

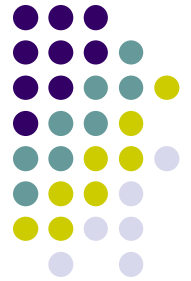
Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

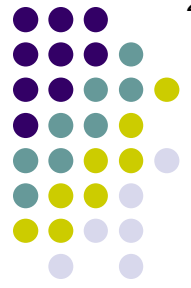
*Basic
clinical
distinction*

?

?



Pediatric Iris Cysts and Nodules



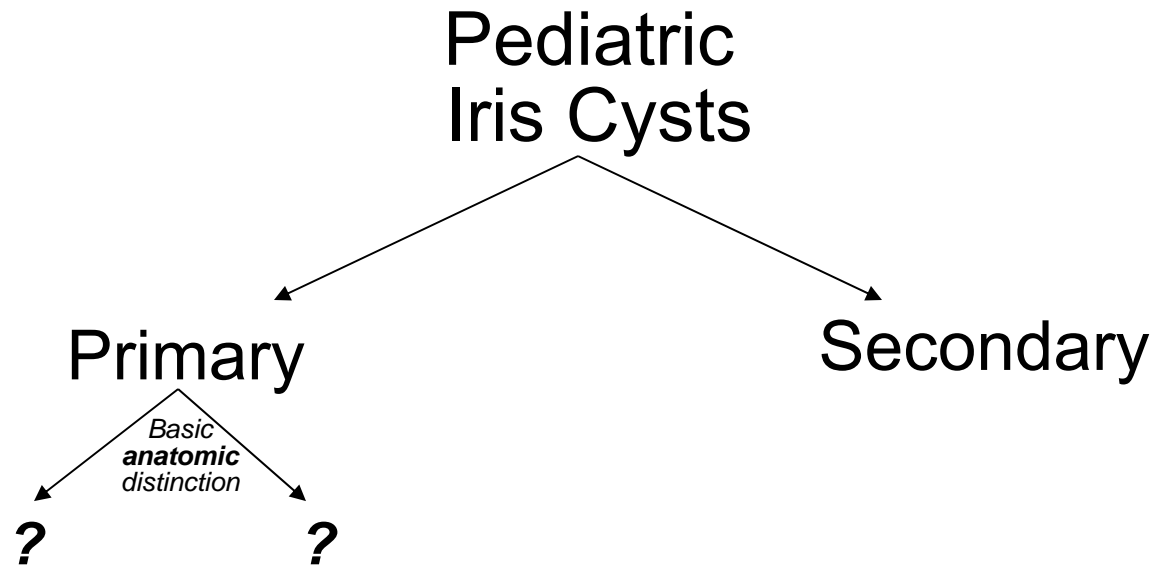
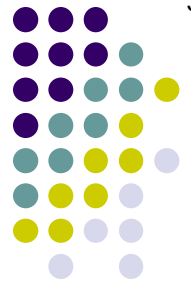
Pediatric Iris Cysts

Basic clinical distinction

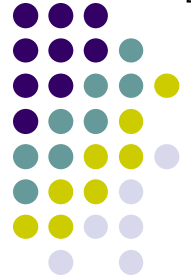
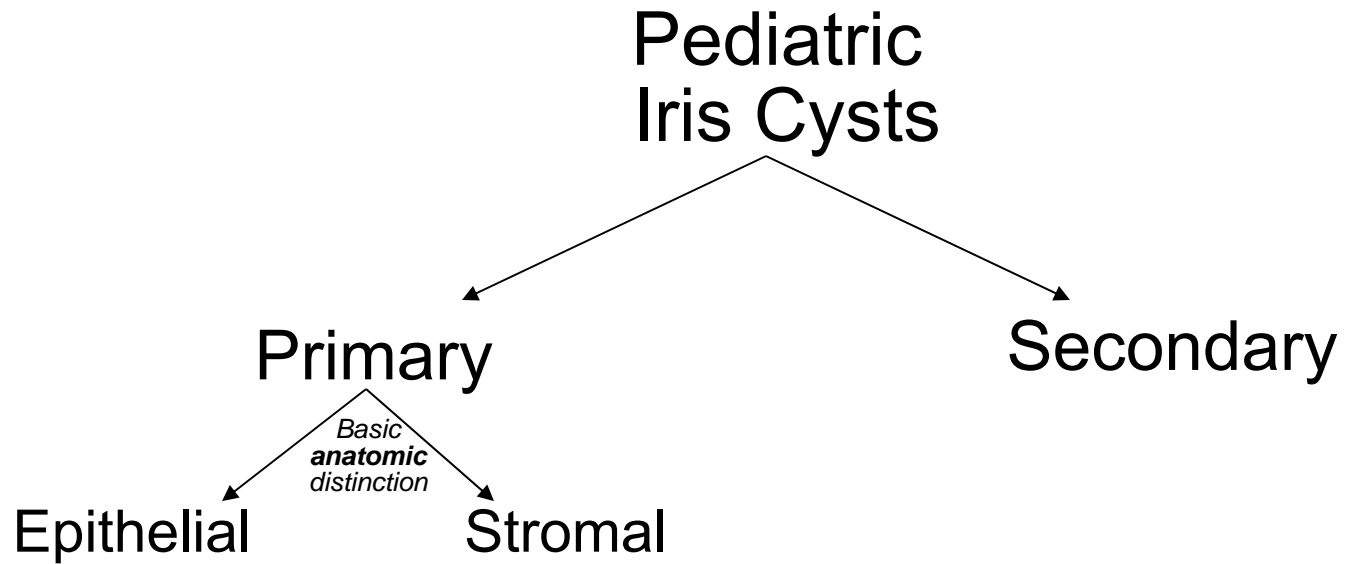
Primary

Secondary

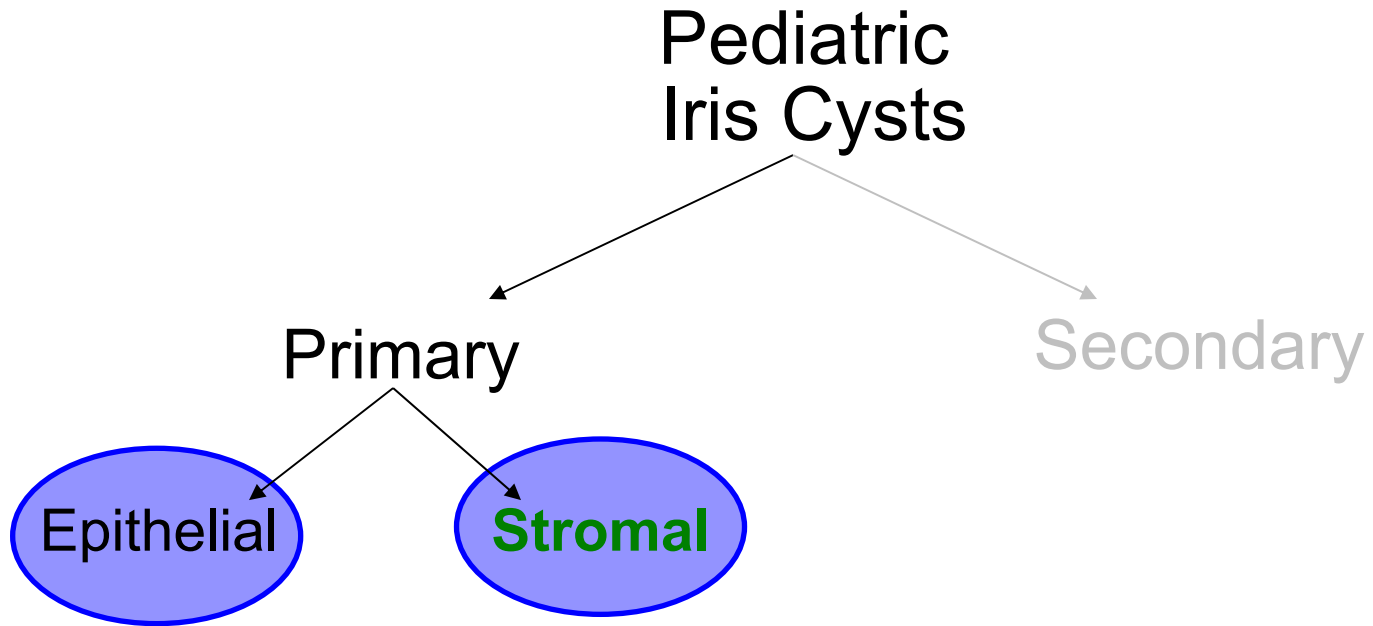
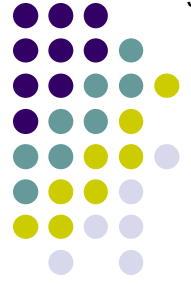
Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts and Nodules

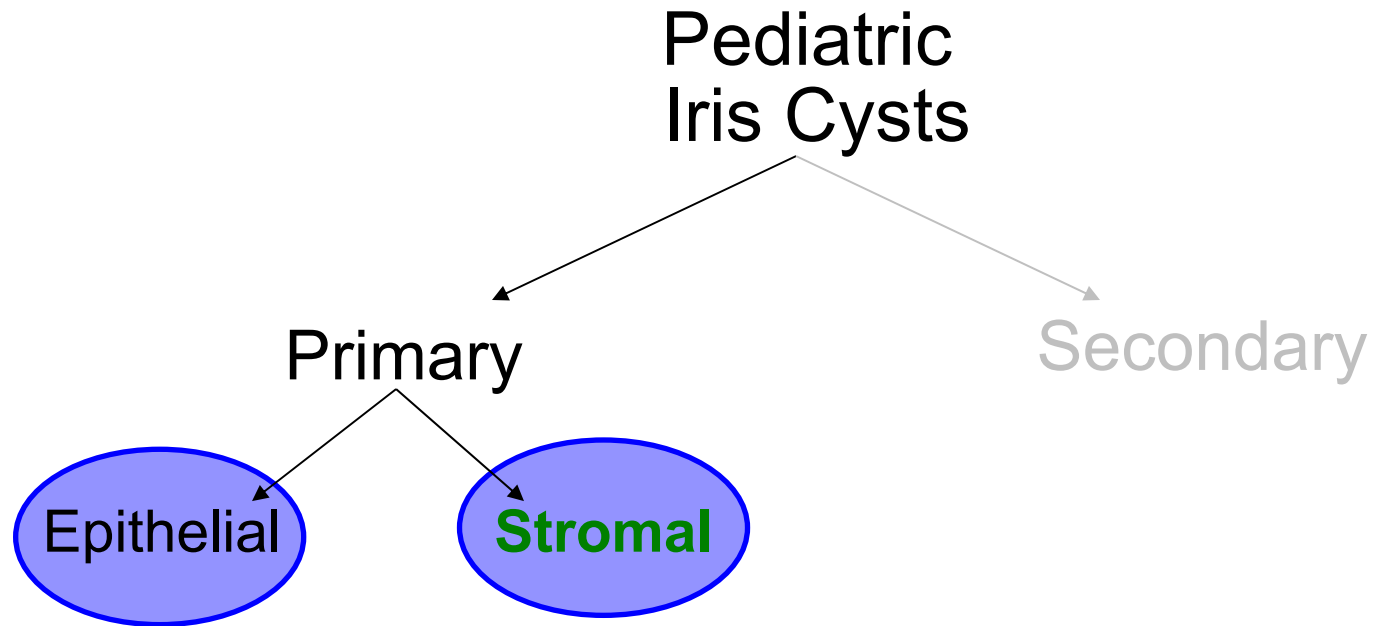


Pediatric Iris Cysts and Nodules



Which is more common--epithelial, or stromal cysts?

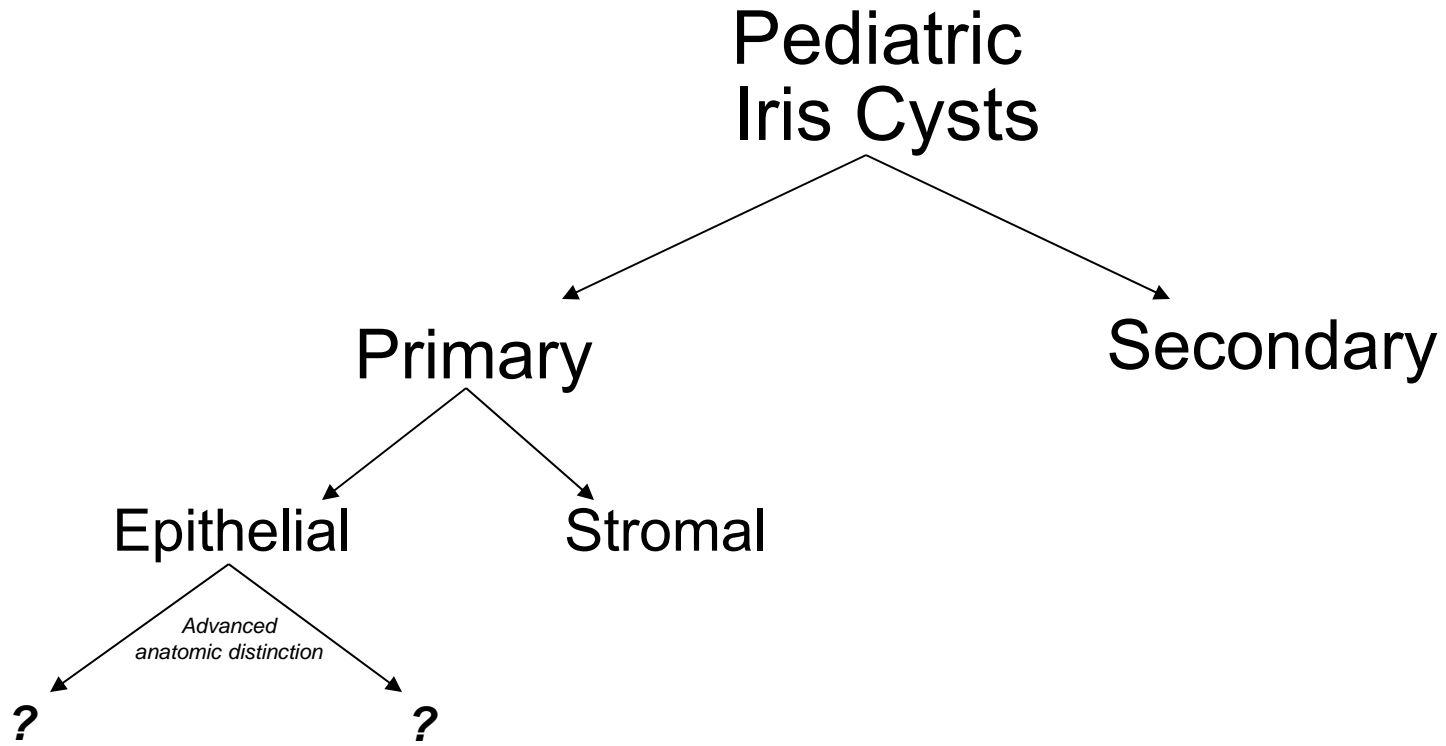
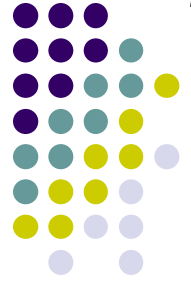
Pediatric Iris Cysts and Nodules



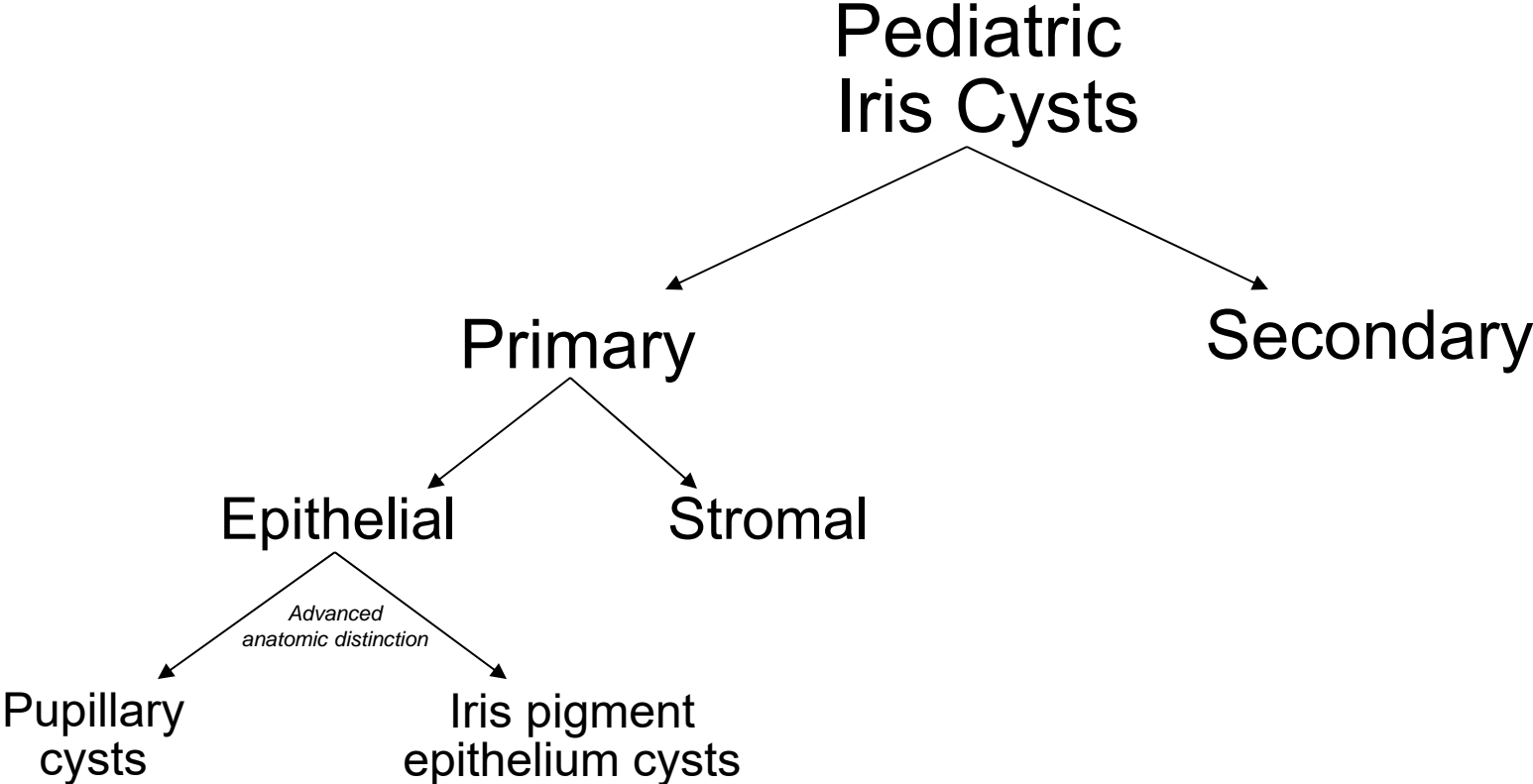
Which is more common--epithelial, or stromal cysts?
Epithelial, by a mile



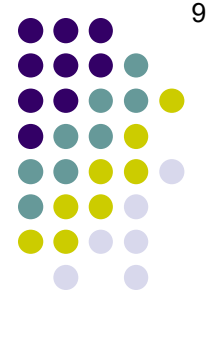
Pediatric Iris Cysts and Nodules



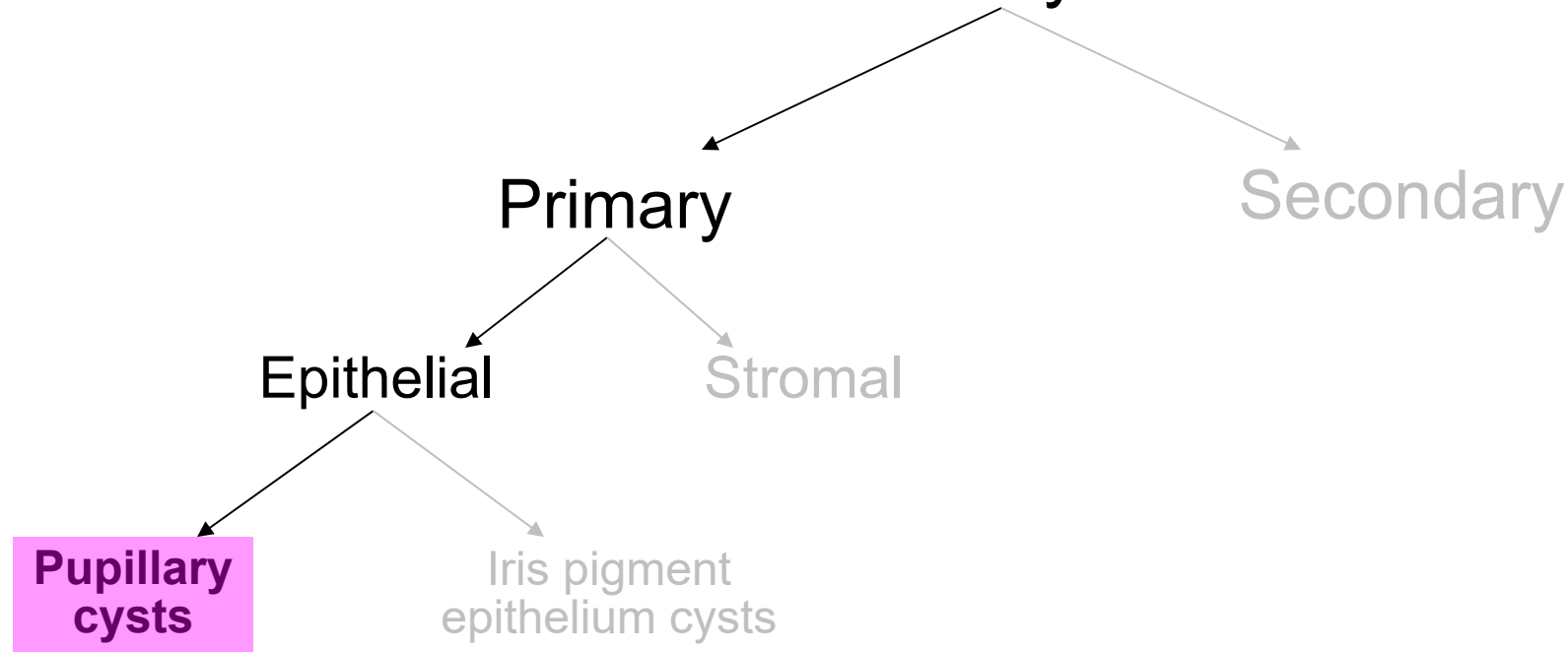
Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

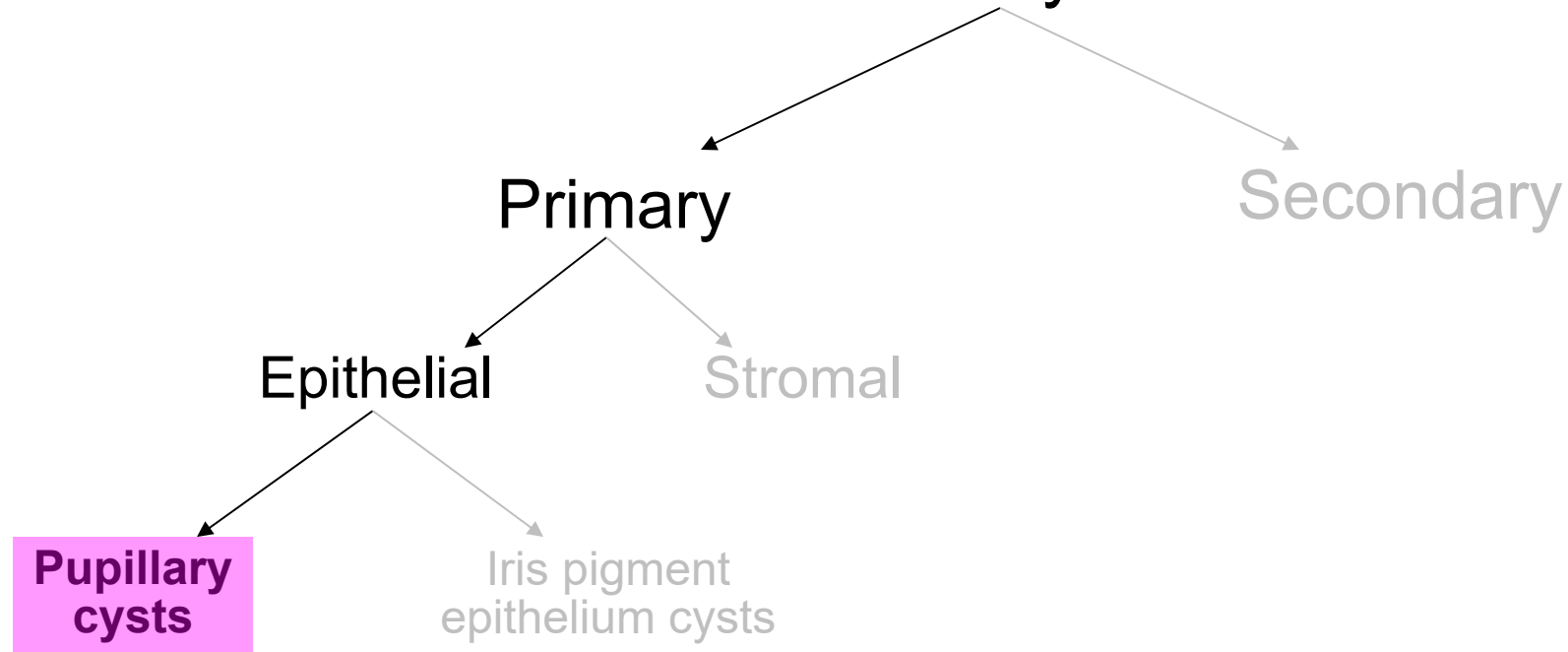


Pupillary cysts
--Pigment epi cysts at the...[location]

Pediatric Iris Cysts and Nodules

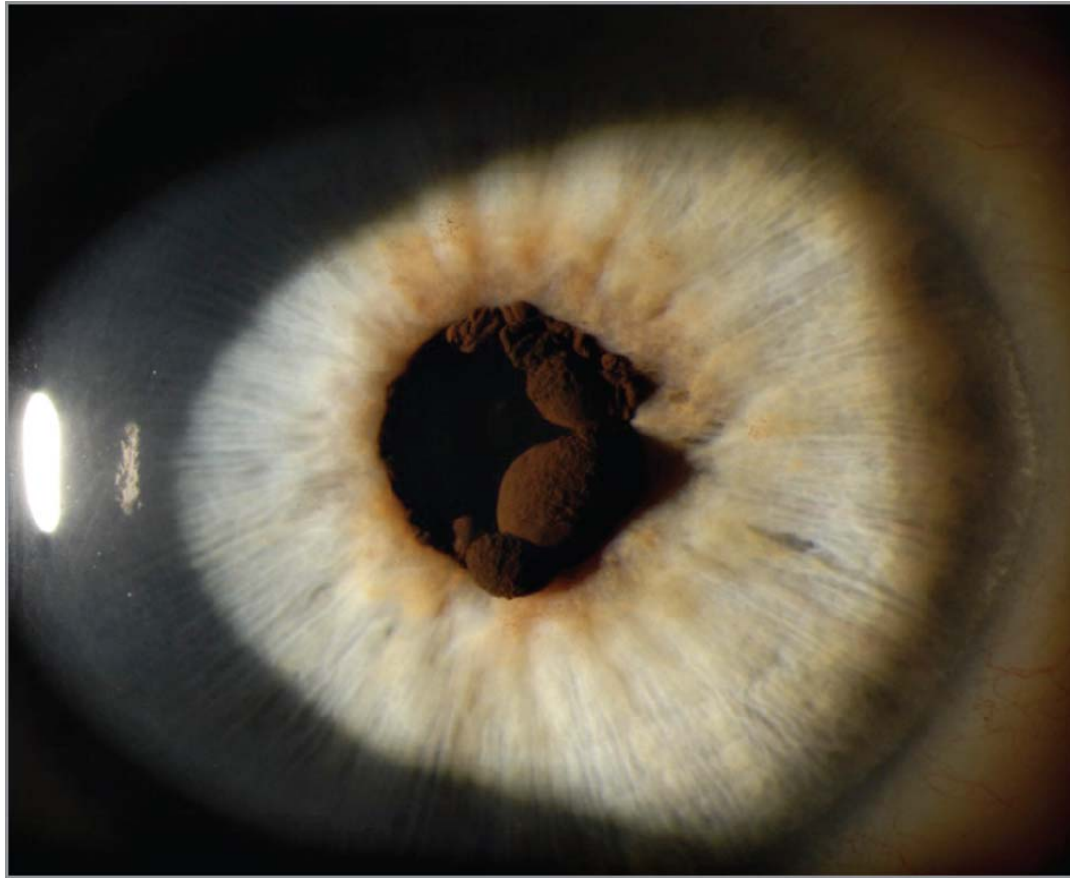
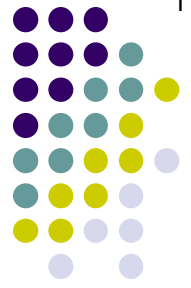


Pediatric Iris Cysts



Pupillary cysts
--Pigment epi cysts at the...pupil border

Pediatric Iris Cysts and Nodules

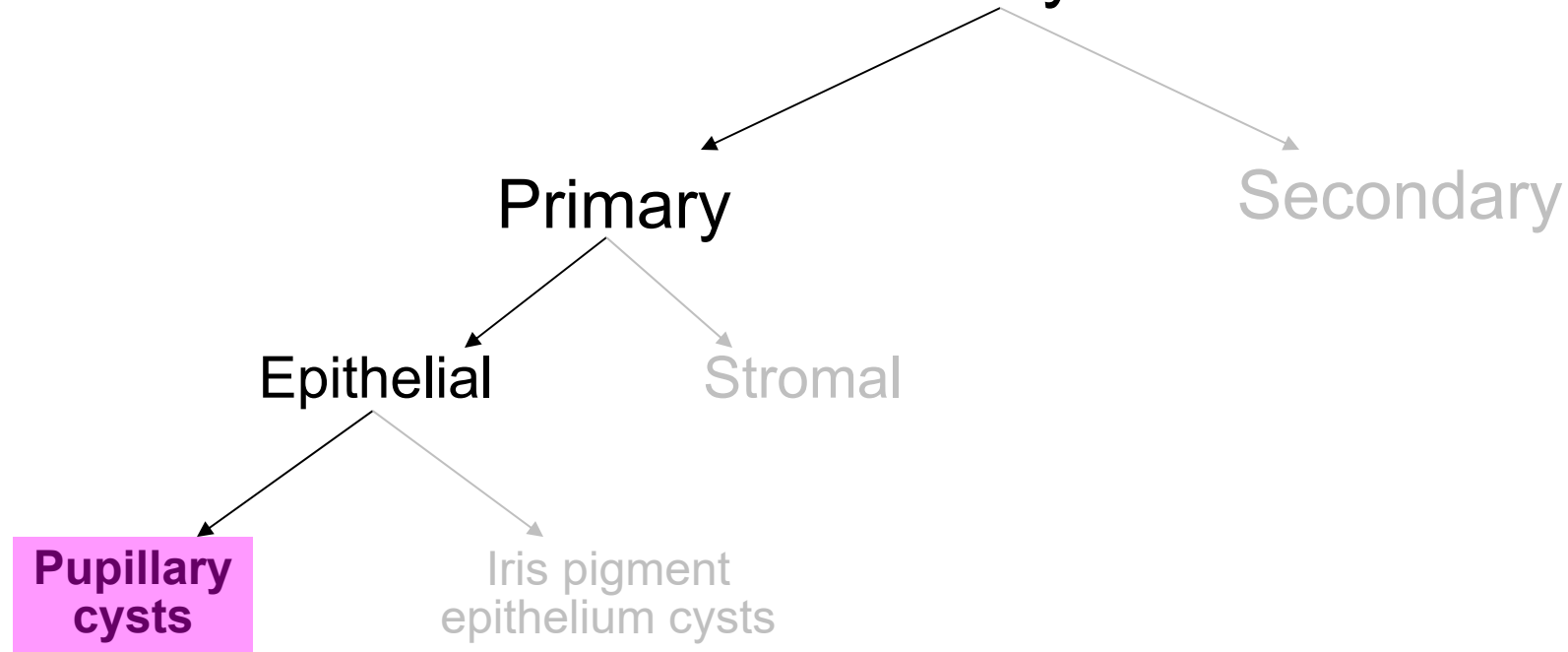


Primary epithelial pupillary cysts

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

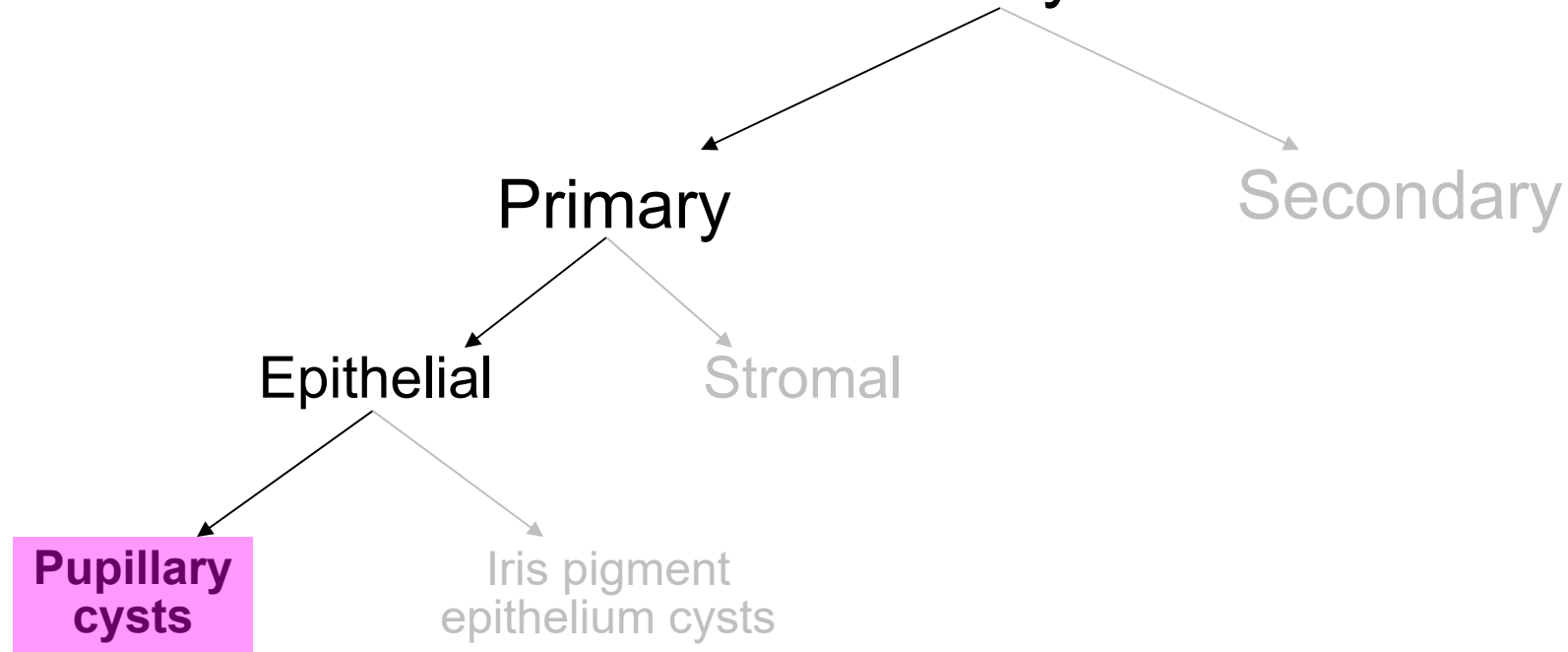


Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...[age]

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts



Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

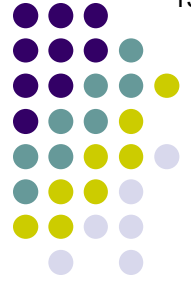
Pupillary cysts

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

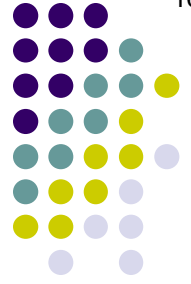
Pupillary cysts

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...[age range]

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

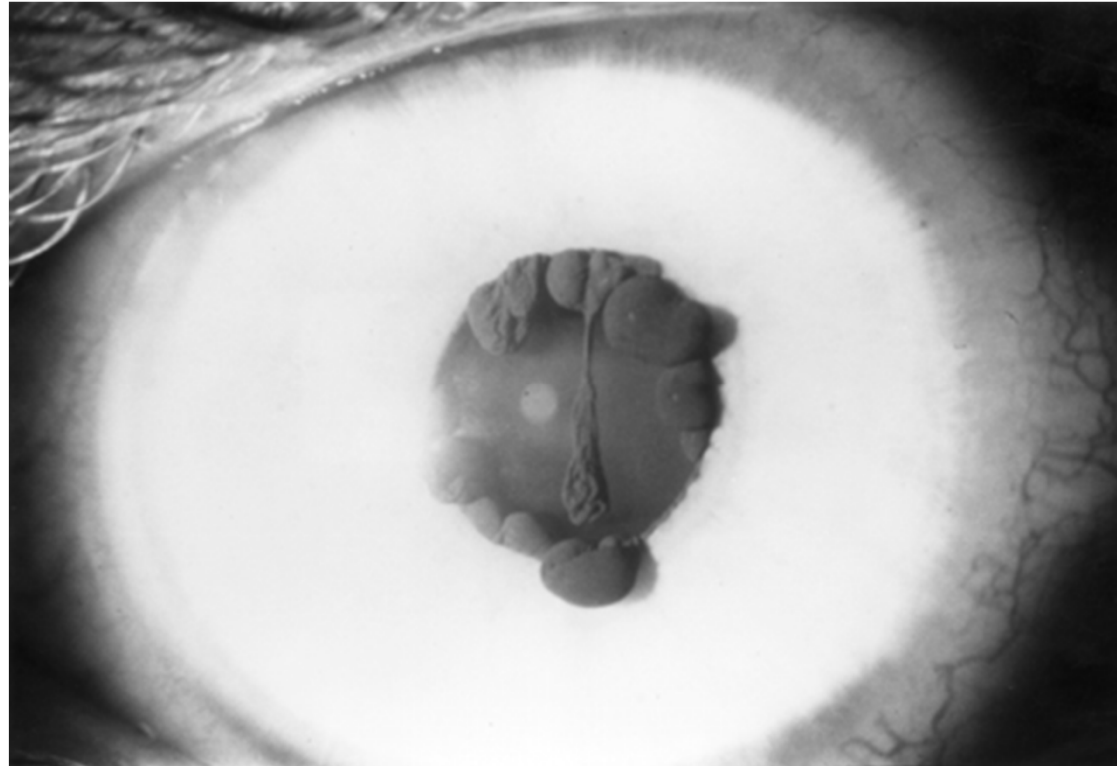
Pupillary cysts

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

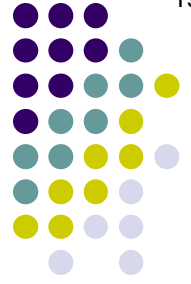
Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens

Pediatric Iris Cysts and Nodules



Primary epithelial pupillary cysts in a teen

Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

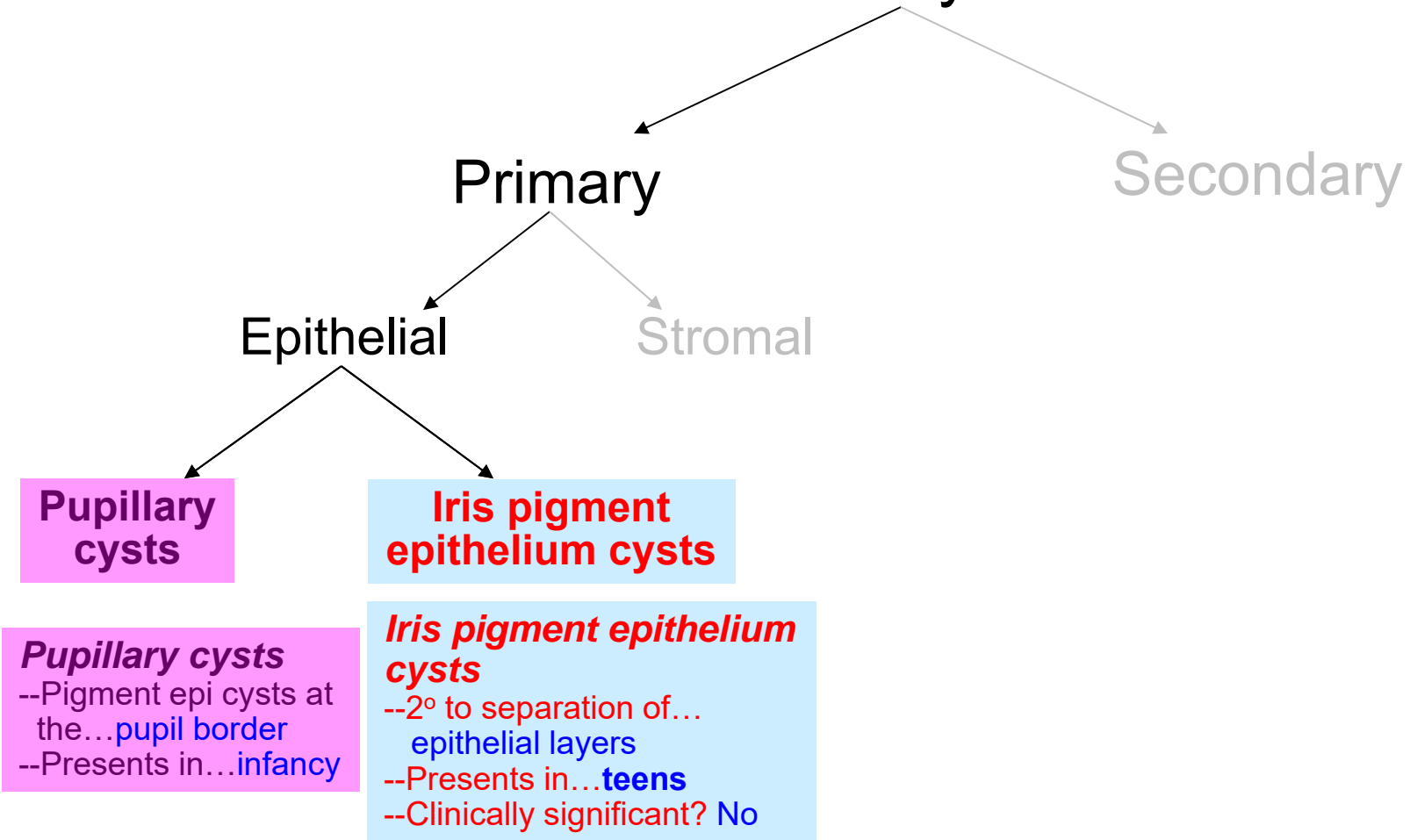
Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? [Y/N]

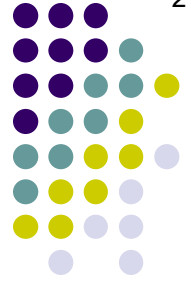
Pediatric Iris Cysts and Nodules



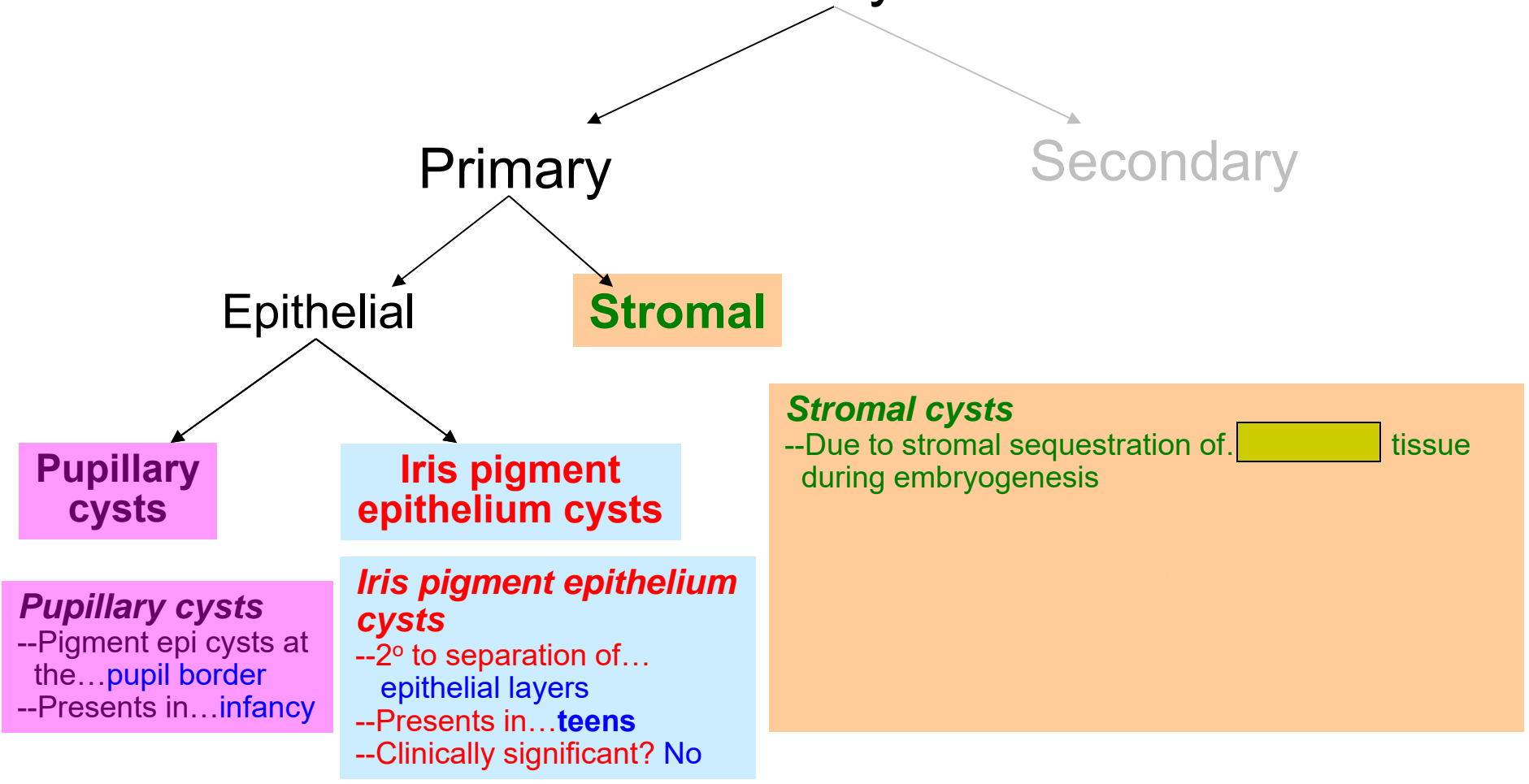
Pediatric Iris Cysts



Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts





Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Primary

Stromal

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Primary

Epithelial

Stromal

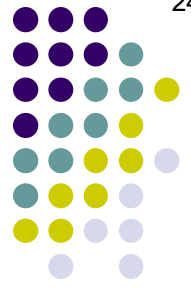
Pupillary cysts

Iris pigment epithelium cysts

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...[age]

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Primary

Epithelial

Stromal

Pupillary cysts

Iris pigment epithelium cysts

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy

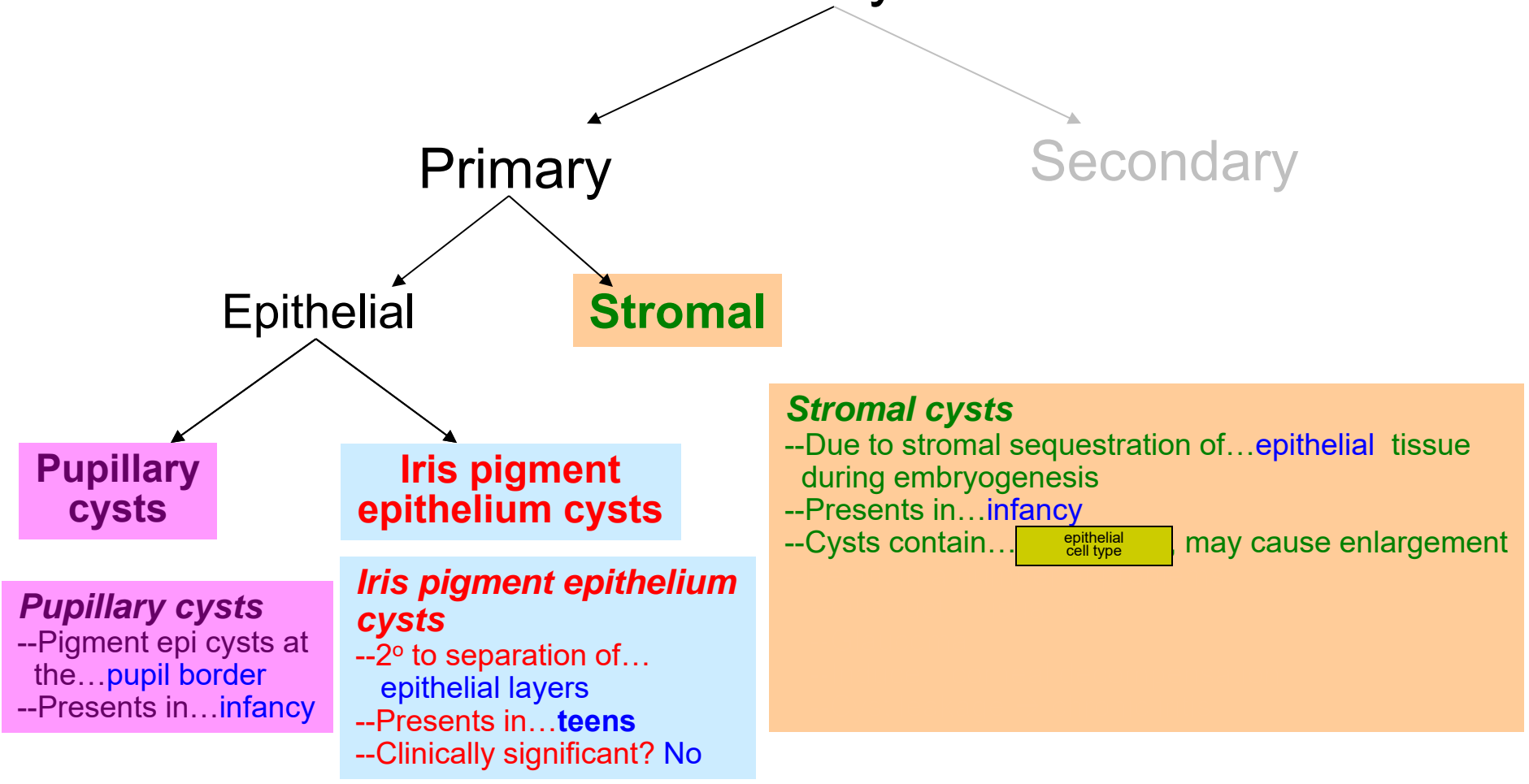
Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

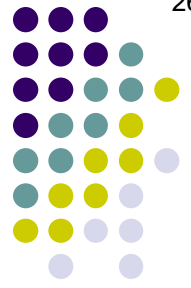
Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No



Pediatric Iris Cysts and Nodules

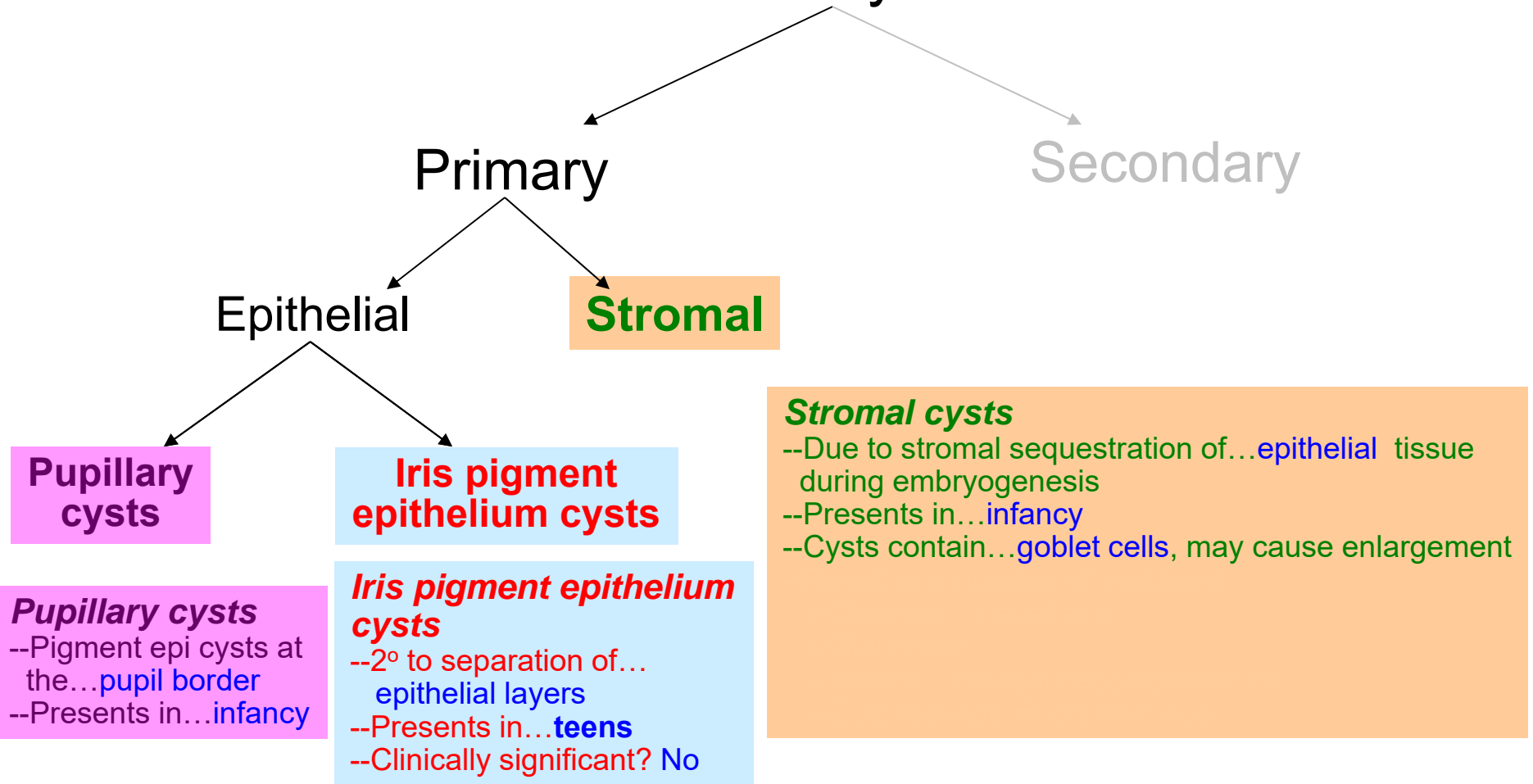
Pediatric Iris Cysts

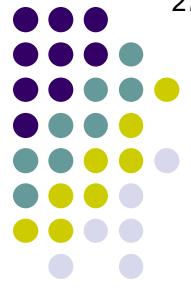




Pediatric Iris Cysts and Nodules

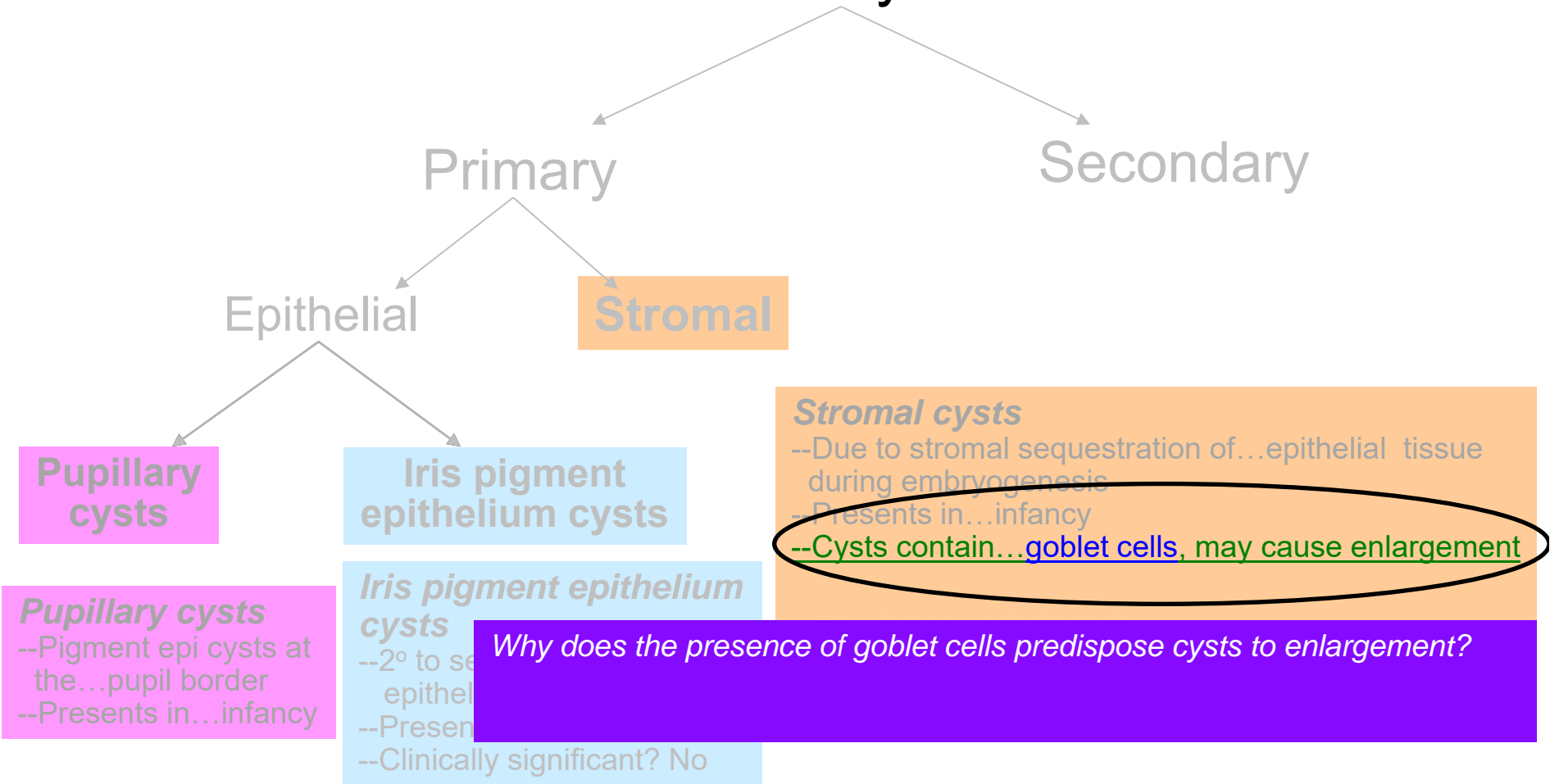
Pediatric Iris Cysts

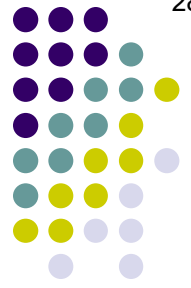




Pediatric Iris Cysts and Nodules

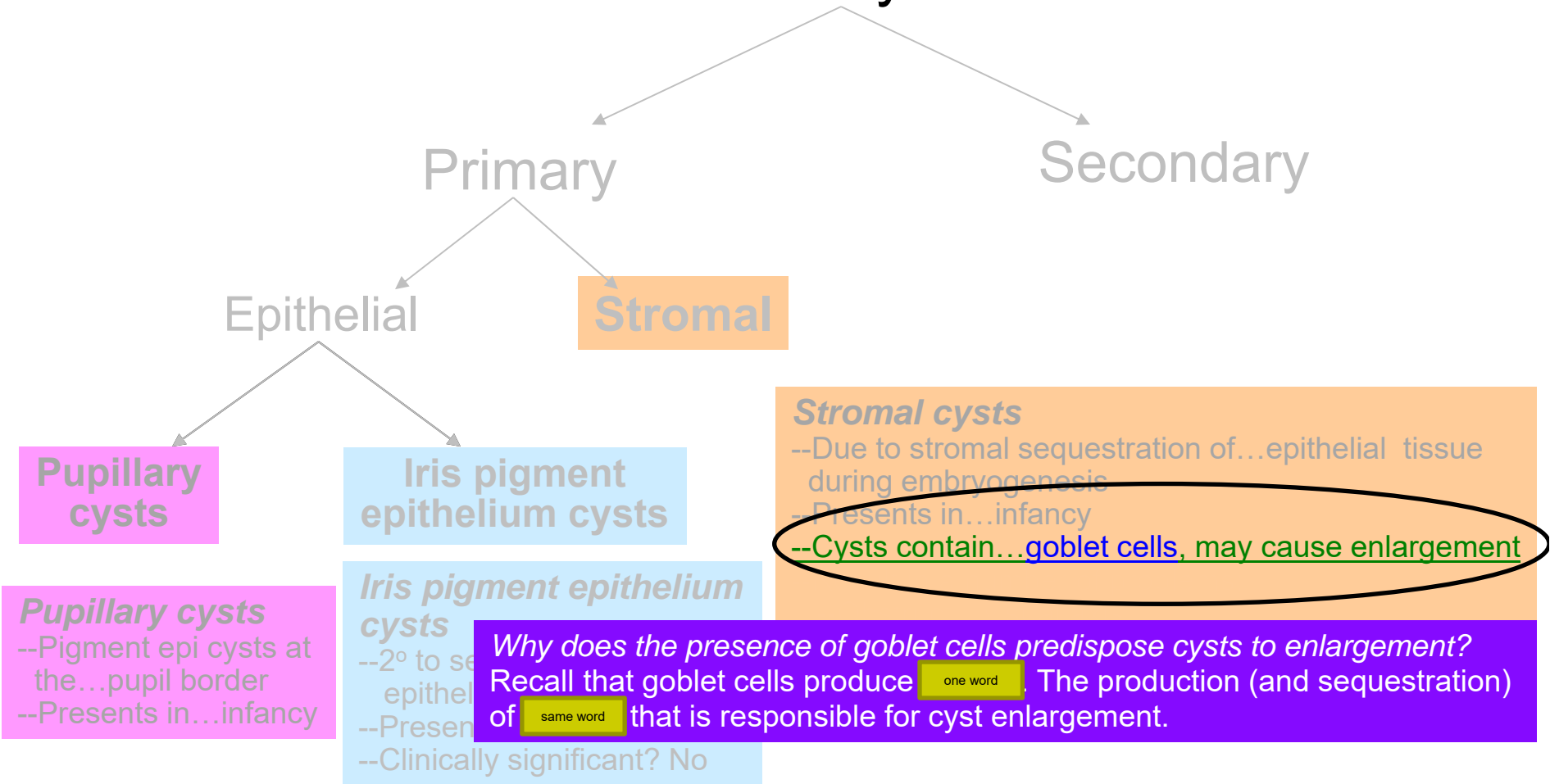
Pediatric Iris Cysts





Pediatric Iris Cysts and Nodules

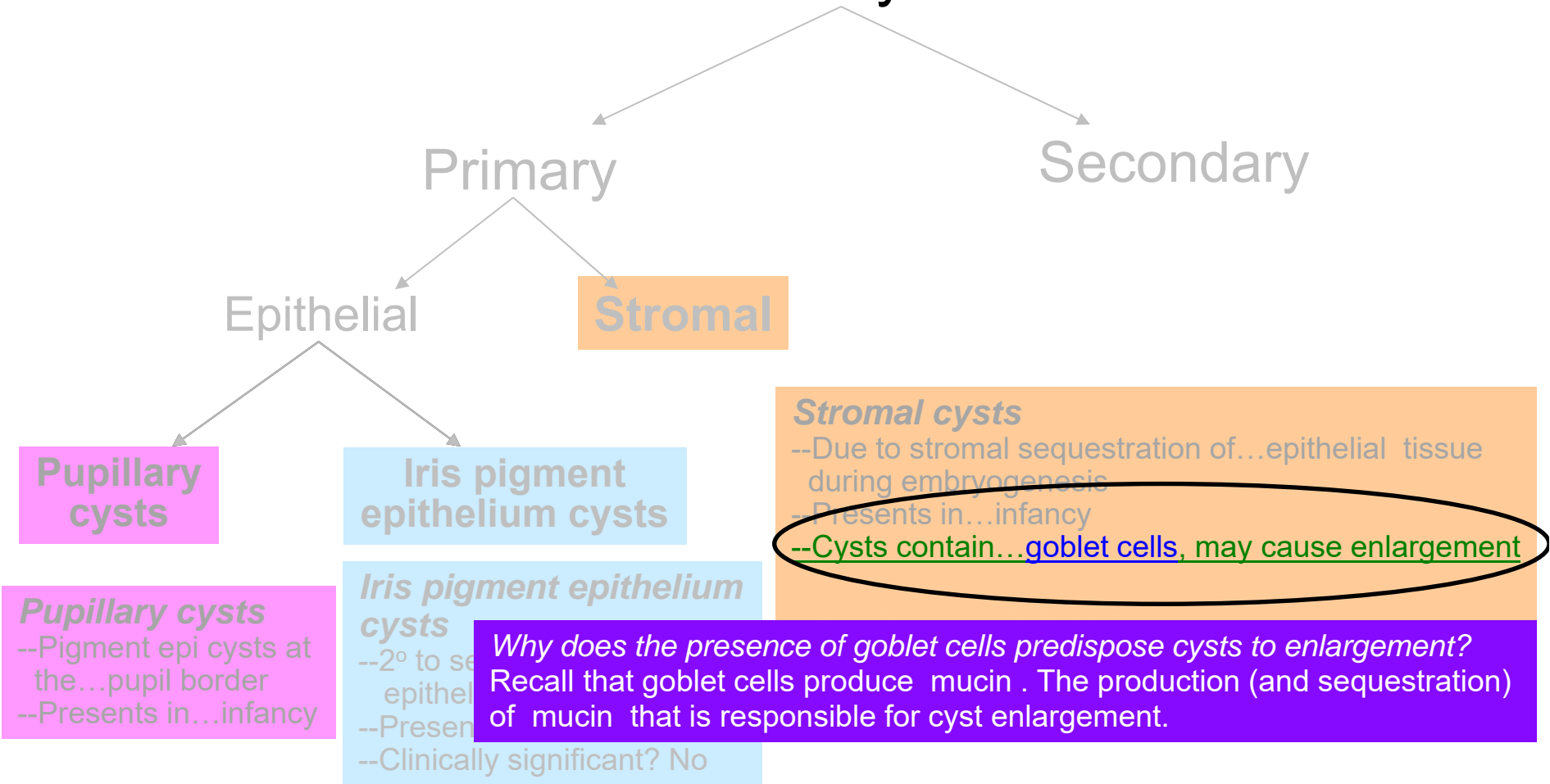
Pediatric Iris Cysts

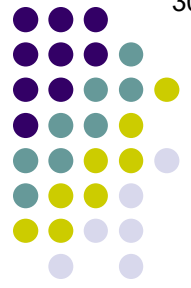




Pediatric Iris Cysts and Nodules

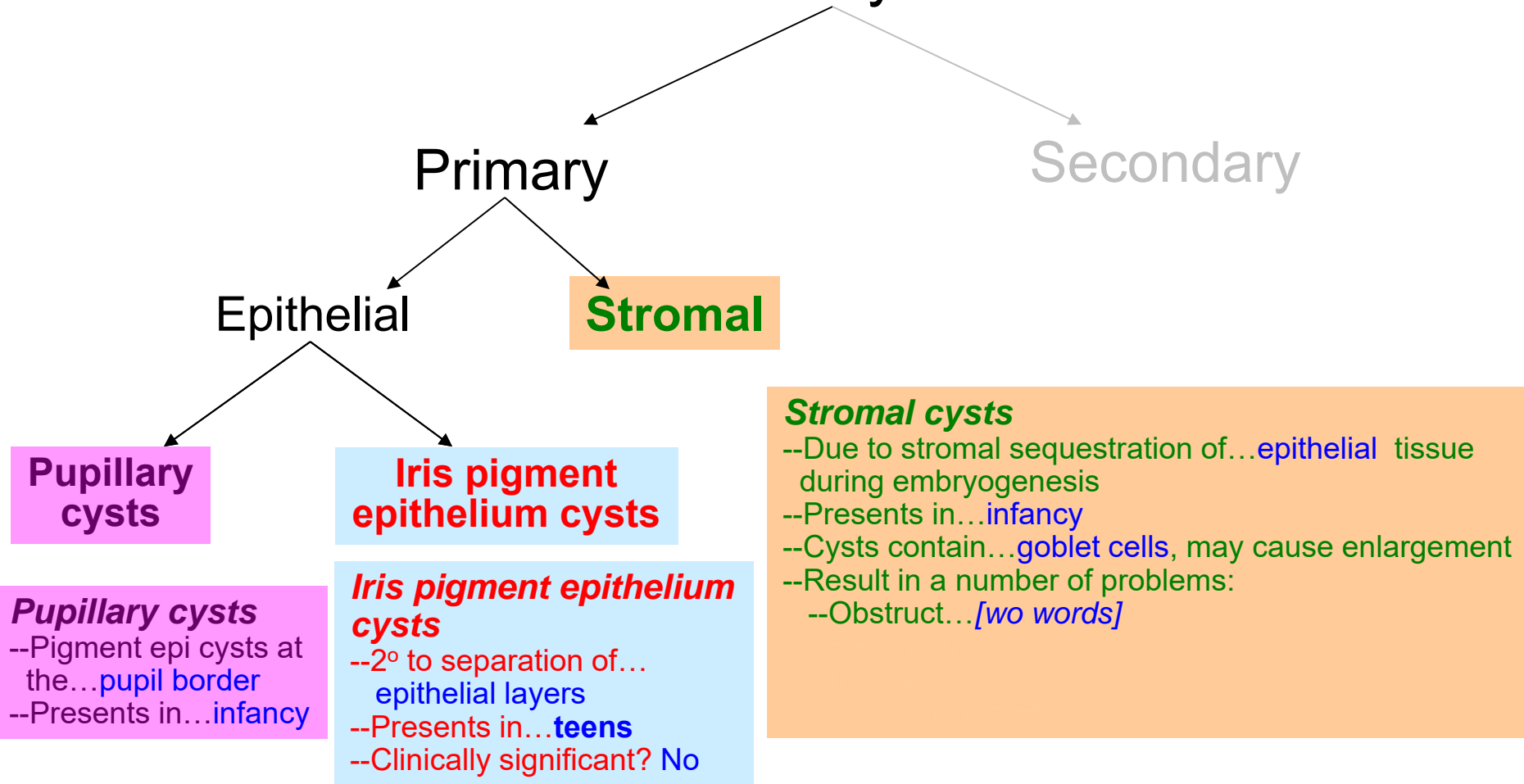
Pediatric Iris Cysts





Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Stromal

Stromal cysts

- Due to stromal sequestration of...epithelial tissue during embryogenesis
- Presents in...infancy
- Cysts contain...goblet cells, may cause enlargement
- Result in a number of problems:
 - Obstruct...[wo words]

Pupillary cysts

- Pigment epi cysts at the...pupil border
- Presents in...infancy

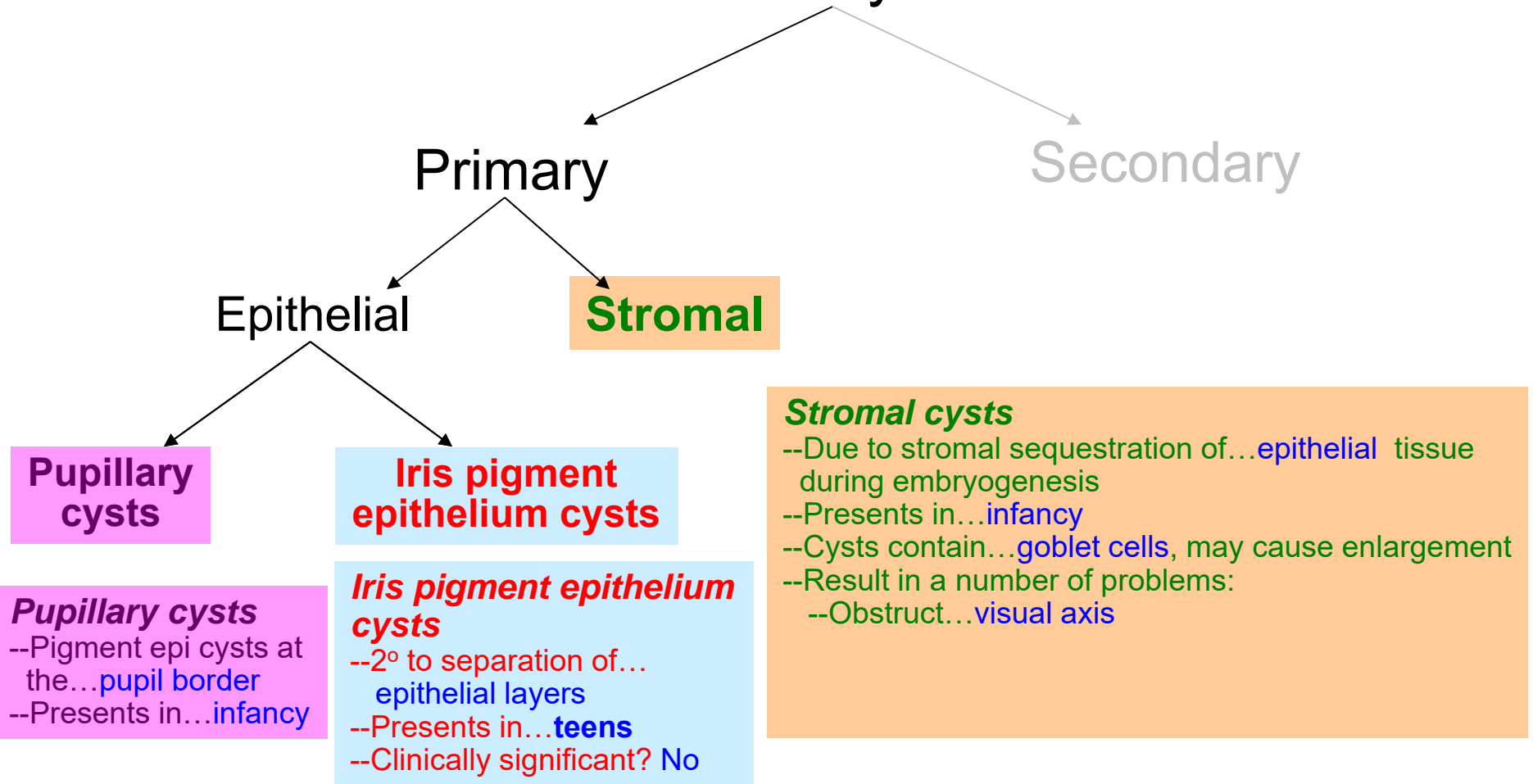
Iris pigment epithelium cysts

- 2° to separation of...epithelial layers
- Presents in...teens
- Clinically significant? No



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Stromal

Stromal cysts

- Due to stromal sequestration of...epithelial tissue during embryogenesis
- Presents in...infancy
- Cysts contain...goblet cells, may cause enlargement
- Result in a number of problems:
 - Obstruct...visual axis

Pupillary cysts

- Pigment epi cysts at the...pupil border
- Presents in...infancy

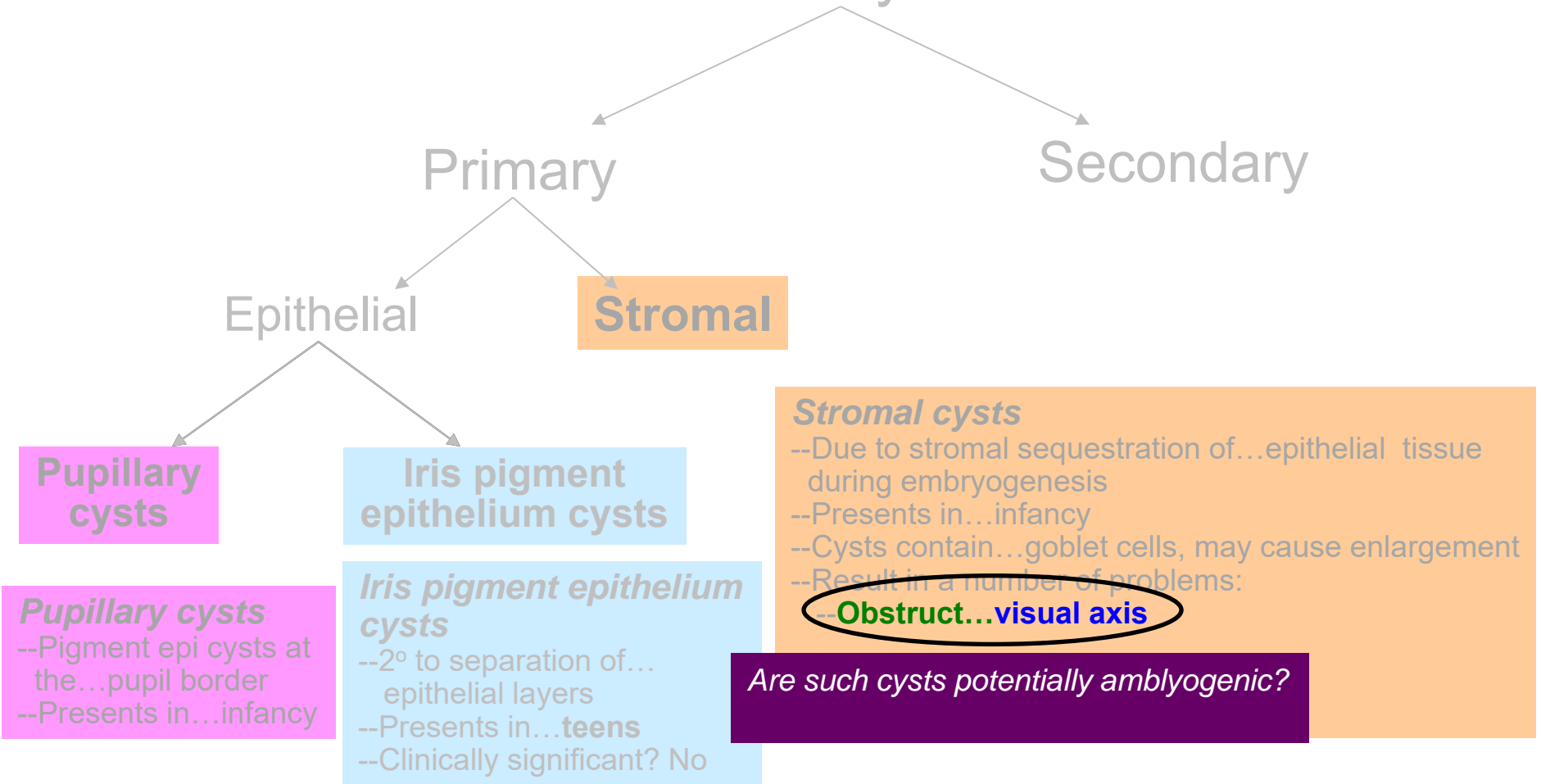
Iris pigment epithelium cysts

- 2° to separation of...epithelial layers
- Presents in...teens
- Clinically significant? No



Pediatric Iris Cysts and Nodules

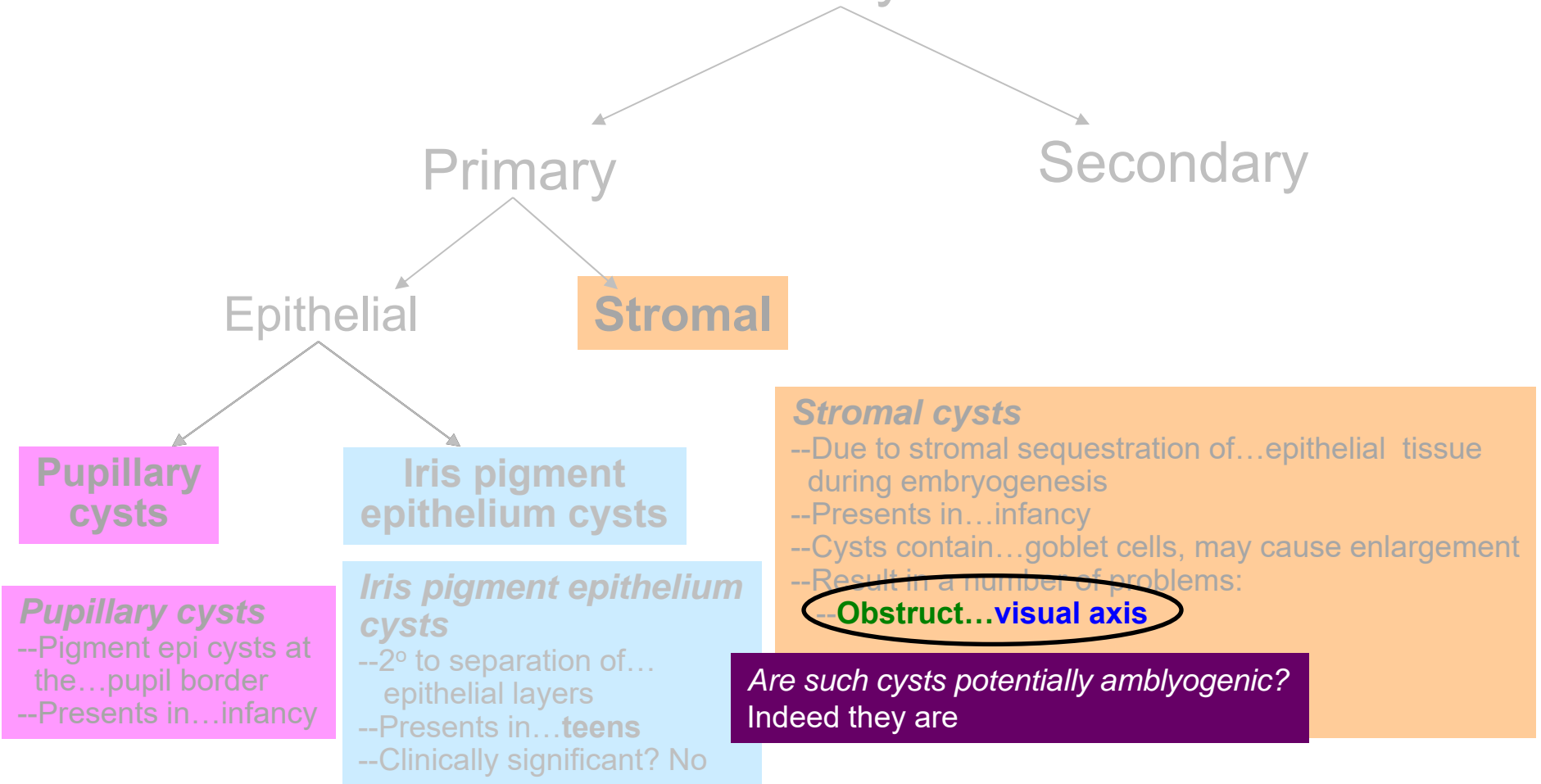
Pediatric Iris Cysts



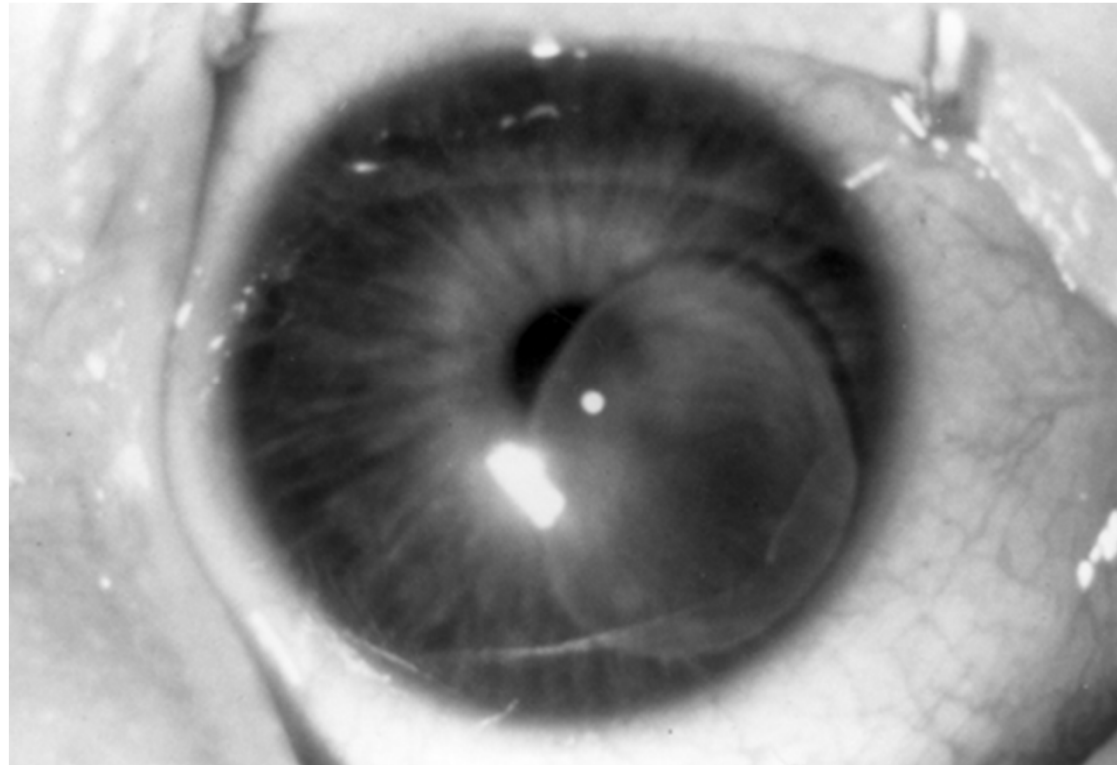


Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Pediatric Iris Cysts and Nodules

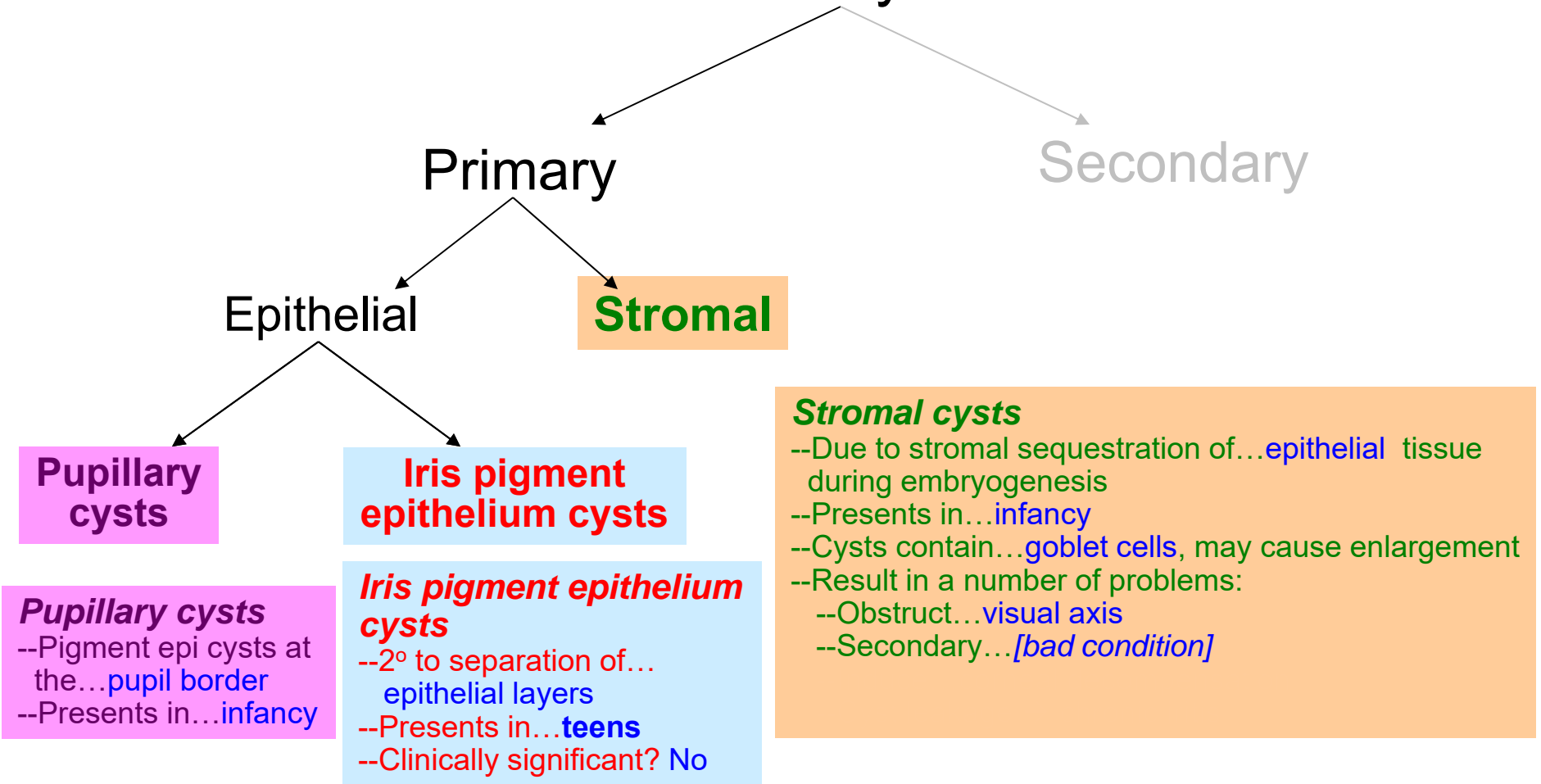


Stromal cyst in an infant, obstructing visual axis



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Stromal

Stromal cysts
 --Due to stromal sequestration of...epithelial tissue during embryogenesis
 --Presents in...infancy
 --Cysts contain...goblet cells, may cause enlargement
 --Result in a number of problems:
 --Obstruct...visual axis
 --Secondary...[bad condition]

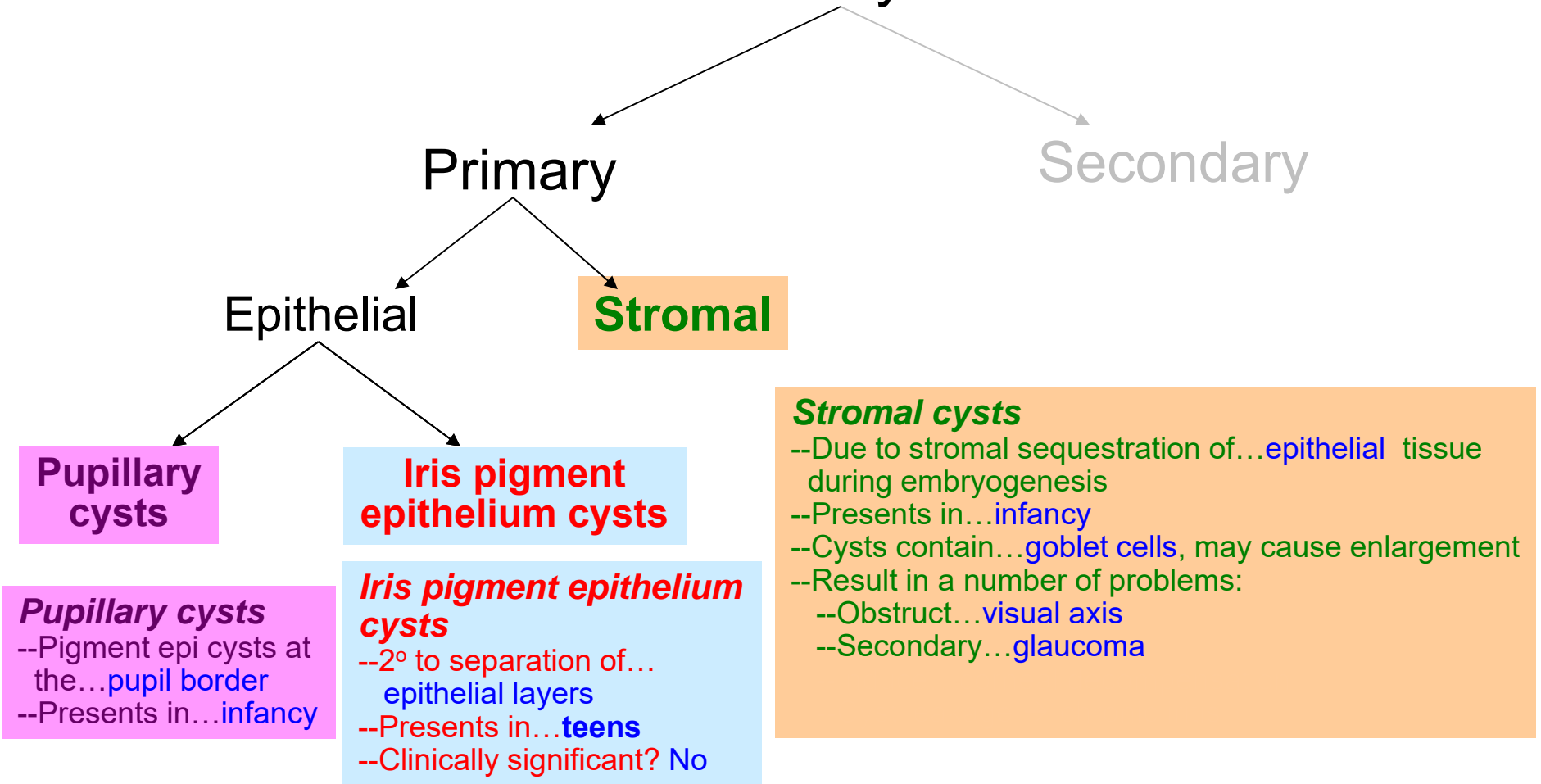
Pupillary cysts
 --Pigment epi cysts at the...pupil border
 --Presents in...infancy

Iris pigment epithelium cysts
 --2° to separation of...epithelial layers
 --Presents in...teens
 --Clinically significant? No



Pediatric Iris Cysts and Nodules

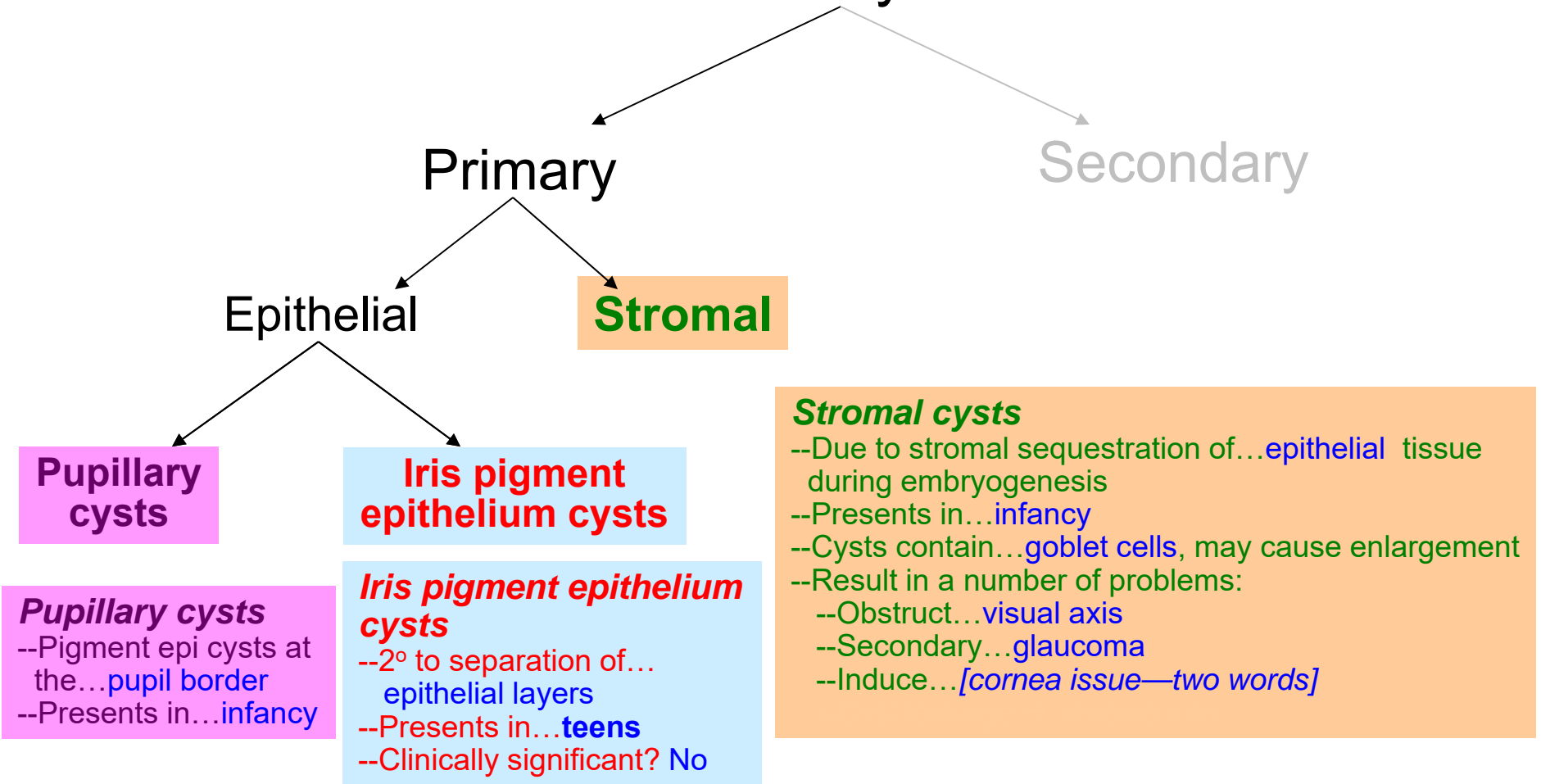
Pediatric Iris Cysts





Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Pupillary cysts
 --Pigment epi cysts at the...pupil border
 --Presents in...infancy

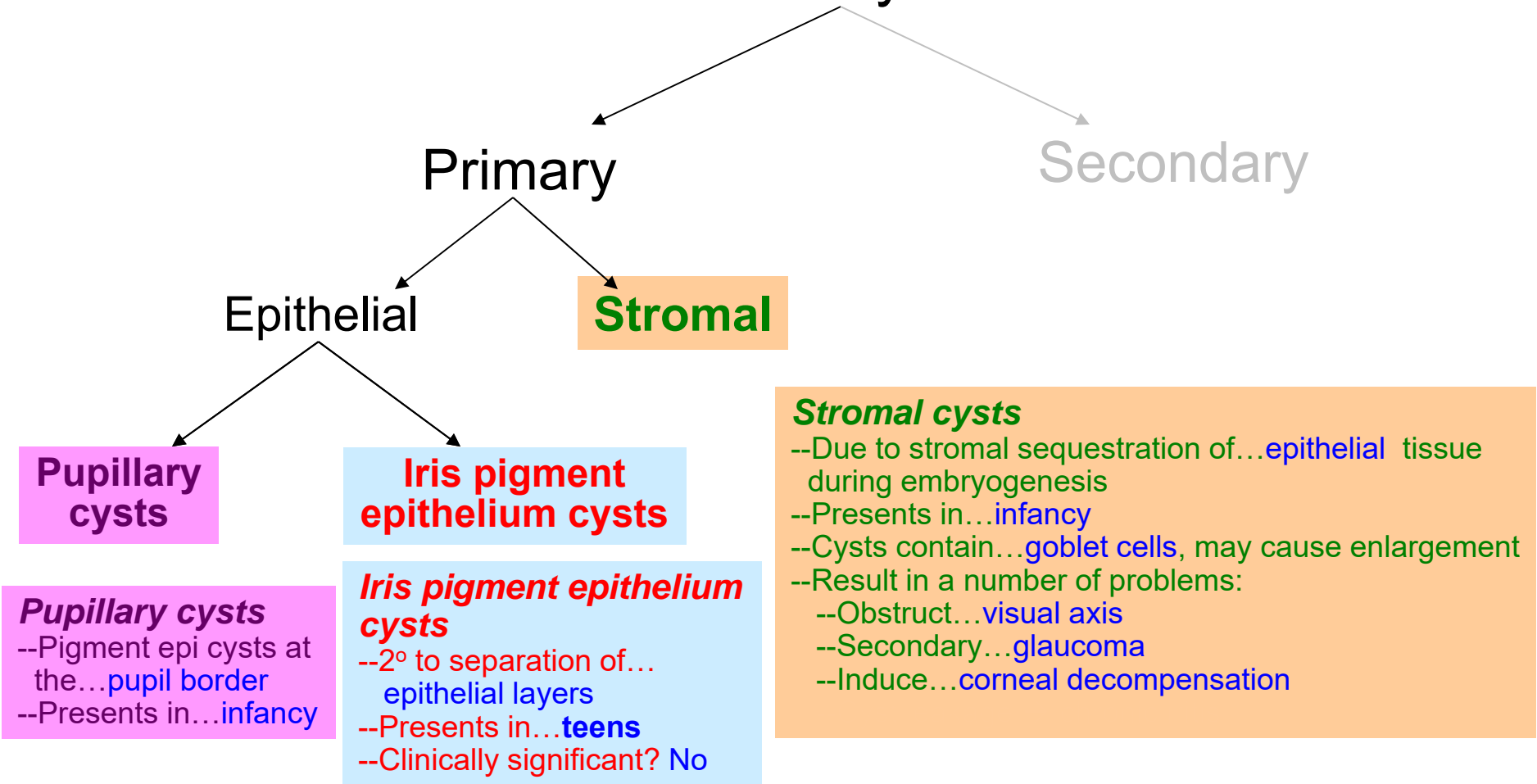
Iris pigment epithelium cysts
 --2° to separation of...epithelial layers
 --Presents in...teens
 --Clinically significant? No

Stromal cysts
 --Due to stromal sequestration of...epithelial tissue during embryogenesis
 --Presents in...infancy
 --Cysts contain...goblet cells, may cause enlargement
 --Result in a number of problems:
 --Obstruct...visual axis
 --Secondary...glaucoma
 --Induce...[cornea issue—two words]



Pediatric Iris Cysts and Nodules

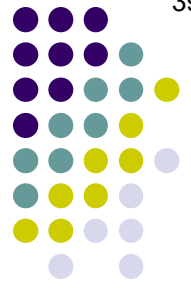
Pediatric Iris Cysts



Pupillary cysts
 --Pigment epi cysts at the...pupil border
 --Presents in...infancy

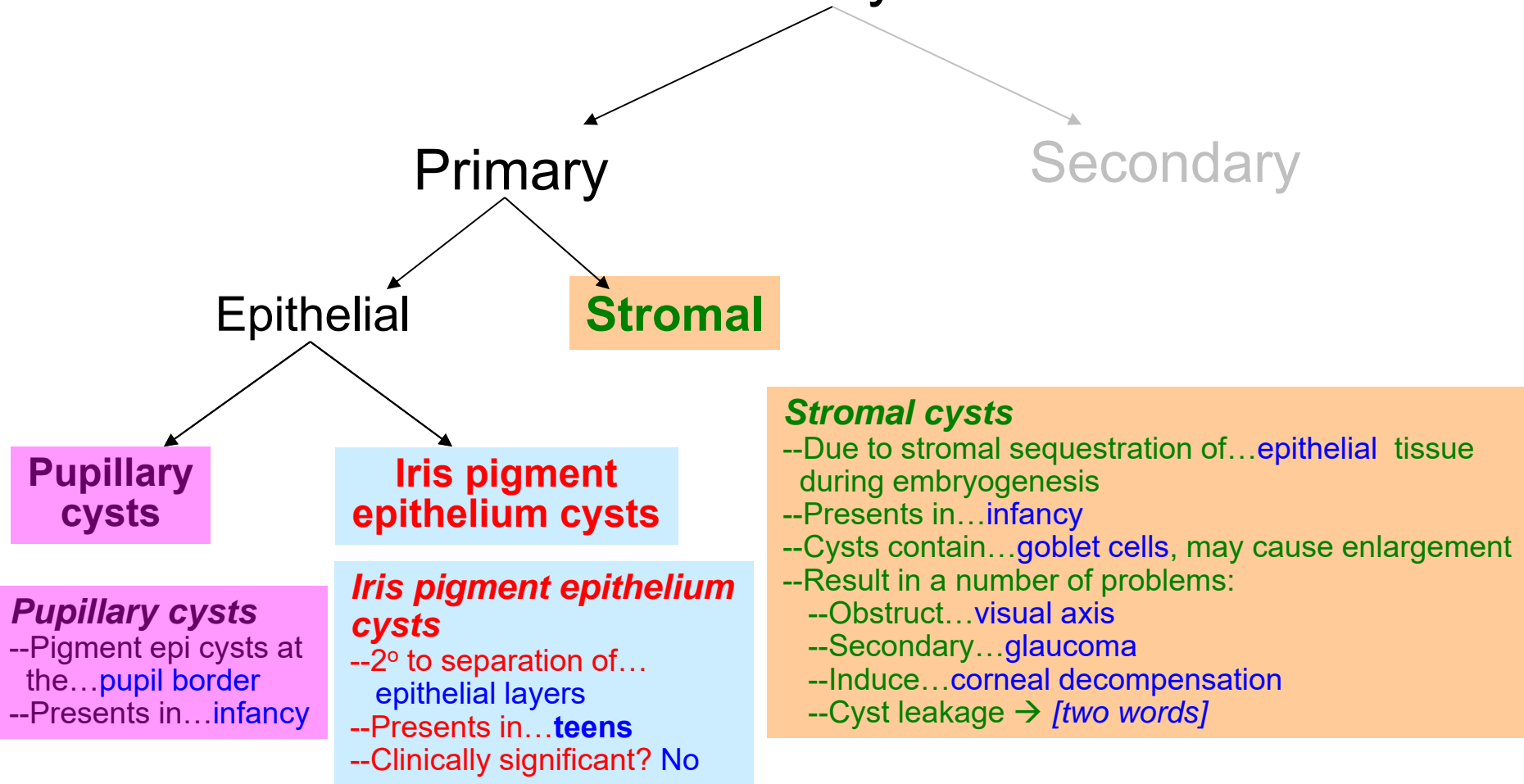
Iris pigment epithelium cysts
 --2° to separation of...epithelial layers
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 --Clinically significant? No

Stromal cysts
 --Due to stromal sequestration of...epithelial tissue during embryogenesis
 --Presents in...infancy
 --Cysts contain...goblet cells, may cause enlargement
 --Result in a number of problems:
 --Obstruct...visual axis
 --Secondary...glaucoma
 --Induce...corneal decompensation



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Pupillary cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

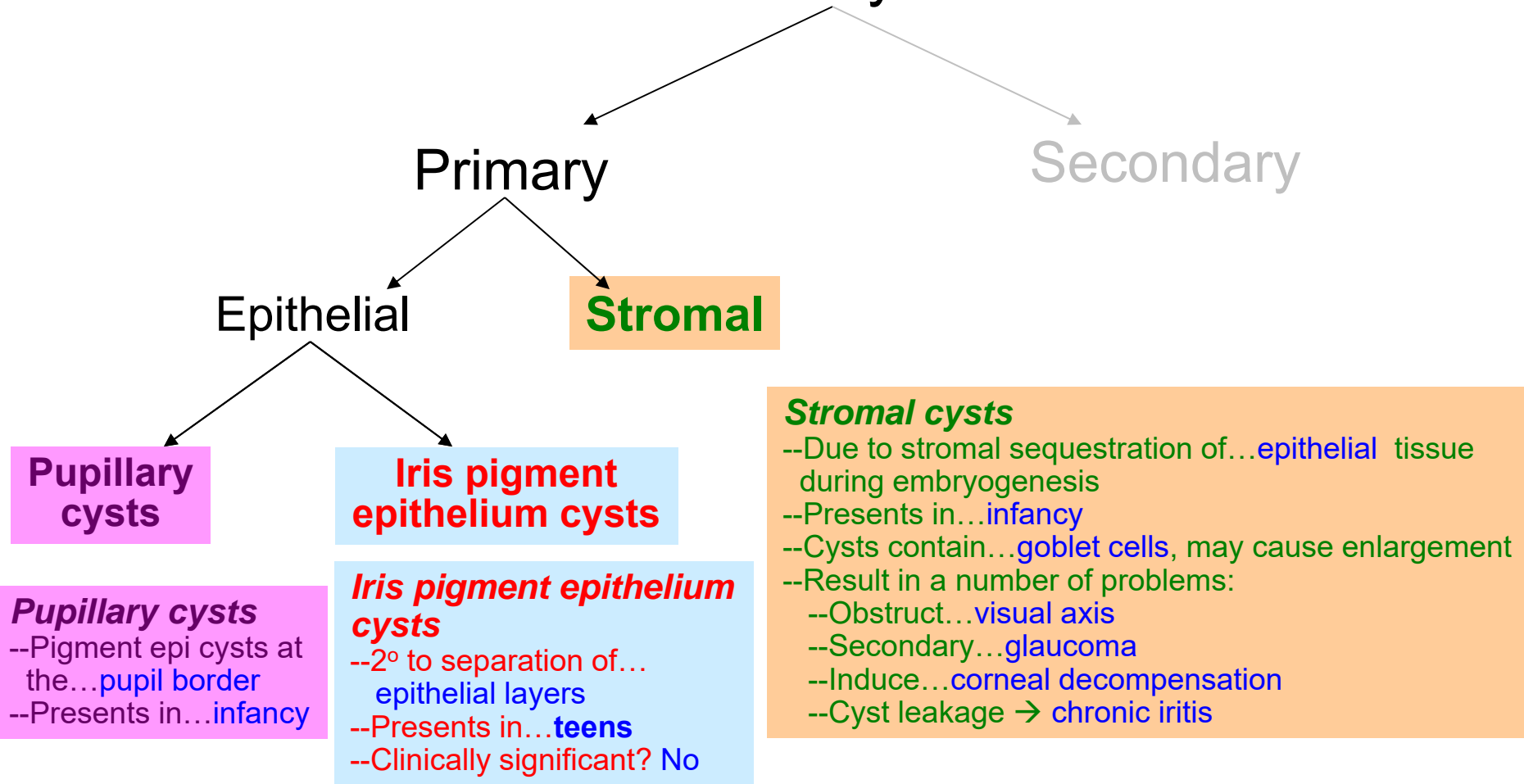
Stromal

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
--Obstruct...visual axis
--Secondary...glaucoma
--Induce...corneal decompensation
--Cyst leakage → [two words]



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Pupillary cysts

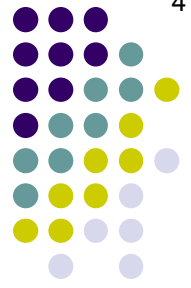
Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
--Obstruct...visual axis
--Secondary...glaucoma
--Induce...corneal decompensation
--Cyst leakage → chronic iritis



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Primary

Secondary

Epithelial

Stromal

Pupillary cysts

Iris pigment epithelium cysts

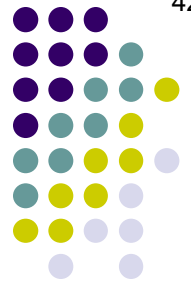
Because of these issues, stromal cysts require treatment in most cases!

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts
--Cysts contain... goblet cells... may cause enlargement

- Obstruct...visual axis
- Secondary...glaucoma
- Induce...corneal decompensation
- Cyst leakage → chronic iritis



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

How are stromal cysts treated?

Pupillary cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

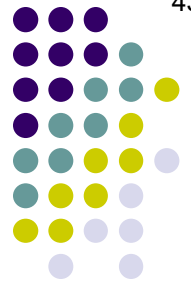
Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts

Because of these issues, stromal cysts require treatment in most cases!

- Obstruct...visual axis
- Secondary...glaucoma
- Induce...corneal decompensation
- Cyst leakage → chronic iritis

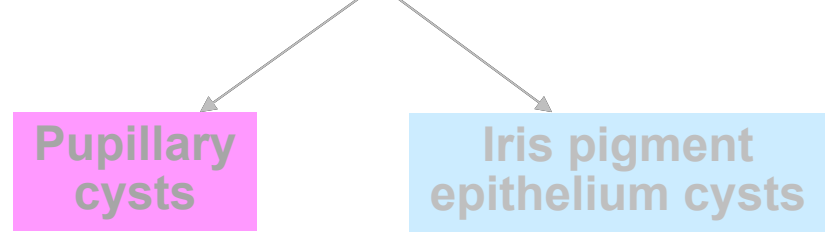
--Cysts contain... goblet cells... may cause enlargement



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

How are stromal cysts treated?
There are multiple approaches; each has its advantages and disadvantages. **Photodisruption** of the cyst is simple and noninvasive, but risks inciting iritis and/or glaucoma when the pro-inflammatory contents of the cyst are released into the AC.

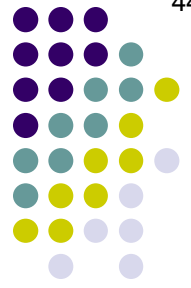


Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of... epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts
Because of these issues, stromal cysts require treatment in most cases!
--Cysts contain... goblet cells... may cause enlargement

- Obstruct...visual axis
- Secondary...glaucoma
- Induce...corneal decompensation
- Cyst leakage → chronic iritis



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

How are stromal cysts treated?
There are multiple approaches; each has its advantages and disadvantages. **Photodisruption** of the cyst is simple and noninvasive, but risks inciting iritis and/or glaucoma when the pro-inflammatory contents of the cyst are released into the AC. Because of this, the *Peds* book recommends **surgical excision**. Other sources recommend the less-invasive **aspiration with absolute alcohol injection**.

Pupillary cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

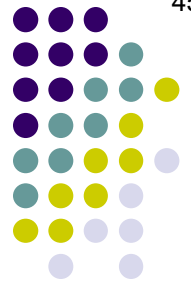
Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts

Because of these issues, stromal cysts require treatment in most cases!

- Obstruct...visual axis
- Secondary...glaucoma
- Induce...corneal decompensation
- Cyst leakage → chronic iritis

-Cysts contain... goblet cells, may cause enlargement

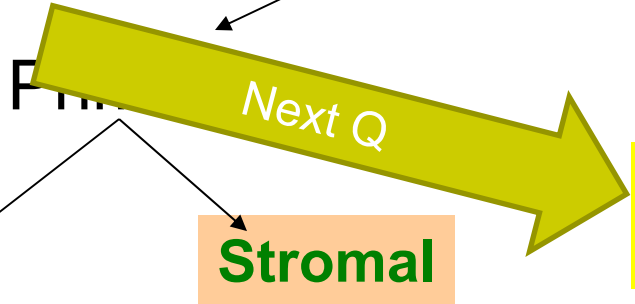


Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Secondary cysts
--Usually due to...[drug]



Epithelial

Stromal

Pupillary cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
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--Result in a number of problems:
--Obstruct...visual axis
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--Induce...corneal decompensation
--Cyst leakage → chronic iritis



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

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Secondary cysts
 --Usually due to...echothiophate

Primary

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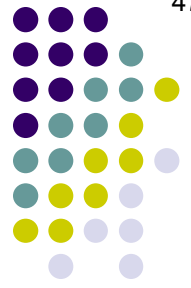
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Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

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What is the trade name for echothiophate?

Primary

Stromal

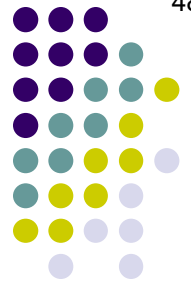
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Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Secondary cysts
--Usually due to...echothiophate

What is the trade name for echothiophate?
Phospholine

Primary

Stromal

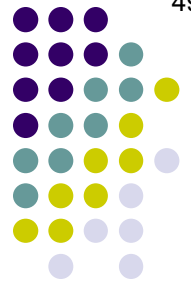
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What is the trade name for echothiophate?
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What sort of drug is it?

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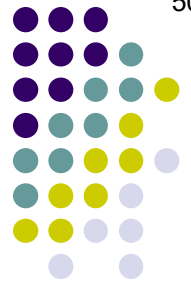
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What sort of drug is it?
A parasympathomimetic

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What is its mechanism of action?

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Pediatric Iris Cysts

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What is the trade name for echothiophate?
Phospholine

What sort of drug is it?
A **parasympathomimetic**

What is its mechanism of action?
It is an **acetylcholinesterase inhibitor**

Primary

Stromal

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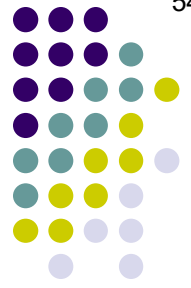
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What ocular condition is echothiophate used to treat?



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

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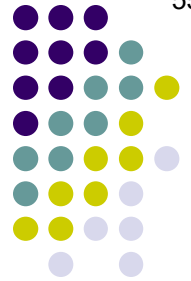
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Phospholine

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What is its mechanism of action?
It is an **acetylcholinesterase inhibitor**

What ocular condition is echothiophate used to treat?
Accommodative ET

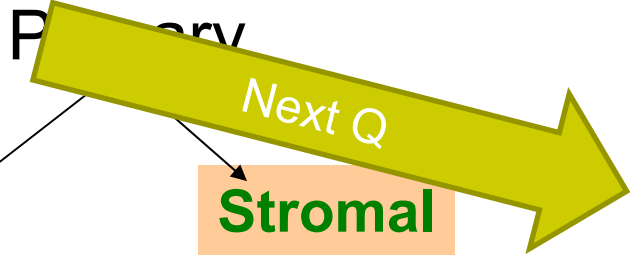


Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

Secondary cysts
 --Usually due to...**echothiophate**
 --Can prophylax with...[drug]



Epithelial

Pupillary cysts

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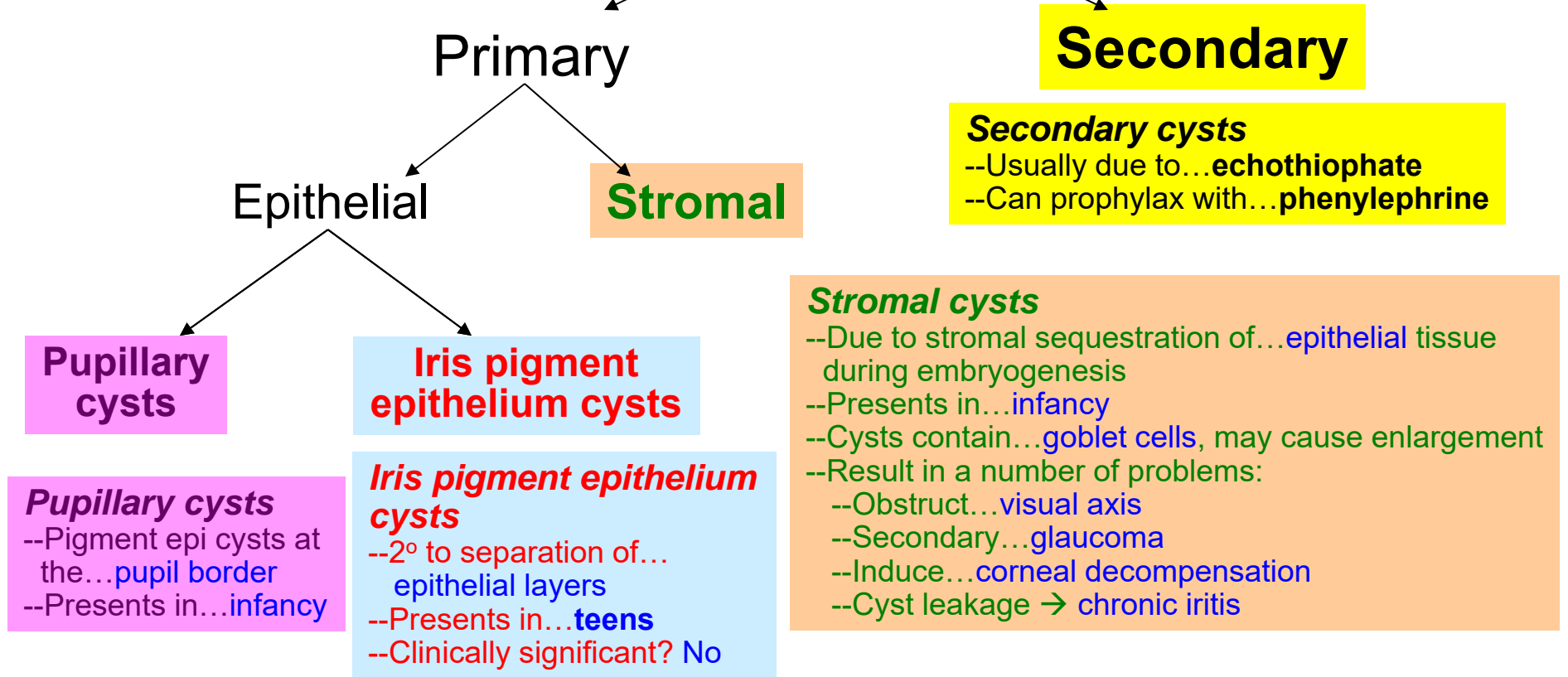
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 --Result in a number of problems:
 --Obstruct...**visual axis**
 --Secondary...**glaucoma**
 --Induce...**corneal decompensation**
 --Cyst leakage → **chronic iritis**

Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts



Pediatric Iris Cysts and Nodules



Pediatric Iris Cysts

Secondary

What about secondary cysts after penetrating trauma?

St

Stromal cysts

- Due to stromal sequestration of...epithelial tissue during embryogenesis
- Presents in...infancy
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Primary

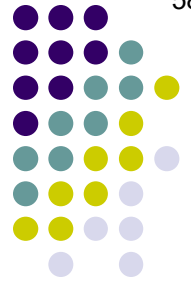
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- Iris pigment epithelium cysts**
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 - Presents in...teens
 - Clinically significant? No



Pediatric Iris Cysts and Nodules

Pediatric Iris Cysts

Secondary

What about secondary cysts after penetrating trauma? That's certainly a thing, **but**—these are more likely to be **epithelial downgrowth cysts** resulting from the implantation of surface epithelium on the iris

Primary

Epithelial

Stromal

Pupillary cysts

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 --Pigment epi cysts at the...pupil border
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Pediatric Iris Cysts and Nodules



Epithelial downgrowth cyst secondary to perforating corneal trauma in a child



Q

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules found in the pediatric population
 -
 -
 -

Mnemonic forthcoming...

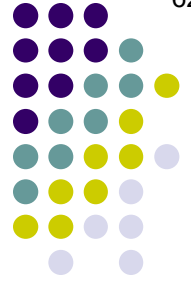


Q

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules found in the pediatric population
 - **L**
 - **B**
 - **J**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



A

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules found in the pediatric population
 - **L**isch nodules
 - **B**rushfield spots
 - **J**XG nodules

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Q

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- Name 3 types of pediatric population
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Lisch nodules are most strongly associated with what congenital condition?

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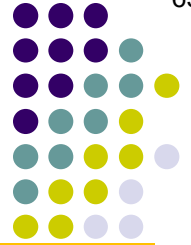
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Q

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

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Q

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Briefly, what is a phakomatosis?

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Q/A

Pediatric Iris Cysts and Nodules

- Name 3 types of pediatric popular

Lisch nodules are most strongly associated with what congenital condition?

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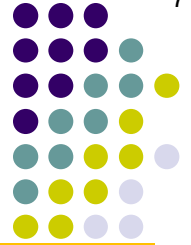
- Brushfi

Briefly, what is a phakomatosis?

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

- JXG no

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A

Pediatric Iris Cysts and Nodules

- Name 3 types of pediatric popula
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- **Brushfi**
- **JXG no**

Lisch nodules are most strongly associated with what congenital condition?

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Q

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

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As **neuro-**

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As **neuro-oculocutaneous** syndromes

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Q

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 - *B*rushfield spots
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Lisch nodules are most strongly associated with what congenital condition?

Neurofibromatosis type 1 (NF1)

Is it associated with NF2?

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What is the prevalence of Lisch nodules in NF1?

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The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

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Are Lisch nodules clinically significant?
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Dome shaped **tan(nish)** masses

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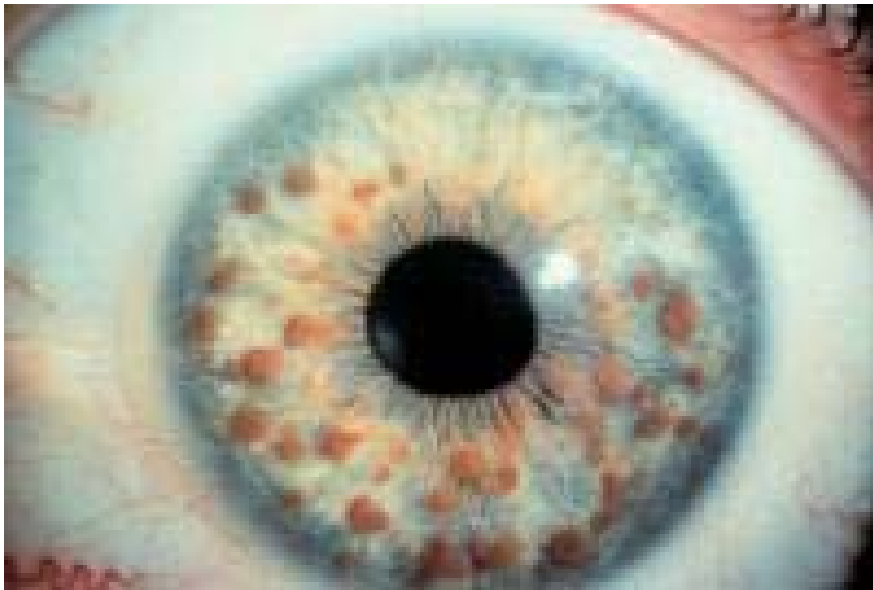
Lisch nodules are **lighter** than the rest of the iris when the iris in question is dark, but **darker** than the rest when the iris is light

Are Lisch nodules clinically significant?

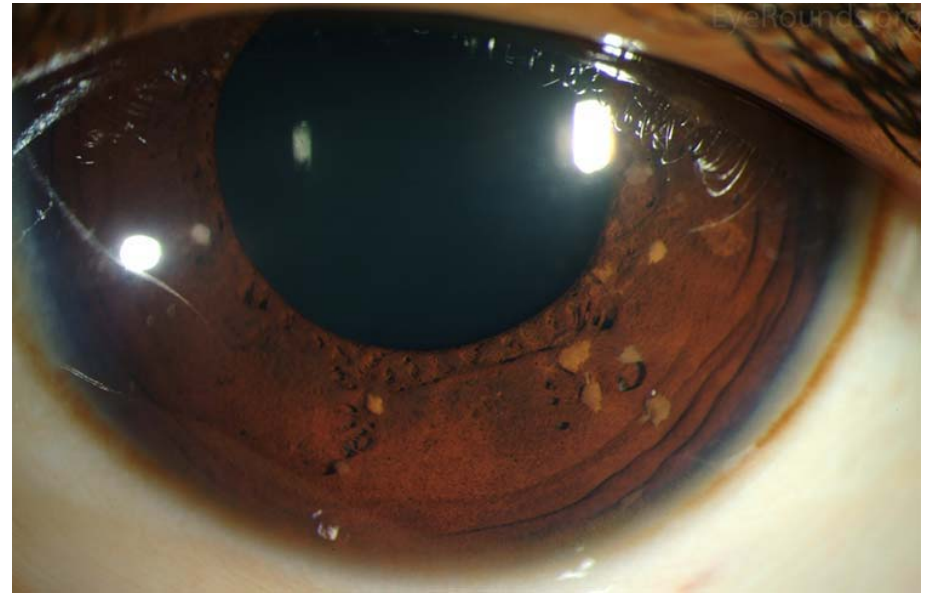
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Pediatric Iris Cysts and Nodules

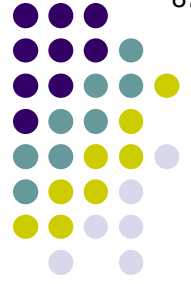


Darker on light iris



Lighter on dark iris

Lisch nodules



Q

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules found in the pediatric population
 - Lisch nodules** *Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?*
 - Brushfield spots
 - JXG nodules
 - Iris one word

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



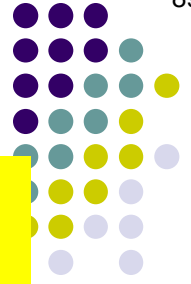
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Pediatric Iris Cysts and Nodules

Mammillations? Aren't those a CNS thingamajig?

- Name 3 types of iris nodules in the pediatric population

- **Lisch nodules**

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Q/A

Pediatric Iris Cysts and Nodules

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You're thinking of the , paired structures that are part of the limbic system

- Name 3 types of iris nodules in pediatric population

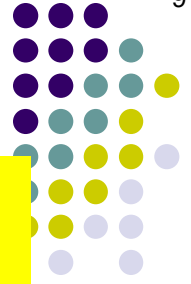
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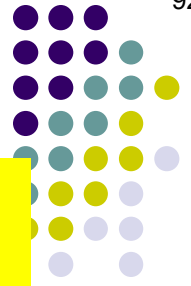
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Pediatric Iris Cysts and Nodules

Mammillations? Aren't those a CNS thingamajig?
You're thinking of the **mammillary bodies**, paired structures that are part of the limbic system

- Name 3 types of iris nodules in pediatric population
 - **Lisch nodules**
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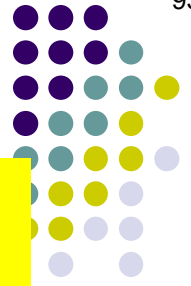
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OK, then what are iris mammillations?

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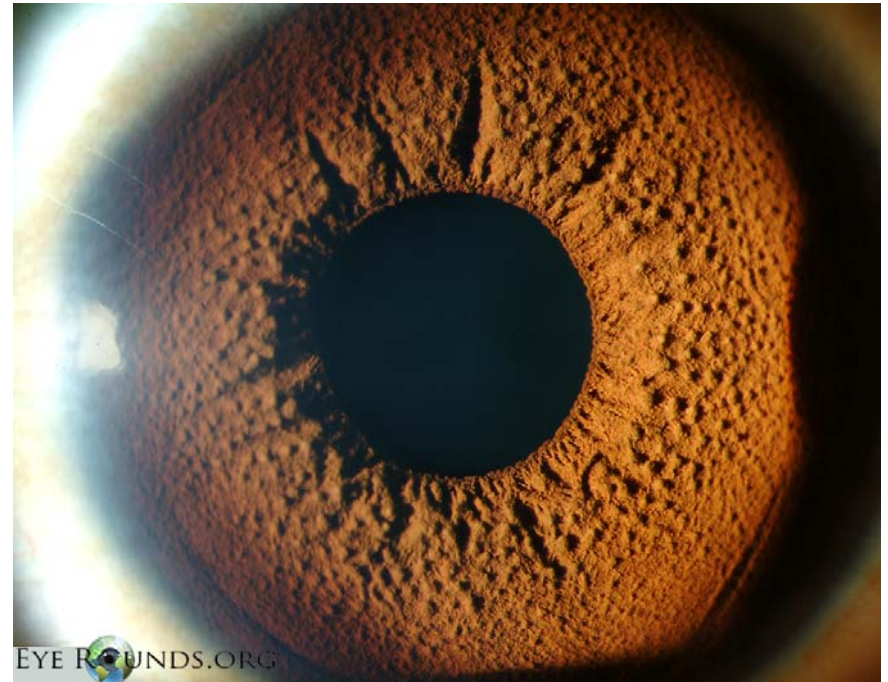
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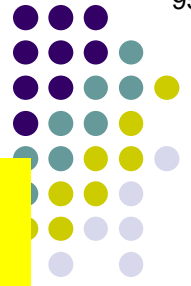
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Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface

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Pediatric Iris Cysts and Nodules



Iris mammilations



Q

Pediatric Iris Cysts and Nodules

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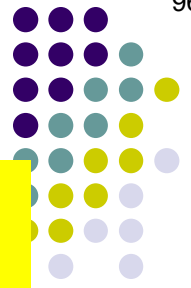
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Are they unilateral, or bilateral?

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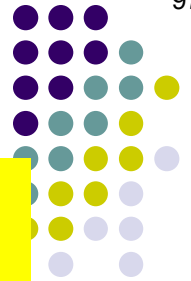
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Are they unilateral, or bilateral?
Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out

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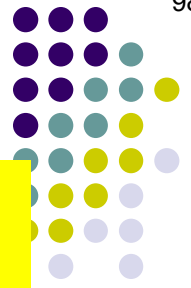
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Are they associated with NF1?

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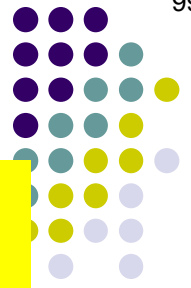
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Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out

Are they associated with NF1?
Yes (albeit not nearly as strongly as Lisch nodules)

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



Pediatric Iris Cysts and Nodules

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● Name 3 types of iris nodules in the pediatric population

'Tiny pigmented iris nodules associated with NF1'--given this, how on earth are you supposed to differentiate between the two?

- **Lisch nodules**
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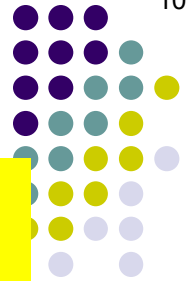
Tiny pigmented nodules which, when present, are

'Tiny pigmented iris nodules associated with NF1'--given this, how on earth are you supposed to differentiate between the two? By appearance. Iris mammillations are always the same color as the rest of the iris. In contrast, Lisch nodules are lighter than the rest of the iris when the iris in question is dark, but darker than the rest when the iris is light.

Are they **associated with NF1**?

Yes (albeit not nearly as strongly as Lisch nodules)

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



Q

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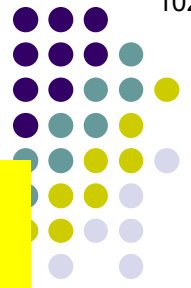
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Are they associated with NF1?
Usually u...
enough t...
In addition to NF1, iris mammillations have another important association. What is it?

Are they associated with NF1 and...
Yes (albeit not nearly as strong)

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



A

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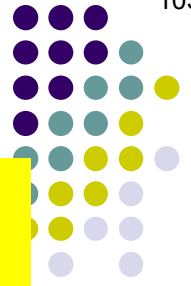
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Oculodermal melanocytosis, aka **nevus of Ota**

Are they associated with NF1 and...oculodermal melanocytosis
Yes (albeit not nearly as strong)

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



Q

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules in pediatric population

- Lisch nodules

What is oculodermal melanocytosis/nevus of Ota?

- JXG nodules

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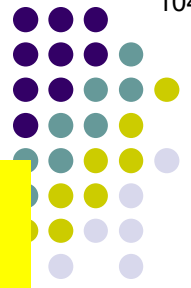
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Yes (albeit not nearly as strong) they **associated with NF1** and...**oculodermal melanocytosis**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...



Pediatric Iris Cysts and Nodules

A

- Name 3 types of iris nodules in pediatric population

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- Lisch nodules

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin

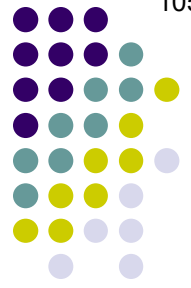
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Pediatric Iris Cysts and Nodules



Oculodermal melanocytosis (nevus of Ota)

Pediatric Iris Cysts and Nodules

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What is
Ocular
Ipsilateral periorbital skin

For more on Nevus of Ota, see slide-set O9

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Q

Pediatric Iris Cysts and Nodules

- Name 3 types of iris nodules found in the pediatric population

- *L*isch nodules
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Brushfield spots are most strongly associated with what congenital condition?

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Pediatric Iris Cysts and Nodules

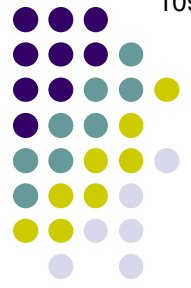
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Pediatric Iris Cysts and Nodules



Brushfield spots



Q

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At least 90%

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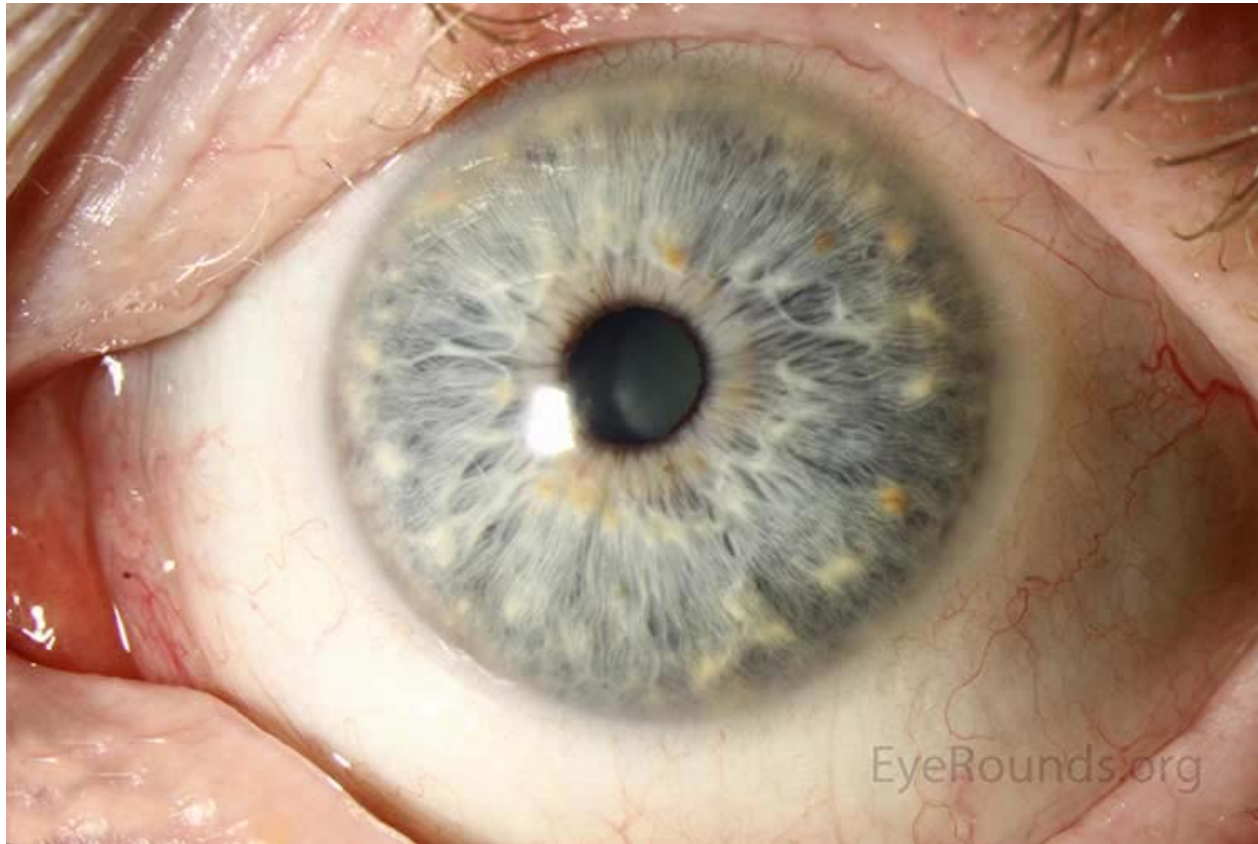
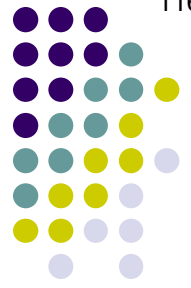
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Pediatric Iris Cysts and Nodules



Wolflin nodules



Q

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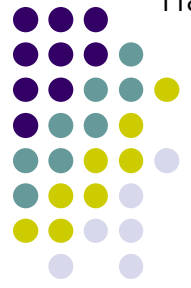
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Q/A

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Yes--they're estimated to be present in about
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- Brushfield spots**

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Brushfield spots are most strongly associated with what congenital condition?

Down syndrome

What is the prevalence of Brushfield spots in the Down population?

At least 90%

Are Wolflin nodules a common finding?

Yes--they're estimated to be present in about 25% of the population

of Brushfield spots?

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

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Q

Pedia

What does JXG stand for?

- Name 3 types of pediatric population
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A

Pedia

What does JXG stand for?
Juvenile xanthogranuloma

- Name 3 types of pediatric population
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 - **JXG nodules**

Think of president Lyndon Baines Johnson

Q

Pedia

What does *JXG* stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?

- Name 3 types of pediatric population
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- *B*rushfield spots
- ***JXG*** nodules

Think of president Lyndon Baines Johnson

A

Pedia

What does *JXG* stand for?
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In three words, what sort of condition is it?
It is a **nonneoplastic histiocytic proliferation**

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Pedia

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Histiocytes are also known as...?

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Histiocytes are also known as...?
Macrophages

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Pediatric

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What does derive mean here? How does a monocyte become a histiocyte?

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Pediatric

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Macrophages

Histiocytes derive from what type of cell?
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What does derive mean here? How does a monocyte become a histiocyte?
Monocytes are found within the vasculature. When a monocyte leaves the circulation and settles down within tissue, it becomes (ie, is re-classified) as a histiocyte.

Think of president Lyndon Baines Johnson

Q

Pedia

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
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What are the two hallmarks of JXG histology?
The presence of...
The presence of...

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- Name 3 types of pediatric population
 - Lisch nodules
 - Brushfield spots
 - **JXG nodules**

What is a giant cell?

Think of president Lyndon Baines Johnson

Q/A

Pedia

What does JXG stand for?
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In three words, what sort of condition is it?
It is a **nonneoplastic histiocytic proliferation**

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What is a giant cell?

It is an aggregate (the formal term is) of histiocytes

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It is an aggregate (the formal term is *syncytium*) of histiocytes.
Put simply, it is a bunch of histiocytes that have glommed together.

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What is the histologic hallmark of a giant cell?

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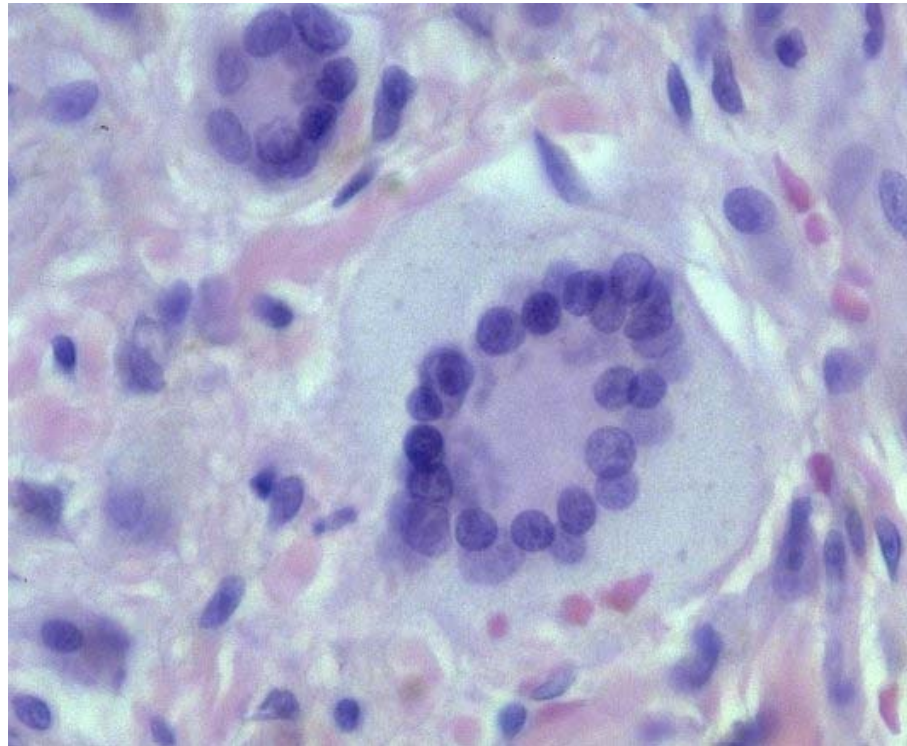
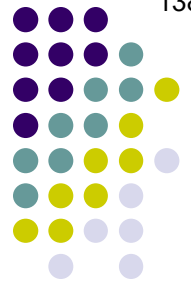
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What is the histologic hallmark of a giant cell?

It is *multinucleated*—the myriad nuclei of the involved histiocytes are all visible within it

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Pediatric Iris Cysts and Nodules



JXG; Touton giant cells

Q

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It's a reference to the macrophage's appearance. They look foamy because they are full of 

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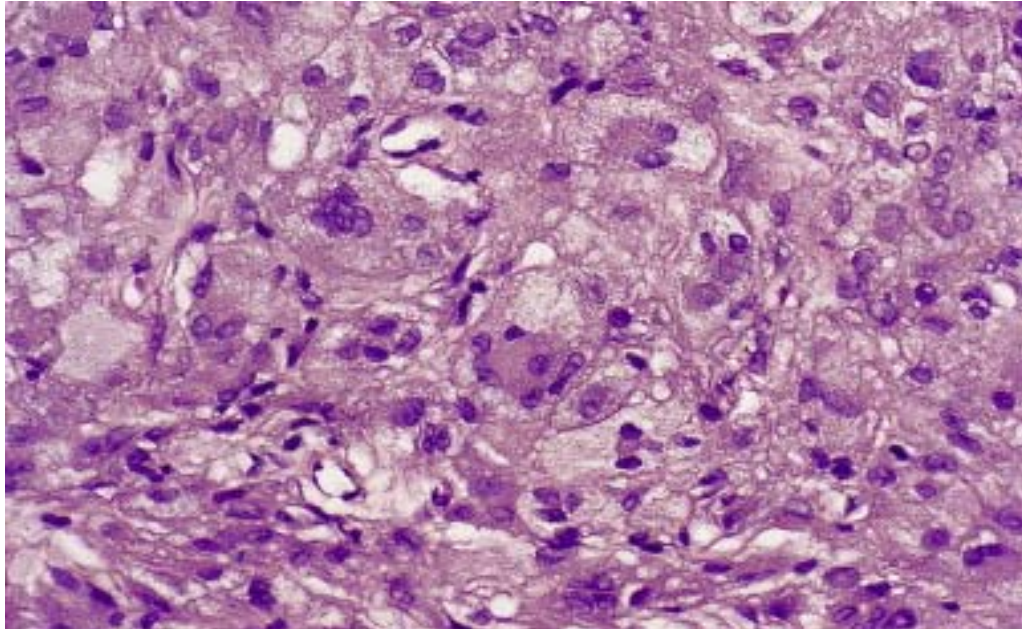
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Pediatric Iris Cysts and Nodules



JXG: Foamy macrophages

Q

Pediatric

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Pediatric Iris Cysts and Nodules



JXG: Skin papules. The orangish color is classic

Q

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Pediatric Iris Cysts and Nodules



JXG: Iris lesion



Q

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In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a 'masquerade syndrome' in peds uveitis
--If enough are present, they will cause heterochromia iridis

- Name 3 types of pediatric population
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Pediatric Iris Cysts and Nodules



JXG: Spontaneous hyphema

Q

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Should they be removed surgically?
Only if the glaucoma is uncontrollable

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What is the natural history of the disease?

- Name 3 types of pediatric population
- *Lisch nodules*
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What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

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- Lis *(Intraocular)*
- Br *(Bilateral)*

For more on JXG, see slide-set P3

For more on histiocytosis, set K20

- JXG nodules

Unilaterally

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Q

Pediatric Iris Cysts and Nodules

- *Congenital iris ectropion syndrome* consists of ectropion uveae plus...

bilateral vs unilateral



A

Pediatric Iris Cysts and Nodules

- *Congenital iris ectropion syndrome* consists of **unilateral** ectropion uveae plus...



Q

Pediatric Iris Cysts and Nodules

- *Congenital iris ectropion syndrome* consists of **unilateral** ectropion uveae plus...
 - ...iris **surface** abnormality--is it *glassy and cryptless*, or *extra corrugated*?

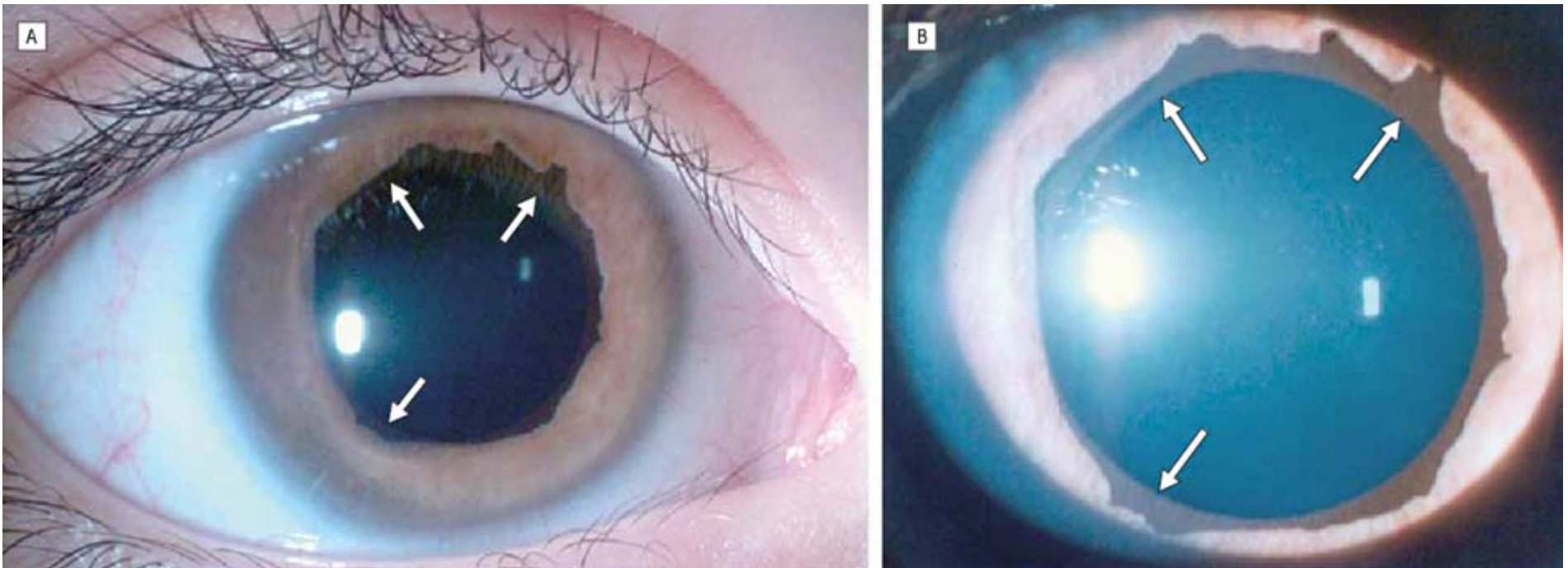
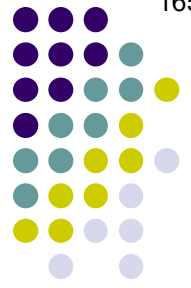


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Pediatric Iris Cysts and Nodules

- *Congenital iris ectropion syndrome* consists of **unilateral** ectropion uveae plus...
 - ...iris **surface** abnormality--is it *glassy and cryptless*, or *extra corrugated*? **Glassy and cryptless**

Pediatric Iris Cysts and Nodules



Congenital ectropion uveae syndrome. A, Wide pupil and striking smoothness of the anterior surface of the iris, absence of the circular contraction furrows, and absence of radial folds are shown. (Arrows indicate the iris ectropion.) B, Higher magnification and illumination of the iris to better differentiate the pupillary margin from the iris ectropion (arrows)



Q

Pediatric Iris Cysts and Nodules

- *Congenital iris ectropion syndrome* consists of **unilateral** ectropion uveae plus...
 - ...iris **surface** abnormality--is it *glassy and cryptless*, or *extra corrugated*? **Glassy** and **cryptless**
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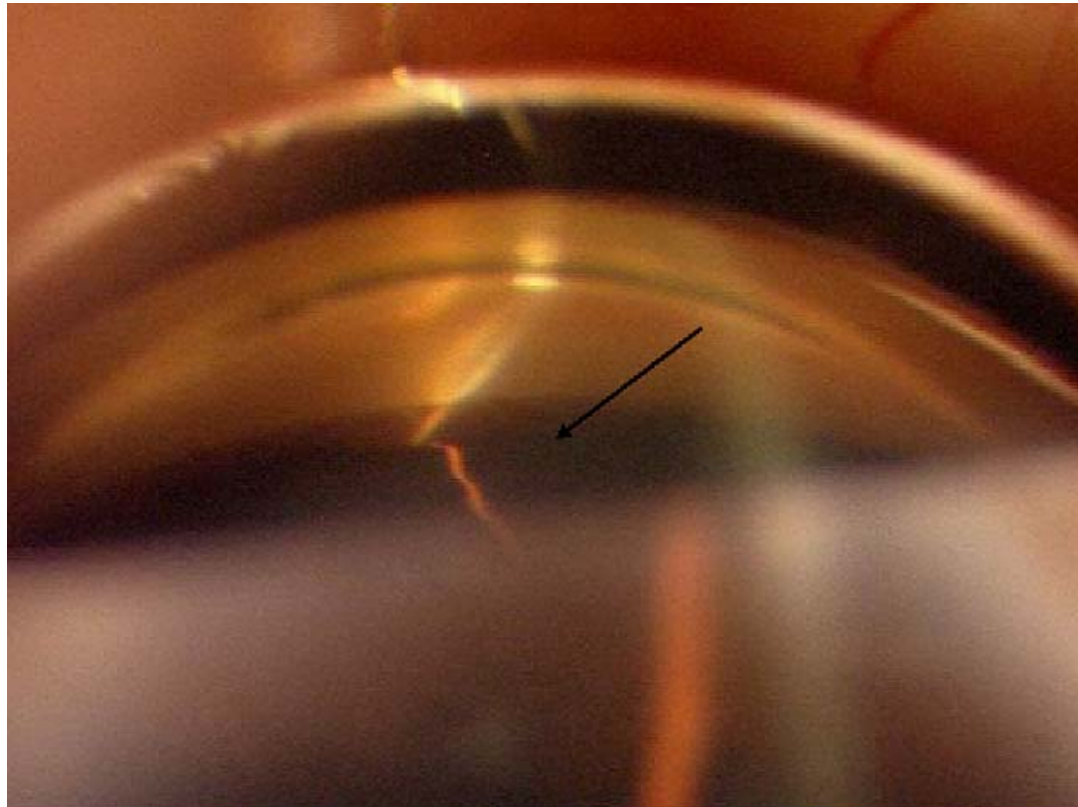


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Pediatric Iris Cysts and Nodules



Congenital ectropion uveae syndrome: Gonioscopy depicting anterior insertion of iris (black arrow)



Q

Pediatric Iris Cysts and Nodules

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 - ...**dysgenesis** of what anterior structure?



A

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 - ...**dysgenesis** of what anterior structure? **The TM** (*trabecular meshwork*)



Q

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- *What does **ectropion uveae** refer to, and why is the term technically a misnomer?*



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 - ...*dysgenesis* of what anterior structure? The **TM**
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- **What does *ectropion uveae* refer to, and why is the term technically a misnomer? *Ectropion uveae* refers to the presence of posterior pigmented iris epithelium on the anterior surface of the iris. It is a misnomer because the posterior pigmented epithelium derives from *neuroectoderm*, not uvea.**

(I guess the term *ectropion neuroectodermeae* didn't scan well...)