

Q

Vascular Lesions of the Orbit

Two
basic lesion types

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Vascular Lesions of the Orbit

*Two
basic lesion types*

Hemangiomas

Malformations



Q

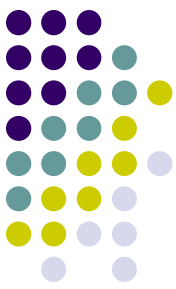
Vascular Lesions of the Orbit

Hemangiomas

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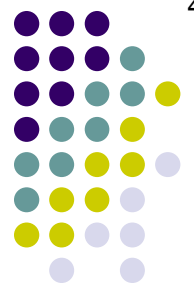
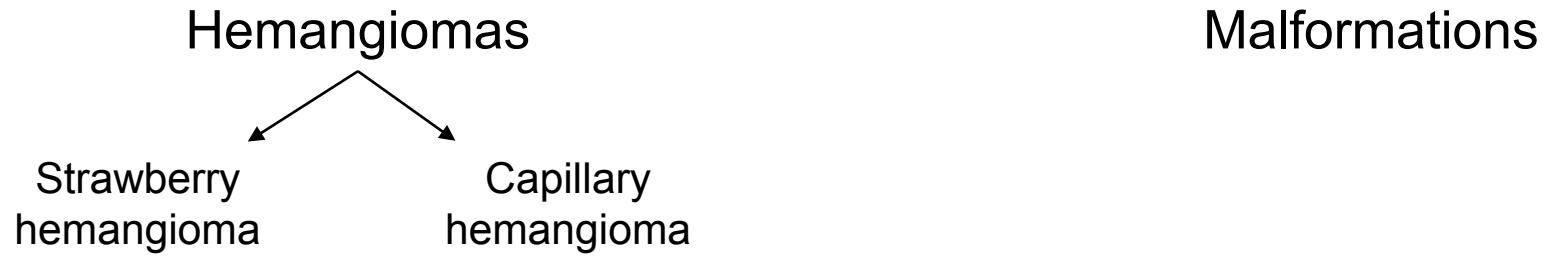
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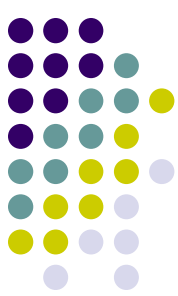
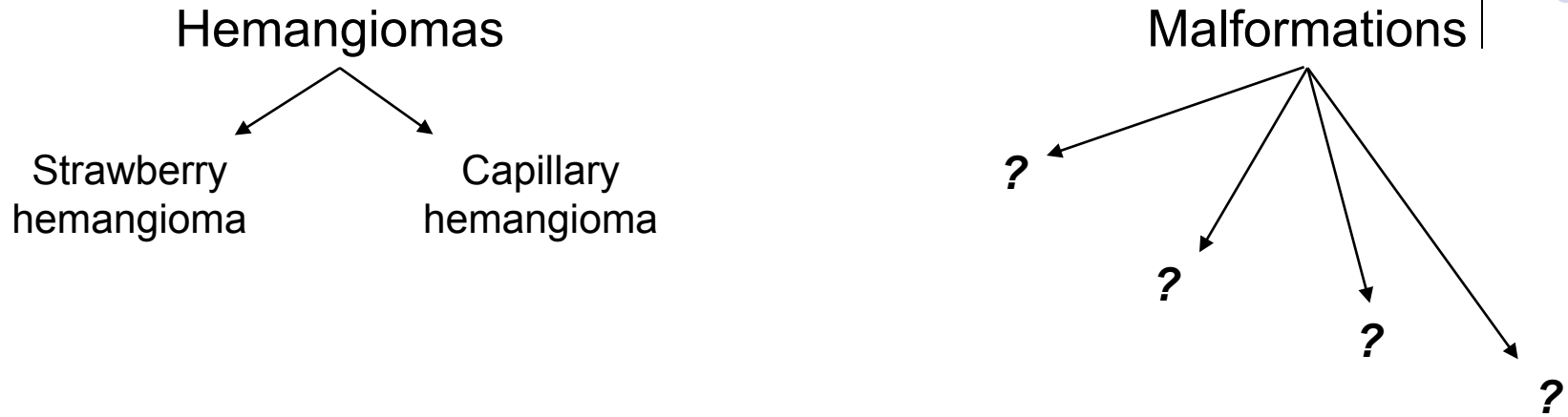
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Vascular Lesions of the Orbit



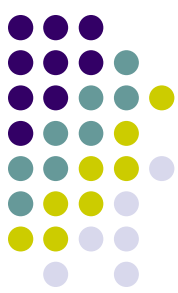
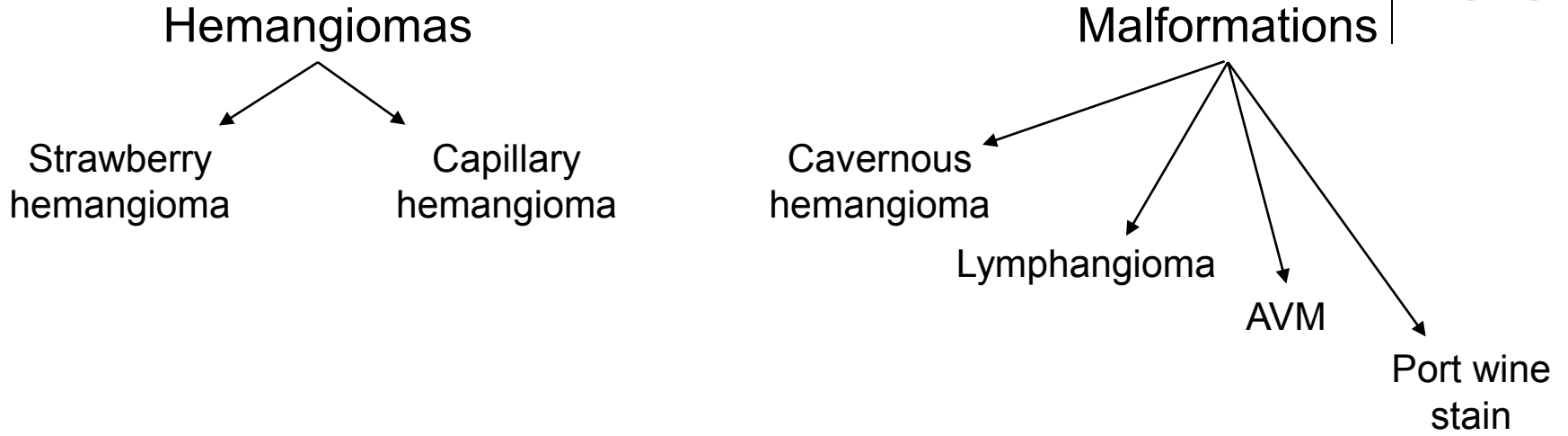
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Vascular Lesions of the Orbit

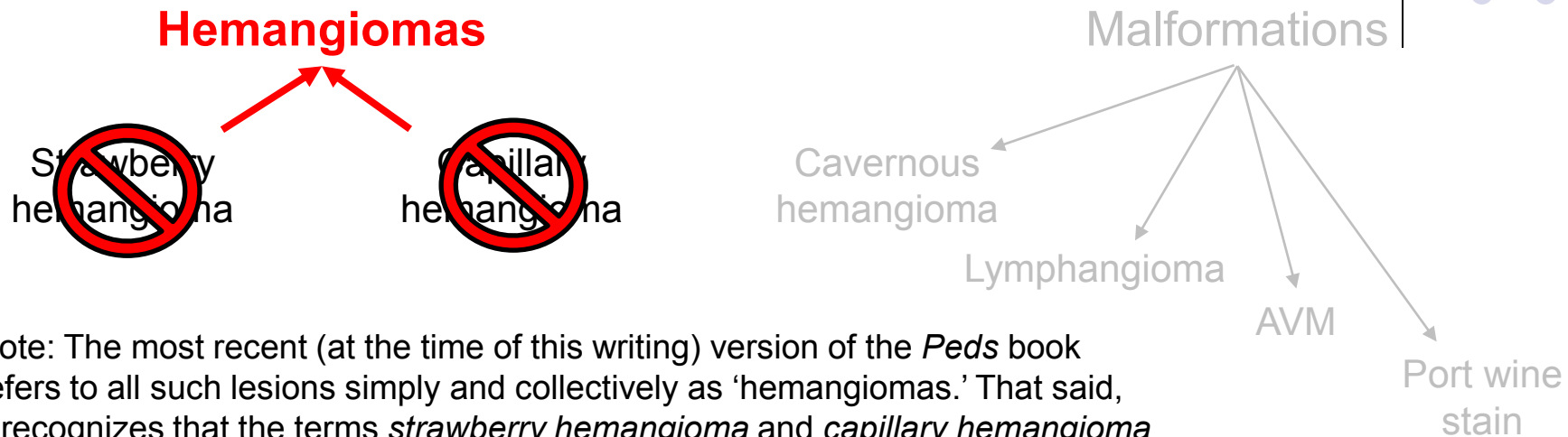


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Vascular Lesions of the Orbit



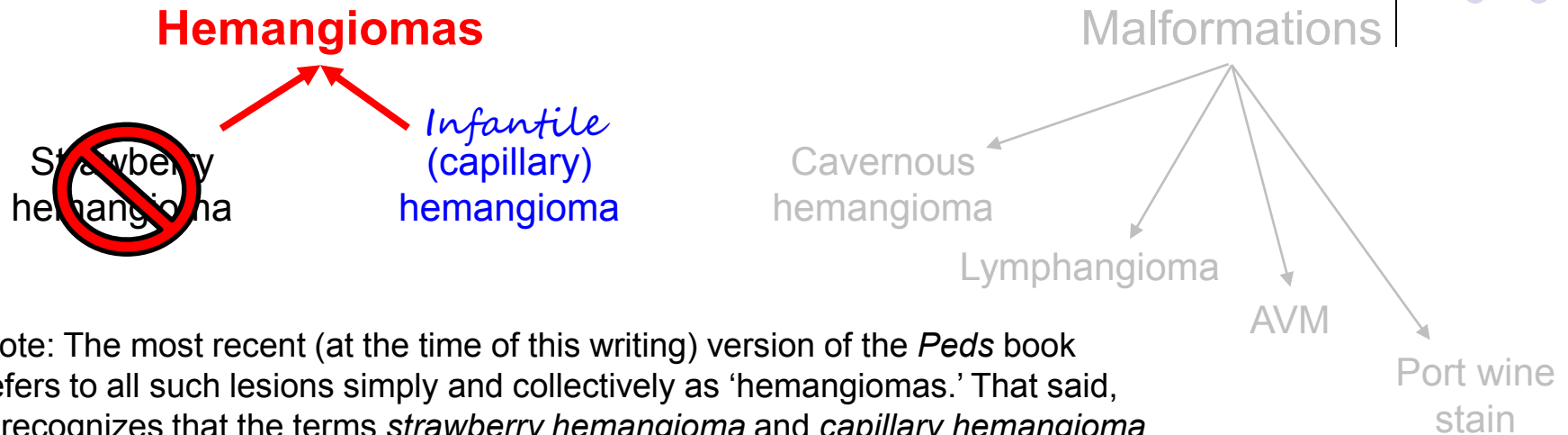
Vascular Lesions of the Orbit



Note: The most recent (at the time of this writing) version of the *Peds* book refers to all such lesions simply and collectively as 'hemangiomas.' That said, it recognizes that the terms *strawberry hemangioma* and *capillary hemangioma* are deeply embedded in the ophthalmic literature.

No question—proceed when ready

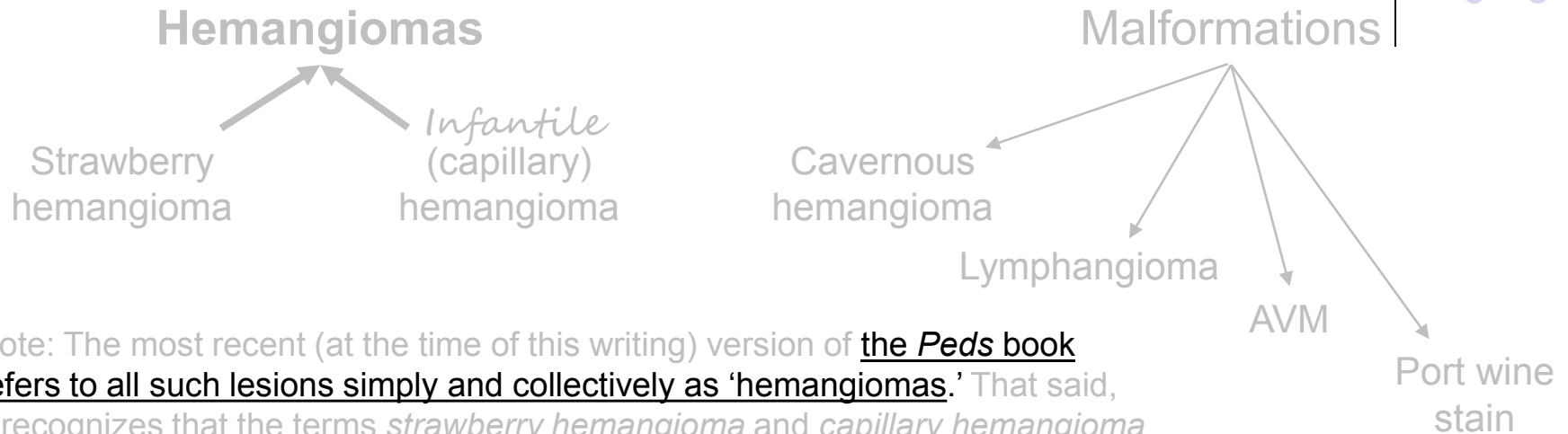
Vascular Lesions of the Orbit



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Vascular Lesions of the Orbit



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Foreshadowing alert: This is the first of several inconsistencies we will encounter among the *BCSC* books regarding hemangiomas (some will not be so trivial as this)

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Vascular Lesions of the Orbit



Note also that the *Peds* (but not *Orbit*) book divvies ophthalmic hemangiomas into three groups based on their location:

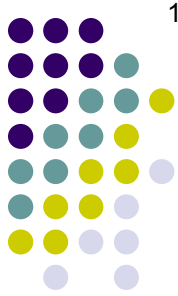
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Vascular Lesions of the Orbit



Hemangiomas

Malformations



Note also that the *Peds* (but not *Orbit*) book divvies ophthalmic hemangiomas into three groups based on their location:

--Those involving the skin and/or preseptal orbit;

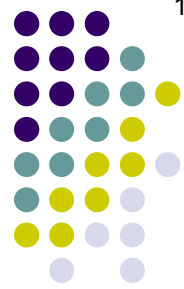
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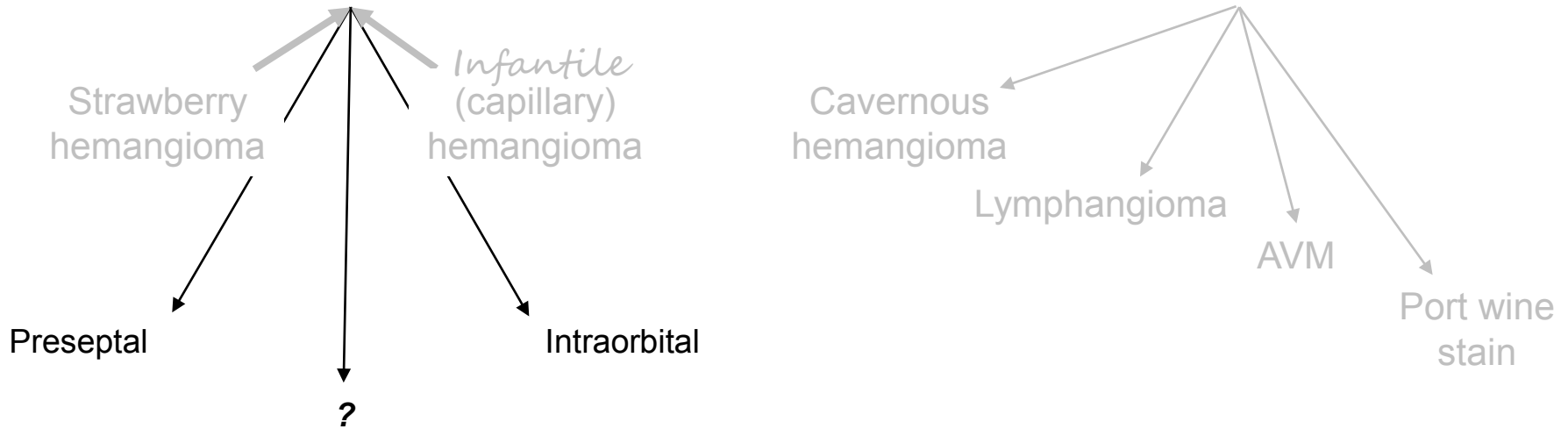
Vascular Lesions of the Orbit

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Hemangiomas

Malformations



Note also that the *Peds* (but not *Orbit*) book divvies ophthalmic hemangiomas into three groups based on their location:

- Those involving the skin and/or preseptal orbit;
- those that are fully posterior to the septum; and
- ?

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Vascular Lesions of the Orbit



Note also that the *Peds* (but not *Orbit*) book divvies ophthalmic hemangiomas into three groups based on their location:

- Those involving the skin and/or preseptal orbit;
- those that are fully posterior to the septum; and
- those that involve **both** sides of the septum.

Vascular Lesions of the Orbit



Note also that the **Peds (but not Orbit) book** divvies ophthalmic hemangiomas into three groups based on their location:

--Those involving the skin and/or preseptal orbit:

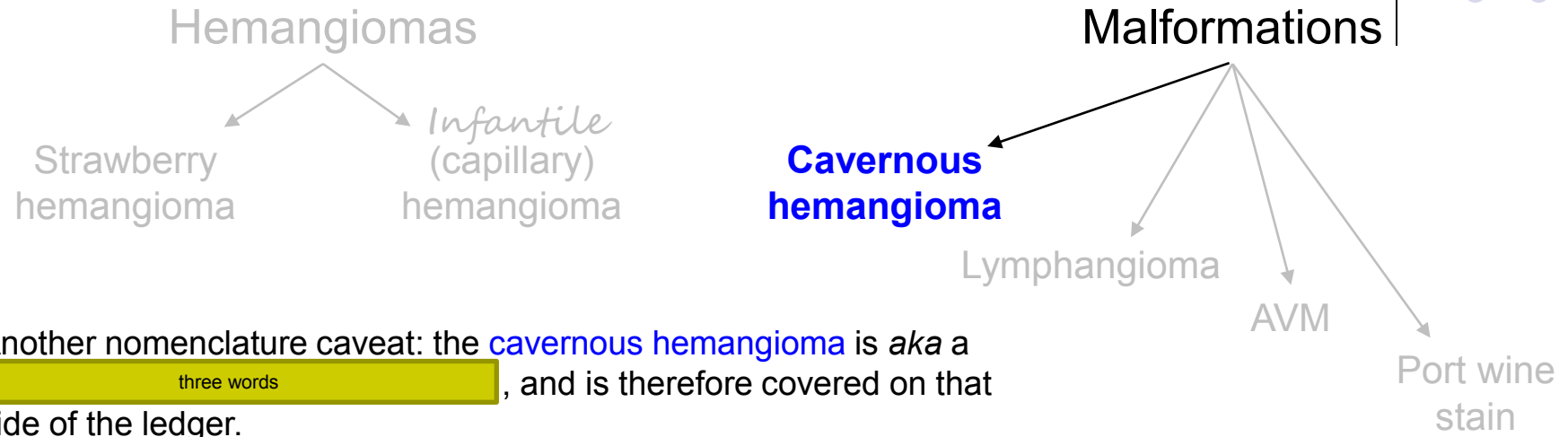
Another, but neither the last nor most important

--those that involve both sides of the septum.

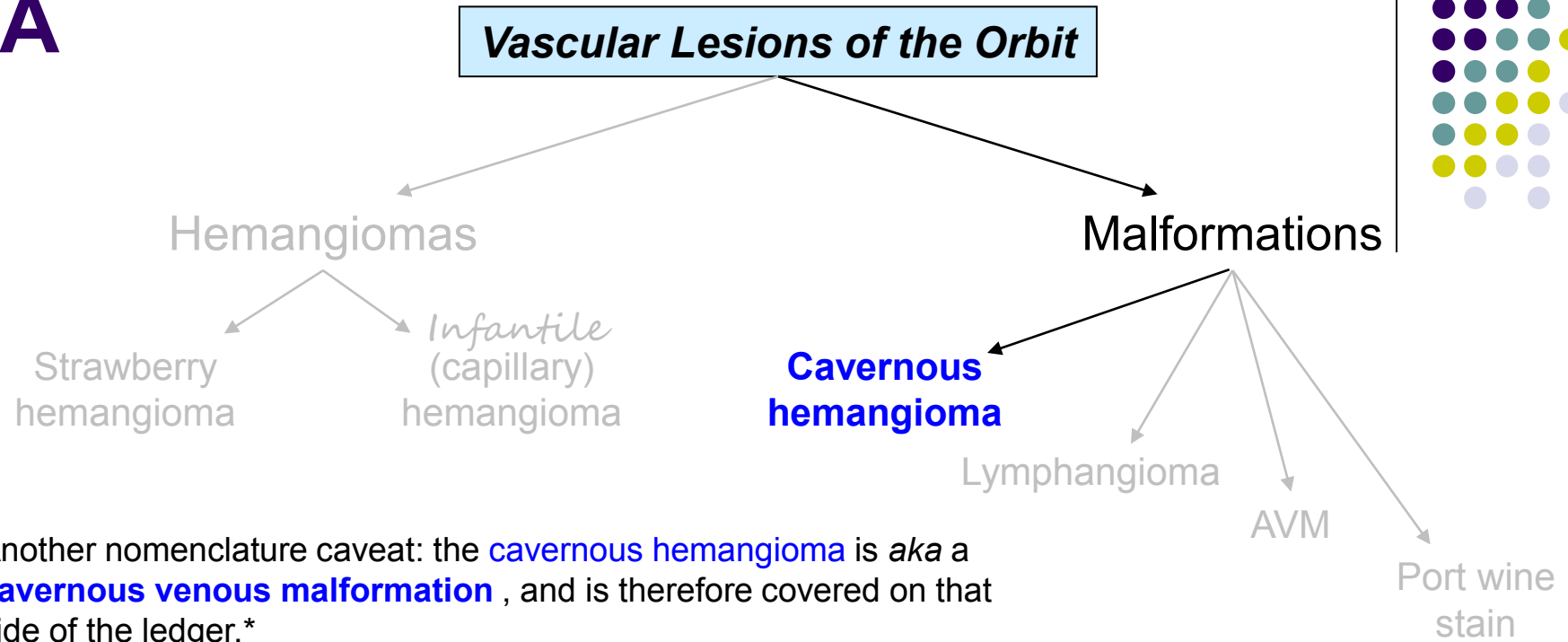
No question—proceed when ready

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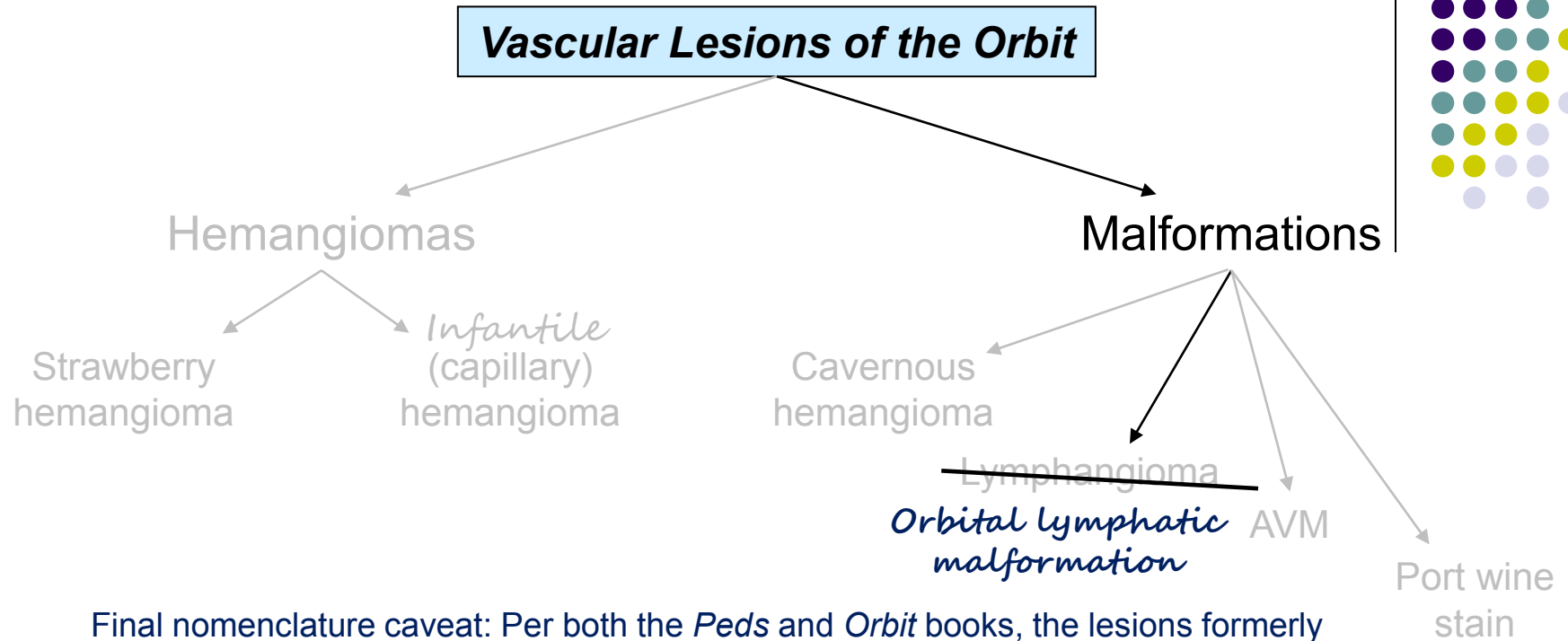
Vascular Lesions of the Orbit



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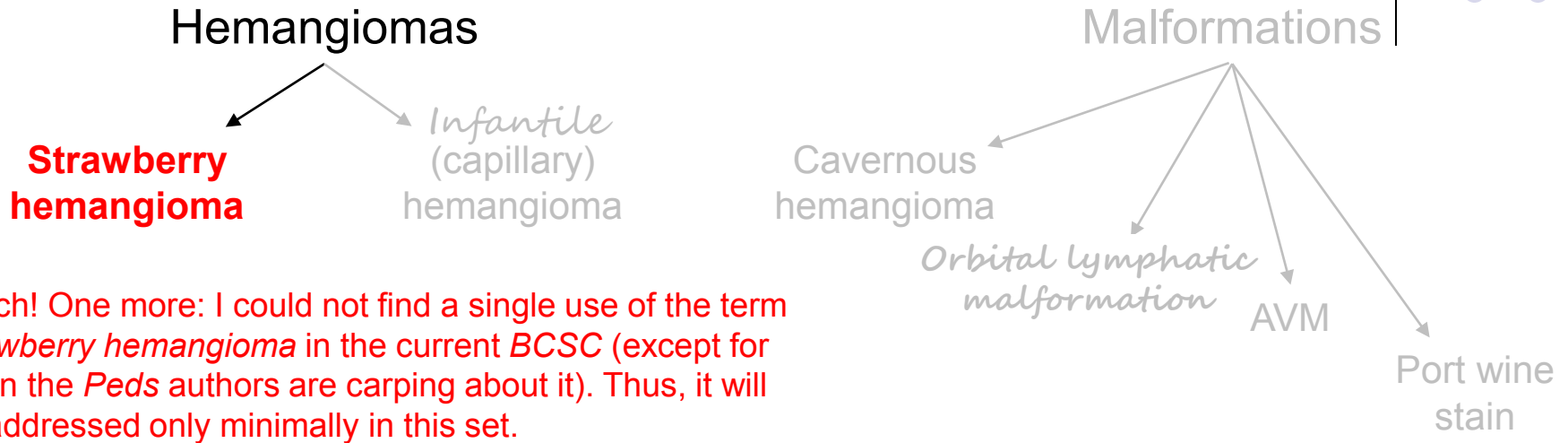
*One section of the most recent (at the time of this writing) version of the *Plastics* book indicates that *cavernous venous malformations* is now the **preferred** term for this lesion. That said, other current *BCSC* volumes continue to use the term cavernous hemangioma—as do other sections of the *Plastics* book, for that matter. So what's the 'proper' term? Beats me. Caveat emptor.



Final nomenclature caveat: Per both the *Peds* and *Orbit* books, the lesions formerly known as *lymphangiomas* are now called **orbital lymphatic malformations**.

No question—proceed when ready

Vascular Lesions of the Orbit

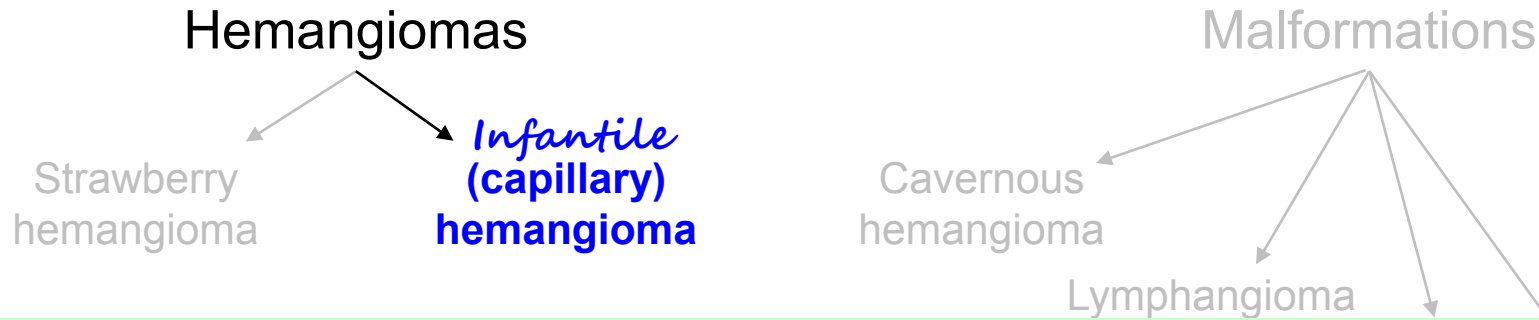


Psych! One more: I could not find a single use of the term *strawberry hemangioma* in the current *BCSC* (except for when the *Peds* authors are carping about it). Thus, it will be addressed only minimally in this set.

No question—proceed when ready

Q

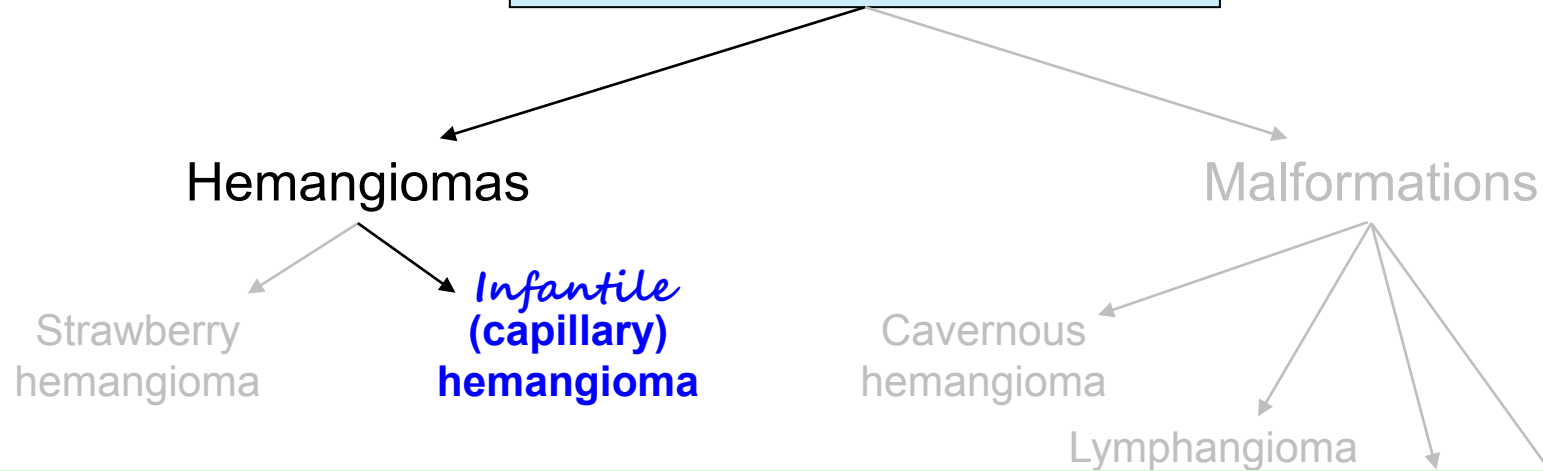
Vascular Lesions of the Orbit



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



Vascular Lesions of the Orbit

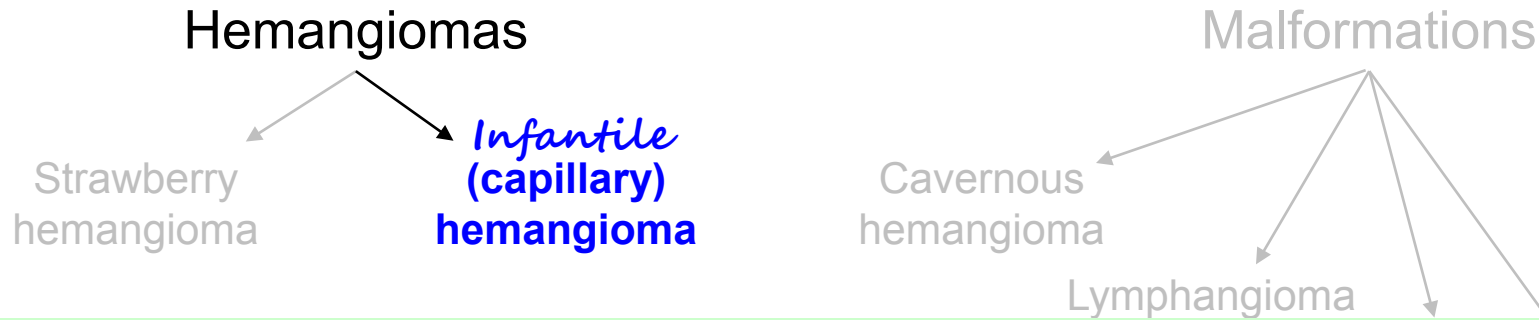


Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

It is the most v least common

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Vascular Lesions of the Orbit

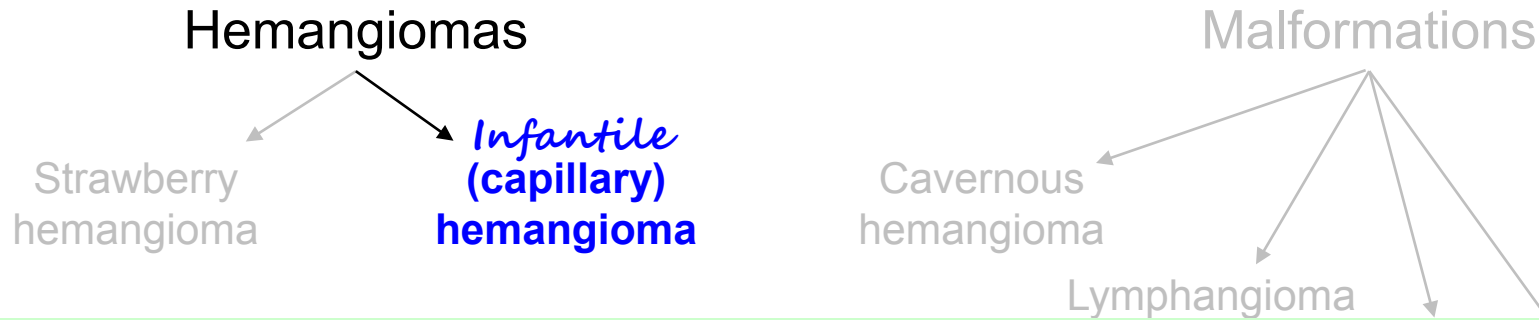


Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?
It is the most common



Q

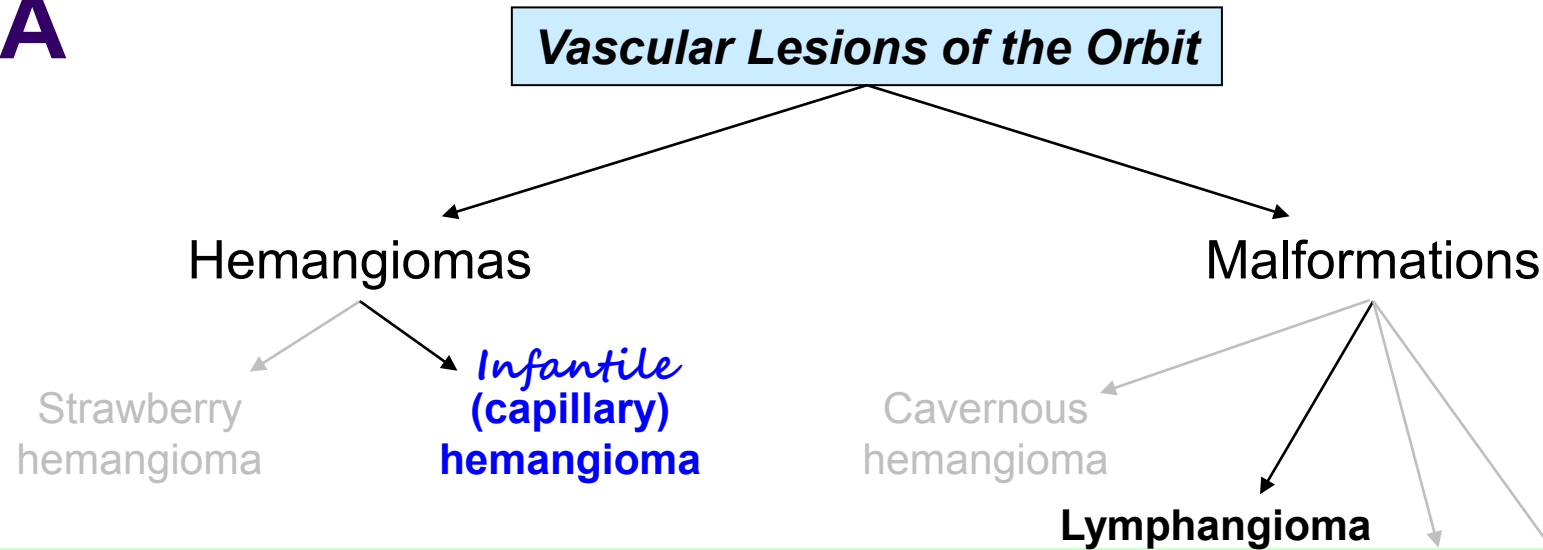
Vascular Lesions of the Orbit



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

What vascular tumor is the next most common?

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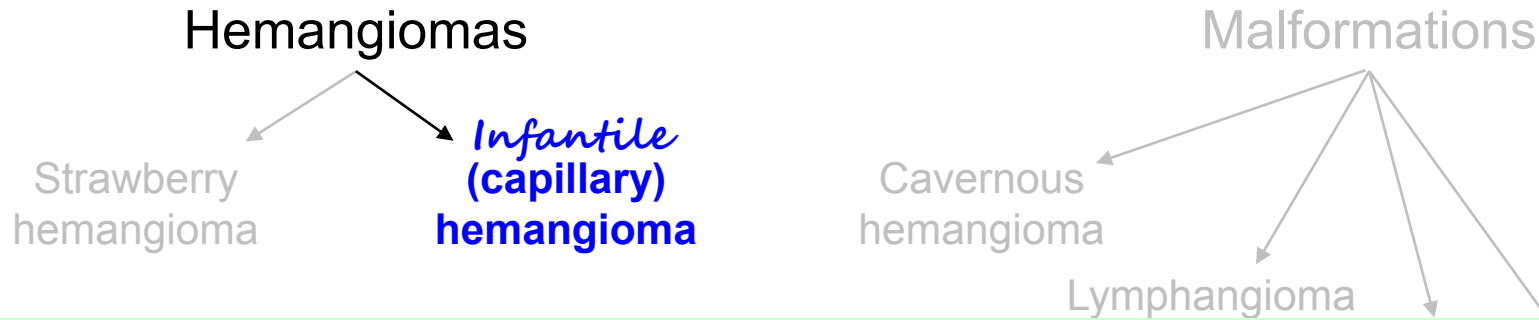


Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?
 It is ^{second} the most common?

What vascular tumor is the next most common?
 Orbital lymphangioma

Q

Vascular Lesions of the Orbit

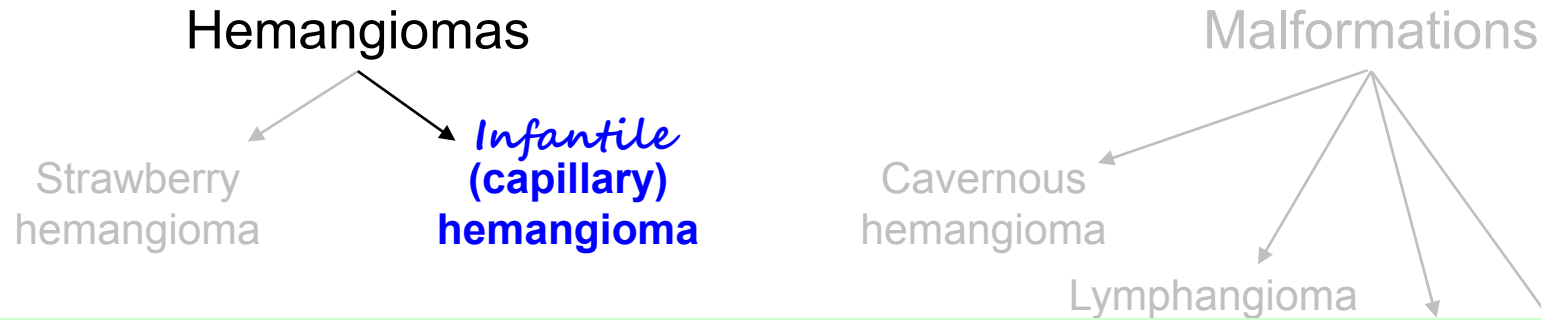


*Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?
It is the most common*

Is there a gender predilection?

Q/A

Vascular Lesions of the Orbit



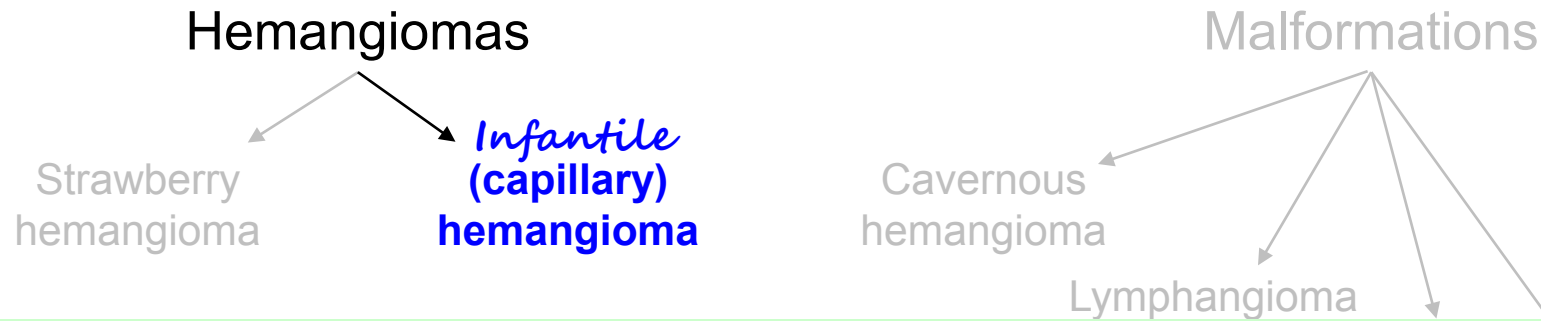
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It is the most common

Is there a gender predilection?

Yes, it is more common in M v F

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Vascular Lesions of the Orbit

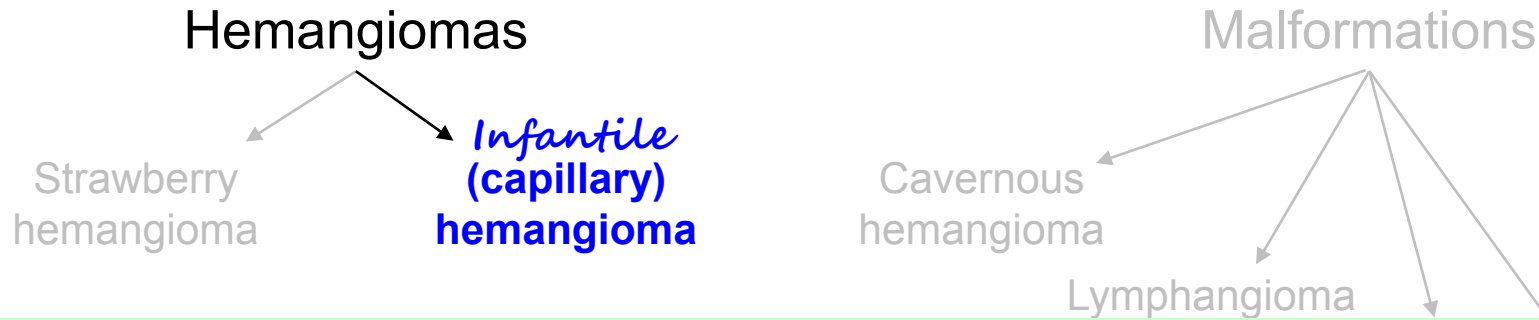


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Is there a gender predilection?
Yes, it is more common in girls

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Vascular Lesions of the Orbit



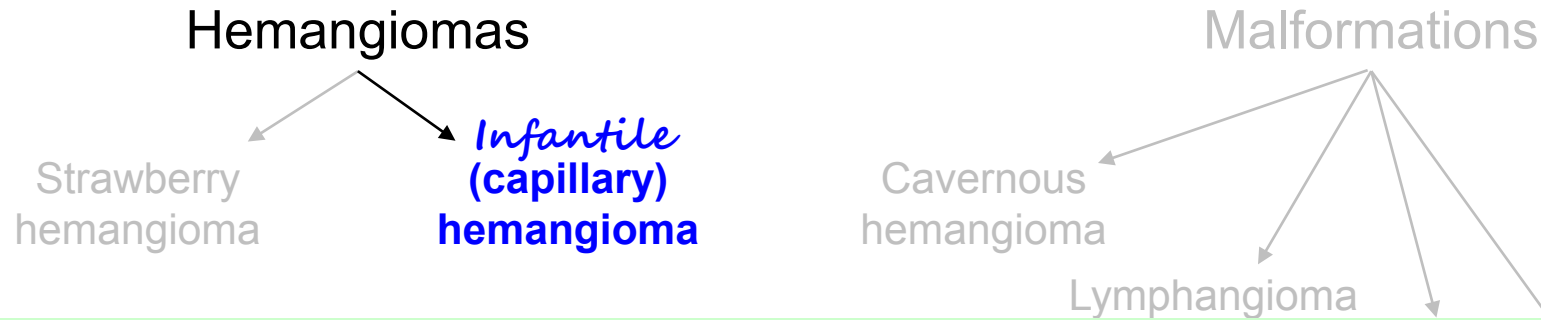
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Female gender is the major risk factor. What is another one?

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Vascular Lesions of the Orbit



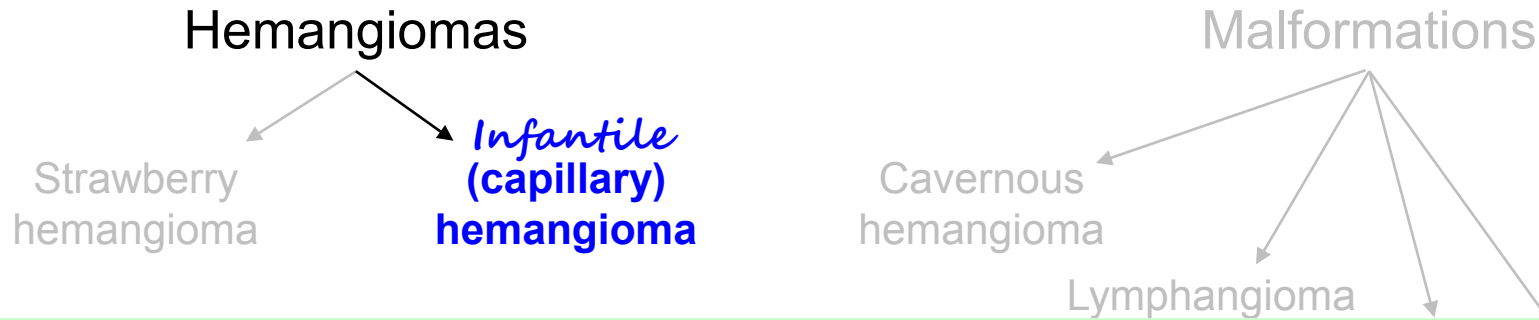
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Female gender is the major risk factor. What is another one?
Prematurity/low birth-weight

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Vascular Lesions of the Orbit

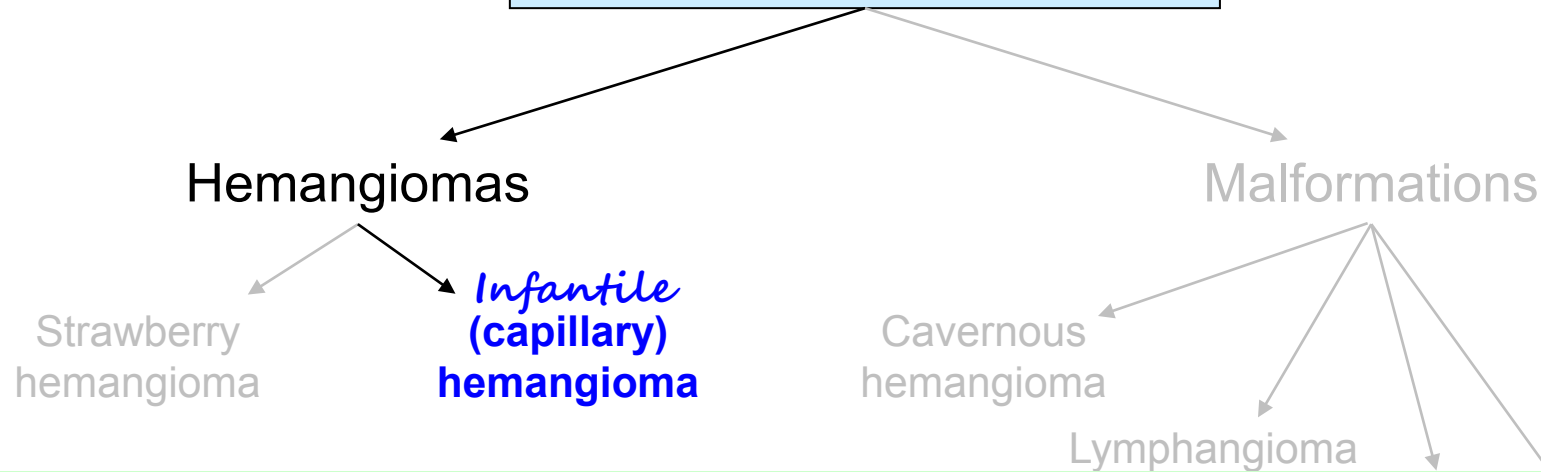


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At what age do orbital capillary hemangiomas present?

Vascular Lesions of the Orbit



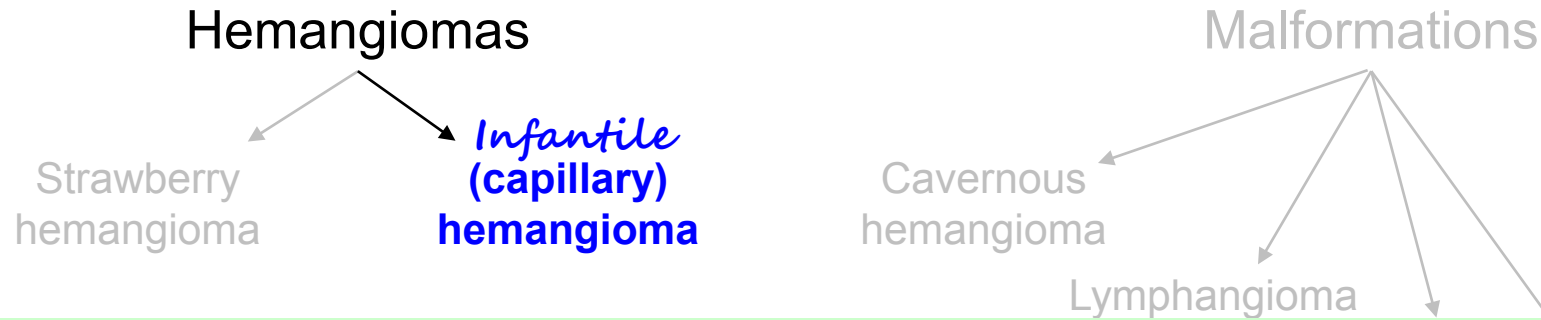
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At what age do orbital capillary hemangiomas present?
Most are not apparent at birth, instead declaring themselves over the first to of life

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Vascular Lesions of the Orbit



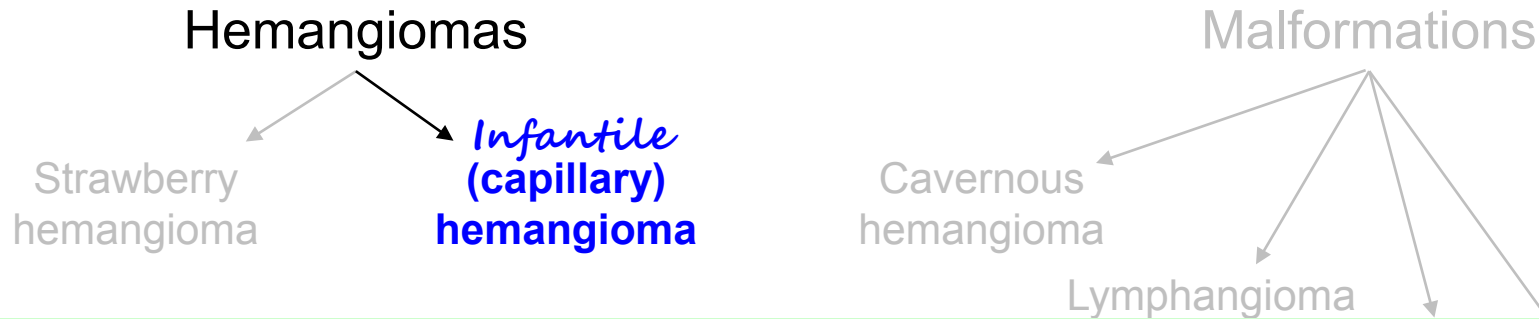
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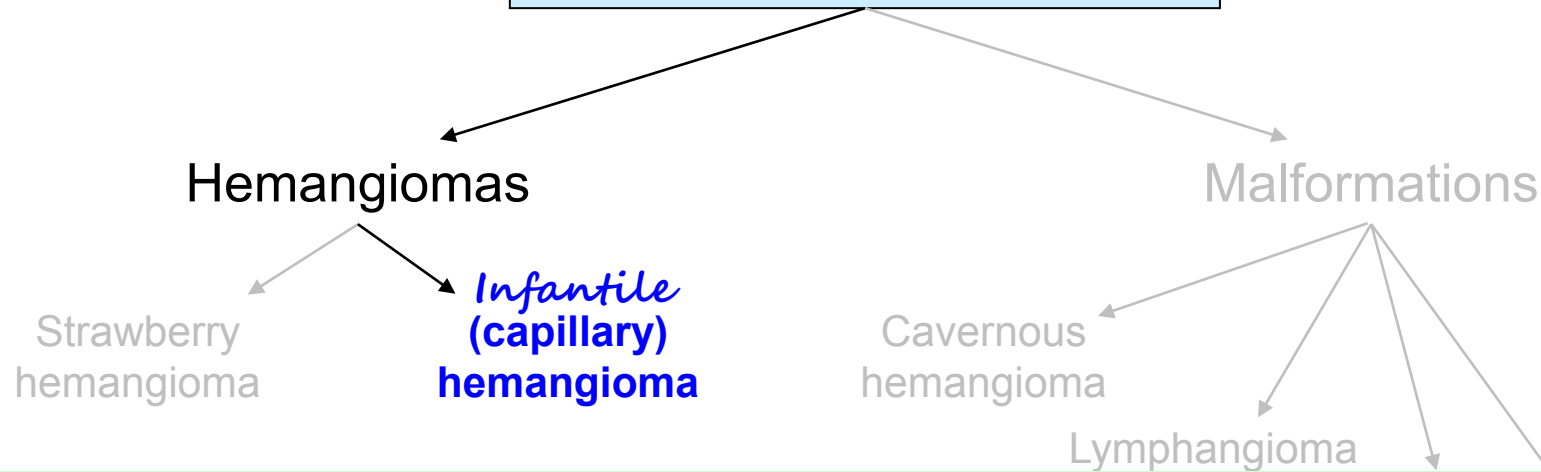
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How does capillary hemangioma present?

Vascular Lesions of the Orbit



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How does capillary hemangioma present?

As a or lesion of the periorbital skin

A

Vascular Lesions of the Orbit

Hemangiomas

Malformations

Strawberry hemangioma

Infantile (capillary) hemangioma

Cavernous hemangioma

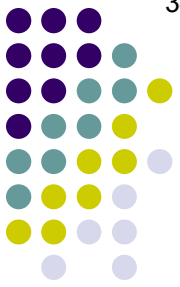
Lymphangioma

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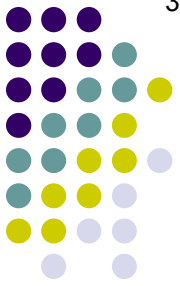
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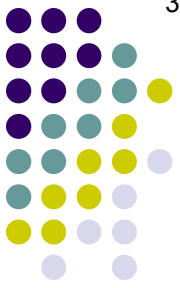
*How does capillary hemangioma present?
As a **bluish** or **strawberry-like** lesion of the periorbital skin*



Vascular Lesions of the Orbit



Hemangioma: Bluish



Vascular Lesions of the Orbit



Hemangioma: Bluish



Hemangioma: Strawberryish

Q

Vascular Lesions of the Orbit

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How does capillary hemangioma present?

As a **bluish** or **strawberry-like** lesion of the periorbital skin

What determines whether it appears **bluish** vs **strawberry-like**?

Q/A

Vascular Lesions of the Orbit

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What determines whether it appears **bluish** vs **strawberry-like**?

It appears **bluish** if it's...*[one depth]*

It appears **strawberry-like** if it's...*[the other depth]*

A

Vascular Lesions of the Orbit

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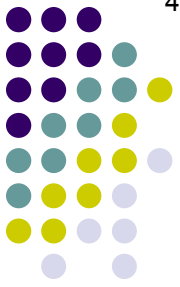
It appears **bluish** if it's...**deep**

It appears **strawberry-like** if it's...**superficial**

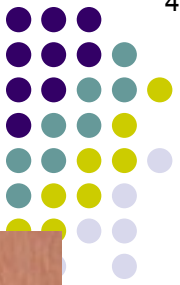
Vascular Lesions of the Orbit



Capillary hemangioma



Vascular Lesions of the Orbit



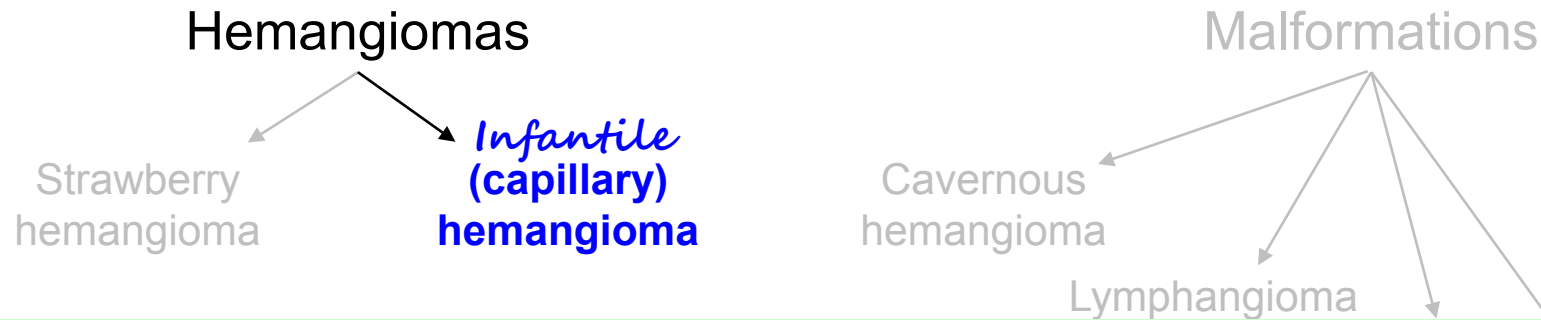
Capillary hemangioma



Stretching the skin of the upper lid clearly shows two distinct portions of the lesion -- an elevated, red area where there is superficial hemangioma and a smooth, bluish-purple area representing a deeper component

Q

Vascular Lesions of the Orbit



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

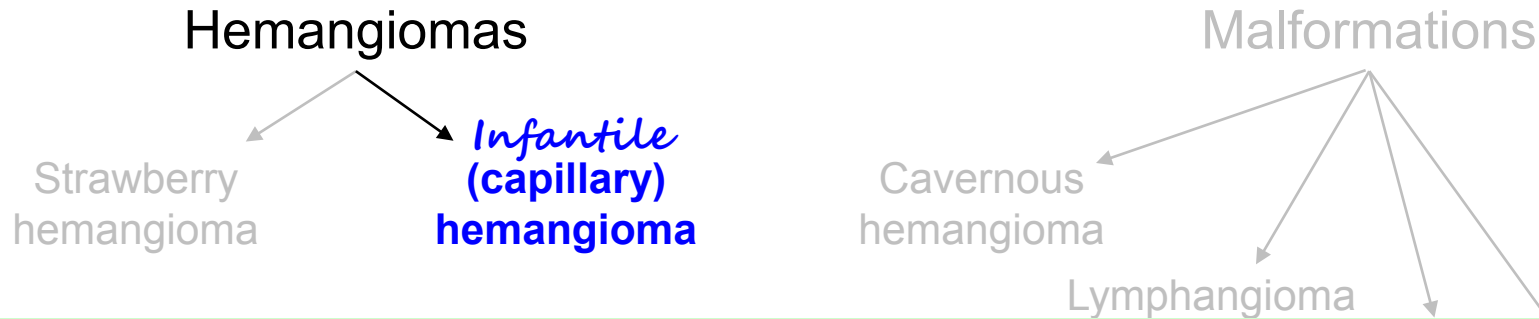
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What is the 'life cycle' of capillary hemangiomas?

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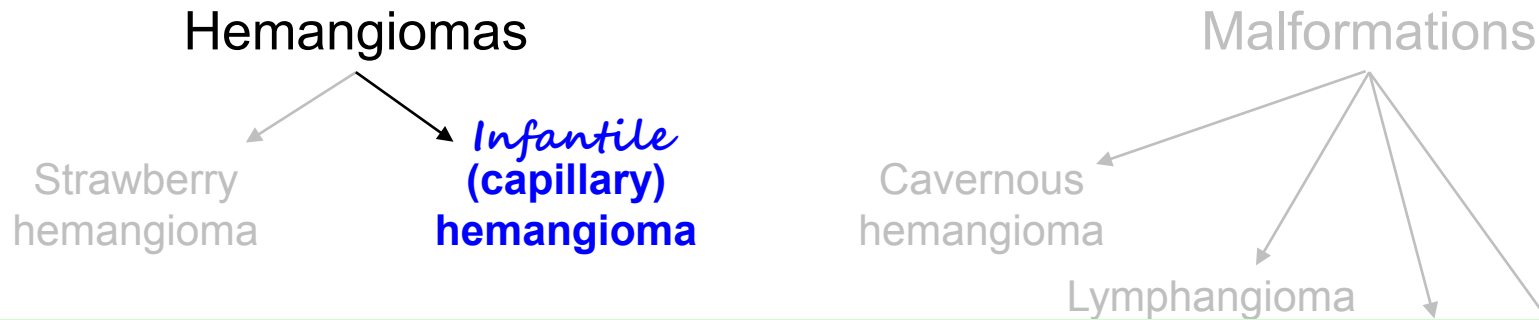
1) They grow rapidly initially, maxing out around age

2)

3)

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Vascular Lesions of the Orbit



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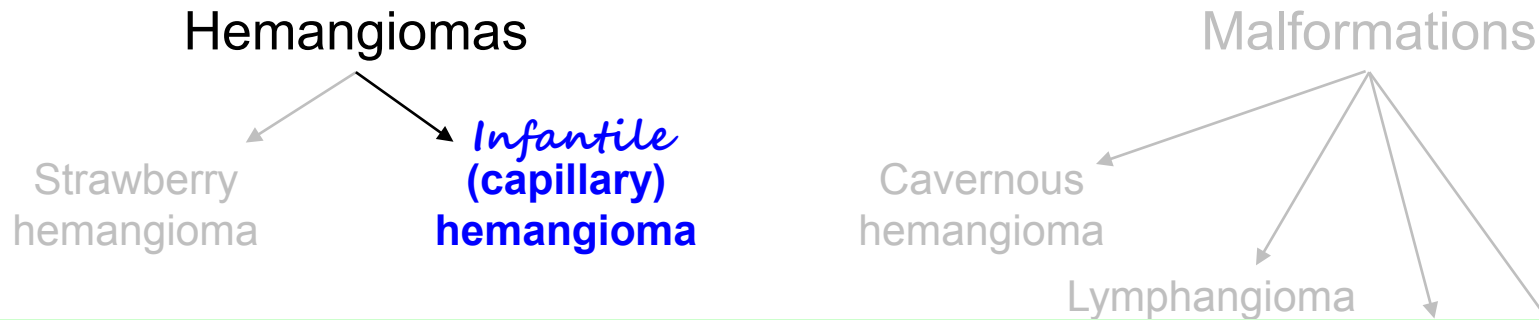
At what age do orbital capillary hemangiomas present?
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What is the 'life cycle' of capillary hemangiomas?

- 1) They grow rapidly initially, maxing out around age 1 year
- 2)
- 3)

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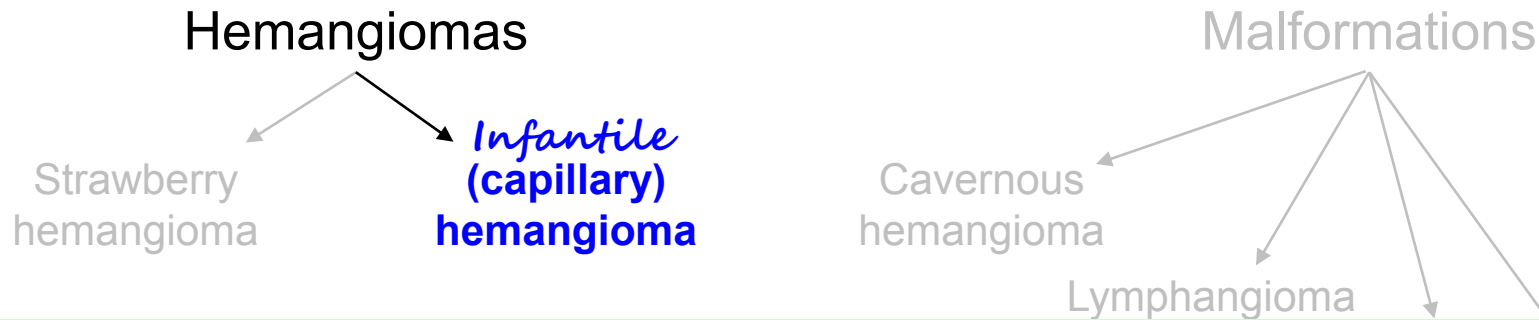
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What is the 'life cycle' of capillary hemangiomas?

- 1) They grow rapidly initially, maxing out around age 1 year
- 2) They then begin to slowly involute
- 3)

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- 1) They grow rapidly initially, maxing out around age 1 year
- 2) They then begin to slowly involute
- 3) Most fully resolve by age range

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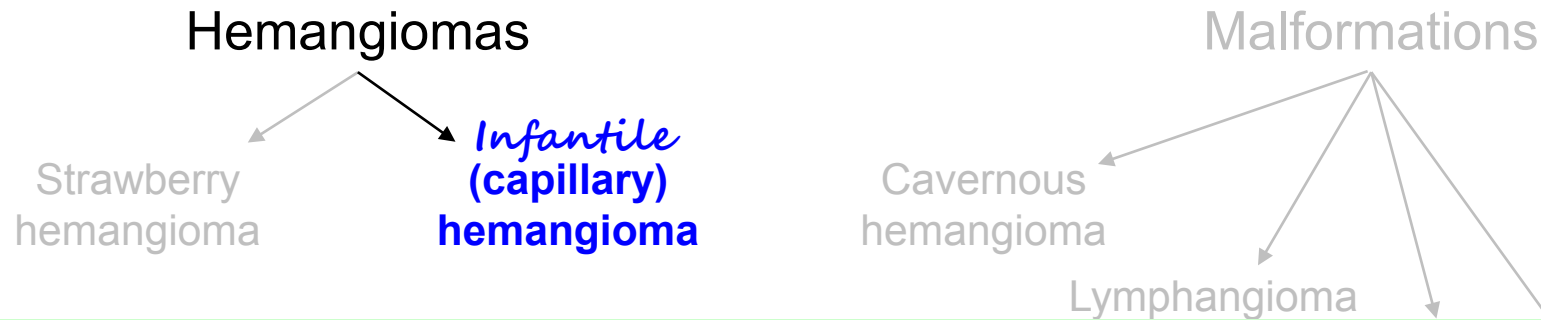
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What is the 'life cycle' of capillary hemangiomas?

- 1) They grow rapidly initially, maxing out around age 1 year
- 2) They then begin to slowly involute
- 3) Most will fully and spontaneously resolve by age 4 to 8

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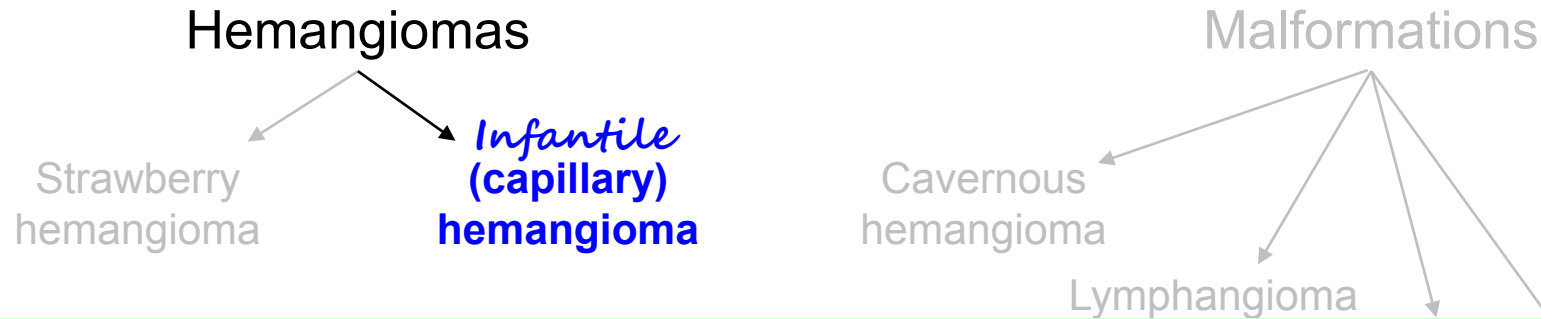
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At what age do orbital capillary hemangiomas present?
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What is the origin of these lesions?

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Vascular Lesions of the Orbit



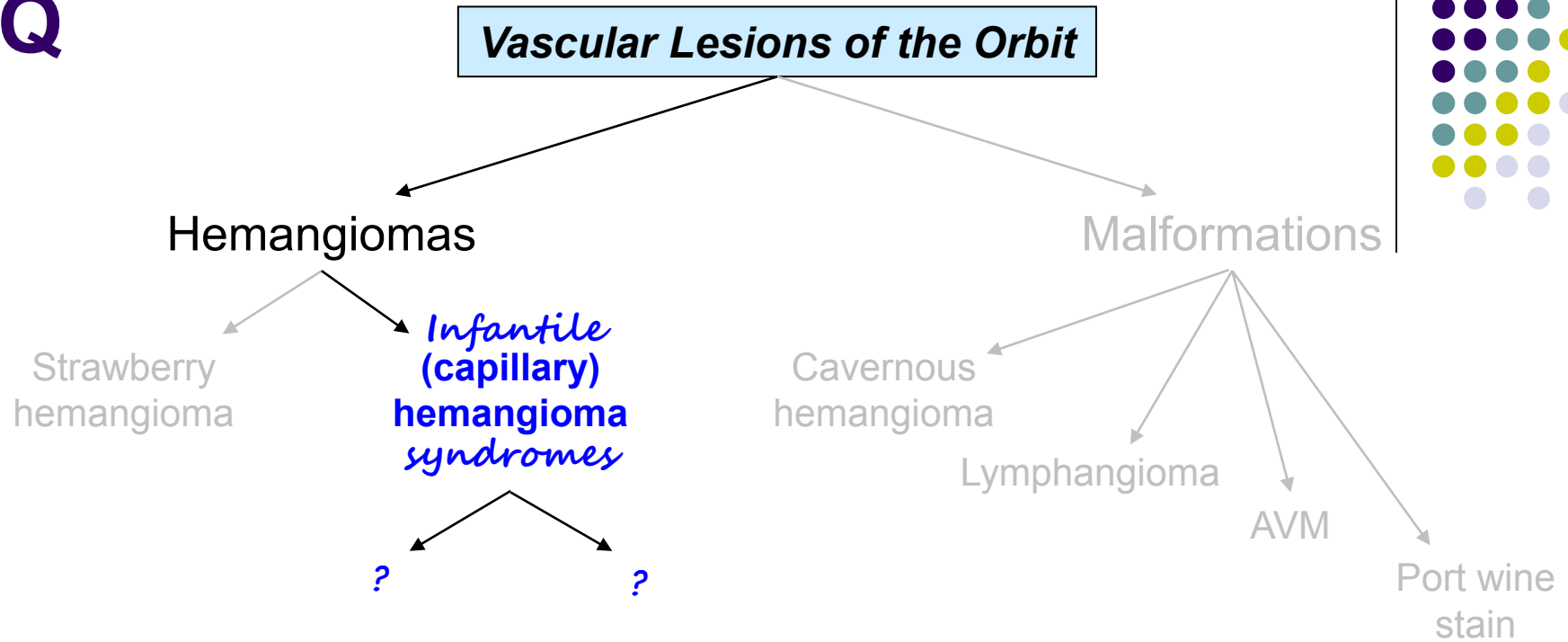
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At what age do orbital capillary hemangiomas present?
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What is the origin of these lesions?
It's not known for certain, but some experts believe they originate as nests of placental cells that 'metastasized' to the fetus

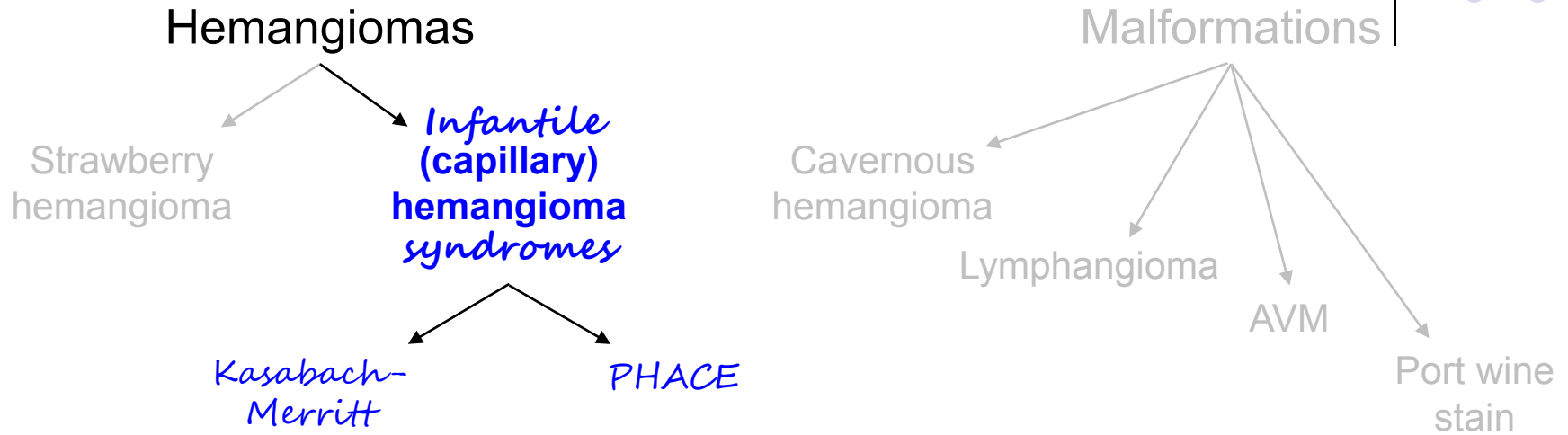
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With respect to capillary hemangiomas, the word syndrome should evoke two conditions. What are they?

A

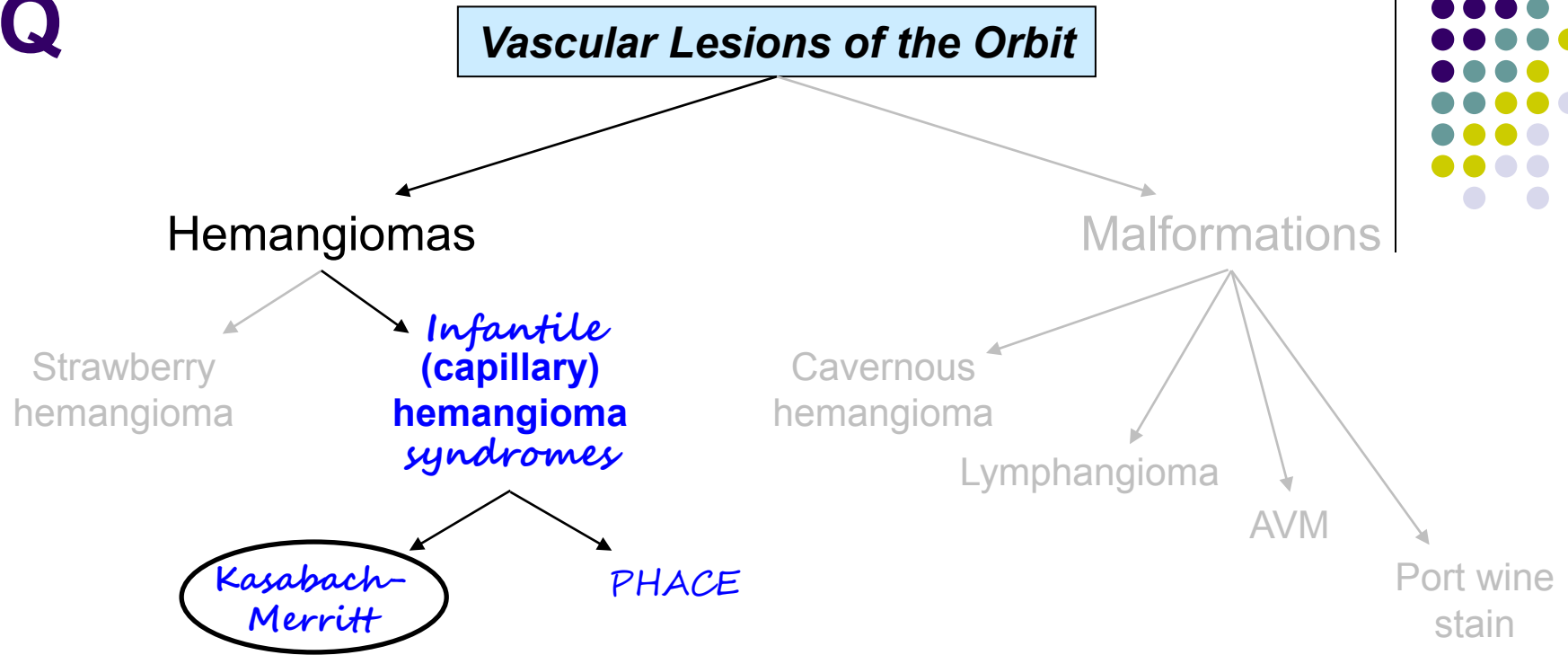
Vascular Lesions of the Orbit



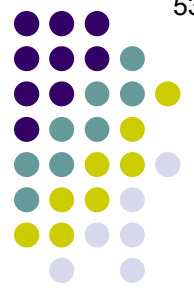
With respect to capillary hemangiomas, the word syndrome should evoke two conditions. What are they?
Kasabach-Merritt, and PHACE



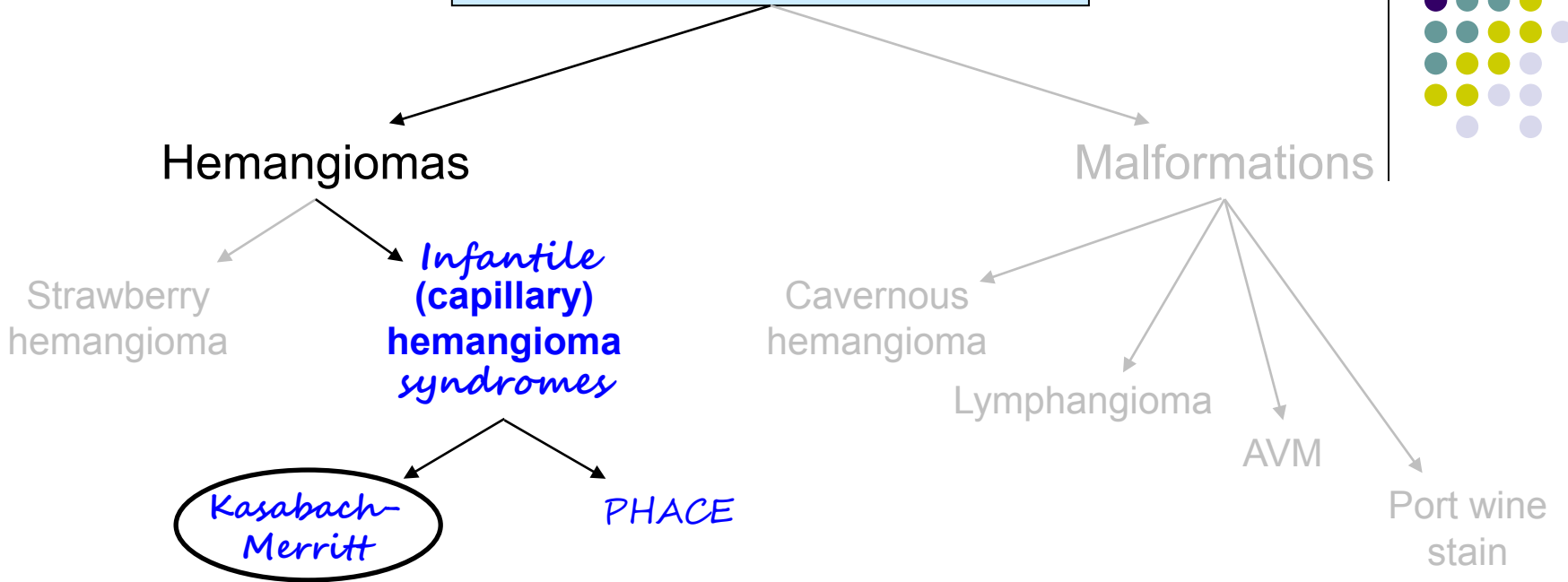
Q



What is the Kasabach-Merritt syndrome?



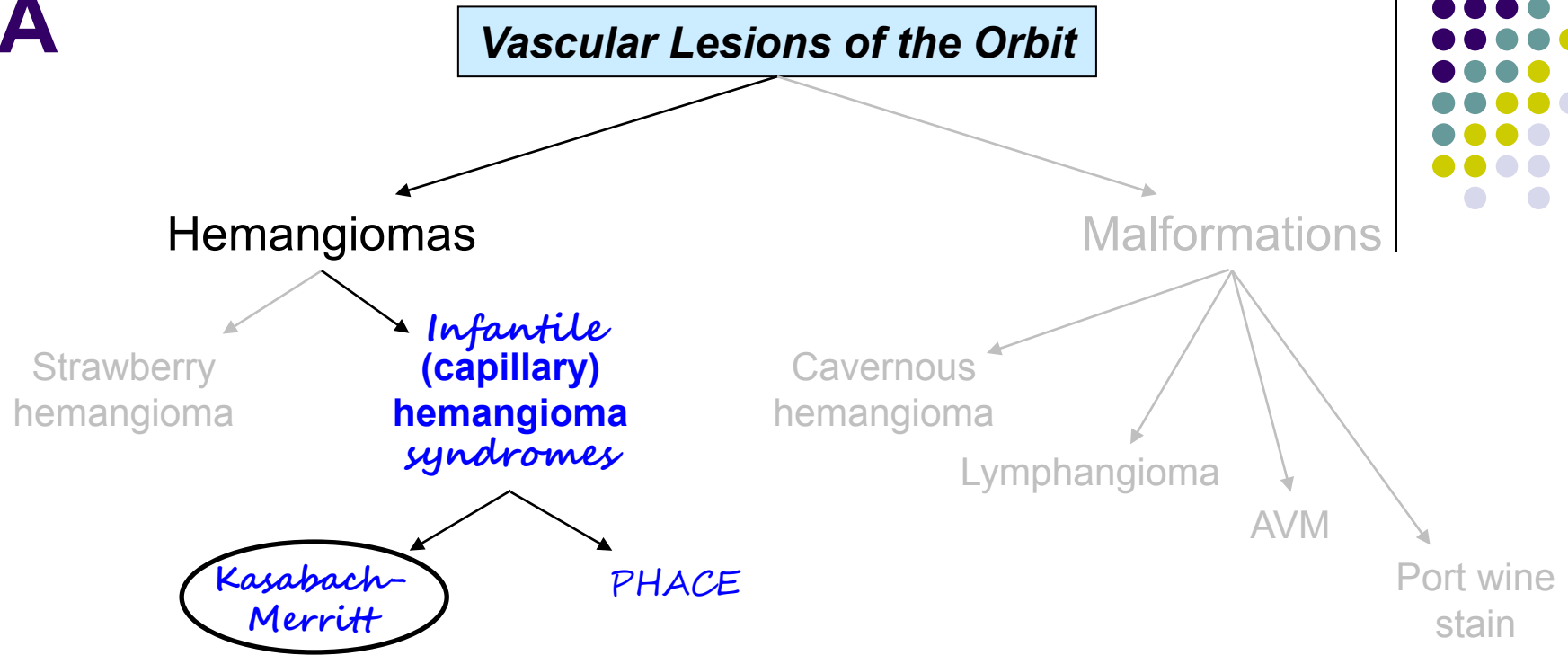
Vascular Lesions of the Orbit



What is the Kasabach-Merritt syndrome?

A hematologic cell type coagulopathy

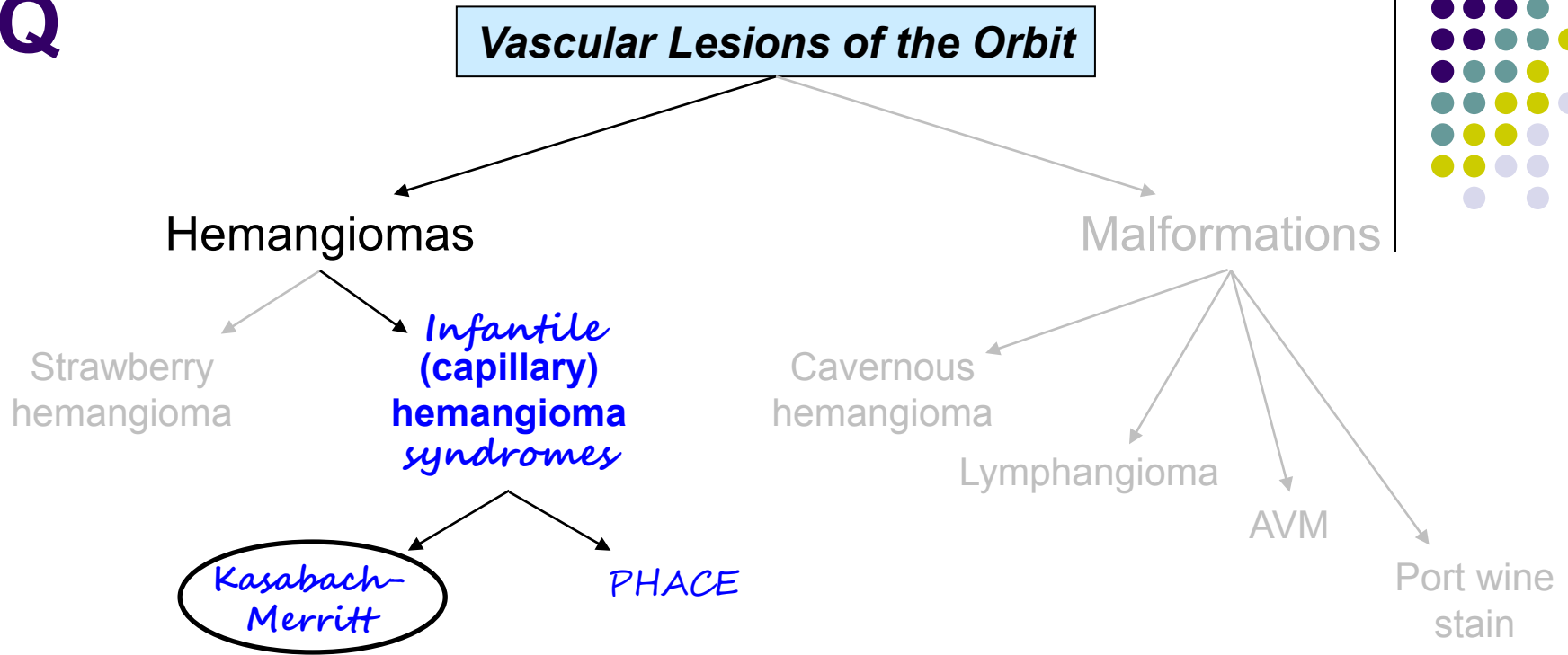
A



What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy

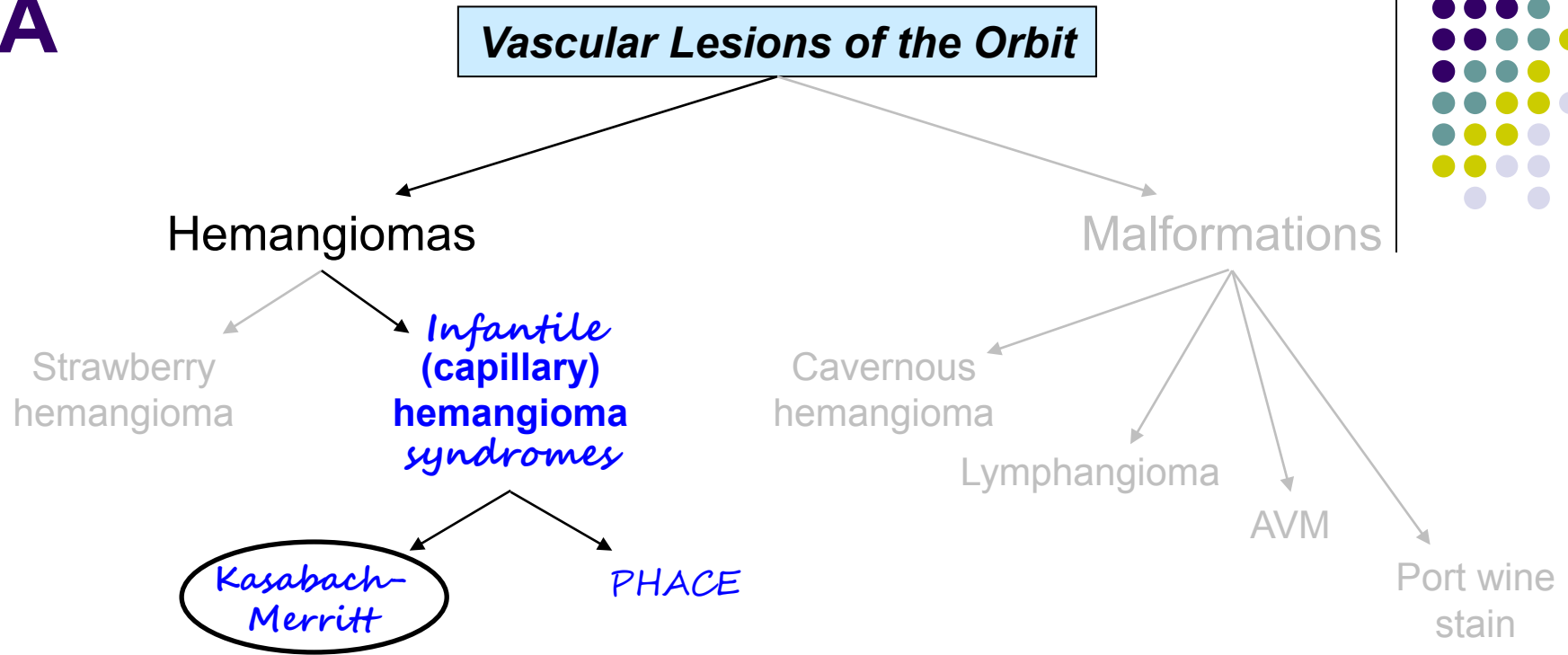
Q



What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to of

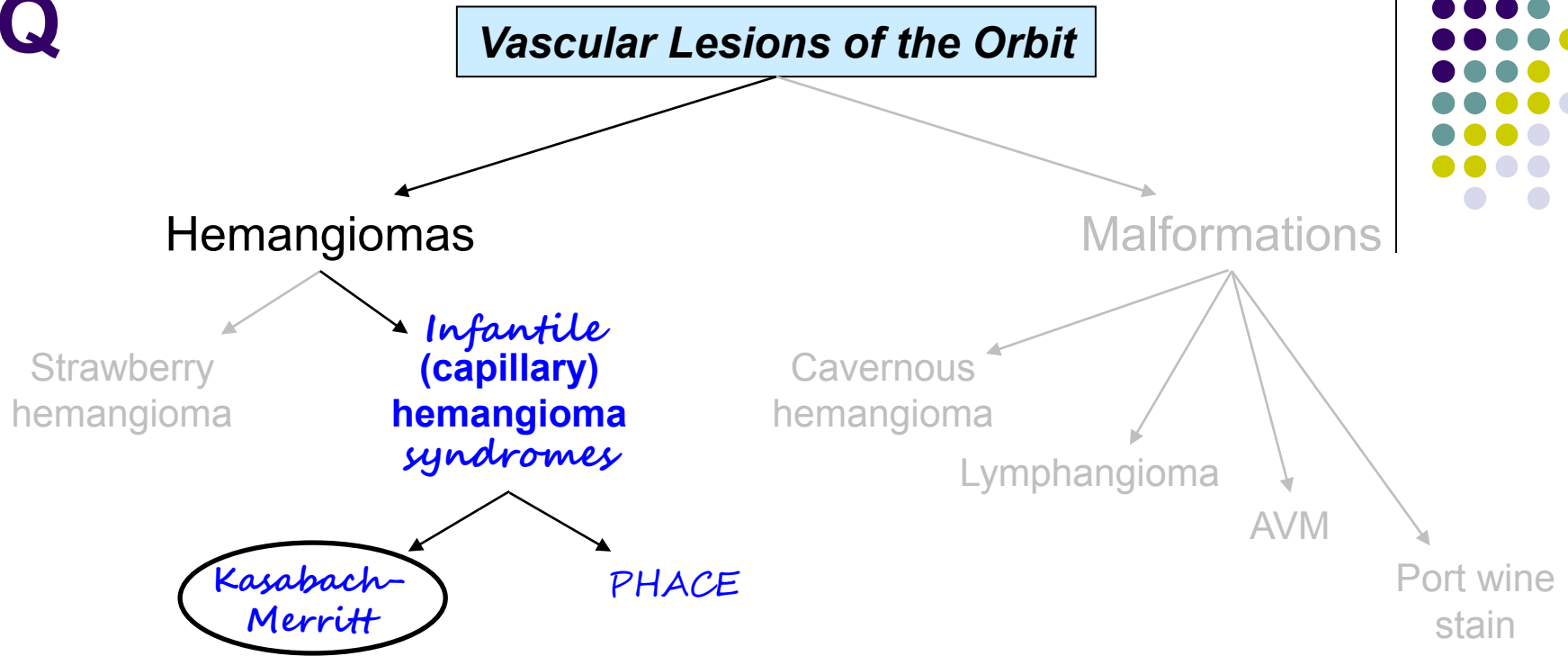
A



What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to sequestration of platelets

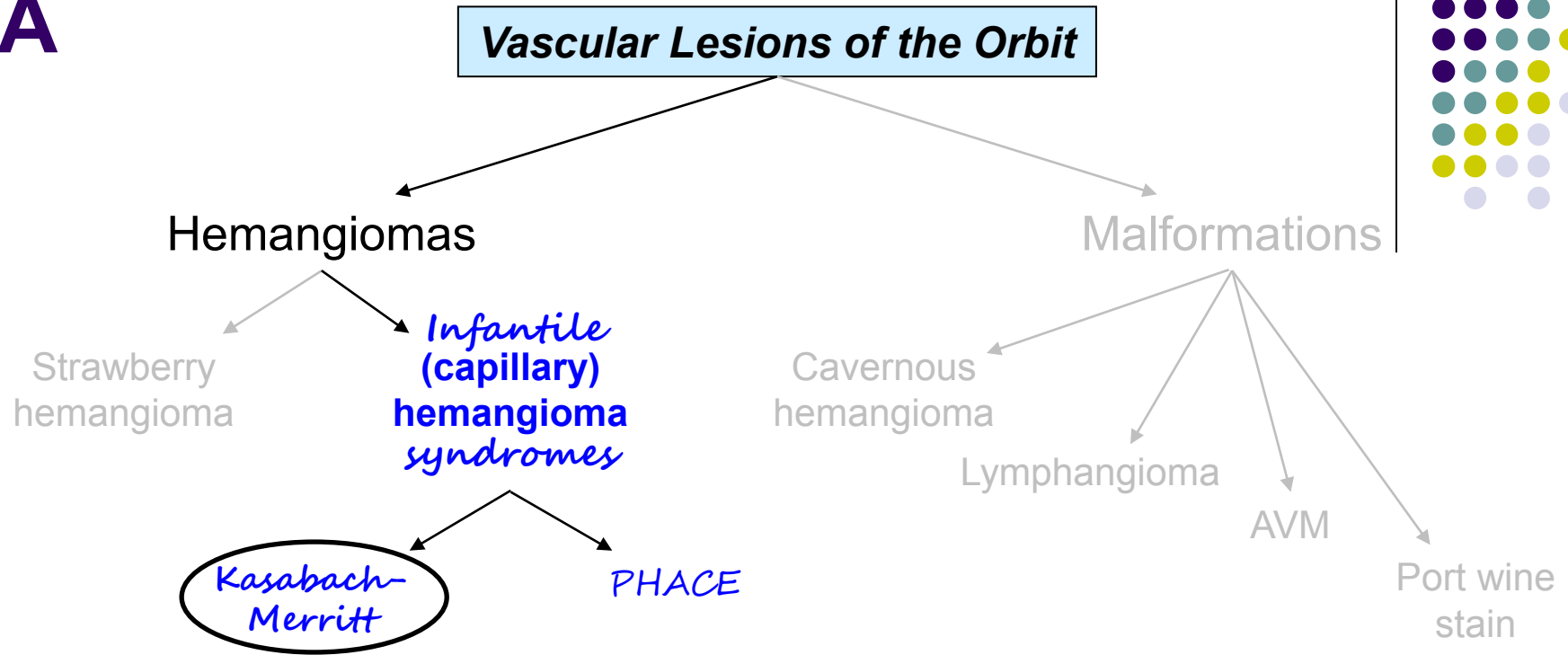
Q



What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to sequestration of platelets in an associated large capillary hemangioma located in the body system

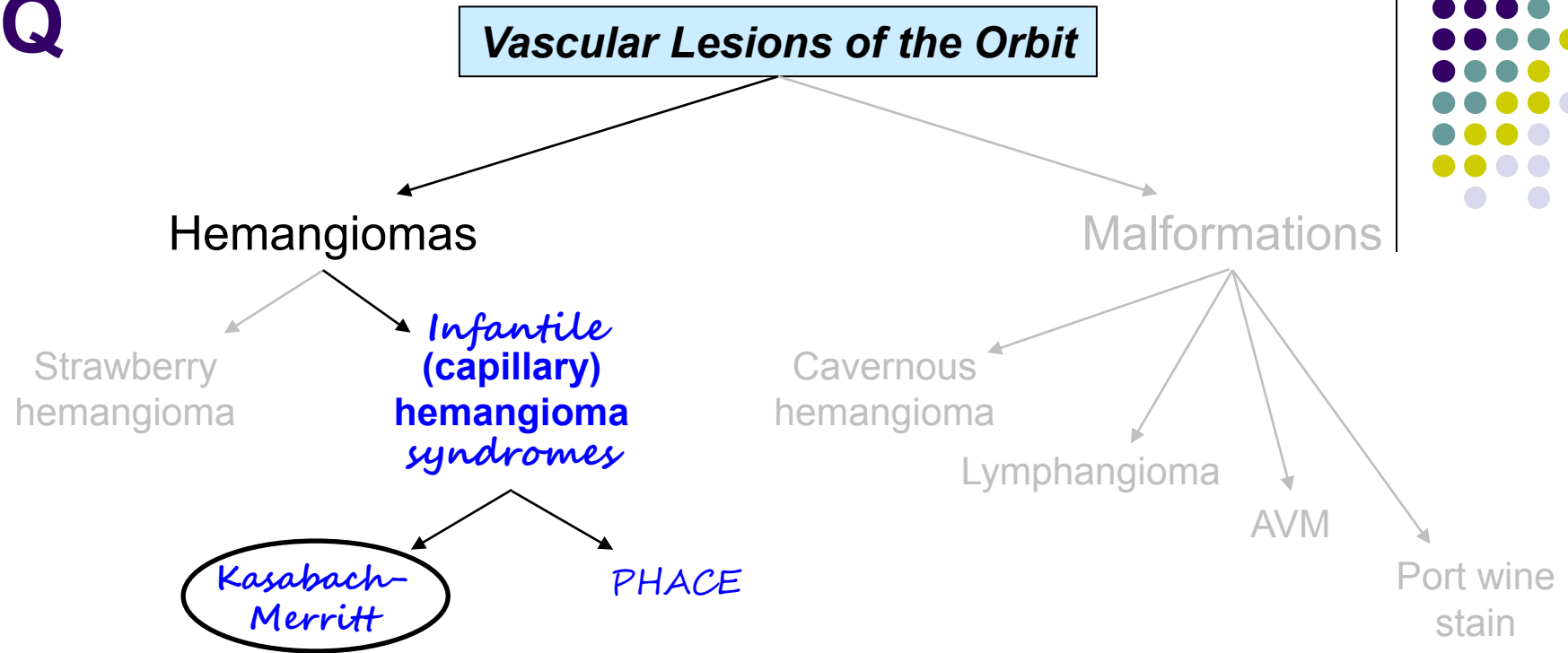
A



What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to sequestration of platelets in an associated large capillary hemangioma located in the GI tract

Q

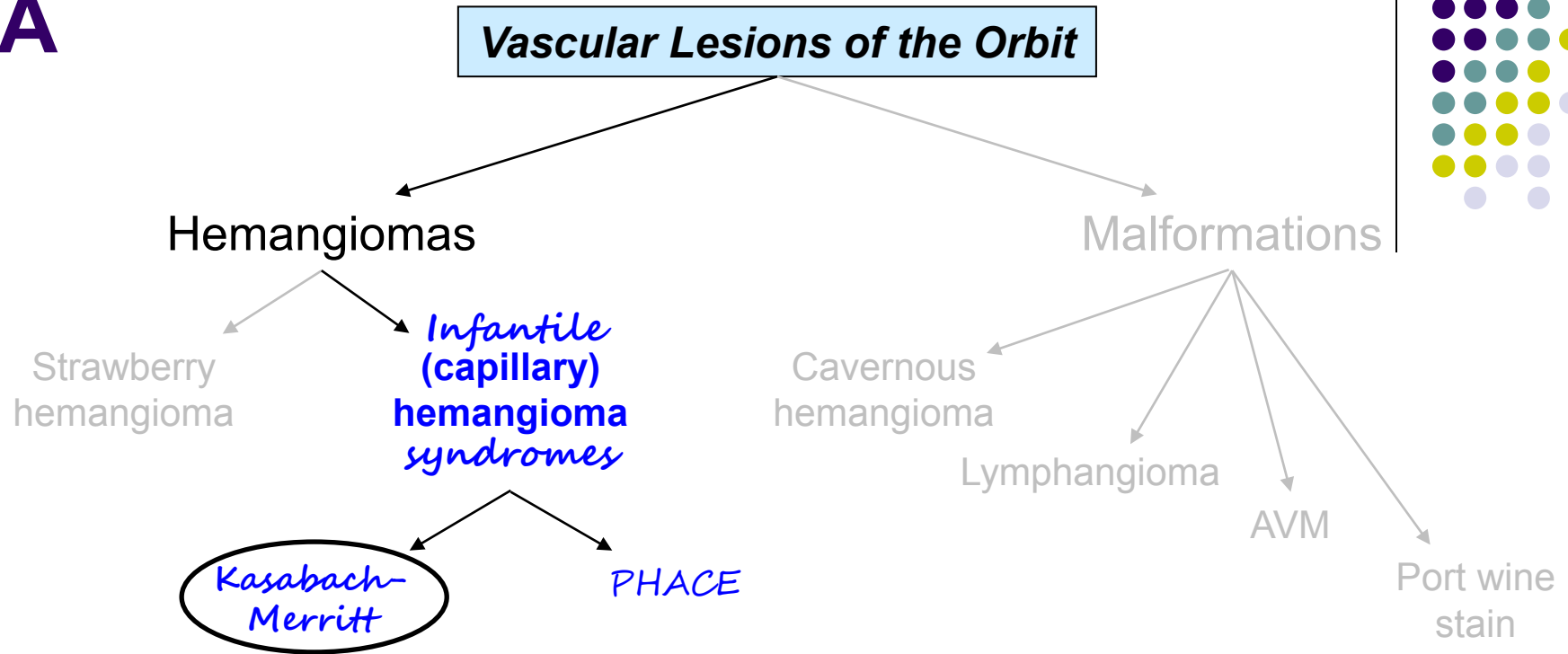


What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to sequestration of platelets in an associated large capillary hemangioma located in the GI tract

Is it serious?

A



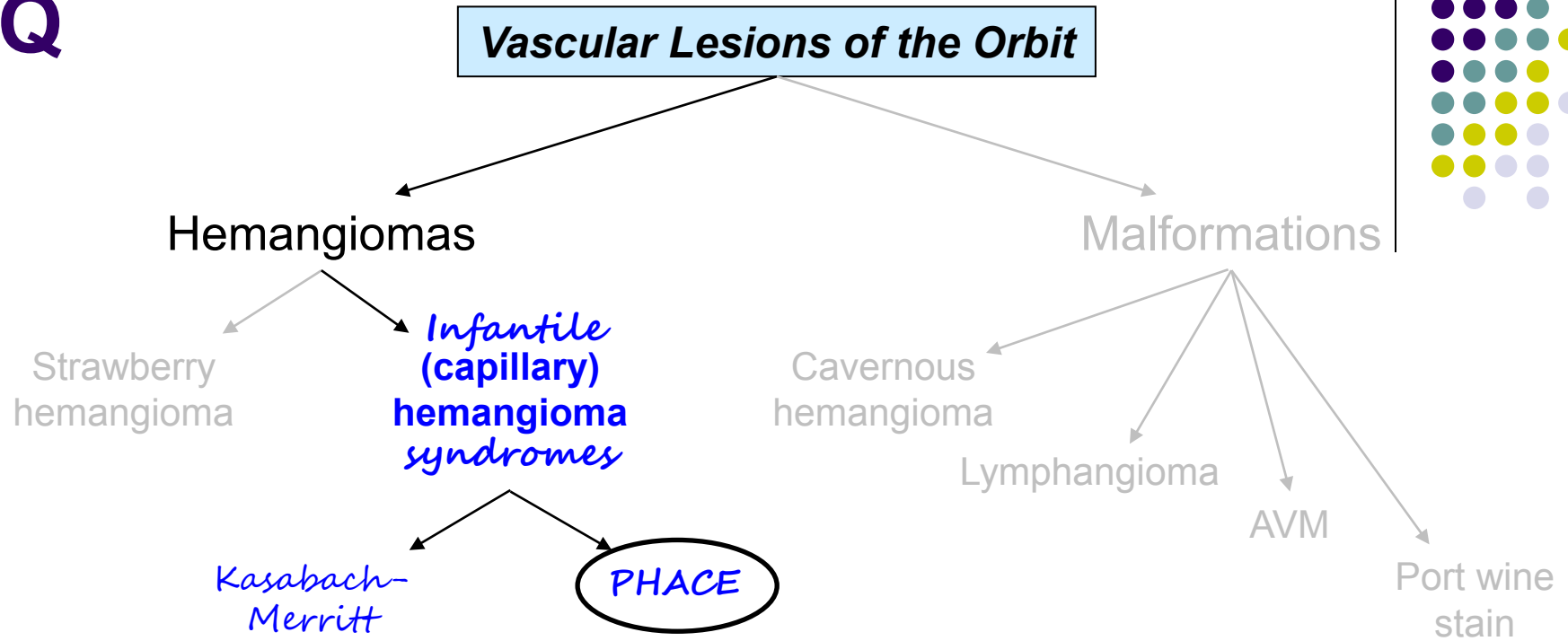
What is the Kasabach-Merritt syndrome?

A thrombocytopenic coagulopathy secondary to sequestration of platelets in an associated large capillary hemangioma located in the GI tract

Is it serious?

Quite—it carries a high mortality rate

Q



I have a hunch PHACE is an acronym, with the 'H' standing for 'hemangiomas.'
Assuming I'm right, what does the rest stand for?

P

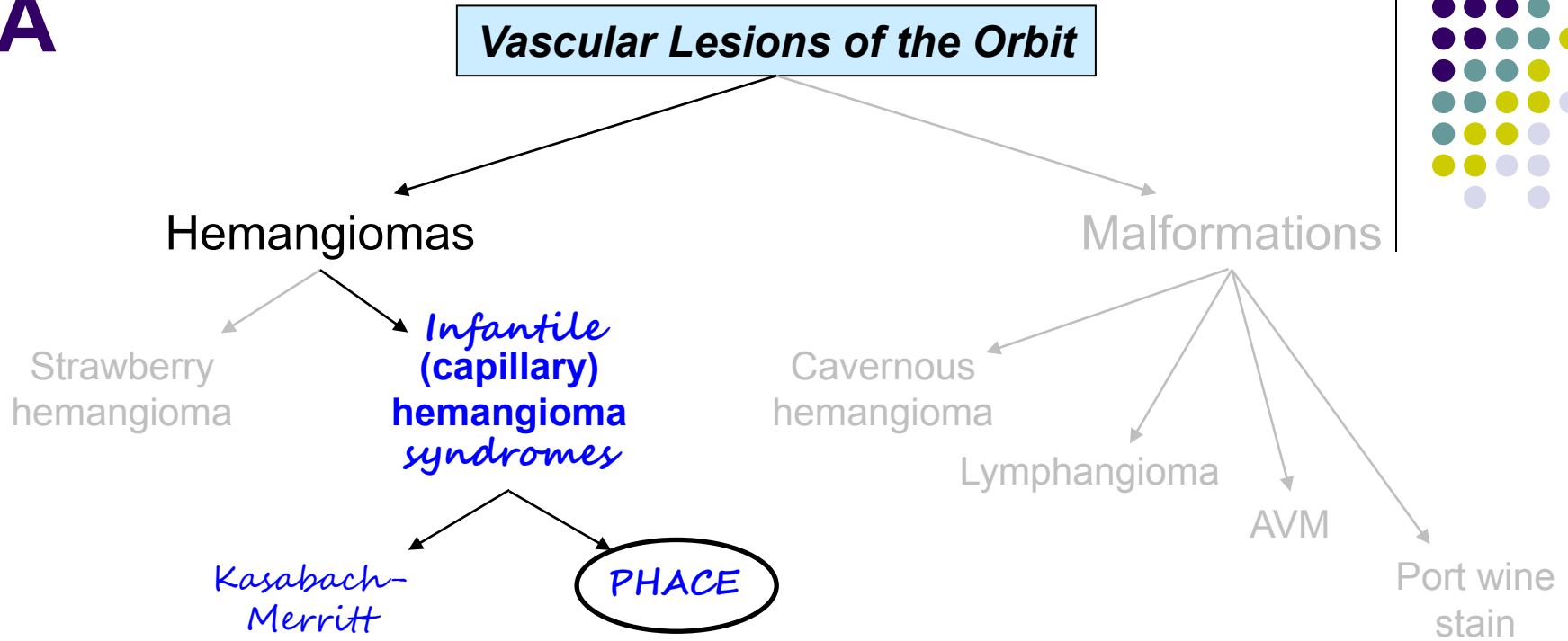
Hemangiomas

A

C

E

A



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Posterior fossa malformations

Hemangiomas

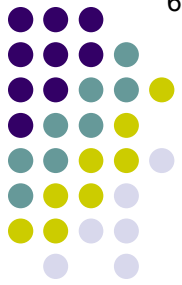
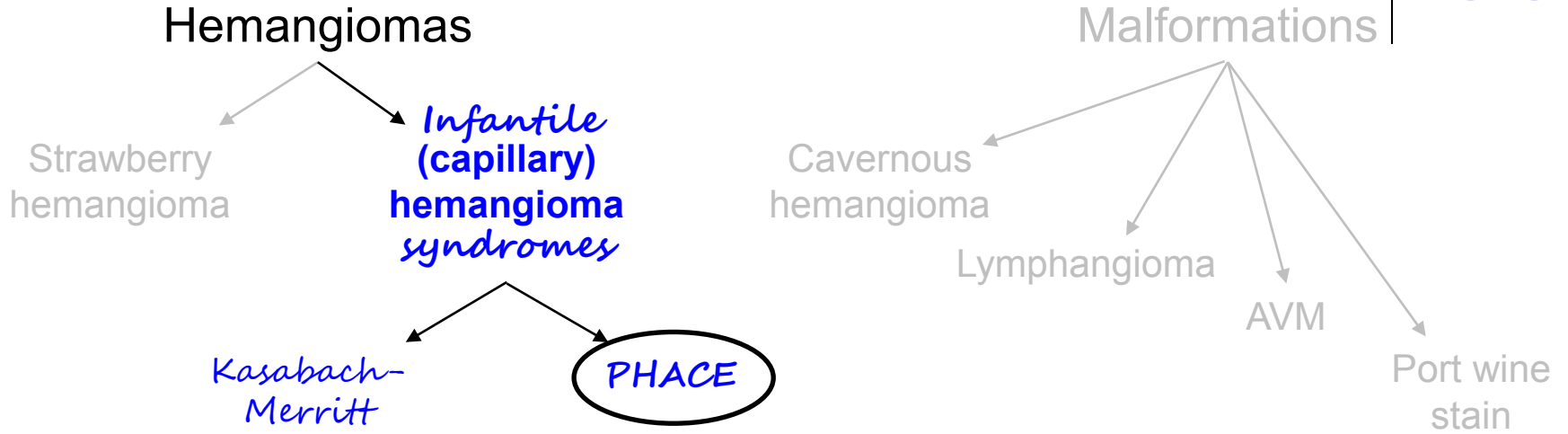
Arterial lesions

Cardiac anomalies

Eye anomalies

Q

Vascular Lesions of the Orbit



I have a hunch PHACE is a hemangioma for 'hemangiomas.'

Assuming I'm right

Posterior fossa malformations

Hemangiomas

Arterial lesions

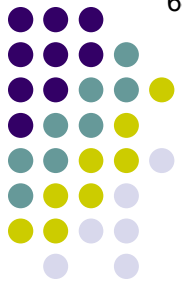
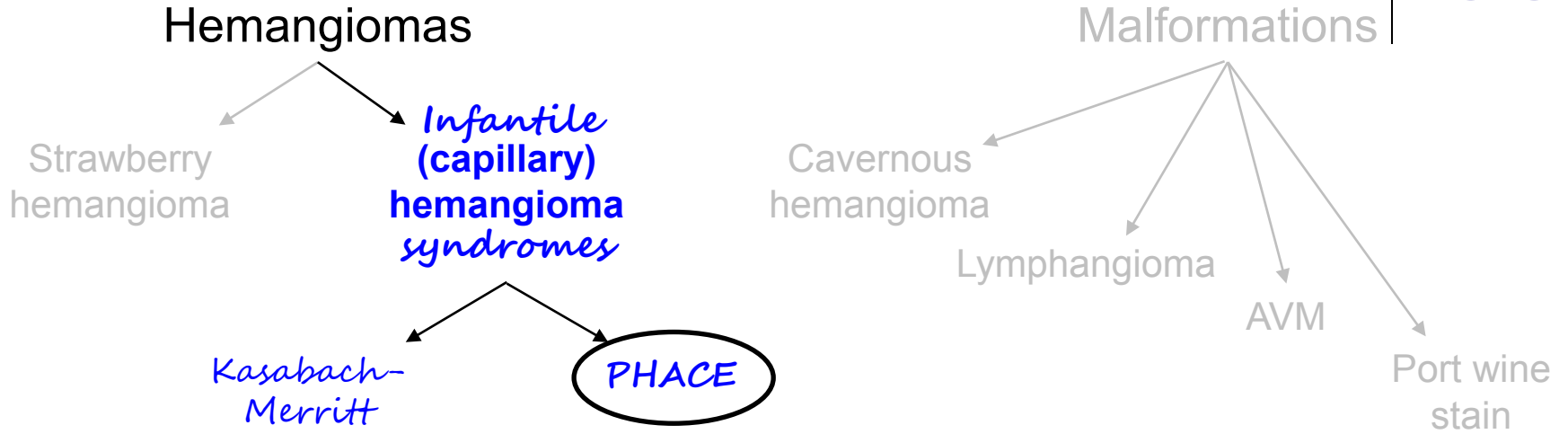
Cardiac anomalies

Eye anomalies

Where is the hemangioma located?

A

Vascular Lesions of the Orbit



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Posterior fossa malformations

Hemangiomas

Arterial lesions

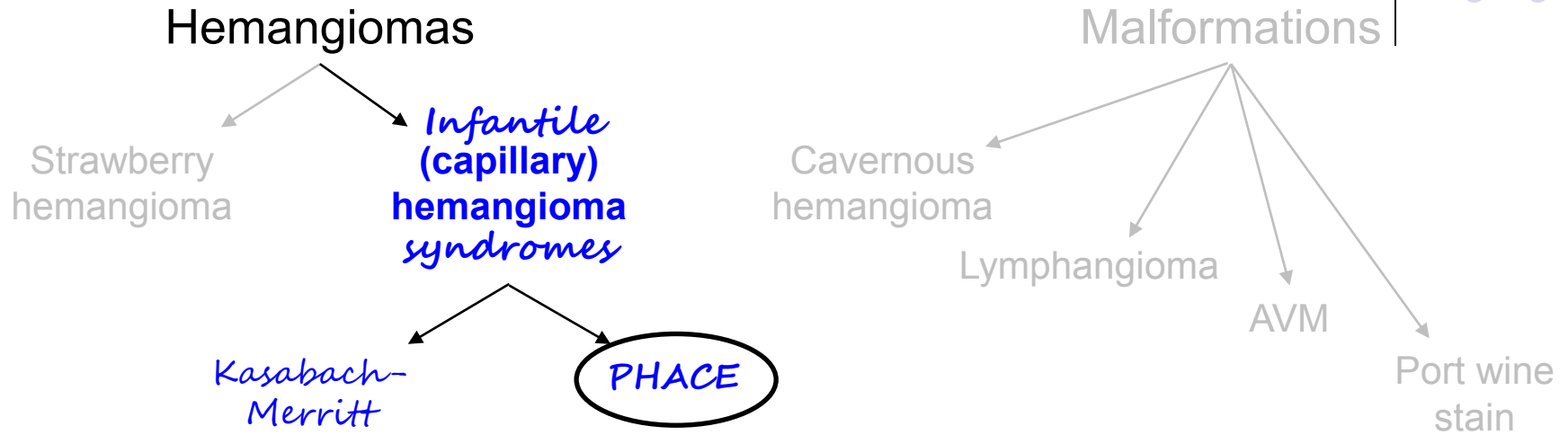
Cardiac anomalies

Eye anomalies

Where is the hemangioma located?
The face, +/- scalp involvement

Q

Vascular Lesions of the Orbit



I have a hunch PH
Assuming I'm right
Posterior fossa ma
Hemangiomas
Arterial lesions
Cardiac anomalies
Eye anomalies

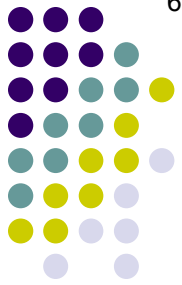
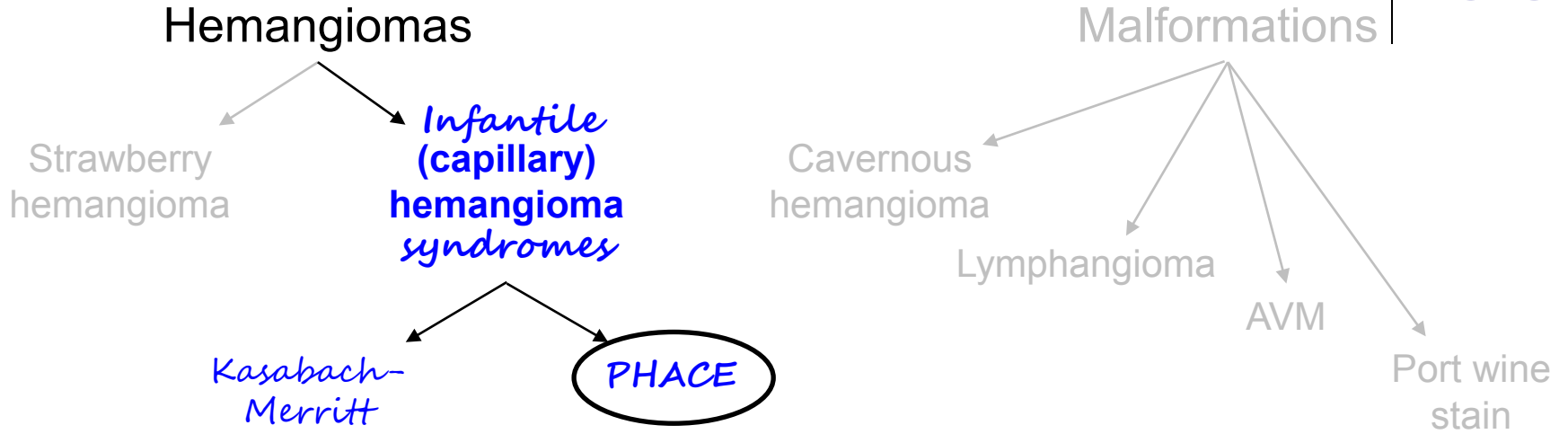
Where is the hemangioma located?
The face, +/- scalp involvement

Is it large, or small?

g for 'hemangiomas.'

A

Vascular Lesions of the Orbit



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Assuming I'm right

Posterior fossa malformations

Hemangiomas

Arterial lesions

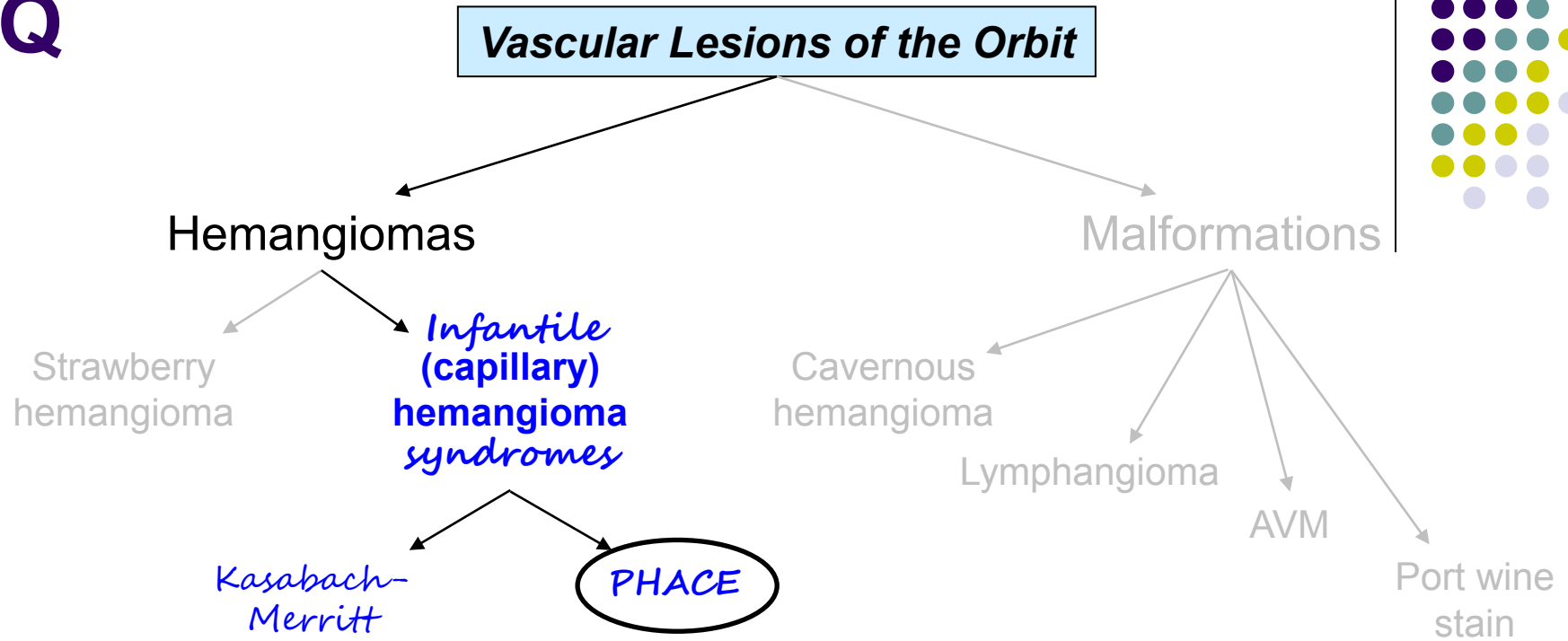
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Where is the hemangioma located?
The face, +/- scalp involvement

Is it large, or small?
Large

Q



I have a hunch PH
Assuming I'm right
Posterior fossa ma
Hemangiomas
Arterial lesions
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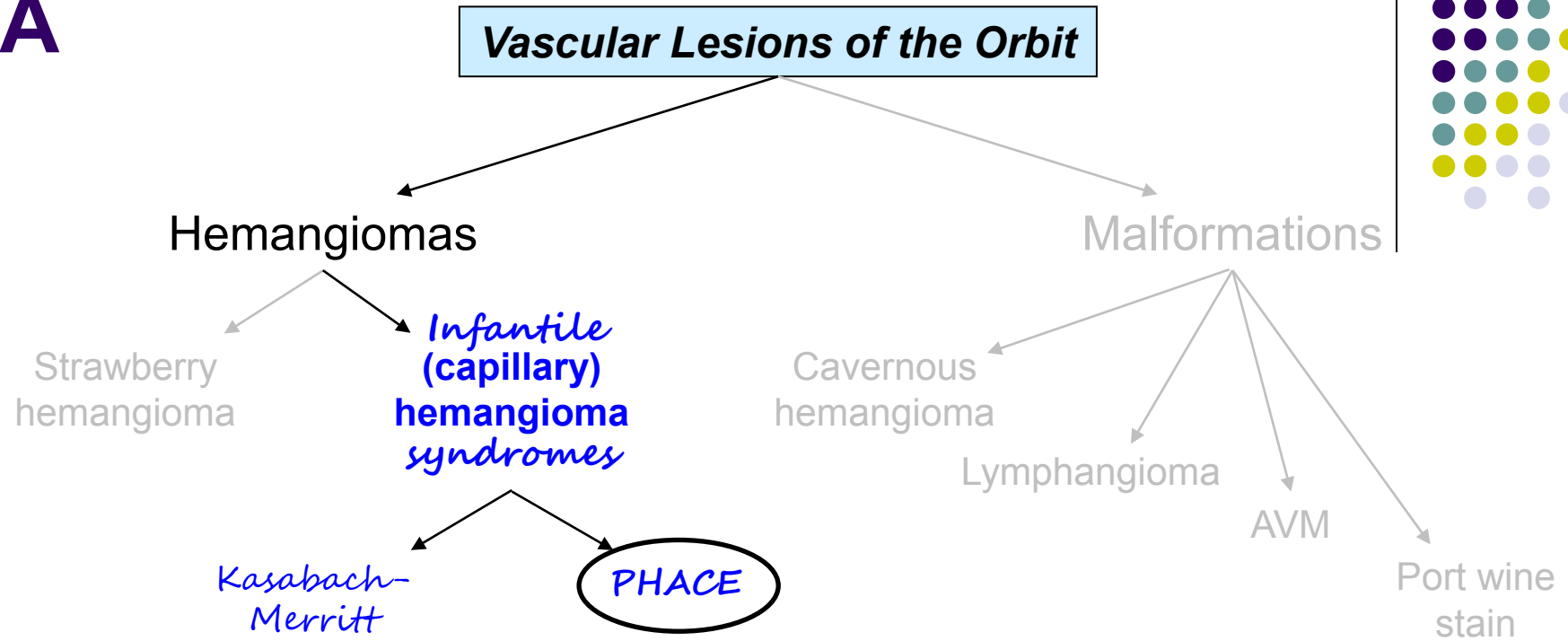
Where is the hemangioma located?
The face, +/- scalp involvement

Is it large, or small?
Large

What is the classic term for its shape?

g for 'hemangiomas.'

A



I have a hunch PH
Assuming I'm right
Posterior fossa ma
Hemangiomas
Arterial lesions
Cardiac anomalies
Eye anomalies

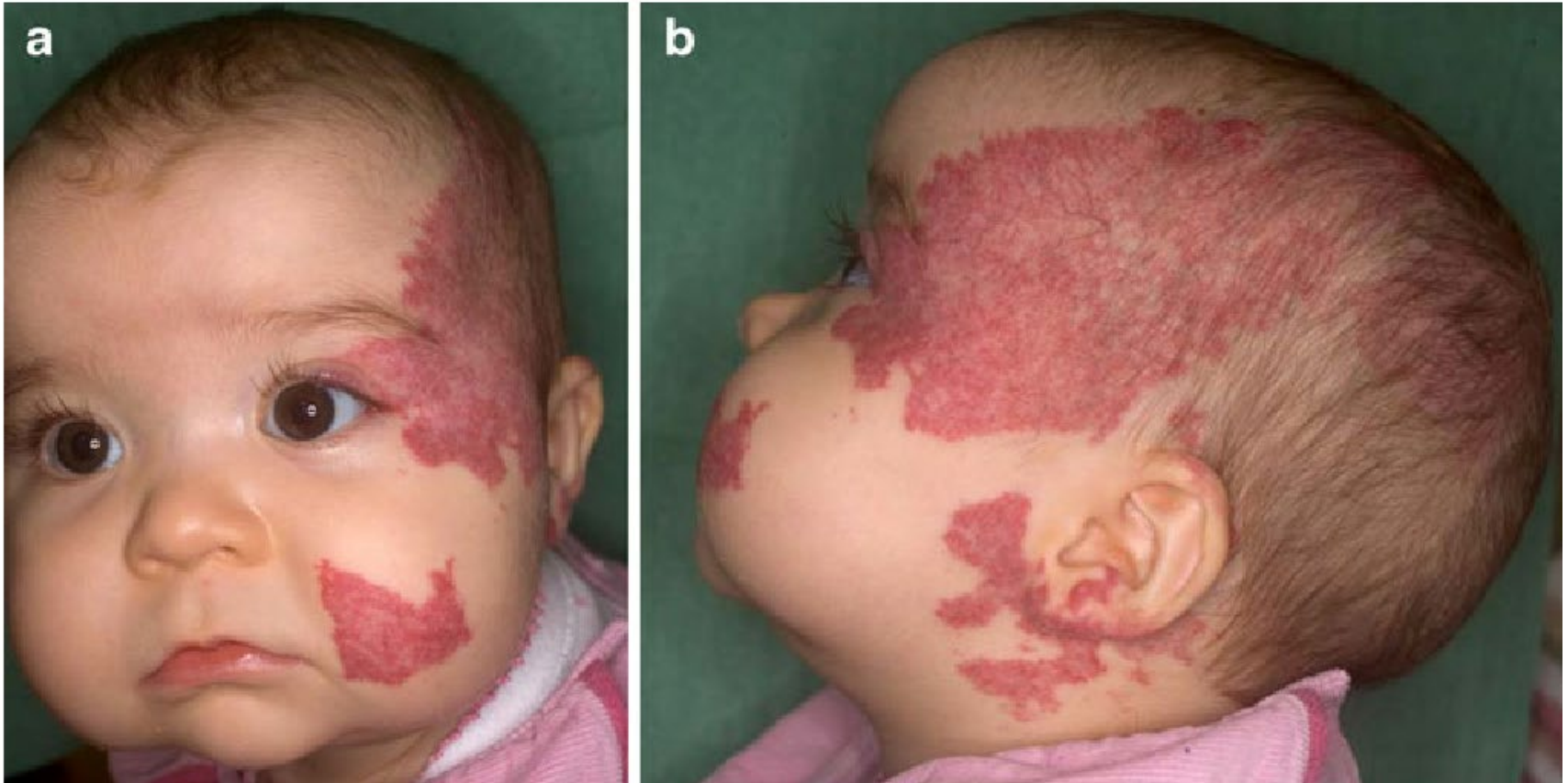
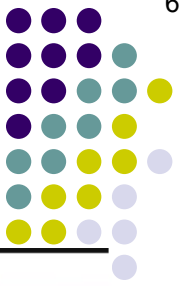
Where is the hemangioma located?
The face, +/- scalp involvement

Is it large, or small?
Large

What is the classic term for its shape?
'Plaquelike'

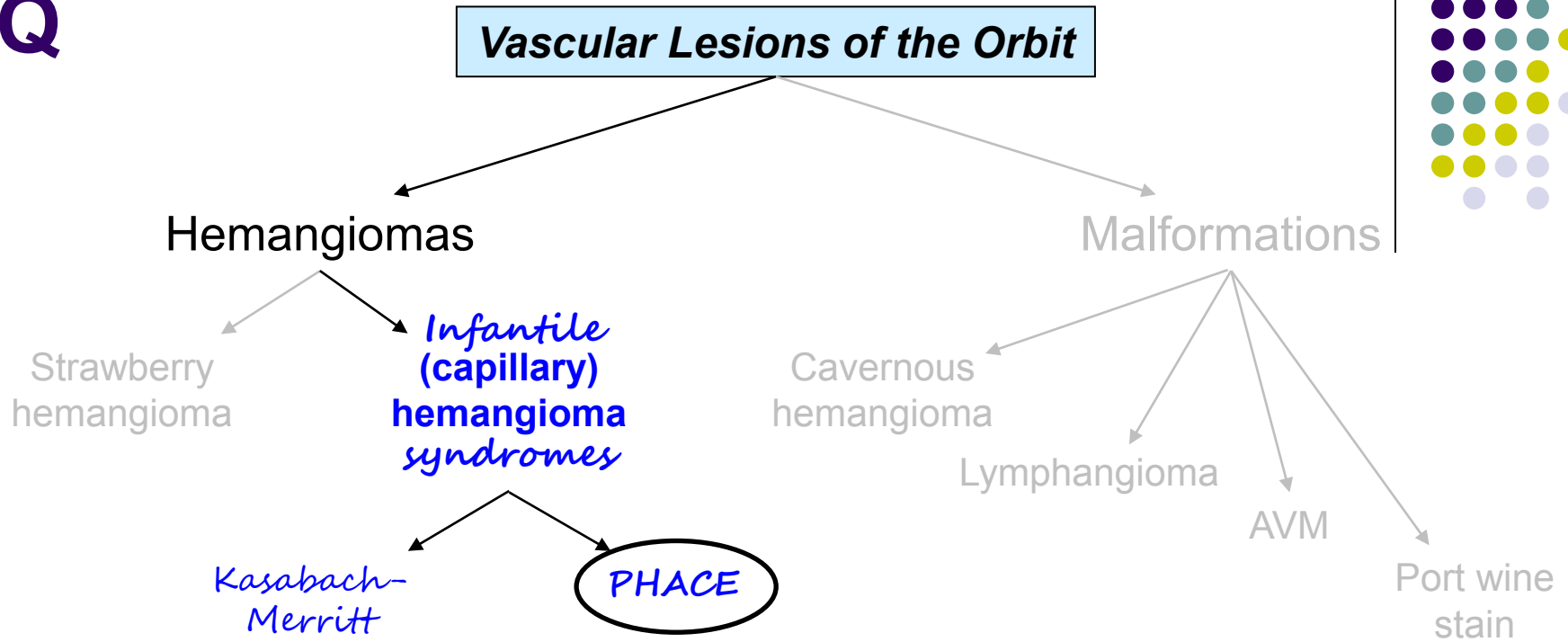
g for 'hemangiomas.'

Vascular Lesions of the Orbit



PHACE syndrome: Plaque-like hemangioma

Q



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Assuming I'm right, what does the rest stand for?

Posterior fossa malformations

Hemangiomas

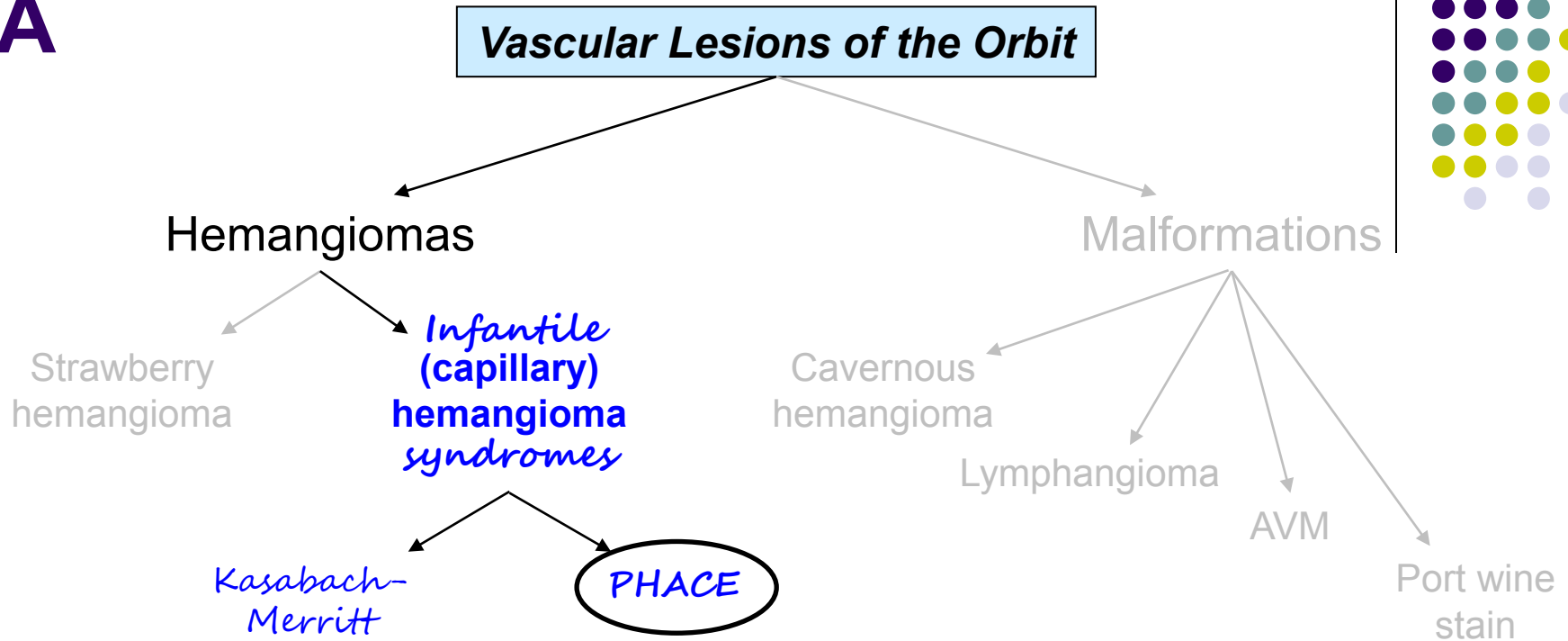
Arterial lesions

Cardiac anomalies

Eye anomalies

What sorts of eye anomalies' may be involved?

A



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Posterior fossa malformations

Hemangiomas

Arterial lesions

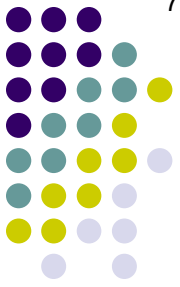
Cardiac anomalies

Eye anomalies

What sorts of eye anomalies' may be involved?

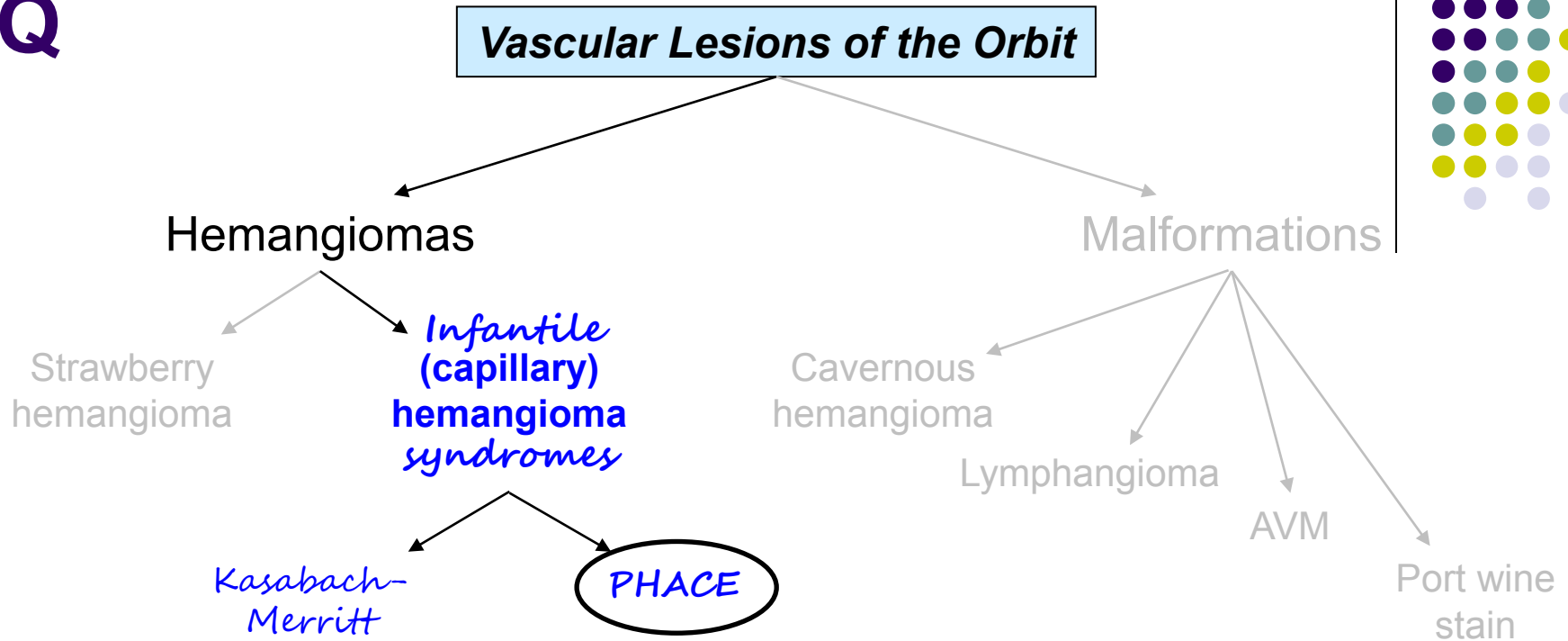
The usual suspects—cataracts; strabismus—along with sequelae of arrested development: colobomas; optic nerve hypoplasia; morning-glory disc; etc.

Vascular Lesions of the Orbit



Morning-glory disc anomaly in a PHACE pt

Q



I have a hunch PHACE is an acronym, with the 'H' standing for 'hemangiomas.'
Assuming I'm right, what does the rest stand for?

Posterior fossa malformations

Hemangiomas

Arterial lesions

Cardiac anomalies

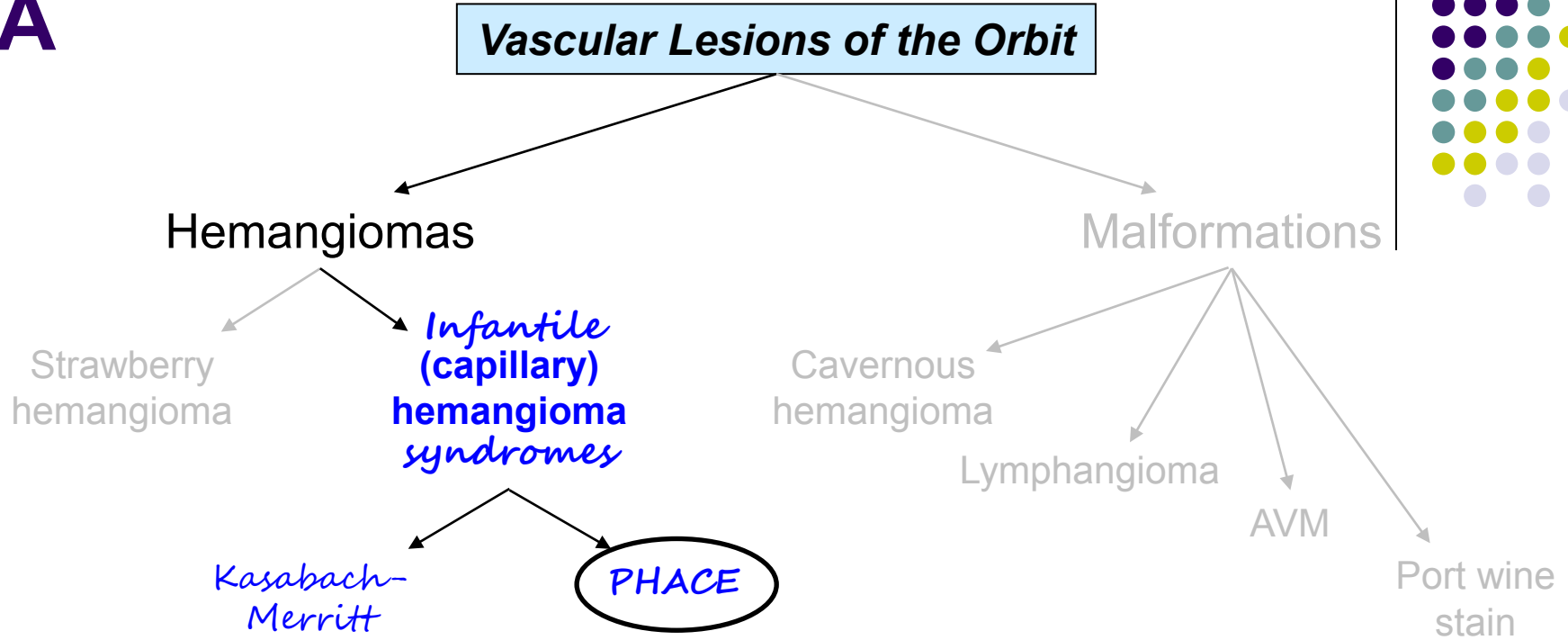
Eye anomalies

What sorts of eye anomalies' may be involved?

The usual suspects—cataracts; strabismus—along with sequelae of arrested development: colobomas; optic nerve hypoplasia; morning-glory disc; etc.

And not surprisingly, of the choroid.

A



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Posterior fossa malformations

Hemangiomas

Arterial lesions

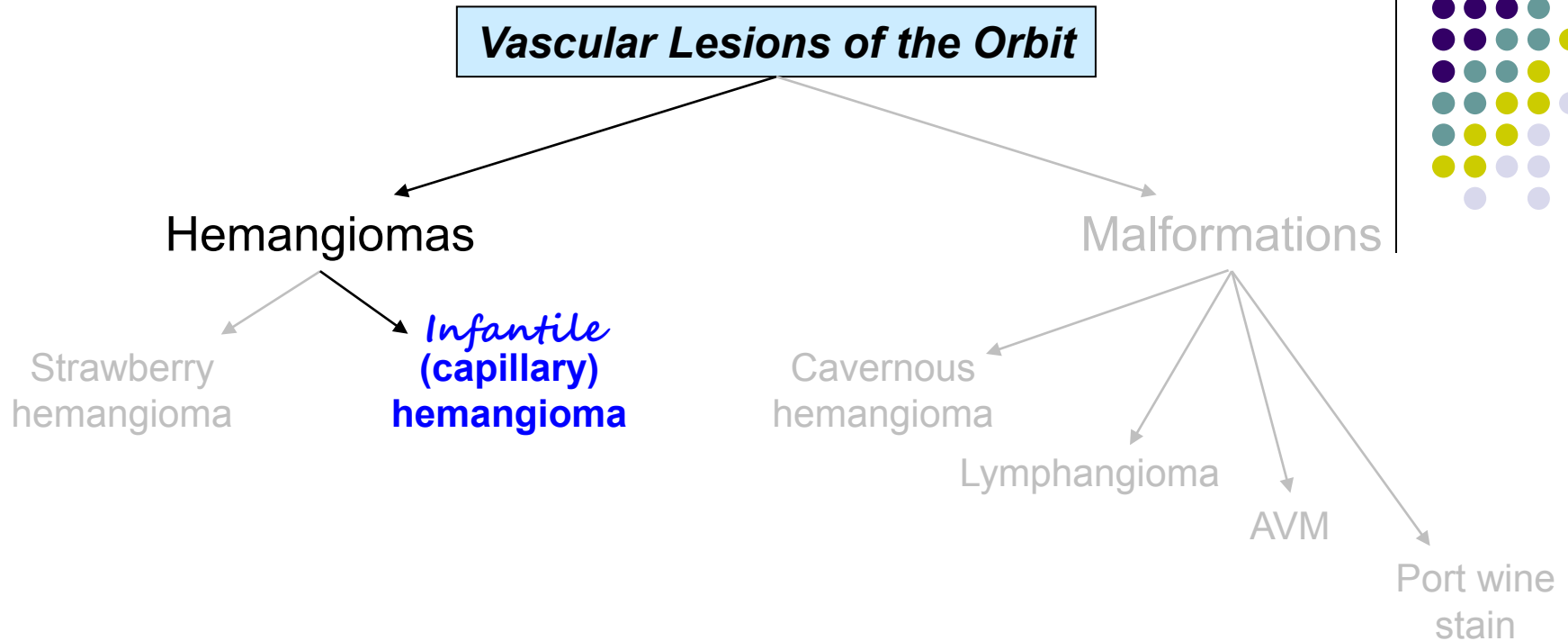
Cardiac anomalies

Eye anomalies

What sorts of eye anomalies' may be involved?

The usual suspects—cataracts; strabismus—along with sequelae of arrested development: colobomas; optic nerve hypoplasia; morning-glory disc; etc.

And not surprisingly, hemangioma of the choroid.



Next we'll address the management of capillary/infantile hemangiomas. Before we start, I must point out that the BCSC books are somewhat inconsistent on this topic, with three separate discussions (two in Orbit, one in Peds) that are not 100% in-sync with one another. We will go through each in turn.

No question—proceed when ready

Q

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

Port wine stain



Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?
For small, clinically un-concerning lesions, is reasonable

Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?
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Port wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Port wine stain

**A**

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Amblyopia—it is strongly associated with them

Port wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Amblyopia—it is strongly associated with them

How might a capillary hemangioma produce amblyopia?

Port wine stain

Q/A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Amblyopia—it is strongly associated with them

How might a capillary hemangioma produce amblyopia?

If it:

--occludes the visual axis

--induces or

Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Amblyopia—it is strongly associated with them

How might a capillary hemangioma produce amblyopia?

If it:

--occludes the visual axis

--induces astigmatism or strabismus

Port wine stain



Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Note that, even if a lesion is clinically concerning, observation *of the lesion itself* may be reasonable so long as the sequelae of concern can be otherwise addressed.

Port wine stain

No question—proceed when ready



Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, **clinically concerning lesions**, observation is reasonable

What is the main clinical concern stemming from capillary hemangiomas?

Note that, even if a lesion is clinically concerning, observation *of the lesion itself* may be reasonable so long as the sequelae of concern can be otherwise addressed. For example, if a lesion induces potentially amblyogenic astigmatism, **but** the astigmatism is correctable with specs, the clinician may choose to just observe the lesion while directly addressing the astigmatism. (Remember, with time essentially all hemangiomas resolve spontaneously.)

Port wine stain

No question—proceed when ready



Q

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, clinically un-concerning lesions, observation is reasonable

For lesions requiring treatment, what class of meds is considered first-line?

Port wine stain

**A*****Vascular Lesions of the Orbit*****Hemangiomas*****Per one section of the Orbit book:***

What are the options for managing infantile/capillary hemangiomas?

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For lesions requiring treatment, what class of meds is considered first-line?

Beta blockers

Port wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

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Beta blockers

There are two first-line first-line beta blockers, each with its own route of delivery. What determines which is selected?

Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

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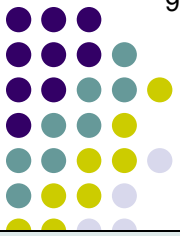
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Lesion depth

Port wine stain



Q

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Lesion depth

--Superficial: ?

--Deep(er)

Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

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Lesion depth

--Superficial: Topical timolol (BTW: It's a gel, not a bottle of the glaucoma med, bro)

--Deep(er)



Port wine stain

Q

Vascular Lesions of the Orbit

Hemangiomas

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Topical beta blockers are very safe, but are the safe enough to be administered by an eye dentist?

Port wine stain

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Vascular Lesions of the Orbit

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--Deep(er)

Topical beta blockers are very safe, but are the safe enough to be administered by an eye dentist?

Yes—even an eye dentist can't screw this up

Port wine stain



Q

Vascular Lesions of the Orbit

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Port wine stain



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--Deep(er): PO propranolol

Port wine stain



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--Deep(er) **PO propranolol**

*Again—very safe. But are systemic beta blockers safe enough for **you** to administer?*

Port wine stain

Q/A

Vascular Lesions of the Orbit

Hemangiomas

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--Deep(er) **PO propranolol**

*Again—very safe. But are systemic beta blockers safe enough for **you** to administer?*

No—although rare, [redacted], [redacted], [redacted] and/or [redacted] may result



Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

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*Again—very safe. But are systemic beta blockers safe enough for **you** to administer?*

No—although rare, hypotension, bradycardia, bronchospasm, and/or hypoglycemia may result



Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

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Port wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

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Again—very safe. But are systemic beta blockers safe enough for you to administer?
 No—although rare, hypotension, bradycardia, bronchospasm, and/or hypoglycemia may result. Let your friends in Peds administer and manage systemic beta blocker tx.

One group of hemangioma pts are particularly vulnerable to severe complication— which one?

Port wine stain

Q/A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

*What are the options for managing infantile/capillary hemangiomas?
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Lesion depth

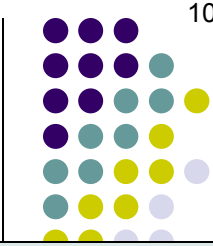
--Superficial: Topical timolol (BTW: It's a gel, not a bottle of the glaucoma med, bro)

--Deep(er) **PO propranolol**

*Again—very safe. But are systemic beta blockers safe enough for you to administer?
No—although rare, hypotension, bradycardia, bronchospasm, and/or hypoglycemia may result. Let your friends in Peds administer and manage systemic beta blocker tx.*

*One group of hemangioma pts are particularly vulnerable to severe complication—
which one?*

PHACE pts—systemic beta blockers put them at significant risk of **abb.**



Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per one section of the Orbit book:

What are the options for managing infantile/capillary hemangiomas?

For small, clinically un-concerning lesions, observation is reasonable

For lesions requiring treatment, what class of meds is considered first-line?

Beta blockers

There are two first-line beta blockers, each with its own route of delivery. What determines which is selected? Which beta blocker (and route) is indicated for each lesion type?

Lesion depth

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--Deep(er) **PO propranolol**

*Again—very safe. But are systemic beta blockers safe enough for **you** to administer?*

No—although rare, hypotension, bradycardia, bronchospasm, and/or hypoglycemia may result. Let your friends in Peds administer and manage systemic beta blocker tx.

*One group of hemangioma pts are particularly vulnerable to severe complication—**which one?***

PHACE pts—systemic beta blockers put them at significant risk of CVA

Port wine stain

Q

Vascular Lesions of the Orbit

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Per one section of the Orbit book:

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--Deep(er): PO propranolol

If beta blockers are ineffective, what is the second-line option and its delivery route(s)?

Port wine stain

A

Vascular Lesions of the Orbit

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Per one section of the Orbit book:

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--Deep(er): PO propranolol

If beta blockers are ineffective, what is the second-line option and its delivery route(s)?

Steroids delivered topically, intralesionally, or systemically

Port wine stain

Q

Vascular Lesions of the Orbit

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Steroids delivered topically, intralesionally, or systemically

And if steroids are ineffective too? What then?

Port wine stain



A

Vascular Lesions of the Orbit

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Steroids delivered topically, intralesionally, or systemically

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)

Port wine stain



Vascular Lesions of the Orbit

Hemangiomas

*Per the **other** Orbit section:*

What are the options for managing infantile/capillary hemangiomas?
For small, clinically un-concerning lesions, observation is reasonable



For lesions requiring treatment, what class of meds is considered first-line?
Beta blockers



There are two first-line first-line beta blockers, each with its own route of delivery. What determines which is selected? Which beta blocker (and route) is indicated for each lesion type?
Lesion depth

--Superficial: Topical timolol (BTW: It's a gel, not a bottle of the glaucoma med, bro)
--Deep(er): PO propranolol



The other *Orbit* section on managing hemangiomas is consistent with these first steps...

Port wine stain

No question on this or the next slide—proceed when ready



Vascular Lesions of the Orbit

Hemangiomas

Per the other Orbit section:

What are the options for managing infantile/capillary hemangiomas?
For small, clinically un-concerning lesions, observation is reasonable



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--Deep(er): PO propranolol



The other *Orbit* section on managing hemangiomas is consistent with these first steps...

As well as in considering incisional surgery the intervention of last resort.

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

Q

Vascular Lesions of the Orbit

Hemangiomas

Per the other Orbit section:

*What are the options for managing infantile/capillary hemangiomas?
For small, clinically un-concerning lesions, observation is reasonable*



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Lesion depth
--Superficial: Topical timolol (BTW: It's a gel, not a bottle of the glaucoma med, bro)
--Deep(er): PO propranolol



If beta blockers are ineffective, what is the second-line option and its delivery route(s)?

However, it differs in its response to *this* question, which proceeds as follows...

*Now answer the question in **green** above*

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

Per the other Orbit section:

*What are the options for managing infantile/capillary hemangiomas?
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If beta blockers are ineffective, what is the second-line option and its delivery route(s)?
Steroids delivered systemically (PO)

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

Per the other Orbit section:

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Steroids delivered systemically (PO)

If PO steroids are ineffective, what is the next option and its delivery route?

And if steroids are ineffective too? What then?

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Port wine stain

Q/A

Vascular Lesions of the Orbit

Hemangiomas

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Steroids delivered systemically (PO)

If PO steroids are ineffective, what is the next option and its delivery route?

Steroids delivered

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

A

Vascular Lesions of the Orbit

Hemangiomas

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If PO steroids are ineffective, what is the next option and its delivery route?

Steroids delivered intralesionally

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

Q

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Hemangiomas

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Steroids delivered systemically (PO)

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Steroids delivered intralesionally

And if intralesional steroids are ineffective too? What then?

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

Q/A

Vascular Lesions of the Orbit

Hemangiomas

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If beta blockers are ineffective, what is the second-line option and its delivery route(s)?
Steroids delivered systemically (PO)

If PO steroids are ineffective, what is the next option and its delivery route?
Steroids delivered intralesionally

And if intralesional steroids are ineffective too? What then?
Consider topical

And if steroids are ineffective too? What then?
Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain



A

Vascular Lesions of the Orbit

Hemangiomas

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If beta blockers are ineffective, what is the second-line option and its delivery route(s)?

Steroids delivered systemically (PO)

If PO steroids are ineffective, what is the next option and its delivery route?

Steroids delivered intralesionally

And if intralesional steroids are ineffective too? What then?

Consider topical clobetasol

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

Q

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Hemangiomas

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Steroids delivered systemically (PO)

If PO steroids are ineffective, what is the next option and its delivery route?

Steroids delivered intralesionally

And if intralesional steroids are ineffective too? What then?

Consider topical clobetasol, or in dire (= life- or sight-threatening) cases, systemic

abb.

And if steroids are ineffective too? What then?

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

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Hemangiomas

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Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain



Vascular Lesions of the Orbit

Hemangiomas

Per the Peds book:

What are the options for managing infantile/capillary hemangiomas?
For small, clinically un-concerning lesions, observation is reasonable



For lesions requiring treatment, what class of meds is considered first-line?
Beta blockers



The *Peds* book on managing hemangiomas is consistent with these first answers...

Port wine stain

No question—proceed when ready



Vascular Lesions of the Orbit

Hemangiomas

Per the Peds book:

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...and is *sorta* consistent about beta-blocker preference (it indicates systemic propranolol is first-line, but 'topical timolol may be effective if the lesion is superficial')

Port wine stain

No question on this or the next slide—proceed when ready



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Port wine stain

It also considers incisional surgery an intervention of last resort.

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Q

Vascular Lesions of the Orbit

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If beta blockers are ineffective, what is the second-line option?

But it differs in its response to *this* question, which is...

Now answer the question in **red** above

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain

A

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If beta blockers are ineffective, what is the second-line option?

Pulsed-dye laser (for superficial lesions)

Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)



Port wine stain



Vascular Lesions of the Orbit

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If beta blockers are ineffective, what is the second-line option?

Pulsed-dye laser (for superficial lesions)

And that's it per the *Peds* book—otherwise, straight to surgery.

No question—proceed when ready



Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)

Port wine stain



Vascular Lesions of the Orbit

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Per the Peds book:

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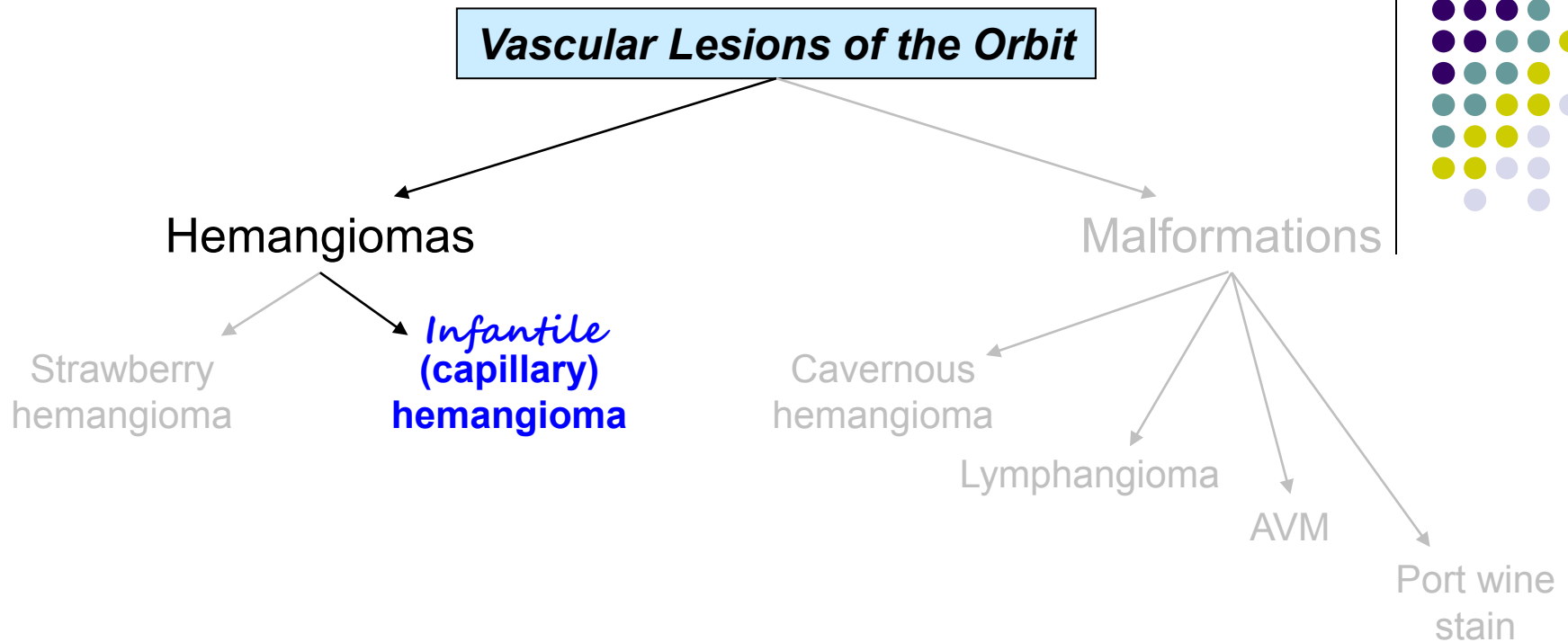
And that's it per the *Peds* book—otherwise, straight to surgery. ***Note that it does not mention steroids as an intervention to be considered.***

No question—proceed when ready



Surgical excision can be considered (but bring extra hemostats, cuz it's gonna bleed)

Port wine stain

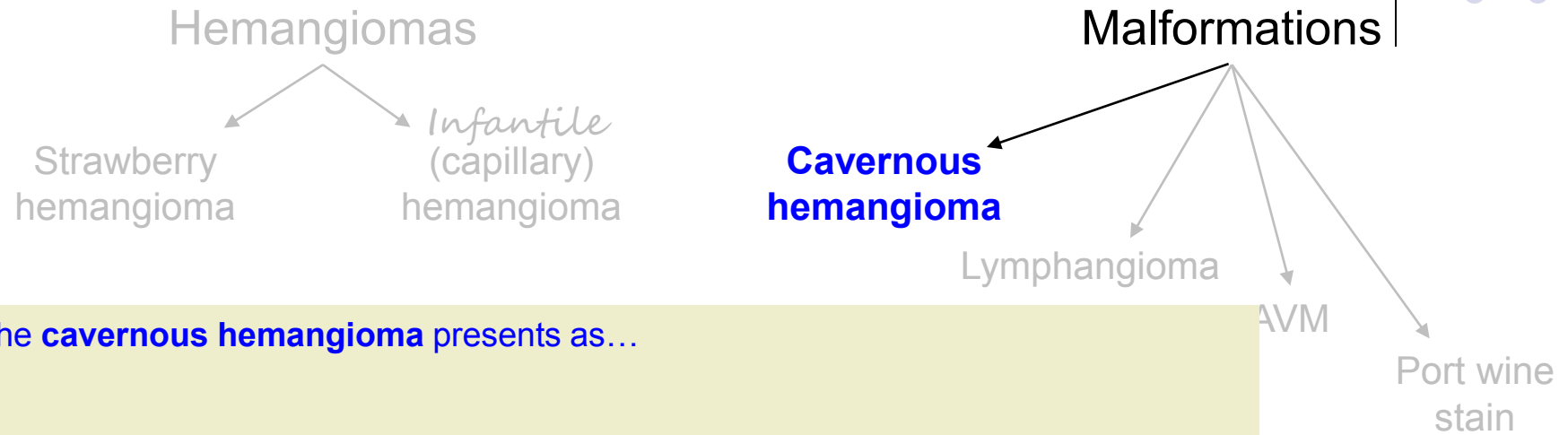


*Next we'll address the management of capillary/infantile hemangiomas. Before we start, I must point out that the BCSC books are somewhat inconsistent on this topic, with three separate discussions (two in Orbit, one in Peds) that are not 100% in-sync with one another. We will go through each in turn. **Given the inconsistencies just discussed, how should you answer questions about hemangioma management on the OKAP and Boards? Very carefully. Good luck.***

No question—proceed when ready

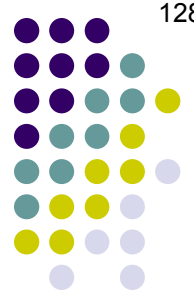
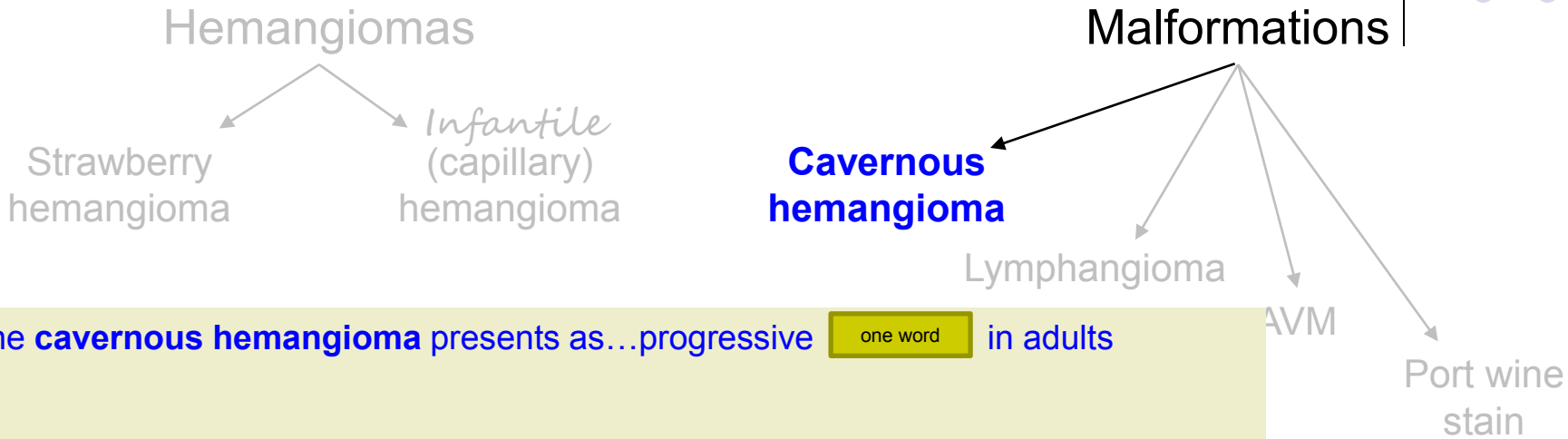
Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...

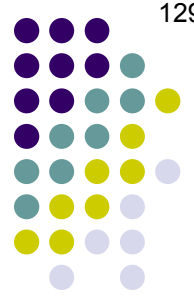
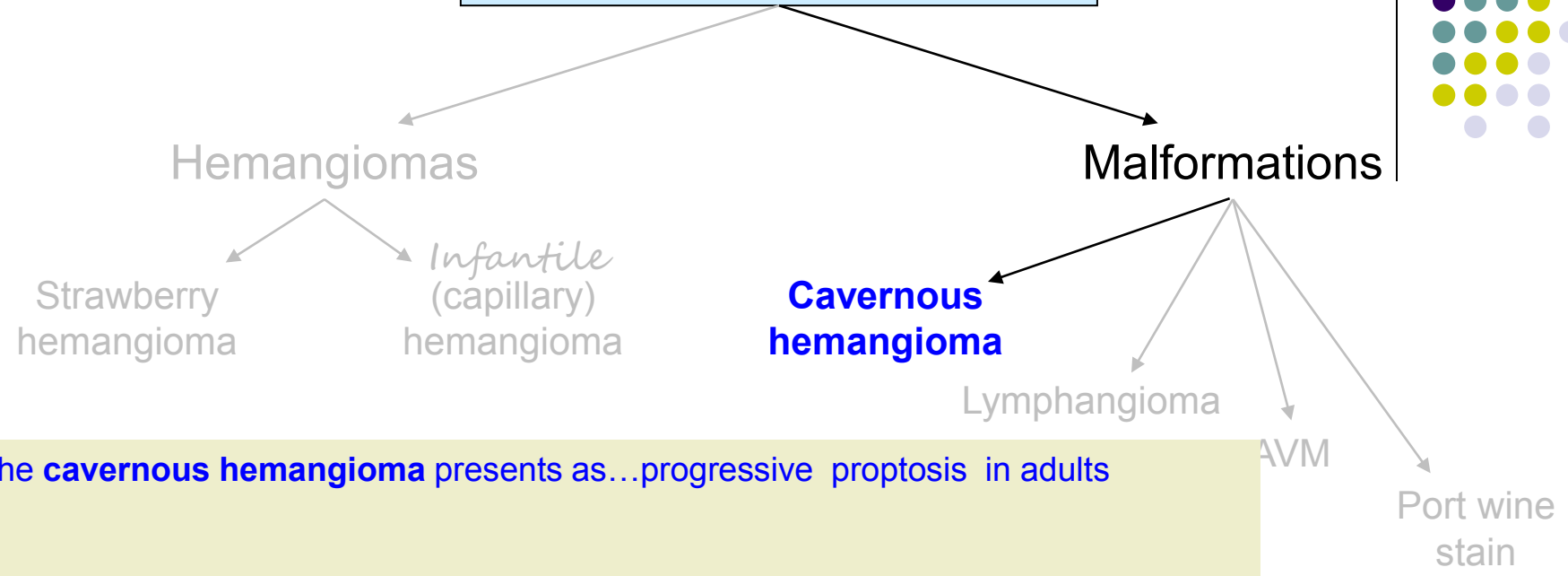
Vascular Lesions of the Orbit



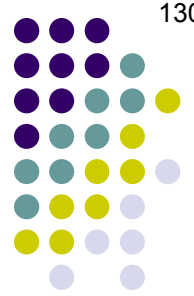
The **cavernous hemangioma** presents as...progressive one word in adults

A

Vascular Lesions of the Orbit

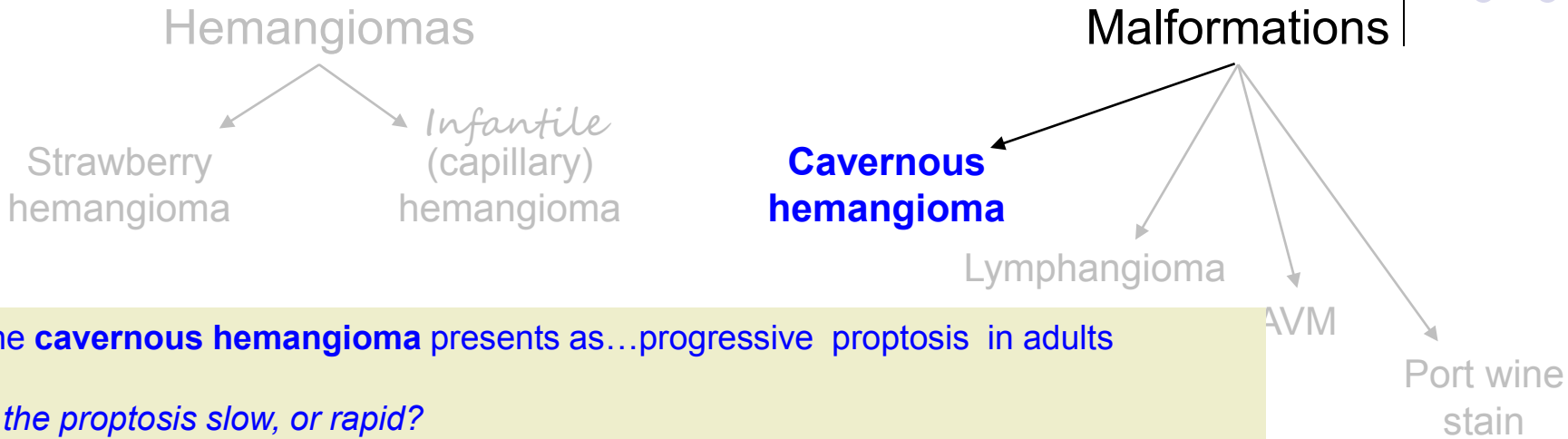


The **cavernous hemangioma** presents as...progressive proptosis in adults



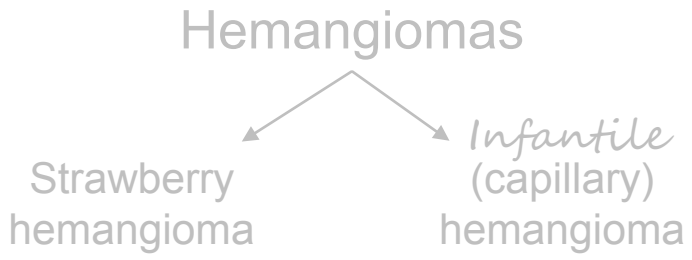
Q

Vascular Lesions of the Orbit



A

Vascular Lesions of the Orbit



Malformations

Cavernous hemangioma

Lymphangioma

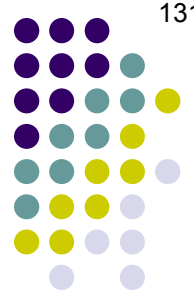
AVM

Port wine stain

The **cavernous hemangioma** presents as...progressive proptosis in adults

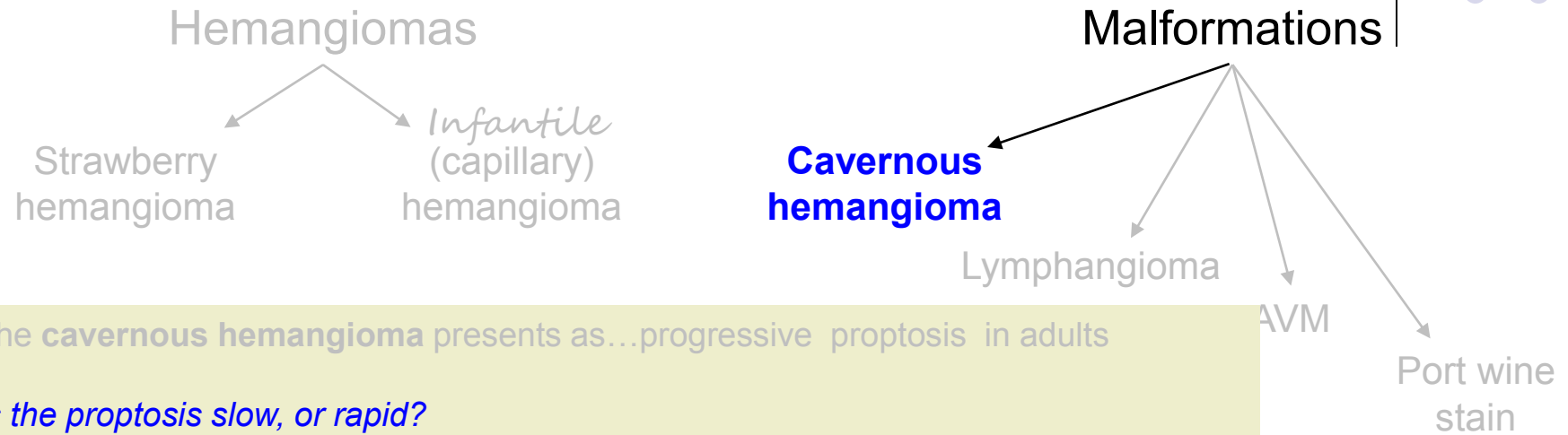
Is the proptosis slow, or rapid?

Slow



Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

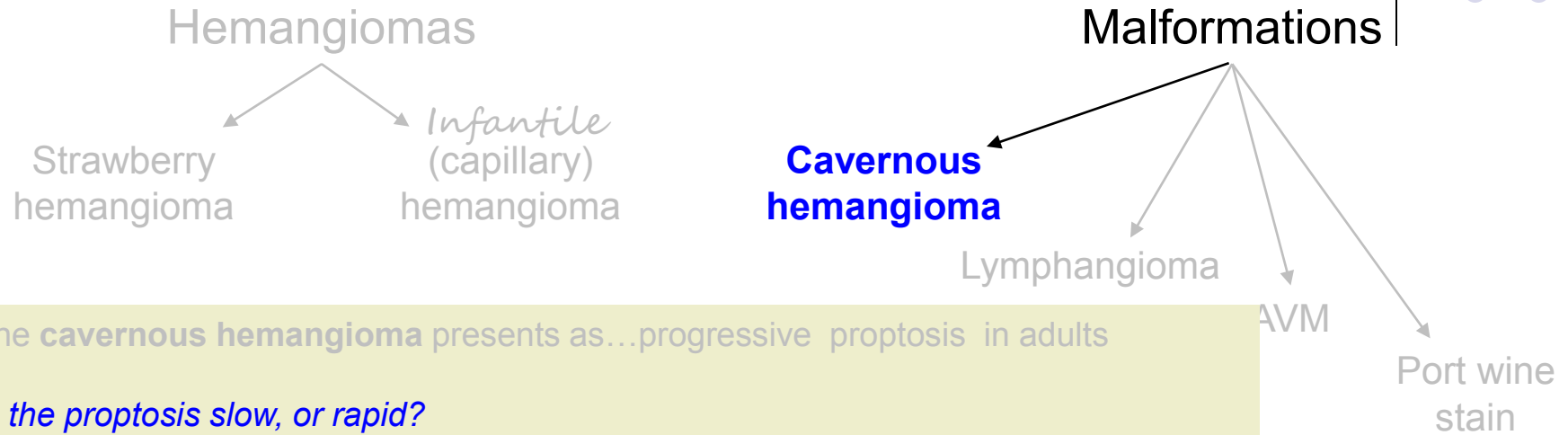
Is the proptosis slow, or rapid?

~~Slow~~ Rapid during...

What common medical status is associated with more rapid proptosis?

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

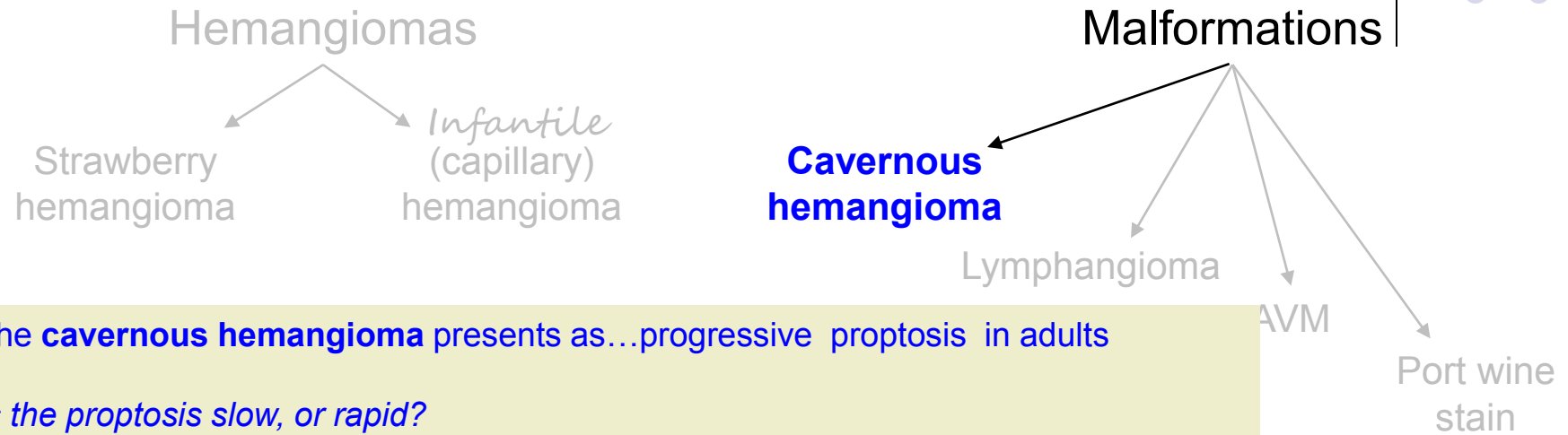
~~Slow~~ Rapid during...pregnancy

What common medical status is associated with more rapid proptosis?

Pregnancy

Q

Vascular Lesions of the Orbit



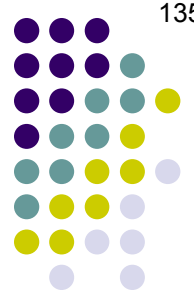
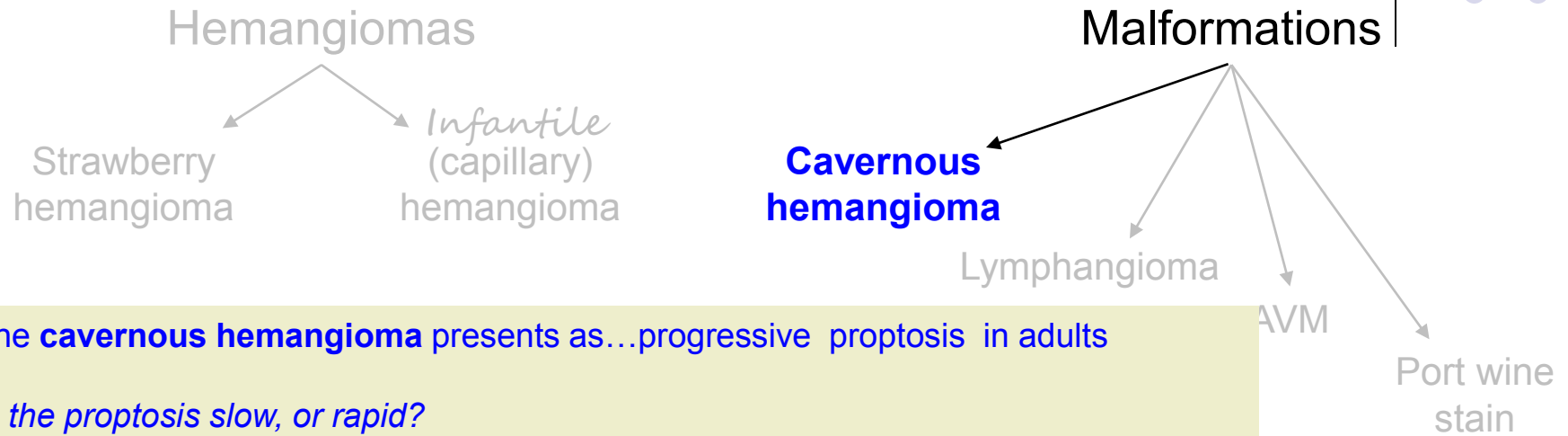
The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Vascular Lesions of the Orbit



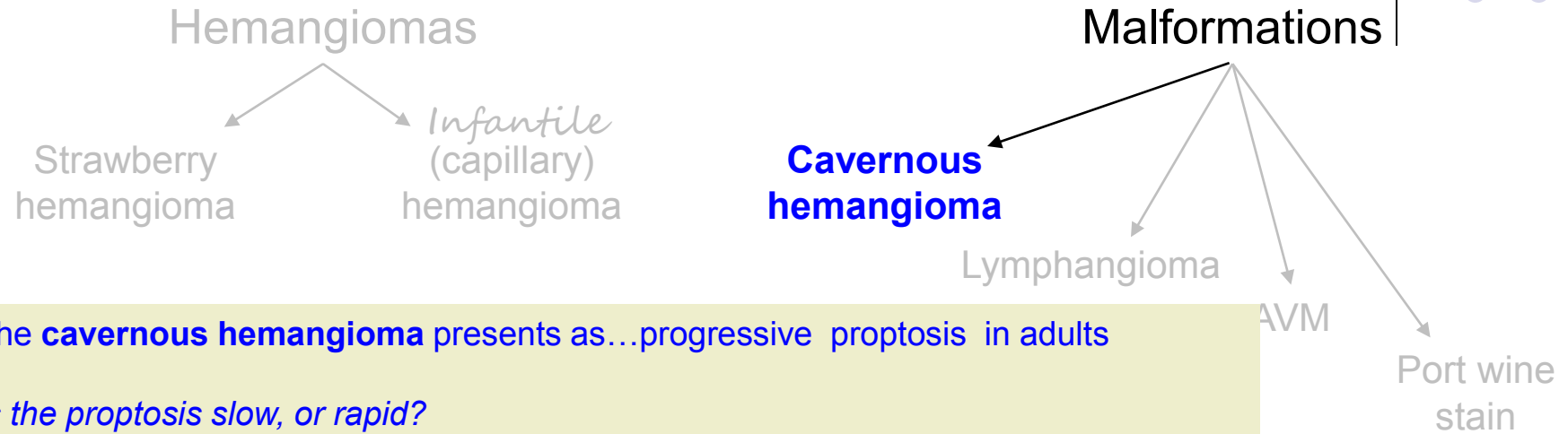
The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?
Slow

Is there a gender predilection?
Yes, it is more common in M v F

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

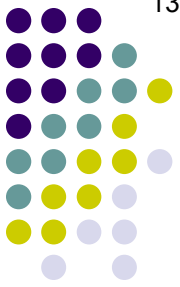
Is the proptosis slow, or rapid?

Slow

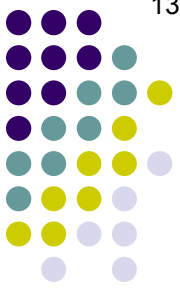
Is there a gender predilection?

Yes, it is more common in women

Vascular Lesions of the Orbit



62-year-old female with painless proptosis noticeable over the last year.



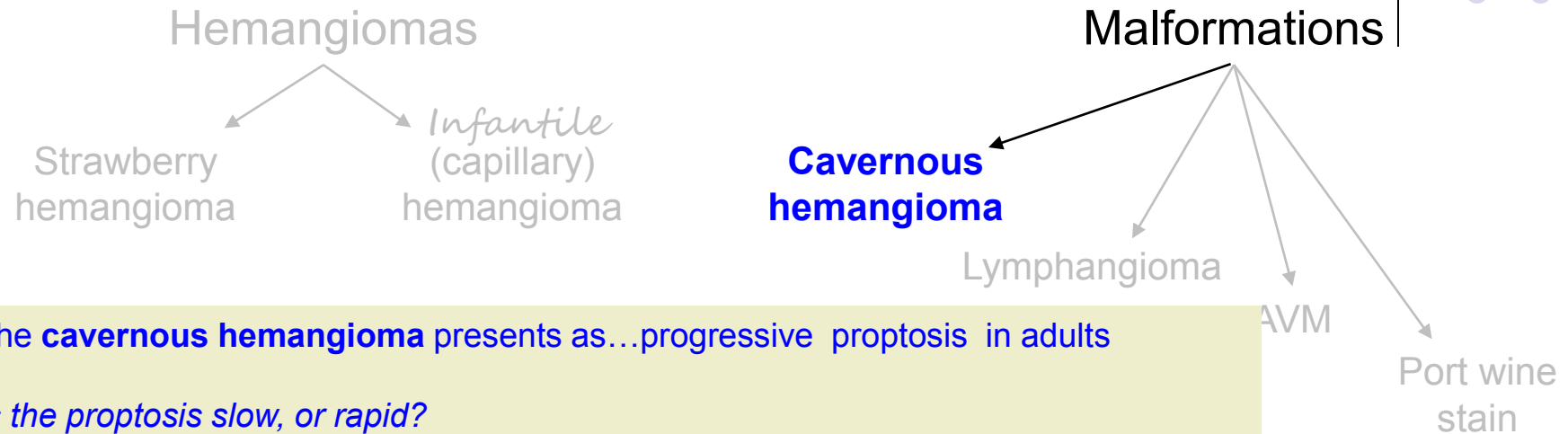
Vascular Lesions of the Orbit



62-year-old female with painless proptosis noticeable over the last year.
Right hyperglobus leading to inferior scleral show is evident.

Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

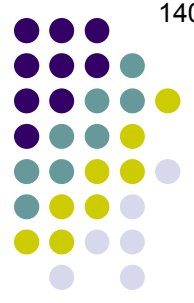
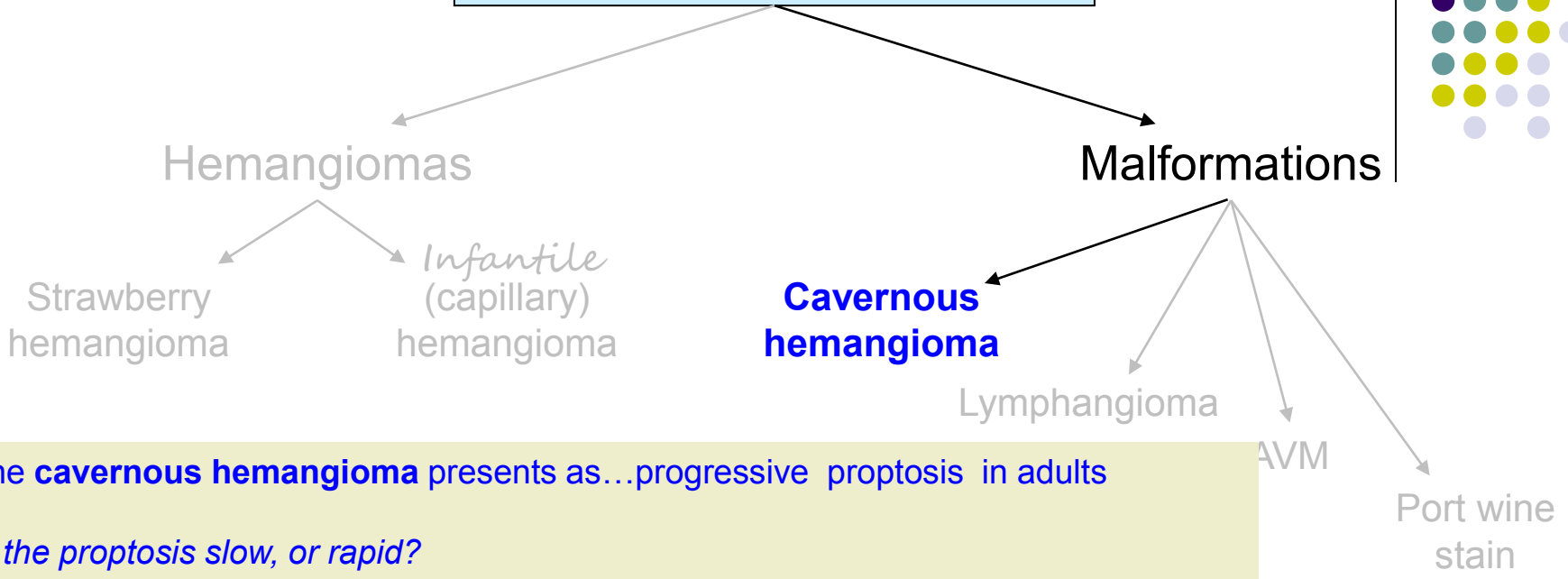
Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

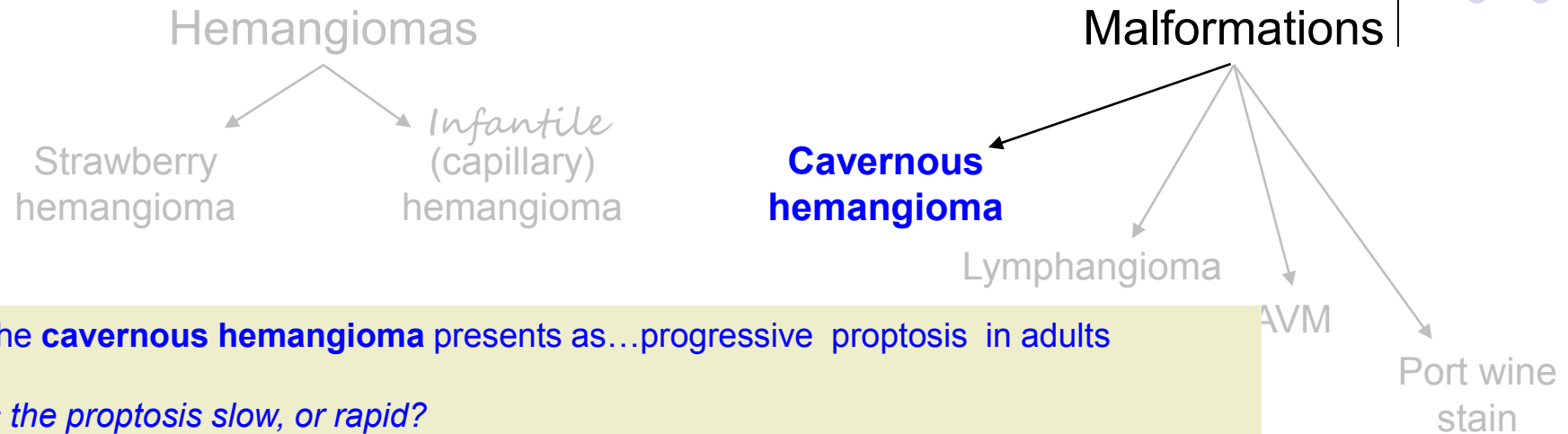
Is the proptosis slow, or rapid?
Slow

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

Is it benign, or malignant?
Benign

Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

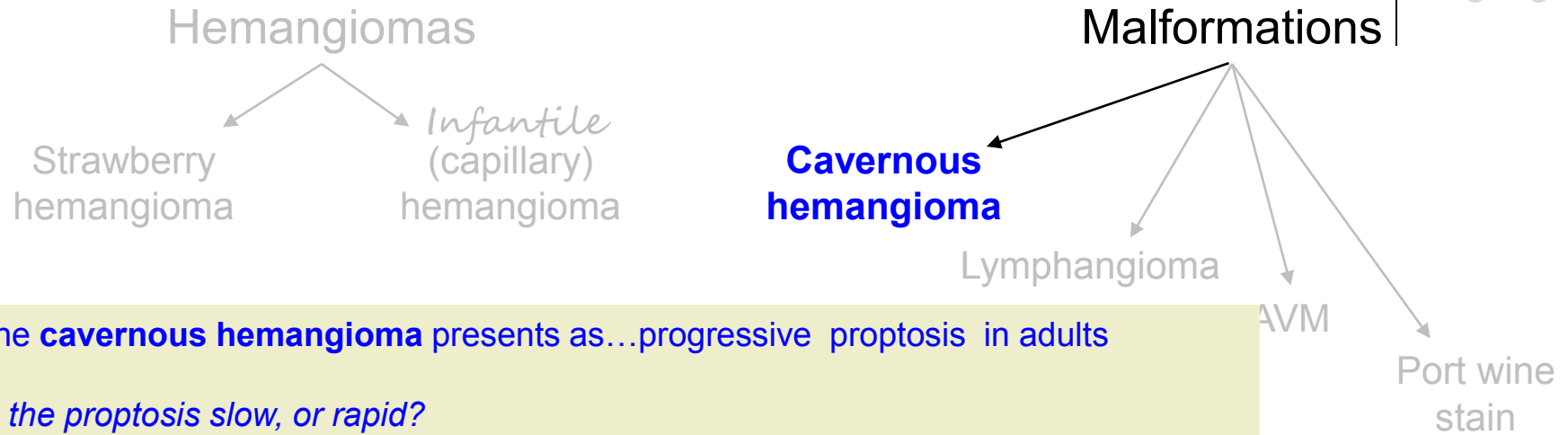
Yes, it is more common in women

Is it benign, or malignant?

Benign

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

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

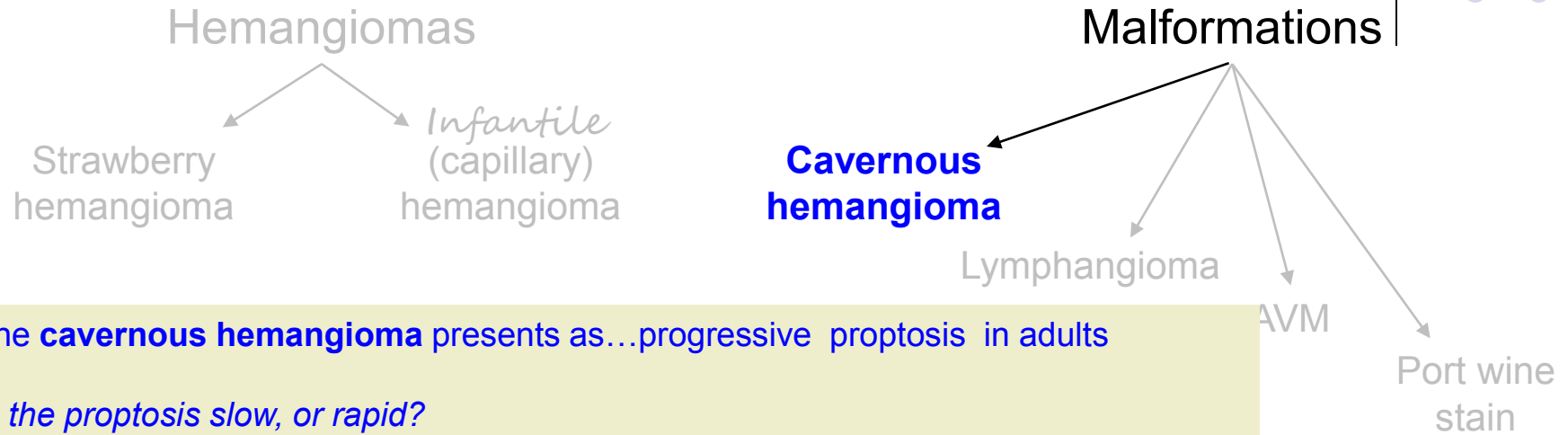
Benign

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

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

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

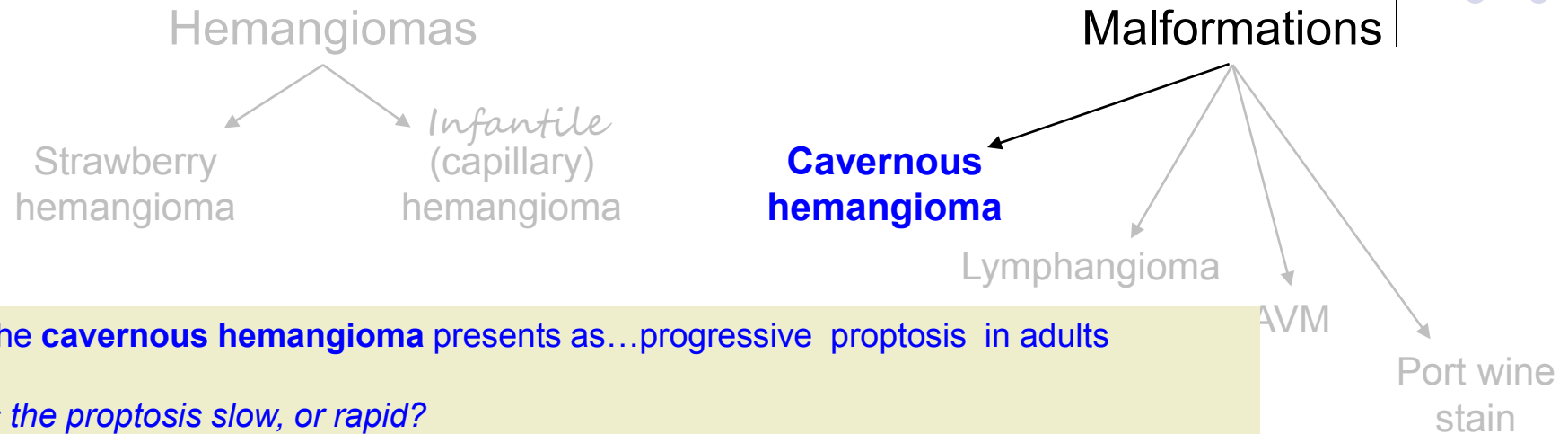
Benign

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

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

Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

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Is it benign, or malignant?

Benign

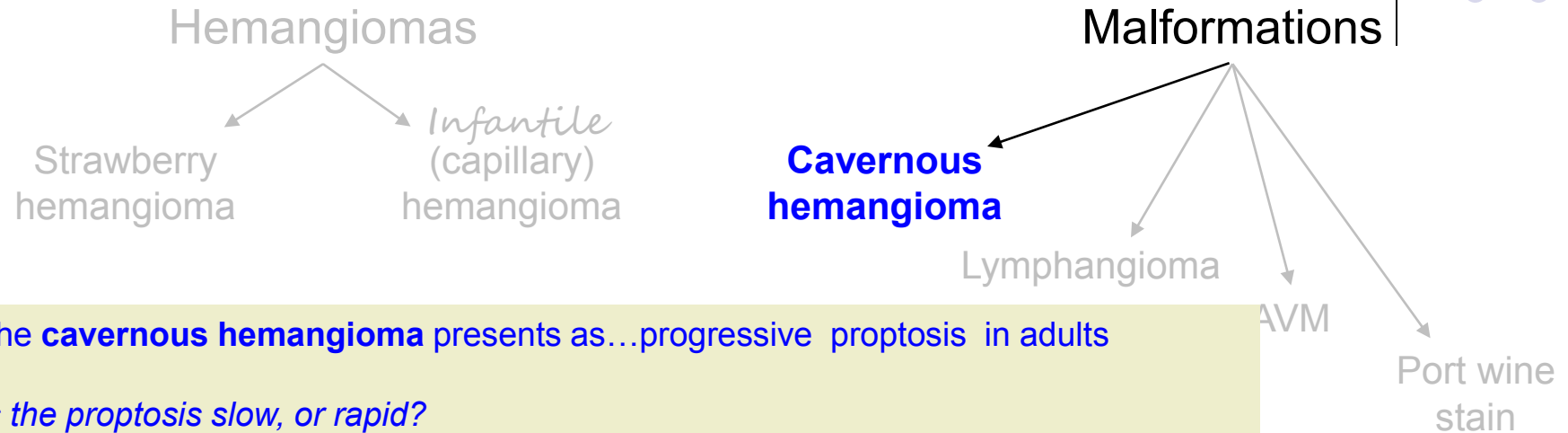
Where does the cavernous hemangioma rank among benign orbital tumors in adults?

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

Does it require surgical excision?

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

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

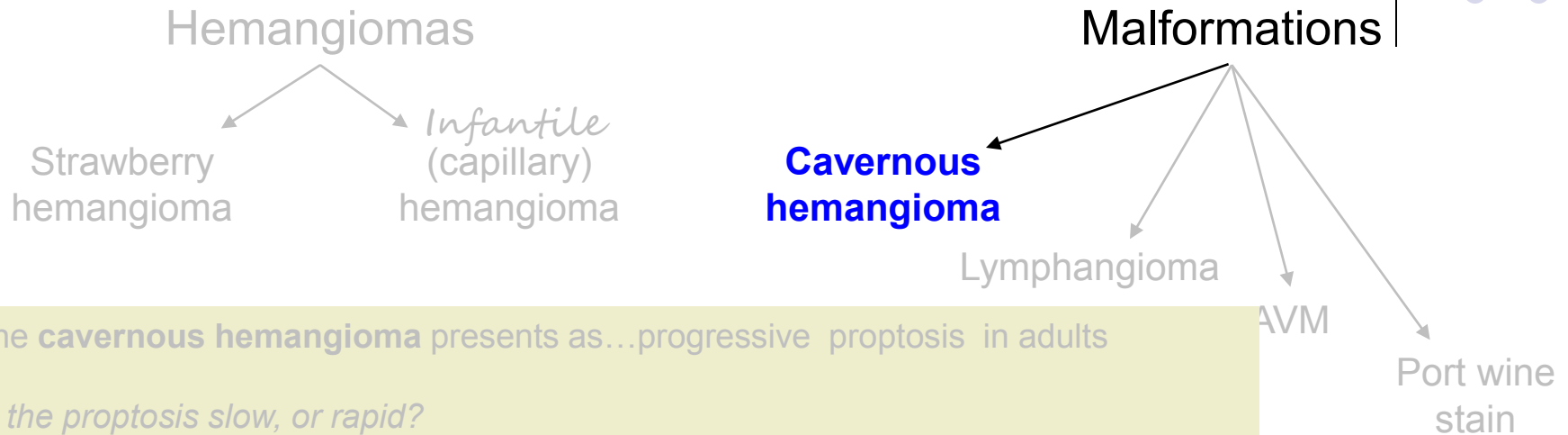
It is the **most** common benign orbital tumor of adults

Does it require surgical excision?

Yes, if it is clinically significant

Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma be clinically significant?

--?

--?

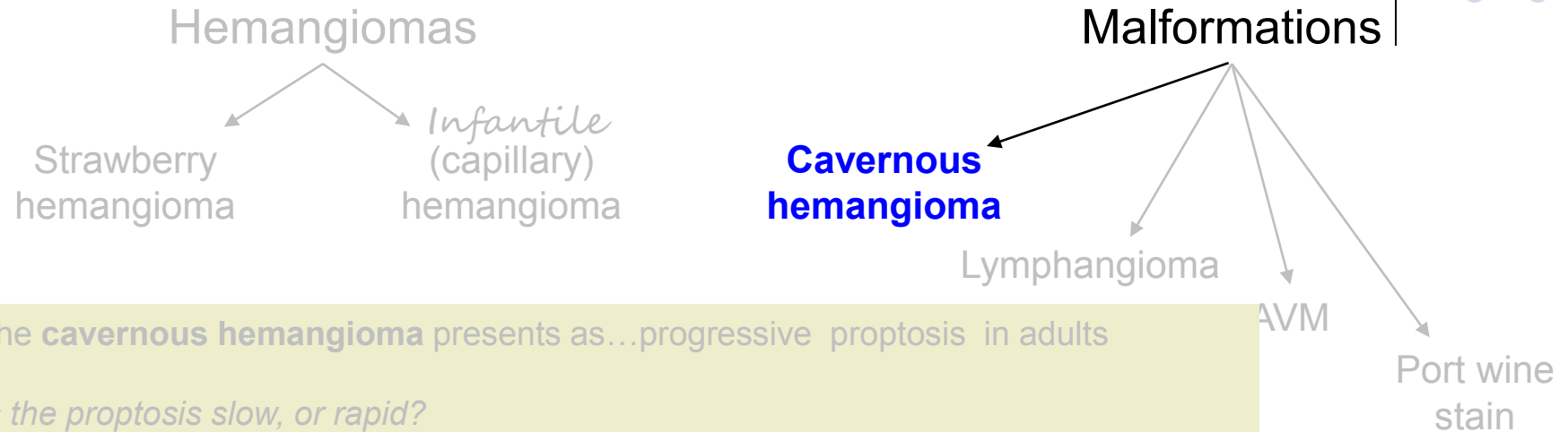
--?

Does it require surgical excision?

Yes, if it is **clinically significant**

Q/A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma be clinically significant?

--By affecting

--By increasing

--By causing

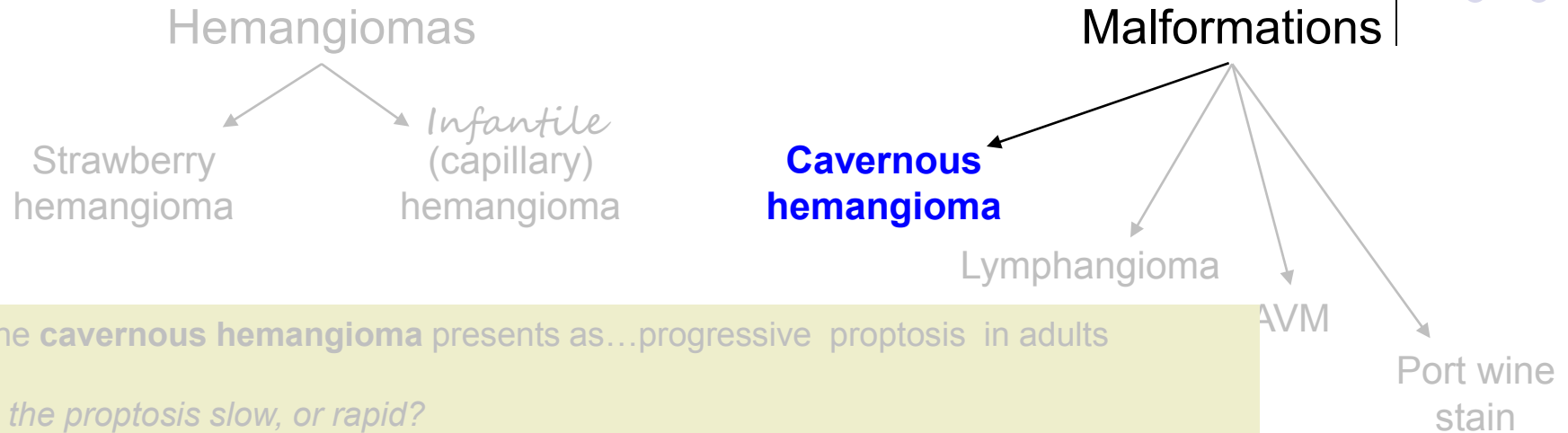
Does it require surgical excision?

Yes, if it is **clinically significant**

How might a cavernous hemangioma be clinically significant?

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma be clinically significant?

--By affecting vision

--By increasing IOP

--By causing strabismus

Does it require surgical excision?

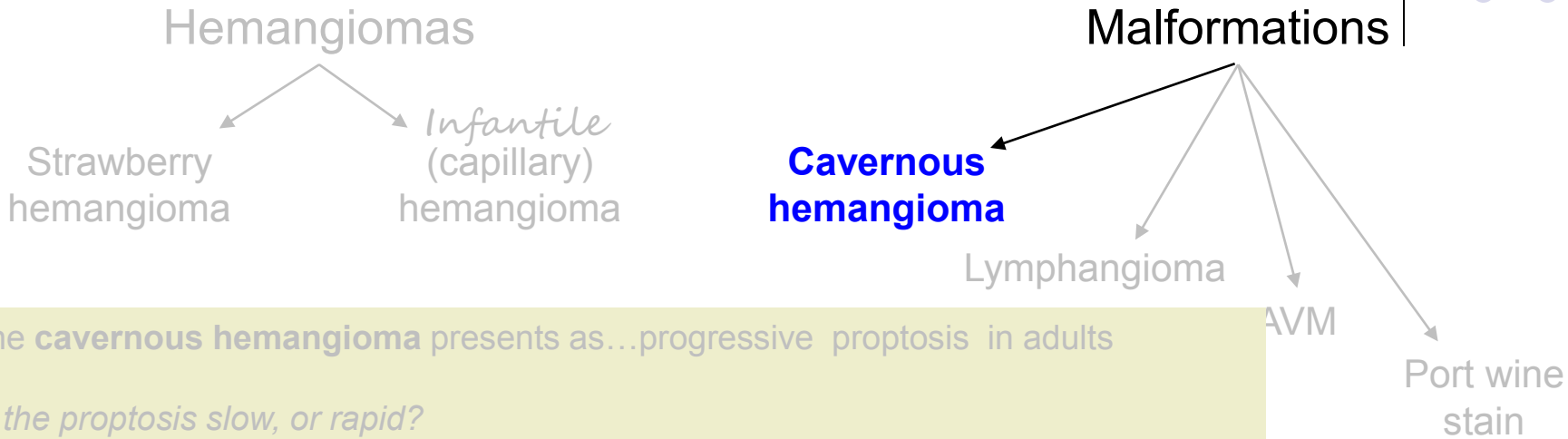
Yes, if it is **clinically significant**

How might a cavernous hemangioma be clinically significant?



Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?
Slow

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

Is it benign, or malignant?
Benign

How might a cavernous hemangioma affect vision?

- By **affecting vision**
- By increasing IOP
- By causing strabismus

Does it require surgical excision?
Yes, if it is **clinically significant**

How might a cavernous hemangioma affect vision?

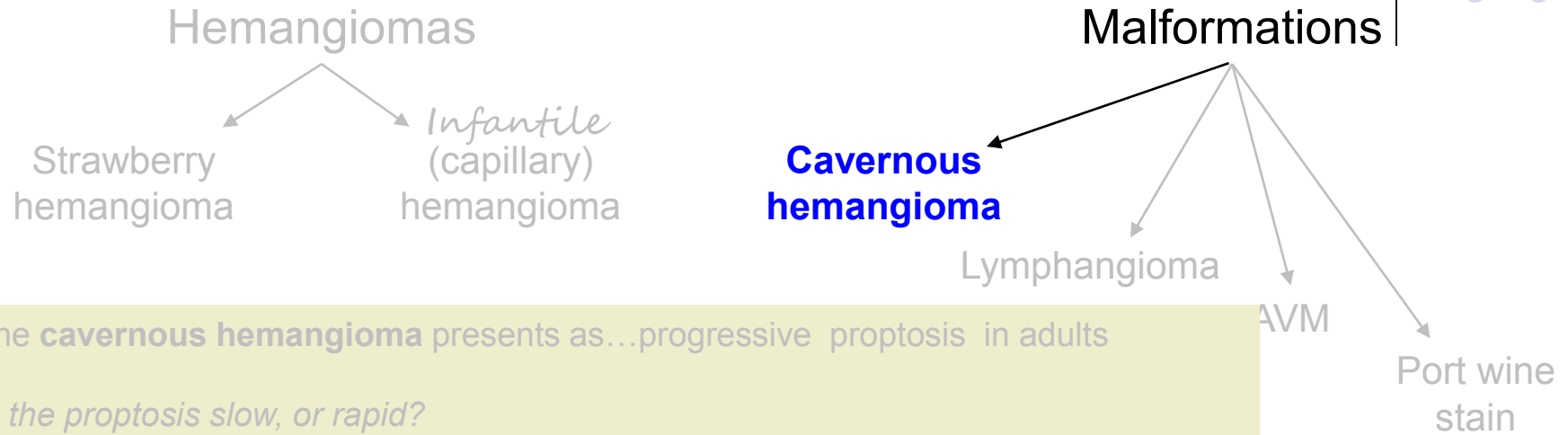
--?

--?

...s in adults?

Q/A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma affect vision?

--By **affecting vision**

--By increasing IOP

--By causing strabismus

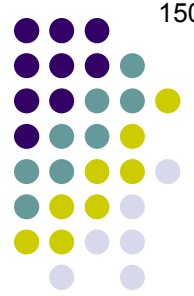
How might a cavernous hemangioma affect vision?

--By causing retinal [] or []

--?

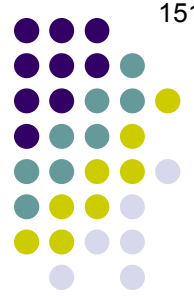
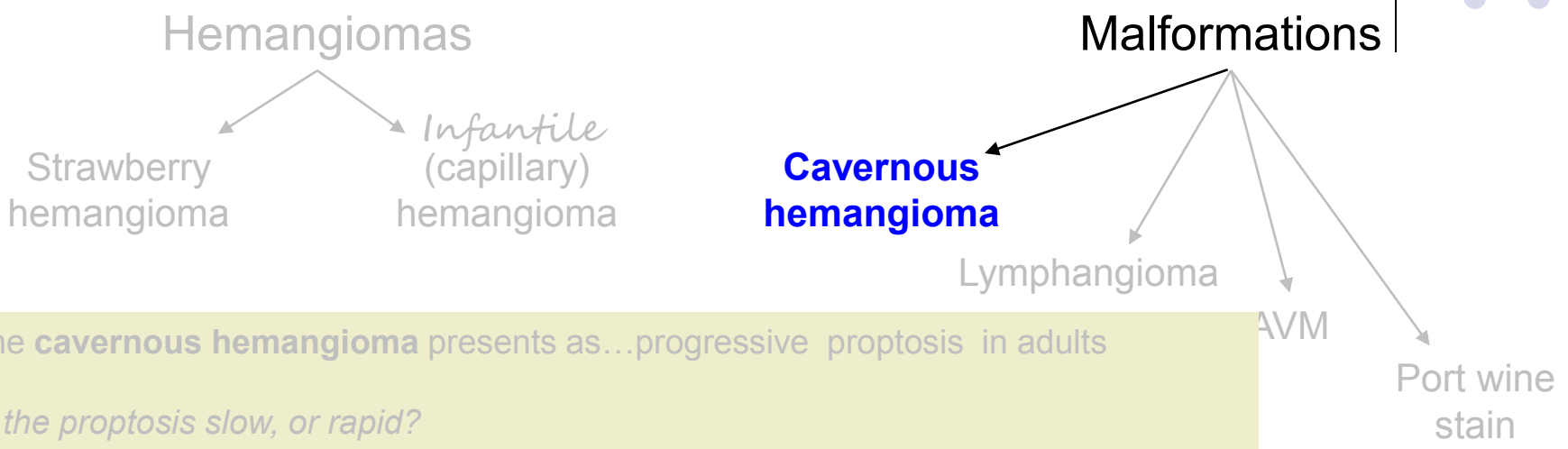
Does it require surgical excision?

Yes, if it is **clinically significant**



A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?
Slow

Is there a gender predilection?
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How might a cavernous hemangioma affect vision?
 --By **affecting vision**
 --By increasing IOP
 --By causing strabismus

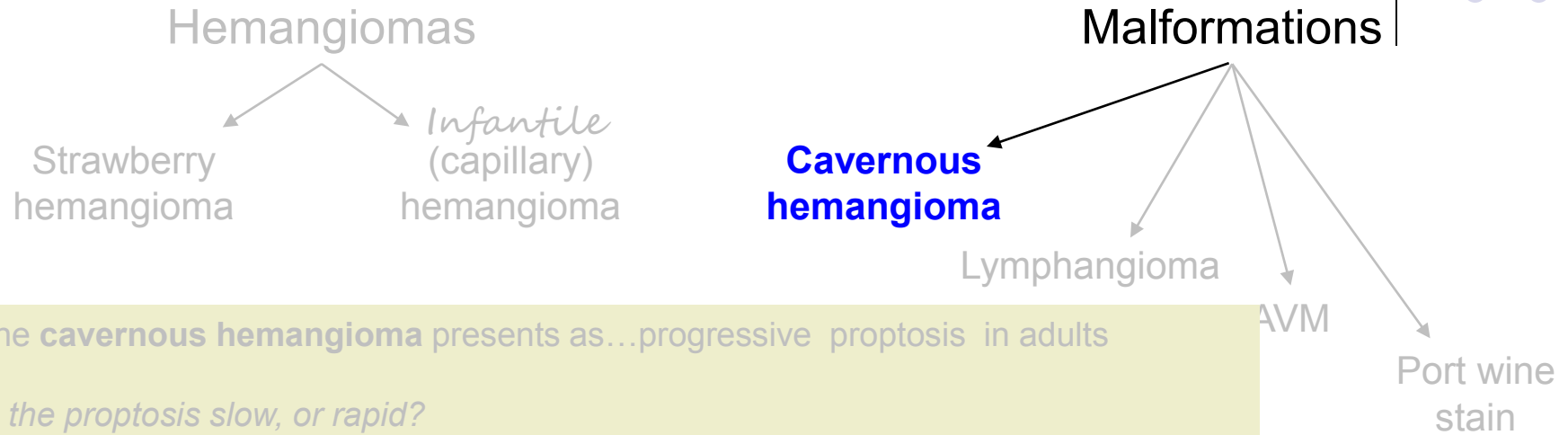
Does it require surgical excision?
Yes, if it is **clinically significant**

How might a cavernous hemangioma affect vision?
 --By causing retinal striae, or hyperopia
 --?

...s in adults?

Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?
Slow

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

Is it benign, or malignant?
Benign

How might a cavernous hemangioma affect vision?
--By **affecting vision**
--By increasing IOP
--By causing strabismus

Does it require surgical excision?
Yes, if it is **clinically significant**

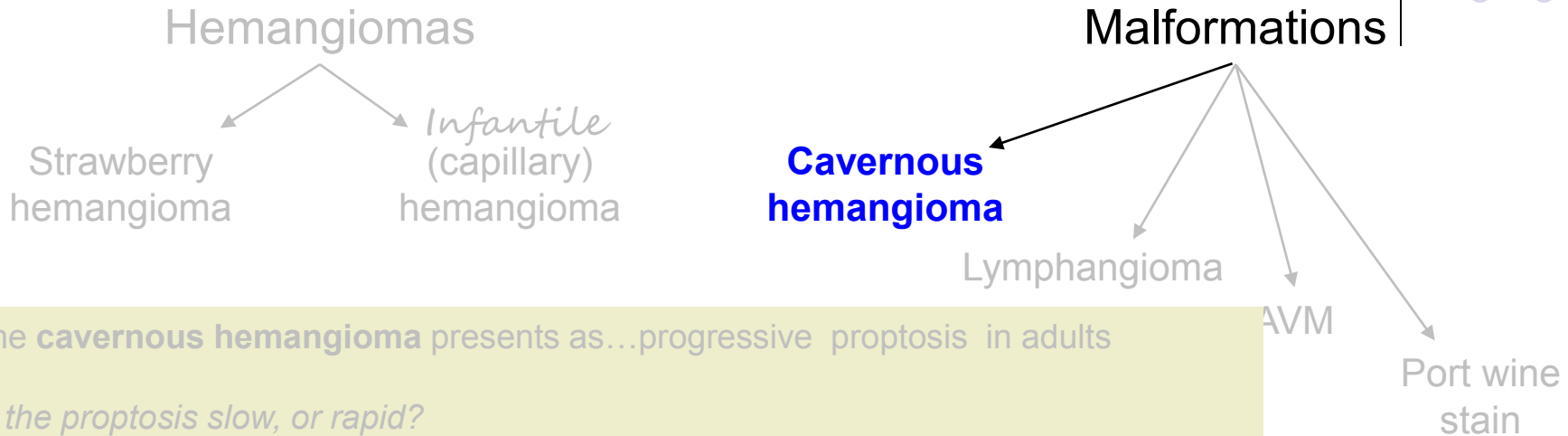
How could a cavernous hemangioma cause striae and/or hyperopia?

How might a cavernous hemangioma affect vision?
--By **causing retinal striae, or hyperopia**
--?

...s in adults?

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

Is there a gender predilection?

Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma affect vision?

--By **affecting vision**

--By increasing IOP

--By causing strabismus

How could a cavernous hemangioma cause striae and/or hyperopia?

By compressing the back of the globe

How might a cavernous hemangioma affect vision?

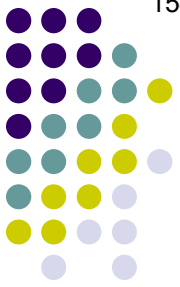
--By **causing retinal striae, or hyperopia**

--?

Does it require surgical excision?

Yes, if it is **clinically significant**

Vascular Lesions of the Orbit

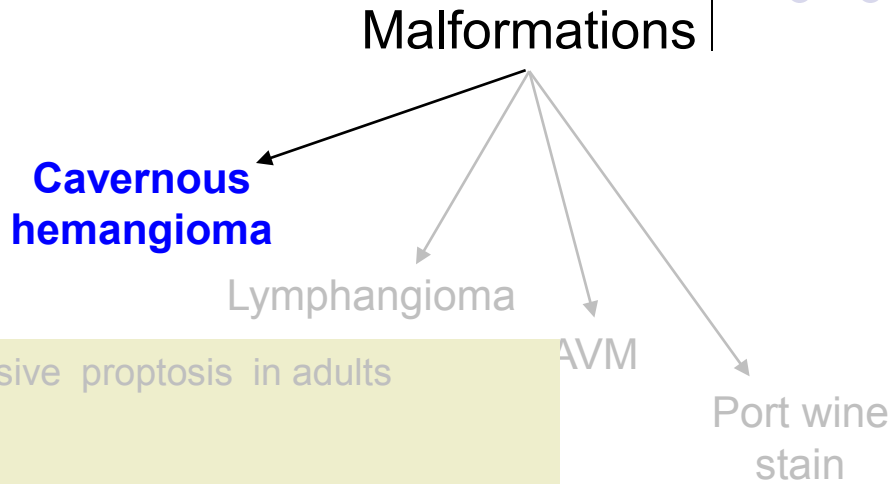
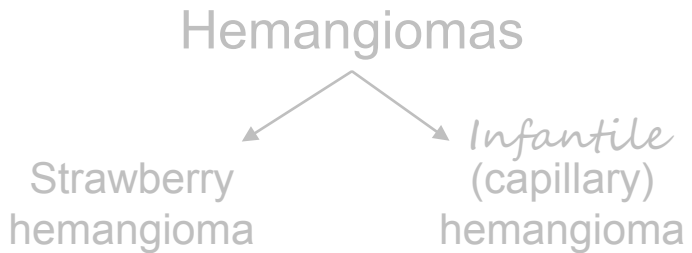


Color fundus photograph of the right eye demonstrating retinal striae in a pt with an intraconal cavernous hemangioma compressing the posterior pole



Q

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?
Slow

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

Is it benign, or malignant?
Benign

How might a cavernous hemangioma affect vision?
 --By **affecting vision**
 --By increasing IOP
 --By causing strabismus

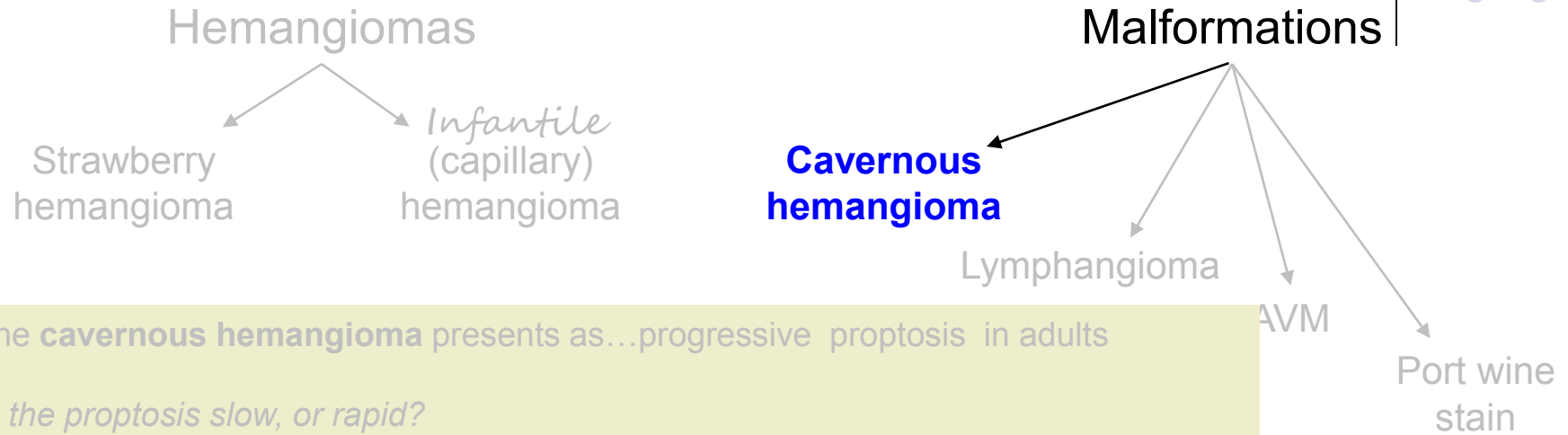
How might a cavernous hemangioma affect vision?
 --By causing retinal striae, or hyperopia
 --By compressing the two words

Does it require surgical excision?
Yes, if it is **clinically significant**

How could a cavernous hemangioma cause striae and/or hyperopia?
By compressing the back of the globe

A

Vascular Lesions of the Orbit



The **cavernous hemangioma** presents as...progressive proptosis in adults

Is the proptosis slow, or rapid?

Slow

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Yes, it is more common in women

Is it benign, or malignant?

Benign

How might a cavernous hemangioma affect vision?

--By **affecting vision**

--By increasing IOP

--By causing strabismus

How could a cavernous hemangioma cause striae and/or hyperopia?

By compressing the back of the globe

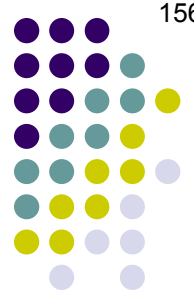
How might a cavernous hemangioma affect vision?

--By **causing retinal striae, or hyperopia**

--By compressing the optic nerve

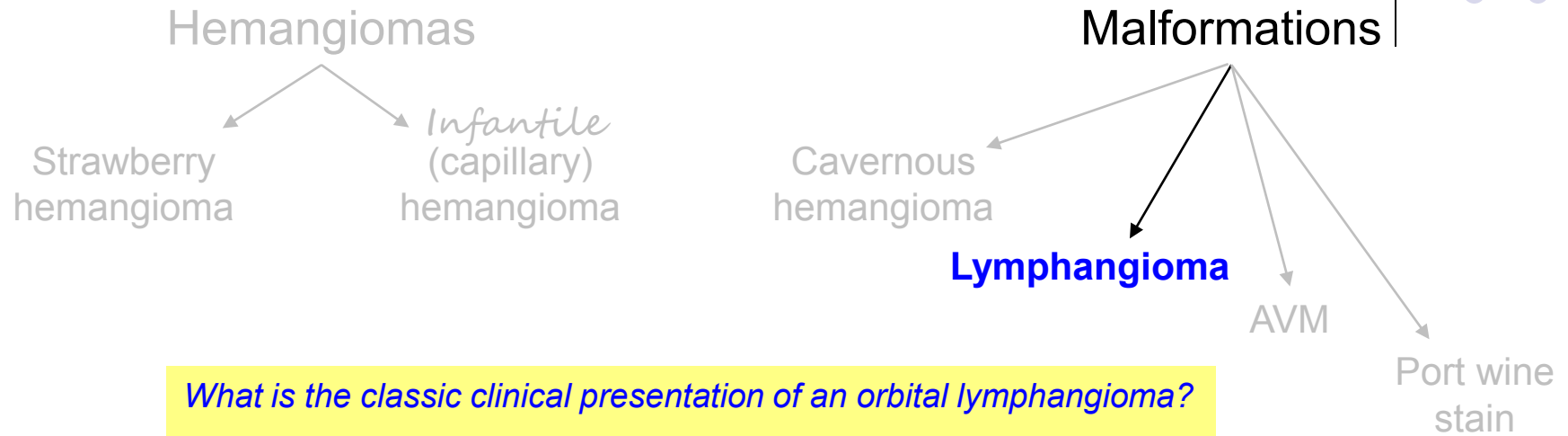
Does it require surgical excision?

Yes, if it is **clinically significant**



Q

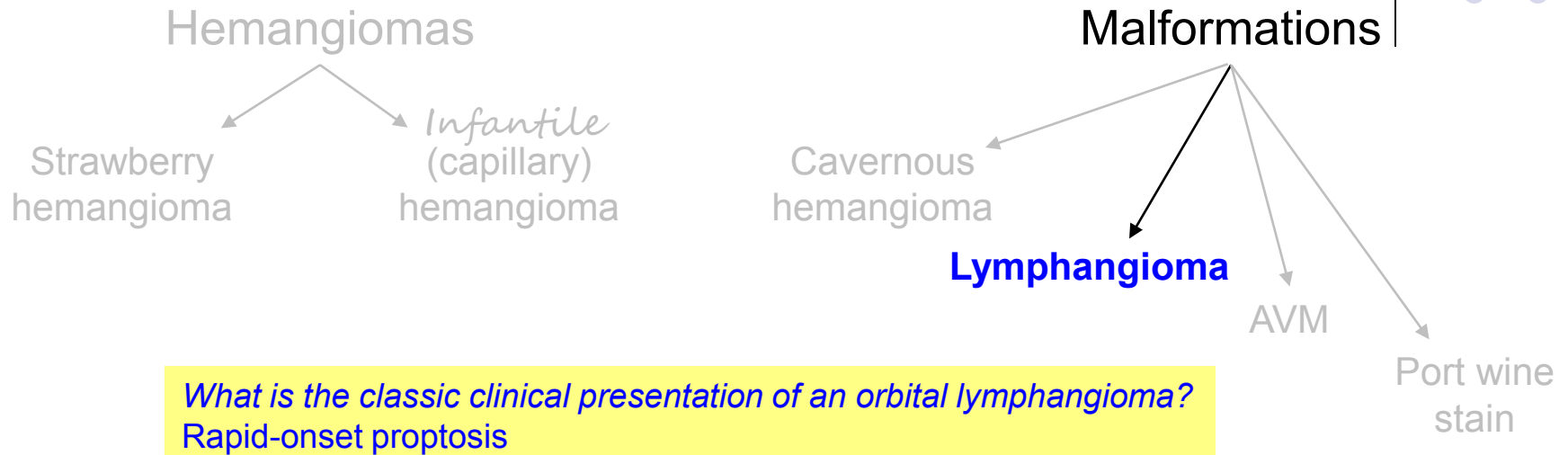
Vascular Lesions of the Orbit



What is the classic clinical presentation of an orbital lymphangioma?

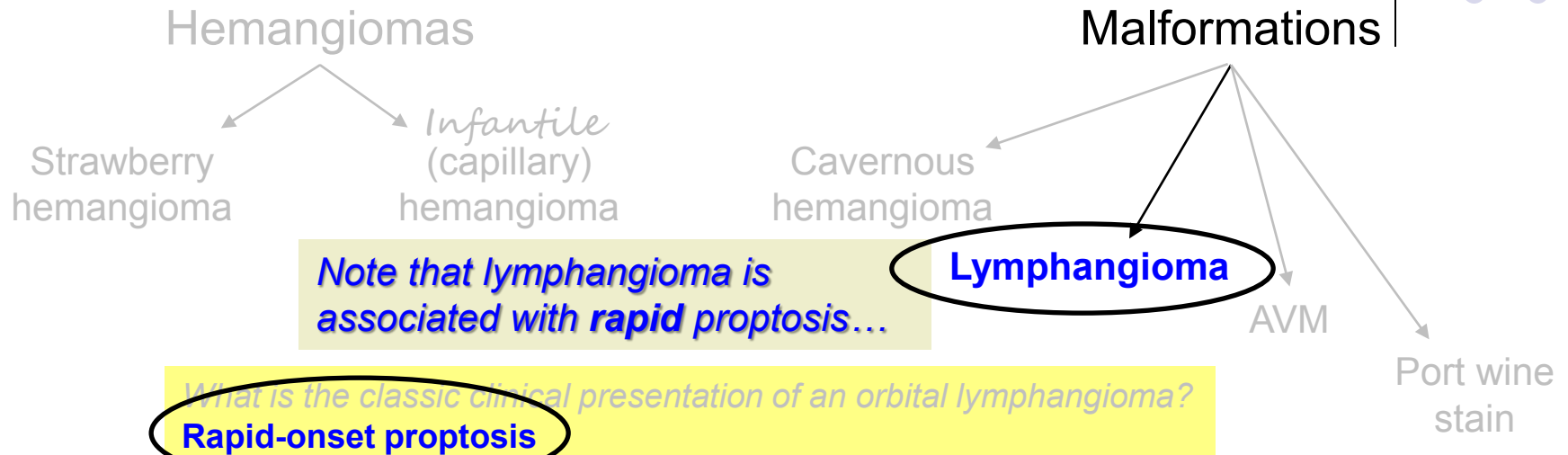
A

Vascular Lesions of the Orbit

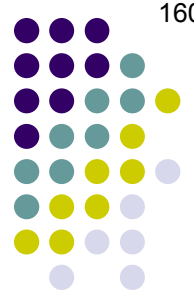


What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

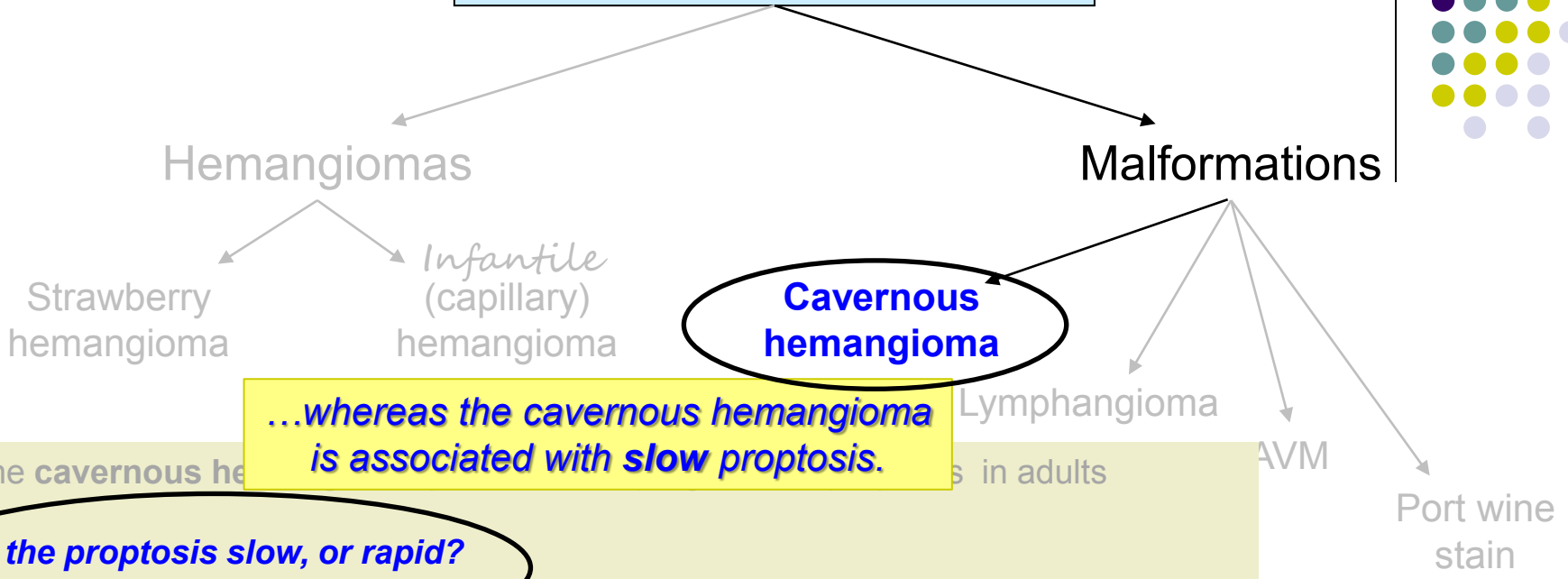
Vascular Lesions of the Orbit



No question on this or the next slide—proceed when ready



Vascular Lesions of the Orbit



...whereas the cavernous hemangioma is associated with slow proptosis.

Is the proptosis slow, or rapid?
Slow

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

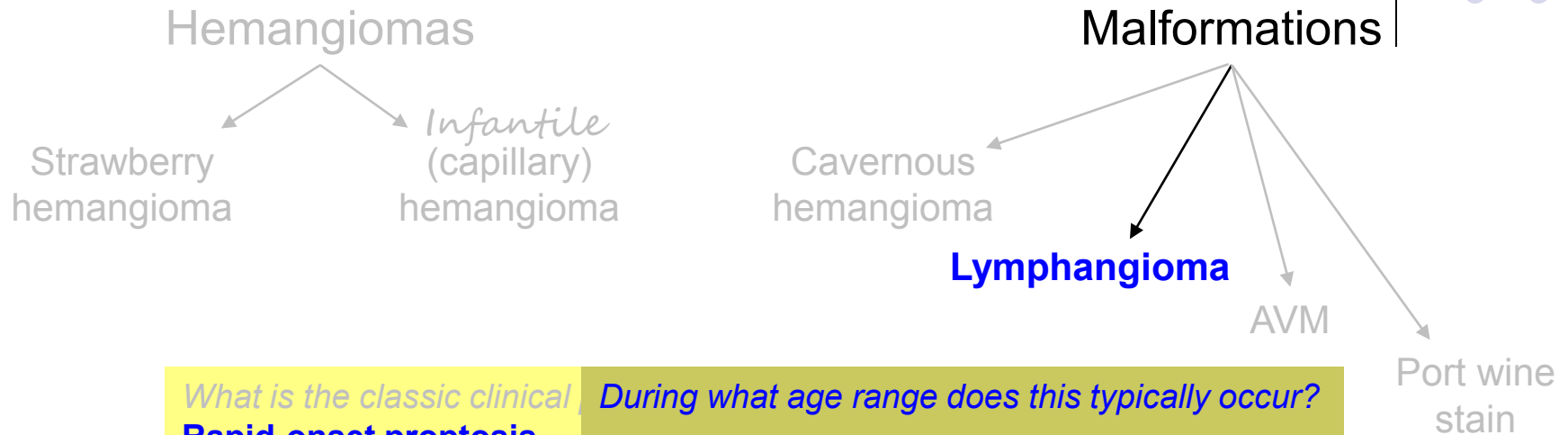
Is it benign, or malignant?
Benign

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

Does it require surgical excision?
Yes, if it is clinically significant

Q

Vascular Lesions of the Orbit



What is the classic clinical **Rapid-onset proptosis** *During what age range does this typically occur?*

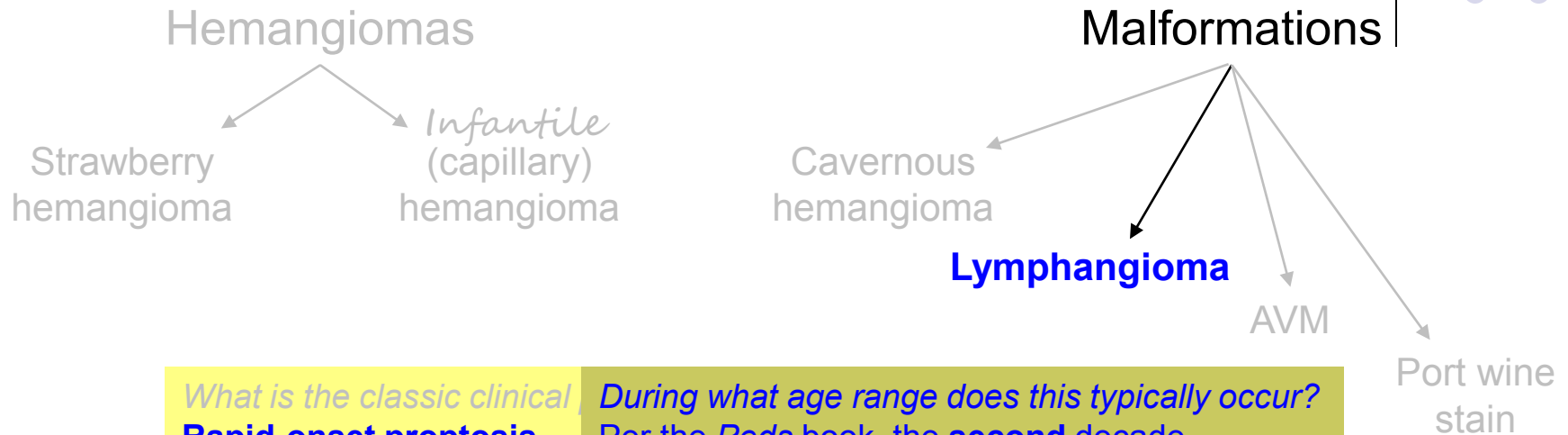
What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

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

- Lymphoid hyperplasia
- Intralesional hemorrhage

A

Vascular Lesions of the Orbit



What is the classic clinical
Rapid-onset proptosis

During what age range does this typically occur?
Per the *Peds* book, the **second** decade
Per the *Orbit* book, the **first** decade

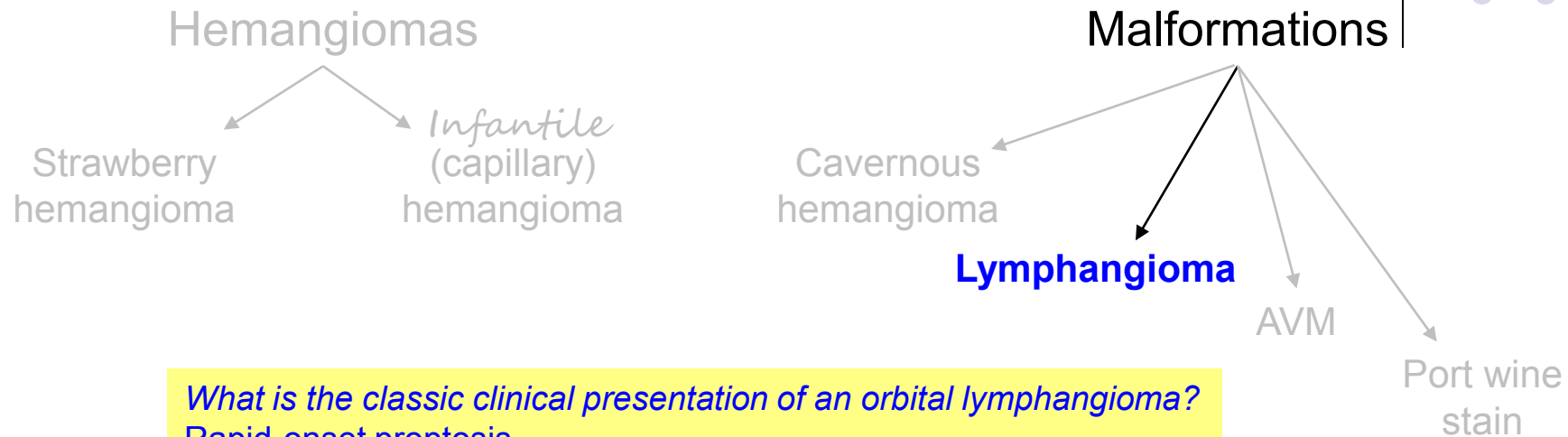
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Vascular Lesions of the Orbit

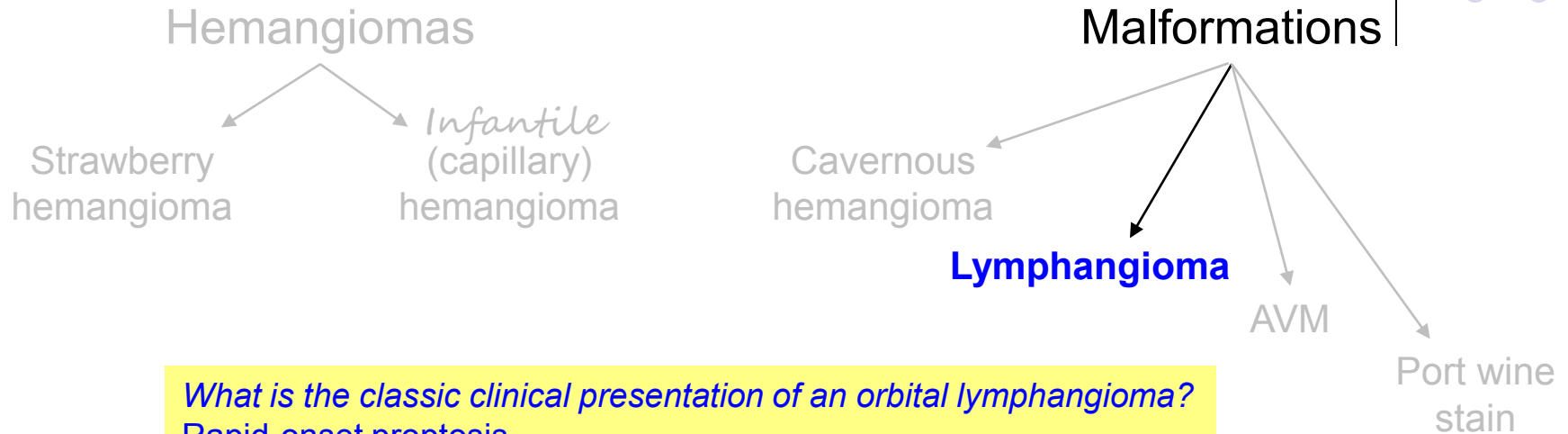


What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?

A

Vascular Lesions of the Orbit

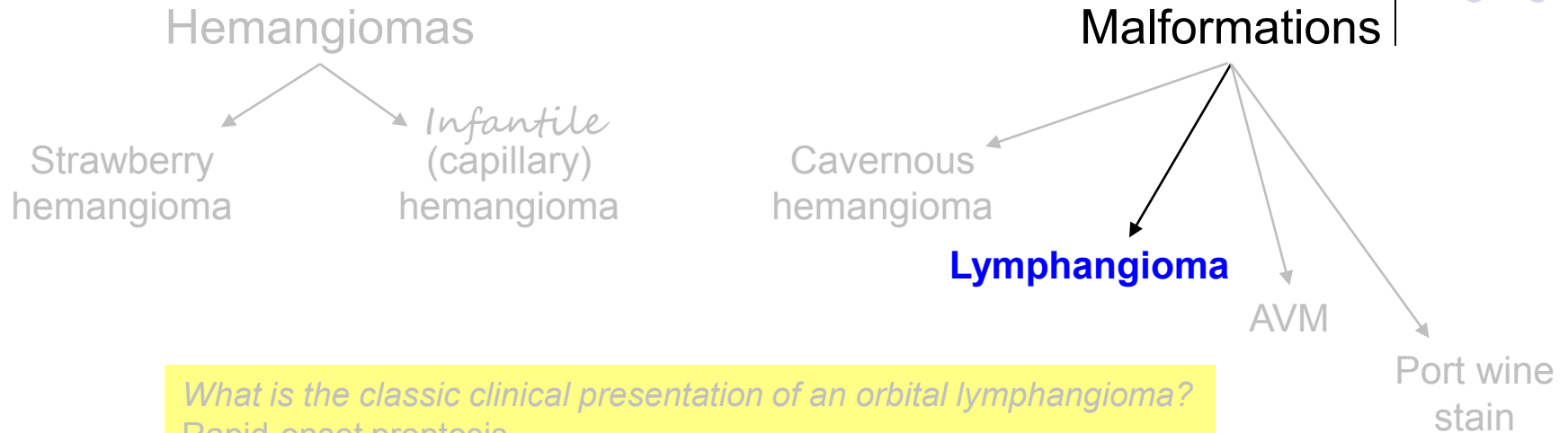


What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

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Q

Vascular Lesions of the Orbit



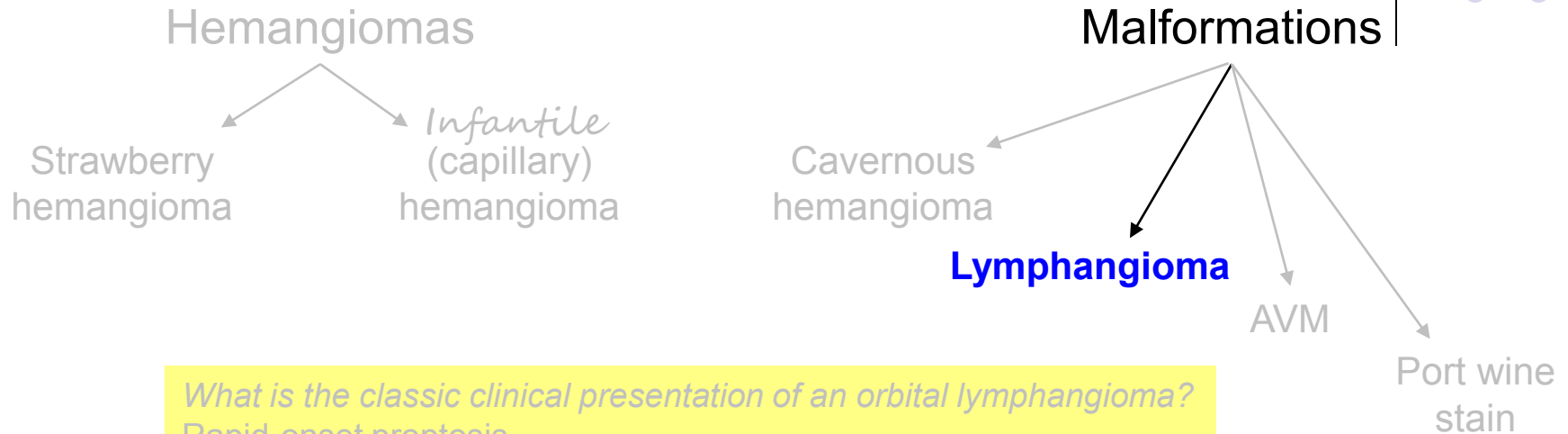
What is the classic clinical presentation of an orbital lymphangioma?
Rapid-onset proptosis

What does histologic examination of a lymphangioma reveal?
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Are lymphangiomas encapsulated?

A

Vascular Lesions of the Orbit



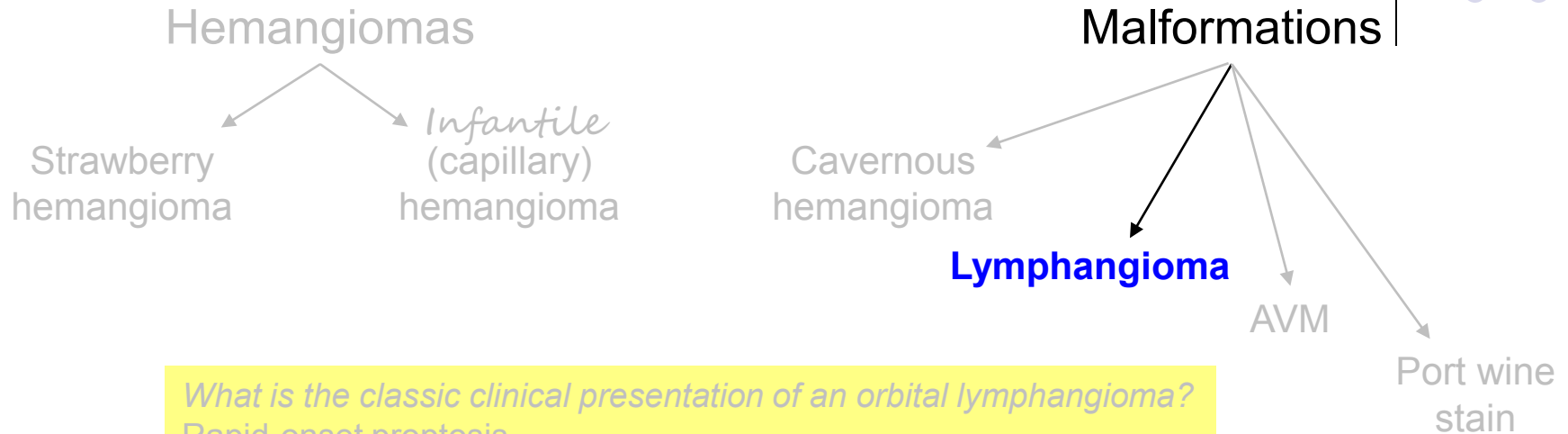
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What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

Are lymphangiomas encapsulated?
No, and this is an important point to remember

Q

Vascular Lesions of the Orbit



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

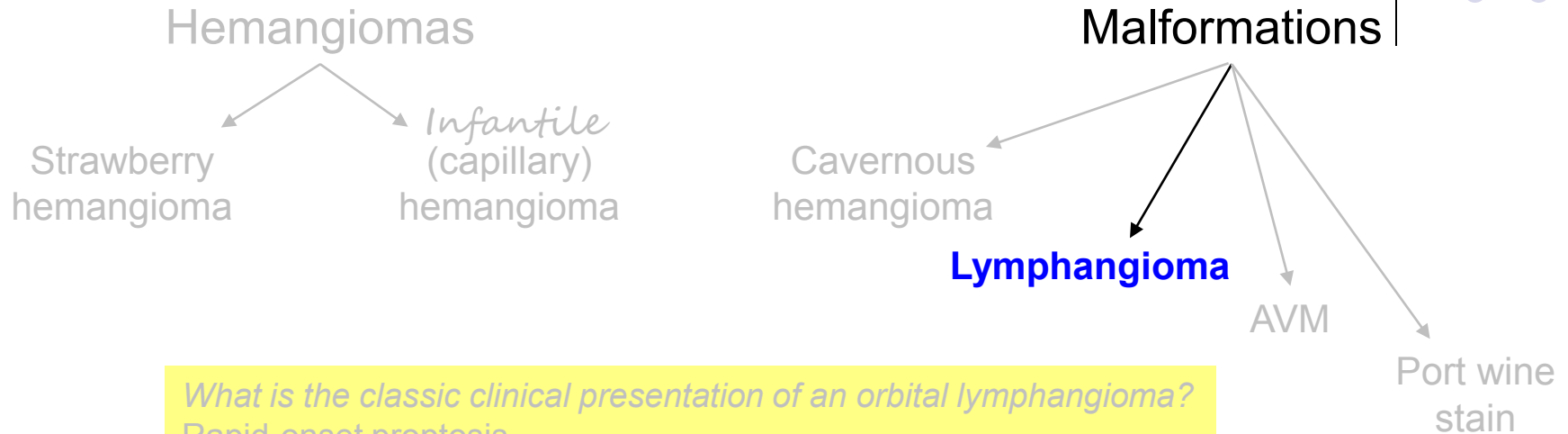
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Are lymphangiomas encapsulated?
No, and this is an important point to remember

Why is it important?

A

Vascular Lesions of the Orbit



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

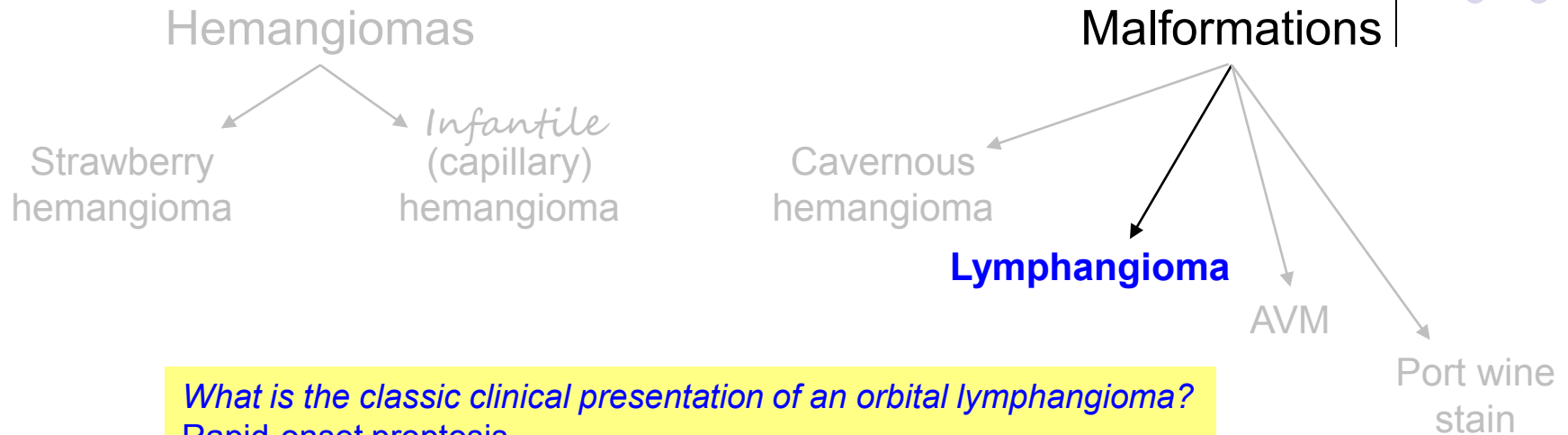
What does histologic examination of a lymphangioma reveal?
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Are lymphangiomas encapsulated?
No, and this is an important point to remember

Why is it important?
Un momento, por favor

Q

Vascular Lesions of the Orbit



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

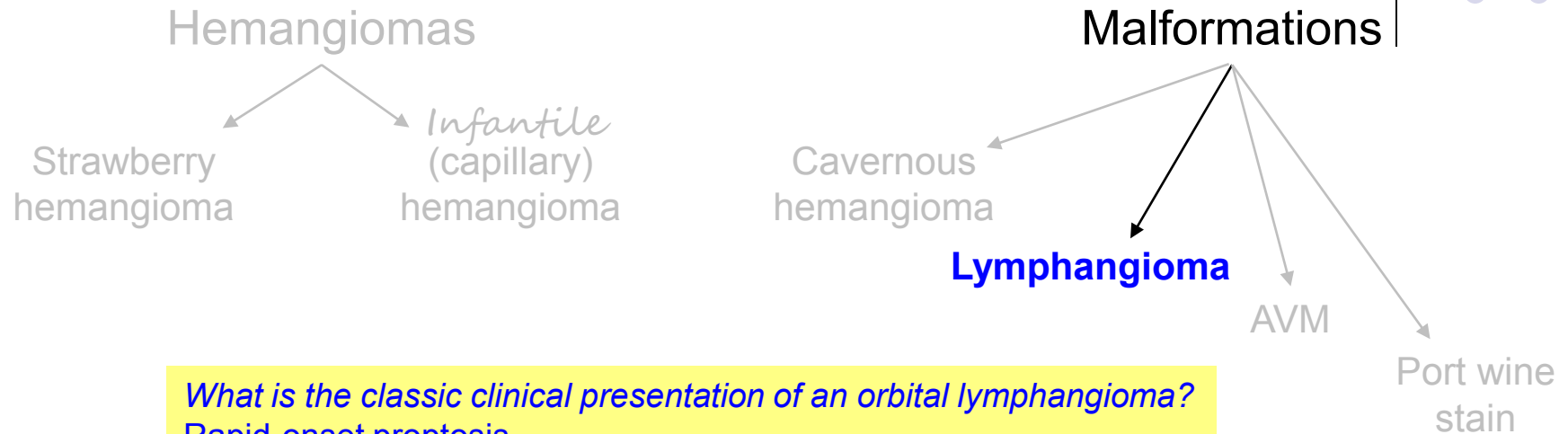
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--?

--?

A

Vascular Lesions of the Orbit



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

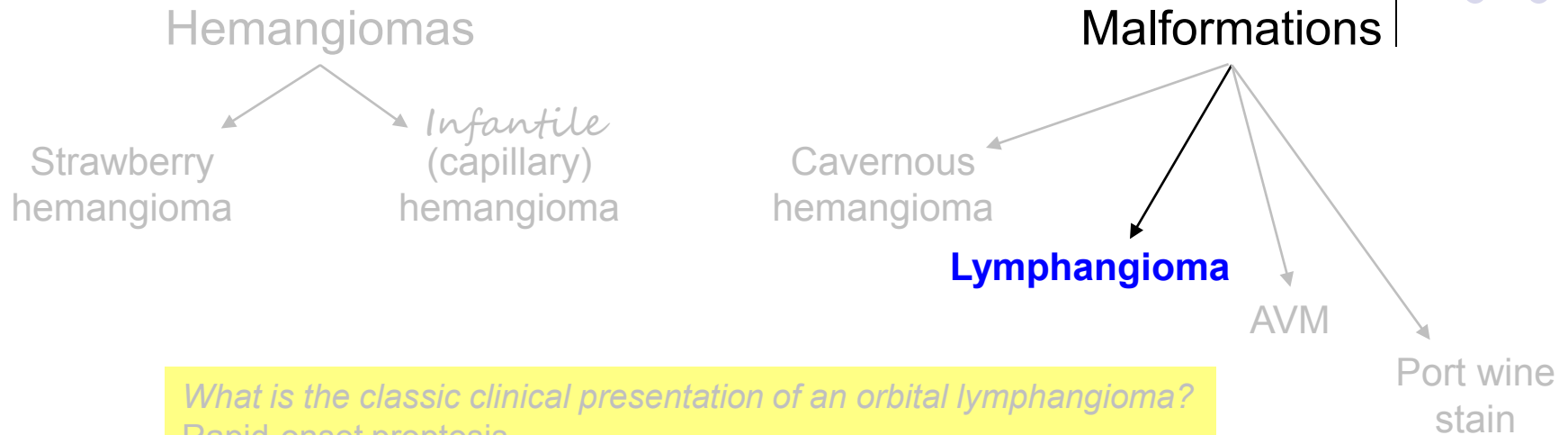
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Q

Vascular Lesions of the Orbit



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Rapid-onset proptosis

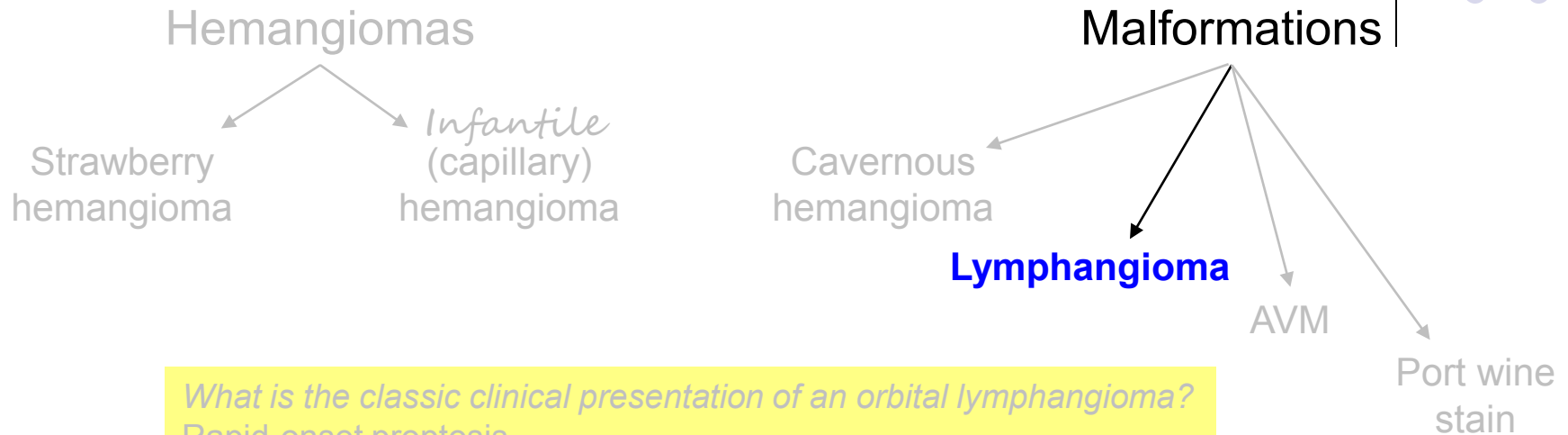
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--Lymphoid hyperplasia
--Intralesional hemorrhage
--Endothelial proliferation?

What about endothelial-cell proliferation? Couldn't that cause lesion enlargement (and thus proptosis)?

A

Vascular Lesions of the Orbit



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

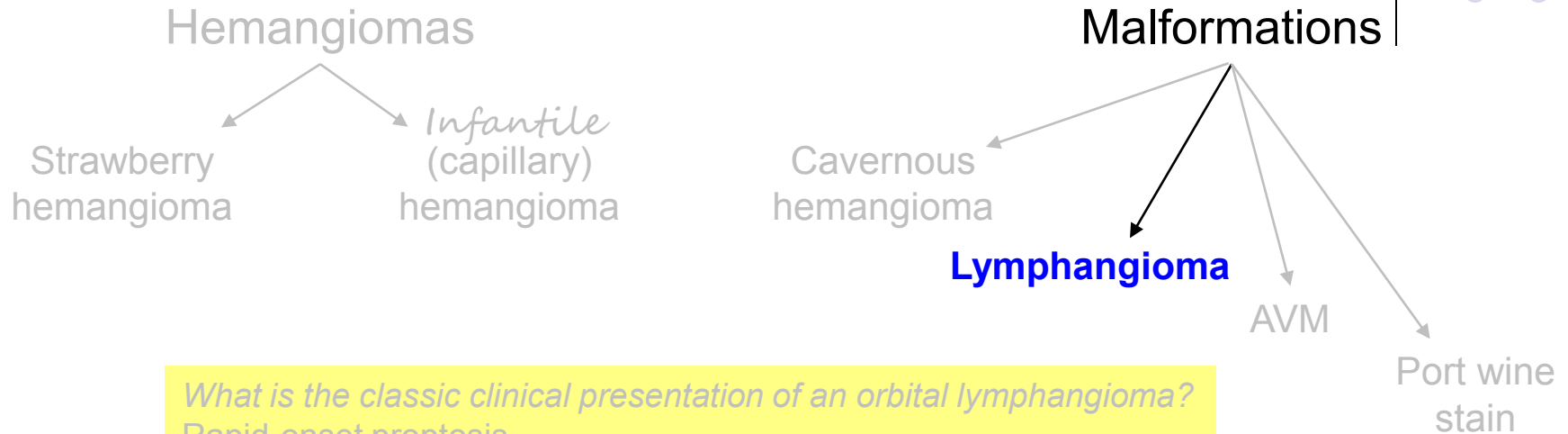
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Two distinct sorts of events can cause that leads to rapid-onset proptosis. What are they?
--Lymphoid hyperplasia
--Intralesional hemorrhage
--Endothelial proliferation? No!

What about endothelial-cell proliferation? Couldn't that cause lesion enlargement (and thus proptosis)?
No, for the simple reason that the endothelial cells of a lymphangioma do not proliferate

Q

Vascular Lesions of the Orbit



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

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

Two distinct sorts of events can cause that leads to rapid-onset proptosis. What are they?
--Lymphoid hyperplasia
--Intralesional hemorrhage
--Endothelial proliferation? *No!*

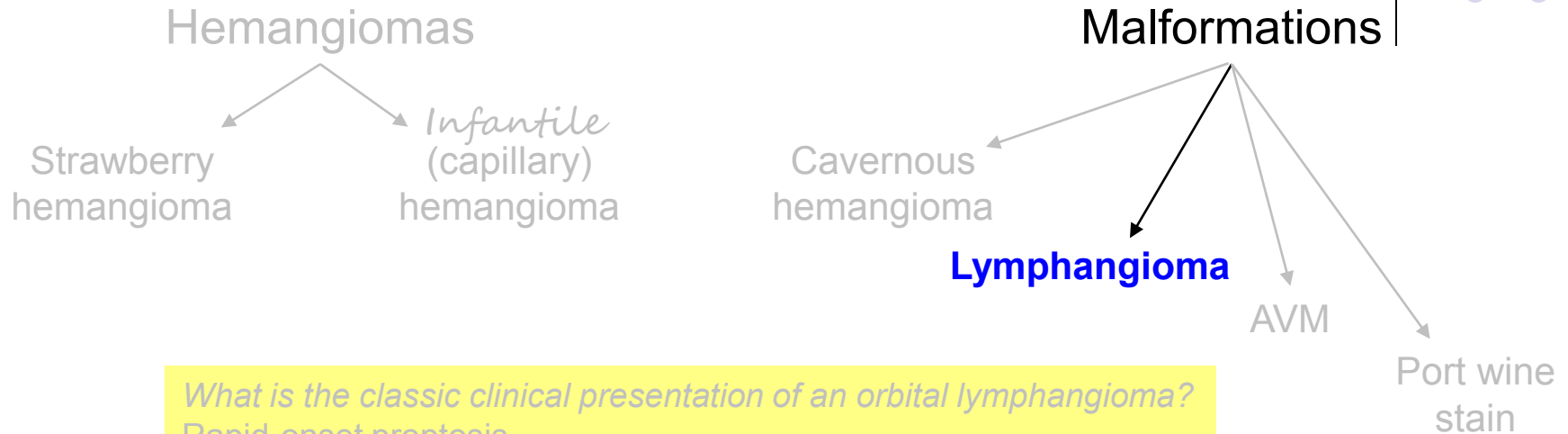
What about endothelial-cell proliferation? Couldn't that cause lesion enlargement (and thus proptosis)?

No, for the simple reason that the endothelial cells of a lymphangioma **do not proliferate**

'The endothelial cells do not proliferate'--what does this fact indicate about the clinical/histologic status of lymphangioma?

A

Vascular Lesions of the Orbit



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

What does histologic examination of a lymphangioma reveal?
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Two distinct sorts of events can cause that leads to rapid-onset proptosis. What are they?
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--Intralesional hemorrhage
--Endothelial proliferation? *No!*

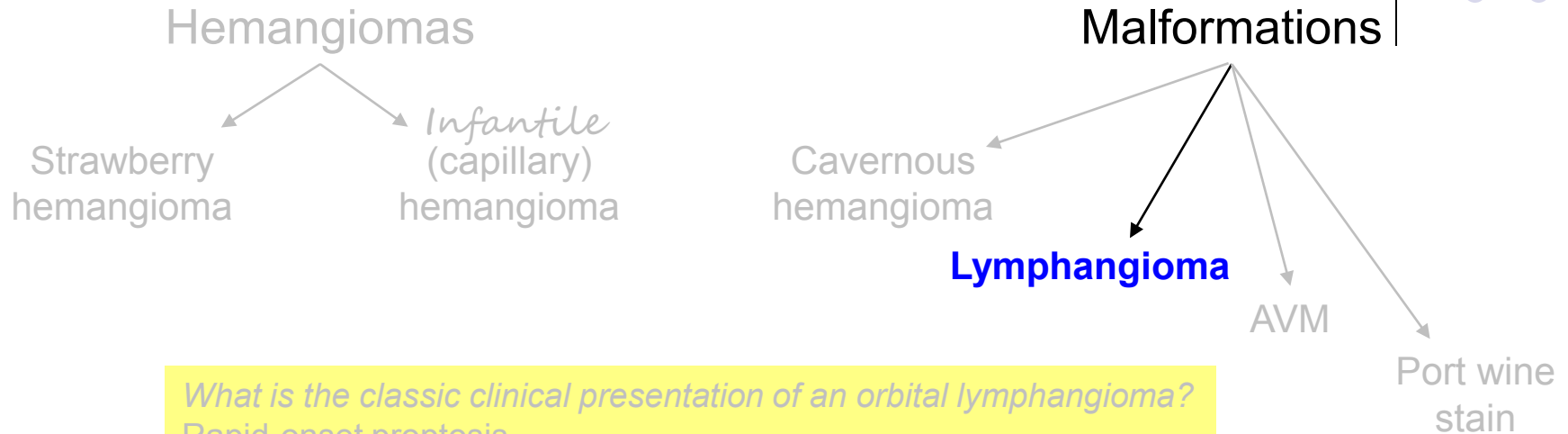
What about endothelial-cell proliferation? Couldn't that cause lesion enlargement (and thus proptosis)?

No, for the simple reason that the endothelial cells of a lymphangioma **do not proliferate**

'The endothelial cells do not proliferate'--what does this fact indicate about the clinical/histologic status of lymphangioma?
It indicates that lymphangiomas are **not** neoplasms

Q

Vascular Lesions of the Orbit



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

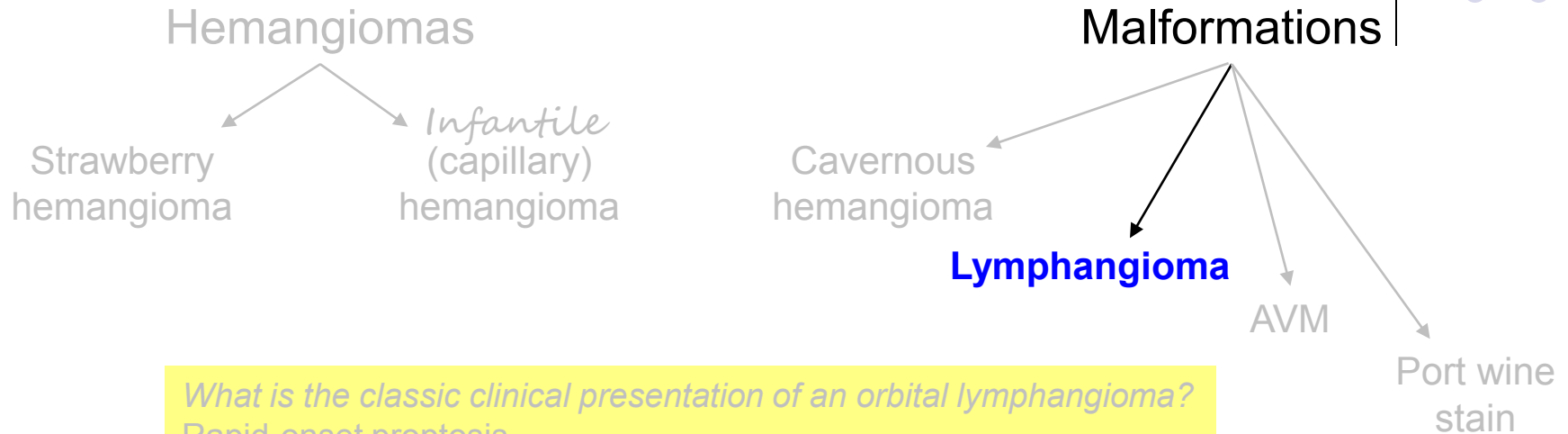
What does histologic examination of a lymphangioma reveal?
Lymphatic-type endothelial cells lining lymph-filled spaces

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

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

A

Vascular Lesions of the Orbit



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

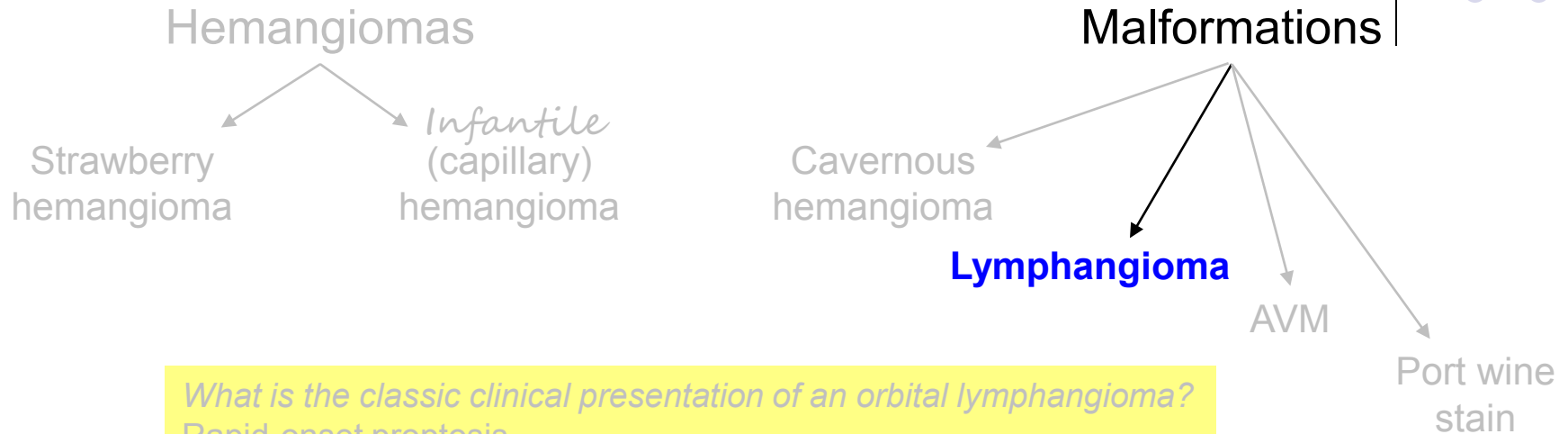
What does histologic examination of a lymphangioma reveal?
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What is the classic health event for precipitating lymphoid hyperplasia in these patients?
Upper respiratory tract infection

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

Q

Vascular Lesions of the Orbit



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

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

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

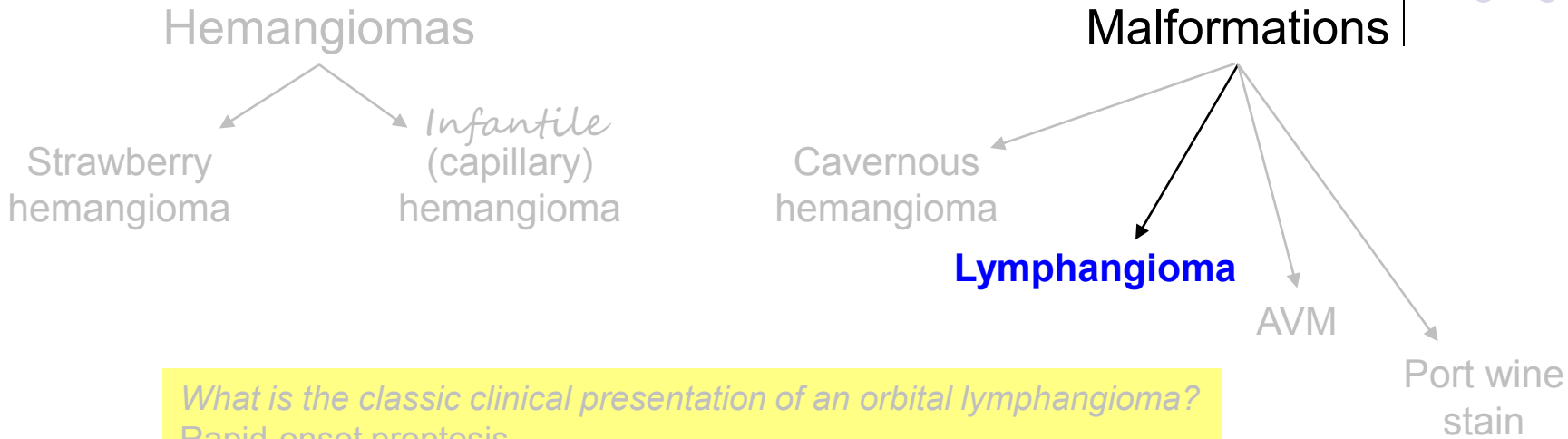
--Lymphoid hyperplasia

--**Intralesional hemorrhage**

What is the name for a lymphangioma in which blood becomes loculated?



Vascular Lesions of the Orbit



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

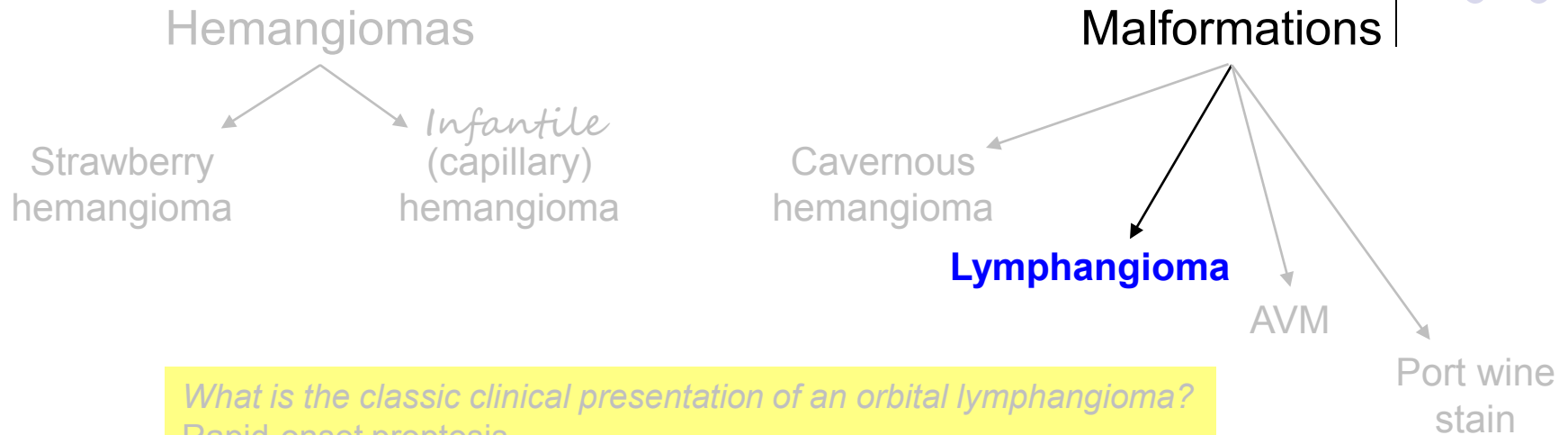
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--Lymphoid hyperplasia
--Intralesional hemorrhage

What is the name for a lymphangioma in which blood becomes loculated?
A **hemorrhagic** cyst

A

Vascular Lesions of the Orbit



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

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

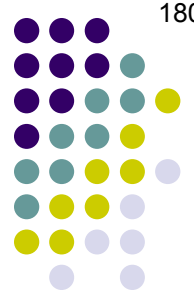
Two distinct sorts of events can cause the rapid lesion enlargement that leads to rapid-onset proptosis. What are these events?

--Lymphoid hyperplasia

--**Intralesional hemorrhage**

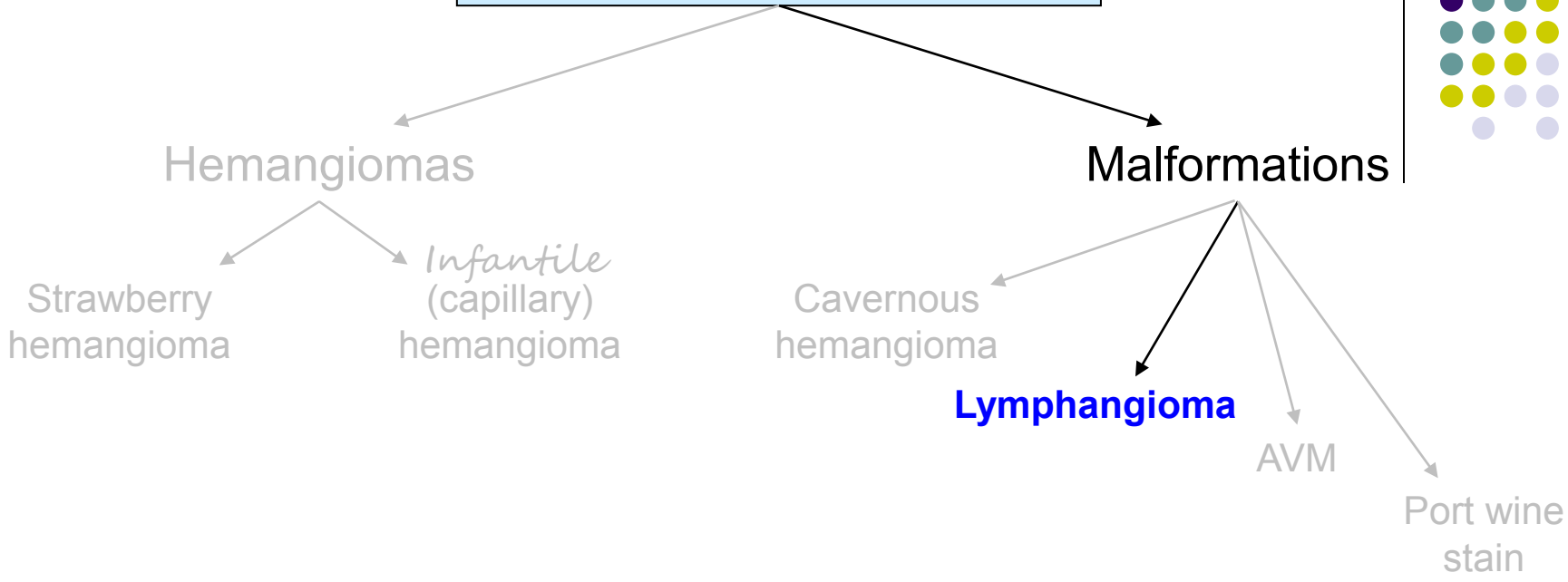
What is the name for a lymphangioma in which blood becomes loculated?

A **chocolate** cyst

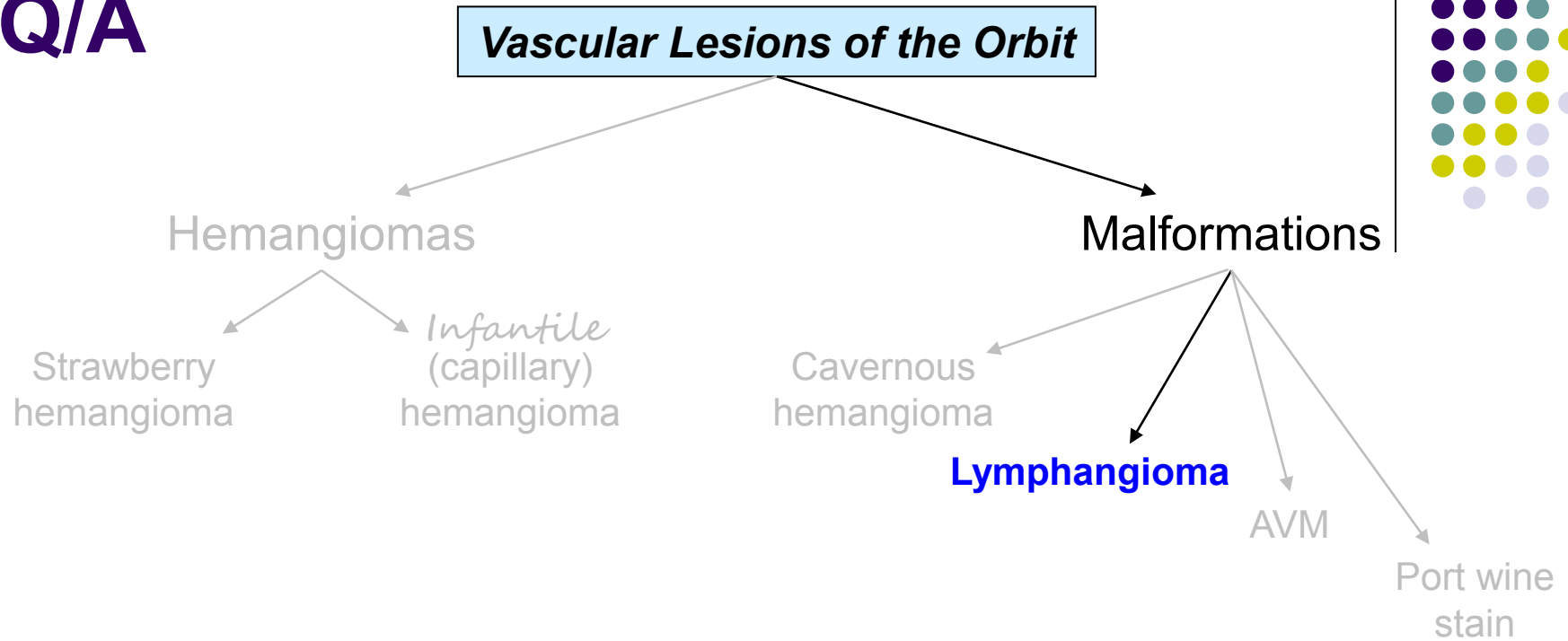


Q

Vascular Lesions of the Orbit



How are lymphangiomas managed?

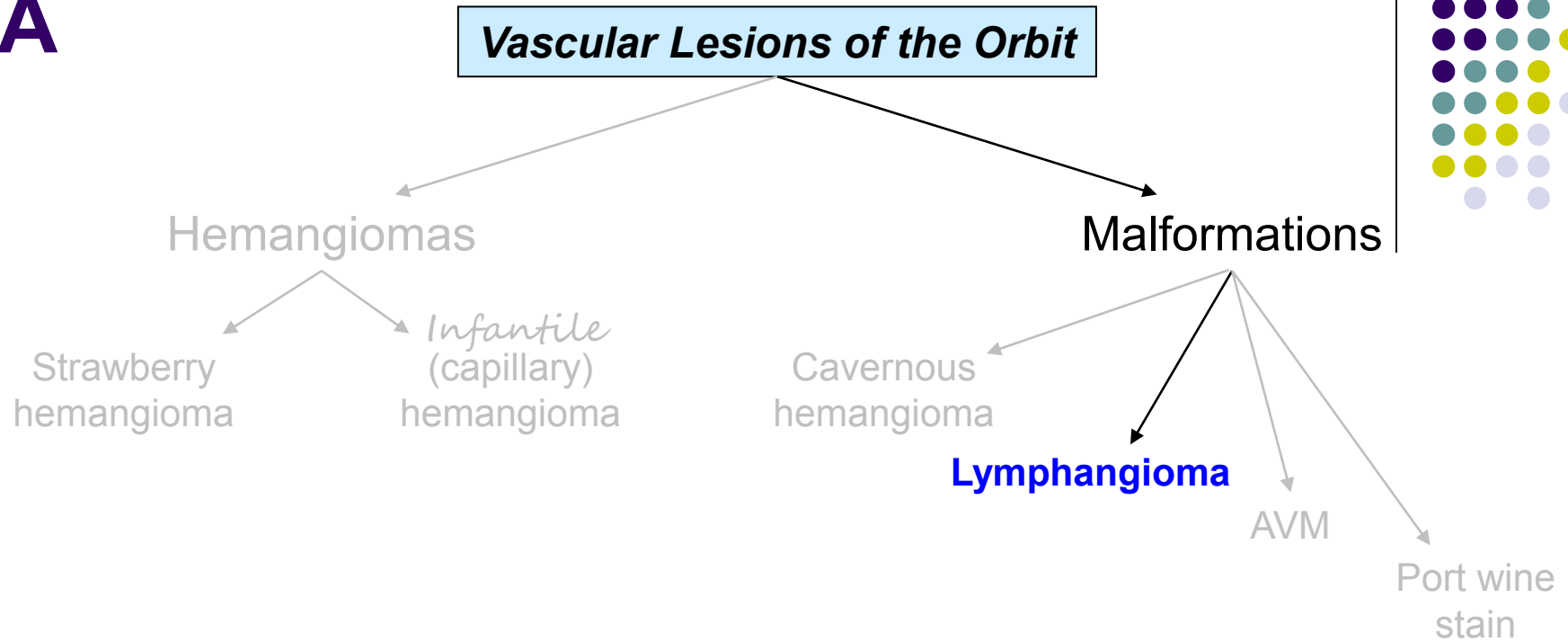


How are lymphangiomas managed?

If threatening vision or producing unacceptable cosmesis— .

Otherwise, conservatively.

A



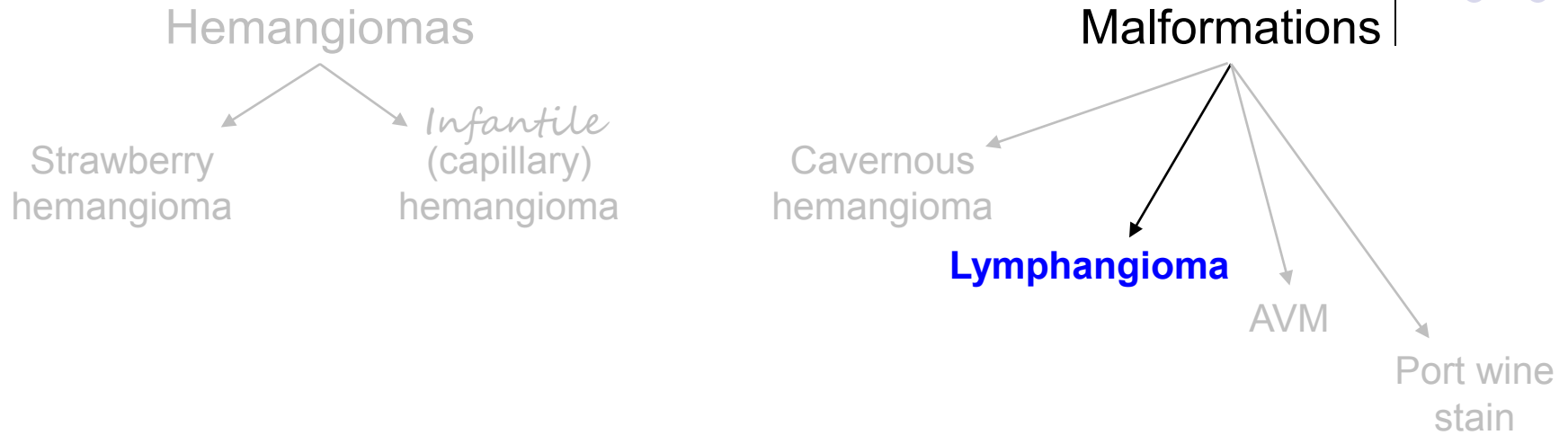
How are lymphangiomas managed?

If threatening vision or producing unacceptable cosmesis—resection .

Otherwise, conservatively.

Q

Vascular Lesions of the Orbit



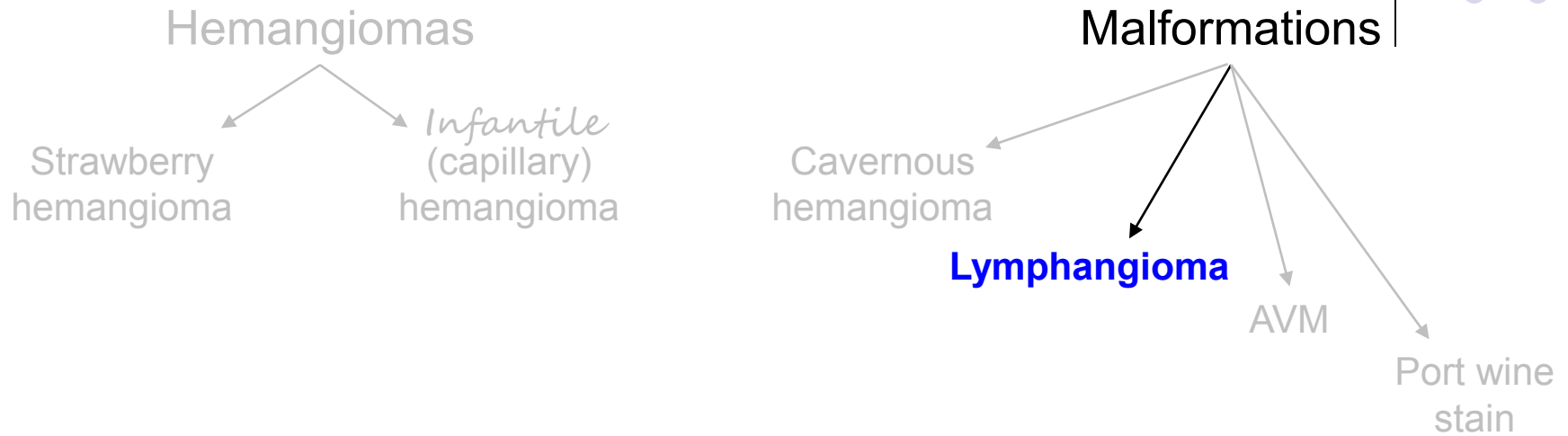
How are lymphangiomas managed?

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Is complete excision of the lesion the surgical goal?

A

Vascular Lesions of the Orbit



How are lymphangiomas managed?

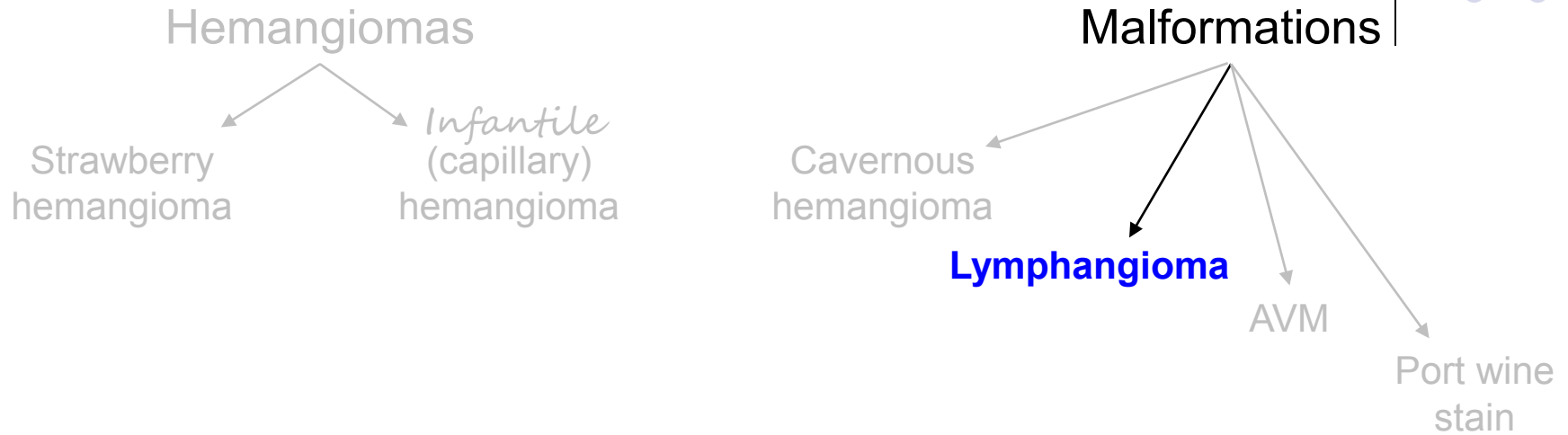
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Is complete excision of the lesion the surgical goal?

No, that would be an impossible task

Q

Vascular Lesions of the Orbit



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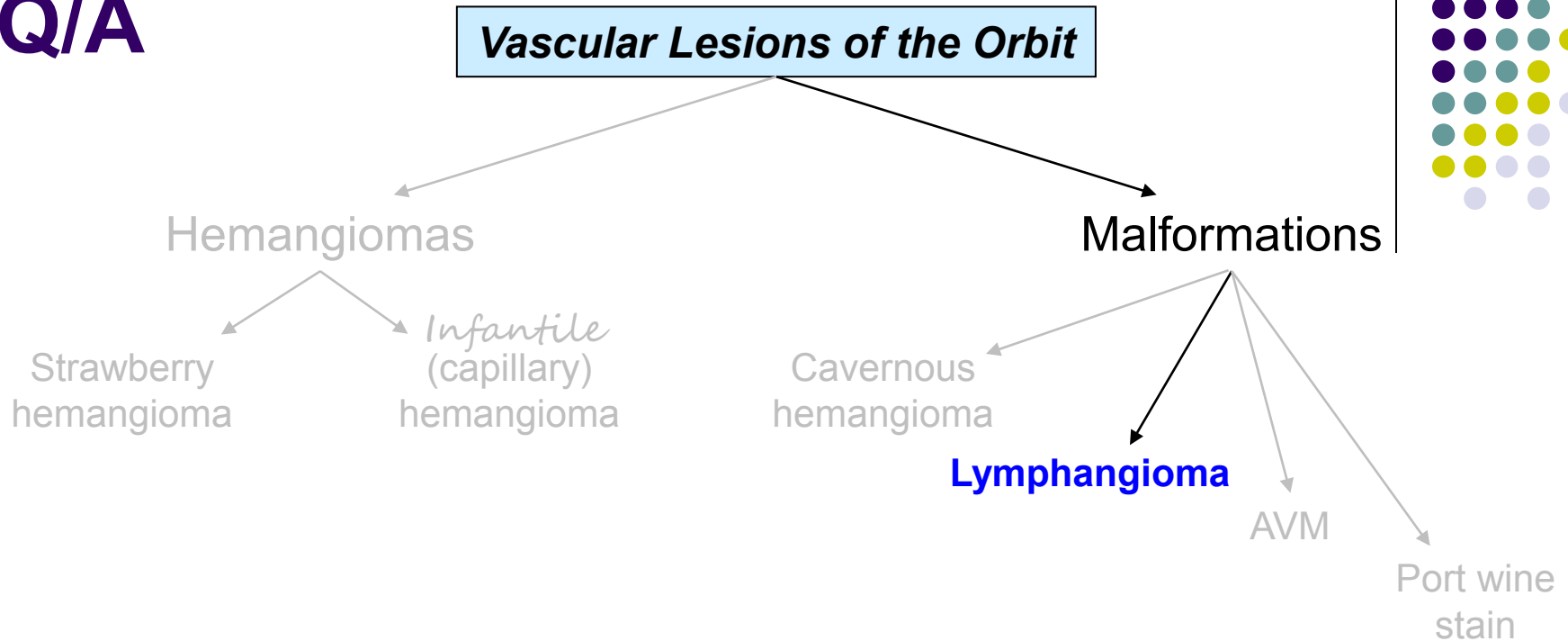
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Why is excision impossible?

Q/A



How are lymphangiomas managed?

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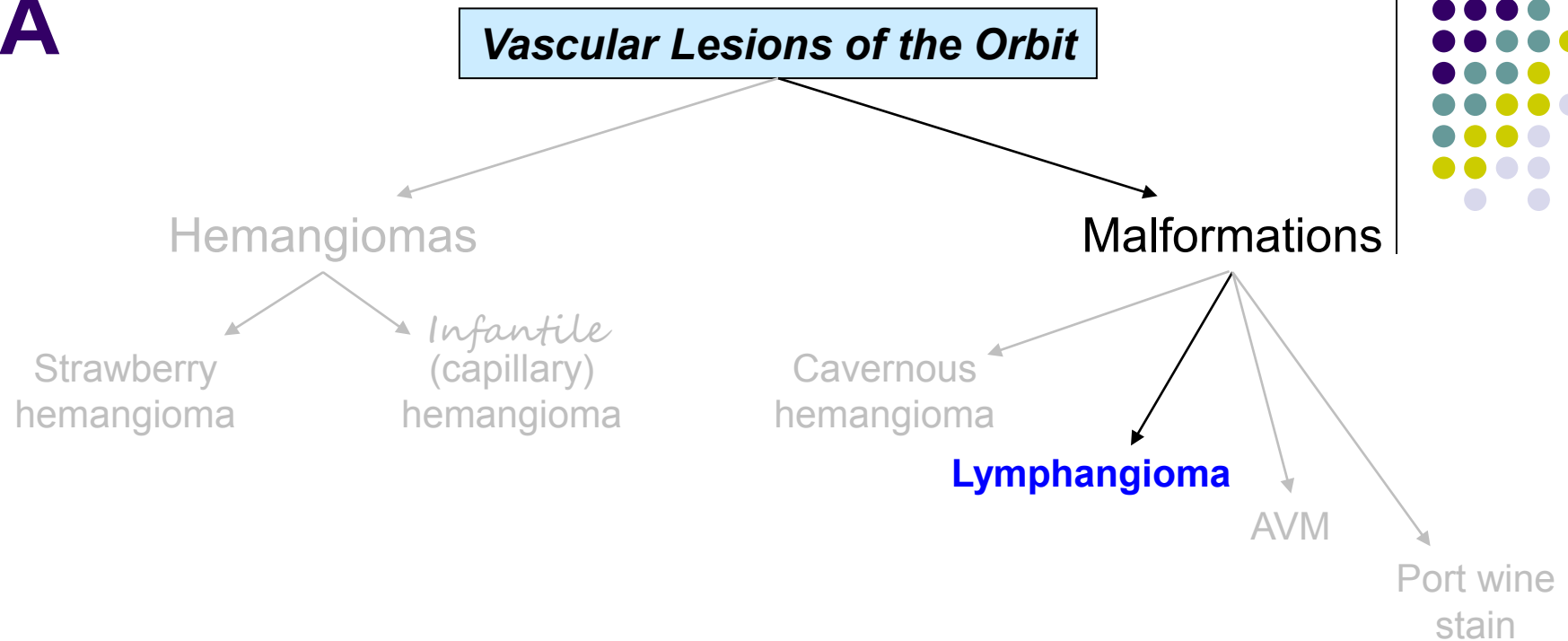
No, that would be an impossible task

Why is excision impossible?

Because lymphangiomas are not encapsulated, ..

(at last we'll see why this fact is important)

A



How are lymphangiomas managed?

If threatening vision or producing unacceptable cosmesis — **resection**
 Otherwise, conservatively.

Is complete excision of the lesion the surgical goal?

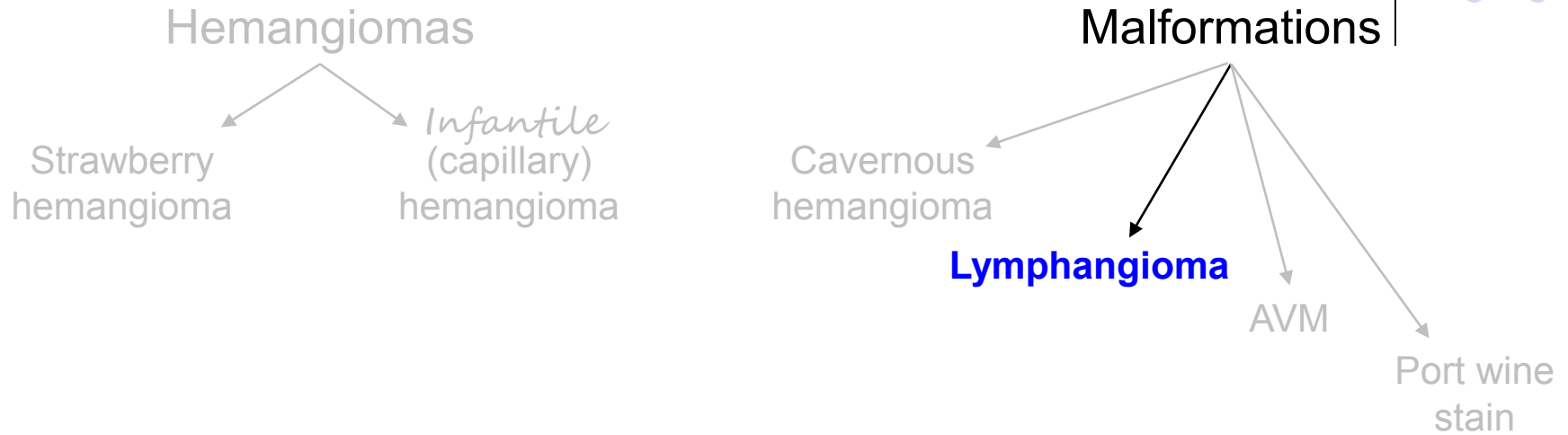
No, that would be an impossible task

Why is excision impossible?

Because lymphangiomas are not encapsulated...they tend to be highly infiltrative, and this renders them essentially un-exciseable

Q

Vascular Lesions of the Orbit

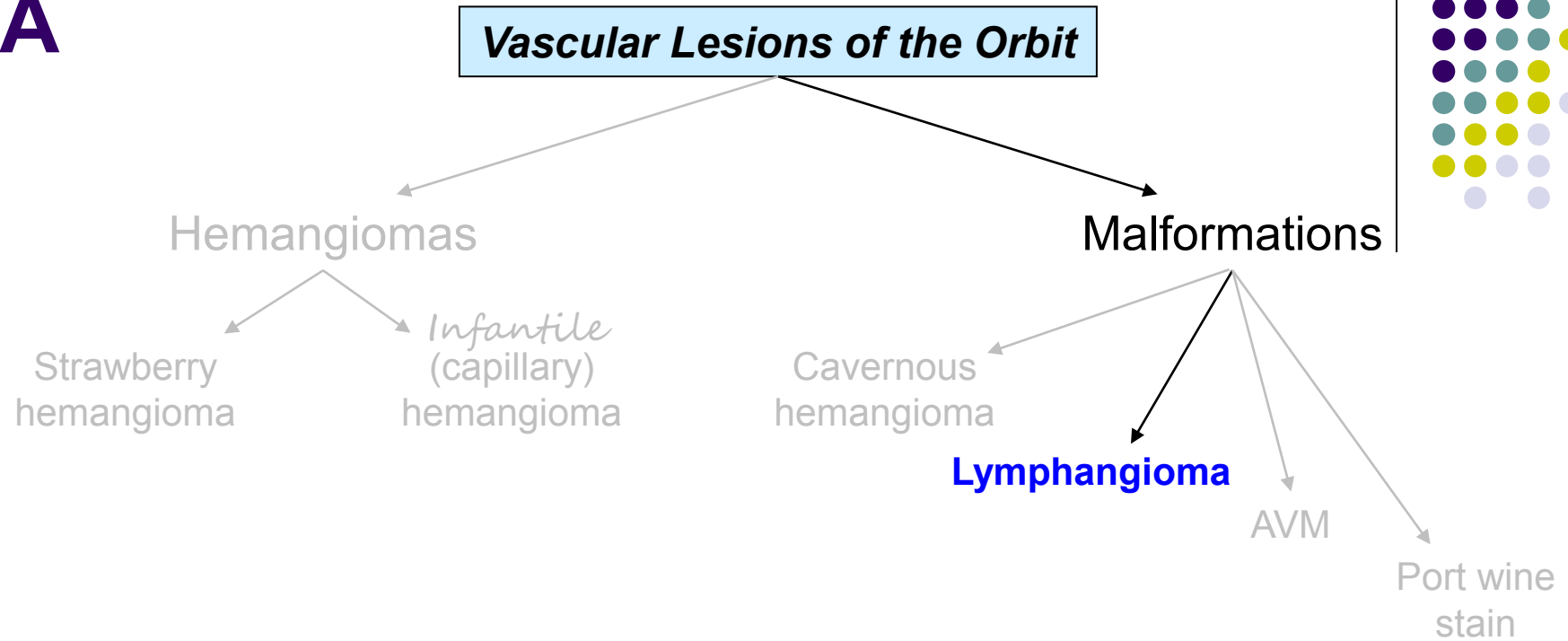


How are lymphangiomas managed?

If threatening vision or producing unacceptable cosmesis—~~resection~~.
Otherwise, conservatively.

Is there a less-drastic surgical intervention that can be tried first?

A



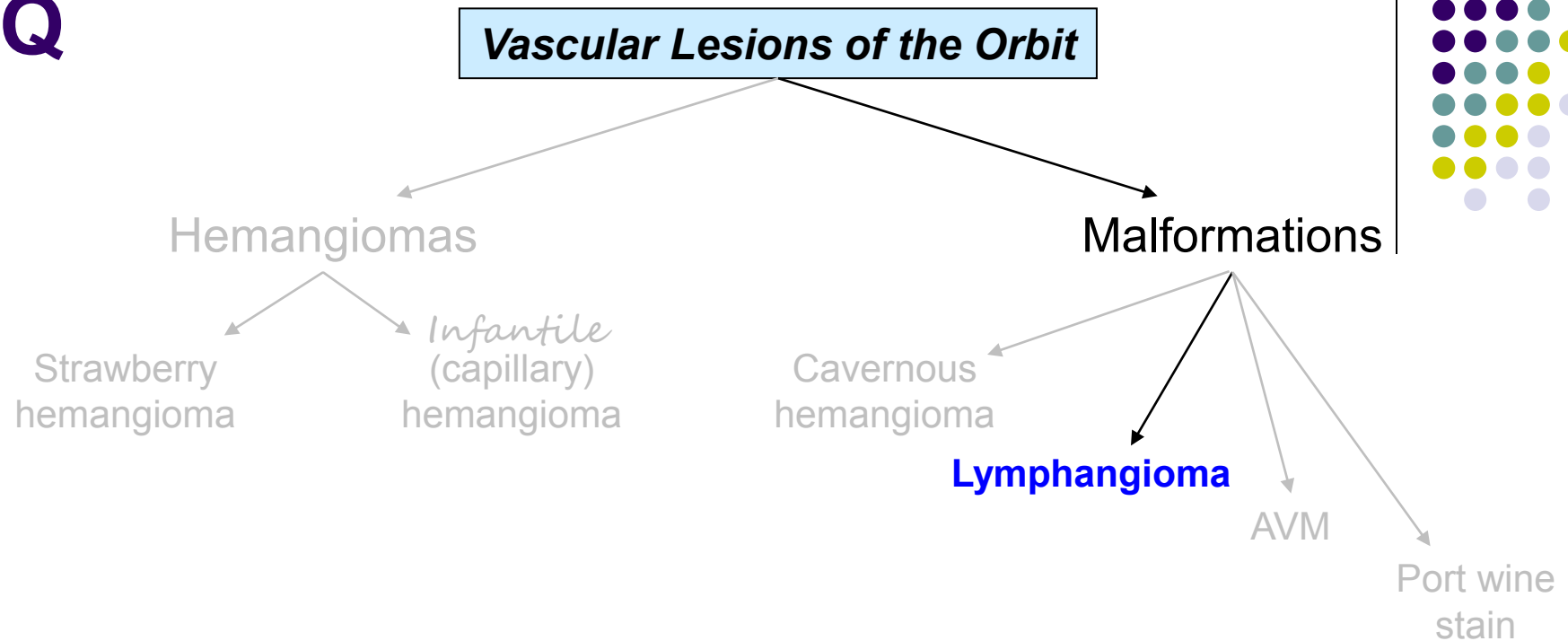
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Yes, a sclerosing agent can be injected into the lesion

Q



How are lymphangiomas managed?

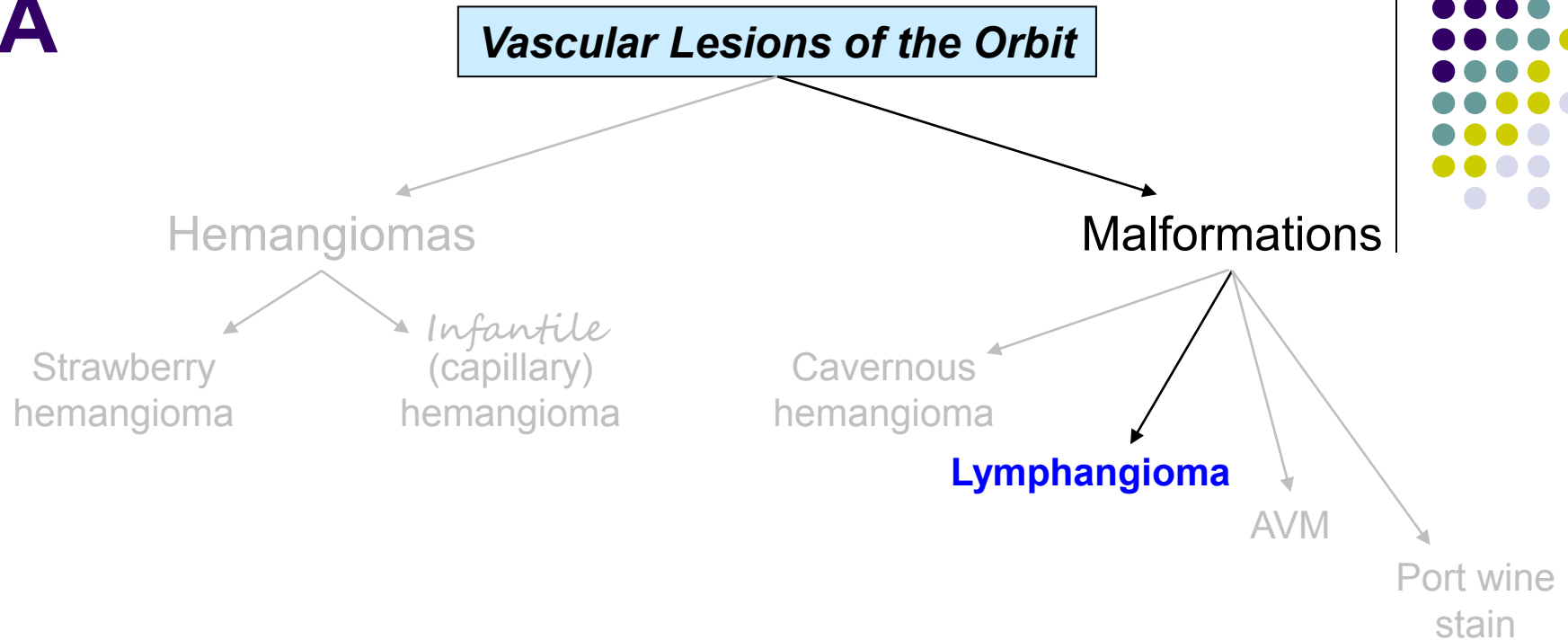
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What potentially devastating complication can arise from this injection?

A



How are lymphangiomas managed?

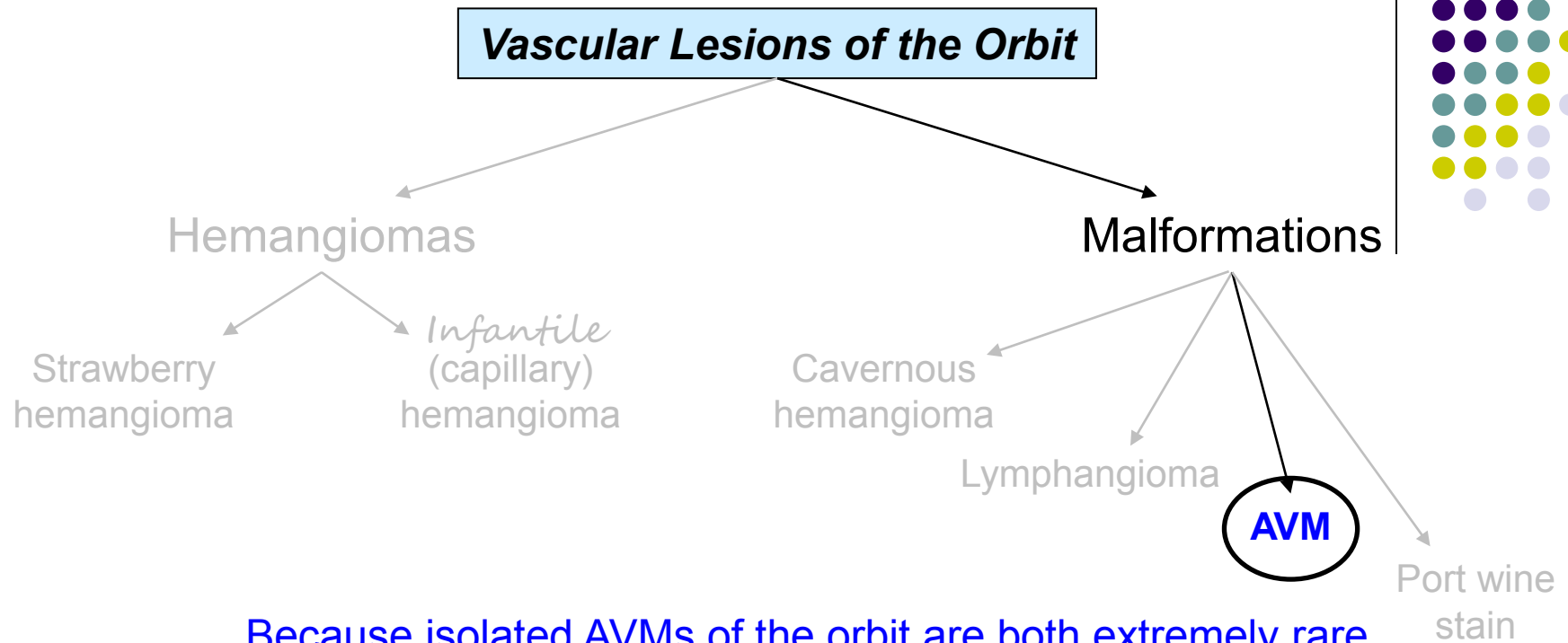
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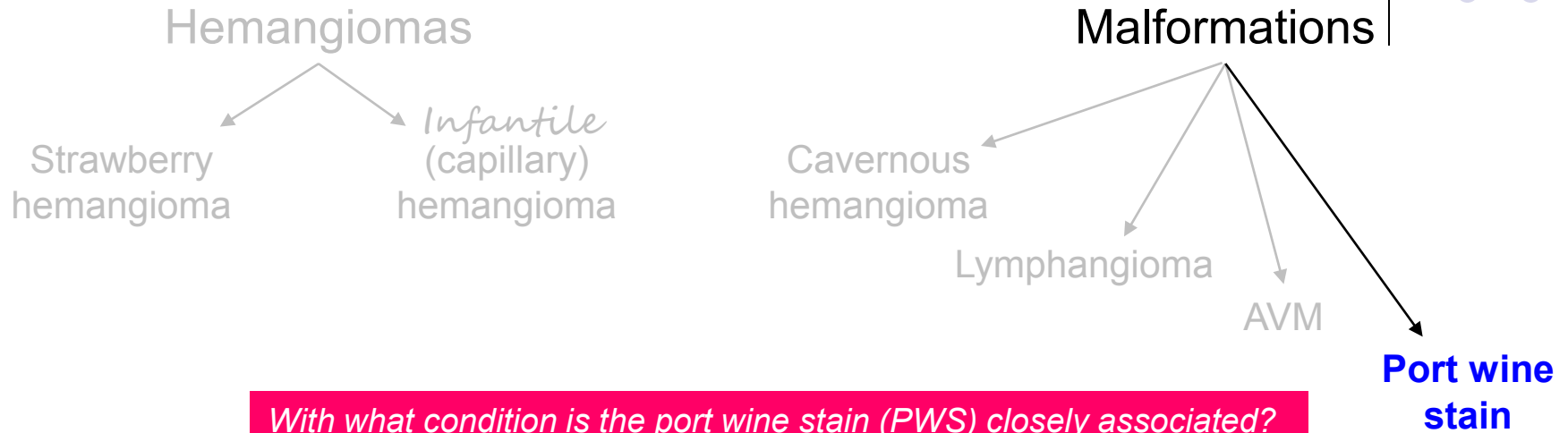
Sclerosis of the optic nerve leading to an iatrogenic optic neuropathy



Because isolated AVMs of the orbit are both extremely rare and only lightly covered in the *BCSC* books, they do not warrant further attention here

Q

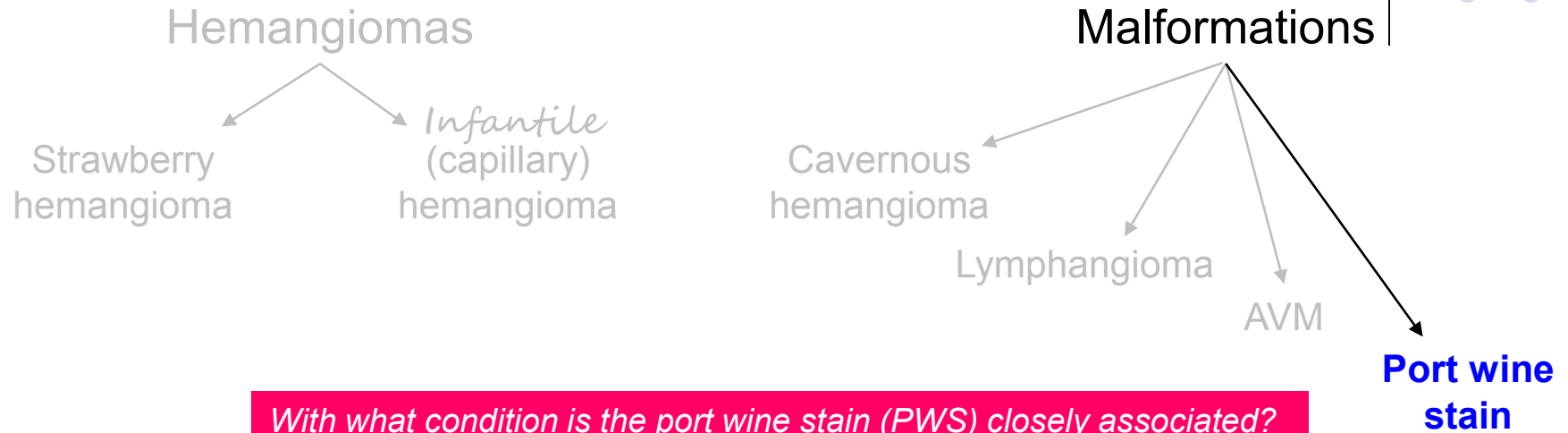
Vascular Lesions of the Orbit



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

A

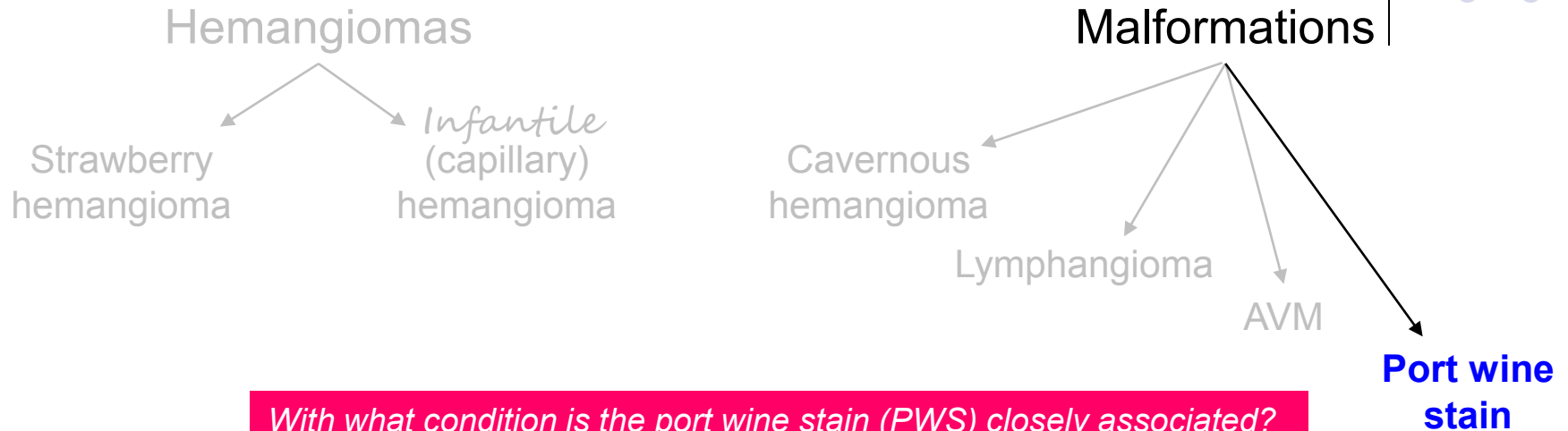
Vascular Lesions of the Orbit



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

Q

Vascular Lesions of the Orbit

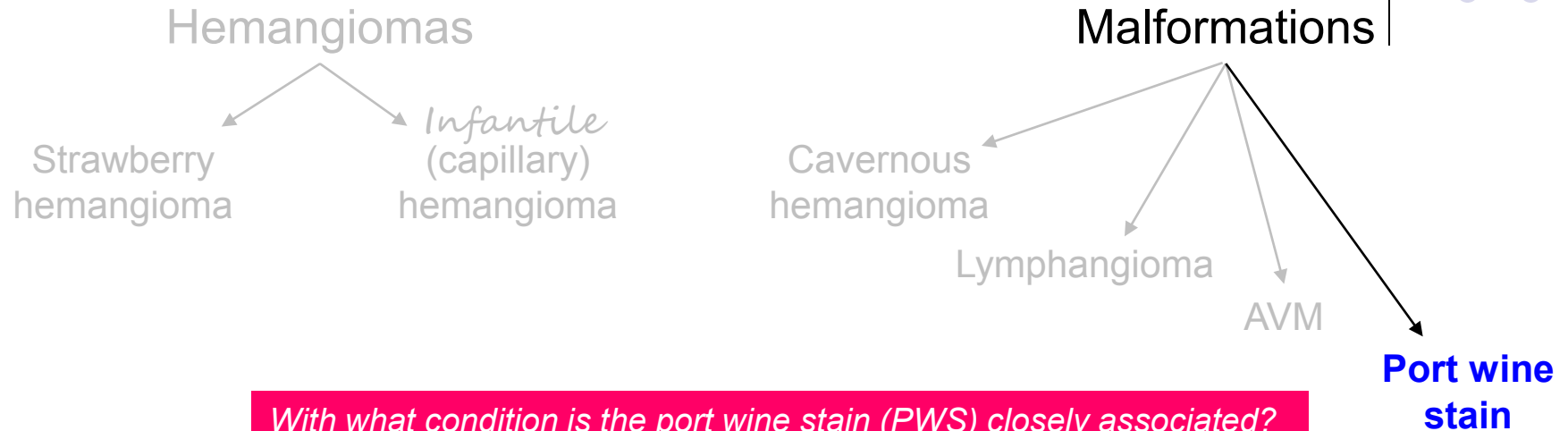


*With what condition is the port wine stain (PWS) closely associated?
Sturge-Weber syndrome*

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

A

Vascular Lesions of the Orbit



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

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

Q

Vascular Lesions of the Orbit



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Briefly, what is a phakomatosis?

Q/A

Vascular Lesions of the Orbit



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Briefly, what is a phakomatosis?

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

A

Vascular Lesions of the Orbit



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Vascular Lesions of the Orbit



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

A

Vascular Lesions of the Orbit



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As **neuro...**

A

Vascular Lesions of the Orbit



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As **neuro-oculo...**

A

Vascular Lesions of the Orbit



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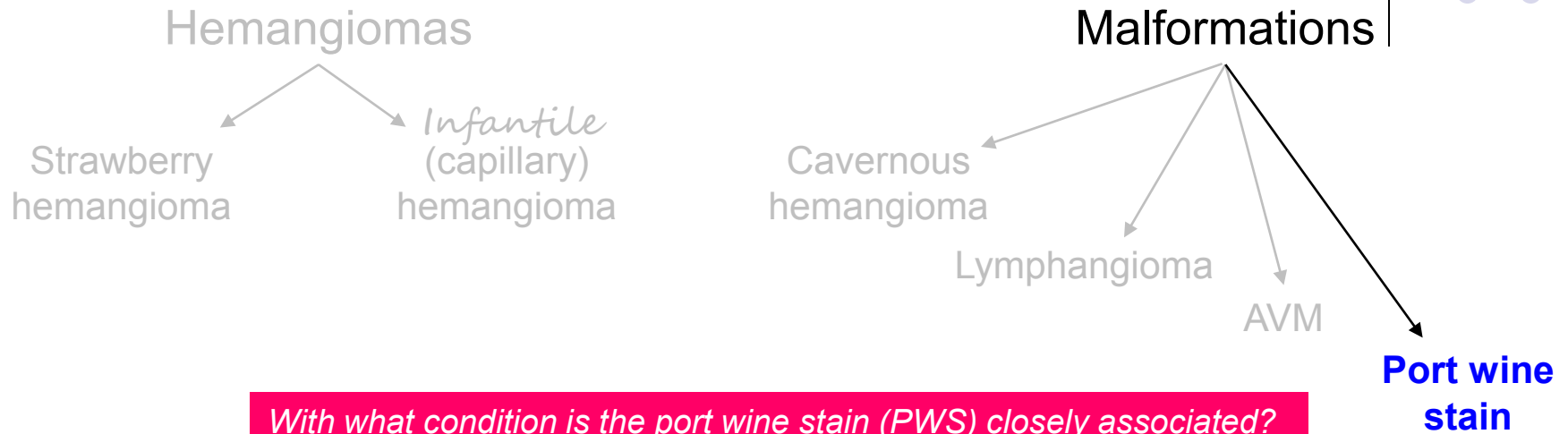
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the **CNS**, **eyes** and **skin**

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

As **neuro-oculocutaneous** syndromes

Q

Vascular Lesions of the Orbit



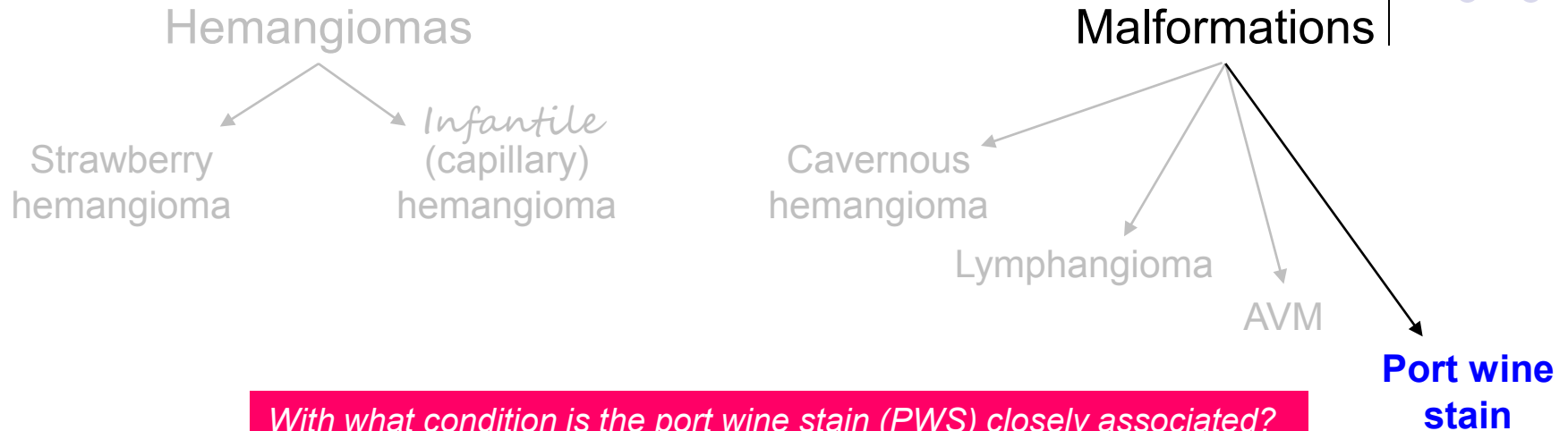
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Sturge-Weber syndrome*

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

By what noneponymous name is Sturge-Weber known?

A

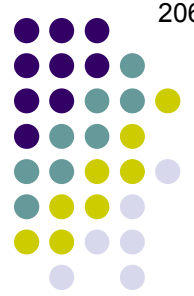
Vascular Lesions of the Orbit



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Encephalotrigeminal angiomatosis



Q

Vascular Lesions of the Orbit

Hemangiomas

Malformations

Strawberry hemangioma

Infantile (capillary) hemangioma

Cavernous hemangioma

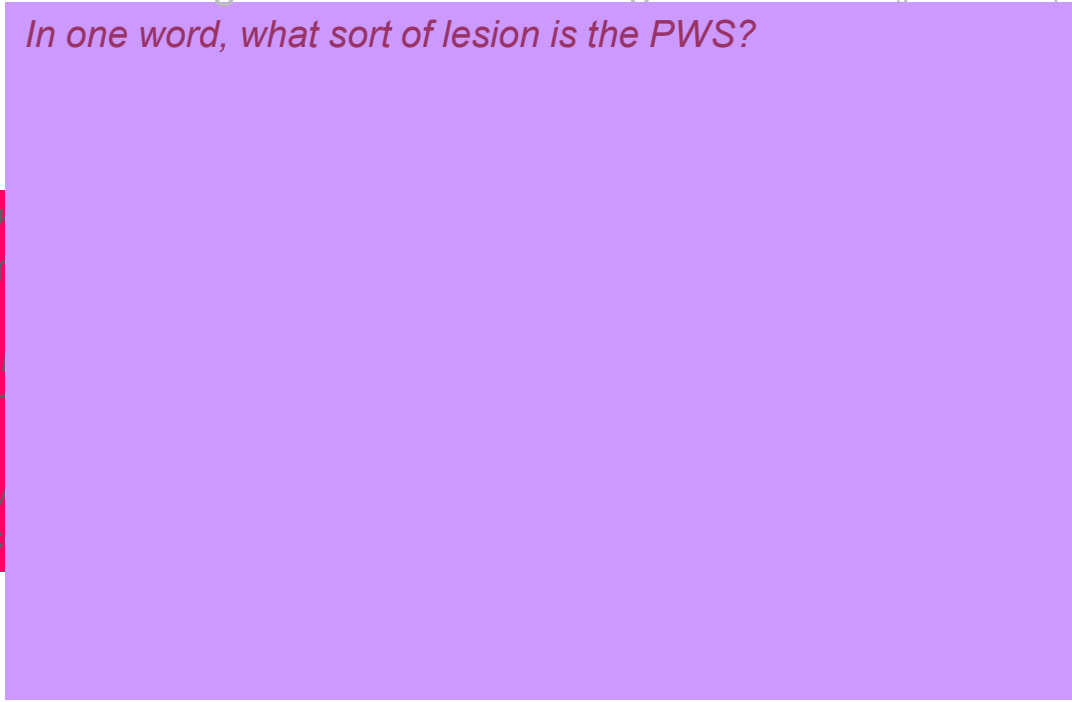
In one word, what sort of lesion is the PWS?

Port wine stain

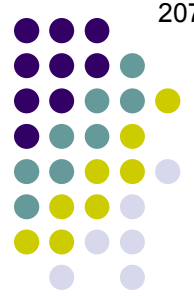
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Vascular Lesions of the Orbit

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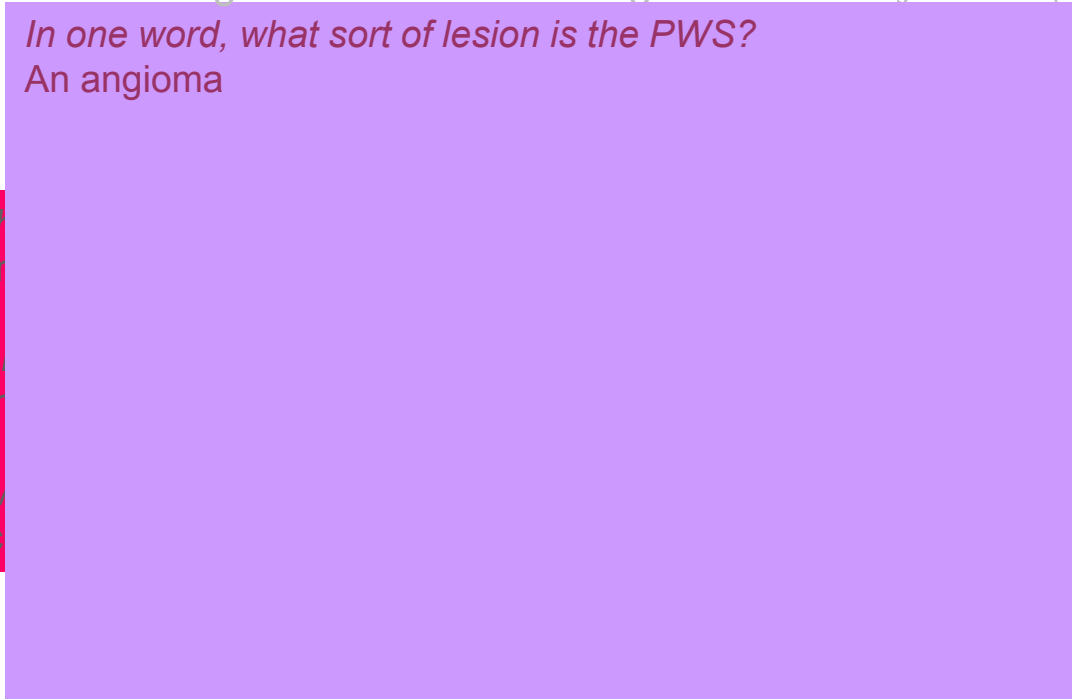
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Port wine stain

With
Stur

In o
Apt

By v
End



YM



Q

Vascular Lesions of the Orbit

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Malformations

Strawberry hemangioma

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Cavernous hemangioma

In one word, what sort of lesion is the PWS?
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By what 'official' name is it known?

Port wine stain

With
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In o
Apt

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YM

A

Vascular Lesions of the Orbit

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Cavernous hemangioma

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Port wine stain

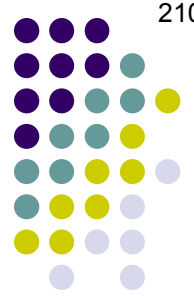
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Q

Vascular Lesions of the Orbit

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When does it present?

Port wine stain

With
Stur

In o
Apt

By v
Enc



A

Vascular Lesions of the Orbit

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

When does it present?
 At birth

Port wine stain

With
Stur
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Enc



Vascular Lesions of the Orbit



Sturge-Weber: Port-wine stain



Q

Vascular Lesions of the Orbit

Hemangiomas

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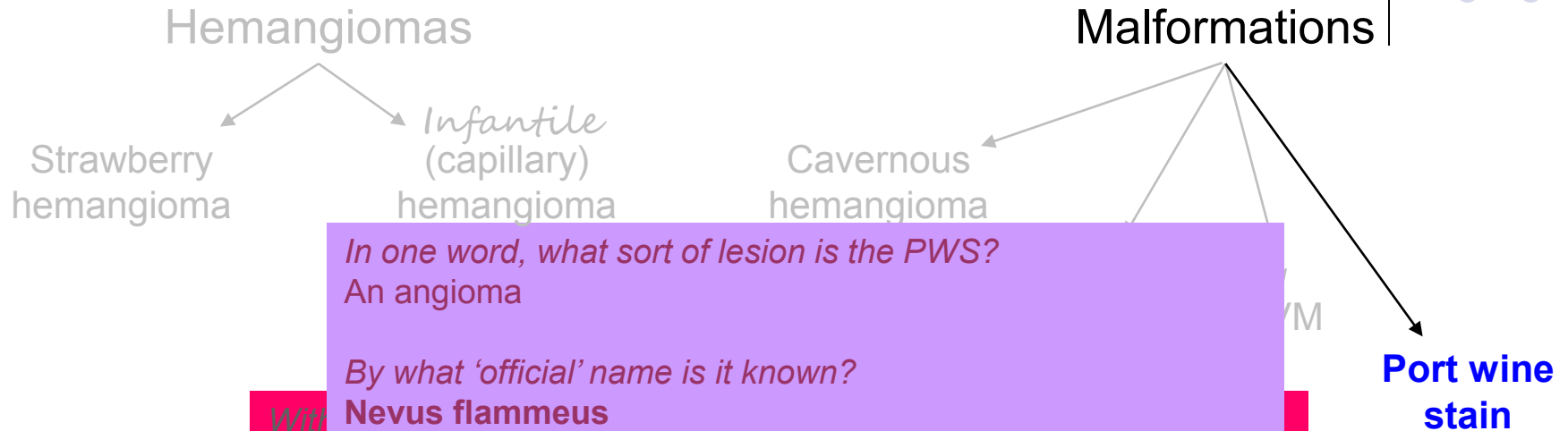
When does it present?
 At birth

What is the typical pattern of distribution?

Port wine stain

A

Vascular Lesions of the Orbit



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

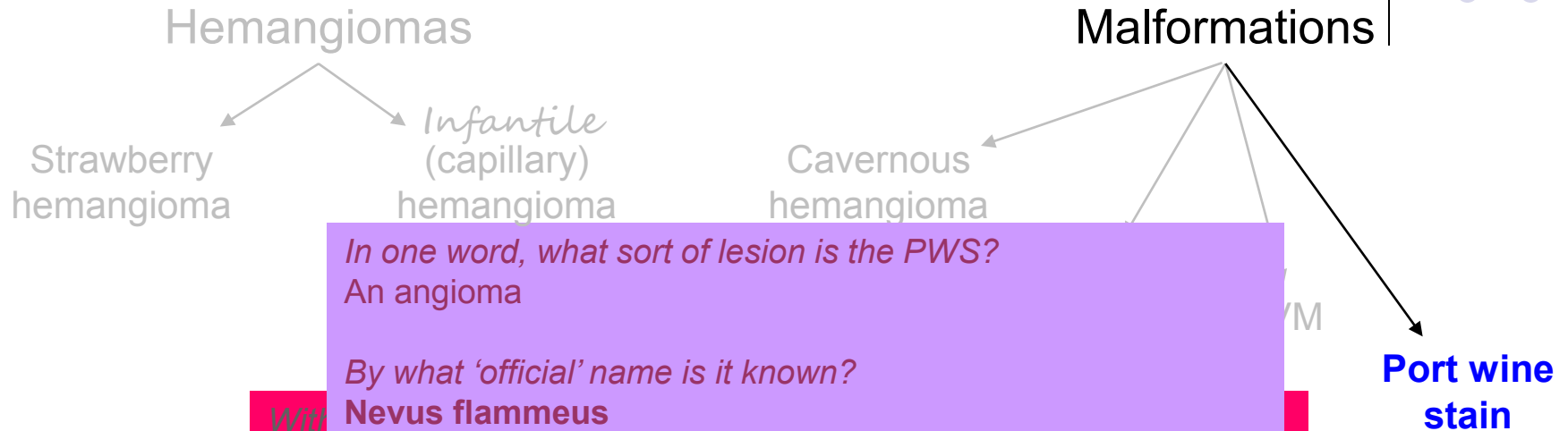
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When does it present?
At birth

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

Q

Vascular Lesions of the Orbit



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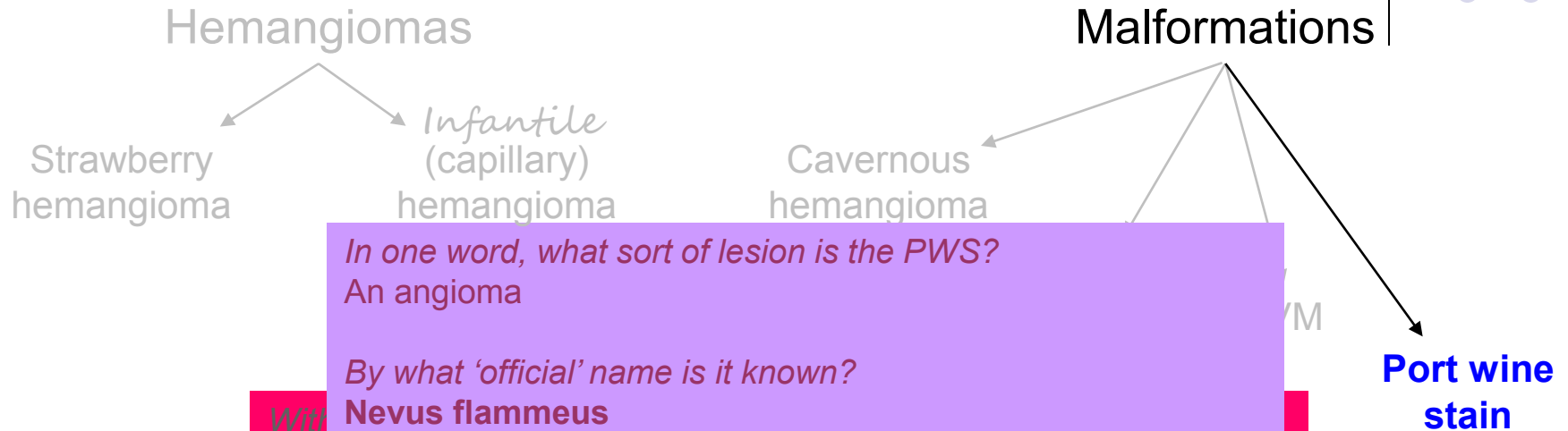
When does it present?
At birth

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

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

A

Vascular Lesions of the Orbit



In one word, what sort of lesion is the PWS?
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By what 'official' name is it known?
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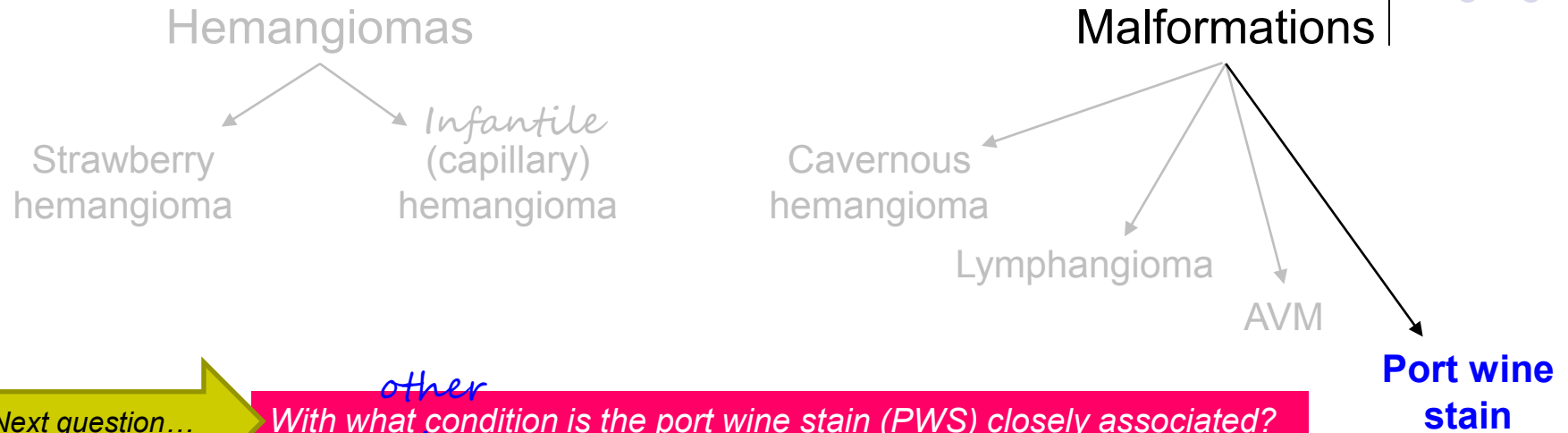
When does it present?
At birth

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

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

Q

Vascular Lesions of the Orbit



Next question...

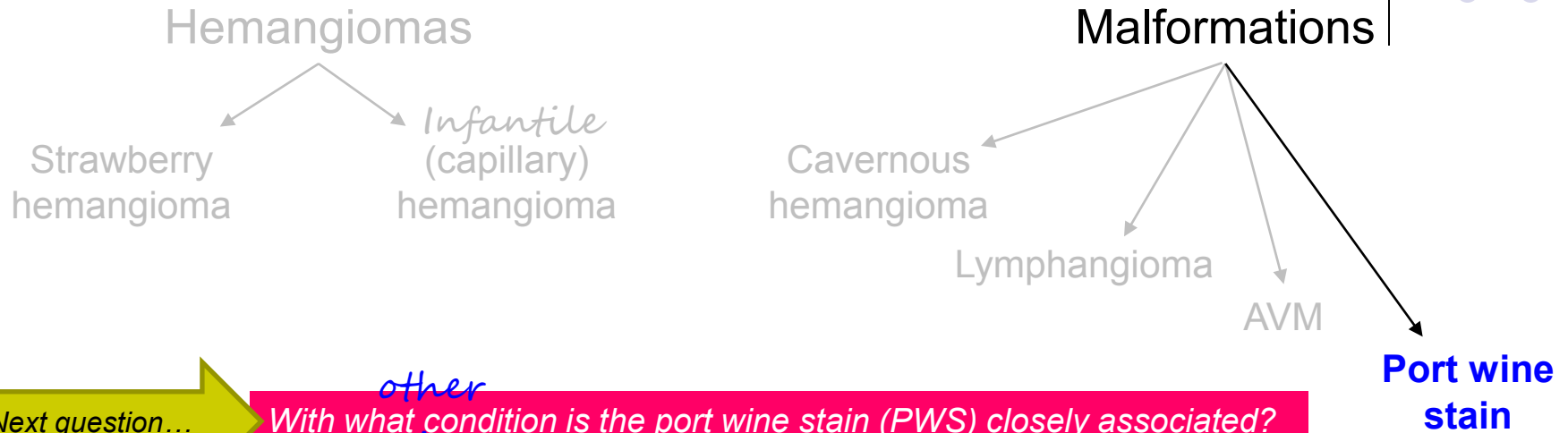
With what ^{other} condition is the port wine stain (PWS) closely associated?
~~Sturge-Weber syndrome~~

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

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

A

Vascular Lesions of the Orbit



Next question...

With what ^{other} condition is the port wine stain (PWS) closely associated?
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Vascular Lesions of the Orbit



Klippel-Trénaunay syndrome

Q

Vascular Lesions of the Orbit



other
 With what condition is the port wine stain (PWS) closely associated?
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In one word, what is a phakomatosis?
 How is Klippel-Trénaunay pronounced?

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A

Vascular Lesions of the Orbit



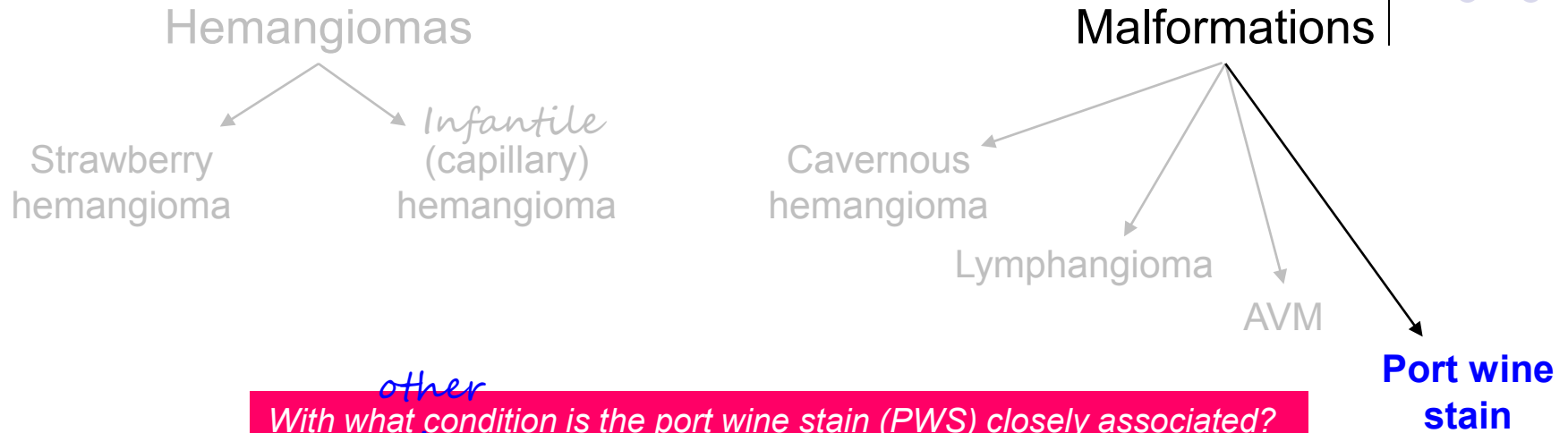
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 CLIP-el treh-NOW-nay

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Q

Vascular Lesions of the Orbit

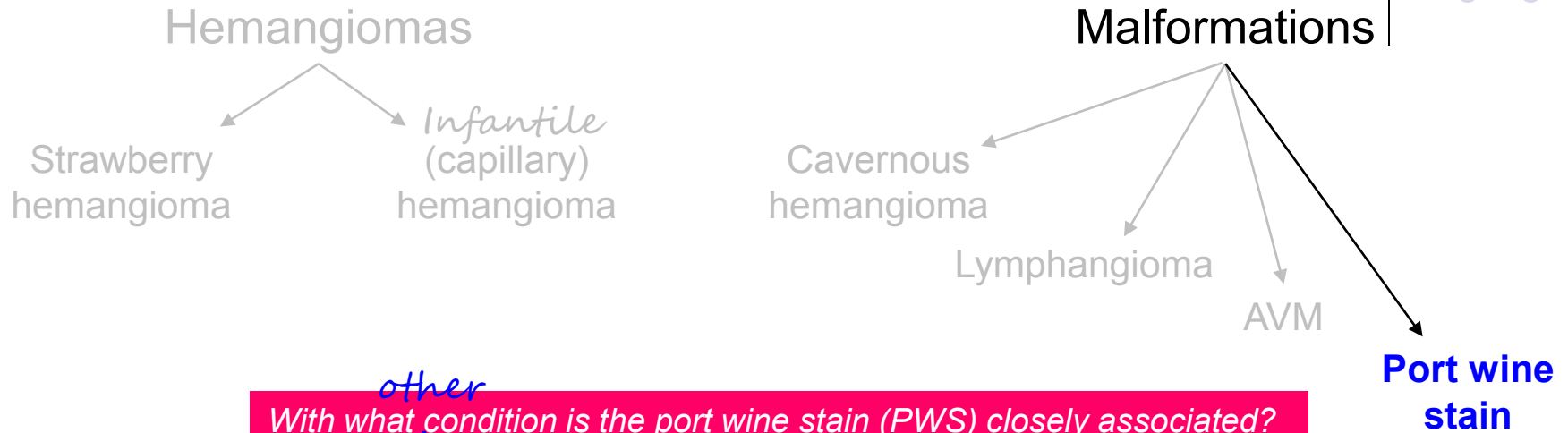


other
 With what condition is the port wine stain (PWS) closely associated?
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Is KTS a phakomatosis?

A

Vascular Lesions of the Orbit

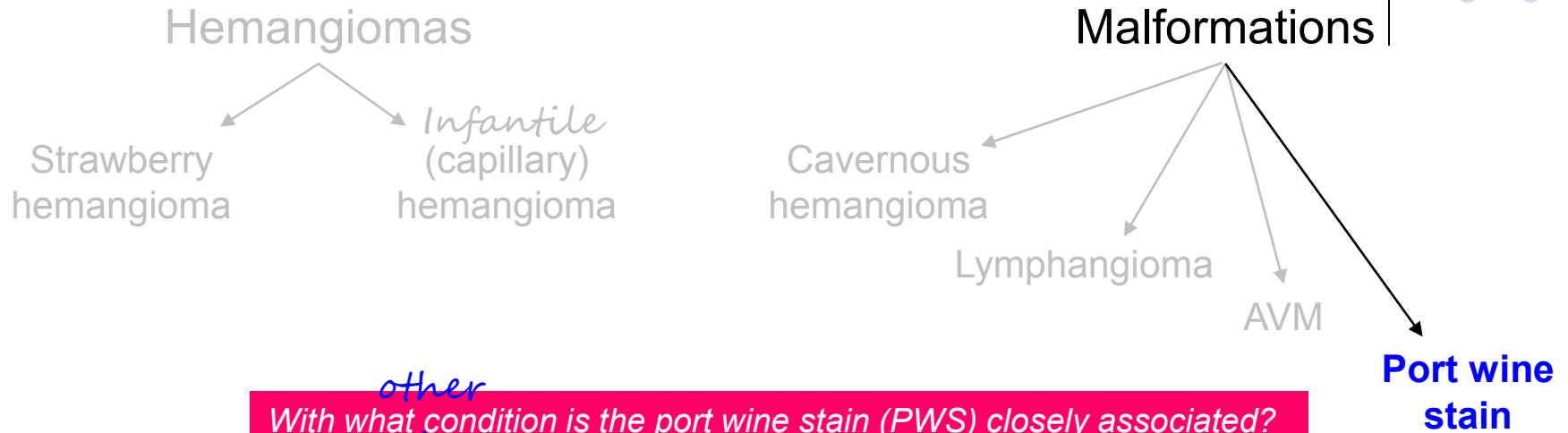


other
 With what condition is the port wine stain (PWS) closely associated?
~~Sturge-Weber syndrome~~ Klippel-Trénaunay syndrome (KTS)

Is KTS a phakomatosis?
 Yes

Q

Vascular Lesions of the Orbit

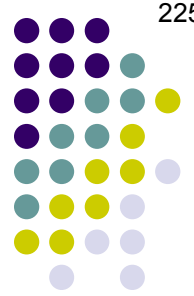


other
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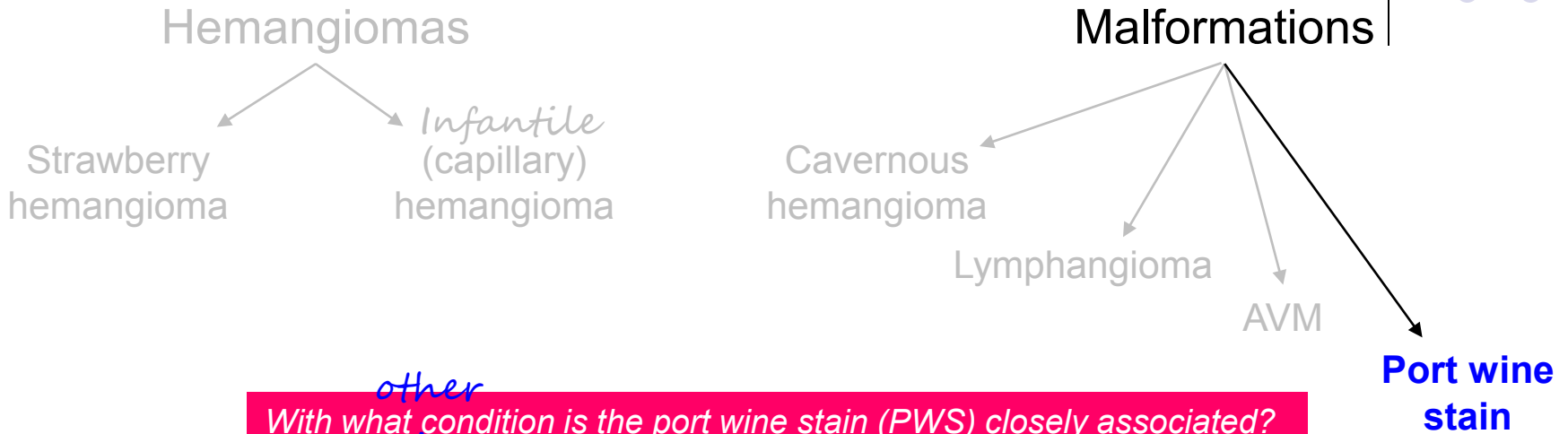
Is KTS a phakomatosis?

Yes

Is the PWS in KTS limited to the face, as in Sturge-Weber?



Vascular Lesions of the Orbit



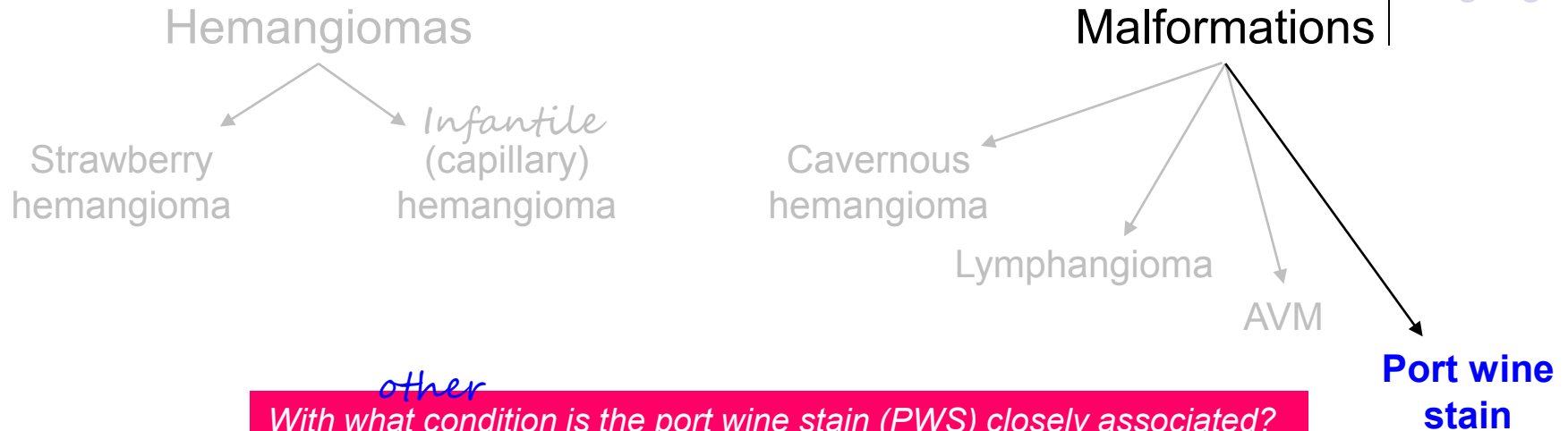
other
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Is KTS a phakomatosis?
Yes

Is the PWS in KTS limited to the face, as in Sturge-Weber?
No, it is also found on the [] as well as a []

A

Vascular Lesions of the Orbit



other
 With what condition is the port wine stain (PWS) closely associated?
~~Sturge-Weber syndrome~~ Klippel-Trénaunay syndrome (KTS)

Is KTS a phakomatosis?

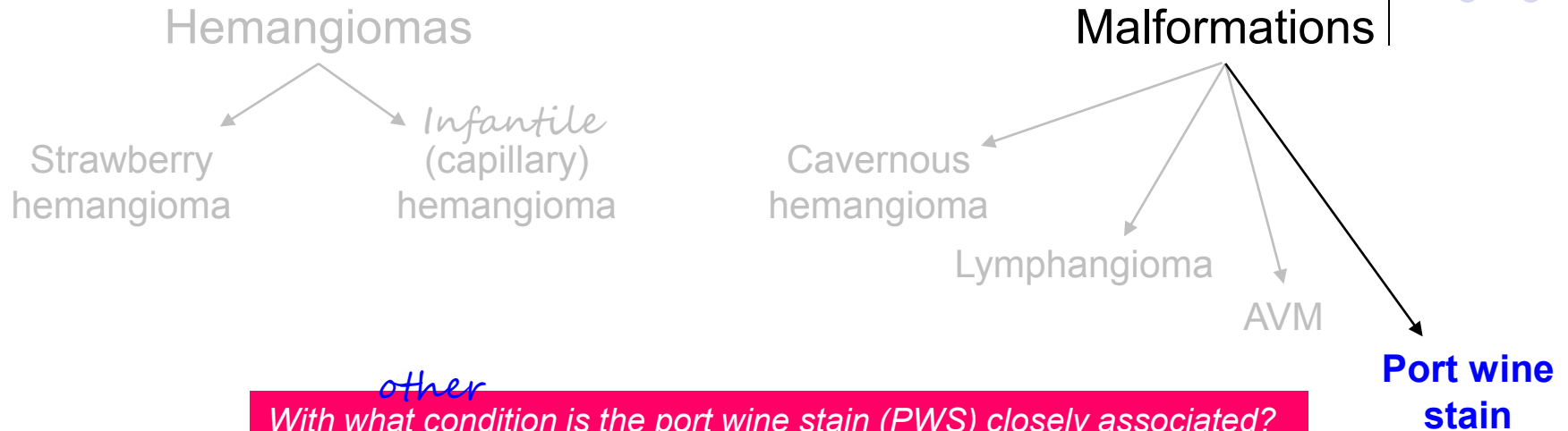
Yes

Is the PWS in KTS limited to the face, as in Sturge-Weber?

No, it is also found on the trunk as well as a limb

Q

Vascular Lesions of the Orbit



other
 With what condition is the port wine stain (PWS) closely associated?
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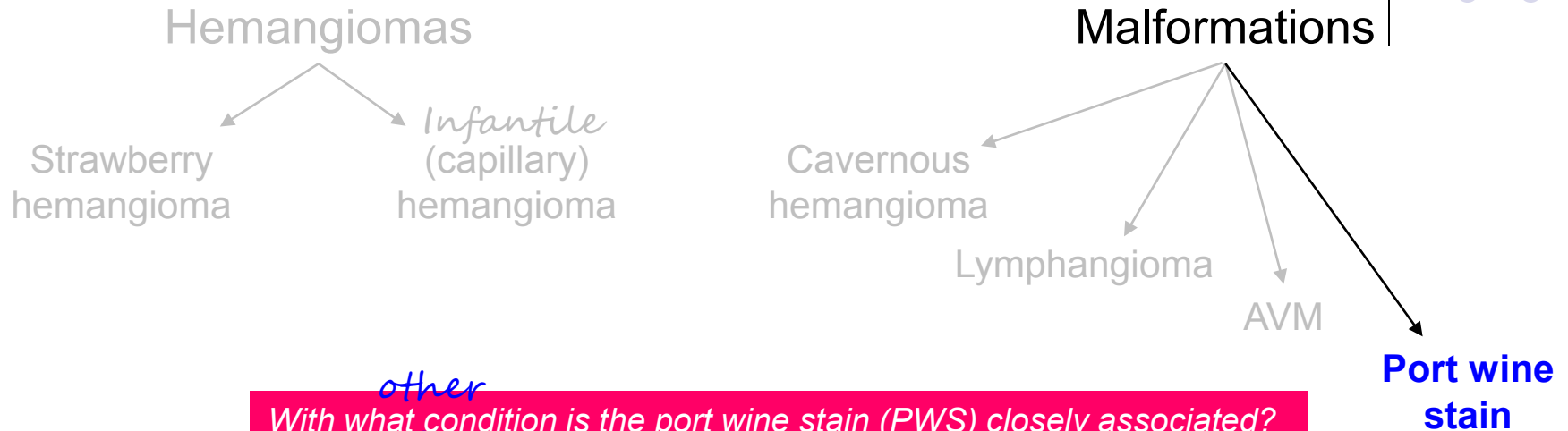
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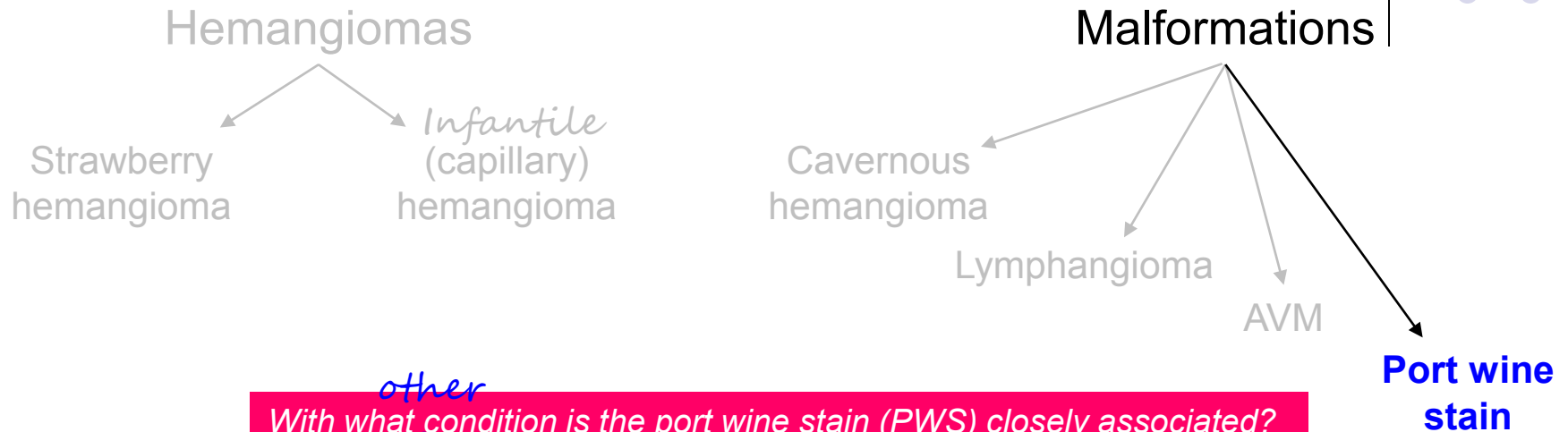
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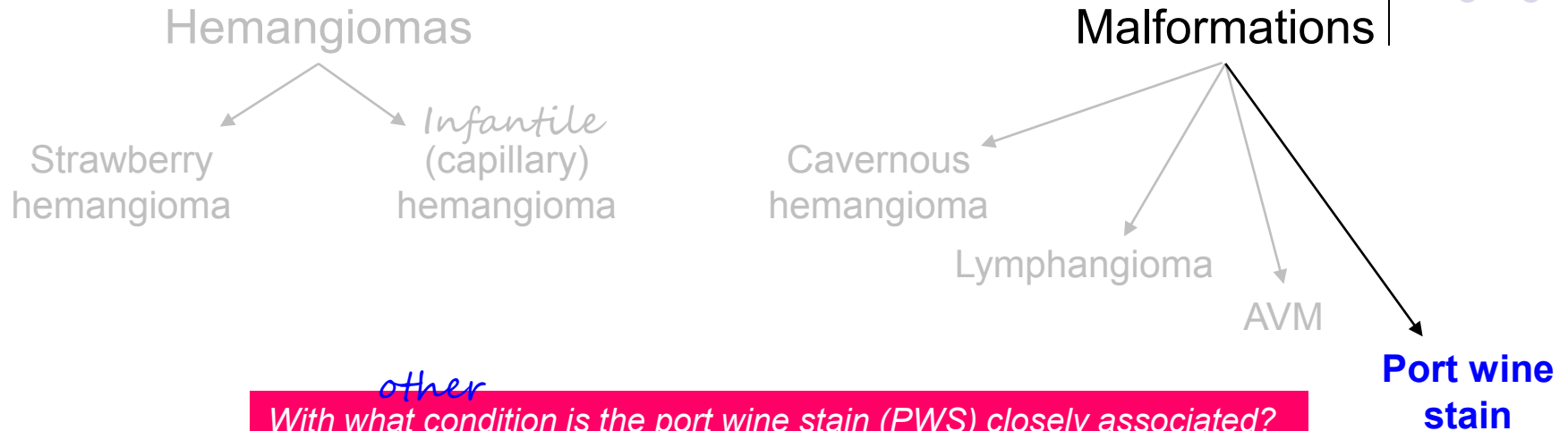
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Vascular Lesions of the Orbit



Klippel-Trénaunay syndrome