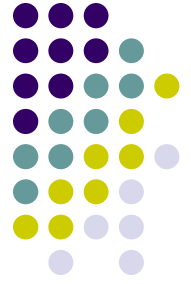
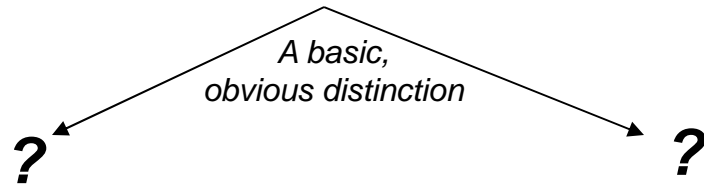
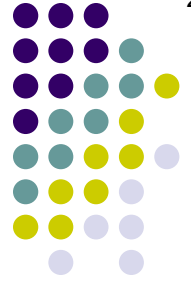
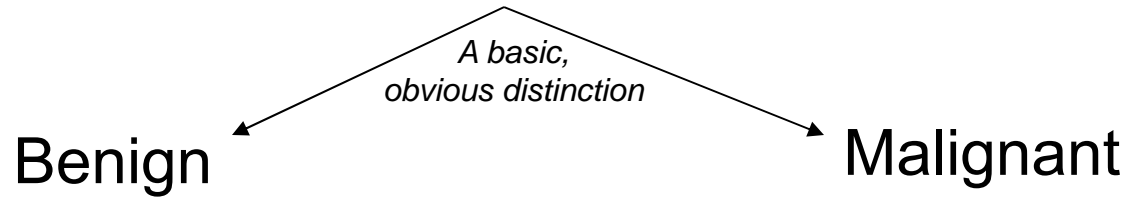


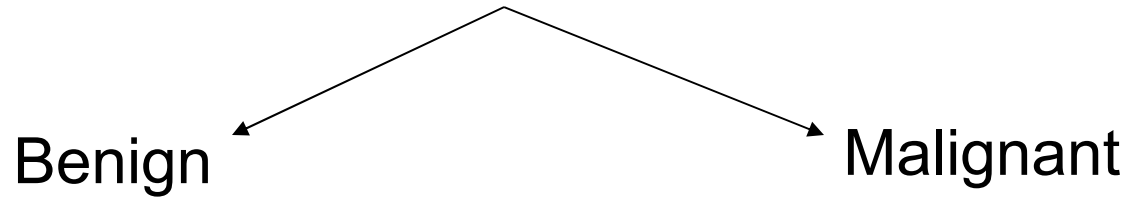
Pediatric Orbital Tumors



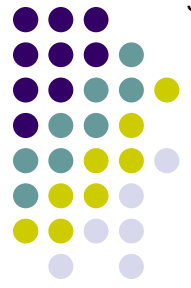
Pediatric Orbital Tumors



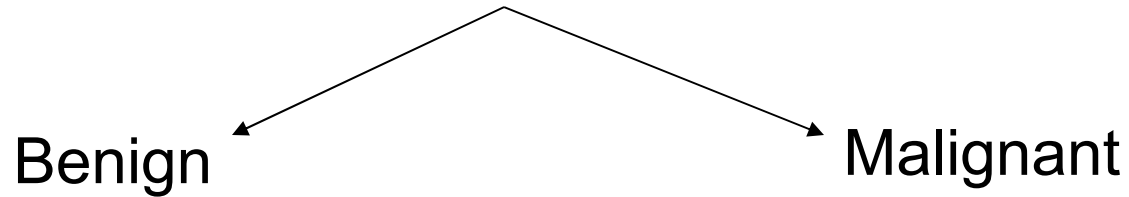
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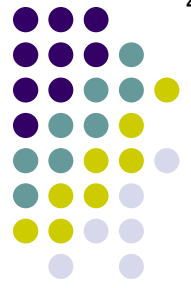
Which enlarge more rapidly in children--benign, or malignant tumors?



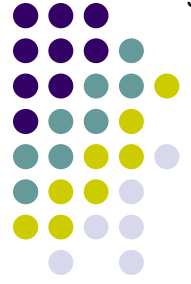
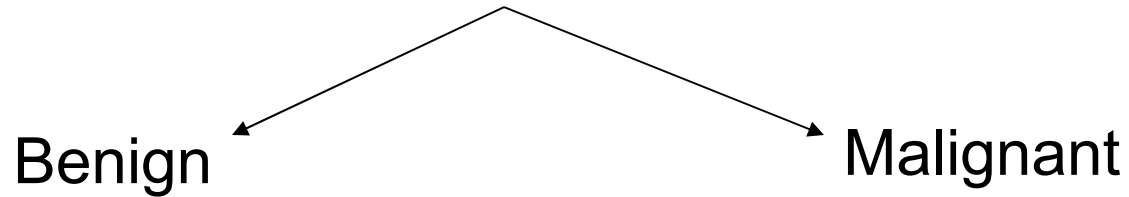
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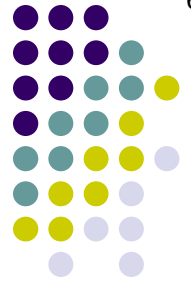
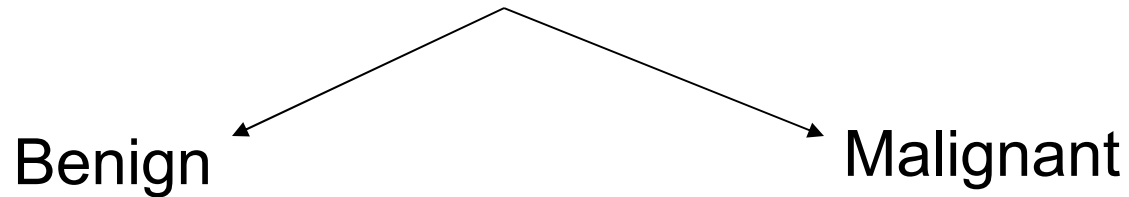
Pediatric Orbital Tumors



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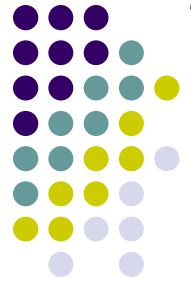
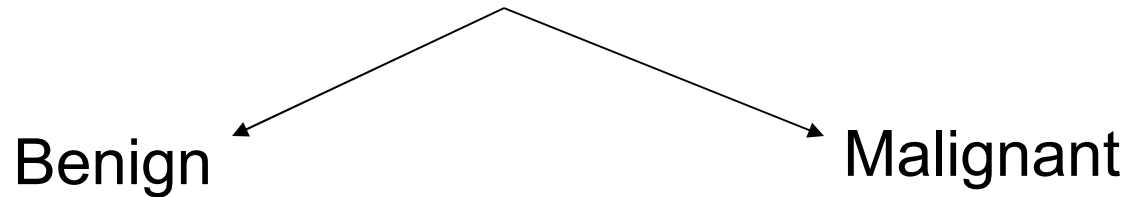
Pediatric Orbital Tumors



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

Pediatric Orbital Tumors

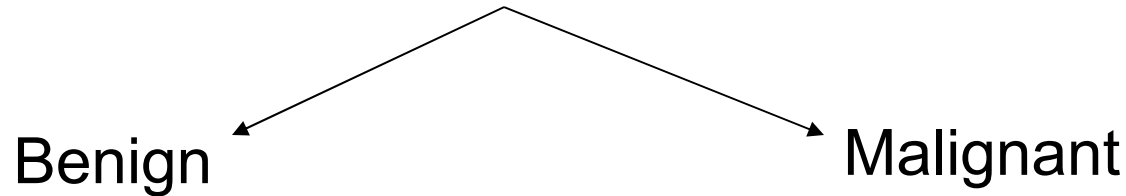


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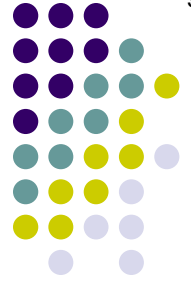
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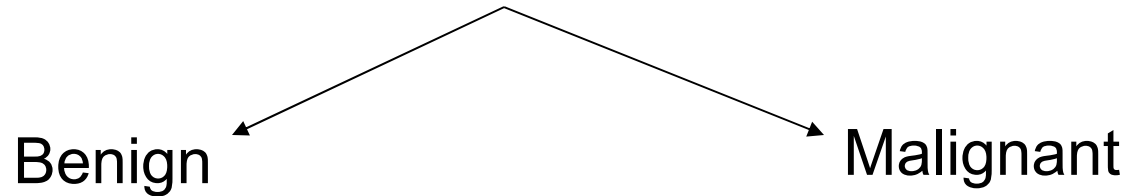
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Pediatric Orbital Tumors



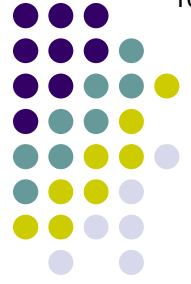
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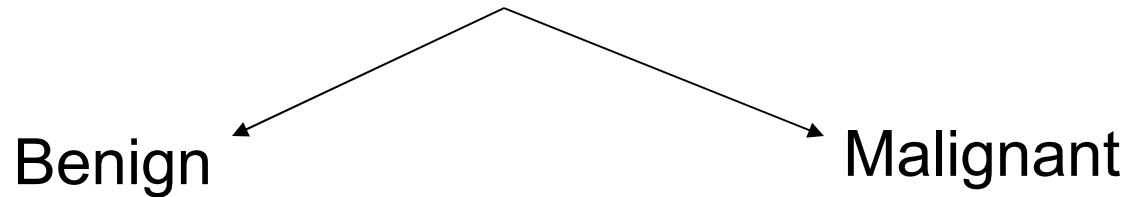
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What signs should make you consider a malignant orbital process?

- 1)
- 2)



Pediatric Orbital Tumors



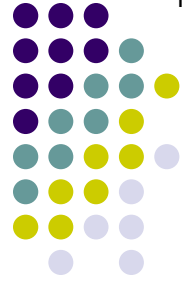
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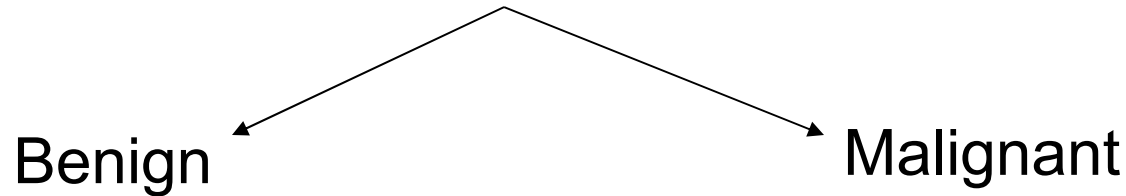
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What signs should make you consider a malignant orbital process?

- 1) A 'cellulitis' that is...
- 2)



Pediatric Orbital Tumors

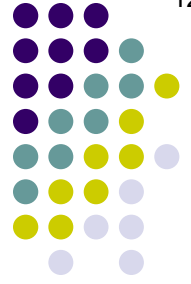


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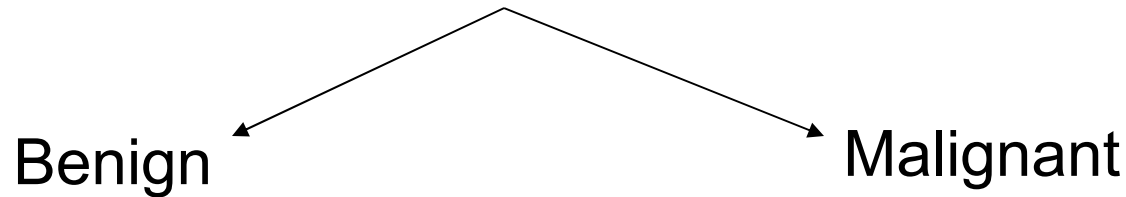
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Pediatric Orbital Tumors

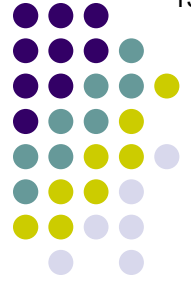


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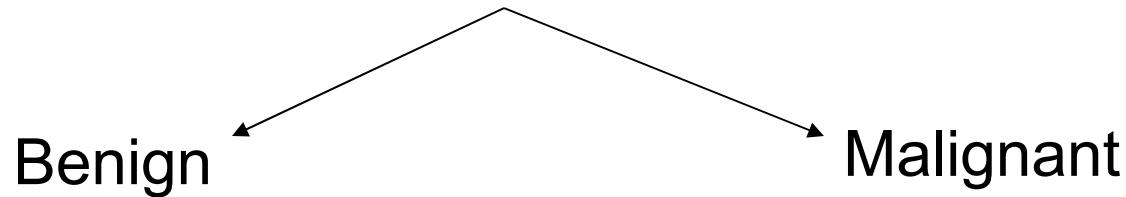
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2) Periorbital ecchymosis and/or hematoma absent...



Pediatric Orbital Tumors



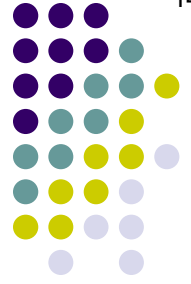
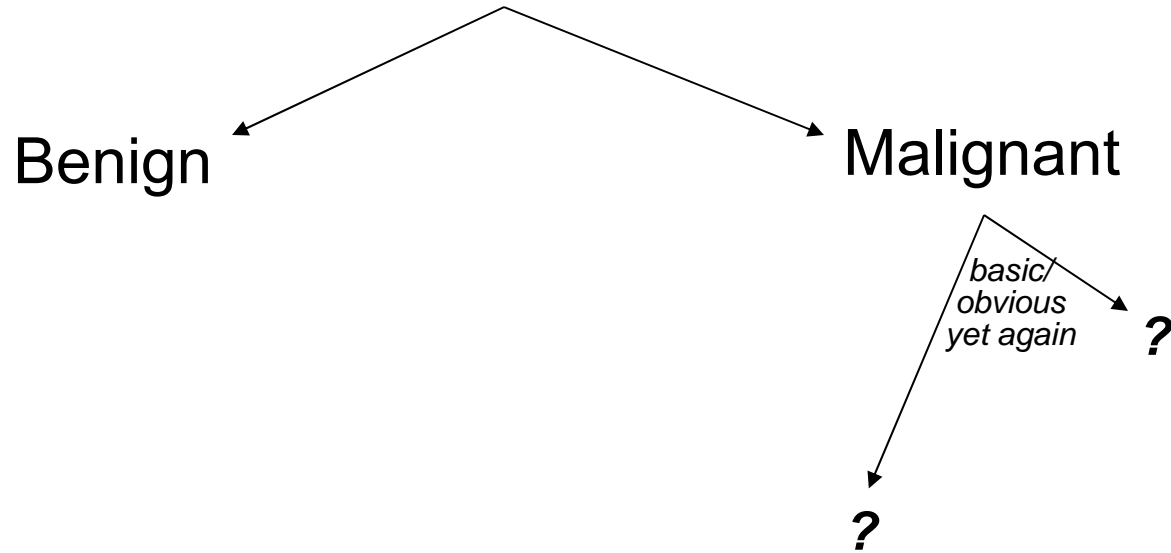
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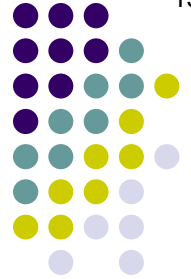
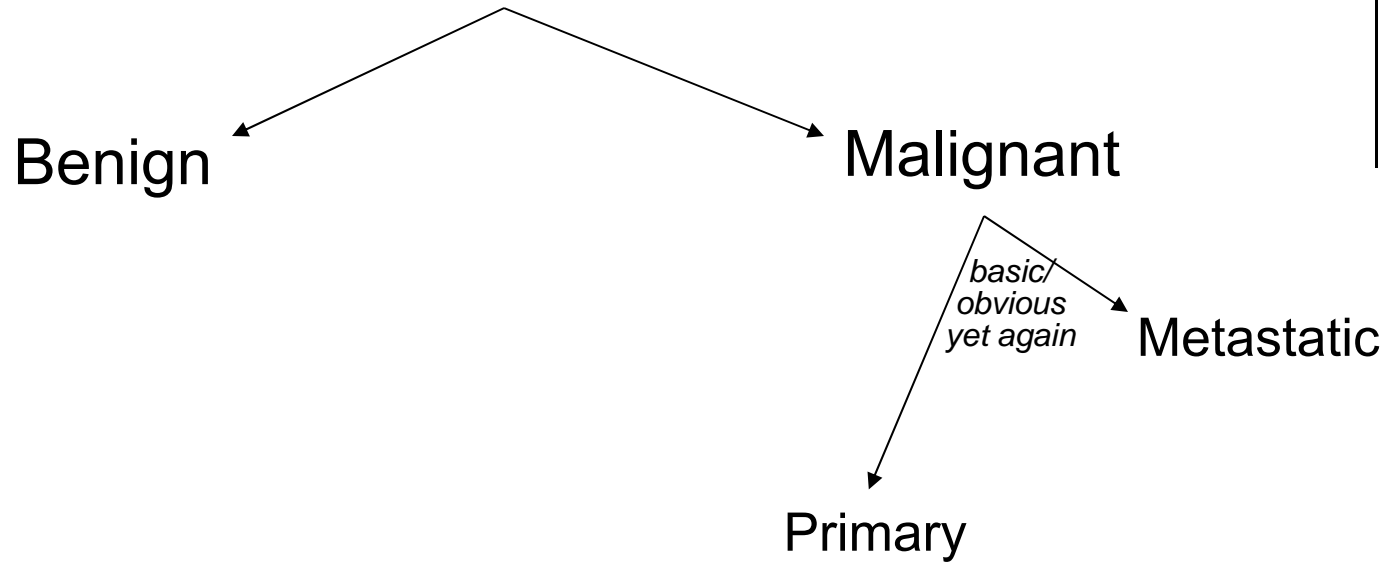
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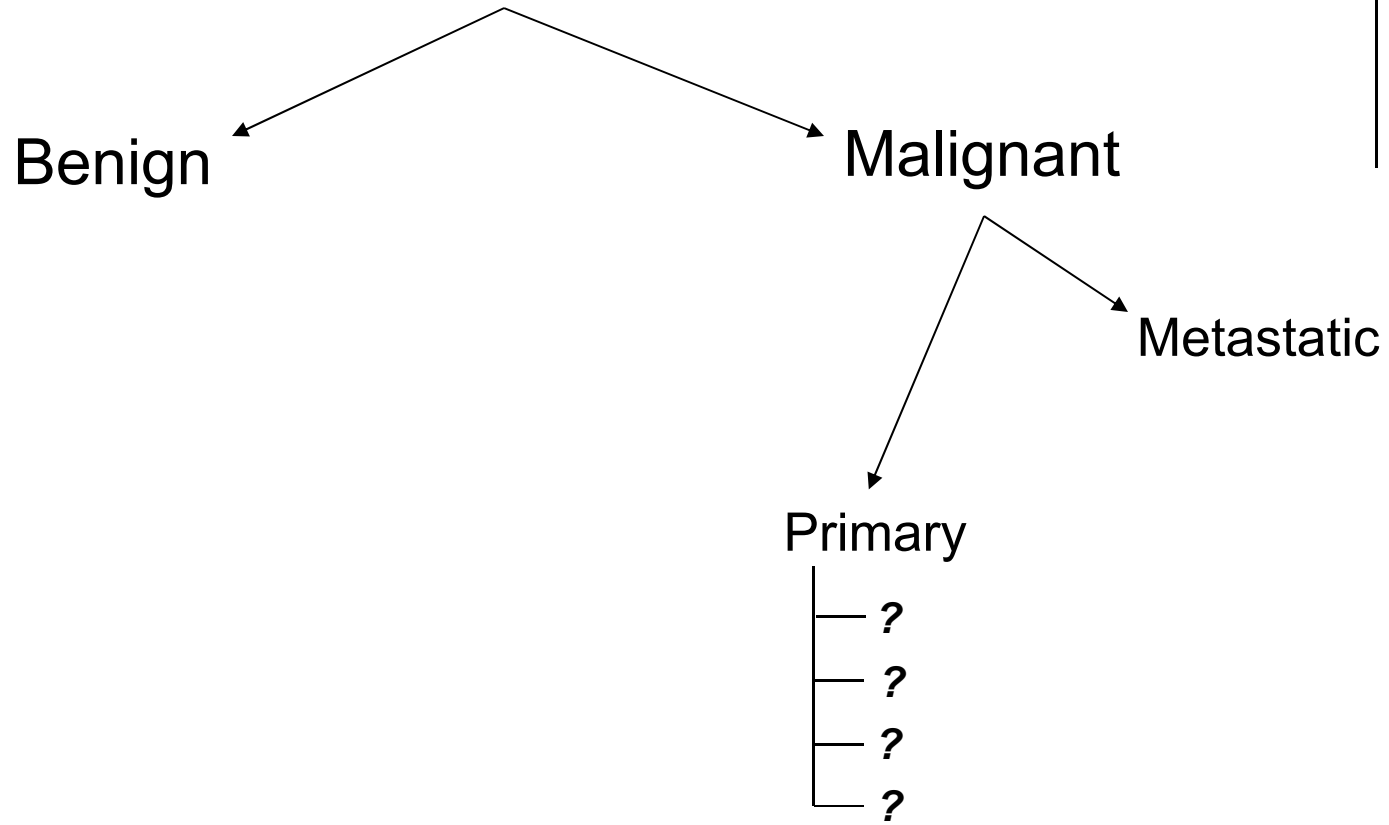
Pediatric Orbital Tumors



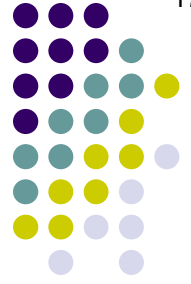
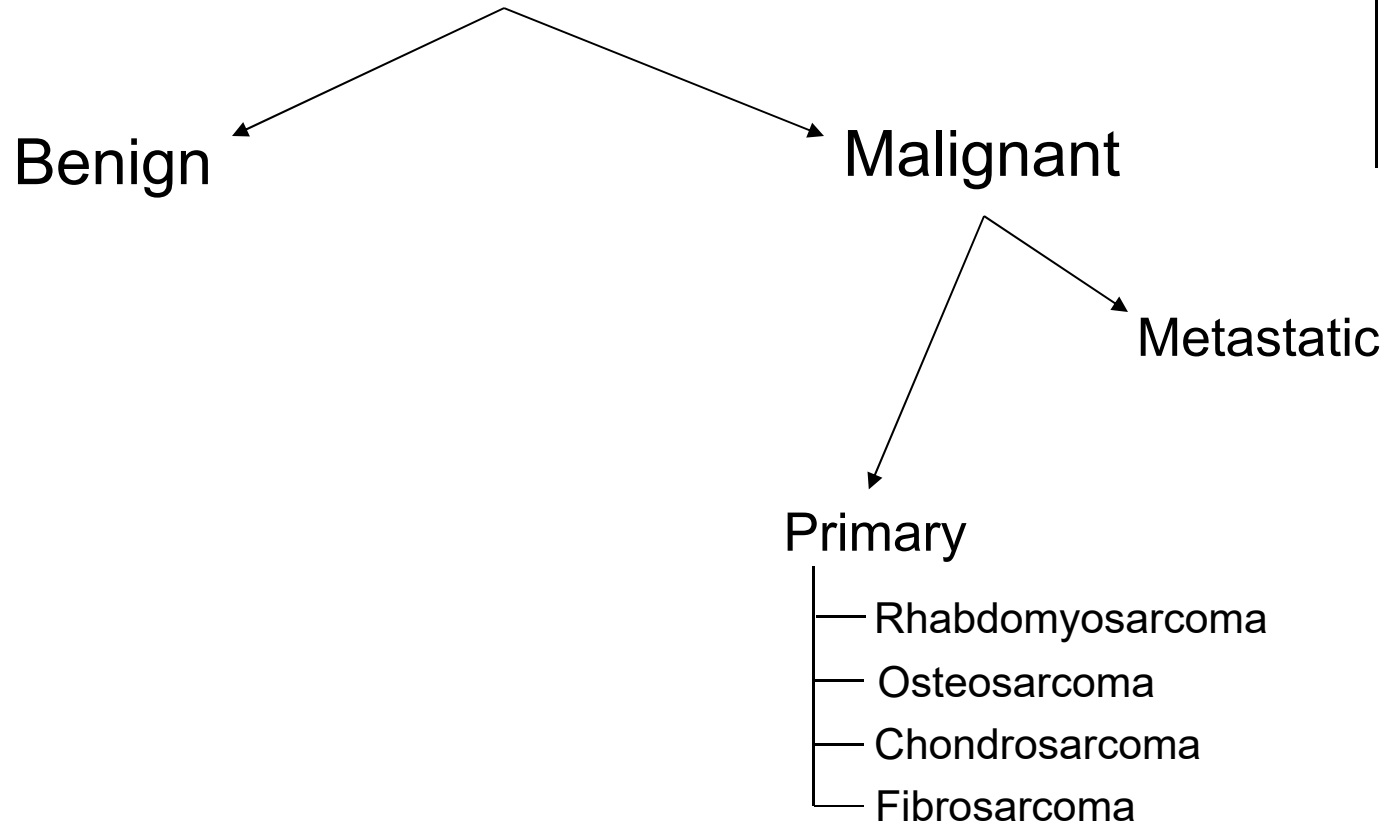
Pediatric Orbital Tumors

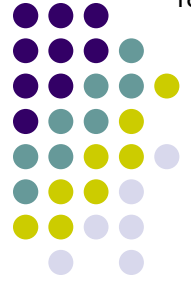


Pediatric Orbital Tumors

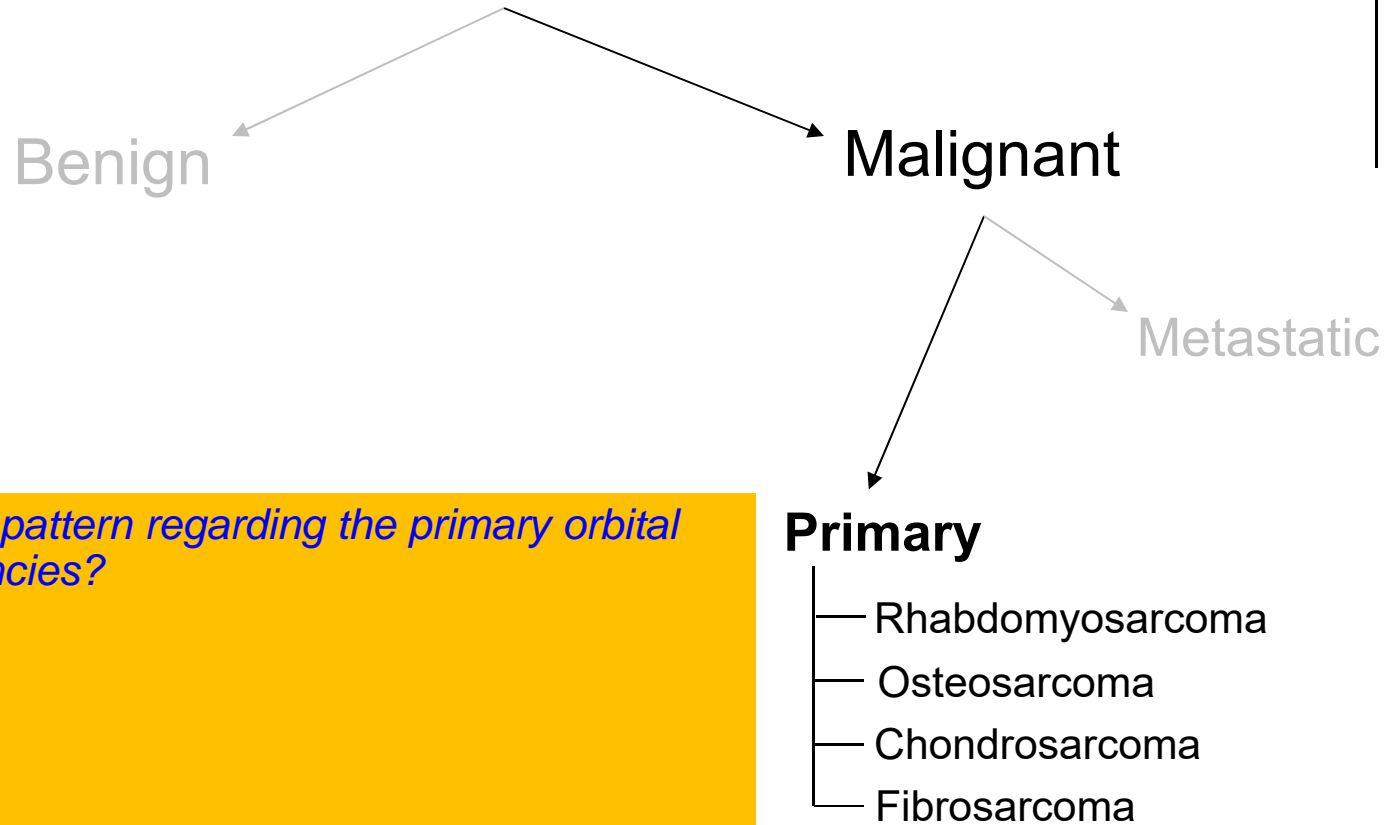


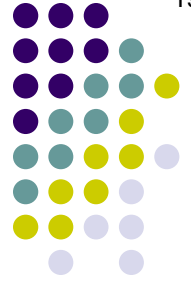
Pediatric Orbital Tumors



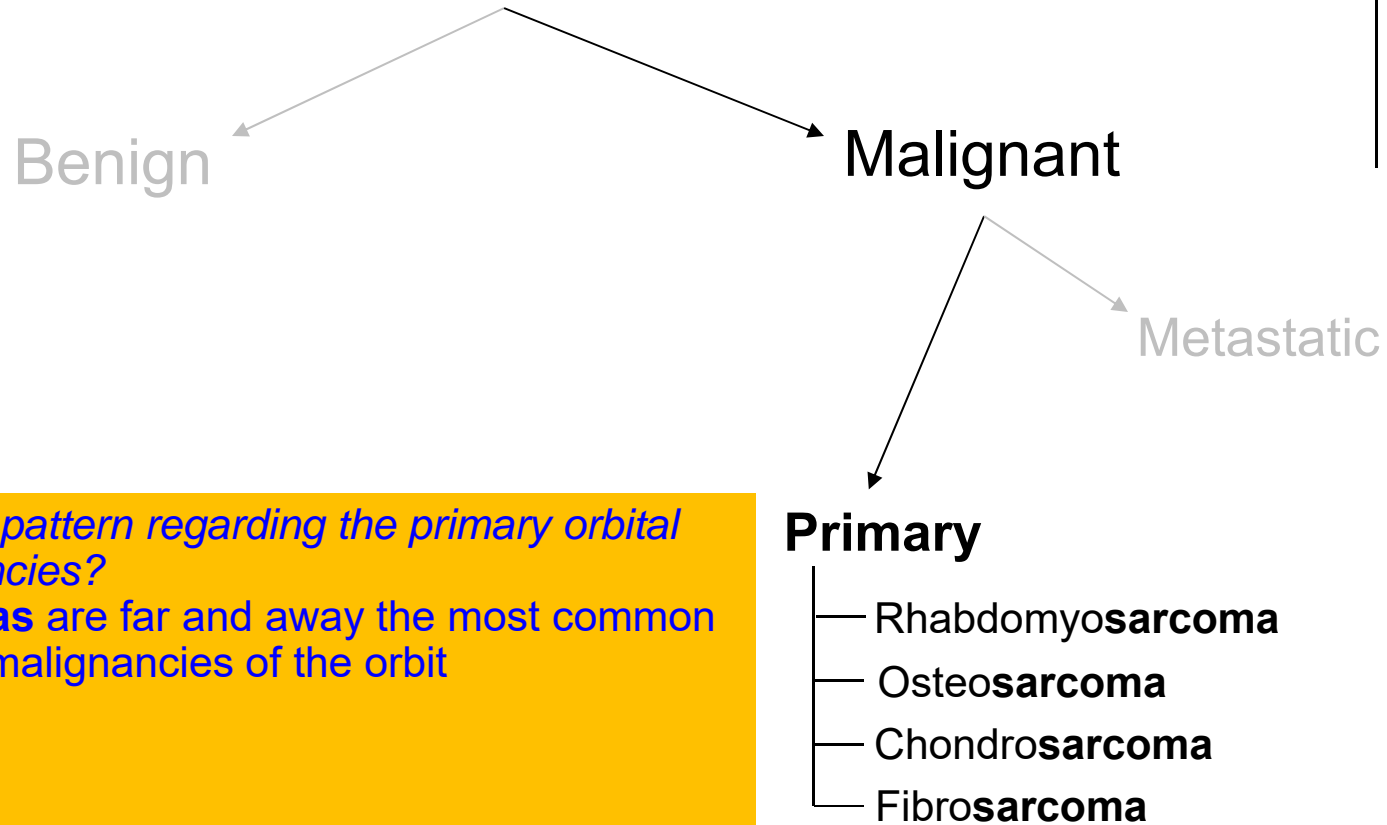


Pediatric Orbital Tumors



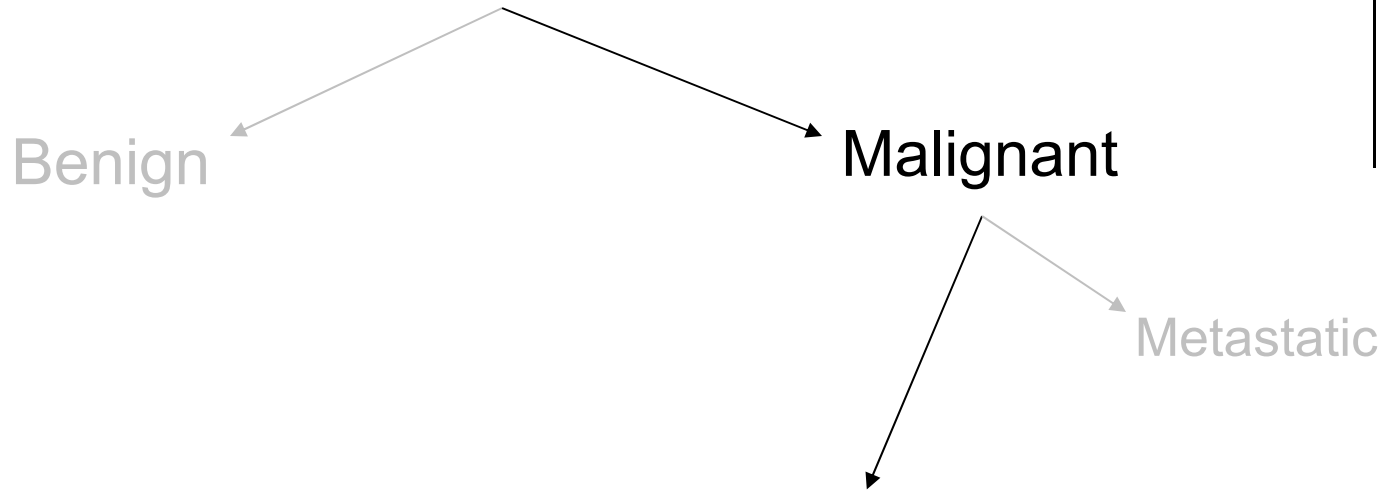


Pediatric Orbital Tumors





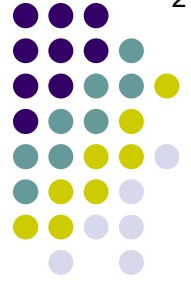
Pediatric Orbital Tumors



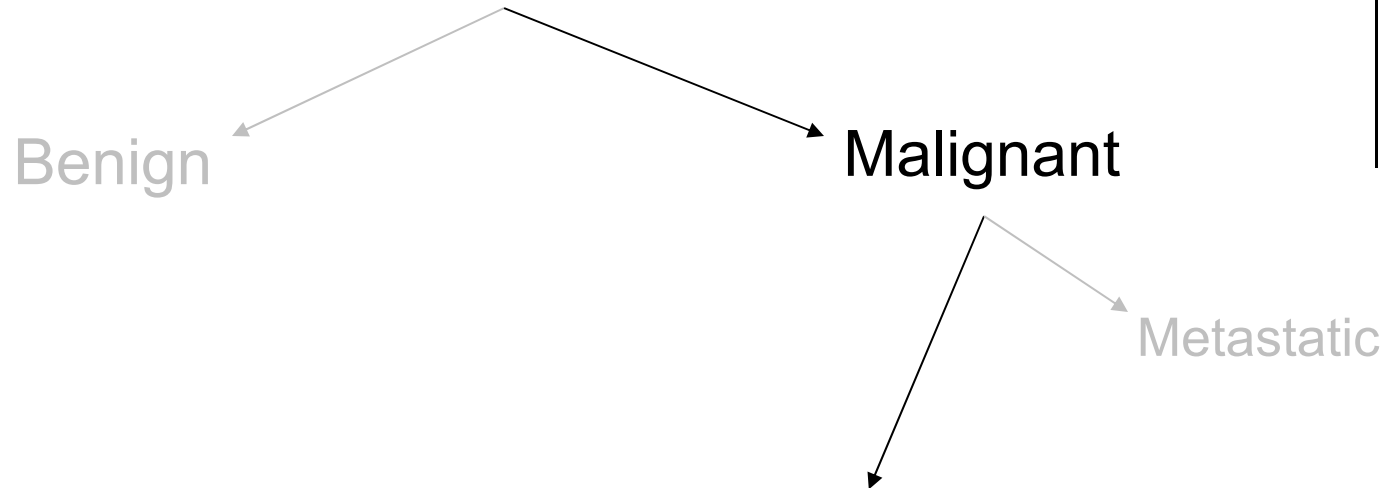
Notice a pattern regarding the primary orbital malignancies?
Sarcomas are far and away the most common primary malignancies of the orbit
What about orbital carcinomas?

Primary

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
- Carcinoma?



Pediatric Orbital Tumors



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

Primary

- Rhabdomyosarcoma
- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma
- Carcinoma? Nah



Pediatric Orbital Tumors

Benign

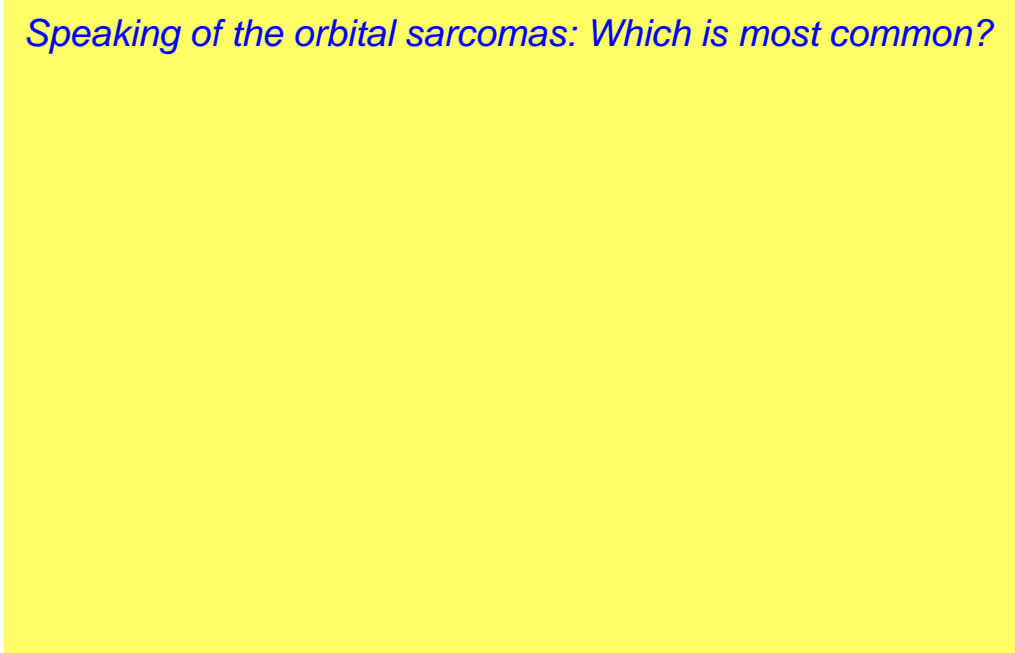
Malignant

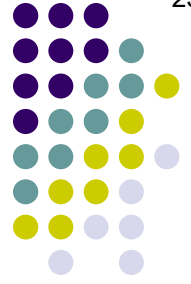
Metastatic

Primary

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- Osteosarcoma
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Speaking of the orbital sarcomas: Which is most common?





Pediatric Orbital Tumors

Benign

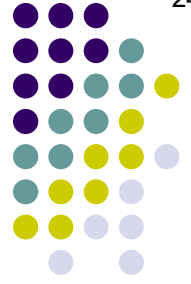
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Rhabdomyosarcoma



Pediatric Orbital Tumors

Benign

Malignant

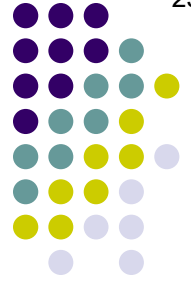
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Primary

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

How much more common is it?



Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

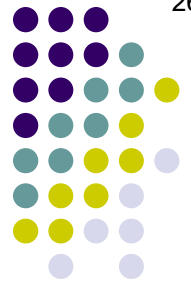
Osteosarcoma

Chondrosarcoma

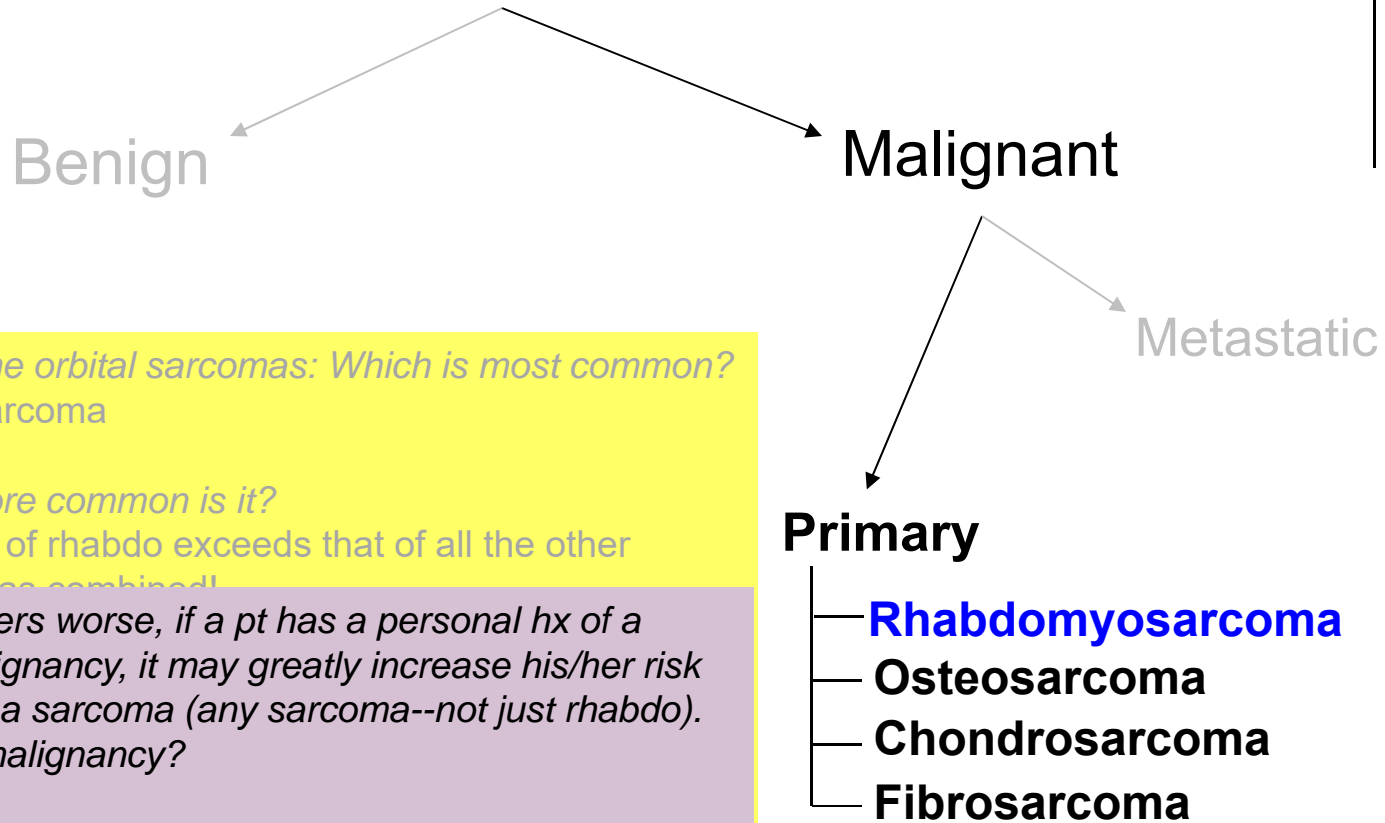
Fibrosarcoma

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!



Pediatric Orbital Tumors



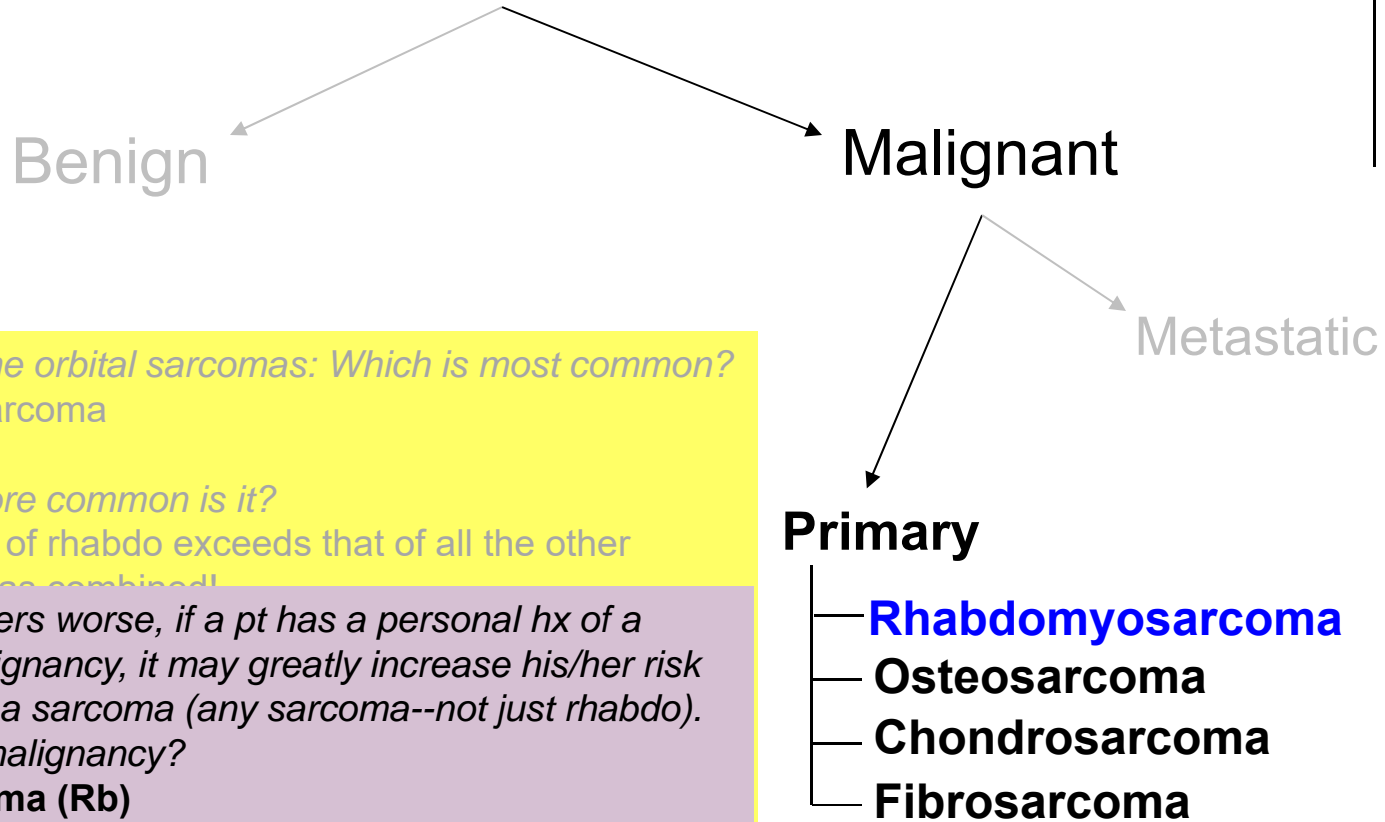
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What is that malignancy?*



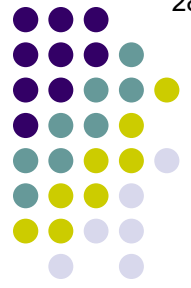
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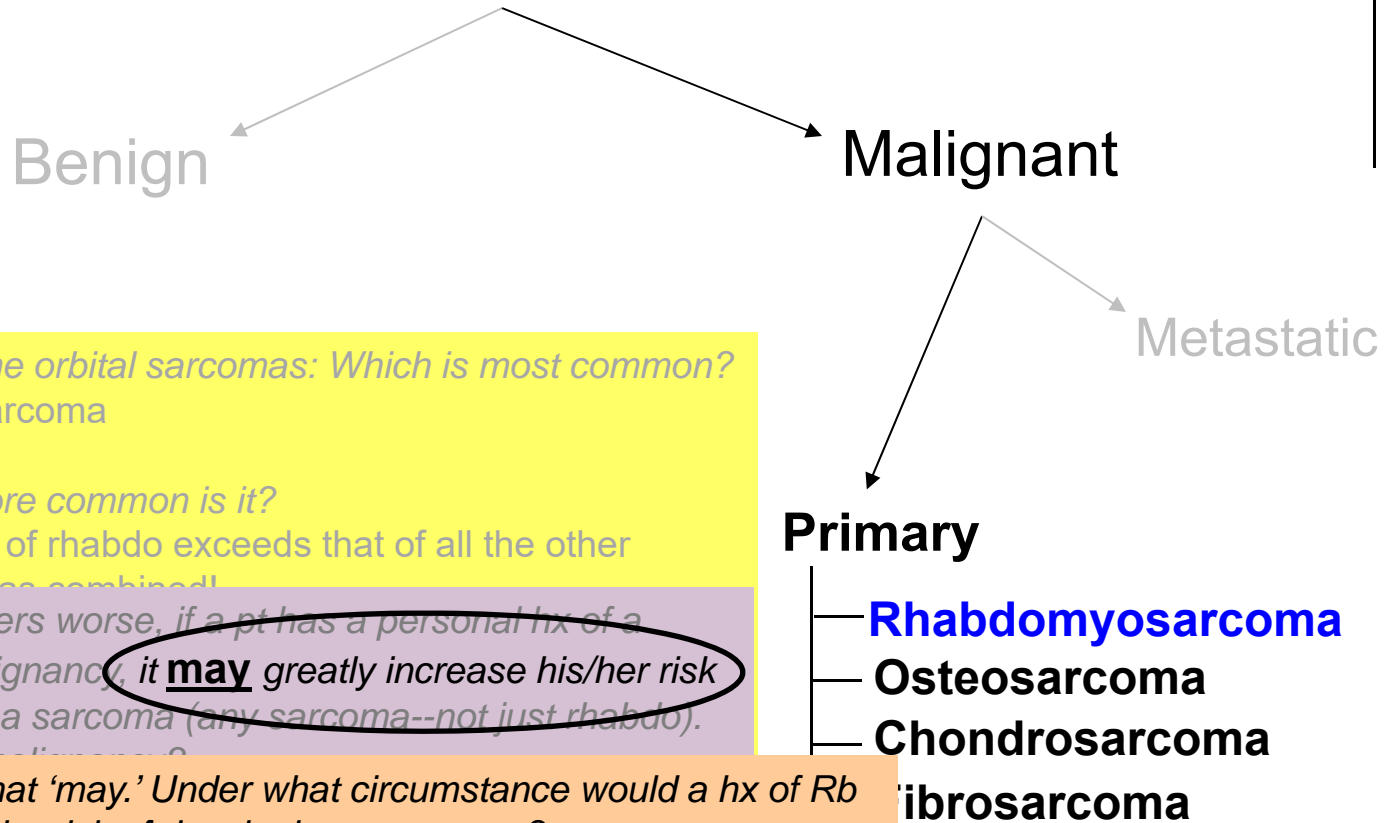
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Retinoblastoma (Rb)



Pediatric Orbital Tumors



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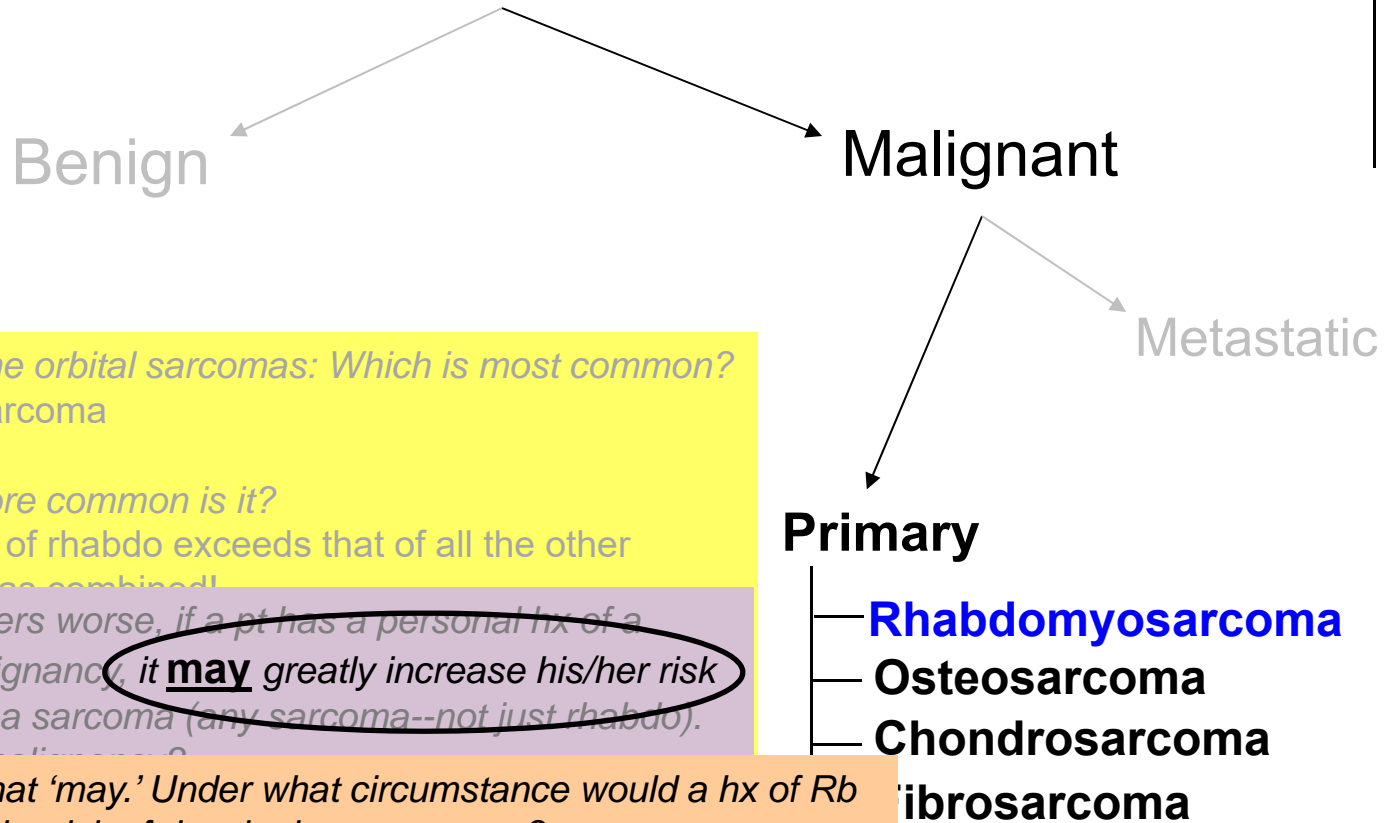
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Re*

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

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Pediatric Orbital Tumors

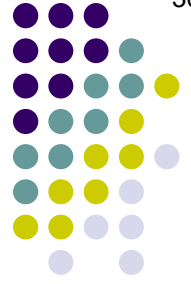


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 When the Rb mutation is heritable (ie, is in the germline)

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Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

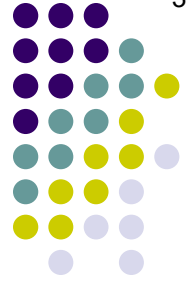
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'When the Rb is heritable'--that means the mutation was inherited, right?



Pediatric Orbital Tumors

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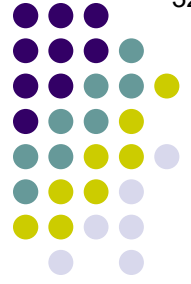
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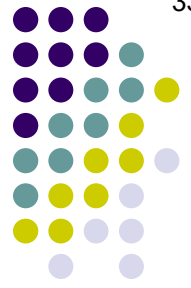
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Unpa
Re incre
When

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

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When is this risk higher?
Re: Unpa
incre
When External-beam radiation therapy

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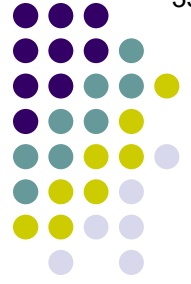
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Pediatric Orbital Tumors

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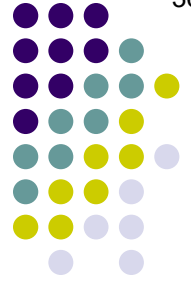
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5 and 7 years



Pediatric Orbital Tumors

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Malignant

Metastatic

Primary

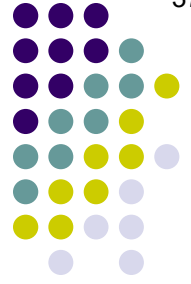
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Between what ages?
5 and 7 years

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

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

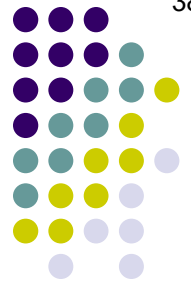
*Speaking of the orbital sarcomas: Which is most common?
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How much more common is it?

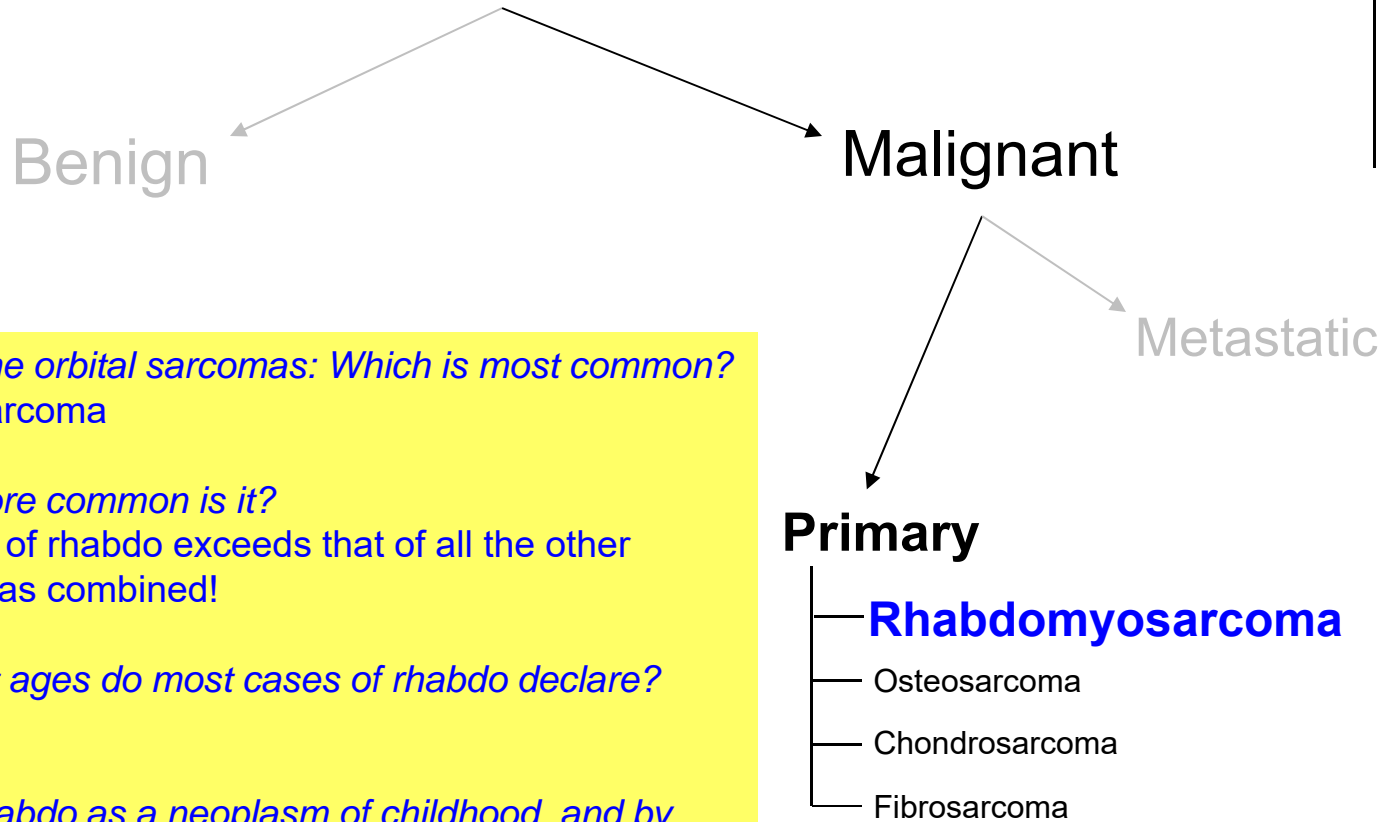
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Pediatric Orbital Tumors



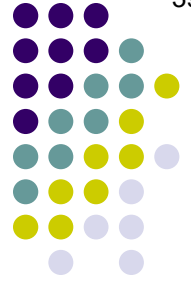
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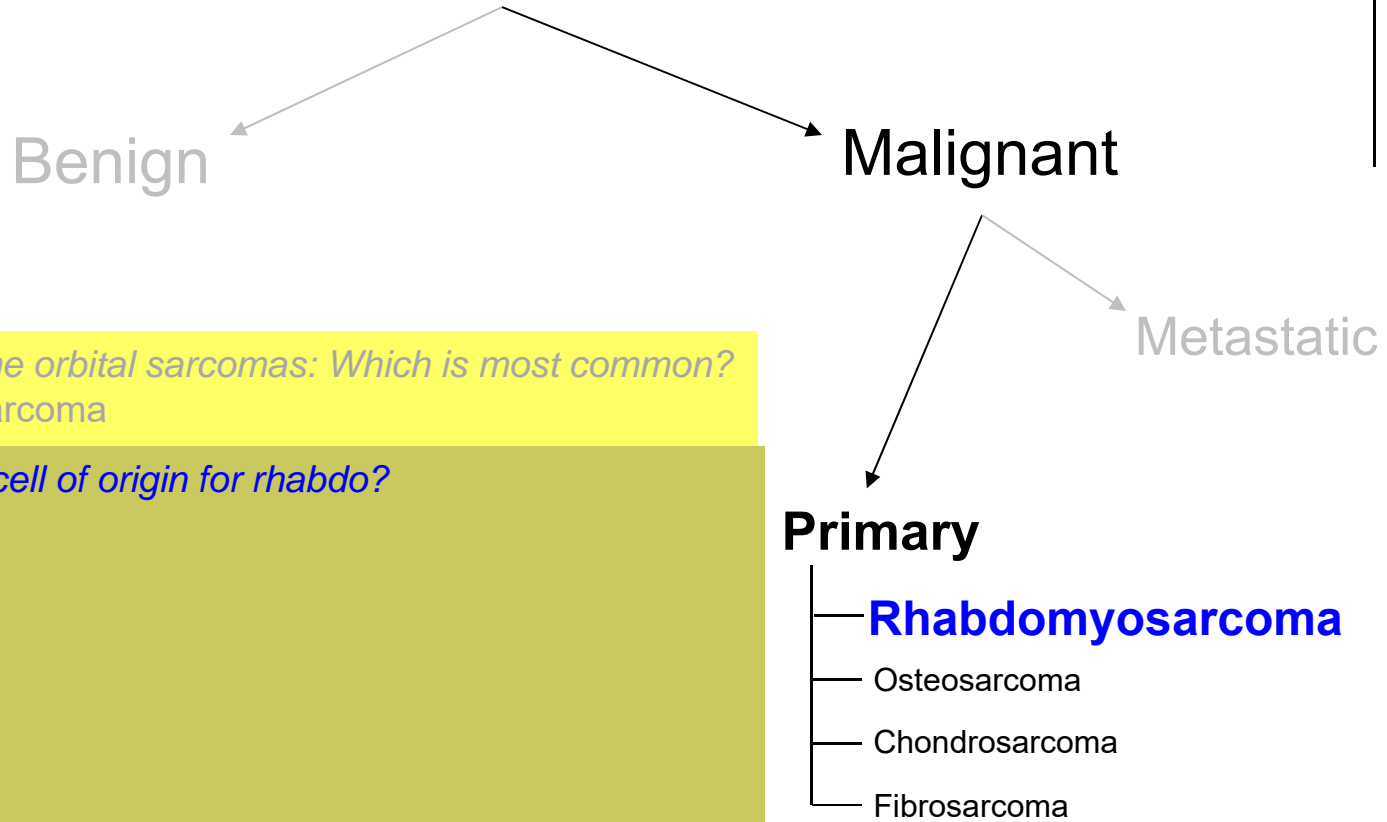
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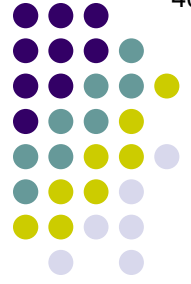
Pediatric Orbital Tumors



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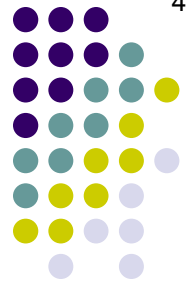
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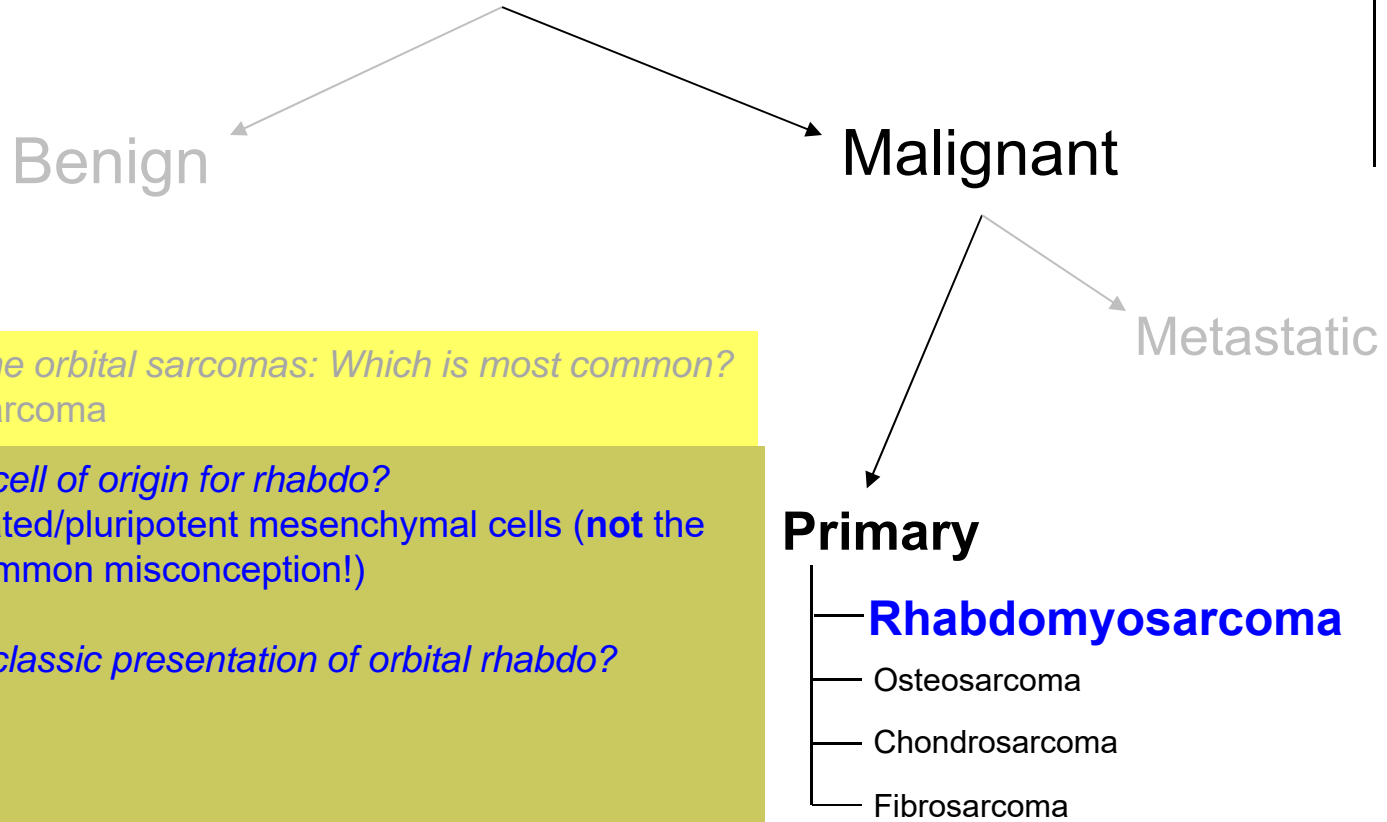
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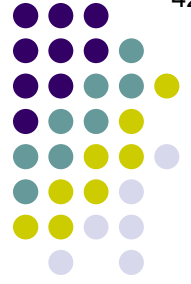


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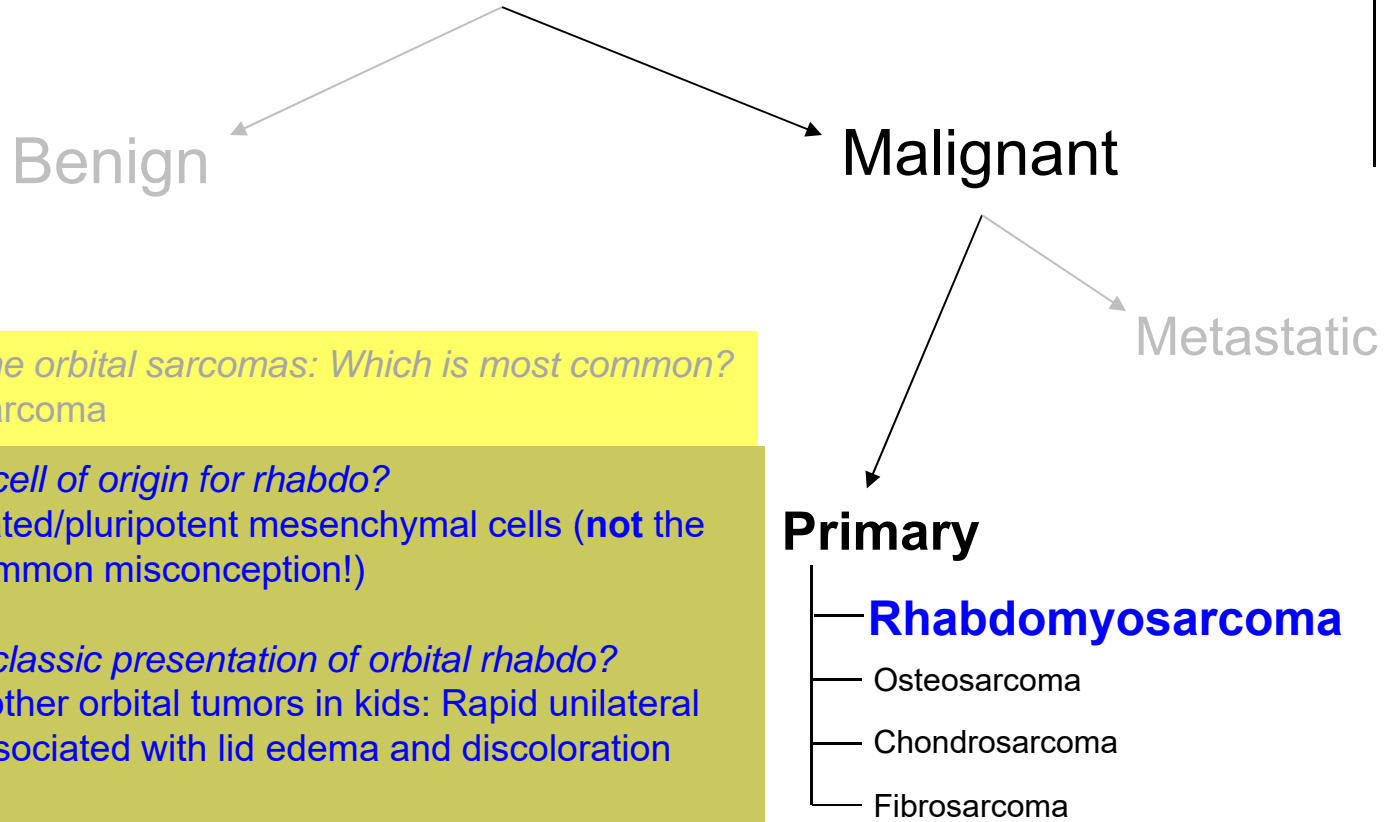
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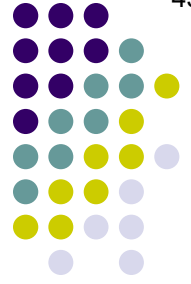


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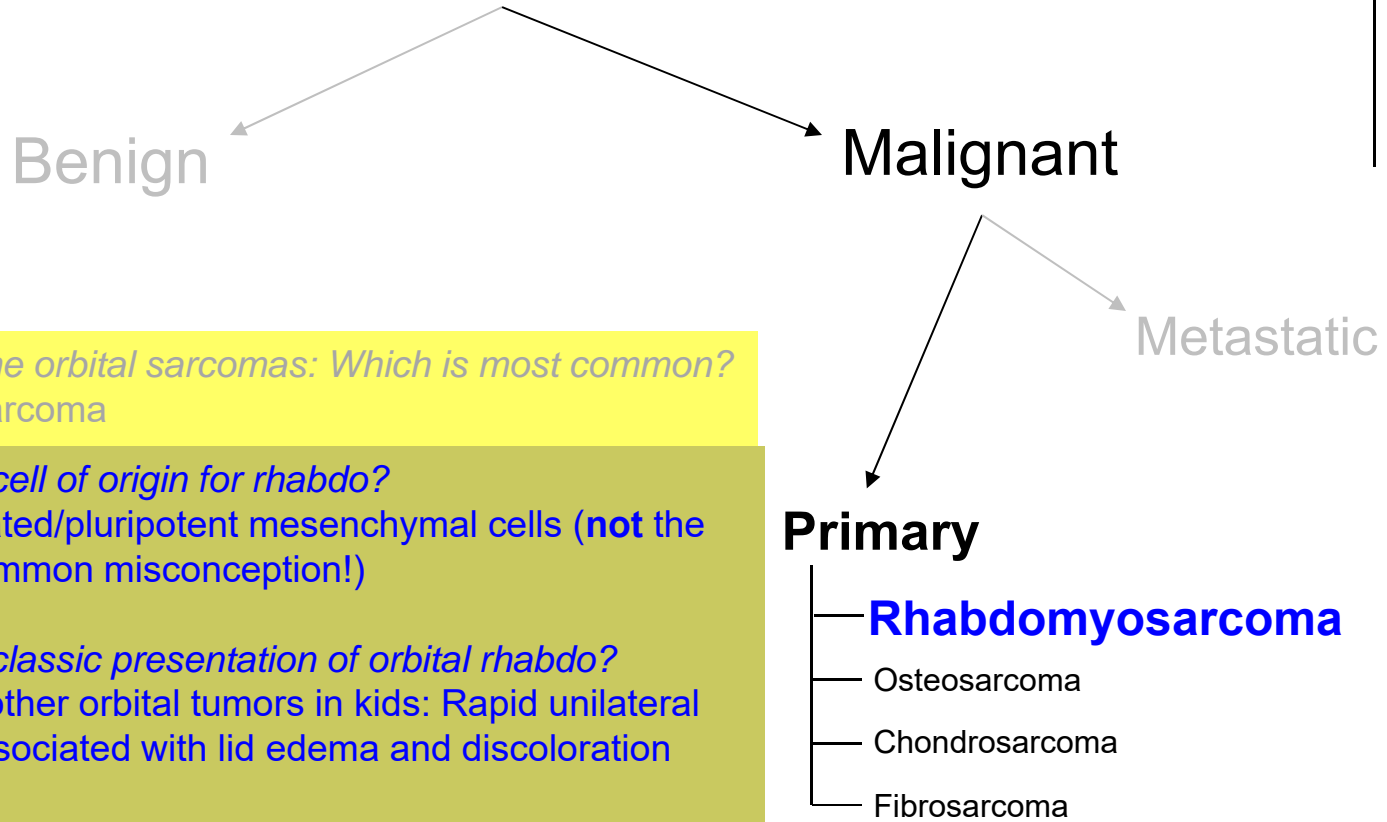
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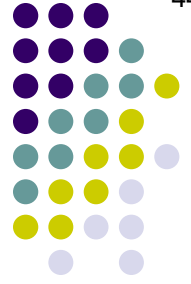
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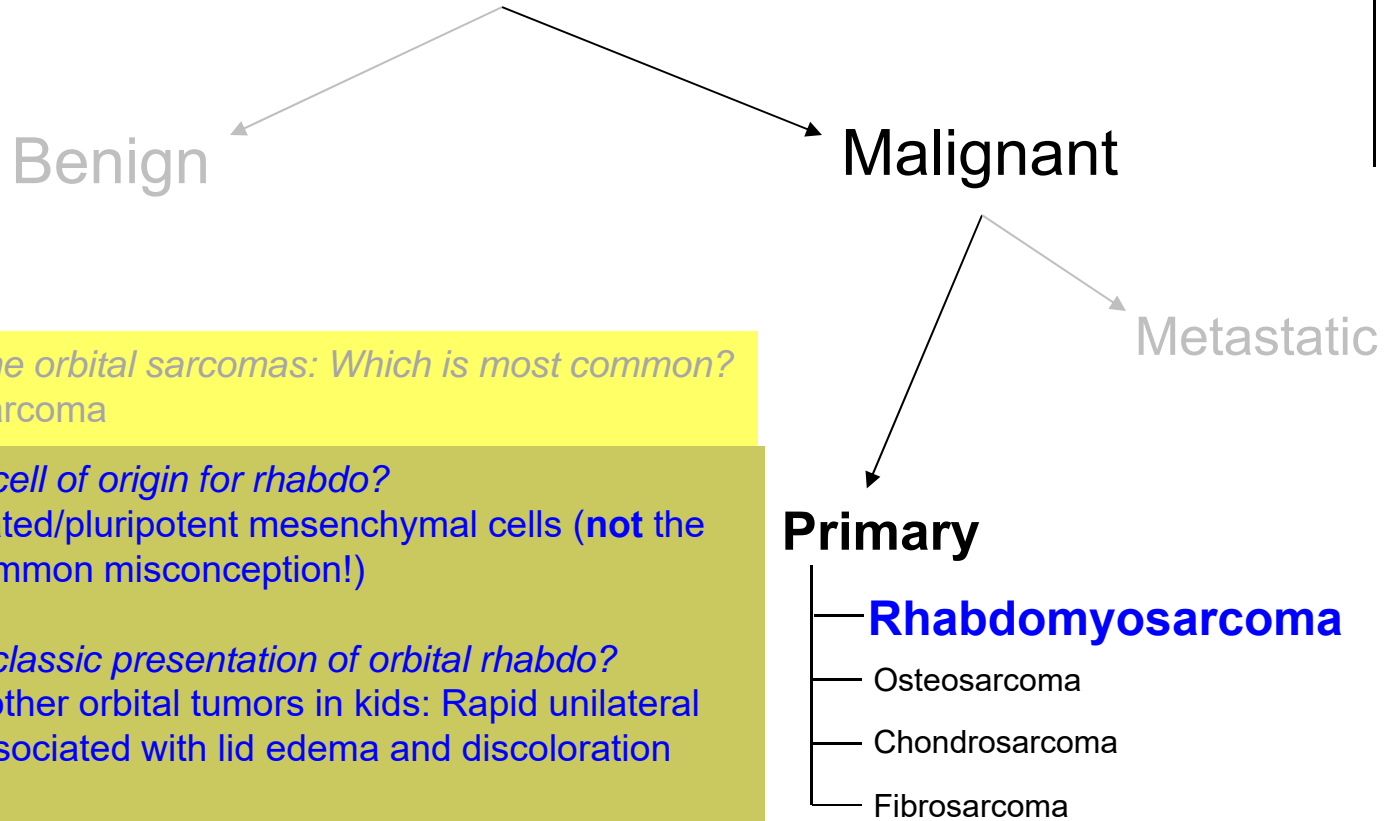
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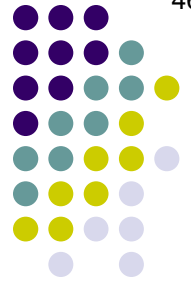
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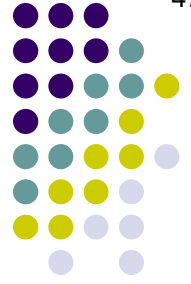
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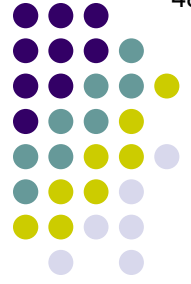
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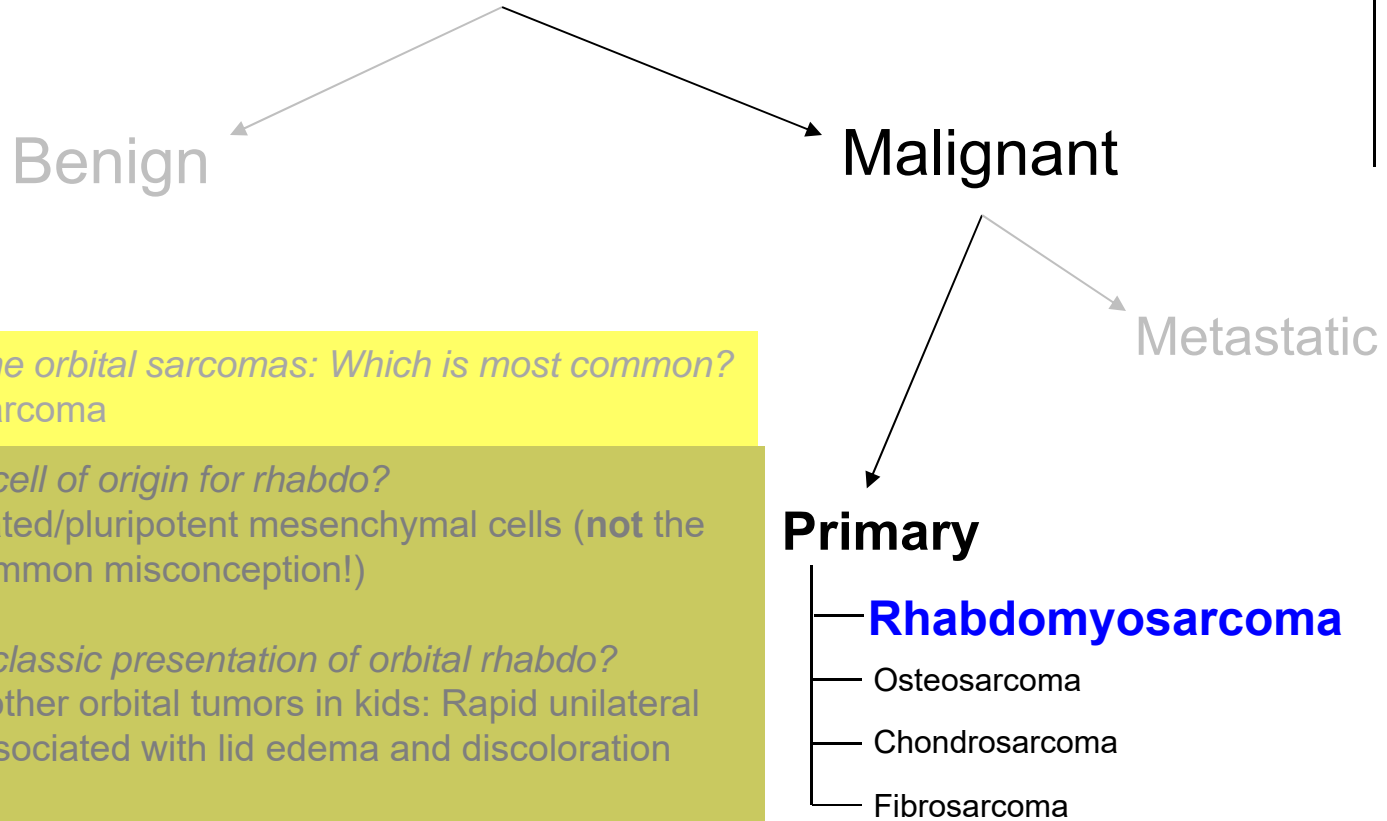
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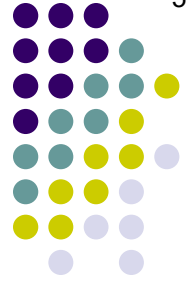
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If not as a primary, then how does botryoid get in the orbit?



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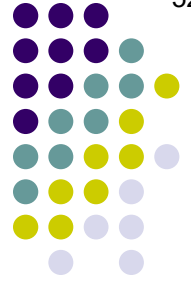
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Via extension from a sinus, or the conj



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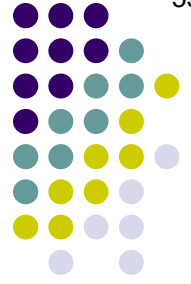
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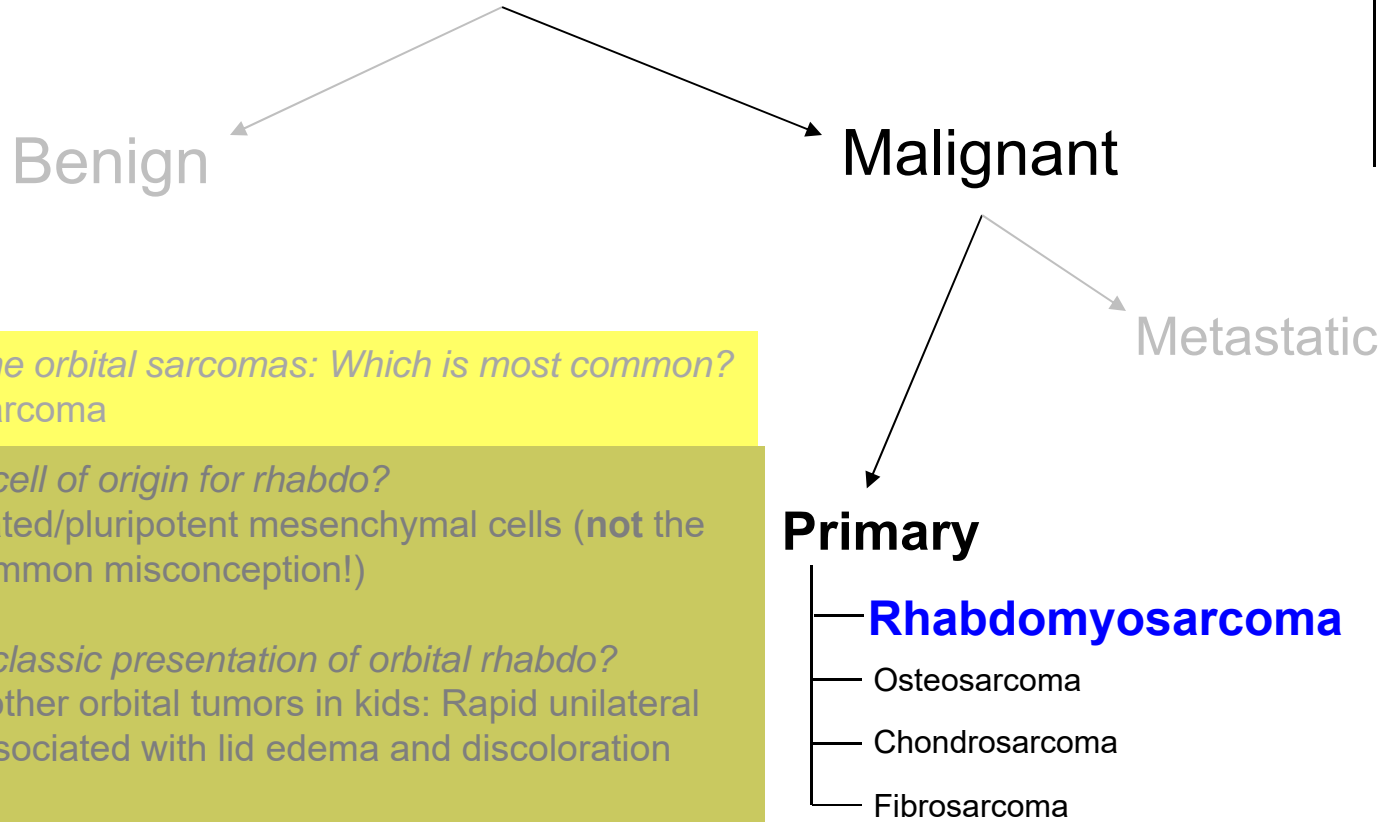
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Pediatric Orbital Tumors



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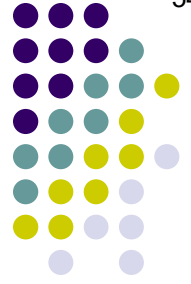
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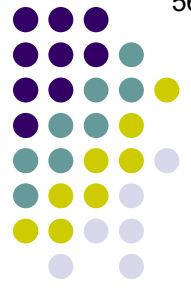
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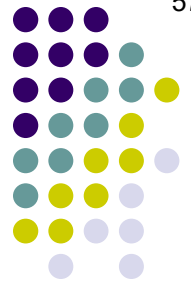
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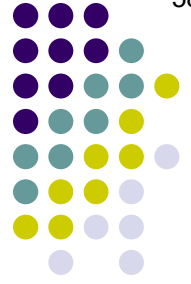
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Per the Peds book, 75%



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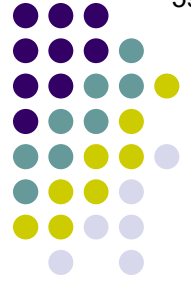
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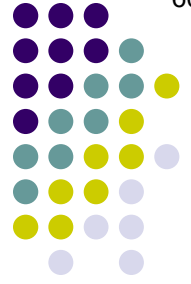
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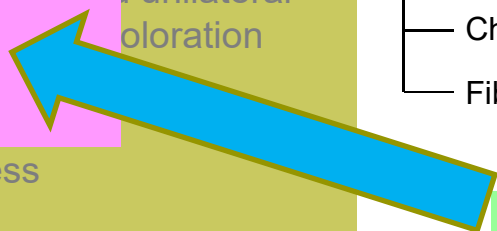
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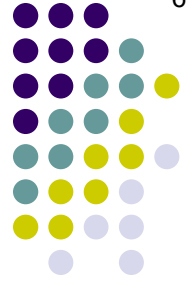
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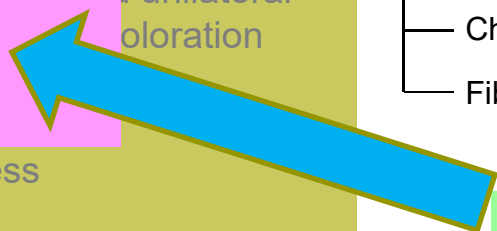
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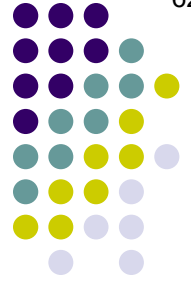
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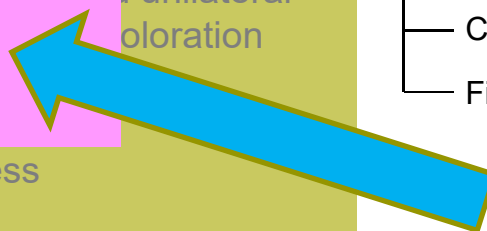
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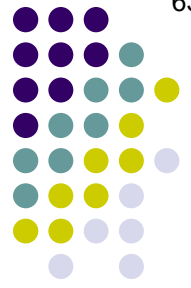
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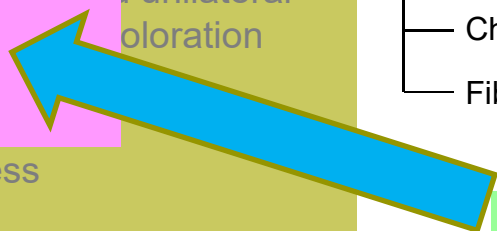
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Neither book addresses this



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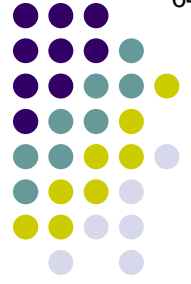
What are the four histological subtypes of rhabdo?

- Embryonal
- Alveolar
- Pleomorphic
- Botryoid

Which subtype carries the poorest prognosis?
Alveolar

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

What is the 10-year survival rate?
Per the Orbit book, only 10%!



Pediatric Orbital Tumors

Benign

Malignant

Metastatic

Primary

Rhabdomyosarcoma

Osteosarcoma

Chondrosarcoma

Fibrosarcoma

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

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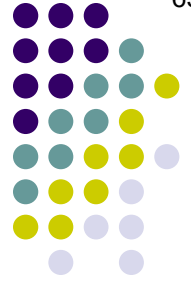
--Embryonal

--Alveolar

--**Pleomorphic**

--Botryoid

Under what circumstances is the pleomorphic subtype encountered?



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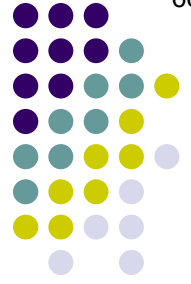
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--Alveolar

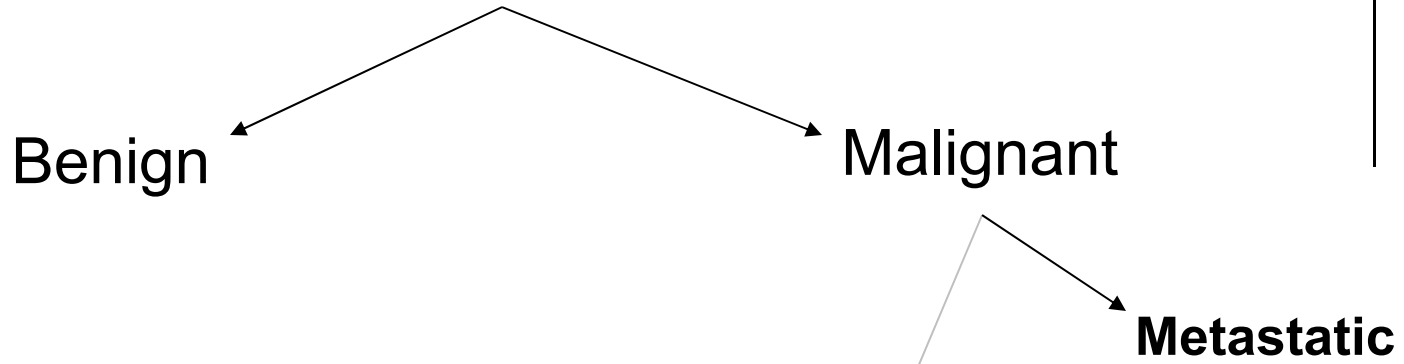
--Pleomorphic

--Botryoid

Under what circumstances is the pleomorphic subtype encountered?
When the pt is an adult (it essentially never occurs in kids)



Pediatric Orbital Tumors

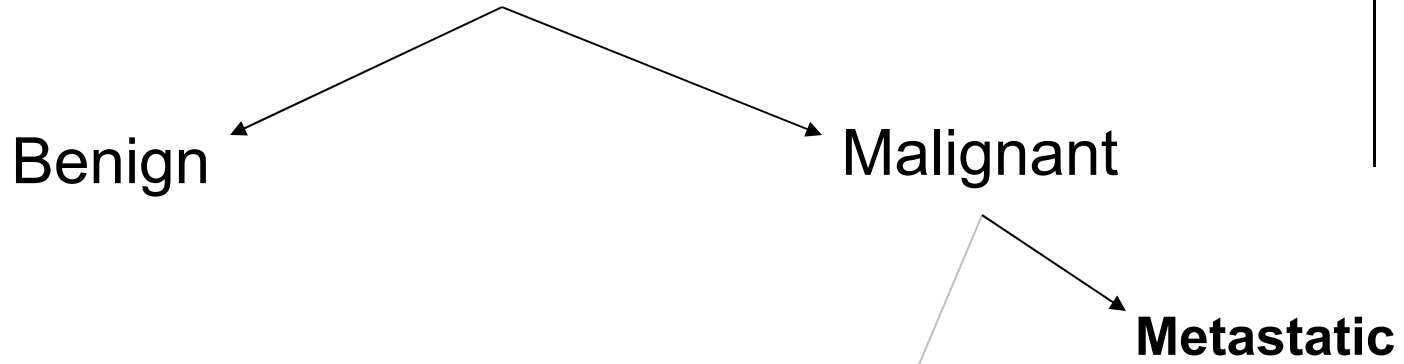


Regarding metastatic dz to the eye or orbit: In what key manner does the pediatric population differ from the adult?
In adults,
In children,

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma



Pediatric Orbital Tumors

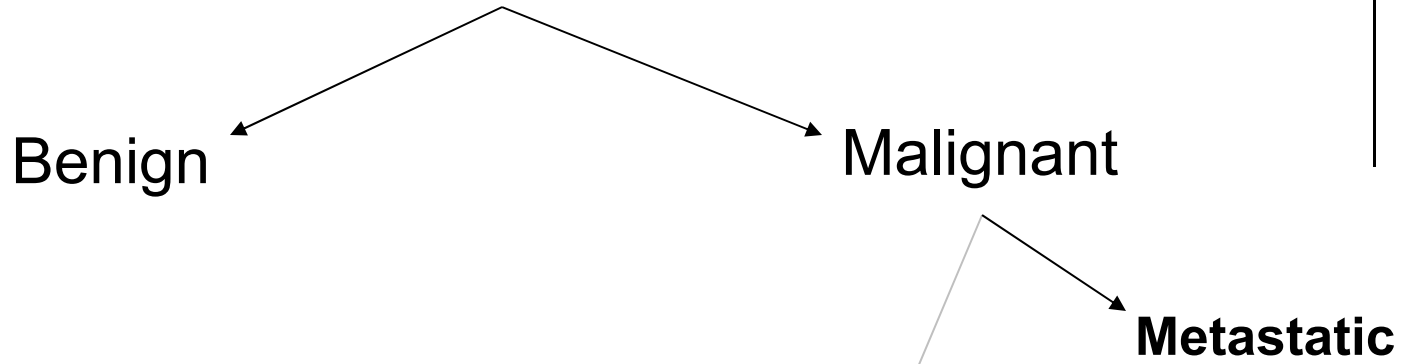


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In adults, mets tend to go to the **eye**; but...
In children,*

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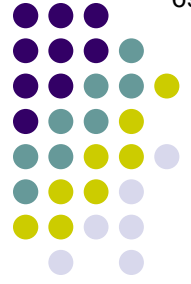


Pediatric Orbital Tumors

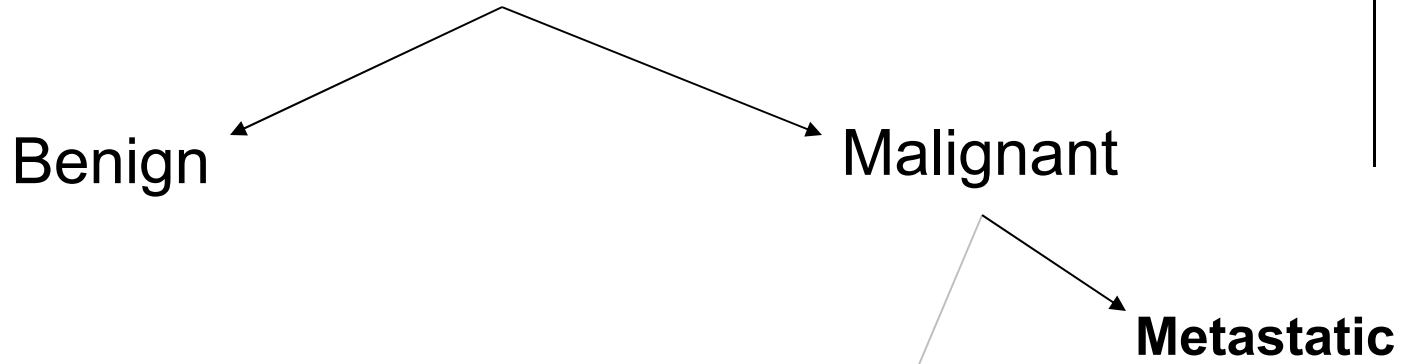


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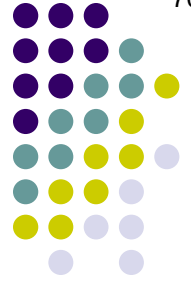


Pediatric Orbital Tumors

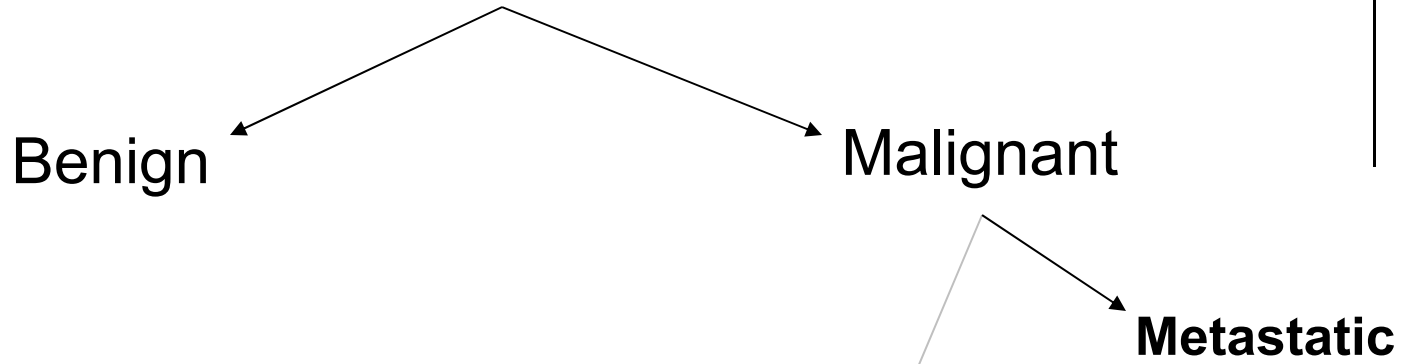


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In children, they go to the **orbit***

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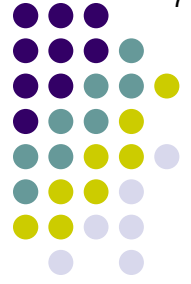
Pediatric Orbital Tumors



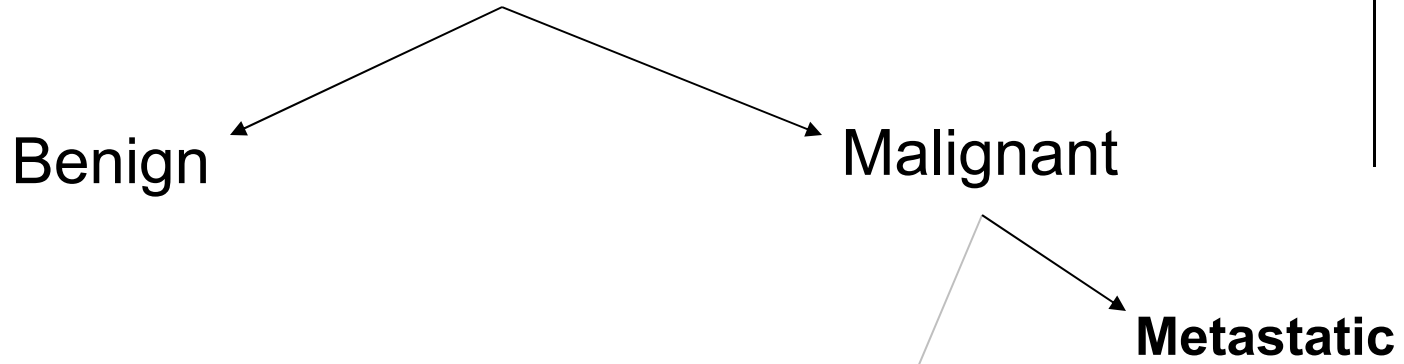
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Where specifically in the adult eye do mets tend to go?

- Chondrosarcoma
- Fibrosarcoma



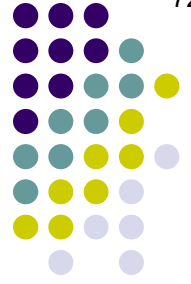
Pediatric Orbital Tumors



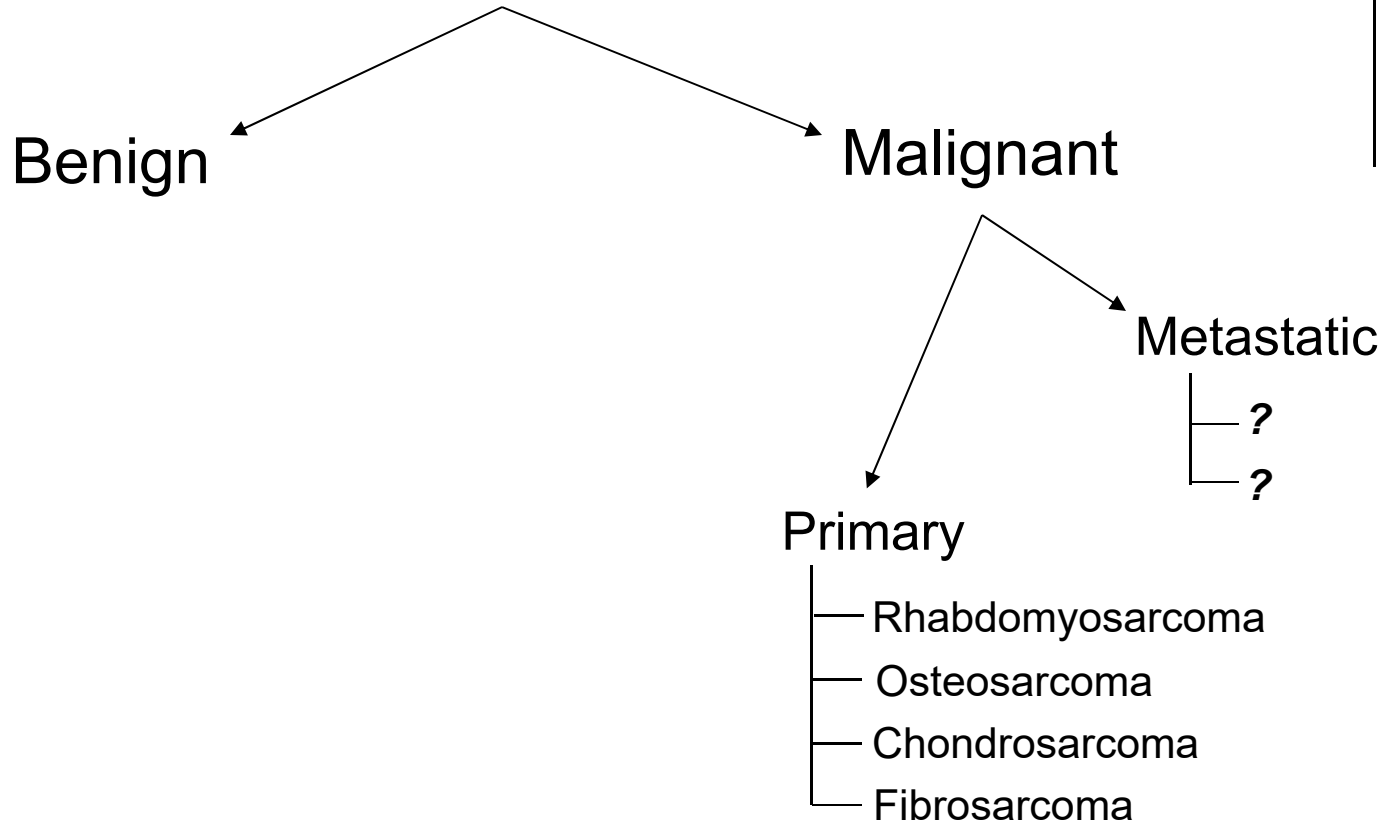
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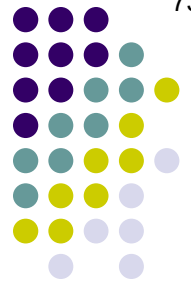
*Where specifically in the adult eye do mets tend to go?
 The choroid*

- Chondrosarcoma
- Fibrosarcoma

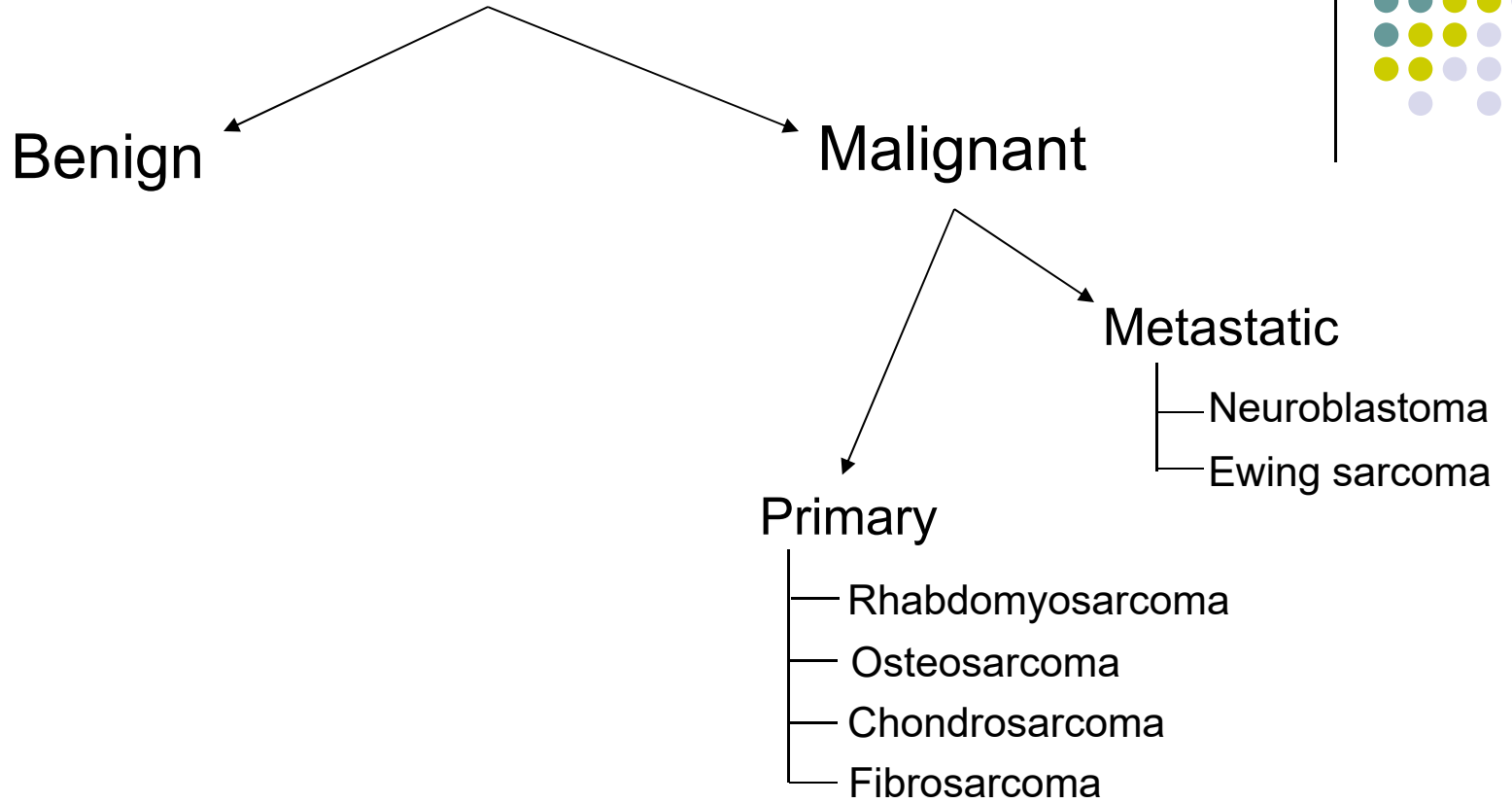


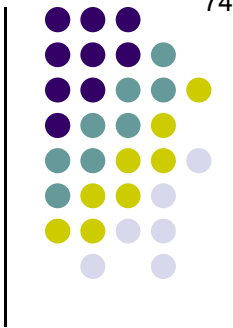
Pediatric Orbital Tumors





Pediatric Orbital Tumors





Pediatric Orbital Tumors

Benign

Malignant

Briefly, what is neuroblastoma (Nb)?

Metastatic

Neuroblastoma

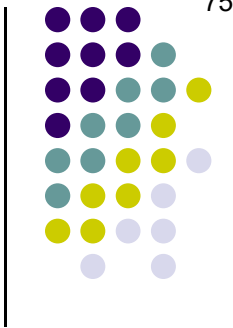
Ewing sarcoma

Myosarcoma

Liposarcoma

Rhabdomyosarcoma

Fibrosarcoma



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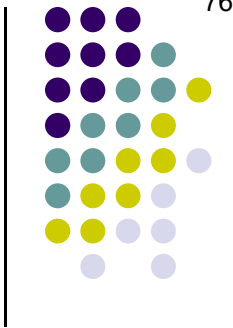
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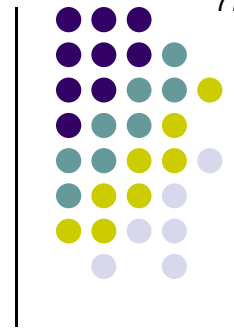
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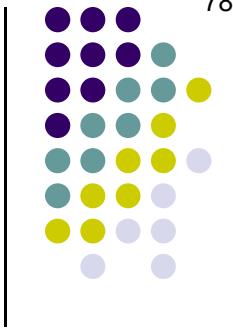
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Metastatic

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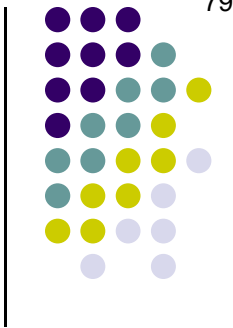
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It is the most common cause of extracranial solid cancer in childhood

Metastatic

Neuroblastoma

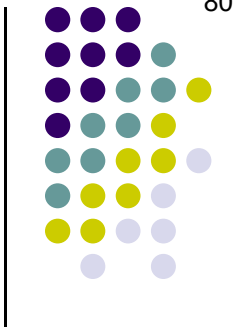
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Ewing sarcoma

Myosarcoma

Liposarcoma

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It is #1

Metastatic

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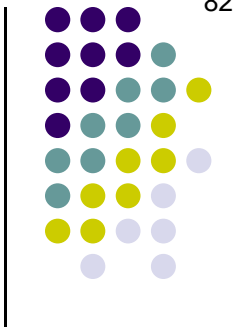
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What proportion of Nb cases metastasize to the orbit?

Where does it rank as a cause of orbital mets in childhood?

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Metastatic

Neuroblastoma

Ewing sarcoma

Myosarcoma

Liposarcoma

Rhabdomyosarcoma

Fibrosarcoma



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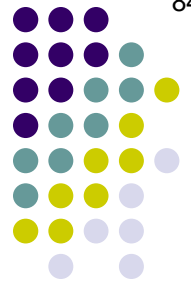
What proportion of Nb cases metastasize to the orbit?
 In About 20%

Where does it rank as a cause of orbital mets in childhood?
It is #1

Metastatic



...yosarcoma
 ...coma
 ...arcoma
 Fibrosarcoma



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Neuroblastoma

Ewing sarcoma

lyosarcoma

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arcoma

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What proportion of all orbital mets in children are due to Nb?

It About 90%!

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Metastatic

Neuroblastoma

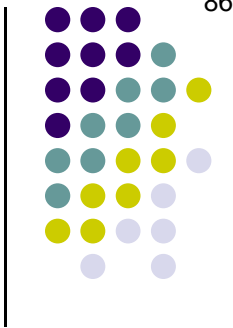
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Where
In the s

What is the average age at diagnosis?

Where
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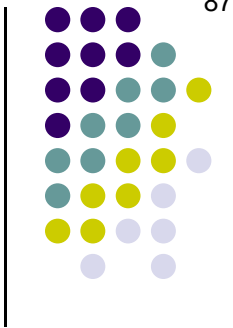
Where
It is #1

Metastatic

Neuroblastoma

Ewing sarcoma

Childhood
rhabdomyosarcoma
osteosarcoma
osteoid osteoma
osteosarcoma
osteosarcoma



Pediatric Orbital Tumors

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Where? In the s
What is the average age at diagnosis?
Two years

Where?
It is the

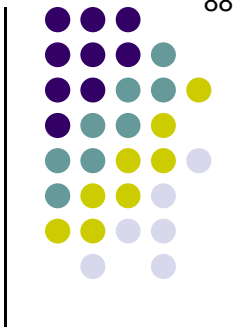
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In the s Two years

Where? What proportion of cases are diagnosed by age 5?

It is the

Where?

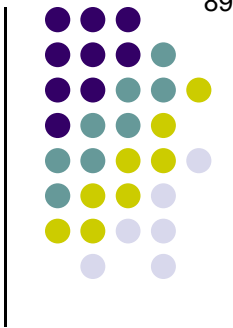
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Neuroblastoma

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Lymphoma
Sarcoma
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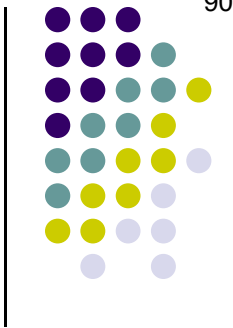
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Fibrosarcoma



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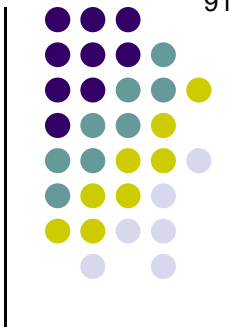
Where? Can it present in infancy, ie, before age 12 months?
It is #1

Metastatic

Neuroblastoma

Ewing sarcoma

Childhood rhabdomyosarcoma
Lymphoma
Sarcoma
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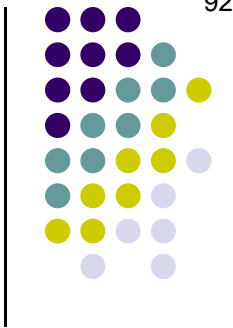
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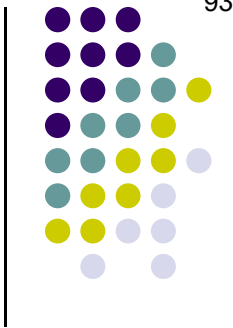
Can it be congenital, ie, present at birth?

Metastatic

Neuroblastoma

Ewing sarcoma

Childhood rhabdomyosarcoma
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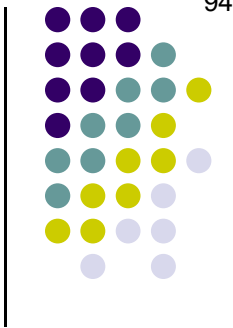
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 Yes

Can it be congenital, ie, present at birth?
 Yes

Metastatic



Childhood rhabdomyosarcoma
 Embryonal rhabdomyosarcoma
 Alveolar rhabdomyosarcoma
 Pleomorphic rhabdomyosarcoma



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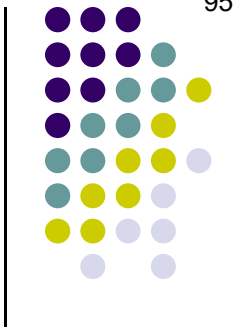
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Metastatic

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d yosarcoma
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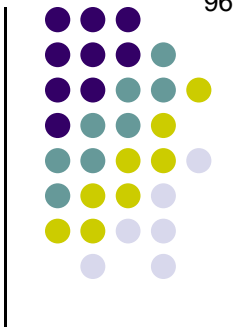
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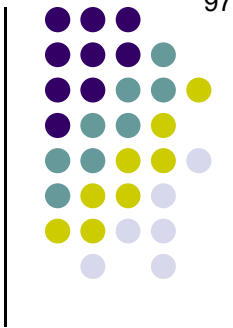
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lyosarcoma
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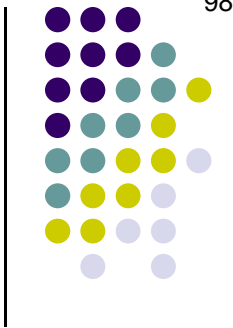
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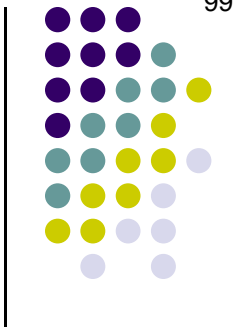
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lyosarcoma
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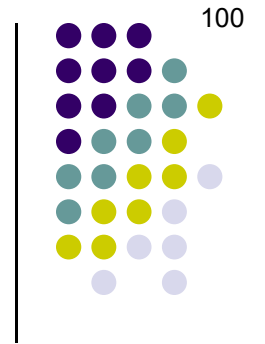
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'Raccoon eyes'

Metastatic



...yosarcoma
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Lid ecchymosis +/- proptosis ("An abrupt ecchymotic proptosis," the *Orbit* book calls it)

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

Metastatic

Neuroblastoma

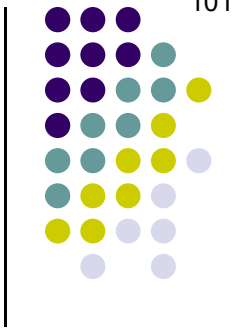
Ewing sarcoma

lyosarcoma

coma

arcoma

coma



Pediatric Orbital Tumors

Benign

Malignant

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? In the s
What is the average age at diagnosis?
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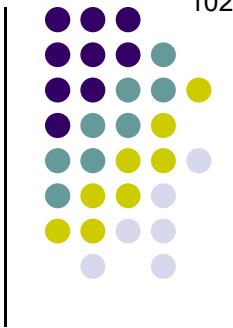
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In two words, what sort of condition is opsoclonus?

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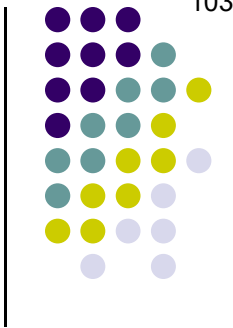
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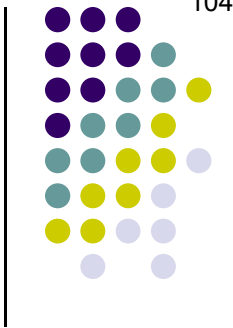
In two words, what sort of condition is opsoclonus?
It is a saccadic intrusion

Metastatic

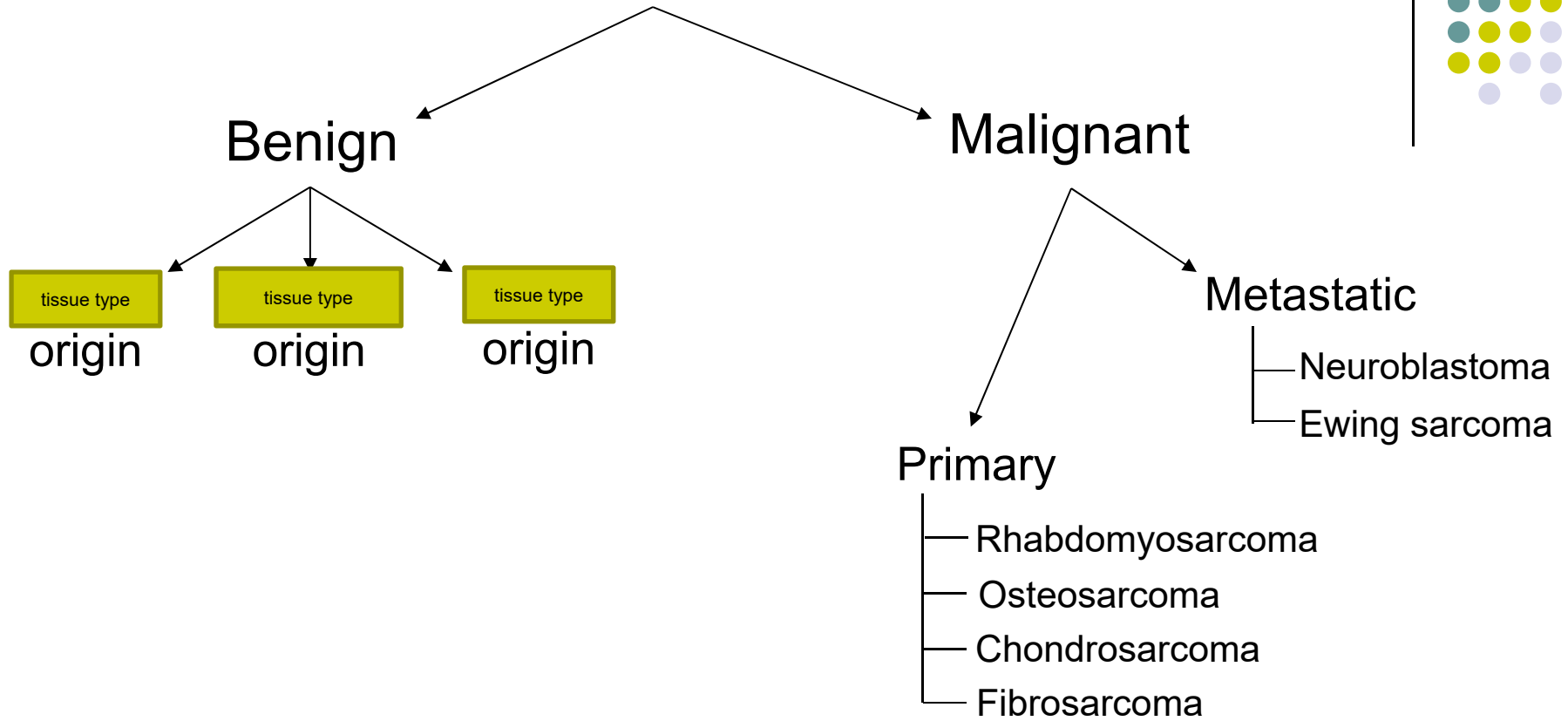
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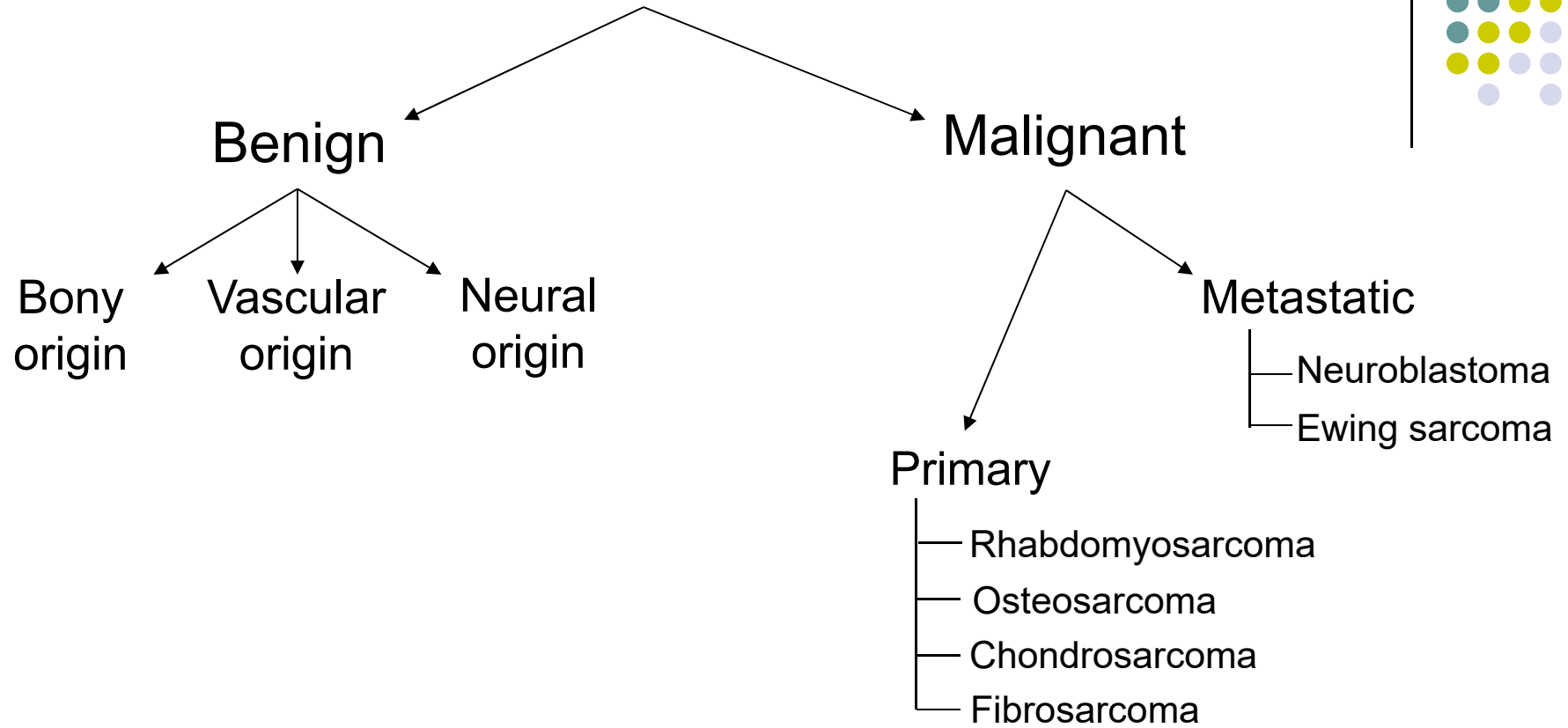


Pediatric Orbital Tumors



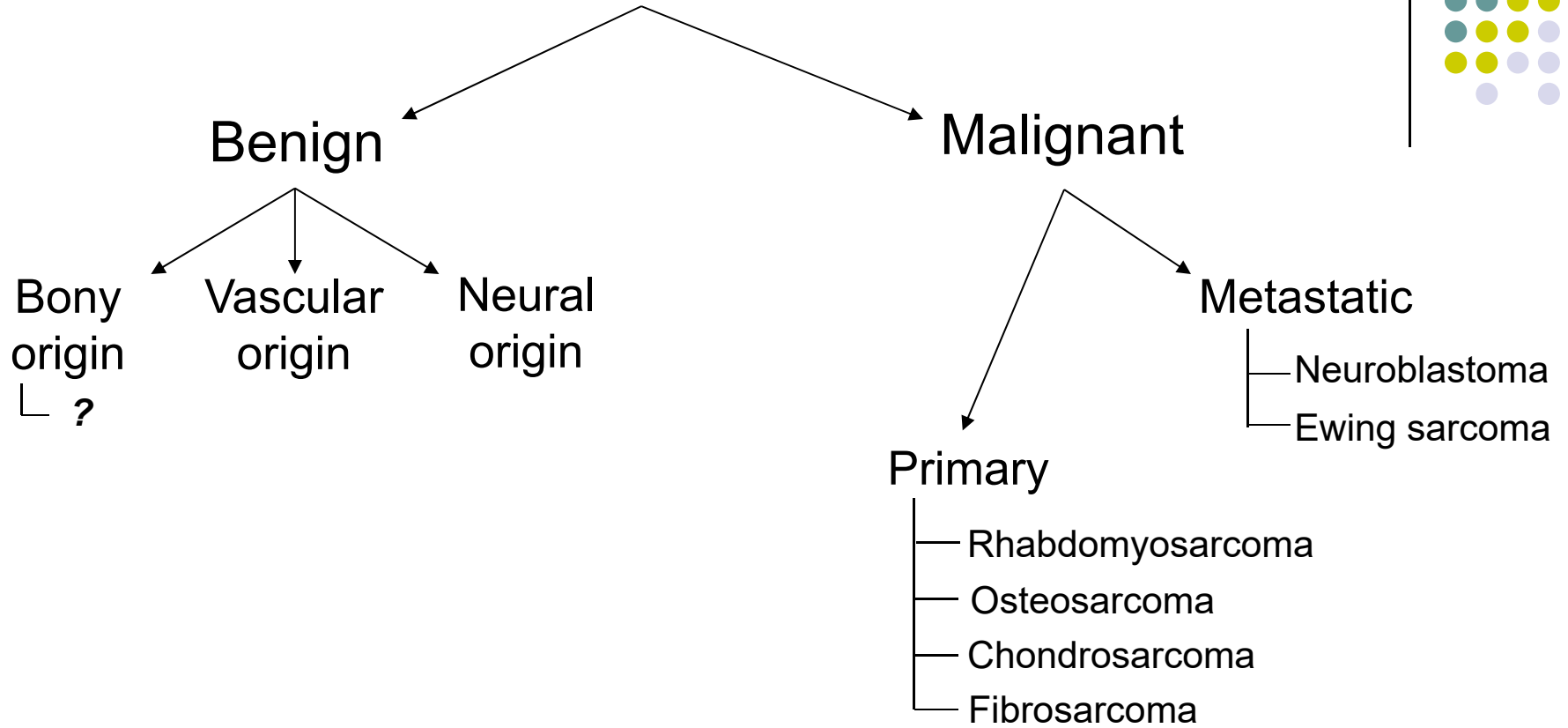


Pediatric Orbital Tumors



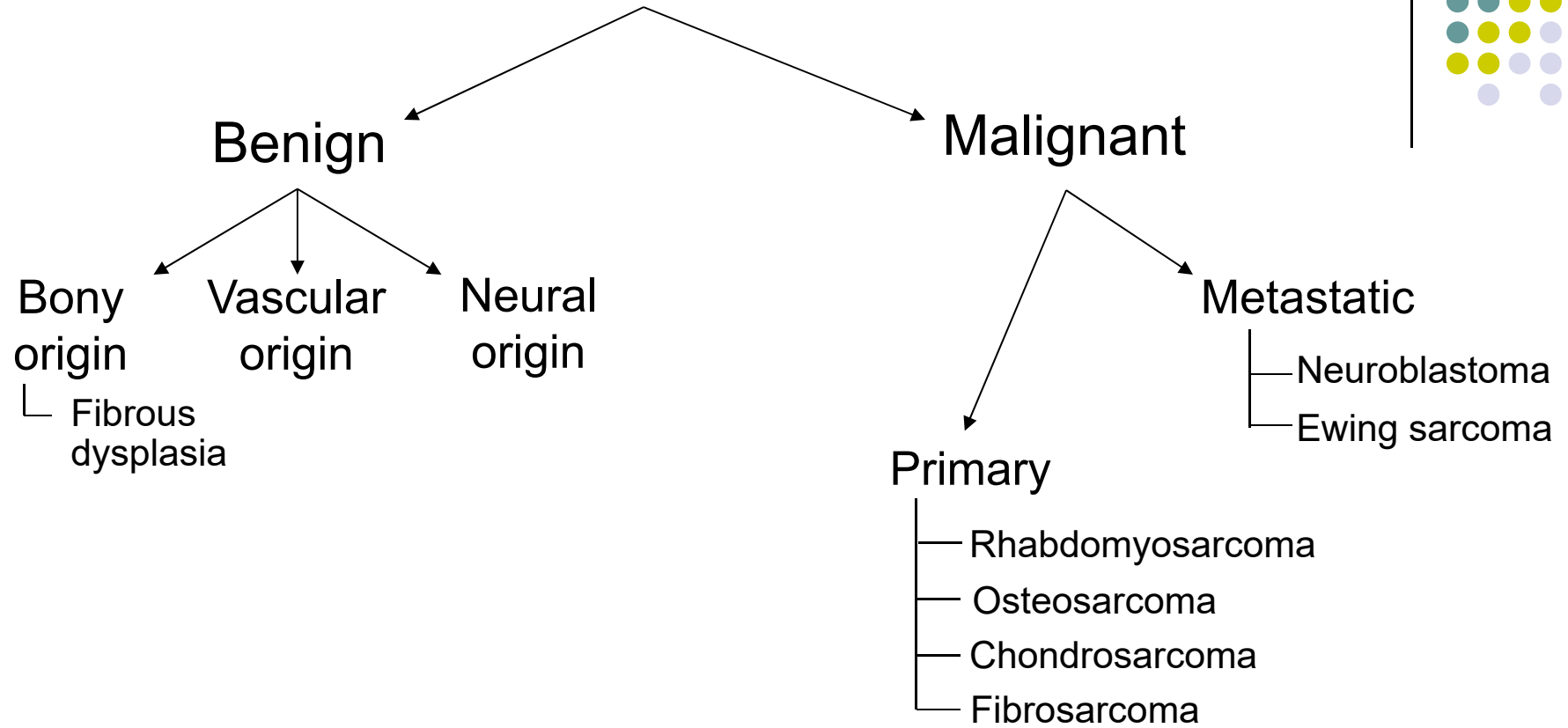


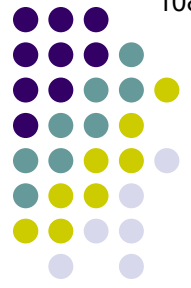
Pediatric Orbital Tumors



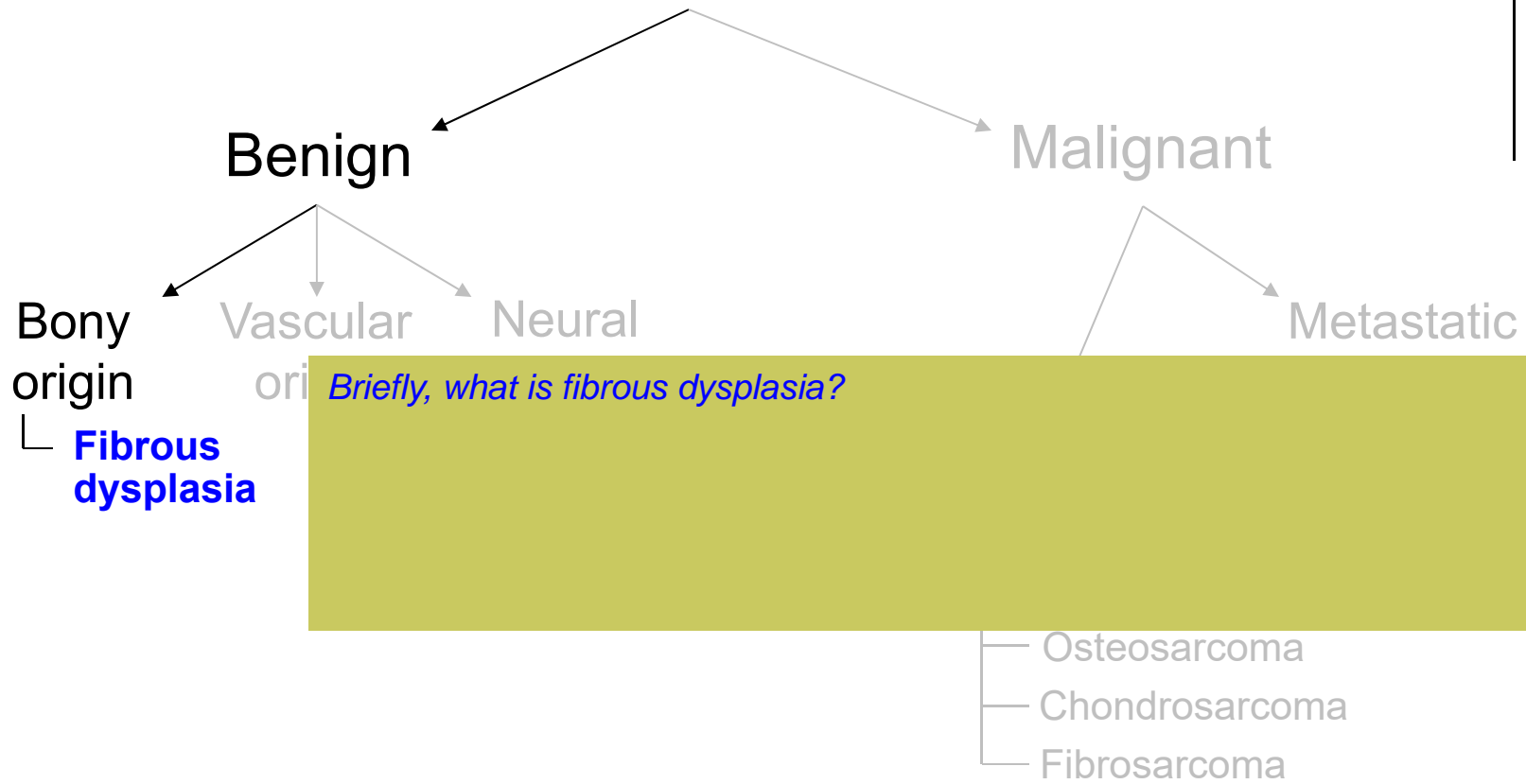


Pediatric Orbital Tumors





Pediatric Orbital Tumors



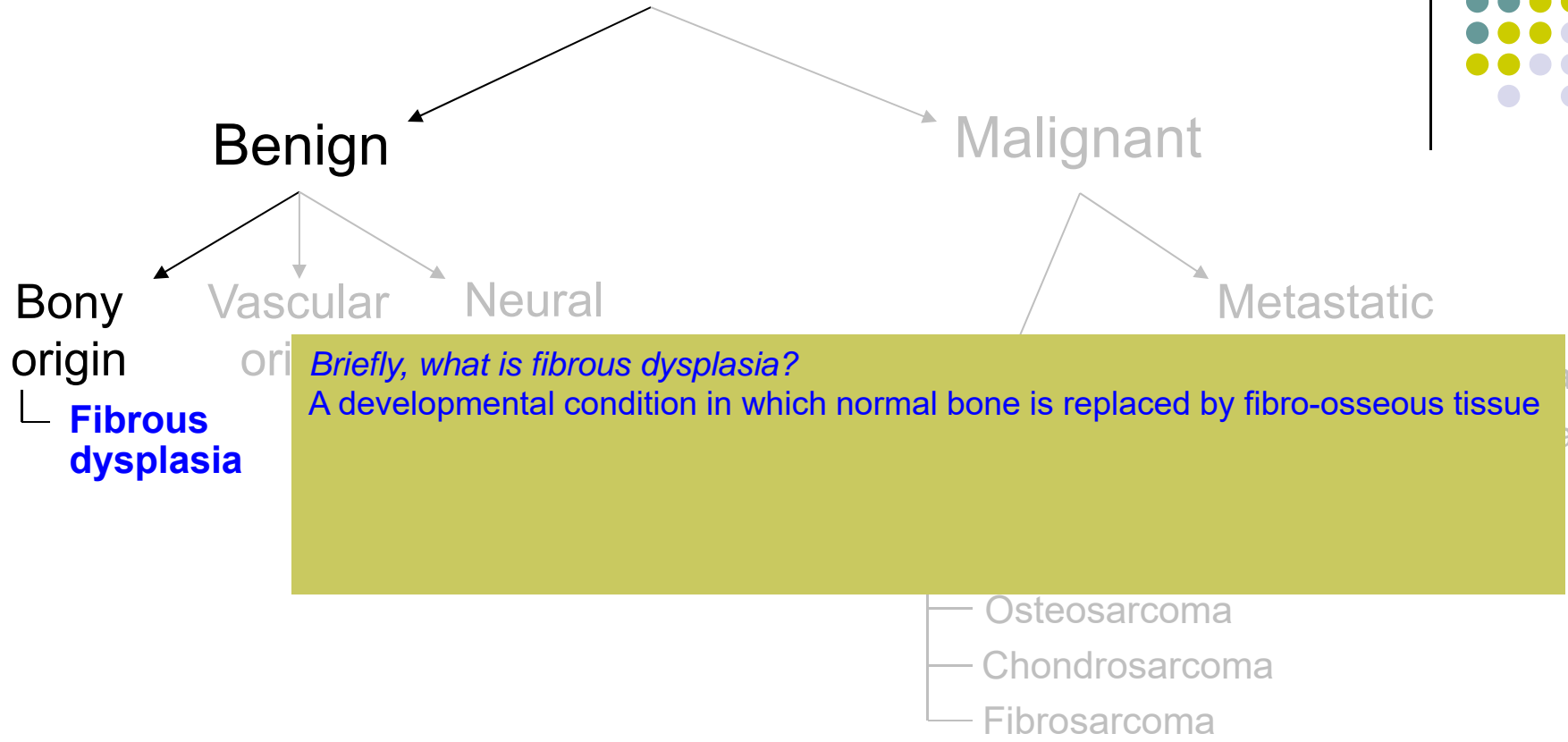
Briefly, what is fibrous dysplasia?

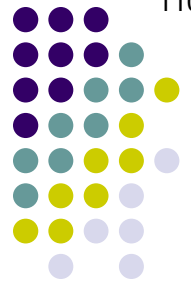
Bony origin
└ Fibrous dysplasia

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

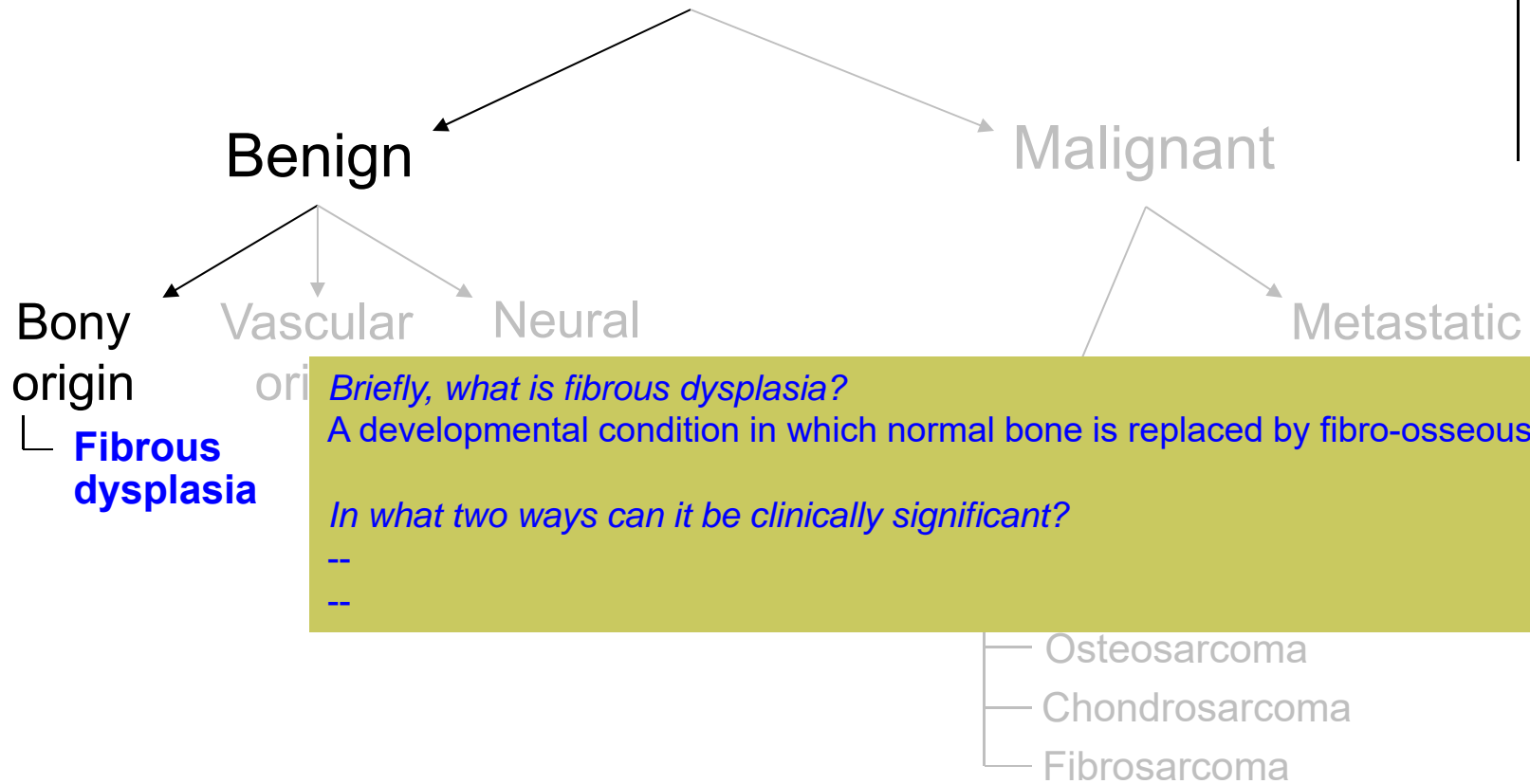


Pediatric Orbital Tumors





Pediatric Orbital Tumors



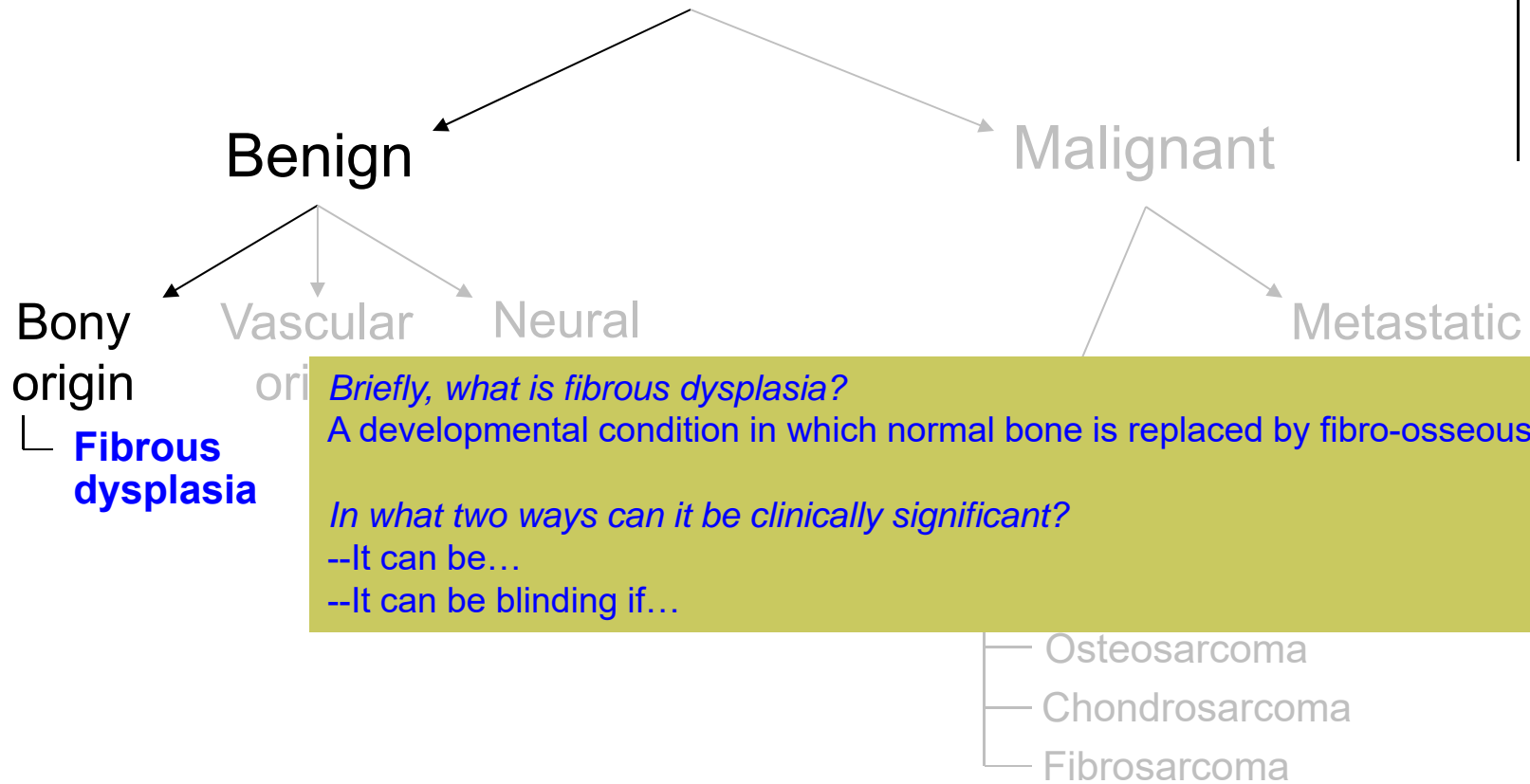
Briefly, what is fibrous dysplasia?
 A developmental condition in which normal bone is replaced by fibro-osseous tissue

In what two ways can it be clinically significant?
 --
 --

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma



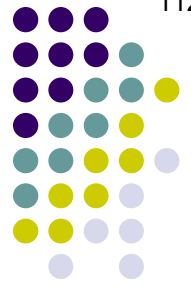
Pediatric Orbital Tumors



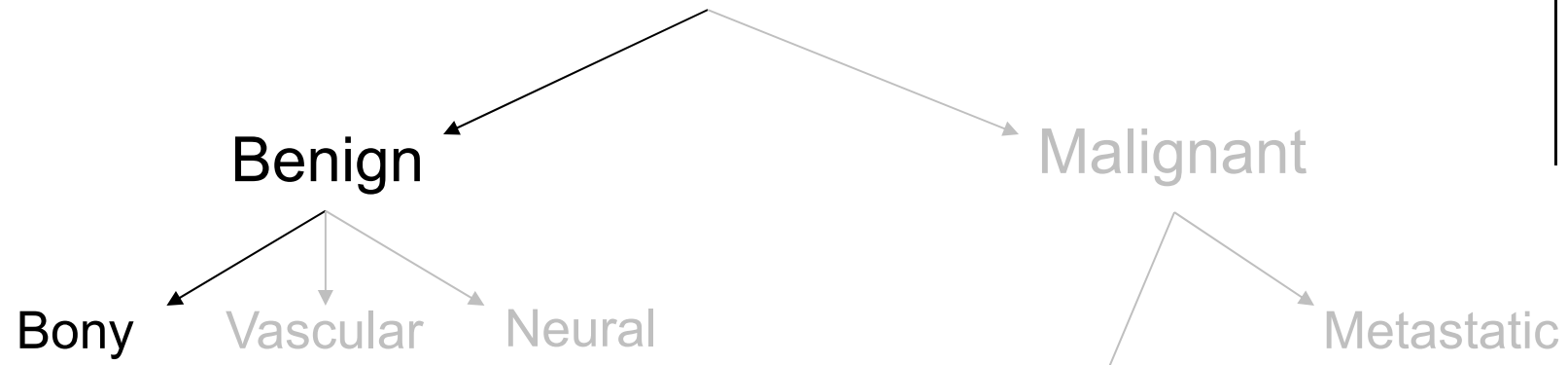
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 --It can be blinding if...

- Osteosarcoma
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Pediatric Orbital Tumors



Bony origin
└ **Fibrous dysplasia**

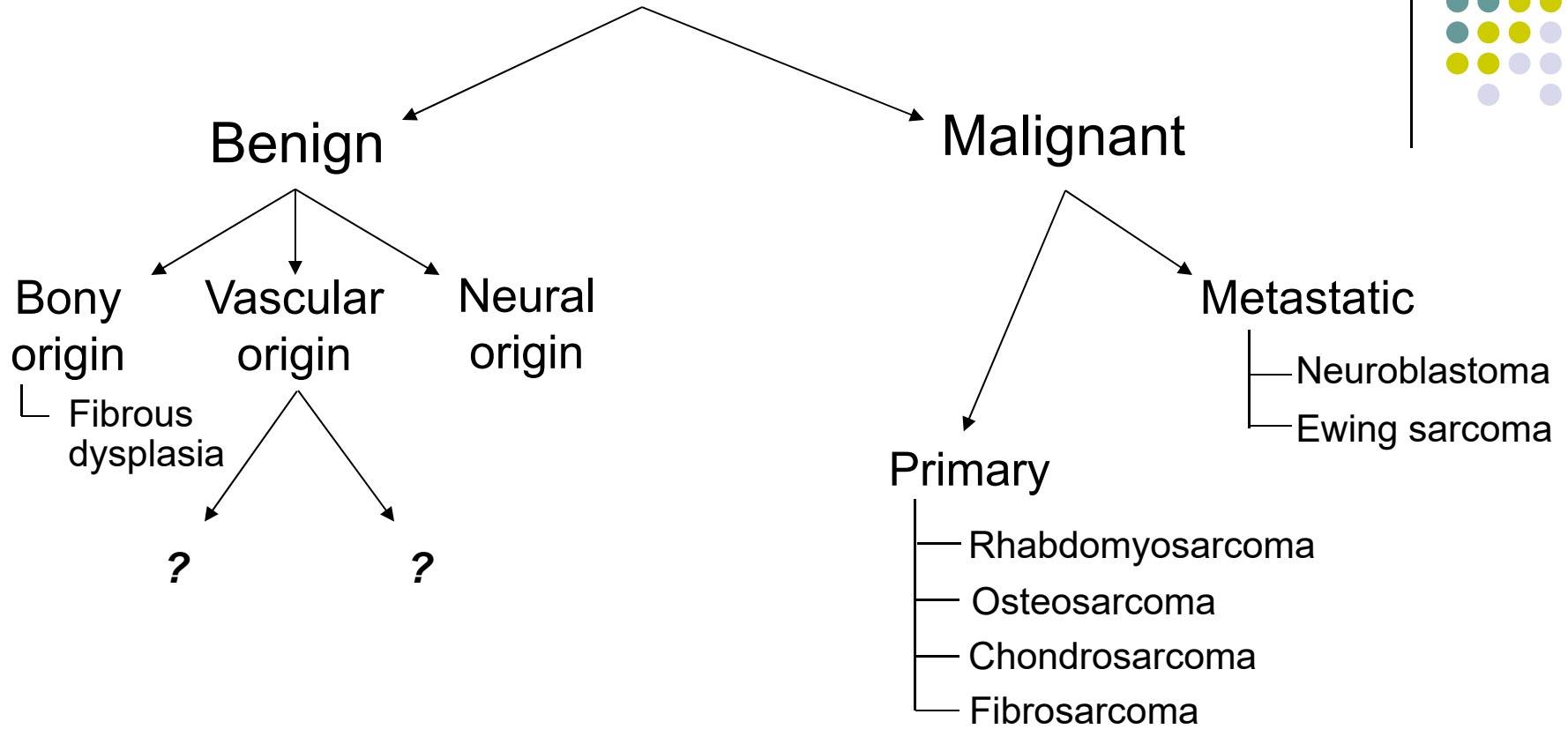
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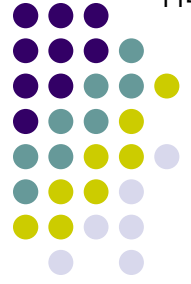
In what two ways can it be clinically significant?
 --It can be...cosmetically disfiguring
 --It can be blinding if...the optic nerve becomes compressed

- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma

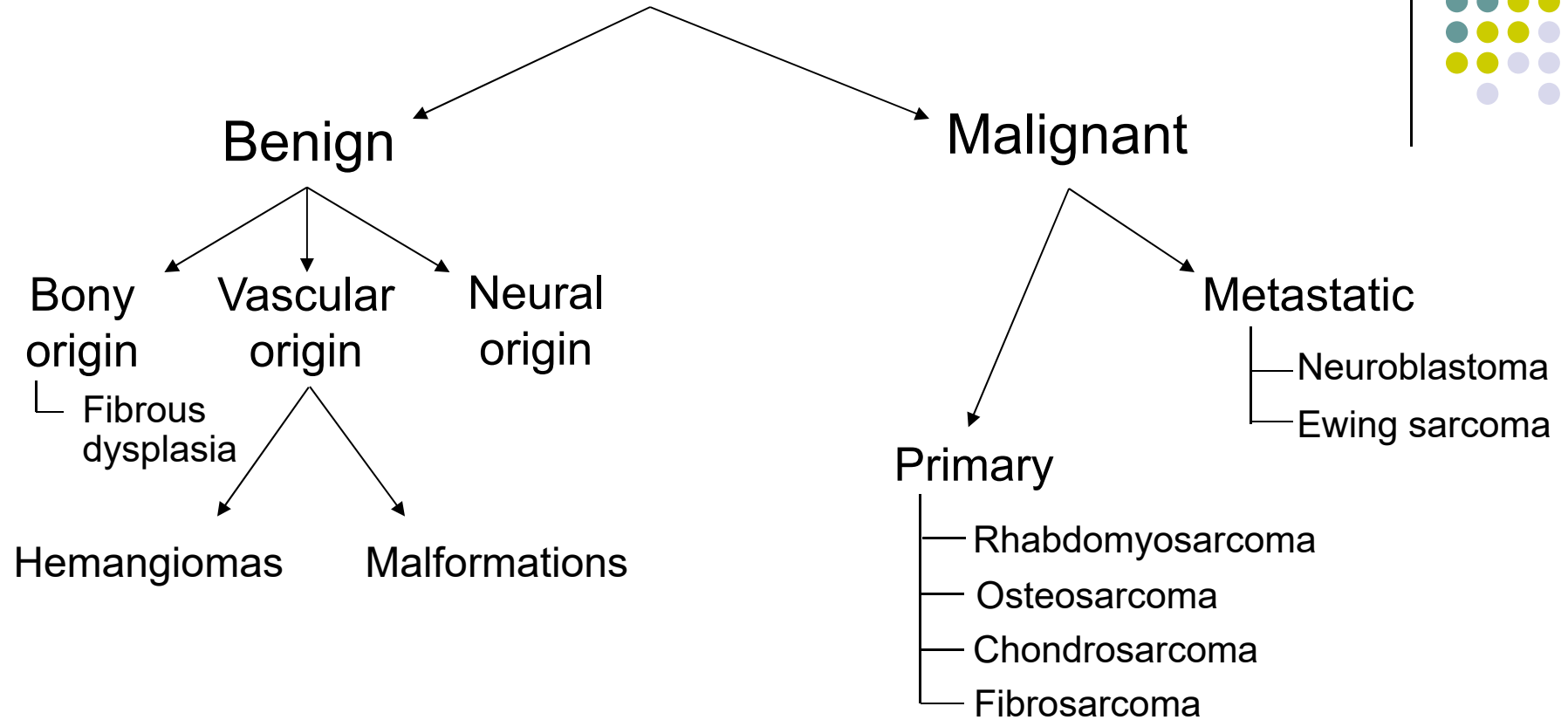


Pediatric Orbital Tumors



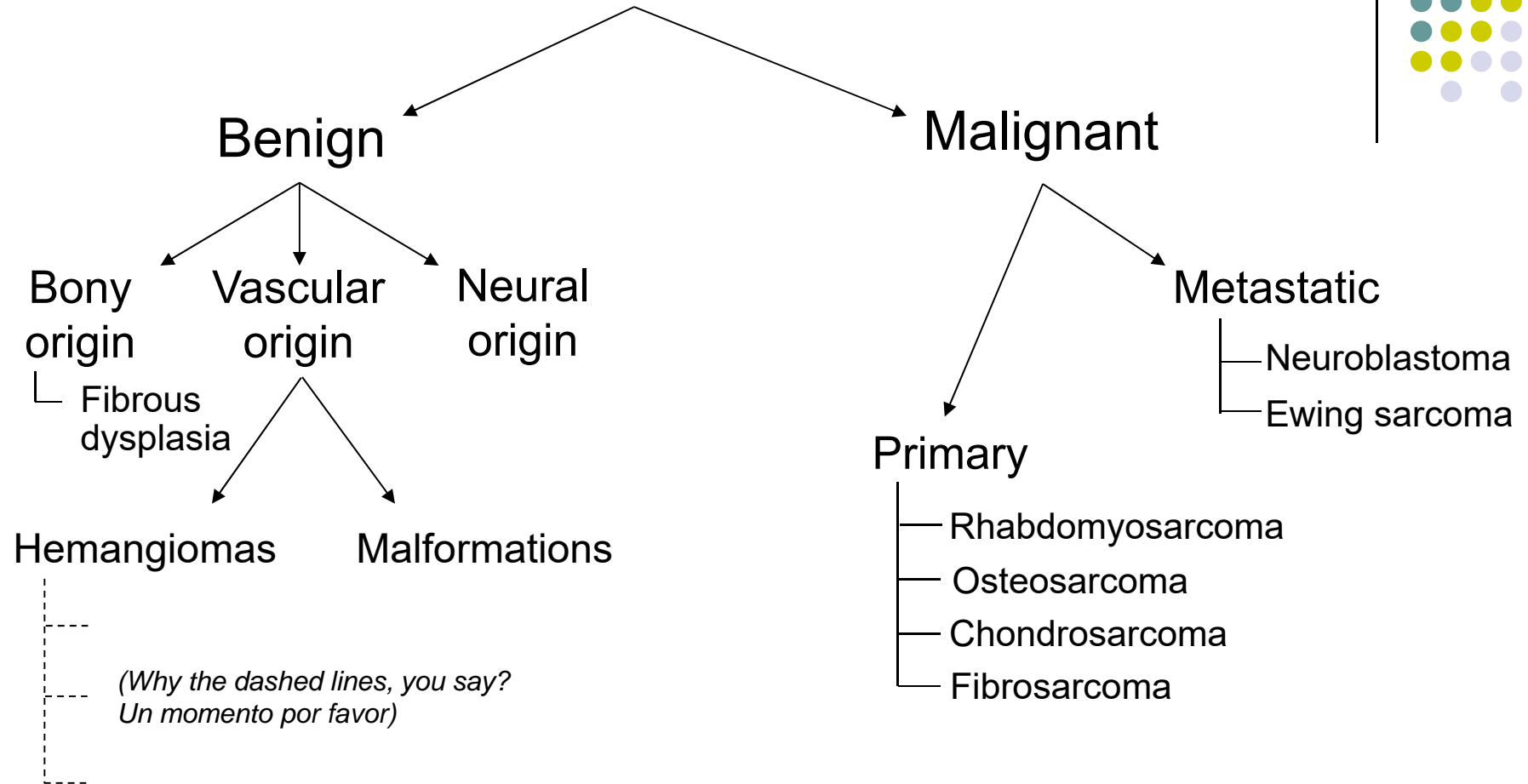


Pediatric Orbital Tumors

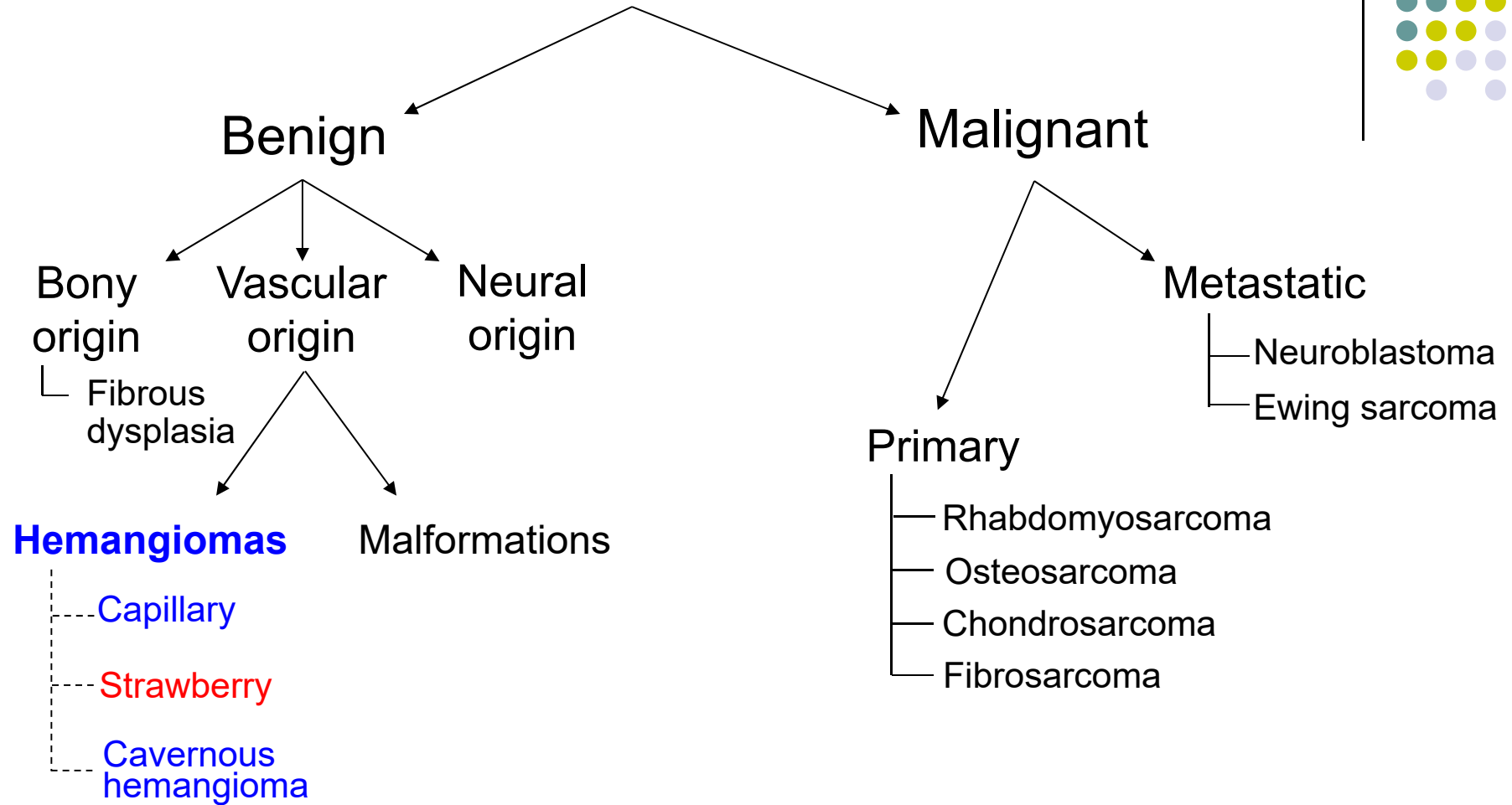




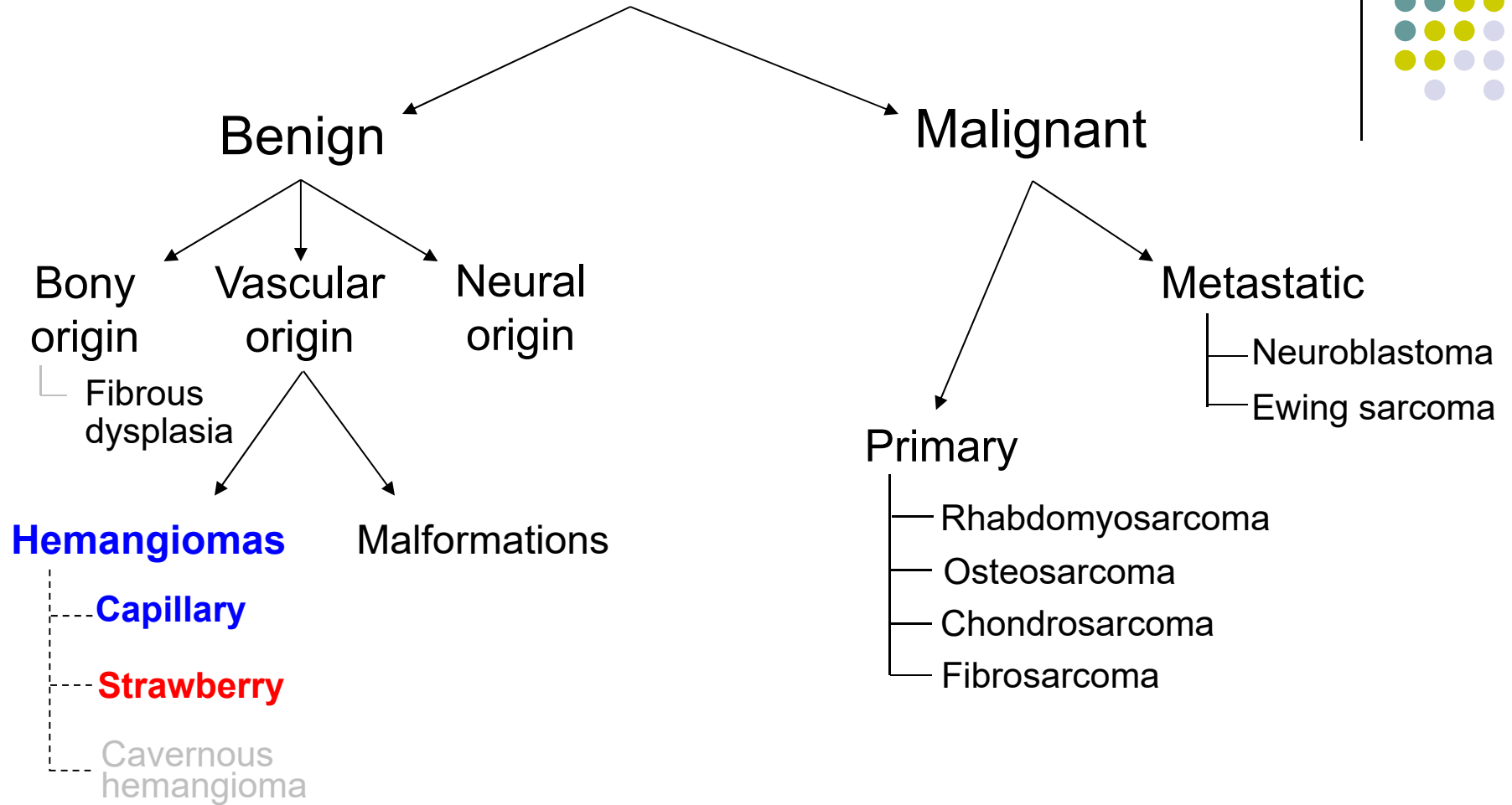
Pediatric Orbital Tumors



Pediatric Orbital Tumors

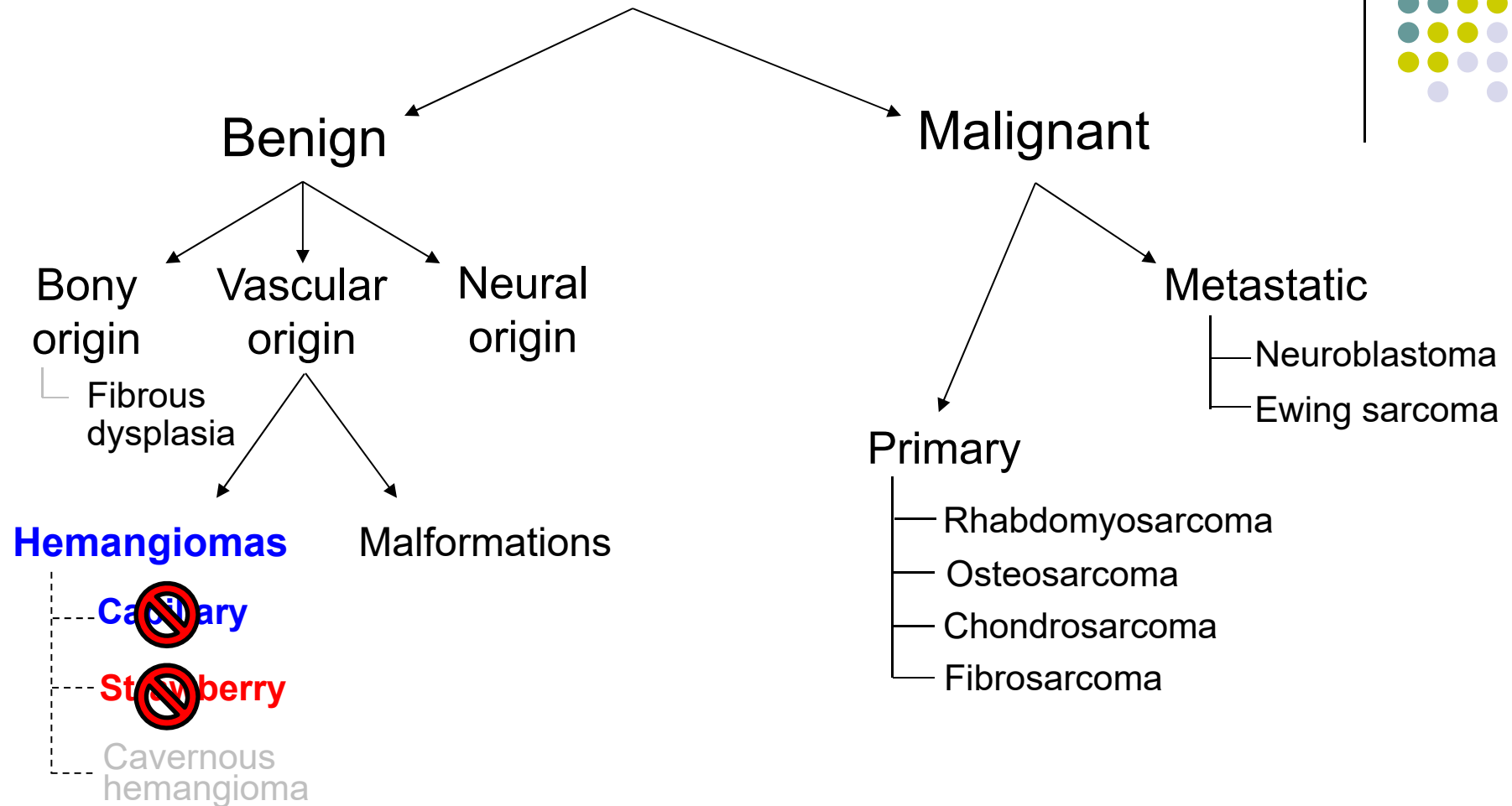


Pediatric Orbital Tumors

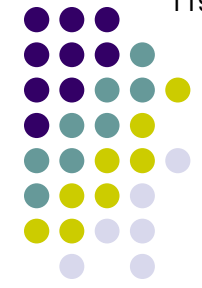


Re the terms *capillary hemangioma* and *strawberry hemangioma*:

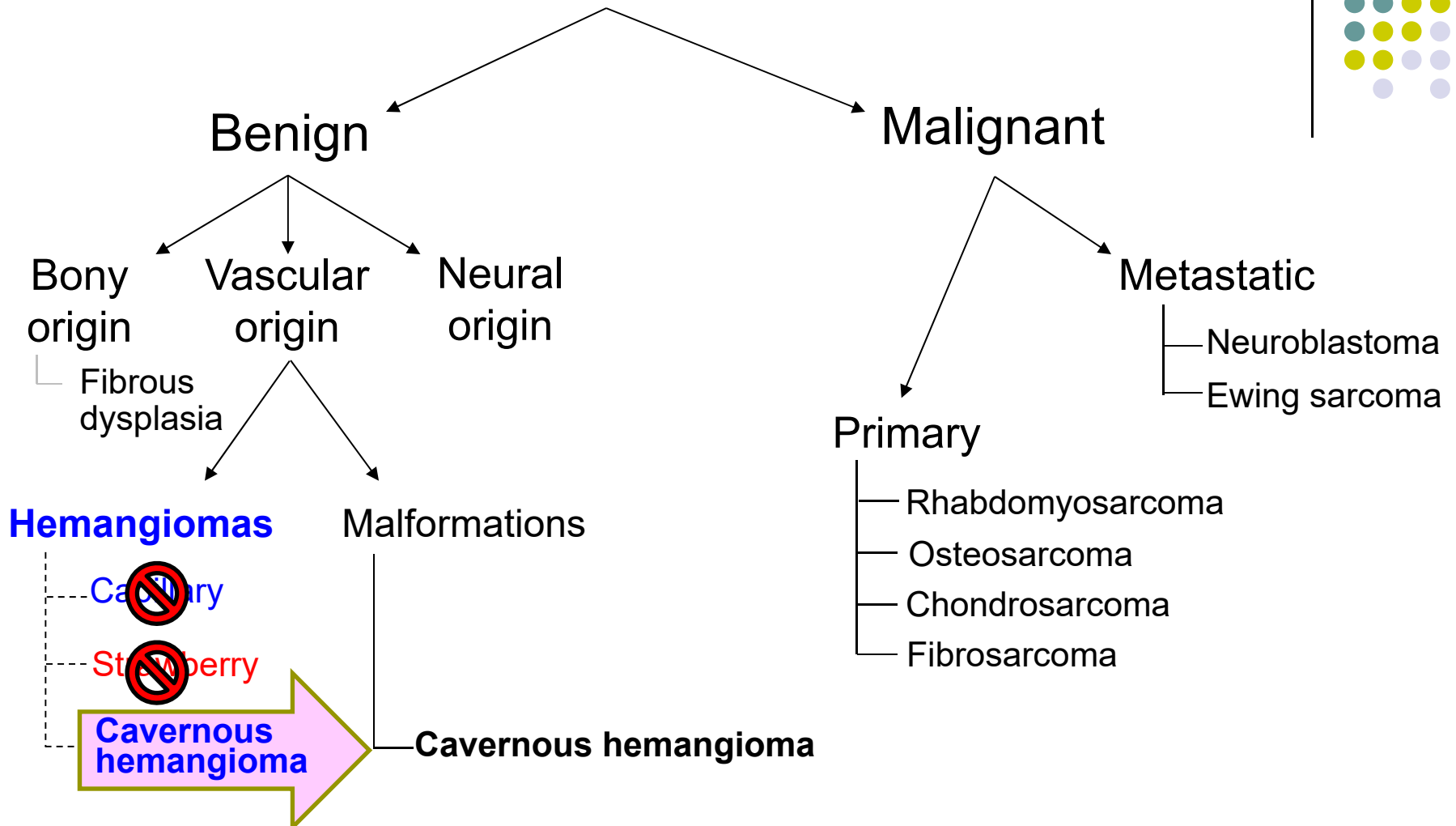
Pediatric Orbital Tumors



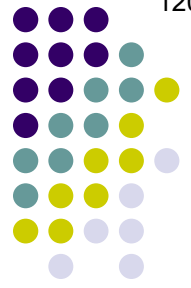
Re the terms *capillary hemangioma* and *strawberry hemangioma*: The 2018-19 edition of the BCSC *Peds* book indicates they are outdated, and such lesions should be referred to simply as 'hemangiomas.' However, it also acknowledges that these terms persist in the ophthalmic literature, so they will be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



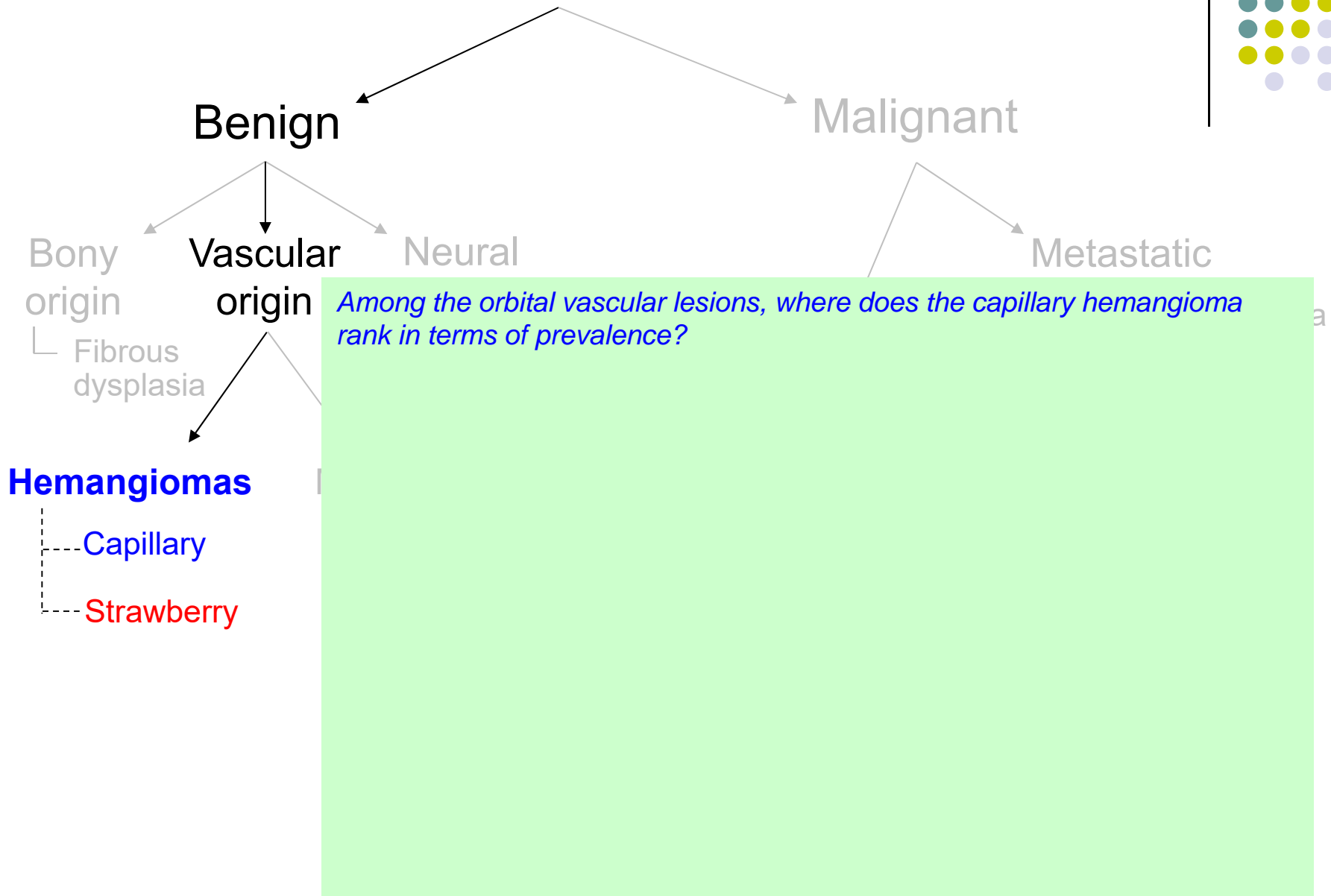
Pediatric Orbital Tumors



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.**

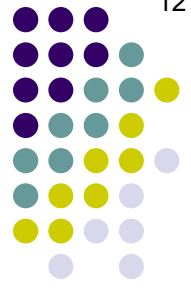


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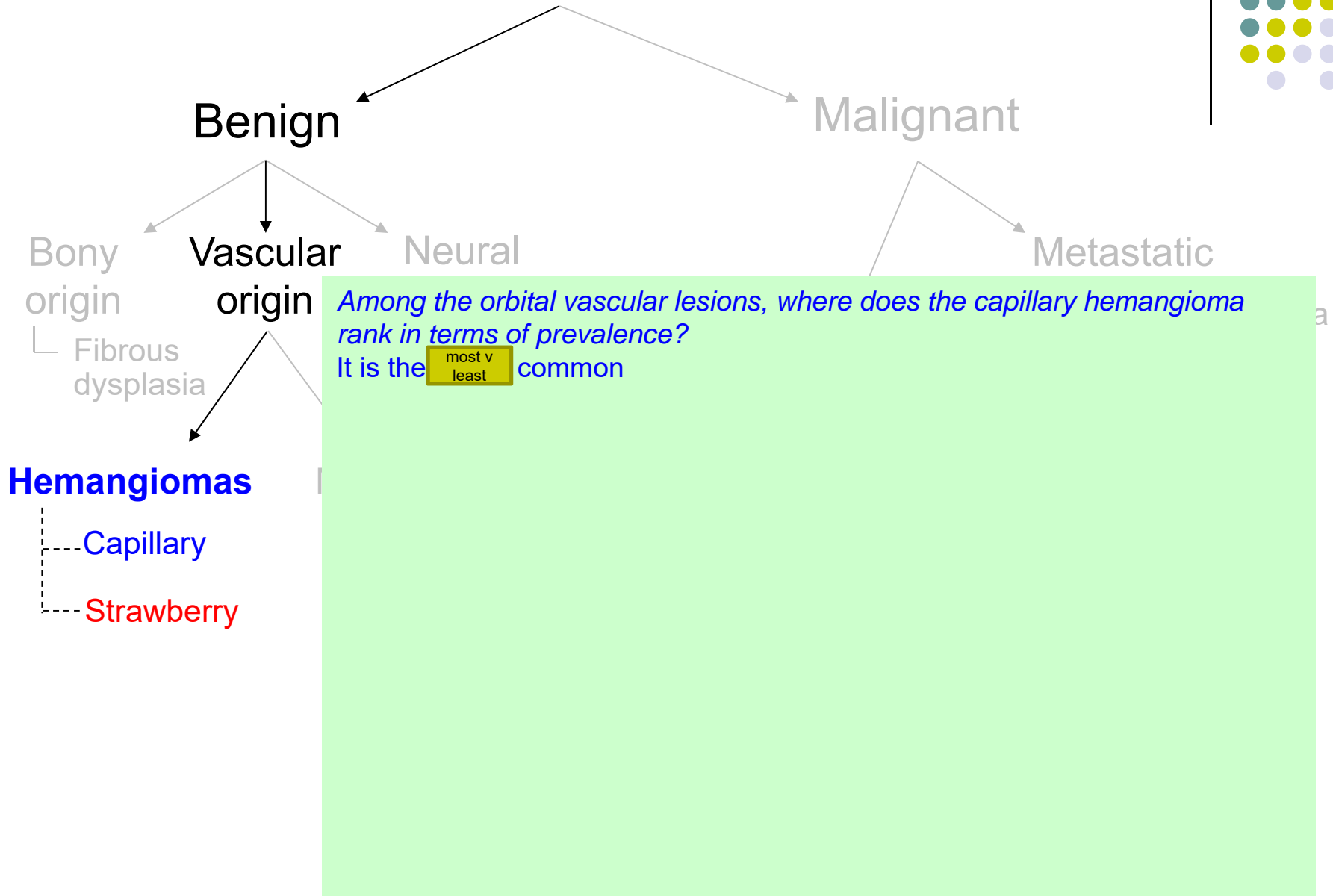


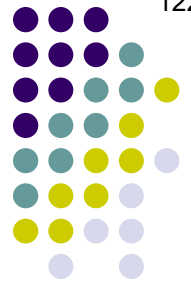
Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

- Hemangiomas**
 - Capillary
 - Strawberry

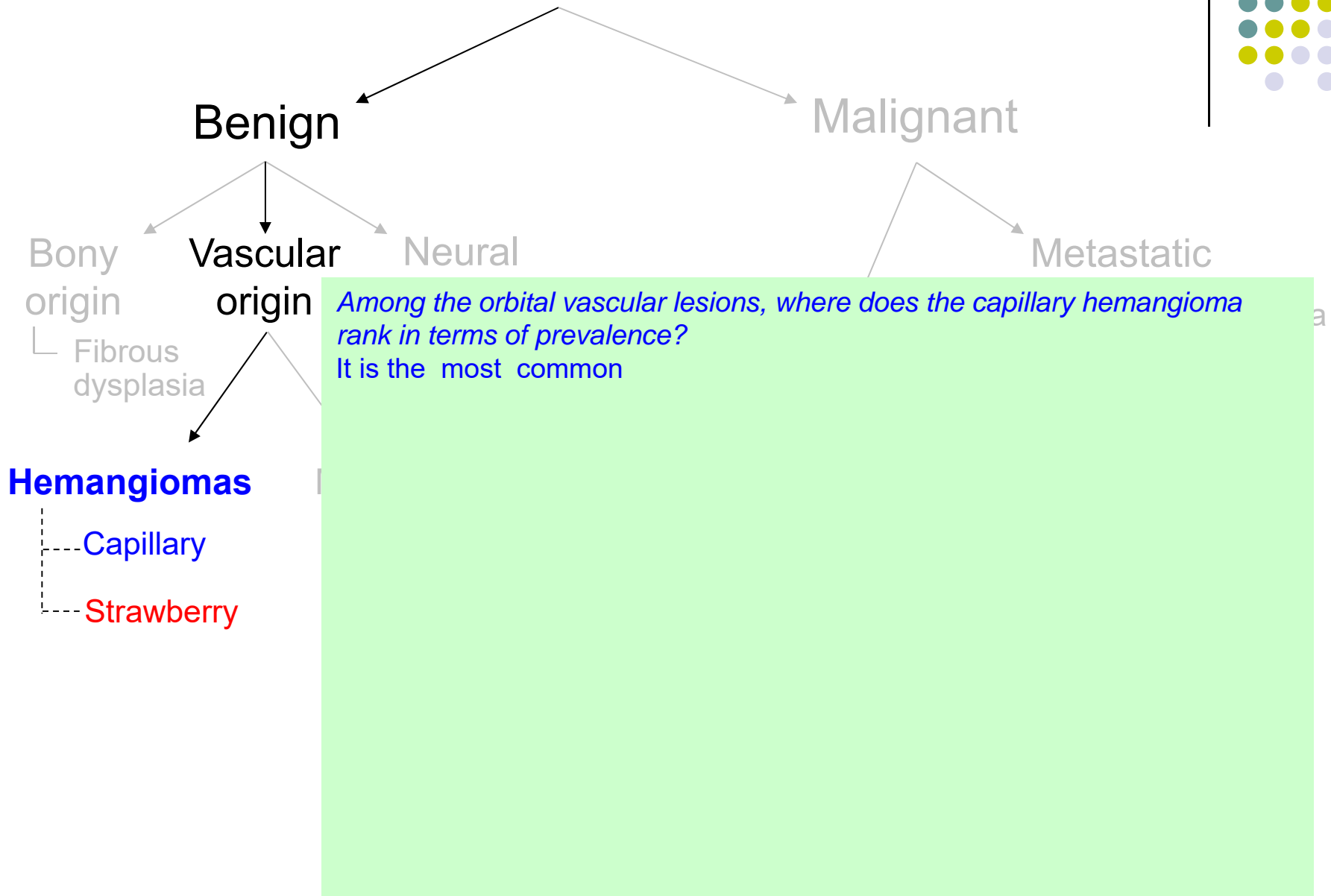


Pediatric Orbital Tumors



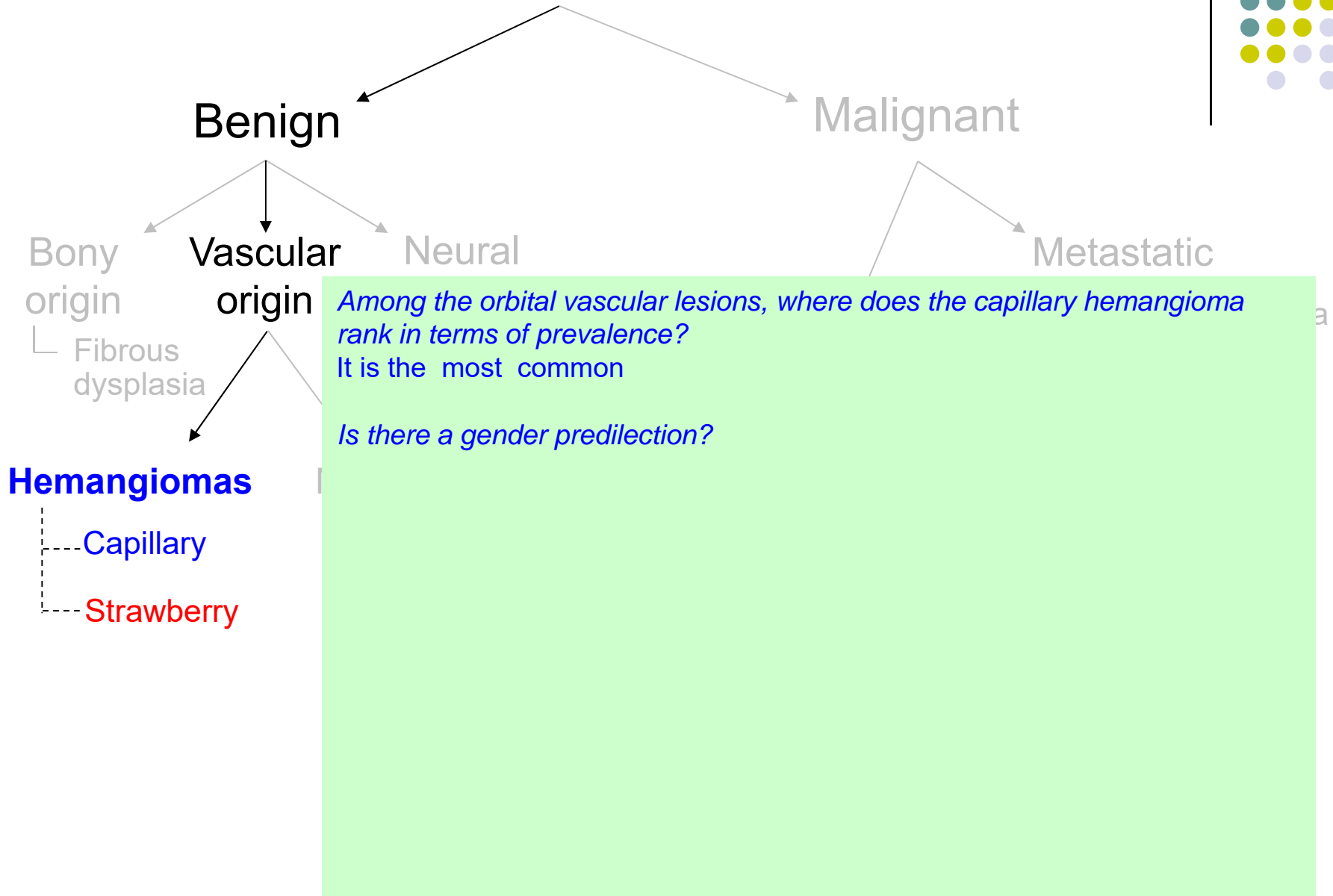


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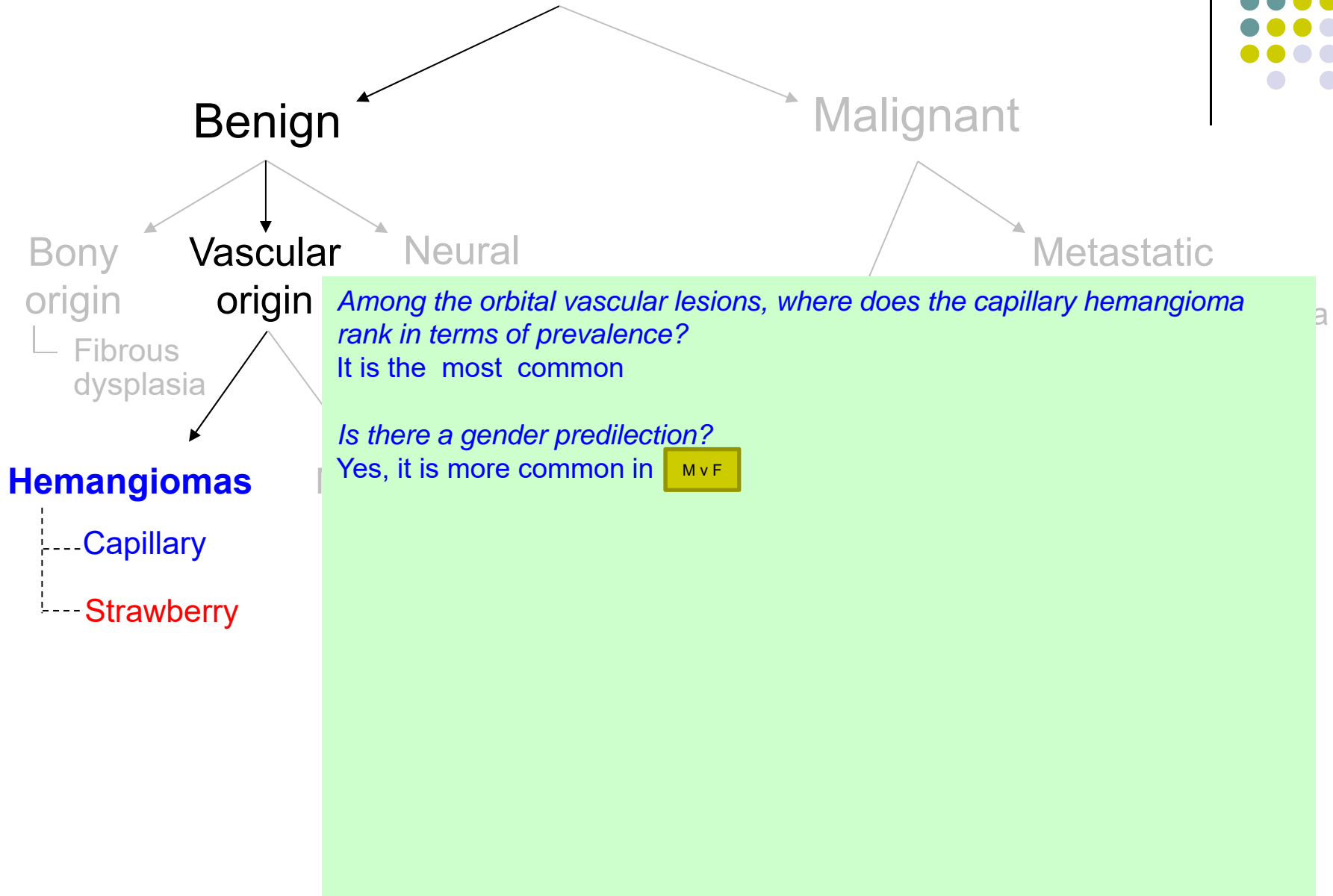


Pediatric Orbital Tumors



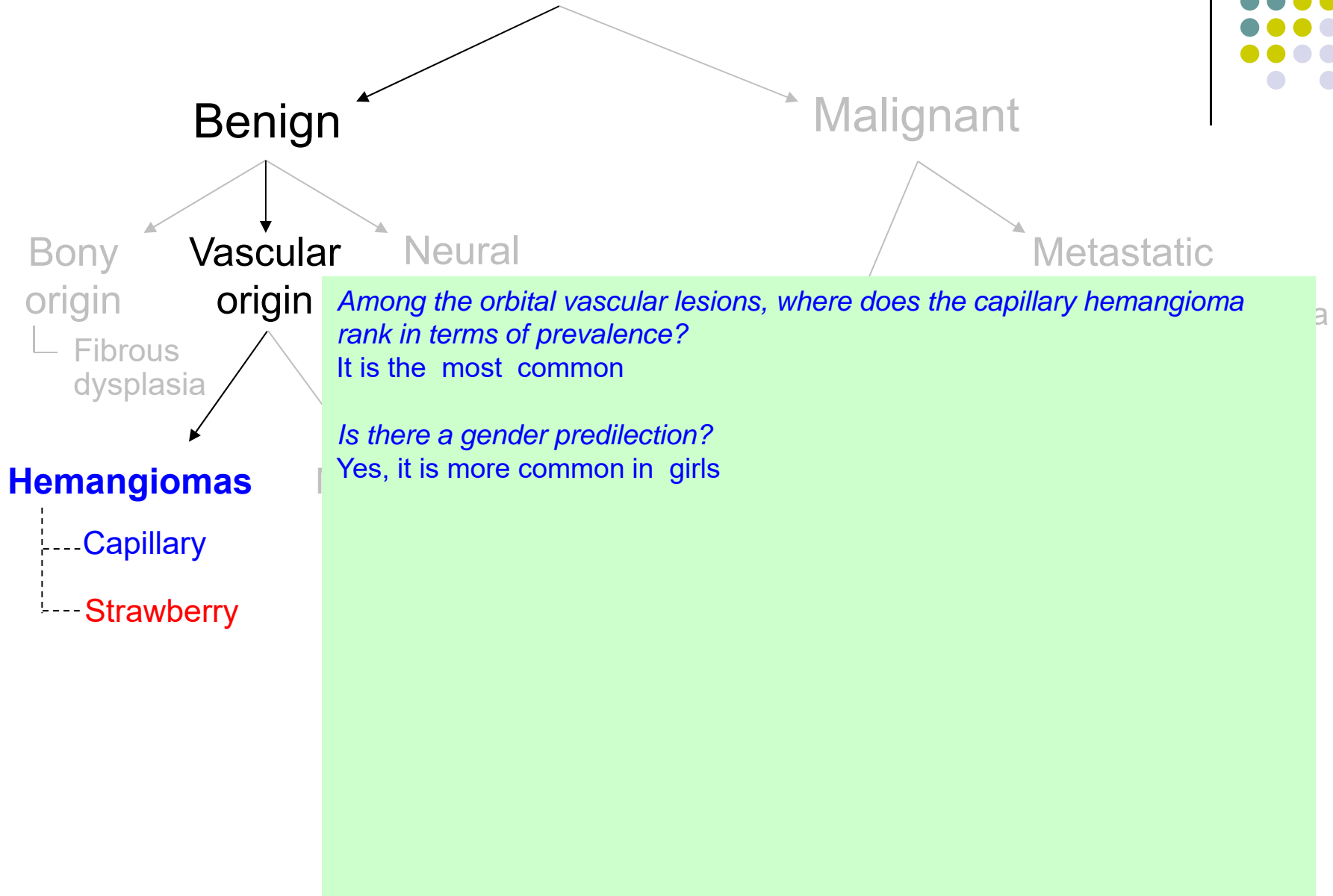


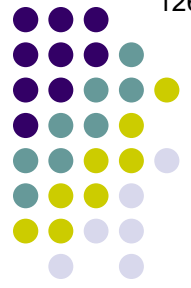
Pediatric Orbital Tumors



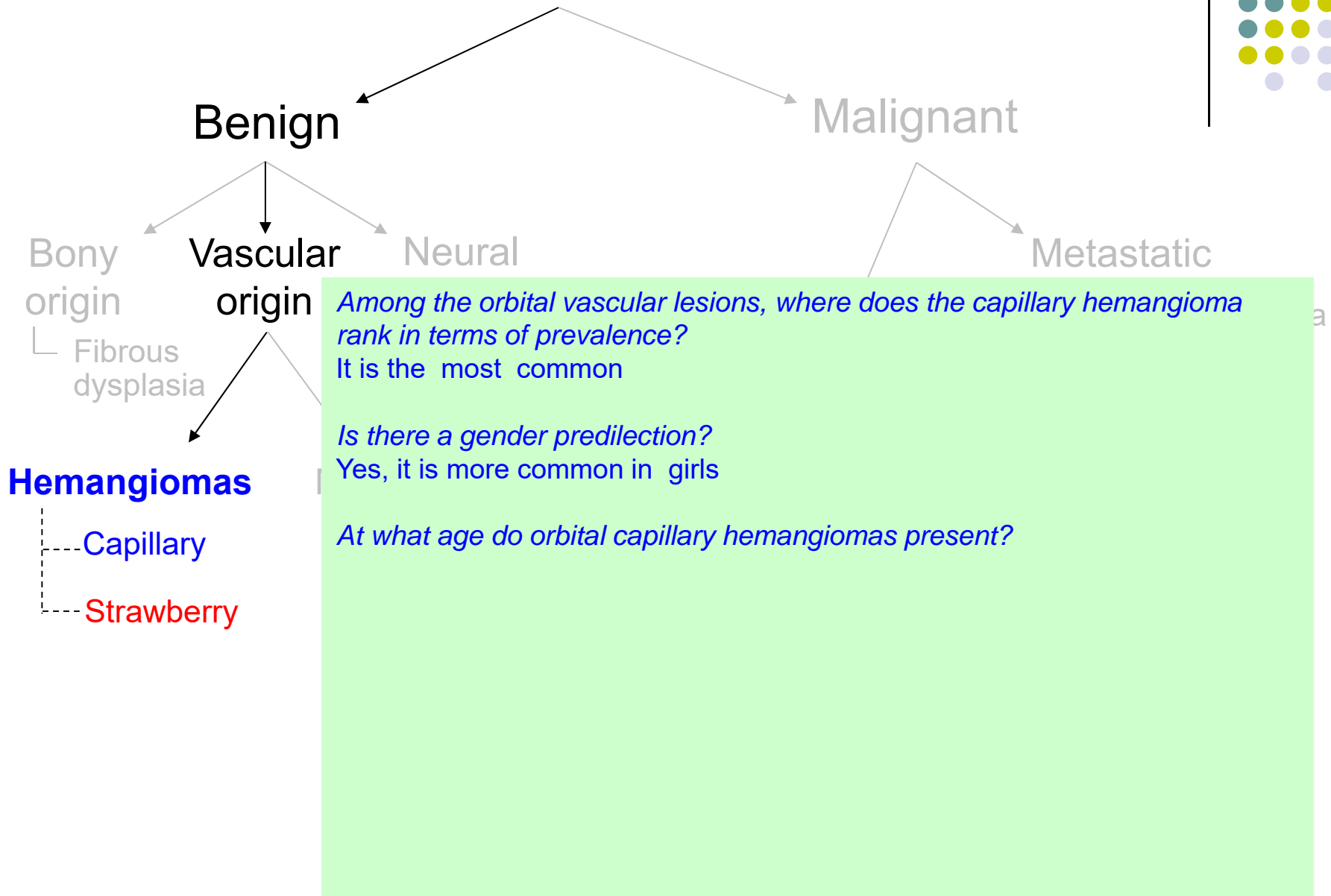


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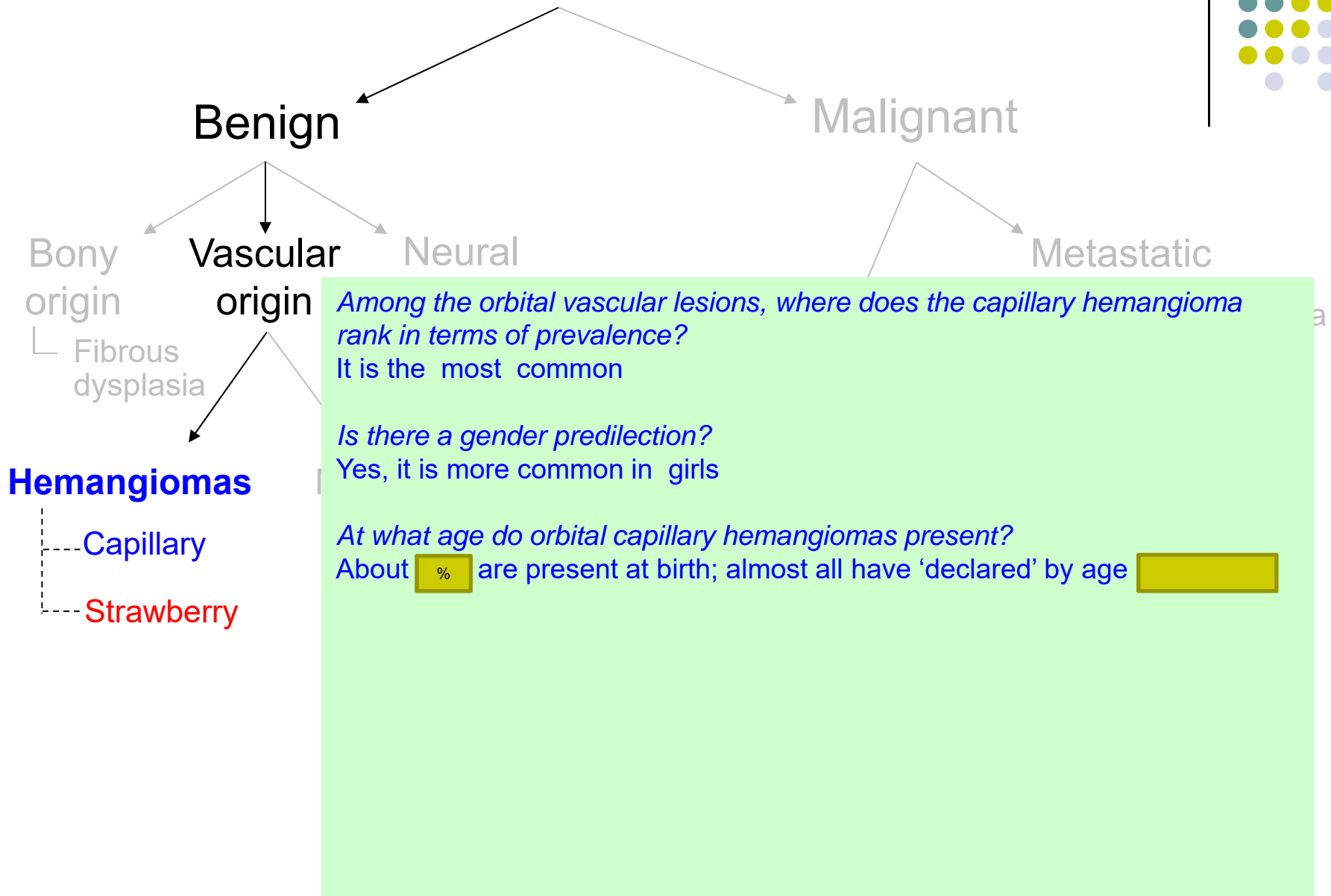


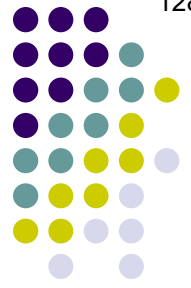
Pediatric Orbital Tumors



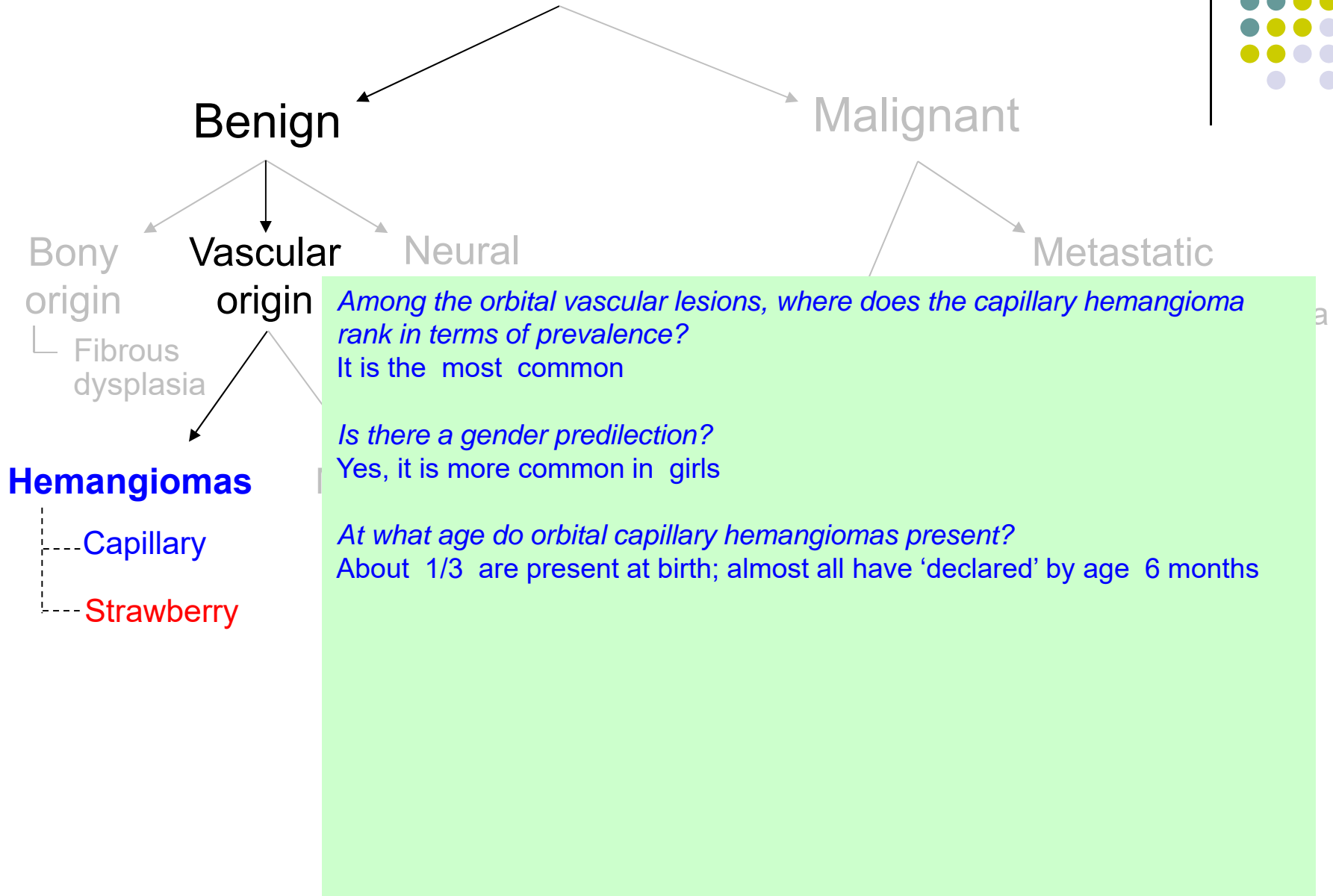


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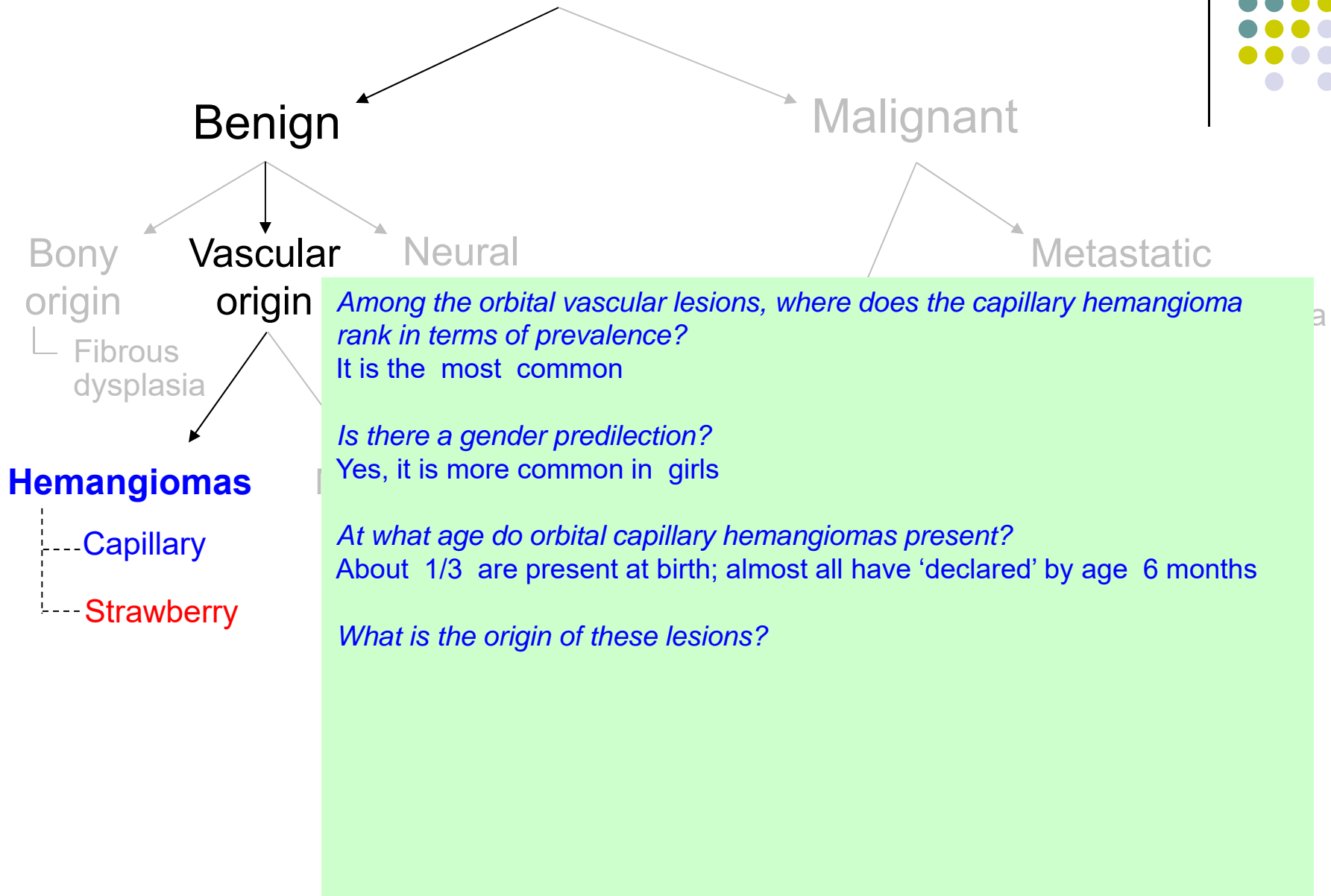


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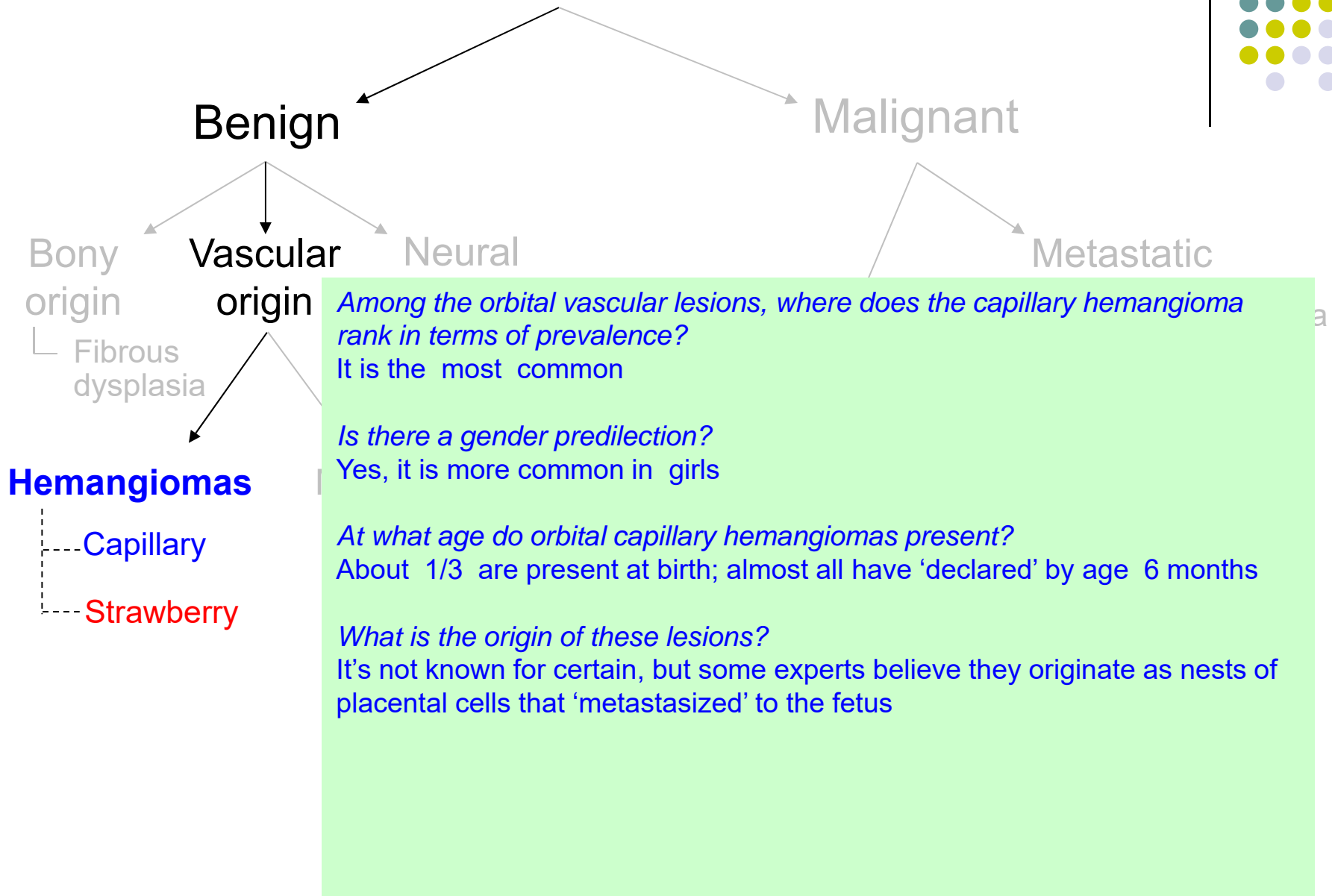


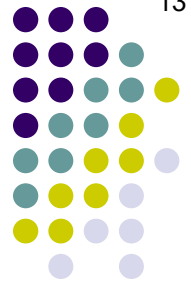
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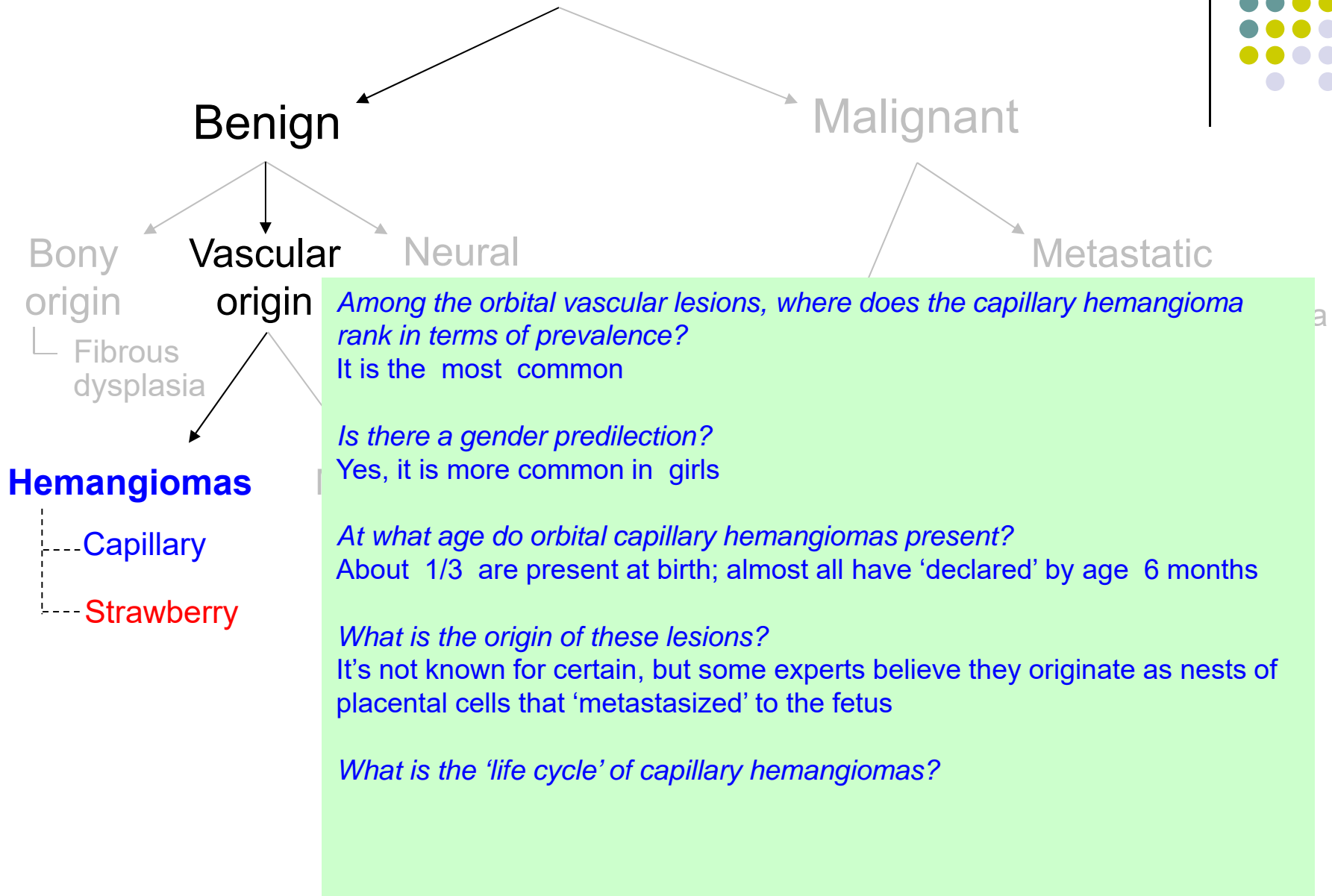


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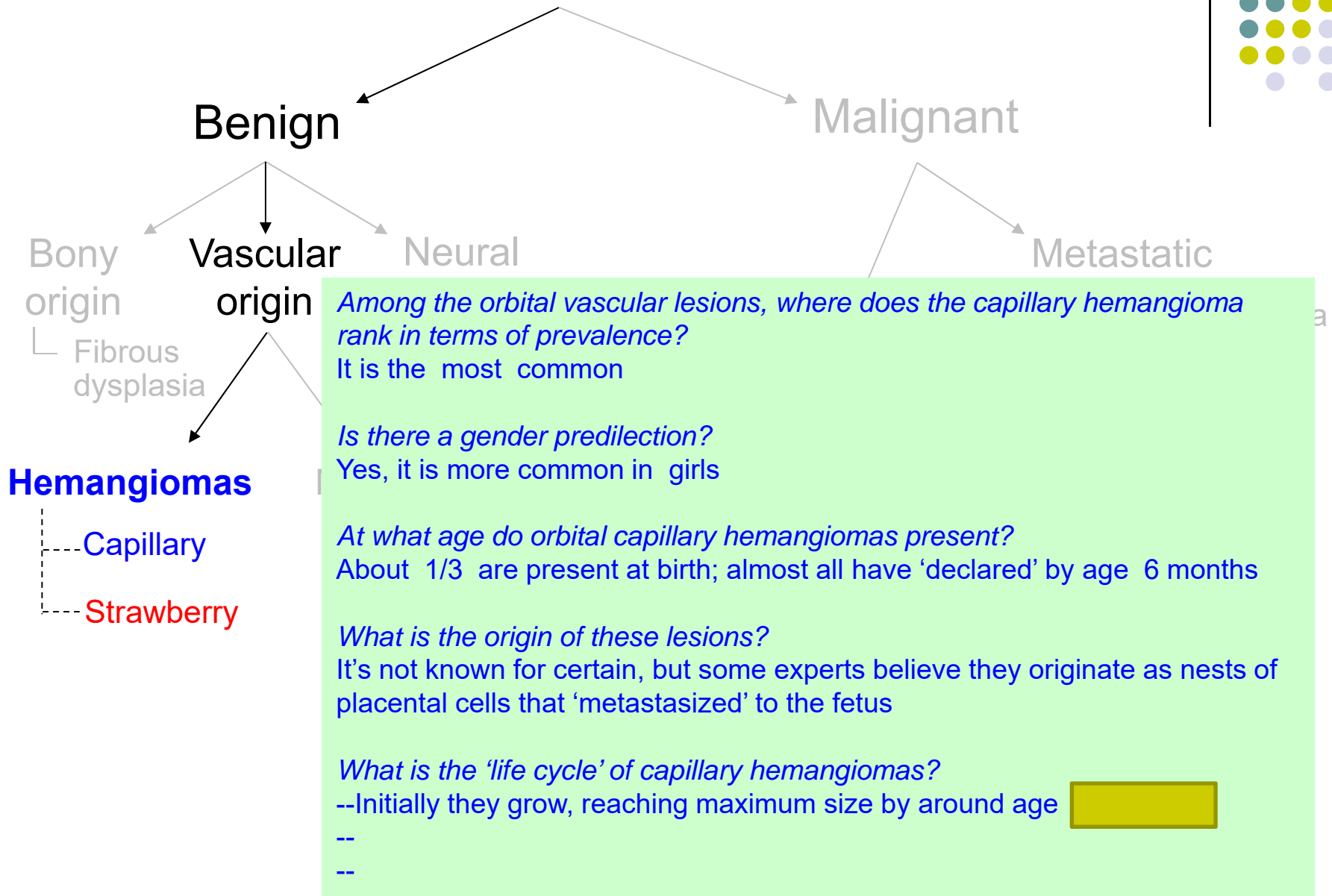


Pediatric Orbital Tumors



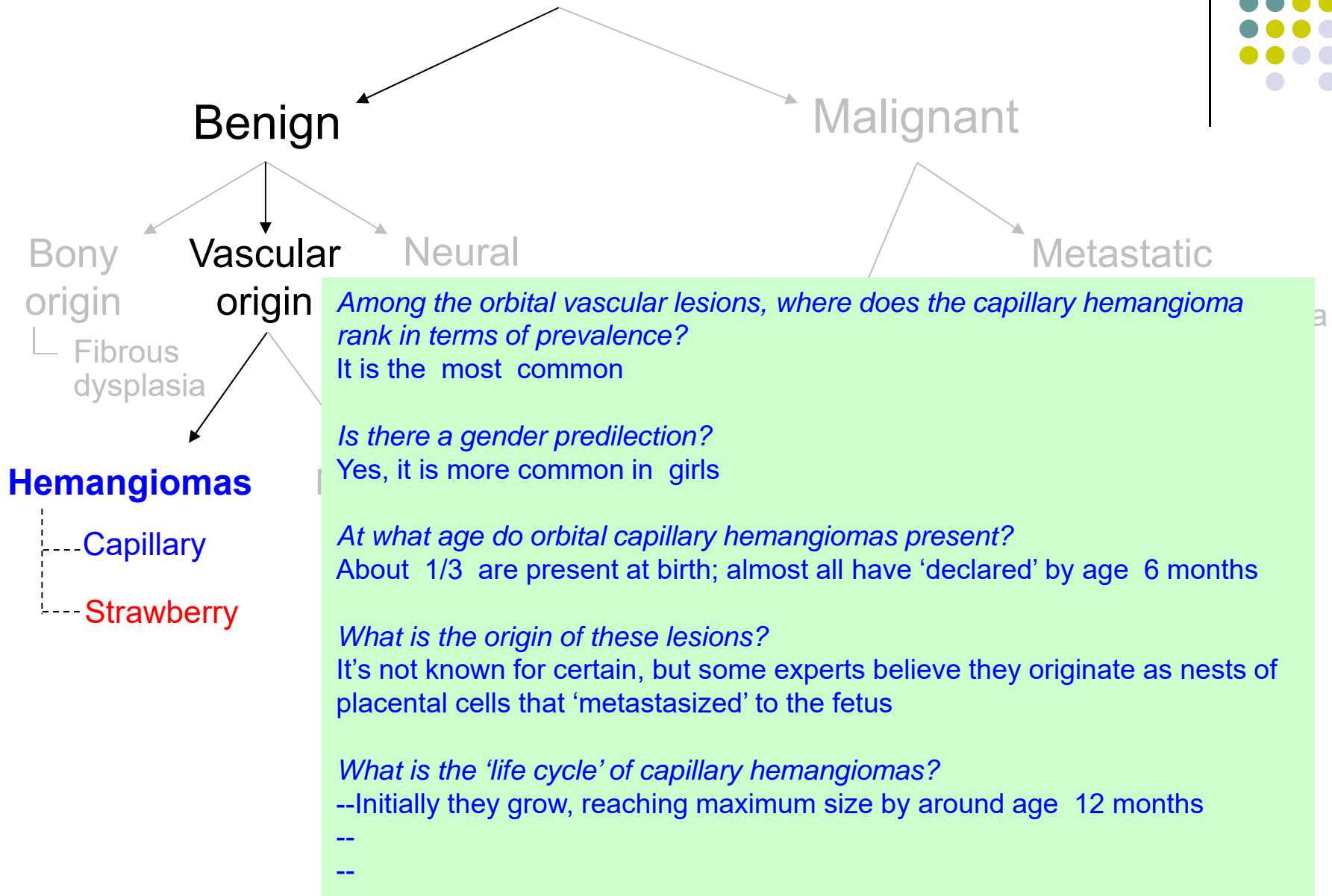


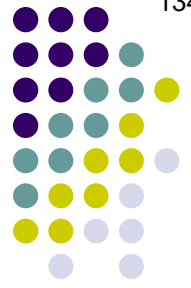
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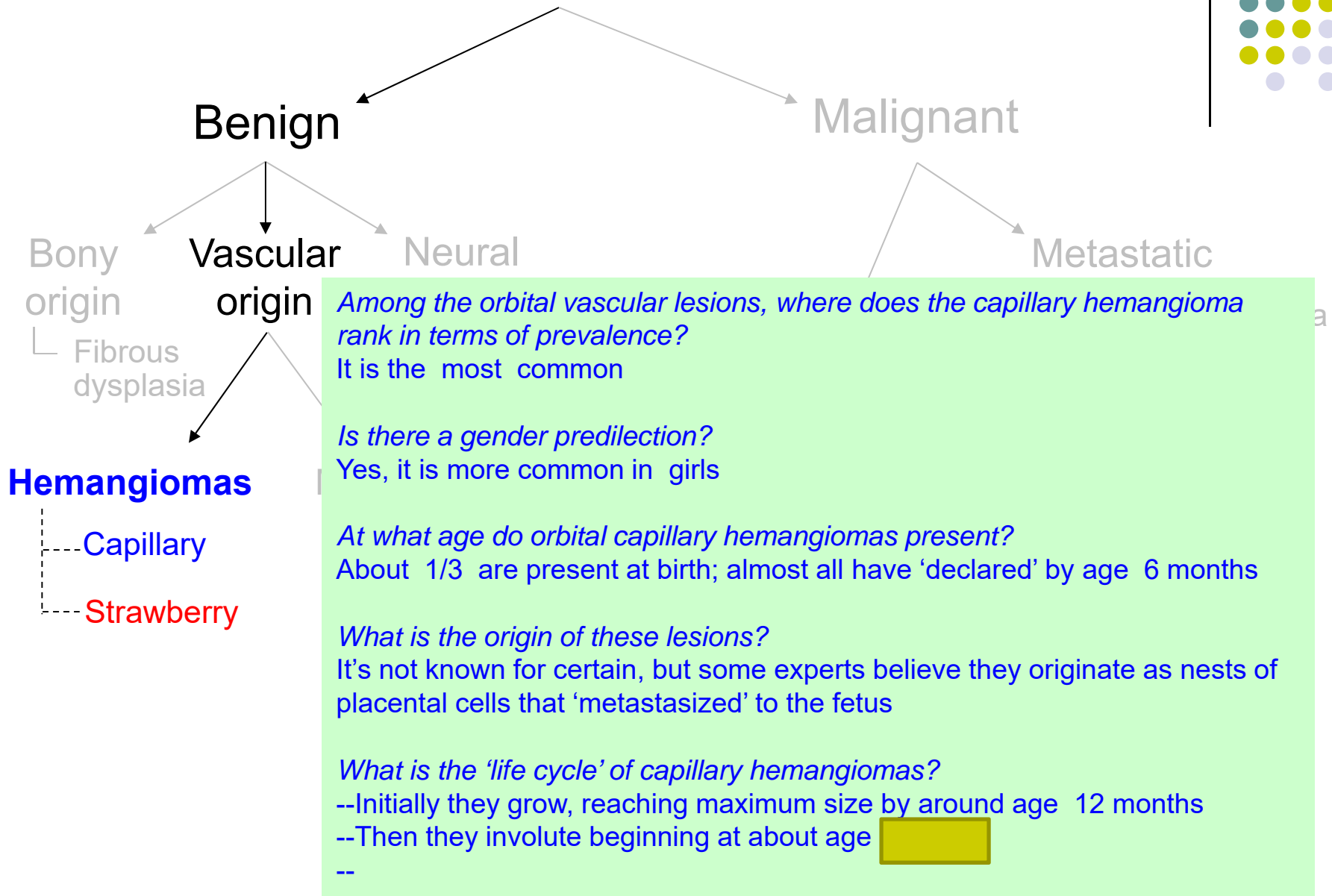


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Pediatric Orbital Tumors



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

Is there a gender predilection?
 Yes, it is more common in girls

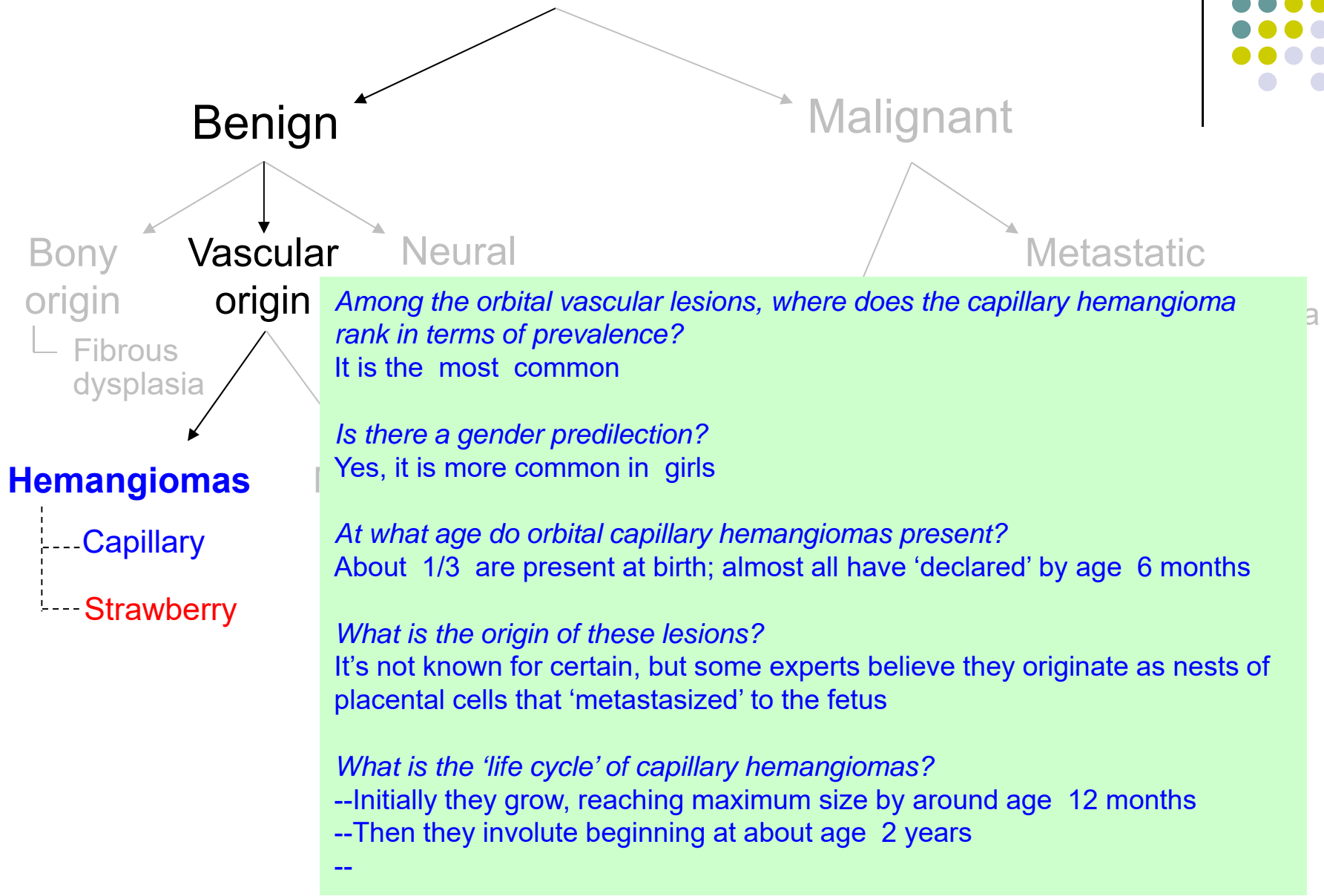
At what age do orbital capillary hemangiomas present?
 About 1/3 are present at birth; almost all have 'declared' by age 6 months

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?
 --Initially they grow, reaching maximum size by around age 12 months
 --Then they involute beginning at about age [redacted]
 --

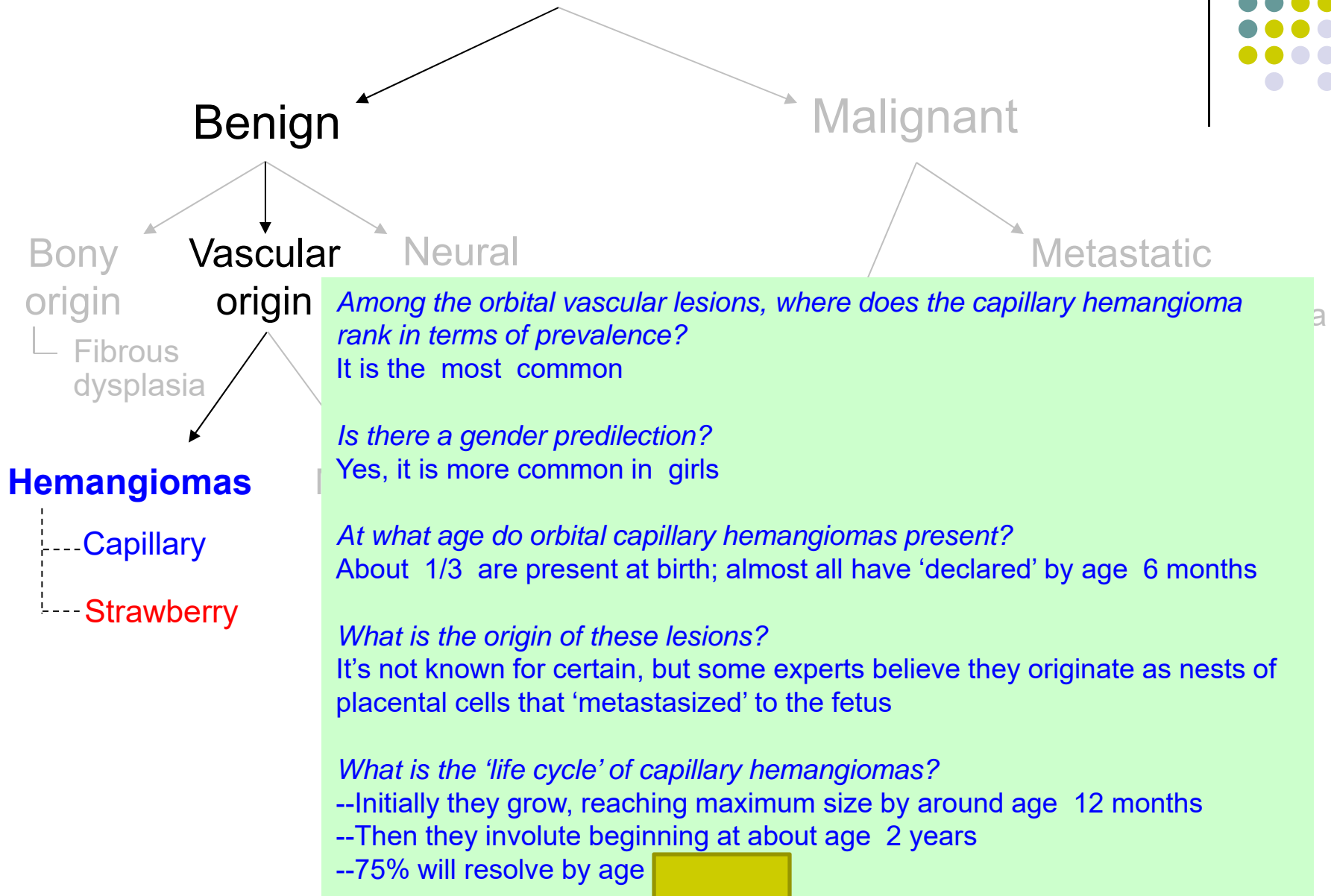


Pediatric Orbital Tumors



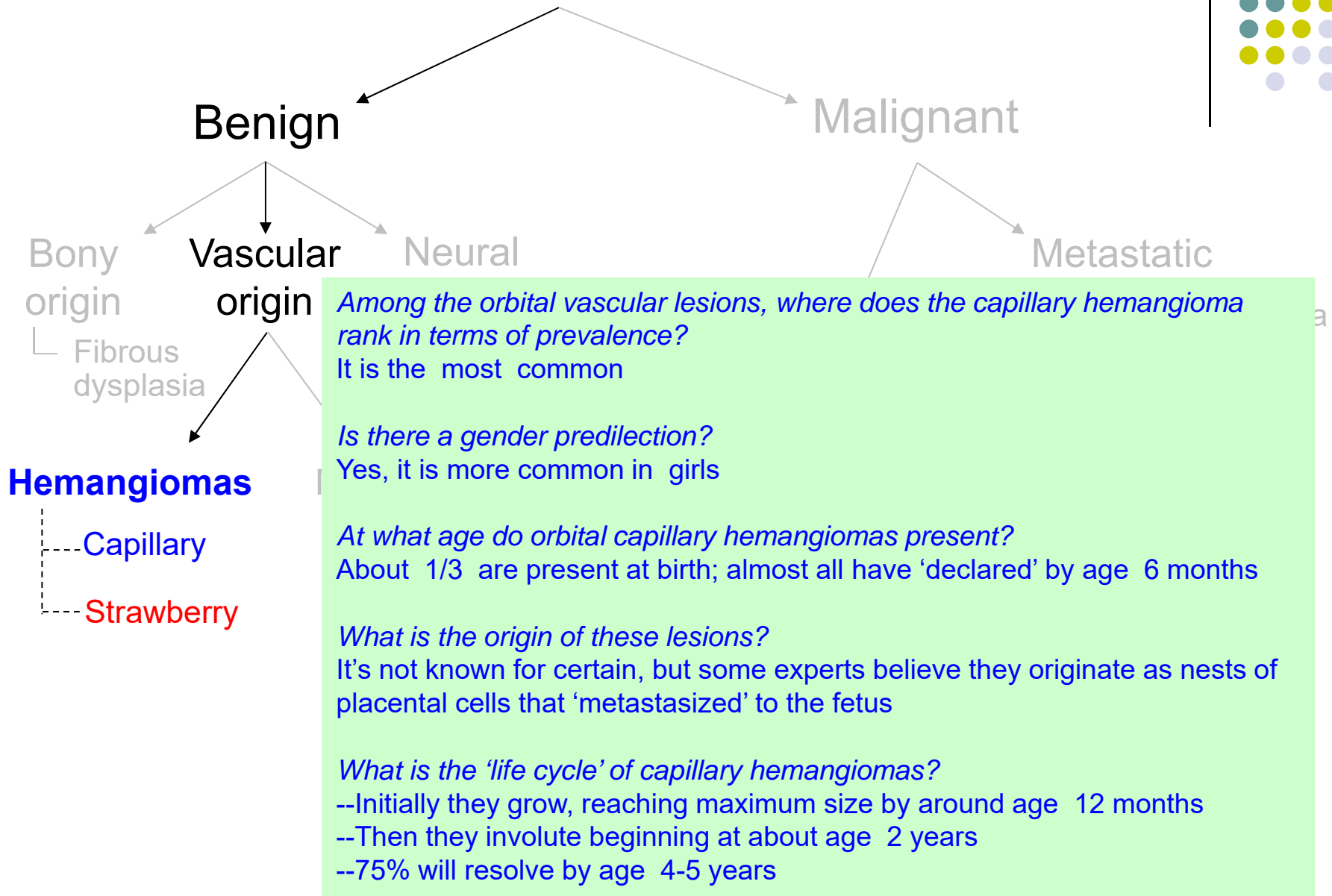


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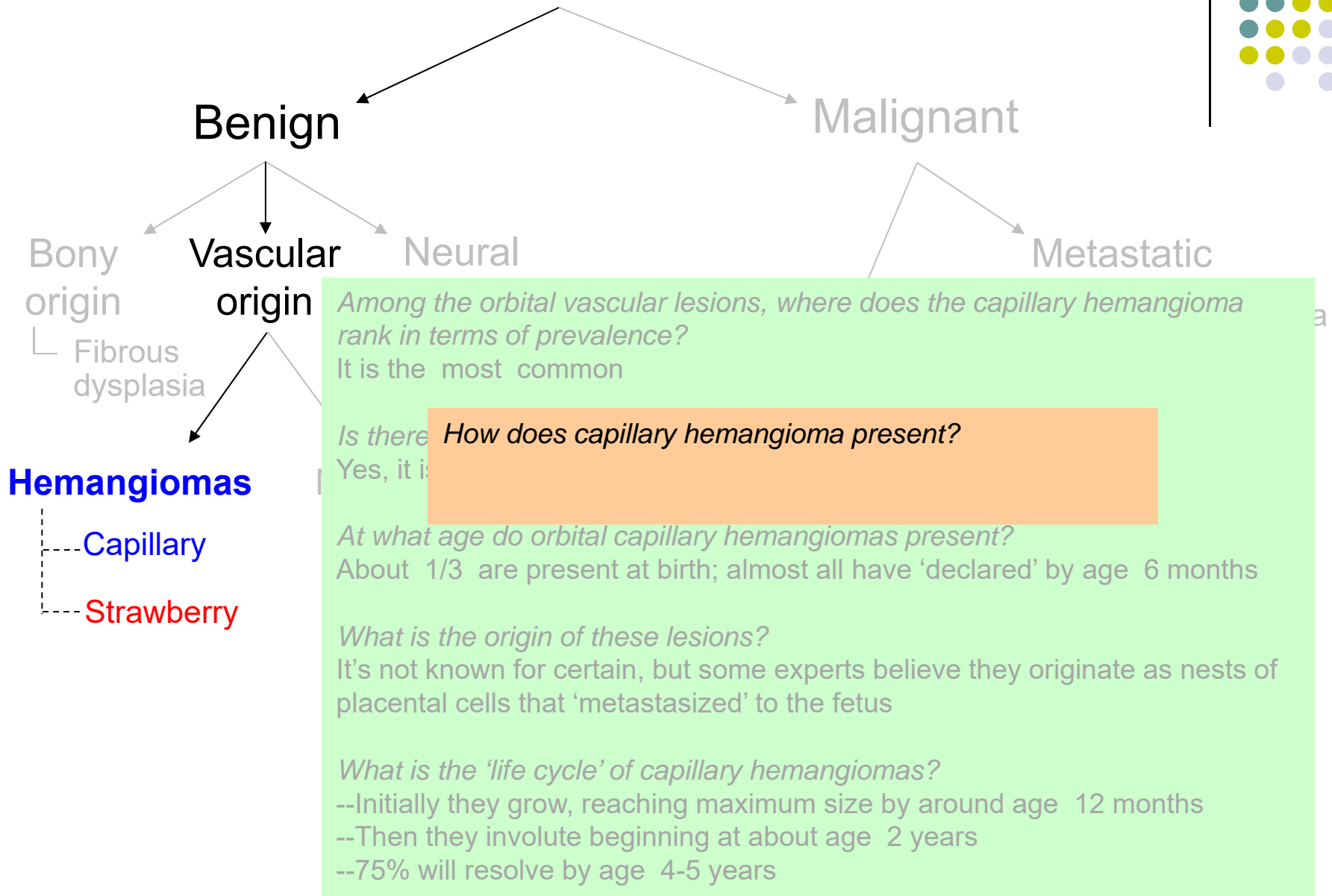


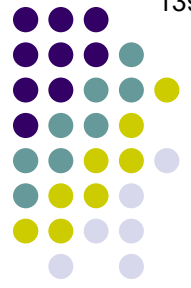
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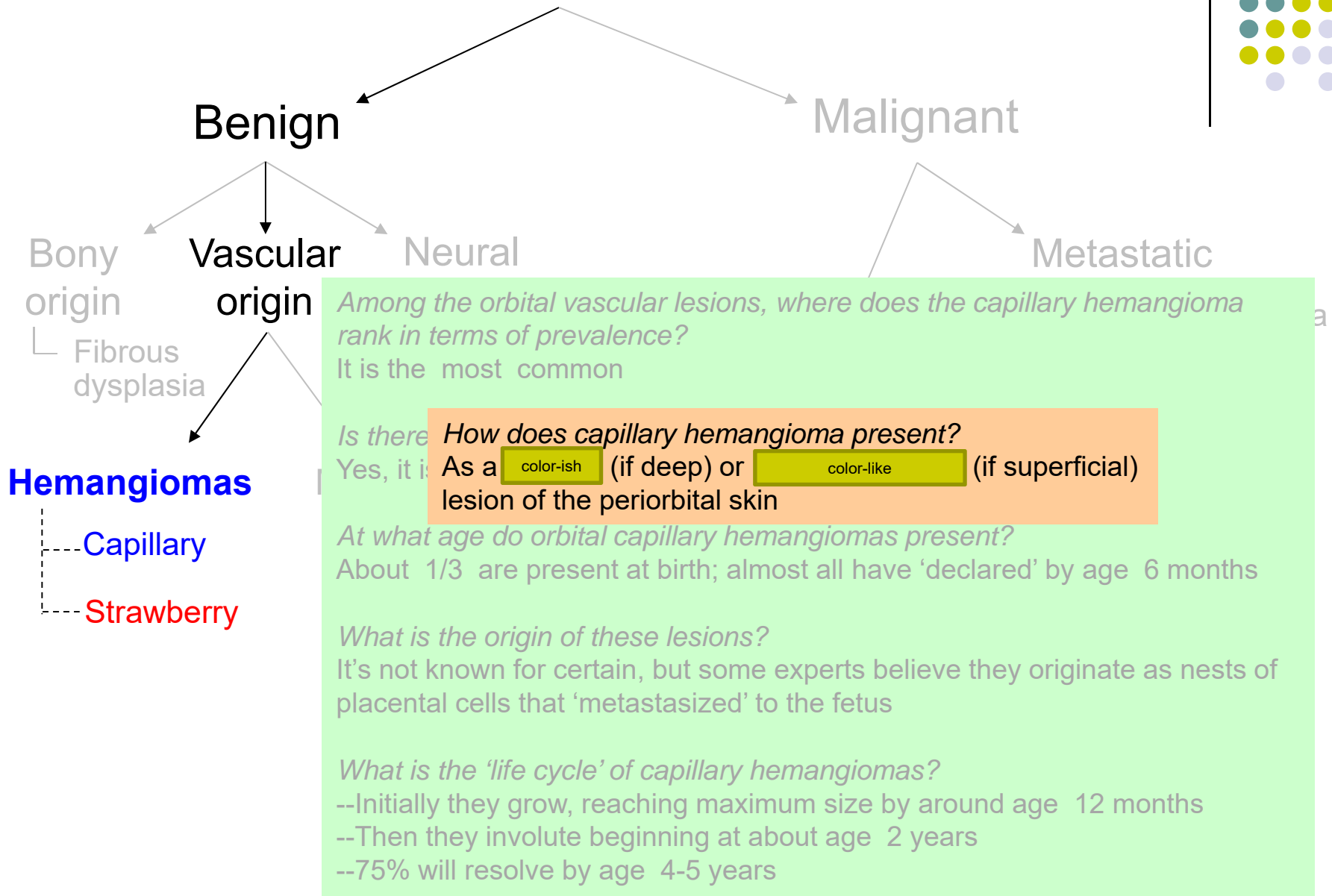


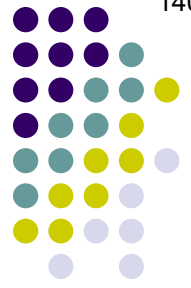
Pediatric Orbital Tumors



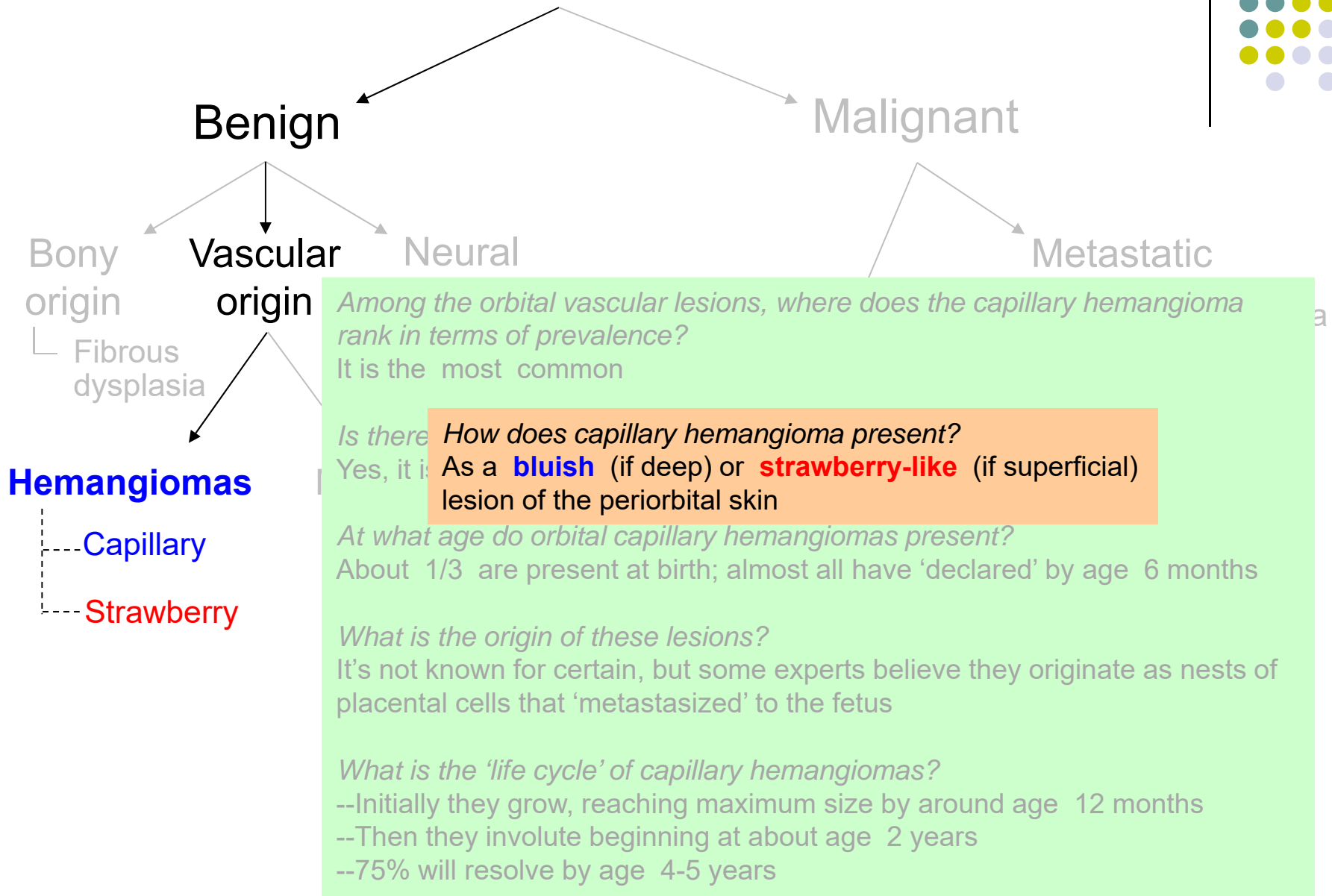


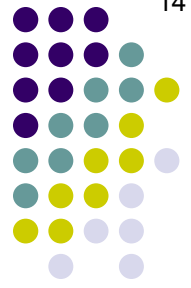
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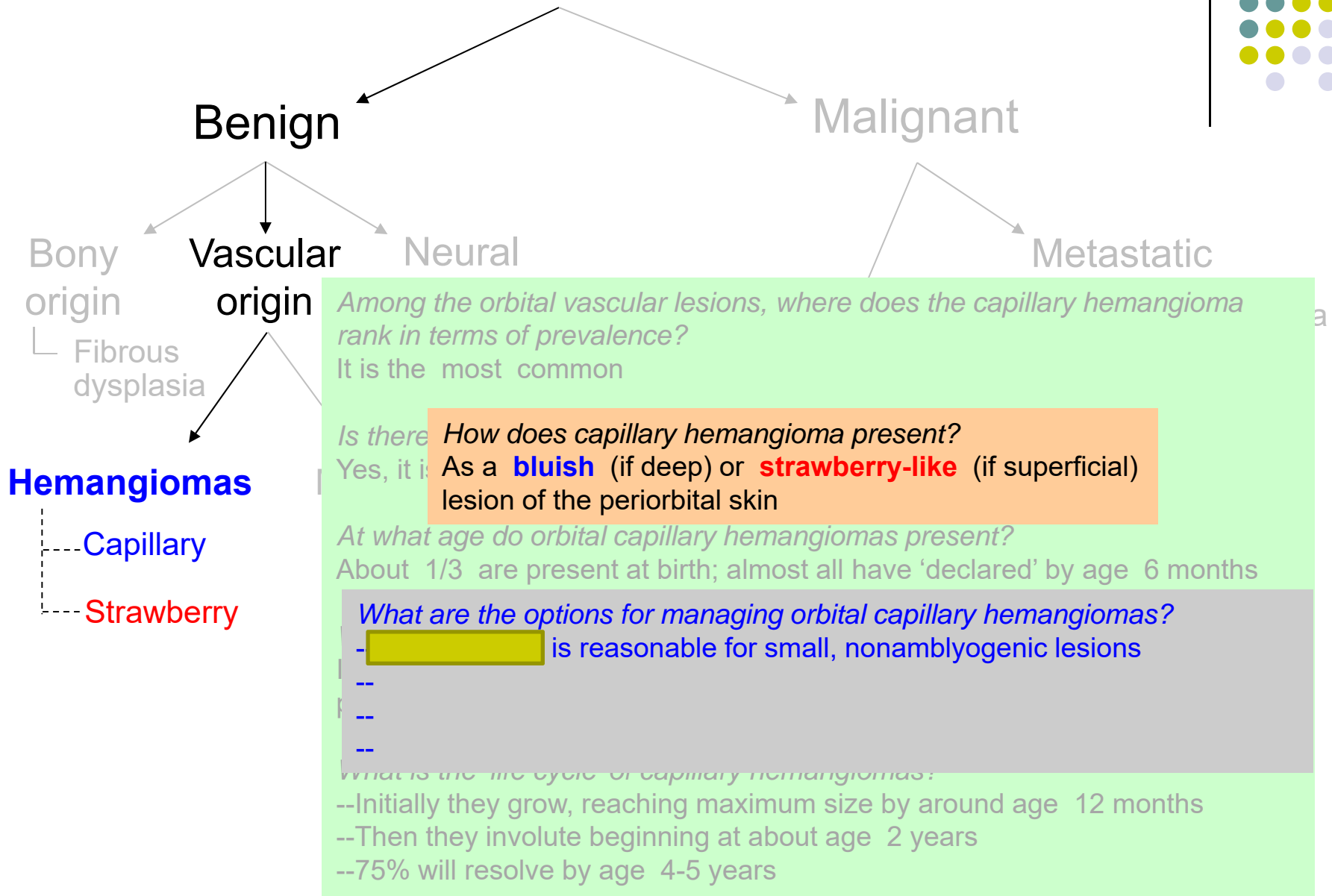


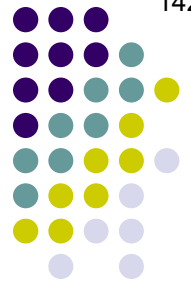
Pediatric Orbital Tumors



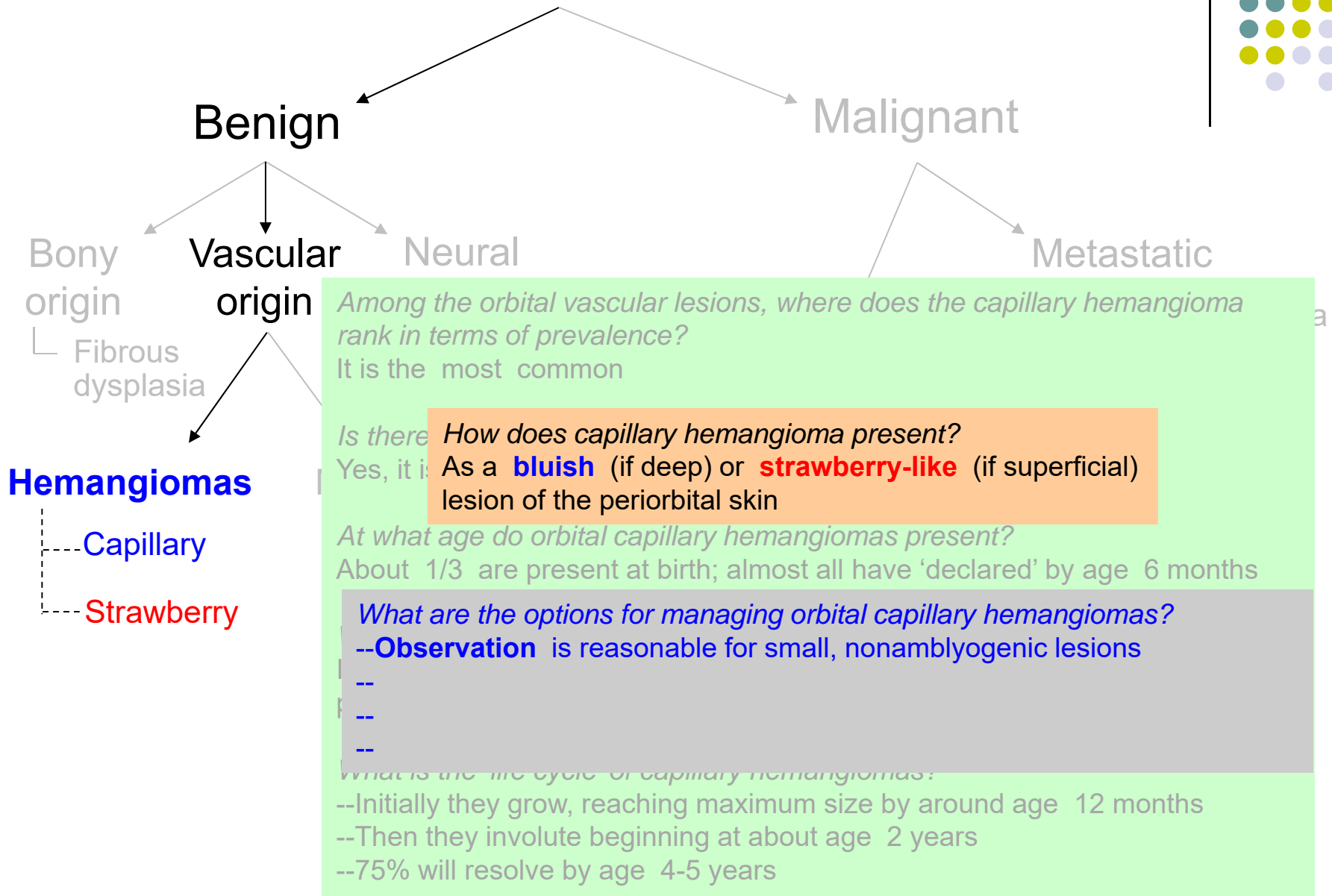


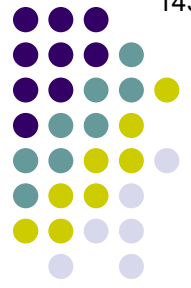
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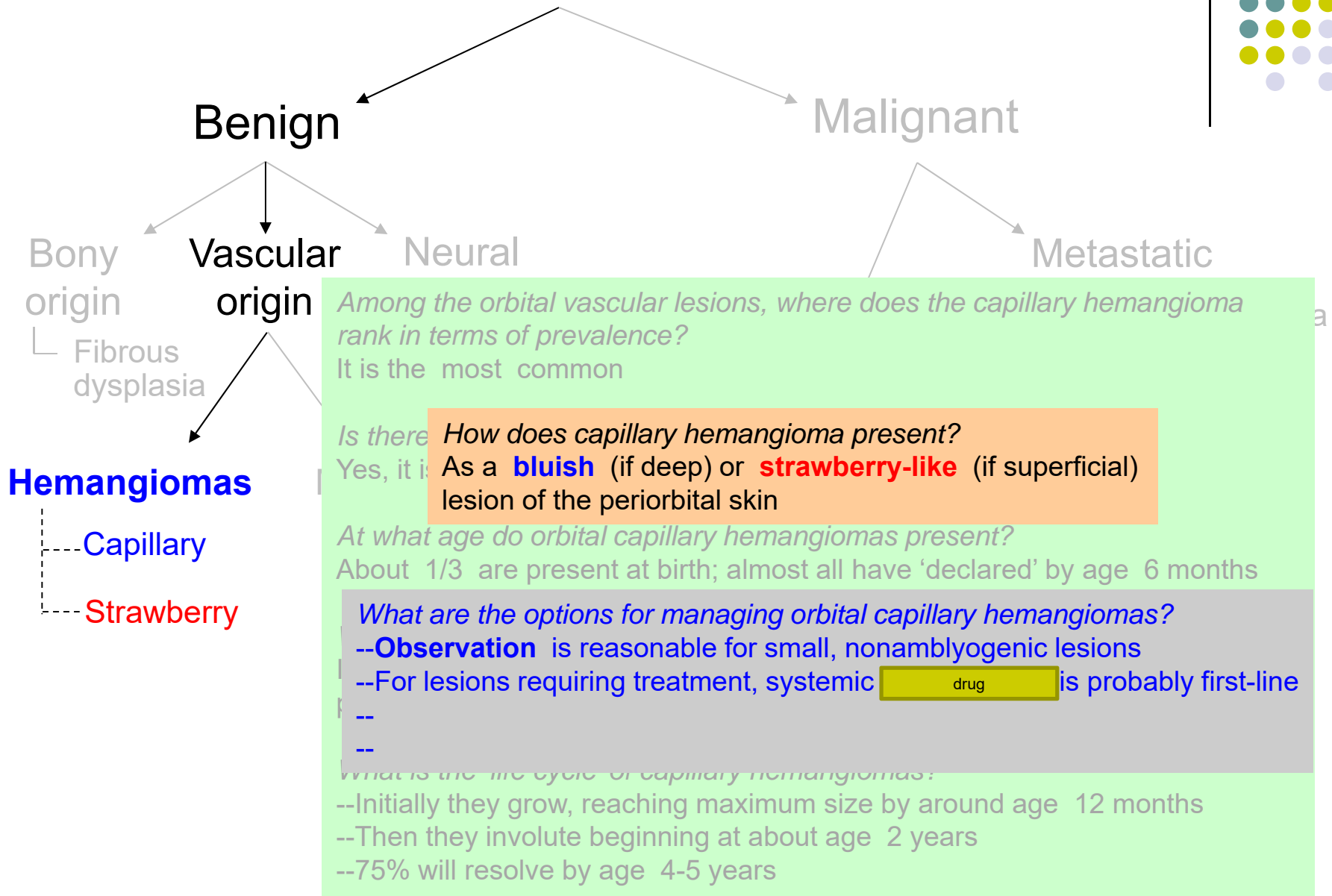


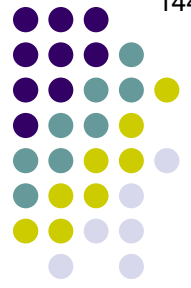
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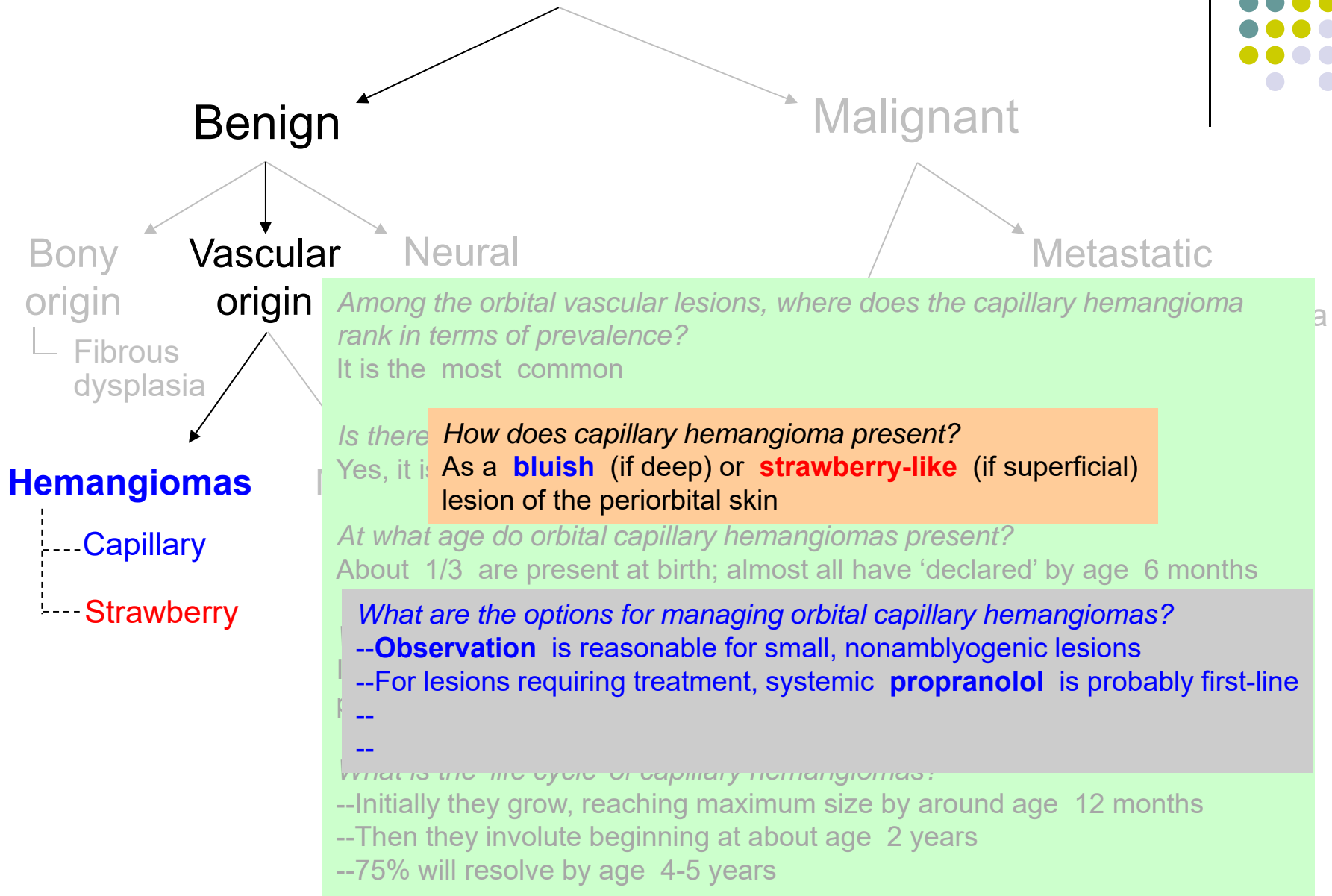


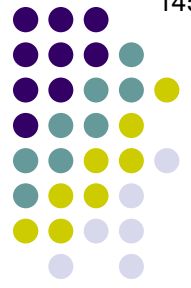
Pediatric Orbital Tumors



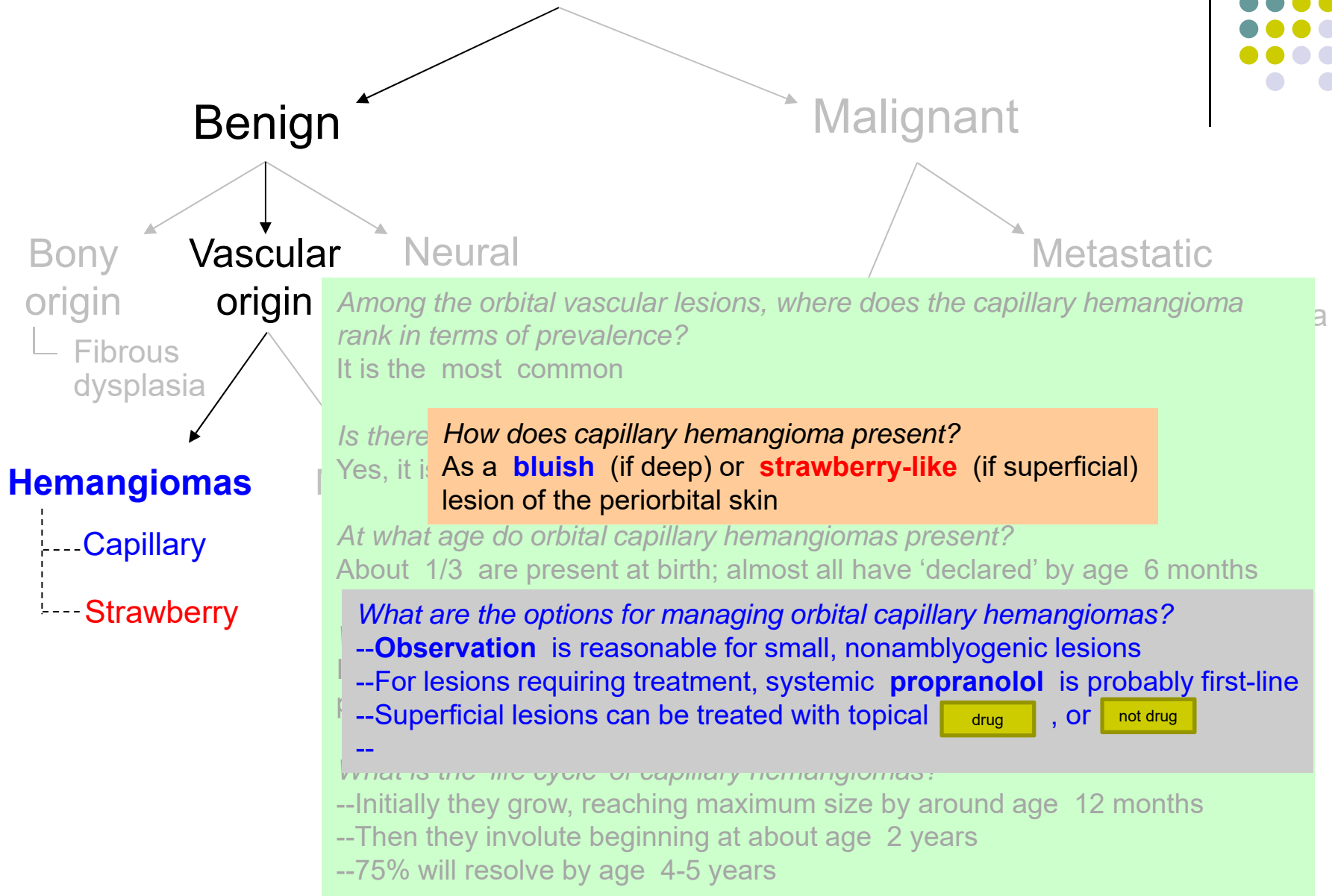


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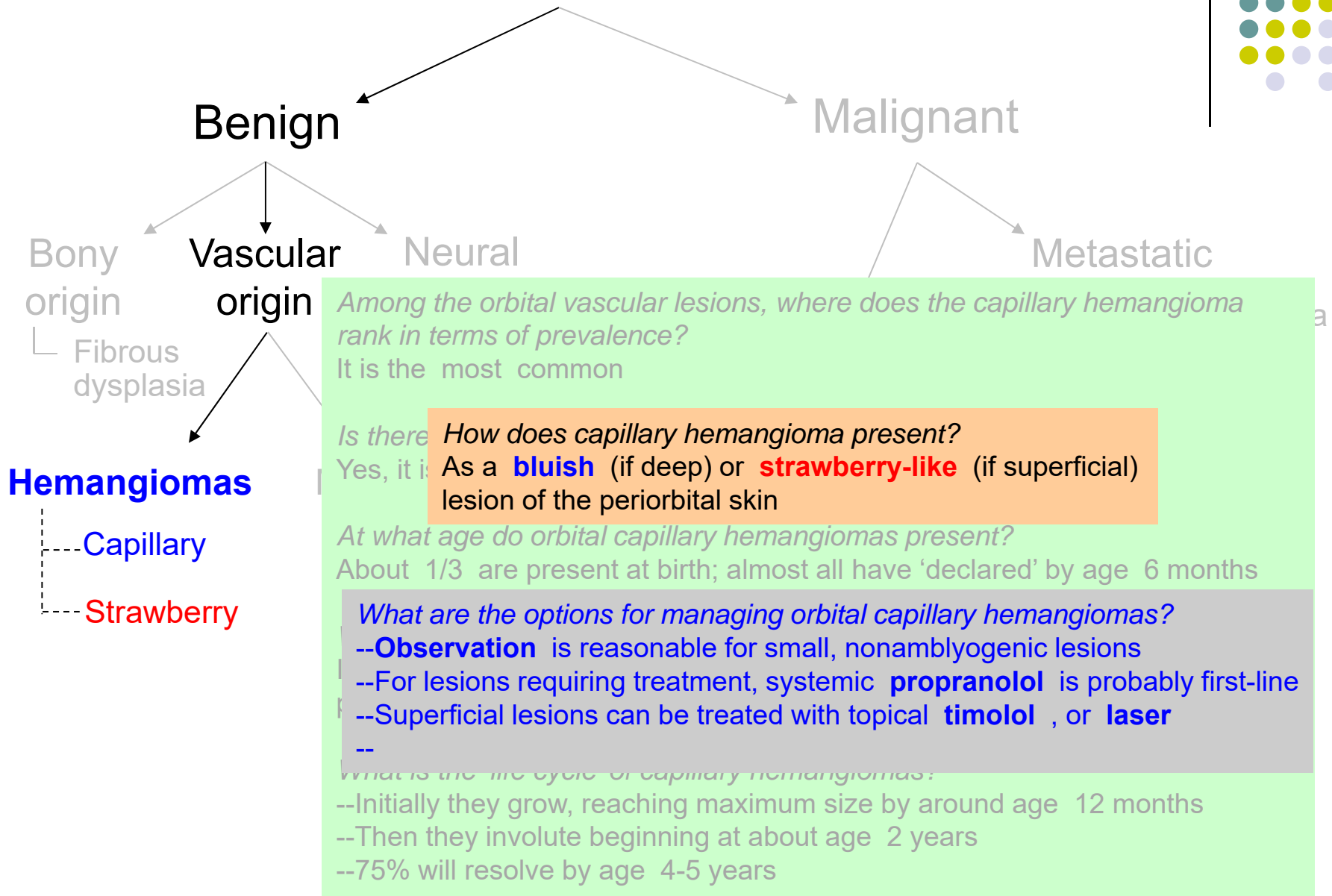


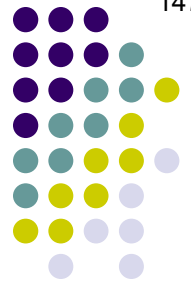
Pediatric Orbital Tumors



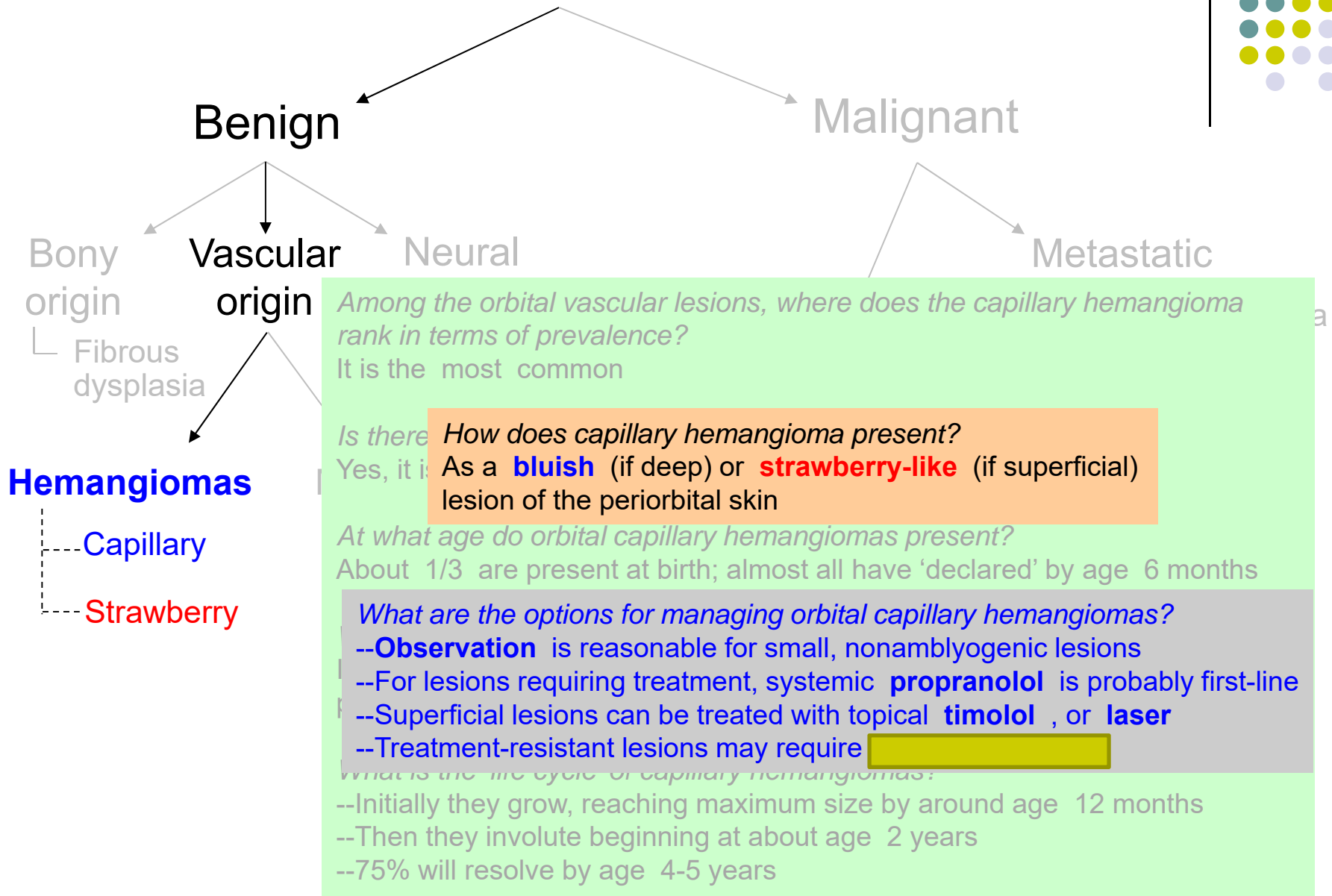


Pediatric Orbital Tumors



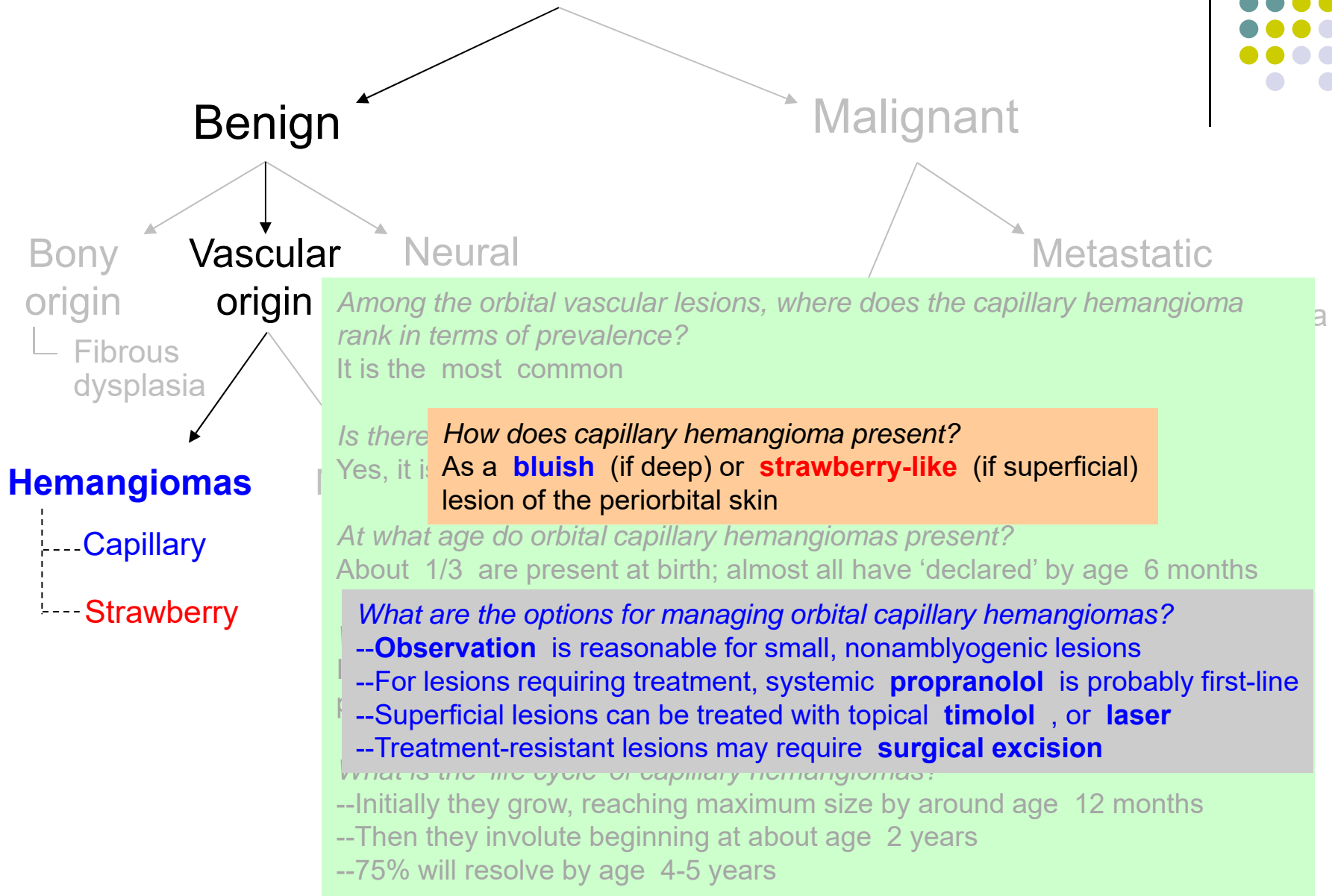


Pediatric Orbital Tumors



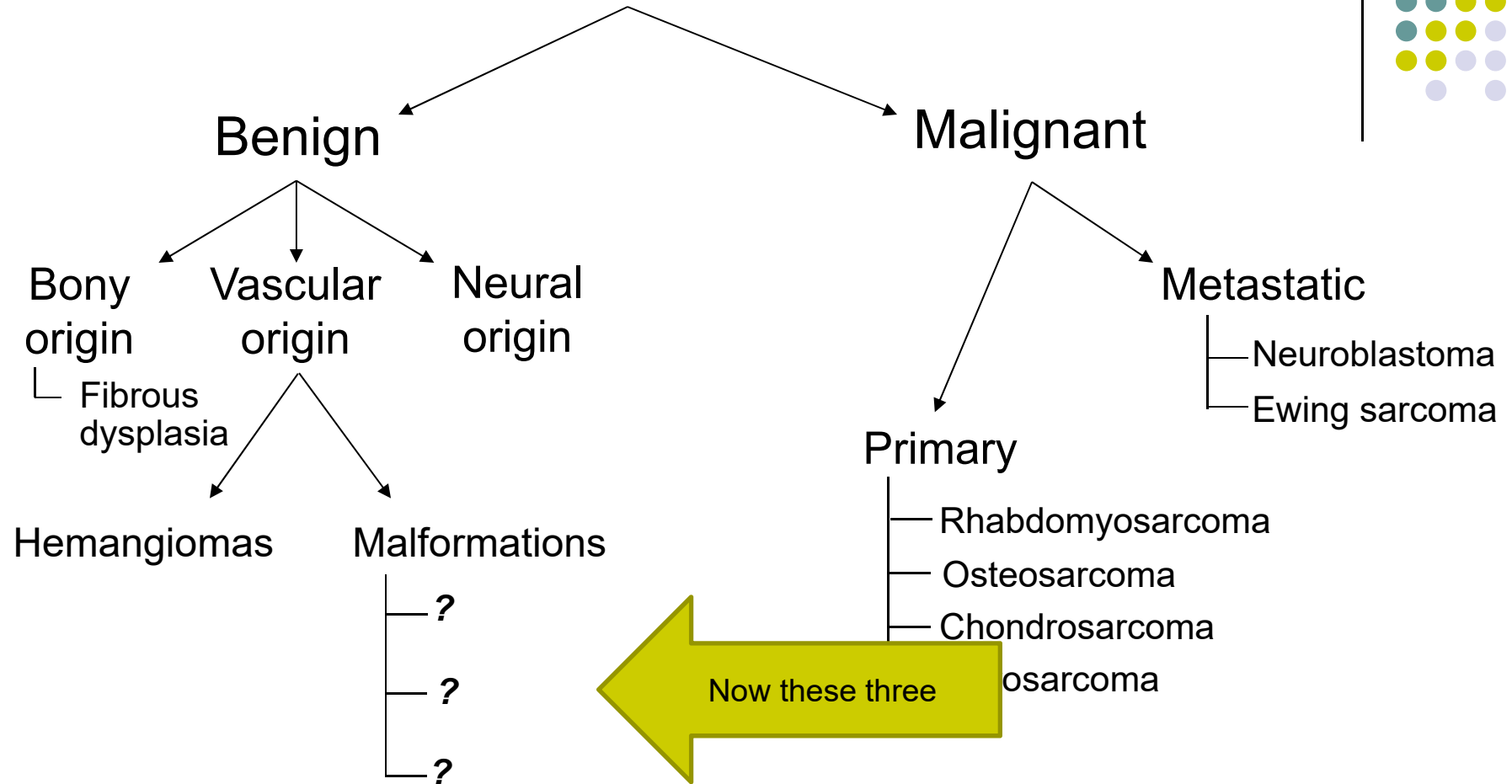


Pediatric Orbital Tumors



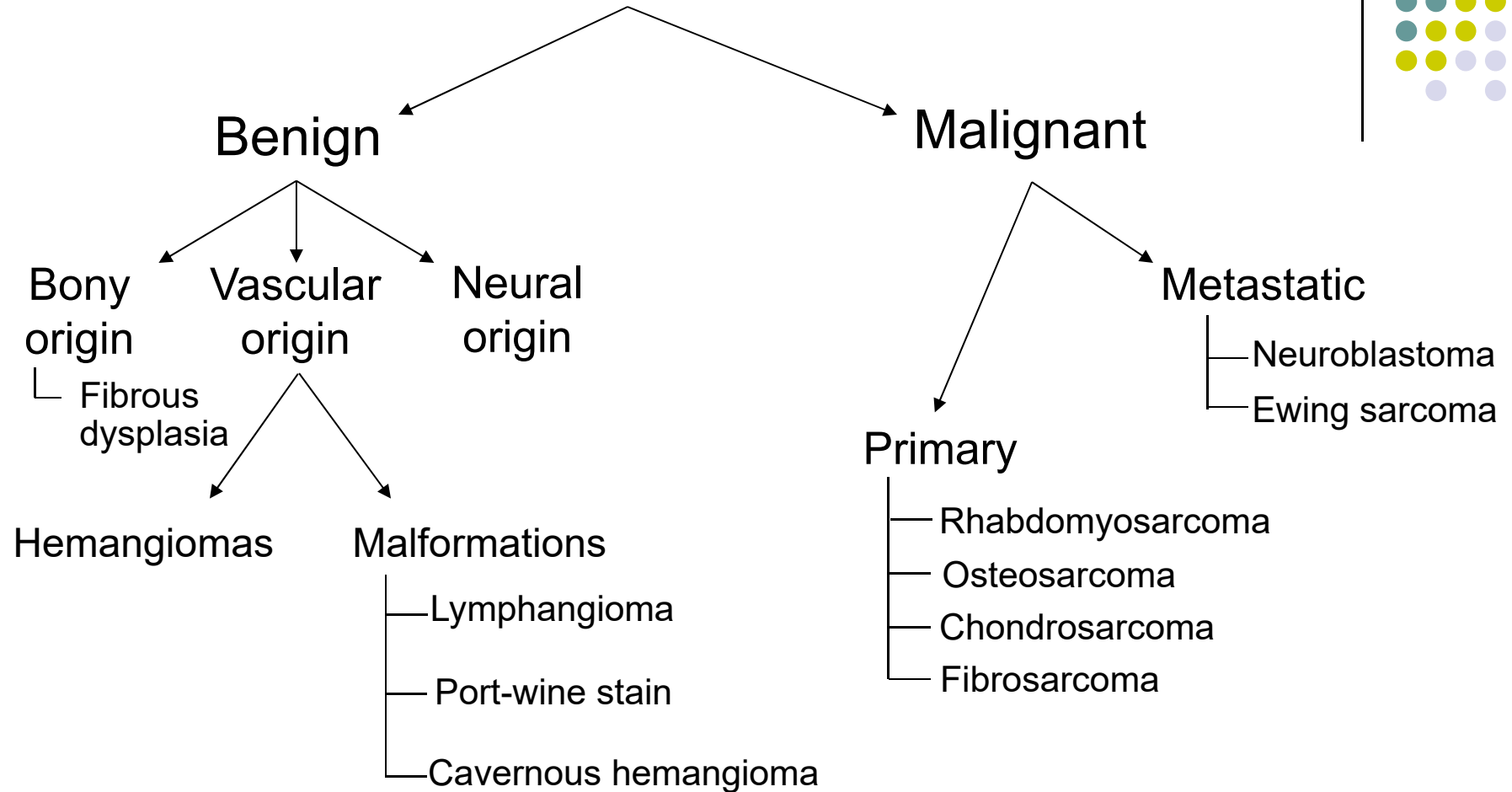


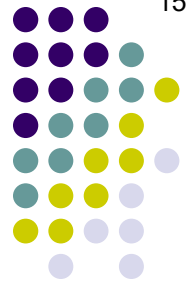
Pediatric Orbital Tumors



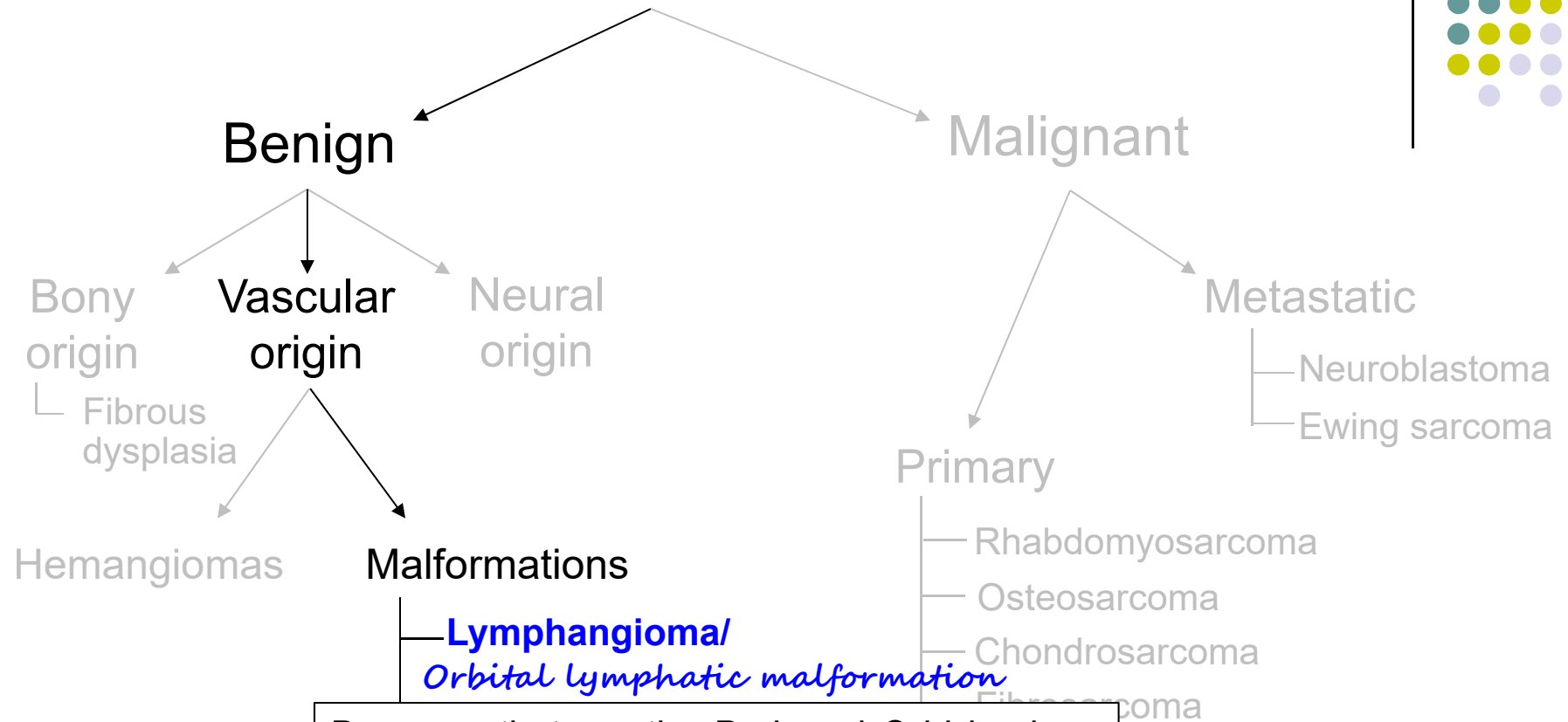


Pediatric Orbital Tumors

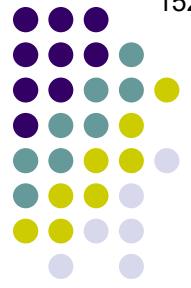




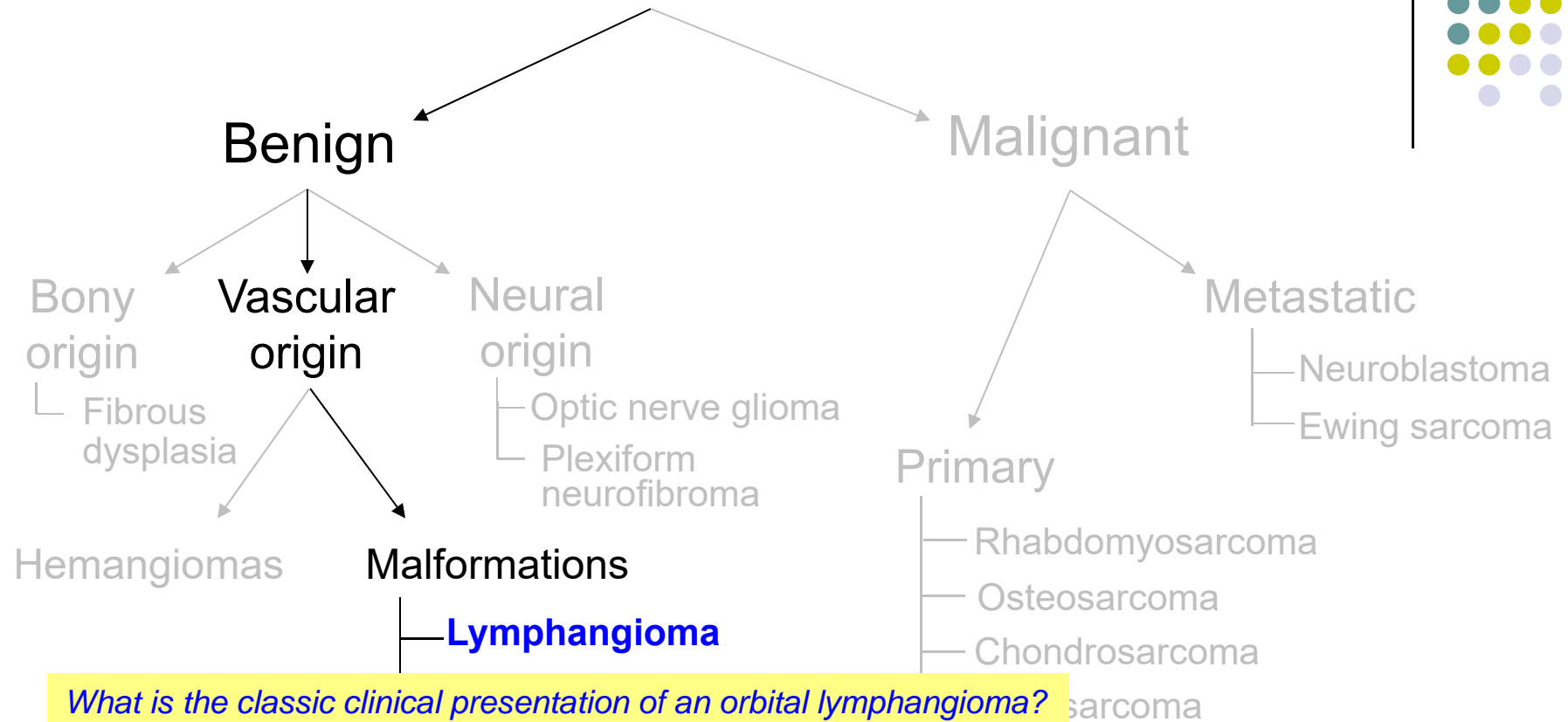
Pediatric Orbital Tumors



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



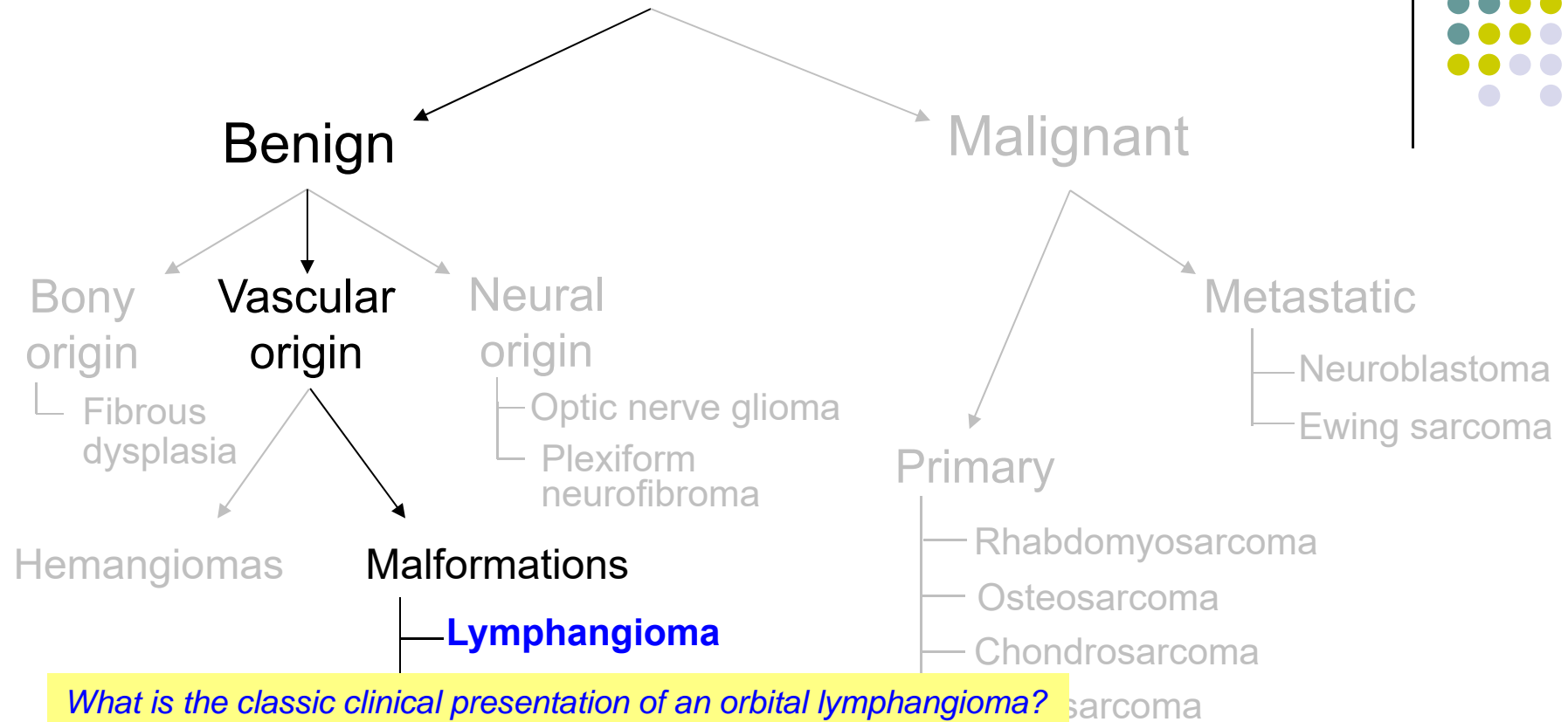
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What is the classic clinical presentation of an orbital lymphangioma?



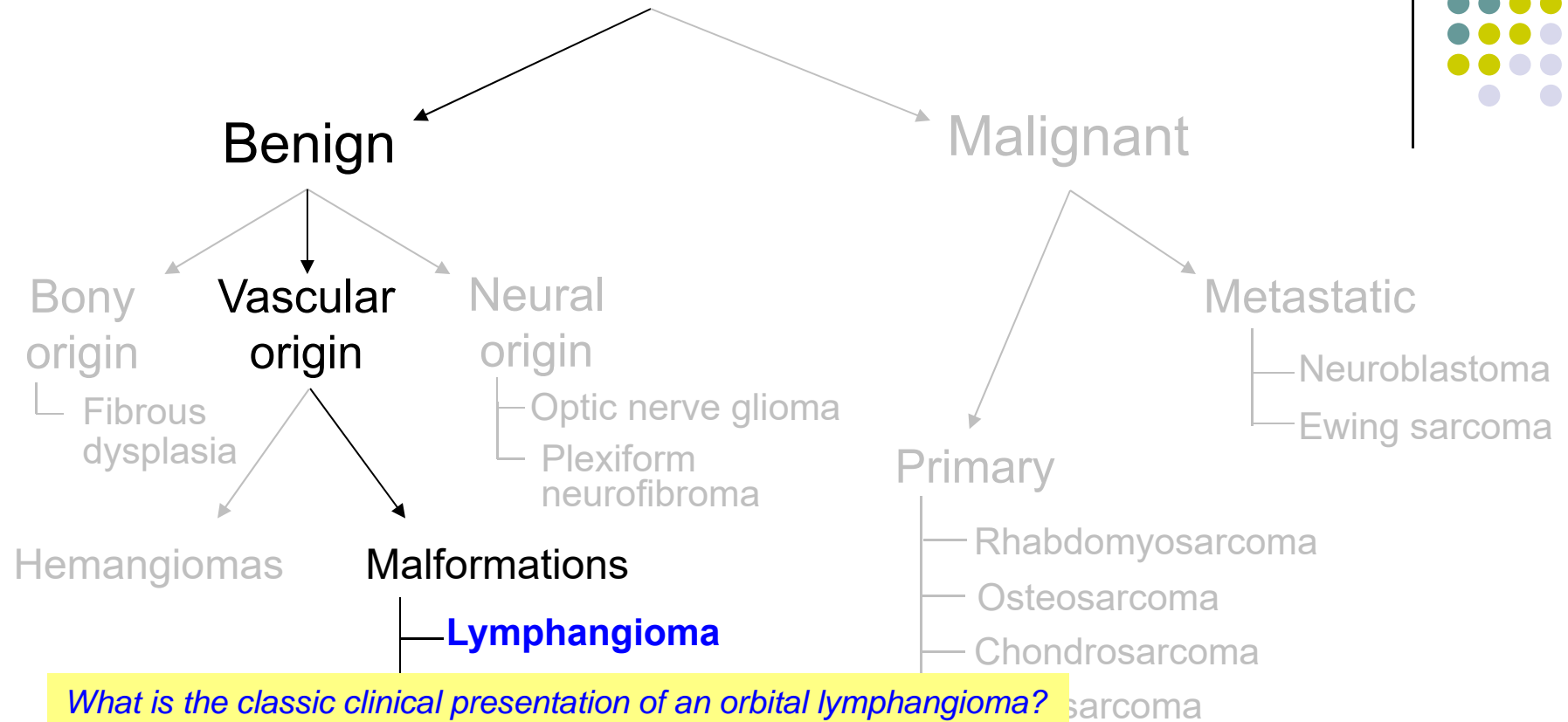
Pediatric Orbital Tumors



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



Pediatric Orbital Tumors

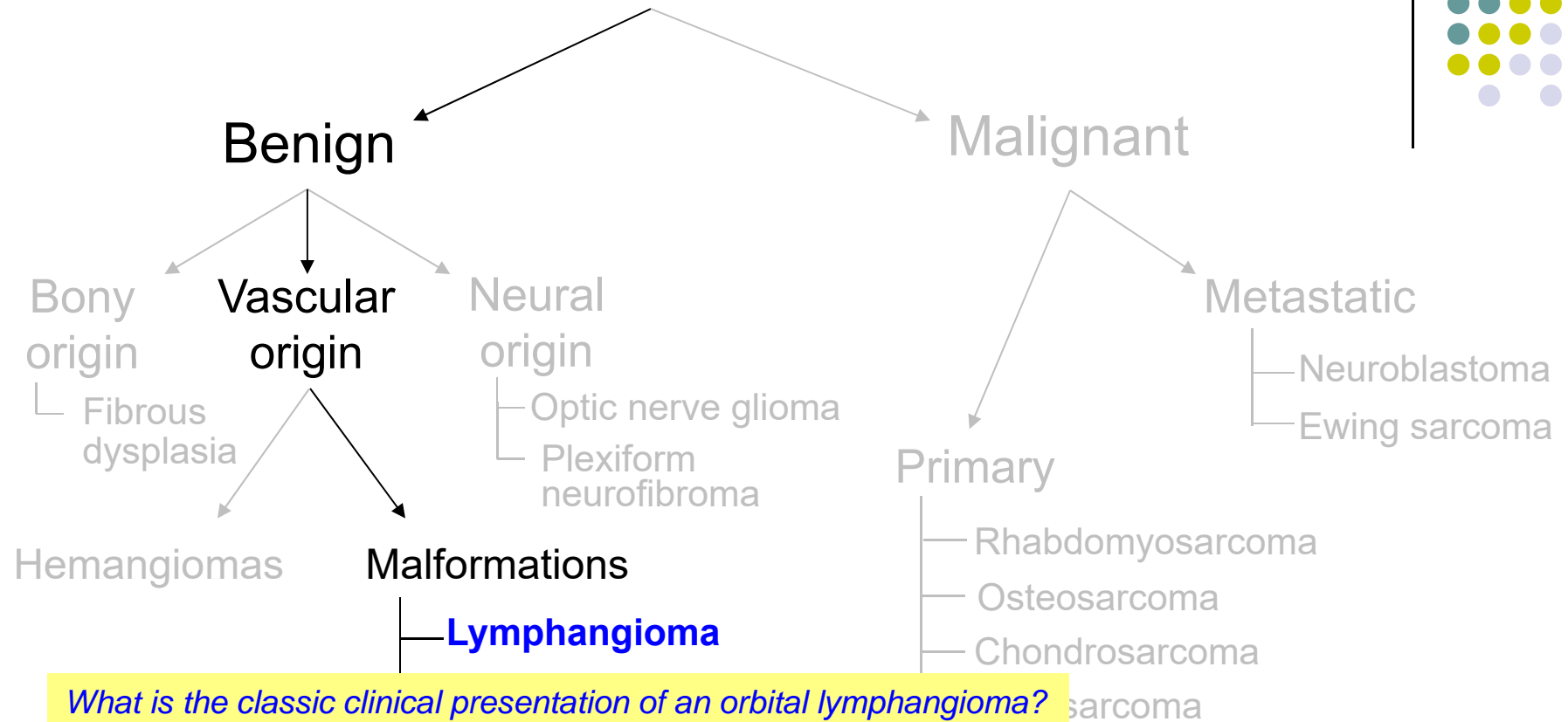


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What does histologic examination of a lymphangioma reveal?



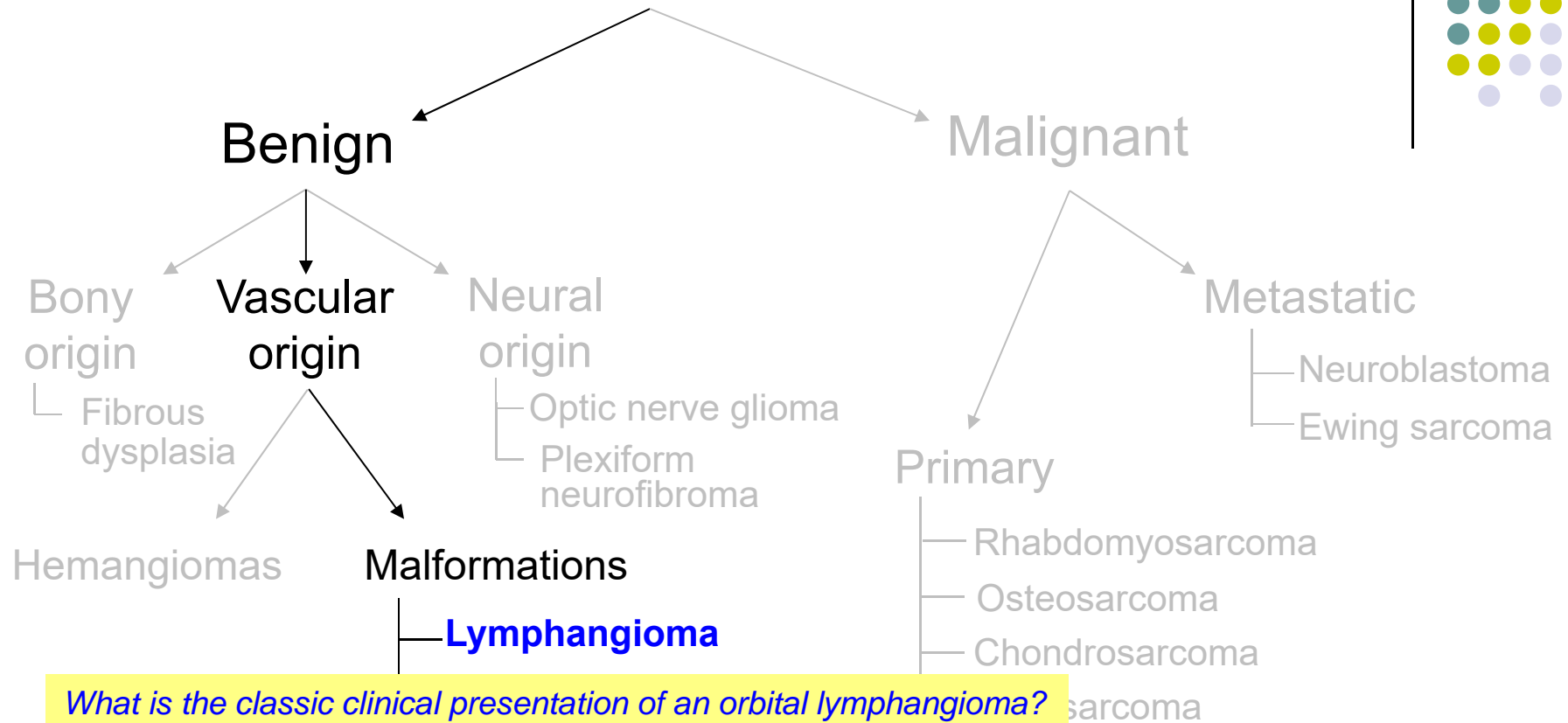
Pediatric Orbital Tumors



What is the classic clinical presentation of an orbital lymphangioma?
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What does histologic examination of a lymphangioma reveal?
 Lymphatic-type endothelial cells lining lymph-filled spaces

Pediatric Orbital Tumors



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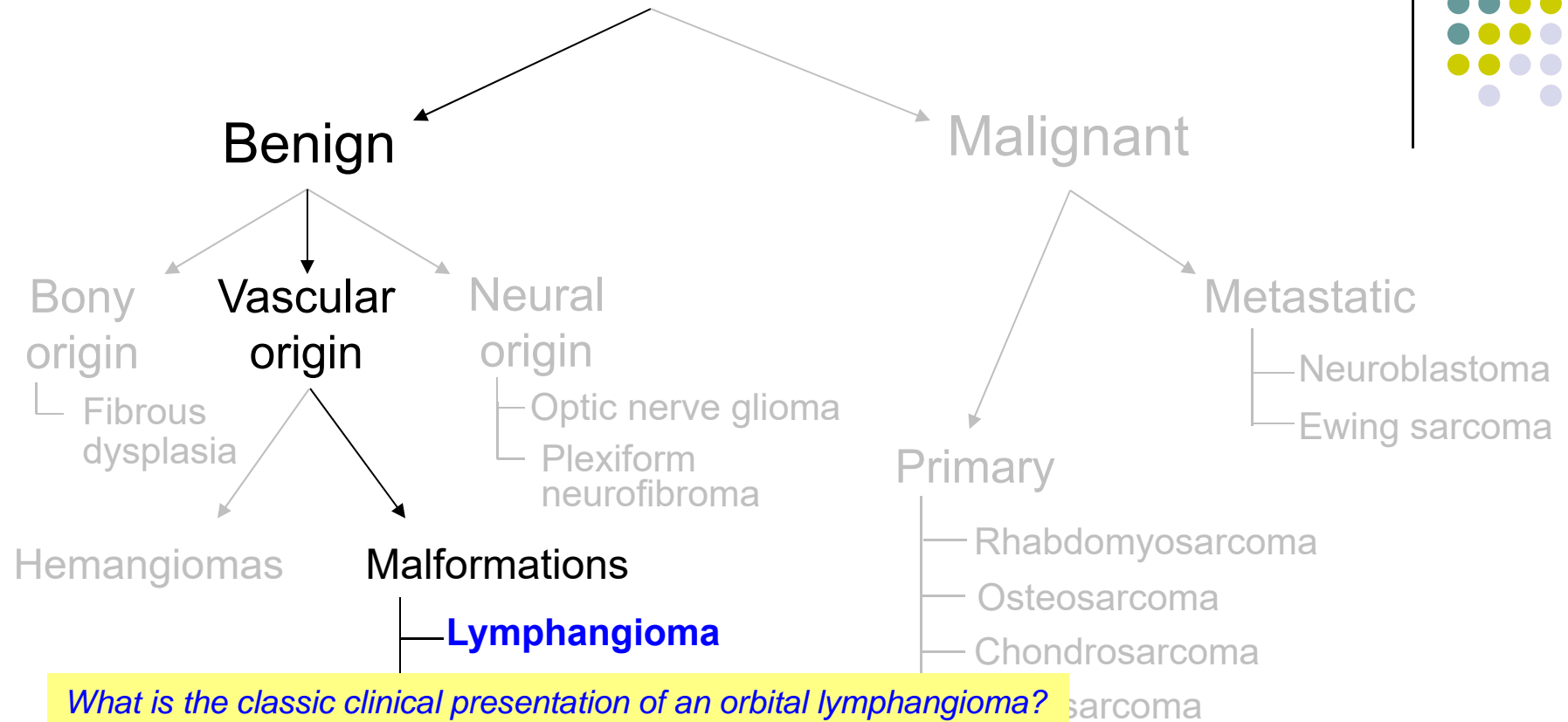
Lymphatic-type endothelial cells lining lymph-filled spaces

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



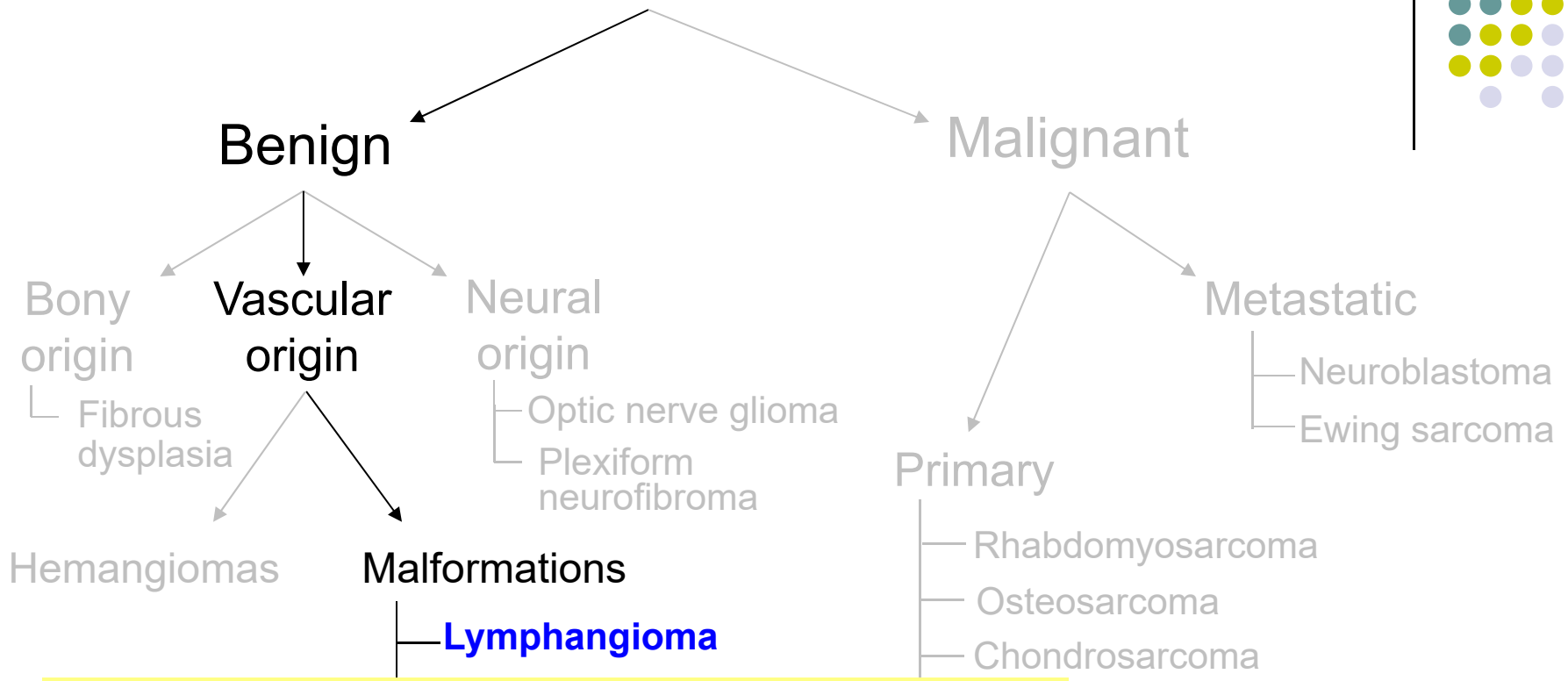
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 --Lymphoid hyperplasia
 --Intralesional hemorrhage



Pediatric Orbital Tumors



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 Rapid-onset proptosis

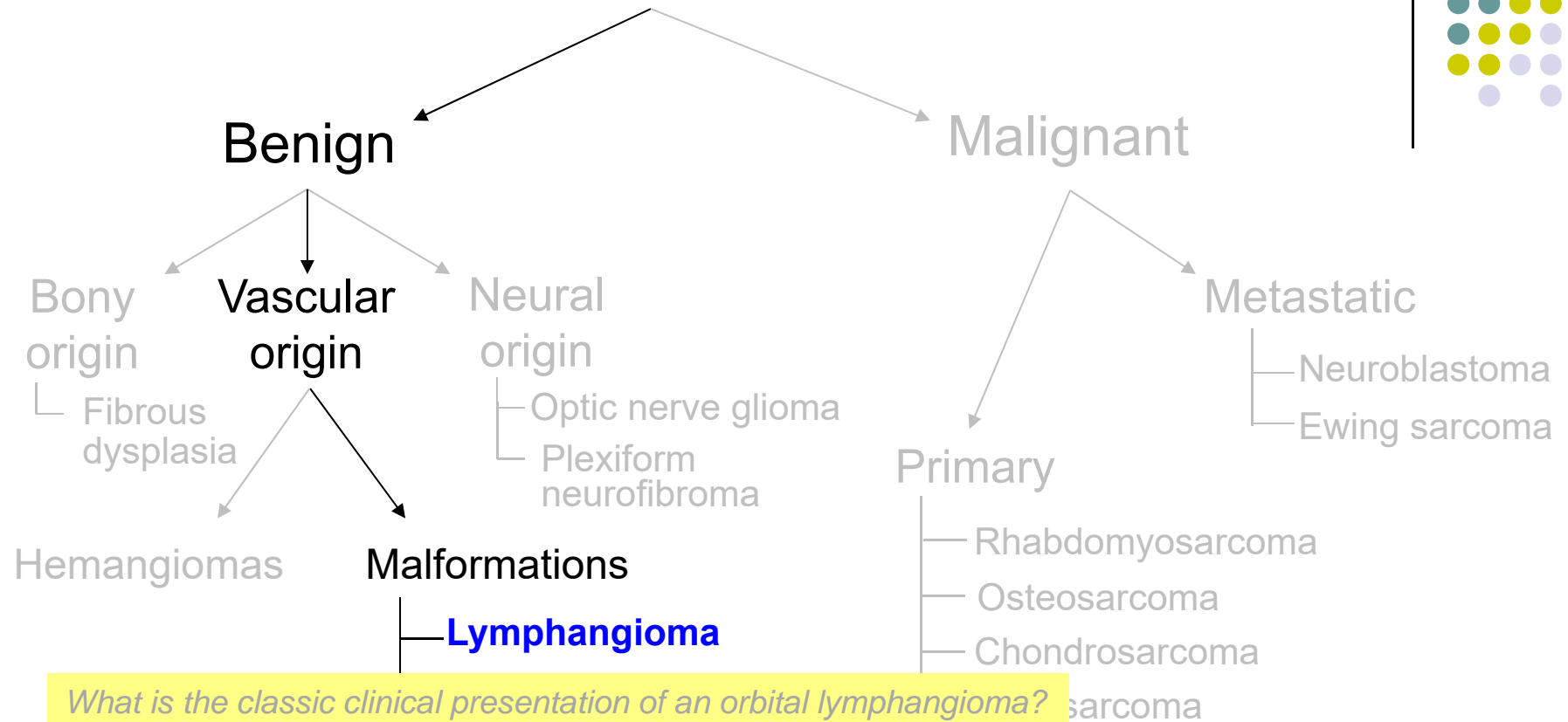
What does histologic examination of a lymphangioma reveal?
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What is the classic health event for precipitating lymphoid hyperplasia in these patients?

--**Lymphoid hyperplasia**
 --Intralesional hemorrhage



Pediatric Orbital Tumors



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

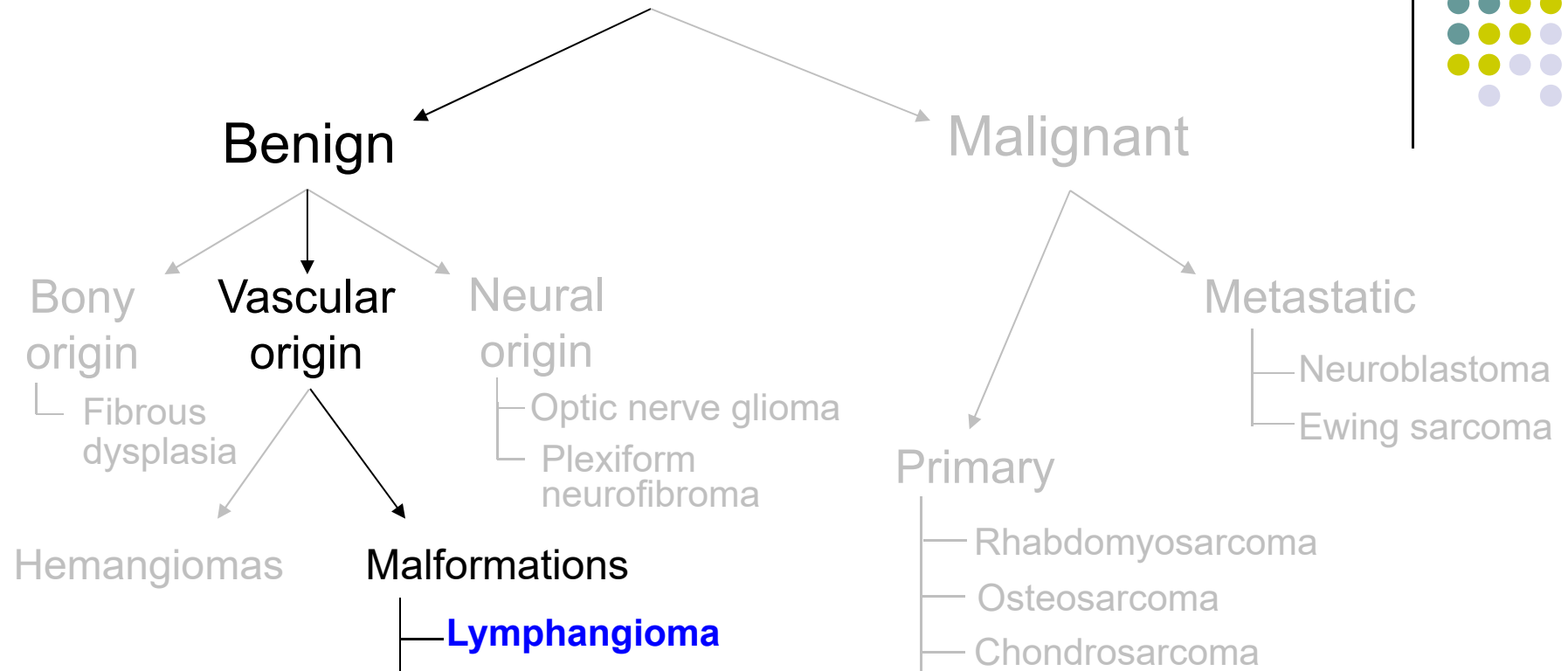
What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

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

--**Lymphoid hyperplasia**
--Intralesional hemorrhage



Pediatric Orbital Tumors



What is the classic clinical presentation of an orbital lymphangioma?

Rapid-onset

How are lymphangiomas managed?

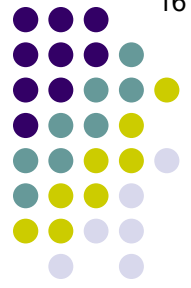
What does

Lymphatic-type endothelial cells lining lymph-filled spaces

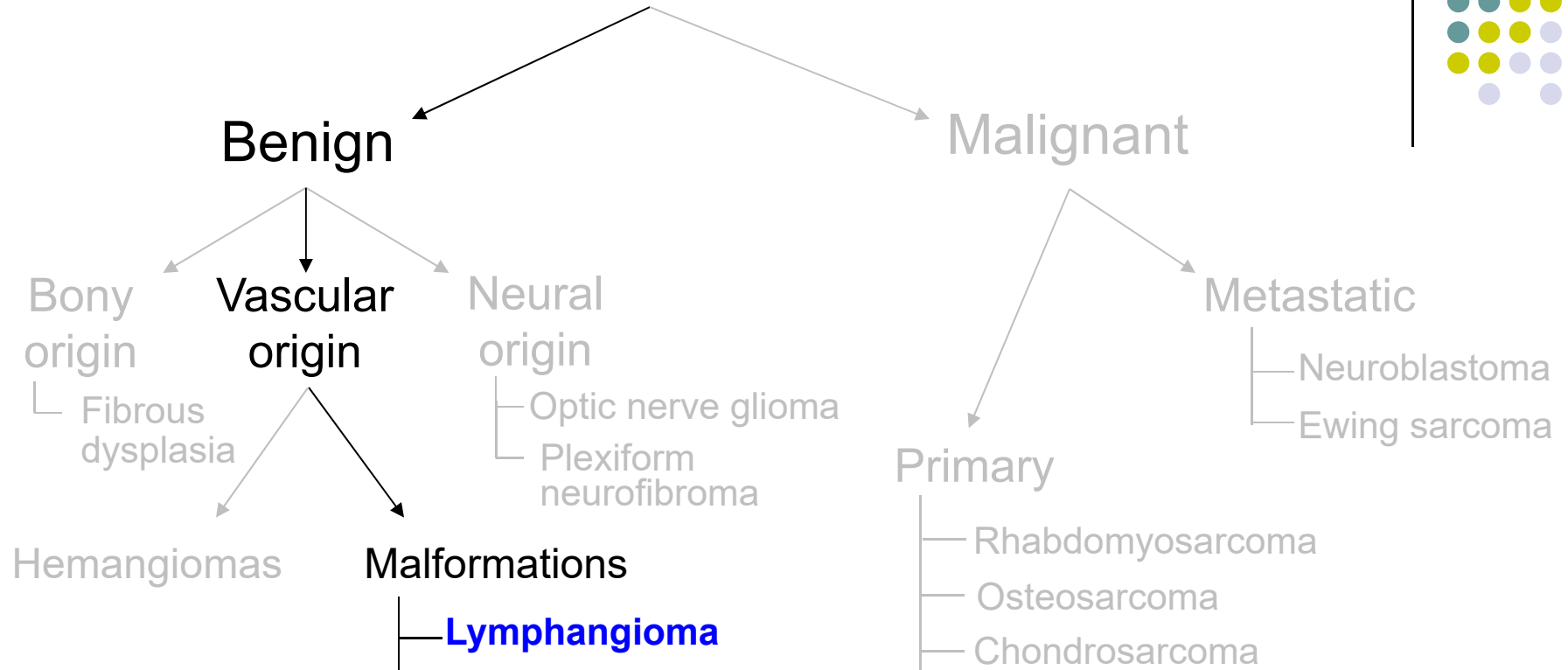
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Pediatric Orbital Tumors



What is the classic clinical presentation of an orbital lymphangioma? Proptosis

Rapid-onset

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

What does

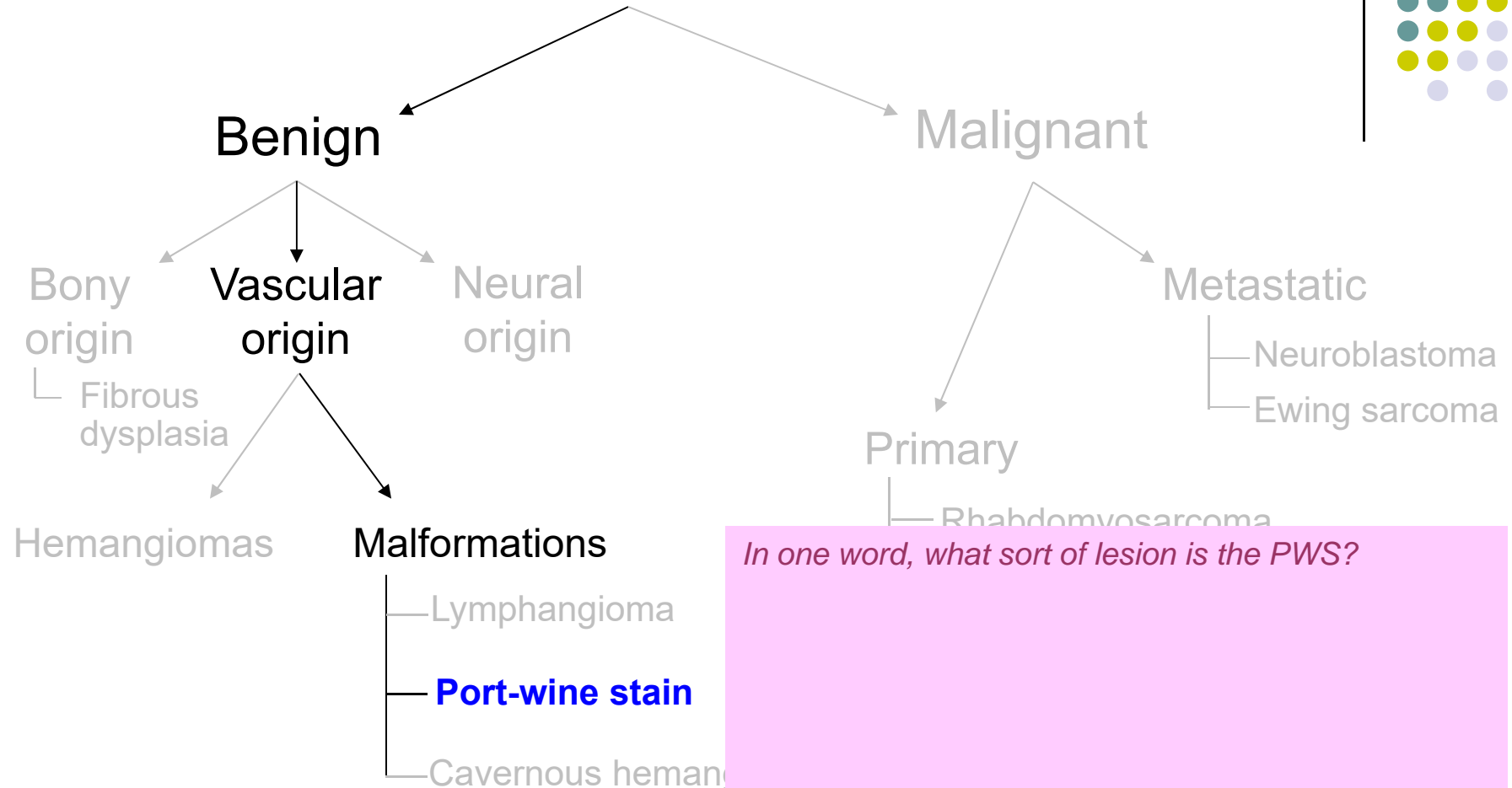
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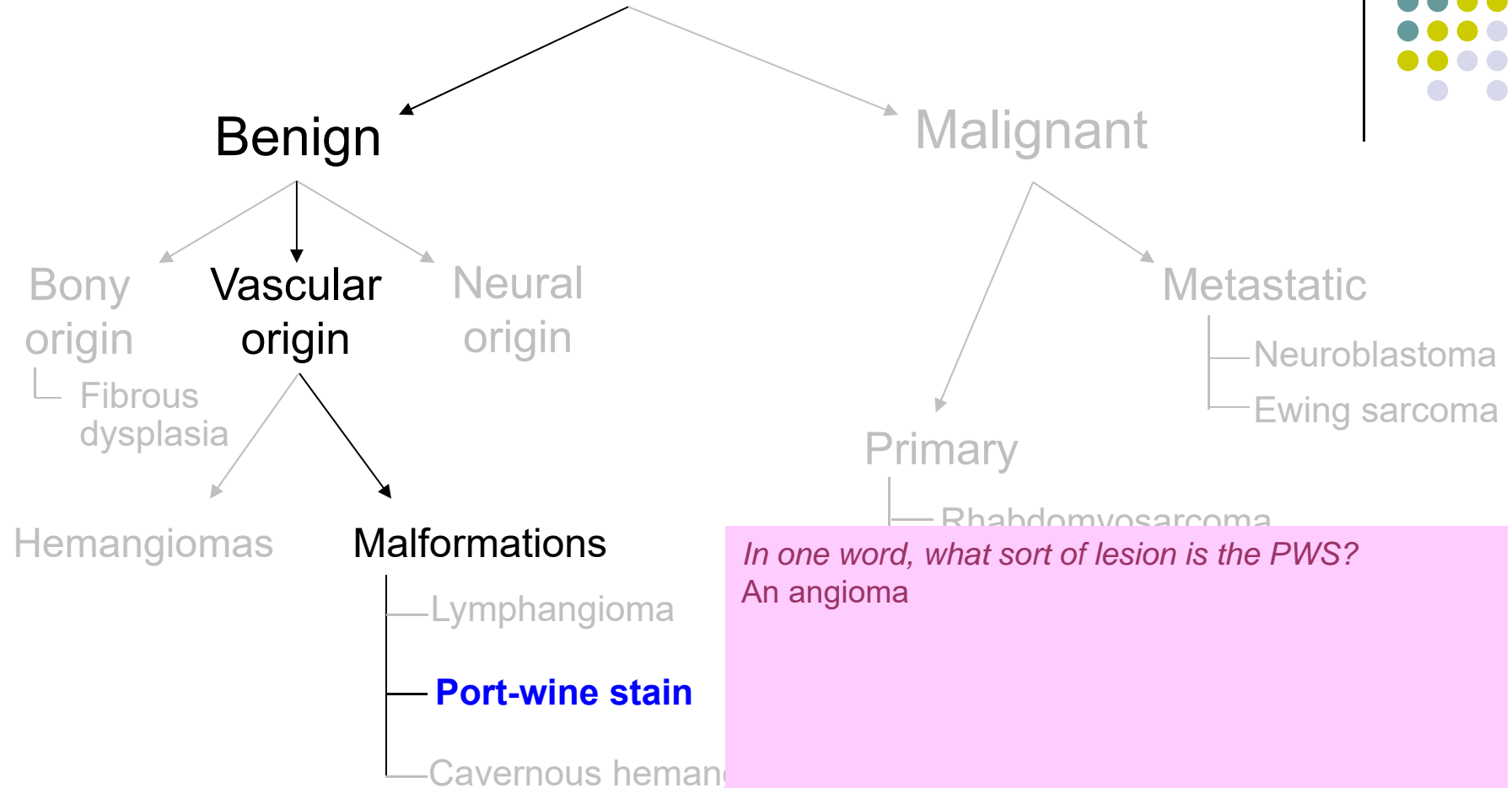
Pediatric Orbital Tumors



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



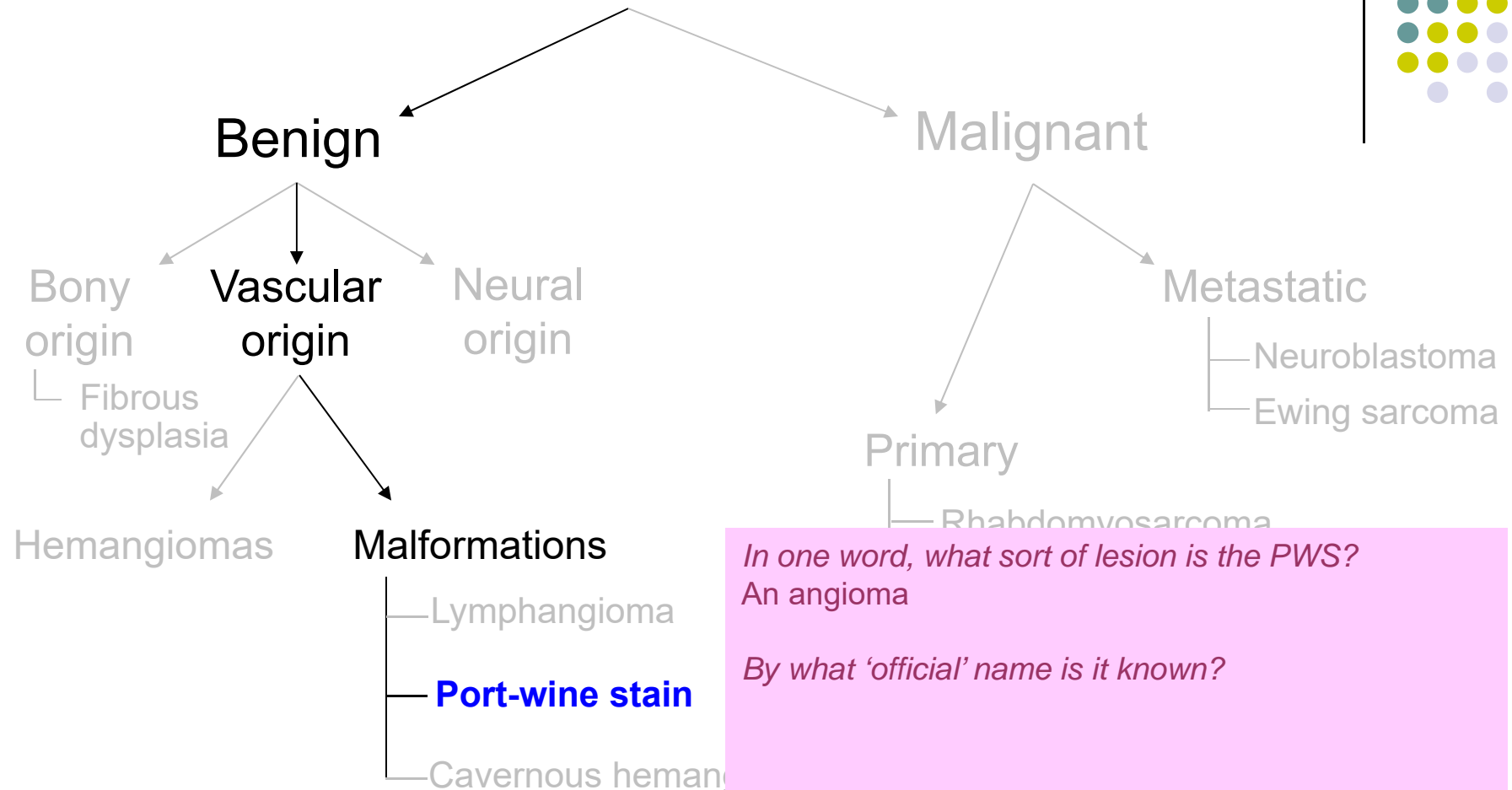
Pediatric Orbital Tumors



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



Pediatric Orbital Tumors

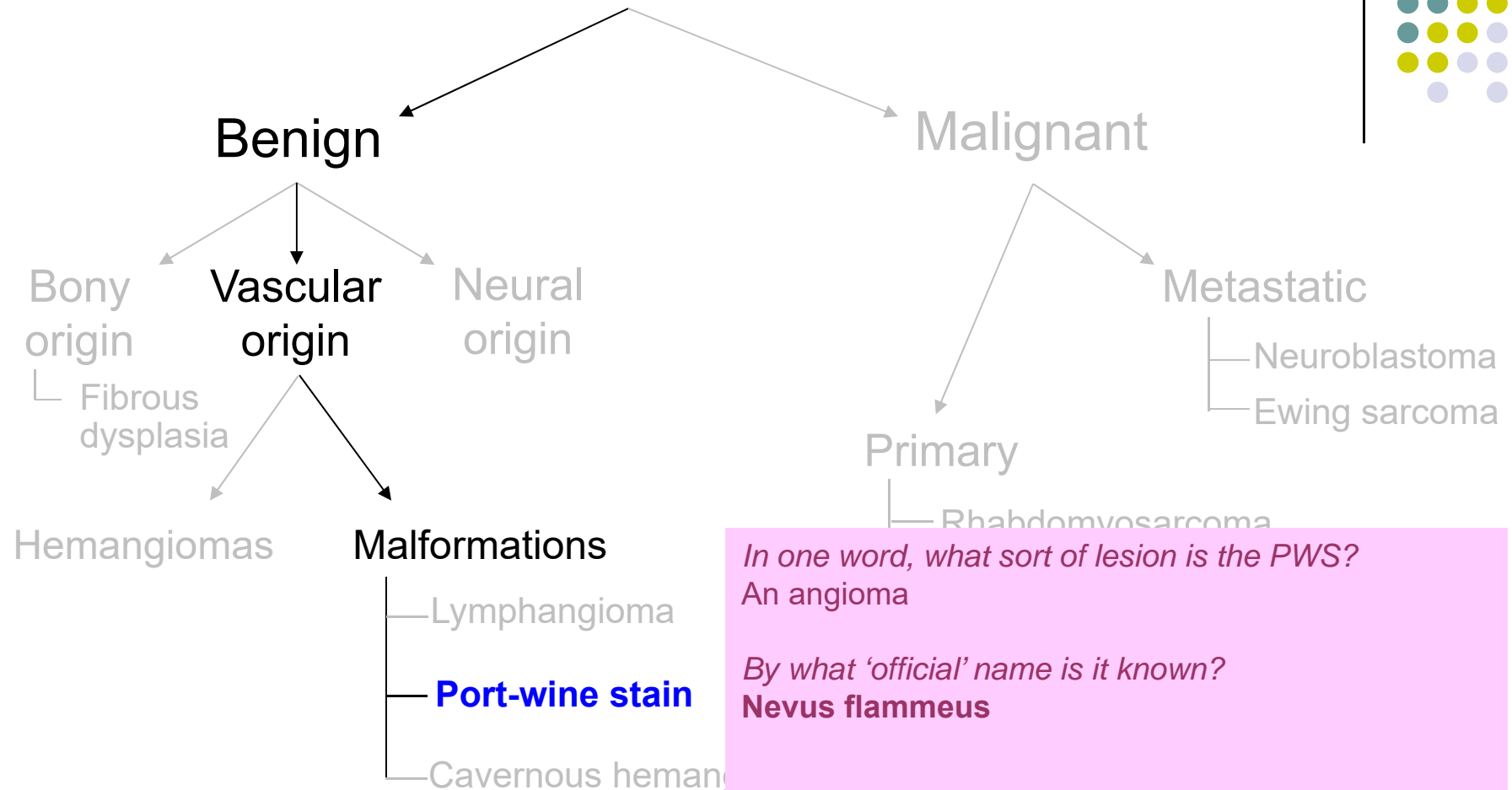


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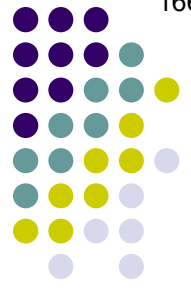


Pediatric Orbital Tumors

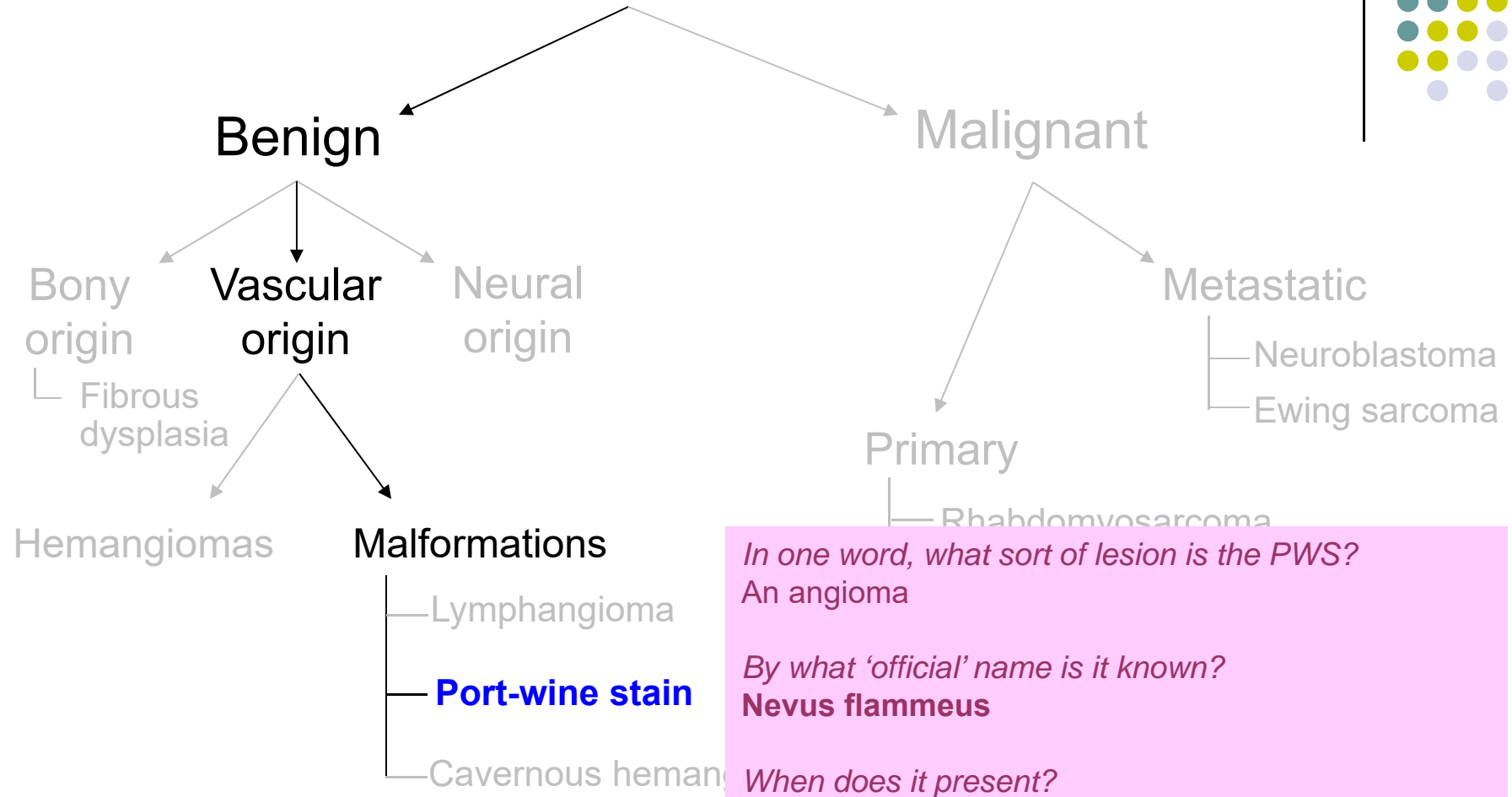


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Nevus flammeus



Pediatric Orbital Tumors



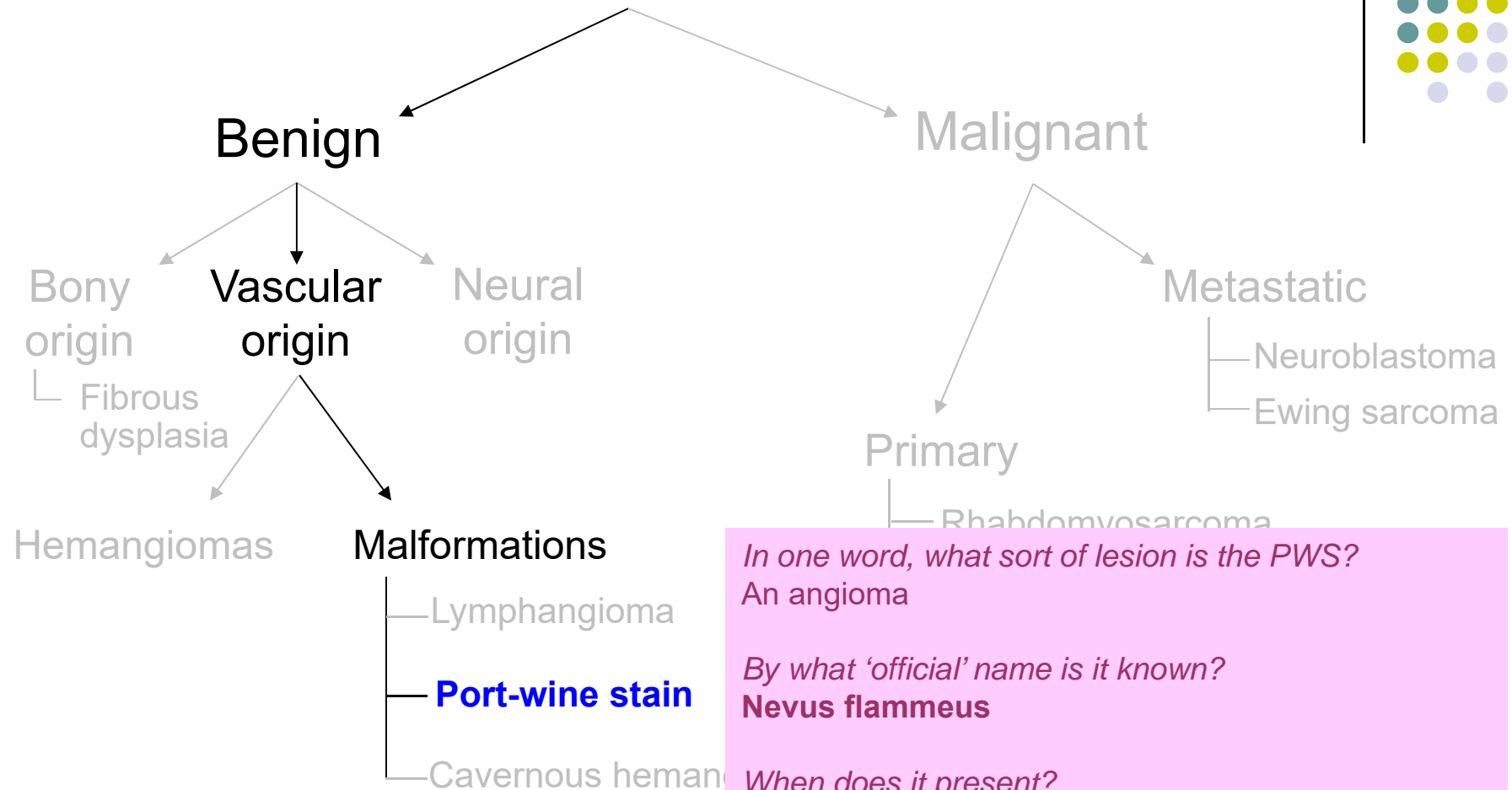
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When does it present?



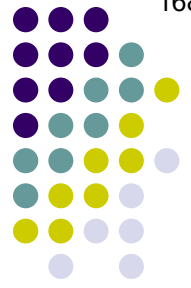
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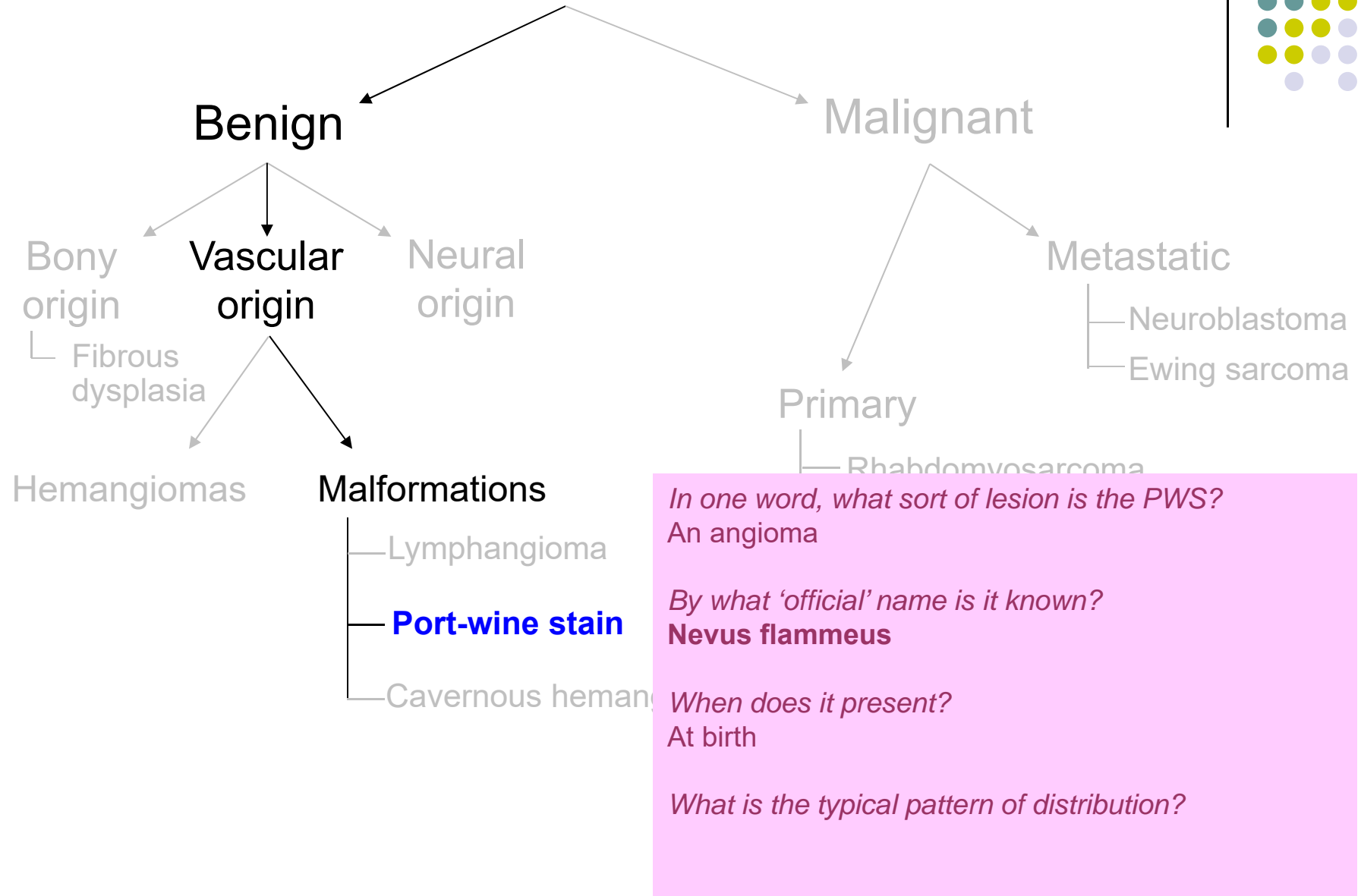
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Pediatric Orbital Tumors



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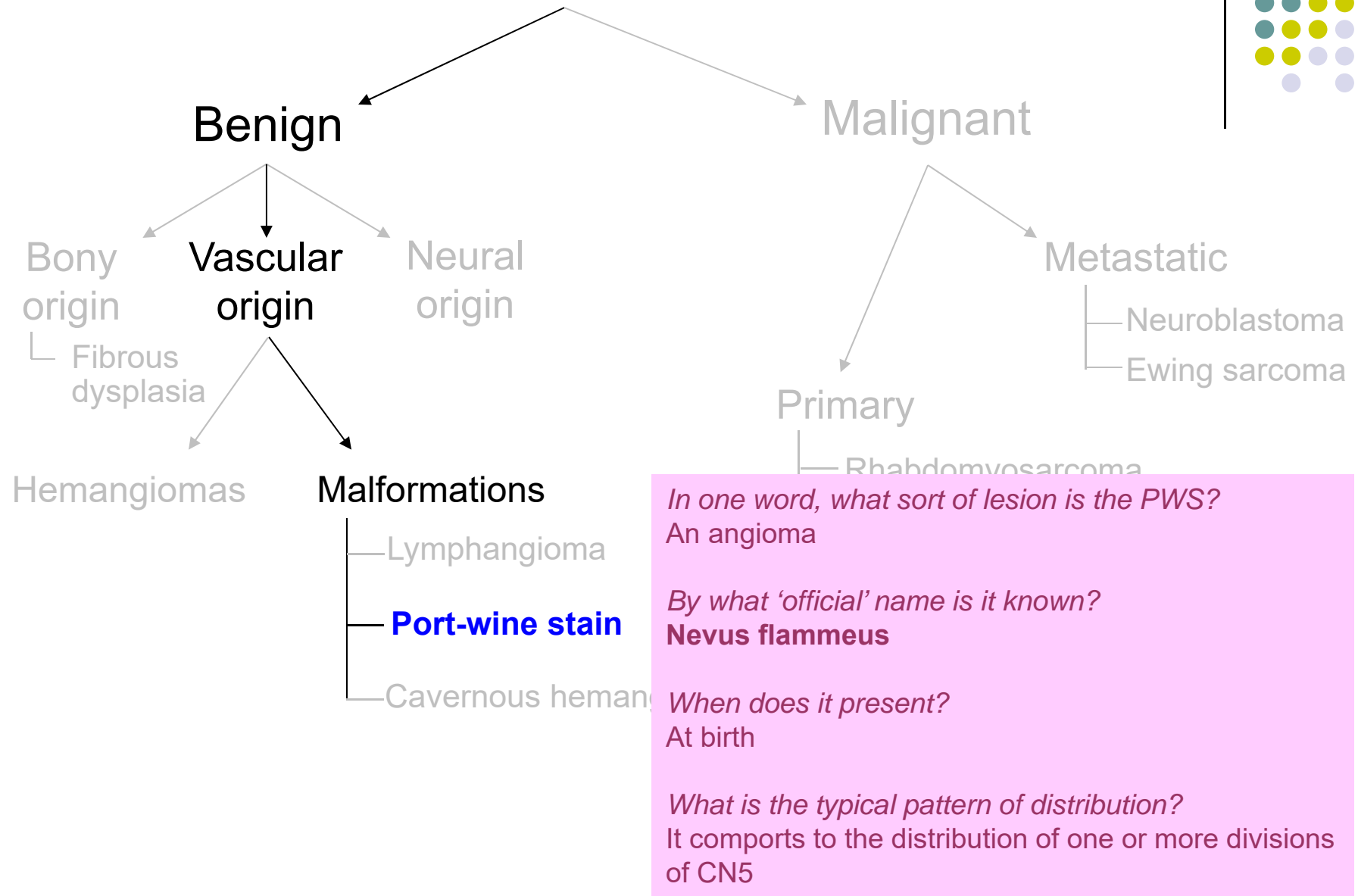
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What is the typical pattern of distribution?



Pediatric Orbital Tumors



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



Pediatric Orbital Tumors

Benign

Malignant

With what condition is the port wine stain (PWS) closely associated?

Metastatic

- Neuroblastoma
- Ewing sarcoma

sarcoma
ion is the PWS?

Port-wine stain

Cavernous hemangioma

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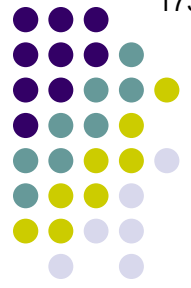
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Port-wine stain

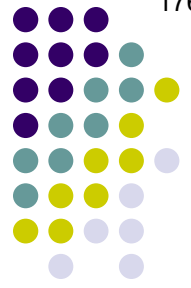
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Briefly, what is a phakomatosis?

In one word, what is it?
A **phakomatosis**

Metastatic

— Neuroblastoma

— Port-wine stain

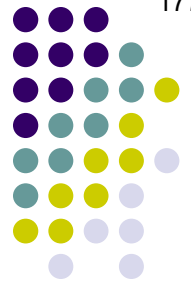
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When does it present?
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Bo
orig
L p
c
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Pediatric Orbital Tumors

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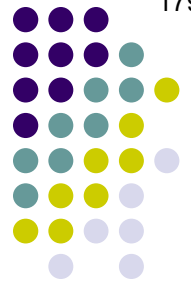
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By what more-descriptive name does the BCSC Peds book refer to them?

Port-wine stain

Cavernous hemangioma

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Nevus flammeus

When does it present?

At birth

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Metastatic

Neuroblastoma



Pediatric Orbital Tumors

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As **neuro-**

— Port-wine stain

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— Cavernous hemangioma

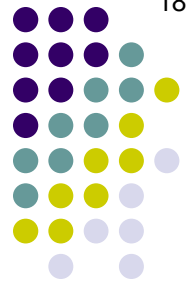
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Pediatric Orbital Tumors

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

Metastatic

— Neuroblastoma

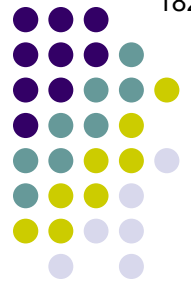
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Pediatric Orbital Tumors

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Port-wine stain

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Pediatric Orbital Tumors

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

Metastatic

- Neuroblastoma
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Port-wine stain

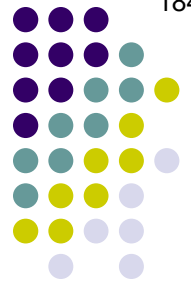
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Port-wine stain

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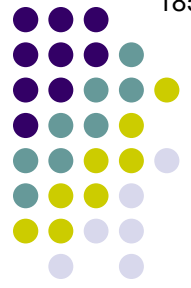
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How is this pronounced?

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TRIP-pel tray-NO-NAY

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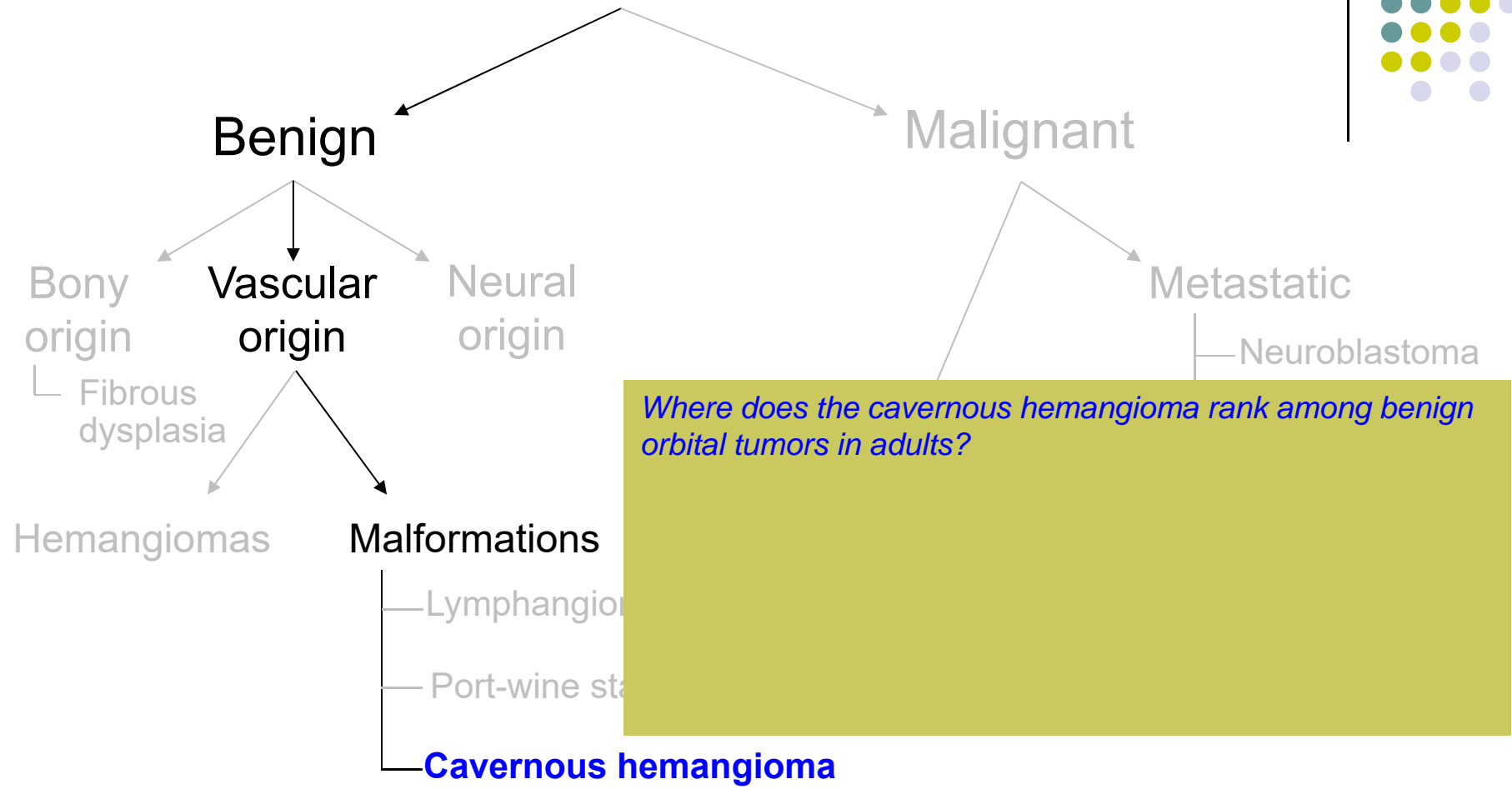
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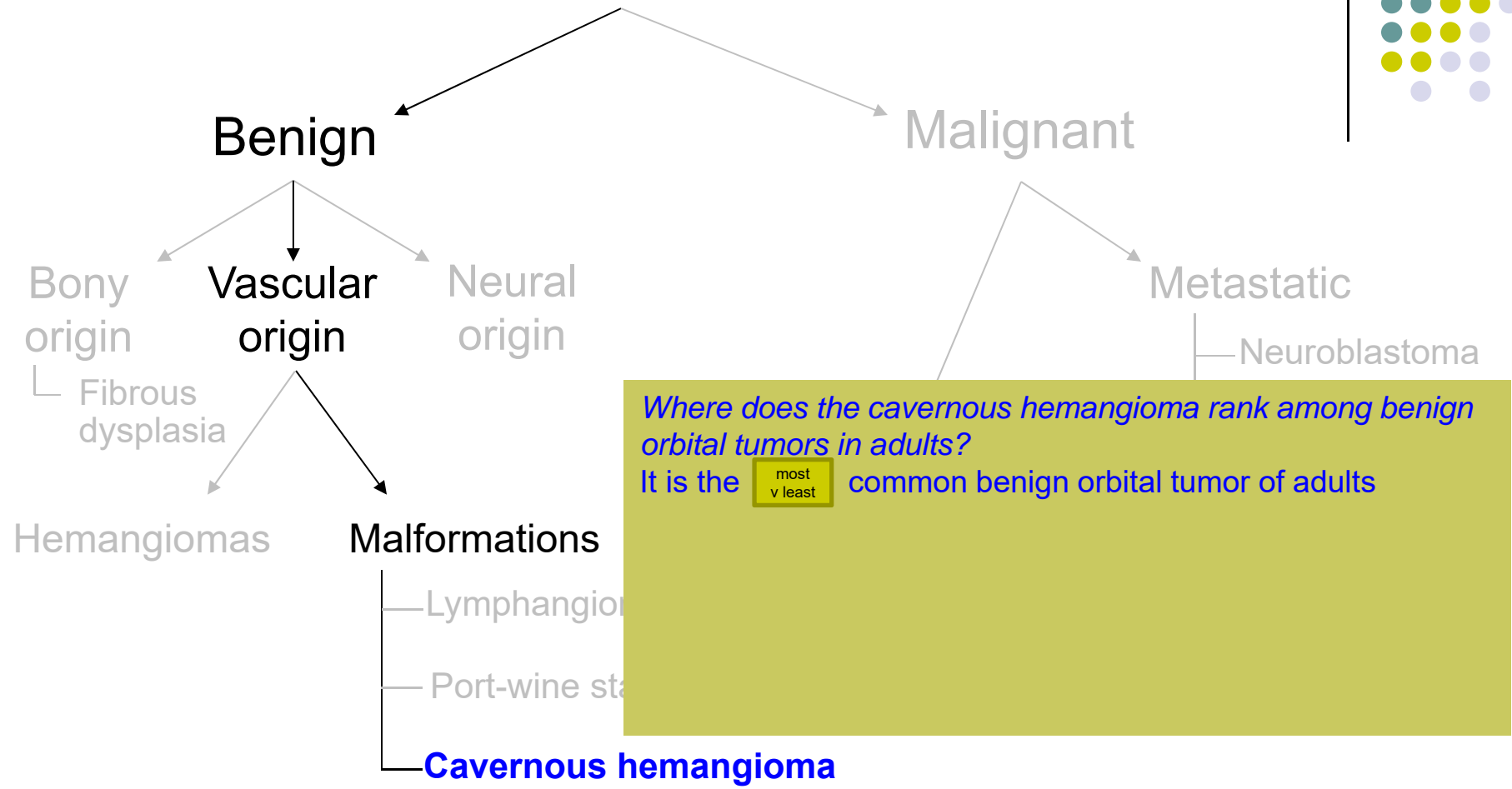


Pediatric Orbital Tumors



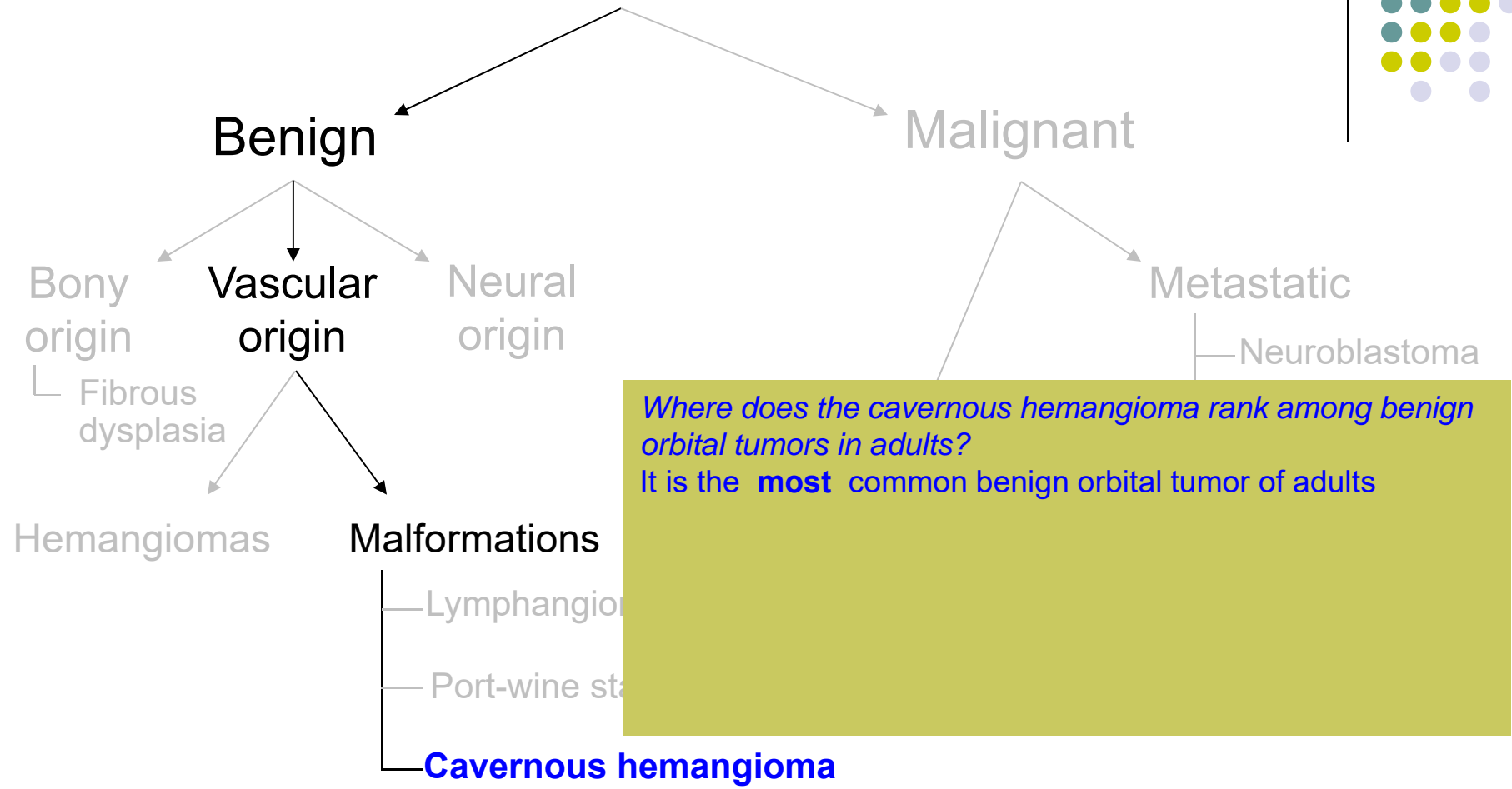


Pediatric Orbital Tumors





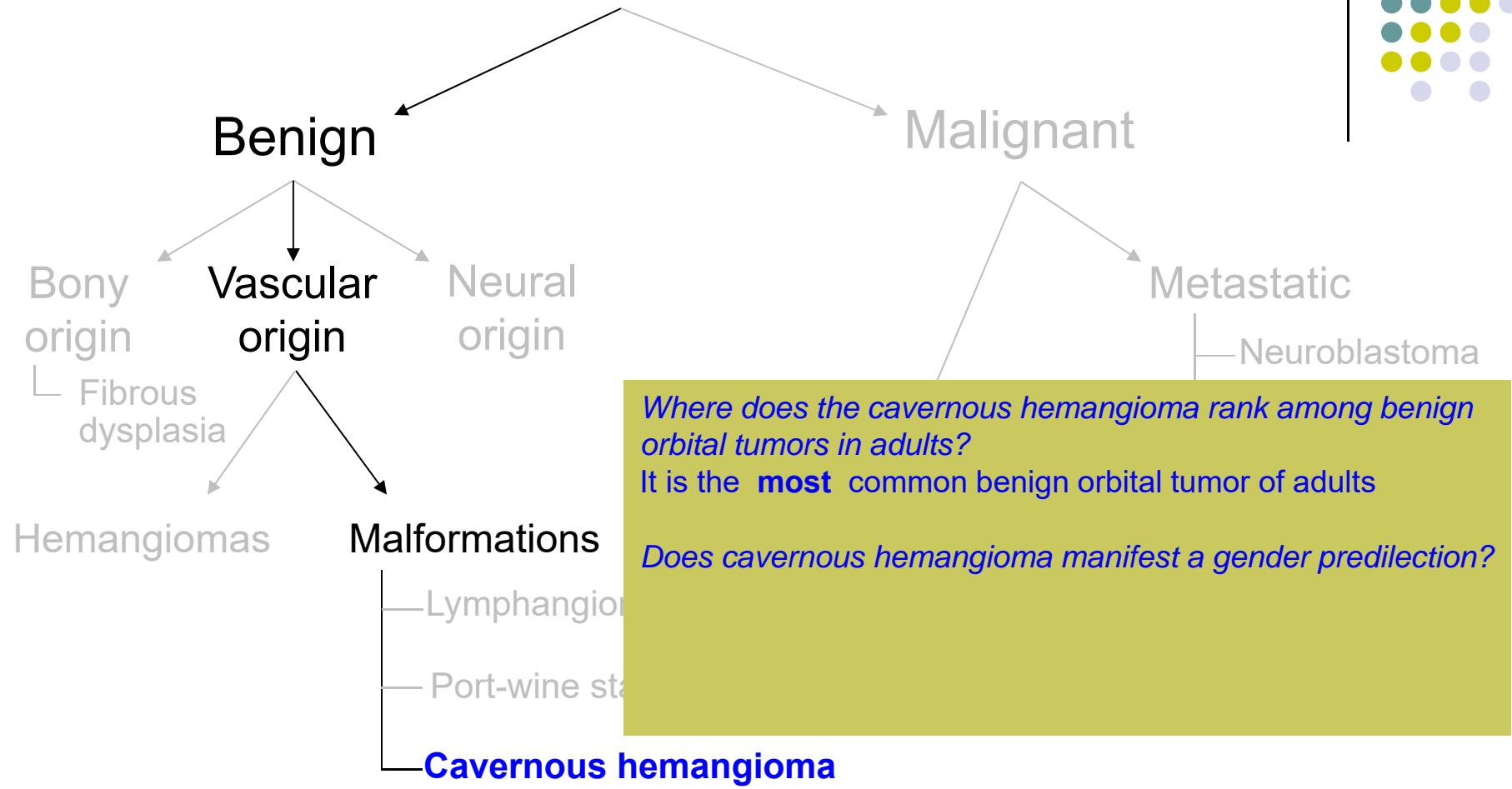
Pediatric Orbital Tumors



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

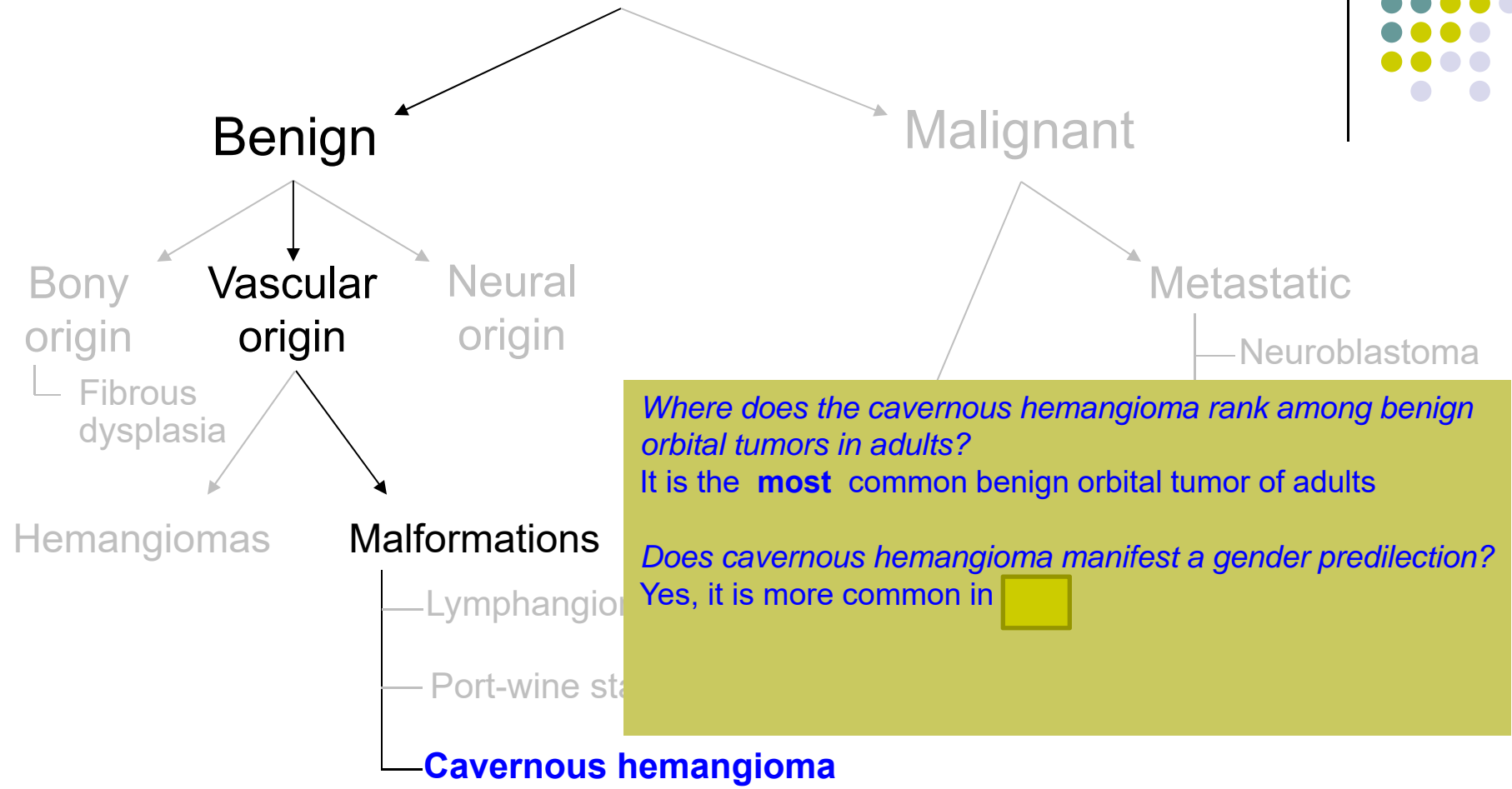


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Does cavernous hemangioma manifest a gender predilection?

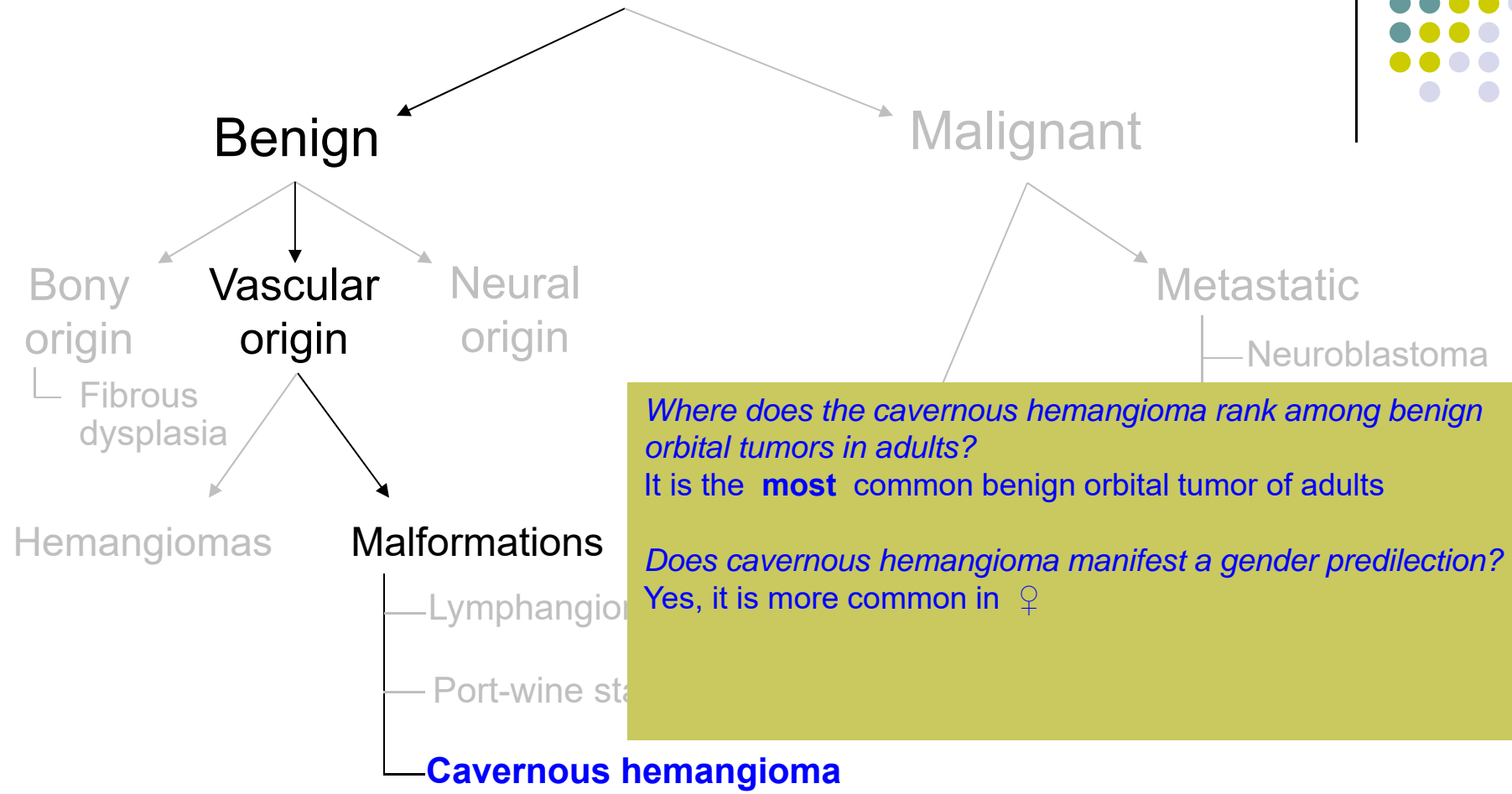


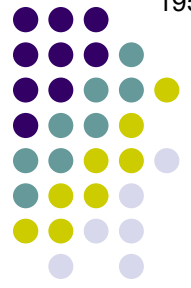
Pediatric Orbital Tumors



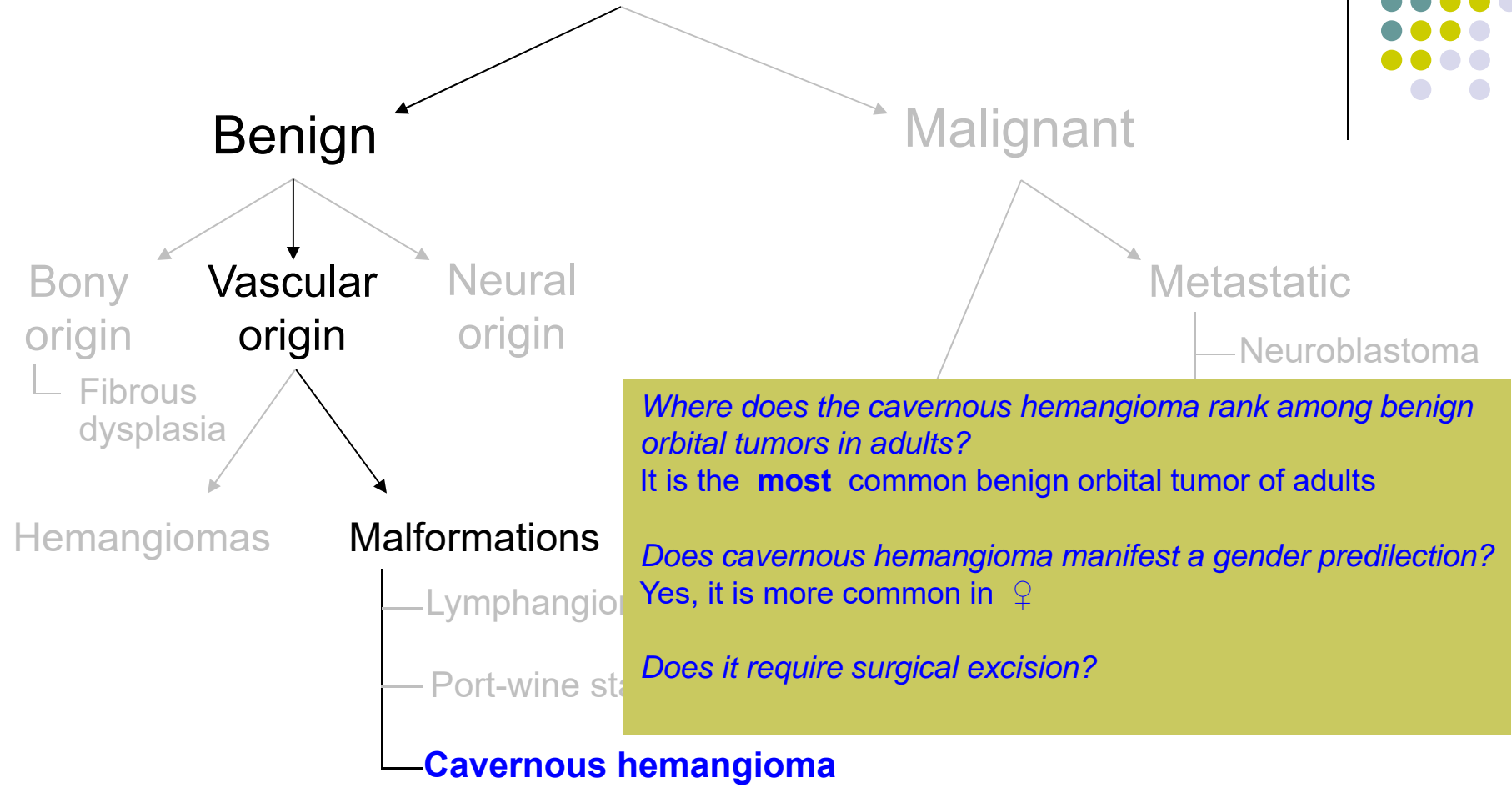


Pediatric Orbital Tumors





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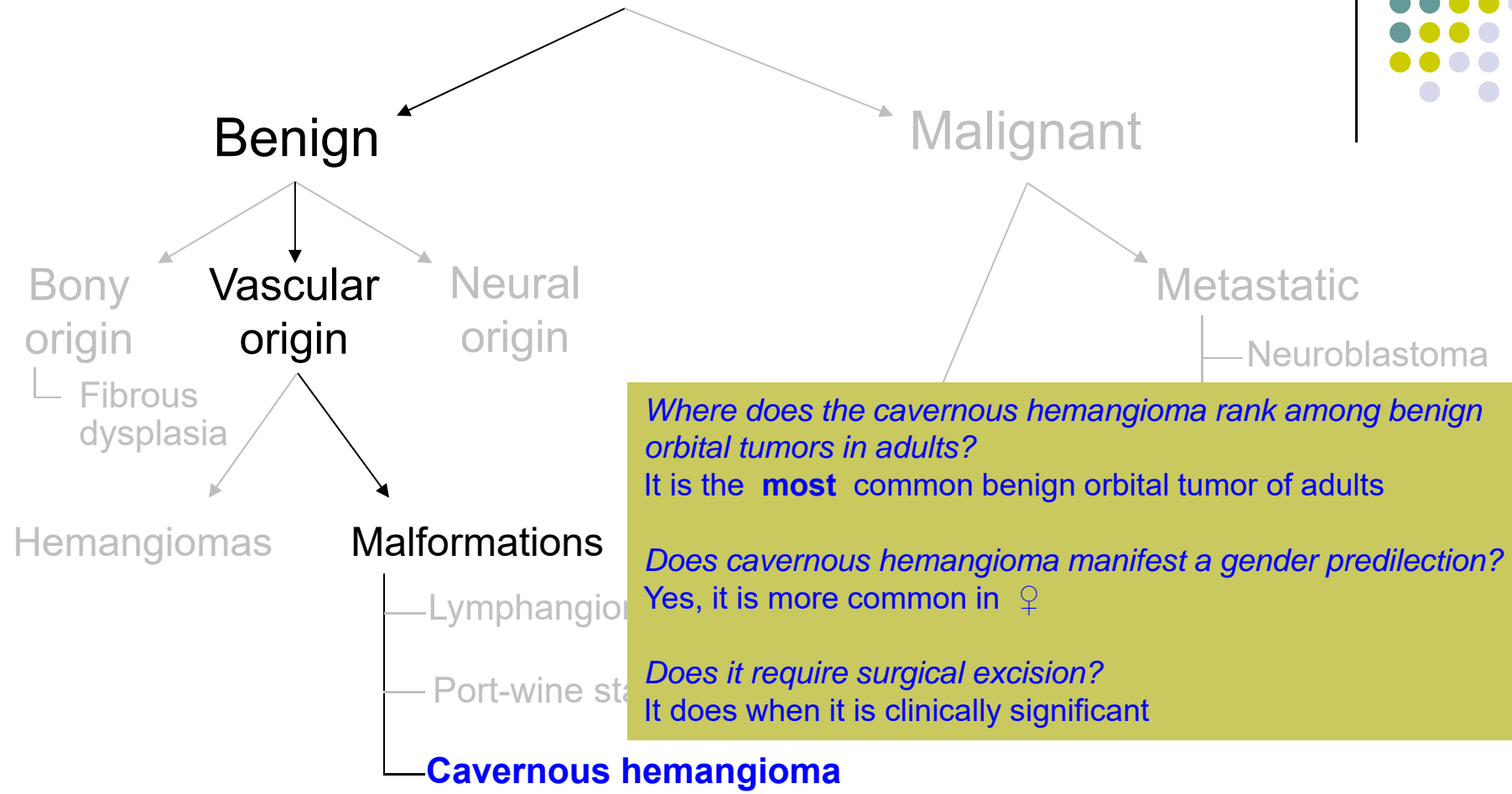
Where does the cavernous hemangioma rank among benign orbital tumors in adults?
 It is the **most** common benign orbital tumor of adults

Does cavernous hemangioma manifest a gender predilection?
 Yes, it is more common in ♀

Does it require surgical excision?



Pediatric Orbital Tumors



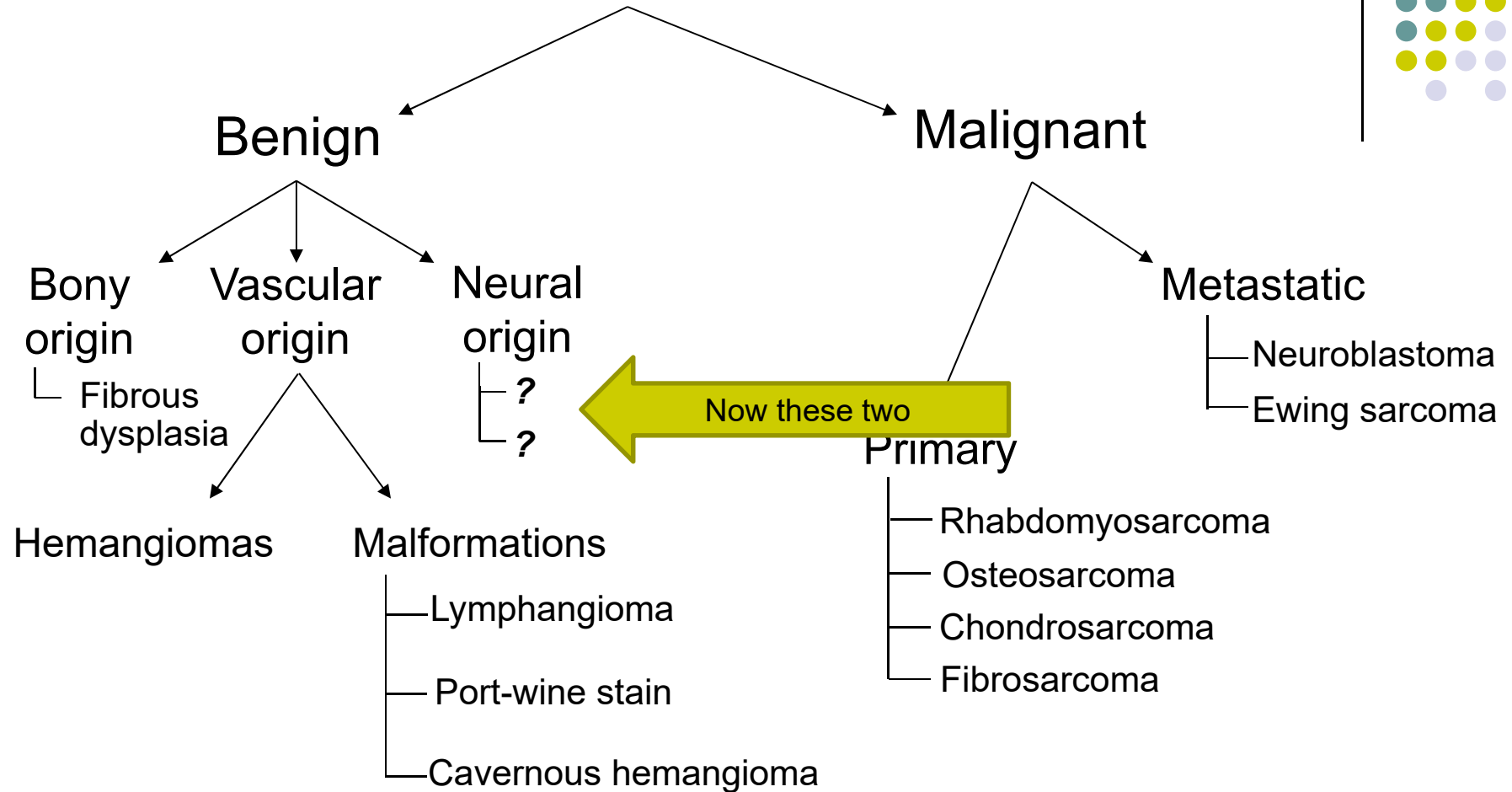
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Does it require surgical excision?
 It does when it is clinically significant

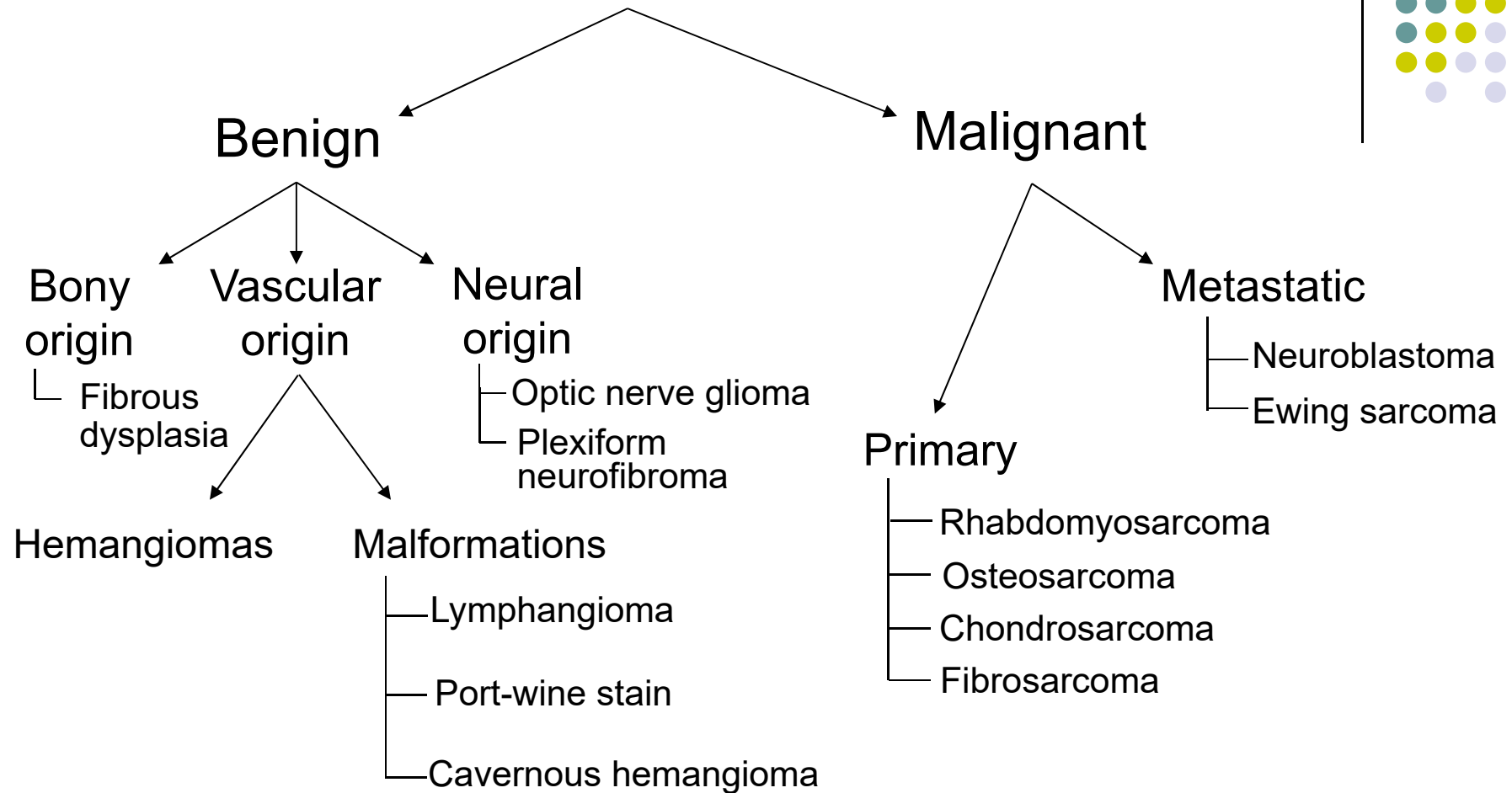


Pediatric Orbital Tumors



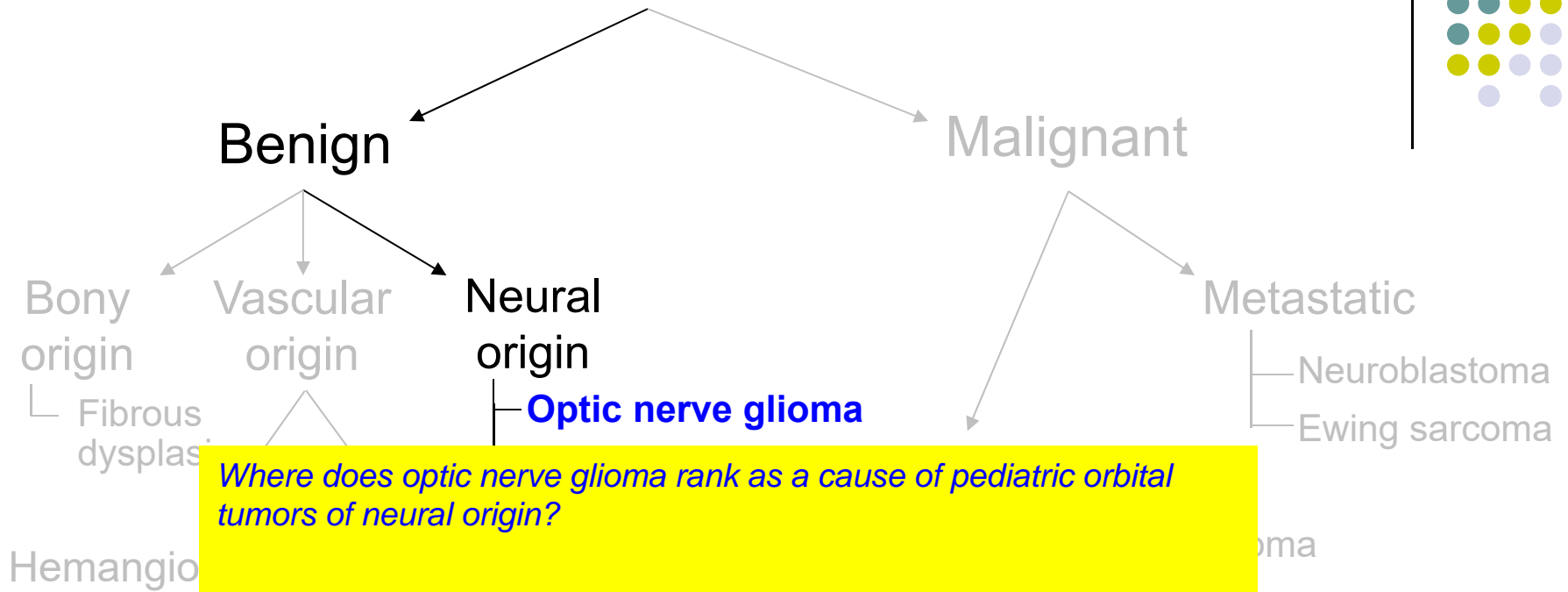


Pediatric Orbital Tumors





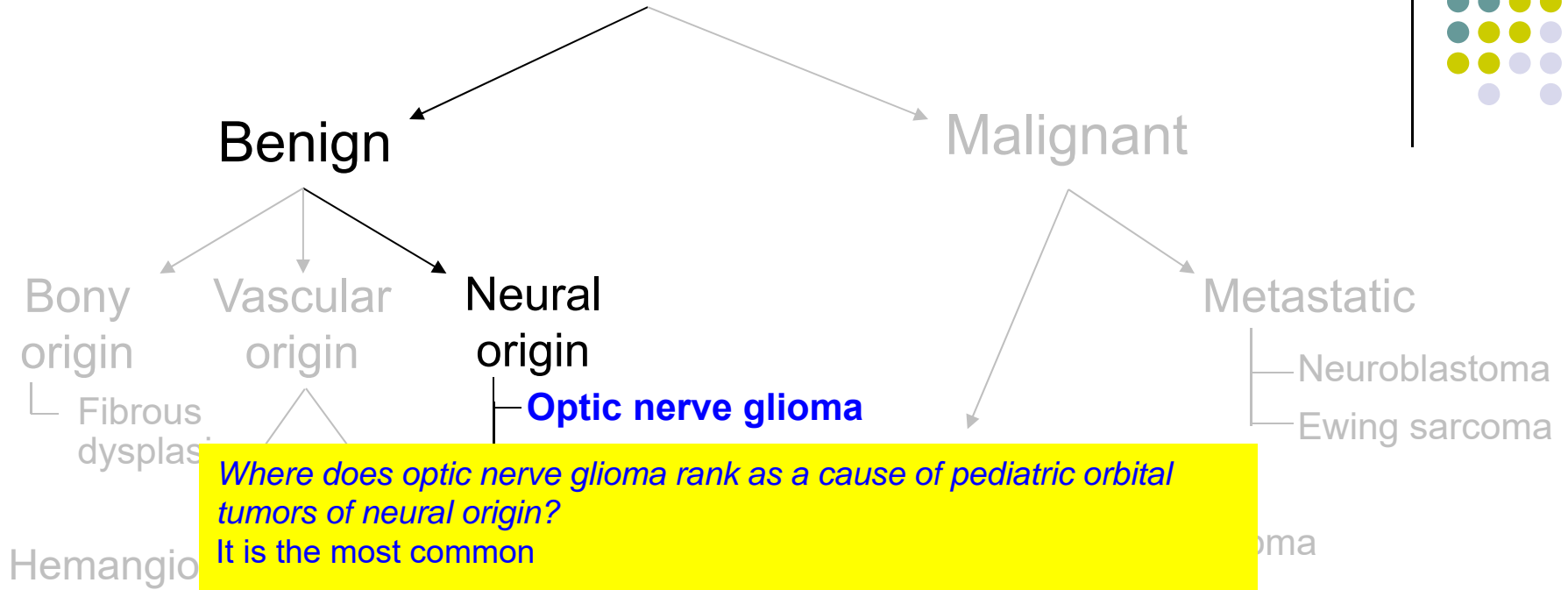
Pediatric Orbital Tumors



Where does optic nerve glioma rank as a cause of pediatric orbital tumors of neural origin?



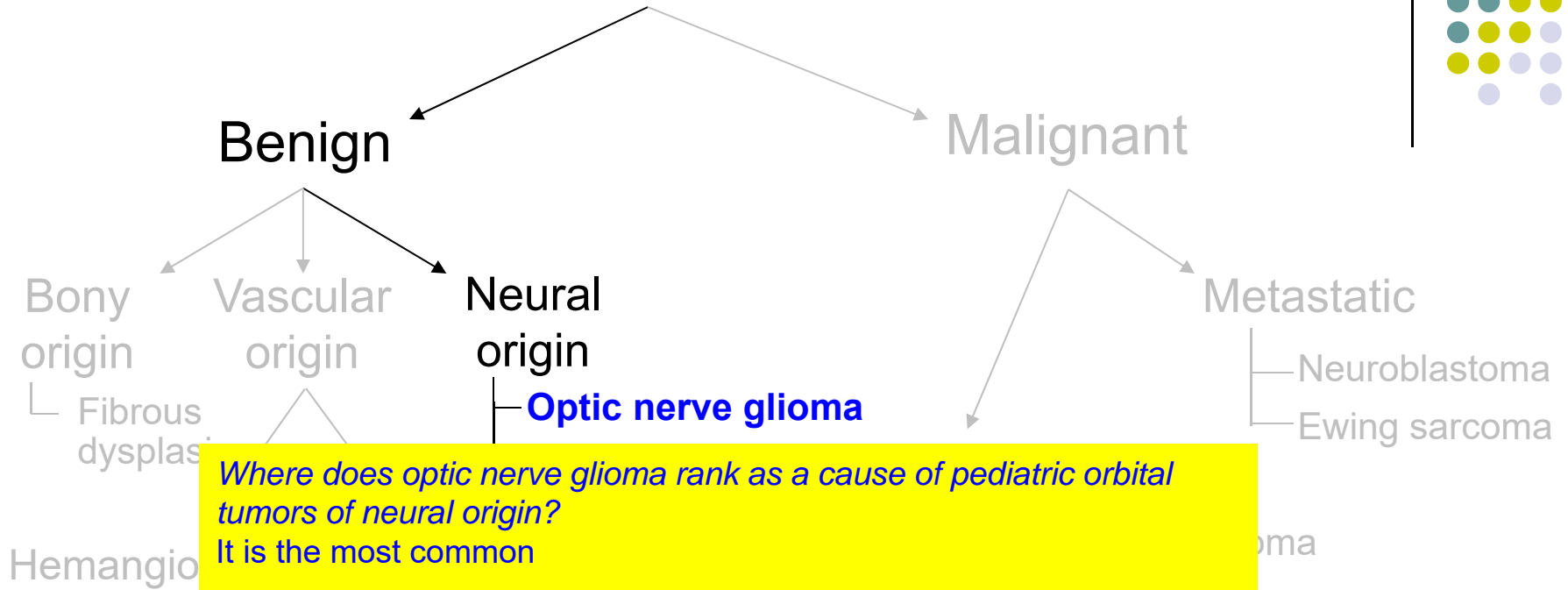
Pediatric Orbital Tumors



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Pediatric Orbital Tumors

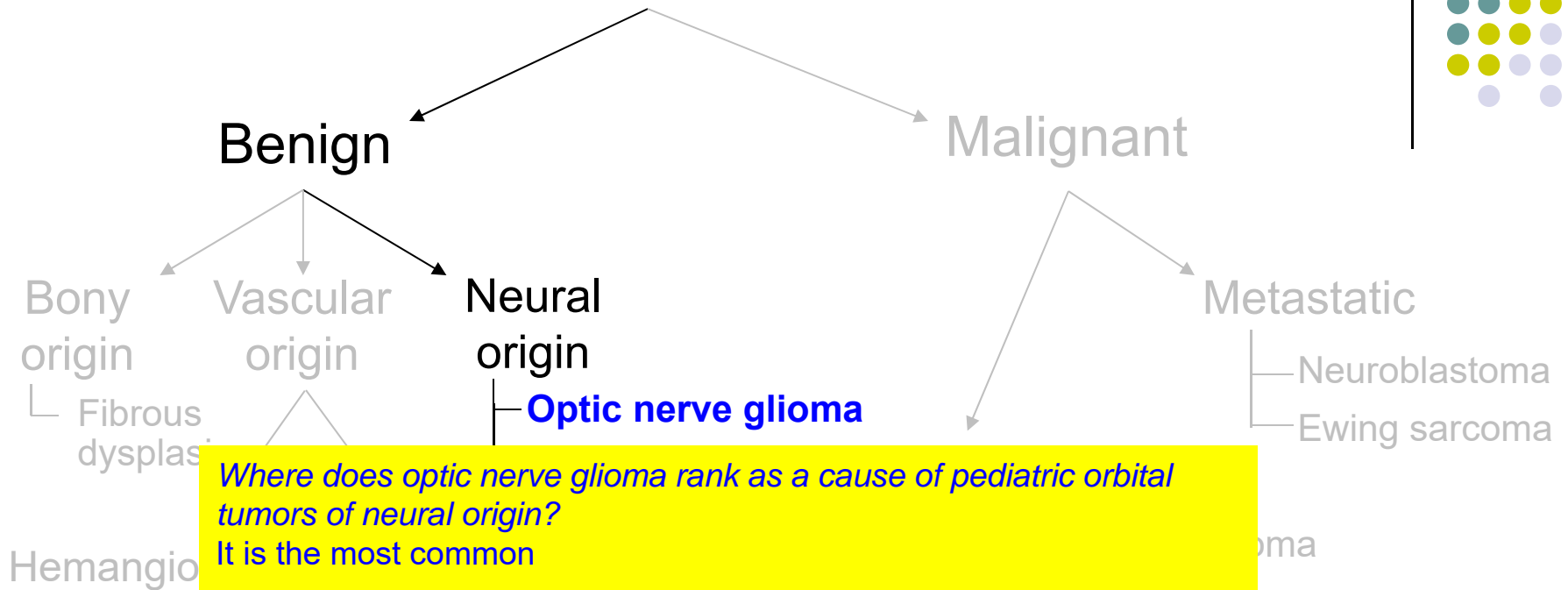


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50% are associated with a syndrome--which one?

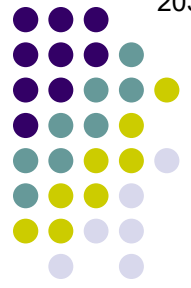


Pediatric Orbital Tumors

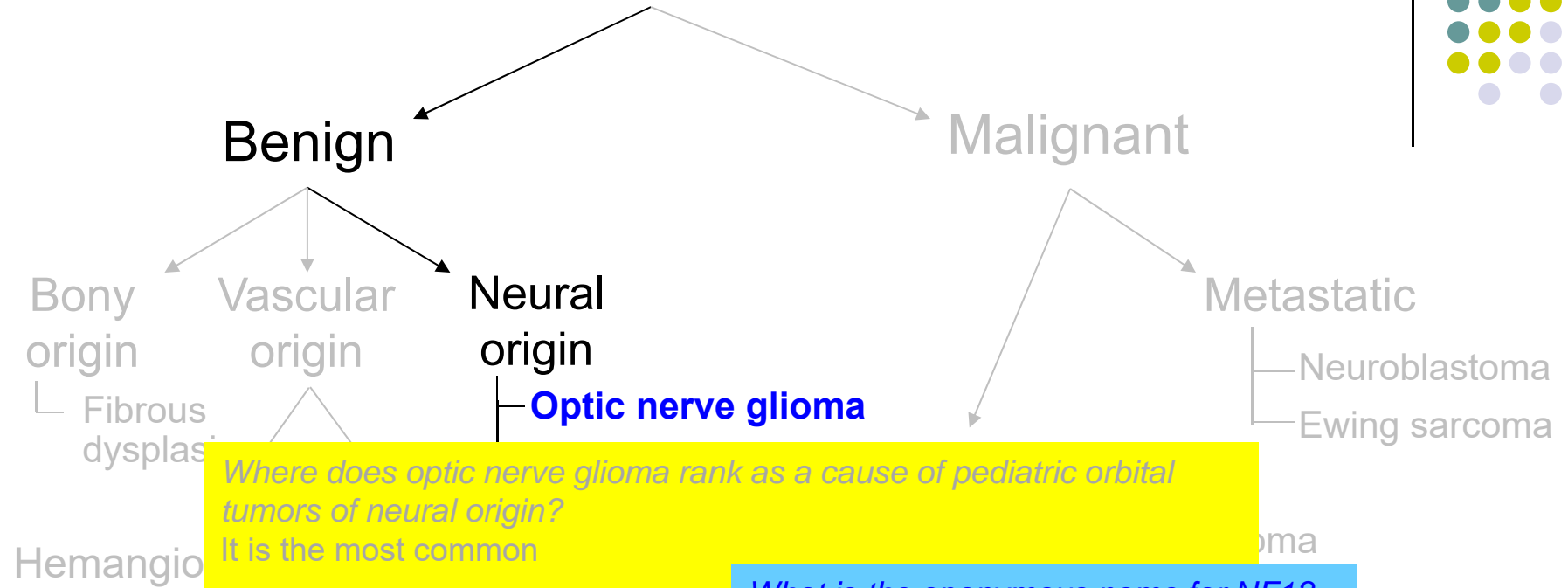


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Pediatric Orbital Tumors



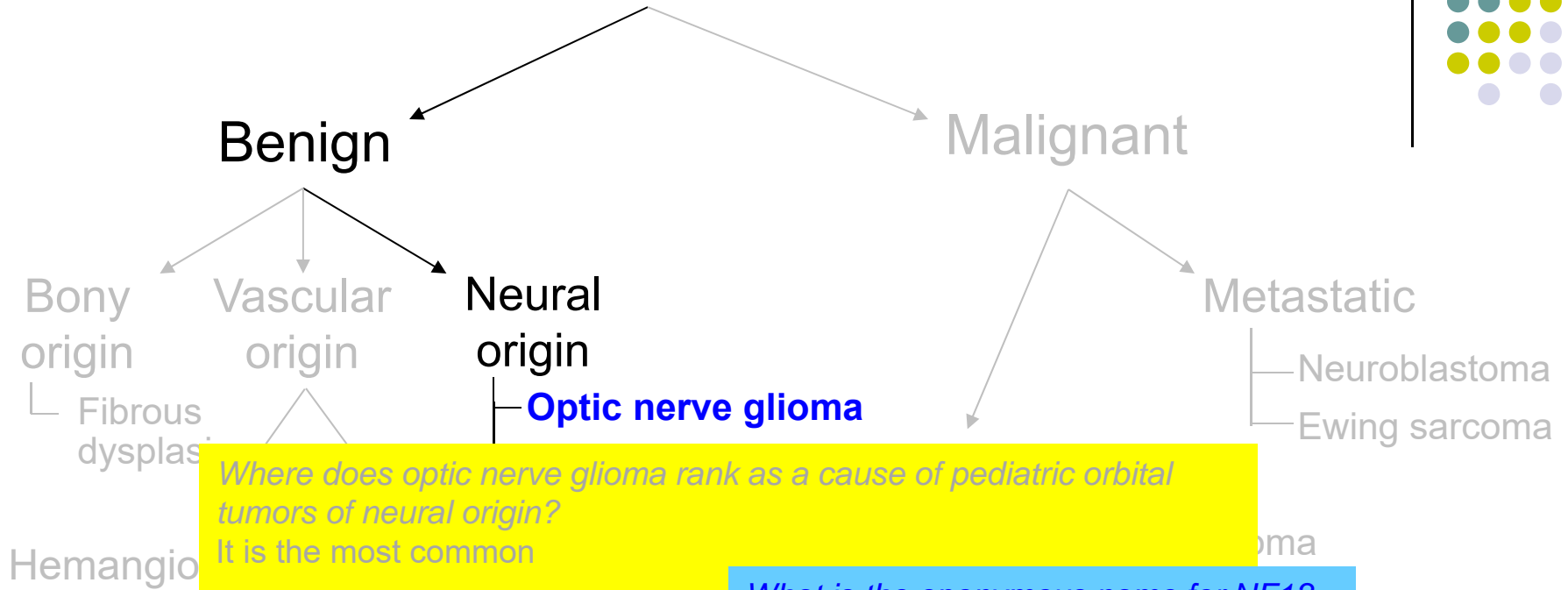
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Pediatric Orbital Tumors



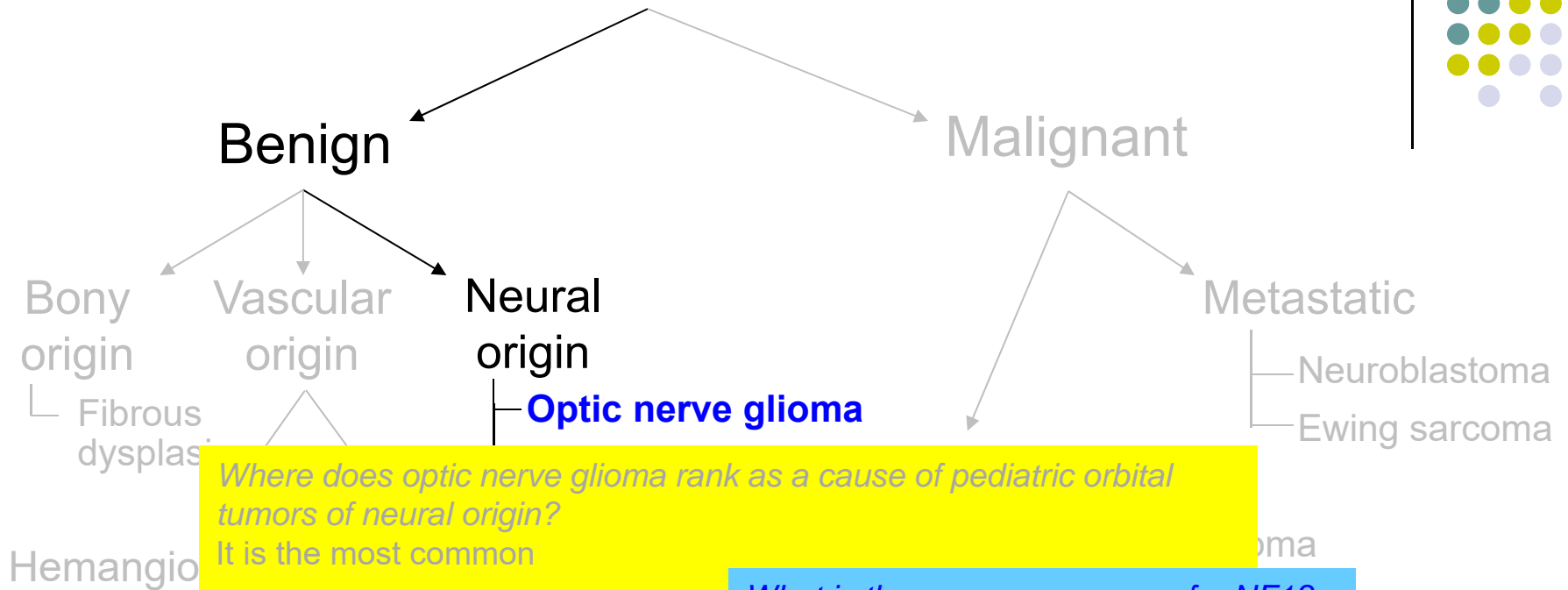
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Pediatric Orbital Tumors

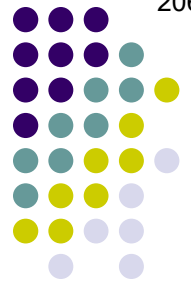


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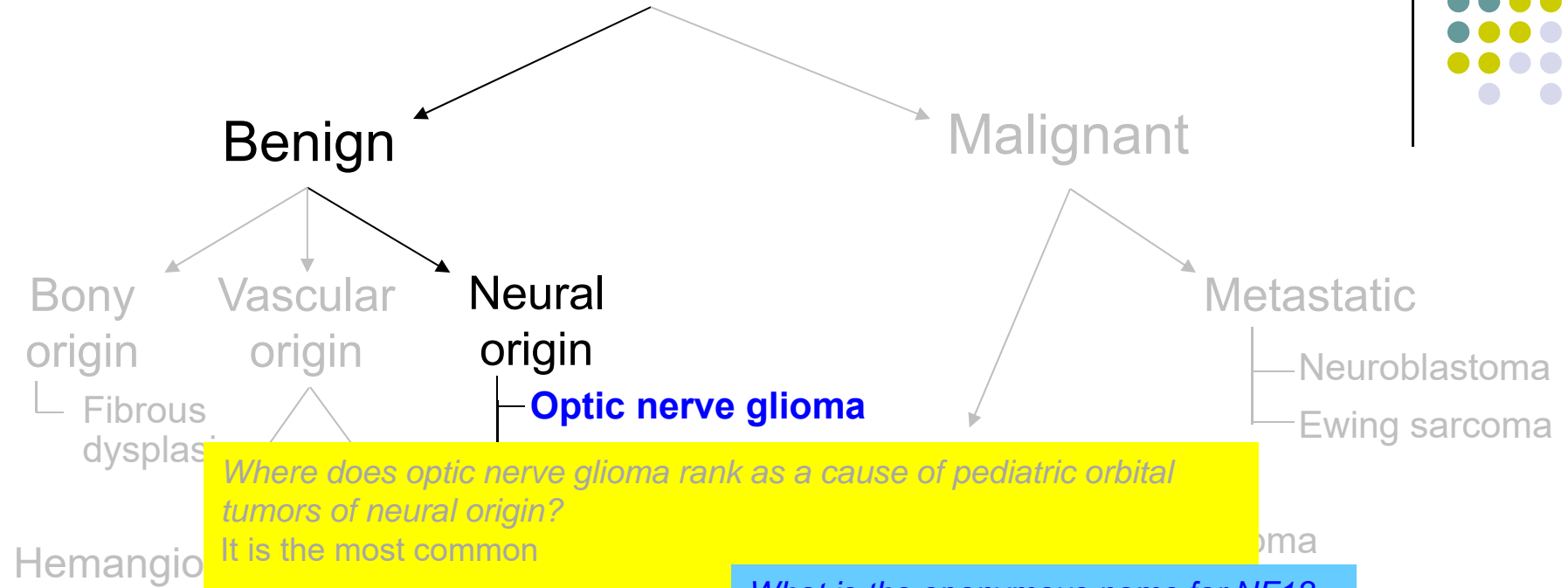
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Pediatric Orbital Tumors



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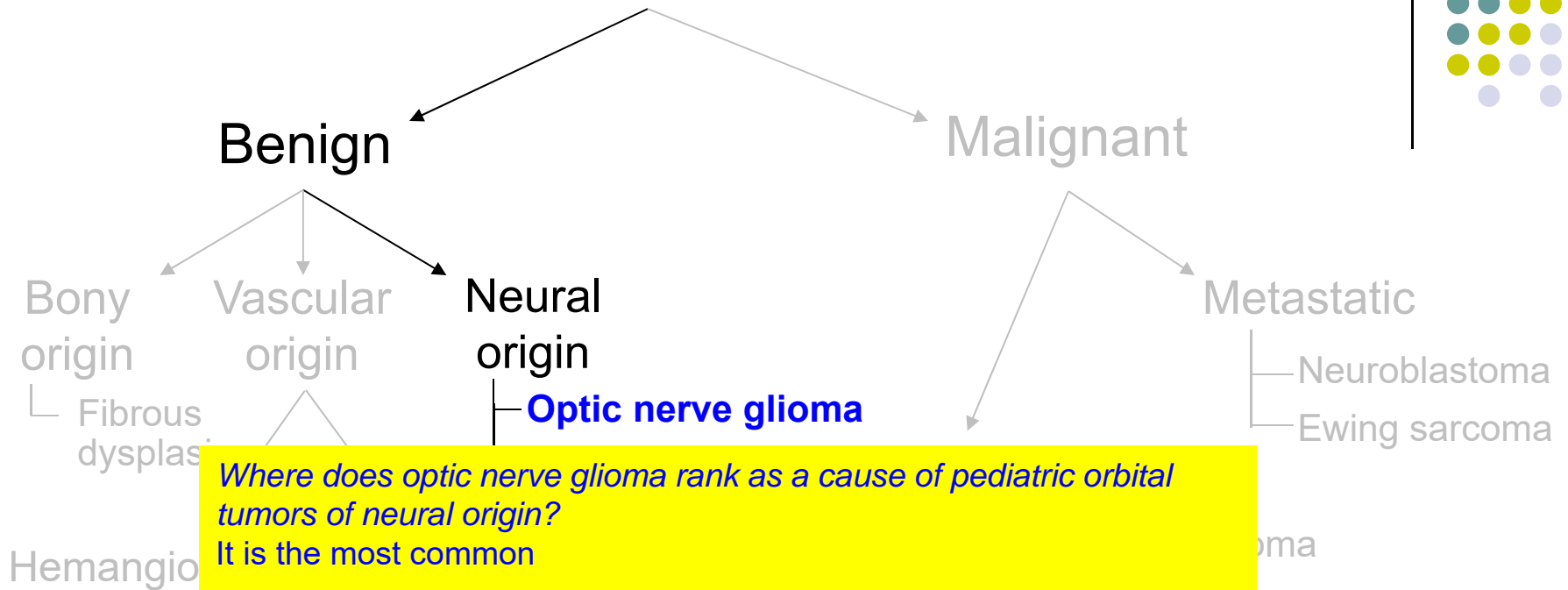
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 A phakomatosis



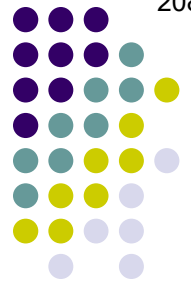
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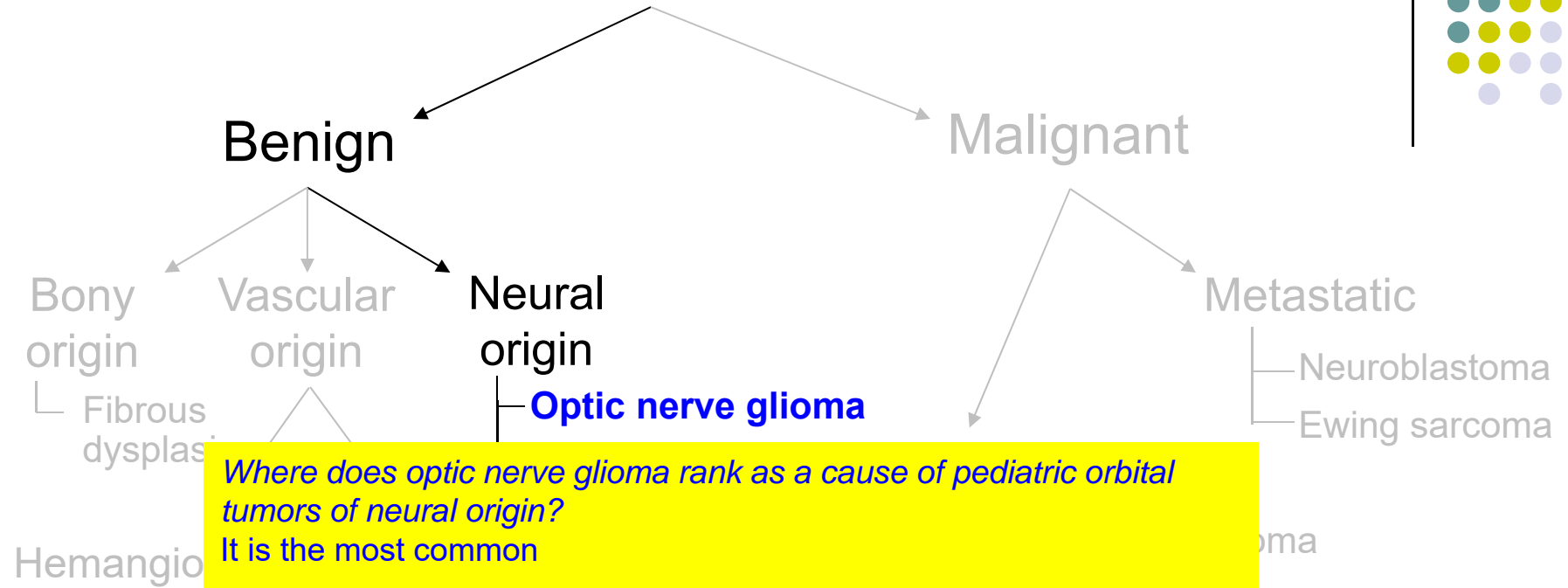
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Would you rather have an optic nerve glioma as a child or as an adult?



Pediatric Orbital Tumors



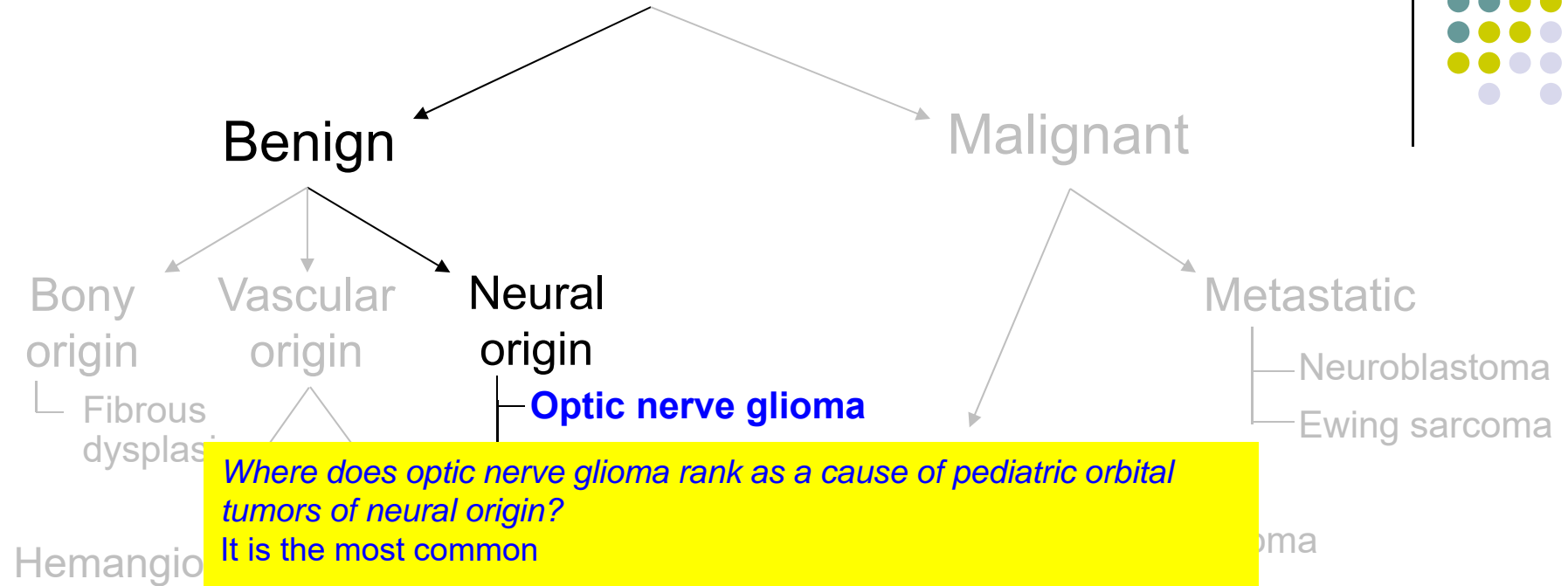
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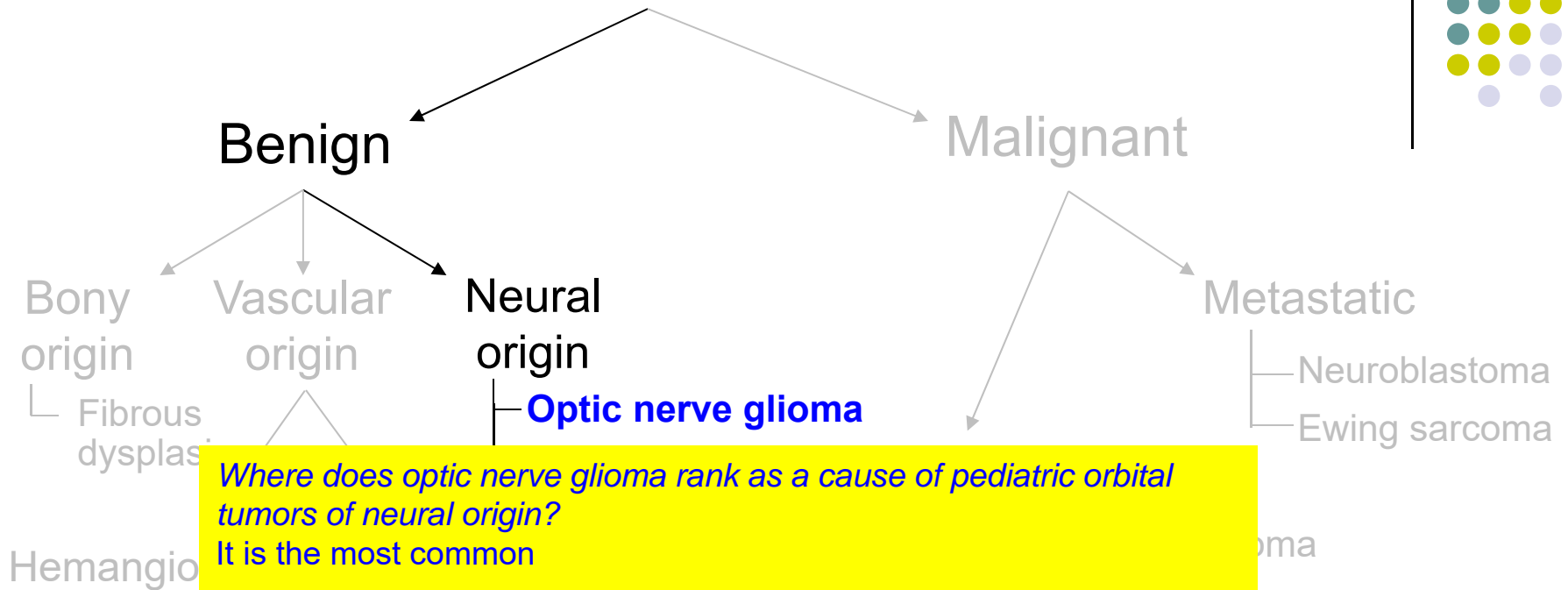
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What finding significantly worsens the prognosis in pediatric cases?



Pediatric Orbital Tumors



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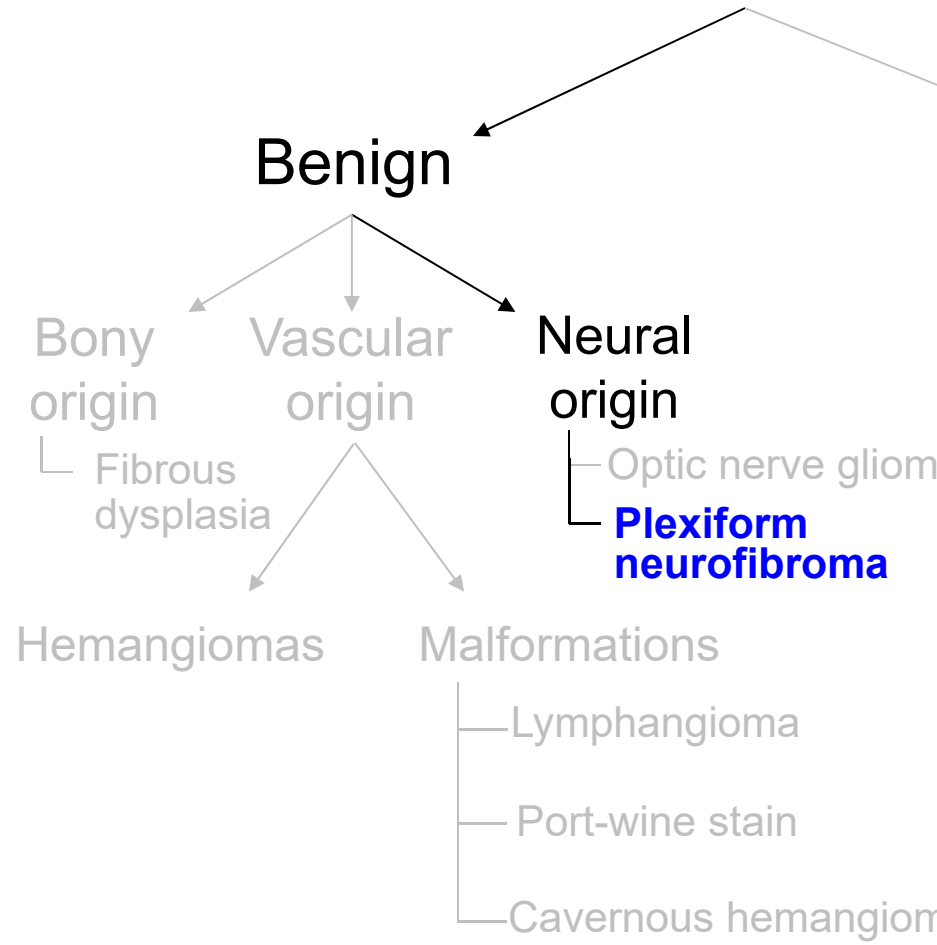
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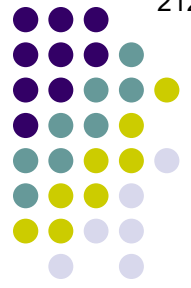
What finding significantly worsens the prognosis in pediatric cases?
 Intracranial extension



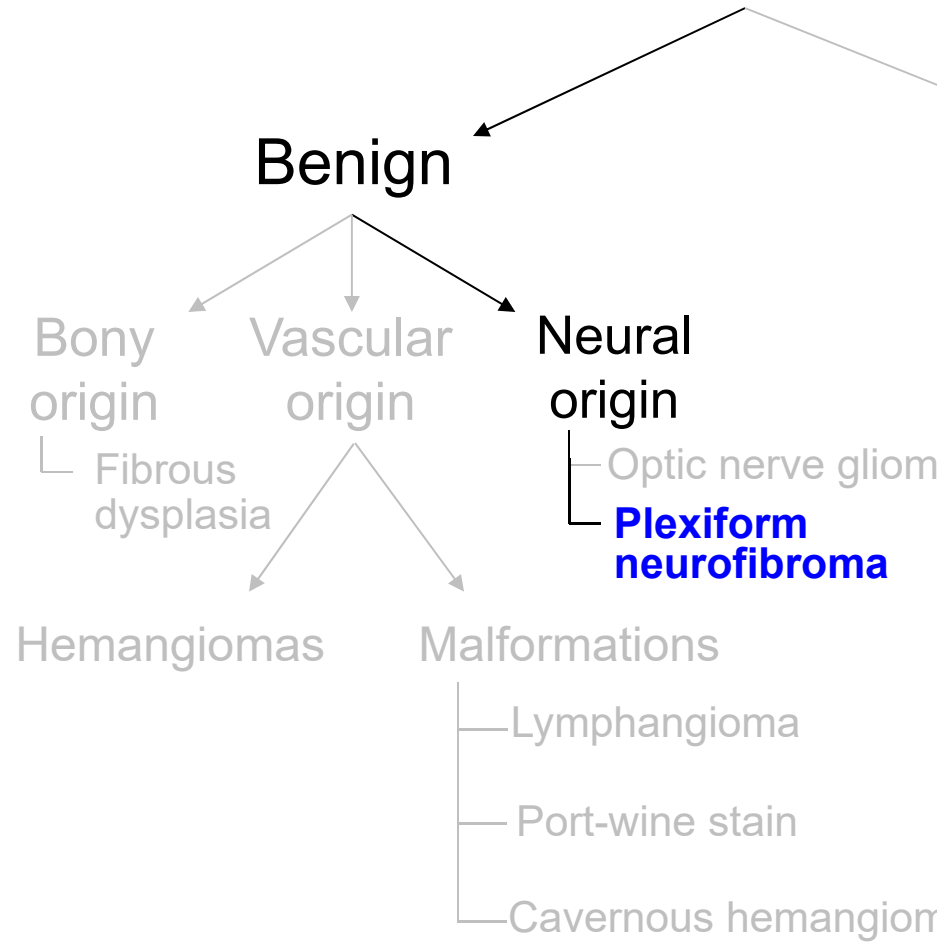
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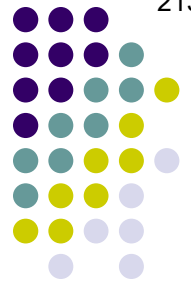
What two specific cell types are implicated in plexiform neurofibroma formation?



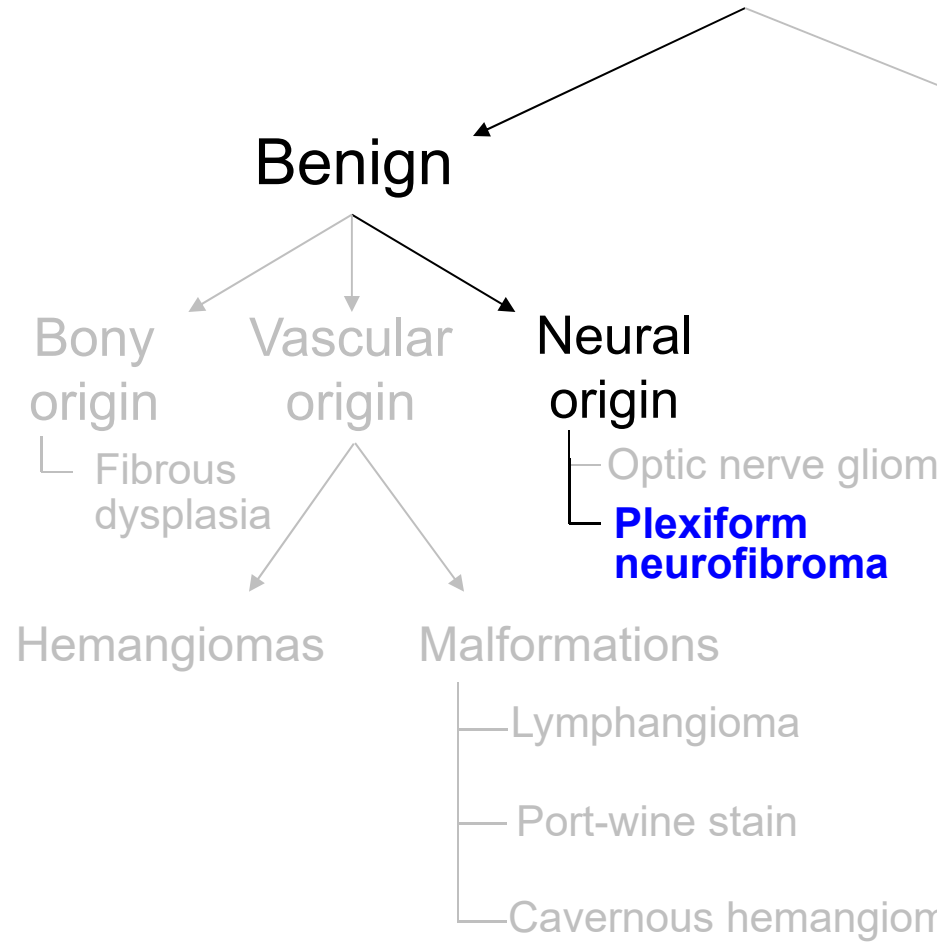
Pediatric Orbital Tumors



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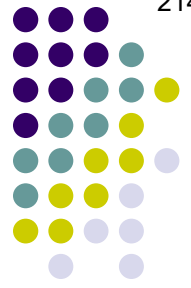


Pediatric Orbital Tumors

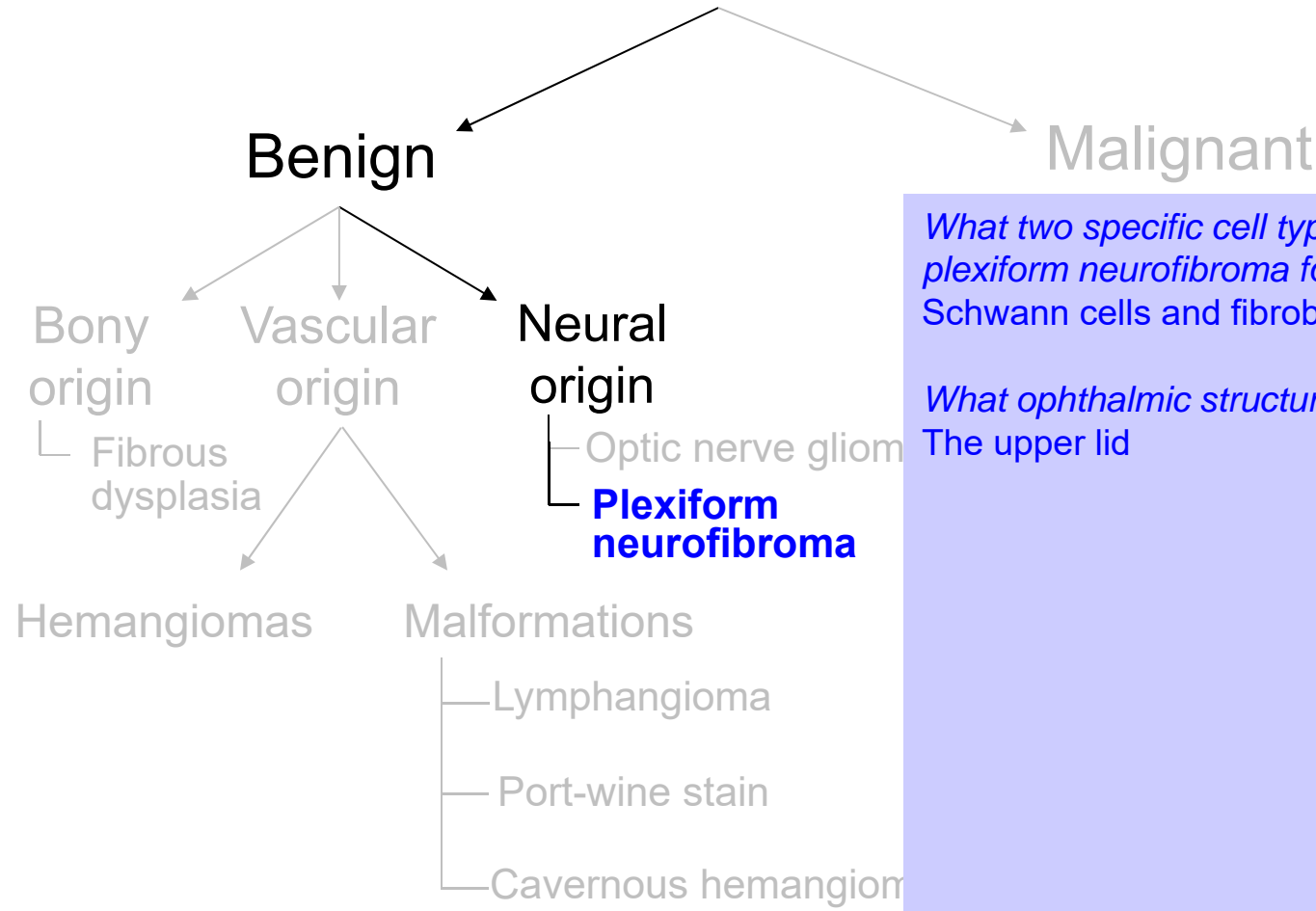


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What ophthalmic structure is typically involved?



Pediatric Orbital Tumors

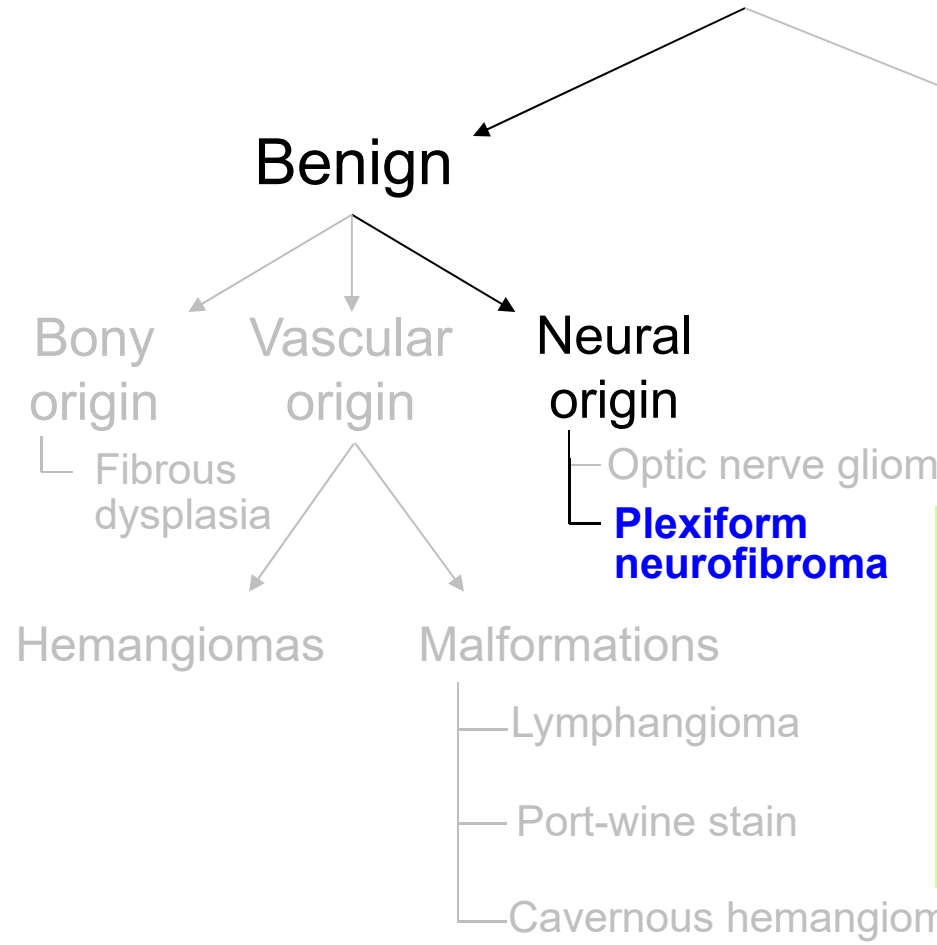


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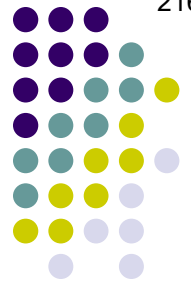
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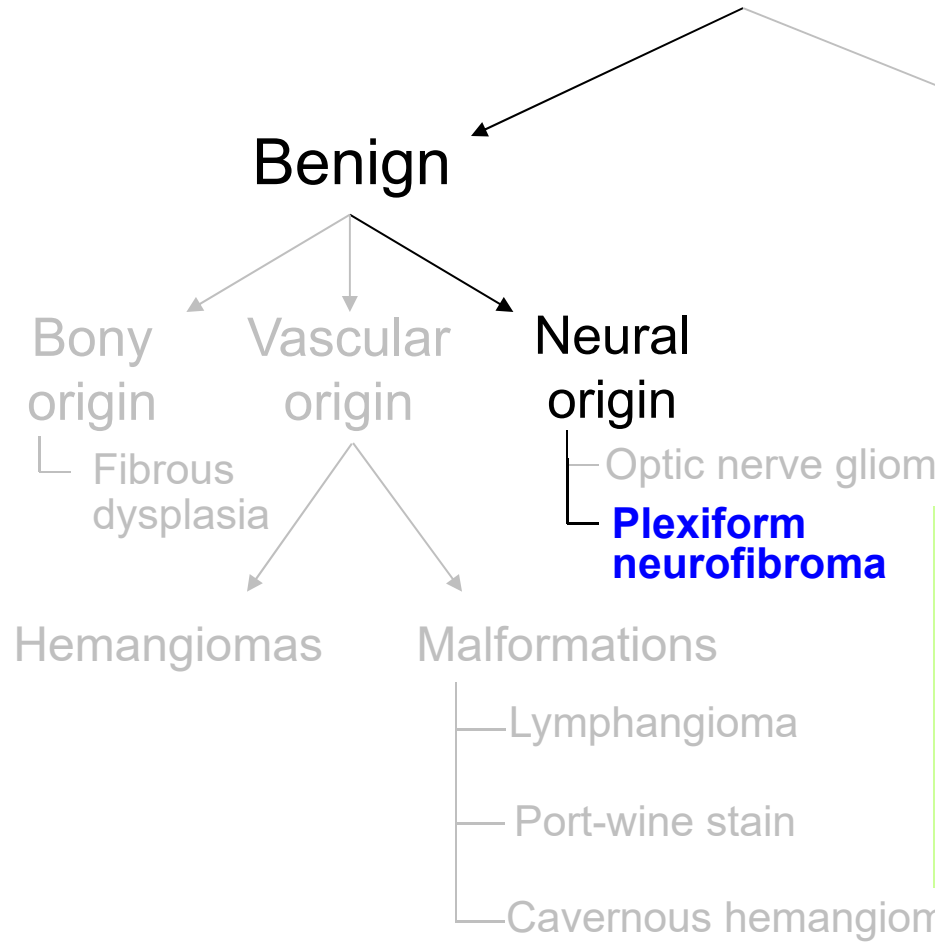
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Their predilection for the temporal aspect of the upper lid produces a specific configuration of the involved lid. What is it?



Pediatric Orbital Tumors



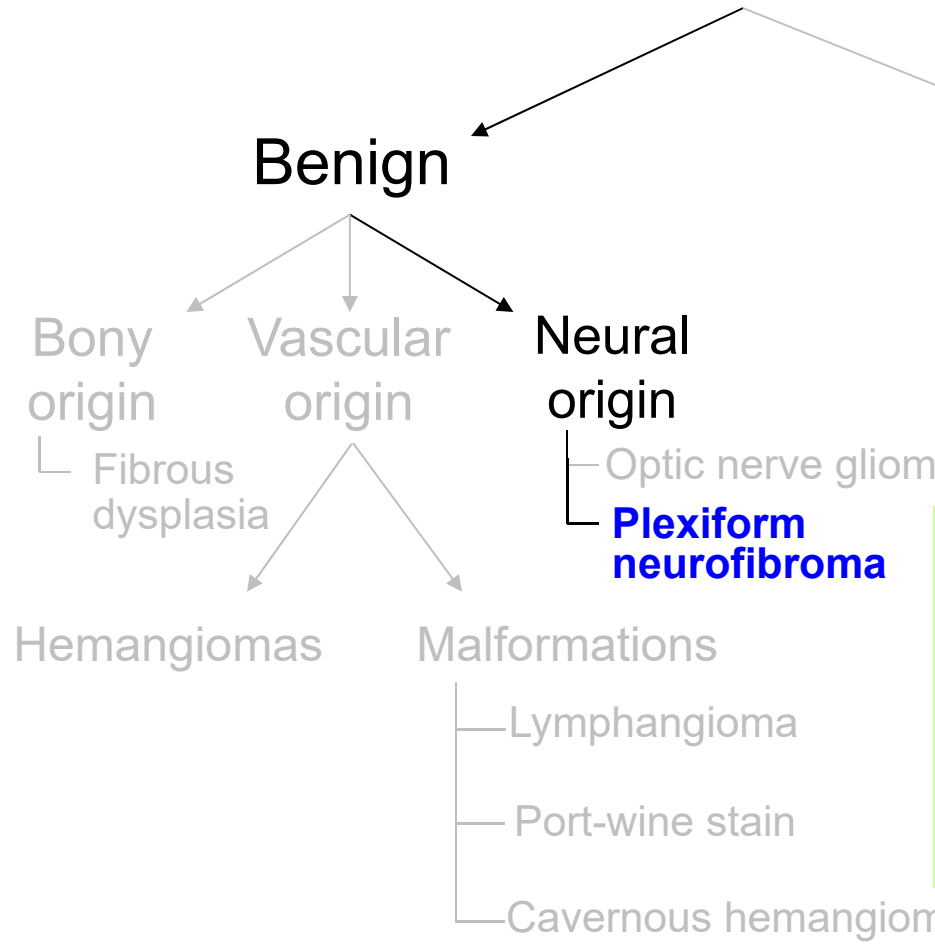
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The tumor gives rise to an 'S-shaped lid'



Pediatric Orbital Tumors

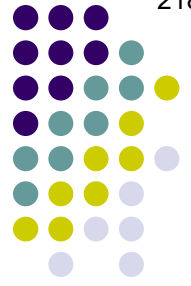


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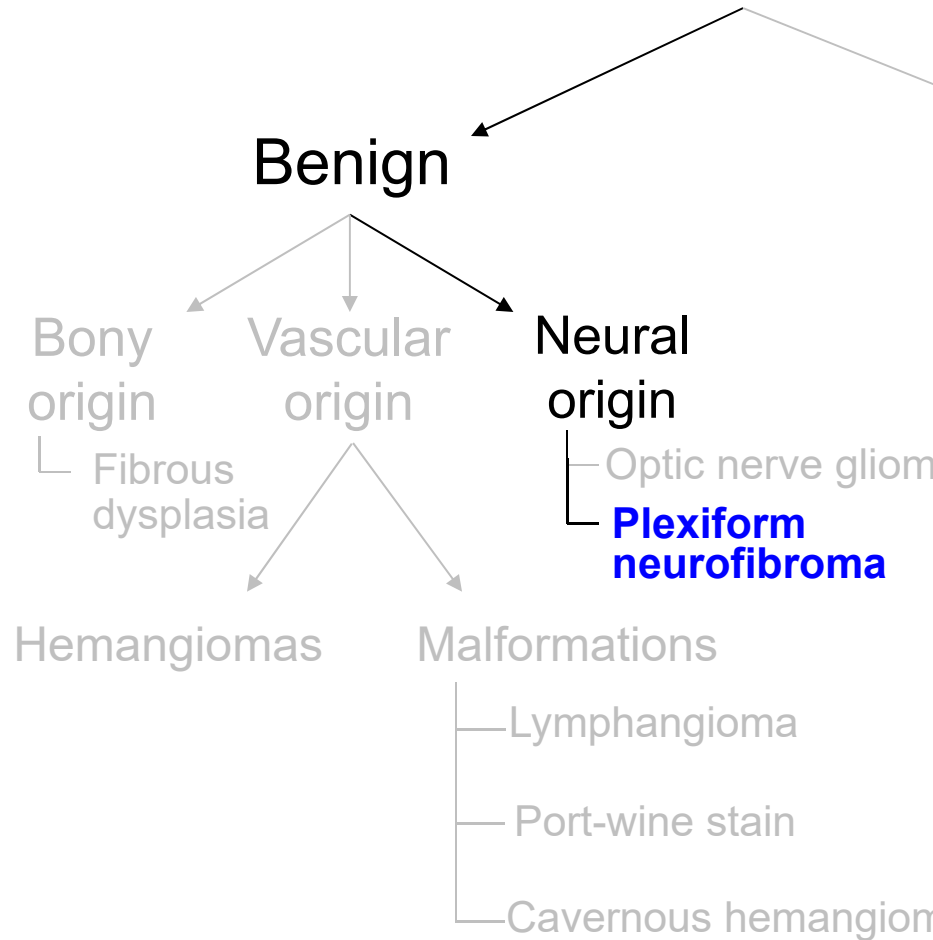
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Are upper-lid plexiform neurofibromas potentially amblyogenic?



Pediatric Orbital Tumors

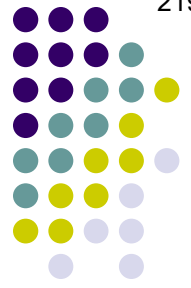


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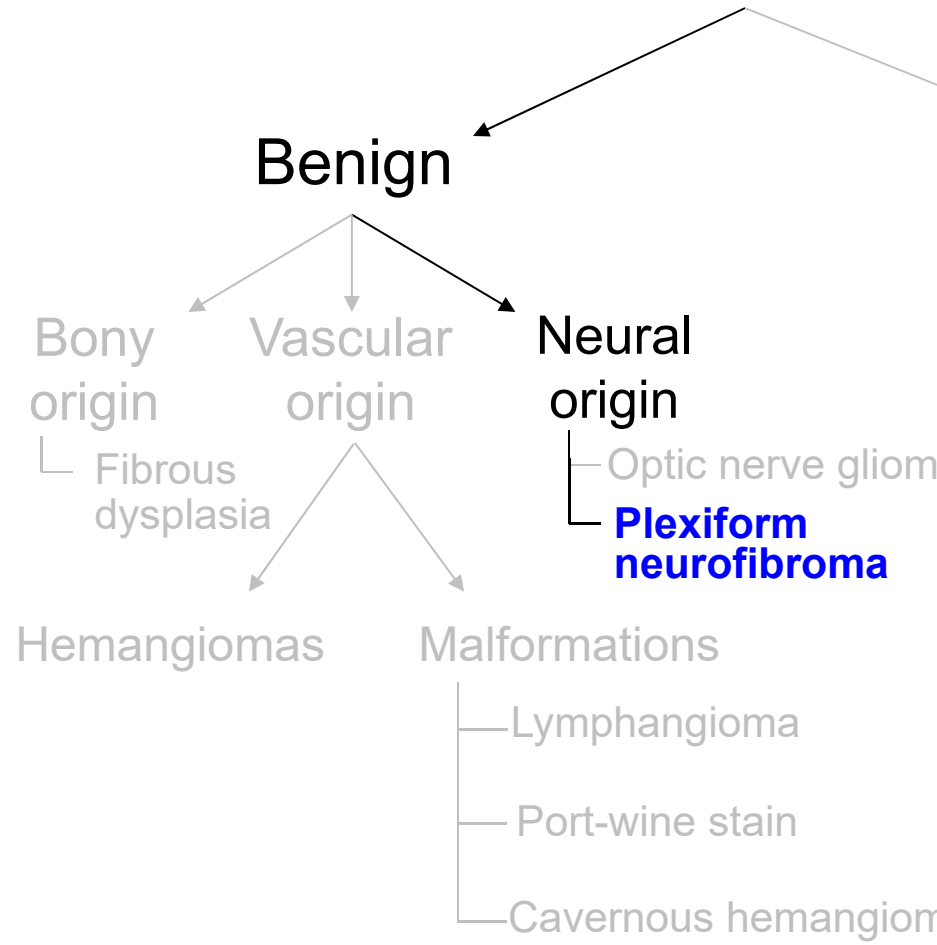
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Pediatric Orbital Tumors



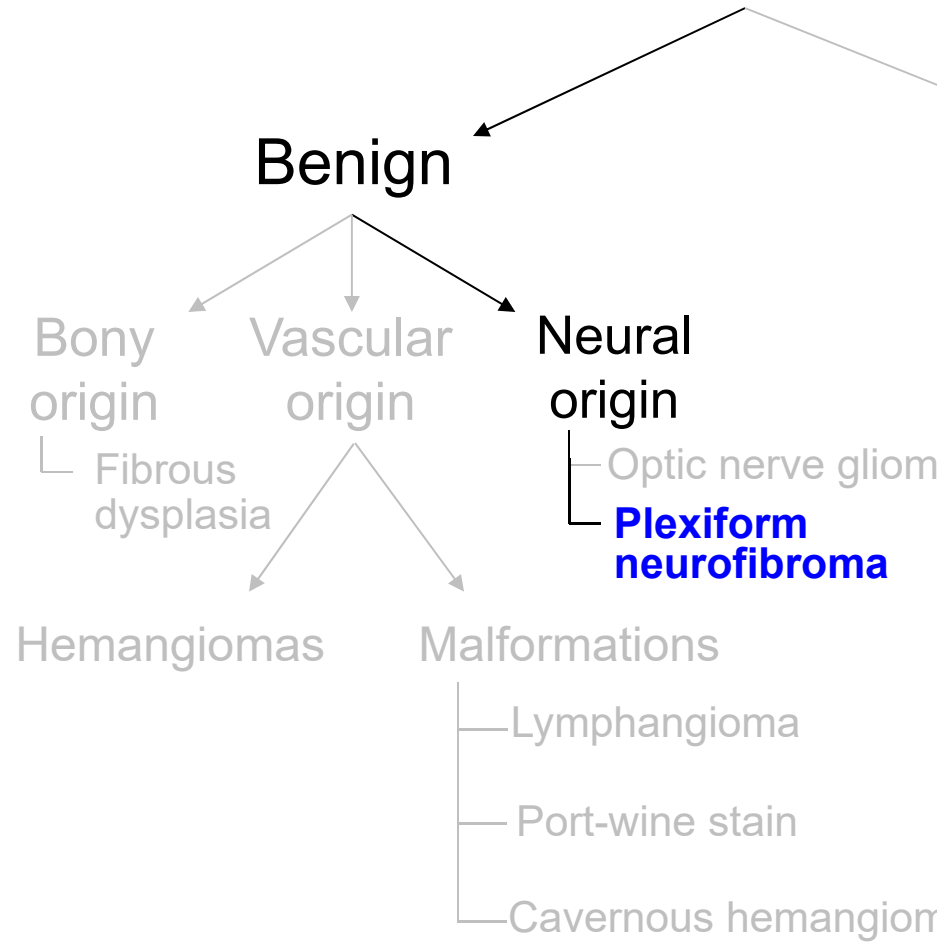
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Pediatric Orbital Tumors



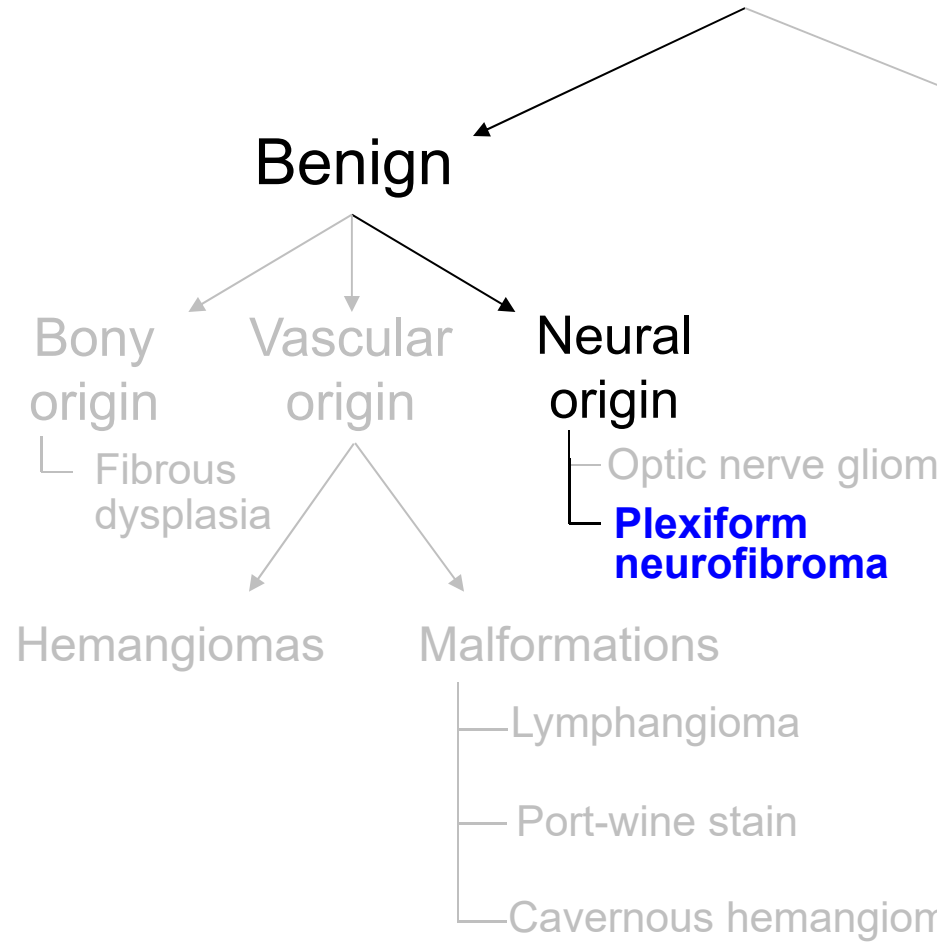
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Pediatric Orbital Tumors

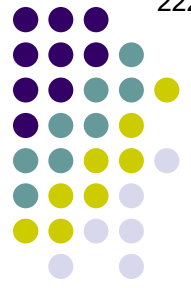


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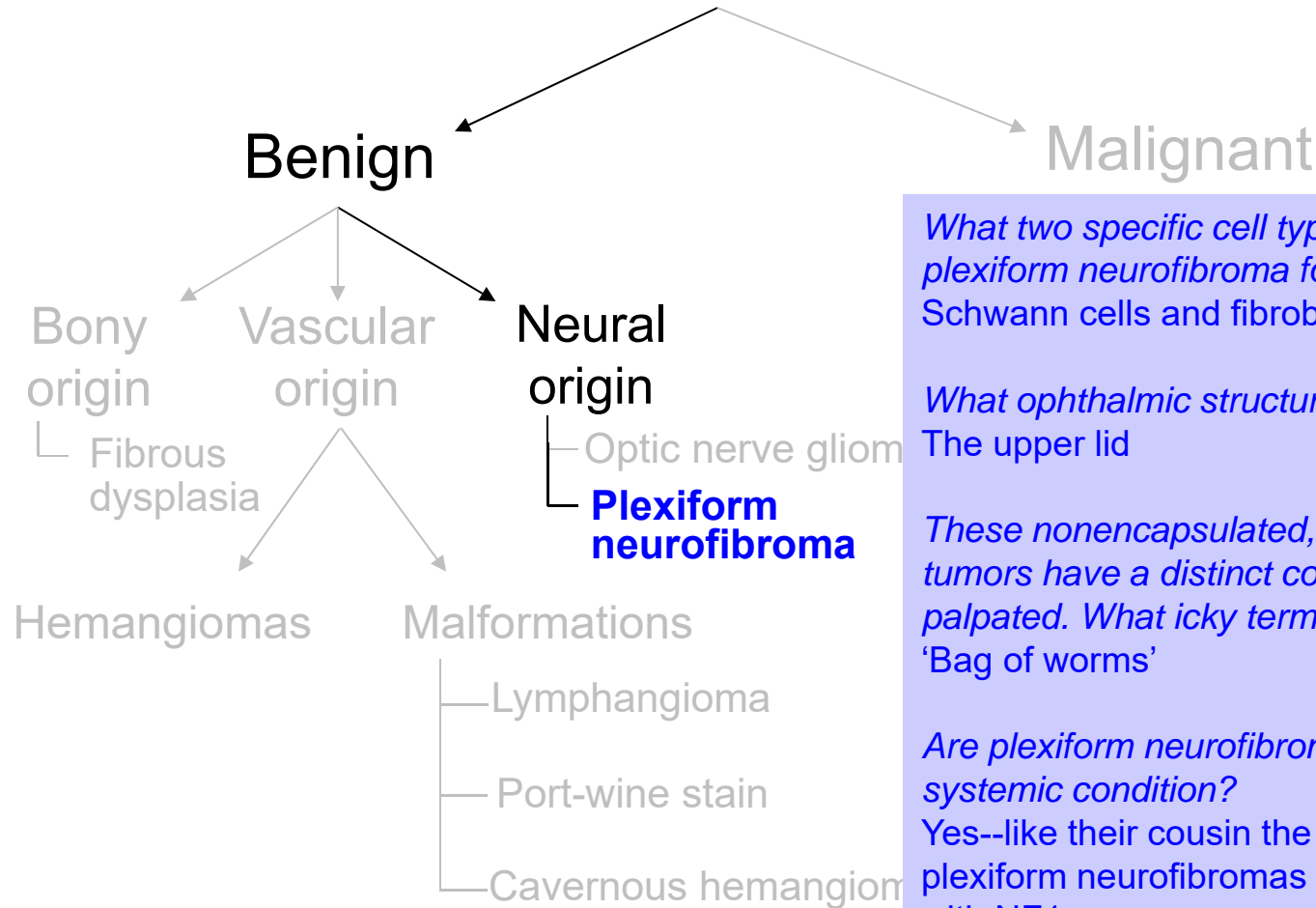
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Are plexiform neurofibromas associated with a systemic condition?



Pediatric Orbital Tumors

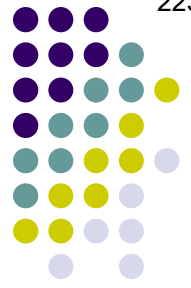


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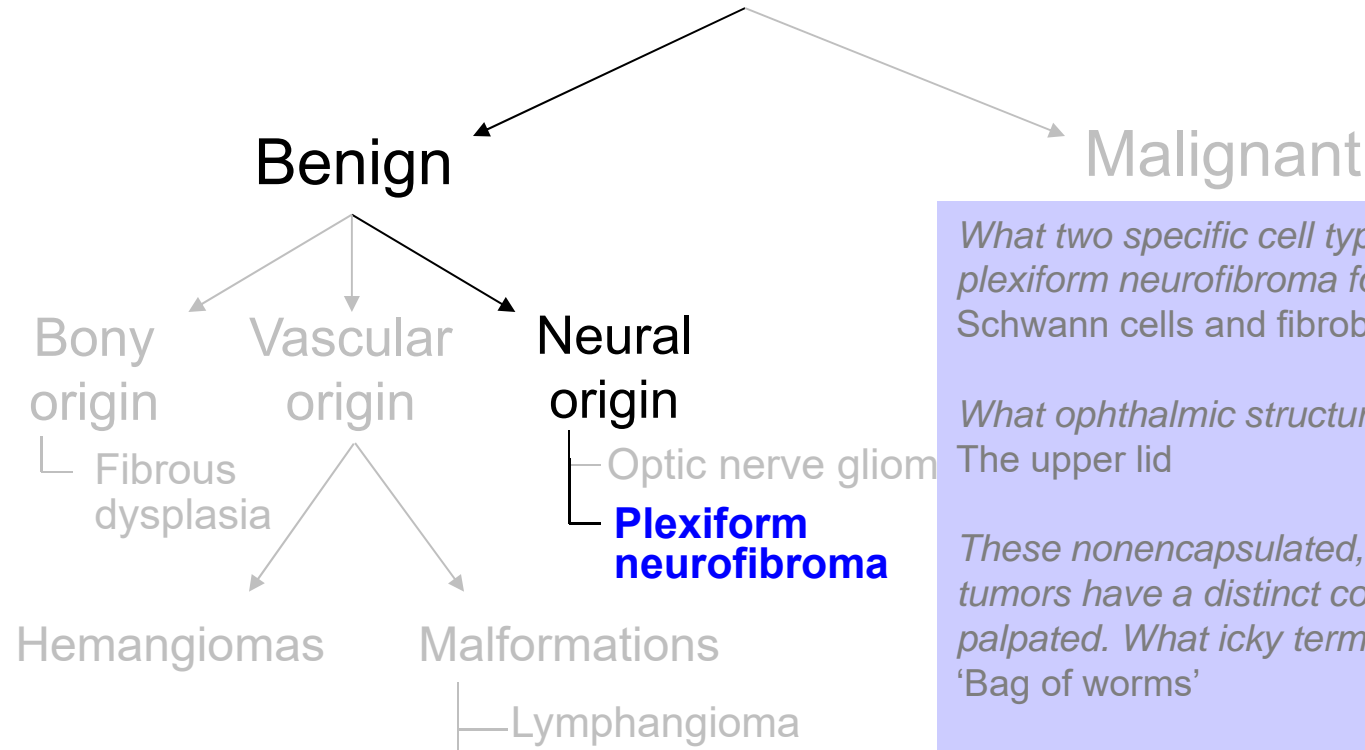
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Pediatric Orbital Tumors



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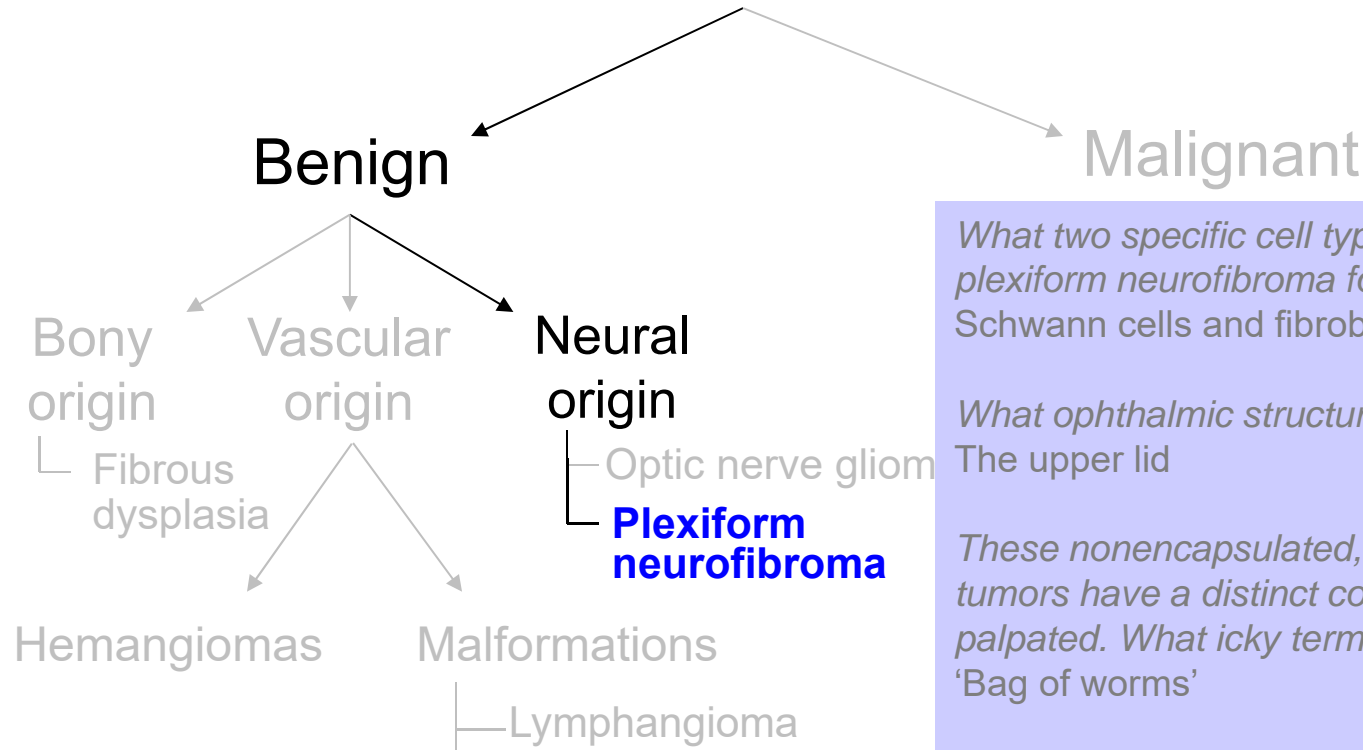
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What proportion of NF1 pts develop plexiform neurofibromas?



Pediatric Orbital Tumors



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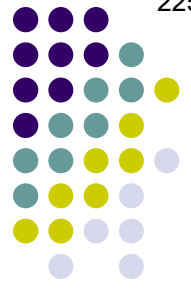
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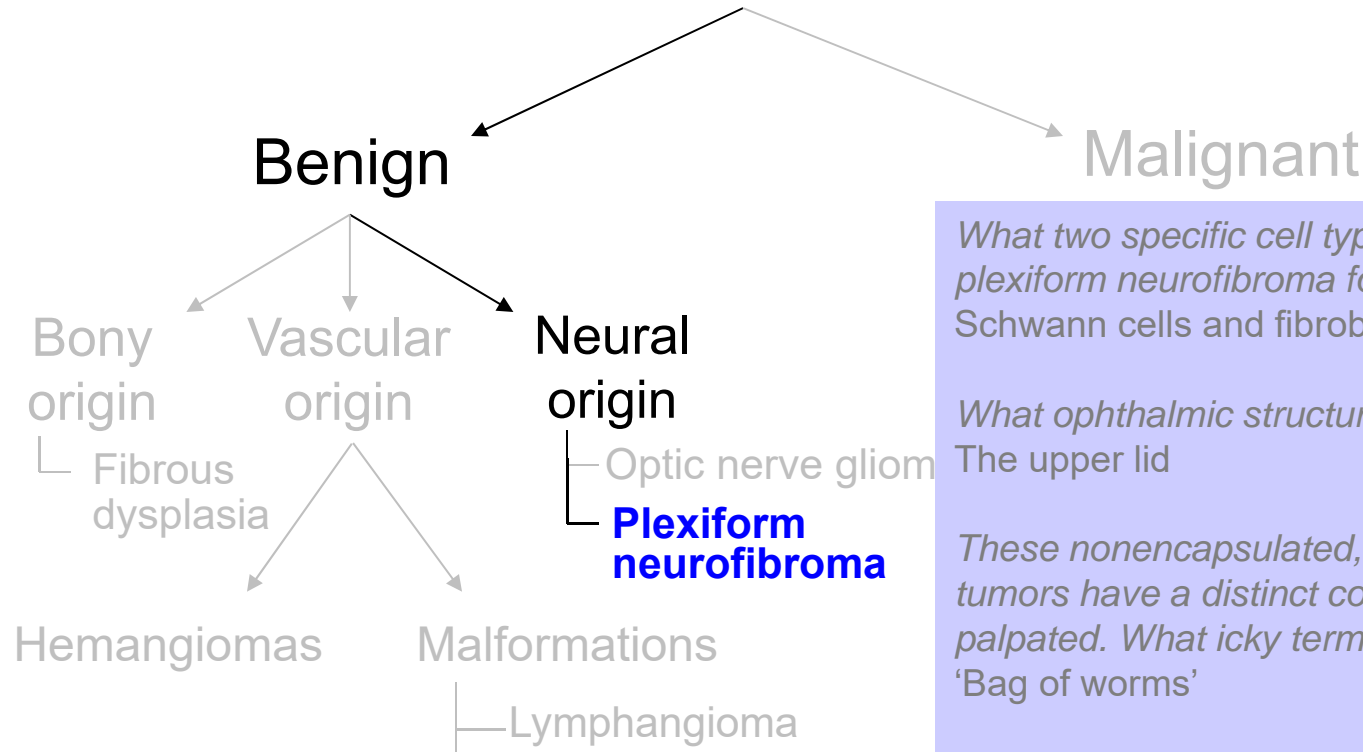
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Pediatric Orbital Tumors



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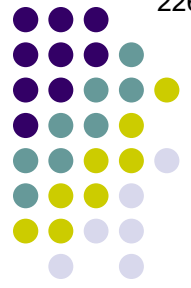
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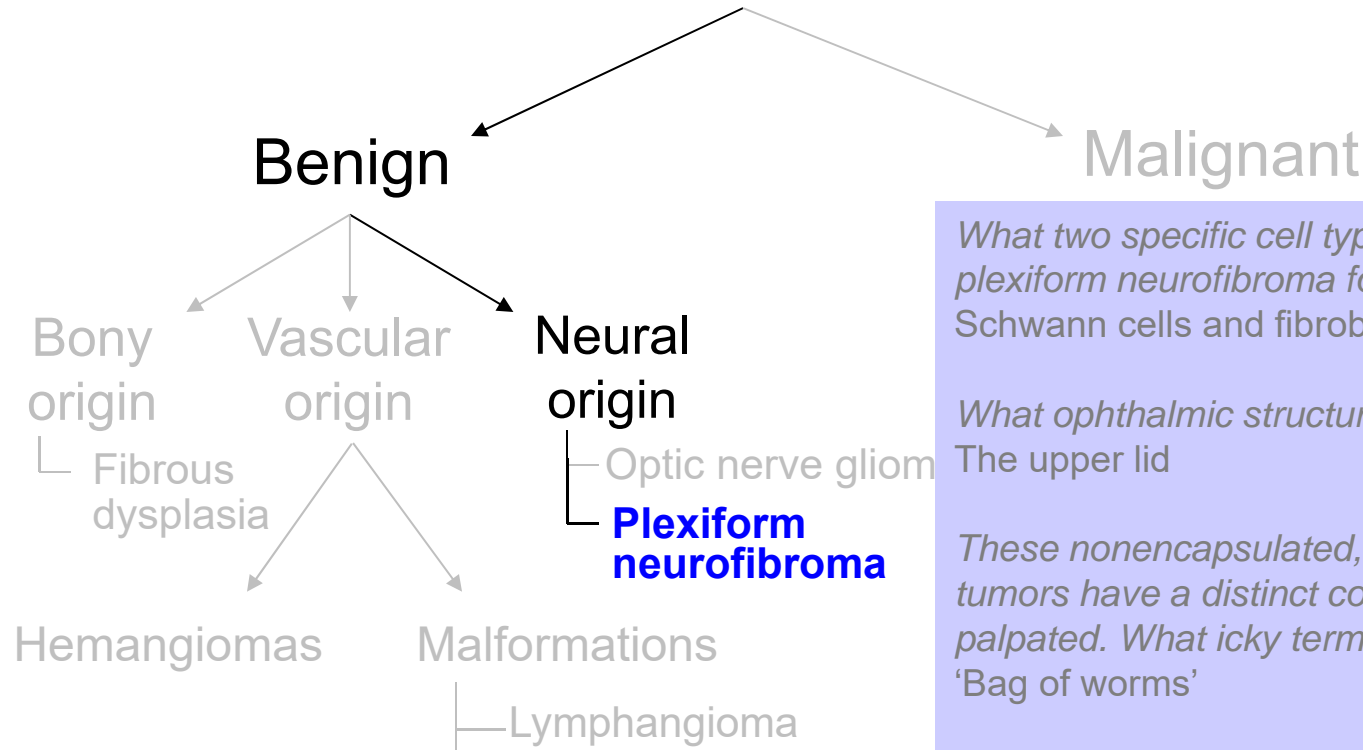
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Pediatric Orbital Tumors



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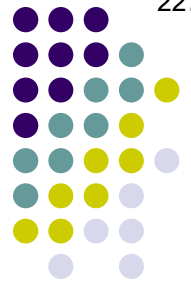
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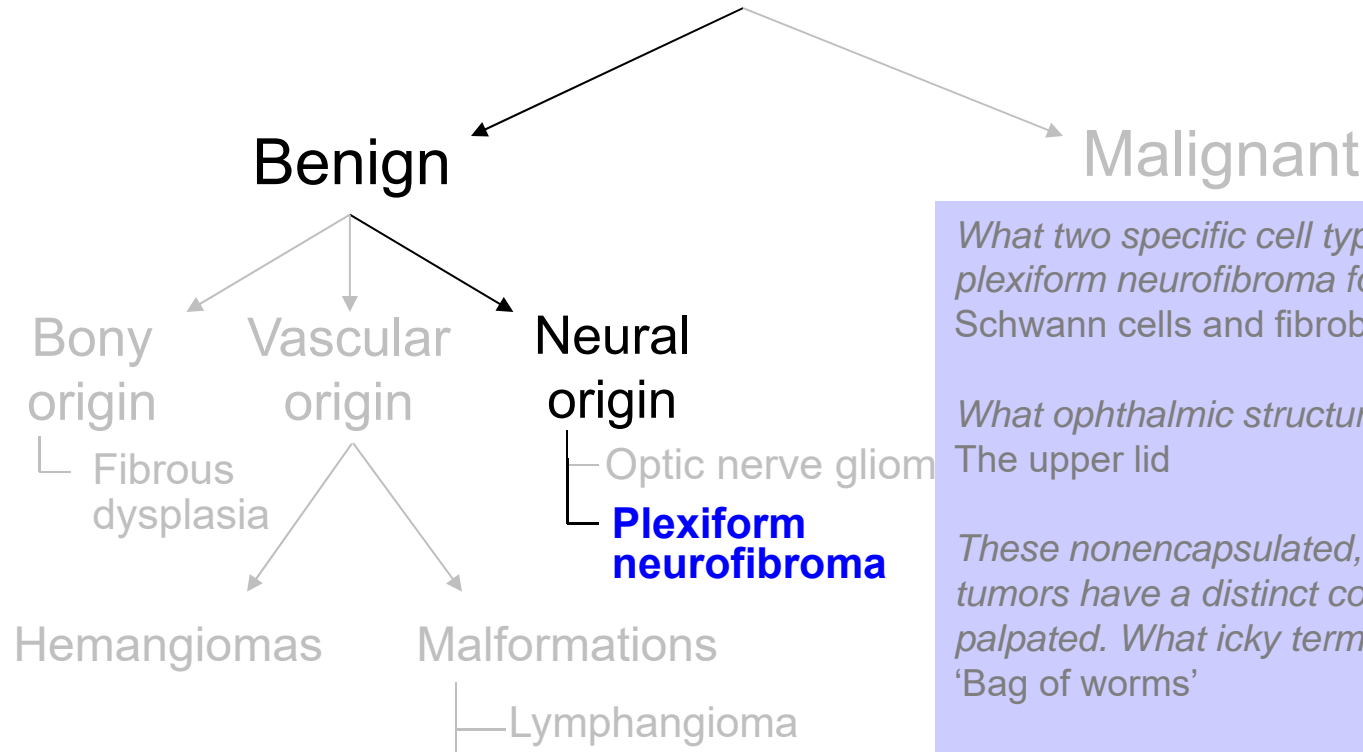
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What proportion of plexiform neurofibroma pts have NF1?
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NF1



Pediatric Orbital Tumors



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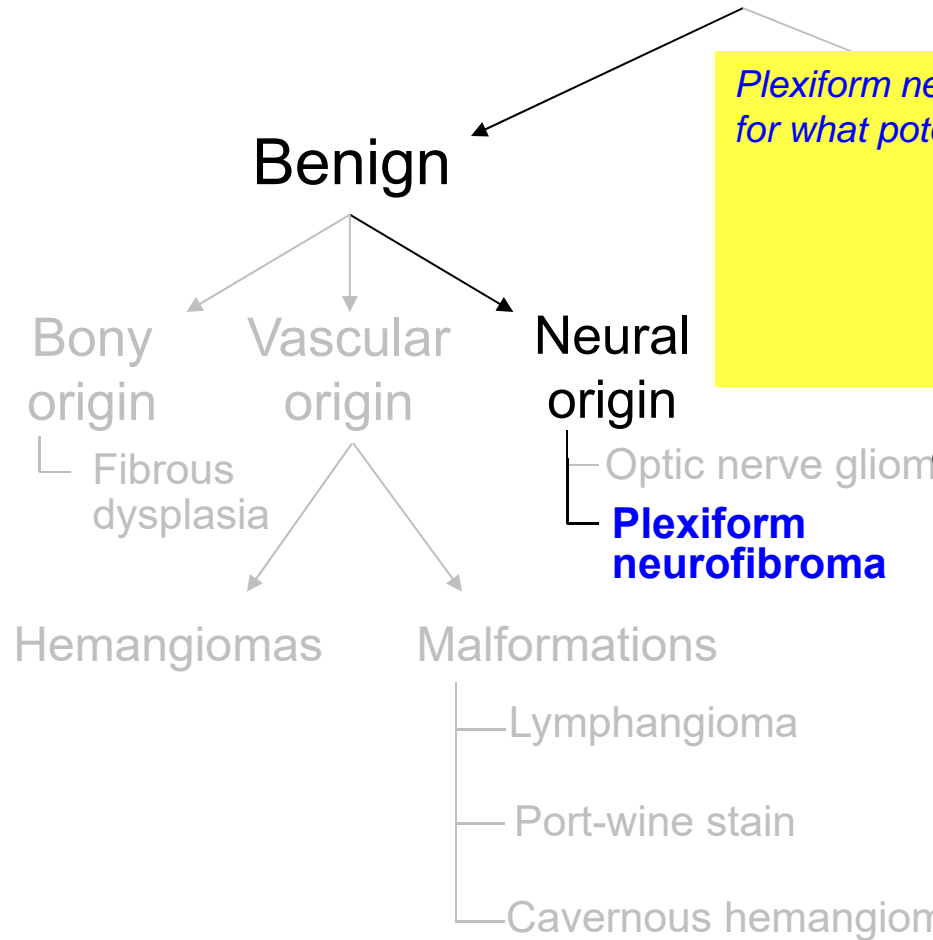
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tl;dr Not all NF1 pts get plexiform neurofibromas, but everyone with plexiform neurofibromas has NF1



Pediatric Orbital Tumors



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

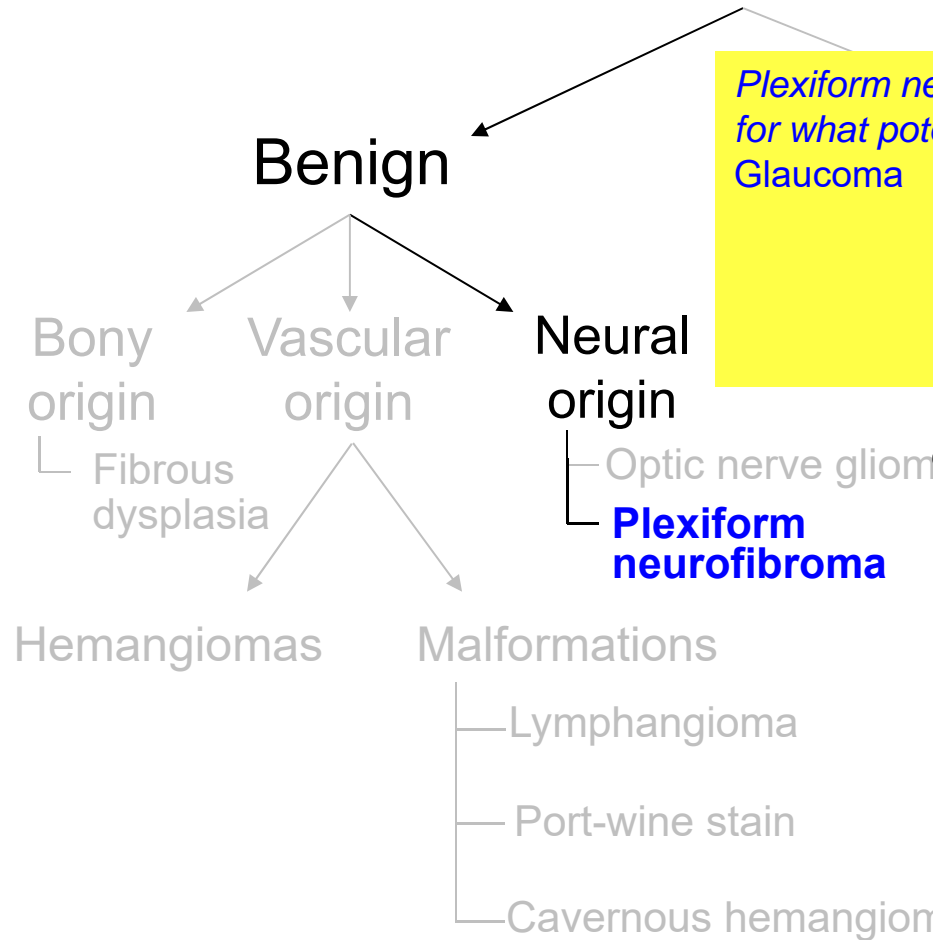
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Pediatric Orbital Tumors



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Glaucoma

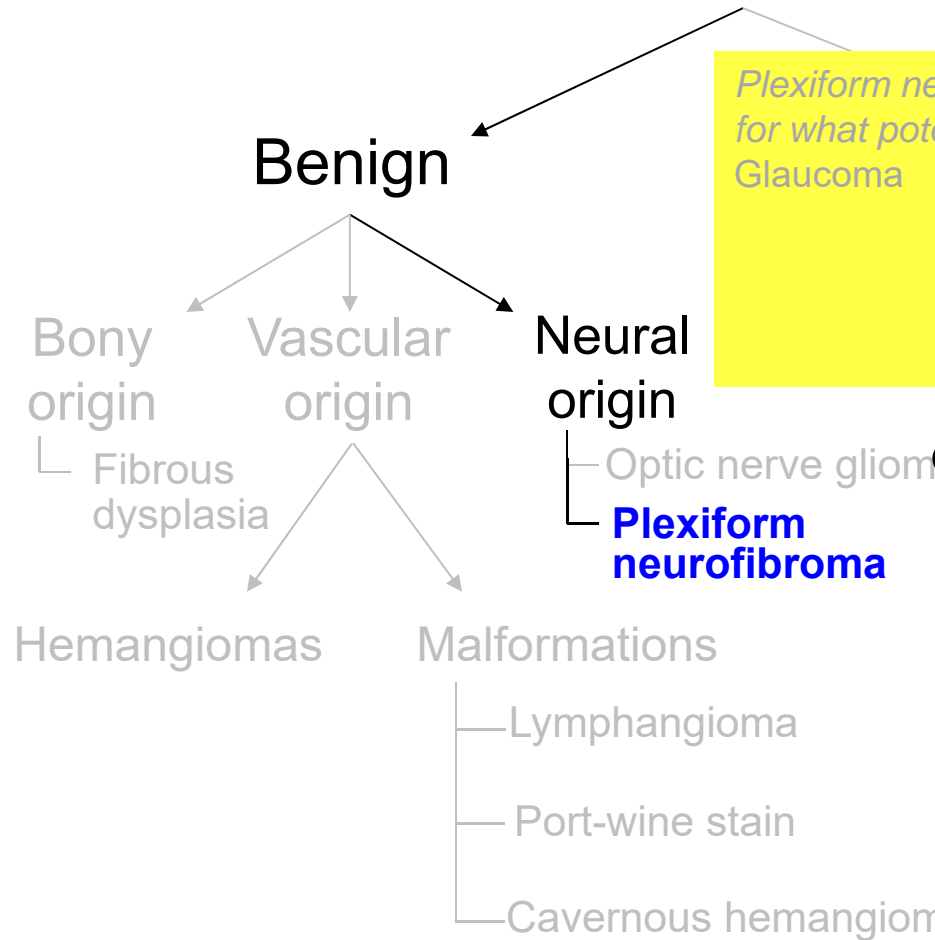
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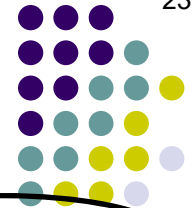
*Plexiform neurofibromas of the upper lid area are a **strong** risk factor for what potentially blinding ocular condition?*
 Glaucoma

How strong of a risk factor?

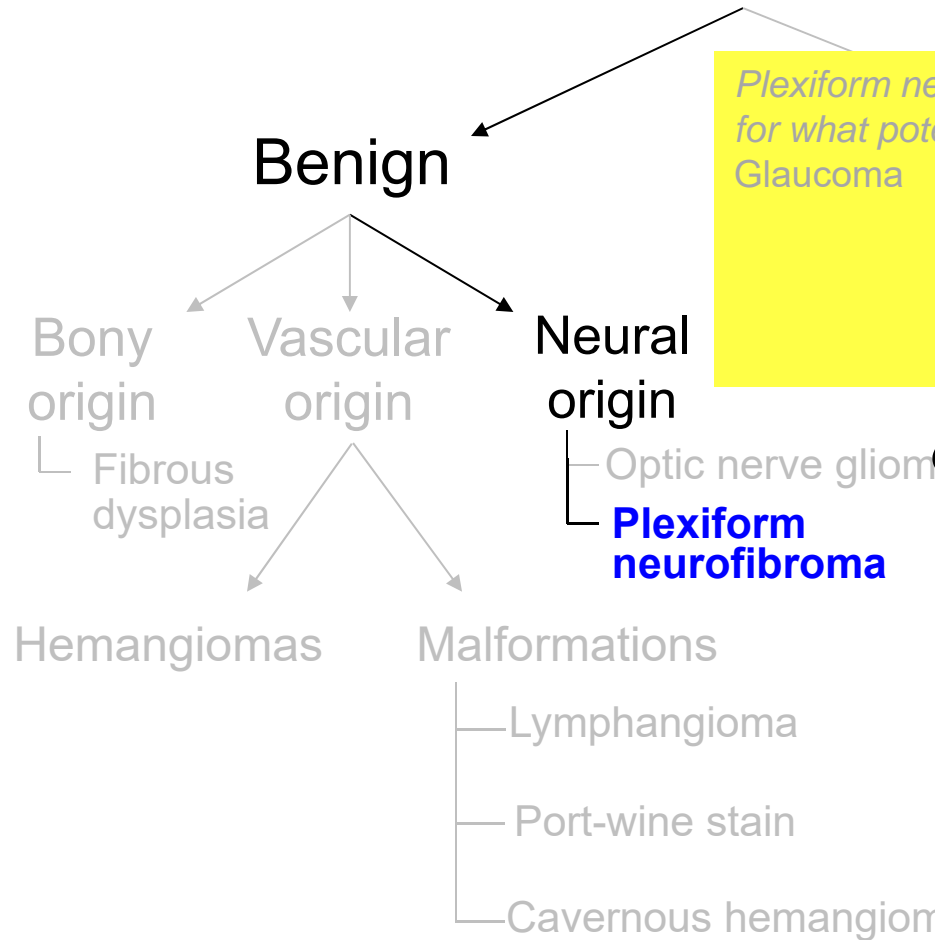
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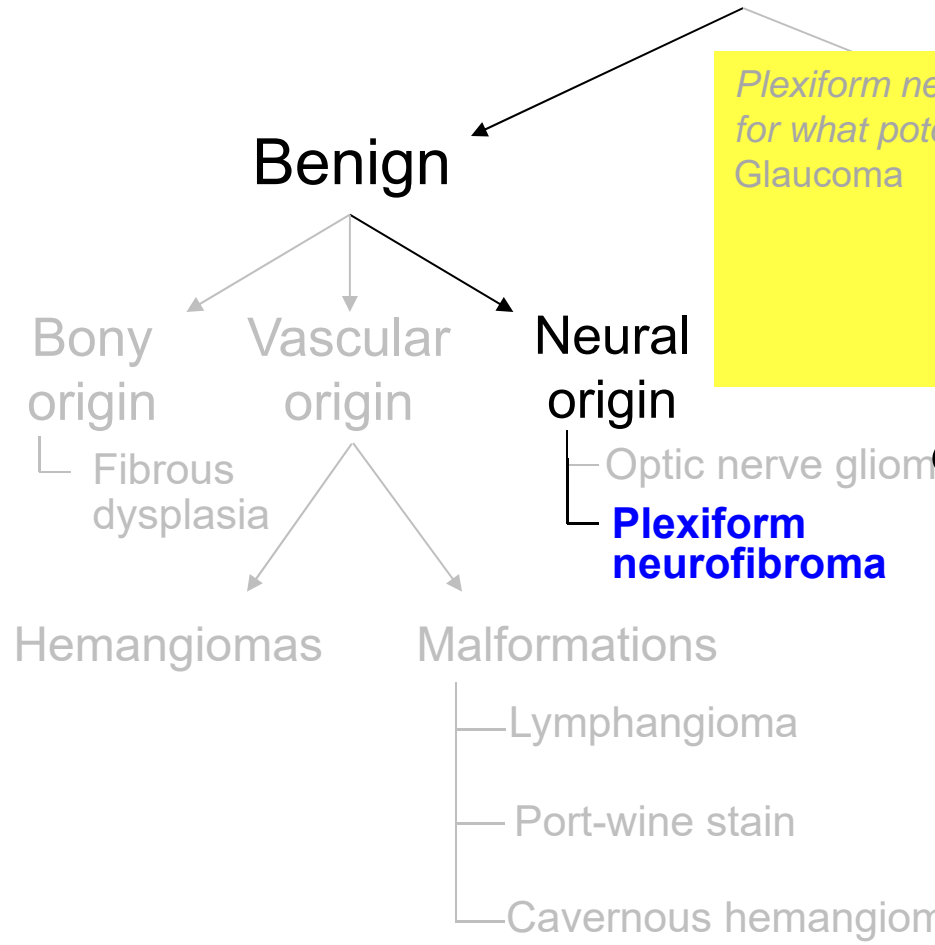
How strong of a risk factor?
Very strong. % of these eyes have glaucoma!

What ophthalmic structure is typically involved?
The upper lid

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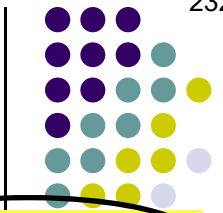
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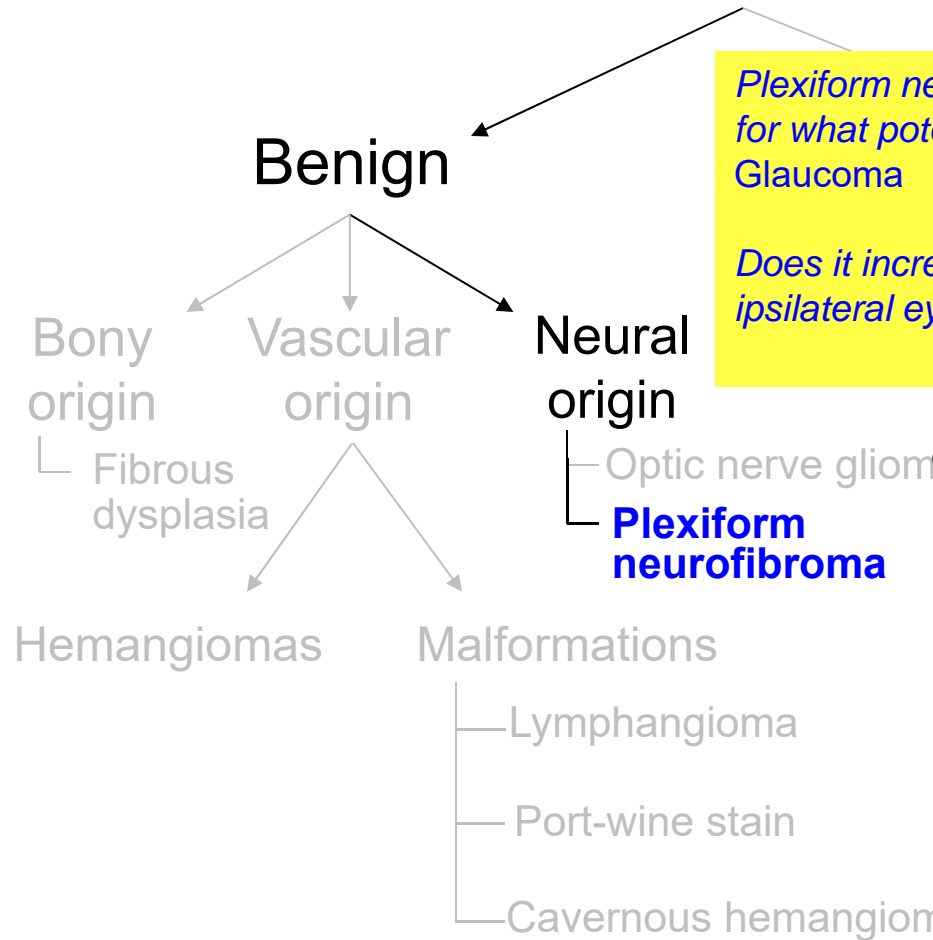
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Glaucoma*

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

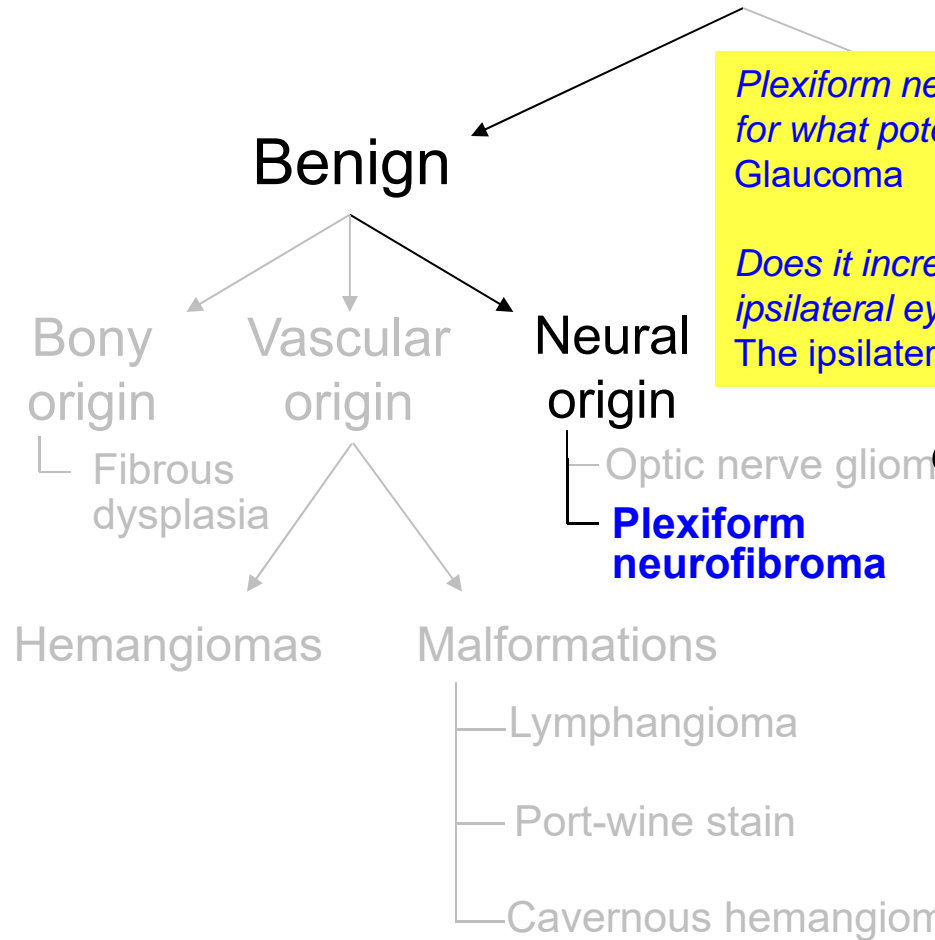
*What ophthalmic structure is typically involved?
The upper lid*

*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



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 only

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