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

- 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 ectasias.

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

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

What is the incidence rate of KCN?
Which of the following are true concerning keratoconus (KCN)?

- 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** ectasias

*What is the incidence rate of KCN?*

1 in 2000

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

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

**What is the incidence rate of KCN?**
1 in 2000

**Very broadly (like, in one line), what is the pathologic 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
- 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 **weak** hereditary component **F**
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**

*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 **F**
- It has a strong hereditary component **F**

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*
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**
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  \( \text{F} \)
- It has a strong hereditary component  \( \text{F} \)
- Fragmentation of Bowman’s is present  \( \text{T} \)

\textit{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
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
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?
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
- Fold/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? 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

Where (relative to the cone) is the Fleischer ring typically found?
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**

**Koncerning Keratoconus**

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

Where (relative to the cone) is the Fleischer ring typically found?
Along its inferior extent
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  \(\checkmark\)

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

Where (relative to the cone) is the Fleischer ring typically found? Along its inferior extent

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  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

Where (relative to the cone) is the Fleischer ring typically found?
Along its inferior extent

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
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: 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
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

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
- 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

How do you pronounce this?  
TEAL BEN-key

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**

**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?*
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

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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

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 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?

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- It is one of the most common corneal dystrophies **F**
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**Fragmentation of Bowman’s is a histologic hallmark** of KCN. What are some others?

- Iron deposition at the base of the cone
- Corneal thinning
- 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-Beinhke**

In the most recent (2018-19) edition of the BCSC *Cornea* book, in what ‘major category’ are Reis-Bückler and Thiel-Beinhke 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 was formerly known as **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 ‘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-Beihne**

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

**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 histlogic 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-Beinhke

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

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 \( \text{T} \)
- 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

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’ 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: 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?*
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 membrane.
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
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 a weak indication for urgent PK (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?**
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
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 do so with, without sequelae?*
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}
- \textbf{Acute hydrops} \textit{\textbf{not}} an indication for urgent PK \textbf{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; \textit{two words} 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 not an indication for urgent PK **F**

**Koncerning Keratoconus**

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

**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

‘Apical scarring’—that sounds like a bad thing. Is it?

When it resolves, does it? With...
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  \( F \)

**Koncerning Keratoconus**

*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 have **apical scarring**?”

‘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
- 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
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**

\( \text{Koncerning Keratoconus} \)
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 fairly common: **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
- Fragmentation of Bowman’s is present
- Acute hydrops is an indication for urgent PK
- The incidence is higher in South Asia and the Middle East
- Onset typically occurs during adolescence
- Spontaneous rupture is 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 adolescence [F]
- Spontaneous rupture is fairly common [F]
- KCN is strongly associated with Marfan syndrome
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 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?*
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)?

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

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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)?

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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
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**With what conditions is KCN associated?**

**Ocular only:**
- --
- --

**Systemic:**
- --
- --
- --

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

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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

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

With what conditions is KCN associated?

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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

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

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

Summary slide--no questions
What are the 5 classic signs of keratoconus? Which is the first to appear?
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- Scissoring of the retinoscopic reflex (earliest sign)
- Rizzuti’s sign
- Munson’s sign
- Fleischer ring
- Vogt lines
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Define Rizzuti’s sign:

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

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Displacement of the central lower lid by the cone in downgaze
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How can you improve visualization of Munson’s sign?

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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?

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

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Displacement of the central lower lid by the cone in downgaze.

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

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

**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 7 classic associations of keratoconus?

Mnemonic is…
What are the 7 classic associations of keratoconus?

F - floppy eyelid syndrome
L - Leber's congenital amaurosis
A - Atopic disease
M - Marfan's
E - Ehlers-Danlos
D - Down syndrome

Mnemonic is...FLAMED
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
Management of KCN often follows a pattern:

_Early KCN: ?_
Koncerning Keratoconus

Management of KCN often follows a pattern:

*Early KCN*: Refractive error corrected via spectacles or soft CLs
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:** ?
Management of KCN often follows a pattern:

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*Moderate KCN*: Rigid gas-permeable (RGP) CLs
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  (KCN progresses → cornea too steep to support RGP)
  
  or
  
  (Pt becomes CL-intolerant)

*Advanced/RGP-intolerant KCN:* ?
Koncerning Keratoconus

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
Management of KCN often follows a pattern:

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or

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

- PK (Penetrating keratoplasty)
- DALK (Deep anterior lamellar keratoplasty)
- ICRS (intracorneal ring segments)
- CXL (Collagen crosslinking)
Management of KCN often follows a pattern:

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_or_

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_Advanced/RGP-intolerant KCN:_ **Surgery**

PK → DALK → ICRS → CXL

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

--

--
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---

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

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

**DALK**

**ICRS**

**CXL**

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

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**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?**

Estimates run as high as 40%

---

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

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**Proven efficacy**

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**Protracted post-op course**

---

**Briefly, how is DALK performed?**
Koncerning Keratoconus

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**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.

---

PK

DALK

ICRS

CXL

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--Familiar skill set
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Koncerning Keratoconus

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*Advanced/RGP-intolerant KCN:* Surgery

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

**DALK**

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

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**Koncerning 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 endo rejection
--Proven efficacy

**ICRS**

**CXL**

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

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

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**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
Koncerning Keratoconus

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*Advanced/RGP-intolerant KCN:* Surgery

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--Familiar skill set
--Lifetime risk of endothelial rejection
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DALK

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

ICRS

--Proven efficacy
--Technically difficult
--Unfamiliar skill-set

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:

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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
- DALK
- ICRS
- CXL

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

**Briefly, how is ICRS surgery performed?**

**PK**

--No risk of endo 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 risk of endo rejection

**CXL**

---

**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

Management of KCN often follows a pattern:

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   \[(Pt \text{ becomes CL-intolerant)}\]

*Advanced/RGP-intolerant KCN: Surgery*

---

**PK**

--Proven efficacy
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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?
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**Advanced/RGP-intolerant KCN:** Surgery

---

PK

--Proven efficacy

--Familiar skill set

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CXL

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)
Management of KCN often follows a pattern:

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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

---

PK  DALK  ICRS  CXL

-- It is reversible (ie, the ICRSs can be removed)
--- It is unlikely to result in good UCVA by itself
Koncerning Keratoconus

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

---

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

---

**ICRS**

--No tissue removed
--Reversible

---

**PK**

**DALK**

**CXL**

---

ICRS have several things going for them, including:

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

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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
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Koncerning Keratoconus

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

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

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

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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.
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*Advanced/RGP-intolerant KCN*: Surgery

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**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 disease progression.

Briefly, how is CXL performed?

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.
Management of KCN often follows a pattern:

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*Advanced/RGP-intolerant KCN*: Surgery

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**Briefly, how is CXL performed?**

After removal of the corneal epithelium, the stroma is suffused with a 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.
Koncerning Keratoconus

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*Advanced/RGP-intolerant KCN:* Surgery

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**Briefly, how is CXL performed?**

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Koncerning 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 endothelial rejection

**ICRS**

--Proven efficacy
--Technical
--Unfamiliar

**CXL**

*CXL has things going for it, including:*

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

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

--It is unlikely to result in good UCVA by itself
**Koncerning Keratoconus**

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
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DALK
--- No risk of --- No corneal tissue is removed
--- Proven efficacy
--- Technical
--- Unfamiliar

ICRS
--- No tissue removed

CXL
--- No tissue removed

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

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

ICRS

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

CXL

--No tissue removed
--No risk of endothelial rejection
--Proven efficacy
--Technically difficult
--Unfamiliar

CXL has things going for it, including:

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

--It is unlikely to result in good UCVA by itself
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**Advanced/RGP-intolerant KCN:** Surgery

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**DALK**

--No risk of

--Proven efficacy

--Technical

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**ICRS**

CXL has things going for it, including:

--No corneal tissue is removed

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

--It is unlikely to result in good UCVA by itself

**CXL**

--No tissue removed

--Unlikely to produce excellent UCVA

In fairness to CXL: As with ICRS placement, its goal is not excellent UCVA; rather, what is it?
Koncerning 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 graft failure
--Proven efficacy
--Technical
--Unfamiliar

ICRS

--No risk of graft failure
--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:

--It is unlikely to result in good UCVA by itself

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