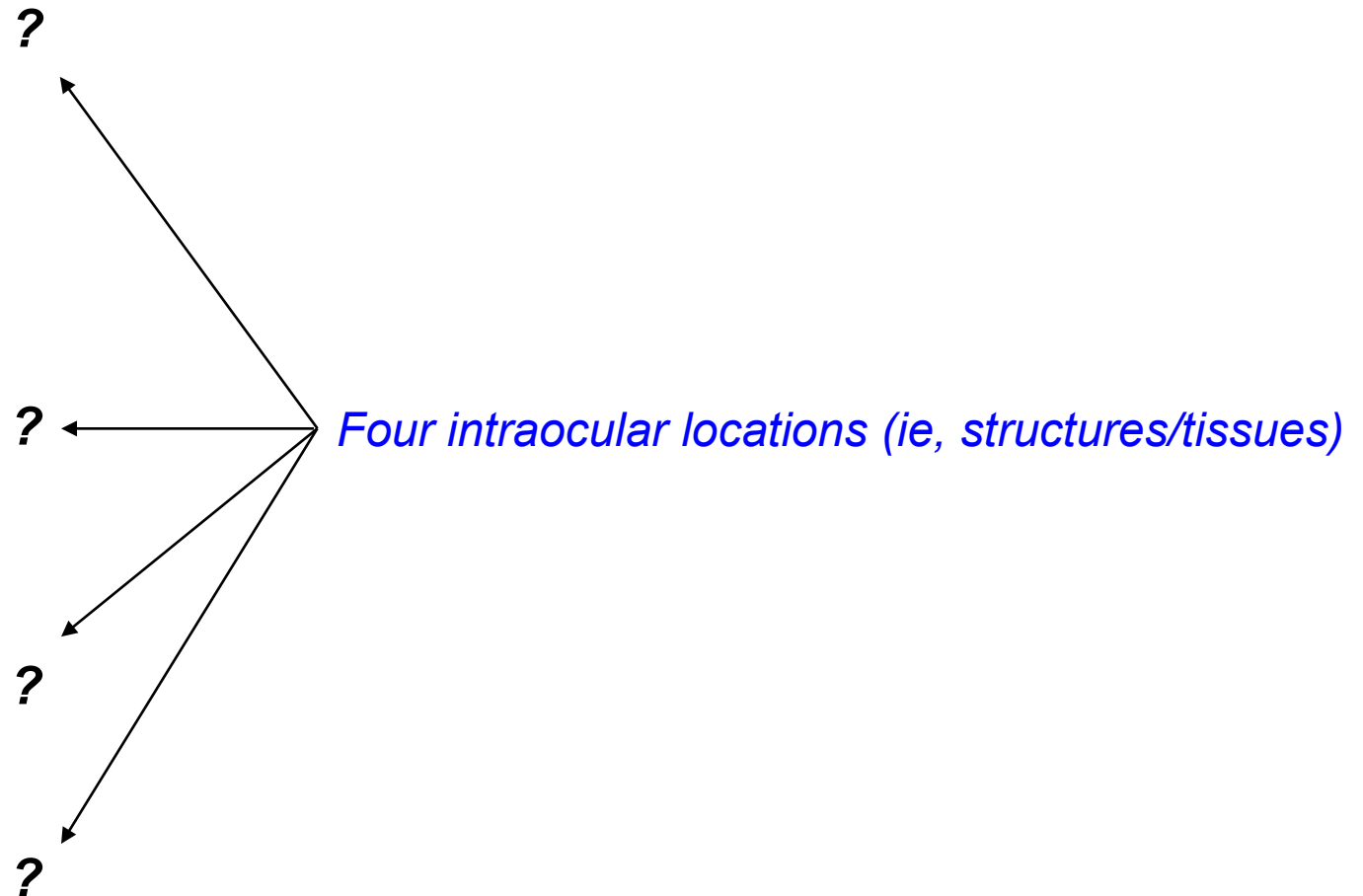


Before you begin: This is a big topic, and big topics beget big slide-sets.  
There's a natural break near the halfway mark (slide 213); I placed a  
*break time!* slide at that point to mark it.

# Intraocular Tumors of Childhood



# Intraocular Tumors of Childhood



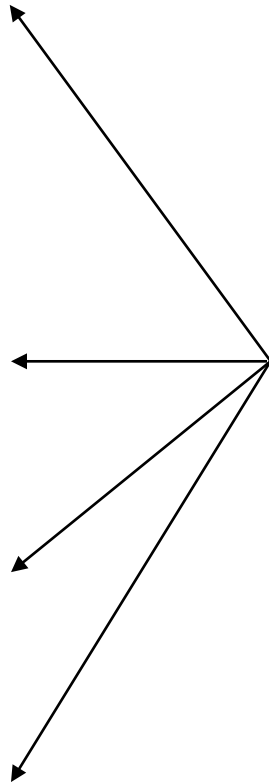
**Iris/Ciliary Body**

**Choroid**

**RPE**

**Retina**

*Four intraocular locations (ie, structures/tissues)*





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

1) ?

2) ?

3) ?

4) ?

5) ?

6) ?

*Six tumors of the iris/ciliary body*

Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) Juvenile xanthogranuloma
- 2) Medulloepithelioma
- 3) Lisch nodules
- 4) Brushfield spots
- 5) Iris mammillations
- 6) Iris cysts

*Six tumors of the iris/ciliary body*

Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

*In three words, what sort of condition is JXG?*  
It is a...

Iris/Ciliary Body

Choroid

RPE

Retina

# Intraocular Tumors of Childhood



## 1) Juvenile xanthogranuloma

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

Iris/Ciliary Body

Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

Choroid

RPE

Retina





# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

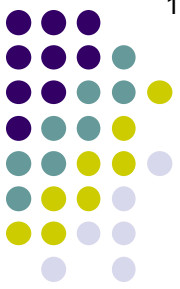
As orangish skin papules

Choroid

RPE

Retina

# Intraocular Tumors of Childhood



JXG: Skin papules. The orangish color is classic



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The majority before age 1 year, and almost all by

Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

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Choroid

RPE

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

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As orangish skin papules

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The majority before age 1 year, and almost all by 2

*When JXG iris nodules are present, are they uni-, or bilateral?*

### Choroid

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

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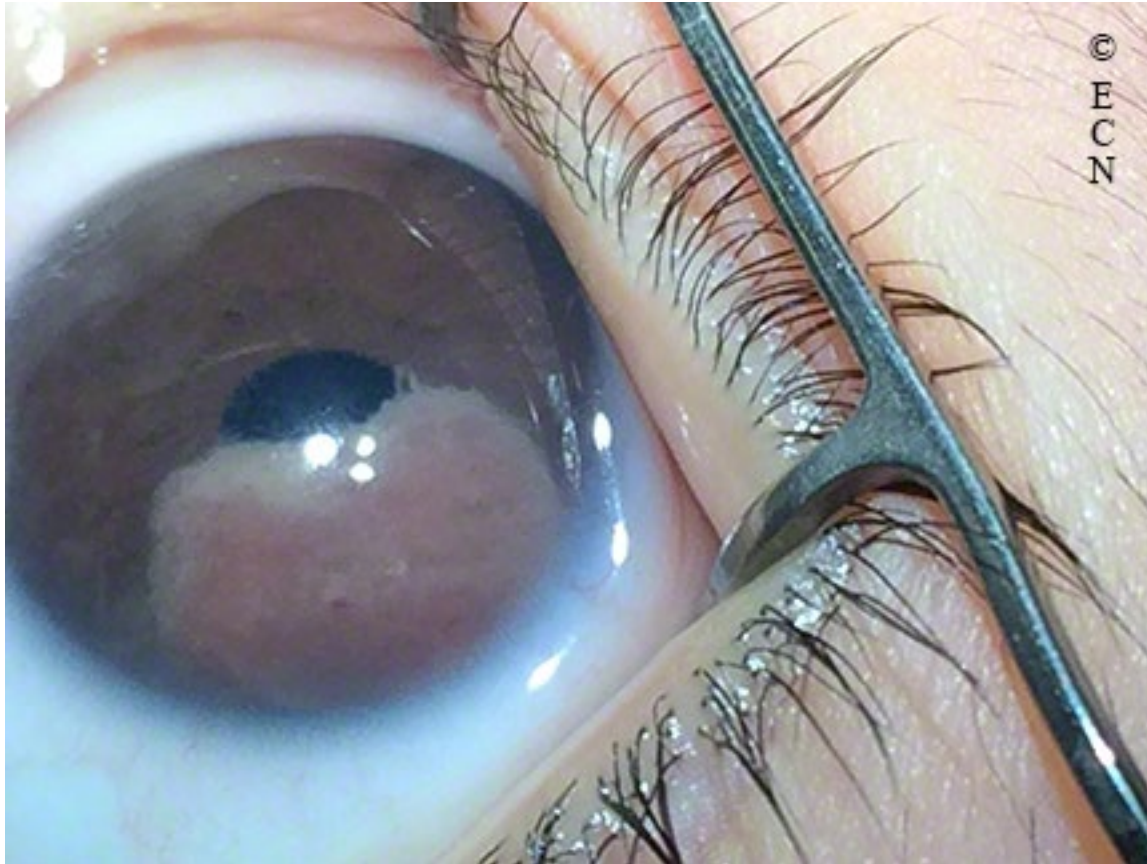
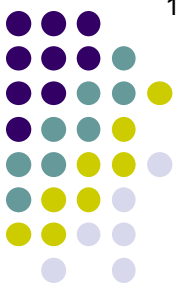
Unilateral

### Choroid

### RPE

### Retina

# Intraocular Tumors of Childhood



JXG: Iris lesion





# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The majority before age 1 year, and almost all by 2

*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

--?

--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

--They are prone to spontaneous  

--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The majority before age 1 year, and almost all by 2

*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

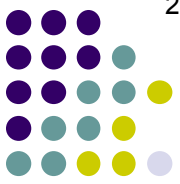
--They are prone to spontaneous bleeding

--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

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*At what age does it present?*

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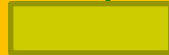
*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

--They are prone to spontaneous bleeding, with subsequent

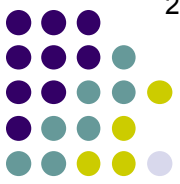


--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

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*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

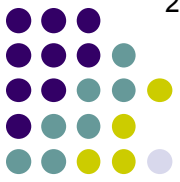
--They are prone to spontaneous bleeding , with subsequent hyphema

--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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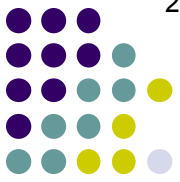
--They are prone to spontaneous bleeding , with subsequent hyphema and

--?

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

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### Choroid

*In what three ways are the iris nodules clinically significant?*

--They are prone to spontaneous bleeding, with subsequent hyphema **and secondary glaucoma**

--?

--?

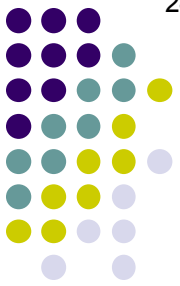
### RPE

### Retina

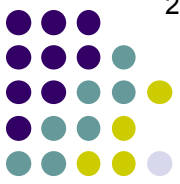
# Intraocular Tumors of Childhood



JXG: Spontaneous hyphema







# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

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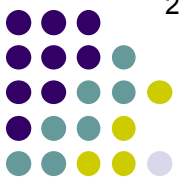
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma

--They are in the DDx as a two words in peds uveitis

--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

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Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

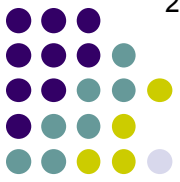
--They are prone to spontaneous bleeding, with subsequent hyphema **and secondary glaucoma**

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--?

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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*In three words, what sort of condition is JXG?*

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*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

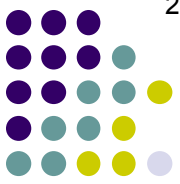
--They are prone to spontaneous bleeding, with subsequent hyphema and **secondary glaucoma**

--They are in the DDx as a *masquerade syndrome* in peds uveitis

--If enough nodules are present, two words will result

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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*In three words, what sort of condition is JXG?*

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Unilateral

### Choroid

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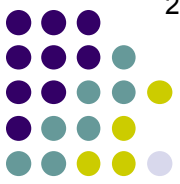
--They are prone to spontaneous bleeding, with subsequent hyphema **and secondary glaucoma**

--They are in the DDx as a *masquerade syndrome* in peds uveitis

--If enough nodules are present, heterochromia iridis will result

### RPE

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

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*At what age does it present?*

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*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

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*In what three ways are the iris nodules clinically significant?*

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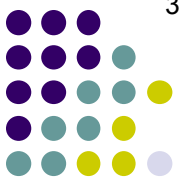
--They are in the DDx as a *masquerade syndrome* in peds uveitis

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### RPE

*What is the natural history of JXG?*

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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Unilateral

### Choroid

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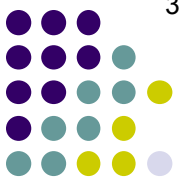
--If enough nodules are present, heterochromia iridis will result

### RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a...**nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The majority before age 1 year, and almost all by 2

*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

--They are prone to spontaneous bleeding, with subsequent hyphema **and secondary glaucoma**

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--If enough nodules are present, heterochromia iridis will result

### RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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The majority before age 1 year, and almost all by 2

*When JXG iris nodules are present, are they uni-, or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

—They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma

***Should JXG nodules be removed surgically?***

—Juvenile xanthogranuloma (JXG) can cause a severe form of uveitis called "Mutter's syndrome" in which the iris nodules can lead to heterochromia iridis. This will result in a change in the color of the iris.

### RPE

*What is the natural history of JXG?*

**It is self-limited**, usually resolving by age 5 years

### Retina





# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

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Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

—They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma

***Should JXG nodules be removed surgically?***

**Only if the glaucoma is uncontrollable**

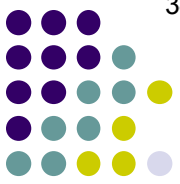
Werde syndrome in peds uveitis  
heterochromia iridis will result

### RPE

*What is the natural history of JXG?*

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### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The most common age is 1-2 years. **What are the two hallmarks of JXG histology?**

*When is it unilateral/bilateral?*

Unilateral

### Choroid

*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 DDx as a *masquerade syndrome* in peds uveitis

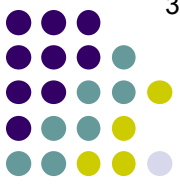
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*What is the natural history of JXG?*

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### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The most common question: **What are the two hallmarks of JXG histology?**

The presence of **eponym** giant cells

*When does it present? The presence of... (we'll come back to this one) lateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

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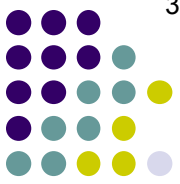
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### RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The most common age is 1 year. **What are the two hallmarks of JXG histology?**

The presence of Touton giant cells

*When does it present? Unilateral or bilateral?*

Unilateral

### Choroid

*In what three ways are the iris nodules clinically significant?*

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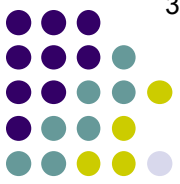
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### RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The most common age is 1-2 years

*What are the two hallmarks of JXG histology?*

The presence of Touton giant cells and foamy xanthoma cells

**giant cells**

*What is a giant cell?*

C

RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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**giant cells**

*What is a giant cell?*

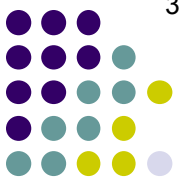
It is an aggregate of

### RPE

*What is the natural history of JXG?*

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### Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

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*At what age does it present?*

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The presence of Touton giant cells

**giant cells**

*What is a giant cell?*

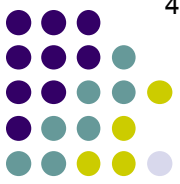
It is an aggregate of histiocytes

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It is self-limited, usually resolving by age 5 years

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# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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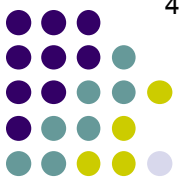
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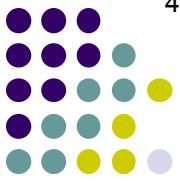
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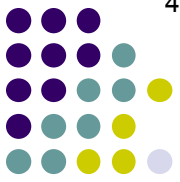
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It is *multinucleated*—the myriad nuclei of the involved histiocytes are all visible within it

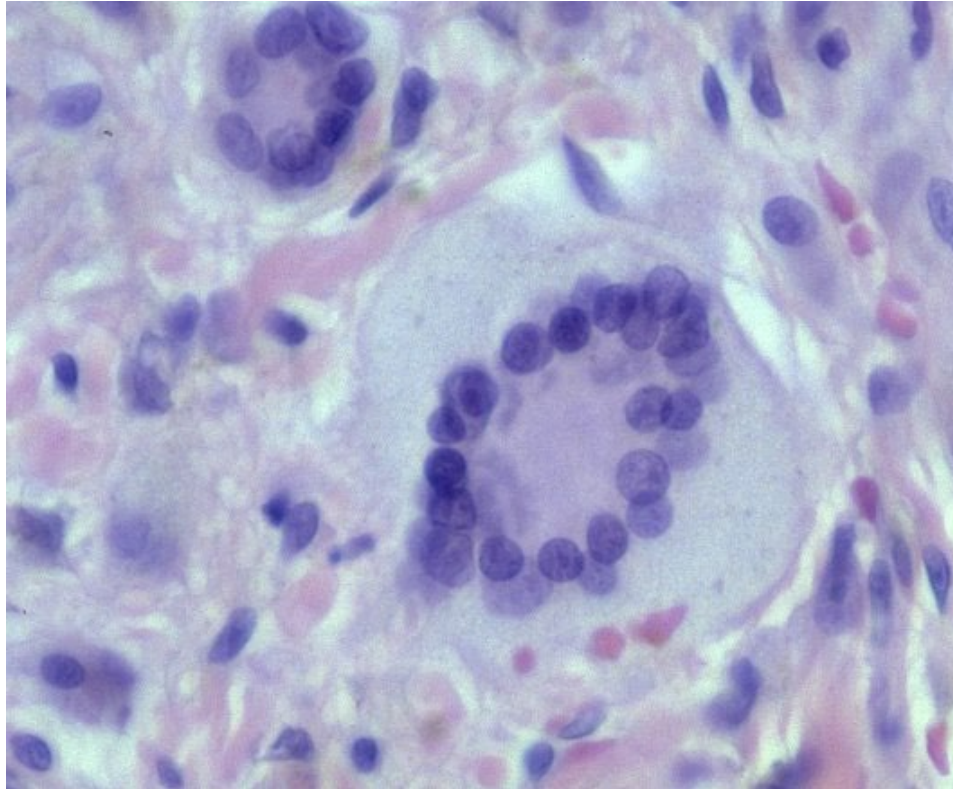
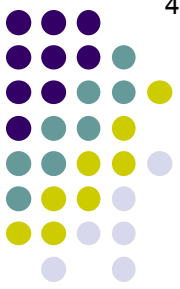
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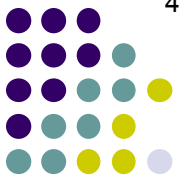
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# Intraocular Tumors of Childhood



JXG: Touton giant cells



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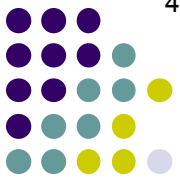
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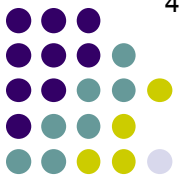
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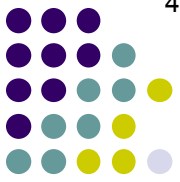
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Monocytes are found within the vasculature. **When a monocyte leaves the circulation and settles down within tissue, it becomes (ie, is re-classified) as a histiocyte.**

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The presence of Touton giant cells

When does it present? **The presence of...** **two words** **lateral?**

Unilateral

### Choroid

*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 DDx as a *masquerade syndrome* in peds uveitis

—If enough nodules are present, heterochromia iridis will result

### RPE

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The presence of... **Tanaka giant cells**

When does it present? The presence of... **'foamy macrophages'** lateral?

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**Choroid** This histology—'foamy macrophages'—is often described with other, equivalent terms. What are they?

Foamy = ?

Macrophages = ?

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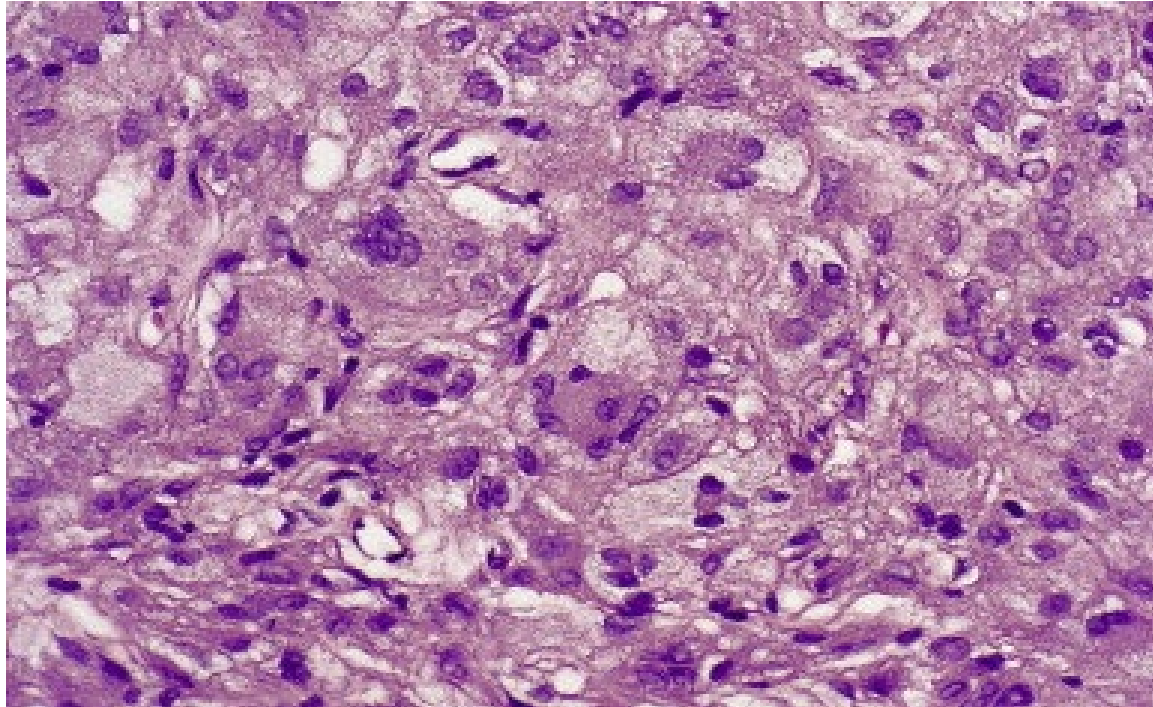
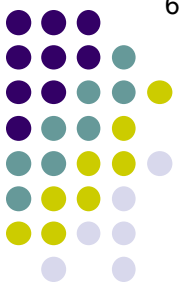
**Macrophages = 'histiocytes'**

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*The point being, the terms 'foamy macrophages,' 'lipid-filled (or -laden) macrophages,' 'foamy histiocytes,' etc, all mean the same thing, so don't be misled if you see one term when you're expecting another*

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JXG: Foamy macrophages



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*Associated with chronic diarrhea? ← Need another?*

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*And CNS symptoms--seizures, dementia, coma? ← Last chance—answer is next!*

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**Whipple's disease**

**Retina**



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**Whipple's disease**

*Broadly speaking, what sort of condition is Whipple's?*

It is an infection

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*Speaking of 'foamy macrophages'...*

*What dz comes to mind if, instead of a toddler*

*with iris nodules, the pt in question was a*

*middle-aged white guy with bilateral panuveitis?*

*And a hx of chronic migratory arthritis?*

*Associated with chronic...?*

*And CNS symptoms--s...*

**Whipple's disease**

### Retina

*Broadly speaking, what sort of condition is Whipple's?*

It is an infection

*What infection agent is responsible for Whipple's?*



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

### Iris/Ciliary Body

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*How does JXG usually present? (Hint: It's not ophthalmic)*

As orangish skin papules

*At what age does it present?*

The most common age is... **What are the two hallmarks of JXG histology?**

The presence of Touton giant cells

When does it present? **'foamy macrophages'** lateral?

Unilateral

### Choroid

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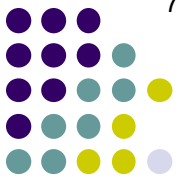
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*What infection agent is responsible for Whipple's?*

The bacterium *Tropheryma whipplei*



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with iris nodules, the pt is a  
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And a hx of chronic migraines  
Associated with chronic inflammation  
And CNS symptoms--s

**Whipple's disease**

### Retina

*When foamy macrophages are found in a biopsy performed on a Whipple's pt, from what site was the biopsy collected?*

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# Intraocular Tumors of Childhood

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*And a hx of chronic mi*

*Associated with chroni*

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**Whipple's disease**

### Retina

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The duodenum (remember, they have GI issues)

*Broadly speaking, what sort of condition is Whipple's?*

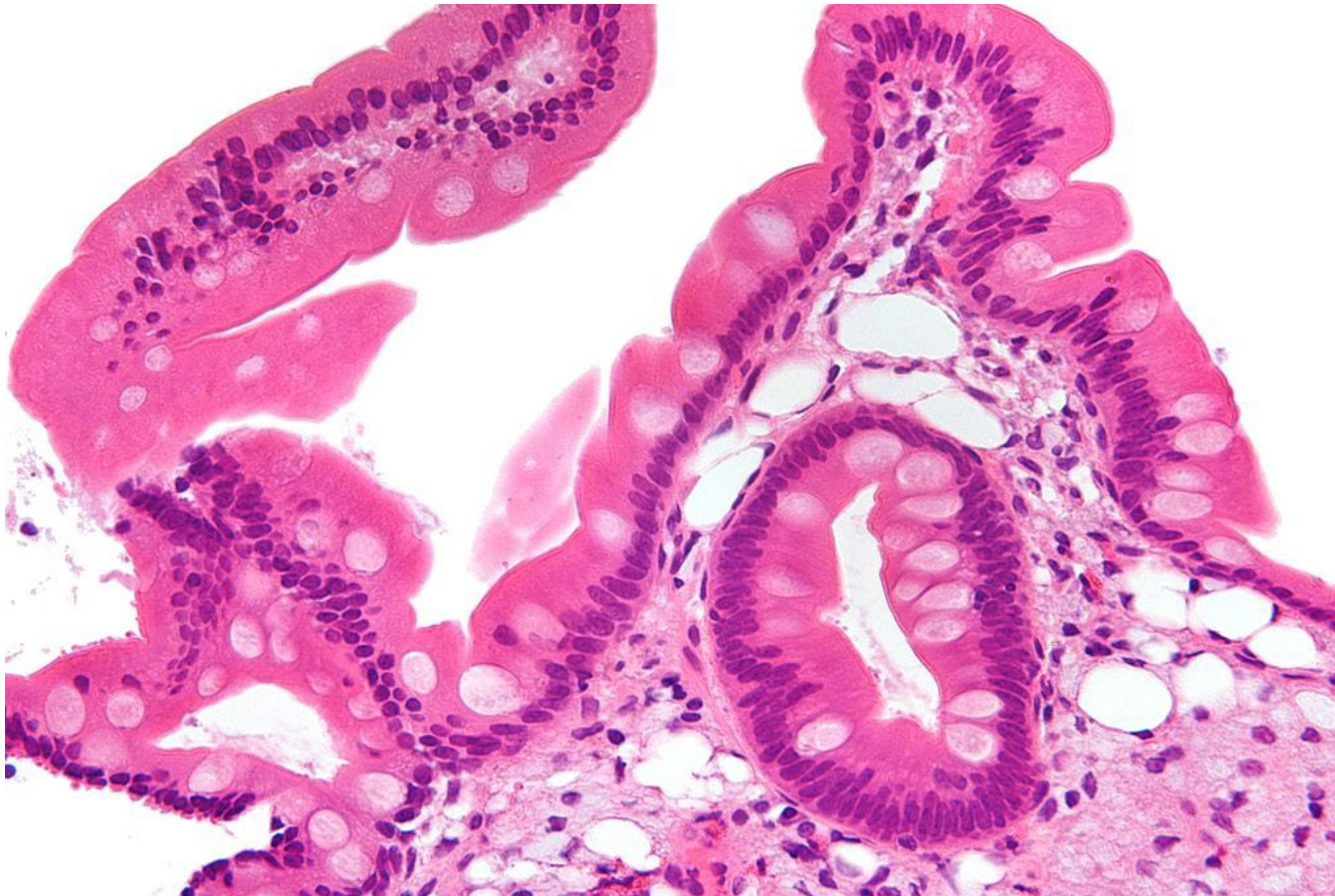
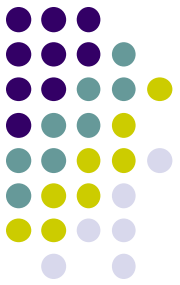
It is an infection

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# Intraocular Tumors of Childhood



Whipple's disease: Duodenal biopsy, high mag. The image shows the characteristic feature of foamy macrophages in the lamina propria.



# Intraocular Tumors of Childhood

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And a hx of chronic migraines

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And CNS symptoms--s

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The presence of **stain** bacteria within macrophages located in intestinal villi

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### Choroid

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*And a hx of chronic mi*

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**Whipple's disease**

### Retina

*When foamy macrophages are found in a biopsy performed on a Whipple's pt, from what site was the biopsy collected?*

The duodenum (remember, they have GI issues)

*What other finding will a duodenal biopsy reveal?*

The presence of PAS+ bacteria within macrophages located in intestinal villi

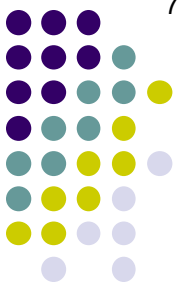
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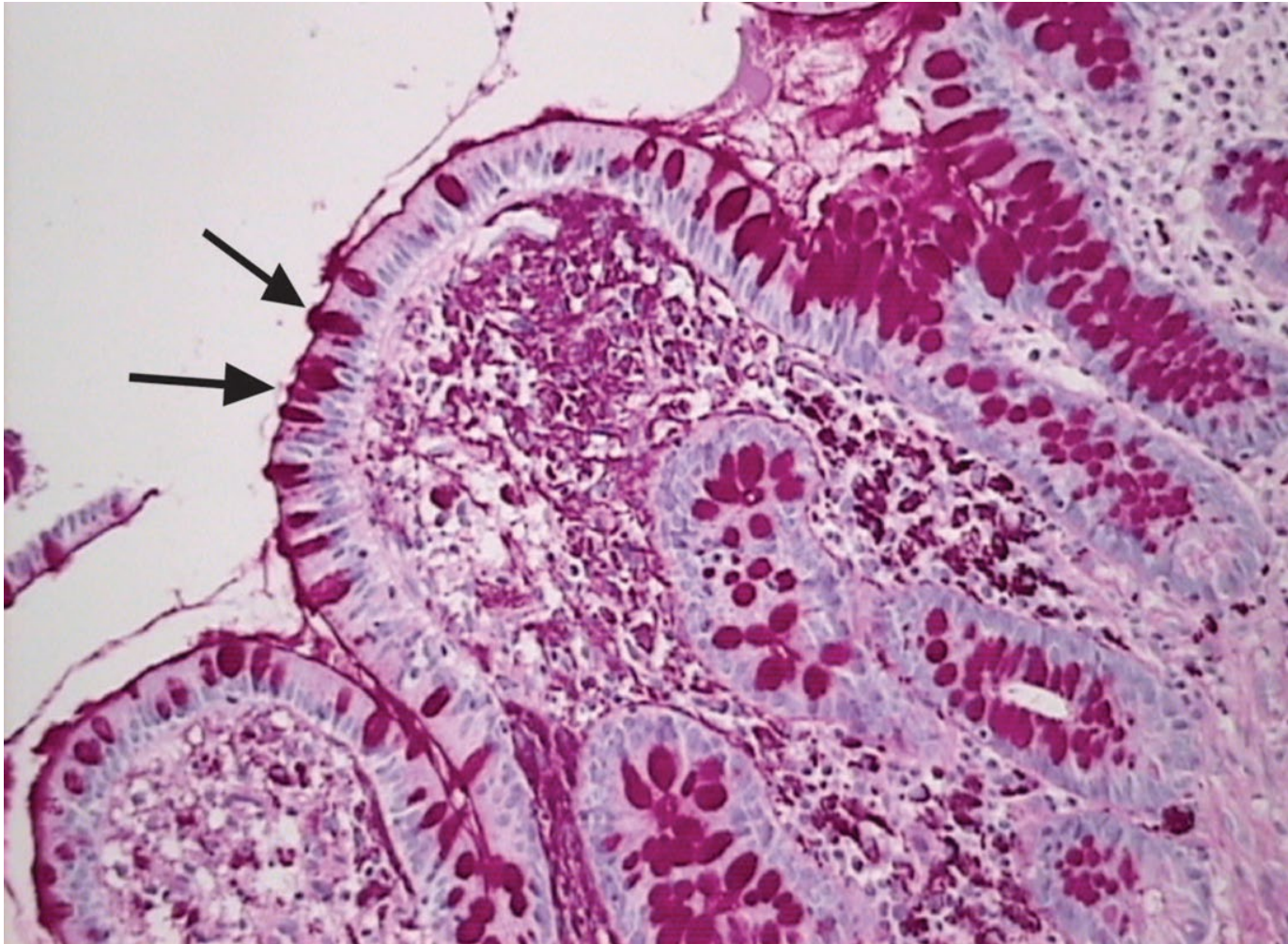
*What infection agent is responsible for Whipple's?*

**The bacterium *Tropheryma whipplei***





# Intraocular Tumors of Childhood



Small-intestine biopsy stained with periodic acid-Schiff. Note the numerous macrophages in the lamina propria (arrows).



# Intraocular Tumors of Childhood

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***For more on Whipple's dz, see slide-set U24***

Chorioid

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# Intraocular Tumors of Childhood

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*What dz comes to mind if, instead of a toddler with iris nodules, the pt in question was a*

*Adult with bilateral upper-lid yellow lesions?*

*(Pic forthcoming—give the dx after seeing it)*

The presence of Touton giant cells  
When The presence of **'foamy macrophages'** lateral?  
Unilateral

**Choroid**

*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 DDx as a 'masquerade syndrome' in peds uveitis

—If enough nodules are present, heterochromia iridis will result

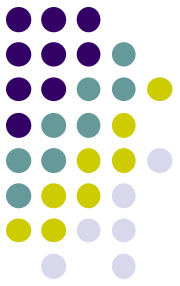
**RPE**

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

**Retina**

# Intraocular Tumors of Childhood



***Condition?***





# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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**Xanthelasma**

The presence of Touton giant cells

When The presence of **'foamy macrophages'** lateral?  
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**Xanthelasma**

The presence of Touton giant cells  
When The presence of **'foamy macrophages'** lateral?  
Unilateral

**Choroid**

*Are xanthelasmas a harbinger of elevated serum lipids?*

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## Choroid

*Are xanthelasmas a harbinger of elevated serum lipids?*

They can be, but in most cases the individual has normal lipid panels

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*Can they be congenital?*

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**Xanthelasma**

*When The presence of Touton giant cells is unilateral?*

**'foamy macrophages'**

## Choroid

*Are xanthelasmas a harbinger of elevated serum lipids?*

They can be, but in most cases the individual has normal lipid panels

*Can they be congenital?*

Yes, and when they are, they usually **are** a sign of lipid derangement

## RPE

*What is the natural history of JXG?*

It is self-limited, usually resolving by age 5 years

## Retina



# Intraocular Tumors of Childhood

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The presence of Touton giant cells  
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*Speaking of foamy macrophages part deux deux...*

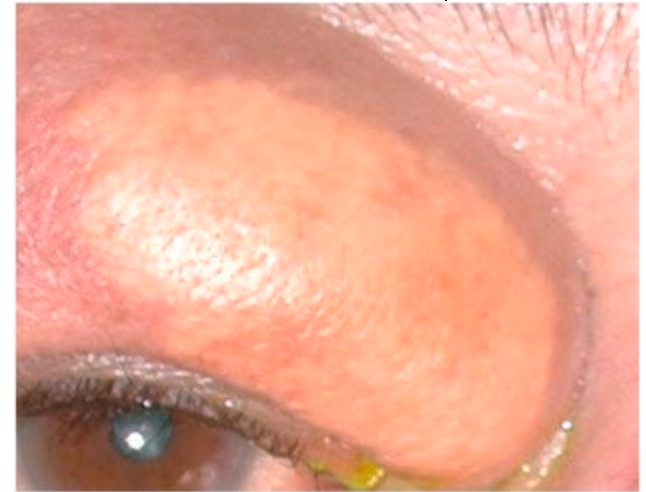
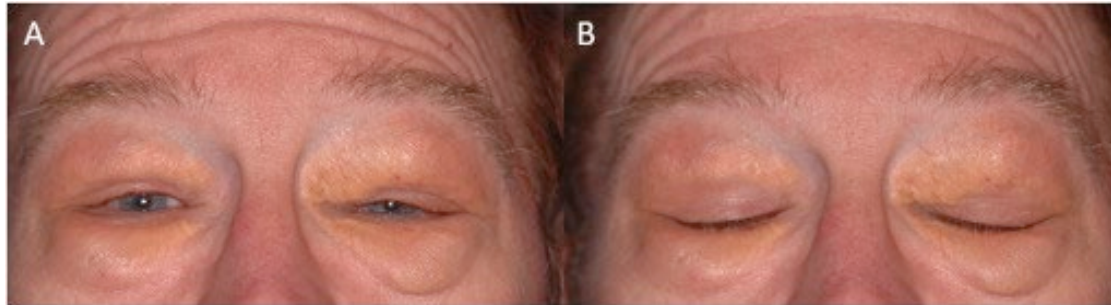
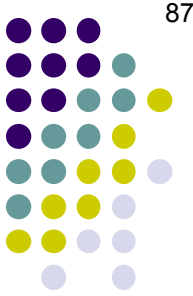
*What if the adult has what could only be described*

*as crazy-bad xanthelasma (pic next slide)*

*(No question yet—advance to the pic)*

**Retina**

# Intraocular Tumors of Childhood





# Intraocular Tumors of Childhood

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**Xanthelasma**



*When The presence of Touton giant cells*  
*Unilateral* **'foamy macrophages'** *bilateral?*

*Speaking of foamy macrophages part deux deux...*

*What if the adult has what could only be described*

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*No other issues whatsoever?*

← *Give the diagnosis*

**Retina**





# Intraocular Tumors of Childhood

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**Xanthelasma**



The presence of Touton giant cells  
When The presence of **'foamy macrophages'** lateral?  
Unilateral

*Speaking of foamy macrophages part deux deux...*

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Retina



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**Xanthelasma**



*When The presence of Touton giant cells is bilateral?*  
Unilateral

**'foamy macrophages'**

*Speaking of foamy macrophages part deux deux...*

*What if the adult has what could only be described as crazy-bad xanthelasma (pic next slide), **and:***

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*Recently diagnosed asthma?*

← Ditto

Retina



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**Retina**



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*Recently diagnosed asthma? **Adult-onset asthma with periocular xanthogranuloma***

*The 'xanthelasma' are ulcerated?*

← Ditto

**Retina**



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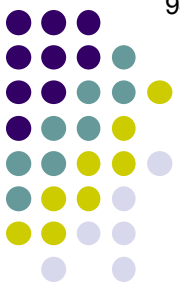
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*Recently diagnosed asthma? **Adult-onset asthma with periocular xanthogranuloma***

*The 'xanthelasma' are ulcerated? **Necrobiotic xanthogranuloma***

**Retina**



# Intraocular Tumors of Childhood



Necrobiotic xanthogranuloma



# Intraocular Tumors of Childhood

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**Xanthelasma**



*When The presence of Touton giant cells*

*Unilateral? The presence of 'foamy macrophages' lateral?*

*Speaking of foamy macrophages part deux deux...*

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*Recently diagnosed asthma? **Adult-onset asthma with periocular xanthogranuloma***

*The 'xanthelasma' are ulcerated? **Necrobiotic xanthogranuloma***

*Proptosis and/or terrible systemic symptoms are present?*

← Last one

**Retina**



# Intraocular Tumors of Childhood

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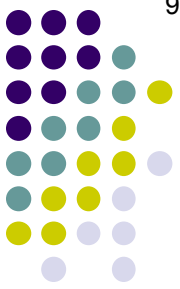
*The 'xanthelasma' are ulcerated? **Necrobiotic xanthogranuloma***

*Proptosis and/or terrible systemic symptoms are present? **Erdheim-Chester dz***

**Retina**



# Intraocular Tumors of Childhood



Erdheim-Chester disease



# Intraocular Tumors of Childhood

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**Xanthelasma**



*When The presence of Touton giant cells is bilateral?*  
Unilateral

**'foamy macrophages'**

*Speaking of foamy macrophages part deux deux...*

*What if the adult has what could only be described*

*as crazy-bad xanthelasma (pic next slide), and:*

*Collectively, these conditions are known as the...*

**Adult-onset xanthogranuloma**

**Adult-onset asthma with periocular xanthogranuloma**

**Atopic dermatitis? Necrobiotic xanthogranuloma**

**Systemic symptoms are present? Erdheim-Chester dz**

**Retina**



# Intraocular Tumors of Childhood

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**Adult xanthogranulomas**

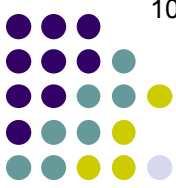
**Adult-onset xanthogranuloma**

**Adult-onset asthma with periocular xanthogranuloma**

**Atypical? Necrobiotic xanthogranuloma**

**Systemic symptoms are present? Erdheim-Chester dz**

**Retina**



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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**Xanthelasma**

*When The presence of Touton giant cells*

*lateral?* The presence of **'foamy macrophages'**

*In three words, what sort of condition are the AXGs?*

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*Collectively, these conditions are known as the...*

**Adult xanthogranulomas**

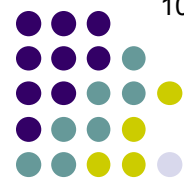
**Adult-onset xanthogranuloma**

**Adult-onset asthma with periocular xanthogranuloma**

**Infected? Necrobiotic xanthogranuloma**

**Systemic symptoms are present? Erdheim-Chester dz**

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

*In three words, what sort of condition is JXG?*

It is a... **nonneoplastic histiocytic proliferation**

*Speaking of foamy macrophages part deux...*

*What dz comes to mind if, instead of a toddler*

*with iris nodules, the pt in question was a*

*Adult with bilateral upper-lid yellow lesions?*

**Xanthelasma**

*When The presence of Touton giant cells*

*is seen The presence of*

**'foamy macrophages'**

*bilateral?*

*In three words, what sort of condition are the AXGs?*

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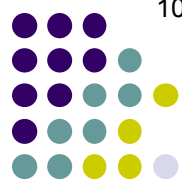
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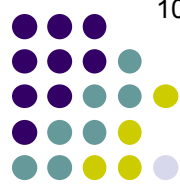
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*Proptosis and/or terrible systemic symptoms are present?*

**Juvenile xanthogranuloma**

**Retina**

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*Do the AXGs have Touton giant cells like their juvenile cousin?*

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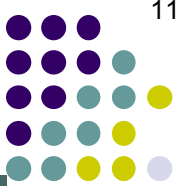
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Indeed they do*



# Intraocular Tumors of Childhood

That there is something called a non-Langerhans cell histiocytosis implies that Langerhans histiocytosis is a thing. Is there?

This implies  
Indeed it is

What is the nature of this relationship?

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'Nonneoplastic histiocytic proliferations'... Why does that sound so familiar?

Because that's the phrase we used to describe/define JXG

Adult-onset xanthogranuloma with peripheral cholesterol crystals?

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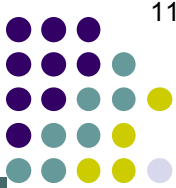
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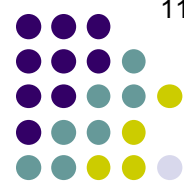
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# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

Iri

Speaking of foamy macrophages part three...  
 What dz comes to mind if, instead of a toddler  
 with iris nodules, the pt in question was a  
 8 y.o. with with a superotemporal orbital mass? ← Clue 1 of 2

Choroid

RPE

Retina

What are the two histologic or histology?  
 The presence of Touton giant cells  
 When The presence of 'foamy macrophages'  
 Unilateral? lateral?

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 DDx as a 'masquerade syndrome' in peds uveitis  
 --If enough nodules are present, heterochromia iridis will result

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



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

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*Speaking of foamy macrophages part three ...*

*What dz comes to mind if, instead of a toddler*

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*Imaging → soft tissue mass + lytic lesions? ← Clue 1 of 2*

What are the two histologic findings?

The presence of Touton giant cells

When The presence of **'foamy macrophages'** lateral?

Unilateral

Choroid

*In what three ways are the iris nodules clinically significant?*

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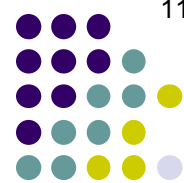
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# Intraocular Tumors of Childhood

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 Imaging → soft tissue mass + lytic lesions?  
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Choroid

RPE

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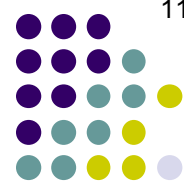
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# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

*Unilateral, nodular, foamy macrophages (JXG)*

Iris

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What dz comes to mind if, instead of a toddler  
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8 y.o. with a superotemporal orbital mass?  
Imaging → soft tissue mass + lytic lesions?*

***For more on the Langerhans (and non-Langerhans)  
histiocytoses, see slide-set K20***

*Unilateral*

Choroid

*In what three ways are the iris nodules clinically significant?*  
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**Langerhans-cell histiocytosis** (and there it is!)

*What are the two hallmarks of LCH histology?*

The presence of Touton-giant cells

When The presence of **'foamy macrophages'** lateral?

Unilateral

Choroid

*Speaking of foamy macrophages part whatever ...*

*What dz comes to mind if the pt in question was a*

*6 y.o. boy with with unilateral leukocoria? ← Clue #1*

Retina



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*No family hx for anything like this? ← Clue #2*

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**DFE → Exudative RD?** ← Clue #3

Retina





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**DFE → Exudative RD?**

**DFE also → Retinal vascular microaneurysms, telangiectasias, dilatation? ← Clue last**

Retina



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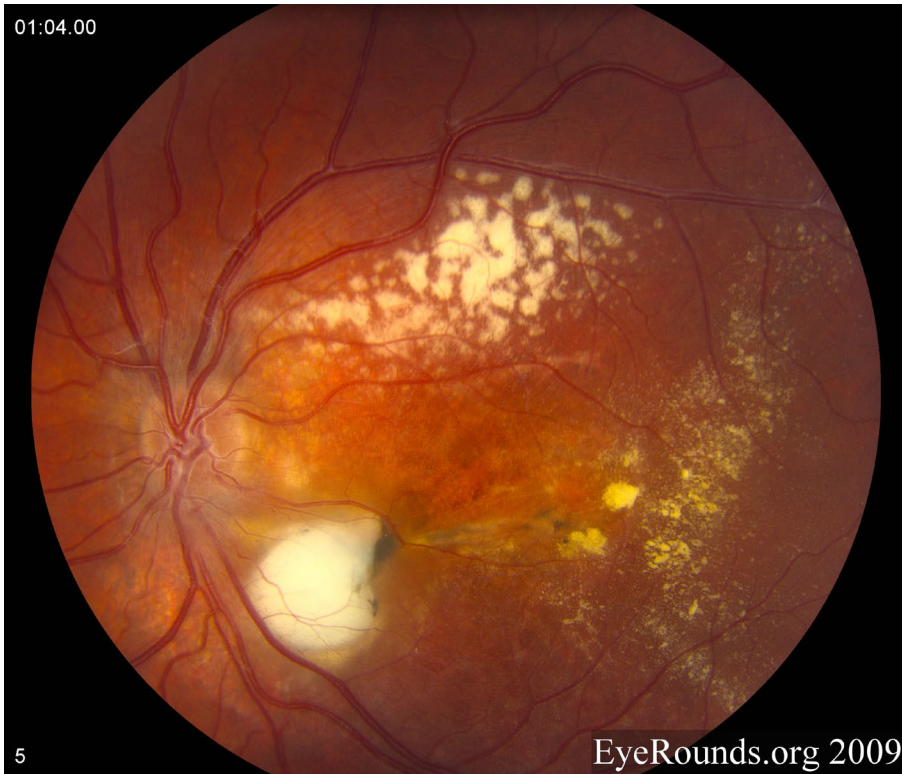
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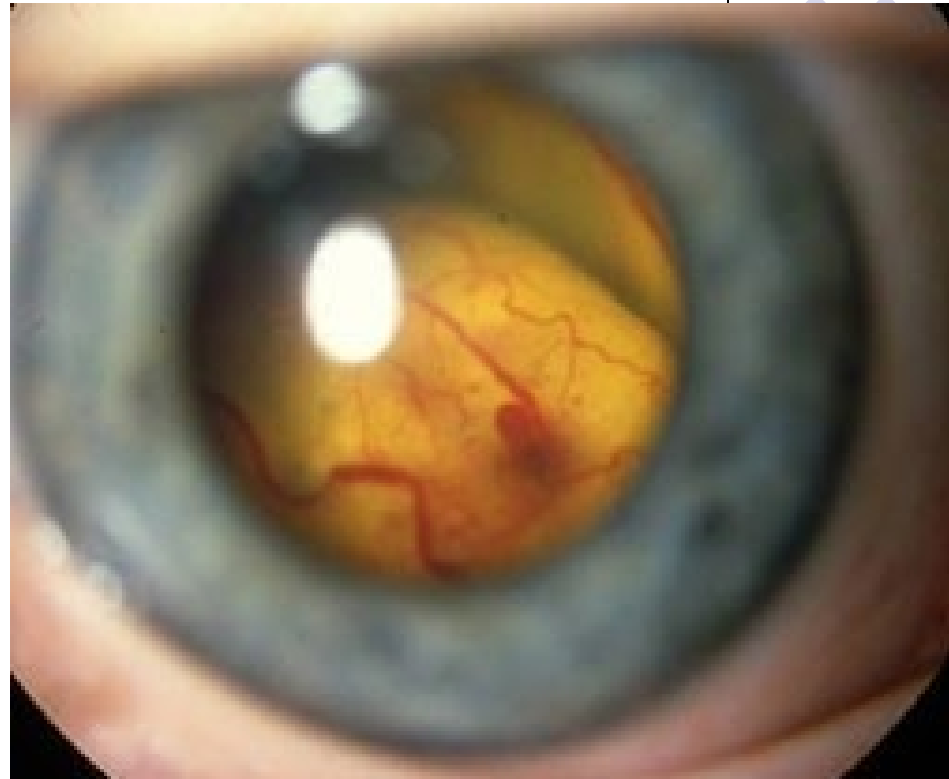
**Coats dz**

Retina

# Intraocular Tumors of Childhood

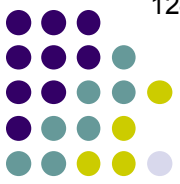


Exudates; MAs; telangiectasias



Exudative RD (note also the vasc abnormalities)

Coats disease



# Intraocular Tumors of Childhood

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Where are the foamy macrophages found in Coats dz?

Retina



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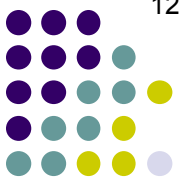
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**Coats dz**

Where are the foamy macrophages found in Coats dz?  
 In the subretinal exudate

Retina



# Intraocular Tumors of Childhood

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What dz comes to mind if, instead of a toddler with iris nodules, the pt in question was a 8 y.o. with with a superotemporal orbital mass? Imaging → soft tissue mass + lytic lesions?

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When The presence of **'foamy macrophages'** lateral?  
Unilateral

Choroid

Speaking of foamy macrophages part whatever ...

What dz comes to mind if the pt in question was a 6 y.o. boy with with unilateral leukocoria?

No family hx for anything like this?

**DFE → Exudative RD?**

DFE also → Retinal vascular microaneurysms, telangiectasias, dilatation?

**Coats dz**

Where are the foamy macrophages found in Coats dz?

In the subretinal exudate (also present: substance crystals)

Retina



# Intraocular Tumors of Childhood

## 1) Juvenile xanthogranuloma

Iri

*Speaking of foamy macrophages part three ...*

*What dz comes to mind if, instead of a toddler with iris nodules, the pt in question was a 8 y.o. with with a superotemporal orbital mass?*

*Imaging → soft tissue mass + lytic lesions?*

**Langerhans-cell histiocytosis** (and there it is!)

*What are the two hallmarks of LCH histology?*

The presence of Touton giant cells

When The presence of **'foamy macrophages'** lateral?

Unilateral

Choroid

*Speaking of foamy macrophages part whatever ...*

*What dz comes to mind if the pt in question was a 6 y.o. boy with with unilateral leukocoria?*

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**DFE → Exudative RD?**

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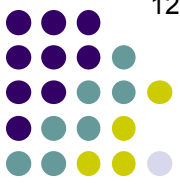
**Coats dz**

*Where are the foamy macrophages found in Coats dz?*

*In the subretinal exudate (also present: cholesterol crystals)*

Retina

# Intraocular Tumors of Childhood



1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells

2) **Medulloepithelioma**

## Iris/Ciliary Body

*What is the other name by which medulloepithelioma is known?*

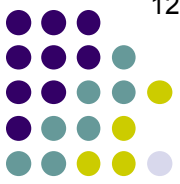
Choroid

RPE

Retina



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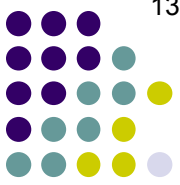
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Diktyoma

Choroid

RPE

Retina



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Choroid

RPE

Retina



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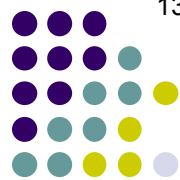
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The nonpigmented epithelium of the ciliary body

Choroid

RPE

Retina



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*What extremely important function does the nonpigmented epi of the CB perform?*

Choroid

RPE

Retina



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Choroid

RPE

Retina



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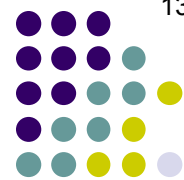
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It is responsible for the creation of aqueous humor

Choroid

RPE

Retina



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*How does it present?*

Choroid

RPE

Retina



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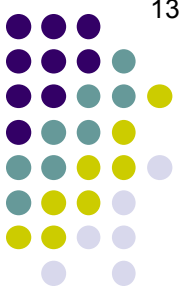
As an iris mass

Choroid

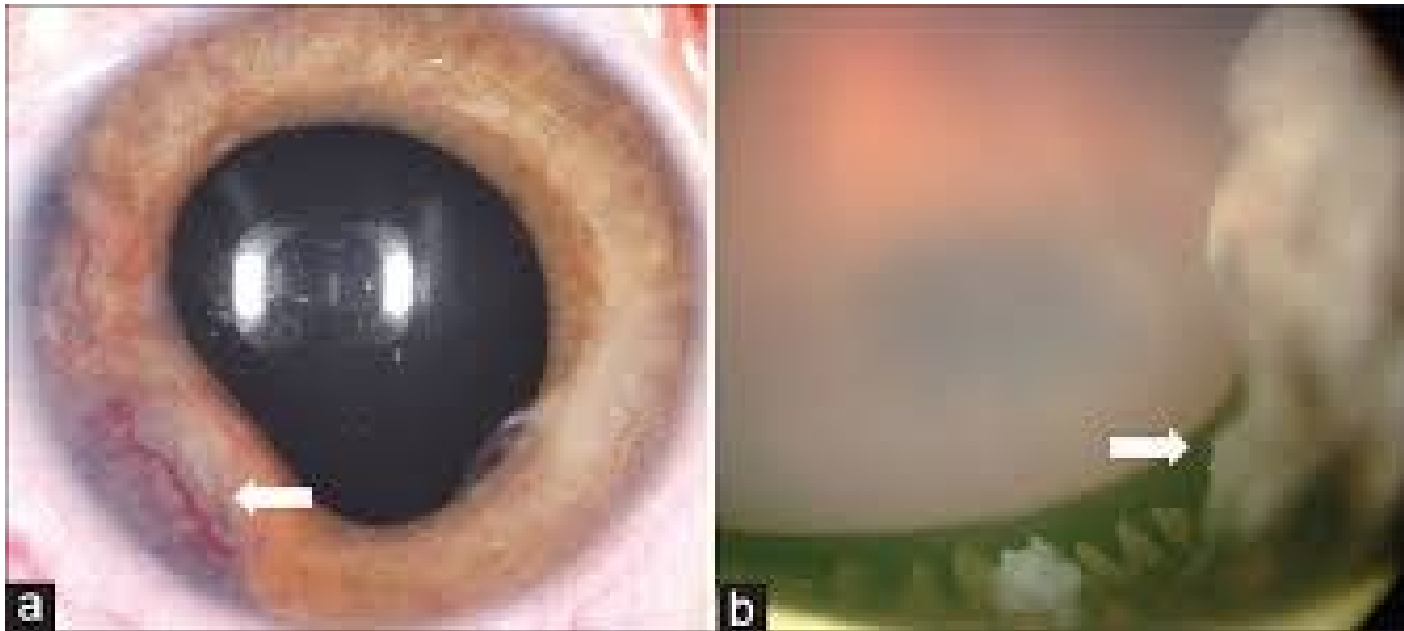
RPE

Retina

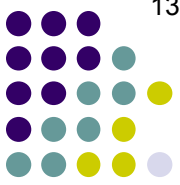




# Intraocular Tumors of Childhood



Medulloepithelioma/diktyoma



# Intraocular Tumors of Childhood

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The nonpigmented epithelium of the ciliary body

*How does it present?*

As an iris mass along with one or more of the following:

--?

--?

--?

Choroid

RPE

Retina



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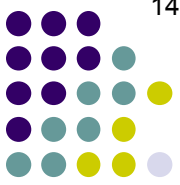
--Hyphema

--Sectoral cataract

Choroid

RPE

Retina



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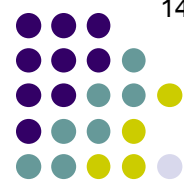
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- Hyphema
- Sectoral cataract

*Is it common, or rare?*

Choroid

RPE

Retina



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- Hyphema
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*Is it common, or rare?*

Very rare

*Is it a benign, or malignant lesion?*

Choroid

RPE

Retina



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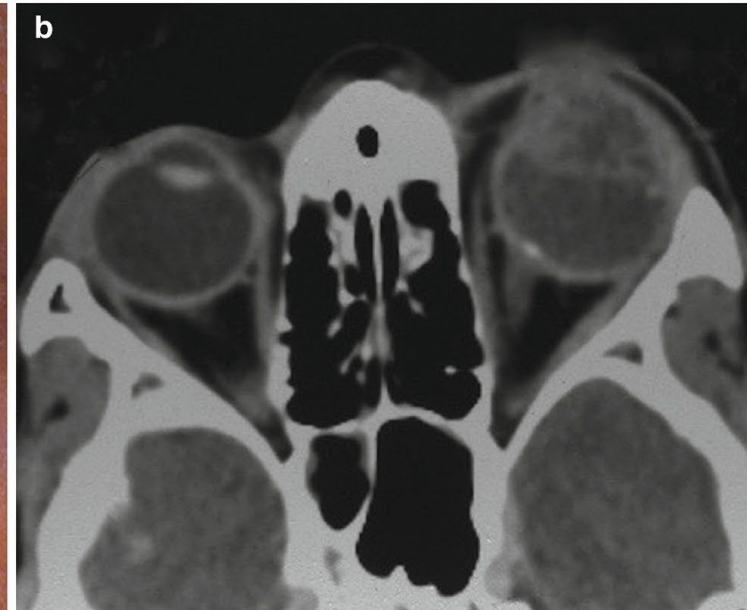
It can have features of both. Either way, it is very locally aggressive.

Choroid

RPE

Retina

# Intraocular Tumors of Childhood



Medulloepithelioma/diktyoma





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## Choroid

*Is it common, or rare?*

Very rare

## RPE

*Is it a benign, or **malignant lesion**?*

**It can have features of both.** Either way, it is very locally aggressive.

*Is a tendency to metastasize one of its 'malignant features'?*



# Intraocular Tumors of Childhood

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## Choroid

*Is it common, or rare?*

Very rare

## RPE

*Is it a benign, or **malignant lesion**?*

**It can have features of both.** Either way, it is very locally aggressive.

*Is a tendency to metastasize one of its 'malignant features'?*  
No, this lesion rarely metastasizes; it does its damage locally



# Intraocular Tumors of Childhood

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*Is it common, or rare?*

Very rare

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*Is it a benign, or malignant lesion?*

It can have features of both. Either way, it is **very locally aggressive**.

## Retina

*How aggressive is 'very aggressive'?*



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*Is it common, or rare?*

Very rare

## RPE

*Is it a benign, or malignant lesion?*

It can have features of both. Either way, it is **very locally aggressive**.

## Retina

*How aggressive is 'very aggressive'?*

Aggressive enough to result in death



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*Is it common, or rare?*

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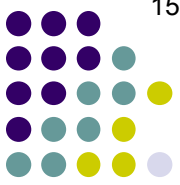
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*How is it managed?*

Choroid

RPE

Retina



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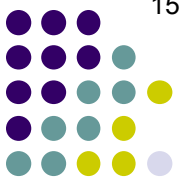
*How is it managed?*

Enucleation is usually required

Choroid

RPE

Retina



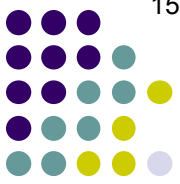
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- 3) **Lisch nodules**
- 4) **Brushfield spots**

*Lisch nodules are most strongly associated with what congenital condition?*

Retina



# Intraocular Tumors of Childhood

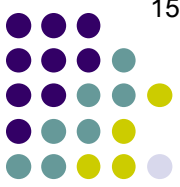
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*Lisch nodules are most strongly associated with what congenital condition?*  
NF1

Retina





# Intraocular Tumors of Childhood

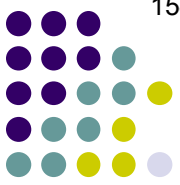
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*Lisch nodules are most strongly associated with what congenital condition?*  
**NF1**

*In this context, what does NF1 stand for?*

Retina



# Intraocular Tumors of Childhood

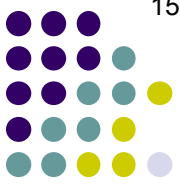
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**NF1**

*In this context, what does NF1 stand for?*  
 Neurofibromatosis type 1

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

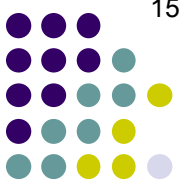
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*What is the eponymous name for NF1?*

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

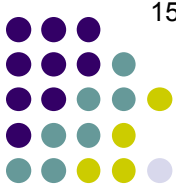
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- 4) **Brushfield spots**

*Lisch nodules are most strongly associated with what congenital condition?*  
**NF1**

*In this context, what does NF1 stand for?*  
 Neurofibromatosis type 1

*What is the eponymous name for NF1?*  
 von Recklinghausen's disease

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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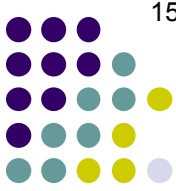
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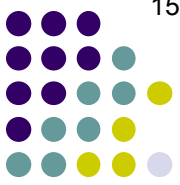
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## Retina



# Intraocular Tumors of Childhood

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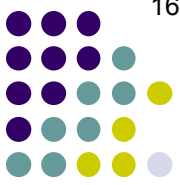
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*Phakomatoses are known also as what sort of syndrome?*

## Retina



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**Neurocutaneous syndromes**

## Retina





# Intraocular Tumors of Childhood

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*Phakomatoses are known also as what sort of syndrome?*  
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*In general terms, how do phakomatoses present?*

## Retina



# Intraocular Tumors of Childhood

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*In general terms, how do phakomatoses present?*  
 With multiple lesions in two or more organ systems, including the  and  abb.

## Retina



# Intraocular Tumors of Childhood

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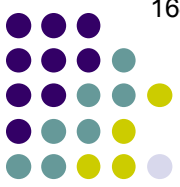
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*In general terms, how do phakomatoses present?*  
 With multiple lesions in two or more organ systems, including the skin and CNS

## Retina



# Intraocular Tumors of Childhood

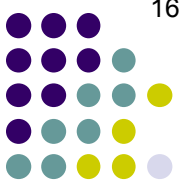
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NF1

*Are Lisch nodules associated with NF2?*

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

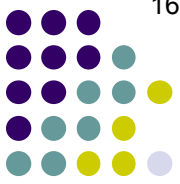
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*Lisch nodules are most strongly associated with what congenital condition?*  
NF1

*Are Lisch nodules associated with NF2?*

Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

Retina



# Intraocular Tumors of Childhood

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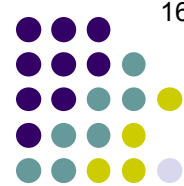
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NF1

*Are Lisch nodules associated with NF2?*

Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

*What is the prevalence of Lisch nodules in NF1?*

## Retina



# Intraocular Tumors of Childhood

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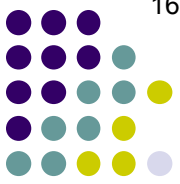
*Are Lisch nodules associated with NF2?*

Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

*What is the prevalence of Lisch nodules in NF1?*

The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

## Retina



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*Are Lisch nodules clinically significant?*

## Retina





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*Are Lisch nodules clinically significant?*

No; their only significance is as a diagnostic marker for NF1

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*Most NF1 lesions are associated with one of two cell types. What are they?*

neoplasia of  
s. Can  
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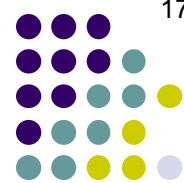
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## Melanocytic lesions

--?  
--?  
--?  
--?

Give four classic examples of each:  
(YMMV of course)

## Neuroglial lesions

--?  
--?  
--?  
--?

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## Melanocytic lesions

- Café au lait spots
- Axillary/inguinal freckles
- Lisch nodules
- Choroidal lesions

*Give four classic examples of each: (YMMV of course)*

## Neuroglial lesions

- Nodular neurofibromas
- Plexiform neurofibromas
- Optic glioma
- Prominent corneal nerves

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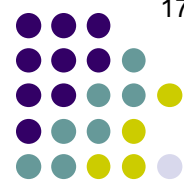
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*In what fundamental way do these lesions differ (other than the cell type of origin, duh)?*

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No; their only significance is as a diagnostic marker for NF1

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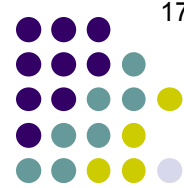
*In what fundamental way do these lesions differ (other than the cell type of origin, duh)?*

The **m'cytic v N-G** lesions are of no clinical significance beyond establishing the diagnosis, whereas the **m'cytic v N-G** lesions are associated with significant ocular and/or systemic morbidity

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*Give four classic examples of each: (YMMV of course)*

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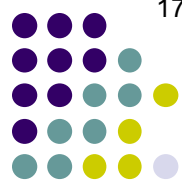
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Retina





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## Melanocytic lesions

--Lisch nodules

## Neuroglial lesions

--Nodular neurofibromas  
--Plexiform neurofibromas  
--Optic glioma  
--Prominent corneal nerves

*Give four classic examples of each: (YMMV of course)*

*What is the prevalence of Lisch nodules in NF1?*

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*Are Lisch nodules clinically significant?*

No; their only significance is as a diagnostic marker for NF1

Retina

(No question—proceed when ready)



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**
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*Lisch nodules are most strongly associated with what congenital condition?*  
NF1

*Are Lisch nodules dark, or light?*  
Are  
Ye  
sp

*What is the prevalence of Lisch nodules in NF1?*

The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

*Are Lisch nodules clinically significant?*

No; their only significance is as a diagnostic marker for NF1

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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- 3) **Lisch nodules**
- 4) **Brushfield spots**

*Lisch nodules are most strongly associated with what congenital condition?*  
NF1

*Are Lisch nodules dark, or light?*

It depends. Lisch nodules are lighter v darker than the rest of the iris when the iris in question is dark, but lighter v darker than the rest when the iris is light.

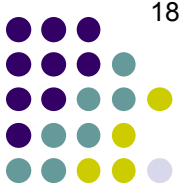
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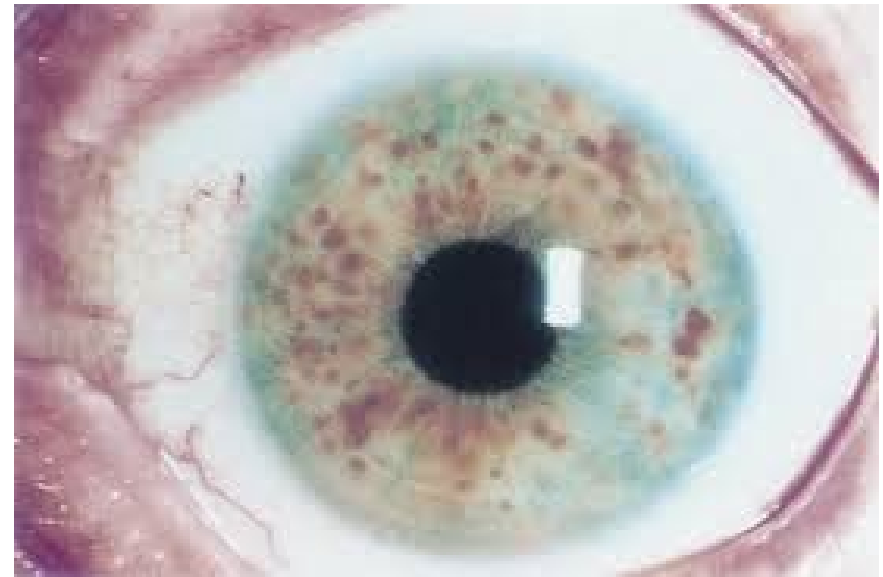
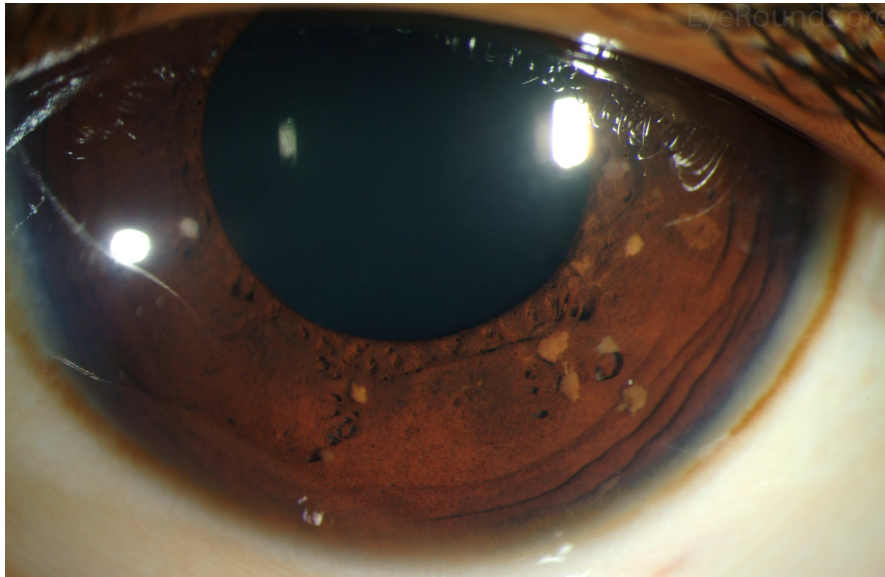
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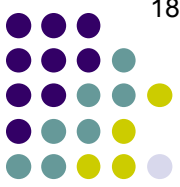
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## Retina

# Intraocular Tumors of Childhood



Lisch nodules



# Intraocular Tumors of Childhood

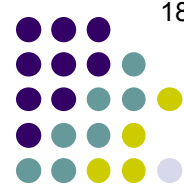
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- 5) **Iris mammillations**

*Brushfield spots are most strongly associated with what congenital condition?*

Ch

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

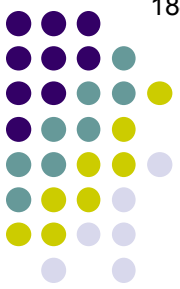
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*Brushfield spots are most strongly associated with what congenital condition?*  
Down syndrome

Ch

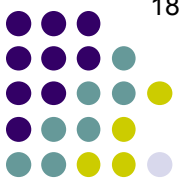
Retina

# Intraocular Tumors of Childhood



Brushfield spots





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

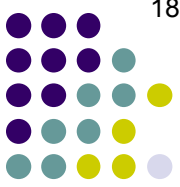
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Down syndrome

*What is the prevalence of Brushfield spots in the Down population?*

Ch

Retina



# Intraocular Tumors of Childhood

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*What is the prevalence of Brushfield spots in the Down population?*  
At least 90%

Ch

Retina



# Intraocular Tumors of Childhood

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*What is the prevalence of Brushfield spots in the Down population?*  
At least 90%

*What is the clinical significance of Brushfield spots?*

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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*What is the prevalence of Brushfield spots in the Down population?*

At least 90%

*What is the clinical significance of Brushfield spots?*

They have none

## Retina



# Intraocular Tumors of Childhood

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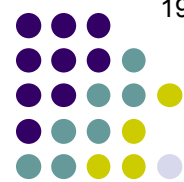
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*What is the clinical significance of Brushfield spots?*

They have none

*When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called?*

## Retina



# Intraocular Tumors of Childhood

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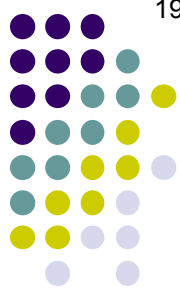
*What is the clinical significance of Brushfield spots?*

They have none

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**Wolfflin nodules**

## Retina



# Intraocular Tumors of Childhood



Wolfflin nodules



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*Mammillations? Aren't those a CNS thingamajig?*

Ch

Retina





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

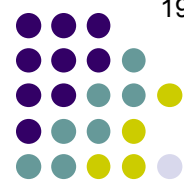
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You're thinking of the , paired structures that are part of the limbic system

Ch

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

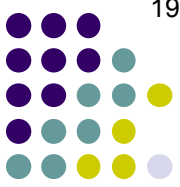
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Ch

Retina



# Intraocular Tumors of Childhood

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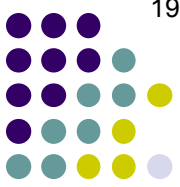
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You're thinking of the **mammillary bodies**, paired structures that are part of the limbic system

*OK, then what are iris mammillations?*

Retina



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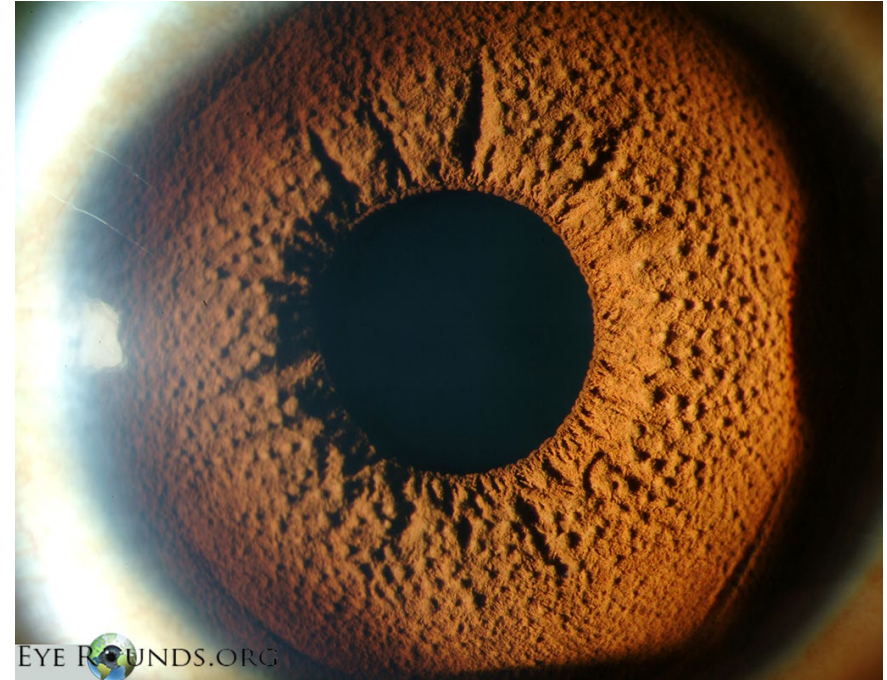
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*OK, then what are iris mammillations?*

Tiny pigmented iris nodules which, when present, are found in vast numbers diffusely scattered across the iris surface

Retina

# Intraocular Tumors of Childhood



Iris mammilations



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*Are they unilateral, or bilateral?*

Retina



# Intraocular Tumors of Childhood

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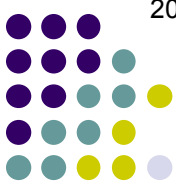
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Retina



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Retina





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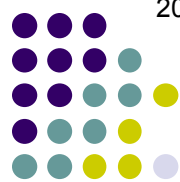
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# Intraocular Tumors of Childhood

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By appearance. As stated previously, Lisch nodules are lighter when the iris is dark, but darker when the iris is light.

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# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**: Strong association with NF1. Lighter on dark irides; darker on light
- 4) **Brushfield spots**: Strong association with Down syndrome
- 5) **Iris mammillations**

*Mammillations? Aren't those a CNS thingamajig?*

You're thinking of the **mammillary bodies**, paired structures that are part of the limbic system

*OK, then what are iris mammillations?*

**Tiny pigmented iris nodules** which, when present, are found in vast numbers diffusely scattered across the iris surface

*'Tiny pigmented iris nodules associated with NF1'--given this, how on earth are you supposed to differentiate between Lisch nodules and mammillations?*

By appearance. As stated previously, Lisch nodules are lighter when the iris is dark, but darker when the iris is light. In contrast, iris mammillations are always

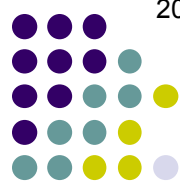
the rest of the iris.

light than?  
darker than?  
same color as?

*With what phakomatosis are they associated?*

**NF1** (albeit not nearly as strongly as Lisch nodules)

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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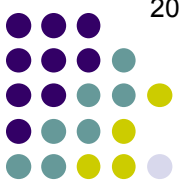
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# Intraocular Tumors of Childhood

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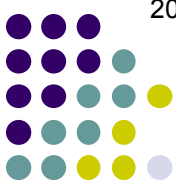
*Are they unilateral, or bilateral?*

Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out

*With what phakomatosis are they associated?*

NF1 (albeit not really as strongly as Lisch nodules)

*In addition to NF1, iris mammillations have another important association. What is it?*



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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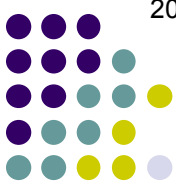
NF1 (albeit not really as strongly as Lisch nodules)

*In addition to NF1, iris mammillations have another important association.*

*What is it?*

Oculodermal melanocytosis, aka

three words



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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*What is it?*

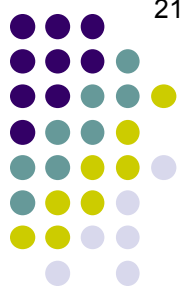
Oculodermal melanocytosis, aka **nevus of Ota**



# Intraocular Tumors of Childhood



Oculodermal melanocytosis (nevus of Ota).



# Intraocular Tumors of Childhood



Oculodermal melanocytosis (nevus of Ota).

Note the presence of non-eye structure pigmentation in addition to that of the eye structure





# Intraocular Tumors of Childhood



Oculodermal melanocytosis (nevus of Ota).

Note the presence of periorcular pigmentation in addition to that of the episclera.



# Intraocular Tumors of Childhood

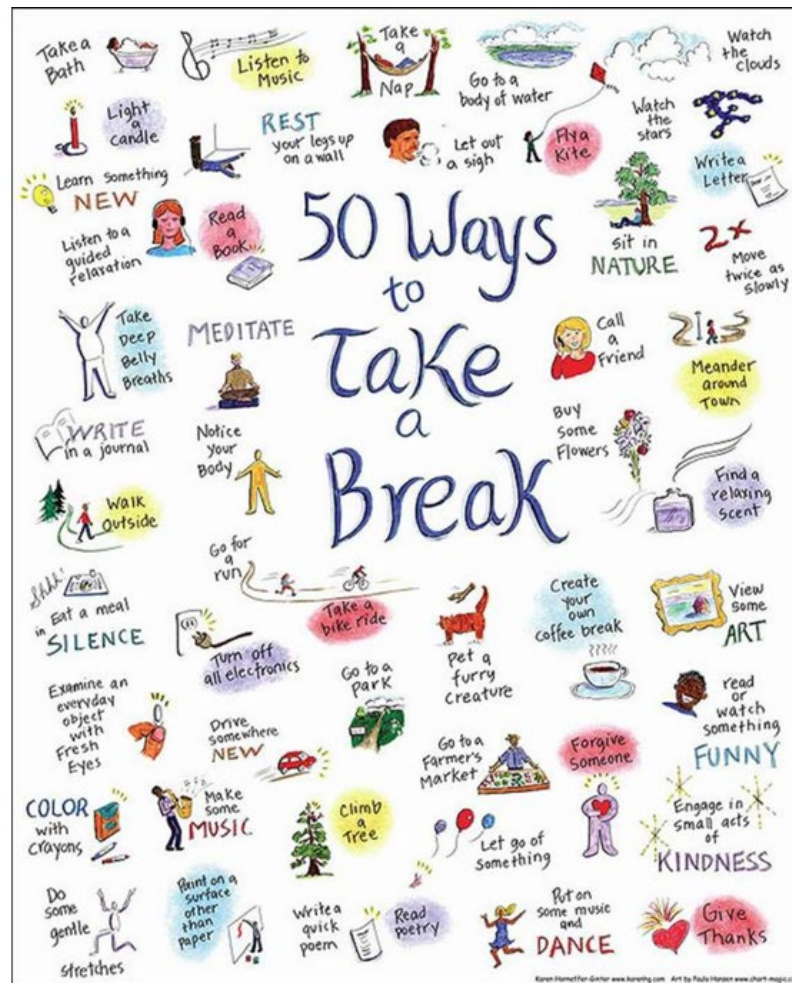
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- 6) **Iris cysts**: Can be pupillary, stromal, secondary (see the *Iris issues is kids* slide-set)

Choroid

RPE

Retina



(This is a good point in the set to take a break)



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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## Choroid

- 1) ?
- 2) ?
- 3) ?
- 4) ?
- 5) ?

} *Five tumors of the choroid*

RPE

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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## Choroid

- 1) **Nevus**
- 2) **Melanocytoma**
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- 4) **Isolated/focal choroidal hemangioma**
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*Five tumors of the choroid*

RPE

Retina



# Intraocular Tumors of Childhood

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But not 6) ?

*Five tumors of the choroid*

*What common sort of choroidal tumor—common in adults—is absent from this list?*

Retina





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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But not 6) Metastases

*Five tumors of the choroid*

*What common sort of choroidal tumor—common in adults—is absent from this list?*

Choroidal tumors arising as **metastases** from a nonocular primary. In adults, metastasis of solid tumors to the uveal tract is common. It almost *never* happens in children.

Retina



# Intraocular Tumors of Childhood

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*If a child does suffer an ophthalmic metastasis, where does it tend to occur?*

Retina



# Intraocular Tumors of Childhood

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*Five tumors of the choroid*

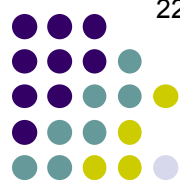
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The **orbit**

Retina



# Intraocular Tumors of Childhood

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Note the factoids, then proceed

## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**: Striae of hamman-rich
- 4) **Brushfield spots**: Pigment clumps
- 5) **Iris mammillations**: Nevus of Ota
- 6) **Iris cysts**: Can be

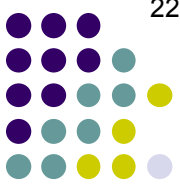
*Melanocytoma is a variant of what common choroidal finding?*

## Choroid

- 1) **Nevus**: Common. Benign
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- 5) **Diffuse choroidal melanoma**

## RPE

## Retina



# Intraocular Tumors of Childhood

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- 3) **Lisch nodules**: Striae of hamman
- 4) **Brushfield spots**: Striae of hamman
- 5) **Iris mammillations**: Striae of hamman
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

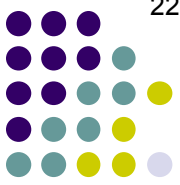
It is a particular sort of **choroidal nevus**

## Choroid

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## RPE

## Retina



# Intraocular Tumors of Childhood

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- 3) **Lisch nodules**: Striae of hamman-Richardson
- 4) **Brushfield spots**: Striae of hamman-Richardson
- 5) **Iris mammillations**: Striae of hamman-Richardson
- 6) **Iris cysts**: Can be congenital or acquired

*Melanocytoma is a variant of what common choroidal finding?*

*It is a particular sort of **choroidal nevus***

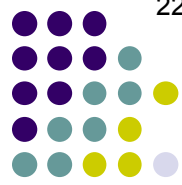
*From what structure does it commonly arise?*

## Choroid

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## RPE

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# Intraocular Tumors of Childhood

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- 3) **Lisch nodules**: Striae
- 4) **Brushfield spots**: Striae
- 5) **Iris mammillations**: Striae
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

## Choroid

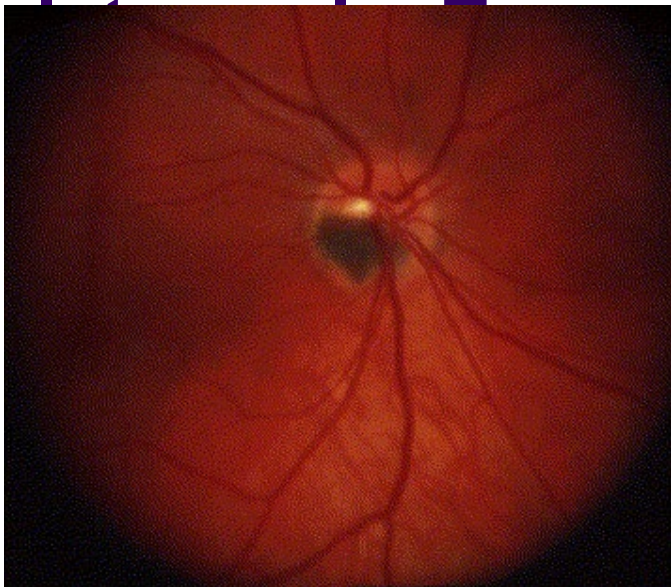
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## RPE

## Retina

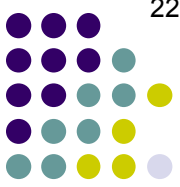


# Lesions of Childhood



Melanocytoma





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**: Striae
- 4) **Brushfield spots**: Striae
- 5) **Iris mammillations**: Striae
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

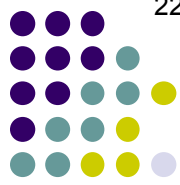
*Does it have a unilateral/bilateral predilection?*

## Choroid

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## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 2) **Medulloepithelioma** (aka **diktyoma**): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed → hyphema → inc IOP → glaucoma. Treat with enucleation. Path: Ependymoma-like
- 3) **Lisch nodules**: Striae of hamman
- 4) **Brushfield spots**: Striae of hamman
- 5) **Iris mammillations**: Striae of hamman
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

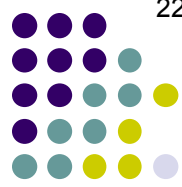
*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal choroidal melanoma**
- 5) **Diffuse choroidal melanoma**

## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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- 3) **Lisch nodules**: Striae of hamman-Richardson
- 4) **Brushfield spots**: Striae of hamman-Richardson
- 5) **Iris mammillations**: Nevus of Ota
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

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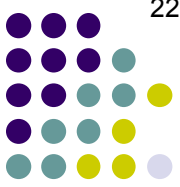
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## Choroid

- 1) **Nevus**: Common. Benign
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## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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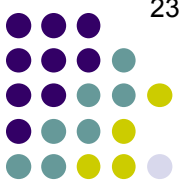
*Is there a racial predilection?*

## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**
- 3) **Osteoma**
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## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**: Striae
- 4) **Brushfield spots**: Striae
- 5) **Iris mammillations**: Striae
- 6) **Iris cysts**: Can be

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

## Choroid

- 1) **Nevus**: Common. Benign
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## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 3) **Lisch nodules**: Str
- 4) **Brushfield spots**:
- 5) **Iris mammillations**: Nevus of Ota
- 6) **Iris cysts**: Can be

## Choroid

- 1) **Nevus**: Common. E
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal cho**
- 5) **Diffuse choroidal**

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

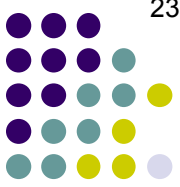
*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

*Does it affect visual acuity?*

## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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*From what structure does it commonly arise?*  
The optic disc

*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

*Does it affect visual acuity?*  
Only in a minority of cases. But in almost all cases, it does affect

two words

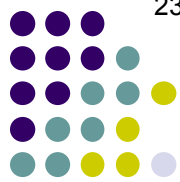
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## RPE

## Retina





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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The optic disc

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Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

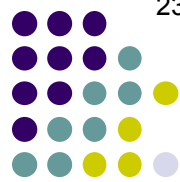
*Does it affect visual acuity?*  
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## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal choroidal melanoma**
- 5) **Diffuse choroidal melanoma**

## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 4) **Brushfield spots**:
- 5) **Iris mammillations**: Nevus of Ota
- 6) **Iris cysts**: Can be

## Choroid

- 1) **Nevus**: Common. B
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal cho**
- 5) **Diffuse choroidal**

## RPE

## Retina

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

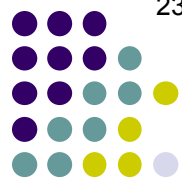
*From what structure does it commonly arise?*  
The optic disc

*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

*Does it affect visual acuity?*  
Only in a minority of cases. But in almost all cases, it does affect visual fields.

*Does melanocytoma have the potential to undergo malignant transformation?*



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 6) **Iris cysts**: Can be

## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal choroidal melanoma**
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## RPE

## Retina

*Melanocytoma is a variant of what common choroidal finding?*

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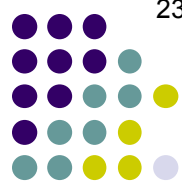
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The optic disc

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Only in a minority of cases. But in almost all cases, it does affect visual fields.

*Does melanocytoma have the potential to undergo malignant transformation?*  
Yes



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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- 3) **Lisch nodules**: Str
- 4) **Brushfield spots**:
- 5) **Iris mammillations**: It is a particular sort of **choroidal nevus**
- 6) **Iris cysts**: Can be

## Choroid

- 1) **Nevus**: Common. E
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal cho**
- 5) **Diffuse choroidal**

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

*Does it have a unilateral/bilateral predilection?*  
Yes, it is virtually always unilateral

*Is there a racial predilection?*  
No

*Does it affect visual acuity?*  
Only in a minority of cases. But in almost all cases, it does affect visual fields.

*Does melanocytoma have the potential to undergo malignant transformation?*

*Approximately what percent of cases will transform?*

## RPE

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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- 3) **Lisch nodules**: Striae of hamman-Richardson
- 4) **Brushfield spots**: Striae of hamman-Richardson
- 5) **Iris mammillations**: Striae of hamman-Richardson
- 6) **Iris cysts**: Can be congenital or acquired

## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**
- 3) **Osteoma**
- 4) **Isolated/focal choroidal melanoma**
- 5) **Diffuse choroidal melanoma**

## RPE

## Retina

*Melanocytoma is a variant of what common choroidal finding?*

It is a particular sort of **choroidal nevus**

*From what structure does it commonly arise?*  
The optic disc

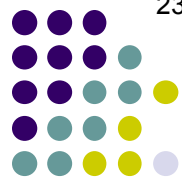
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*Is there a racial predilection?*  
No

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*Does melanocytoma have the potential to undergo malignant transformation?*

*Approximately what percent of cases will transform?*  
1-2



*In a word, what is a choroidal osteoma composed of?*

Int

Iris/C

ation. <2 years old.

oma. Self-limited;

s

neoplasia of

s. Can

Tx: Enucleate

darker on light

ciation with NF1,

s kids slide-set)

emely rare

**Choroid**

3) **Osteoma**

4) Isolated/focal choroidal hemangioma

5) Diffuse choroidal hemangioma

**RPE**

**Retina**



# Int

*In a word, what is a choroidal osteoma composed of?*

Bone

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ation. <2 years old.

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neoplasia of

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Tx: Enucleate

darker on light

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emely rare

## Choroid

3) **Osteoma**

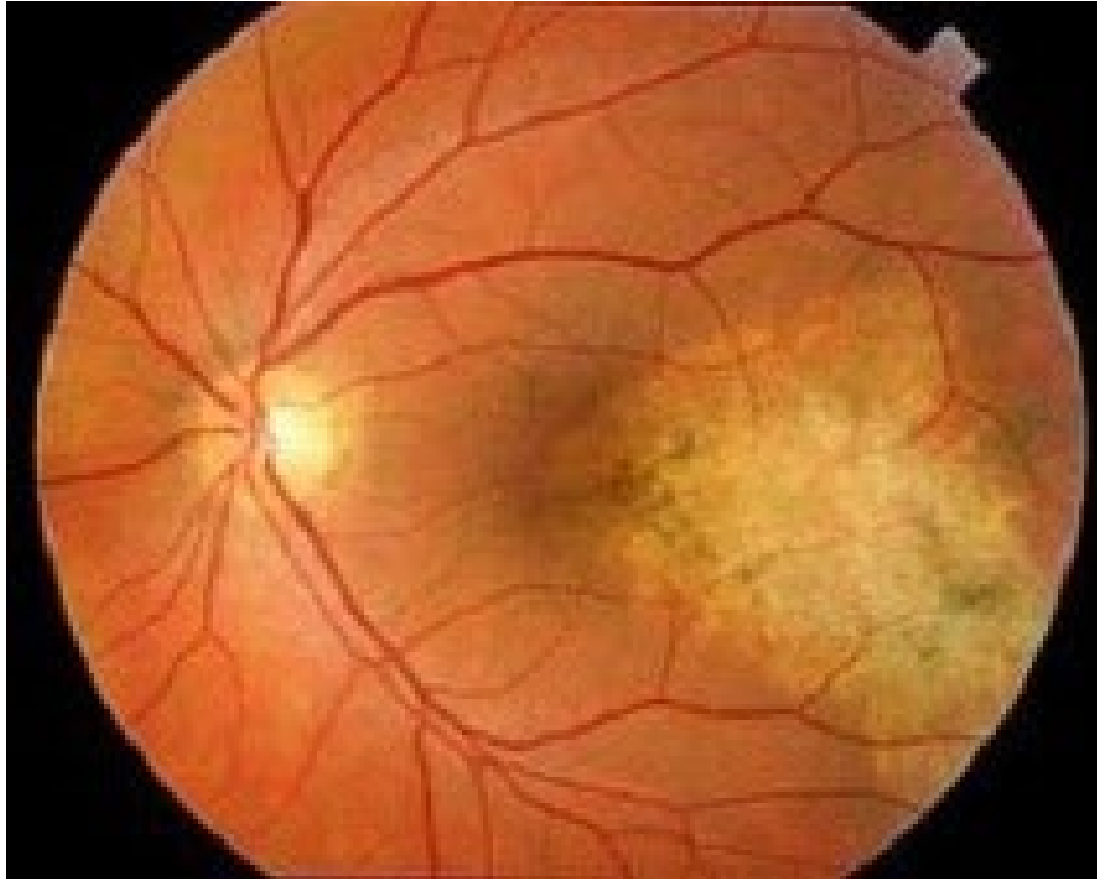
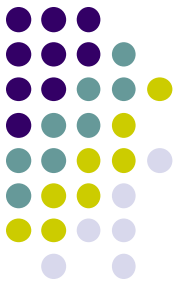
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5) Diffuse choroidal hemangioma

## RPE

## Retina

# Intraocular Tumors of Childhood

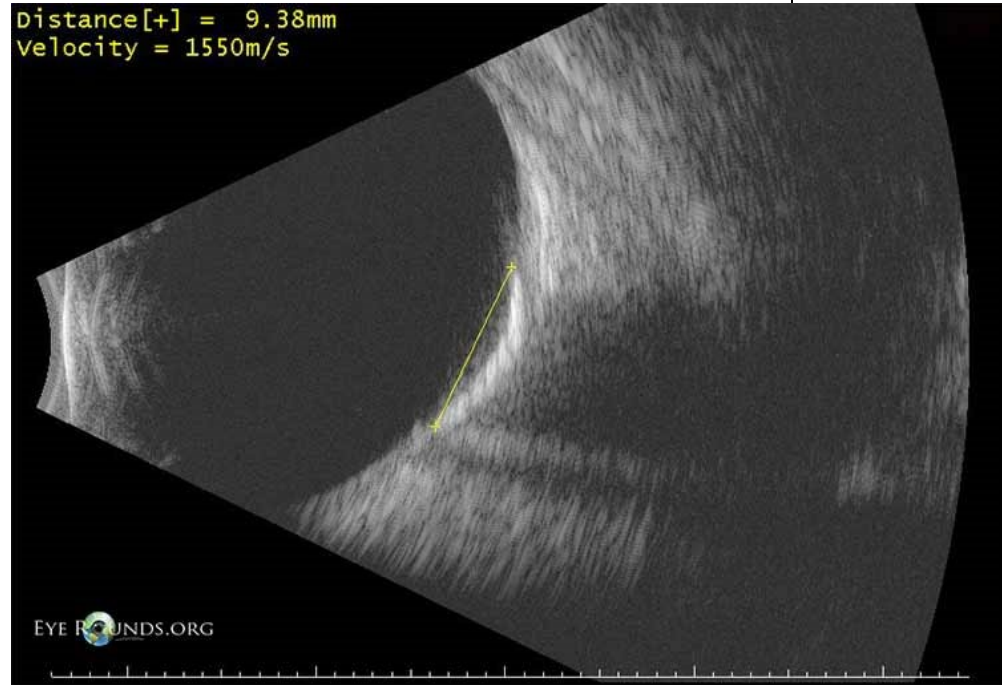
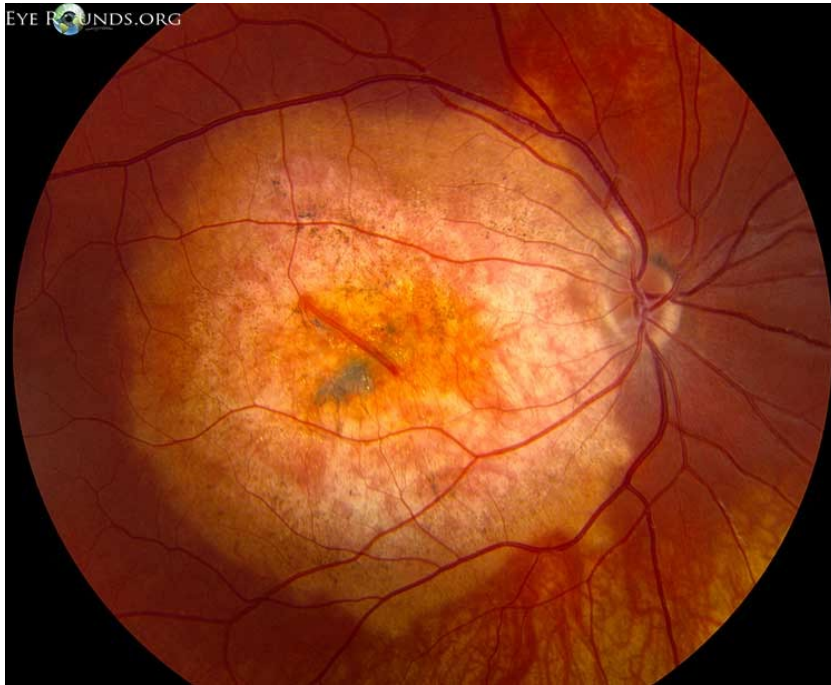


Osteoma





# Intraocular Tumors of Childhood

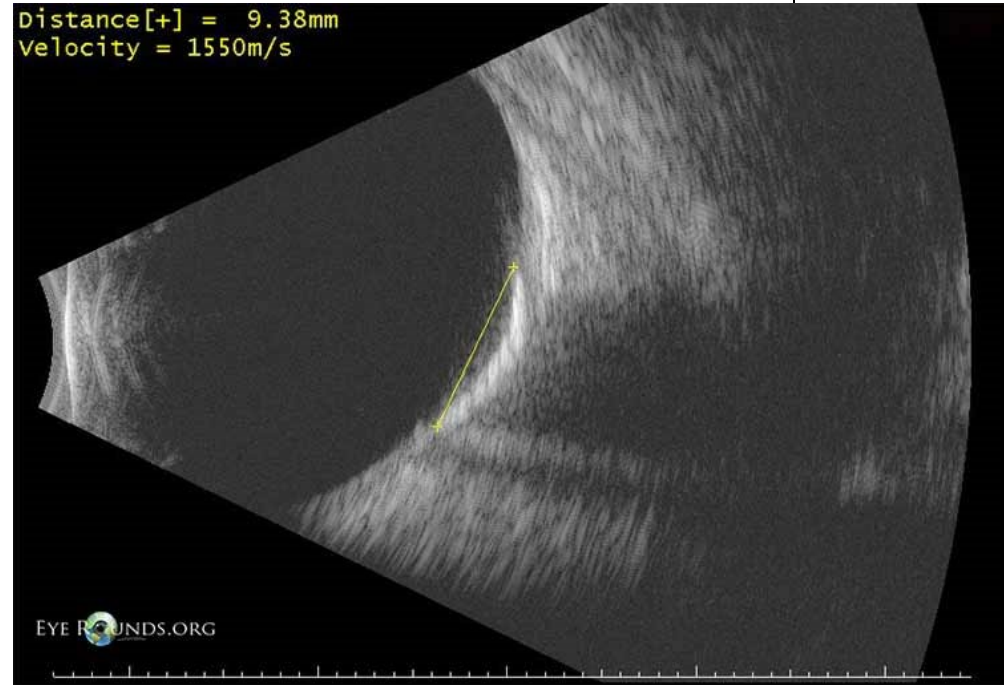
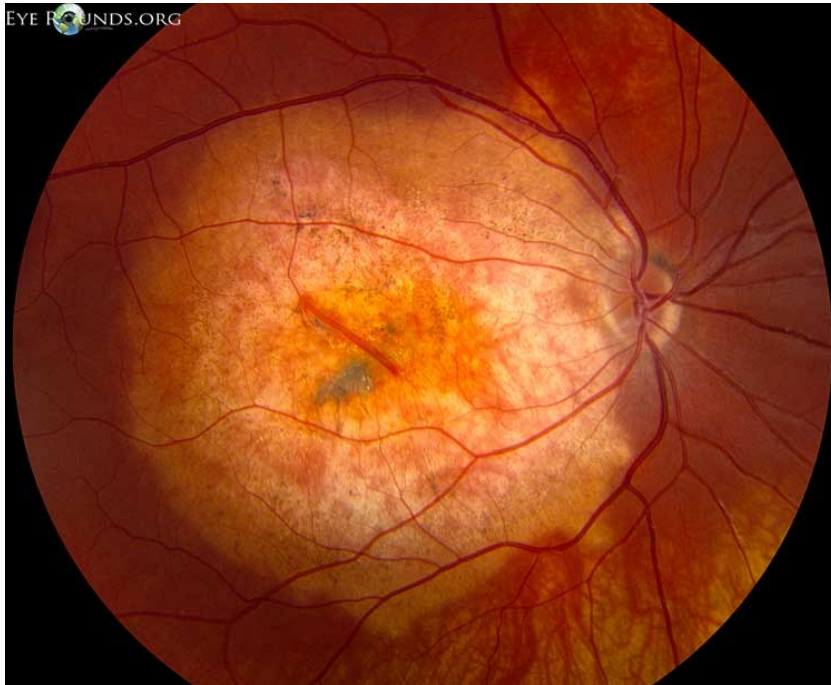


Osteoma: FP, and *b*-scan demonstrating

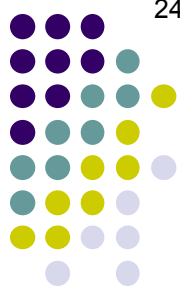
buzzword describing a *b*-scan finding illustrated above



# Intraocular Tumors of Childhood



Osteoma: FP, and *b*-scan demonstrating 'shadowing'

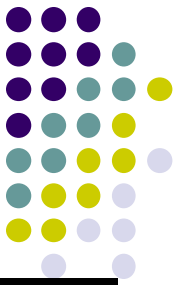


# Intraocular Tumors of Childhood



Osteoma: Another example

# Intraocular Tumors of Childhood



Osteomas (same pt, different cuts).  
Note how bright the lesions are

# Int

*In a word, what is a choroidal osteoma composed of?*

Bone

*Is it common or rare?*



Iris/C

ation. <2 years old.

oma. Self-limited;

s

neoplasia of

s. Can

Tx: Enucleate

darker on light

ciation with NF1,

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4) Isolated/focal choroidal hemangioma

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**RPE**

**Retina**

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Very rare

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*In a word, what is a choroidal osteoma composed of?*

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*Is it common or rare?*

Very rare

*Is it benign or malignant?*

Benign

Iris/C

*Does there a gender predilection?*



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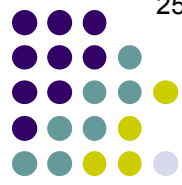
*Is it benign or malignant?*

Benign

*Does there a gender predilection?*

Yes, it is more common in

M v F



ation. <2 years old.

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neoplasia of

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Iris/C

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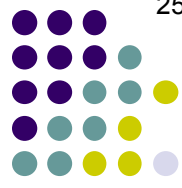
*Is it benign or malignant?*

Benign

## Iris/C

*Does there a gender predilection?*

Yes, it is more common in females



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# Int

*In a word, what is a choroidal osteoma composed of?*

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*Is it benign or malignant?*

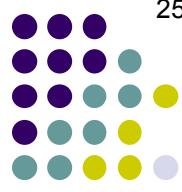
Benign

## Iris/C

*Does there a gender predilection?*

Yes, it is more common in females

*Is it more typically found in pre-teens, or teens?*



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oma. Self-limited;

s

neoplasia of

s. Can

Tx: Enucleate

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*Is it benign or malignant?*

Benign

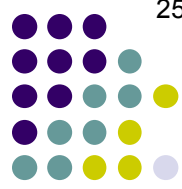
## Iris/C

*Does there a gender predilection?*

Yes, it is more common in females

*Is it more typically found in pre-teens, or teens?*

Teens



ation. <2 years old.

oma. Self-limited;

s

neoplasia of

s. Can

Tx: Enucleate

darker on light

ciation with NF1,

s kids slide-set)

emely rare

## Choroid

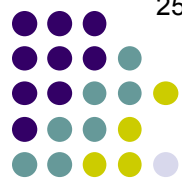
3) **Osteoma**

4) Isolated/focal choroidal hemangioma

5) Diffuse choroidal hemangioma

## RPE

## Retina



# Int

*In a word, what is a choroidal osteoma composed of?*

Bone

*Is it common or rare?*

Very rare

*Is it benign or malignant?*

Benign

# Iris/C

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Yes, it is more common in females

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Teens

*If significant vision loss occurs, what osteoma complication is usually the culprit?*

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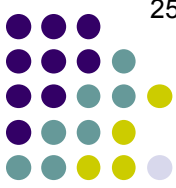
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Choroidal neovascular membrane

## Choroid

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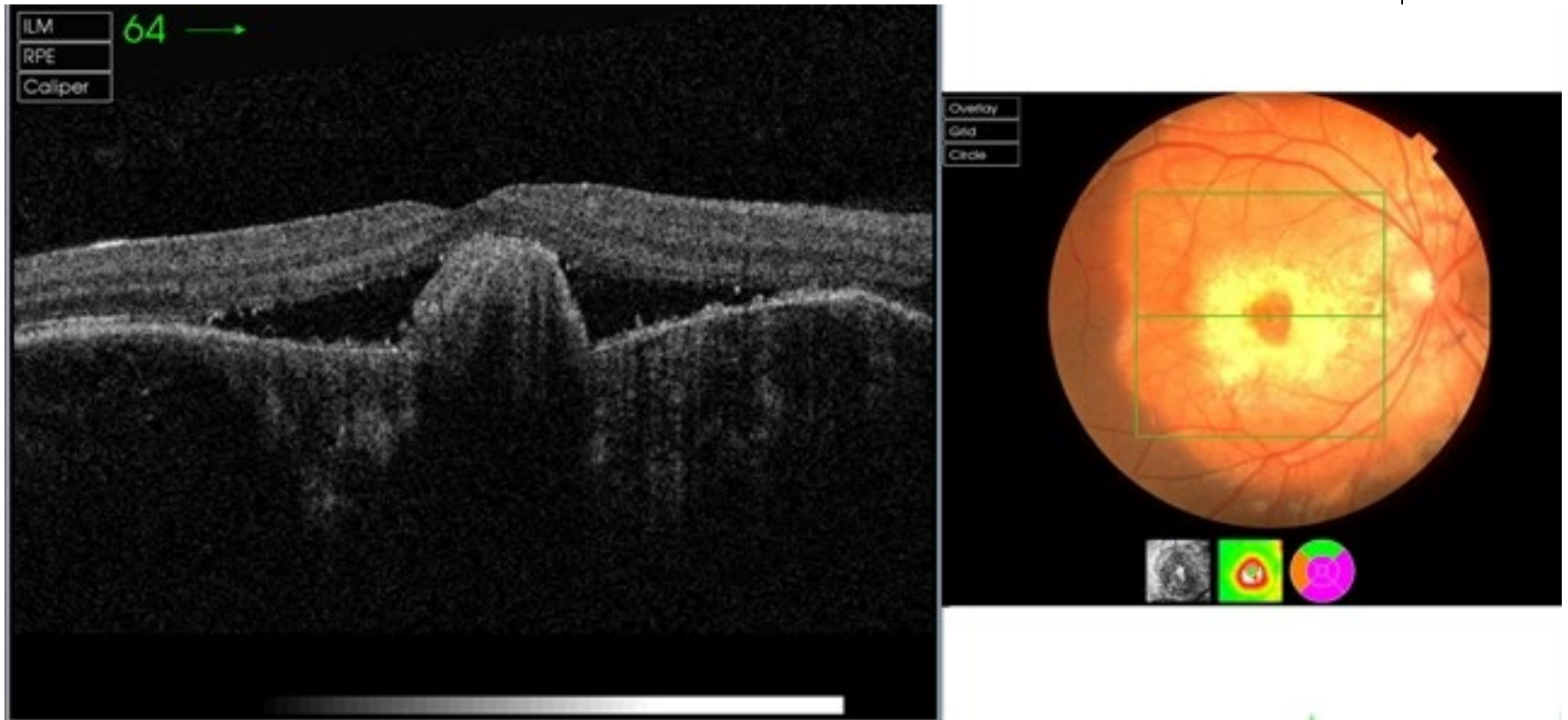
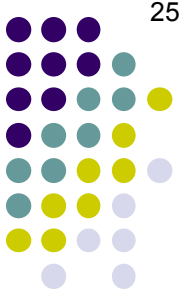
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# Intraocular Tumors of Childhood



Osteoma with CNVM in a 13 y.o. female



# Int

*In a word, what is a choroidal osteoma composed of?*

Bone

*Is it common or rare?*

Very rare

*True osteomas are indeed rare; however, secondary osteoma-like lesions can be found in eyes with what sorts of history?*

*Does there a gender predilection?*

Yes, it is more common in females

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Teens

*If significant vision loss occurs, what osteoma complication is usually the culprit?*

Choroidal neovascular membrane

## Choroid

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Bone

*Is it common or rare?*

Very rare

*True osteomas are indeed rare; however, secondary osteoma-like lesions can be found in eyes with what sorts of history?*

Eyes that have suffered severe chronic inflammation (especially if they become phthisical)

*Does there a gender predilection?*

Yes, it is more common in females

*Is it more typically found in pre-teens, or teens?*

Teens

*If significant vision loss occurs, what osteoma complication is usually the culprit?*

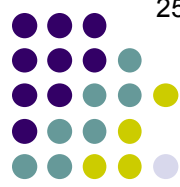
Choroidal neovascular membrane

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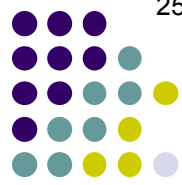
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# Intraocular Tumors of Childhood

Iris/Ciliary Body

**Choroid**

RPE

Retina

*By what other name is this lesion known?*

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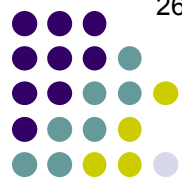


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# Intraocular Tumors of Childhood

Iris/Ciliary Body

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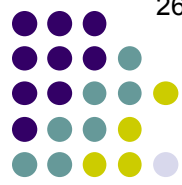


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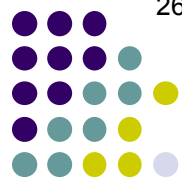


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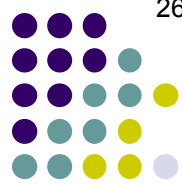


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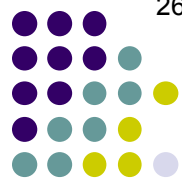


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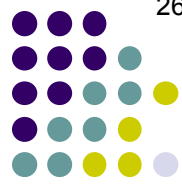
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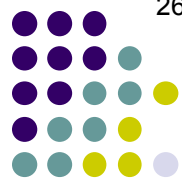


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# Intraocular Tumors of Childhood

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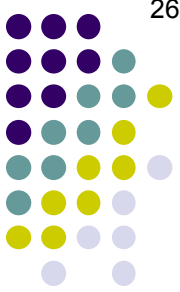
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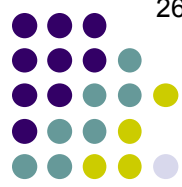
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# Intraocular Tumors of Childhood



Circumscribed choroidal hemangioma



# Intraocular Tumors of Childhood

Iris/Ciliary Body

Choroid

RPE

Retina

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*Is it common, or rare?*

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*Is it associated with a systemic condition, ie, is it syndromic?*

No

*How does it present?*

As a reddish-orange mass in the macula

*What is its characteristic pattern on a-scan ultrasonography?*

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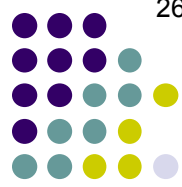
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*What is its characteristic pattern on a-scan ultrasonography?*

It is one of 'high internal reflectivity'

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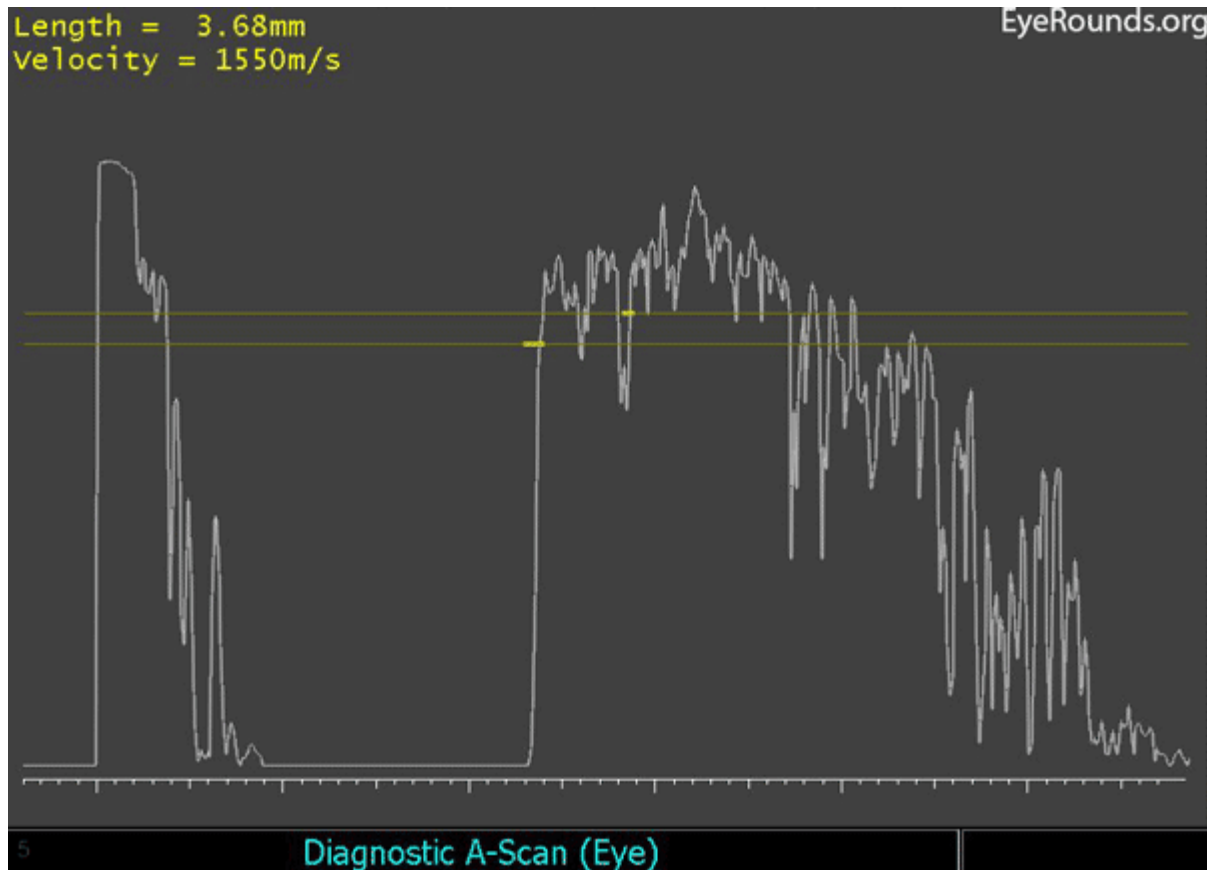
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# Intraocular Tumors of Childhood



Circumscribed choroidal hemangioma: High internal reflectivity on a-scan



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

*By what other name is this lesion known?*

**Circumscribed** choroidal hemangioma

*Is it common, or rare?*

Rare

*Is it associated with a systemic condition, ie, is it syndromic?*

No

*How does it present?*

*'High internal reflectivity'--what other choroidal lesion's a-scan is described the same way?*

## Choroid

It is one of **'high internal reflectivity'**

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# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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No

*How does it present?*

*'High internal reflectivity'--what other choroidal lesion's a-scan is described the same way?*

Choroidal nevus

It is one of **'high internal reflectivity'**

## Choroid

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## RPE

## Retina



# Intraocular Tumors of Childhood

*With what condition is the diffuse choroidal hemangioma associated?*



Iris/C

ration. <2 years old.

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scan pattern

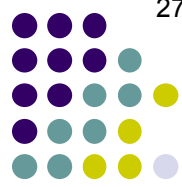
5) **Diffuse choroidal hemangioma**

RPE

Retina

# Intraocular Tumors of Childhood

*With what condition is the diffuse choroidal hemangioma associated?*  
Sturge-Weber syndrome (SWS)



Iris/C

ration. <2 years old.  
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5) **Diffuse choroidal hemangioma**

RPE

Retina

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

*In a word, what sort of condition is SWS?*

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RPE

Retina

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

In a word, what sort of condition is SWS?

A phakomatosis

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RPE

Retina

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

*In a word, what sort of condition is SWS?*

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*What is the noneponymous name for SWS?*

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5) **Diffuse choroidal hemangioma**

RPE

Retina

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

In a word, what sort of condition is SWS?

A phakomatosis

What is the noneponymous name for SWS?

**Encephalotrigeminal angiomatosis** (you might also see **encephalofacial or cerebrofacial angiomatosis**)

ration. <2 years old.

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5) **Diffuse choroidal hemangioma**

RPE

Retina

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

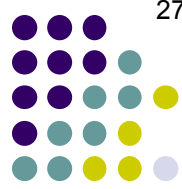
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What is the noneponymous name for SWS?

**Encephalotrigeminal angiomatosis** (you might also see **encephalofacial** or **cerebrofacial angiomatosis**)

What is the hallmark skin finding in SWS?



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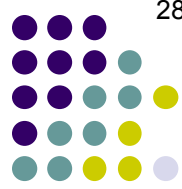
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The **port-wine stain**



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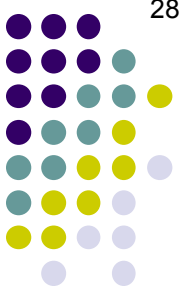
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# Intraocular Tumors of Childhood



Sturge-Weber: Port-wine stain

# Intraocular Tumors of Childhood

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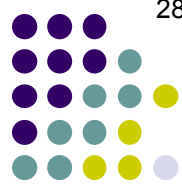
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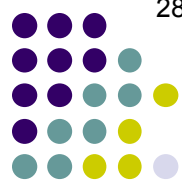
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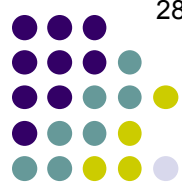
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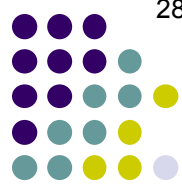
The **port-wine stain**

In one word, what sort of lesion is the port-wine stain?

An angioma

When does it present?

At birth



ration. <2 years old.

oma. Self-limited;

ls

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rs. Can

Tx: Enucleate

darker on light

ciation with NF1,

s kids slide-set)

remely rare

. Risk of CNVM

scan pattern

# Intraocular Tumors of Childhood



Sturge-Weber: Port-wine stain

# Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

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In a word, what sort of condition is SWS?

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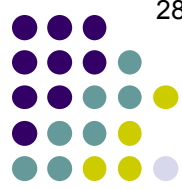
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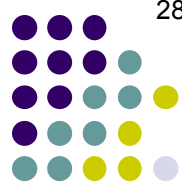
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What is the typical pattern of distribution?

It comports to the distribution of one or more divisions of CN5



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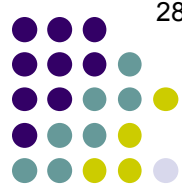
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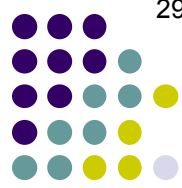
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Does it always present in this manner?

No, some cases will cross the midline of the face



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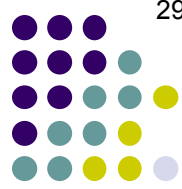
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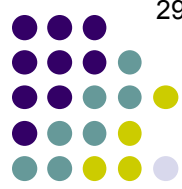
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# Intraocular Tumors of Childhood

*With what condition is the diffuse choroidal hemangioma associated?  
Sturge-Weber syndrome (SWS)*

*Diffuse choroidal hemangioma is present in what percent of SWS?*



Iris/C

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5) **Diffuse choroidal hemangioma**

RPE

Retina

# Intraocular Tumors of Childhood

*With what condition is the diffuse choroidal hemangioma associated?*  
Sturge-Weber syndrome (SWS)

*Diffuse choroidal hemangioma is present in what percent of SWS?*  
About half

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## 5) Diffuse choroidal hemangioma

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Retina

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*What does the fundus look like in an eye with a diffuse choroidal hemangioma?*

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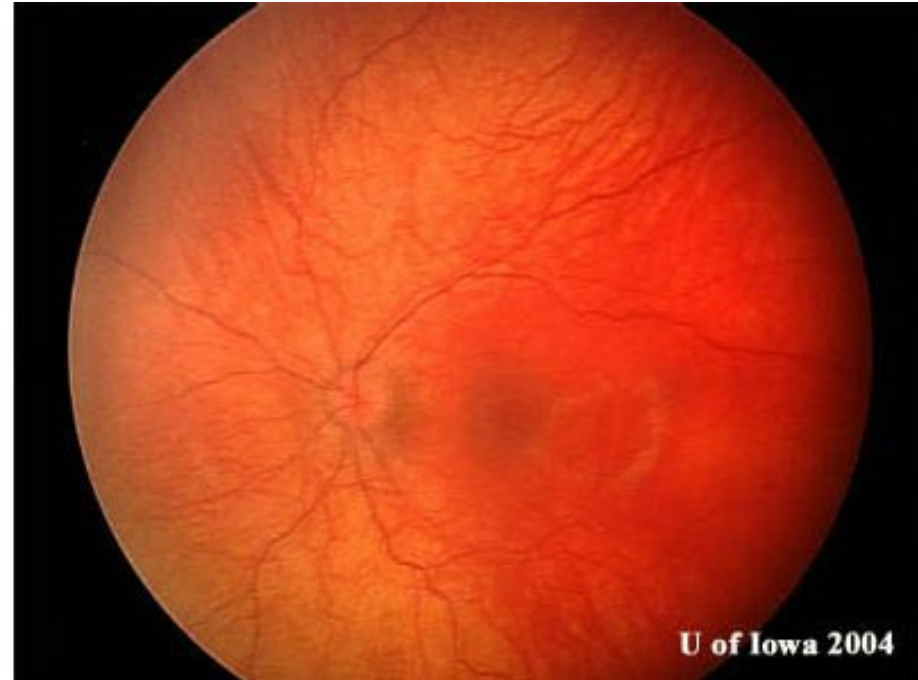
RPE

Retina





# Intraocular Tumors of Childhood



Sturge-Weber: Tomato catsup fundus OD

# Intraocular Tumors of Childhood

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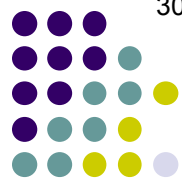
*Can the choroidal hemangioma be present bilaterally?*

Iris/C

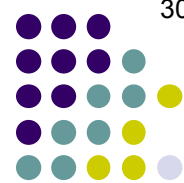
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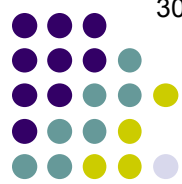
*What food-related term is used to describe the fundus appearance in SWS?*  
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*Can the choroidal hemangioma be present bilaterally?*  
Yes, but it's uncommon

## 5) Diffuse choroidal hemangioma

RPE

Retina



# Intraocular Tumors of Childhood

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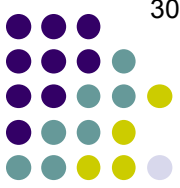
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Yes, but it's uncommon

*Does the choroidal hemangioma have malignant potential?*

## 5) Diffuse choroidal hemangioma

RPE

Retina



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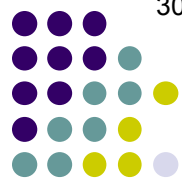
*Does the choroidal hemangioma have malignant potential?*  
No

## 5) Diffuse choroidal hemangioma

RPE

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***For more on SWS, see slide-set P10***

1) Isolated focal choroidal hemangioma. Very rare. Characteristic a  
5) Diffuse choroidal hemangioma

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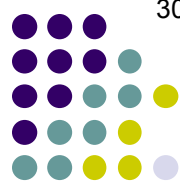
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# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed→hyphema→increased IOP→glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
- 2) **Medulloepithelioma** (aka **diktyoma**): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed→hyphema→increased IOP→glaucoma. Locally invasive→death. Tx: Enucleate
- 3) **Lisch nodules**: Strong association with NF1. Lighter on dark irides; darker on light
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- 5) **Iris mammillations**: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota
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## Choroid

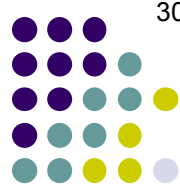
- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**: Usually juxtapapillary. Malignant transformation extremely rare
- 3) **Osteoma**: Benign bony tumor, most common in teen years, females. Risk of CNVM
- 4) **Isolated/focal choroidal hemangioma**: Very rare. Characteristic a-scan pattern
- 5) **Diffuse choroidal hemangioma**: Unilateral. Found in Sturge-Weber syndrome

## RPE

- 1) ?



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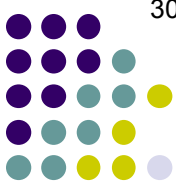
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## RPE

- 1) **Congenital hypertrophy of the RPE (CHRPE)**



## Retina



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*What is the clinical appearance of CHRPE?*

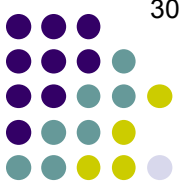
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*What is the clinical appearance of CHRPE?*

Flat, mainly black lesion(s) ranging in size from a 1 mm up to ~10

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**RPE**

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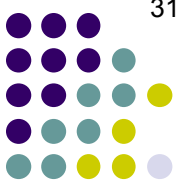
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# Intraocular Tumors of Childhood



CHRPE





# Intraocular Tumors of Childhood

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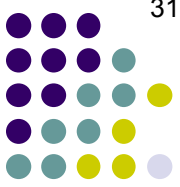
Ch *Is it common, or rare?*  
Common

on extremely rare  
males. Risk of CNVM  
stic a-scan pattern  
Weber syndrome

## RPE

- 1) **Congenital hypertrophy of the RPE (CHRPE)**

## Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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*What is the clinical appearance of CHRPE?*

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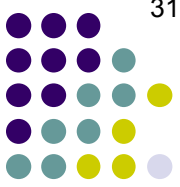
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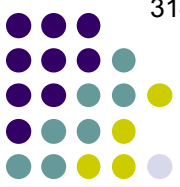
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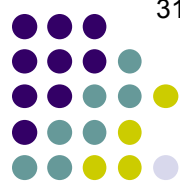
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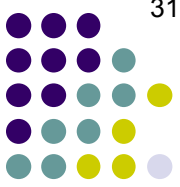
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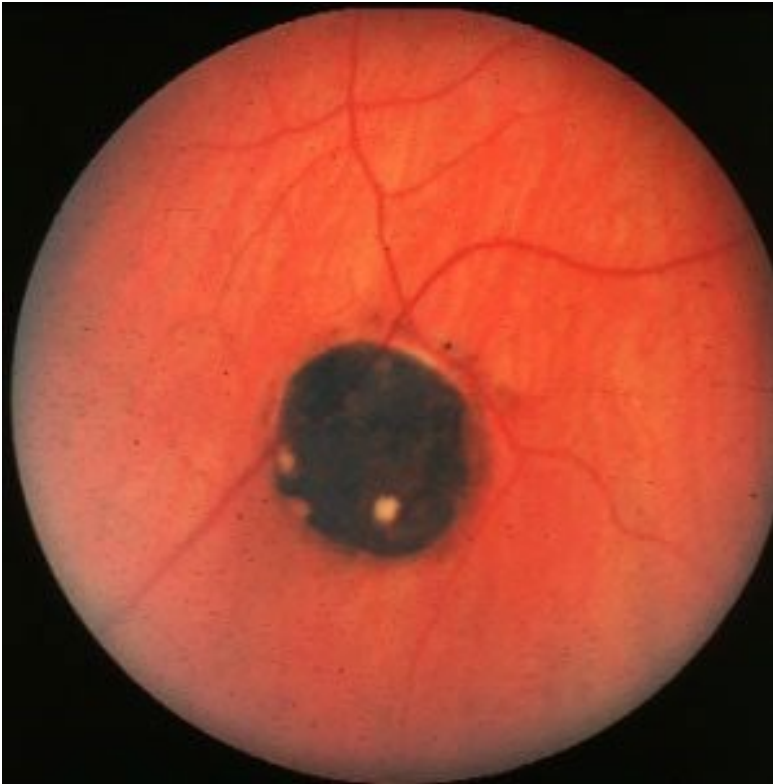
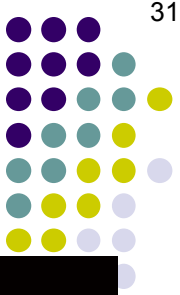
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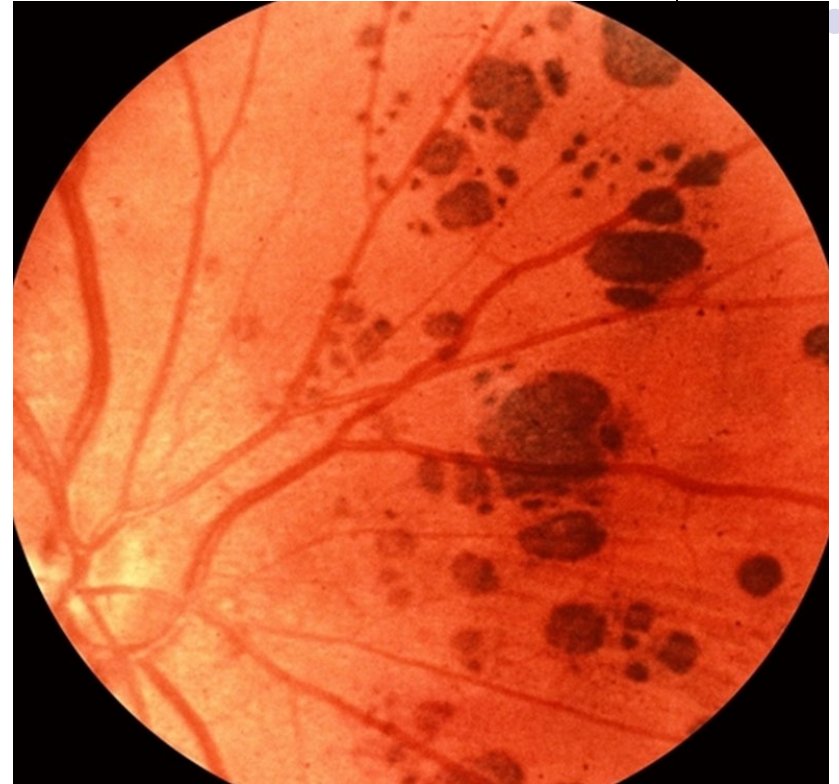
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Retina

# Intraocular Tumors of Childhood



Solitary



Grouped

CHRPE



# Intraocular Tumors of Childhood

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*What descriptive name is used with regard to the appearance of Multifocal/Grouped CHRPE?*



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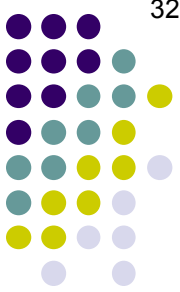
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**'Bear tracks'**

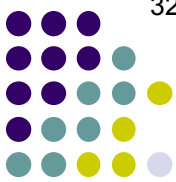
# Intraocular Tumors of Childhood



CHRPE: Bear tracks







# Intraocular Tumors of Childhood

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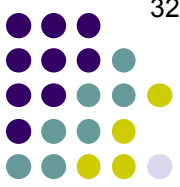
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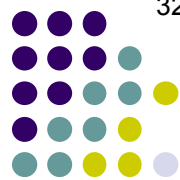
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A **CHRPE-like** lesion is

(both eponymous and non-eponymous)

Familial adenomatous polyposis, aka Gardner syndrome

Take careful note of the modifier 'like' here, because while CHRPE and the lesions associated with Gardner syndrome are ophthalmoscopically similar, they are **not** the same!

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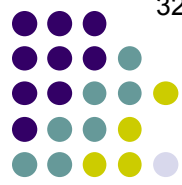
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--If it is **bi- v unilateral** (regular CHRPE is almost always **bi- v unilateral**)

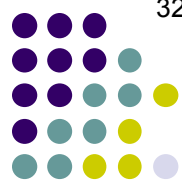
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--If it is bilateral (regular CHRPE is almost always unilateral)

## RPE

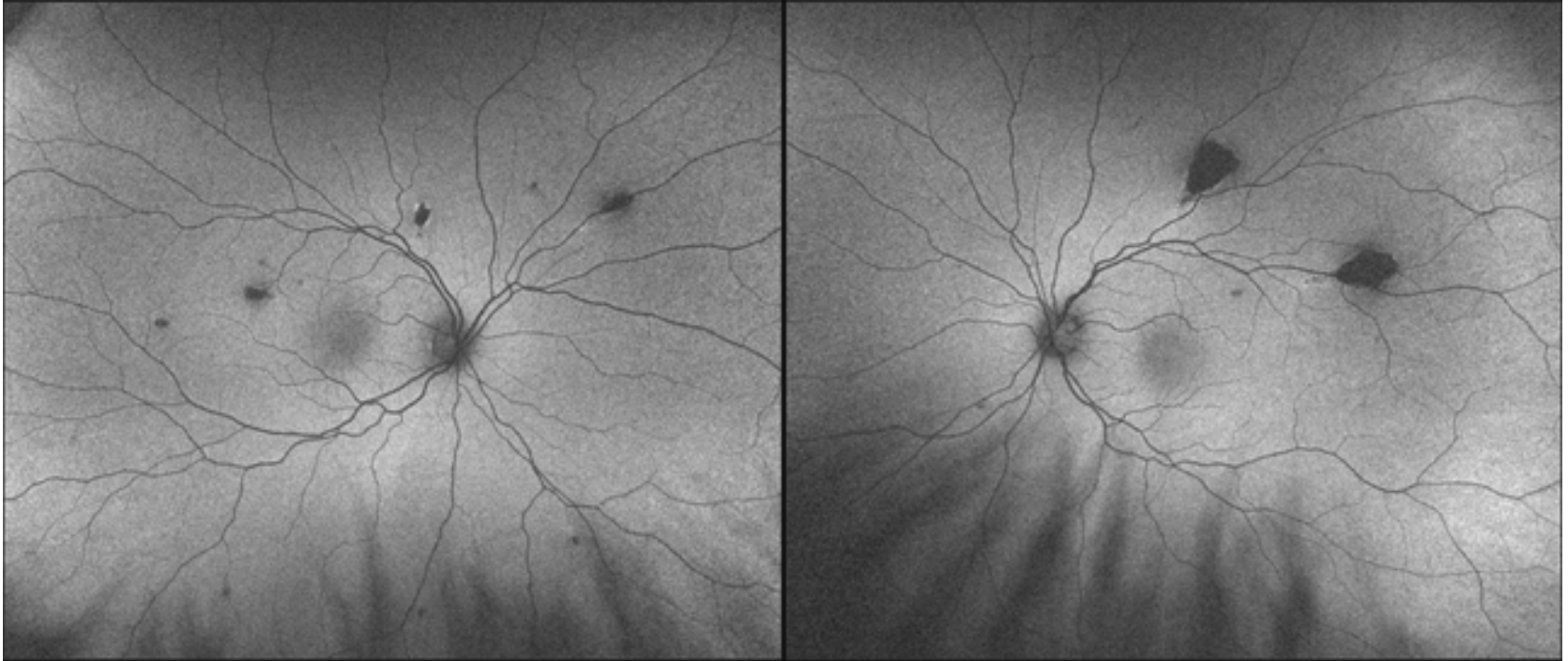
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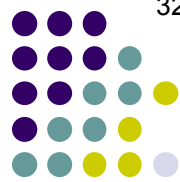
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# Intraocular Tumors of Childhood



CHRPE-like lesions of Gardner syndrome: Bilateral presentation



# Intraocular Tumors of Childhood

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--If it is bilateral (regular CHRPE is almost always unilateral)

--If the lesions are

distribution pattern

## RPE

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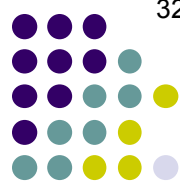
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*What characteristics of a CHRPE-like presentation increase the likelihood that it is a component of Gardner syndrome?*

--If it is bilateral (regular CHRPE is almost always unilateral)

--If the lesions are scattered throughout multiple sectors of the eyes (ie, not 'grouped')

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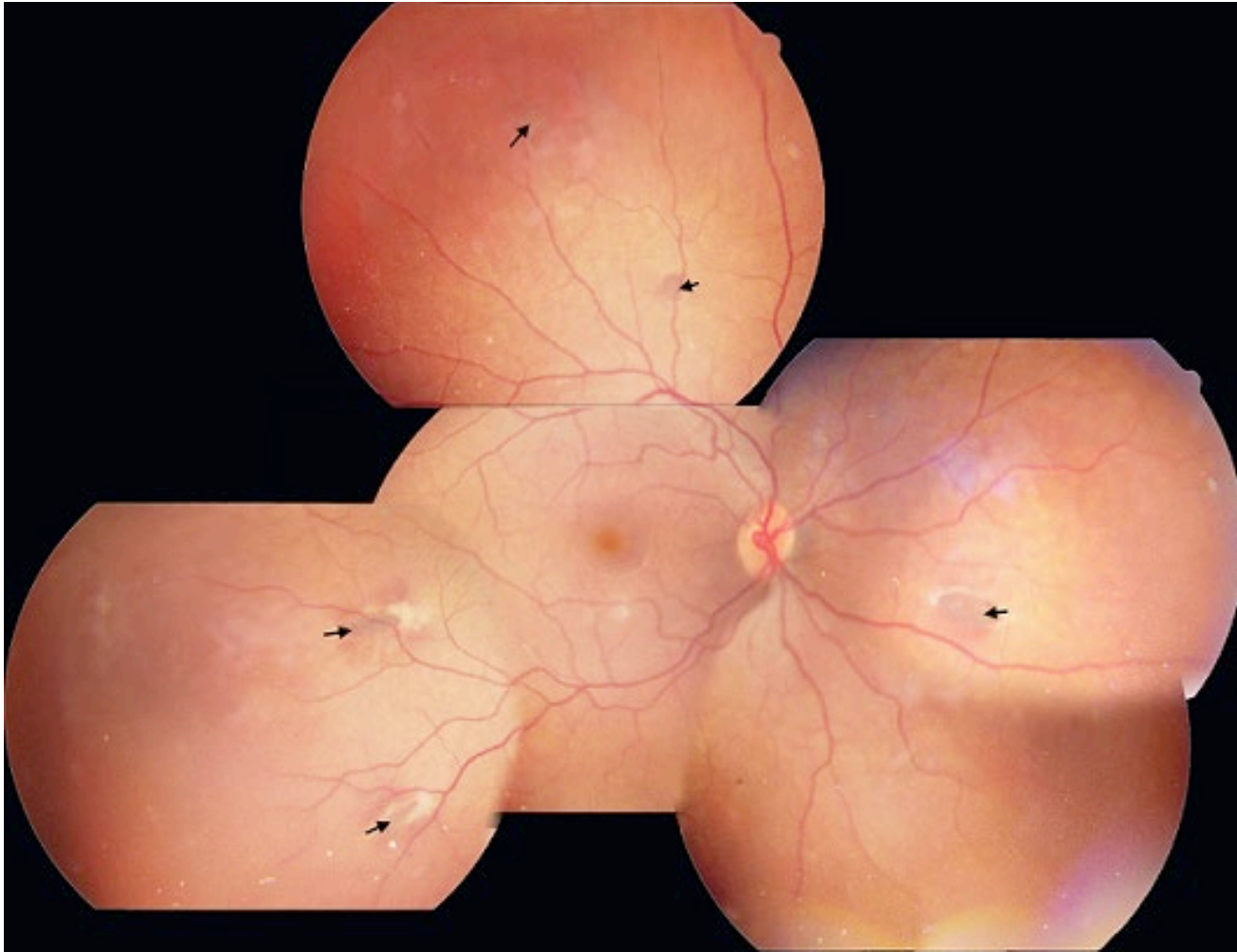
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# Intraocular Tumors of Childhood



CHRPE-like lesions of Gardner syndrome: Scattered distribution



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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*A CHRPE-like lesion is associated with a potentially fatal inherited syndrome. What is the name (both eponymous and noneponymous) of this syndrome?*

Familial adenomatous polyposis, aka **Gardner syndrome**

*What characteristics of a CHRPE-like presentation increase the likelihood that it is a component of Gardner syndrome?*

- If it is bilateral (regular CHRPE is almost always unilateral)
- If the lesions are scattered throughout multiple sectors of the eyes (ie, not 'grouped')
- If the shape of the lesions is rectangle

## RPE

- 1) **Congenital hypertrophy of the RPE (CHRPE)**

*CHRPE is characterized according to its presentation. In what two ways does it present?*

- Solitary** CHRPE
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*What does pisciform mean?*

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*What does pisciform mean?*

It means 'fish-shaped'

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CHRPE-like lesions of Gardner syndrome: Pisciform shape



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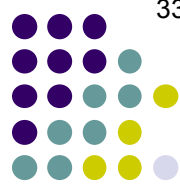
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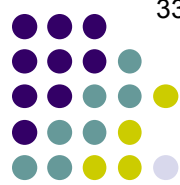
hypo- vs hyperpigmented

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- They are **hypopigmented**
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(CHRPE)

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*The tails of these fish-shaped lesions have two telltale (tell-tail?) characteristics—what are they?*

- They are **hypopigmented**
- They point towards

location in eye

(CHRPE)

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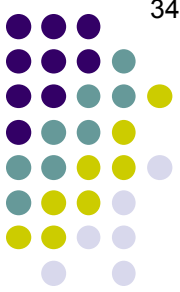
- They are **hypopigmented**
- They point towards the optic nerve head

(CHRPE)

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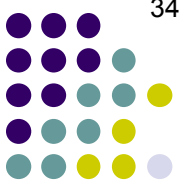
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CHRPE-like lesions of Gardner syndrome: Hypopigmented tail pointing toward ONH



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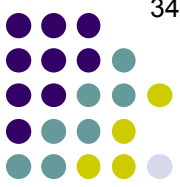
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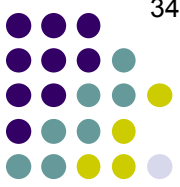
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Gardner syndrome: Colonic polyps





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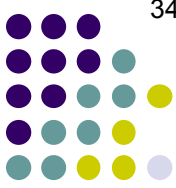
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**40, maybe a little later**

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What proportion of **untreated Gardner syndrome** pts will develop colon cancer?

**All of them**

**What is the treatment of choice?**

By what age will this occur?

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Pts develop thousands of **colonic polyps, a significant number of which are malignant**

What proportion of **untreated Gardner syndrome** pts will develop colon cancer?

All of them

What is the treatment of choice?

Prophylactic colectomy

By what age will this occur?

40, maybe a little later

CHRPE is characterized according to its presentation. In what two ways does it present?

--**Solitary** CHRPE

--**Multifocal** or **Grouped** CHRPE: Large lesion(s) surrounded by a few smaller ones



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
- 2) **Medulloepithelioma** (aka **diktyoma**): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed → hyphema → increased IOP → glaucoma. Locally invasive → death. Tx: Enucleate
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A CHRPE-like lesion is associated with a potentially fatal inherited syndrome. What is the name (both eponymous and noneponymous) of this syndrome?

Familial adenomatous polyposis, aka **Gardner syndrome**

What characterizes Gardner syndrome?

--If it is bilateral

--If the lesions are numerous

--If the lesions are large

**What is the most clinically important (and ominous) component to Gardner syndrome?**

Pts develop thousands of colonic polyps, a significant number of which are malignant

**Other than the colonic and RPE lesions, what are the findings in Gardner syndrome?**

--?

--?

--?

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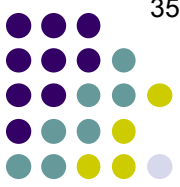
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- Benign tumors of the [redacted]
- Benign tumors of [redacted]
- ?

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Familial adenomatous polyposis, aka **Gardner syndrome**

What characterizes Gardner syndrome?

- If it is benign
- If the lesion is malignant
- If the syndrome is hereditary

**What is the most clinically important (and ominous) component to Gardner syndrome?**

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**Other than the colonic and RPE lesions, what are the findings in Gardner syndrome?**

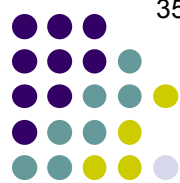
- Benign tumors of the **skin**
- Benign tumors of **bone**

--?

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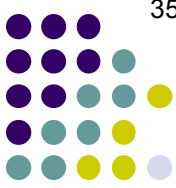
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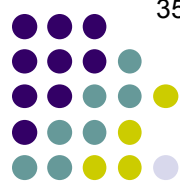
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- Benign tumors of **bone**
- Dental** anomalies

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Speaking of eye dentistry: When a pt has teeptus issues, several conditions should spring immediately to mind. One is Gardners; what are the other three?

--Gardner syndrome

--?

--?

--?

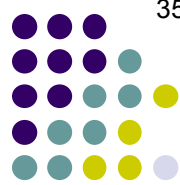
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- Gardner syndrome
- Congenital syphilis
- Axenfeld-Reiger
- Incontinentia pigmenti

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A CHRPE-like lesion is associated with a potentially fatal inherited syndrome. What is the name (both eponymous and noneponymous) of this syndrome? (slide-set)

Familial adenomatous polyposis

Speaking of eye dentistry:

What should spring immediately

of Gardner syndrome

--If it's **Congenital syphilis**

--If it's Axenfeld-Reiger

--If it's Incontinentia pigmenti

What is the eponym for the dental abnormalities in congenital syphilis?

--Rieger tumors of bone

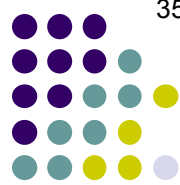
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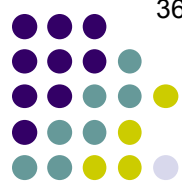
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What is the eponym for the dental abnormalities in congenital syphilis?  
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What is the classic description re the shape of Hutchinson teeth?

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**Dental anomalies**

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Speaking of eye dentistry:

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What is the eponym for the dental abnormalities in congenital syphilis?  
Hutchinson teeth

What is the classic description re the shape of Hutchinson teeth?  
'Peg-shaped'

(For more on congenital syphilis, see slide-set K4)

--Benign tumors of bone

**Dental anomalies**

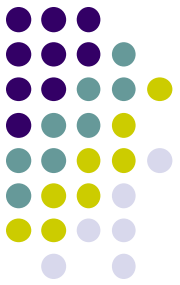
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# Intraocular Tumors of Childhood



Congenital syphilis: Hutchinson teeth



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In three words, what sort of condition is A-R?

An

syphilis, see slide-set K4)

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An anterior-segment dysgenesis

(for more on A-R, see slide-set FELT7)

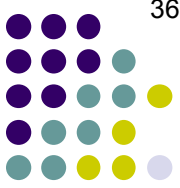
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Congenital syphilis, see slide-set K4)

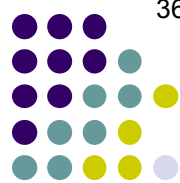
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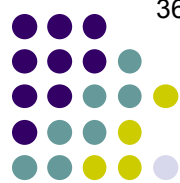
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Familial adenomatous polyposis

Speaking of eye dentistry:

What should spring immediately

--Gardner syndrome

--Congenital syphilis

--Axenfeld-Reiger

--Incontinentia pigmenti

What is the eponym for the dental abnormalities in congenital syphilis?  
Hutchinson teeth

What is the classic description re the shape of Hutchinson teeth?

In three words, what sort of condition is A-R?

What is the eponymous name of IP?

Bloch-Sulzberger syndrome

In one word, what sort of condition is IP?

A

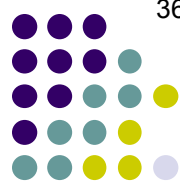
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Retina

**Dental anomalies**



# Intraocular Tumors of Childhood

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(For more on IP, see slide-set P10)

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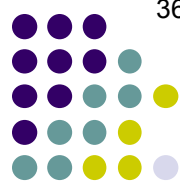
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## RPE

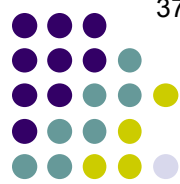
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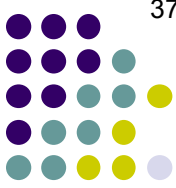
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How is Muir-Torre pronounced?

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mure (rhymes with 'pure') tore-ay

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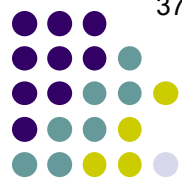
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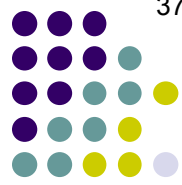
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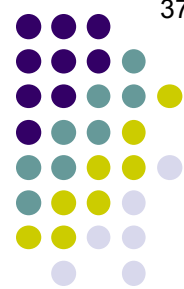
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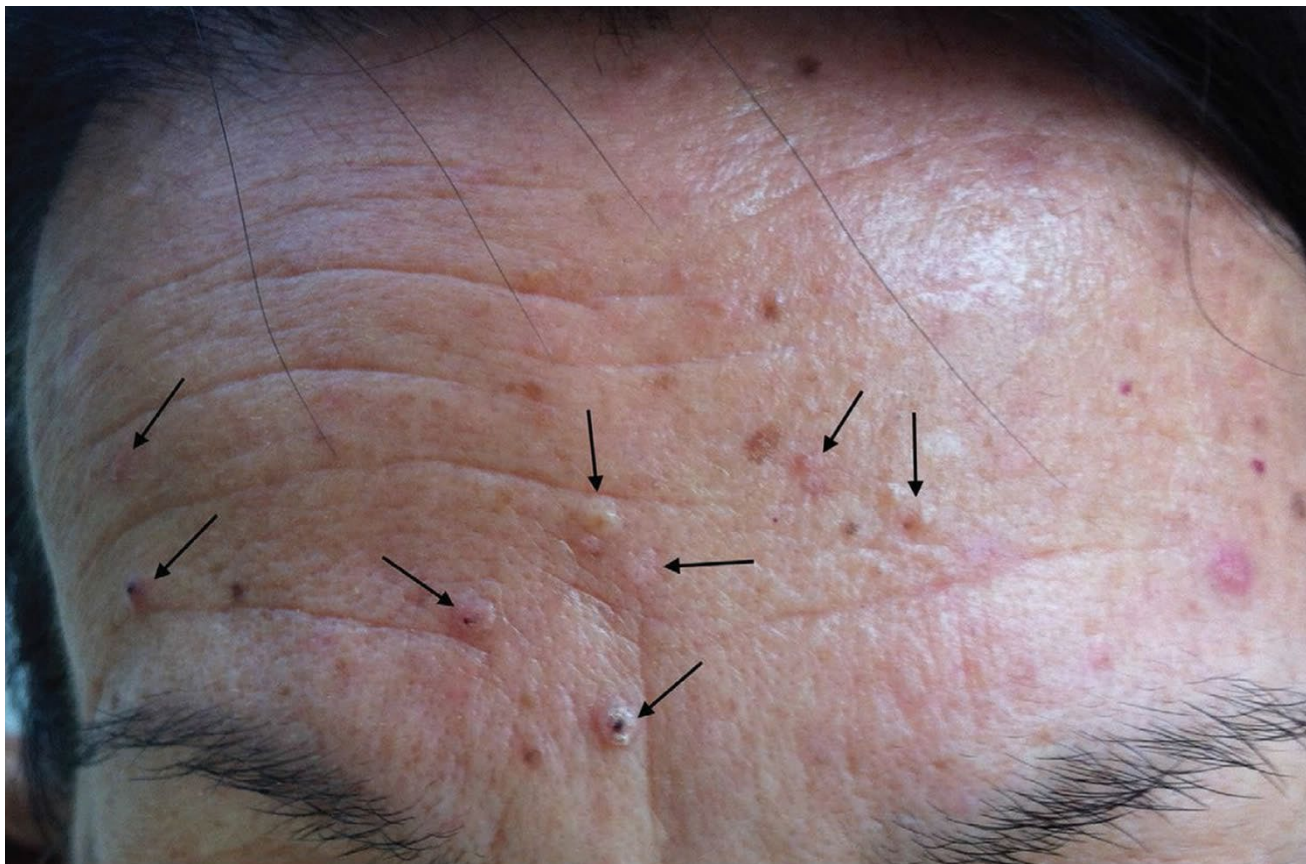
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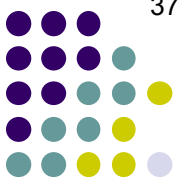


# Intraocular Tumors of Childhood



Multiple skin-colored to yellow-pink papules (arrows) on the face of a 64-year-old woman with a history of colon and cervical cancer. A skin biopsy confirmed a diagnosis of sebaceous adenoma resulting from Muir-Torre syndrome





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What sorts of sebaceous lesions?

--?  
--?  
--?

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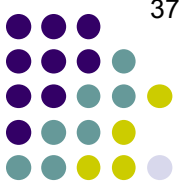
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- Sebaceous-cell carcinomas
- Sebaceous-cell adenomas
- Basal-cell carcinomas with sebaceous differentiation

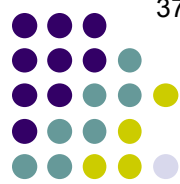
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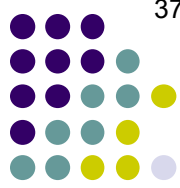
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No; Muir-Torre is an example of a disease spectrum called Hereditary **Non-Polyposis Colorectal Cancer**

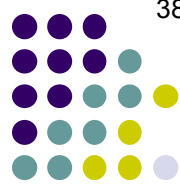
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How is Peutz-Jeghers pronounced?

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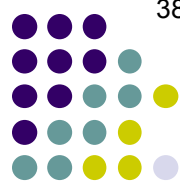
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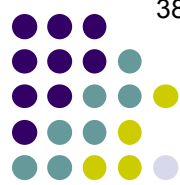
What is the main ophthalmic manifestation of Peutz-Jeghers syndrome?

CHRPE is it present?

--Solitary CHRPE

--**Multifocal** or **Grouped CHRPE**: Large lesion(s) surrounded by a few smaller ones

Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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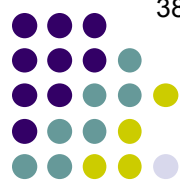
Simple lentiginos of (but not necessarily limited to) the eyelids

Cherry-red spot is it present?

--Century CHRPE

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Retina



# Intraocular Tumors of Childhood

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**Simple lentigines**

What are simple lentigines?

C

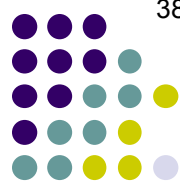
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Flat melanocytic lesions histologically similar to ephelides

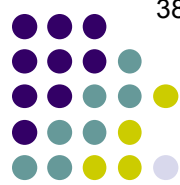
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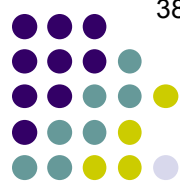
What are ephelides (singular, ephelis)?

C

--Century 19th C

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Retina



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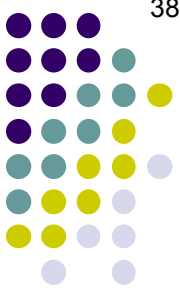
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Freckles

C

--Century 19th C

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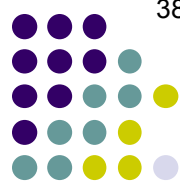
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# Intraocular Tumors of Childhood



Peutz-Jeghers syndrome: Eyelid simple lentigines



# Intraocular Tumors of Childhood

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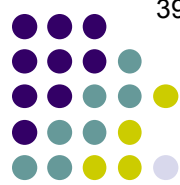
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--Century CFM

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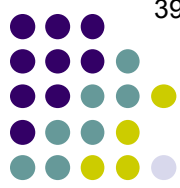
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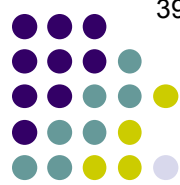
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Does lentigo simplex have malignant potential?



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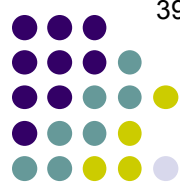
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I coulda sworn lentigo simplex had malignant potential. You sure about this?

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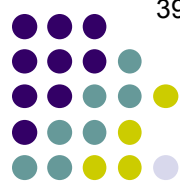
--Century 19th C

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Retina

**Does lentigo simplex have malignant potential?**

No



# Intraocular Tumors of Childhood

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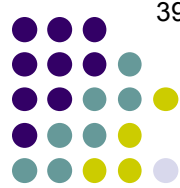
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You sure about this?

Yes, I'm sure. You're thinking of **two words**,  
a pre-malignant melanocytic lesion of the skin.

**Does lentigo simplex have malignant potential?**  
**No**



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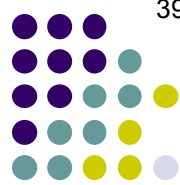
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You sure about this?

Yes, I'm sure. You're thinking of **lentigo maligna**, a pre-malignant melanocytic lesion of the skin.

**Does lentigo simplex have malignant potential?**

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**Simple lentigines** of (but not necessarily limited to) the eyelids

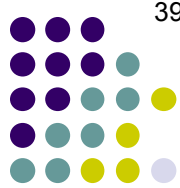
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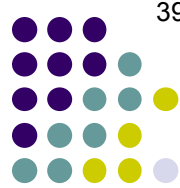
No, pigmented lesions of the [ ] region are the classic/most common finding

Is it present?

--Century 19th C

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Retina



# Intraocular Tumors of Childhood

## Iris/Ciliary Body

- 1) **Juvenile xanthogranuloma (JXG)**: Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed → hyphema → increased IOP → glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells
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- 3) **Lisch nodules**: Strong association with NF1. Lighter on dark irides; darker on light
- 4) **Brushfield spots**: Strong association with Down syndrome
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A CHRPE-like lesion is associated with a potentially fatal inherited syndrome. What is the name (both eponymous and noneponymous) of this syndrome?

Familial adenomatous polyposis, aka **Gardner syndrome**

When 'colon cancer + ophthalmic issue' is mentioned, three syndromes should come to mind.

One is Gardner syndrome. What is the other?

Muir-Torre syndrome and **Peutz-Jeghers syndrome**

--If it's bilateral (regular CHRPE is almost always unilateral)

--If the lesion

--If the shape

What is the main ophthalmic manifestation of Peutz-Jeghers syndrome?

**Simple lentigines** of (but not necessarily limited to) the eyelids

Are lentigo simplex eyelid lesions the classic harbinger of Peutz-Jeghers syndrome?

No, pigmented lesions of the perioral region are the classic/most common finding

What is the classic ophthalmic finding in Peutz-Jeghers syndrome?

--Unilateral CHRPE

--**Multifocal** or **Grouped CHRPE**: Large lesion(s) surrounded by a few smaller ones

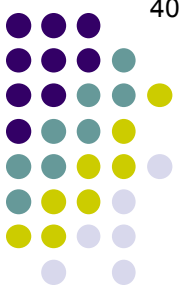
Retina



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Characteristic circumoral pigmentation in a patient with Peutz-Jeghers syndrome

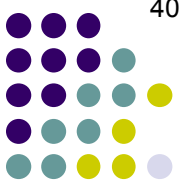


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Speaking of: Did you notice the pigmented lip lesions in this pic?





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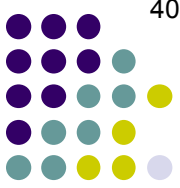
**Does Peutz-Jeghers present with multiple adenomatous polyps of the colon a la Gardner syndrome?**

Yes, it is present?

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Does Peutz-Jeghers present with multiple adenomatous polyps of the colon a la Gardner syndrome?

Yes

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## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**: Usually juxtapapillary. Malignant transformation extremely rare
- 3) **Osteoma**: Benign bony tumor, most common in teen years, females. Risk of CNVM
- 4) **Isolated/focal choroidal hemangioma**: Very rare. Characteristic a-scan pattern
- 5) **Diffuse choroidal hemangioma**: Unilateral. Found in Sturge-Weber syndrome

## RPE

- 1) **Congenital hypertrophy of the RPE (CHRPE)**: Common. Deeply pigmented. A CHRPE-like finding is associated with Gardner syndrome

## Retina

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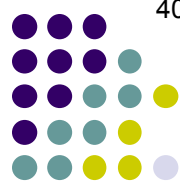
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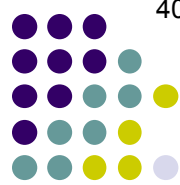
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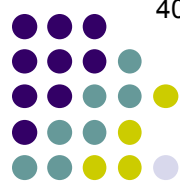
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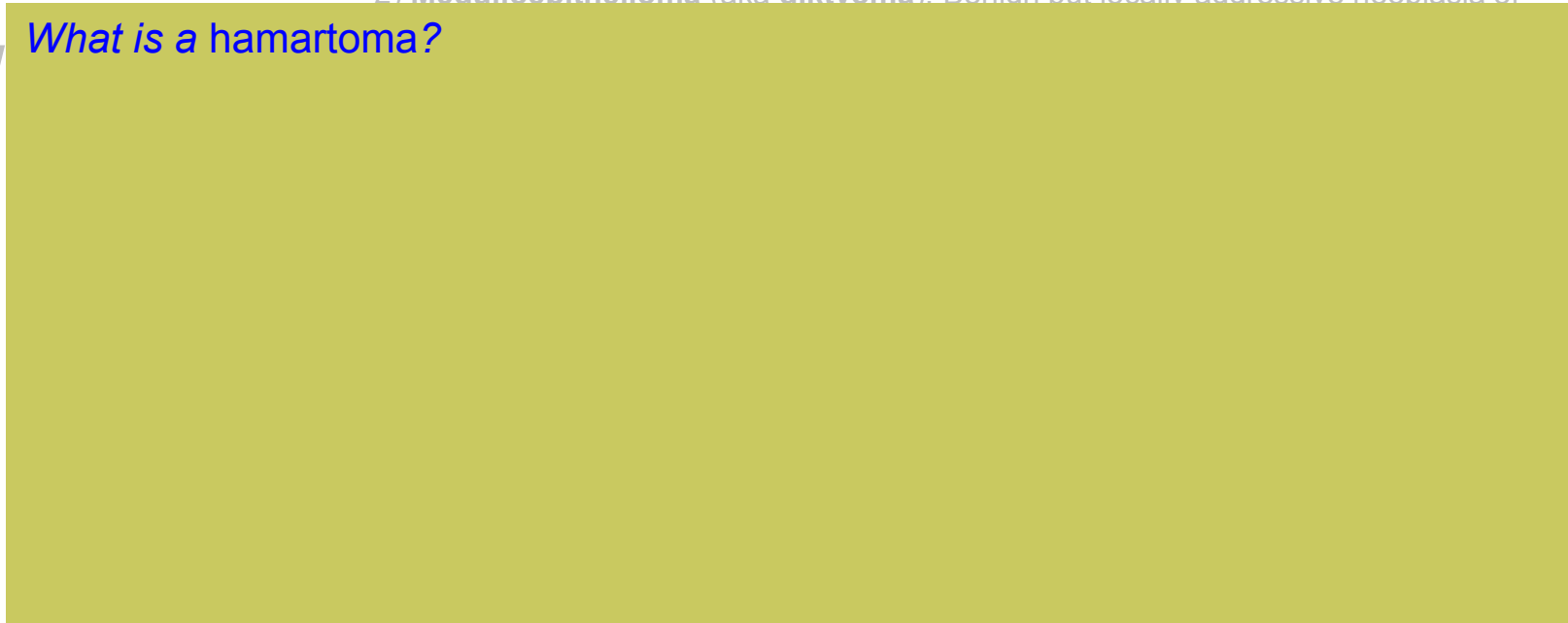
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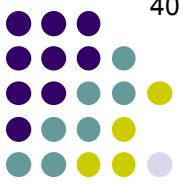
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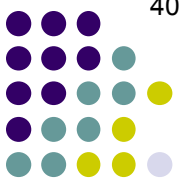
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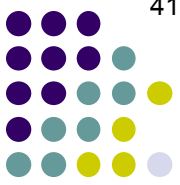


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# Intraocular Tumors of Childhood

What is the name of the reverse clinical entity, ie, one with normal cells found in an abnormal location?

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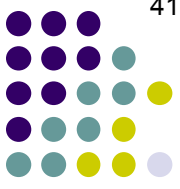
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*That a lesion is a hamartoma (or choristoma) indicates what about its onset?*

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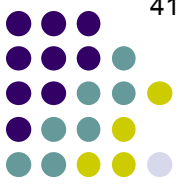
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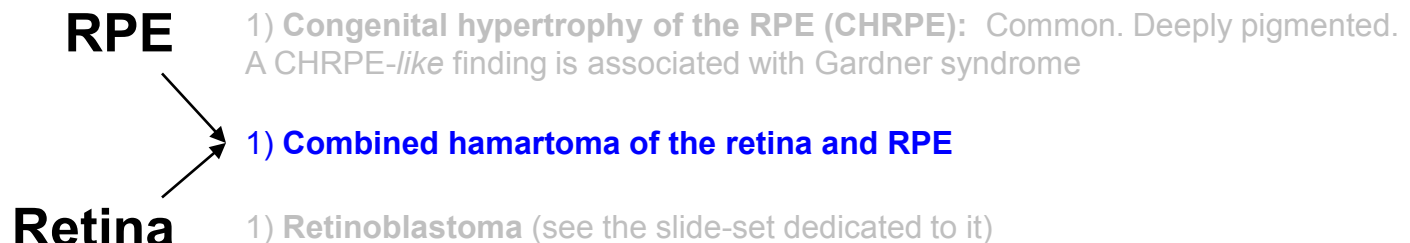
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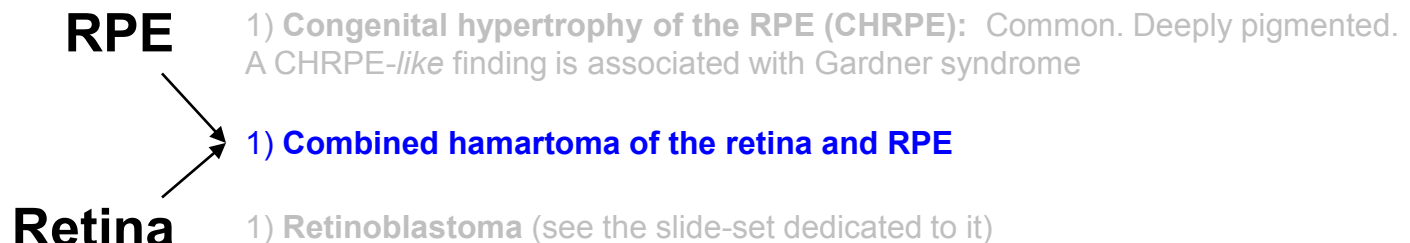
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*How does it present clinically?*

As a variably pigmented, slightly elevated retinal mass of the area retina

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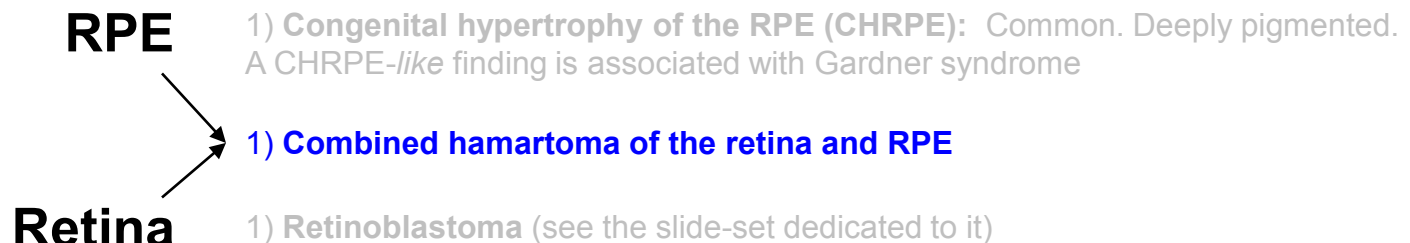
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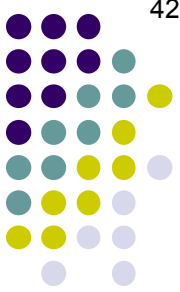
RPE cells (duh) and retinal glial cells

*How does it present clinically?*

As a variably pigmented, slightly elevated retinal mass of the peripapillary retina



# Intraocular Tumors of Childhood



Combined hamartoma of retina and RPE



# Intraocular Tumors of Childhood

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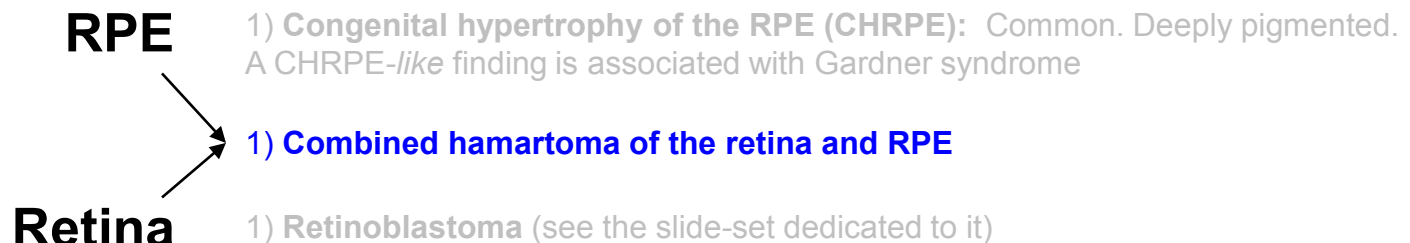
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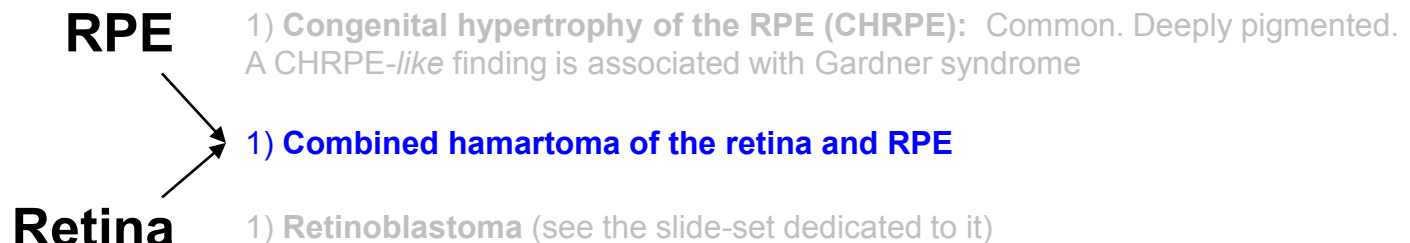
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Choroidal melanoma—eyes have been enucleated because of this misdiagnosis





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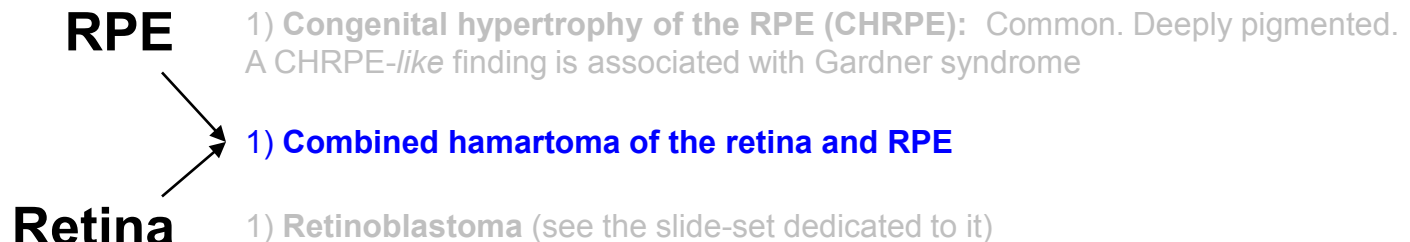
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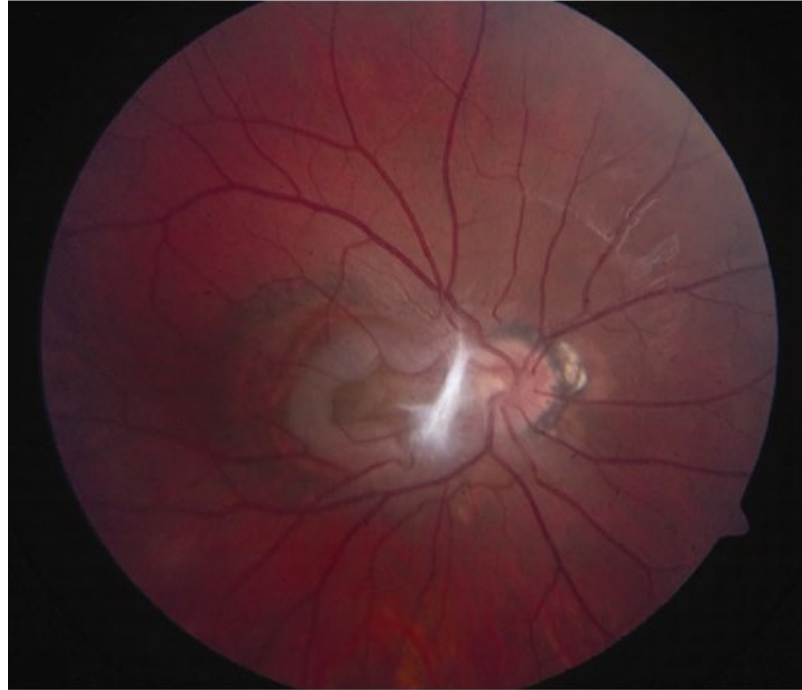
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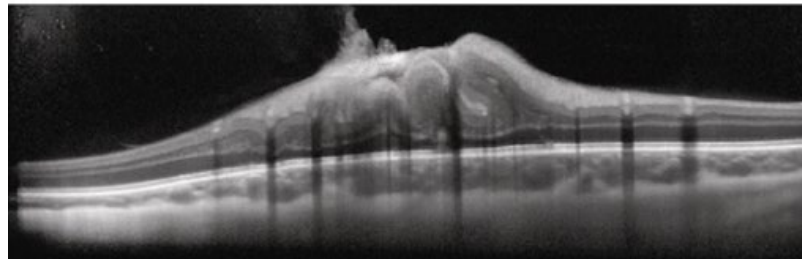
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# Intraocular Tumors of Childhood



( a )



( b )

Combined hamartoma of retina and RPE.  
Note the entire lesion is *above* Bruchs





# Intraocular Tumors of Childhood

## Iris/Ciliary Body

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- 2) **Medulloepithelioma** (aka **diktyoma**): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed→hyphema→increased IOP→glaucoma. Locally invasive→death. Tx: Enucleate
- 3) **Lisch nodules**: Strong association with NF1. Lighter on dark irides; darker on light
- 4) **Brushfield spots**: Strong association with Down syndrome
- 5) **Iris mammillations**: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota
- 6) **Iris cysts**: Can be pupillary, stromal, secondary (see the *Iris issues is kids* slide-set)

## Choroid

- 1) **Nevus**: Common. Benign
- 2) **Melanocytoma**: Usually juxtapapillary. Malignant transformation extremely rare
- 3) **Osteoma**: Benign bony tumor, most common in teen years, females. Risk of CNVM
- 4) **Isolated/focal choroidal hemangioma**: Very rare. Characteristic a-scan pattern
- 5) **Diffuse choroidal hemangioma**: Unilateral. Found in Sturge-Weber syndrome

## RPE

- 1) **Congenital hypertrophy of the RPE (CHRPE)**: Common. Deeply pigmented. A CHRPE-like finding is associated with Gardner syndrome

## Retina

- 1) **Combined hamartoma of the retina and RPE**: Benign, congenital retinal lesion
- 1) **Retinoblastoma** (see the slide-set dedicated to it)