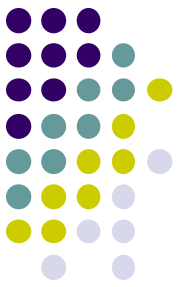


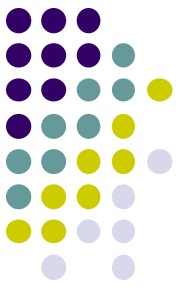
Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal dystrophies



Concerning Keratoconus

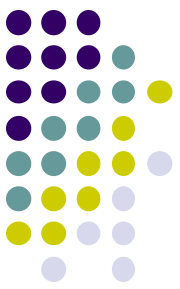
- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ~~dystrophies~~ *ectasias* **F**



Concerning Keratoconus

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How common is KCN, ie, what is the incidence?

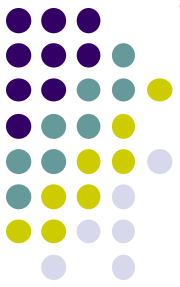


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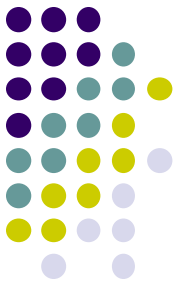
1 in 2000



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal **ectasias** F

Let's back up a step. Briefly, what is an ectasia?



Concerning Keratoconus

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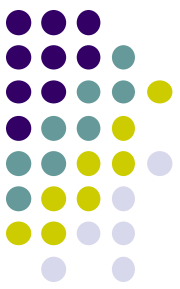
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Concerning Keratoconus



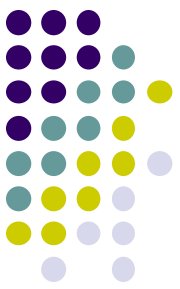
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A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal dystrophies **F**

ectasias

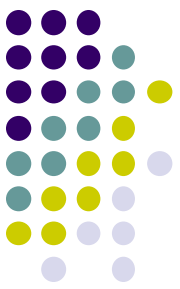
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A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

Very broadly (like, in one line), what is the ectatic process in KCN?

Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?

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ectasias

Let's back up a step. Briefly, what is an ectasia?

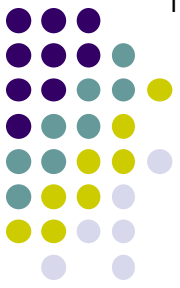
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A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

Very broadly (like, in one line), what is the ectatic process in KCN?

Progressive thinning of the **area** and/or **para-area** cornea leads to **shape** bulging of the cornea

Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal *ectasias* dystrophies F

Let's back up a step. Briefly, what is an ectasia?

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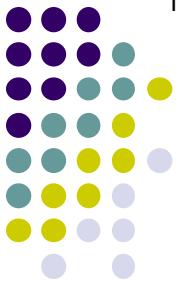
A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

Very broadly (like, in one line), what is the ectatic process in KCN?

Progressive thinning of the central and/or paracentral cornea leads to cone-like bulging of the cornea



Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal **dystrophies** F

Let's back up a step. Briefly, what is an ectasia?

To my friend **OK then: What is a dystrophy?**

in the E

following

A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

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Concerning Keratoconus



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Let's back up a step. Briefly, what is an ectasia?

To my f

OK then: What is a dystrophy?

in the E

A dystrophy is an inherited condition characterized by bilateral symmetric changes independent of environmental or systemic processes

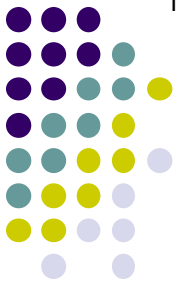
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Very broadly (like, in one line), what is the ectatic process in KCN?

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Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?

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Let's back up a step. Briefly, what is an ectasia?

To my friend *OK then: What is a dystrophy?*

in the E *A dystrophy is an inherited condition characterized by bilateral symmetric changes independent of*

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 A noninflammatory condition characterized by progressive thinning
 resulting in co

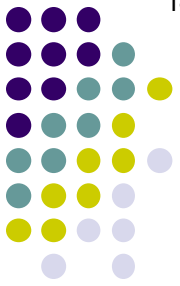
Very broadly

Progressive thinning of the central and/or paracentral cornea leads to cone-like bulging of the cornea

As we will soon see: Unlike a dystrophy, KCN has environmental and systemic associations aplenty!

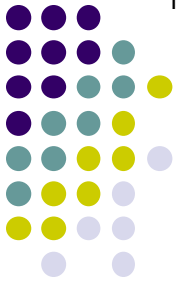
Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ~~dystrophies~~ *ectasias* **F**
 - It has a strong hereditary component



Concerning Keratoconus

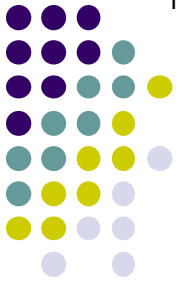
- Which of the following are true concerning keratoconus (KCN)?
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Concerning Keratoconus

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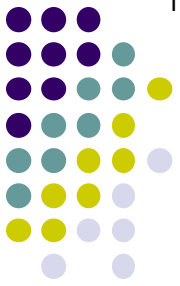
What percent of KCN cases have a positive family history?



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal *ectasias* ~~dystrophies~~ **F**
 - It has a ~~strong~~ *weak* hereditary component **F**

What percent of KCN cases have a positive family history?
5-10%



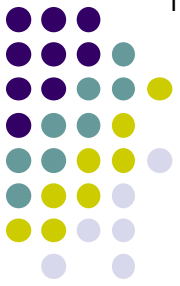
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 - It is one of the most common corneal ~~dystrophies~~ ^{ectasias} **F**
 - It has a ~~strong~~ ^{weak} hereditary component **F**
 - Fragmentation of Bowman's is present



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
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 - It has a ~~strong~~ ^{weak} hereditary component **F**
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Koncerning Keratoconus



Keratoconus: Bowman's is fragmented (3,4)



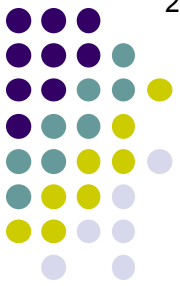
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Fragmentation of Bowman's is a histologic hallmark of KCN.

What are some others?

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Concerning Keratoconus

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What are some others?

- ^{stuff} deposition at the base of the cone
- Corneal ^{said this already}
- Folds/breaks in ^{layer}

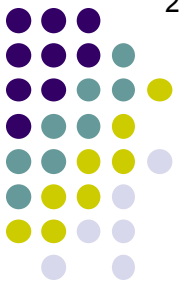
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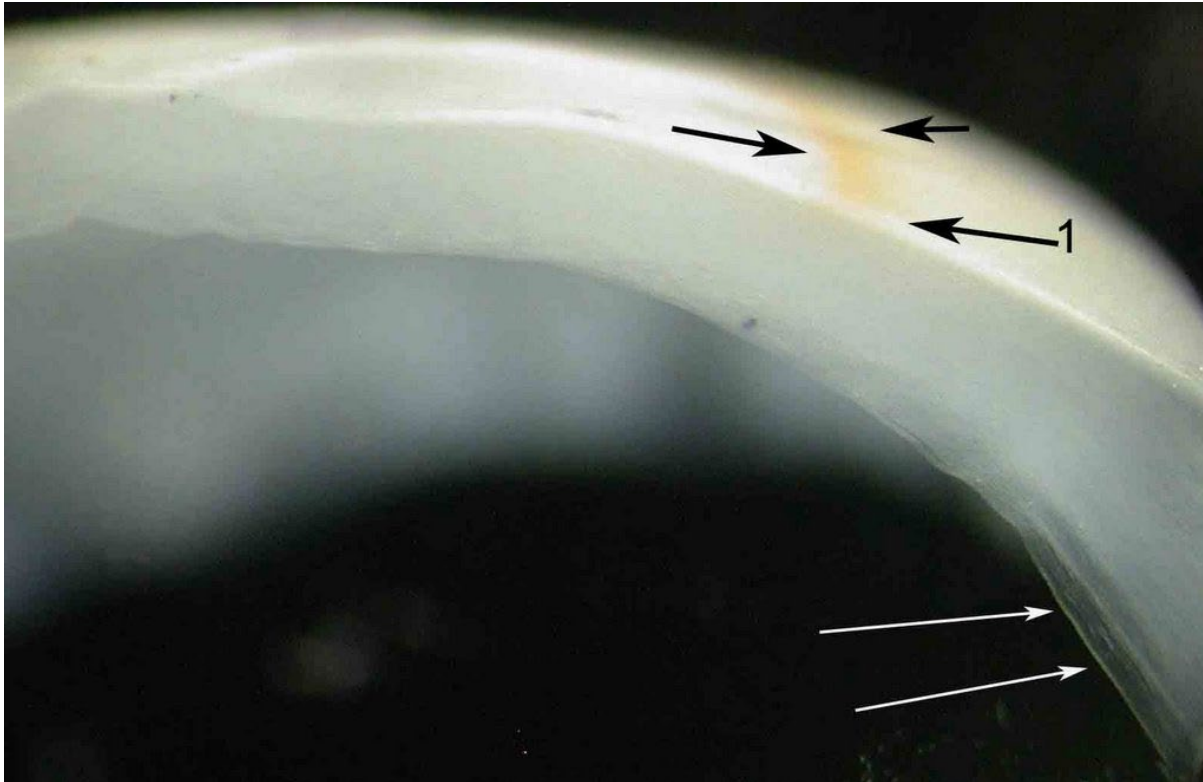
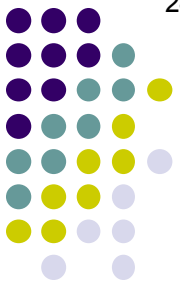
Fragmentation of Bowman's is a histologic hallmark of KCN.

What are some others?

- Iron deposition at the base of the cone
- Corneal thinning
- Folds/breaks in Descemet's

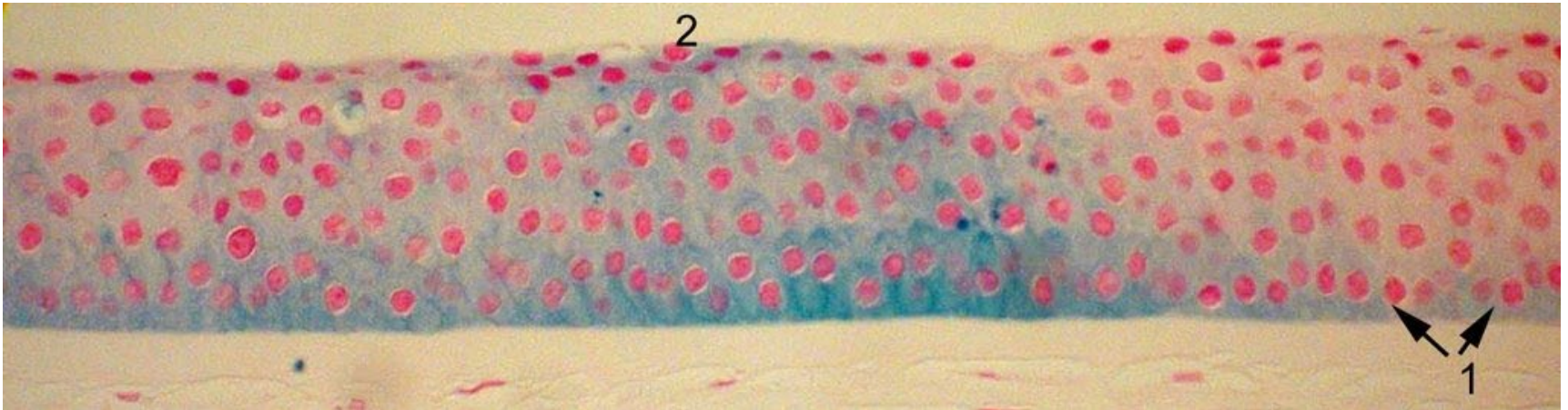


Koncerning Keratoconus



Keratoconus: Iron deposits in the epi (the brown stuff)

Concerning Keratoconus



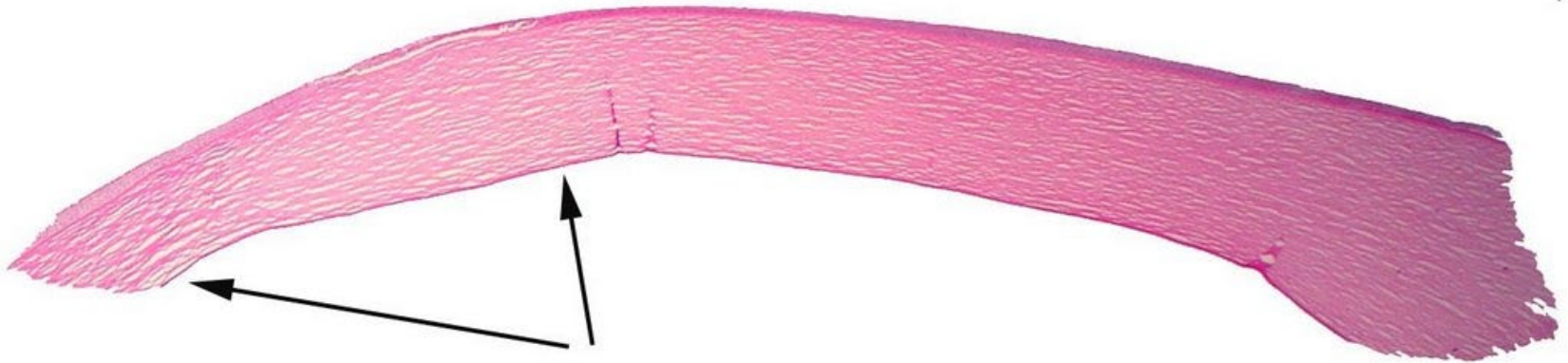
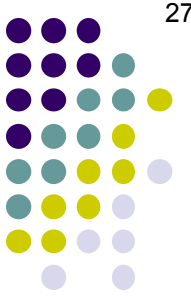
Keratoconus: Iron deposits in the epi (the blue stuff)

Concerning Keratoconus



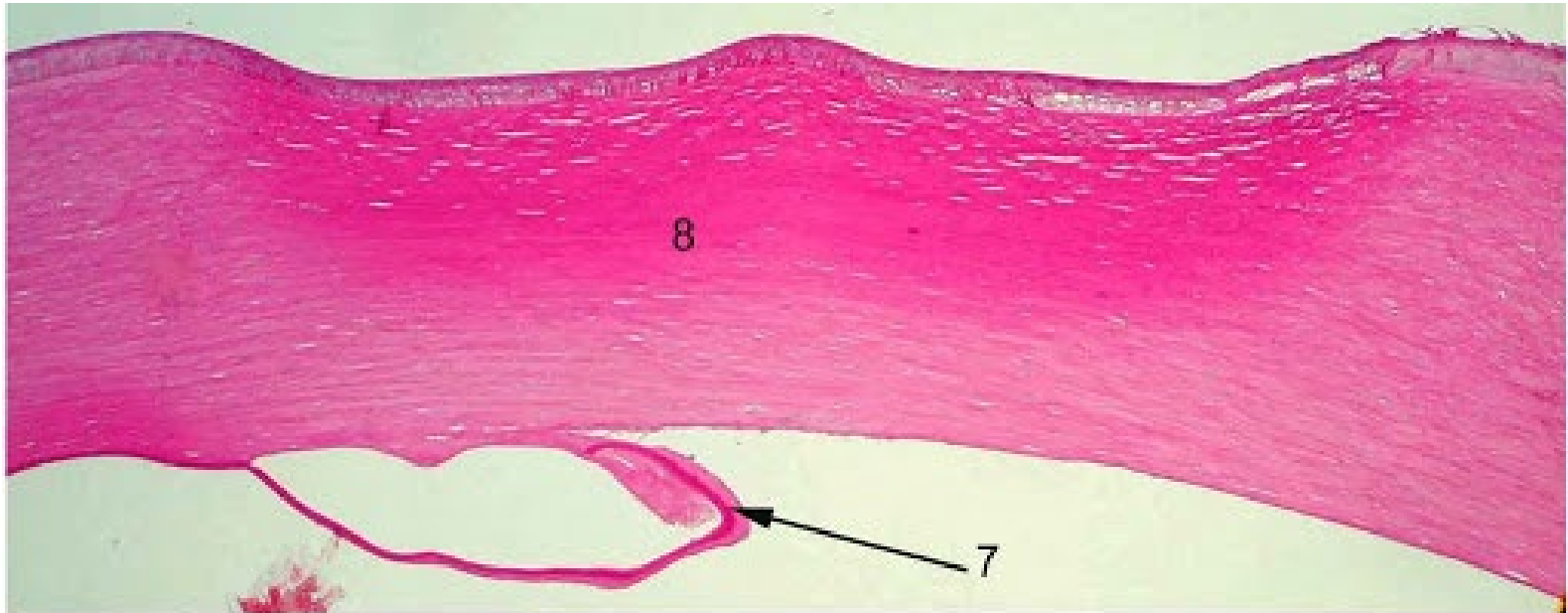
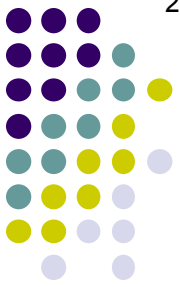
Keratoconus: Corneal thinning (take note of the central portion of the parallelepiped)

Koncerning Keratoconus

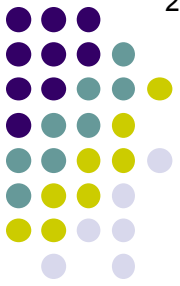


Keratoconus: Corneal thinning (at the arrows)

Concerning Keratoconus



Keratoconus: Descemet's membrane is disrupted and folded back to rejoin the cornea in the wrong orientation (7)



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ^{ectasias} dystrophies **F**
 - It has a ^{weak} ~~strong~~ hereditary component **F**
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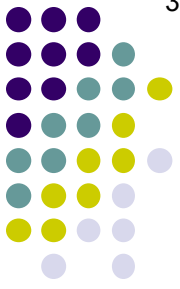
Fragmentation of Bowman's is a histologic hallmark of KCN.

What are some others?

-- **Iron deposition at the base of the cone**

-- Corneal thinning

*Iron deposition at the base of the cone leads to an important clinical sign of KCN.
What is the eponymous name of this sign?*



Concerning Keratoconus

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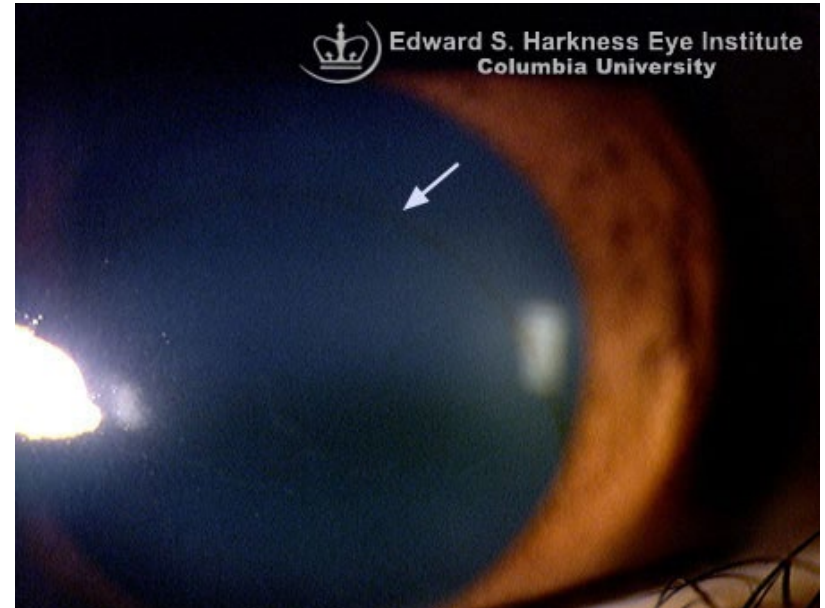
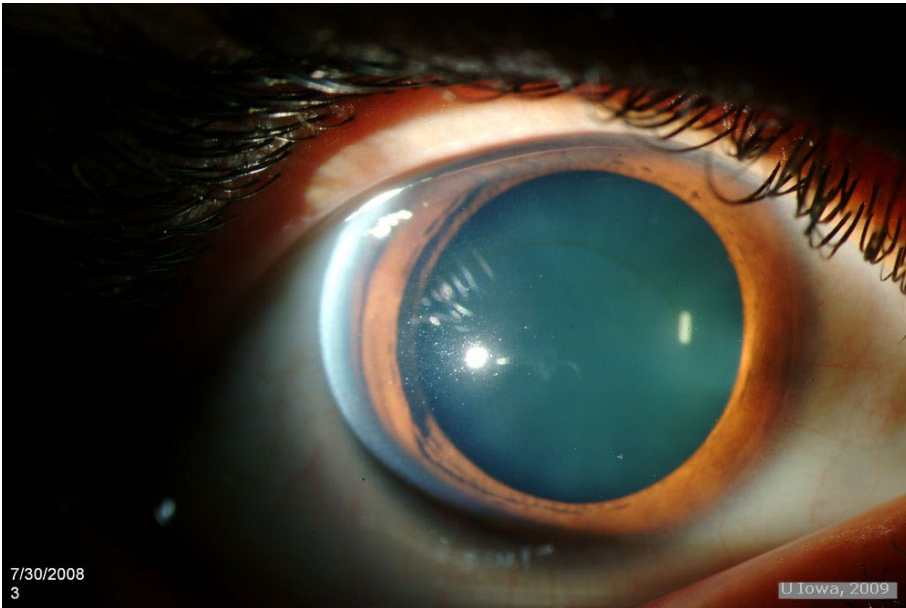
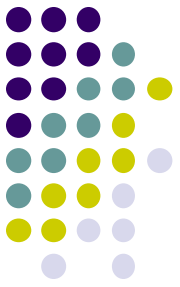
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What is the eponymous name of this sign?

Fleischer ring

Koncerning Keratoconus



KCN: Fleischer ring



Concerning Keratoconus

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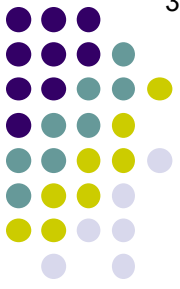
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What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring?



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What is the eponymous name of this sign?

Fleischer ring

What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring?

Examine the cornea with the **cobalt-blue light**



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-- Corneal thinning

Iron deposition at the base of the cone leads to an important clinical sign of KCN.

What is the eponymous name of this sign?

Fleischer ring

What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring?

Examine the cornea with the **cobalt-blue light**

Let's talk more generally about corneal iron lines...



- *There are four **corneal iron lines**. Name them.*

1) ?

2) **Fleischer line (ring)**

3) ?

4) ?



- *There are four **corneal iron lines**. Name them.*
 - 1) Stocker line
 - 2) Fleischer line (ring)
 - 3) Ferry line
 - 4) Hudson-Stähli line



- *There are four corneal iron lines. Name them. With what condition are they associated?*
 - 1) *Stocker line is associated with...*
 - ?
 - 2) Fleischer line (ring)
 - Keratoconus
 - 3) Ferry line
 - 4) Hudson-Stähli line



- *There are four **corneal iron lines**. Name them. With what condition are they associated?*
 - 1) *Stocker line is associated with...*
 - Pterygium
 - 2) Fleischer line (ring)
 - Keratoconus
 - 3) Ferry line
 - 4) Hudson-Stähli line

Koncerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) **Stocker line** *is associated with...*

- **Pterygium**

With respect to its associated pterygium, where is the Stocker line found?

4) Hudson-Stähli line

Koncerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

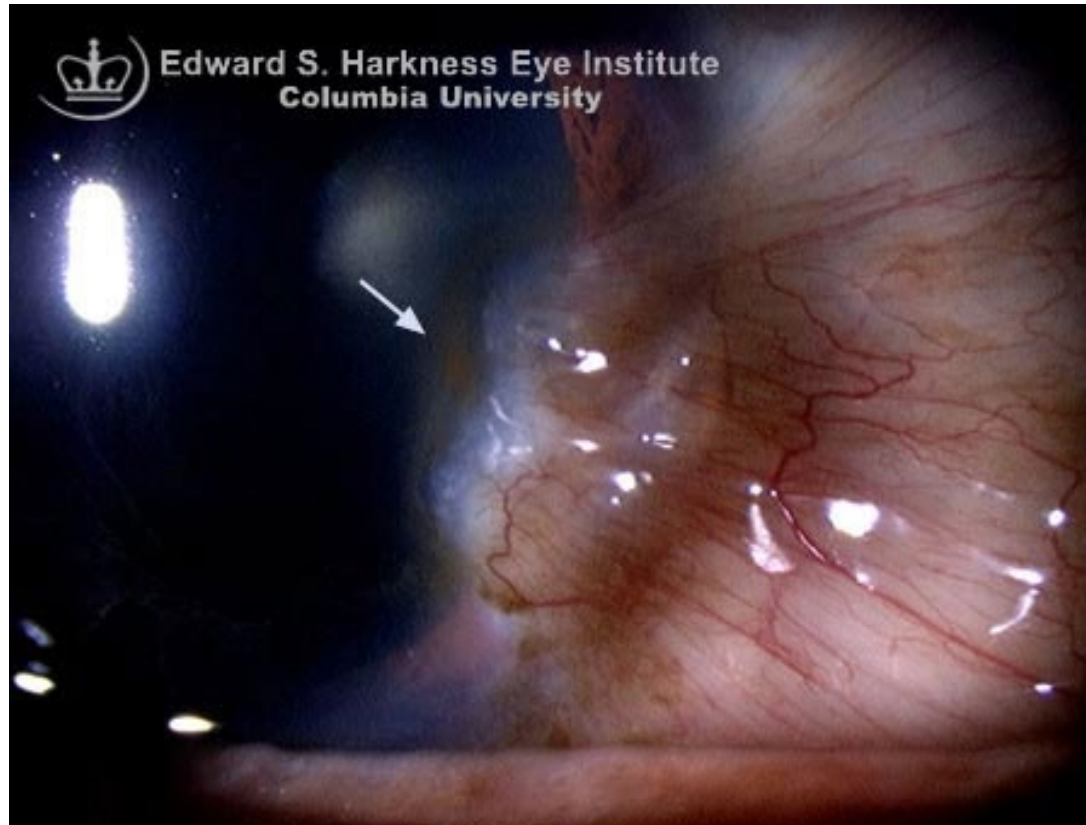
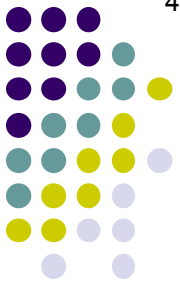
1) **Stocker line** *is associated with...*

- **Pterygium**

With respect to its associated pterygium, where is the Stocker line found?
Just anterior to the leading edge of the pterygium

4) Hudson-Stähli line

Koncerning Keratoconus



Stocker line

Koncerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) **Stocker line** *is associated with...*

- **Pterygium**

With respect to its associated pterygium, where is the Stocker line found?
Just anterior to the leading edge of the pterygium

Is the presence of a Stocker line an indication for pterygium removal?

4) Hudson-Stähli line



Koncerning Keratoconus

- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) **Stocker line** *is associated with...*

- **Pterygium**

With respect to its associated pterygium, where is the Stocker line found?
Just anterior to the leading edge of the pterygium

Is the presence of a Stocker line an indication for pterygium removal?
No—just the opposite. A Stocker line forms when the pterygium is stable; i.e., it indicates a *lack* of growth.

4) Hudson-Stähli line

Koncerning Keratoconus



- *There are four **corneal iron lines**. Name them. With what condition are they associated?*
 - 1) **Stocker line**
 - Pterygium
 - 2) **Fleischer line (ring)**
 - Keratoconus
 - 3) **Ferry line is associated with...**
 - ?
 - 4) Hudson-Stähli line

Koncerning Keratoconus



- *There are four **corneal iron lines**. Name them. With what condition are they associated?*
 - 1) **Stocker line**
 - Pterygium
 - 2) **Fleischer line (ring)**
 - Keratoconus
 - 3) **Ferry line is associated with...**
 - Filtering bleb
 - 4) Hudson-Stähli line

Concerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) Stocker line

- Pterygium

2) Fleischer line (ring)

- Keratoconus

3) **Ferry line** *is associated with...*

- Filtering bleb

4) Hudson-Jones line

With respect to the bleb, where is the Ferry line located?



Concerning Keratoconus

- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) Stocker line

- Pterygium

2) Fleischer line (ring)

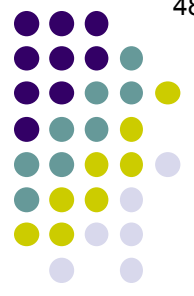
- Keratoconus

3) **Ferry line** *is associated with...*

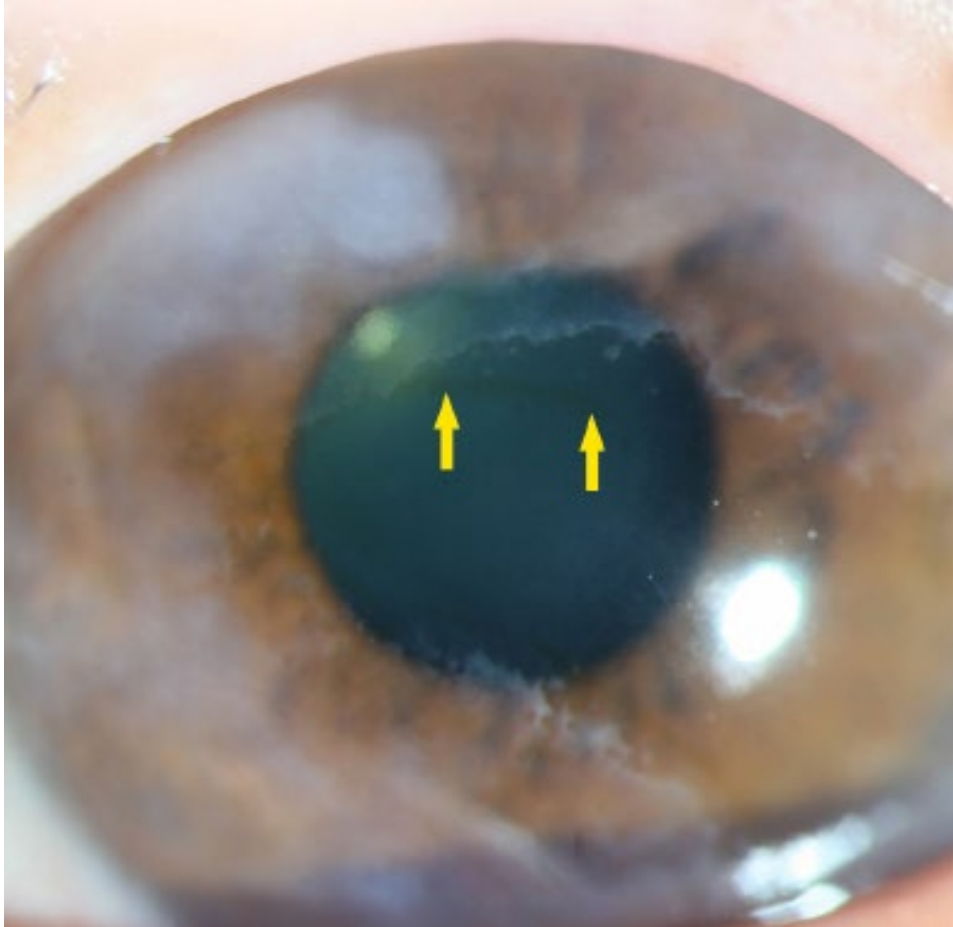
- Filtering bleb

4) Hudson-Sulzberger line

*With respect to the bleb, where is the Ferry line located?
Anterior to it*



Koncerning Keratoconus



Ferry line

Koncerning Keratoconus



- *There are four **corneal iron lines**. Name them. With what condition are they associated?*
 - 1) **Stocker line**
 - Pterygium
 - 2) **Fleischer line (ring)**
 - Keratoconus
 - 3) **Ferry line**
 - Filtering bleb
 - 4) **Hudson-Stähli line** *is associated with...*
 - ?



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 - 1) **Stocker line**
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 - 3) **Ferry line**
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 - 4) **Hudson-Stähli line** *is associated with...*
 - Normal and common finding in the elderly



- *There are four corneal iron lines. Name them. With what condition are they associated?*

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2) Fleischer line (ring)

- Keratoconus

3) Ferry line

- Filtering bleb

4) **Hudson-Stähli line** *is associated with...*

- Normal and common finding in the elderly

Where is the Hudson-Stähli line located?

Concerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

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2) Fleischer line (ring)

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4) **Hudson-Stähli line** *is associated with...*

- Normal and common finding in the elderly

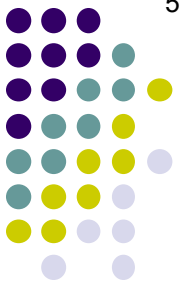
Where is the Hudson-Stähli line located?

At the junction of the lower- and middle-thirds of the cornea

Koncerning Keratoconus



Hudson-Stähli line



Concerning Keratoconus



- *There are four corneal iron lines. Name them. With what condition are they associated?*

1) Stocker line

For more on corneal iron lines, see slide-set K30

2) Fleischer line (ring)

- Keratoconus

3) Ferry line

- Filtering bleb

4) Hudson-Stähli line *is associated with...*

- Normal and common finding in the elderly



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ^{ectasias} dystrophies **F**
 - It has a ^{weak} ~~strong~~ hereditary component **F**
 - Fragmentation of Bowman's is present **T**

Fragmentation of Bowman's is a histologic hallmark of

KCN. What are some others?

KCN is not the only condition for which disruption of Bowman's is a histologic hallmark. For example: Which two corneal dystrophies bear disruption of Bowman's as their sine qua non?



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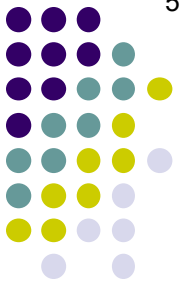
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How do you pronounce this?

How do you pronounce this?



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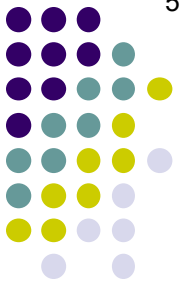
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Reis-Bückler and **Thiel-Behnke**

How do you pronounce this?
TEAL BEN-key

How do you pronounce this?
RICE BOO-kler



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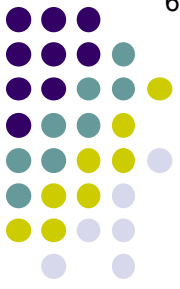
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What does TGFB1 stand for in this context?

In the most recent (2018-19) edition of the WHO Classification of Corneal Dystrophies, in what 'major category' are Reis-Bückler and Thiel-Behnke dystrophies classified? The 'Epithelial-stromal **TGFB1** dystrophies'



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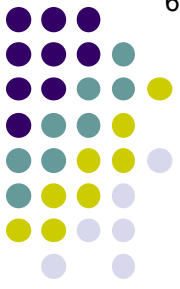
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TGFBI

What does TGFBI stand for in this context?
'Transforming growth factor beta induced'

What is TGFBI's chromosomal location?
5q31



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What does TGFBI stand for in this context?
'Transforming growth factor beta induced'

What is TGFBI's chromosomal location?
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The TGFBI gene was formerly known as what?



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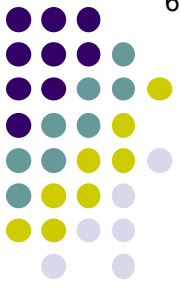
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The TGFBI gene was formerly known as what?
BIGH3 (this factoid is important because you might encounter this name in the older literature)



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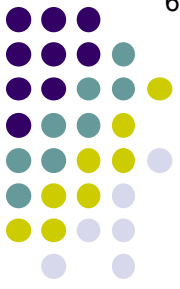
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The corneal-dystrophy section underwent a major revision for this version of the Cornea book. In what category were Reis-Bückler and Thiel-Behnke placed in previous editions?

In the most recent (2018-19) edition of the EBM, in what 'major category' are Reis-Bückler and Thiel-Behnke placed?

The 'Epithelial-stromal TGFBI dystrophies' formerly known as the...



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The CDBs---the 'Corneal Dystrophies of Bowmans'

In the most recent (2018-19) edition of the Cornea, in what 'major category' are Reis-Bückler and Thiel-Behnke placed?

The 'Epithelial-stromal TGFB1 dystrophies' formerly known as the...

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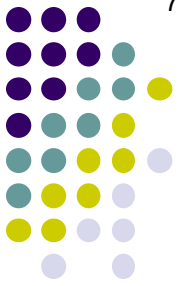
(PK = Penetrating keratoplasty, ie, a corneal transplant)

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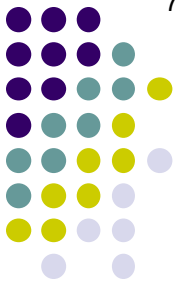
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The sudden development of severe
2ndry to a break in

two words

layer

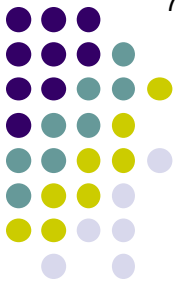


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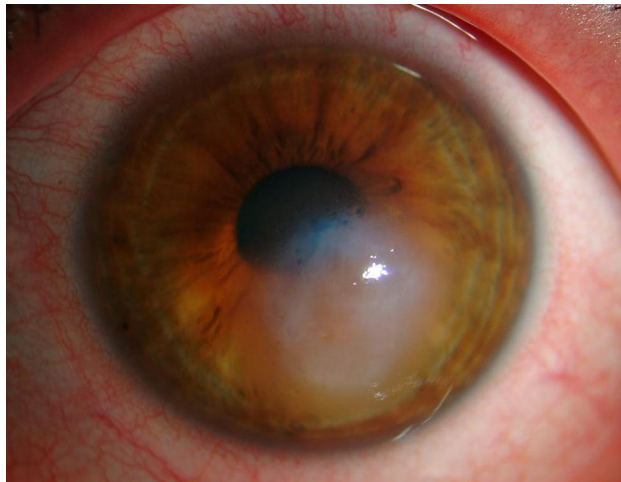
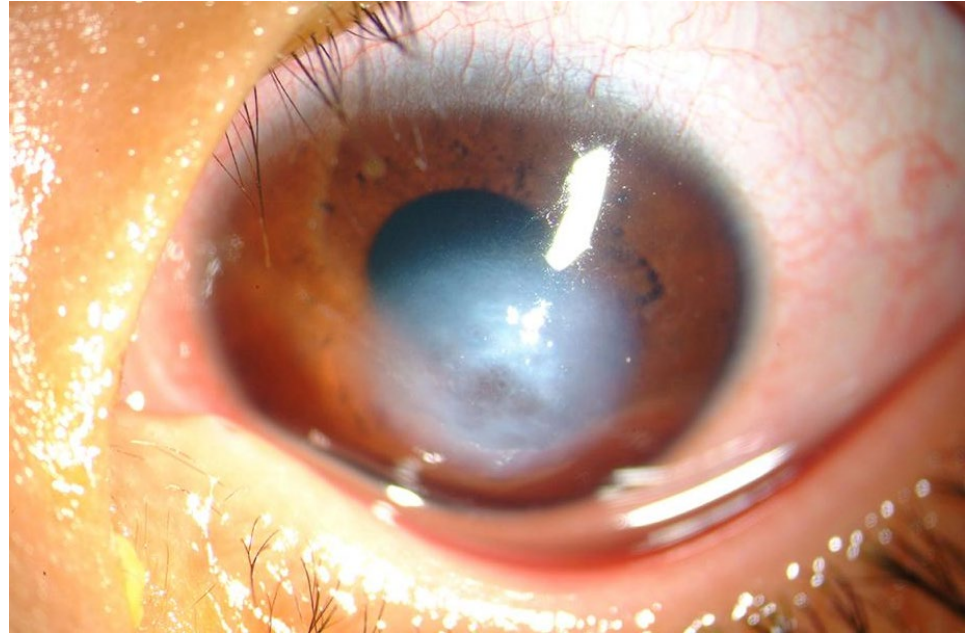
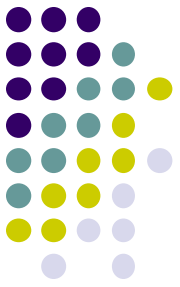
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What is acute hydrops?

The sudden development of severe corneal edema
 2ndry to a break in Descemet's



Koncerning Keratoconus



KCN: Acute hydrops

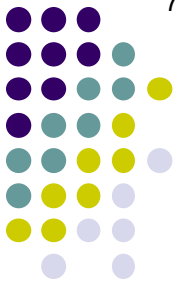
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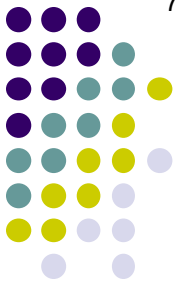
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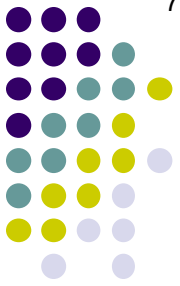
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With; two words usually occurs

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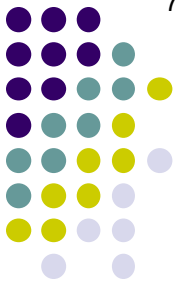
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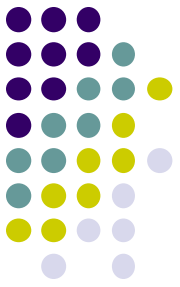
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With; apical scarring usually occurs

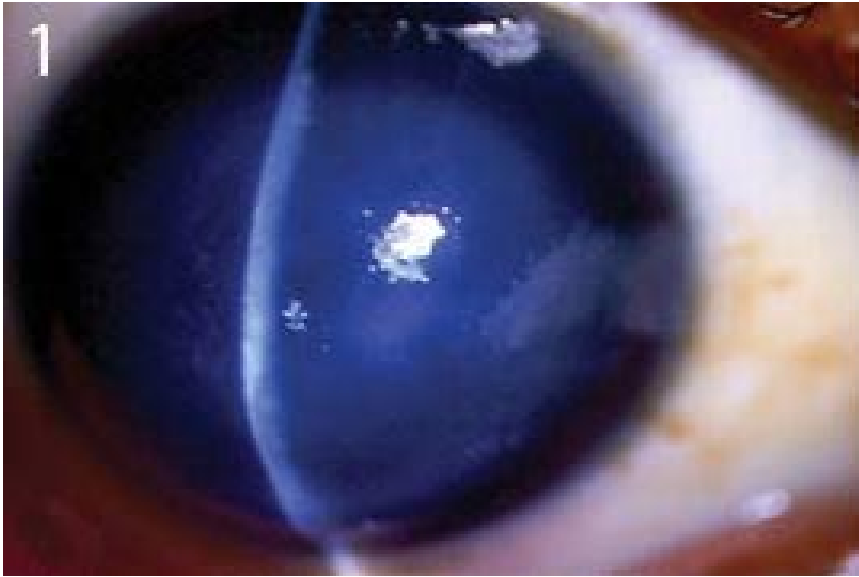


Koncerning Keratoconus



KCN: Apical scarring after acute hydrops

Koncerning Keratoconus

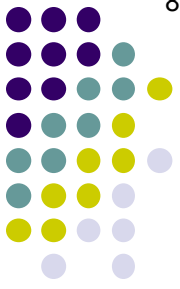


3 days after break



Scarring, same eye, 2+ months later

KCN: Acute hydrops



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'Apical scarring'--that sounds like a bad thing. Is it?



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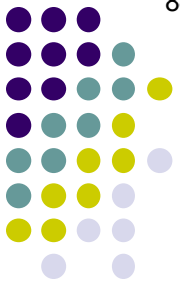
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Not necessarily—in some cases, the scarring flattens the cone, thereby reducing myopia and/or astigmatism

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Concerning Keratoconus



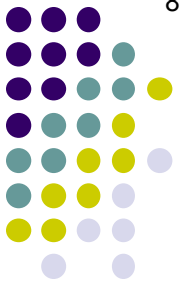
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During what period of life does KCN progress the fastest?



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During what period of life does KCN progress the fastest?
 Early on—teens to 20s



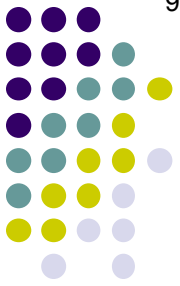
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 - Acute hydrops is ^{not} an indication for urgent PK **F**
 - The incidence is higher in South Asia and the Middle East **T**
 - Onset typically occurs during adolescence ~~childhood~~ **F**

During what period of life does KCN progress the fastest?
 Early on—teens to 20s

By what age does progression typically cease?

Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ^{ectasias} dystrophies **F**
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During what period of life does KCN progress the fastest?
 Early on—teens to 20s

By what age does progression typically cease?
 40



Concerning Keratoconus

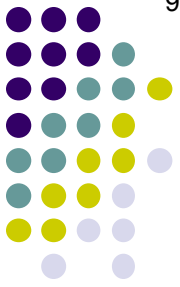
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There is a subset of KCN pts who do progress after 40. Regarding these pts, the Cornea book notes they tend to share a common systemic finding—one that would seem to be related to their KCN. What is that finding?

By what age does progression typically cease?

40

Concerning Keratoconus



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There is a subset of KCN pts who do progress after 40. Regarding these pts, the Cornea book notes they tend to share a common systemic finding—one that would seem to be related to their KCN. What is that finding?

About half of these late progressors are said to have "hyperelastic joints"

By what age does progression typically cease?

40



Concerning Keratoconus



- Which of the following are true concerning keratoconus (KCN)?
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 - Spontaneous rupture is fairly common

Concerning Keratoconus



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Concerning Keratoconus



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Concerning Keratoconus



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What is the most common pathologic corneal finding in Marfan's?

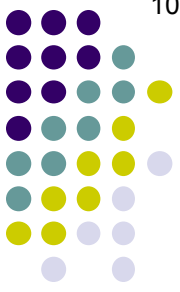


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What is the most common pathologic corneal finding in Marfan's?

An abnormally steep vs flat cornea



Concerning Keratoconus

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An abnormally flat cornea



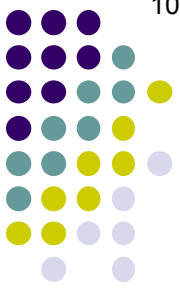
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How flat (in diopters) are we talking about here?



Concerning Keratoconus

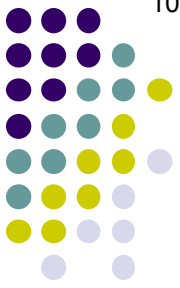
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Values in the 35D range are common



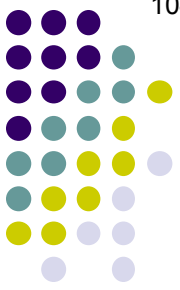
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What is the dioptric power of a typical 'normal' cornea?

most common pathologic corneal
Marfan's?
usually flat cornea

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Concerning Keratoconus

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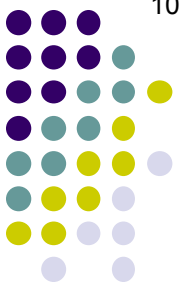
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Concerning Keratoconus

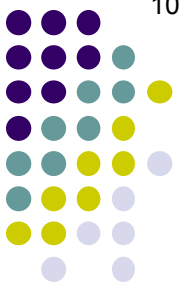
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What is the dioptric power of an advanced KCN cornea (at the cone)?

the 35D range



Concerning Keratoconus

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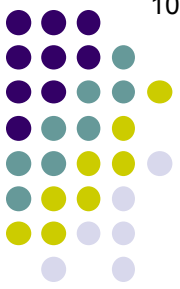
Around 43D

What is the dioptric power of an advanced KCN cornea (at the cone)?

Values >50D are the rule, and >60D are not uncommon

Values in **the 35D range** are common

Concerning Keratoconus



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With what conditions is KCN associated?

Ocular only:

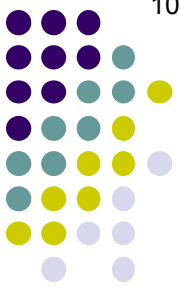
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Systemic:

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--

List not exhaustive,
obviously

Concerning Keratoconus



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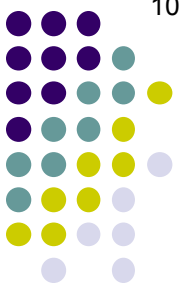
Ocular only:

- Leber's congenital amaurosis
- Vernal keratoconjunctivitis

Systemic:

- Down syndrome
- Ehler's-Danlos syndrome
- Osteogenesis imperfecta

List not exhaustive,
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Concerning Keratoconus

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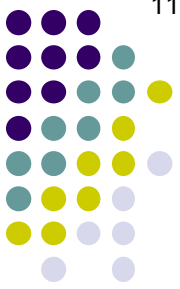
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What is the common thread among these?

Concerning Keratoconus



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Systemic:

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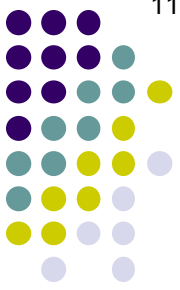
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Eye rubbing

Concerning Keratoconus



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Ocular only:

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--**Vernal keratoconjunctivitis**

Systemic:

--**Down syndrome**

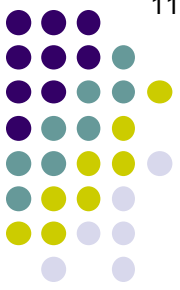
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Why do VKC and Down pts rub their eyes?



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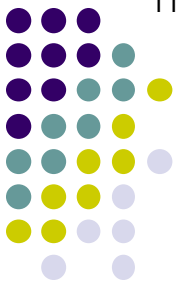
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Eye rubbing

Why do VKC and Down pts rub their eyes?

Because they itch

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● *Why do Leber's pts rub their eyes—do they itch as well?*

No, their motivation is very different. Recall that Leber's pts have extremely low vision from a very young age. Because of this lack of visual stimulation, Leber's pts will rub their eyes in order to mechanically stimulate the retina, thereby producing the false visual impression of lights known as .

What is the common thread among these?

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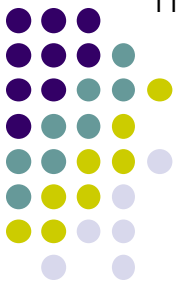
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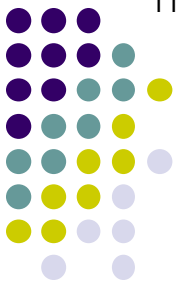
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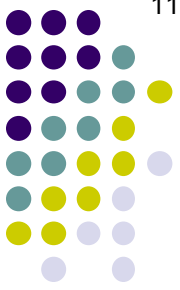
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What is the common thread among these?

Abnormal connective tissue



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What is the common thread among these?

Eye rubbing

Eye rubbing is one of the 'environmental factors' referred to early on in the slide-set. The Cornea book lists four others—what are they?

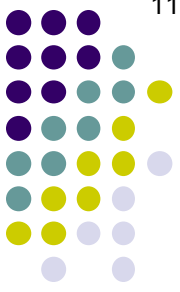
--Eye rubbing

--

--

--

--



Concerning Keratoconus

- Which of the following are true concerning keratoconus (KCN)?
 - It is one of the most common corneal ~~dystrophies~~ ^{ectasias} **F**
 - It has a ~~strong~~ ^{weak} hereditary component **F**
 - Fragmentation of Bowman's is present **T**
 - Acute hydrops is ~~an~~ ^{not} indication for urgent PK **F**
 - The incidence is higher in South Asia and the Middle East **T**
 - Onset typically occurs during ~~childhood~~ ^{adolescence} **F**
 - Spontaneous rupture is ~~fairly common~~ ^{extremely rare} **F**
 - KCN is ~~strongly~~ ^{uncommonly} associated with Marfan syndrome **F**

With what conditions is KCN associated?

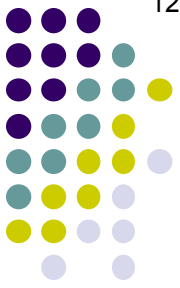
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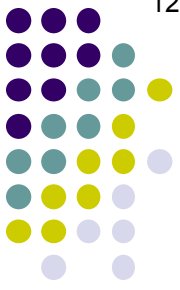
- Eye rubbing
- Atopy
- Rigid CL wear
- Inflammation
- Oxidative stress

Concerning Keratoconus



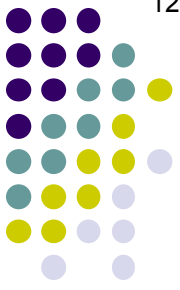
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 - Females are more likely to be affected than males

Concerning Keratoconus



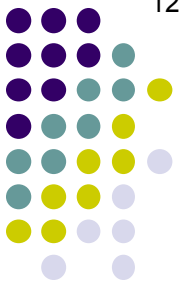
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Concerning Keratoconus



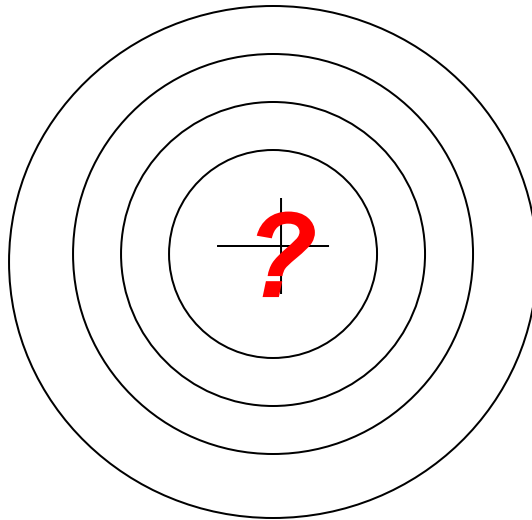
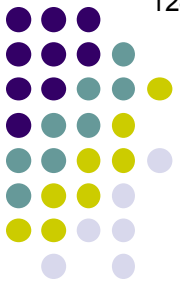
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 - Corneal topography reveals superior steepening

Concerning Keratoconus



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 - Females are more likely to be affected than males **T**
 - Corneal topography reveals ~~superior~~ ^{inferior} steepening **F**

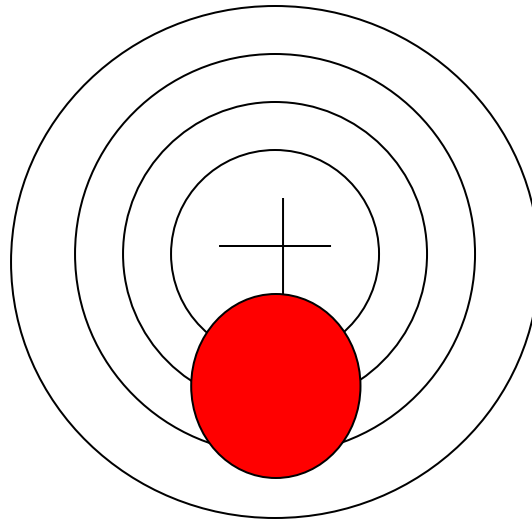
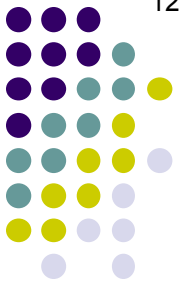
Concerning Keratoconus



Topography in keratoconus reveals

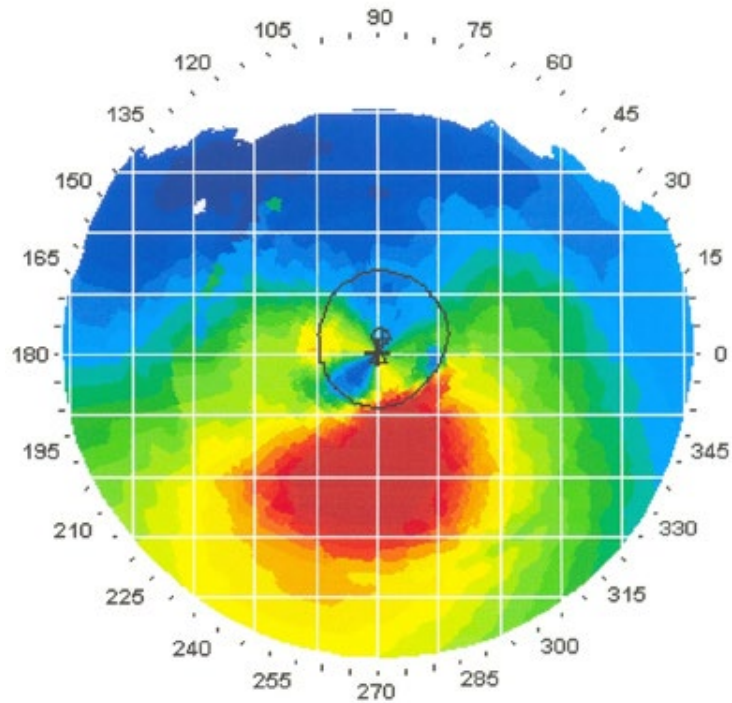
three words

Concerning Keratoconus



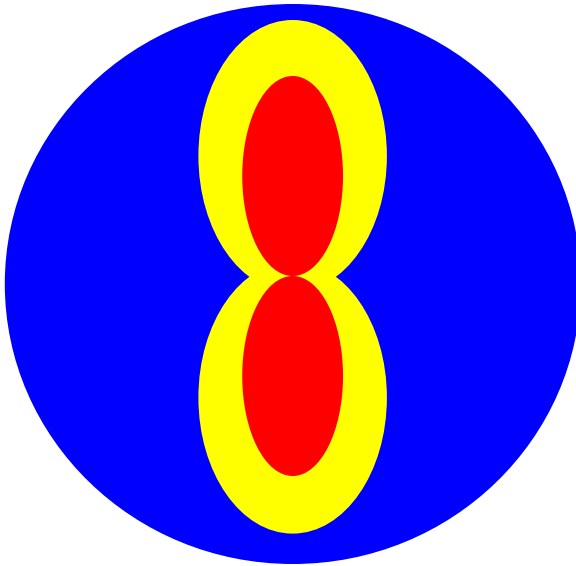
Topography in keratoconus reveals
inferior corneal steepening

Concerning Keratoconus

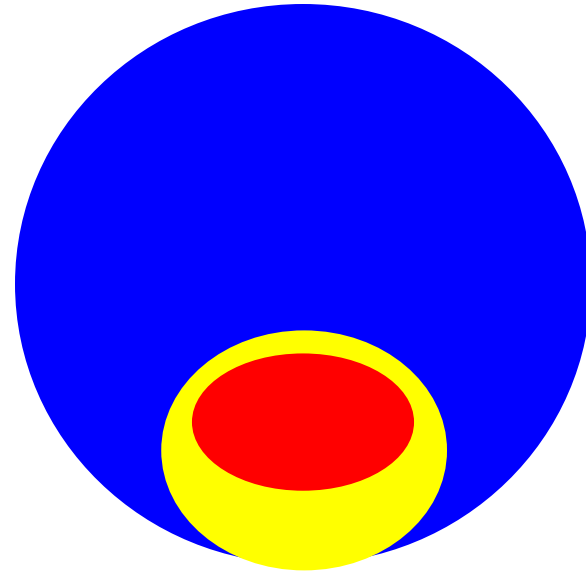


Topography in keratoconus reveals
inferior corneal steepening

Concerning Keratoconus



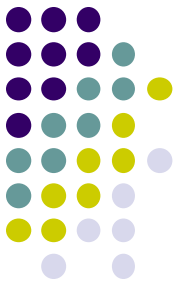
?



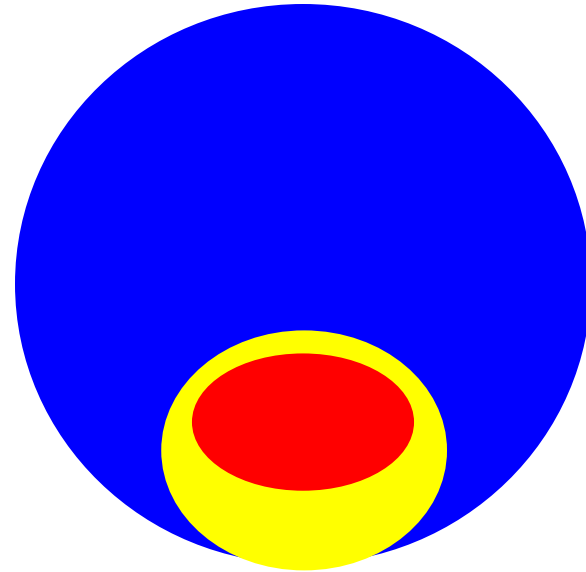
?

What corneal shape is represented?

Koncerning Keratoconus



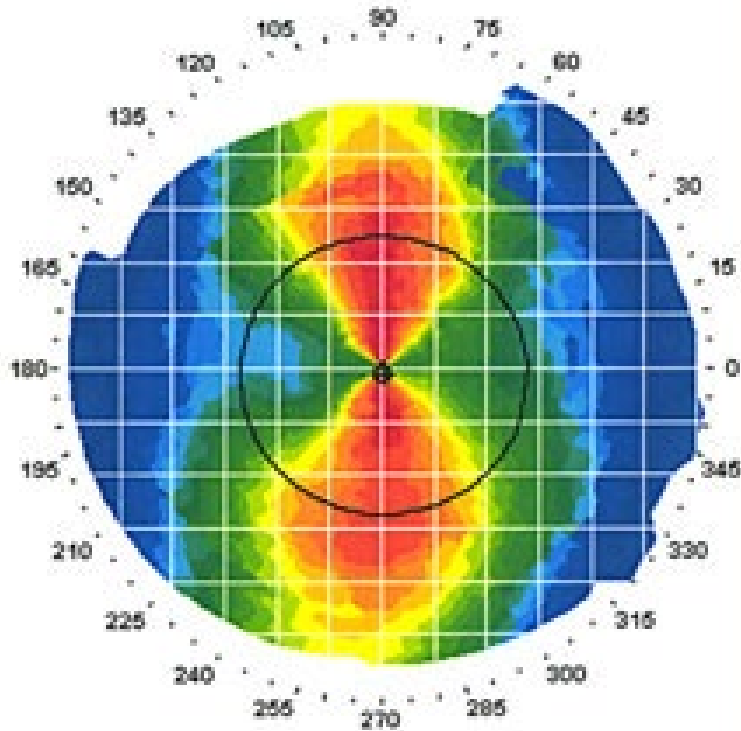
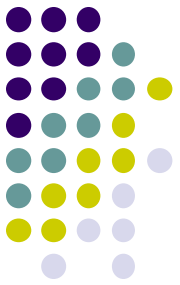
With-the-rule astigmatism



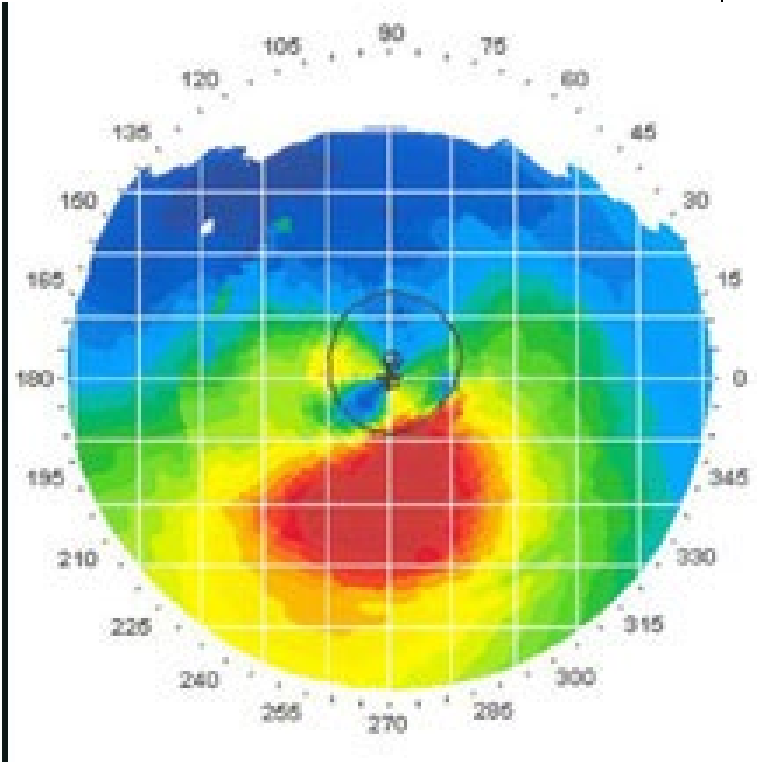
KCN

What corneal shape is represented?

Koncerning Keratoconus



With-the-rule astigmatism



KCN

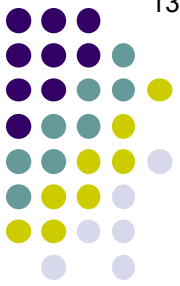
Koncerning Keratoconus



KCN is a fairly common (~1/2,000) noninflammatory ectasia of the cornea. It displays a weak hereditary pattern, with a positive family history in 5-10% of cases. The central and/or paracentral cornea thins progressively and bulges out like a cone. Extreme irregular astigmatism eventually results. Progression usually occurs during adolescence; the cornea tends to stabilize in early adulthood. Histology is characterized by fragmentation of Bowman's, thinning of the stroma and overlying epithelium, and folds or frank breaks in Descemet's. Disruption of Descemet's allows ingress of aqueous, resulting in the acute opacification of the cornea known as *hydrops*. With time, the endothelium will seal the breach and deturgesce the cornea. Scarring post-hydrops is common and may necessitate PK. Occasionally, however, the scarring flattens the central cornea, thereby reducing astigmatism and *improving* vision.

Management is dictated by the status of the cornea. Early in the disease course the astigmatism may be correctable with spectacles. At some point RGP CLs will be needed to neutralize the ever-worsening astigmatism. Many corneas go to PK as the disease progresses further or the patient becomes CL-intolerant. PK is highly successful. KCN has been reported to recur in the graft, but it is unclear whether this represents true recurrence vs progression in the residual host bed. Intrastromal corneal rings (Intacs) show promise as a less-invasive surgical correction, especially when coupled with corneal cross-linking.

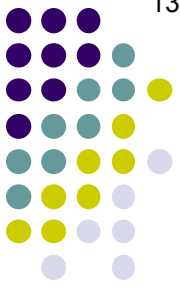
Summary slide--no questions



● What are the 5 classic signs of keratoconus?
Which is the first to appear?

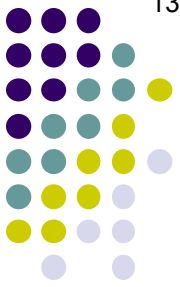
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Concerning Keratoconus



- What are the 5 classic signs of keratoconus?
Which is the first to appear?
 - Scissoring of the retinoscopic reflex (earliest sign)
 - Rizzuti's sign
 - Munson's sign
 - Fleischer ring
 - Vogt lines

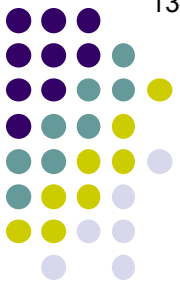
Koncerning Keratoconus



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Define Rizzuti's sign:

Concerning Keratoconus



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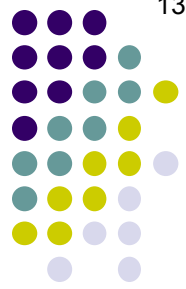
Define Rizzuti's sign:

A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

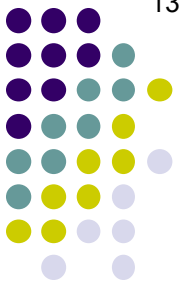
Koncerning Keratoconus



Keratoconus: Rizzuti's sign



Koncerning Keratoconus



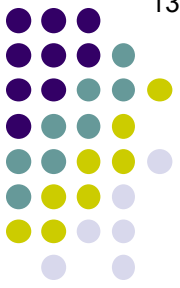
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Define Munson's sign:

Koncerning Keratoconus



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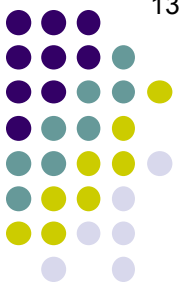
Define Rizzuti's sign:

A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

Define Munson's sign:

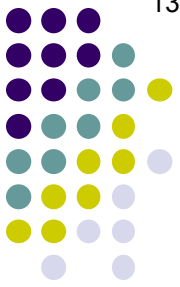
Displacement of the central lower lid by the cone in downgaze

Koncerning Keratoconus



Keratoconus: Munson's sign

Koncerning Keratoconus



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Define Rizzuti's sign:

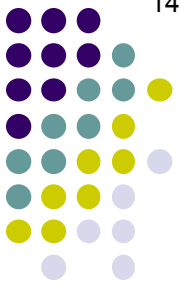
A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

How can you improve visualization of Munson's sign?

Define Munson's sign:

Displacement of the central lower lid by the cone in downgaze

Concerning Keratoconus



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 - **Munson's sign**
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Define Rizzuti's sign:

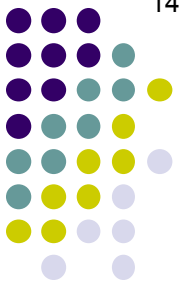
A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

How can you improve visualization of Munson's sign?
By viewing the pt from **above and behind** the exam chair

Define Munson's sign:

Displacement of the central lower lid by the cone in downgaze

Koncerning Keratoconus



- What are the 5 classic signs of keratoconus?
Which is the first to appear?
 - Scissoring of the retinoscopic reflex (earliest sign)
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 - Munson's sign
 - **Fleischer ring**
 - Vogt lines

Define Fleischer ring:

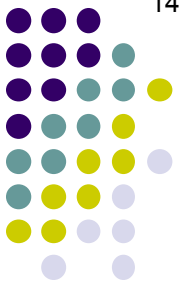
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A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

Define Munson's sign:

Displacement of the central lower lid by the cone in downgaze

Koncerning Keratoconus



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 - Munson's sign
 - **Fleischer ring**
 - Vogt lines

Define Fleischer ring:

Corneal iron line, usually along the lower limit of the cone

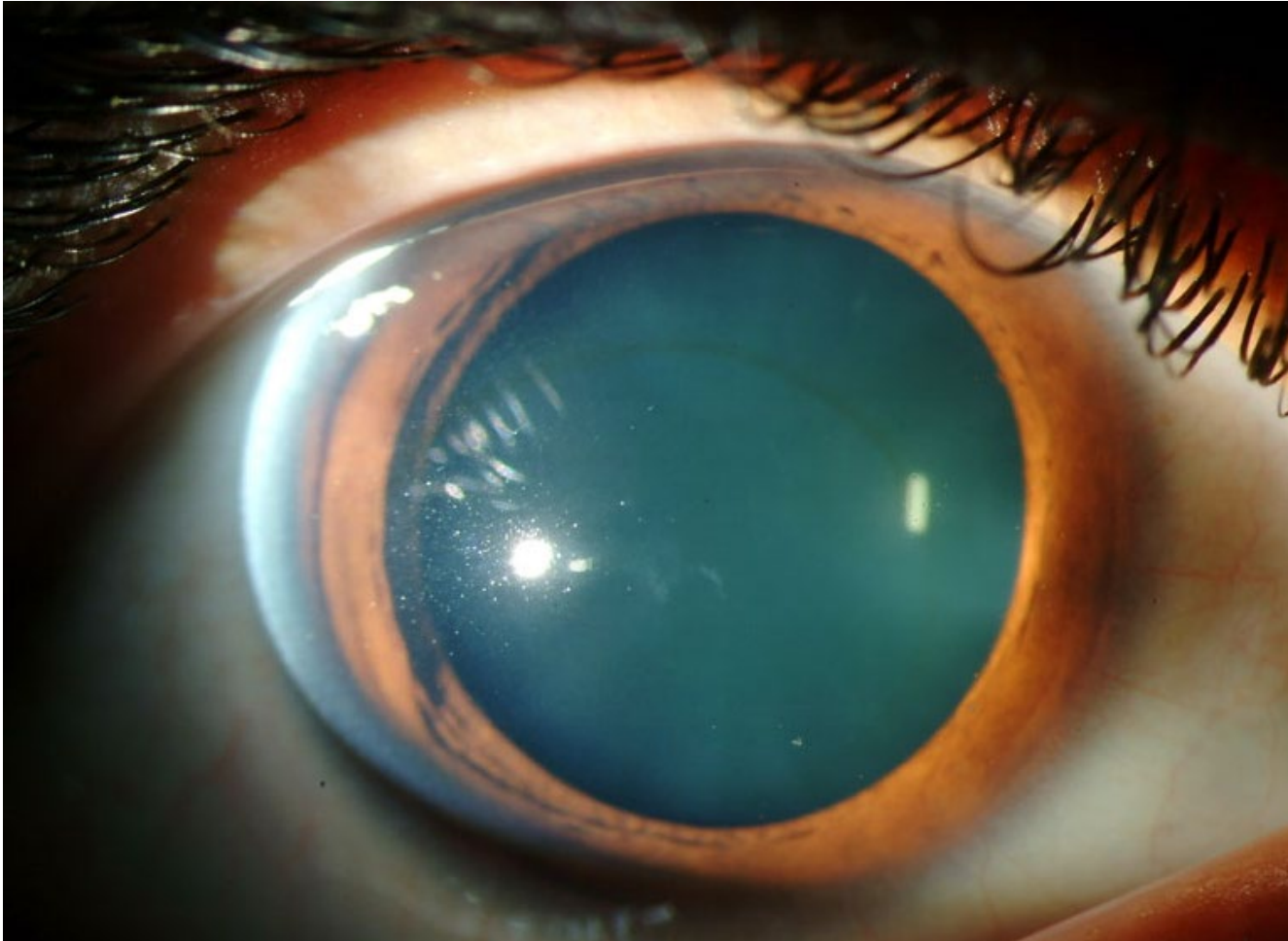
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A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

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Displacement of the central lower lid by the cone in downgaze

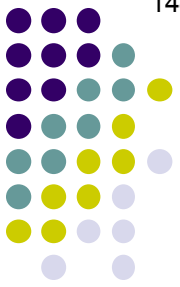
Koncerning Keratoconus



Keratoconus: Fleischer ring



Concerning Keratoconus



- What are the 5 classic signs of keratoconus?
Which is the first to appear?
 - Scissoring of the retinoscopic reflex (earliest sign)
 - Rizzuti's sign

How can you improve visualization of the Fleischer ring at the slit lamp?

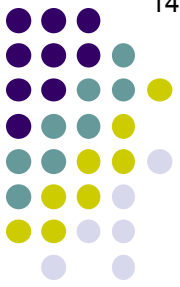
Define Fleischer ring:
Corneal iron line, usually along the lower limit of the cone

- **Fleischer ring**
- Vogt lines

Define Rizzuti's sign:
A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

Define Munson's sign:
Displacement of the central lower lid by the cone in downgaze

Concerning Keratoconus



- What are the 5 classic signs of keratoconus?
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 - Scissoring of the retinoscopic reflex (earliest sign)
 - Rizzuti's sign

*How can you improve visualization of the Fleischer ring at the slit lamp?
By using the **cobalt blue** light*

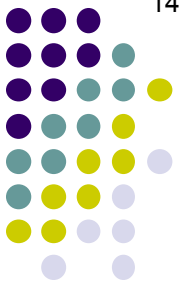
Define Fleischer ring:
Corneal iron line, usually along the lower limit of the cone

- **Fleischer ring**
- Vogt lines

Define Rizzuti's sign:
A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

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Koncerning Keratoconus



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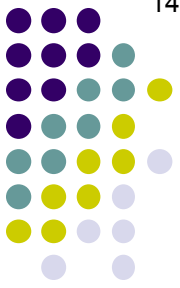
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Define Vogt's lines:

Define Munson's sign:
Displacement of the central lower lid by the cone in downgaze

Koncerning Keratoconus



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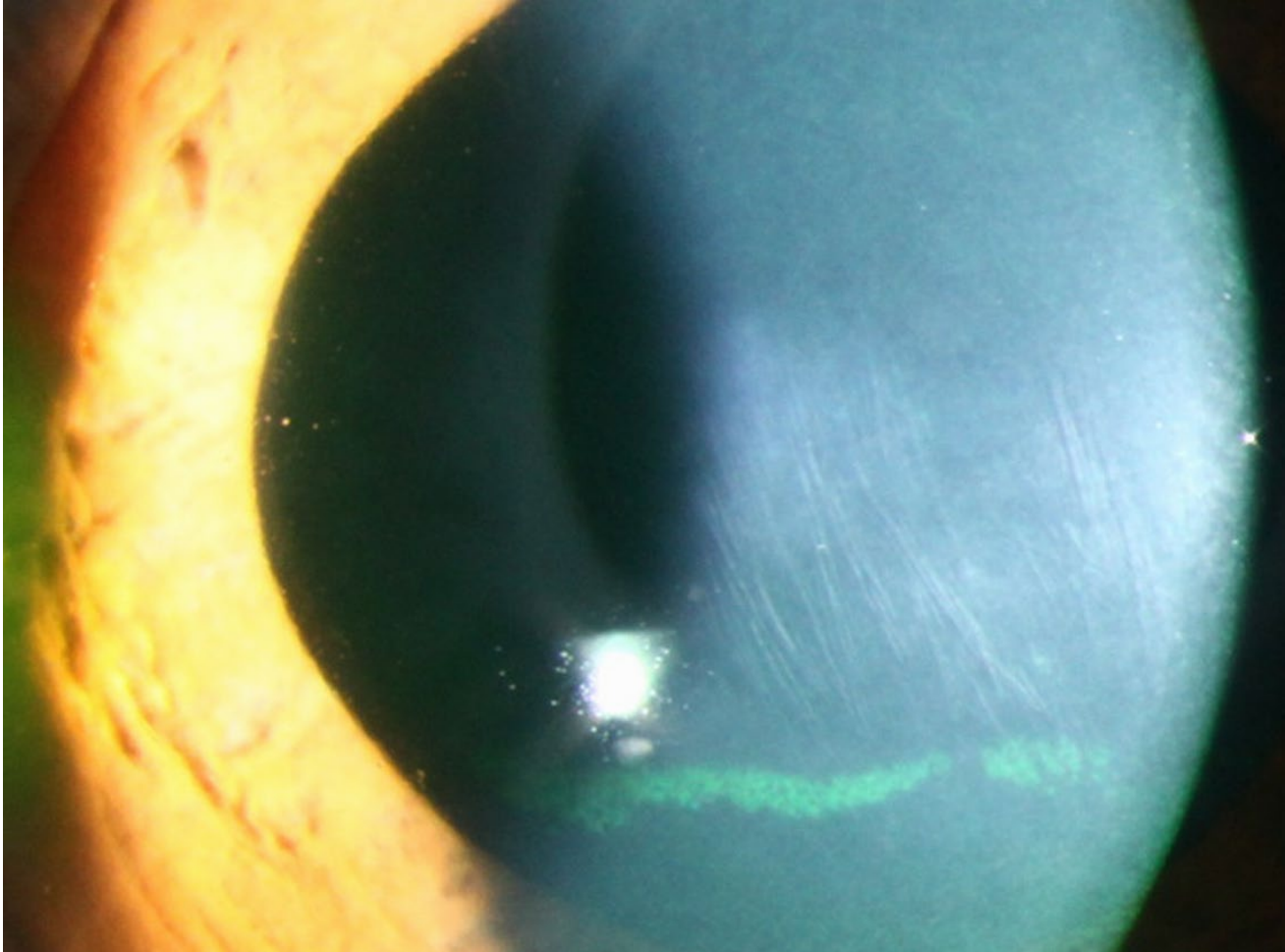
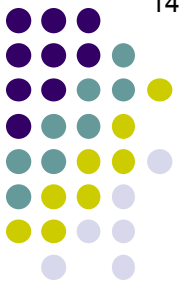
Define Fleischer ring:
Corneal iron line, usually along the lower limit of the cone

Define Rizzuti's sign:
A cone-shaped reflection that appears on the nasal side of the cornea when a light is shone from the temporal side

Define Vogt's lines:
Vertical stress lines in the cornea; disappear with gentle pressure

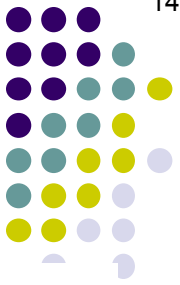
Define Munson's sign:
Displacement of the central lower lid by the cone in downgaze

Koncerning Keratoconus



Keratoconus: Vogt's lines

Koncerning Keratoconus



- What are the 7 classic associations of keratoconus?



While these two are ocular...

(I know, there are only 6 dots. Wait for it.)



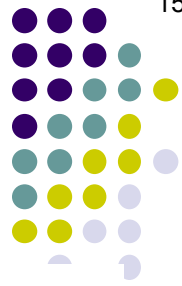
- What are the 7 classic associations of keratoconus?

-
-
-
-
-
-

While these two are ocular...

...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)

Concerning Keratoconus



- What are the 7 classic associations of keratoconus?



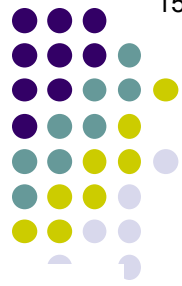
While these two are ocular...

...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)

★ *Mnemonic is...*



Concerning Keratoconus



- What are the 7 classic associations of keratoconus?

● **F**

● **L**

*There are
two 'As'*

● **A**

● **M**

● **E**

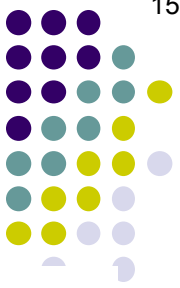
● **D**

While these two are ocular...

...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)

★ Mnemonic is...**FLAMED**★

Concerning Keratoconus



- What are the 7 classic associations of keratoconus?

- **F**loppy eyelid syndrome
- **L**eber's congenital amaurosis
- **A**topic disease (including **A**KC)
- **M**itral valve prolapse
- **E**hlers-Danlos
- **D**own syndrome

There are
two 'As'

While these two are ocular...

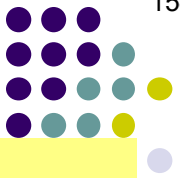
...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)



Mnemonic is...**FLAMED**



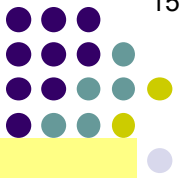
Concerning Keratoconus



In a nutshell, what is floppy eyelid syndrome (FES)?

- What is floppy eyelid syndrome (FES)?
- keratoconus
- **Floppy eyelid syndrome**
- Le
- At
- Mi
- Eh
- Do

Concerning Keratoconus



In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) upper-lid [redacted] and 2) chronic [redacted] of the ocular surface

- What
kerat

- **Floppy eyelid syndrome**

- Le

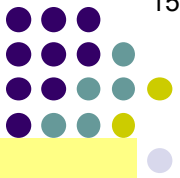
- Atc

- Mi

- Eh

- Do

Koncerning Keratoconus



In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) upper-lid laxity and 2) chronic inflammation of the ocular surface

- What is floppy eyelid syndrome?
- keratoconus
- **Floppy eyelid syndrome**
- Le
- At
- Mi
- Eh
- Do



Concerning Keratoconus

In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) **upper-lid laxity** and 2) chronic inflammation of the ocular surface.

How does one determine if the upper lid is lax?

- What
kerat

- **Floppy eyelid syndrome**

- Le

- Atc

- Mi

- Eh

- Do



Concerning Keratoconus

In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) **upper-lid laxity** and 2) chronic inflammation of the ocular surface.

How does one determine if the upper lid is lax?

By attempting to evert it. In a normal eye, upper-lid eversion is tricky and difficult, whereas in FES it occurs with minimal effort on the part of the examiner.

- What
kerat

- **Floppy eyelid syndrome**

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Concerning Keratoconus

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A condition characterized by 1) upper-lid laxity and 2) chronic **inflammation** of the ocular surface

How does this inflammation manifest on exam?

- What is floppy eyelid syndrome?
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Concerning Keratoconus

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inflammation

How does this inflammation manifest on exam?

The eye will be erythematous, and a pap vs foll reaction will be present on the upper palpebral conj

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Concerning Keratoconus

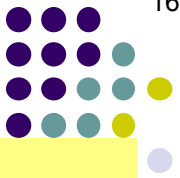
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Concerning Keratoconus

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What do FES pts complain of?

● What
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- **Floppy eyelid syndrome**

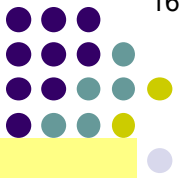
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FBS and mucous discharge that are worse in the

morning vs evening

- **Floppy eyelid syndrome**

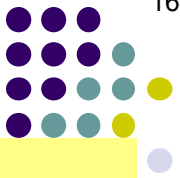
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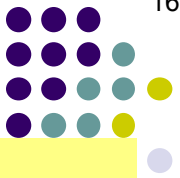
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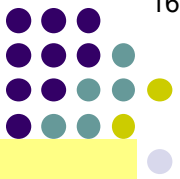
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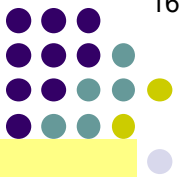
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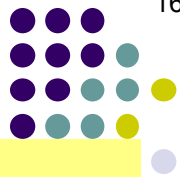
- Atc

Is FES unilateral, or bilateral?

- Mi

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- Do



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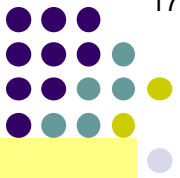
Usually bilateral, but it can be highly asymmetric if the pt manifests a strong predilection for sleeping on one side

- Mi

- Eh

- Do

Concerning Keratoconus



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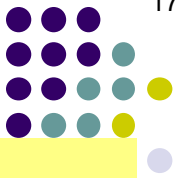
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- *What is the main risk factor for FES?*

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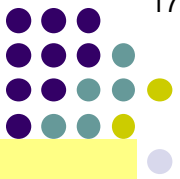
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- Obesity

- Eh

- Do

Koncerning Keratoconus



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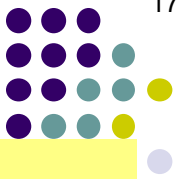
● *How is FES managed initially?*

--?

--?

● Do

Concerning Keratoconus



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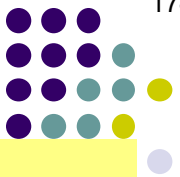
- *What is the main risk factor for FES?*

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--Apply ointment to the involved eye(s) at qHS, and

--?



Concerning Keratoconus

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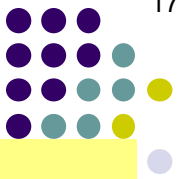
● *At* *What is the main risk factor for FES?*

Obesity

● *Mi* *How is FES managed initially?*

● *Do* --Apply ointment to the involved eye(s) at qHS, and
--Prevent eversion by either the eye(s) or

Concerning Keratoconus



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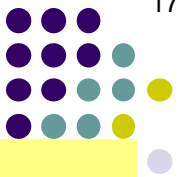
Obesity

- *How is FES managed initially?*

--Apply ointment to the involved eye(s) at qHS, and

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- Do



Koncerning Keratoconus

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- Atc

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Obesity

- Eh

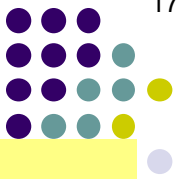
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- Do

If FES fails to respond to the above, what's next?

Concerning Keratoconus



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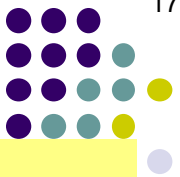
- *How is FES managed initially?*

--Apply ointment to the involved eye(s) at qHS, and
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- Do

If FES fails to respond to the above, what's next?

Surgical tightening of the lax upper lid(s)



Concerning Keratoconus

In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) upper-lid laxity and 2) chronic inflammation of the ocular surface

- What is floppy eyelid syndrome?
- What are the symptoms of floppy eyelid syndrome?

What do FES pts complain of?

FBS and mucous discharge that are worse in the morning

- **Floppy eyelid syndrome**

With what potentially lethal systemic condition is FES strongly associated?

- **At** *between the ocular surface/palpebral conj and the bedding, and this contact traumatizes the involved ocular epithelia.*

- **Mi** *What is the main risk factor for FES?*

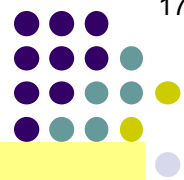
Obesity

- **Eh** *How is FES managed initially?*

--Apply ointment to the involved eye(s) at qHS, and
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Concerning Keratoconus

In a nutshell, what is floppy eyelid syndrome (FES)?

A condition characterized by 1) upper-lid laxity and 2) chronic inflammation of the ocular surface

- What is floppy eyelid syndrome (FES)?
- What is keratoconus?

What do FES pts complain of?

FBS and mucous discharge that are worse in the morning

- **Floppy eyelid syndrome**

With what potentially lethal systemic condition is FES strongly associated?

Obstructive sleep apnea (OSA)

- Atelectasis
- Mitral regurgitation
- Ehlers-Danlos syndrome
- Doxycycline

against a... in contact... and this contact traumatizes the involved ocular epithelia.

What is the main risk factor for FES?

Obesity

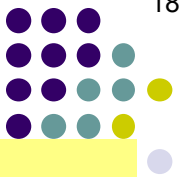
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What do FES pts complain of?

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- **Floppy eyelid syndrome**

With what potentially lethal systemic condition is FES strongly associated?

Obstructive sleep apnea (OSA). In addition to keratoconus, the BCSC states that **all FES pts should be evaluated for OSA.**

- What is the pathophysiology of FES?
- What is the main risk factor for FES?
- How is FES managed initially?
- What is the next step if FES fails to respond to the above?

between the ocular surface/palpebral conj and the bedding, and this contact traumatizes the involved ocular epithelia.

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Koncerning Keratoconus

Management of KCN often follows a pattern:

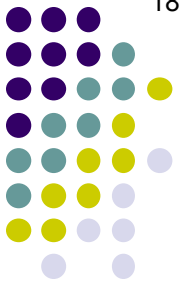
Early KCN: ?



Koncerning Keratoconus

Management of KCN often follows a pattern:

Early KCN: Refractive error corrected via spectacles or soft CLs

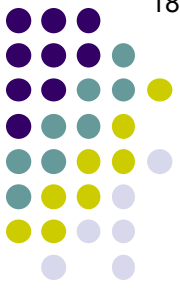


Koncerning Keratoconus

Management of KCN often follows a pattern:

Early KCN: Refractive error corrected via spectacles or soft CLs

KCN progresses → specs/soft CLs no longer adequate...



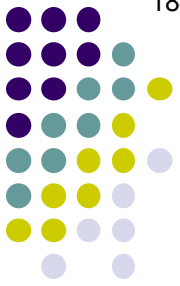
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Moderate KCN: ?



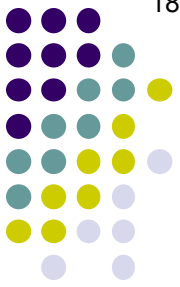
Koncerning Keratoconus

Management of KCN often follows a pattern:

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KCN progresses → specs/soft CLs no longer adequate...

Moderate KCN: **Rigid gas-permeable (RGP) CLs**



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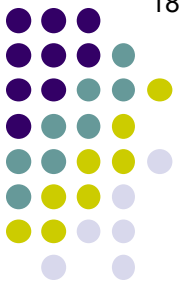
KCN progresses → specs/soft CLs no longer adequate...

Moderate KCN: **Rigid gas-permeable (RGP) CLs**

KCN progresses → cornea too steep to support RGP...

or

Pt becomes CL-intolerant...



Koncerning Keratoconus

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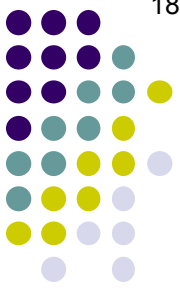
Moderate KCN: Rigid gas-permeable (RGP) CLs

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Advanced/RGP-intolerant KCN: ?



Concerning Keratoconus

Management of KCN often follows a pattern:

Early KCN: **Refractive error corrected via spectacles or soft CLs**

KCN progresses → specs/soft CLs no longer adequate...

Moderate KCN: **Rigid gas-permeable (RGP) CLs**

KCN progresses → cornea too steep to support RGP...

or

Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: **Surgery**



Concerning Keratoconus

Management of KCN often follows a pattern:

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Advanced/RGP-intolerant KCN: Surgery



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Management of KCN often follows a pattern:

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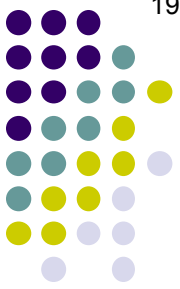
or

Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: Surgery



What does PK stand for?



Concerning Keratoconus

Management of KCN often follows a pattern:

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Moderate KCN: Rigid gas-permeable (RGP) CLs

KCN progresses → cornea too steep to support RGP...

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Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: Surgery



What does PK stand for?

Penetrating keratoplasty

What does DALK stand for?

Concerning Keratoconus

Management of KCN often follows a pattern:

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Advanced/RGP-intolerant KCN: Surgery



What does PK stand for?

Penetrating keratoplasty

What does ICRS stand for?

What does DALK stand for?
Deep anterior lamellar keratoplasty

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KCN progresses → cornea too steep to support RGP...

or

Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: Surgery



What does PK stand for?
Penetrating keratoplasty

What does ICRS stand for?
Intracorneal ring segments

What does DALK stand for?
Deep anterior lamellar keratoplasty

What does CXL stand for?

Concerning Keratoconus

Management of KCN often follows a pattern:

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Advanced/RGP-intolerant KCN: Surgery



What does PK stand for?
Penetrating keratoplasty

What does ICRS stand for?
Intracorneal ring segments

What does DALK stand for?
Deep anterior lamellar keratoplasty

What does CXL stand for?
Collagen crosslinking

Concerning Keratoconus

Management of KCN often follows a pattern:

Early KCN: **Refractive error corrected via spectacles or soft CLs**

KCN progresses → specs/soft CLs no longer adequate...

Moderate KCN: **Rigid gas-permeable (RGP) CLs**

KCN progresses → cornea too steep to support RGP...

or

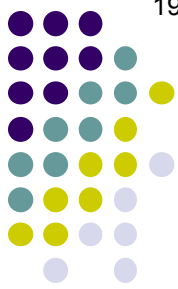
Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: **Surgery**



PK has several things going for it, including:

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--



Concerning Keratoconus

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or

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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

- Proven efficacy
- Familiar skill set

PK has several things going for it, including:

- Proven efficacy with excellent visual results
- The skill-set needed to perform it is familiar to most ophthalmologists



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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

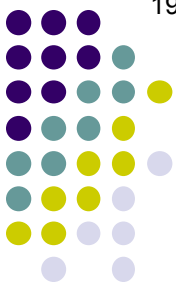
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- Familiar skill set
- Lifetime risk of endothelial rejection
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That said, it has disadvantages as well, including:

- Lifetime risk of endothelial graft rejection
- Post-op management requires frequent visits for an extended time



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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

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What is the lifetime risk of endothelial rejection for someone who undergoes PK as a young adult?

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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

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CXL

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PK has several disadvantages for its indication:

What is the lifetime risk of endothelial rejection for someone who undergoes PK as a young adult?

Estimates run as high as **40%**

ologists

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What is the classic exam finding in endothelial rejection?

someone

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Advanced/RGP-intolerant KCN: Surgery

PK

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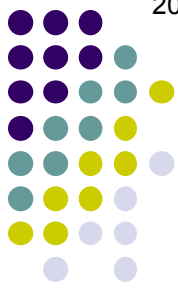
What is the classic exam finding in endothelial rejection?
A line of inflammatory precipitates on the endothelial surface

someone

ologists

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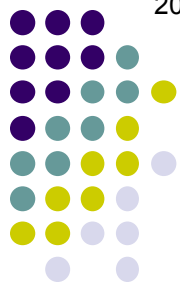
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ologists

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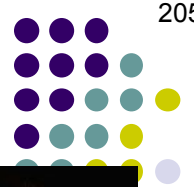
What is the classic exam finding in endothelial rejection?
A line of inflammatory precipitates on the endothelial surface

What is the eponymous name for this finding?
A Khodadoust line

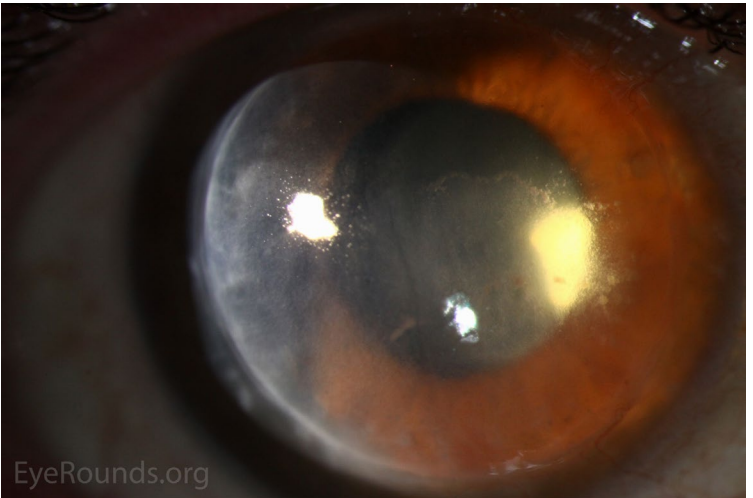
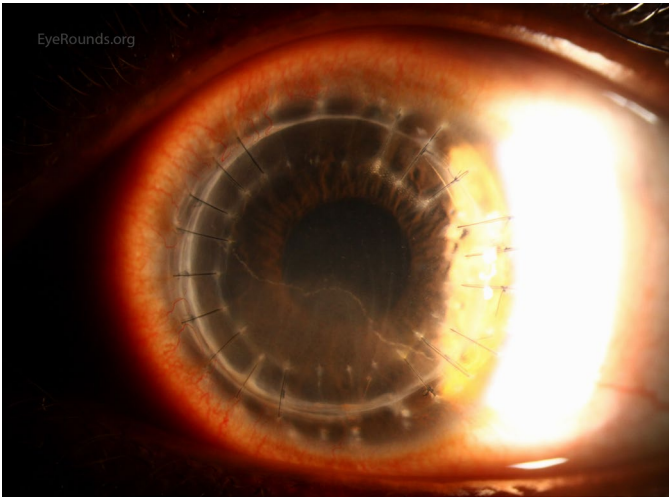
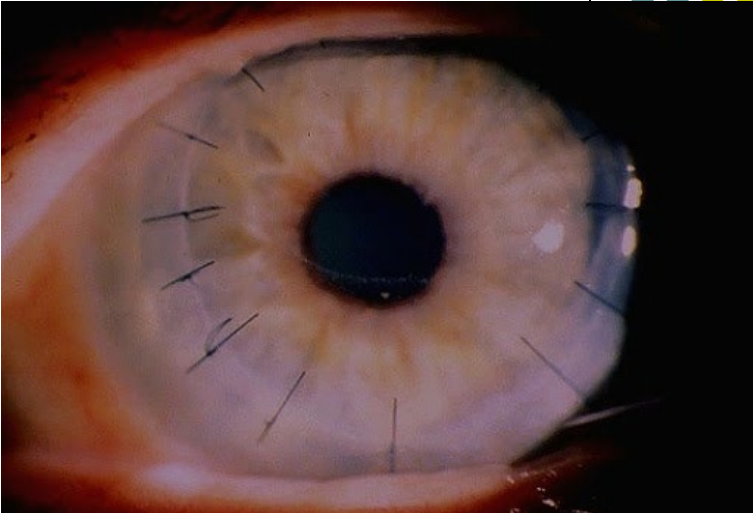
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Koncerning Keratoconus



Khodadoust line

Concerning Keratoconus

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Advanced/RGP-intolerant KCN: Surgery



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Briefly, how is DALK performed?

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Advanced/RGP-intolerant KCN: Surgery



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Briefly, how is DALK performed?

The corneal stroma is trephined to a depth of ~80%, and this portion of stroma is dissected off. The surgeon then *carefully* dissects down to Descemet's membrane, and injects an air bubble between the remaining stroma and Descemet's, thereby separating the two. (Hence the name 'the big bubble technique.')

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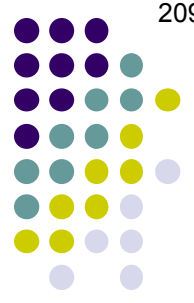
Advanced/RGP-intolerant KCN: Surgery



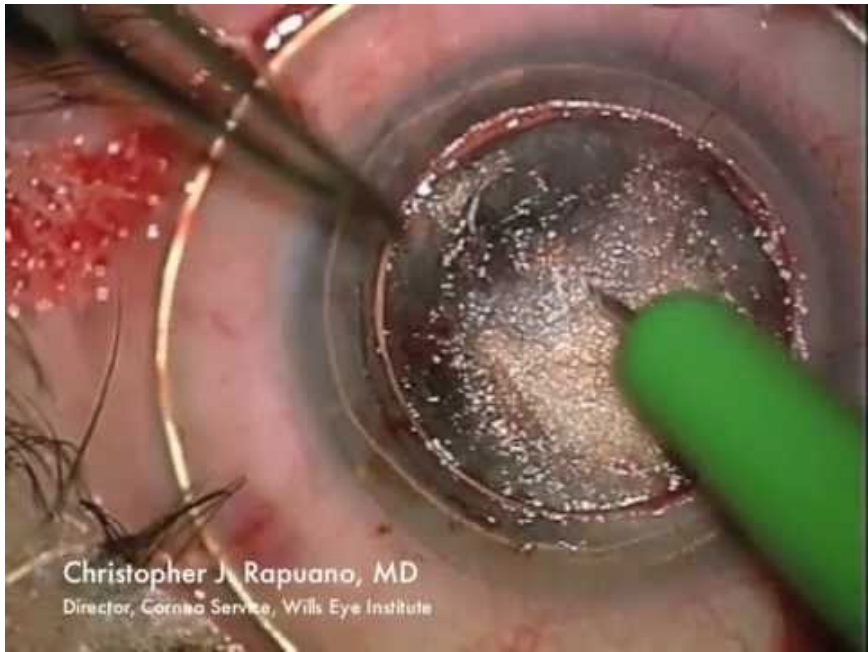
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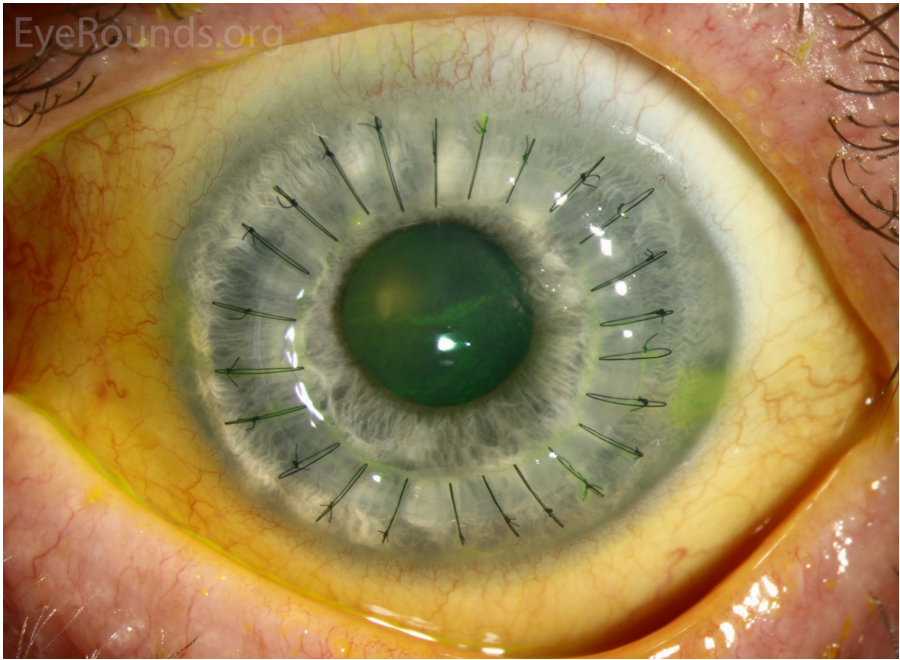
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Concerning Keratoconus



Christopher J. Rapuano, MD
Director, Cornea Service, Wills Eye Institute



EyeRounds.org

DALK

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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

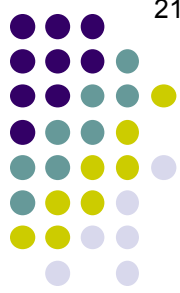
ICRS

CXL

--Proven efficacy
--Familiar skill set
--Lifetime risk of endothelial rejection
--Protracted post-op course

--No risk of endo rejection
--Proven efficacy

DALK has several things going for it, including:
--It obviates the possibility of endothelial rejection
--Proven efficacy with excellent visual results



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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

--Proven efficacy
--Familiar skill set
--Lifetime risk of endothelial rejection
--Protracted post-op course

--No risk of endo rejection
--Proven efficacy
--Technically difficult
--Unfamiliar skill-set

DALK has several things going for it, including:
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That said, it has disadvantages as well, including:
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--The skill-set is unfamiliar to most ophthalmologists



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Advanced/RGP-intolerant KCN: Surgery

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DALK

- No risk of endo rejection
- Proven efficacy
- Technically difficult
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ICRS

CXL

How does the visual outcome of DALK compare to that of PK?

- it obviates the possibility of endothelial rejection
- Proven efficacy with excellent visual results

That said, it has disadvantages as well, including:

- Technically challenging
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Advanced/RGP-intolerant KCN: Surgery

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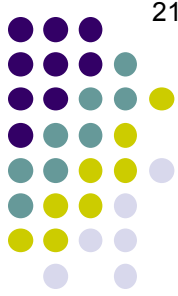
ICRS

CXL

**How does the visual outcome of DALK compare to that of PK?
In skilled/experienced hands, they are equal**

- it obviates the possibility of endothelial rejection
- Proven efficacy with excellent visual results**

- That said, it has disadvantages as well, including:*
- Technically challenging
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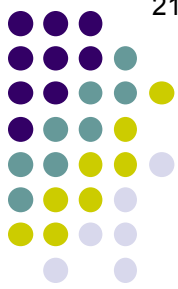
Advanced/RGP-intolerant KCN: Surgery



--Proven efficacy
--Familiar skill set
--Lifetime risk of endothelial rejection
--Protracted post-op course

--No risk of endo rejection

Briefly, how is ICRS surgery performed?



Concerning Keratoconus

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Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

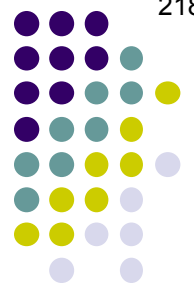
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Briefly, how is ICRS surgery performed?

One or two circular tunnels are created in the mid-peripheral stroma of the cornea, and PMMA semicircular segments are slipped into the tunnels

Koncerning Keratoconus



ICRS

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How does placement of the ICRSs improve VA?

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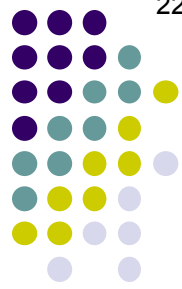
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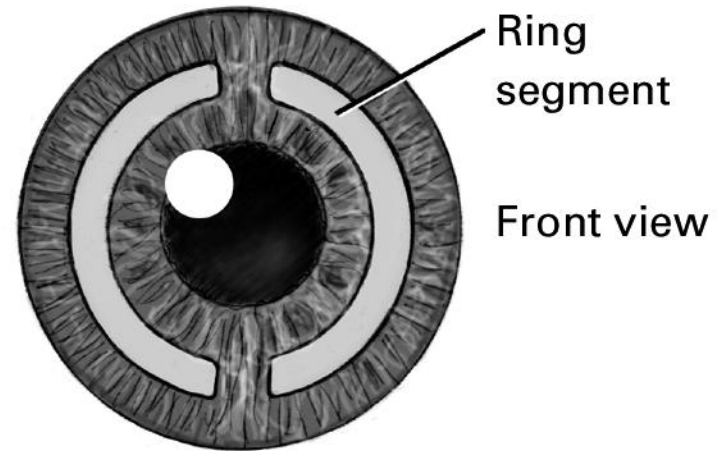
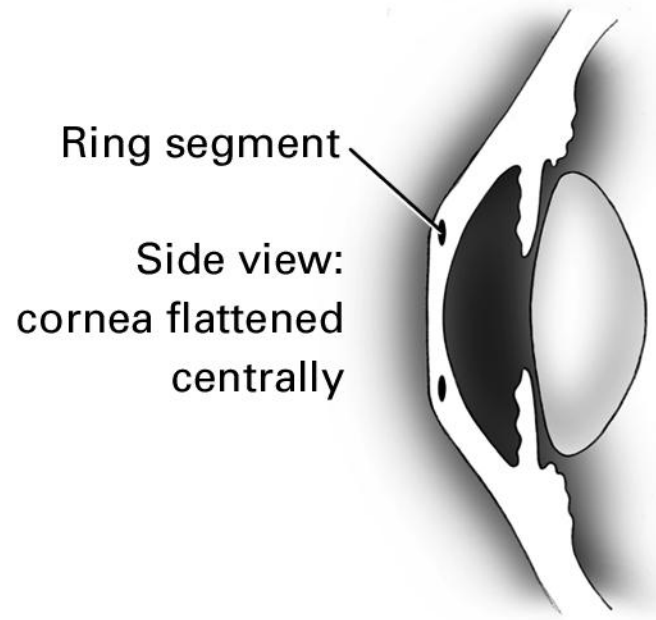
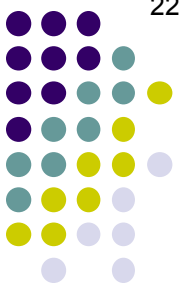
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How does placement of the ICRSs improve VA?

By flattening the central cornea. Also, the number, size and location of the segments can be adjusted to counteract corneal astigmatism (including irregular astigmatism)



Concerning Keratoconus



ICRS

Concerning Keratoconus

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Advanced/RGP-intolerant KCN: **Surgery**

PK

DALK

ICRS

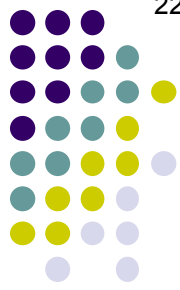
CXL

ICRS have several things going for them, including:

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--Life
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--Pro
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Advanced/RGP-intolerant KCN: Surgery

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CXL

ICRS have several things going for them, including:

- No corneal tissue is removed
- It is reversible (ie, the ICRSs can be removed)

- No tissue removed
- Reversible



Concerning Keratoconus

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Advanced/RGP-intolerant KCN: Surgery

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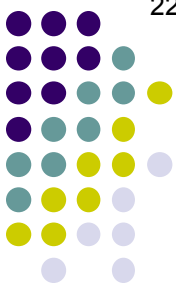
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Advanced/RGP-intolerant KCN: Surgery

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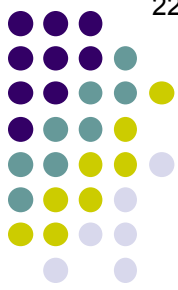
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That said, the procedure has disadvantages as well, including:

- It is unlikely to result in good UCVA by itself

- No tissue removed
- Reversible
- Unlikely to produce excellent UCVA



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In fairness, the goal of ICRS placement is not excellent UCVA; rather, what is it?



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ICRS

CXL

ICRS have several things going for them, including:

- No corneal tissue is removed
- It is reversible (ie, the ICRSs can be removed)

That said, the procedure has disadvantages as well, including:

- It is unlikely to result in good UCVA by itself

- No tissue removed
- Reversible
- Unlikely to produce excellent UCVA

In fairness, the goal of ICRS placement is not excellent UCVA; rather, what is it?

By flattening the cornea and reducing astigmatism (especially irregular astigmatism), the hope is that the pt can once again have his/her refractive error adequately corrected by RGPs, or even spectacles



Concerning Keratoconus

Management of KCN often follows a pattern:

Early KCN: Refractive error corrected via spectacles or soft CLs

KCN progresses → specs/soft CLs no longer adequate...

Moderate KCN: Rigid gas-permeable (RGP) CLs

KCN progresses → cornea too steep to support RGP...

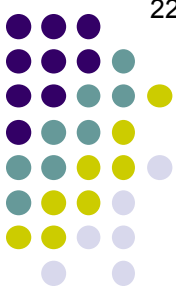
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Pt becomes CL-intolerant...

Advanced/RGP-intolerant KCN: Surgery



What corneal problem, fundamental to KCN, is addressed by CXL?



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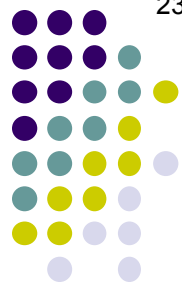


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Briefly, how is CXL performed?

After removal of the corneal epithelium, the stroma is suffused with substance, then subjected to UV vs IR radiation.



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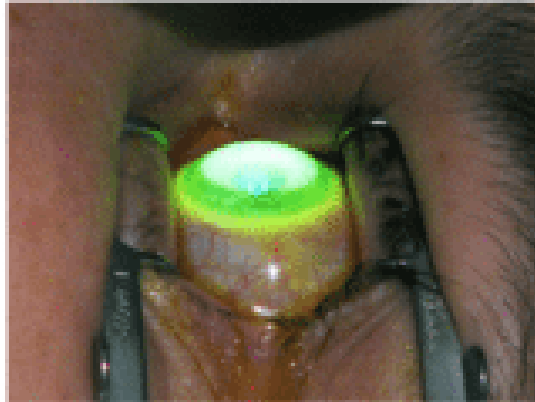
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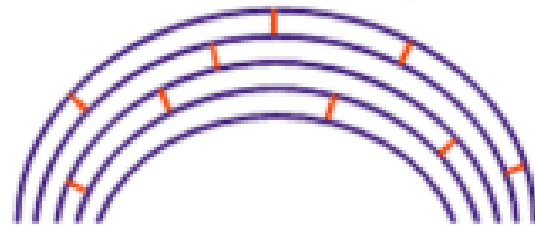
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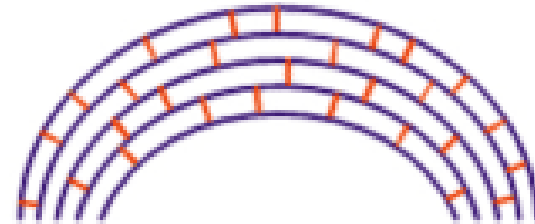
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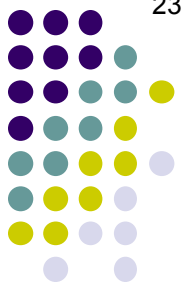
Before CXL (weaker)



After CXL (stronger)



CXL



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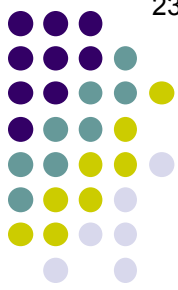


--Proven efficacy
--Familiar skill set
--Lifetime risk of endothelial rejection
--Protracted post-op course

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--Proven e
--Technical
--Unfamilia

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- Technical difficulty
- Unfamiliar

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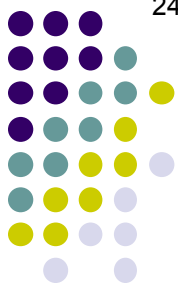
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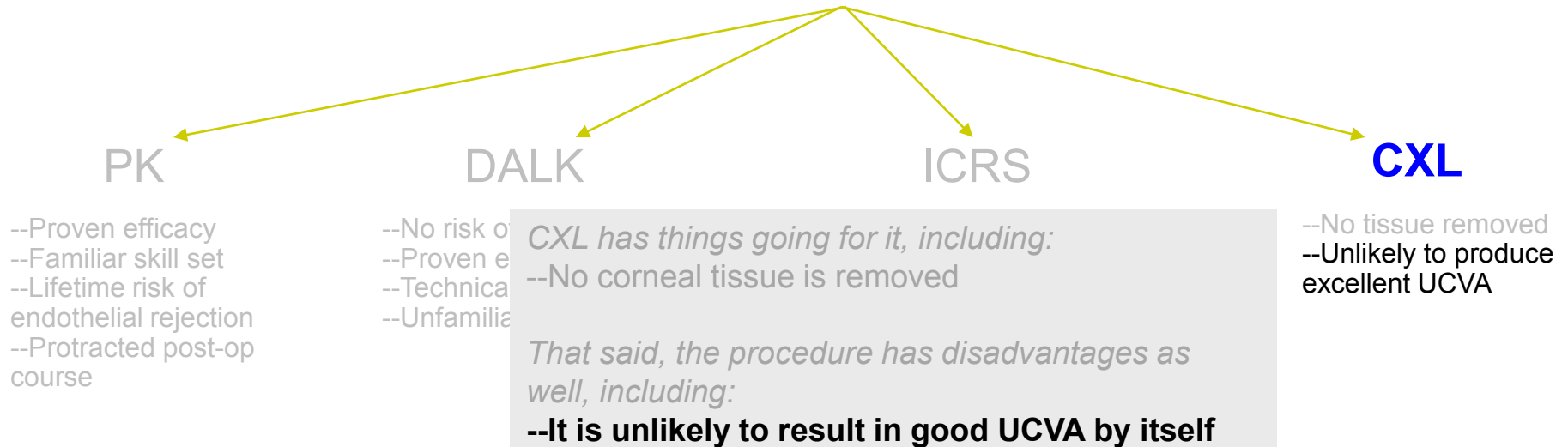
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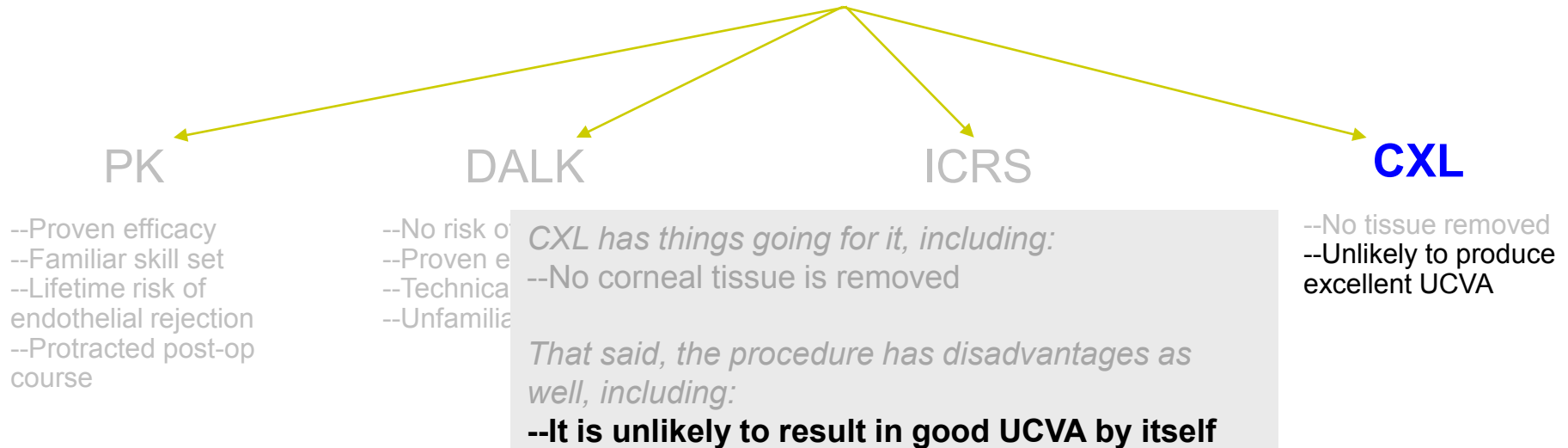
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In fairness to CXL: As with ICRS placement, its goal is not excellent UCVA; rather, what is it? Prevention of disease progression

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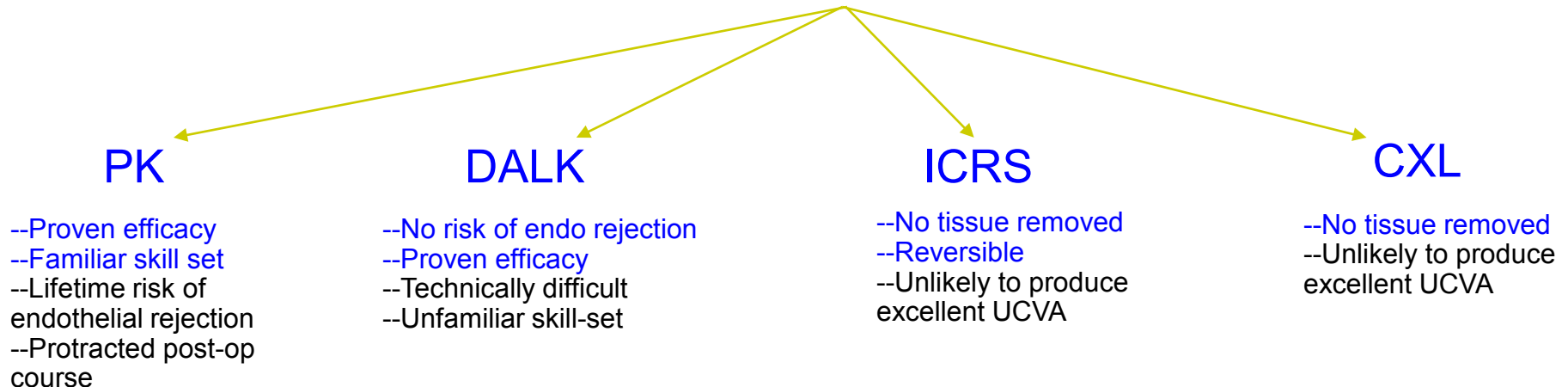
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Summary slide--no questions