Intraocular Tumors of Childhood

Four intraocular locations (i.e., structures/tissues)
Intraocular Tumors of Childhood

Four intraocular locations (i.e., structures/tissues)

Iris/Ciliary Body

Choroid

RPE

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

1) ?

2) ?

1) ?

3) ?

4) ?

5) ?

6) ?

Six tumors of the iris/ciliary body

Choroid

RPE

Retina
Intraocular Tumors of Childhood

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

Iris/Ciliary Body

Six tumors of the 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?*
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

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

1) **Juvenile xanthogranuloma**

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Choroid

RPE

Retina
Intraocular Tumors of Childhood

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Iris/Ciliary Body

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

1) Juvenile xanthogranuloma

Iris/Ciliary Body

Choroid

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Iris/Ciliary Body

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It is self-limited, usually resolving by age 5 years
Iris/Ciliary Body

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## Intraocular Tumors of Childhood

### Iris/Ciliary Body

1) **Juvenile xanthogranuloma**

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  - *nonneoplastic histiocytic proliferation*

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

### Choroid

### RPE

### Retina
Intraocular Tumors of Childhood

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Unilateral

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--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a ‘masquerade syndrome’ in pediatrics uveitis
--If enough nodules are present, *heterochromia iridis* will result

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

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

1) **Juvenile xanthogranuloma**

- **In three words, what sort of condition is JXG?**
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- **When JXG iris nodules are present, are they uni-, or bilateral?**
  - Unilateral

- **In what three ways are the iris nodules clinically significant?**
  - They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
  - ‘Masquerade syndrome’ in peds uveitis
  - Heterochromia iridis will result

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Intraocular Tumors of Childhood

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*When are the two hallmarks of JXG histology?*
Touton giant cells

*What are the two hallmarks of JXG histology?*
'foamy macrophages'

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Intraocular Tumors of Childhood

Iris/Ciliary Body

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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

Choroid

RPE

Retina
**Intraocular Tumors of Childhood**

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**Choroid**


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Intraocular Tumors of Childhood

Iris/Ciliary Body

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

*When unilateral, is it 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?  
First clue--more forthcoming

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

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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?*  
Lateral

*When bilateral?*  
Lateral?

'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
Intraocular Tumors of Childhood

Iris/Ciliary Body

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*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? Need another?
Intraocular Tumors of Childhood

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

*When are the JXG iris nodules unilateral or bilateral?*
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?*  

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Last chance--answer is next!
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The presence 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

*Speaking of ‘foamy macrophages’…*
Intraocular Tumors of Childhood

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*When Unilateral*

When Unilateral

*‘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?

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

Retina

Choroid
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The presence of ‘foamy macrophages’ unilateral?

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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?
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**Whipple’s disease**

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

Retina

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

*When unilateral, are JXG clinically significant?*  
Unilateral

*What are the inclusions of JXG histology?*  
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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?  
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**Whipple’s disease**

*Broadly speaking, what sort of condition is Whipple’s?*  
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*What infection agent is responsible for Whipple’s?*  
The bacterium Tropheryma whipplei
Intraocular Tumors of Childhood

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*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

*When?*  
The presence of ‘foamy macrophages’ lateral?

**Speaking of ‘foamy macrophages’…**

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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 and ‘foamy macrophages’.

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

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.

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

What infection agent is responsible for Whipple’s? The bacterium *Tropheryma whipplei*.
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The duodenum (remember, they have GI issues)

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Intraocular Tumors of Childhood

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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’.

When foamy macrophages are found in a biopsy performed on a Whipple’s pt, from what site was the biopsy collected?
The duodenum (remember, they have GI issues).

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

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

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

What is the other name by which medulloepithelioma is known?

- Diktyoma

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 benign, or malignant?

- It is benign, but very aggressive locally

How is it managed?

- Enucleation is usually required
Intraocular Tumors of Childhood

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Iris/Ciliary Body

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**Choroid**

*How aggressive is ‘very aggressive’?*
Aggressive enough to result in death
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Intraocular Tumors of Childhood

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3) Lisch nodules
4) Brushfield spots

Lisch nodules are most strongly associated with what congenital condition?

NF1

Is it 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

Retina

Intraocular Tumors of Childhood
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3) **Lisch nodules:** Strong association with NF1. Lighter on dark irides; darker on light

4) **Brushfield spots:** Strong association with Down syndrome; 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

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**Iris/Ciliary Body**

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

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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

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In a word, what sort of condition is it?

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Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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*In a word, what sort of condition is NF1?*  
A **phakomatosis**

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

A **neurocutaneous syndromes**

*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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Intraocular Tumors of Childhood

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2) Medulloepithelioma (aka diktyoma): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed → hyphema → increased IOP → glaucoma. Locally invasive → death. Tx: Enucleate

3) Lisch nodules

4) Brushfield spots

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

NF1

**Are Lisch nodules associated with NF2?**

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

**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
Iris/Ciliary Body

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

Are Lisch nodules dark, or light?

Yes, 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

Iris/Ciliary Body

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### Iris/Ciliary Body

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**Intraocular Tumors of Childhood**

**Iris/Ciliary Body**

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

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

Iris/Ciliary Body

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Retina

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Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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

You’re thinking of the **two words**, paired structures that are part of the limbic system
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Tiny pigmented iris nodules which, when present, are found in vast numbers diffusely scattered across the iris surface
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NF1 (albeit not nearly as strongly as Lisch nodules)
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You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

**OK, then what are iris mammillations?**

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

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

With what phakomatosis are they associated?

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**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?

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?*

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(albeit not nearly as strongly as Lisch nodules)
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In addition to NF1, iris mammillations have another important association. What is it?
Intraocular Tumors of Childhood

Iris/Ciliary Body

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NF1 (albeit not nearly as strongly as Lisch nodules)

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

Oculodermal melanocytosis, aka *three words*
Intraocular Tumors of Childhood

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Oculodermal melanocytosis, aka nevus of Ota
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3) **Lisch nodules:** Strong association with NF1. Lighter on dark irides; darker on light

4) **Brushfield spots:** Strong association with Down syndrome

5) **Iris mammillations:** 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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4) **Brushfield spots:** Strong association with Down syndrome
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**Iris/Ciliary Body**

**Choroid**

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

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**Five tumors of the choroid**

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**RPE**

**Retina**
Intraocular Tumors of Childhood

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Iris/Ciliary Body

**Choroid**

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

Five tumors of the choroid

RPE

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?*

**Five tumors of the choroid**

*What common sort of choroidal tumor—common in adults—is absent from this list?*
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**
2) **Melanocytoma**
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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.
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

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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?**
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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**Choroid**

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

*But not 6) Metastases*

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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.

*If a child does suffer an ophthalmic metastasis, where does it tend to occur?* The orbit
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

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Intraocular Tumors of Childhood

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Melanocytoma is a variant of what common choroidal finding?

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**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**

1) **Nevus:** Common. Benign

2) **Melanocytoma**

3) **Osteoma**

4) **Isolated/focal choroidal hemangioma**

5) **Diffuse choroidal hemangioma**

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**Choroid**

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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1) **Nevus:** Common. Benign

2) **Melanocytoma**

3) **Osteoma**

4) **Isolated/focal choroidal hemangioma**

5) **Diffuse choroidal hemangioma**

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**RPE**

**Retina**
Intraocular Tumors of Childhood

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1) Nevus: Common. Benign

2) Melanocytoma: A variant of choroidal nevus

3) Osteoma

4) Isolated/focal choroidal hemangioma

5) Diffuse choroidal hemangioma

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
**Intraocular Tumors of Childhood**

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**Choroid**

1) **Nevus:** Common. Benign. From what structure does it commonly arise? The optic disc

2) **Melanocytoma**

3) **Osteoma**

4) **Isolated/focal choroidal hemangioma**

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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? The optic disc

- Does it have a unilateral/bilateral predilection?
Intraocular Tumors of Childhood

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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

Does it have a unilateral/bilateral predilection?
Yes, it is virtually always
Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

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

- Nevus: Common. Benign
- Melanocytoma
- Osteoma
- Isolated/focal choroidal hemangioma
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**RPE**

**Retina**
Intraocular Tumors of Childhood

Iris/Ciliary Body

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Choroid

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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
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Yes, it is virtually always unilateral
Is there a racial predilection?
Intraocular Tumors of Childhood

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Choroid

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RPE

Retina

**Melanocytoma is a variant of what common choroidal finding?**
It is a particular sort of choroidal nevus

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

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

**Is there a racial predilection?**
No
Iris/Ciliary Body

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2) Medulloepithelioma (aka diktyoma): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed→hyphema→increased IOP→glaucoma. Path: From CB→island of epithelial tissue→hemorrhage→glaucoma
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Choroid

1) Nevis: Common. Benign
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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

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

Is there a racial predilection?
No

Does it affect visual acuity?

Retina
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**Iris/Ciliary Body**

1) **Nevus:** Common. Benign

2) **Melanocytoma**

3) **Osteoma**

4) **Isolated/focal choroidal hemangioma**

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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

**Does it have a unilateral/bilateral predilection?**

Yes, it is virtually always unilateral

**Is there a racial predilection?**

No

**Does it affect visual acuity?**

Only in a minority of cases. But in almost all cases, it does affect **two words**
Iris/Ciliary Body

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Choroid

1) Nevus: Common. Benign. Bimodal age distribution (ages 1-3 and 9-10 years)
2) Melanocytoma: It is a particular sort of choroidal nevus. From what structure does it commonly arise? The optic disc
3) Osteoma
4) Isolated/focal choroidal hemangioma
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RPE

Retina

Melanocytoma is a variant of what common choroidal finding?

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Yes, it is virtually always unilateral

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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**

**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?**
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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

**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
Intraocular Tumors of Childhood

Iris/Ciliary Body

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

Choroid

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

RPE

Melanocytoma is a variant of what common choroidal finding? It is a particular sort of choroidal nevus

From what structure does it commonly arise? The optic disc

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

Is there a racial predilection? No

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

Does melanocytoma have the potential to undergo malignant transformation?

Approximately what percent of cases will transform?
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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?

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

Is it common or rare?
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Is it benign or malignant?
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Is it more typically found in pre-teens, or teens?
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If significant vision loss occurs, what osteoma complication is usually the culprit?
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Intraocular Tumors of Childhood
123

Retina

Intraocular Tumors of Childhood
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Iris/Ciliary Body

Choroid

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**Path:** Touton giant cells

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**Intraocular Tumors of Childhood**

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RPE

Retina

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**Intraocular Tumors of Childhood**

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True osteomas are indeed rare; however, secondary osteoma-like lesions are not uncommon found in what sorts of eyes?
Eyes that have suffered severe chronic inflammation, especially after they become phthisical

Intraocular Tumors of Childhood

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### Choroid

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**By what other name is this lesion known?**

**Circumscribed choroidal hemangioma**
**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

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

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

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**Intraocular Tumors of Childhood**

**Retina**

- **By what other name is this lesion known?**  
  C**ircumscribed** 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'

---

**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  
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**Intraocular Tumors of Childhood**

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**Choroid**

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As a reddish-orange mass in the macula

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**Intraocular Tumors of Childhood**

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In a word, what sort of condition is SWS?

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?

A phakomatosis

What is the noneponymous name for SWS?

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

Iris/Ciliary Body

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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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Sturge-Weber syndrome (SWS) is a phakomatosis that is associated with diffuse choroidal hemangioma. In SWS, the choroidal hemangioma is present in about 50% of cases. The fundus appearance in SWS is described as 'tomato catsup fundus', which is a very red coloration much more so than an unaffected fundus. It is uncommon for the choroidal hemangioma to be present bilaterally, and it is not malignant. Sturge-Weber syndrome is also known as Encephalotrigeminal angiomatosis. You might also see it as encephalofacial or cerebrofacial angiomatosis.
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**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?

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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 in kids slide-set)

**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)

What is the hallmark skin finding in SWS?

The port-wine stain

Intraocular Tumors of Childhood

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

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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:** 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:**

*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)

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

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

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**Intraocular Tumors of Childhood**

With what condition is the diffuse choroidal hemangioma associated?

**Sturge-Weber syndrome (SWS)**

**Retina**

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

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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**

- **Sturge-Weber syndrome (SWS)**
  - About 50%
  - The coloration is a very red—much more so than an unaffected fundus
  - 'Tomato catsup fundus'
  - Yes, but it’s uncommon
  - No
- **A phakomatosis**
- **Encephalotrigeminal angiomatosis** (you might also see encephalofacial or cerebrofacial angiomatosis)
- **The port-wine stain**
  - An angioma
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)

*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?*
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
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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?**

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**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?**

**Sturge-Weber syndrome (SWS)**

**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)

**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?**
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

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

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: Associated with 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? 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
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

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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: Associated with Sturge-Weber syndrome (SWS). About 50% of cases present bilaterally. The coloration is a very red—much more so than an unaffected fundus. Fundus appearance in SWS is described as 'Tomato catsup fundus'. Choroidal hemangioma is not malignant. Sturge-Weber syndrome is a phakomatosis (you might also see encephalofacial or cerebrofacial angiomatosis). The hallmark skin finding in SWS is the port-wine stain. It presents at birth and comports to the distribution of one or more divisions of CN5. Not all infants with SWS have a port-wine stain; some cases can cross the midline of the face. Not all infants with a port-wine stain have 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)

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
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.

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.

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, Neves of Ota.

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. Risk of CNVM.

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

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


What condition is the diffuse choroidal hemangioma associated with? 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).

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?
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: 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)

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

2) **Medulloepithelioma** (aka diktyoma): Benign but locally aggressive neoplasia of nonpigmented epithelium of CB. Presents: Iris mass before age 10 years. Can bleed → hyphema → increased IOP → glaucoma. Locally invasive → death. Tx: Enucleate

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

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

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

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1) **Nevus**: Common. Benign

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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**

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**Intraretinal 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
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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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 +/−-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
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) 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

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'

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Yes, but it’s uncommon

Does the choroidal hemangioma have malignant potential?
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Isolated/focal choroidal hemangioma: Very rare. Characteristic a-scan pattern

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

With what 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
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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

- **Diffuse choroidal hemangioma**

  5) Associated with: Sturge-Weber syndrome. Risk of CNVM

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

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

  2) Melanocytoma: Usually juxtapapillary. Malignant transformation extremely rare

  1) Nevus: Common. Benign

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**Iris/Ciliary Body**

- **Intraocular Tumors of Childhood**

- Iris/Ciliary Body

- Choroid

- RPE

- Retina
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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4) 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

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’
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.


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.

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: 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 fundus looks very red, much more so than an unaffected fundus. 'Tomato catsup fundus'. Can be present bilaterally but uncommon. No malignant potential.

Sturge-Weber syndrome (SWS): Associated with diffuse choroidal hemangioma. About half of SWS have diffuse choroidal hemangioma. The fundus appearance looks like 'Tomato catsup fundus'.
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**

**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?**
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

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**5) Diffuse choroidal hemangioma**
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:** 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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**Intraocular Tumors of Childhood**

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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? 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
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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**Iris/Ciliary Body**

**Choroid**

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**Intraocular Tumors of Childhood**

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

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

3. **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

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

5. **Can the choroidal hemangioma be present bilaterally?**
   - Yes, but it’s uncommon

6. **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?**

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

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**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

- **Iris/Ciliary Body**

- **Choroid**
Intraocular Tumors of Childhood

Iris/Ciliary Body

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3) **Osteoma:** Benign bony tumor, most common in teen years, females. Risk of CNVM
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5) **Diffuse choroidal hemangioma:** Unilateral. Found in Sturge-Weber syndrome

RPE

1) ?

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

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RPE

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

Retina
Intraocular Tumors of Childhood

Iris/Ciliary Body

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What is the clinical appearance of CHRPE?

RPE

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

Retina
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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**
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**Iris/Ciliary Body**

**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?**

**RPE**

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

Iris/Ciliary Body

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RPE

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

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

Retina
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?
Common

Is it a hamartoma or a choristoma?

RPE

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

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**Iris/Ciliary Body**

**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

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**RPE**

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

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**Retina**
Intraocular Tumors of Childhood

Iris/Ciliary Body

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

**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

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

Choroid

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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
- Or Grouped or Multifocal CHRPE: Large lesion(s) surrounded by a few smaller ones
Intraocular Tumors of Childhood

**Iris/Ciliary Body**

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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

Iris/Ciliary Body

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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

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

Iris/Ciliary Body

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Choroid

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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

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

Iris/Ciliary Body

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

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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

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**

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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**Iris/Ciliary Body**

A **CHRPE-like lesion** is: (both eponymous and non-eponymous)

Familial adenomatous polyposis, aka Gardner syndrome

**A CHRPE-like lesion** is associated with a potentially fatal inherited syndrome. What is the name 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

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

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**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

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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?
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RPE

1) Congenital hypertrophy of the RPE (CHRPE)

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

-- If the lesions are distributed pattern

-- If the shape of the lesions is pisciform bi- v unilateral distribution pattern
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

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

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**What does pisciform mean?** *Fish-shaped*

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

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   - Retina
   - Choroid

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--They are hypo- vs hyperpigmented
--They point towards the optic nerve head

Retina

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**Intraocular Tumors of Childhood**

**Iris/Ciliary Body**

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

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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

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

**What is the treatment of choice?**

Prophylactic colectomy by what age will this occur? 40, maybe a little later

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- Benign tumors of bone
- Dental anomalies

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

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

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

Familial adenomatous polyposis, aka **Gardner syndrome**

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

- 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

---

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

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

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

Familial adenomatous polyposis, aka **Gardner syndrome**

When ‘colon cancer + ophthalmic issue’ is mentioned, two syndromes should come to mind. What is the other?

**Muir-Torre syndrome**

RPE

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**Muir-Torre syndrome**

How is Muir-Torre pronounced?

---Muir-Torre syndrome

Retina

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RPE
**Intraocular Tumors of Childhood**

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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’)
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Muir-Torre syndrome

**How is Muir-Torre pronounced?**

Mure (rhymes with ‘pure’) tore-AY

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Iris/Ciliary Body

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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

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

Muir-Torre syndrome

RETINA

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

RPE

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RPE

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**Intraocular Tumors of Childhood**

Iris/Ciliary Body

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

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--- What sorts of sebaceous lesions?

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Muir-Torre syndrome

---

**What sorts of sebaceous lesions?**

--Sebaceous-cell carcinomas

--Sebaceous-cell adenomas

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

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

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

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**Iris/Ciliary Body**

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

Iris/Ciliary Body

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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

RPE

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

1) Congenital hypoplasia of the fovea

Fovea is characterized according to its presentation. In what two ways does it present?
-- Hypoplasia of fovea
-- Atrophy of fovea: Fovea is absent or atrophic
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**Iris/Ciliary Body**

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

**Choroid**

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

**RPE**

**Retina**

1) ?
## Intraocular Tumors of Childhood

### Iris/Ciliary Body

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### RPE

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### Retina

1. **Retinoblastoma** (see the slide-set dedicated to it)
Iris/Ciliary Body

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**Iris/Ciliary Body**

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**RPE**

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

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**Retina**

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

1) **Retinoblastoma** (see the slide-set dedicated to it)
Intraocular Tumors of Childhood

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1) Retinoblastoma (see the slide-set dedicated to it)

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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

---

What is a hamartoma?

RPE

1) Combined hamartoma of the retina and RPE

Retina

1) Retinoblastoma (see the slide-set dedicated to it)
Intraocular Tumors of Childhood

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---

**What is a hamartoma?**

A tumor composed of histologically abnormal cells found in their normal location

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

- RPE cells (duh)
- Retinal glial cells

**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) **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**

1) **Retinoblastoma** (see the slide-set dedicated to it)
**Intraocular Tumors of Childhood**

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**Iris/Ciliary Body**

What is a hamartoma? A tumor composed of histologically normal cells found in their normal location but (aka choristoma)

---

**RPE**

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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 in kids slide-set)

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


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RPE

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

Retina

1) Combined hamartoma of the retina and RPE

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

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

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? That it is benign
Intraocular Tumors of Childhood

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RPE
Retina

What is the name of the reverse clinical entity, ie, one with normal cells found in an abnormal location?
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What is a hamartoma?
A tumor composed of histologically normal cells found in their normal location

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1) Combined hamartoma of the retina and RPE

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

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Intraocular Tumors of Childhood

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**Iris/Ciliary Body**

**Choroid**

**RPE**

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**Iris/Ciliary Body**

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:**
   - **RPE cells (duh) and retinal glial cells**

   **How does it present clinically?**
   - As a variably pigmented, slightly elevated retinal mass of the retina

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**Retina**

1) **Retinoblastoma** (see the slide-set dedicated to it)
Intraocular Tumors of Childhood

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7) Nevox: Common. Benign

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


Retinal Tumors

1) Retinoblastoma (see the slide-set dedicated to it)

RPE


- 1) Combined hamartoma of the retina and RPE

Retina

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Intraocular Tumors of Childhood

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Iris/Ciliary Body

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RPE

Retina

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1) Retinoblastoma (see the slide-set dedicated to it)

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?
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
Intraocular Tumors of Childhood

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1) Retinoblastoma (see the slide-set dedicated to it)

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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

Retina

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1) 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?

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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Retinoblastoma (see the slide-set dedicated to it)

Iris/Ciliary Body

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- \(RPE\)
- \(Retina\)
- \(1)\) Combined hamartoma of the retina and RPE

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Intraocular Tumors of Childhood

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

RPE

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

Retina

1) **Combined hamartoma of the retina and RPE:** Benign, congenital retinal lesion

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