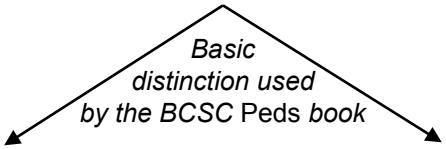
# Developmental Abnormalities of the Cornea

*Basic distinction used by the BCSC* Peds *book*

Abnormalities of Corneal [redacted] or [redacted]

Abnormalities of Corneal [redacted]

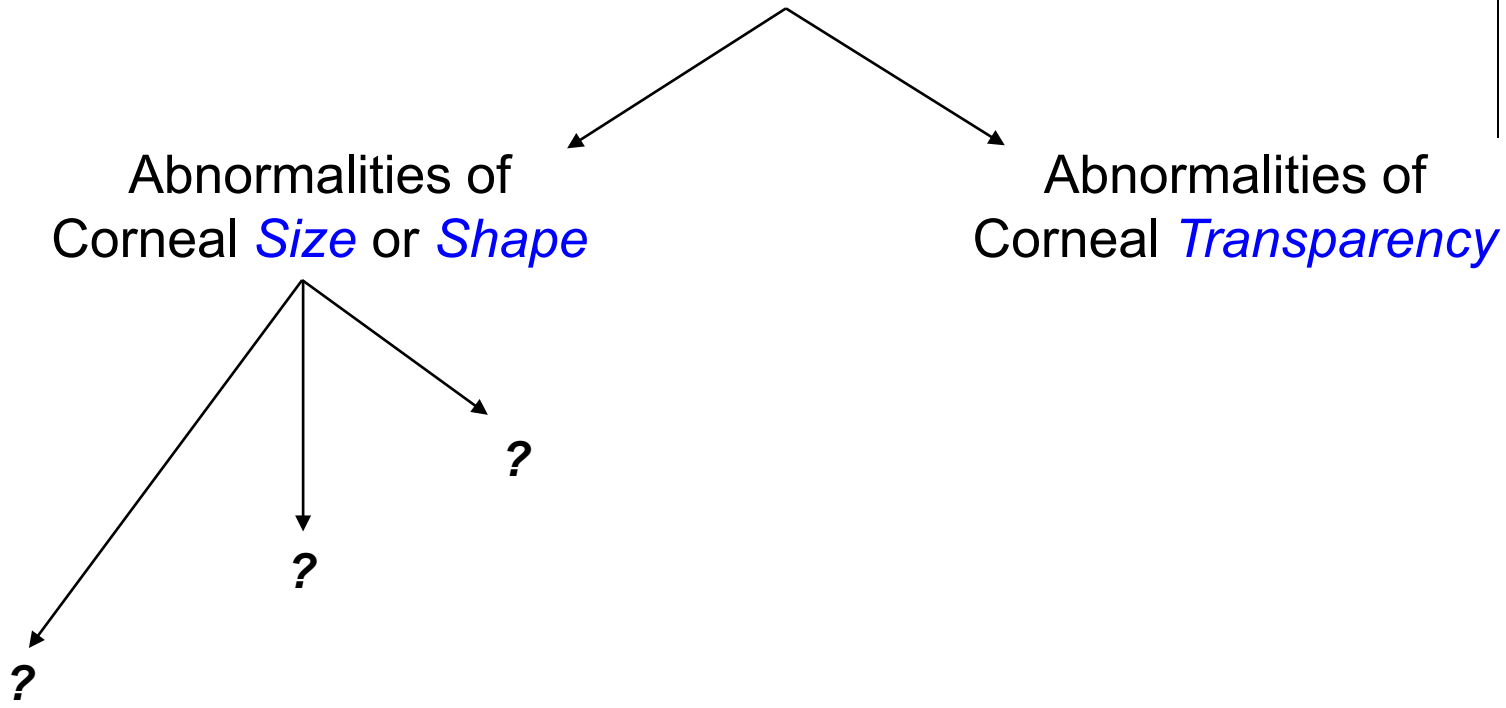
# Developmental Abnormalities of the Cornea




Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

# Developmental Abnormalities of the Cornea



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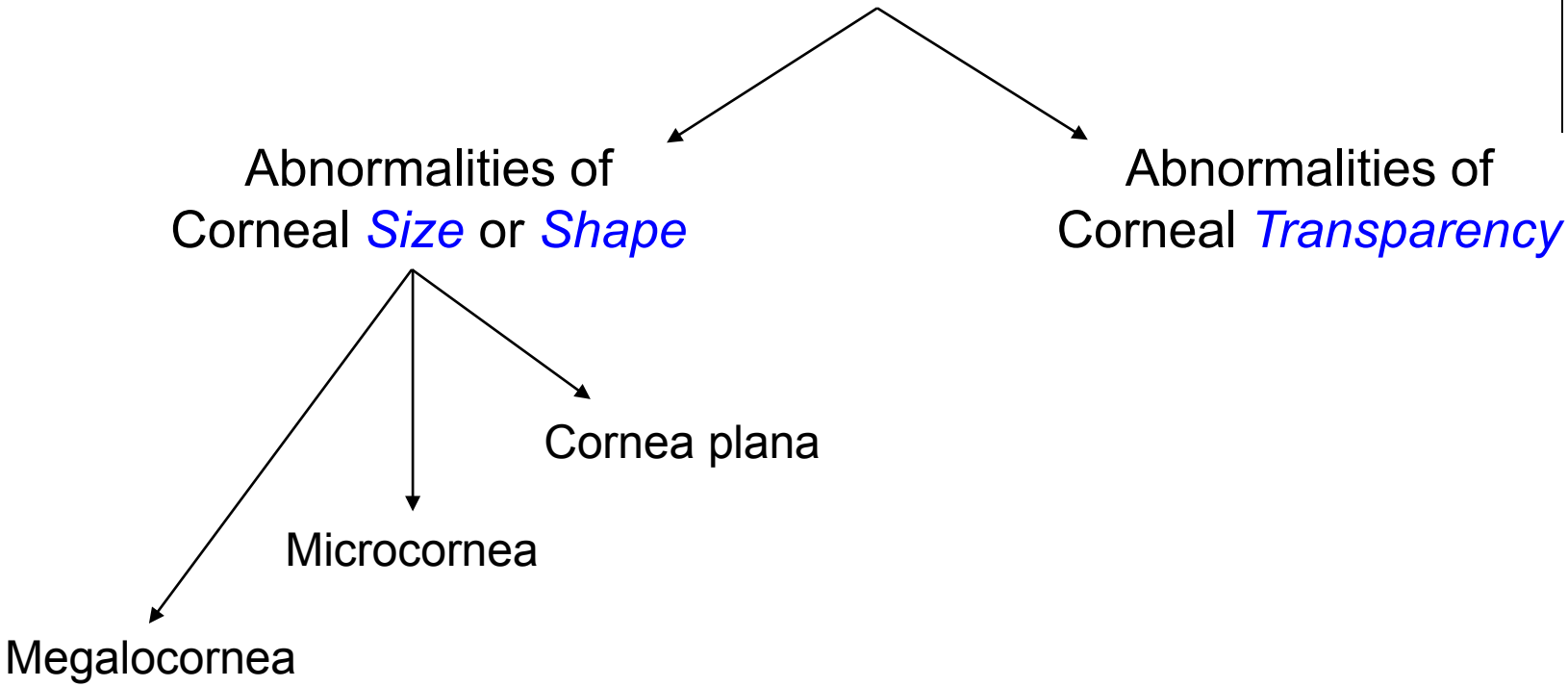


# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

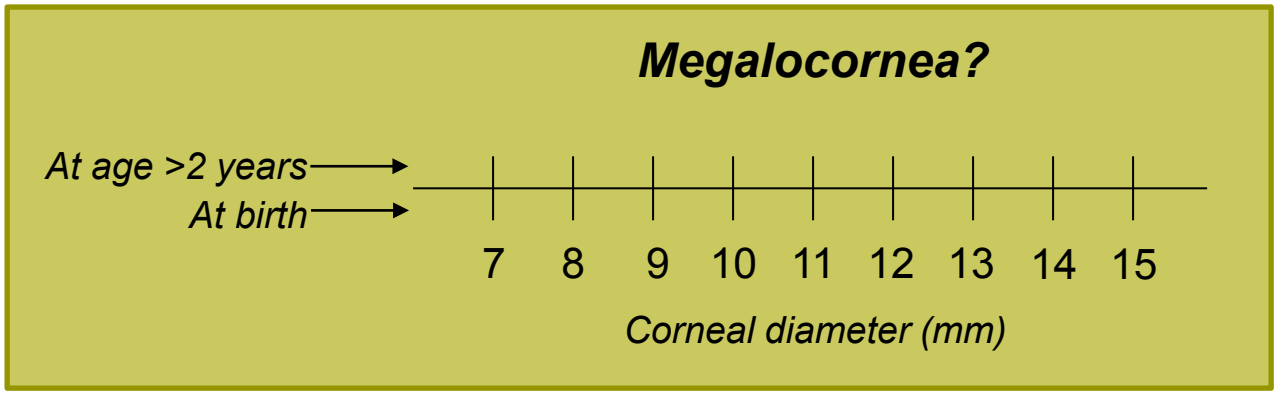
Abnormalities of Corneal *Transparency*

Cornea plana

Microcornea

**Megalocornea**

*What is the definition of megalocornea, ie, how 'megalo' does it have to be?*



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Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

Cornea plana

Microcornea

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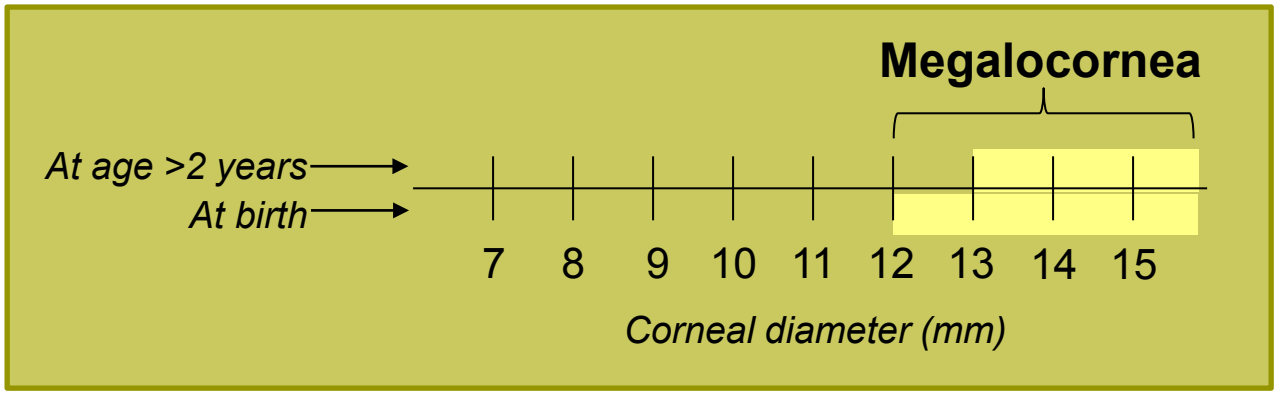
*What is the definition of megalocornea, ie, how 'megalo' does it have to be?*
Corneal diameter > 12 mm at birth, or > 13 mm at age 2 years or older



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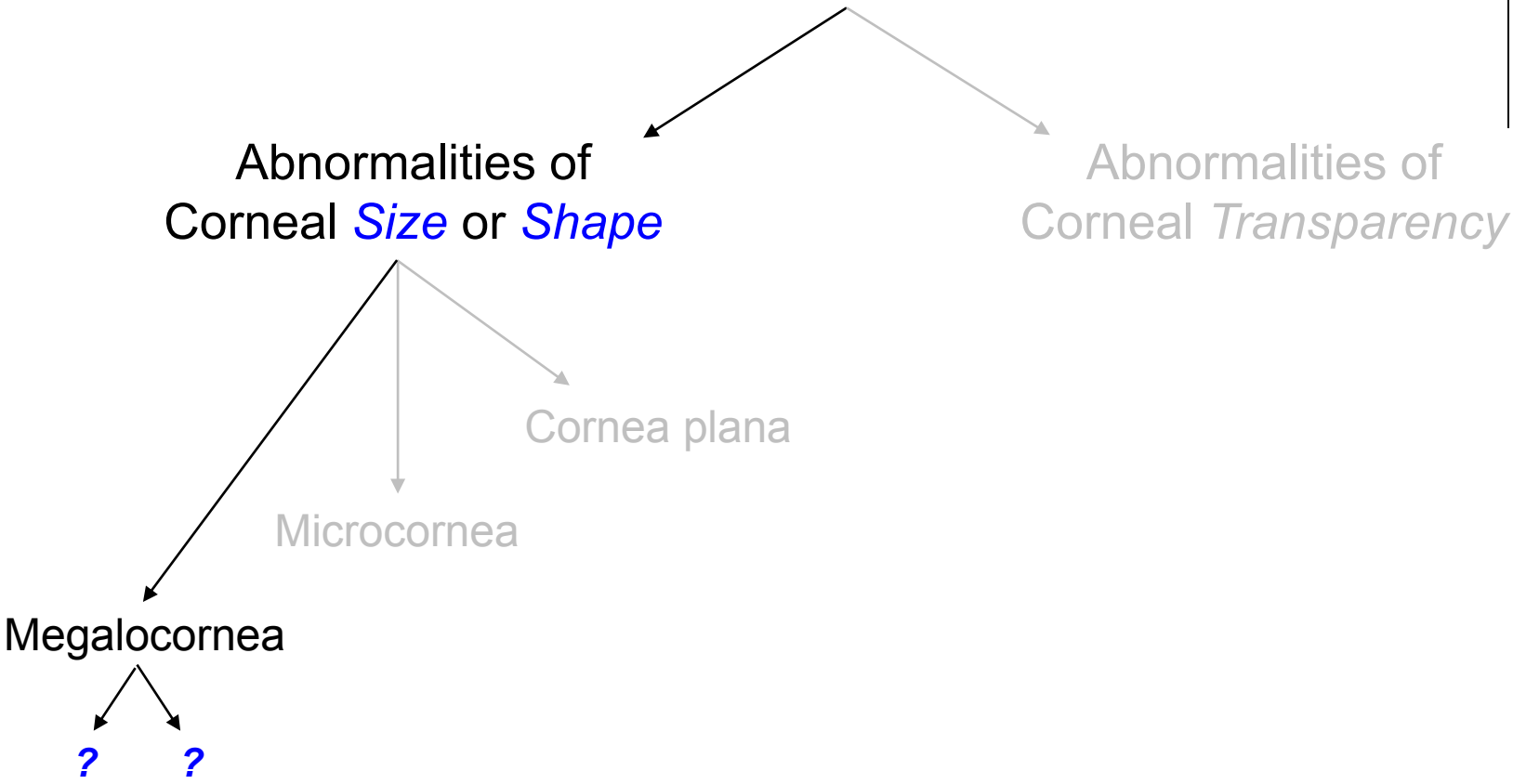

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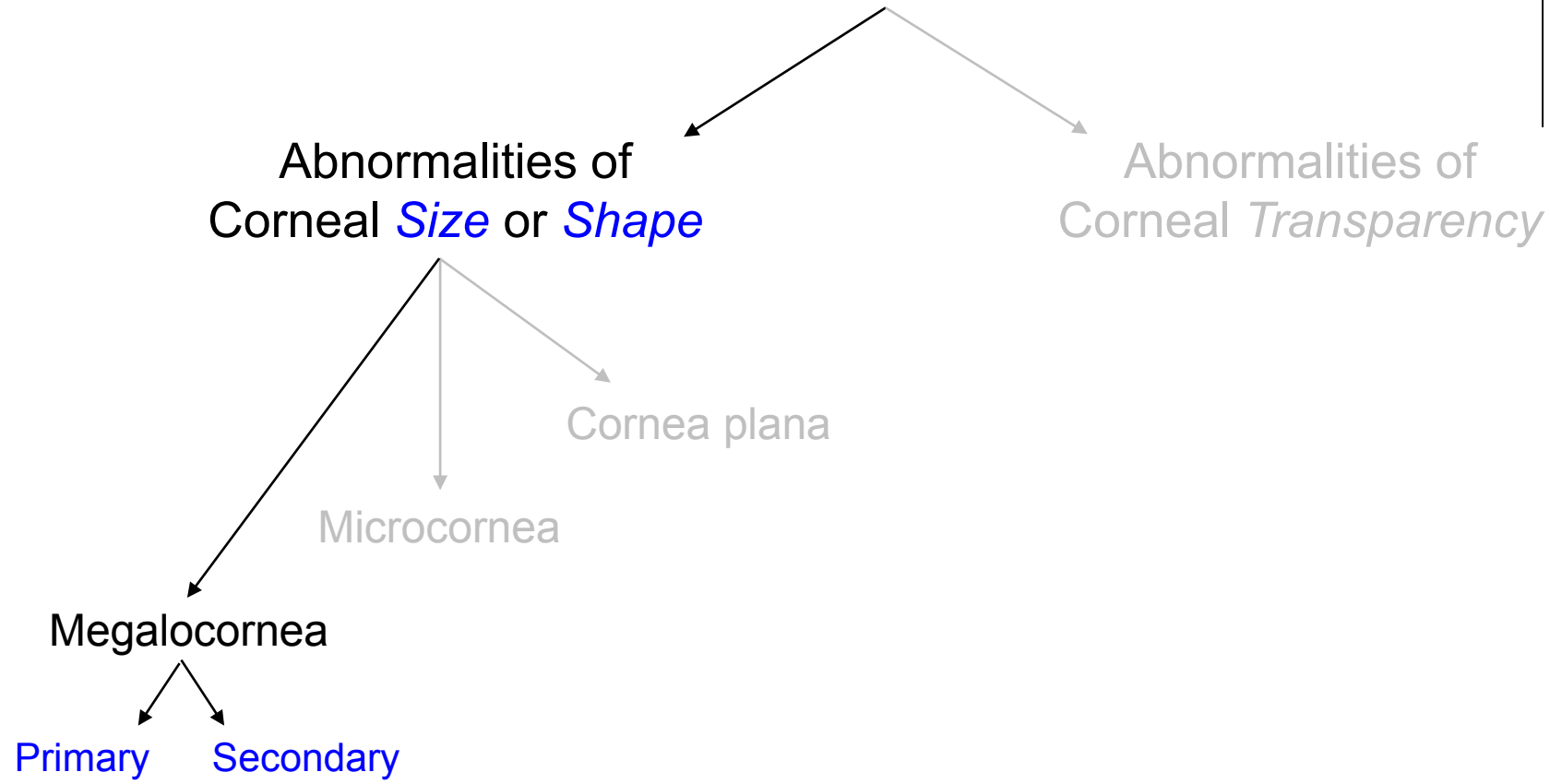

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal

Abnormalities of

*Is primary megalocornea a congenital, or acquired condition?*

Mic

Megalocornea

**Primary**

Secondar

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal

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Congenital—always, and by definition

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Primary megalocornea

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--Lens abnormalities: Presenile word, ( two words
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--Iris abnormalities: word word

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*How does 'iris translucency' manifest during slit-lamp exam?*

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*How does 'iris translucency' manifest during slit-lamp exam?*
As iris transillumination during retroillumination

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Abnormalities of Corneal

Abnormalities of

*Is primary megalocornea a congenital, or acquired condition?*

*Speaking of cataracts: Is primary megalocornea associated with an increased risk of intraoperative complications during cataract surgery?*

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*What is it about megalocornea eyes that predisposes them to complications? (Hint: It's* **not** *a corneal issue)*

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These eyes can have "poor zonular integrity," with all the intra-op problems that entails.

Megalocornea

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Indeed it is

*What is it about megalocornea eyes that predisposes them to complications? (Hint: It's* **not** *a corneal issue)*
These eyes can have "poor zonular integrity," with all the intra-op problems that entails. (Note: The BCSC *Cornea* book has made this fact about megalocornea a chapter "highlight." The point being, it is probably worthy of memorization.)

Megalocornea

**Primary** Secondar

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*What is* ectopia lentis?

*With what othe*
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**ectopia lentis**

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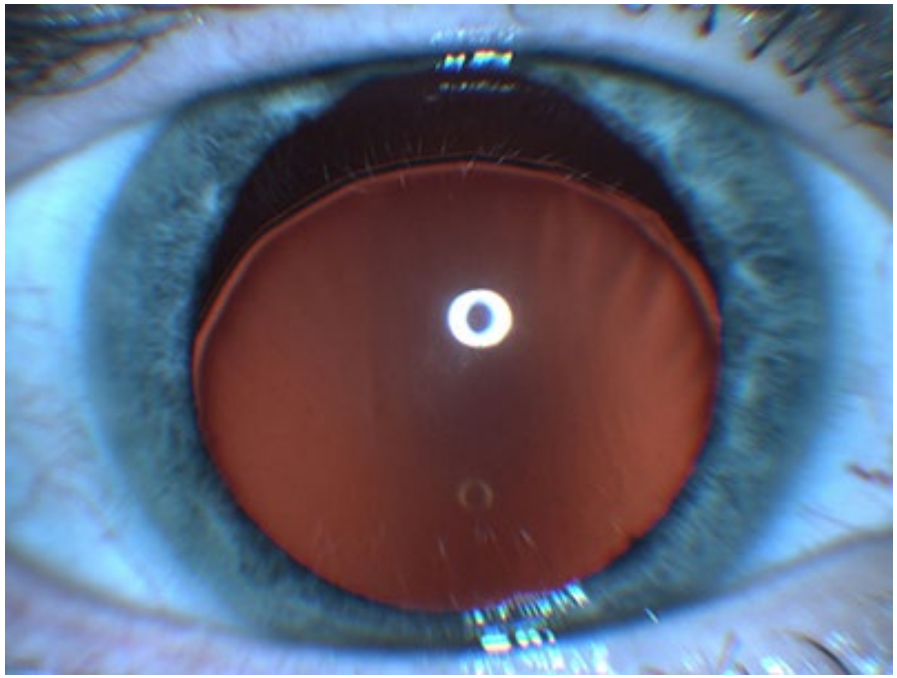
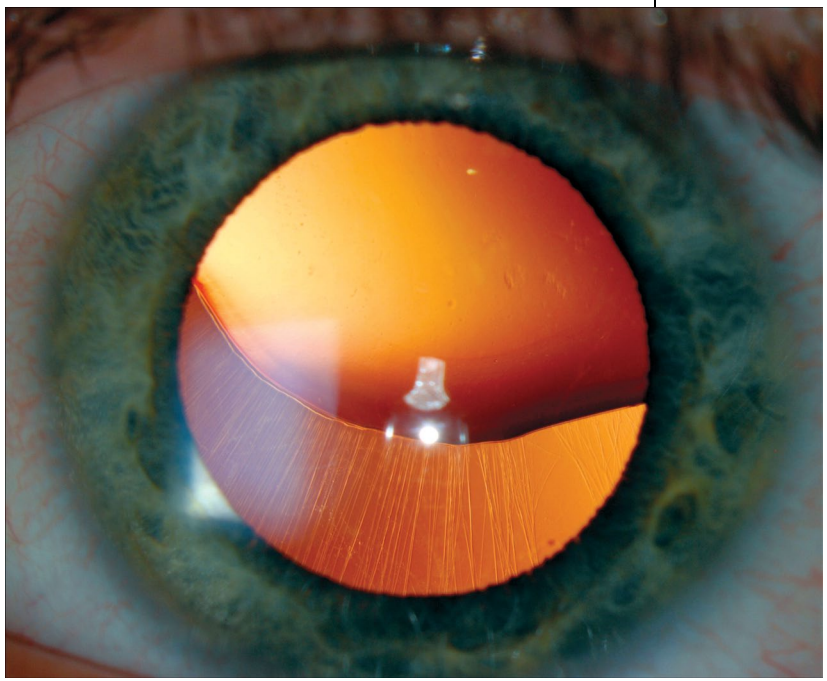
*What is* ectopia lentis?
Displacement of the lens from its normal anatomic position

**ectopia lentis**

# Developmental Abnormalities of the Cornea





Ectopia lentis

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*It can be associated with systemic conditions. Which ones?*

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*Another, potentially blinding, ocular abnormality is a late-onset association with primary megalocornea—what is it?*

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Glaucoma

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There are many; they include:
-- [redacted] syndrome
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--Down syndrome
--Marfan syndrome
--Alport syndrome

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*What three structures/systems manifest abnormalities in Marfan's?*
--The eye (duh)
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--The musculoskeletal

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*What proportion of Marfan pts manifest ocular abnormalities?*

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*Is megalocornea a common ocular manifestation?*

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*Is megalocornea a common ocular manifestation?*
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*OK then, what ocular abnormalities* **are** *common?*
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*Is megalocornea a common ocular manifestation?*
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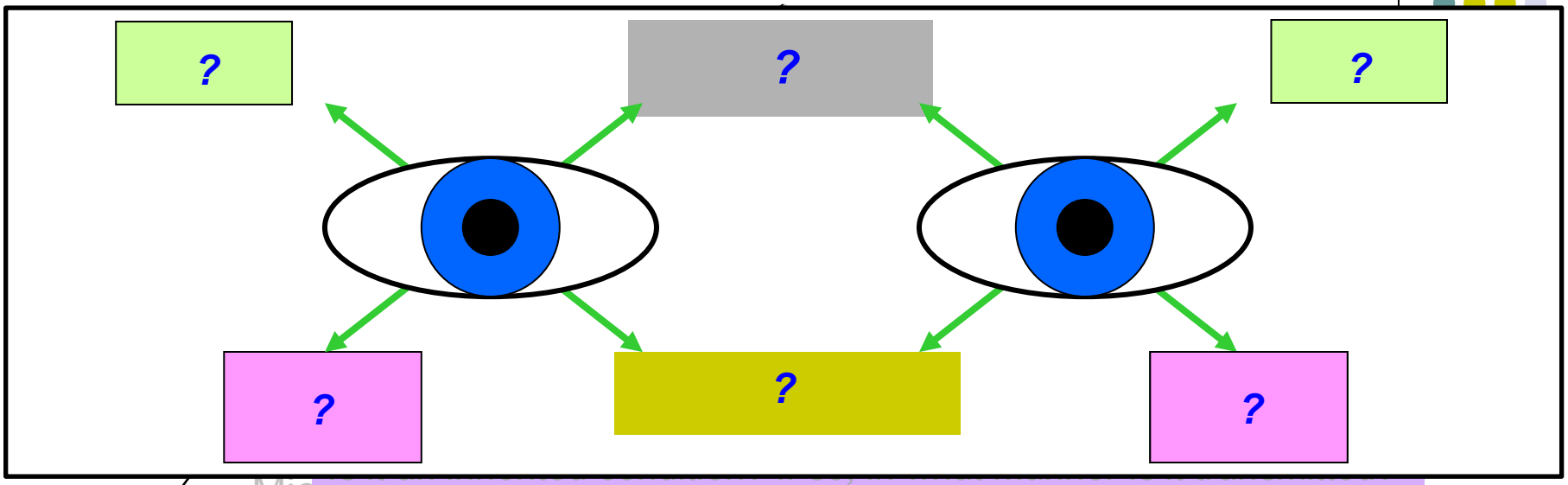
*OK then, what ocular abnormalities* **are** *common?*
--High myopia
--Ectopia lentis

# Developmental Abnormalities of the Cornea




Megalocornea

**Primary** Secondar

Yes; it is X-linked recessive (so is more common in males )

*What is th*
Fibrillin

*What three*
--**The eye**
--The card
--The mus
--Down syndrome
--**Marfan syndrome**
--Alport syndrome

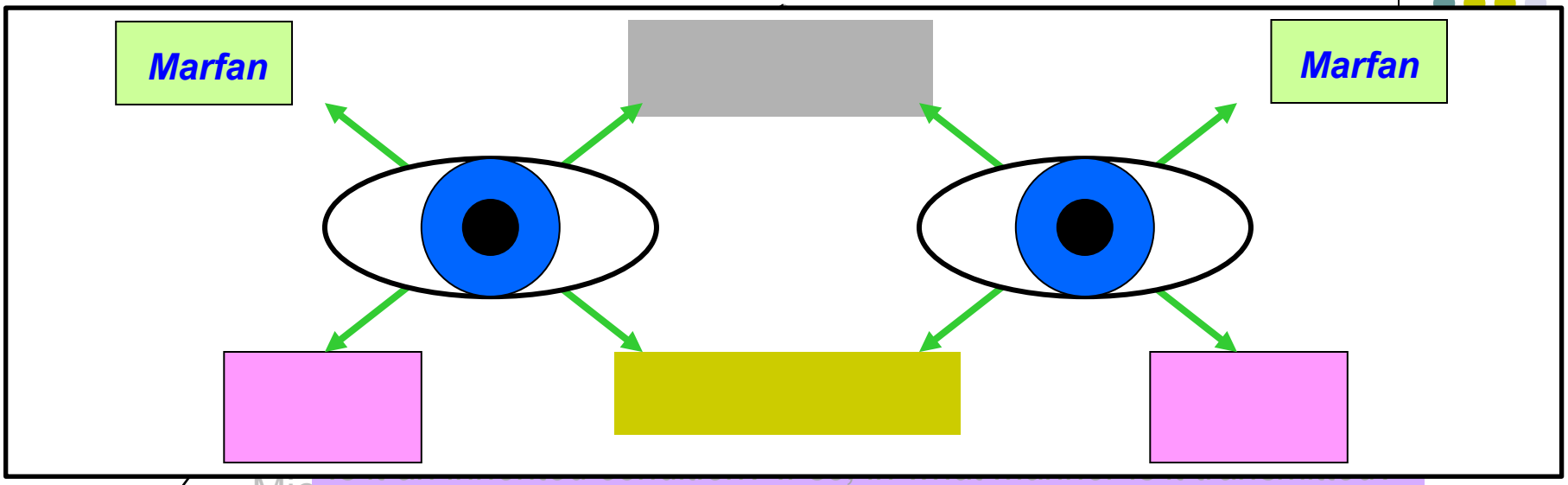
*What proportion of Marfan pts manifest ocular abnormalities?*
At least 80%

*Is megalocornea a common ocular manifestation?*
No

*OK then, what ocular abnormalities **are** common?*
--High myopia
--**Ectopia lentis**

*In which direction do the lenses tend to displace in Marfan's?*

# Developmental Abnormalities of the Cornea

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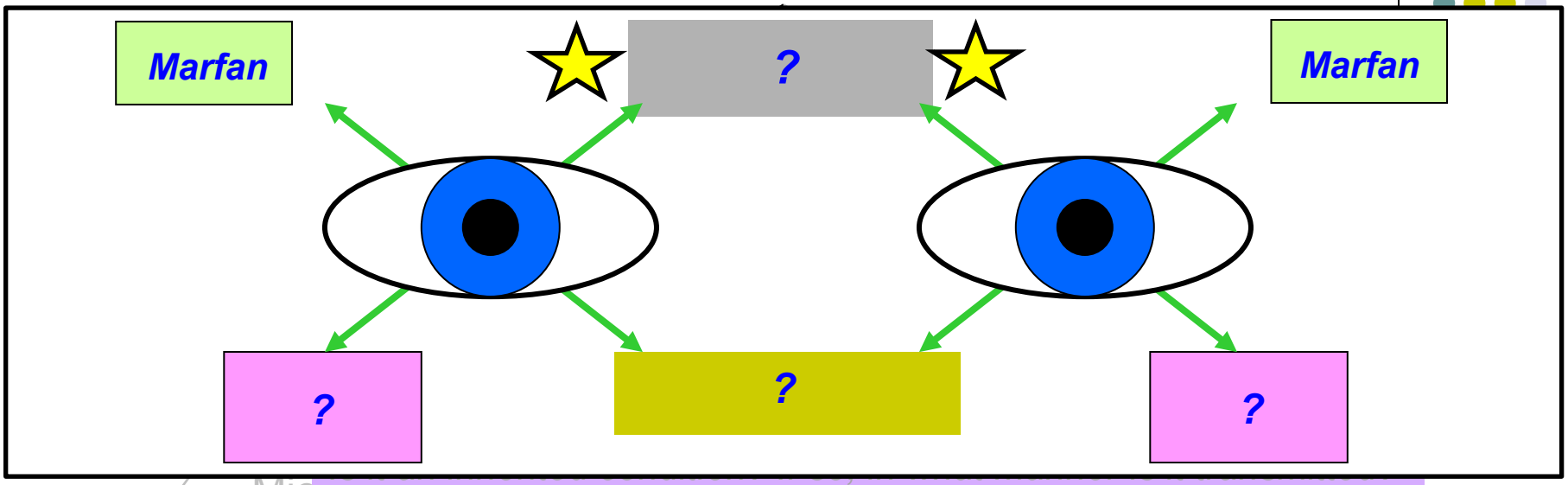
**Superotemporal**

# Developmental Abnormalities of the Cornea




*What condition is associated with displacement supero***nasally***?*

Primary Secondar

*What three*
--**The eye**
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--The mus
--Down syndrome
--**Marfan syndrome**
--Alport syndrome

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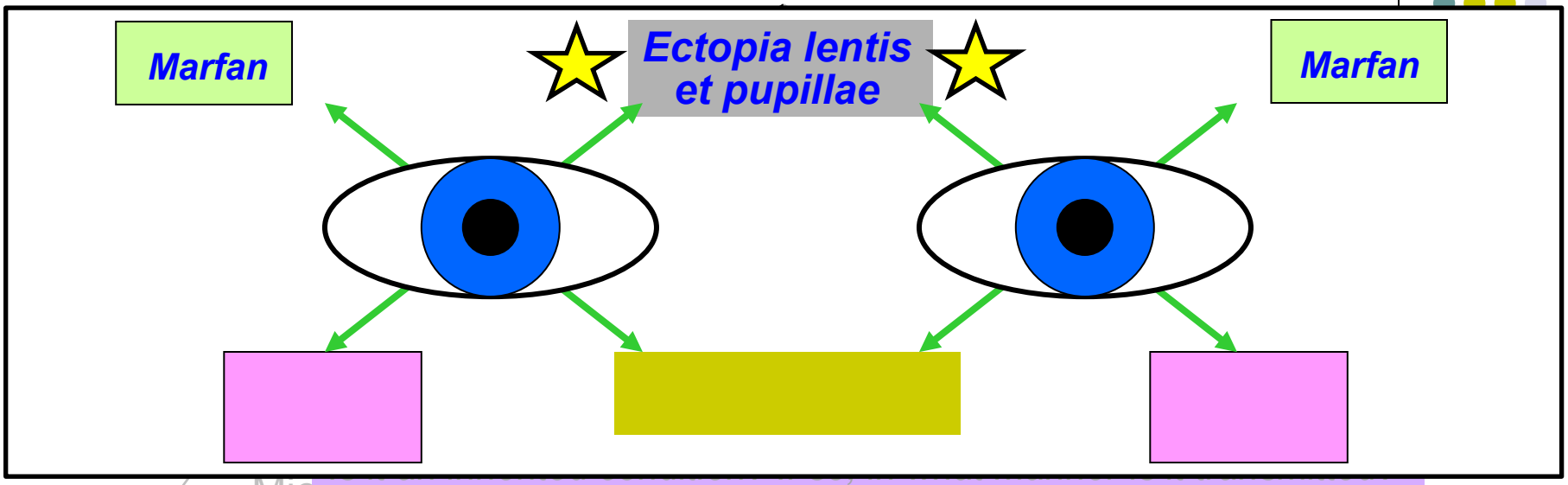


*What condition is associated with displacement superonasally?* Ectopia lentis et pupillae

Primary Secondar

*What three*
--**The eye**
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--The mus

*Is megalocornea a common ocular manifestation?*
No

*OK then, what ocular abnormalities **are** common?*
--High myopia
--
**Ectopia lentis**

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--**Marfan syndrome**
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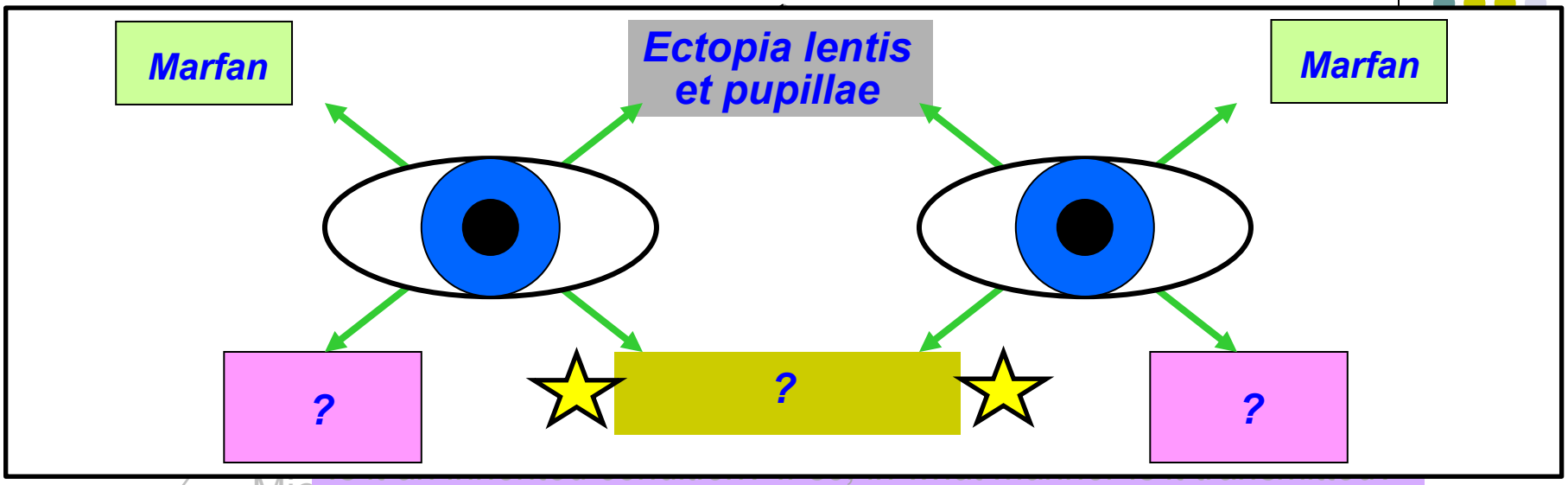


*What condition is associated with displacement supero***nasally***?* Ectopia lentis et pupillae
*What condition is associated with displacement* ***infero****nasally?*

Primary Secondar

*What three*
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--The mus
--Down syndrome
--**Marfan syndrome**
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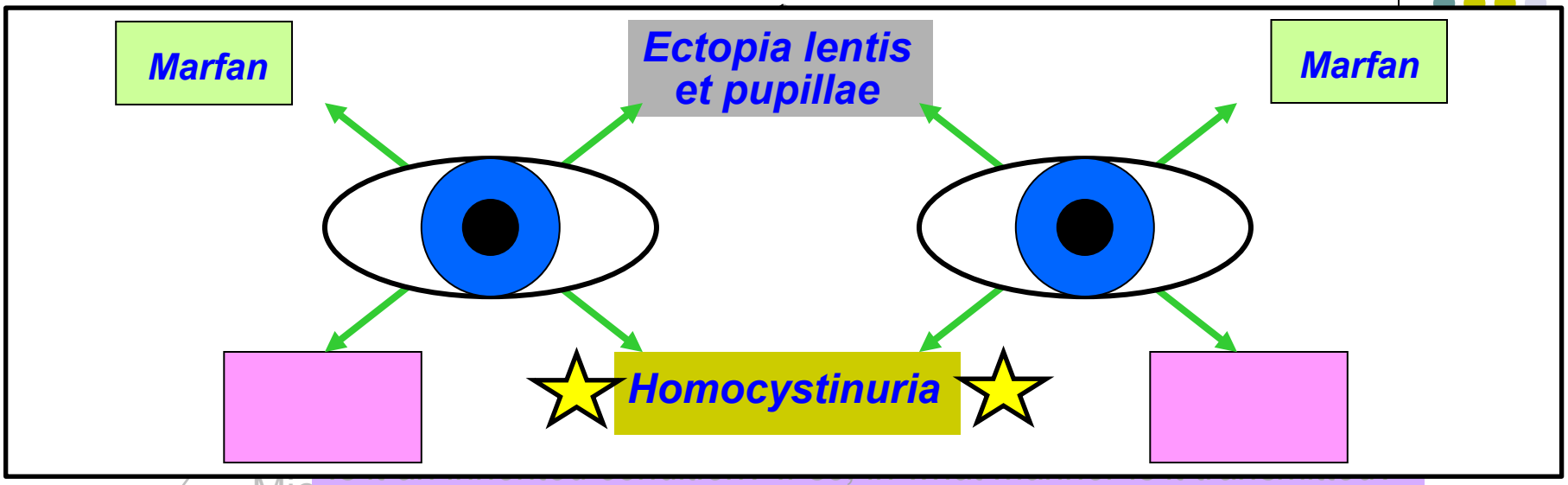


*What condition is associated with displacement supero***nasally***?* Ectopia lentis et pupillae
*What condition is associated with displacement* ***infero****nasally?* Homocystinuria

Primary Secondar

*What three*
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*Is megalocornea a common ocular manifestation?*
No

*OK then, what ocular abnormalities* **are** *common?*
--High myopia
--
**Ectopia lentis**
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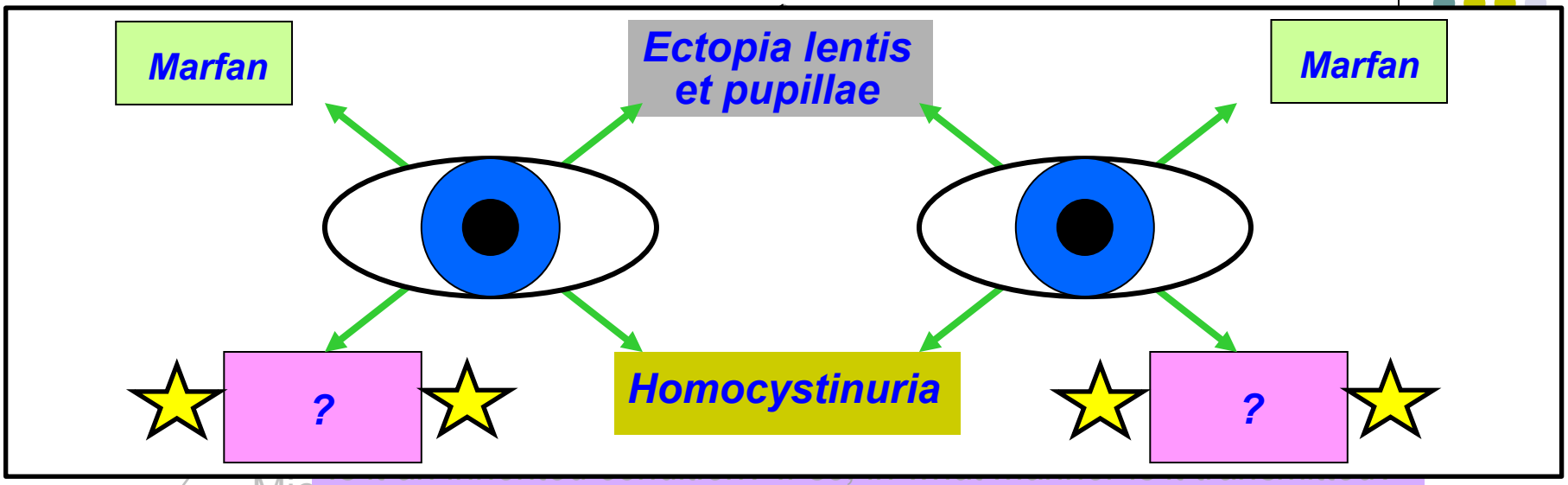


*What condition is associated with displacement supero***nasally***?* Ectopia lentis et pupillae
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What condition is associated with displacement *infero****temporally***?

Primary Secondar

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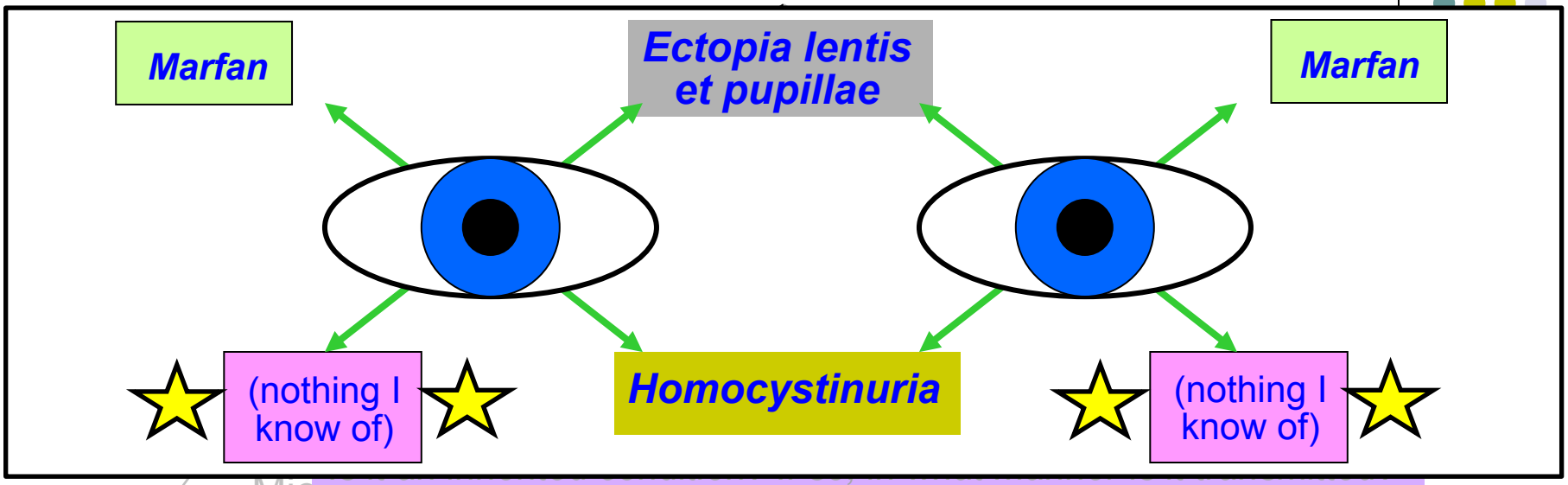


*What condition is associated with displacement supero***nasally***?* Ectopia lentis et pupillae
*What condition is associated with displacement* ***infero****nasally?* Homocystinuria
What condition is associated with displacement *infero****temporally***? Nothing I know of

Primary Secondar

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# Developmental Abnormalities of the Cornea

Abnormalities of Corneal

Abnormalities of

*Is primary megalocornea a congenital, or acquired condition?*
Congenital—always, and by definition

*Does it usually present unilaterally, or bilaterally?*
Bilaterally—always, and by definition

*What sort of condition is Alport syndrome?*
A [two words] syndrome

*nsmitted?*
*es )*

Megaloco

**Primary** S

*es?*

--Alport syndrome

# Developmental Abnormalities of the Cornea

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A familial oculorenal syndrome

Megaloco

**Primary**

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Megaloco

**Primary**

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Megaloco

**Primary**

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*What is the classic presenting sign of the familial oculorenal syndromes (hint: It's nonocular)?*

Megaloco

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Megaloco

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**Lenticonus**

Megaloco

**Primary**

**Alport syndrome**

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
  - Megalocornea
    - Primary
    - **Secondary**
  - Microcornea
  - Cornea plana
- Abnormalities of Corneal *Transparency*

*Is secondary megalocornea a congenital, or acquired condition?*

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*What is the cause?*
Elevated IOP

*Wait a minute—I thought an enlarged globe secondary to elevated IOP is* word *. What's the difference between secondary megalocornea and* same word *?*

Megalocornea

Primary **Secondary**

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In buphthalmos, the entire globe (including the cornea) is enlarged, whereas in secondary megalocornea, only the cornea is—the rest of the eye is normally sized

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*With regard to AC depth in secondary megalocornea, what proportion of total eye length are we talking about?*

Megalocornea

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*With regard to AC depth in secondary megalocornea, what proportion of total eye length are we talking about?*
If AC depth is greater than about % of total eye length, it's secondary megalocornea

Megalocornea

Primary **Secondary**

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Acquired—always, and by definition

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***What is the cause?***
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*With regard to AC depth in secondary megalocornea, what proportion of total eye length are we talking about?*
If AC depth is greater than about 20% of total eye length, it's secondary megalocornea

Megalocornea

Primary **Secondary**

*Is secondary megalocornea a congenital, or acquired condition?*
Acquired—always, and by definition

*Does it usually present unilaterally, or bilaterally?*
It can be either

***What is the cause?***
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*With regard to AC depth in secondary megalocornea, what proportion of total eye length are we talking about?*
If AC depth is greater than about 20% of total eye length, it's secondary megalocornea

**Think of secondary megalocornea as 'arrested buphthalmos,' ie, the IOP affected the anterior segment, but not the rest of the globe**

Megalocornea

Primary **Secondary**

*Is secondary megalocornea a congenital, or acquired condition?*
Acquired—always, and by definition

*Does it usually present unilaterally, or bilaterally?*
It can be either

***What is the cause?***
**Elevated IOP**

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

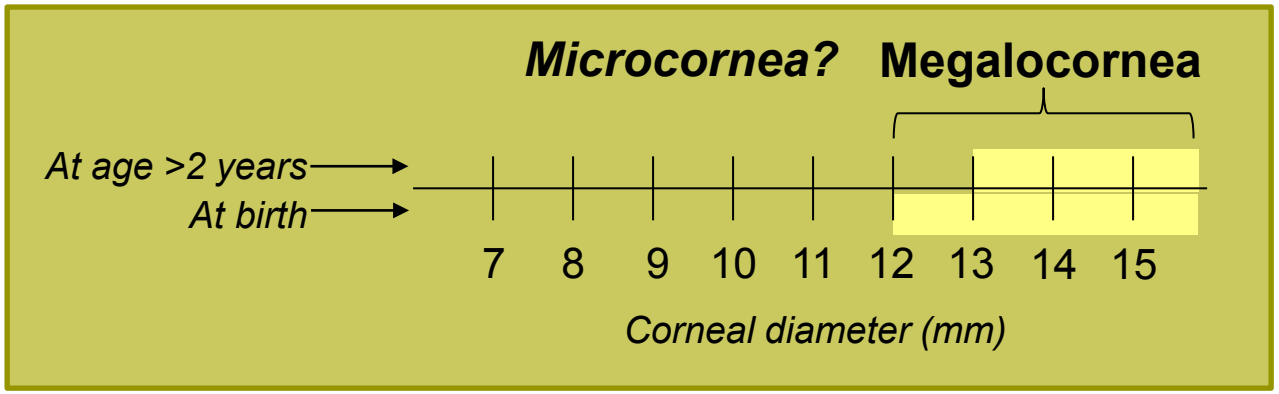
Abnormalities of Corneal *Transparency*

Cornea plana

**Microcornea**

Megalocornea

*What is the definition of microcornea, ie, how 'micro' does it have to be?*



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Abnormalities of Corneal *Size* or *Shape*

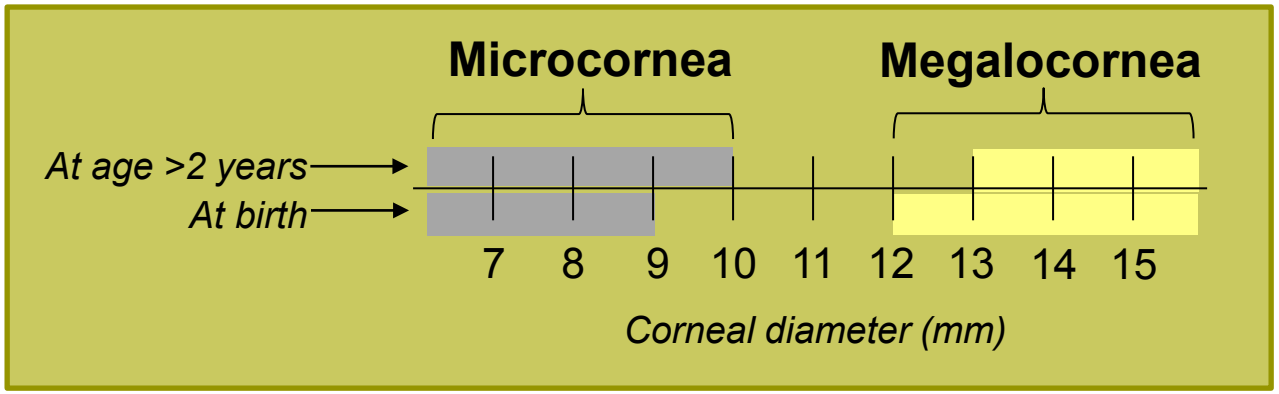
Abnormalities of Corneal *Transparency*

Cornea plana

**Microcornea**

Megalocornea

*What is the definition of microcornea, ie, how 'micro' does it have to be?*
Corneal diameter < 9 mm at birth, or < 10 mm at age 2 years or older



# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

# Developmental Abnormalities of the Cornea


Unilateral


Bilateral

Microcornea

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

*In addition to being small, can the cornea also be:*
*--Too thick?*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

*In addition to being small, can the cornea also be:*
*--Too thick?* No—by definition, the thickness is normal

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

*In addition to being small, can the cornea also be:*
*--Too thick?* No—by definition, the thickness is normal
*--Hazy?*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

*In addition to being small, can the cornea also be:*
*--Too thick?* No—by definition, the thickness is normal
*--Hazy?* No—by definition, the cornea is clear

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
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*In addition to being small, can the cornea also be:*
*--Too thick?* No—by definition, the thickness is normal
*--Hazy?* No—by definition, the cornea is clear
*--Too flat?*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Microcornea**

Megalocornea

*Does microcornea present unilaterally, or bilaterally?*
It can be either

*In addition to being small, can the cornea also be:*
*--Too thick?* No—by definition, the thickness is normal
*--Hazy?* No—by definition, the cornea is clear
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*What about* **open**-*angle glaucoma--does cornea plana convey an increased risk of it?*

*A shallow AC implies increased risk of angle-closure glaucoma. Is microcornea in fact associated with an increased risk of* open-angle *glaucoma?*
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Yes. Of the cornea-plana pts who manage to avoid developing angle-closure glaucoma, % will go on to develop the open-angle version.

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*What about* **open**-*angle glaucoma--does cornea plana convey an increased risk of it?*
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--Persistent fetal vasculature (PFV, aka PHPV)
--Congenital

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*With what ocular conditions is microcornea associated?*
--Persistent fetal vasculature (PFV, aka PHPV)
--Congenital cataracts

*With what* systemic *conditions is microcornea associated?*
--Ehlers-Danlos syndrome
--Myotonic dystrophy

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

*Basic distinction used by the BCSC* Peds *book*

***?*** ***?***

Cornea plana

Microcornea

Megalocornea

*Hol up—you're skipping cornea plana! What's up with that?*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

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***?*** ***?***

Cornea plana

Microcornea

Megalocornea

*Hol up—you're skipping cornea plana! What's up with that?*

Patience, Grasshopper—all will be made clear soon

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

- Cornea plana
- Microcornea
- Megalocornea

Abnormalities of Corneal *Transparency*

*Basic distinction used by the BCSC* Peds *book*

- ?
- ?

*(OK, now answer this)*

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

- Cornea plana
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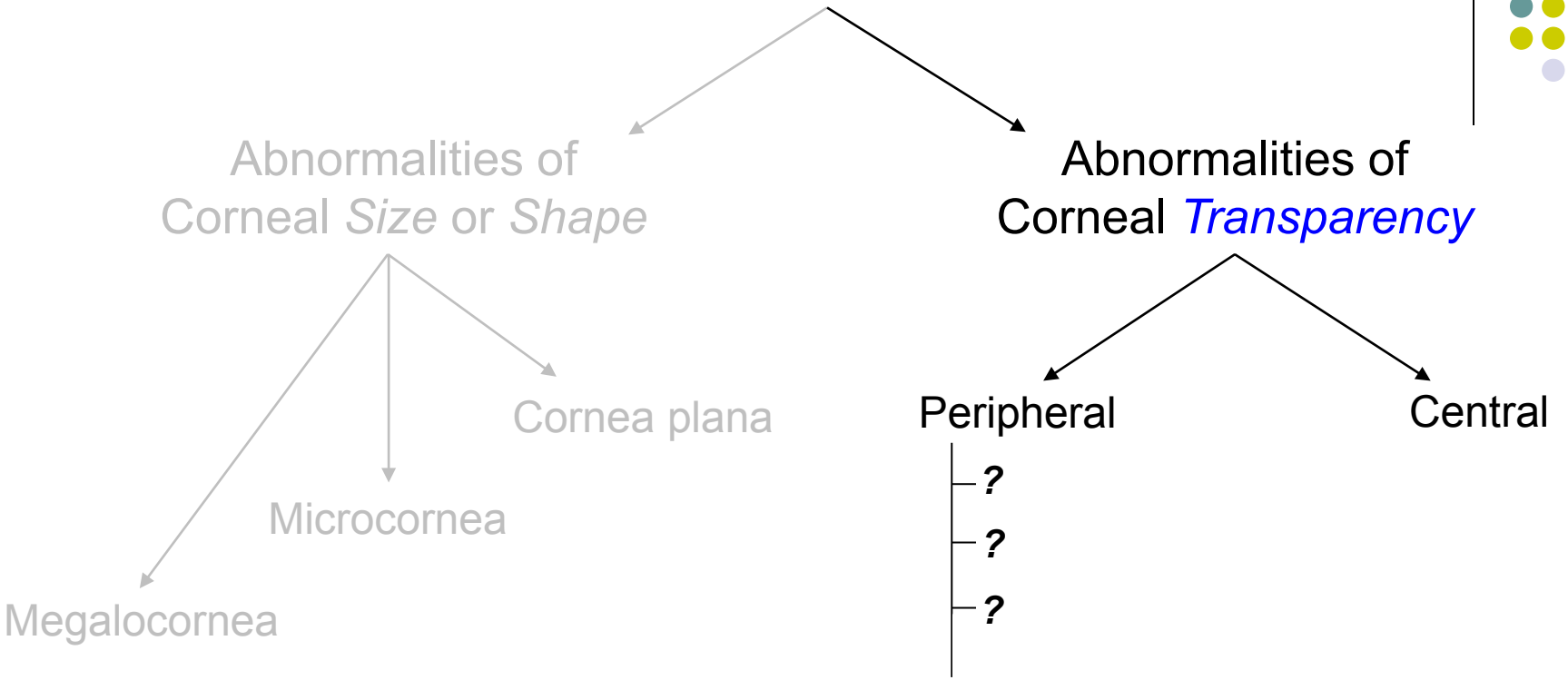
Abnormalities of Corneal *Transparency*

*Basic distinction used by the BCSC* Peds *book*

- Peripheral
- Central

*(OK, now answer this)*

# Developmental Abnormalities of the Cornea



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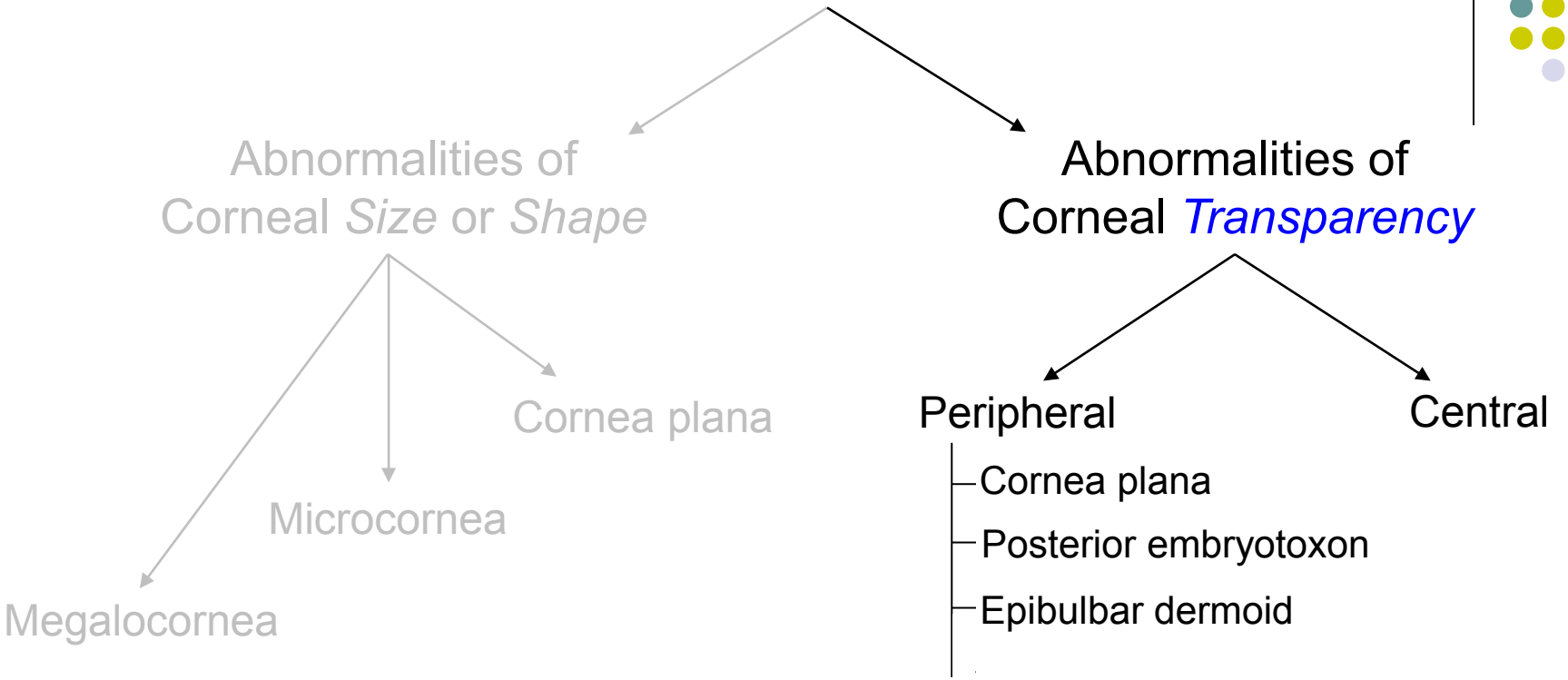

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Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Cornea plana**

Microcornea

Me

Peripheral

**Cornea plana**

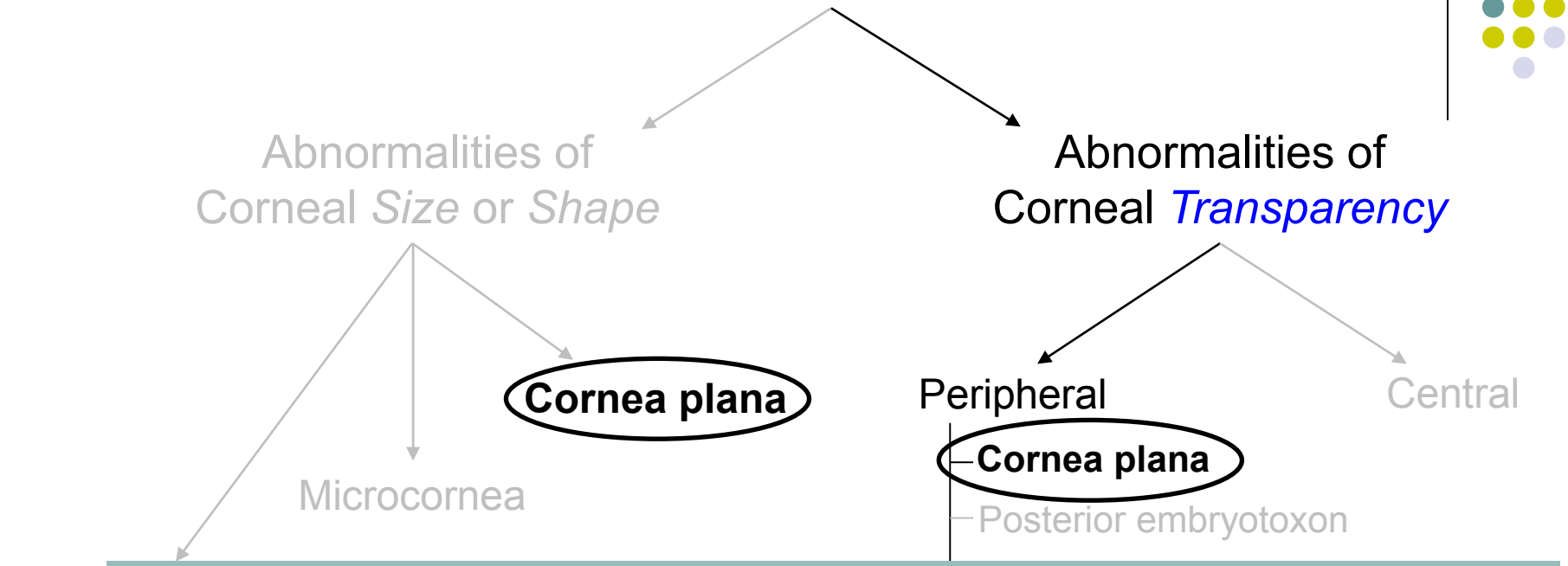
Posterior embryotoxon

Central

Cornea plana belongs in *both* groups, is why it was saved until now…

*No question—carry on*

# Developmental Abnormalities of the Cornea




Cornea plana belongs in *both* groups, is why it was saved until now…

*Cornea plana has five characteristics, one of which is flat Ks. What are the other four?*

--Flat Ks

--

*(The reason there are only three more slots will become apparent on the next slide)*

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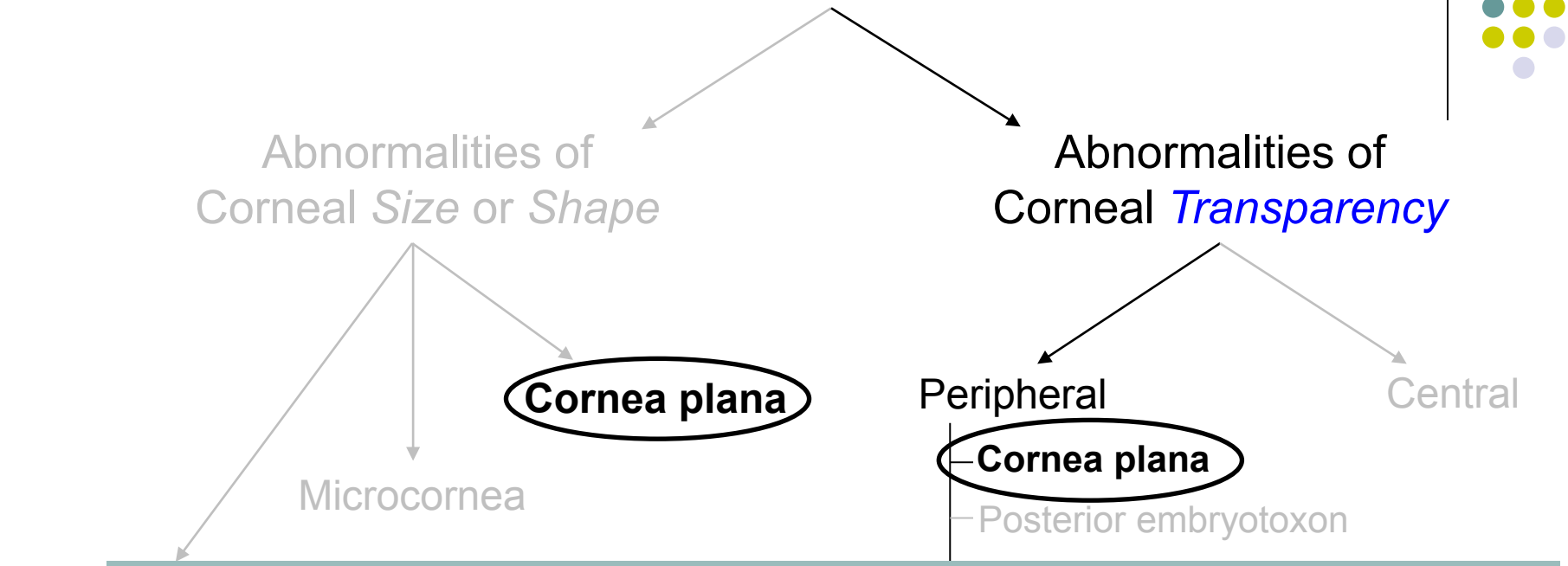
# Developmental Abnormalities of the Cornea

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*Cornea plana has five characteristics, one of which is flat Ks. What are the other four?*

--Flat Ks

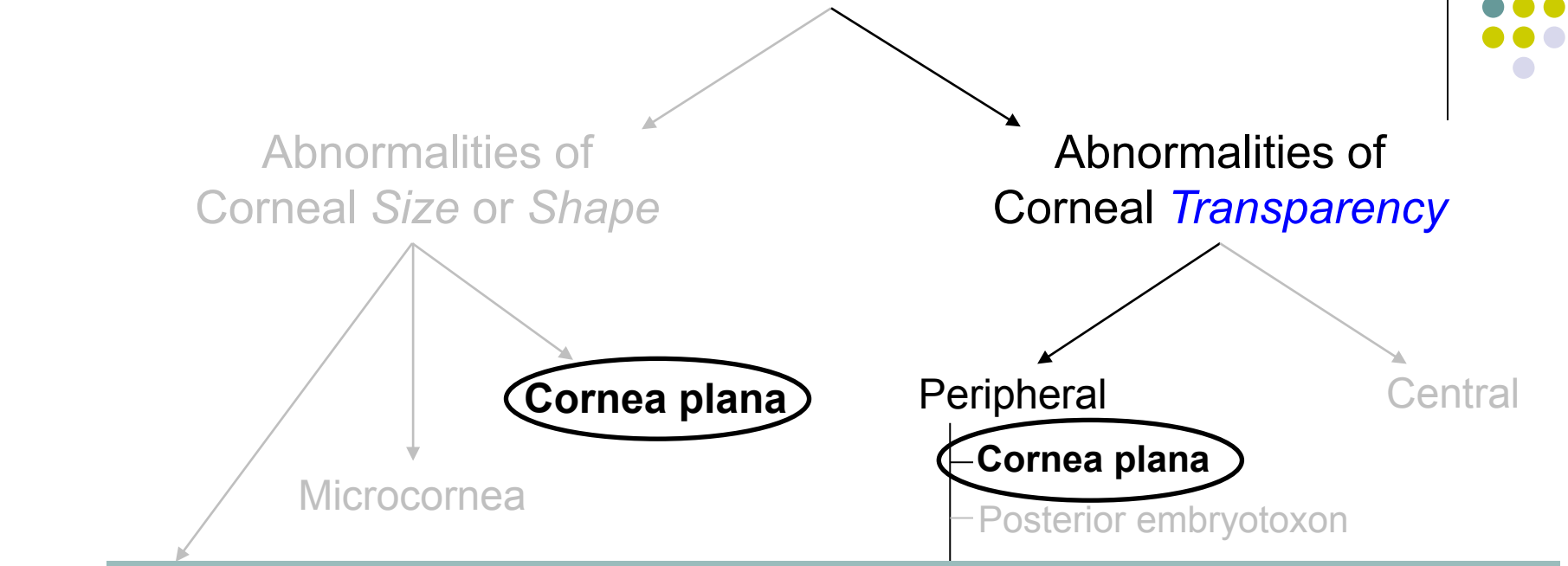
--Poorly developed ("indistinct") limbus

--Shallow AC

--High refractive status with associated two words

*#4* *#5*

# Developmental Abnormalities of the Cornea

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--Poorly developed ("indistinct") limbus
--Shallow AC
--High hyperopia with associated accommodative esotropia

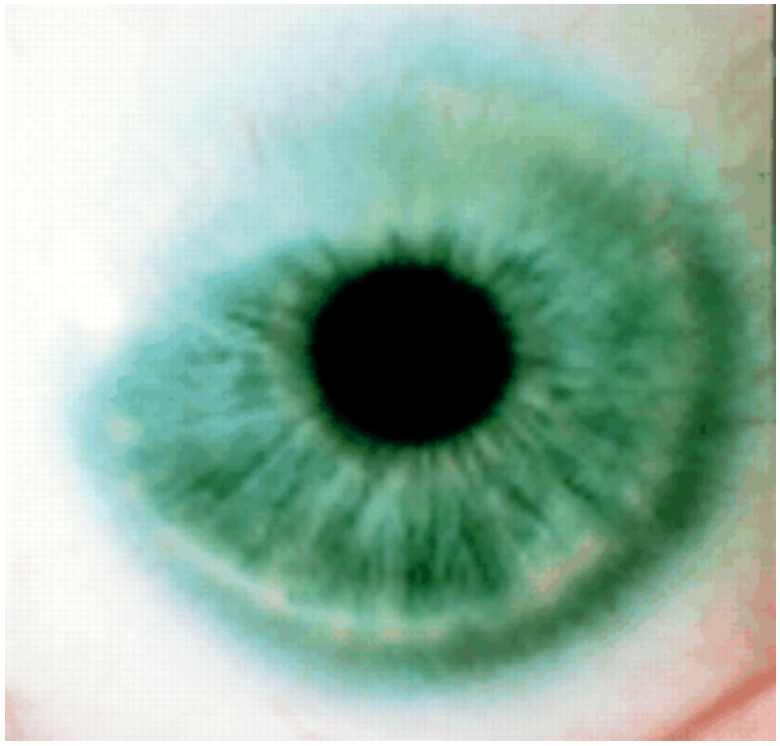
# Developmental Abnormalities of the Cornea

Yikes—that **is** pretty flat, innit?

Cornea plana

# Developmental Abnormalities of the Cornea

Yikes—that **is** pretty flat, innit?



Note the ‘indistinct’ limbus

Cornea plana

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Cornea plana**

Microcornea

Peripheral

Central

**Cornea plana**

Me Cornea plana

*Cornea plana*

--**Flat Ks**

--Poorly deve

--Shallow AC

--High hyper

*our?*

*What is the average central corneal power of the normal adult cornea?*

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About 43D

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*What is the average central corneal power of the normal adult cornea?*
About 43D

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Technically, less than 43D, but don't get it twisted--most corneas within shouting distance of 43D are not 'plana.' Plana cornea are **much** flatter.

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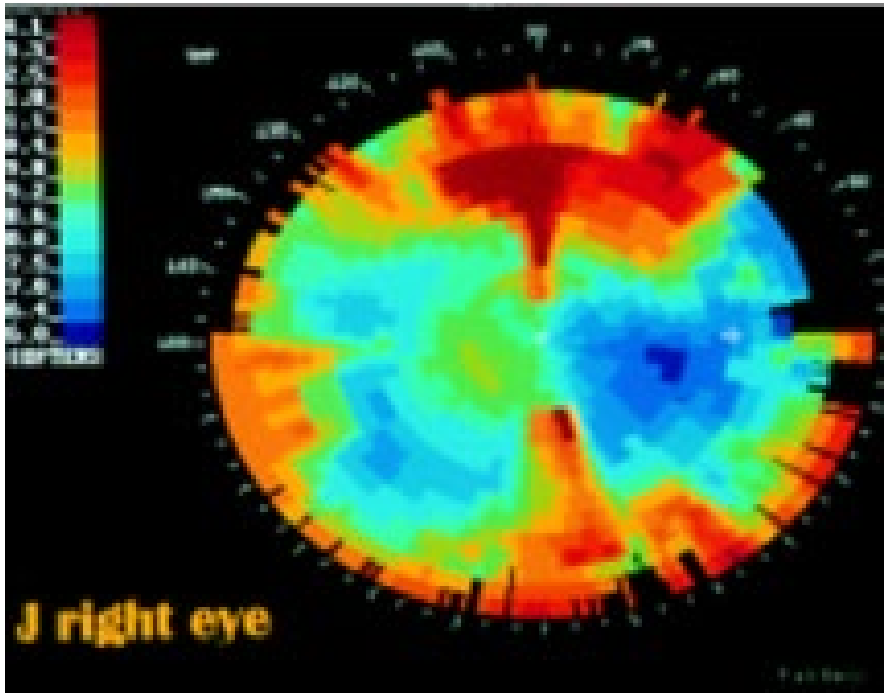
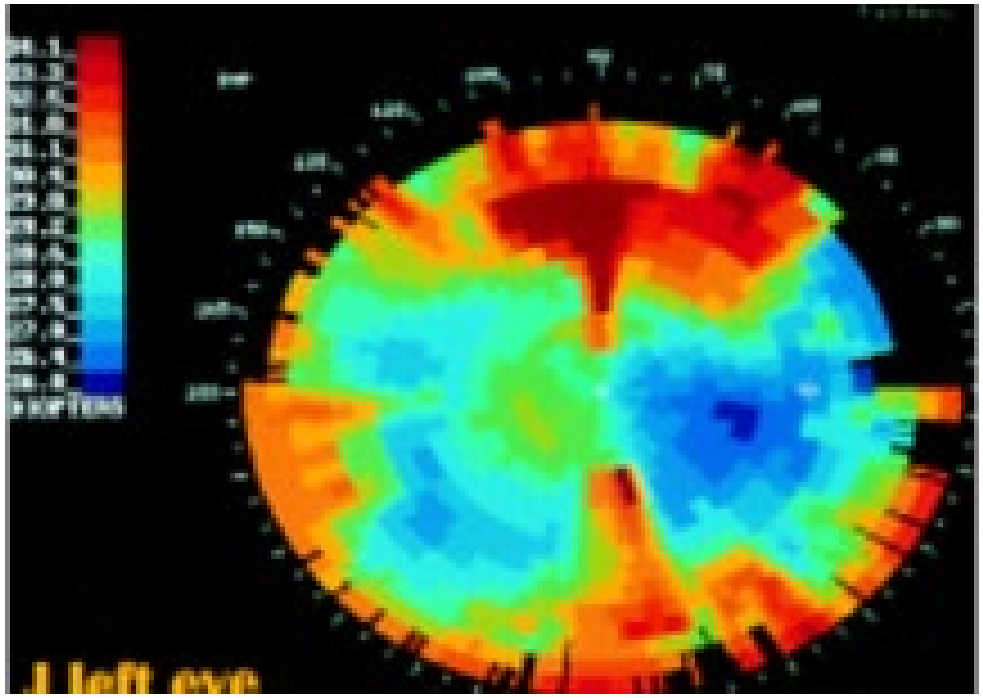
Values in the 30-35D range are common

# Developmental Abnormalities of the Cornea




Cornea plana: Keratometry (I know, it's really blurry)

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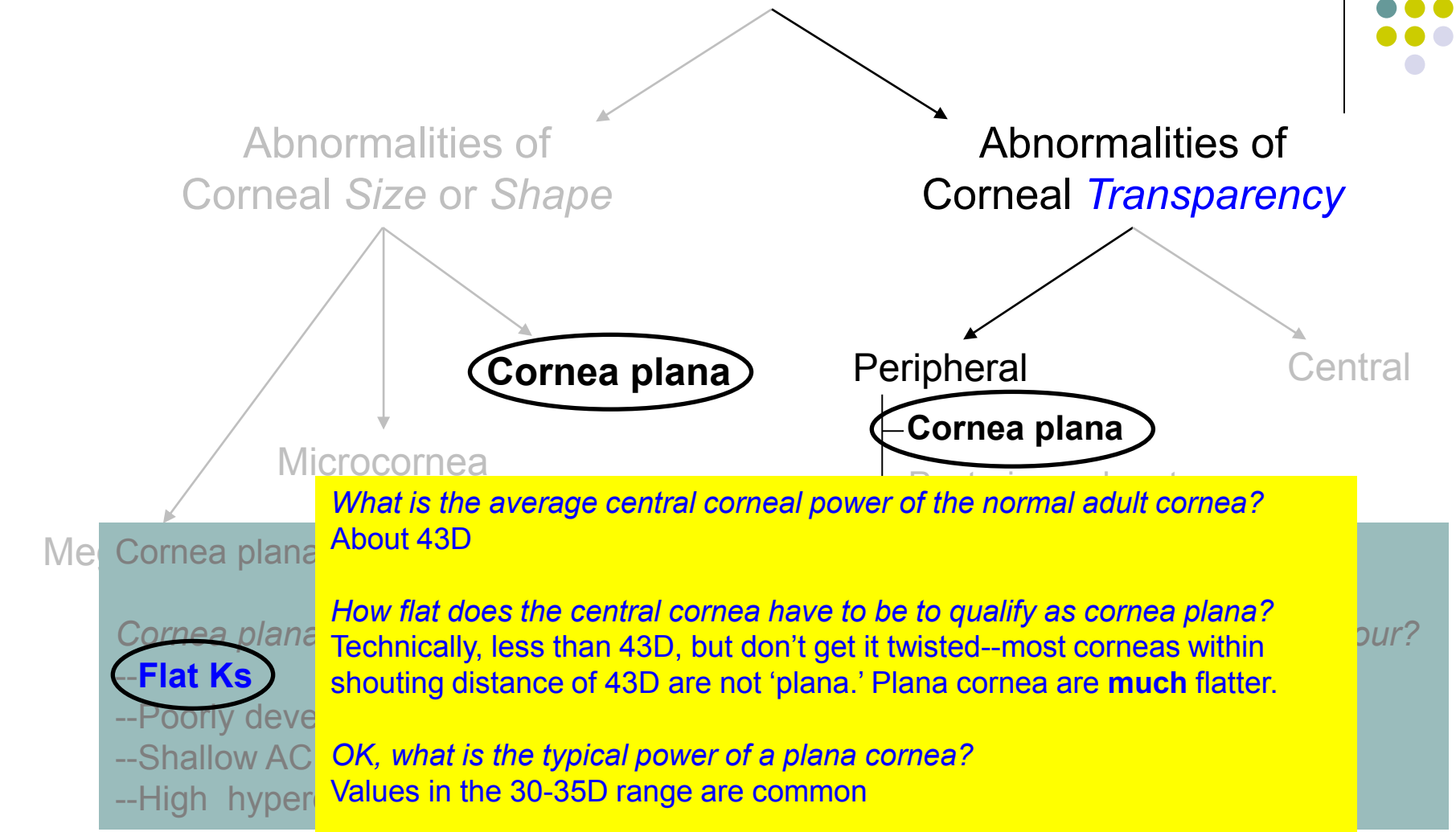


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The key difference is one of corneal transparency :
In sclerocornea, the cornea is [a word]
In cornea plana, the cornea is [its antonym]

*There a pathognomonic corneal curvature--what is it? (Hint: It's not a specific numeric value.)*
If the cornea is the same curvature as the adjacent sclera ...e eye is plana

# Developmental Abnormalities of the Cornea

Abnormalities of Corneal *Size* or *Shape*

Abnormalities of Corneal *Transparency*

**Cornea plana**

Microcornea

Peripheral

**Cornea plana**

Central

Me Cornea plana

*Cornea plana*

**Flat Ks**

--Poorly deve

--Shallow AC

--High hyper

*What is the average central corneal power of the normal adult cornea?*

About 43D

*How flat does the central cornea have to be to qualify as cornea plana?*

Techn

shouti

*OK, w*

Value

*But if the cornea is the same curvature as the adjacent sclera, how is that not simply sclerocornea? How do you differentiate between these conditions?*

The key difference is one of corneal transparency :

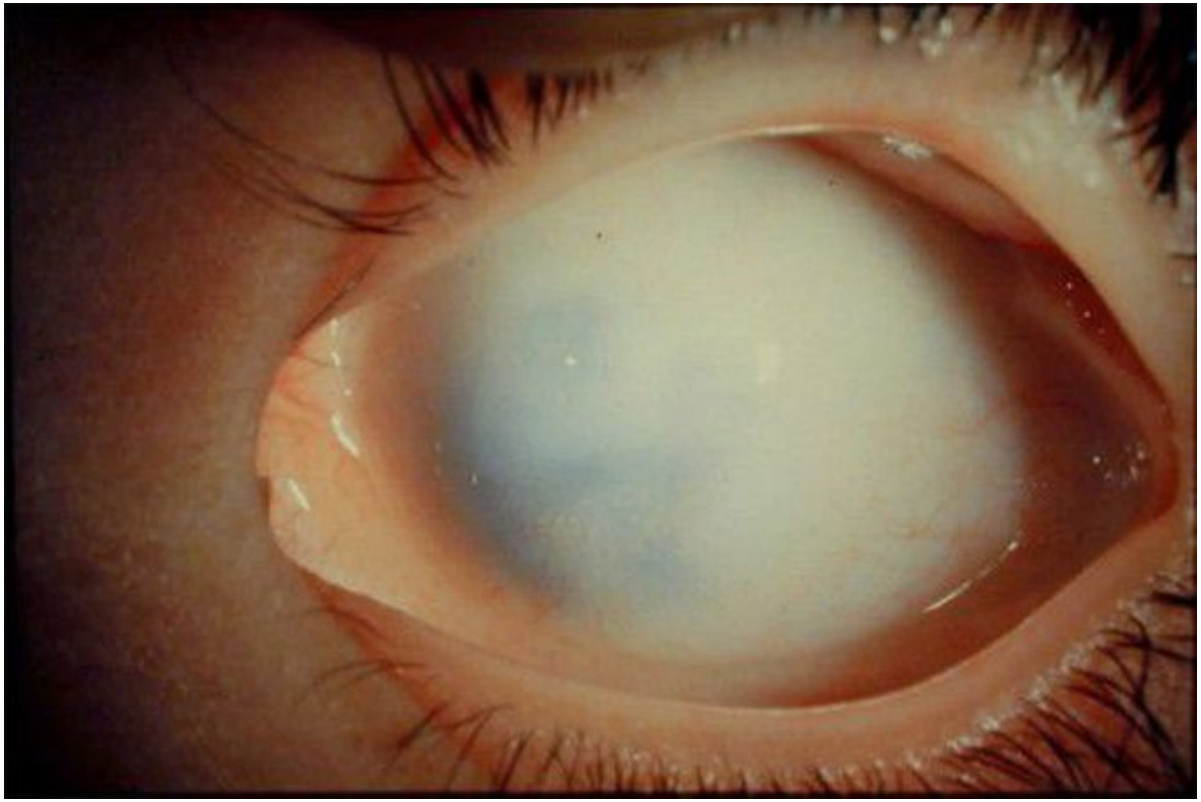
In sclerocornea, the cornea is opaque

In cornea plana, the cornea is clear (relatively speaking)

*There a pathognomonic corneal curvature--what is it? (Hint: It's not a specific numeric value.)*

If the cornea is the same curvature as the adjacent sclera, the eye is plana

# Developmental Abnormalities of the Cornea



Sclerocornea

# Developmental Abnormalities of the Cornea

*What is a posterior embryotoxon?*

Abnormalities of
Corneal *Size* or *Sha*

Co

Microcornea

Megalocornea

Cornea plana

**Posterior embryotoxon**

Epibulbar dermoid

# Developmental Abnormalities of the Cornea

*What is a posterior embryotoxon?*
An anteriorly displaced and thickened Schwalbe's line/ring

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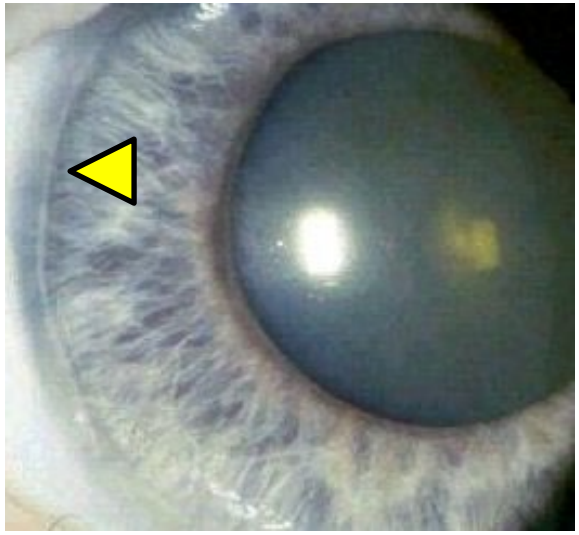

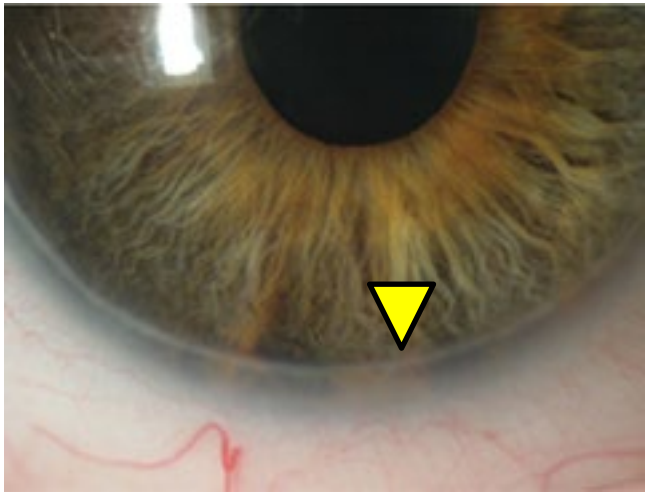

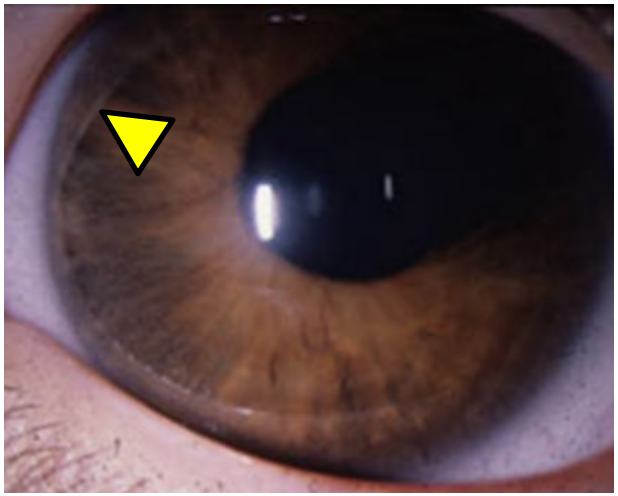

Posterior embryotoxon

# Developmental Abnormalities of the Cornea

*What is Schwalbe's line/ring?*

?

ened **Schwalbe's line/ring**

Corneal *Size* or *Sha*

Co

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# Developmental Abnormalities of the Cornea

*What is Schwalbe's line/ring?*
The edge or termination of Descemet's layer

?

ened **Schwalbe's line/ring**

Corneal *Size* or *Sha*

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**Posterior embryotoxon**

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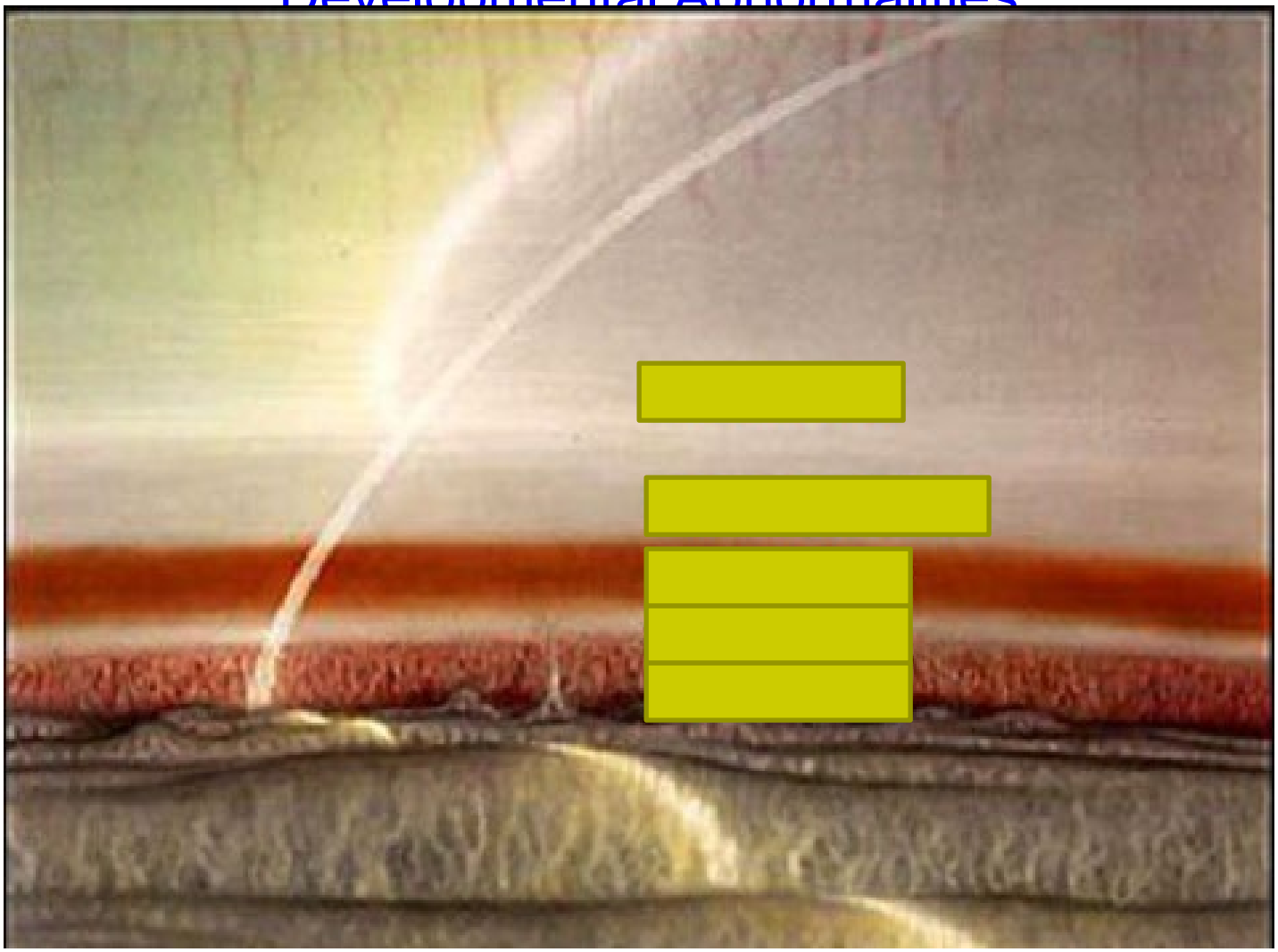
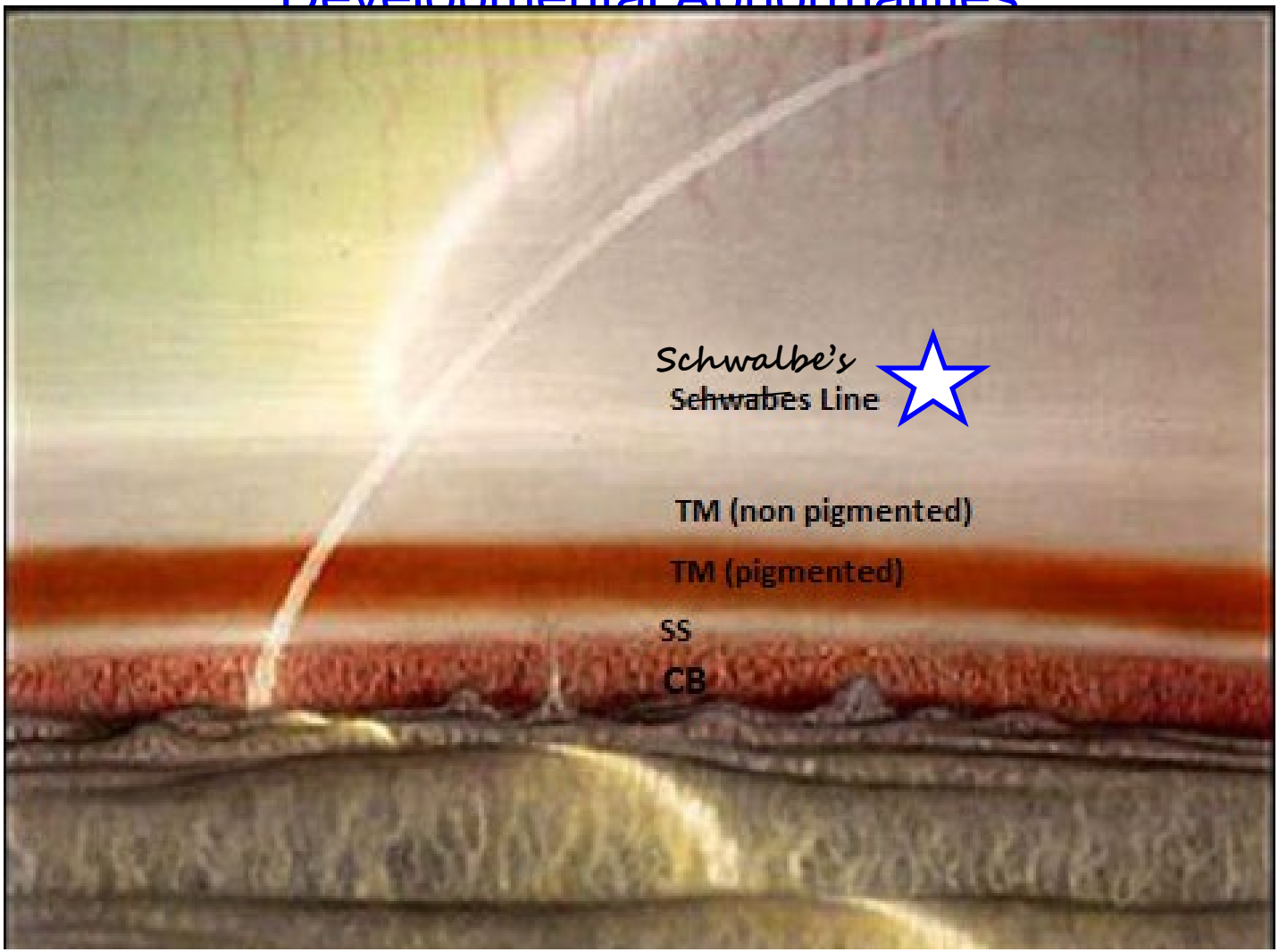


Normal angle anatomy: Identify the structures

# Developmental Abnormalities




Normal angle anatomy: Identify the structures

# Developmental Abnormalities of the Cornea

*What is Schwalbe's line/ring?*
The edge or termination of Descemet's layer

*Is it normally apparent during slit-lamp examination?*

?

ened **Schwalbe's line/ring**

Corneal *Size* or *Sha*

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Microcornea

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# Developmental Abnormalities of the Cornea

*What is Schwalbe's line/ring?*
The edge or termination of Descemet's layer

*Is it normally apparent during slit-lamp examination?*
No--it is usually too thin and posterior to be seen

**Schwalbe's line/ring**

Corneal *Size* or *Sha*

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*In what three situations is it a significant finding?*
1)
2)
3)

*Hints forthcoming…*

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# Developmental Abnormalities of the Cornea

Abnormalities of
Corneal *Size* or *Sha*

*Interestingly, all three of these begin with the letter 'A'*

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1) When it is part of the eponym-eponym **syndrome**
2) When it is associated with not an eponym
3) When it is associated with eponym **syndrome**

Cornea plana

**Posterior embryotoxon**

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1) When it is part of the **Axenfeld-Rieger syndrome**
2) When it is associated with **aniridia**
3) When it is associated with **Alagille syndrome**

Abnormalities of
Corneal *Size* or *Sha*

*Interestingly, all three of these begin with the letter 'A'*

Microcornea

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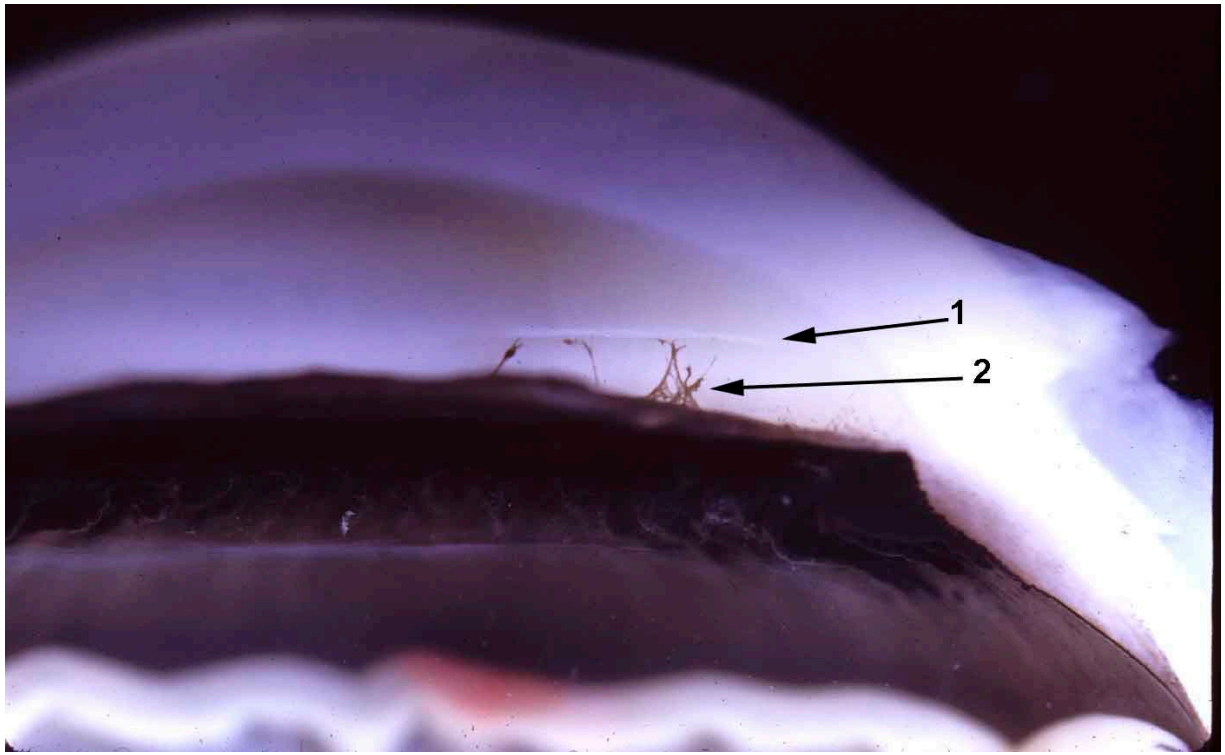


Axenfeld-Reiger: Note the posterior embryotoxon (1) with attached iris strands (2)

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*'Angle abnormalities' suggests an increased risk of glaucoma. Does ARS in fact convey such a risk?*

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It does indeed

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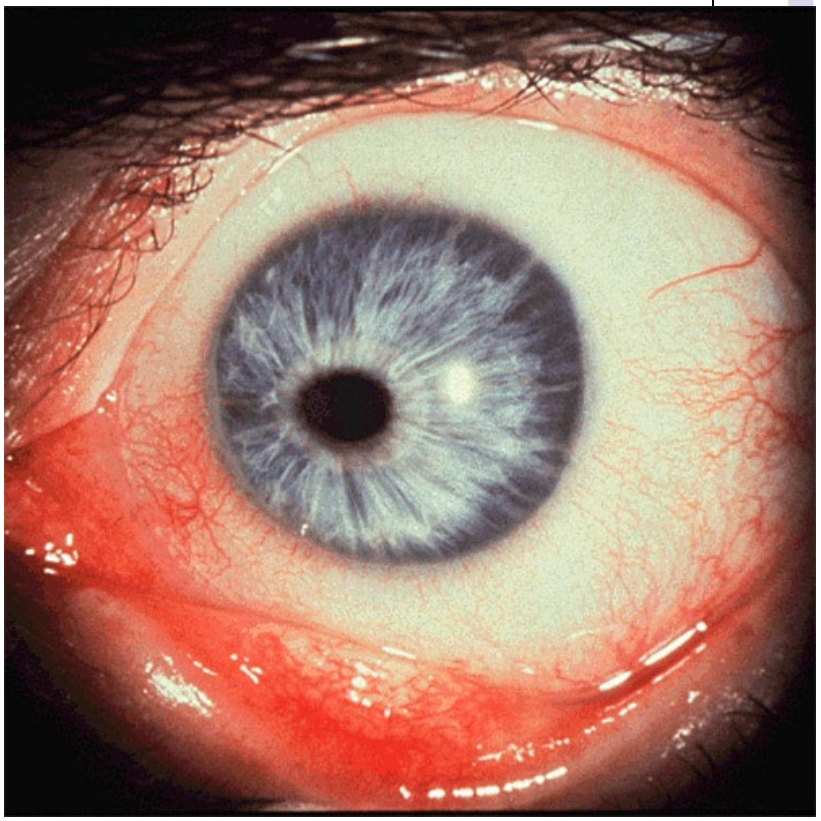

Corectopia

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Abnormalities of

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The displacement of the pupil from its normal central-ish location

*Why central-***ish***?*
Deviation from centrality of 1/2 mm is common, and up to 1 mm is considered normal

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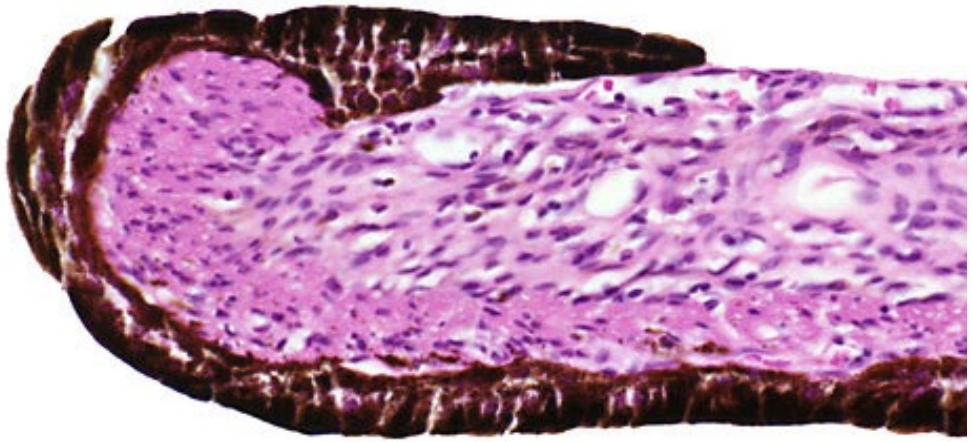
*What does the term* **ectropion uveae** *refer to?*
The presence of posterior pigmented iris epithelium on the anterior surface of the iris

# Developmental Abnormalities of the Cornea



Ectropion uveae

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} *Not simultaneously, obviously*

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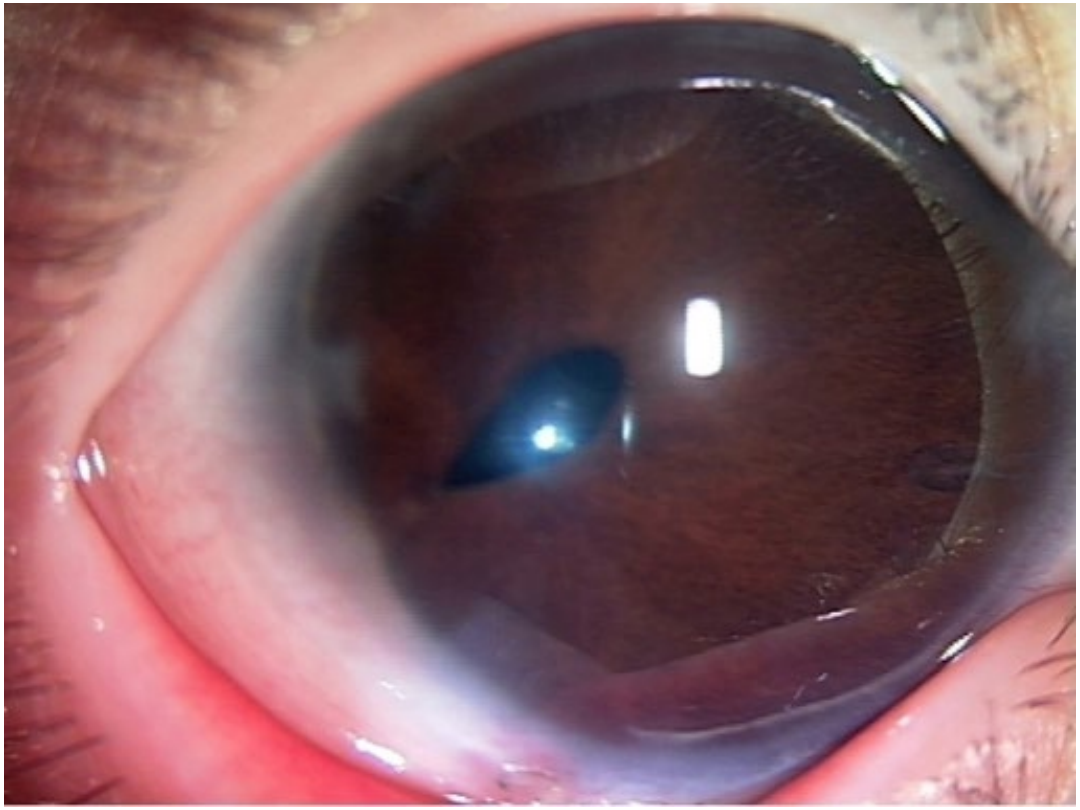

3 y.o. girl who presented at three months of age with hazy megalocornea, posterior embryotoxon, iris hypoplasia, corectopia, and early-onset severe glaucoma. The horizontal/vertical corneal diameters were 13.0/12.5 mm.

# Developmental Abnormalities of the Cornea

Axenfeld-Reiger with microcornea (8.5 mm)

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*What* nonocular *abnormalities may be present?*
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2)
3)
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Abnormalities of
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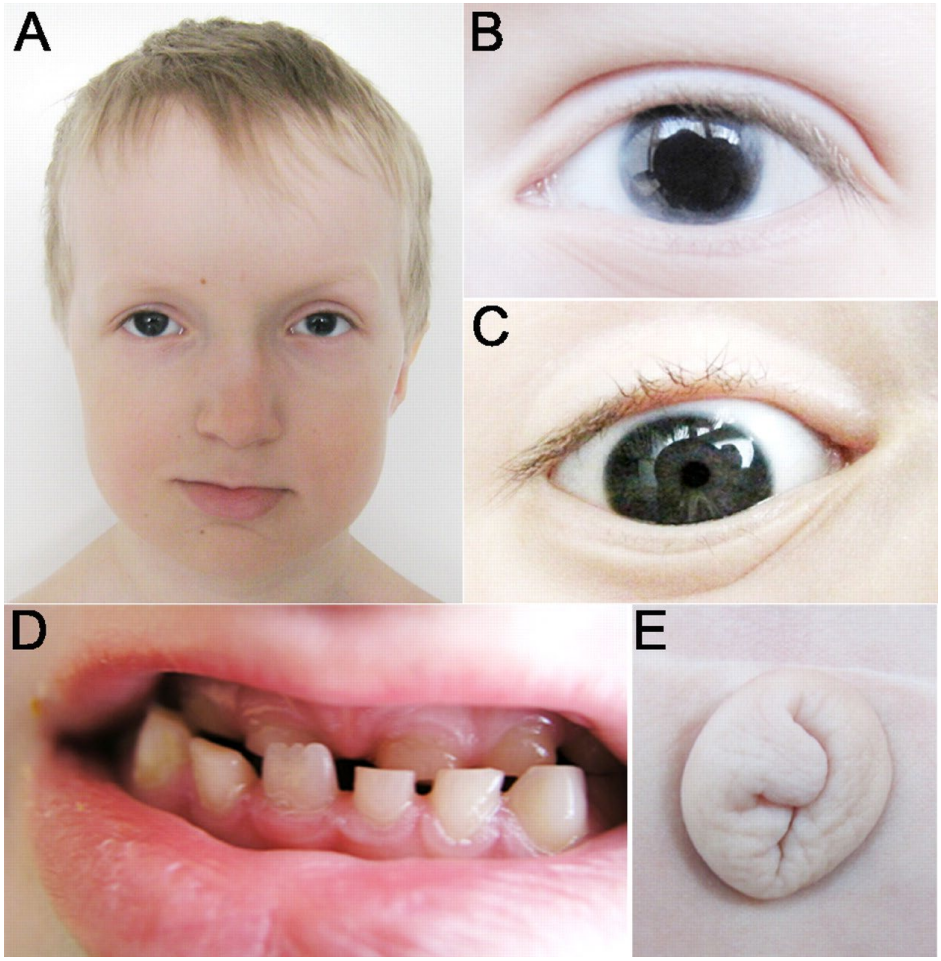
*What* nonocular *abnormalities may be present?*
1) Abnormal dentition
2) Characteristic facies
3) Periumbilical skin folds
4) Cardiac valve problems

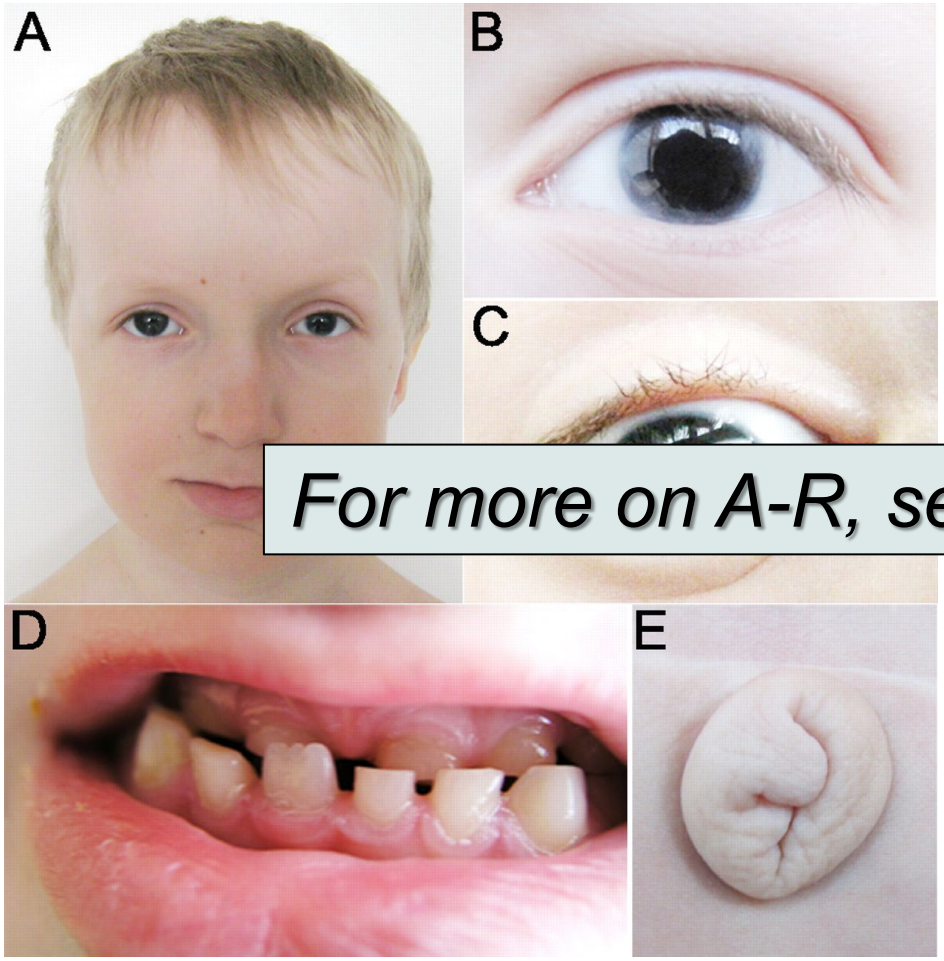
# Developmental Abnormalities of the Cornea




(A) Facial photograph showing maxillary hypoplasia, thin upper lip, and broad nasal bridge
(B) Left eye with corectopia
(C) Right eye with posterior embryotoxon
(D) Dental anomalies, including maxillary hypodontia
(E) Redundant periumbilical skin

Axenfeld-Reiger syndrome

# Developmental Abnormalities of the Cornea




(A) Facial photograph showing maxillary hypoplasia, thin upper lip, and broad nasal bridge

xon

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*For more on A-R, see slide-set* FELT7

Axenfeld-Reiger syndrome

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*Why is the term 'aniridia' technically a misnomer?*
Because a rudimentary iris root is always present

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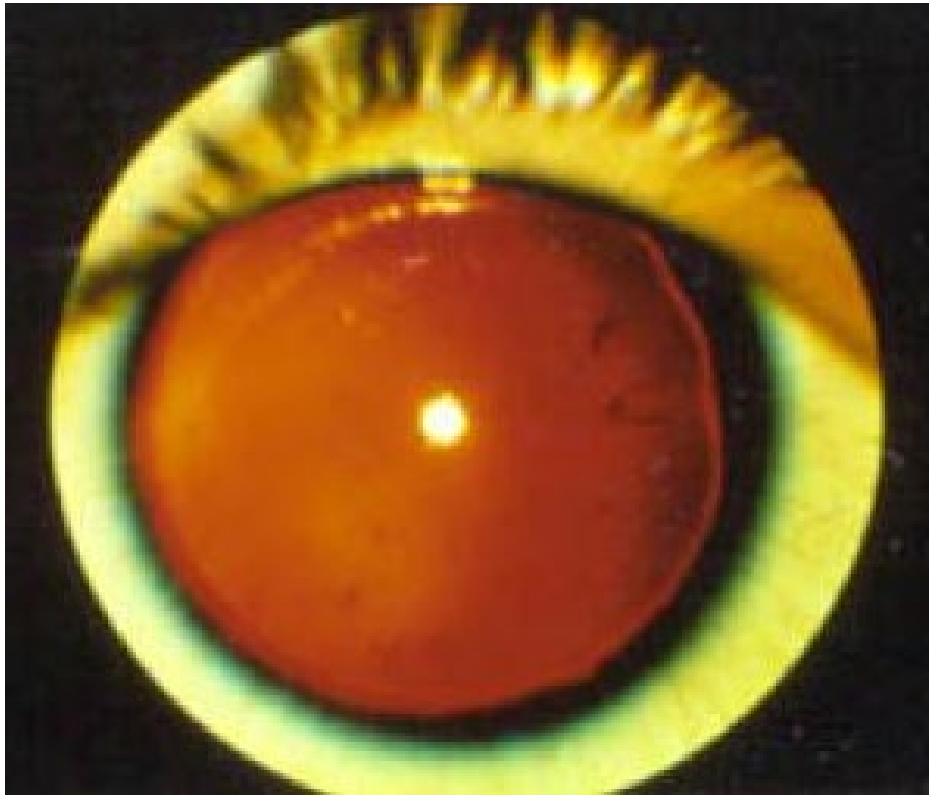
# Developmental Abnormalities of the Cornea



Aniridia. Note the presence of an iris stub/root

# Developmental Abnormalities of the Cornea

*What is a posterior embryotoxon?*
An anteriorly displaced and thickened Schwalbe's line/ring

*Why is the term 'aniridia' technically a misnomer?*
Because a rudimentary iris root is always present

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WAGR complex consists of:
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**G**
**R**

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WAGR complex consists of:
**W**ilms tumor
**A**niridia
**G**enitourinary abnormalities
**R**etardation

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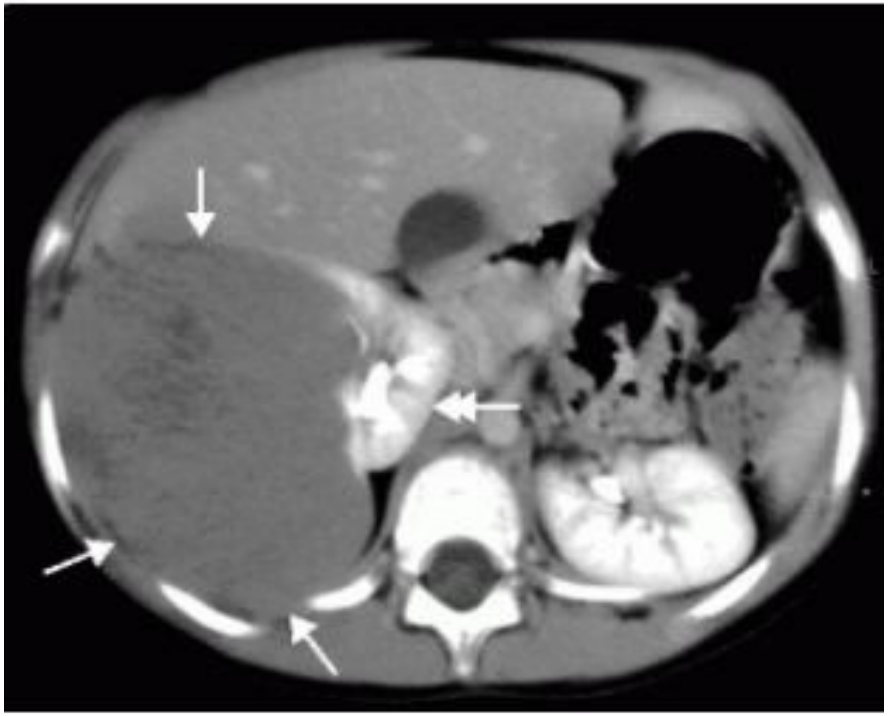

WAGR complex: Wilm's tumor

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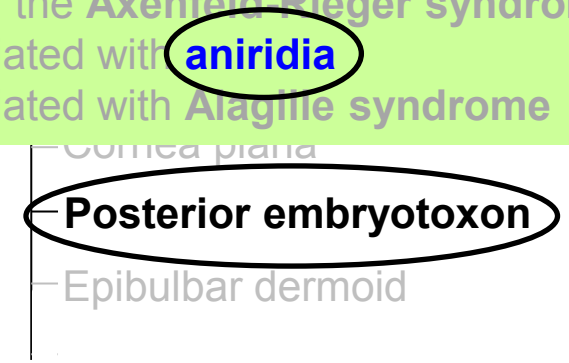

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*For more on aniridia, see slide-set P17*

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*To be clear—what is being asked of the eye service, ie, what are you supposed to check for in a 'rule out Alagille' consult?*
You are being asked to determine whether the infant has a posterior embryotoxon

**Alagille syndrome**

**Posterior embryotoxon**

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Alagille syndrome: Facies

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--The skeleton: The classic finding is 'butterfly vertebrae'

(Renal, neurologic and vascular abnormalities are also common)

# Developmental Abnormalities of the Cornea

Alagille syndrome: Butterfly vertebrae

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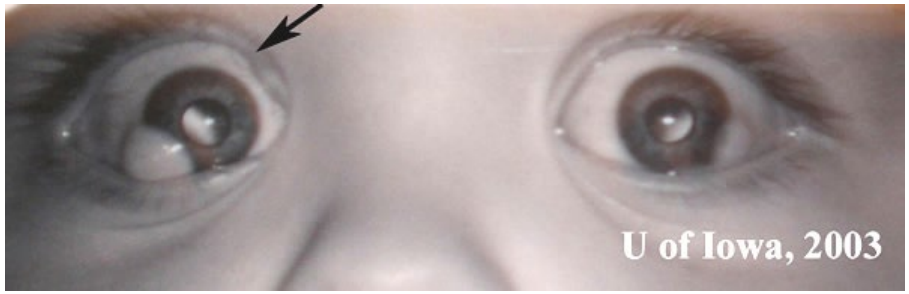
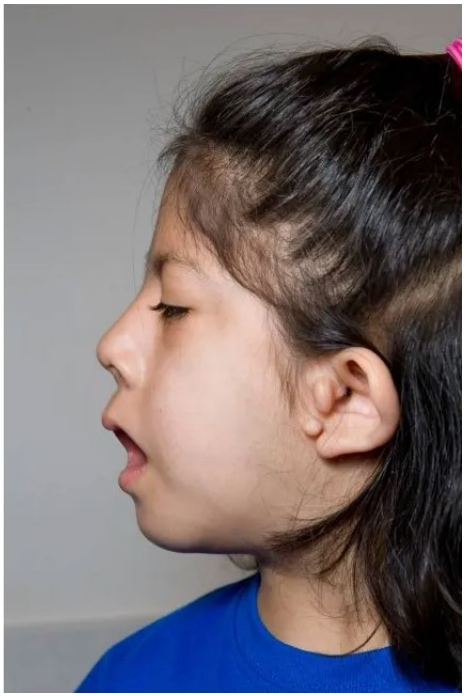
(Renal, neurologic and vascular abnormalities are also common)

# Developmental Abnormalities of the Cornea



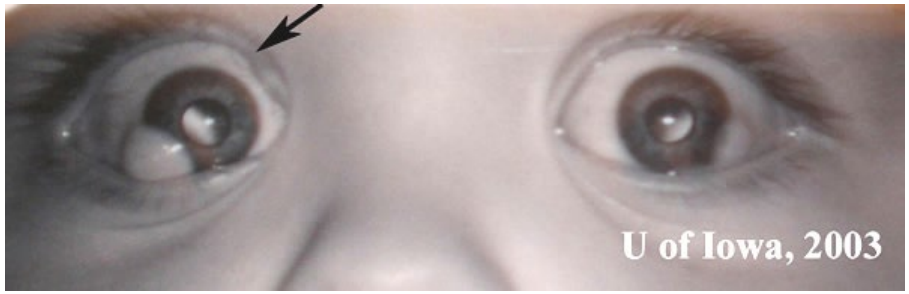

Goldenhar: Limbal (epibulbar) dermoids; lid coloboma



Goldenhar syndrome: Hemifacial microsomia

Goldenhar: Ear abnormalities

# Developmental Abnormalities of the Cornea

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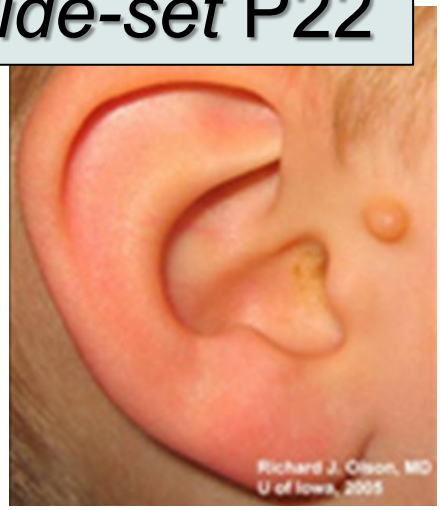
*For more on Goldenhar, see slide-set P22*

Goldenhar syndrome: Hemifacial microsomia




Goldenhar: Ear abnormalities

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
  - Megalocornea
  - Microcornea
  - Cornea plana
- Abnormalities of Corneal *Transparency*
  - Peripheral
    - Cornea plana
    - Posterior embryotoxon
    - **Epibulbar dermoid**
  - Central

*Are epibulbar dermoids hamartomas, or choristomas?*

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*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*What is the difference between a* hamartoma *and* choristoma*?*

# Developmental Abnormalities of the Cornea

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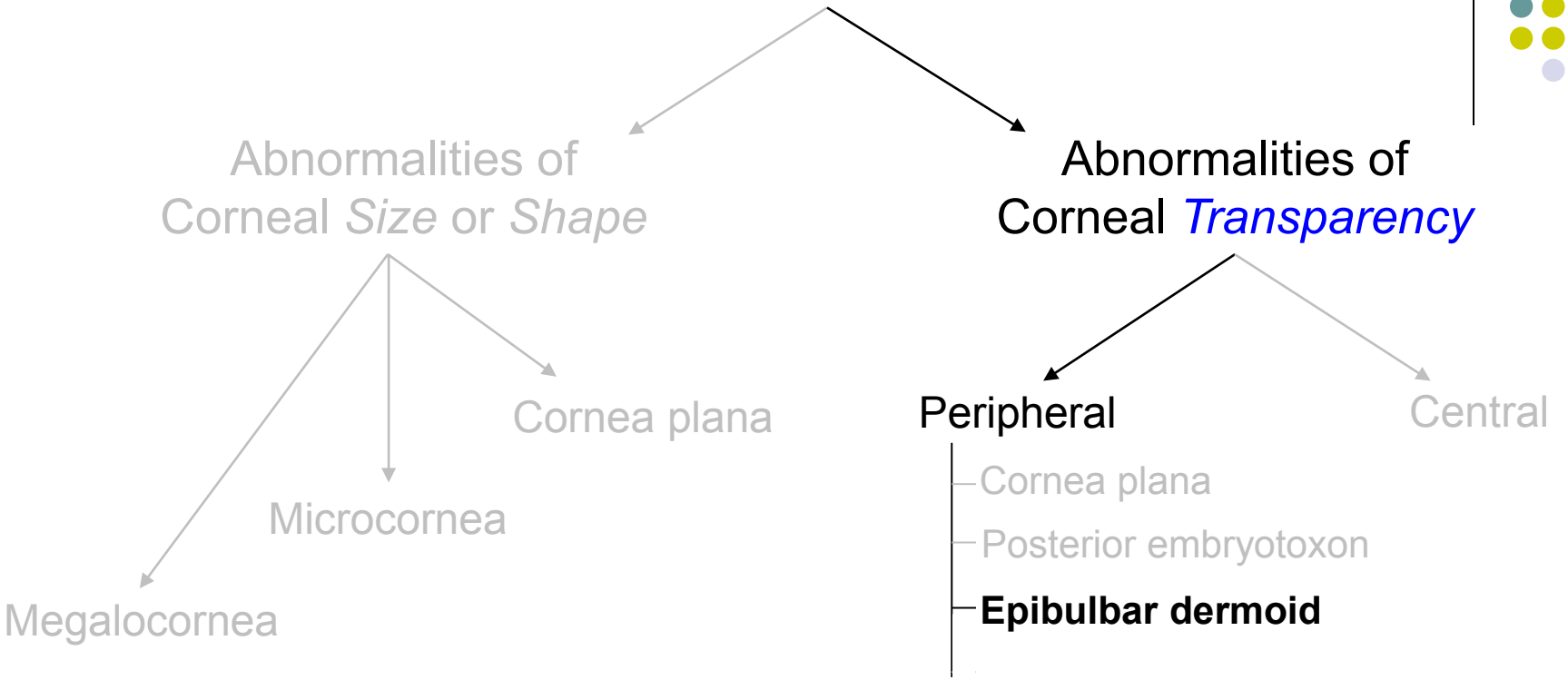
  - Central

*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*What is the difference between a* hamartoma *and* choristoma*?*
A one of them is a nest of abnormal cells in their normal location, whereas a the other is the opposite--normal cells in an abnormal location

# Developmental Abnormalities of the Cornea




*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*What is the difference between a* hamartoma *and* choristoma*?*
A hamartoma is a nest of abnormal cells in their normal location, whereas a choristoma is the opposite--normal cells in an abnormal location

# Developmental Abnormalities of the Cornea

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Choristomas

*What is the difference between a* hamartoma *and* choristoma*?*
A hamartoma is a nest of abnormal cells in their normal location, whereas a choristoma is the opposite--normal cells in an abnormal location

*So then, from what structure do the 'normal' choristoma cells of an epibulbar dermoid derive?*

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*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*What is the difference between a* hamartoma *and* choristoma*?*
A hamartoma is a nest of abnormal cells in their normal location, whereas a choristoma is the opposite--normal cells in an abnormal location

*So then, from what structure do the 'normal' choristoma cells of an epibulbar dermoid derive?*
The eyelid (the embryologic eyelid, that is)

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*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*With what syndrome are epibulbar dermoids associated?*

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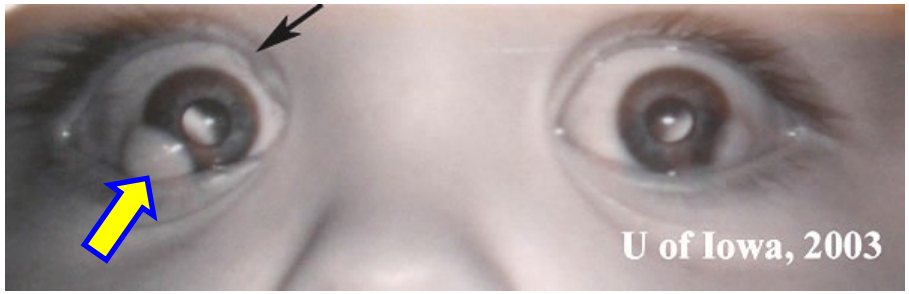
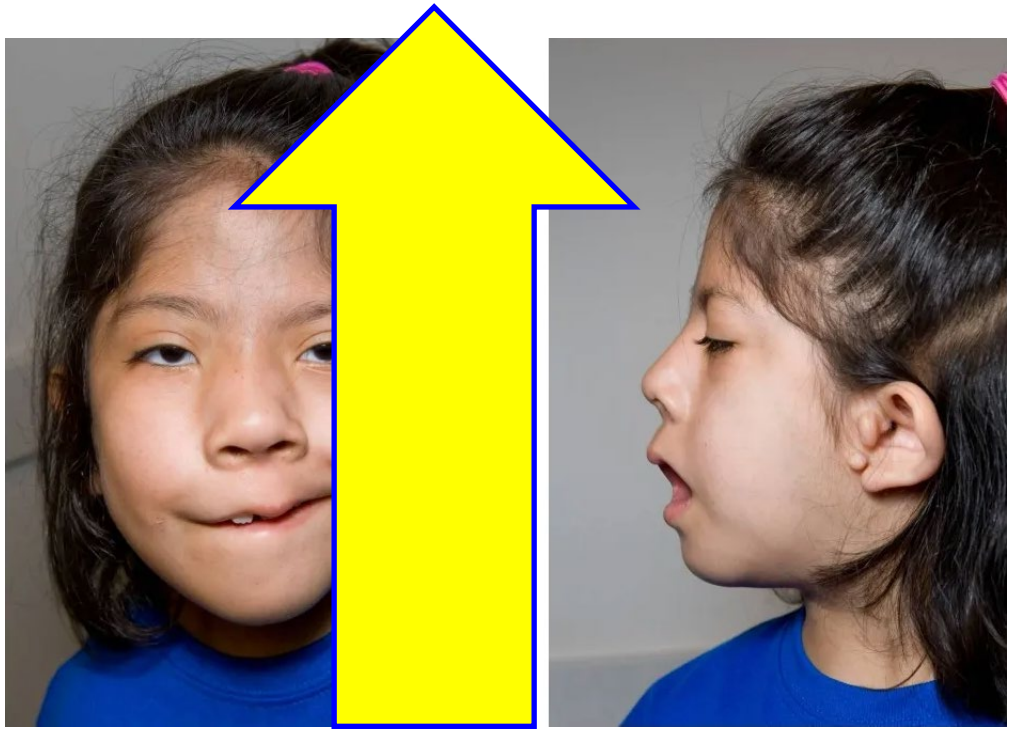
Choristomas

*With what syndrome are epibulbar dermoids associated?*
Goldenhar

# Developmental Abnormalities of the Cornea




**Goldenhar: Limbal (epibulbar) dermoids**; lid coloboma



Goldenhar syndrome: Hemifacial microsomia

Goldenhar: Ear abnormalities

# Developmental Abnormalities of the Cornea

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*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*With what syndrome are epibulbar dermoids associated?*
Goldenhar

*Where on the ocular surface are epibulbar dermoids typically located?*

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Choristomas

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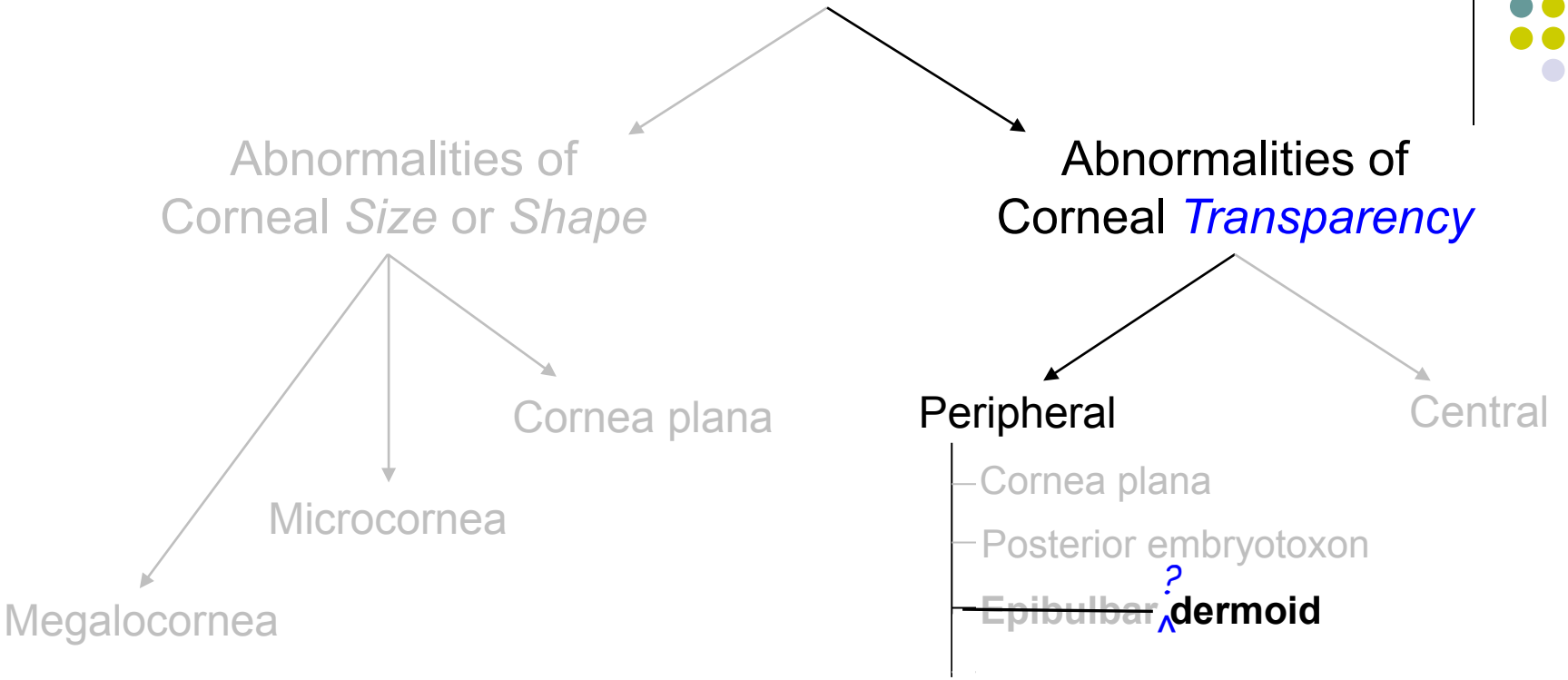
Goldenhar

*Where on the ocular surface are epibulbar dermoids typically located?*
At the limbus

# Developmental Abnormalities of the Cornea




*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*With what syndrome are epibulbar dermoids associated?*
Goldenhar

*Where on the oc*
At the limbus

*By what other name are epibulbar dermoids commonly known?*

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
  - Megalocornea
  - Microcornea
  - Cornea plana
- Abnormalities of Corneal *Transparency*
  - Peripheral
    - Cornea plana
    - Posterior embryotoxon
    - ~~Epibulbar~~ limbal dermoid
  - Central

*Are epibulbar dermoids hamartomas, or choristomas?*
Choristomas

*With what syndrome are epibulbar dermoids associated?*
Goldenhar

*Where on the oc*
**At the limbus**

*By what other name are epibulbar dermoids commonly known?*
**Limbal** dermoids

# Developmental Abnormalities of the Cornea

Goldenhar syndrome: Epibulbar (aka ‘limbal’) dermoid

# Developmental Abnormalities of the Cornea

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  - Peripheral
    - Cornea plana
    - Posterior embryotoxon
    - Epibulbar dermoid
  - Central
    - ?
    - ?
    - ?
    - ?
    - ?
    - ?
    - ?

# Developmental Abnormalities of the Cornea

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  - Peripheral
    - Cornea plana
    - Posterior embryotoxon
    - Epibulbar dermoid
  - Central
    - S
    - T
    - U
    - M
    - P
    - E
    - D

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
  - Megalocornea
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    - Cornea plana
    - Posterior embryotoxon
    - Epibulbar dermoid
  - Central
    - **S**
    - **T**
    - **U**
    - **M**
    - **P**
    - **E**
    - **D**

*STUMPED? What that even mean?*

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
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    - Epibulbar dermoid
  - Central
    - **S**
    - **T**
    - **U**
    - **M**
    - **P**
    - **E**
    - **D**

*STUMPED? What that even mean?*

It's a reference to the infamous STUMPED mnemonic for remembering the DDx for a two words in an infant.

# Developmental Abnormalities of the Cornea

- Abnormalities of Corneal *Size* or *Shape*
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  - Central
    - **S**
    - **T**
    - **U**
    - **M**
    - **P**
    - **E**
    - **D**

*STUMPED? What that even mean?*
It's a reference to the infamous STUMPED mnemonic for remembering the DDx for a cloudy cornea in an infant.

# Developmental Abnormalities of the Cornea

- **S**
- T
- U
- M
- P
- E
- D

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**
- **U**
- **M**
- **P**
- **E**
- **D**

*Next*

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**
- **M**
- **P**
- **E**
- **D**

(***T****ears in Descemet's membrane* works too)

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**
- M
- P
- E
- D

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**
- P
- E
- D

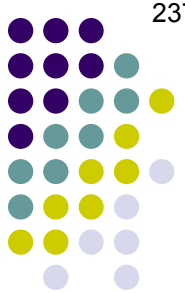
# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**
- E
- D

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**
- D

# Developmental Abnormalities of the Cornea



- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**ndothelial dystrophy (CHED)

*(CHED = congenital hereditary endothelial dystrophy)*

- D (***E****dema* works too)

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
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- **D**

# Developmental Abnormalities of the Cornea

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
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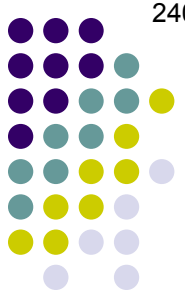
- **M**etabolic disorders
- **P**eters anomaly
- **E**ndothelial dystrophy (CHED)
- **D**ermoid of the cornea

# Developmental Abnormalities of the Cornea



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*For more on the STUMPED mnemonic, see slide-set* K9