Intraocular Tumors of Childhood

Four intraocular locations (ie, structures/tissues)
Intraocular Tumors of Childhood

Four intraocular locations (ie, structures/tissues)

- 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) ?

Choroid

RPE

Retina

Six tumors of the iris/ciliary body
Intraocular Tumors of Childhood

1) Juvenile xanthogranuloma
   - Nonneoplastic histiocytic proliferation
   - +/− 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
   - 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; 15% of non-Down pop

5) Iris mammillations
   - Tiny, numerous
   - Same color as iris
   - Weak association with NF1, Nevus of Ota

6) Iris cysts

Six tumors of the iris/ciliary body
1) **Juvenile xanthogranuloma**

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

It is a...

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**Iris/Ciliary Body**

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

2. **Medulloepithelioma**: Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed \(\rightarrow\) hyphema \(\rightarrow\) increased IOP \(\rightarrow\) glaucoma. Locally invasive \(\rightarrow\) 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; 15% of non-Down pop.

5. **Iris mammillations**: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota.

6. **Iris cysts**: Can be pupillary, stromal, secondary.
1) **Juvenile xanthogranuloma**

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

**Iris/Ciliary Body**

**Choroid**

**RPE**

**Retina**

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1) **Juvenile xanthogranuloma**: 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**: 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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6) **Iris cysts**: Can be pupillary, stromal, secondary.

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

1) Juvenile xanthogranuloma

*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)*
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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

Iris mammillations

In three words, what sort of condition is Iris mammillations?

It is a __tiny, numerous. Same color with iris__

Iris cysts

In three words, what sort of condition is Iris cysts?

It is a __can be pupillary, stromal, secondary__

Intraocular Tumors of Childhood

1) **Juvenile xanthogranuloma**

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Iris mammillations

In three words, what sort of condition is Iris mammillations?

It is a __tiny, numerous. Same color with iris__

Iris cysts

In three words, what sort of condition is Iris cysts?

It is a __can be pupillary, stromal, secondary__
Intraocular Tumors of Childhood

JXG: Skin papules. The orangish color is classic
Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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?*

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?
Intraocular Tumors of Childhood

1) **Juvenile xanthogranuloma**

*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 age 2

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Iris/Ciliary Body

Choroid

RPE

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

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

RPE

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

   *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?*

2) **Medulloepithelioma**

   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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   Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota.

6) **Iris cysts**

   Can be pupillary, stromal, secondary.
Iris/Ciliary Body

1) Juvenile xanthogranuloma

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

Intraocular Tumors of Childhood
Intraocular Tumors of Childhood

JXG: Iris lesion
Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

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

*In what three ways are the iris nodules clinically significant?*
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Intraocular Tumors of Childhood
Intraocular Tumors of Childhood

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

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

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*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

*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
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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)*
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Choroid

RPE

Retina
Intraocular Tumors of Childhood

JXG: Spontaneous hyphema
1) **Juvenile xanthogranuloma**

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*What is the natural history of JXG?*
It is self-limited, usually resolving by age 5 years
# Intraocular Tumors of Childhood

## Iris/Ciliary Body

1. **Juvenile xanthogranuloma**

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

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## Choroid

## RPE

## Retina
Intraocular Tumors of Childhood

1) **Juvenile xanthogranuloma**

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

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

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Intraocular Tumors of Childhood

Iris/Ciliary Body

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*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 are the two hallmarks of JXG histology?*
Touton giant cells
'foamy macrophages'

*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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Intraocular Tumors of Childhood

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

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As orangish skin papules

*At what age does it present?*

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

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*What are the two hallmarks of JXG histology?*
- The presence of **giant cells**
- The presence of **Touton giant cells**

*When Unilateral?*

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*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
### Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*In three words, what sort of condition is JXG?*
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*What are the two hallmarks of JXG histology?*
The presence of Touton giant cells

*In what three ways are the iris nodules bleeding clinically significant?*
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*What is the natural history of JXG?*
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### Choroid

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### RPE

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### Retina
Intraocular Tumors of Childhood

Touton giant cells

Foamy macrophages

JXG
Intraocular Tumors of Childhood

1) **Juvenile xanthogranuloma**

*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 years

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

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

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**Iris/Ciliary Body**

1. **Juvenile xanthogranuloma**

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

- **What are the two hallmarks of JXG histology?**
  - Touton giant cells
  - ‘Foamy macrophages’

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

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**Choroid**

**RPE**

**Retina**
1) **Juvenile xanthogranuloma**

*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

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

*When Unilateral?*  

*This histology—'foamy macrophages'—is often described with other, equivalent terms. What are they?*  
Foamy = ‘lipid filled’  
Macrophages = ‘histiocytes’

---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

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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)*
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*At what age does it present?*
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*What are the two hallmarks of JXG histology?*
The presence of Touton giant cells
The presence of ‘foamy macrophages’

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

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

*This histology—‘foamy macrophages’—is often described with other, equivalent terms. What are they?*
Foamy = ‘lipid filled’
Macrophages = ‘histiocytes’

*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*
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) Juvenile xanthogranuloma

*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

*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

*What are the two hallmarks of JXG histology?*

The presence of Touton giant cells

The presence of ‘foamy macrophages’

Speaking of ‘foamy macrophages’...

What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis? First clue--more forthcoming

Retina

Choroid
Intraocular Tumors of Childhood

**Iris/Ciliary Body**

1) **Juvenile xanthogranuloma**

*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

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

**Retina**

Speaking of ‘foamy macrophages’…  
What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?  
And a hx of chronic migratory arthritis?  
Clue #2
1) **Juvenile xanthogranuloma**

*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

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

*When are these lesions unilateral or bilateral?*
Unilateral

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**Speaking of ‘foamy macrophages’...**

*What dz comes to mind if, instead of a young child 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 diarrhea?*  

Need another?
1) **Juvenile xanthogranuloma**

*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

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

*Speaking of ‘foamy macrophages’…*
What dz comes to mind if, instead of a young child 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 diarrhea?
And CNS symptoms--seizures, dementia, coma?  

*Last chance--answer is next!*
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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

*What are the two hallmarks of JXG histology?*

The presence of Touton giant cells

*When unilateral?*

When unilateral

Unilateral

*When bilateral?*

Bilateral

*‘foamy macrophages’*

Speaking of ‘foamy macrophages’…

*What dz comes to mind if, instead of a young child 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 diarrhea?*

*And CNS symptoms--seizures, dementia, coma?*

*Whipple’s disease*

Retina
1) **Juvenile xanthogranuloma**

*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 years

*What are the two hallmarks of JXG histology?*

- The presence of Touton giant cells
- The presence of ‘foamy macrophages’

*When unilateral?*

Unilateral

*Speaking of ‘foamy macrophages’…*

What dz comes to mind if, instead of a young child 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 diarrhea? And CNS symptoms—seizures, dementia, coma?

**Whipple’s disease**

Broadly speaking, what sort of condition is Whipple’s?
Intraocular Tumors of Childhood

1) Juvenile xanthogranuloma

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

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

When
Unilateral

In three words, what sort of condition is Whipple’s?
It is infectious

Speaking of ‘foamy macrophages’...
What dz comes to mind if, instead of a young child
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 diarrhea?
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Whipple’s disease
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

*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.

*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.

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

Speaking of ‘foamy macrophages’…  
*What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?*  
*And a hx of chronic migratory arthritis?*  
*And CNS symptoms—seizures, dementia, coma?*  
**Whipple’s disease**.

Broadly speaking, what sort of condition is Whipple’s?  
It is infectious.

*What infection agent is responsible for Whipple’s?*  
**The bacterium Tropheryma whipplei**.
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) Juvenile xanthogranuloma

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

What are the two hallmarks of JXG histology?
The presence of Touton giant cells
The presence of ‘foamy macrophages’

When unilateral?

Speaking of ‘foamy macrophages’...
What dz comes to mind if, instead of a young child
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 diarrhea?
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*When unilateral?*  
Bilateral

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

*What other finding will a duodenal biopsy reveal?*  
The presence of acid-fast bacteria within macrophages located in intestinal villi

**Whipple’s disease**

*Broadly speaking, what sort of condition is Whipple’s?*  
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Intraocular Tumors of Childhood

Iris/Ciliary Body

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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

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

What are the two hallmarks of JXG histology? The presence of Touton giant cells
- ‘foamy macrophages’

Speaking of ‘foamy macrophages’...

What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with chronic migratory arthritis? Associated with chronic diarrhea? And CNS symptoms—seizures, dementia, coma?

Whipple’s disease

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Whipple’s disease: Duodenal biopsy, high mag. The image shows the characteristic feature of foamy macrophages in the lamina propria.
**Intraocular Tumors of Childhood**

**Iris/Ciliary Body**

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Speaking of ‘foamy macrophages’…

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Associated with chronic migratory arthritis and CNS symptoms—seizures, dementia, coma

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Iris/Ciliary Body

Intraocular Tumors of Childhood

1) Juvenile xanthogranuloma

- Nonneoplastic histiocytic proliferation
- Present in <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

- 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
- 15% of non-Down population

5) Iris mammillations

- Tiny, numerous
- Same color as iris
- Weak association with NF1, Nevus of Ota

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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

1) **Juvenile xanthogranuloma**

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

**Nonneoplastic histiocytic proliferation**

**Speaking of foamy macrophages part deaux...**

What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a **Adult with bilateral upper-lid yellow lesions?**

--- Pic forthcoming

**The presence of Touton giant cells**

**The presence of 'foamy macrophages'**

**When unilateral?**

--- Pic forthcoming

**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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**What is the natural history of JXG?**

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

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2) **Medulloepithelioma**

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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Strong association with NF1. Lighter on dark irides; darker on light.

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Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota.

6) **Iris cysts**

Can be pupillary, stromal, secondary.

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Intraocular Tumors of Childhood
Intraocular Tumors of Childhood

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*In three words, what sort of condition is JXG?* 
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Speaking of foamy macrophages part deaux…

*What dz comes to mind if, instead of a young child with iris nodules, the pt in question was an Adult with bilateral upper-lid yellow lesions?* 
Z Xanthelasma

*Are xanthelasmas a harbinger of elevated serum lipids?* 
Yes, and when they are, they usually are a sign of lipid derangement.

*Can they be congenital?* 
Yes, and when they are, they usually are a sign of lipid derangement.

*Pic forthcoming*

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

- **What is the natural history of JXG?**
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- **When is the presence of 'foamy macrophages' unilateral?**

Choroid

- **Are the xanthelasmas a harbinger of elevated serum lipids?**

RPE

- **What is the natural history of JXG?**

Retina

- **The presence of Touton giant cells?**

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Intraocular Tumors of Childhood

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**Xanthelsasma**

The presence of Touton giant cells
The presence of 'foamy macrophages'

When unilateral?
When bilateral?

**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?**

**RPE**

What is the natural history of JXG?  
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**Xanthelsasma**

- The presence of **Touton giant cells**
- The presence of **‘foamy macrophages’**

- When **Unilateral**
- When **Bilateral?**

**Choroid**

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What is the natural history of JXG?  
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**1) Juvenile xanthogranuloma**

- **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**: Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed
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**Iris/Ciliary Body**

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**Iris/Ciliary Body**

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

**What is it?**

**Retina**

**Choroid**

**RPE**

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It is responsible for the creation of aqueous humor.
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*How does it present?*

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

- Glaucoma
- Hyphema
- Sectoral cataract

*Is it common, or rare?*

Very rare

*Is it benign, or malignant?*

It is benign, but very aggressive locally

*How is it managed?*

Enucleation is usually required
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Which specific component of the iris/CB is involved in medulloepithelioma?
The nonpigmented epithelium of the ciliary body

How does it present?
As an iris mass along with one or more of the following:
--Glaucoma
--Hyphema
--Sectoral cataract

Is it common, or rare?
Very rare

Is it a benign, or malignant lesion?
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

Medulloepithelioma

Lisch nodules: Strong association with NF1. Lighter on dark irides; darker on light

Brushfield spots: Strong association with Down syndrome; 15% of non-Down pop.

Iris mammillations: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

Iris cysts: Can be pupillary, stromal, secondary

Iris/Ciliary Body

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Intraocular Tumors of Childhood

Medulloepithelioma/diktyoma
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2) **Medulloepithelioma**

**Iris/Ciliary Body**

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

No, this lesion rarely metastasizes; it does its damage locally
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How does it present?
As an iris mass along with one or more of the following:
--Glaucoma
--Hyphema
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Is it common, or rare?
Very rare

Choroid

Is it a benign, or malignant lesion?
It can have features of both. Either way, it is very locally aggressive.

RPE

Retina

How aggressive is ‘very aggressive’?
Intraocular Tumors of Childhood

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Is it a benign, or malignant lesion?
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RPE

How aggressive is ‘very aggressive’?
Aggressive enough to result in death

Retina
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**RPE**

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

**Retina**

*How is it managed?*  
Enucleation is usually required
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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**

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Retina
Intraocular Tumors of Childhood

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3) **Lisch nodules**

4) **Brushfield spots**

In this context, what does NF1 stand for?

- NF1

NF1 is the eponymous name for Neurofibromatosis type 1. In a word, what sort of condition is it?

- A phakomatosis
Intraocular Tumors of Childhood

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

**NF1**

Neurofibromatosis type 1

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

**NF1**

Intraocular Tumors of Childhood

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NF1

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

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

Intraocular Tumors of Childhood

Iris/Ciliary Body

Retina
Intraocular Tumors of Childhood

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NF1
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3) **Lisch nodules**

4) **Brushfield spots**

5) **Iris mammillations**

6) **Iris cysts**

**Iris/Ciliary Body**

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**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?*

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

A phakomatosis

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

Phakomatoses are known also as neurocutaneous syndromes.

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**Retina**
Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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

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

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

A phakomatosis

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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

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

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

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Lisch nodules

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

Are Lisch nodules associated with NF2?
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**What is the prevalence of Lisch nodules in NF1?**

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**Intraocular Tumors of Childhood**

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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.

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**Iris/Ciliary Body**
Intraocular Tumors of Childhood

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**Are Lisch nodules clinically significant?**
Intraocular Tumors of Childhood

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*Are Lisch nodules clinically significant?*

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

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6) Iris cysts: Can be pupillary, stromal, secondary

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

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

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Melanocytic lesions
--?
--?
--?
--?

Neuroglial lesions
--?
--?
--?
--?
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4) Brushfield spots: Strong association with Down syndrome; 15% of non-Down pop.

5) Iris mammillations: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

6) Iris cysts: Can be pupillary, stromal, secondary

Intraocular Tumors of Childhood

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

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

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? 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
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

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Retina

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

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

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

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**Neuroglial lesions**
- Nodular neurofibromas
- Plexiform neurofibromas
- Optic glioma
- Prominent corneal nerves

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

The *melanocytic* lesions are of no clinical significance beyond establishing the diagnosis, whereas the *neuroglial* lesions are associated with significant ocular and/or systemic morbidity

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Most NF1 lesions are associated with one of two cell types. What are they? Melanocytes and neuroglial cells

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

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

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

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

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(No question—proceed when ready)
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3) **Lisch nodules**

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**Iris/Ciliary Body**

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

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

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

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**Retina**
Intraocular Tumors of Childhood

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**Brushfield spots are most strongly associated with what congenital condition?** Down syndrome
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**Iris/Ciliary Body**

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Intraocular Tumors of Childhood

Brushfield spots
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4) **Brushfield spots**

5) **Iris mammillations**

---

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

Down syndrome

**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

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

Wolfflin nodules
Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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Wolfflin nodules
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Mammillations? Aren’t those a CNS thingamajig?
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Iris/Ciliary Body

Choroid

Retina
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**OK, then what are iris mammillations?**
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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.
Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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*Are they unilateral, or bilateral?*
Intraocular Tumors of Childhood

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With what phakomatosis are they associated?
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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 nearly as strongly as Lisch nodules)
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

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

‘Tiny pigmented iris nodules associated with NF1’—given this, how on earth are you supposed to differentiate between Lisch nodules and mammillations?

With what phakomatosis are they associated?

NF1 (albeit not nearly as strongly as Lisch nodules)
Intraocular Tumors of Childhood

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OK, then what are iris mammillations?

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‘Tiny pigmented iris nodules associated with NF1’—given this, how on earth are you supposed to differentiate between Lisch nodules and mammillations?

By appearance. Iris mammillations are always the same color as the rest of the iris. In contrast and as stated previously, Lisch nodules are lighter when the iris is dark, but darker when the iris is light.

With what phakomatosis are they associated?

NF1 (albeit not nearly as strongly as Lisch nodules)
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With what phakomatosis are they associated?
NF1 (albeit not nearly as strongly as Lisch nodules)

In addition to NF1, iris mammillations have another important association. What is it?
Intraocular Tumors of Childhood

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Oculodermal melanocytosis, aka three words
Intraocular Tumors of Childhood

Iris/Ciliary Body

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With what phakomatosis are they associated?
NF1 (albeit not nearly as strongly as Lisch nodules)

In addition to NF1, iris mammillations have another important association. What is it?
Oculodermal melanocytosis, aka nevus of Ota
Intraocular Tumors of Childhood

Oculodermal melanocytosis (nevus of Ota)
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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)
Intraocular Tumors of Childhood

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Iris/Ciliary Body

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

Choroid

Five tumors of the choroid

RPE

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

1) **Nevus**
2) **Melanocytoma**
3) **Osteoma**
4) **Isolated/focal choroidal hemangioma**
5) **Diffuse choroidal hemangioma**

RPE

Retina

Five tumors of the choroid
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

1) **Nevus**
2) **Melanocytoma**
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What common sort of choroidal tumor—common in adults—is absent from this list?

Retina
**Iris/Ciliary Body**

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**Choroid**

1) **Nevus**
2) **Melanocytoma**
3) **Osteoma**
4) **Isolated/focal choroidal hemangioma**
5) **Diffuse choroidal hemangioma**

*But not 6) Metastases*

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**Retina**

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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.
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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Choroid

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3) Osteoma
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5) Diffuse choroidal hemangioma
6) Metastases

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.

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

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

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2) **Melanocytoma**
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4) **Isolated/focal choroidal hemangioma**
5) **Diffuse choroidal hemangioma**

But not 6) **Metastases**

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.

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

Retina
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Melanocytoma is a variant of what common choroidal finding?

1) Nevus: Common. Benign
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4) Isolated/focal choroidal hemangioma
5) Diffuse choroidal hemangioma

Melanocytoma is a variant of what common choroidal finding?

Melanocytoma is a variant of choroidal nevus. From what structure does it commonly arise? The optic disc. Is it unilateral or bilateral? 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? Yes.
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**Melanocytoma**

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

**It is a particular sort of choroidal nevus**
Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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

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

**From what structure does it commonly arise?**

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**Choroid**

1) Nevus: Common. Benign

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**RPE**

**Retina**
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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

Retina

RPE
Intraocular Tumors of Childhood

Melanocytoma
Intraocular Tumors of Childhood

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From what structure does it commonly arise?
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Does it have a unilateral/bilateral predilection?

Retina

RPE

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**Iris/Ciliary Body**

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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

---

**Choroid**

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**RPE**

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**Retina**
Intraocular Tumors of Childhood

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Iris/Ciliary Body

Choroid

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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

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Yes, it is virtually always unilateral

Is there a racial predilection?
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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
**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 (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 in kids slide-set)

---

**Iris/Ciliary Body**

**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?**

Yes

---

**Choroid**

1) **Nevus:** Common. Benign

2) **Melanocytoma**

3) **Osteoma**

4) **Isolated/focal choroidal hemangioma**

5) **Diffuse choroidal hemangioma**

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**RPE**

**Retina**
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Intraocular Tumors of Childhood

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Nevus: Common. Benign

Melanocytoma: Usually juxtapapillary. Malignant transformation extremely rare

Osteoma: Bone

Very rare

Benign

Yes, it is more common in females

Teens

Choroidal neovascular membrane

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

Bone

Common. Benign

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Choroidal neovascular membrane
Intraocular Tumors of Childhood

Osteoma
Intraocular Tumors of Childhood

Osteoma: FP, and b-scan demonstrating...
Intraocular Tumors of Childhood

Osteoma: FP, and b-scan demonstrating ‘shadowing’
Osteoma: Another example
Intraocular Tumors of Childhood

Osteomas (same pt, different cuts).
Note how bright the lesions are
**In a word, what is a choroidal osteoma composed of?**

Bone

**Is it common or rare?**

Very rare

**Is it benign or malignant?**

Benign

**Does it have 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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Intraretinal Tumors

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Choroid

RPE

Retina
Intraocular Tumors of Childhood

Osteoma with CNVM in a 13 y.o. female
Juvenile xanthogranuloma (JXG): Nonneoplastic histiocytic proliferation. <2 years old. +/− skin papules. Iris nodules bleed \(\rightarrow\) hyphema \(\rightarrow\) increased IOP \(\rightarrow\) glaucoma. Self-limited; regresses by age 5. Treat inflammation and IOP. Path: Touton giant cells

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Lisch nodules: Strong association with NF1. Lighter on dark irides; darker on light

Brushfield spots: Strong association with Down syndrome

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

Nevus: Common. Benign

Melanocytoma: Usually juxtapapillary. Malignant transformation extremely rare

Osteoma

Isolated/focal choroidal hemangioma

Diffuse choroidal hemangioma

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

Is it common or rare?
Very rare

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

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

Choroid

3) Osteoma

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**Iris/Ciliary Body**

**Choroid**

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By what other name is this lesion known? **Circumscribed choroidal hemangioma**

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**Intraocular Tumors of Childhood**

**Iris/Ciliary Body**

**Choroid**

**RPE**

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Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

- Iris/Ciliary Body
- Choroid

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?
As a reddish-orange mass in the macula

What is its characteristic pattern on a -scan ultrasonography?
It is one of 'high internal reflectivity'

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Intraocular Tumors of Childhood

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How does it present? As a reddish-orange mass in the macula

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Intraocular Tumors of Childhood

Circumscribed choroidal hemangioma: High internal reflectivity on a-scan
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**Retina**

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‘High internal reflectivity’--what other choroidal lesion’s a-scan is described the same way?
Choroidal nevus

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3) Osteoma: benign bony tumor, most common in teen years, females. Risk of CNVM
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RPE
Intraocular Tumors of Childhood

By what other name is this lesion known?
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high internal reflectivity

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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?

About 50%

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

The coloration is a very red—much more so than an unaffected fundus

What food-related term is used to describe the fundus appearance in SWS?

'Tomato catsup fundus'

Can the choroidal hemangioma be present bilaterally?

Yes, but it's uncommon

Does the choroidal hemangioma have malignant potential?

No

5) **Diffuse choroidal hemangioma**
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With what condition is the diffuse choroidal hemangioma associated? Sturge-Weber syndrome (SWS)
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**MEDULLOEPITHELIOMA**
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**NEVUS:**
Common. Benign

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Usually juxtapapillary. Malignant transformation extremely rare

**OSTEOMA:**
Benign bony tumor, most common in teen years, females. Risk of CNVM

**ISOLATED/FOCAL CHOROIDAL HEMANGIOMA:**
Very rare. Characteristic a-scan pattern

**DIFFUSE CHOROIDAL HEMANGIOMA**

**STURGE-WEBER SYNDROME (SWS):**
In a word, what sort of condition is SWS?

- A phakomatosis
- Encephalotrigeminal angiomatosis
  - Also you might also see encephalofacial or cerebrofacial angiomatosis

---

1) **INTRAOCULAR TUMORS OF CHILDHOOD**
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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11) Diffuse choroidal hemangioma:

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In a word, what sort of condition is SWS?
A phakomatosis
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

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 in kids slide-set)

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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**

*Sturge-Weber syndrome (SWS)*

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

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**RPE**

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**Retina**
**Intraocular Tumors of Childhood**

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

About 50%

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

The coloration is a very red--much more so than an unaffected fundus

**What food-related term is used to describe the fundus appearance in SWS?**

'Tomato catsup fundus'

**Can the choroidal hemangioma be present bilaterally?**

Yes, but it's uncommon

**Does the choroidal hemangioma have malignant potential?**

No

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)

What is the hallmark skin finding in SWS?

The port-wine stain

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

An angioma

When does it present?

At birth

What is the typical pattern of distribution?

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

Does it always present in this manner?

No. Some cases will cross the midline of the face

All infants with SWS have a port-wine stain. Do all infants with a port-wine stain have SWS?

No
Intraocular Tumors of Childhood

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5) Diffuse choroidal hemangioma: Sturge-Weber syndrome (SWS)

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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Intraocular Tumors of Childhood

Sturge-Weber: Port-wine stain
Intraocular Tumors of Childhood

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Nevus

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Osteoma

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Isolated/focal choroidal hemangioma

- Very rare. Characteristic a−scan pattern

Diffuse choroidal hemangioma

- Associated with Sturge-Weber syndrome (SWS). Present in about 50% of SWS cases. The coloration is a very red—much more so than an unaffected fundus. The hallmark skin finding in SWS is the port-wine stain, an angioma. It presents at birth and comports to the distribution of one or more divisions of CN5. It does not always present in this manner. Some cases will cross the midline of the face.

Sturge-Weber syndrome (SWS)

- A phakomatosis

- Noneponymous name: Encephalotrigeminal angiomatosis (you might also see encephalofacial or cerebrofacial angiomatosis)

- Hallmark skin finding: The port-wine stain

- Fundus appearance: The coloration is a very red—much more so than an unaffected fundus

- TOMATO CATSUP FUNDUS

- Can be present bilaterally

- No malignant potential

- A phakomatosis

- Noneponymous name: Encephalotrigeminal angiomatosis (you might also see encephalofacial or cerebrofacial angiomatosis)
Intraocular Tumors of Childhood

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Diffuse choroidal hemangioma: Sturge-Weber syndrome (SWS)

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**Diffuse choroidal hemangioma**

*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?**
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**What is the hallmark skin finding in SWS?**
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**Intraocular Tumors of Childhood**

With what condition is the diffuse choroidal hemangioma associated?

- Sturge-Weber syndrome (SWS)

**What is the hallmark skin finding in SWS?**

- The port-wine stain

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

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**When does it present?**

- At birth

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Intraocular Tumors of Childhood

Sturge-Weber: Port-wine stain
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Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern.

Diffuse choroidal hemangioma: Associated with Sturge-Weber syndrome (SWS).

Diffuse choroidal hemangioma is present in about 50% of SWS.

What does the fundus look like in an eye with a diffuse choroidal hemangioma?
The coloration is very red—much more so than an unaffected fundus.

What food-related term is used to describe the fundus appearance in SWS?
'Tomato catsup fundus'

Can the choroidal hemangioma be present bilaterally?
Yes, but it's uncommon.

Does the choroidal hemangioma have malignant potential?
No.

In a word, what sort of condition is SWS?
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In a word, what sort of condition is SWS? A phakomatosis

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What is the hallmark skin finding in SWS? The port-wine stain

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**When does it present?**
At birth

**What is the typical pattern of distribution?**
It comports to the distribution of one or more divisions of CN5

**Does it always present in this manner?**
No, some cases will cross the midline of the face
Intraocular Tumors of Childhood

With what condition is the diffuse choroidal hemangioma associated?

Sturge-Weber syndrome (SWS)

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

What food-related term is used to describe the fundus appearance in SWS? 'Tomato catsup fundus'

Can the choroidal hemangioma be present bilaterally? Yes, but it's uncommon

Does the choroidal hemangioma have malignant potential? No

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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?**
***About 50%***

**What does the fundus look like in an eye with a diffuse choroidal hemangioma?**
The coloration is a very red—much more so than an unaffected fundus

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**Can the choroidal hemangioma be present bilaterally?**
Yes, but it's uncommon

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

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 in kids slide-set)

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**Intracocular 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

**What does the fundus look like in an eye with a diffuse choroidal hemangioma?**
The coloration is a very red—much more so than an unaffected fundus

**What food-related term is used to describe the fundus appearance in SWS?**
'Tomato catsup fundus'

**Can the choroidal hemangioma be present bilaterally?**
Yes, but it's uncommon

**Does the choroidal hemangioma have malignant potential?**
No

**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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**Iris/Ciliary Body**

**Choroid**

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### Intraocular Tumors of Childhood

**Iris/Ciliary Body**

**Choroid**

**Retina**

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

Sturge-Weber syndrome (SWS)

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

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

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**Can the choroidal hemangioma be present bilaterally?**

Yes, but it's uncommon

**Does the choroidal hemangioma have malignant potential?**

No

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5) **Diffuse choroidal hemangioma**

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**RPE**

**Retina**
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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

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

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

**Retina**
Intraocular Tumors of Childhood

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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?**

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

The coloration is a very red, much more so than an unaffected fundus

**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

Does the choroidal hemangioma have malignant potential? No

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**Iris/Ciliary Body**

**Choroid**

**RPE**

**Retina**

**Retina**

5) Diffuse choroidal hemangioma
Intracocular Tumors of Childhood

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

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Iris/Ciliary Body

RPE

Retina

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**Iris/Ciliary Body**

Nevus: Common. Benign

Melanocytoma: Usually juxtapapillary. Malignant transformation extremely rare

Osteoma: Benign bony tumor, most common in teen years, females. Risk of CNVM

Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern

5) **Diffuse choroidal hemangioma**

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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‘Tomato catsup fundus’

**Retina**

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Sturge-Weber syndrome (SWS)

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Diffuse choroidal hemangioma

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

What does the fundus look like in an eye with a diffuse choroidal hemangioma? The coloration is a very red, much more so than an unaffected fundus

What food-related term is used to describe the fundus appearance in SWS? ‘Tomato catsup fundus’

Can the choroidal hemangioma be present bilaterally? Yes, but it’s uncommon

What condition is associated with diffuse choroidal hemangioma? Sturge-Weber syndrome (SWS)

About half of eyes with SWS have diffuse choroidal hemangioma

The coloration is a very red, much more so than an unaffected fundus

‘Tomato catsup fundus’

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**Iris/Ciliary Body**

**RPE**

**Retina**

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**Intracocular 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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Yes, but it’s uncommon

**Does the choroidal hemangioma have malignant potential?**
No

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4) Isolated/local choroidal hemangioma: Very rare. Characteristic a-scan pattern

5) Diffuse choroidal hemangioma

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RPE

Retina
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Osteoma: Benign bony tumor, most common in teen years, females. Risk of CNVM

Isolated/focal choroidal hemangioma: Very rare. Characteristic a- scan pattern

Diffuse choroidal hemangioma: Associated with Sturge-Weber syndrome (SWS). Present in about half of SWS. The coloration is a very red, much more so than an unaffected fundus. What food-related term is used to describe the fundus appearance in SWS? ‘Tomato catsup fundus’

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With what condition is the diffuse choroidal hemangioma associated? Sturge-Weber syndrome (SWS)
Intraocular Tumors of Childhood

Iris/Ciliary Body

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5) **Diffuse choroidal hemangioma:** Unilateral. Found in Sturge-Weber syndrome

RPE

1) ?

Retina
Intraocular Tumors of Childhood

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RPE

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

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**Iris/Ciliary Body**

**Choroid**

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

**Retina**
Iris/Ciliary Body

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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

RPE

1) **Congenital hypertrophy of the RPE (CHRPE)**
Intraocular Tumors of Childhood

CHRPE
Intraocular Tumors of Childhood

Iris/Ciliary Body

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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

Is it common, or rare?

Ch

Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern
Diffuse choroidal hemangioma: Unilateral. Found in Sturge-Weber syndrome

RPE

1) Congenital hypertrophy of the RPE (CHRPE)

Retina

What is the clinical appearance of CHRPE?
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RPE

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Intraocular Tumors of Childhood

Iris/Ciliary Body

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

Is it common, or rare?
Common

Retina

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Iris/Ciliary Body

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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

**Is it common, or rare?**
Common

**Is it a hamartoma or a choristoma?**

**Retina**

1) **Congenital hypertrophy of the RPE (CHRPE)**
Intraocular Tumors of Childhood

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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

Is it common, or rare?
Common

Is it a hamartoma or a choristoma?
It is neither

RPE
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**Iris/Ciliary Body**

**Choroid**

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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

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**RPE**

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

*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

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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
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)**

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

- **CHRPE**

- **or**

*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
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Choroid

1) Nevus: Common. Benign
2) Melanocytoma: Usually juxtapapillary. Malignant transformation extremely rare
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4) Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern

RPE

1) Congenital hypertrophy of the RPE (CHRPE)

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

Solitary

Grouped

CHRPE
Iris/Ciliary Body

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5) **Diffuse choroidal hemangioma**: Unilateral. Found in Sturge-Weber syndrome

RPE

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

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

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**Solitary** CHRPE

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

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

1) **Nevus:** Common. Benign
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5) **Diffuse choroidal hemangioma:** Unilateral. Found in Sturge-Weber syndrome

RPE

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

*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

*What descriptive name is used with regard to the appearance of Multifocal/Grouped CHRPE? ‘Bear tracks’*
Intraocular Tumors of Childhood

CHRPE: Bear tracks
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

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

Lisch nodules: Strong association with NF1. Lighter on dark irides; darker on light

Brushfield spots: Strong association with Down syndrome

Iris mammillations: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

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

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
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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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**
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
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Isolated/focal choroidal hemangioma: Very rare. Characteristic \( ^a \)-scan pattern
Diffuse choroidal hemangioma: Unilateral. Found in Sturge-Weber syndrome

1) Congenital hypertrophy of the RPE (CHRPE)

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

A CHRPE-like lesion is:
(both eponymous and non)
Familial adenomatomatous 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!
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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5) **Iris mammillations**: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

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?**

RPE

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

**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 characteristics of a CHRPE-like presentation increase the likelihood that it is a component of Gardner syndrome?
--If it is bi- v unilateral (regular CHRPE is almost always bi- v unilateral)

RPE

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

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
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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5) **Iris mammillations:** Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

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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)

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**RPE**

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

**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

CHRPE-like lesions of Gardner syndrome: Bilateral presentation
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) Juvenile xanthogranuloma (JXG): Nonneoplastic histiocytic proliferation. <2 years old. +/- skin papules. Iris nodules bleed \(\rightarrow\) hyphema \(\rightarrow\) increased IOP \(\rightarrow\) 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 \(\rightarrow\) hyphema \(\rightarrow\) increased IOP \(\rightarrow\) glaucoma. Locally invasive \(\rightarrow\) death. Tx: Enucleate
3) Lisch nodules: Strong association with NF1. Lighter on dark irides; darker on light
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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' distribution pattern)

RPE

1) Congenital hypertrophy of the RPE (CHRPE)

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

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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4) Brushfield spots: Strong association with Down syndrome
5) Iris mammillations: Tiny, numerous. Same color as iris. Weak association with NF1, Nevus of Ota

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’)

1) Congenital hypertrophy of the RPE (CHRPE)

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
CHRPE-like lesions of Gardner syndrome: Scattered distribution
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

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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 pisciform

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**RPE**

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

**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
**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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---

**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 pisciform

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**RPE**

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**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

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Nevus: Common. Benign

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Osteoma: Benign bony tumor, most common in teen years, females. Risk of CNVM

Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern

Diffuse choroidal hemangioma: Unilateral. Found in Sturge-Weber syndrome

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 (i.e., not ‘grouped’)
--If the shape of the lesions is pisciform

What does pisciform mean?

'Fish-shaped'

1) Congenital hypertrophy of the RPE (CHRPE)

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
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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Iris/Ciliary Body

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

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 eye (i.e., not ‘grouped’)
-- If the shape of the lesions is pisciform

What does pisciform mean?
It means ‘fish-shaped’
Intraocular Tumors of Childhood

CHRPE-like lesions of Gardner syndrome: Pisciform shape
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 pisciform

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

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--

**CHRPE**

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
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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 pisciform

The tails of these fish-shaped lesions have two telltale (tell-tail?) characteristics--what are they?
--They are hypopigmented vs hyperpigmented
--They point towards the optic nerve head

---Multifocal or Grouped CHRPE: Large lesion(s) surrounded by a few smaller ones
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
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Iris/Ciliary Body

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 pisciform

The tails of these fish-shaped lesions have two telltale (tell-tail?) characteristics--what are they?
--They are hypopigmented
--They point towards the optic nerve head

Retina

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-

RPE
CHRPE-like lesions of Gardner syndrome: Hypopigmented tail pointing toward ONH
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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What is the most clinically important (and ominous) component to Gardner syndrome?
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--Benign tumors of skin
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Gardner syndrome: Colonic polyps
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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

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

All of them

By what age will this occur? Age 40, maybe a little later

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Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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

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4) **Isolated/focal choroidal hemangioma:** Very rare. Characteristic a-scan pattern

5) **Diffuse choroidal hemangioma:** Unilateral. Found in Sturge-Weber syndrome

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**What is the treatment of choice?**
Prophylactic colectomy

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**Retina**

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**Intraocular Tumors of Childhood**

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Intraocular Tumors of Childhood

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When ‘colon cancer + ophthalmic issue’ is mentioned, two syndromes should come to mind. One is Gardner syndrome. What is the other? **Muir-Torre syndrome**

RPE

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

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How is Muir-Torre pronounced?
mure (rhymes with ‘pure’) tore-AY

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**Intraocular Tumors of Childhood**

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Intraocular Tumors of Childhood

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What is the main ophthalmic manifestation of Muir-Torre syndrome? What is Muir-Torre syndrome pronounced? What is the main ophthalmic manifestation of Muir-Torre syndrome? Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

When 'colon cancer + ophthalmic issue' is mentioned, two syndromes should come to mind. One is Gardner syndrome. What is the other?

Muir-Torre syndrome (Mure (rhymes with 'pure') tore-AY)

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Muir-Torre syndrome
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.
Intraocular Tumors of Childhood

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**RPE**

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

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**Retina**

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

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

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**Iris/Ciliary Body**

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**

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When 'colon cancer + ophthalmic issue' is mentioned, two syndromes should come to mind. One is Gardner syndrome. What is the other?

Muir-Torre syndrome

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What is the main ophthalmic manifestation of Muir-Torre syndrome?

Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

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Does Muir-Torre present with multiple adenomatous polyps of the colon a la Gardner syndrome?

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

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What sorts of sebaceous lesions?

- Basal-cell carcinomas with sebaceous differentiation
Intraocular Tumors of Childhood

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# Intraocular Tumors of Childhood

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1. ?
Intraocular Tumors of Childhood

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**Intraocular Tumors of Childhood**

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Iris/Ciliary Body

What is a hamartoma?
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**Iris/Ciliary Body**

**Choroid**

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

That a lesion is a hamartoma (or choristoma) indicates what about its status vis a vis malignancy?
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Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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**Intraocular Tumors of Childhood**

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**Path:**

- Touton giant cells
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**What is a hamartoma?**

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**So, what combination of hamartomatous cells of the retina and RPE are involved in a combined hamartoma of the retina and RPE?**

- **RPE**
  - 1) Combined hamartoma of the retina and RPE
  - 1) Retinoblastoma (see the slide-set dedicated to it)
**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** (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)

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**Iris/Ciliary Body**

**RPE**

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

2) **Combined hamartoma of the retina and RPE**

**Retina**

1) **Retinoblastoma** (see the slide-set dedicated to it)
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1) **Nevus:** Common. Benign

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5) **Diffuse choroidal hemangioma:** Unilateral. Found in Sturge-Weber syndrome

6) **Retinoblastoma:** (see the slide-set dedicated to it)

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**What is a hamartoma?**
A tumor composed of histologically abnormal cells found in their normal location

**So, what combination of hamartomatous cells of the retina and RPE are involved in a combined hamartoma of the retina and RPE?**
RPE cells (duh) and retinal glial cells

**How does it present clinically?**

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Iris

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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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RPE

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**How does it present clinically?**

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Intraocular Tumors of Childhood

Combined hamartoma of retina and RPE
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**Iris/Ciliary Body**

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   **How does it present clinically?**

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

   **With what more sinister dz entity is it often confused?**

   Choroidal melanoma--eyes have been enucleated because of this misdiagnosis

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1) **Combinhed hamartoma of the retina and RPE**

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**Iris/Ciliary Body**

**Choroid**

**RPE**

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**How can one avoid making such a disastrous mistake?**

By taking pains to carefully determine the anatomic location of the tumor in question—choroidal melanomas originate behind Bruch’s membrane, whereas combined hamartomas of the retina and RPE are located wholly in front of it

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**Intraocular Tumors of Childhood**

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**Iris/Ciliary Body**

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**Choroid**

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Intraocular Tumors of Childhood

Combined hamartoma of retina and RPE. Note the entire lesion is above Bruchs
Intraocular Tumors of Childhood

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*No question—summary/review slide*