Pediatric Orbital Tumors

A basic, obvious distinction
Pediatric Orbital Tumors

Benign

A basic, obvious distinction

Malignant
Pediatric Orbital Tumors

Which enlarge more rapidly in children--benign, or malignant tumors?
Pediatric Orbital Tumors

Which enlarge more rapidly in children--benign, or malignant tumors? Both can enlarge rapidly, so don’t use this to differentiate between them.
Pediatric Orbital Tumors

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What is the typical presentation of an orbital tumor?
Pediatric Orbital Tumors

Which enlarge more rapidly in children--benign, or malignant tumors?
Both can enlarge rapidly, so don’t use this to differentiate between them

What is the typical presentation of an orbital tumor?
Rapid unilateral proptosis +/- lid edema
Pediatric Orbital Tumors

Which enlarge more rapidly in children--benign, or malignant tumors? Both can enlarge rapidly, so don’t use this to differentiate between them.

What is the typical presentation of an orbital tumor? Rapid unilateral proptosis +/- lid edema.

With what non-neoplastic process is tumor presentation often confused?
Which enlarge more rapidly in children--benign, or malignant tumors?
Both can enlarge rapidly, so don’t use this to differentiate between them

What is the typical presentation of an orbital tumor?
Rapid unilateral proptosis +/- lid edema

With what non-neoplastic process is tumor presentation often confused?
Orbital cellulitis
**Pediatric Orbital Tumors**

- **Benign**
- **Malignant**

**Which enlarge more rapidly in children--benign, or malignant tumors?**
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**What is the typical presentation of an orbital tumor?**
Rapid unilateral proptosis +/- lid edema.

**With what non-neoplastic process is tumor presentation often confused?**
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**What signs should make you consider a malignant orbital process?**
1) 
2)
Pediatric Orbital Tumors

Benign

Malignant

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Rapid unilateral proptosis +/- lid edema.

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What signs should make you consider a malignant orbital process?
1) A ‘cellulitis’ that is…
2)
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Rapid unilateral proptosis +/- lid edema.

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What signs should make you consider a malignant orbital process?
1) A ‘cellulitis’ that is…not accompanied by erythema and warmth
2)
Pediatric Orbital Tumors

Which enlarge more rapidly in children—benign, or malignant tumors? Both can enlarge rapidly, so don’t use this to differentiate between them.

What is the typical presentation of an orbital tumor? Rapid unilateral proptosis +/- lid edema.

With what non-neoplastic process is tumor presentation often confused? Orbital cellulitis.

What signs should make you consider a malignant orbital process?
1) A ‘cellulitis’ that is… not accompanied by erythema and warmth
2) Periorbital ecchymosis and/or hematoma absent…
Pediatric Orbital Tumors

Which enlarge more rapidly in children--benign, or malignant tumors? Both can enlarge rapidly, so don’t use this to differentiate between them.

What is the typical presentation of an orbital tumor? Rapid unilateral proptosis +/- lid edema.

With what non-neoplastic process is tumor presentation often confused? Orbital cellulitis.

What signs should make you consider a malignant orbital process?  
1) A ‘cellulitis’ that is…not accompanied by erythema and warmth  
2) Periorbital ecchymosis and/or hematoma absent…a history of trauma.
Pediatric Orbital Tumors

Benign

Malignant

basic/obvious yet again

?
Pediatric Orbital Tumors

- Benign
- Malignant
  - Primary
  - Metastatic
  - basic/obvious yet again
Pediatric Orbital Tumors

- Benign
- Malignant
  - Metastatic
  - Primary
    - ?
    - ?
    - ?
    - ?
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary
  Rhabdomyosarcoma
  Osteosarcoma
  Chondrosarcoma
  Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

Notice a pattern regarding the primary orbital malignancies?
Notice a pattern regarding the primary orbital malignancies?

**Sarcomas** are far and away the most common primary malignancies of the orbit.
**Pediatric Orbital Tumors**

- **Benign**
- **Malignant**
  - **Metastatic**
  - **Primary**
    - Rhabdomyosarcoma
    - Osteosarcoma
    - Chondrosarcoma
    - Fibrosarcoma
    - Carcinoma?

Notice a pattern regarding the primary orbital malignancies? **Sarcomas** are far and away the most common primary malignancies of the orbit.

What about orbital carcinomas?
Pediatric Orbital Tumors

- Benign
- Malignant
  - Primary
    - Rhabdomyosarcoma
    - Osteosarcoma
    - Chondrosarcoma
    - Fibrosarcoma
    - Carcinoma? Nah
  - Metastatic

Notice a pattern regarding the primary orbital malignancies?
**Sarcomas** are far and away the most common primary malignancies of the orbit.

What about orbital carcinomas? These are very rare in the pediatric population.
Pediatric Orbital Tumors

**Benign**

**Malignant**

**Primary**
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

**Metastatic**

Speaking of the orbital sarcomas: Which is most common?

Rhabdomyosarcoma

How much more common is it?

The incidence of rhabdo exceeds that of all the other orbital sarcomas combined

Between what ages do most cases of rhabdo declare?

5 and 7 years

We think of rhabdo as a neoplasm of childhood, and by and large it is. What percent of cases present at age 16 years or older?

10% Make sure to keep rhabdo in mind in non-children.
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

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Pediatric Orbital Tumors

- Benign

- Malignant
  - Primary
    - Rhabdomyosarcoma
    - Osteosarcoma
    - Chondrosarcoma
    - Fibrosarcoma
  - Metastatic

Speaking of the orbital sarcomas: Which is most common? Rhabdomyosarcoma

How much more common is it?
Pediatric Orbital Tumors

**Benign**

**Malignant**

**Metastatic**

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- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Speaking of the orbital sarcomas: Which is most common? Rhabdomyosarcoma

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To make matters worse, if a pt has a personal hx of a particular malignancy, it may greatly increase his/her risk of developing a sarcoma (any sarcoma—not just rhabdo). What is that malignancy?

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

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To make matters worse, if a pt has a personal hx of a particular malignancy, it may greatly increase his/her risk of developing a sarcoma (any sarcoma--not just rhabdo). What is that malignancy? Retinoblastoma (Rb)
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

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To make matters worse, if a pt has a personal hx of a particular malignancy, it **may** greatly increase his/her risk of developing a sarcoma (any sarcoma—not just rhabdo). What is that malignancy?

Unpack that ‘may.’ Under what circumstance would a hx of Rb increase the risk of developing a sarcoma?
Pediatric Orbital Tumors

Benign

Malignant

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Repack that ‘may.’ Under what circumstance would a hx of Rb increase the risk of developing a sarcoma? When the Rb mutation is heritable (ie, is in the germline)
Pediatric Orbital Tumors

Benign

Malignant

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Metastatic

Speaking of the orbital sarcomas: Which is most common? Rhabdomyosarcoma

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What is that malignancy?

Ret.

Unpack that ‘may.’ Under what circumstance would a hx of Rb increase the risk of developing a sarcoma?

When the Rb mutation is heritable (ie, is in the germline)

‘When the Rb is heritable’--that means the mutation was inherited, right?
Pediatric Orbital Tumors

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‘When the Rb is heritable’—that means the mutation was inherited, right? Nope. While all inherited Rb is heritable, all heritable Rb was not inherited. (If this is confusing, review the slide-set entitled Concerning retinoblastoma.)
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
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What is that malignancy? Retinoblastoma (Rb)

Unpack that 'may.' Under what circumstance would a hx of Rb increase the risk of developing a sarcoma?

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The risk of a later sarcoma is even higher if the Rb was managed with a particular treatment modality—which one?

External-beam radiation therapy
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The risk of a later sarcoma is even higher if the Rb was managed with a particular treatment modality—which one? External-beam radiation therapy
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Pediatric Orbital Tumors

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Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

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Metastatic

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We think of rhabdo as a neoplasm of childhood, and by and large it is. What percent of cases present at age 16 years or older? 10%

Note: The ‘5-7’ range is from the most recent edition of the *Peds* book. OTOH, the *Orbit* book gives a range of 8-10. (FWIW, at the time of this writing the *Peds* book is the more recent of the two.) *EyeWiki* gives a range of 7-8. The truth is out there. Caveat emptor.
Pediatric Orbital Tumors

**Benign**

**Malignant**

**Metastatic**

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Pediatric Orbital Tumors

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Speaking of the orbital sarcomas: Which is most common?
Rhabdomyosarcoma

What is the cell of origin for rhabdo?

Undifferentiated/pluripotent mesenchymal cells (not the EOMs--a common misconception!)

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

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Is it painful, or painless?
In the vast majority (90%), it is painless

What are the four histological subtypes of rhabdo?

--- Embryonal--- Alveolar--- Pleomorphic--- Botyroid
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

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Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
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**Primary**

- **Rhabdomyosarcoma**
- Osteosarcoma
- Chondrosarcoma
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Pediatric Orbital Tumors

**Benign**

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Rhabdomyosarcoma

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

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What are the four histological subtypes of rhabdo?
--Embryonal
--Alveolar
--Pleomorphic
--Botryoid

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

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--Alveolar?
--Pleomorphic?
--Botryoid?

Which subtype does not occur as a primary in the orbit?
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

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Which subtype does not occur as a primary in the orbit? Botryoid

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

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--Pleomorphic
--Botryoid

Which subtype does not occur as a primary in the orbit? Botryoid

If not as a primary, then how does botryoid get in the orbit?

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Speaking of the orbital sarcomas: Which is most common? Rhabdomyosarcoma

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What are the four histological subtypes of rhabdo? --Embryonal --Alveolar --Pleomorphic --Botryoid

Which subtype does not occur as a primary in the orbit? Botryoid

If not as a primary, then how does botryoid get in the orbit? Via extension from a sinus, or the conjunctiva.
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

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Which subtype is most common?
Embryonal
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

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Chondrosarcoma

Fibrosarcoma

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--Alveolar
--Pleomorphic
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Which subtype is most common?
Embryonal
Pediatric Orbital Tumors

Benign

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--Embryonal?
--Alveolar?
--Pleomorphic?
--Botryoid

Which subtype carries the poorest prognosis?
Alveolar

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

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Pediatric Orbital Tumors

Benign

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Primary

Rhabdomyosarcoma

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--Embryonal
--Alveolar
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--Botryoid

Which subtype carries the poorest prognosis?
Alveolar
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma
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--Embryonal
--Alveolar
--Pleomorphic
--Botryoid

What subtype carries the poorest prognosis?
Alveolar

What is the 5-year survival rate for the alveolar subtype?
Per the Peds book, 75%
Per the Orbit book, only 10%
Pediatric Orbital Tumors

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Rhabdomyosarcoma

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--Embryonal
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Which subtype carries the poorest prognosis?
Alveolar

What is the 5-year survival rate for the alveolar subtype?
Per the Peds book, 75%
Pediatric Orbital Tumors

Speaking of the orbital sarcomas: Which is most common? Rhabdomyosarcoma

What is the cell of origin for rhabdo? Undifferentiated/pluripotent mesenchymal cells (not the EOMs—a common misconception!)

What is the classic presentation of orbital rhabdo? Like that of other orbital tumors in kids: Rapid unilateral proptosis associated with lid edema and discoloration

Is it painful, or painless? In the vast majority (90%), it is painless

What are the four histological subtypes of rhabdo? --Embryonal --Alveolar --Pleomorphic --Botryoid

Which subtype carries the poorest prognosis? Alveolar

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

What is the 5-year survival rate for the alveolar subtype? Per the Peds book, 75%

What is the 10-year survival rate?
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

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Per the Orbit book, only 10%!
Pediatric Orbital Tumors

**Benign**

- Hemangiomas
- Malformations
- Lymphangioma
- Cavernous hemangioma
- Port-wine stain
- Fibrous dysplasia
- Optic nerve glioma
- Plexiform neurofibroma

**Malignant**

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
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**Metastatic**

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Neither book addresses this

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Malignant

- Metallic

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Under what circumstances is the pleomorphic subtype encountered? When the pt is an adult (it essentially never occurs in kids)
Regarding metastatic dz to the eye or orbit: In what key manner does the pediatric population differ from the adult? In adults, In children,
Pediatric Orbital Tumors

Regarding metastatic dz to the eye or orbit: In what key manner does the pediatric population differ from the adult? In adults, mets tend to go to the **eye**; but…
In children,

- Osteosarcoma
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Regarding metastatic dz to the eye or orbit: In what key manner does the pediatric population differ from the adult? In adults, mets tend to go to the **eye**; but...

Where specifically in the adult eye do mets tend to go?
Pediatric Orbital Tumors

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In adults, mets tend to go to the **eye**; but…
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**Where specifically in the adult eye do mets tend to go?**
The choroid
Pediatric Orbital Tumors

- Benign
- Malignant
  - Metastatic
    - Primary
      - Rhabdomyosarcoma
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Pediatric Orbital Tumors

- Benign
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    - Neuroblastoma
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Briefly, what is neuroblastoma (Nb)?

A malignancy arising from precursor cells that give rise to the sympathetic chain and a portion of the adrenal medulla.

Where does the primary arise?

In the sympathetic chain, or the adrenal medulla.

Where does Nb rank as a cause of cancer in childhood?

It is the most common cause of extracranial solid cancer in childhood.

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**What proportion of Nb cases metastasize to the orbit?**
About 20%

**Of all orbital mets in children, what proportion are due to Nb?**
About 90%!
Pediatric Orbital Tumors

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It is #1.

**What is the average age at diagnosis?**
Two years.

**What proportion of cases are diagnosed by age 5?**
About 90%.

**Can it present in infancy, ie, before age 12 months?**
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**Malignant**

Metastatic Neuroblastoma

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What critter-based description is used for the appearance of bilateral ecchymoses in Nb?
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**Pediatric Orbital Tumors**

**Benign**

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With regard to Nb, what is the other ocular manifestation of note? (It’s paraneoplastic, not metastatic.)

Opsoclonus
It is a saccadic intrusion.
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In two words, what sort of condition is opsoclonus?
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It is a saccadic intrusion
Pediatric Orbital Tumors

Benign

Malignant

Metastatic
- Neuroblastoma
- Ewing sarcoma

Primary
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
Pediatric Orbital Tumors

**Benign**
- Bony origin
- Vascular origin
- Neural origin

**Malignant**
- Metastatic
  - Neuroblastoma
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- Primary
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  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma
Pediatric Orbital Tumors

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Pediatric Orbital Tumors

Benign
- Bony origin
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- Neural origin

Malignant
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- Primary
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  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma
Briefly, what is fibrous dysplasia?

A developmental condition in which normal bone is replaced by fibro-osseous tissue.

In what two ways can it be clinically significant?

1. It can be cosmetically disfiguring.
2. It can be blinding if the optic nerve becomes compressed.
Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin

Malignant
- Metastatic
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Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia

Malignant

- Vascular origin
- Neural origin
- Metastatic
  - Osteosarcoma
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Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia

- Vascular origin

- Neural origin

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- Metastatic origin
  - Osteosarcoma
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Briefly, what is fibrous dysplasia?
A developmental condition in which normal bone is replaced by fibro-osseous tissue

In what two ways can it be clinically significant?
--It can be...
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Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin

Malignant
- Metastatic origin
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  - Chondrosarcoma
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**Briefly, what is fibrous dysplasia?**
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Pediatric Orbital Tumors

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    - Fibrous dysplasia
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Malignant
  - Metastatic
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Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
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  - Hemangiomas
  - Malformations

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(Why the dashed lines, you say? Un momento por favor)
Pediatric Orbital Tumors

Benign
- Bony origin
- Vascular origin
- Neural origin
  - Fibrous dysplasia
  - Hemangiomas
    - Capillary
    - Strawberry
    - Cavernous hemangioma
  - Malformations

Malignant
- Metastatic
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Pediatric Orbital Tumors

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Hemangiomas
- Capillary
- Strawberry
- Cavernous hemangioma

Malformations

Malignant
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Re the terms *capillary hemangioma* and *strawberry hemangioma*: 
**Pediatric Orbital Tumors**

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**Hemangiomas**

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- Cavernous hemangioma

Malformations

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Re the terms *capillary hemangioma* and *strawberry hemangioma*: The 2018-19 edition of the BCSC *Peds* book indicates they are outdated, and such lesions should be referred to simply as ‘hemangiomas.’ However, it also acknowledges that these terms persist in the ophthalmic literature, so they will be included in this slide-set.
**Pediatric Orbital Tumors**

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- Bony origin
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Re the terms *capillary hemangioma* and *strawberry hemangioma*: The 2018-19 edition of the BCSC Peds book indicates they are outdated, and such lesions should be referred to simply as ‘hemangiomas.’ However, it also acknowledges that these terms persist in the ophthalmic literature, so they will be included in this slide-set. Also per the Peds book, and despite its name, the *cavernous hemangioma* is classified as a ‘vascular malformation,’ not a hemangioma.
Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

Capillary

Strawberry

Hemangiomas

Bony origin

Fibrous dysplasia

Vascular origin

Neural origin

Metastatic origin

Optic nerve glioma

Plexiform neurofibroma

Lymphangioma

Cavernous hemangioma

Port-wine stain

Fibrous dysplasia

Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

It is the most common.

Yes, it is more common in girls.

About 1/3 are present at birth; almost all have declared by age 6 months.

It's not known for certain, but some experts believe they originate as nests of placental cells that ‘metastasized’ to the fetus.

-- Initially they grow, reaching maximum size by around age 12 months
-- Then they involute beginning at about age 2 years--75% will resolve by age 4-5 years
Pediatric Orbital Tumors

Benign
- Bony origin
  - Hemangiomas
    - Capillary
    - Strawberry
  - Fibrous dysplasia

Vascular origin
- Vascular malformations
  - Hemangiomas
    - Capillary
    - Strawberry
  - Lymphangioma
  - Cavernous hemangioma
  - Port-wine stain

Neural origin
- Neural malformations
  - Optic nerve glioma
  - Plexiform neurofibroma

Malignant
- Metastatic
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Is there a gender predilection? Yes, it is more common in girls.

At what age do orbital capillary hemangiomas present? About 1/3 are present at birth; almost all have declared by age 6 months.

What is the origin of these lesions? It’s not known for certain, but some experts believe they originate as nests of placental cells that ‘metastasized’ to the fetus.

What is the ‘life cycle’ of capillary hemangiomas?
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Capillary
- Strawberry

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Pediatric Orbital Tumors

Benign
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Malignant
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Pediatric Orbital Tumors

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As a bluish (if deep) or strawberry-like (if superficial) lesion of the periorbital skin.

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What does capillary hemangioma present?
As a bluish (if deep) or strawberry-like (if superficial) lesion of the periorbital skin.

What are the options for managing orbital capillary hemangiomas?
- Observation is reasonable for small, nonamblyogenic lesions
- For lesions requiring treatment, systemic propranolol is probably first-line.
- Superficial lesions can be treated with topical timolol, or laser.
- Treatment-resistant lesions may require surgical excision.

What is the ‘life cycle’ of capillary hemangiomas?
--Initially they grow, reaching maximum size by around age 12 months.
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Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia

- Vascular origin
  - Capillary
  - Strawberry

- Neural origin
  - Hemangiomas
  - Optic nerve glioma
  - Plexiform neurofibroma

Malignant

- Metastatic origin
  - Neuroblastoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

Metastatic

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How does capillary hemangioma present?

As a bluish (if deep) or strawberry-like (if superficial) lesion of the periorbital skin.

At what age do orbital capillary hemangiomas present?

About 1/3 are present at birth; almost all have 'declared' by age 6 months.

What are the options for managing orbital capillary hemangiomas?

-- Observation is reasonable for small, nonamblyogenic lesions
-- Other treatments include systemic propranolol, topical timolol, or laser treatment for superficial lesions.
-- Treatment-resistant lesions may require surgical excision.
Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia
- Vascular origin
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    - Capillary
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Primary
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Pediatric Orbital Tumors

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Pediatric Orbital Tumors

Benign

Metastatic

Neural

Vascular

Bony origin

Fibrous dysplasia

Hemangiomas

Capillary

Strawberry

1. Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence? It is the most common.

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Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin
  - Hemangiomas
  - Malformations
    - ?
    - ?
    - ?

Malignant
- Metastatic
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Pediatric Orbital Tumors

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Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Hemangiomas
- Malformations
  - Lymphangioma
  Orbital lymphatic malformation

Malignant
- Neural origin
- Malformations
- Lymphangioma/Cavernous hemangioma
  - Port-wine stain
- Bony origin
- Malformations
  - Hemangioma
- Vascular origin
- Malformations
- Lymphangioma/Cavernous hemangioma
  - Port-wine stain

Metastatic
- Primary
  - Neuroblastoma
  - Ewing sarcoma
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

Be aware that, per the Peds and Orbit books, the term *lymphangioma* has given way to *orbital lymphatic malformation*
Pediatric Orbital Tumors

Benign
- Vascular origin
  - Hemangiomas
- Neural origin
  - Malformations
    - Lymphangioma

Malignant
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
- Primary
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma

What is the classic clinical presentation of an orbital lymphangioma?

- Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?

- Lymphatic-type endothelial cells lining lymph-filled spaces

Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?

- Lymphoid hyperplasia
- Intralesional hemorrhage
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces
Pediatric Orbital Tumors

Benign
- Vascular origin
  - Hemangiomas
- Neural origin
  - Malformations
    - Lymphangioma

Malignant
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
- Primary
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?
--
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**Pediatric Orbital Tumors**

**Benign**
- Vascular origin
  - Hemangiomas
- Neural origin
  - Optic nerve glioma
  - Plexiform neurofibroma
- Malformations
  - Lymphangioma

**Malignant**
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma

**Primary**
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma

*What is the classic clinical presentation of an orbital lymphangioma?*
Rapid-onset proptosis

*What does histologic examination of a lymphangioma reveal?*
Lymphatic-type endothelial cells lining lymph-filled spaces

*Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?*
--Lymphoid hyperplasia
--Intralesional hemorrhage
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

What is the classic health event for precipitating lymphoid hyperplasia in these patients?

- Lymphoid hyperplasia
- Intrallesional hemorrhage
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

What is the classic health event for precipitating lymphoid hyperplasia in these patients?
Upper respiratory tract infection

--Lymphoid hyperplasia
--Intralesional hemorrhage
Pediatric Orbital Tumors

**Benign**
- Vascular origin:
  - Hemangiomas
- Neural origin:
  - Malformations
  - Lymphangioma
  - Other: Fibrous dysplasia, Optic nerve glioma, Plexiform neurofibroma

**Malignant**
- Metastatic:
  - Neuroblastoma
  - Ewing sarcoma
- Primary:
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma

**What is the classic clinical presentation of an orbital lymphangioma?**
Rapid-onset proptosis

**What does histologic examination of a lymphangioma reveal?**
Lymphatic-type endothelial cells lining lymph-filled spaces

**How are lymphangiomas managed?**
- If threatening vision or producing unacceptable cosmesis: resection.
- Otherwise, conservatively.

**Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?**
- Lymphoid hyperplasia
- Intralesional hemorrhage
Pediatric Orbital Tumors

**Benign**
- Vascular origin
  - Hemangiomas
  - Malformations
    - Lymphangioma
- Bony origin
  - Fibrous dysplasia
- Neural origin
  - Optic nerve glioma
  - Plexiform neurofibroma

**Malignant**
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
- Primary
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma

**What is the classic clinical presentation of an orbital lymphangioma?** Rapid-onset proptosis.

**How are lymphangiomas managed?**
- If threatening vision, or producing unacceptable cosmesis--resection.
- Otherwise, conservatively.

**What does histologic examination of a lymphangioma reveal?** Lymphatic-type endothelial cells lining lymph-filled spaces.

**Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?**
- Lymphoid hyperplasia
- Intralesional hemorrhage
Pediatric Orbital Tumors

Benign

Vascular origin
- Hemangiomas
- Fibrous dysplasia

Bony origin
- Hemangiomas

Neural origin
- Malformations
  - Lymphangioma
  - Port-wine stain
  - Cavernous hemangioma

Malignant

Primary
- Neuroblastoma
- Ewing sarcoma
- Rhabdomyosarcoma

Metastatic

In one word, what sort of lesion is the PWS?

An angioma

By what 'official' name is it known?

Nevus flammeus

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
In one word, what sort of lesion is the PWS?
An angioma
Pediatric Orbital Tumors

**Benign**
- Vascular origin
  - Hemangiomas
    - Lymphangioma
    - Cavernous hemangioma
  - Malformations
    - Port-wine stain
    - Lymphangioma
    - Cavernous hemangioma

**Malignant**
- Neural origin
  - Benign
  - Malignant
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma

In one word, what sort of lesion is the PWS?
An angioma

By what ‘official’ name is it known?
Nevus flammeus
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas
  - Fibrous dysplasia
- Hemangiomas
- Malformations
  - Lymphangioma
  - Port-wine stain
  - Cavernous hemangioma

Malignant

- Neural origin
  - Neural malformations
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
- Metastatic
  - Rhabdomyosarcoma

In one word, what sort of lesion is the PWS? An angioma

By what ‘official’ name is it known? Nevus flammeus
Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin

- Hemangiomas
- Malformations
  - Lymphangioma
  - **Port-wine stain**
  - Cavernous hemangioma

Malignant

- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma

Primary

In one word, what sort of lesion is the PWS? An angioma

By what ‘official’ name is it known? **Nevus flammeus**

When does it present?
Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Hemangiomas

- Neural origin
  - Malformations
    - Lymphangioma
    - Port-wine stain
    - Cavernous hemangioma

Malignant
- Malformations
- Primary
  - Rhabdomyosarcoma
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma

In one word, what sort of lesion is the PWS? An angioma

By what ‘official’ name is it known? Nevus flammeus

When does it present? At birth
In one word, what sort of lesion is the PWS?
An angioma

By what ‘official’ name is it known?
Nevus flammeus

When does it present?
At birth

What is the typical pattern of distribution?
Pediatric Orbital Tumors

**Benign**
- Bony origin
  - Fibrous dysplasia
- Vascular origin
  - Hemangiomas
- Neural origin
  - Malformations
    - Lymphangioma
    - Port-wine stain
    - Cavernous hemangiomas

**Malignant**
- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
- Primary
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

*In one word, what sort of lesion is the PWS?*
An angioma

*By what ‘official’ name is it known?*
Nevus flammeus

*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
Pediatric Orbital Tumors

**Benign**
- Hemangiomas
- Malformations
- Lymphangioma
- Cavernous hemangioma
- Port-wine stain
- Nevus flammeus
- Fibrous dysplasia

**Malignant**
- Primary
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

**Metastatic**
- Neuroblastoma
- Ewing sarcoma

With what condition is the port wine stain (PWS) closely associated?
- Sturge-Weber syndrome
- Encephalotrigeminal angiomatosis

By what ‘official’ name is it known?
- Nevus flammeus

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

Do all infants with a PWS have SWS?
- No
Pediatric Orbital Tumors

**Benign**
- Hemangiomas
  - Vascular origin
- Neuronal malformations
  - Lymphangiomas
  - Cavernous hemangiomas
  - Port-wine stain
- Fibrous dysplasia

**Malignant**
- Primary
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

**Metastatic**
- Neuroblastoma
- Ewing sarcoma

*With what condition is the port wine stain (PWS) closely associated?* Sturge-Weber syndrome

*By what ‘official’ name is it known?* Nevus flammeus

*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

*With what condition is the PWS closely associated?* Sturge-Weber syndrome

*By what noneponymous name is Sturge-Weber known?* Encephalotrigeminal angiomatosis

*In one word, what sort of condition is Sturge-Weber?* A phakomatosis

*All infants with SWS have a PWS. Do all infants with a PWS have SWS?* No
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas

- Bony origin

- Neural origin
  - Malformations
    - Lymphangioma
    - Cavernous hemangioma

- Fibrous dysplasia

- Primary origin

Malignant

- Metastatic
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

1. With what condition is the port wine stain (PWS) closely associated? Sturge-Weber syndrome
2. By what noneponymous name is Sturge-Weber known? Encephalotrigeminal angiomatosis
3. In one word, what sort of condition is Sturge-Weber? A phakomatosis
4. All infants with SWS have a PWS. Do all infants with a PWS have SWS? No

- Port-wine stain
  - By what ‘official’ name is it known? Nevus flammeus
  - 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
Pediatric Orbital Tumors

Benign

Vascular origin

Hemangiomas

Bony origin

Neural origin

Malformations

Hemangiomas

Primary

Metastatic

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

Port-wine stain

By what ‘official’ name is it known?
Nevus flammeus

Cavernous hemangioma

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

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

Port-wine stain

By what ‘official’ name is it known?
Nevus flammeus

Cavernous hemangioma

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
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas
  - Lymphangioma
  - Cavernous hemangioma
  - Port-wine stain
  - Fibrous dysplasia

Malignant

- Bony origin
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

- Neural origin

Metastatic

- Neuroblastoma
- Ewing sarcoma

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber?
A phakomatosis

All infants with SWS have a PWS. Do all infants with a PWS have SWS?
No

By what ‘official’ name is it known?
Nevus flammeus

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
Pediatric Orbital Tumors

Benign

With what condition is the port wine stain (PWS) closely associated? Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known? Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber? A phakomatosis

Port-wine stain

Cavernous hemangioma

Malignant

Vascular origin

Hemangiomas

Bony origin

Neural origin

Malformations

Lymphangioma

Cavernous hemangioma

Port-wine stain

Fibrous dysplasia

Metastatic

Primary

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

In one word, what sort of lesion is the PWS? An angioma

By what ‘official’ name is it known? Nevus flammeus

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

With what condition is the port wine stain (PWS) closely associated? Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known? Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber? A phakomatosis

Port-wine stain

Cavernous hemangioma

Malignant

Vascular origin

Hemangiomas

Bony origin

Neural origin

Malformations

Lymphangioma

Cavernous hemangioma

Port-wine stain

Fibrous dysplasia

Metastatic

Primary

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

In one word, what sort of lesion is the PWS? An angioma

By what ‘official’ name is it known? Nevus flammeus

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
Pediatric Orbital Tumors

Benign

Malignant

Metastatic

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber?
A phakomatosis

All infants with SWS have a PWS. Do all infants with a PWS have SWS?
No

Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin

By what ‘official’ name is it known?
Nevus flammeus

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
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas
- Bony origin
- Neural origin
  - Malformations
  - Lymphangioma
  - Cavernous hemangioma
  - Port-wine stain
- Fibrous dysplasia

Malignant

- Primary
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

Metastatic

- Neuroblastoma

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of lesion is the PWS?
An angioma

By what ‘official’ name is it known?
Nevus flammeus

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

Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the

Abb., , and

A phakomatosis
Pediatric Orbital Tumors

Benign

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber?
A phakomatosis

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin

By what ‘official’ name is it known?
Nevus flammeus

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

Malignant

Metastatic

Vascular origin

Hemangiomas

Bony origin

Neural origin

Malformations

Lymphangioma

Cavernous hemangioma

Port-wine stain

Fibrous dysplasia

Primary

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

All infants with SWS have a PWS. Do all infants with a PWS have SWS?
No

Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin
Pediatric Orbital Tumors

Benign

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of lesion is the PWS?
An angioma

By what ‘official’ name is it known?
Nevus flammeus

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

By what more-descriptive name does the BCSC Peds book refer to them?
A phakomatosis

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin

Malignant

Metastatic

Neuroblastoma

Bony origin

Fibrous dysplasia

Primary

Metastatic

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Malformations

Lymphangioma

Cavernous hemangioma

Port-wine stain

Vascular origin

Hemangiomas
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas
- Bony origin
- Neural origin
  - Malformations
  - Lymphangioma
  - Cavernous hemangioma
  - Port-wine stain
- Fibrous dysplasia

Malignant

- Primary
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma
- Metastatic

With what condition is the port wine stain (PWS) closely associated?
- Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
- Encephalotrigeminal angiomatosis

In one word, what sort of lesion is the PWS?
- An angioma

By what ‘official’ name is it known?
- Nevus flammeus

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

By what more-descriptive name does the BCSC Peds book refer to them?
- As neuro-
**Pediatric Orbital Tumors**

**Benign**

- Hemangioma
- Lymphangioma
- Cavernous hemangioma
- Port-wine stain
- Fibrous dysplasia

**Malignant**

- Neuroblastoma
- Ewing sarcoma
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

**Metastatic**

- Neuroblastoma

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**Port-wine stain (PWS)**

- **With what condition is the port wine stain (PWS) closely associated?** Sturge-Weber syndrome
- **By what noneponymous name is Sturge-Weber known?** Encephalotrigeminal angiomatosis
- **In one word, what sort of condition is Sturge-Weber?** A phakomatosis
- **By what more-descriptive name does the BCSC Peds book refer to them?** As neuro-oculo

- **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

**By what ‘official’ name is it known?** Nevus flammeus
With what condition is the port wine stain (PWS) closely associated? Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known? Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber? A phakomatosis

Briefly, what is a phakomatosis? A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes, and skin.

By what more-descriptive name does the BCSC Peds book refer to them? As neuro-oculocutaneous syndromes

By what ‘official’ name is it known? Nevus flammeus

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
Pediatric Orbital Tumors

Benign

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber?
A phakomatosis

All infants with SWS have a PWS. Do all infants with a PWS have SWS?

Metastatic

By what ‘official’ name is it known?
Nevus flammeus

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
Pediatric Orbital Tumors

Benign

- Vascular origin
  - Hemangiomas
  - Malformations
    - Lymphangioma
    - Cavernous hemangioma
    - Port-wine stain

- Neural origin
  - Fibrous dysplasia

Malignant

- Bony origin
  - Primary
    - Neuroblastoma
    - Ewing sarcoma
    - Rhabdomyosarcoma
    - Osteosarcoma
    - Chondrosarcoma
    - Fibrosarcoma

- Metastatic
  - Neuroblastoma
  - Ewing sarcoma

**Port-wine stain**

*With what condition is the port wine stain (PWS) closely associated?*
- Sturge-Weber syndrome

*By what noneponymous name is Sturge-Weber known?*
- Encephalotrigeminal angiomatosis

*In one word, what sort of condition is Sturge-Weber?*
- A phakomatosis

*All infants with SWS have a PWS. Do all infants with a PWS have SWS?*
- No, it can occur in non-SWS infants

*By what ‘official’ name is it known?*
- Nevus flammeus

*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
Pediatric Orbital Tumors

Benign

Vascular

Hemangiomas
Bony

Neural

Malformations

Fibrous dysplasia

Primary

Metastatic

Neuroblastoma
Ewing sarcoma
Rhabdomyosarcoma
Osteosarcoma
Chondrosarcoma
Fibrosarcoma

With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known?
Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber?
A phakomatosis

All infants with SWS have a PWS. Do all infants with a PWS have SWS?
No, it can occur in non-SWS infants

Port-wine stain

PWS is associated with another, less well-known phakomatosis--which one?

By what ‘official’ name is it known?
Nevus flammeus

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

In one word, what sort of lesion is the PWS?
An angioma

By what ‘official’ name is it known?
Nevus flammeus

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
Pediatric Orbital Tumors

**Benign**

*Vascular origin*
- Hemangiomas
- Malformations
  - Lymphangioma
  - Cavernous hemangioma
  - Port-wine stain
- Fibrous dysplasia

**Malignant**

*Bone origin*
- Neuroblastoma
- Ewing sarcoma
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

*Neural origin*

**Metastatic**

- By what condition is the port wine stain (PWS) closely associated?
  - Sturge-Weber syndrome

- By what noneponymous name is Sturge-Weber known?
  - Encephalotrigeminal angiomatosis

- In one word, what sort of condition is Sturge-Weber?
  - A phakomatosis

- All infants with SWS have a PWS. Do all infants with a PWS have SWS?
  - No, it can occur in non-SWS infants

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

- PWS is associated with another, less well-known phakomatosis--which one?
  - Klippel-Trénaunay syndrome

- By what ‘official’ name is it known?
  - Nevus flammeus
Pediatric Orbital Tumors

Benign

With what condition is the port wine stain (PWS) closely associated? Sturge-Weber syndrome

By what noneponymous name is Sturge-Weber known? Encephalotrigeminal angiomatosis

In one word, what sort of condition is Sturge-Weber? A phakomatosis

All infants with SWS have a PWS. Do all infants with a PWS have SWS? No, it can occur in non-SWS infants

By what ‘official’ name is it known? Nevus flammeus

PWS is associated with another, less well-known phakomatosis--which one? Klippel-Trénaunay syndrome

How is this pronounced? TRIPpel tray-NO-NAY

Metastatic

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Vascular origin

Hemangiomas

Nevus flammeus

Bony origin

Lymphangioma

Cavernous hemangioma

Primary

Fibrous dysplasia

Secondary

Secondary malignancy

Malignant

Neuroblastoma

Ewing sarcoma

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Secondary malignancy

Metastatic

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

**Benign**
- Hemangiomas
- Lymphangiomas
- Cavernous hemangiomas
- Port-wine stain
- Fibrous dysplasia

**Malignant**
- Neuroblastoma
- Ewing sarcoma
- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

**Metastatic**
- Neuroblastoma
- Ewing sarcoma

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**Port-wine stain (PWS)**
- With what condition is the port wine stain (PWS) closely associated?
  - Sturge-Weber syndrome

- By what nonepithelial name is Sturge-Weber known?
  - Encephalotrigeminal angiomatosis

- In one word, what sort of condition is Sturge-Weber?
  - A phakomatosis

- All infants with SWS have a PWS. Do all infants with a PWS have SWS?
  - No, it can occur in non-SWS infants

- By what ‘official’ name is it known?
  - Nevus flammeus

- PWS is associated with another, less well-known phakomatosis—which one?
  - Klippel-Trénaunay syndrome

**Klippel-Trénaunay**
- How is this pronounced?
  - TRIP-pel tray-NO-NAY

- What is the typical pattern of distribution?
  - It comports to the distribution of one or more divisions of CN5
Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
  - Hemangiomas
- Neural origin
  - Malformations
    - Lymphangioma
    - Port-wine stain
    - Cavernous hemangioma
- Metastatic
  - Neuroblastoma

Where does the cavernous hemangioma rank among benign orbital tumors in adults?

Yes, it is more common in women.

It does when it is clinically significant.
Pediatric Orbital Tumors

Benign
- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin
- Malformations
  - Lymphangioma
  - Port-wine stain
  - Cavernous hemangioma

Malignant
- Metastatic
  - Neuroblastoma

Where does the cavernous hemangioma rank among benign orbital tumors in adults? It is the most common benign orbital tumor of adults.
Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia
- Vascular origin
  - Hemangiomas
- Neural origin

- Malformations
  - Lymphangiomata
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Malignant

- Metastatic
  - Neuroblastoma

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Pediatric Orbital Tumors

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- Bony origin
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- Neural origin
  - Malformations
    - Lymphangioma
    - Port-wine stain
    - Cavernous hemangioma

Malignant

- Metastatic
  - Neuroblastoma

- Primary
  - Neuroblastoma
  - Ewing sarcoma
  - Rhabdomyosarcoma
  - Osteosarcoma
  - Chondrosarcoma
  - Fibrosarcoma

Where does the cavernous hemangioma rank among benign orbital tumors in adults?
It is the most common benign orbital tumor of adults

Does cavernous hemangioma manifest a gender predilection?

Yes, it is more common in women.
Where does the cavernous hemangioma rank among benign orbital tumors in adults?
It is the most common benign orbital tumor of adults.

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Does it require surgical excision?
Pediatric Orbital Tumors

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It is the **most** common benign orbital tumor of adults

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Pediatric Orbital Tumors

Benign
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Malignant
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Now these two
Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia
- Vascular origin
- Neural origin
  - Optic nerve glioma
  - Plexiform neurofibroma

Primary

- Hemangiomas
- Malformations
  - Lymphangioma
  - Port-wine stain
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- Metastatic
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Where does optic nerve glioma rank as a cause of pediatric orbital tumors of neural origin?
Pediatric Orbital Tumors

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Malignant
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Where does optic nerve glioma rank as a cause of pediatric orbital tumors of neural origin?
It is the most common
Pediatric Orbital Tumors

Benign

- Neural origin
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Malignant

- Metastatic
  - Neuroblastoma
  - Ewing sarcoma

Vascular origin

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Bony origin

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Hemangiomas

Where does optic nerve glioma rank as a cause of pediatric orbital tumors of neural origin?
It is the most common

50% are associated with a syndrome— which one?
Pediatric Orbital Tumors

Benign

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Pediatric Orbital Tumors

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Where does optic nerve glioma rank as a cause of pediatric orbital tumors of neural origin?
It is the most common.

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

In a word, what sort of condition is NF1?
A phakomatosis
Pediatric Orbital Tumors

Benign

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As a child; the adult version carries a very poor prognosis.

What finding significantly worsens the prognosis in pediatric cases? Intracranial extension.
Pediatric Orbital Tumors

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Would you rather have an optic nerve glioma as a child or as an adult?
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  - Fibrous dysplasia

- **Malignant**
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What two specific cell types are implicated in plexiform neurofibroma formation?

What ophthalmic structure is typically involved?
The upper lid

These nonencapsulated, diffusely infiltrative tumors have a distinct consistency when palpated. What icky term is used to describe it?

'Bag of worms'

Are plexiform neurofibromas associated with a systemic condition?
Yes—like their cousin the optic nerve glioma, plexiform neurofibromas are strongly associated with NF1
**Pediatric Orbital Tumors**

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**Questions:**
- What two specific cell types are implicated in plexiform neurofibroma formation?
  - Schwann cells and fibroblasts
Pediatric Orbital Tumors

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Their predilection for the temporal aspect of the upper lid produces a specific configuration of the involved lid. What is it? 
The 'S-shaped lid'
**Pediatric Orbital Tumors**

<table>
<thead>
<tr>
<th>Benign</th>
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The upper lid

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The tumor gives rise to an ‘S-shaped lid’

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Are upper-lid plexiform neurofibromas potentially amblyogenic?

Yes
Pediatric Orbital Tumors

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**Malignant**

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  - The upper lid

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  - The tumor gives rise to an ‘S-shaped lid’

- Are upper-lid plexiform neurofibromas potentially amblyogenic?
  - Yes
Pediatric Orbital Tumors

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'Bag of worms'
Pediatric Orbital Tumors

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Are plexiform neurofibromas associated with a systemic condition?
Pediatric Orbital Tumors

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Yes--like their cousin the optic nerve glioma, plexiform neurofibromas are strongly associated with NF1
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What proportion of NF1 pts develop plexiform neurofibromas? About 1/3.

Are plexiform neurofibromas associated with a systemic condition? Yes—like their cousin the optic nerve glioma, plexiform neurofibromas are strongly associated with NF1.
Pediatric Orbital Tumors

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What proportion of plexiform neurofibroma pts have NF1?
Pretty much all of them

TL;DR Not all NF1 pts get plexiform neurofibromas, but everyone with plexiform neurofibromas has NF1
Pediatric Orbital Tumors

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Malignant

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**What ophthalmic structure is typically involved?**

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**Are plexiform neurofibromas associated with a systemic condition?**

- Yes--like their cousin the optic nerve glioma, plexiform neurofibromas are strongly associated with NF1

**Plexiform neurofibromas of the upper lid are a strong risk factor for what potentially blinding ocular condition?**

- Glaucoma

**Does it increase the risk of glaucoma in both eyes, or just the ipsilateral eye?**

- The ipsilateral eye
Pediatric Orbital Tumors

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  - Fibrous dysplasia

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  - Optic nerve glioma
  - Plexiform neurofibroma

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How strong of a risk factor?
- Very strong. 50% of these eyes have glaucoma

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How strong of a risk factor?
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### Pediatric Orbital Tumors

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- Yes--like their cousin the optic nerve glioma, plexiform neurofibromas are strongly associated with NF1
Pediatric Orbital Tumors

Benign

- Bony origin
  - Fibrous dysplasia

- Vascular origin

- Neural origin
  - Hemangiomas
  - Malformations
    - Lymphangioma
    - Port-wine stain
    - Cavernous hemangioma

Primary

- Neuroblastoma
- Ewing sarcoma
- Vascular origin
  - Hemangiomas
- Bony origin
- Neural origin
  - Optic nerve glioma

Plexiform neurofibroma

What specific cell type is implicated in plexiform neurofibroma formation?

Schwann cells and fibroblasts

What ophthalmic structure is typically involved?
The upper lid

These nonencapsulated, diffusely infiltrative tumors have a distinct consistency when palpated. What icky term is used to describe it?

‘Bag of worms’

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How strong of a risk factor?

Very strong. 50% of these eyes have glaucoma!

How strong of a risk factor?

Strong risk factor

for what potentially blinding ocular condition?

Glaucoma

Plexiform neurofibromas of the upper lid are a strong risk factor for what potentially blinding ocular condition?

Glaucoma

Does it increase the risk of glaucoma in both eyes, or just the ipsilateral eye?
The ipsilateral eye

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  - Plexiform neurofibroma
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What ophthalmic structure is typically involved? The upper lid

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