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What does it mean to say the corneal stroma is inflamed?
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So, there’s pus in the stroma?
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**What does it mean to say the corneal stroma is inflamed?**
It means inflammatory cells are present in the interlamellar stroma.

**So, there’s pus in the stroma?**
No—IK is a nonsuppurative condition.
Interstitial keratitis is an inflammatory condition of the corneal stroma in the absence of primary involvement of either the corneal epithelium or endothelium.

Be sure to take note of this! This is why stromal inflammation resulting from, say, a corneal ulcer eating its way into stroma would not be classified as IK.
Interstitial keratitis is an inflammatory condition of the corneal stroma in the absence of primary involvement of either the corneal epithelium or endothelium.

What are the typical symptoms of IK?

Tearing and photophobia

What are the typical signs of IK, ie, what does it look like at the slit lamp?

Early IK is characterized by perilimbal injection, inflammation of the peripheral stroma, and possibly keratic precipitates. As the disease progresses, deep stromal vessels appear and make their way toward the central cornea.
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- IK? IDK IK! OMG!

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With respect to IK, to what does the term salmon patch refer?
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What are the typical symptoms of IK?
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Early IK is characterized by perilimbal injection, inflammation of the peripheral stroma, and possibly keratic precipitates. As the disease progresses, **deep stromal vessels appear and make their way toward the central cornea.**

*With respect to IK, to what does the term salmon patch refer?*
If the stromal vascularization is particularly dense and the blood flow is exuberant, the resulting color of the cornea has been likened to that of salmon flesh.
Salmon patch in IK
**Interstitial keratitis** is an inflammatory condition of the corneal stroma in the absence of primary involvement of either the corneal epithelium or endothelium.

IK represents a Type hypersensitivity reaction to antigens within the
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Speaking of hypersensitivity reactions…
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How many types of ocular-surface hypersensitivity reactions are there?
Speaking of hypersensitivity reactions…

How many types of ocular-surface hypersensitivity reactions are there?
Type I reactions involve...
Type II reactions involve...
Type III reactions involve...
Type IV reactions involve...

Speaking of hypersensitivity reactions…
How many types of ocular-surface hypersensitivity reactions are there?
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</tr>
</thead>
<tbody>
<tr>
<td>Anaphylaxis</td>
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<td>Type I reactions involve...</td>
<td>Anaphylaxis</td>
<td>Type II reactions involve...</td>
<td>[Two words capturing this rxn]</td>
</tr>
<tr>
<td>Type III reactions involve...</td>
<td></td>
<td>Type IV reactions involve...</td>
<td></td>
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Type I reactions involve Anaphylaxis
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‘Cell-mediated reaction’…Which sort of immune cell is doing the mediating? T-helper cells
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T-helper cells

In what way are T-helper cells mediating the reaction?
Type I reactions involve...Anaphylaxis
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Type III reactions involve...Immune-complex reactions
Type IV reactions involve...Cell-mediated reactions

‘Cell-mediated reaction’...Which sort of immune cell is doing the mediating?
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In what way are T-helper cells mediating the reaction?
In Type IV reactions, T-helpers interact with antigens, thereby becoming activated. Once activated, the T-helpers release chemotactic factors that recruit and activate macrophages.
Anaphylaxis

Type I

Cytotoxic Ab

Type II

Immune-complex reactions

Type III

Cell-mediated reactions

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24-72 hours, which is why this reaction is often referred to as delayed hypersensitivity
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Note that if you remember Type IV as ‘delayed hypersensitivity’…
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Note that if you remember Type IV as ‘delayed hypersensitivity’... the four forms can be remembered with the mnemonic ACID.
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For more on hypersensitivity reactions of the ocular surface, see slide-set K21

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IK? IDK IK! OMG!
The BCSC addresses IK four times in three volumes (twice in the Cornea book). The combined differential is listed below. It long. Let’s work through it...

The only cause to make all four lists
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The rest receive one shout-out only
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Which one is described (by at least one BCSC book) as the “classic cause” of IK?
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Which is the most common cause of IK?
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*Which is the most common cause of IK? HSV (interesting that the most common cause is not considered the ‘classic’ cause!)*
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Let’s review Chlamydial dz. Chlamydia causes three conditions—what are they?

--?
--?
--?

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Is trachoma a serious ocular condition?

Indeed it is—trachoma is a blinding condition. It is #1 cause worldwide.
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**Is trachoma a serious ocular condition?**
Mos def—it is a blinding condition

**Where does it rank in terms of infectious causes of blindness?**

Is it #1 cause worldwide?
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Is adult inclusion conjunctivitis a serious ocular condition?

Nah—it is mild and transient

Does this bug have serious effects elsewhere?

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Let’s review Chlamydial dz. Chlamydia causes three conditions—what are they?

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--- Lymphogranuloma venereum
--- Cogan syndrome
--- Measles virus
--- C trachomatis

--- Adult inclusion conjunctivitis
--- L1, L2, L3
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--- Onchocerca volvulus
--- Acanthamoeba
--- Mumps
--- Sarcoid
--- Atopic keratitis (ie, AKC)
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Is adult inclusion conjunctivitis a serious ocular condition? Nah—it is mild and transient

Does this bug have serious effects elsewhere? Mos def—it is the cause of the classic chlamydial urethritis/cervicitis, ie, the STD form of Chlamydia disease

How does a sexually-transmitted dz cause conjunctivitis?
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Does this bug have serious effects elsewhere?
Mos def—it is the cause of the classic chlamydial urethritis/cervicitis, ie, the STD form of Chlamydia disease

How does a sexually-transmitted dz cause conjunctivitis?
Um, ask your parents
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Surely lymphogranuloma venereum is an STD as well? I mean, it has venere- in its name.
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Surely lymphogranuloma venereum is an STD as well? I mean, it has venere- in its name. It is an STD too, but it’s not ‘the’ chlamydial STD.
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Let’s review Chlamydial dz. Chlamydia causes three conditions—what are they? What property of a Chlamydial bug determines which condition it will cause?

--Trachoma: ?
--Adult inclusion conjunctivitis: ?
--Lymphogranuloma venereum: ?

--Cogan syndrome
--Measles virus

C trachomatis
--C trachomatis serotypes L1, L2, L3
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Let’s review Chlamydial dz. Chlamydia causes three conditions—what are they? What property of a Chlamydial bug determines which condition it will cause?

Its **serotype**

-- **Trachoma**: Serotype
-- **Adult inclusion conjunctivitis**: Serotype
-- **Lymphogranuloma venereum**: Serotype

--- C **trachomatis**

--- L1, L2, L3
--- Serotypes L1, L2, L3
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**Which chlamydia serotypes produce each condition?**

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--- Serotypes L1, L2, L3

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Its serotype

Which chlamydia serotypes produce each condition?
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Its **serotype**

Which *chlamydia* serotypes produce each condition?

--Trachoma: Serotypes…**A, B, C**
--Adult inclusion conjunctivitis: Serotypes…**D-K**
--Lymphogranuloma venereum: Serotype

--Cogan syndrome
--Measles virus

**C trachomatis**

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*Which chlamydia serotypes produce each condition?*

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--Adult inclusion conjunctivitis: Serotypes... **D-K**
--Lymphogranuloma venereum: Serotypes... ?

finally

---Cogan syndrome
---Measles virus
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---finally
Got a mnemonic for remembering these serotypes?

---Trachoma: Serotypes...A, B, C
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IK? IDK IK! OMG!

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Try these:
--Trachoma is as simple as ABC

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Let's work through it...

--- Syphilis --- HSV --- Lyme --- EBV --- VZV --- TB --- Leprosy/Hansen's dz --- Cogan syndrome --- Measles virus --- *C trachomatis* serotypes L1, L2, L3 --- *Leishmania* spp --- *Onchocerca volvulus* --- Acanthamoeba --- Mumps --- Sarcoid --- Atopic keratitis (ie, AKC) --- Vernal keratitis (ie, VKC)

A

Got a mnemonic for remembering these serotypes?

Try these:

--- *Trachoma* is as simple as ABC

--- Adult inclusion conjunctivitis is ‘the’ sexually-transmitted form of Chlamydia.

Can you think of a sex-related word that starts with a D and ends with a K?

--- *C trachomatis*

--- Adult inclusion conjunctivitis: Serotypes… **D-K**

--- Lymphogranuloma venereum: Serotypes… L1, L2, L3
The BCSC addresses IK four times in three volumes (twice in the Cornea book). The combined differential is listed below. It is long. Let's work through it...

--Syphilis--HSV--Lyme--EBV--VZV--TB--Leprosy/Hansen’s dz--Cogan syndrome--Measles virus--C trachomatis serotypes L1, L2, L3--Leishmania spp--Onchocerca volvulus--Acanthamoeba--Mumps--Sarcoid--Atopic keratitis (ie, AKC)--Vernal keratitis (ie, VKC)

Let's review Chlamydial dz. Chlamydia causes three conditions—what are they? What property of a Chlamydial bug determines which condition it will cause? Its serotype. Which chlamydia serotypes produce each condition?

--Trachoma: Serotypes…A, B, C
--Adult inclusion conjunctivitis: Serotypes…D-K
--Lymphogranuloma venereum: Serotypes…L1, L2, L3

Got a mnemonic for remembering these serotypes? Try these:

--Trachoma is as simple as ABC
--Adult inclusion conjunctivitis is ‘the’ sexually-transmitted form of Chlamydia. Can you think of a sex-related word that starts with a D and ends with a K?

--As for the ‘L’ serotypes causing Lymphogranuloma, I assume you got that one.

--C trachomatis
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--Trachoma: Serotypes…A, B, C
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--Lymphogranuloma venereum: Serotypes…L1, L2, L3

Finally: Take note that it is the lymphogranuloma venereum serotypes which are associated with IK!
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Because HSV is the most common cause of IK...

(No question—proceed when ready)
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Because HSV is the most common cause of IK…and syphilis is its ‘classic’ cause…

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Because HSV is the most common cause of IK... and syphilis is its ‘classic’ cause... it should come as no surprise that the BCSC addresses them in depth—and thus, so shall we.

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Because HSV is the most common cause of IK… and syphilis is its ‘classic’ cause… it should come as no surprise that the BCSC addresses them in depth—and thus, so shall we. Of the remaining causes of IK, only one is covered in detail by any BCSC volume—and thus by this slide-set. Which one?
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Because HSV is the most common cause of IK… and syphilis is its ‘classic’ cause… it should come as no surprise that the BCSC addresses them in depth—and thus, so shall we. Of the remaining causes of IK, only one is covered in detail by any BCSC volume—and thus by this slide-set. Which one?

**Cogan syndrome**

*Let’s start with luetic (ie, syphilitic) IK…*
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  T
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  True
- Symptoms include tearing and photophobia  True
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**

**IK? IDK IK! OMG!**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**

**Rule of thumb regarding congenital syphilis manifestations and age:**
--Manifestations presenting within the first two years of life are secondary to...
--Manifestations presenting later in life are secondary to...an
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**

**Rule of thumb regarding congenital syphilis manifestations and age:**
--- Manifestations presenting within the first two years of life are secondary to… **active infection**
--- Manifestations presenting later in life are secondary to… an **immune-mediated process**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease **T**
- Symptoms include tearing and photophobia **T**
- Usual age of presentation is **F**
- Treatment is topical steroids and cycloplegia

Alternative text: "nine years"
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
- Treatment is topical steroids and cycloplegia  **T**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease **T**
- Symptoms include tearing and photophobia **T**
- Usual age of presentation is late teens to early 20s **F**
- Treatment is topical steroids and cycloplegia **T**
- Most patients have bilateral disease
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
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- Most patients have bilateral disease  **T**
Regarding luetic interstitial keratitis (IK), which are true?

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- Symptoms include tearing and photophobia  T
- Usual age of presentation is late teens to early 20s  F
- Treatment is topical steroids and cycloplegia  T
- Most patients have bilateral disease  T

Re bilateral IK, do both eyes always present simultaneously?
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease **T**
- Symptoms include tearing and photophobia **T**
- Usual age of presentation is late teens to early 20s **F**
- Treatment is topical steroids and cycloplegia **T**
- Most patients have bilateral disease **T**

Re bilateral IK, do both eyes always present simultaneously? No
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
- Treatment is topical steroids and cycloplegia  **T**
- Most patients have bilateral disease  **T**

**Re bilateral IK, do both eyes always present simultaneously?**  
No

**Is the extent of involvement always equal between the eyes?**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease - True
- Symptoms include tearing and photophobia - True
- Usual age of presentation is late teens to early 20s - False
- Treatment is topical steroids and cycloplegia - True
- Most patients have bilateral disease - True

Re bilateral IK, do both eyes always present simultaneously? No

Is the extent of involvement always equal between the eyes? No, involvement can be asymmetric
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease \( \text{T} \)
- Symptoms include tearing and photophobia \( \text{T} \)
- Usual age of presentation is late teens to early 20s \( \text{F} \)
- Treatment is topical steroids and cycloplegia \( \text{T} \)
- Most patients have bilateral disease \( \text{T} \)
- About 50% of congenital syphilis cases manifest IK

IK? IDK IK! OMG!
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is nine years  **F**
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- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
- Treatment is topical steroids and cycloplegia  **T**
- Most patients have bilateral disease  **T**
- About 10% of congenital syphilis cases manifest IK  **F**
- About 50% of congenital syphilis cases manifest IK  **F**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease **T**
- Symptoms include tearing and photophobia **T**
- Usual age of presentation is late teens to early 20s **F**
- Treatment is topical steroids and cycloplegia **T**
- Most patients have bilateral disease **T**
- About 50% of congenital syphilis cases manifest IK **F**

What is the natural course of syphilitic IK if it goes untreated?
- It tends to burn itself out in a matter of weeks to months

Irrespective of whether it was treated, what are the common corneal stigmata of resolved IK that may be seen at the slit lamp?

- Formerly-perfused, now-empty stromal blood vessels (aka ghost vessels)
- Corneal scarring may produce visually significant haze and/or astigmatism
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- About 50% of congenital syphilis cases manifest IK  **F**
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Boo!
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  
  True
- Symptoms include tearing and photophobia  
  True
- Usual age of presentation is late teens to early 20s  
  False
- Treatment is topical steroids and cycloplegia  
  True
- Most patients have bilateral disease  
  True

What is the natural course of syphilitic IK if it goes untreated?
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Irrespective of whether it was treated, what are the common corneal stigmata of resolved IK that may be seen at the slit lamp?
- Formerly-perfused, now-empty stromal blood vessels (aka ghost vessels)
- Corneal scarring may produce visually significant haze and/or astigmatism

About 50% of congenital syphilis cases manifest IK  
False
Ghost vessels

Congenital syphilis: Interstitial keratitis
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
- Treatment is topical steroids and cycloplegia  **T**
- Most patients have bilateral disease  **T**
- About 50% of congenital syphilis cases manifest IK  **F**

What is the natural course of syphilitic IK if it goes untreated?
It tends to burn itself out in a matter of weeks to months

Irrespective of whether it was treated, what are the common corneal stigmata of resolved IK that may be seen at the slit lamp?
- Formerly-perfused, now-empty stromal blood vessels (aka **ghost vessels**)
- Corneal scarring may produce visually significant haze and/or **astigmatism**

About 50% of congenital syphilis cases manifest IK  **F**
Regarding luetic interstitial keratitis (IK), which are true?

- Most cases are secondary to congenital disease  **T**
- Symptoms include tearing and photophobia  **T**
- Usual age of presentation is late teens to early 20s  **F**
- Treatment is topical steroids and cycloplegia  **T**
- Most patients have bilateral disease  **T**
- About 50% of congenital syphilis cases manifest IK  **F**

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Corneal scarring/haze

Congenital syphilis: Interstitial keratitis
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- **Circumoral scars**
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

What is the formal term for these circumoral scars?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- **Circumoral scars**
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

What is the formal term for these circumoral scars? ‘Rhagades’
Congenital syphilis: Rhagades
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- **Hutchinson teeth**
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- **Hutchinson teeth**
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

*What is the classic description of Hutchinson teeth? ‘Peg-shaped’*
Congenital syphilis: Teephus
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:

- Axenfeld-Rieger syndrome
- Incontinentia pigmenti
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:

- Axenfeld-Rieger syndrome
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In three words, what sort of condition is A-R?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
- Axenfeld-Rieger syndrome
- Incontinentia pigmenti

In three words, what sort of condition is A-R?
An anterior-segment dysgenesis
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)'

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
- Axenfeld-Rieger syndrome
- Incontinentia pigmenti

In three words, what sort of condition is A-R?
An anterior-segment dysgenesis

If limited to one word, what sort of condition is A-R?

abnormal teeth...
abnormal teeth...

- Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?
  - Circumoral scars
  - Hutchinson teeth
  - Saddle nose
  - Saber shins
  - Cognitive impairment
  - CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
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In three words, what sort of condition is A-R?
An anterior-segment dysgenesis

If limited to one word, what sort of condition is A-R?
A neurocristopathy
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
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In three words, what sort of condition is A-R?
An anterior-segment dysgenesis

If limited to one word, what sort of condition is A-R?
A neurocristopathy

A ‘neurocristopathy’? What does that mean?
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- Circumoral scars
- Hutchinson teeth
- Saddle nose
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Name two other congenital eye syndromes that are associated with abnormal dentition:
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In three words, what sort of condition is A-R?
An anterior-segment dysgenesis

If limited to one word, what sort of condition is A-R?
A neurocristopathy

A ‘neurocristopathy’? What does that mean?
It means ‘a disorder stemming from abnormal neural-crest cell migration and/or differentiation’
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
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Name two other congenital eye syndromes that are associated with abnormal dentition:
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In one word, what sort of condition is IP?

 IK? IDK IK! OMG!
Absent teeth...

**Speaking of congenital syphilis:** What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:

-- Axenfeld-Rieger syndrome
-- Incontinentia pigmenti

In one word, what sort of condition is IP? A phakomatosis
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
- Axenfeld-Rieger syndrome
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In one word, what sort of condition is IP?

A phakomatosis

Briefly, what is a phakomatosis?

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin.
abnormal teeth...

- Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?
  - Circumoral scars
  - Hutchinson teeth
  - Saddle nose
  - Saber shins
  - Cognitive impairment
  - CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
- Axenfeld-Rieger syndrome
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In one word, what sort of condition is IP?
- Phakomatosis

Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the...
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
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Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin

By what more-descriptive name does the BCSC Peds book refer to them?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?
- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Name two other congenital eye syndromes that are associated with abnormal dentition:
- Axenfeld-Rieger syndrome
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In one word, what sort of condition is IP?
- Phakomatosis

Briefly, what is a phakomatosis?
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes, and skin

By what more-descriptive name does the BCSC Peds book refer to them?
As neuro-
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
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- Axenfeld-Rieger syndrome
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In one word, what sort of condition is IP? Phakomatosis

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By what more-descriptive name does the BCSC Peds book refer to them? Neuro-oculo
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

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- Axenfeld-Rieger syndrome
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In one word, what sort of condition is IP?  

Phakomatosis

Briefly, what is a phakomatosis?  
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes, and skin.

By what more-descriptive name does the BCSC Peds book refer to them?  
As neuro-oculocutaneous syndromes.
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- **Saddle nose**

Speaking of saddle-nose deformity... if a pt with it had peripheral ulcerative keratitis (PUK) rather than IK, what two diagnoses should you consider?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- **Saddle nose**

Speaking of saddle-nose deformity...If a pt with it had peripheral ulcerative keratitis (PUK) rather than IK, what two diagnoses should you consider? Relapsing polychondritis (RP), and granulomatosis with polyangiitis (formerly known as two words).
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- **Saddle nose**

_Speaking of saddle-nose deformity… If a pt with it had peripheral ulcerative keratitis (PUK) rather than IK, what two diagnoses should you consider?_  
Relapsing polychondritis (RP), and granulomatosis with polyangiitis (formerly known as Wegener’s granulomatosis)
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
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*Why formerly? Why is the term Wegener’s no longer preferred?*
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

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- Hutchinson teeth
- **Saddle nose**

Speaking of saddle-nose deformity... If a pt with it had peripheral ulcerative keratitis (PUK) rather than IK, what two diagnoses should you consider? Relapsing polychondritis (RP), and granulomatosis with polyangiitis (**formerly known as** Wegener’s granulomatosis)

Why formerly? Why is the term Wegener’s no longer preferred? Because Dr Wegener was a Nazi
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

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- Hutchinson teeth
- **Saddle nose**

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**Relapsing polychondritis** (RP), and **granulomatosis with polyangiitis** (formerly known as Wegener’s granulomatosis)

With respect to a saddle-nose pt with PUK, what would push you toward a dx of:

--RP?
--Granulomatosis with polyangiitis?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose

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- Relapsing polychondritis (RP), and granulomatosi with polyangiitis (formerly known as Wegener’s granulomatosis)

With respect to a saddle-nose pt with PUK, what would push you toward a dx of:

--RP? The presence of ear-cartilage inflammation and/or deformity
--Granulomatosi with polyangiitis?
Auricular damage in RP

Acute inflammation

Post-inflammation deformity

IK? IDK IK! OMG!
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?
- Circumoral scars
- Hutchinson teeth
- **Saddle nose**

**Speaking of saddle-nose deformity…**If a pt with it had peripheral ulcerative keratitis (PUK) rather than IK, what two diagnoses should you consider?
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*With respect to a saddle-nose pt with PUK, what would push you toward a dx of:*
--RP? The presence of ear-cartilage inflammation and/or deformity
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Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

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With respect to a saddle-nose pt with PUK, what would push you toward a dx of:

--**RP**? The presence of ear-cartilage inflammation and/or deformity
--**Granulomatosis with polyangiitis**? The presence of chronic sinusitis
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
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With respect to a saddle-nose pt with PUK, what would push you toward a dx of:

---RP? The presence of ear-cartilage inflammation and/or deformity
---**Granulomatosis with polyangiitis**? The presence of chronic sinusitis (especially if the nasal discharge is **bloody**.
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- **Saddle nose**

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--RP? The presence of ear-cartilage inflammation and/or deformity
--Granulomatosis with polyangiitis? The presence of chronic sinusitis (especially if the nasal discharge is **bloody** )
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

In the present context, to what does the term Hutchinson’s triad refer?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

In the present context, to what does the term Hutchinson’s triad refer? To the three stigmata of congenital syphilis that are especially common.
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Which three comprise Hutchinson’s triad?

In the present context, to what does the term Hutchinson’s triad refer?
To the three stigmata of congenital syphilis that are especially common.
Speaking of congenital syphilis: What are the classic signs (other than **IK**, duh)?

- Circumoral scars
- **Hutchinson teeth**
- Saddle nose
- Saber shins
- Cognitive impairment
- **CN8 deafness**

Which three comprise Hutchinson’s triad?
- Interstitial keratitis
- Deafness
- Hutchinson teeth

In the present context, to what does the term Hutchinson’s triad refer?
To the three stigmata of congenital syphilis that are especially common.
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

What is the classic retinal finding in congenital lues?
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

What is the classic retinal finding in congenital lues?
Salt-and-pepper retinitis
Congenital syphilis: Salt-and-pepper retinitis
Speaking of congenital syphilis: What are the classic signs (other than IK, duh)?

- Circumoral scars
- Hutchinson teeth
- Saddle nose
- Saber shins
- Cognitive impairment
- CN8 deafness

Next we will turn our attention to HSV
1) ?

2) ?

You should think of anterior HSV eye dz as having two very broad forms. What are they?
1) Primary ocular disease

- Usually a unilateral blepharoconjunctivitis
- Presents with lid margin vesicles/ulcers and bulbar conjunctivitis

2) Recurrent ocular disease

You should think of anterior HSV eye dz as having two very broad forms. What are they?
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected?
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected?  
No! Remember, herpes virus infection is never cleared--rather, it becomes latent within the host. Thus, recurrence means the virus is **reactivated**, not re-acquired.
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected?
No! Remember, herpes virus infection is never cleared—rather, it becomes latent within the host. Thus, recurrence means the virus is reactivated, not re-acquired.

Where in the body do herpesviruses establish their latency?
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected?
No! Remember, herpes virus infection is never cleared—rather, **it becomes latent within the host**. Thus, recurrence means the virus is **reactivated**, not re-acquired.

*Where in the body do herpesviruses establish their latency?*
Different members of the herpesvirus family take up residence in different cell types. HSV-1 and HSV-2 hole up in sensory neural ganglia.*
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected?
No! Remember, herpes virus infection is never cleared—rather, it becomes latent within the host. Thus, recurrence means the virus is reactivated, not re-acquired.

Where in the body do herpesviruses establish their latency?
Different members of the herpesvirus family take up residence in different cell types. 
**HSV-1 and HSV-2 hole up in sensory neural ganglia.**

Which sensory ganglion harbors the virions responsible for recurrent ocular dz?
1) Primary ocular disease

2) Recurrent ocular disease

Does ‘recurrence’ mean the pt gets re-infected? No! Remember, herpes virus infection is never cleared--rather, it becomes latent within the host. Thus, recurrence means the virus is reactivated, not re-acquired.

Where in the body do herpesviruses establish their latency? Different members of the herpesvirus family take up residence in different cell types. HSV-1 and HSV-2 hole up in sensory neural ganglia.

Which sensory ganglion harbors the virions responsible for recurrent ocular dz? The trigeminal (CN5; ‘stellate’) ganglion
1) Primary ocular disease
   --Usually a *unilateral* blepharoconjunctivitis

2) Recurrent ocular disease
1) Primary ocular disease
   --Usually a *unilateral* blepharoconjunctivitis

2) Recurrent ocular disease
1) Primary ocular disease
--Usually a *unilateral* blepharoconjunctivitis
--Presents with lid margin and bulbar

2) Recurrent ocular disease

IK? IDK IK! OMG!
1) Primary ocular disease
   --Usually a unilateral blepharoconjunctivitis
     --Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
HSV blepharoconjunctivitis
1) Primary ocular disease
--Usually a unilateral blepharoconjunctivitis
  --Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
a) 

b) 

c) 

d) 

Four distinct ocular manifestations (think broadly and anatomically)
1) Primary ocular disease
--Usually a *unilateral* blepharoconjunctivitis
    --Presents with lid margin *vesicles/ulcers* and bulbar *conj ulcers*

2) Recurrent ocular disease
   a) *Blepharoconjunctivitis*
   b) *Keratitis*
   c) *Iridocyclitis*
   d) *Trabeculitis*

Four distinct ocular manifestations
(think broadly and anatomically)
1) Primary ocular disease
--Usually a unilateral blepharoconjunctivitis
  --Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
a) Blepharoconjunctivitis
b) Keratitis
   --?
   --?
   Three specific and distinct keratitis subtypes
   --?
c) Iridocyclitis
d) Trabeculitis
1) Primary ocular disease
--Usually a unilateral **blepharoconjunctivitis**
  --Presents with lid margin **vesicles/ulcers** and bulbar **conj ulcers**

2) Recurrent ocular disease
a) **Blepharoconjunctivitis**
b) **Keratitis**
   --Epithelial
   --Stromal
   --Endotheliitis

Three specific and distinct keratitis subtypes

c) **Iridocyclitis**

d) **Trabeculitis**
1) Primary ocular disease
--Usually a unilateral **blepharoconjunctivitis**
  --Presents with lid margin **vesicles/ulcers** and bulbar **conj ulcers**

2) Recurrent ocular disease
a) **Blepharoconjunctivitis**
b) **Keratitis**
   --Epithelial: c/o three words
   --Stromal
   --Endotheliitis
c) **Iridocyclitis**

d) **Trabeculitis**
1) Primary ocular disease
--Usually a unilateral **blepharoconjunctivitis**
  --Presents with lid margin **vesicles/ulcers** and bulbar **conj ulcers**

2) Recurrent ocular disease
a) **Blepharoconjunctivitis**
b) **Keratitis**
   --Epithelial: c/o **foreign body sensation**. Classic sign: **Dendrites**
   --Stromal
   --**Endotheliitis**
c) **Iridocyclitis**
d) **Trabeculitis**
HSV epithelial keratitis
1) Primary ocular disease
   --Usually a unilateral blepharoconjunctivitis
     --Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
   a) Blepharoconjunctivitis
   b) Keratitis
      --Epithelial: c/o foreign body sensation. Classic sign: Dendrites
      --Stromal

      --Endotheliitis (aka two words): Presents as -shaped edematous area with

   c) Iridocyclitis

   d) Trabeculitis
1) Primary ocular disease
--Usually a unilateral blepharoconjunctivitis
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   a) Blepharoconjunctivitis
   b) Keratitis
      --Epithelial: c/o foreign body sensation. Classic sign: Dendrites
      --Stromal
      --Endotheliitis (aka disciform keratitis): Presents as disc-shaped edematous area with KP
   c) Iridocyclitis
   d) Trabeculitis
HSV endotheliitis/disciform keratitis
1) Primary ocular disease
--Usually a *unilateral* blepharoconjunctivitis
--Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
a) *Blepharoconjunctivitis*
b) *Keratitis*
   --Epithelial: c/o foreign body sensation. Classic sign: Dendrites
   --Stromal
   --? [Two subtypes of stromal keratitis]
   --?
   --*Endotheliitis* (aka *disciform keratitis*): Presents as disc-shaped edematous area with KP
c) *Iridocyclitis*
d) *Trabeculitis*
1) Primary ocular disease
--Usually a *unilateral* **blepharoconjunctivitis**
  --Presents with lid margin **vesicles/ulcers** and bulbar **conj ulcers**

2) Recurrent ocular disease
a) **Blepharoconjunctivitis**

b) **Keratitis**
  --Epithelial: c/o **foreign body sensation**. Classic sign: **Dendrites**
  --Stromal
    --**Necrotizing**
    --**Interstitial**

**Two subtypes of stromal keratitis**

--**Endotheliitis** (aka **disciform keratitis**): Presents as **disc-shaped edematous area** with **KP**

c) **Iridocyclitis**

d) **Trabeculitis**
1) Primary ocular disease
--Usually a unilateral blepharoconjunctivitis
--Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
a) Blepharoconjunctivitis
b) Keratitis
   --Epithelial: c/o foreign body sensation. Classic sign: Dendrites
   --Stromal
      --Necrotizing: Looks like an [ ]
      --Interstitial
   c) Iridocyclitis (aka disciform keratitis): Presents as disc-shaped edematous area with KP
   d) Trabeculitis
1) Primary ocular disease
--Usually a unilateral **blepharoconjunctivitis**
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  --Epithelial: c/o **foreign body sensation**. Classic sign: **Dendrites**
  --Stromal
    --**Necrotizing**: Looks like an **ulcer**
  ⭐--Interstitial
    --**Endotheliitis** (aka **disciform keratitis**): Presents as disc-shaped edematous area with **KP**
c) *Iridocyclitis*

d) *Trabeculitis*
1) Primary ocular disease
--Usually a unilateral blepharoconjunctivitis
  --Presents with lid margin vesicles/ulcers and bulbar conj ulcers

2) Recurrent ocular disease
a) Blepharoconjunctivitis
b) Keratitis
   --Epithelial: c/o foreign body sensation. Classic sign: Dendrites
   --Stromal
      --Necrotizing: Looks like an ulcer (ie, suppurative, with an overlying epithelial defect)
      --Interstitial
      --Endotheliitis (aka disciform keratitis): Presents as disc-shaped edematous area with KP
d) Iridocyclitis
d) Trabeculitis
HSV necrotizing keratitis
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For more on anterior HSV dz, see slide-set K23

c) Iridocyclitis
d) Trabeculitis
1) Primary ocular disease
--Usually a **unilateral blepharoconjunctivitis**
  --Presents with lid margin **vesicles/ulcers** and bulbar **conj ulcers**

2) Recurrent ocular disease
a) **Blepharoconjunctivitis**
   --Epithelial: c/o **foreign body sensation**. Classic sign: **Dendrites**
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b) **Keratitis**
   --Epithelial: c/o **foreign body sensation**. Classic sign: **Dendrites**
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c) **Iridocyclitis**
   --Can be granulomatous or non-granulomatous
   --Classic sign: patchy iris transillumination defects

d) **Trabeculitis**

For more on anterior HSV dz, see slide-set K23

**Next we will turn our attention to Cogan syndrome**
Regarding interstitial keratitis (IK) associated with Cogan syndrome, which are true?

- It usually strikes children around age 9 years
Regarding interstitial keratitis (IK) associated with Cogan syndrome, which are true?

- It usually strikes children around age 9 years. **F**

**IK? IDK IK! OMG!**
Regarding interstitial keratitis (IK) associated with Cogan syndrome, which are true?

- It usually strikes children around age 9 years  **F**
- It often follows an URTI by 1-2 weeks or so  
  *(Upper respiratory tract infection)*
Regarding interstitial keratitis (IK) associated with Cogan syndrome, which are true?

- It usually strikes children around age 9 years  F
- It often follows an URTI by 1-2 weeks or so  T
Regarding interstitial keratitis (IK) associated with **Cogan syndrome**, which are true?

- It usually strikes **children around age 9 years** — **F**
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- Some patients have serologic evidence of granulomatosis with polyangiitis
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- Some patients have serologic evidence of granulomatosis with polyangiitis: **Do these pts go on to manifest a systemic vasculitic process?**
- Some patients have serologic evidence of polyarteritis nodosum: **F**

*Q*
Regarding interstitial keratitis (IK) associated with Cogan syndrome, which are true?

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Do these pts go on to manifest a systemic vasculitic process? Some do, and it can be life-threatening
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Does this mean topical steroids play no role in managing Cogan syndrome?

- Not at all; in fact, topical steroids are the mainstay of treatment for the IK component of Cogan syndrome.

What other med(s) is/are indicated in managing Cogan syndrome?

- PO steroids (or on occasion, IMT) are used to treat the CNS manifestations.

How urgent is the need to start systemic steroids?

- Quite. Cogan's tends to progress rapidly, and profound, permanent hearing loss and even death can result if systemic steroid therapy isn't initiated promptly.
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