg syndrome?

--Heterochromia iridis

--**Synophrys**

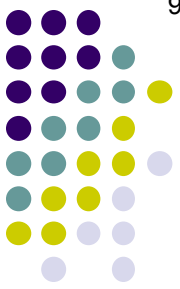
--Dystopia canthorum

What is synophrys?

The formal medical term for a **unibrow**





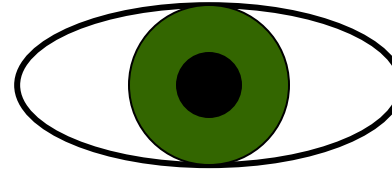
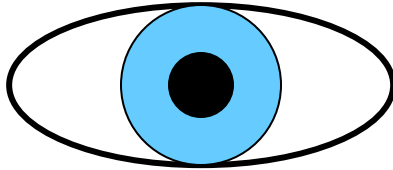


Waardenburg syndrome: Synophrys

Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

Siderosis

Rb

Melanoma

What 3 ophthalmic findings?

--Heterochromia iridis

--Synophrys

--**Dystopia canthorum**

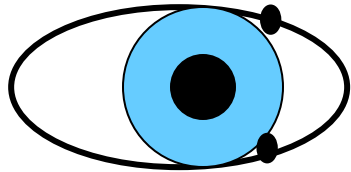
What is dystopia canthorum?

Unilateral Xalatan use

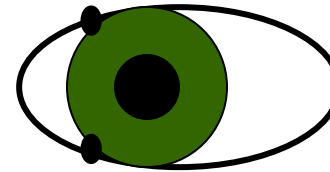
A

Heterochromia iridis:  
Divide these...

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categories



LIGHTER iris abnormal



DARKER iris abnormal

Congenital Horner's

JXG

**Waardenburg syndrome**

Siderosis

Rb

Melanoma

What 3 ophthalmic findings

--Heterochromia iridis

--Synophrys

--**Dystopia canthorum**

What is dystopia canthorum?

Lateral displacement of the canthi (ie, telecanthus) **PLUS** laterally displaced lacrimal puncta



Dystopia canthorum. Note the telecanthus, and laterally displaced lacrimal puncta (also, synophrys)

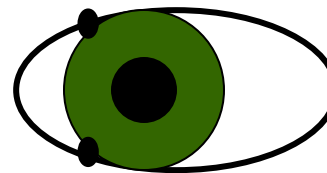
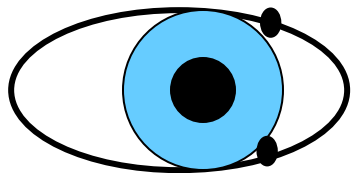


Waardenburg syndrome: Heterochromia iridis,  
synophrys and dystopia canthorum

Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Congenital Horner's

Siderosis  
Rb

Melanoma

JXG

**Waardenburg syndrome**

What 3 ophthalmic findings?

--Heterochromia iridis

--Synophrys

--**Dystopia canthorum**

What is dystopia canthorum?

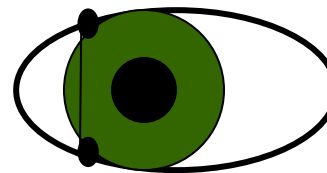
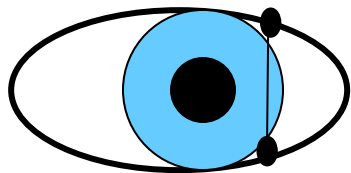
Lateral displacement of the canthi (ie, telecanthus) **PLUS** laterally displaced lacrimal puncta

*How on earth are you supposed to recognize that the puncta are too lateral?*

A

Heterochromia iridis:  
Divide these...

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LIGHTER iris abnormal

DARKER iris abnormal

Congenital Horner's

JXG

Siderosis  
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Melanoma

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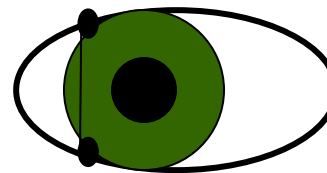
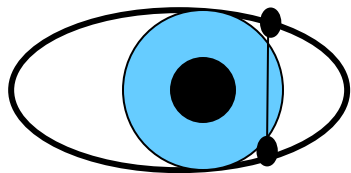
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced. (Next time you examine a pt at the slit-lamp, take note of whether such a line crosses their cornea [it won't].)



Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Siderosis

*What is the difference between telecanthus and hypertelorism?*

*What 3 ophthalmic findings?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

*What is dystopia canthorum?*

Lateral displacement of the canthi (ie, **telecanthus**) PLUS laterally displaced lacrimal puncta

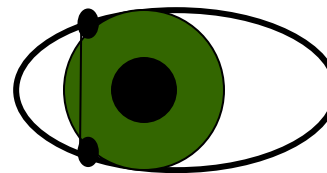
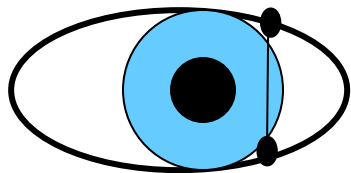
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A/Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Siderosis

*What is the difference between telecanthus and hypertelorism?*

**Telecanthus** refers to an abnormally increased distance between the two words, whereas **hypertelorism** refers to an abnormally increased distance between the three words

*What 3 ophthalmic findings?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

*What is dystopia canthorum?*

Lateral displacement of the canthi (ie, **telecanthus**) **PLUS** laterally displaced lacrimal puncta

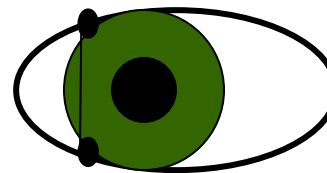
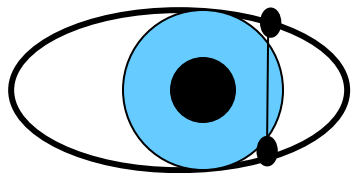
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A

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Siderosis

*What is the difference between telecanthus and hypertelorism?*

**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls

*What 3 ophthalmic findings?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

*What is dystopia canthorum?*

Lateral displacement of the canthi (ie, **telecanthus**) PLUS laterally displaced lacrimal puncta

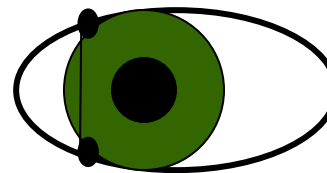
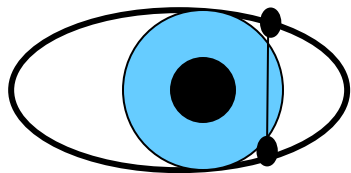
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Q

Heterochromia iridis:  
Divide these...

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categories



LIGHTER iris abnormal

DARKER iris abnormal

Siderosis

*What is the difference between telecanthus and hypertelorism?*

**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls

*Which manifests as an increased interpupillary distance?*

*What 3 ophthalmic findings?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

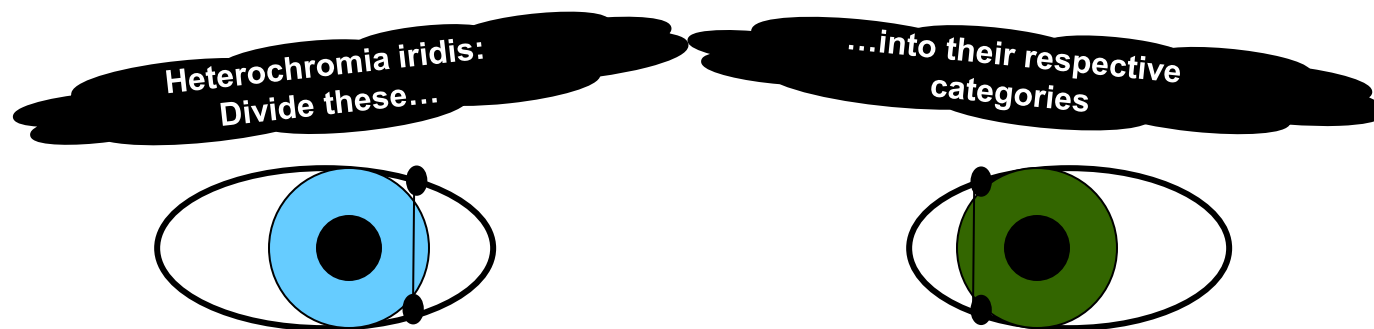
*What is dystopia canthorum?*

Lateral displacement of the canthi (ie, **telecanthus**) PLUS laterally displaced lacrimal puncta

*How on earth are you supposed to recognize that the puncta are too lateral?*

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A



LIGHTER iris abnormal

DARKER iris abnormal

Siderosis

*What is the difference between telecanthus and hypertelorism?*

**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls

*Which manifests as an increased interpupillary distance?*

**Hypertelorism**

*What 3 ophthalmic findings?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

*What is dystopia canthorum?*

Lateral displacement of the canthi (ie, **telecanthus**) PLUS laterally displaced lacrimal puncta

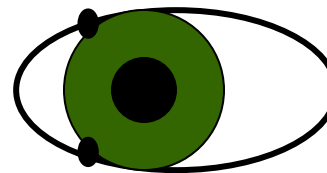
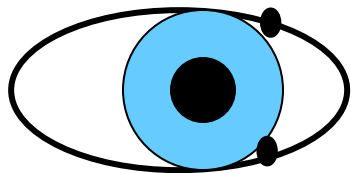
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Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

Congenital Horner's

JXG

Siderosis

Rb

Melanoma

## Waardenburg syndrome

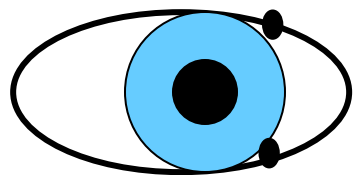
*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

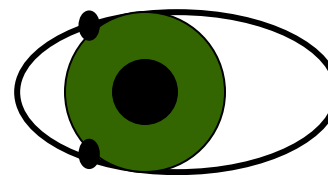
*What non-ophthalmic finding is classic for Waardenburg syndrome?*



A



LIGHTER iris abnormal



DARKER iris abnormal

Congenital Horner's

JXG

Siderosis

Rb

Melanoma

## Waardenburg syndrome

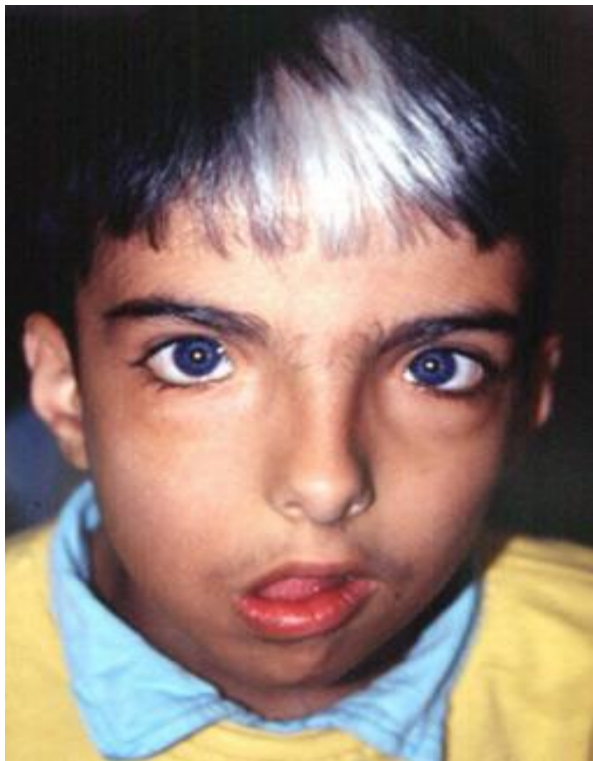
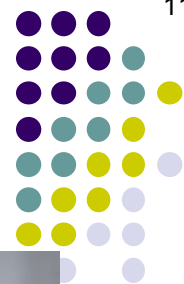
*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

- Heterochromia iridis
- Synophrys
- Dystopia canthorum

*What **non-ophthalmic** finding is classic for Waardenburg syndrome?*

The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

**W**aardenburg syndrome  
hite forelock



Waardenburg syndrome: White forelock



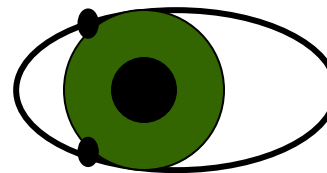
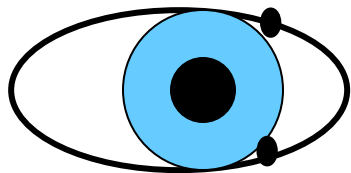
Note that Waardenburg syndrome has forms that do not involve heterochromia



Q

Heterochromia iridis:  
Divide these...

...into their respective  
categories



LIGHTER iris abnormal

DARKER iris abnormal

*What are some of the other non-ophthalmic manifestations of Waardenburg syndrome?*

--?

--?

--?

--(There are a number of others as well)

Siderosis

Rb

Melanoma

## Waardenburg syndrome

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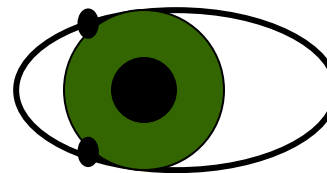
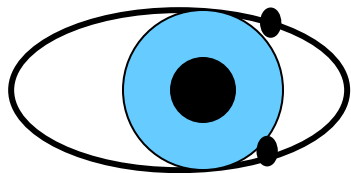


A



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*What are some of the other non-ophthalmic manifestations of Waardenburg syndrome?*

- Hirschprung's disease
- Deafness
- Depigmented patches of skin
- (There are a number of others as well)

Siderosis

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## Waardenburg syndrome

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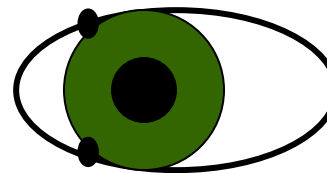
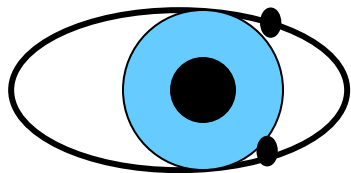
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Unilateral Xalatan use  
normal melanocytosis

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What are some of the other  
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--Hirschprung's disease

--Deafness

--Depigmented patches of skin

--(There are a number of other)

What is the noneponymous name for Hirschprung's disease?

## Waardenburg syndrome

What 3 ophthalmic findings are classic for Waardenburg syndrome?

--Heterochromia iridis

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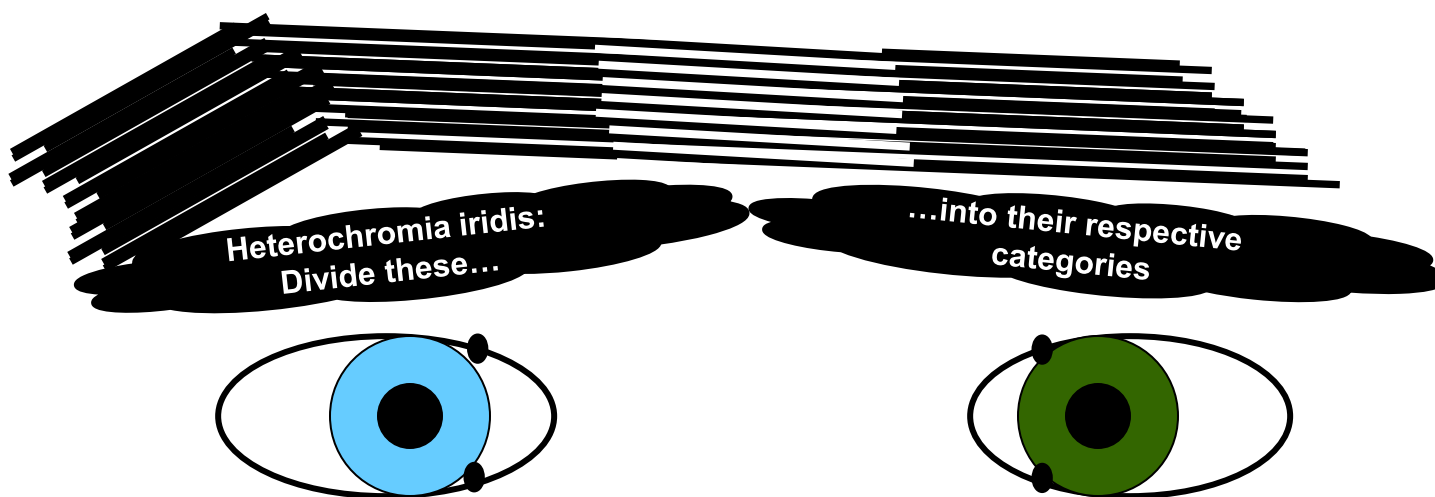
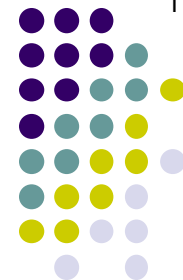
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What non-ophthalmic finding is classic for Waardenburg syndrome?

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A



LIGHTER iris abnormal

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What are some of the other findings in Waardenburg syndrome?

--Hirschprung's disease

--Deafness

--Depigmented patches of skin

--(There are a number of other findings)

What is the noneponymous name for Hirschprung's disease?

Congenital megacolon

## Waardenburg syndrome

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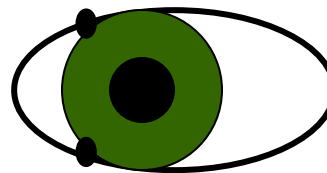
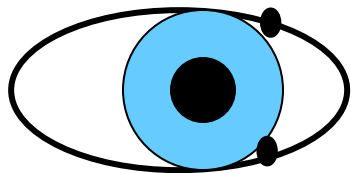
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--(There are a number of other)

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Briefly, what is the pathogenesis and problem?

## Waardenburg syndrome

What 3 ophthalmic findings are classic for Waardenburg syndrome?

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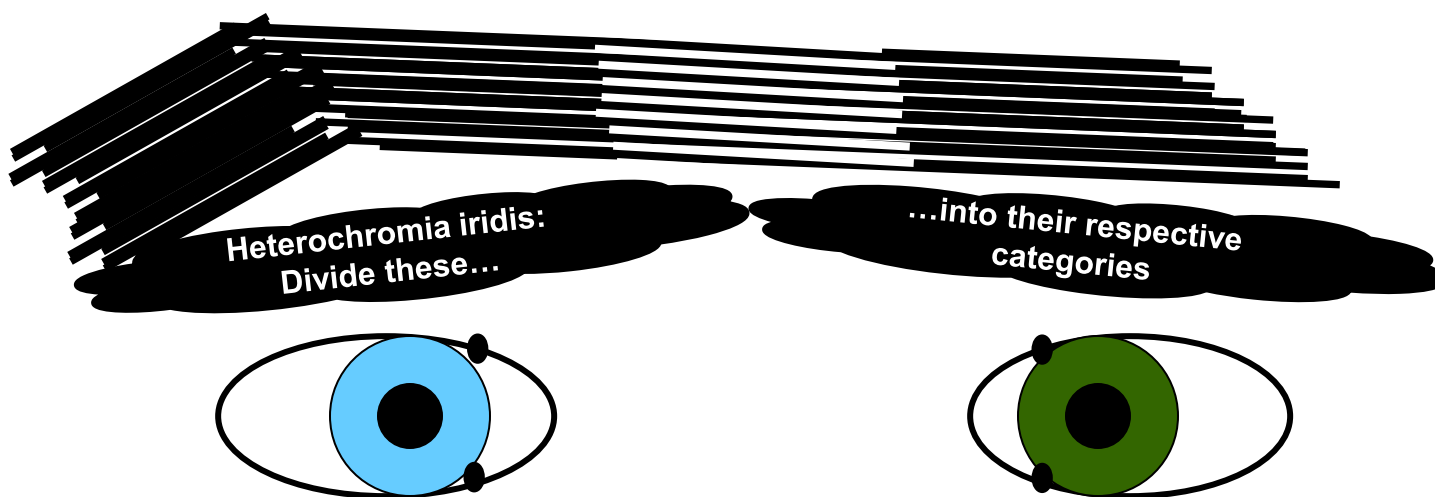
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What are some of the other findings in Waardenburg syndrome?

### --Hirschprung's disease

- Deafness
- Depigmented patches of skin
- (There are a number of other findings)

*What is the noneponymous name for Hirschprung's disease?*

Congenital megacolon

*Briefly, what is the pathogenesis and problem?*

A failure of neural-crest migration leaves a portion of the colon uninnervated and thus nonfunctional. This is a setup for obstruction at that segment.

## Waardenburg syndrome

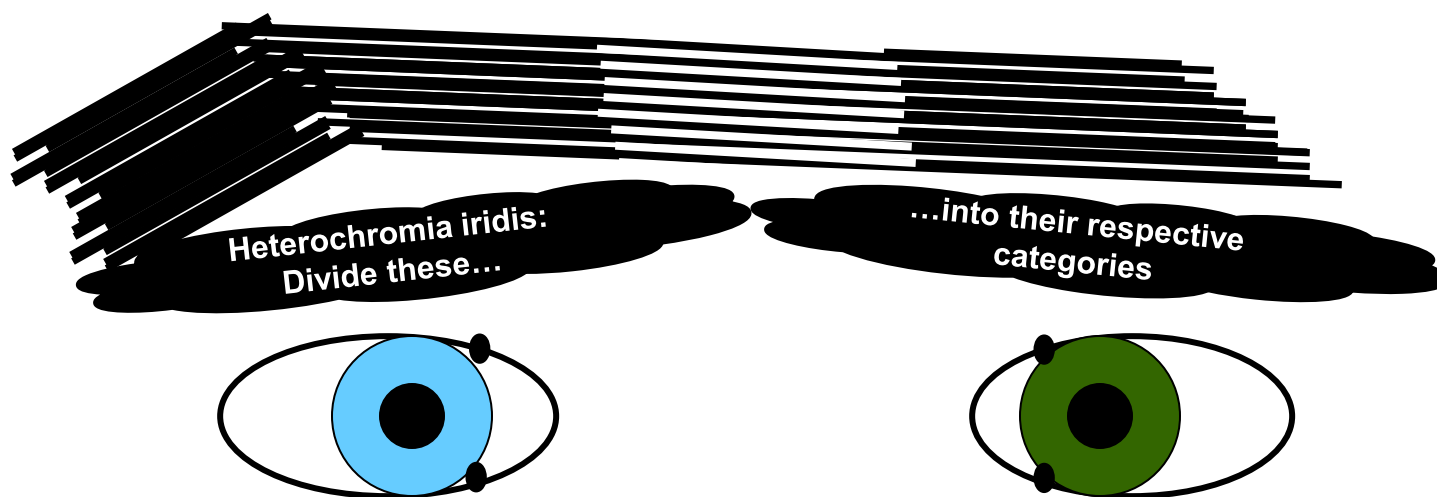
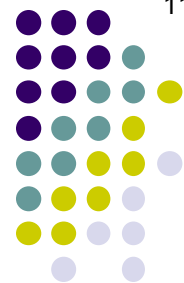
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The presence of a white forelock (i.e., an isolated streak of white hair in the forehead region)

Unilateral Xalatan use  
normal melanocytosis



**LIGHTER** iris abnormal

**DARKER** iris abnormal

What are some of the other non-ophthalmic manifestations of Waardenburg syndrome?

- Hirschprung's disease
- Deafness
- Depigmented patches of skin**
- (There are a number of others as well)

**Waardenburg syndrome**

Siderosis  
Rh

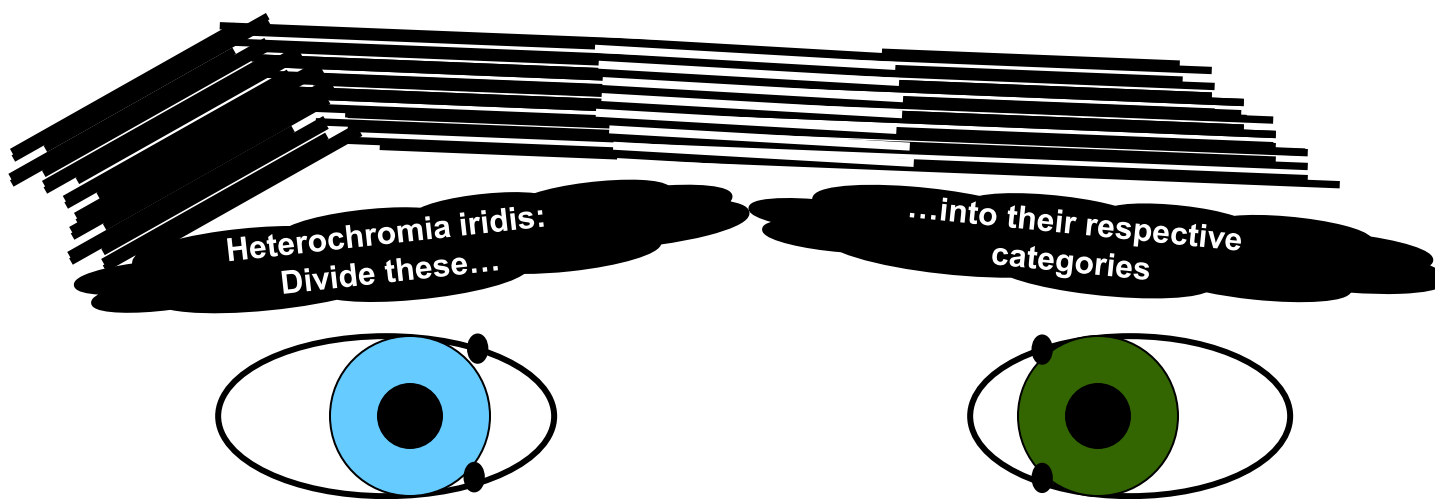
Note that **depigmentation** (ie, of the iris, the skin, the hair, the RPE and choroid) is a dominant and recurring theme in **Waardenburg syndrome**

What 3 ophthalmic findings are classic for Waardenburg syndrome?

- Heterochromia iridis**
- Synophrys
- Dystopia canthorum

**(Some pts have sectoral fundus pigmentation changes)**

What non-ophthalmic finding is classic for Waardenburg syndrome?  
The presence of a white forelock (ie, an isolated streak of **white hair** in the forehead region)



LIGHTER iris abnormal

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## Waardenburg syndrome

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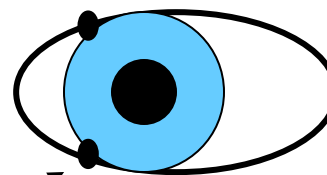
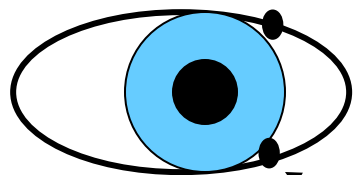
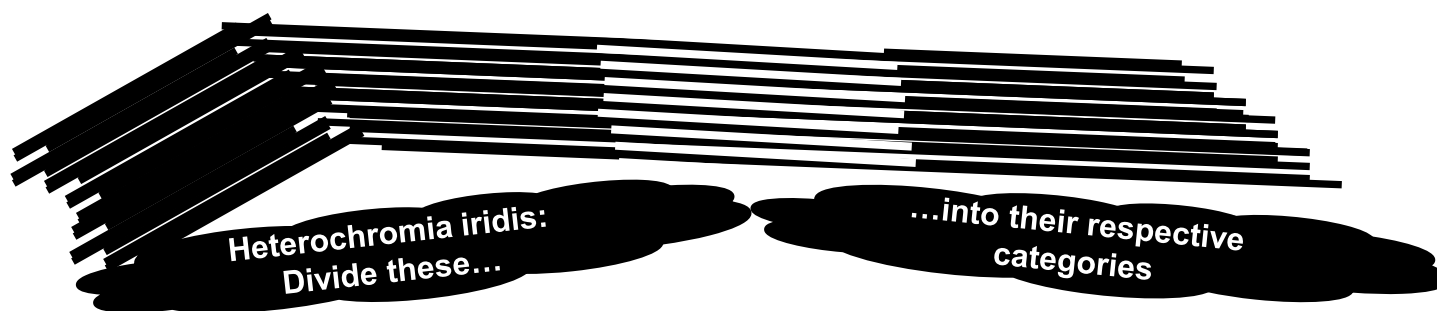
*What non-ophthalmic finding is classic for Waardenburg syndrome?*  
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

Siderosis

And now, a disclaimer:

Waardenburg syndrome is actually a more complex condition than is being presented here. It has four types, two of which each have 2 subtypes. **Not all forms of Waardenburg syndrome include all of the features mentioned here.** That said, after reviewing every mention of the condition in the BCSC books, the info presented in this slide-set is what I *think* a resident is expected to know about the condition. Still, caveat emptor.





LIGHTER iris abnormal

DARKER iris abnormal

**In some forms of Waardenburg syndrome, heterochromia is absent, with light irides present bilaterally**

What are some of the  
Waardenburg syndrome?

- Hirschsprung's disease
- Deafness
- Depigmented patches of skin
- (There are a number of others as well)

## Waardenburg syndrome

What 3 ophthalmic findings are classic for Waardenburg syndrome?

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What non-ophthalmic finding is classic for Waardenburg syndrome?

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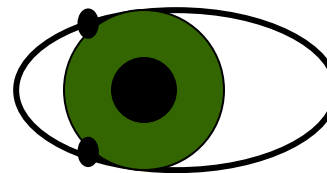
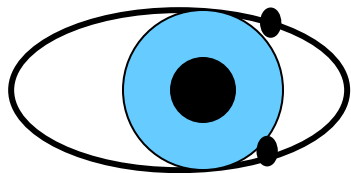
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Q



Heterochromia iridis:  
Divide these...

...into their respective  
categories



**LIGHTER** iris abnormal

**DARKER** iris abnormal

*What are some of the other non-ophthalmic manifestations of Waardenburg syndrome?*

- Hirschprung's disease
- Deafness
- Depigmented patches of skin
- (There are a number of others as well)

Siderosis  
Rb

Melanoma

## Waardenburg syndrome

*What gene is responsible for Waardenburg syndrome?*

*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

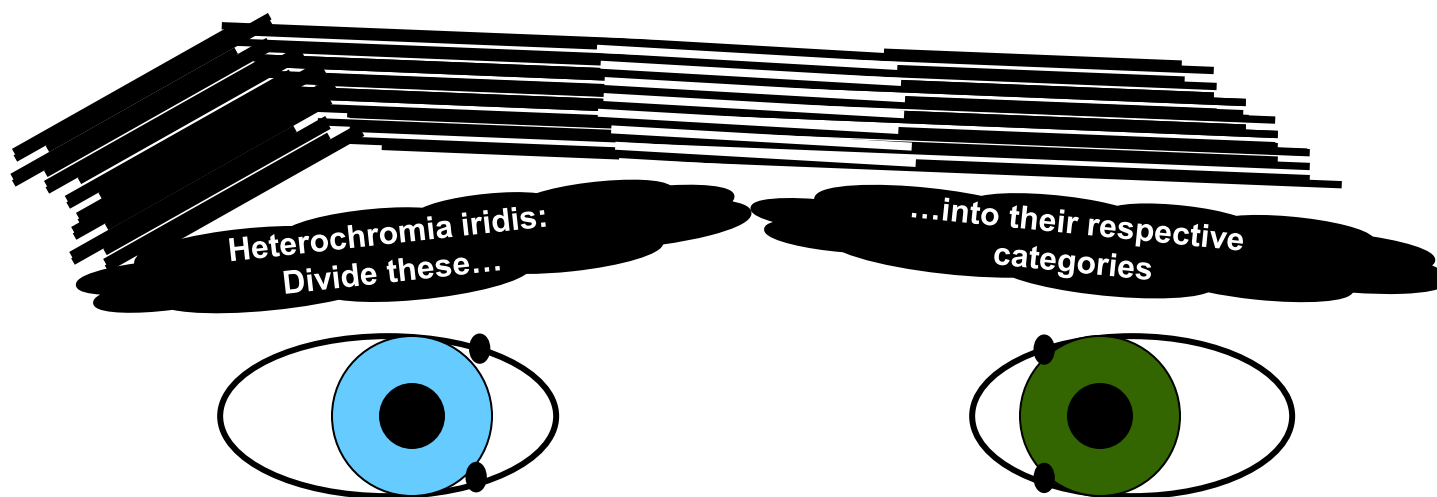
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...oral Xanthan use  
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A



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- Hirschprung's disease
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Siderosis  
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## Waardenburg syndrome

*What gene is responsible for Waardenburg syndrome?*  
PAX3

*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

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- Synophrys
- Dystopia canthorum

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*What non-ophthalmic finding is classic for Waardenburg syndrome?*  
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

Q

*In the present context, what is the origin of the word PAX, ie, where does it come from?*

LIGH

What are some features of Waardenburg syndrome?

- Hirschsprung
- Deafness
- Depigmentation
- (There are)

## Waardenburg syndrome

**PAX3**

*What 3 ophthalmic findings are classic for Waardenburg syndrome?*

- Heterochromia iridis
- Synophrys
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The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

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In the present context, what is the origin of the word PAX, ie, where does it come from?  
 It is a portmanteau of the terms 'PAired homeoboX' and/or 'PAired boX'

LIGH

What are some clinical features of Waardenburg syndrome?  
 --Hirschsprung disease  
 --Deafness  
 --Depigmentation  
 --(There are more)

## Waardenburg syndrome

PAX3

What 3 ophthalmic findings are classic for Waardenburg syndrome?

- Heterochromia iridis
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The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

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*In two words, what is the role of the proteins coded for by PAX genes?*

They are

LIGH

What are so

Waardenbu

--Hirschprun

--Deafness

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## Waardenburg syndrome

PAX3

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LIGH

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## Waardenburg syndrome

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Several variants use  
 normal melanocytes

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## Waardenburg syndrome

**PAX3**

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Other X-linked use  
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Morphogenesis

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Waardenbu

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## Waardenburg syndrome

**PAX3**

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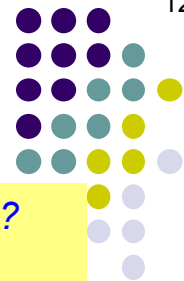
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*What are the three PAX genes of concern to us eye dentists?*

?, PAX3, and ?

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## Waardenburg syndrome

PAX3

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The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

Other clinical findings include  
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PAX2, PAX3, and PAX6

## Waardenburg syndrome

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*Of these three PAX genes, which is most important to the development of the eye?*

## Waardenburg syndrome

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PAX2, PAX3, and PAX6

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**PAX6**. The *Fundamentals* book refers to it as "the master switch for eye development."

The *Peds* book says, "The *PAX6* gene is the master control gene for eye morphogenesis."

## Waardenburg syndrome

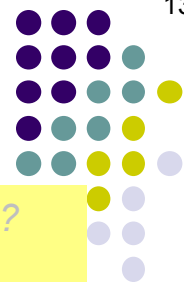
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**For more on PAX genes, see slide-set FELT4**

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