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

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

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

*How common is KCN, ie, what is the incidence?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies.

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

1 in 2000
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies

*Let’s back up a step. Briefly, what is an ectasia?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies

Let’s back up a step. Briefly, what is an ectasia?
To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies.  

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

To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:

A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies

Let’s back up a step. Briefly, what is an ectasia?
To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:
A noninflammatory condition characterized by progressive thinning resulting in corneal warpage.

Very broadly (like, in one line), what is the ectatic process in KCN?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies.  

Let’s back up a step. Briefly, what is an ectasia?
To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:
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 para-areas of the cornea leads to bulging of the cornea.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies

Let’s back up a step. Briefly, what is an ectasia?
To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:
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.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies.

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

To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:

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

**OK then: What is a dystrophy?**

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

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

- It is one of the most common corneal dystrophies.

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.

OK then: What is a dystrophy?

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Progressive thinning of the central and/or paracentral cornea leads to cone-like bulging of the cornea.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies.

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

To my frustration, I have been unable to locate an ‘official’ definition in the BCSC (or EyeWiki, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:

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

OK then: What is a dystrophy?

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

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

Progressive thinning of the central and/or paracentral cornea leads to cone-like bulging of the cornea.
Which of the following are true concerning keratoconus (KCN)?

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

- It is one of the most common corneal dystrophies \( F \)
- It has a strong hereditary component \( F \)
- It has a weak hereditary component \( \text{F} \)
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  
- It has a strong hereditary component  

What percent of KCN cases have a positive family history?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  
- It has a strong hereditary component  
- Ectasias

What percent of KCN cases have a positive family history?

5-10%
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present

**Koncerning Keratoconus**

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

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
Keratoconus: Bowman’s is fragmented (3, 4)
Which of the following are true concerning keratoconus (KCN)?

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

- It is one of the most common corneal dystrophies \( F \)
- It has a 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?

- Iron deposition at the base of the cone
- Corneal layer
- Folds/breaks in Descemet’s layer
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies F
- It has a 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?
-- 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)
Koncerning Keratoconus

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

Keratoconus: Corneal thinning (take note of the central portion of the parallelepiped)
Keratoconus: Corneal thinning (at the arrows)
Keratoconus: Descemet's membrane is disrupted and folded back to rejoin the cornea in the wrong orientation (7)
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a 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?

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

Iron deposition at the base of the cone leads to an important clinical sign of KCN. What is the eponymous name of this sign?
Which of the following are true concerning keratoconus (KCN)?

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

- It is one of the most common corneal dystrophies  F
- It has a 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?
- 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?
Fleischer ring

What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  \textbf{F}
- It has a strong hereditary component \textbf{F}
- Fragmentation of Bowman’s is present \textbf{T}

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? Fleischer ring

What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring? Examine the cornea with the \textbf{cobalt-blue light}
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  **F**
- It has a 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?
- 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?
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
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
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
Stocker line
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
There are four corneal iron lines. Name them. With what condition are they associated?

1) **Stocker line**
   - 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
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
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
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

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

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

Ferry line
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...
   - ?
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...
   - Normal and common finding in the elderly
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...
   - Normal and common finding in the elderly

Where is the Hudson-Stähli line located?
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...
   - 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
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...
   - Normal and common finding in the elderly

For more on corneal iron lines, see slide-set K30
Which of the following are true concerning keratoconus (KCN)?

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

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

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

- It is one of the most common corneal dystrophies ✗
- It has a strong hereditary component ✗
- Fragmentation of Bowman’s is present ✓

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

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? Reis-Bückler and Thiel-Behnke
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies • F
- It has a 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?

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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?

Reis-Bückler and Thiel-Behnke

How do you pronounce this?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a 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?
- Iron deposition at the base of the cone
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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?

**Reis-Bückler** and **Thiel-Behnke**

How do you pronounce this? **Reis-Bückler**

How do you pronounce this? **Thiel-Behnke**

How do you pronounce this? **RICE BOO-kler**
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**

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

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?

**Reis-Bückler and Thiel-Behnke**

*In the most recent (2018-19) edition of the BCSC Cornea book, in what ‘major category’ are Reis-Bückler and Thiel-Behnke placed?*
Which of the following are true concerning keratoconus (KCN)?

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

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

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*?

**Reis-Bückler and Thiel-Behnke**

*In the most recent (2018-19) edition of the BCSC Cornea book, in what ‘major category’ are Reis-Bückler and Thiel-Behnke placed? The ‘Epithelial-stromal TGFBI dystrophies’*
Which of the following are true concerning keratoconus (KCN)?

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

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

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*?

**Reis-Bückler and Thiel-Behnke**

What does **TGFBI** stand for in this context?

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

**Epithelial-stromal TGFBI dystrophies**

What is **TGFBI**’s chromosomal location?

**5q31**

The TGFBI was formerly known as what?

**BIGH3** (this factoid is important because you might encounter this name in the older literature)
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  ❌
- It has a strong hereditary component  ❌
- Fragmentation of Bowman’s is present  ✓

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

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?

- Reis-Bückler and Thiel-Behnke

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

- The ‘Epithelial-stromal TGFBI dystrophies’

What does TGFBI stand for in this context?

- ‘Transforming growth factor beta induced’

The TGFBI was formerly known as what?

- BIGH3 (this factoid is important because you might encounter this name in the older literature)
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  F  
- It has a 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?

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

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*?

**Reis-Bückler and Thiel-Behnke**

*In the most recent (2018-19) edition of the BCSC *Cornea* book, in what ‘major category’ are Reis-Bückler and Thiel-Behnke places?*

The ‘Epithelial-stromal’ dystrophies

**What does TGFBI stand for in this context?**

‘Transforming growth factor beta induced’

**What is TGFBI’s chromosomal location?**

5q31

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

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
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- Iron deposition at the base of the cone
- Corneal thinning
- Folds/breaks in Descemet’s

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?

Reis-Bückler and Thiel-Behnke

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

The ‘Epithelial-stromal TGFBI dystrophies’

What does TGFBI stand for in this context?

‘Transforming growth factor beta induced’

What is TGFBI’s chromosomal location?

5q31
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: \( F \)
- It has a 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?

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

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*?

**Reis-Bückler and Thiel-Behnke**

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

The ‘Epithelial-stromal TGFBI dystrophies’

What does TGFBI stand for in this context?

‘Transforming growth factor beta induced’

What is TGFBI’s chromosomal location?

5q31

The TGFBI gene was formerly known as what?
Which of the following are true concerning keratoconus (KCN)?

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

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

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*?

**Reis-Bückler and Thiel-Behnke**

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

The ‘Epithelial-stromal TGFBI dystrophies’

What does TGFBI stand for in this context?

‘Transforming growth factor beta induced’

What is TGFBI’s chromosomal location?

5q31

The TGFBI gene was formerly known as what?

BIGH3 (this factoid is important because you might encounter this name in the older literature)
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies F
- It has a 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?
- Iron deposition at the base of the cone
- Corneal thinning
- Folds/breaks in Descemet’s

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?

Reis-Bückler and Thiel-Behnke

In the most recent (2018-19) edition of the BCSC Cornea book, in what ‘major category’ are Reis-Bückler and Thiel-Behnke placed? The Corneal-dystrophy section underwent a major revision for this version of the Cornea book. In what category were Reis-Bückler and Thiel-Behnke place in previous editions?

The ‘Epithelial-stromal TGFBI dystrophies’ formerly known as the...
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies \( \text{F} \)
- It has a strong hereditary component \( \text{F} \)
- Fragmentation of Bowman’s is present \( \text{T} \)

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

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?

Reis-Bückler and Thiel-Behnke

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?

Reis-Bückler and Thiel-Behnke were placed in the ‘Nongranulomatous Nonsclerotic’ category in previous editions. In the most recent (2018-19) edition of the BCSC Cornea book, they are placed in the ‘Epithelial-stromal TGFB1 dystrophies’ category.

The ‘Epithelial-stromal TGFB1 dystrophies’ formerly known as the...
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK

*(PK = Penetrating keratoplasty, ie, a corneal transplant)*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies \( \text{T} \)
- It has a strong hereditary component \( \text{F} \)
- Fragmentation of Bowman’s is present \( \text{T} \)
- **Acute hydrops** is an indication for urgent PK \( \text{F} \)

*What is acute hydrops?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- **Acute hydrops** is an indication for urgent PK **F**

*What is acute hydrops?*

The sudden development of severe corneal edema secondary to a break in Descemet’s layer.
Concerning Keratoconus

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

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- **Acute hydrops** is an indication for urgent PK: **F**

*What is acute hydrops?*

The sudden development of severe corneal edema secondary to a break in Descemet’s
KCN: Acute hydrops
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  **F**
- It has a strong hereditary component  **F**
- Fragmentation of Bowman's is present  **T**
- Acute hydrops is not an indication for urgent PK  **F**

*What is acute hydrops?*
The sudden development of severe corneal edema 2ndry to a break in Descemet's

*Why is it not an indication for urgent PK?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  F
- It has a strong hereditary component  F
- Fragmentation of Bowman’s is present  T
- Acute hydrops is an indication for urgent PK  F

**What is acute hydrops?**
The sudden development of severe corneal edema

**Why is it not an indication for urgent PK?**
Because it will resolve on its own in a few months
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies \textcolor{red}{F}
- It has a strong hereditary component \textcolor{red}{F}
- Fragmentation of Bowman’s is present \textcolor{green}{T}
- **Acute hydrops** is an indication for urgent PK \textcolor{red}{F}

*What is acute hydrops?*

The sudden development of severe corneal edema secondary to a break in Descemet’s membrane.

*Why is it not an indication for urgent PK?*

Because it will resolve on its own in a few months.

*When it resolves, does it do so with, without sequelae?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- **Acute hydrops** is an indication for urgent PK: **F**

**What is acute hydrops?**
The sudden development of severe corneal edema 2ndry to a break in Descemet’s membrane.

**Why is it not an indication for urgent PK?**
Because it will resolve on its own in a few months.

**When it resolves, does it do so with, without sequelae?**
With; apical scarring usually occurs.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- **Acute hydrops** is an indication for urgent PK **F**

What is acute hydrops?
The sudden development of severe corneal edema secondary to a break in Descemet’s

Why is it not an indication for urgent PK?
Because it will resolve on its own in a few months

When it resolves, does it do so with, without sequelae?
With; apical scarring usually occurs
KCN: Apical scarring after acute hydrops
Koncerning Keratoconus

3 days after break

Scarring, same eye, 2+ months later

KCN: Acute hydrops
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- **Acute hydrops** is an indication for urgent PK **F**

What is acute hydrops?
The sudden development of severe corneal edema 2ndry to a break in Descemet’s

Why is it not an indication for urgent PK?
Because it will resolve on its own in a few months

When it resolves, does it correct the keratoconus? 
It can. 'Apical scarring'—that sounds like a bad thing. Is it?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- **Acute hydrops** is an indication for urgent PK **F**

What is acute hydrops?
The sudden development of severe corneal edema
2ndry to a break in Descemet’s

Why is it not an indication for urgent PK?
Because it will resolve on its own in a few months

When it resolves, does it... 

'Apical scarring'—that sounds like a bad thing. Is it?
Not necessarily—in some cases, the scarring flattens the cone, thereby reducing myopia and/or astigmatism.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: F
- It has a strong hereditary component: F
- Fragmentation of Bowman’s is present: T
- Acute hydrops is an indication for urgent PK: F
- The incidence is higher in South Asia and the Middle East: T
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
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- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood:
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydrops is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during adolescence **F**

*During what period of life does KCN progress the fastest?*
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  F
- It has a strong hereditary component  F
- Fragmentation of Bowman’s is present  T
- Acute hydrops is an indication for urgent PK  F
- The incidence is higher in South Asia and the Middle East  T
- Onset typically occurs during  F

**During what period of life does KCN progress the fastest?**
Early on—teens to 20s
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  F
- It has a strong hereditary component  F
- Fragmentation of Bowman’s is present  T
- Acute hydrops is an indication for urgent PK  F
- The incidence is higher in South Asia and the Middle East  T
- Onset typically occurs during adolescence  F

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

By what age does progression typically cease?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  \( F \)
- It has a strong hereditary component  \( F \)
- Fragmentation of Bowman’s is present  \( T \)
- Acute hydrops is an indication for urgent PK  \( F \)
- The incidence is higher in South Asia and the Middle East  \( T \)
- Onset typically occurs during adolescence  \( \text{not} \)
  childhood  \( F \)

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

By what age does progression typically cease?
40
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  F
- It has a strong hereditary component  F
- Fragmentation of Bowman’s is present  T
- Acute hydrops is an indication for urgent PK  F
- The incidence is higher in South Asia and the Middle East  T
- Onset typically occurs during adolescence  F

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

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydrops is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during **adolescence** **F**

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

- It is one of the most common corneal dystrophies \textbf{F}
- It has a \textbf{strong} hereditary component \textbf{F}
- Fragmentation of Bowman’s is present \textbf{T}
- Acute hydrops is an indication for urgent PK \textbf{F}
- The incidence is higher in South Asia and the Middle East \textbf{T}
- Onset typically occurs during \textbf{adolescence} \textbf{F}
- Spontaneous rupture is fairly common
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **false**
- It has a strong hereditary component: **false**
- Fragmentation of Bowman’s is present: **true**
- Acute hydrops is an indication for urgent PK: **false**
- The incidence is higher in South Asia and the Middle East: **true**
- Onset typically occurs during adolescence: **false**
- Spontaneous rupture is fairly common: **false**

**Koncerning Keratoconus**
Which of the following are true concerning keratoconus (KCN)?

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

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

- It is one of the most common corneal dystrophies \( \mathbf{F} \)
- It has a strong hereditary component \( \mathbf{F} \)
- Fragmentation of Bowman’s is present \( \mathbf{T} \)
- Acute hydrops is an indication for urgent PK \( \mathbf{F} \)
- The incidence is higher in South Asia and the Middle East \( \mathbf{T} \)
- Onset typically occurs during adolescence \( \mathbf{F} \)
- Spontaneous rupture is fairly common \( \mathbf{F} \)
- KCN is strongly associated with Marfan syndrome \( \mathbf{F} \)

What is the most common pathologic corneal finding in Marfan’s?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

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

An abnormally steep vs flat cornea
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydros is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during childhood **F**
- Spontaneous rupture is fairly common **F**
- KCN is strongly associated with **F**

What is the most common pathologic corneal finding in Marfan’s?
An abnormally flat cornea
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

What is the most common pathologic corneal finding in Marfan’s?
An abnormally flat cornea

How flat (in diopters) are we talking about here?
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydrops is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during childhood **F**
- Spontaneous rupture is fairly common **F**
- KCN is strongly associated with Marfan syndrome **F**

**Marfan syndrome**

What is the most common pathologic corneal finding in Marfan’s?
An abnormally flat cornea

How flat (in diopters) are we talking about here?
Values in the 35D range are common
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

What is the dioptric power of a typical ‘normal’ cornea?

Values in the 35D range are common.

What is the dioptric power of a typical ‘advanced’ KCN cornea (at the cone)?

Values greater than 50D are the rule, and values greater than 60D are not uncommon.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

What is the dioptric power of a typical ‘normal’ cornea? 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.

What is the most common pathologic corneal ectasia? Keratoconus

What form of (corneal) ectasia is most common pathologic corneal ectasia in Marfan’s? Keratoconus

What are we talking about here? Values in the 35D range are extremely rare, and usually flat cornea are extremely rare.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

What is the dioptric power of a typical ‘normal’ cornea?
Around 43D

What is the dioptric power of an advanced KCN cornea (at the cone)?
Values in the 35D range are common.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydrops is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during childhood **F**
- Spontaneous rupture is fairly common **F**
- KCN is strongly associated with Marfan syndrome **F**

What is the dioptric power of a typical ‘normal’ cornea?
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

What is the dioptric power of a most common pathologic corneal ectasia?
(usually flat cornea)

Marfan syndrome

What (diopeters) are we talking about here?
Values in the 35D range are common
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies \textbf{F}
- It has a strong hereditary component \textbf{F}
- Fragmentation of Bowman’s is present \textbf{T}
- Acute hydrops is an indication for urgent PK \textbf{F}
- The incidence is higher in South Asia and the Middle East \textbf{T}
- Onset typically occurs during \textit{adolescence} \textbf{F}
- Spontaneous rupture is fairly common \textbf{F}
- KCN is strongly associated with Marfan syndrome \textbf{F}

\begin{itemize}
  \item \underline{With what conditions is KCN associated?}
  \begin{itemize}
    \item \textbf{Ocular only:}
    \begin{itemize}
      \item --
      \item --
    \end{itemize}
    \item \textbf{Systemic:}
    \begin{itemize}
      \item --
      \item --
      \item --
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Which of the following are true concerning keratoconus (KCN)?

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- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

**With what conditions is KCN associated?**

**Ocular only:**
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

**Systemic:**
- Down syndrome
- Ehler’s-Danlos syndrome
- Osteogenesis imperfecta

List not exhaustive, obviously
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: F
- It has a strong hereditary component: F
- Fragmentation of Bowman’s is present: T
- Acute hydrops is an indication for urgent PK: F
- The incidence is higher in South Asia and the Middle East: T
- Onset typically occurs during adolescence: F
- Spontaneous rupture is fairly common: F
- KCN is strongly associated with Marfan syndrome: F

What is the common thread among these?

With what conditions is KCN associated?

Ocular only:
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

Systemic:
- Down syndrome
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- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

What is the common thread among these?

Eye rubbing

With what conditions is KCN associated?

Ocular only:
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

Systemic:
- Down syndrome
- Ehler’s-Danlos syndrome
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- Acute hydrops is an indication for urgent PK: F
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- Onset typically occurs during adolescence: T
- Spontaneous rupture is fairly common: F
- KCN is strongly associated with Marfan syndrome: F

With what conditions is KCN associated?

**Ocular only:**
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

**Systemic:**
- Down syndrome
- Ehler’s-Danlos syndrome
- Osteogenesis imperfecta

Why do VKC and Down pts rub their eyes?

What is the common thread among these? **Eye rubbing**
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
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With what conditions is KCN associated?

**Ocular only:**
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

**Systemic:**
- Down syndrome
- Ehler’s-Danlos syndrome
- Osteogenesis imperfecta

Why do VKC and Down pts rub their eyes? Because they itch

What is the common thread among these? Eye rubbing
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies \( \text{F} \)
- It has a strong hereditary component \( \text{F} \)
- Fragmentation of Bowman’s is present \( \text{T} \)
- Acute hydrops is an indication for urgent PK \( \text{F} \)
- The incidence is higher in South Asia and the Middle East \( \text{T} \)
- Onset typically occurs during adolescence \( \text{F} \)

Why do Leber’s pts rub their eyes—do they itch as well?

What is the common thread among these?

Eye rubbing

- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis
- Systemic:
  - Down syndrome
  - Ehler’s-Danlos syndrome
  - Osteogenesis imperfecta

Why do VKC and Down pts rub their eyes? Because they itch
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  **F**
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- Onset typically occurs during adolescence  **F**

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 **phosphenes**.

**Ocular only:**

--- Leber’s congenital amaurosis
--- Vernal keratoconjunctivitis
--- Down syndrome
--- Ehler’s-Danlos syndrome
--- Osteogenesis imperfecta

**Systemic:**

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--- Osteogenesis imperfecta

What is the common thread among these?
**Eye rubbing**

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

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- Onset typically occurs during childhood: **F**

---

**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 **phosphenes**.

---

**What is the common thread among these?**

**Eye rubbing**

- **Ocular only:**
  - **Leber’s congenital amaurosis**
  - Vernal keratoconjunctivitis
- **Systemic:**
  - **Down syndrome**
  - Ehler’s-Danlos syndrome
  - Osteogenesis imperfecta

---

**Why do VKC and Down pts rub their eyes?**

Because they itch
Which of the following are true concerning keratoconus (KCN)?

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- Spontaneous rupture is fairly common: F
- KCN is strongly associated with Marfan syndrome: F

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What is the common thread among these?
Which of the following are true concerning keratoconus (KCN)?

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- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during adolescence: **T**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

**With what conditions is KCN associated?**
- **Ocular only:**
  - Leber’s congenital amaurosis
  - Vernal keratoconjunctivitis
- **Systemic:**
  - Down syndrome
  - Ehler’s-Danlos syndrome
  - Osteogenesis imperfecta

*What is the common thread among these?* Abnormal connective tissue
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: **F**
- It has a strong hereditary component: **F**
- Fragmentation of Bowman’s is present: **T**
- Acute hydrops is an indication for urgent PK: **F**
- The incidence is higher in South Asia and the Middle East: **T**
- Onset typically occurs during childhood: **F**
- Spontaneous rupture is fairly common: **F**
- KCN is strongly associated with Marfan syndrome: **F**

With what conditions is KCN associated?

- Ocular only:
  - Leber’s congenital amaurosis
  - Vernal keratoconjunctivitis
- Systemic:
  - Down syndrome
  - Ehler’s-Danlos syndrome
  - Osteogenesis imperfecta

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

- It is one of the most common corneal dystrophies  \( \text{F} \)
- It has a strong hereditary component  \( \text{F} \)
- Fragmentation of Bowman’s is present  \( \text{T} \)
- Acute hydrops is an indication for urgent PK  \( \text{F} \)
- The incidence is higher in South Asia and the Middle East  \( \text{T} \)
- Onset typically occurs during childhood  \( \text{F} \)
- Spontaneous rupture is fairly common  \( \text{F} \)
- KCN is strongly associated with Marfan syndrome  \( \text{F} \)

With what conditions is KCN associated?

Ocular only:
- Leber’s congenital amaurosis
- Vernal keratoconjunctivitis

Systemic:
- Down syndrome
- Ehler’s-Danlos syndrome
- Osteogenesis imperfecta

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
- Atopy
- Rigid CL wear
- Inflammation
- Oxidative stress

What is the common thread among these?  

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

- It is one of the most common corneal dystrophies **F**
- It has a strong hereditary component **F**
- Fragmentation of Bowman’s is present **T**
- Acute hydrops is an indication for urgent PK **F**
- The incidence is higher in South Asia and the Middle East **T**
- Onset typically occurs during adolescence **F**
- Spontaneous rupture is fairly common **F**
- KCN is strongly associated with Marfan syndrome **F**
- Females are more likely to be affected than males
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: F
- It has a strong hereditary component: F
- Fragmentation of Bowman’s is present: T
- Acute hydrops is an indication for urgent PK: F
- The incidence is higher in South Asia and the Middle East: T
- Onset typically occurs during adolescence: F
- Spontaneous rupture is extremely rare: F
- KCN is strongly associated with Marfan syndrome: F
- Females are more likely to be affected than males: T
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies: F
- It has a strong hereditary component: F
- Fragmentation of Bowman’s is present: T
- Acute hydrops is an indication for urgent PK: F
- The incidence is higher in South Asia and the Middle East: T
- Onset typically occurs during adolescence: F
- Spontaneous rupture is extremely rare: F
- KCN is strongly associated with Marfan syndrome: F
- Females are more likely to be affected than males: T
- Corneal topography reveals superior steepening.
Which of the following are true concerning keratoconus (KCN)?

- It is one of the most common corneal dystrophies  **F**
- It has a weak hereditary component  **F**
- Fragmentation of Bowman’s is present  **T**
- Acute hydrops is an indication for urgent PK  **F**
- The incidence is higher in South Asia and the Middle East  **T**
- Onset typically occurs during adolescence  **F**
- Spontaneous rupture is extremely rare  **F**
- KCN is strongly associated with Marfan syndrome  **F**
- Females are more likely to be affected than males  **T**
- Corneal topography reveals superior steepening  **F**
Topography in keratoconus reveals

three words
Topography in keratoconus reveals **inferior corneal steepening**
Topography in keratoconus reveals **inferior corneal steepening**
What corneal shape is represented?
Koncerning Keratoconus

With-the-rule astigmatism

KCN

What corneal shape is represented?
With-the-rule astigmatism

KCN
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?

- Scissoring of the retinoscopic reflex—earliest sign
- Rizzuti's sign
- Munson's sign
- Fleischer ring
- Vogt lines
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
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

*Define Rizzuti’s sign:*
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

**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.
Keratoconus: Rizzuti’s sign
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

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:
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

**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
Keratoconus: Munson’s sign
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

**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.
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

**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.

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

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

**Define Munson’s sign:** Displacement of the central lower lid by the cone in downgaze.
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

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

**Define Munson’s sign:**
Displacement of the central lower lid by the cone in downgaze.
Keratoconus: Fleischer ring
What are the 5 classic signs of keratoconus? Which is the first to appear?

- Scissoring of the retinoscopic reflex (earliest sign)
- Rizzuti’s sign
- 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 Fleischer ring:**
Corneal iron line, usually along the lower limit of the cone

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

**Define Scissoring of the retinoscopic reflex:**
Ears of the reflex are parallel and scissor-like.
What are the 5 classic signs of keratoconus? Which is the first to appear?
- Scissoring of the retinoscopic reflex (earliest sign)
- Rizzuti’s sign
- Fleischer ring
- Vogt lines

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

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
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

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

**Define Munson’s sign:**
Displacement of the central lower lid by the cone in downgaze.
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

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

**Define Munson’s sign:**
Displacement of the central lower lid by the cone in downgaze.

**Define Vogt’s lines:**
Vertical stress lines in the cornea; disappear with gentle pressure.
Keratoconus: Vogt’s lines
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?

- Loopy eyelid syndrome
- Leber's congenital amaurosis
- A topic disease
- Marfan's
- Ehlers-Danlos
- Down syndrome

While these two are ocular...

...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)
What are the 7 classic associations of keratoconus?

- Flexible eyelid syndrome
- Leber's congenital amaurosis
- Atopic disease
- Marfan's
- Ehlers-Danlos
- Down syndrome

Mnemonic is...

While these two are ocular...

...these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)
What are the 7 classic associations of keratoconus?

- Fluffy eyelid syndrome
- Leber's congenital amaurosis
- Akrone disease
- Marfan's
- Ehlers-Danlos
- Down syndrome

Mnemonic is…FLAMED

Koncerning Keratoconus

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)
What are the 7 classic associations of keratoconus?

- Floppy eyelid syndrome
- Leber’s congenital amaurosis
- Atopic disease (including AKC)
- Mitral valve prolapse
- Ehlers-Danlos
- Down syndrome

Mnemonic is…FLAMED

While these two are ocular…

…these five are systemic (and thus constitute the systemic associations alluded to at the beginning of the slide-set)
Management of KCN often follows a pattern:

*Early KCN: ?*
Management of KCN often follows a pattern:

*Early KCN:* Refractive error corrected via spectacles or soft CLs
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…*
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:* ?
Koncerning Keratoconus

Management of KCN often follows a pattern:

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

   *KCN progresses* $\rightarrow$ *specs/soft CLs no longer adequate…*

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

Management of KCN often follows a pattern:

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

\[ \text{KCN progresses} \rightarrow \text{specs/soft CLs no longer adequate} \ldots \]

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

\[ \text{KCN progresses} \rightarrow \text{cornea too steep to support RGP} \ldots \]

or

\[ \text{Pt becomes CL-intolerant} \ldots \]
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: ?
Management of KCN often follows a pattern:

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

\[ KCN \text{ progresses} \rightarrow \text{specs/soft CLs no longer adequate} \ldots \]

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

\[ KCN \text{ progresses} \rightarrow \text{cornea too steep to support RGP} \ldots \]

\[ \text{or} \]

\[ \text{Pt becomes CL-intolerant} \ldots \]

*Advanced/RGP-intolerant KCN*: Surgery
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  DALK  ICRS  CXL
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
  - DALK
  - ICRS
  - CXL

What does PK stand for?
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*  
*DALK*  
*ICRS*  
*CXL*

*What does PK stand for?*  
Penetrating keratoplasty

*What does DALK stand for?*
**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…

**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
- DALK
- ICRS
- CXL

What does PK stand for?
Penetrating keratoplasty

What does ICRS stand for?

What does DALK stand for?
Deep anterior lamellar keratoplasty
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…*

**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
- DALK
- ICRS
- CXL

**What does PK stand for?**
Penetrating keratoplasty

**What does DALK stand for?**
Deep anterior lamellar keratoplasty

**What does ICRS stand for?**
Intracorneal ring segments

**What does CXL stand for?**
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

What does PK stand for?
Penetrating keratoplasty

What does DALK stand for?
Deep anterior lamellar keratoplasty

What does ICRS stand for?
Intracorneal ring segments

What does CXL stand for?
Collagen crosslinking
Management of KCN often follows a pattern:

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

KCN progresses $\rightarrow$ specs/soft CLs no longer adequate…

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

KCN progresses $\rightarrow$ cornea too steep to support RGP…

or

Pt becomes CL-intolerant…

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

PK

DALK

ICRS

CXL

---

PK has several things going for it, including:

--

--
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**

---Proven efficacy
---Familiar skill set

**DALK**

**ICRS**

**CXL**

---Proven efficacy

---The skill-set needed to perform it is familiar to most ophthalmologists
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**
--Proven efficacy
--Familiar skill set

**DALK**

**ICRS**

**CXL**

---

*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*

*That said, it has disadvantages as well, including:*

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

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

That said, it has disadvantages as well, including:
--- Lifetime risk of endothelial graft rejection
--- Post-op management requires frequent visits for an extended time

--- DALK
--- ICRS
--- CXL
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**
- Proven efficacy
- Familiar skill set
- Lifetime risk of endothelial rejection
- Protracted post-op course

**DALK**

**ICRS**

**CXL**

---

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

*That said, it has disadvantages as well, including:*

- Lifetime risk of endothelial graft rejection
- Post-op management requires frequent visits for an extended time
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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- KCN progresses → cornea too steep to support RGP…
  - or
  - Pt becomes CL-intolerant…

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

- PK
- DALK
- ICRS
- CXL

PK has several things going for it, including:

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

That said, it has disadvantages as well, including:

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

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

Estimates run as high as 40%
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**
  - Proven efficacy
  - Familiar skill set
  - Lifetime risk of endothelial rejection
  - Protracted post-op course

- **DALK**

- **ICRS**

- **CXL**

*What is the classic exam finding in endothelial rejection?*

*That said, it has disadvantages as well, including:*

--Lifetime risk of endothelial graft rejection

--Post-op management requires frequent visits for an extended time

---

A line of inflammatory precipitates on the endothelial surface

---

A Khodadoust line
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…*

*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**

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

**DALK**

**ICRS**

**CXL**

What is the classic exam finding in endothelial rejection?

A line of inflammatory precipitates on the endothelial surface

That said, it has disadvantages as well, including:

--Lifetime risk of endothelial graft rejection
--Post-op management requires frequent visits for an extended time
Early KCN: Refractive error corrected via spectacles or soft CLs

Moderate KCN: Rigid gas-permeable (RGP) CLs

Advanced/RGP-intolerant KCN: Surgery

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

That said, it has disadvantages as well, including:
- Lifetime risk of endothelial graft rejection
- Post-op management requires frequent visits for an extended time

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?
Khodadoust line
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
- DALK
- ICRS
- CXL

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

**DALK**

**ICRS**

**CXL**

---

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

That said, it has disadvantages as well, including:

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

Khodadoust line
**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…

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

- **DALK**

- **ICRS**

- **CXL**

---

*Briefly, how is DALK performed?*
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

---

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.’) The remaining stroma is carefully dissected away, leaving a recipient bed composed of Descemet’s and endothelium. Descemet’s and the endothelium are stripped from the donor button, which is then sutured in place atop the recipient bed.
Koncerning Keratoconus

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

**DALK has several things going for it, including:**
- --
- --
Koncerning Keratoconus

Management of KCN often follows a pattern:

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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

KCN progresses → cornea too steep to support RGP…
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**Advanced/RGP-intolerant KCN:** Surgery

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

**DALK**
- No risk of endo rejection
- Proven efficacy

**ICRS**

**CXL**

---DALK has several things going for it, including:
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Management of KCN often follows a pattern:

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*or*

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

---

**PK**

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

**DALK**

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

**ICRS**

**CXL**

---

**DALK has several things going for it, including:**

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

That said, it has disadvantages as well, including:

--Technically challenging
--The skill-set is unfamiliar to most ophthalmologists
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**Advanced/RGP-intolerant KCN:** Surgery

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

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

- **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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Management of KCN often follows a pattern:

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

---

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

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

**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  
--The skill-set is unfamiliar to most ophthalmologists*
Management of KCN often follows a pattern:

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

---

**PK**

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

**DALK**

**ICRS**

--No risk of endo rejection

**CXL**

---

**Briefly, how is ICRS surgery performed?**
Management of KCN often follows a pattern:

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

---

**PK**

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

**DALK**

**ICRS**

**CXL**

--No risk of endo rejection

*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
Management of KCN often follows a pattern:

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

---

**PK**

--Proven efficacy

**DALK**

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**ICRS**

--Lifetime risk of endothelial rejection

**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

**How does placement of the ICRSs improve VA?**
Management of KCN often follows a pattern:

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

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

*Advanced/RGP-intolerant KCN:* Surgery

---

**PK**

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

**DALK**

---

**ICRS**

--No risk of endo rejection

**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.

**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).
Koncerning Keratoconus

Ring segment

Side view: cornea flattened centrally

Front view

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

---

ICRS have several things going for them, including:

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

ICRS have several things going for them, including:

--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
Management of KCN often follows a pattern:

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

---

**PK**

--Proven efficacy

--Familiar skill set

--Lifetime risk of endothelial rejection

--Protracted post-op course

**DALK**

**ICRS**

--No tissue removed

--Reversible

--No risk of endothelial rejection

--Proven efficacy

--Technically difficult

--Unfamiliar skill-set

**CXL**

*ICRS have several things going for them, including:*

--No corneal tissue is removed

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

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**CXL**
**Koncerning Keratoconus**

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

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PK

DALK

ICRS

CXL

--No tissue removed

--Reversible

--Unlikely to produce excellent UCVA

--Proven efficacy

--Familiar skill set

--Lifetime risk of endothelial rejection

--Protracted post-op course

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--Unfamiliar skill-set

--No risk of endothelial rejection

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

---

**PK**

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

**DALK**

**ICRS**

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

**CXL**

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

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**ICRS have several things going for them, including:**

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

In fairness, the goal of ICRS placement is not excellent UCVA; rather, what is it?
Early KCN: Refractive error corrected via spectacles or soft CLs

Moderate KCN: Rigid gas-permeable (RGP) CLs

Advanced/RGP-intolerant KCN: Surgery

PK

DALK

ICRS

CXL

**Koncerning Keratoconus**

Management of KCN often follows a pattern:

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  - or
  - Pt becomes CL-intolerant…

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

ICRS have several things going for them, including:

- No tissue removed
- Reversible (i.e., the ICRSs can be removed)
- Less likely to produce excellent UCVA

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

- It is unlikely to result in good UCVA by itself

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
Management of KCN often follows a pattern:

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

---

**Koncerning Keratoconus**

What corneal problem, fundamental to KCN, is addressed by CXL?
Management of KCN often follows a pattern:

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

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

In normal corneal stroma, collagen fibrils are arranged in tightly packed, orderly lattices. These lattices are disrupted in KCN, which allows the cornea to progressively warp. CXL tightens the bonds among corneal fibrils, thereby preventing further warpage and thus halting dz progression.
**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*

---

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

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**Briefly, how is CXL performed?**
Management of KCN often follows a pattern:

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

---

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

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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 radiation. The [substance] acts as a photosensitizer, absorbing the radiation and producing reactive oxygen species. The reactive oxygen species cause cross-linking to occur among corneal fibrils.
Managements of KCN often follows a pattern:

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

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*Advanced/RGP-intolerant KCN: Surgery*

---

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After removal of the corneal epithelium, the stroma is suffused with riboflavin, then subjected to UV radiation. The riboflavin acts as a photosensitizer, absorbing the radiation and producing reactive oxygen species. The reactive oxygen species cause cross-linking to occur among corneal fibrils.
Koncerning Keratoconus

Before CXL (weaker)

After CXL (stronger)

CXL
**Kerncerning Keratoconus**

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

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

- **DALK**
  -- No risk of rejection
  -- Proven efficacy
  -- Technical
  -- Unfamiliar

- **ICRS**
  -- CXL has things going for it, including:
    --

- **CXL**
Management of KCN often follows a pattern:

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

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**PK**

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--Lifetime risk of endothelial rejection
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**ICRS**

--Proven efficacy
--Technical
--Unfamiliar

**CXL**

--No tissue removed

---

*CXL has things going for it, including:

--No corneal tissue is removed*
Management of KCN often follows a pattern:

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

---PK

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--Familiar skill set
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--Technical
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---CXL

--No tissue removed

CXL has things going for it, including:

--No corneal tissue is removed

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

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

---

**PK**  
--Proven efficacy  
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--No risk of endo rejection

**ICRS**  
--Proven efficacy  
--Technical  
--Unfamiliar

**CXL**  
--No tissue removed  
--Unlikely to produce excellent UCVA

_CXL has things going for it, including:_  
--No corneal tissue is removed

*That said, the procedure has disadvantages as well, including:_  
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*KCN progresses → cornea too steep to support RGP… or Pt becomes CL-intolerant…*

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

- PK
  - Proven efficacy
  - Familiar skill set
  - Lifetime risk of endothelial rejection
  - Protracted post-op course
- DALK
- ICRS
  - No risk of endo rejection
  - Proven efficacy
  - Technically difficult
  - Unfamiliar skill-set
- CXL
  - No tissue removed
  - Unlikely to produce excellent UCVA

---

**CXL has things going for it, including:**

- No corneal tissue is removed

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

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In fairness to CXL: As with ICRS placement, its goal is not excellent UCVA; rather, what is it?
Management of KCN often follows a pattern:

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  or

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

---

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--Proven efficacy
--Familiar skill set
--Lifetime risk of endothelial rejection
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**DALK**

**ICRS**

**CXL**

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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
Management of KCN often follows a pattern:

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

---

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

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

**ICRS**:
- No tissue removed
- Reversible
- Unlikely to produce excellent UCVA

**CXL**:
- No tissue removed
- Unlikely to produce excellent UCVA

---

**Summary slide -- no questions**