Corneal Dystrophies

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What are the four categories of corneal dystrophies?

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

What are the four categories of corneal dystrophies?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

Endothelial Dystrophies

1) ***?***
2) ***?***
3) ***?***

What are the three endothelial dystrophies?

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

What are the three endothelial dystrophies?

Endothelial Dystrophies

1) Fuchs endothelial corneal dystrophy
2) Posterior polymorphous corneal dystrophy
3) Congenital hereditary endothelial dystrophy

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does FECD begin to manifest?

Endothelial Dystrophies

1) **Fuchs endothelial corneal dystrophy**
2) Posterior polymorphous corneal dystrophy
3) Congenital hereditary endothelial dystrophy

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does FECD begin to manifest?

Usually in the 30s, if not later. (There is a rare 'early onset' variant that declares in childhood.)

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How does it present? What is seen at the slit lamp?

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one word (wartlike excrescences on Descemet's) are noted, first in the central region, and later across the entire cornea.

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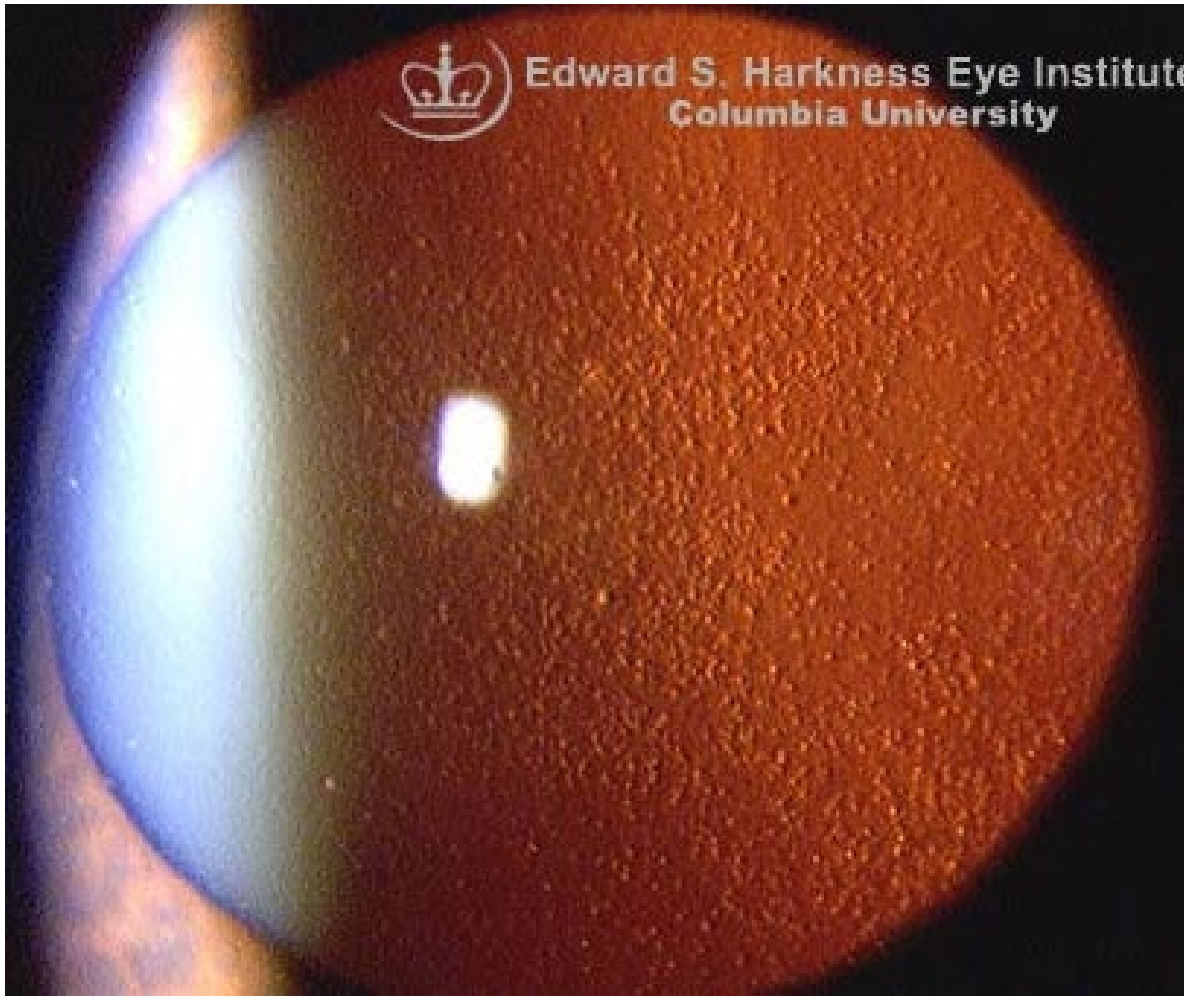
Corneal Dystrophies

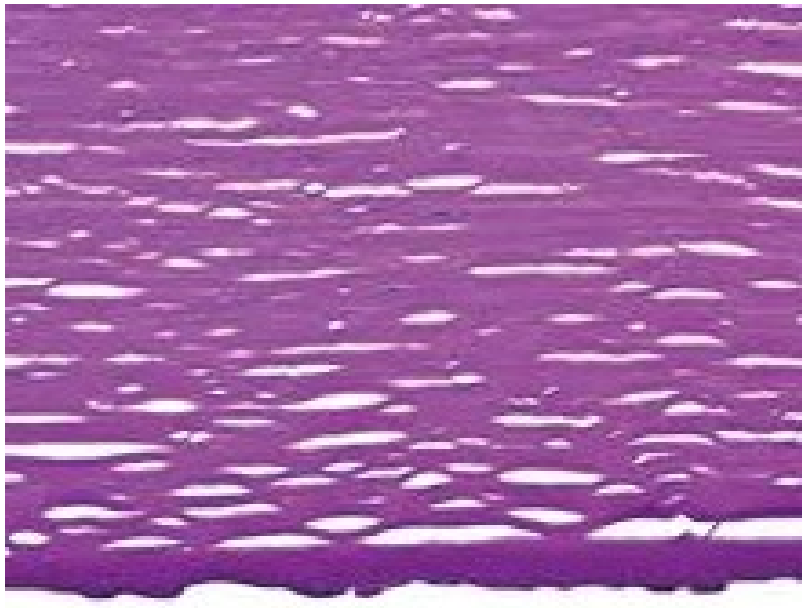
Fuchs endothelial corneal dystrophy. Cornea guttata, slit-lamp view

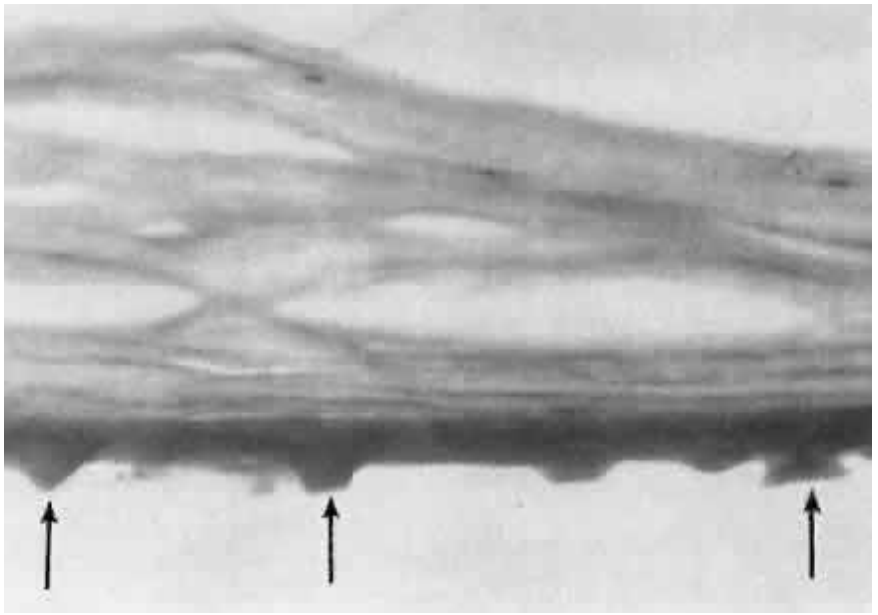
Corneal Dystrophies



Fuchs endothelial corneal dystrophy. Cornea guttata in retroillumination

Corneal Dystrophies





Fuchs endothelial corneal dystrophy. Light microscopy: cornea guttata in the form of focal excrescences at the level of the endothelium.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Usually in the 30s, if not later. (There is a rare 'early onset' variant that declares in childhood.)

How does it present? What is seen at the slit lamp?
Guttata (wartlike excrescences on Descemet's) are noted, first in the central region, and later across the entire cornea. The abnormal endothelial layer takes on a metallic appearance (keywords: two words).

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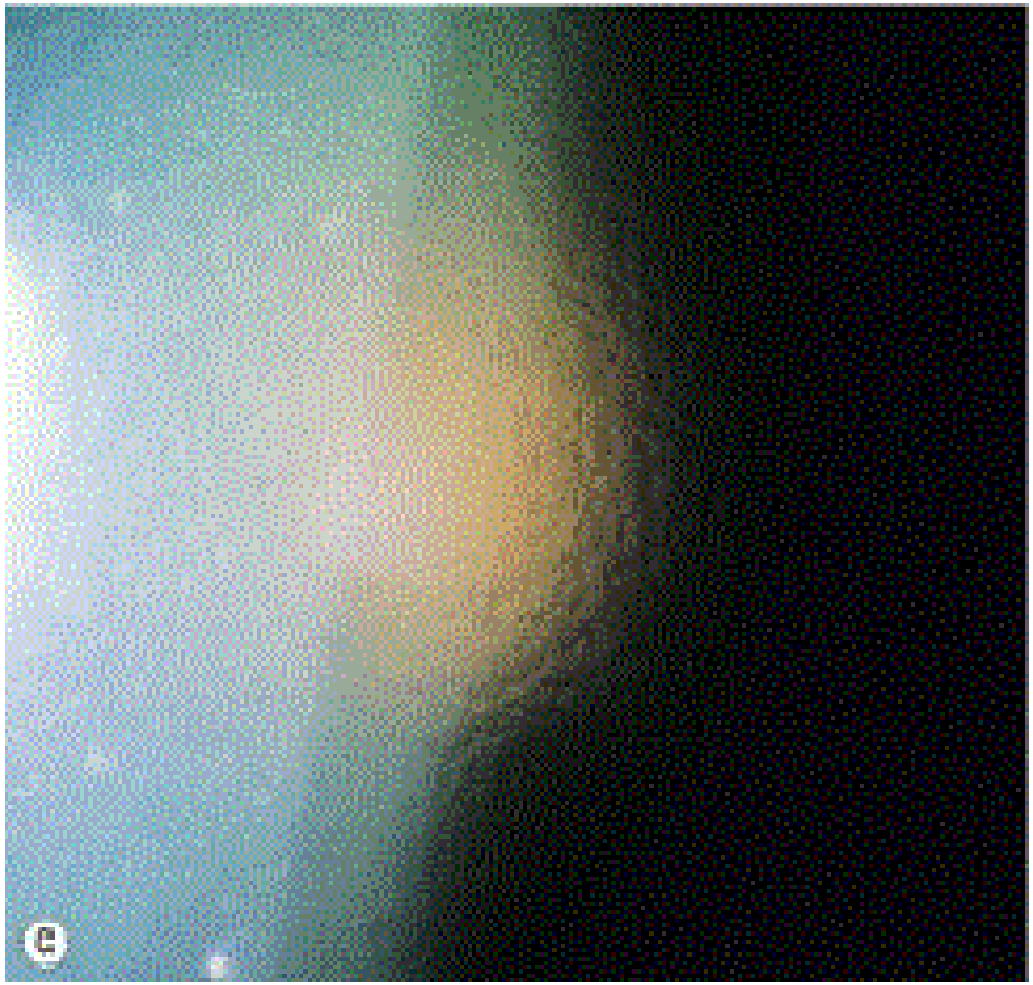

Fuchs endothelial corneal dystrophy. The appearance wrought by dense guttata has been likened to that of '**beaten bronze**.'

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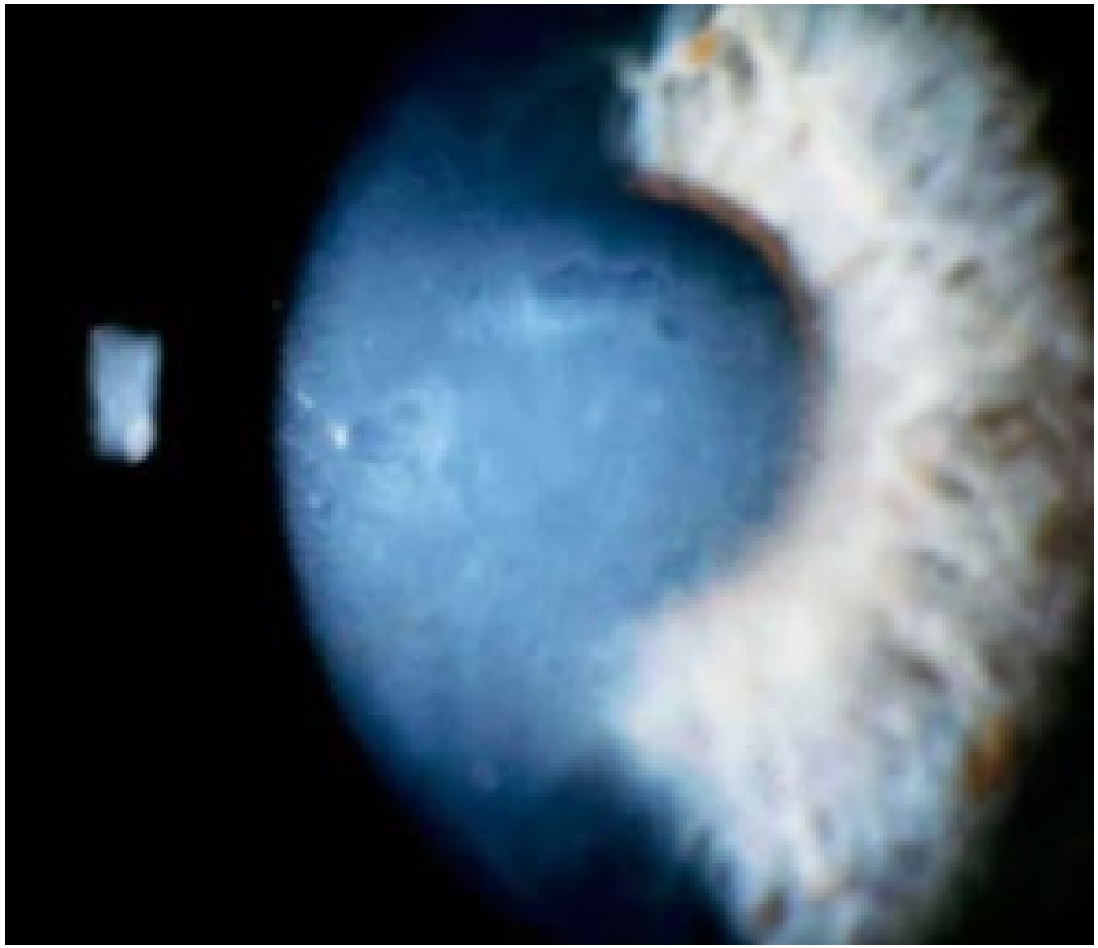
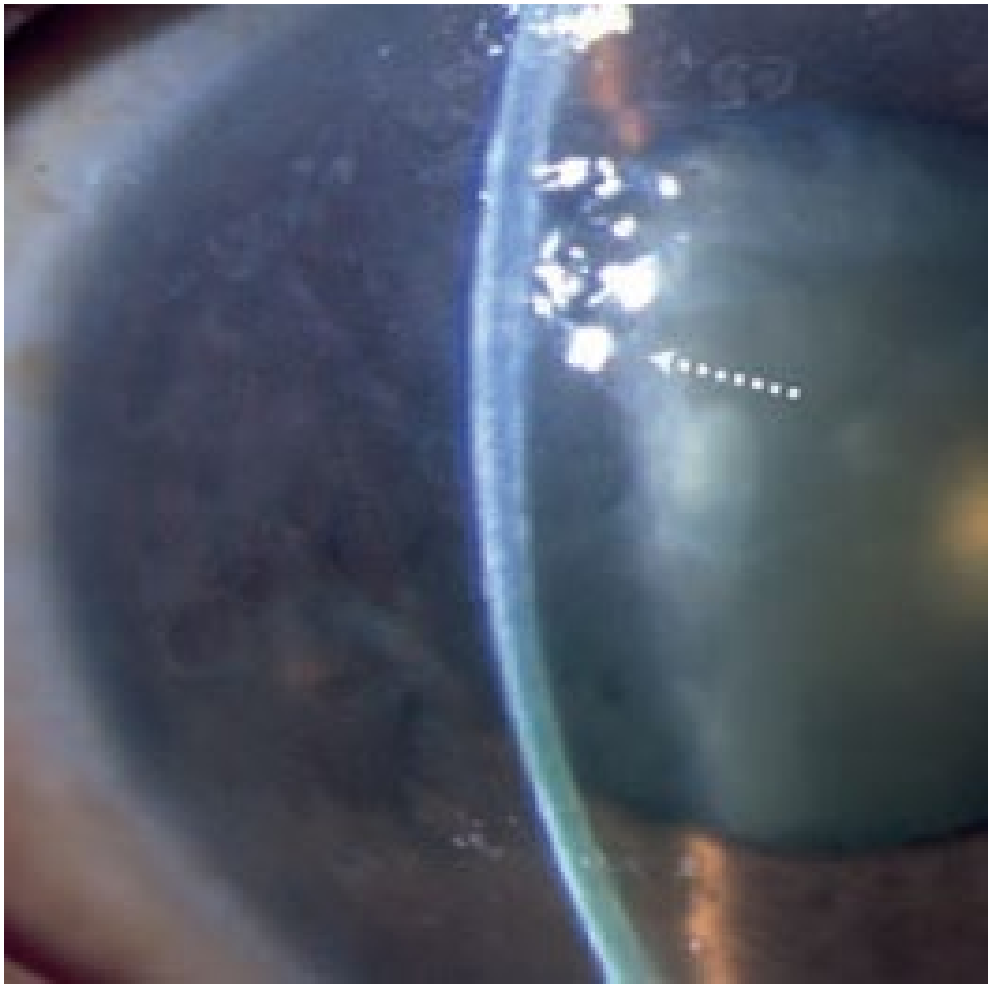


Fuchs endothelial corneal dystrophy. Bullous keratopathy due to endothelial decompensation.

Corneal Dystrophies



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Is it painful?

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If/when epithelial bullae rupture, yes

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Does it affect vision?

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If/when epithelial bullae rupture, yes

Does it affect vision?
Stromal/epithelial edema causes decreased vision, which is usually worse in the morning

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Why is vision worse in the morning?

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Why is vision worse in the morning?
Because the eyelids are closed overnight, corneal edema (and thus vision) is at its worst in the morning

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What is the histologic hallmark of FECD on confocal microscopy?
1)

2)

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1) Endothelial cell abnormalities, including:
--Decreased density (ie, there are fewer cells than there should be)
--Size abnormalities

2) The presence of guttata

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What is the histologic hallmark of FECD on confocal microscopy?
1) Endothelial cell abnormalities, including:
--Decreased density (ie, there are fewer cells than there should be)
--Size abnormalities, including the presence of cells that are much too large (known as one word)

2) The presence of guttata

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2) The presence of guttata

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--Size abnormalities, including the presence of cells that are much too large (known as **polymegathism**)
and an increase in the cell-to-cell variability in size (known as different word)
2) The presence of guttata

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--Size abnormalities, including the presence of cells that are much too large (known as **polymegathism**) and an increase in the cell-to-cell variability in size (known as **pleomorphism**)
2) The presence of guttata

Endothelial Dystrophies

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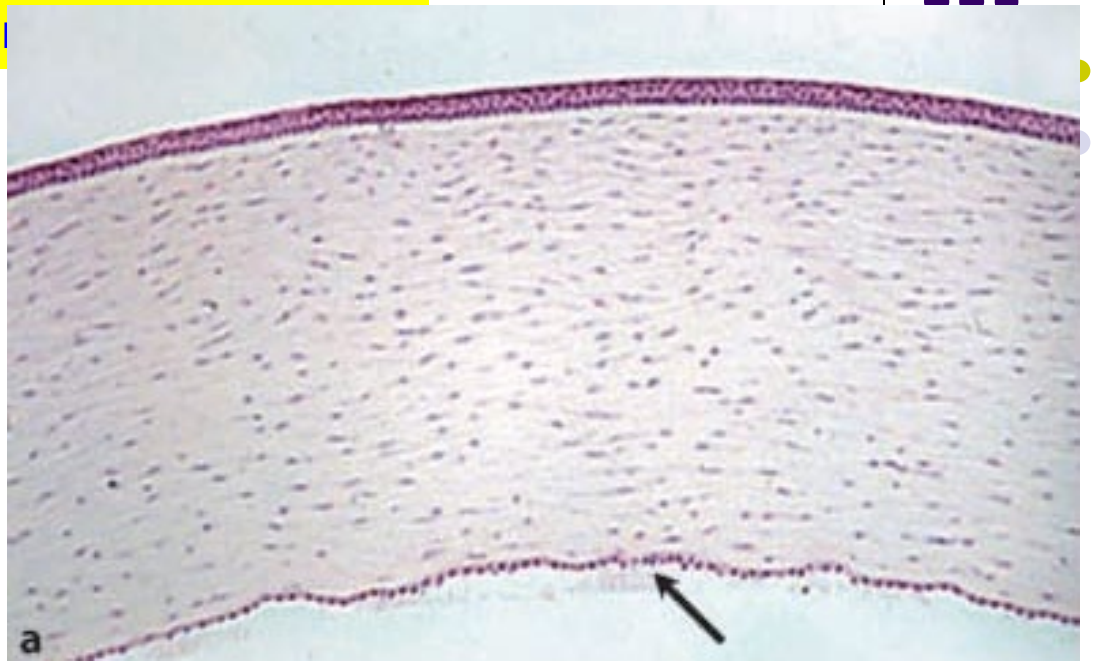
Normal human cornea. Note numerous endothelial cell nuclei lining the posterior surface (*arrow*).



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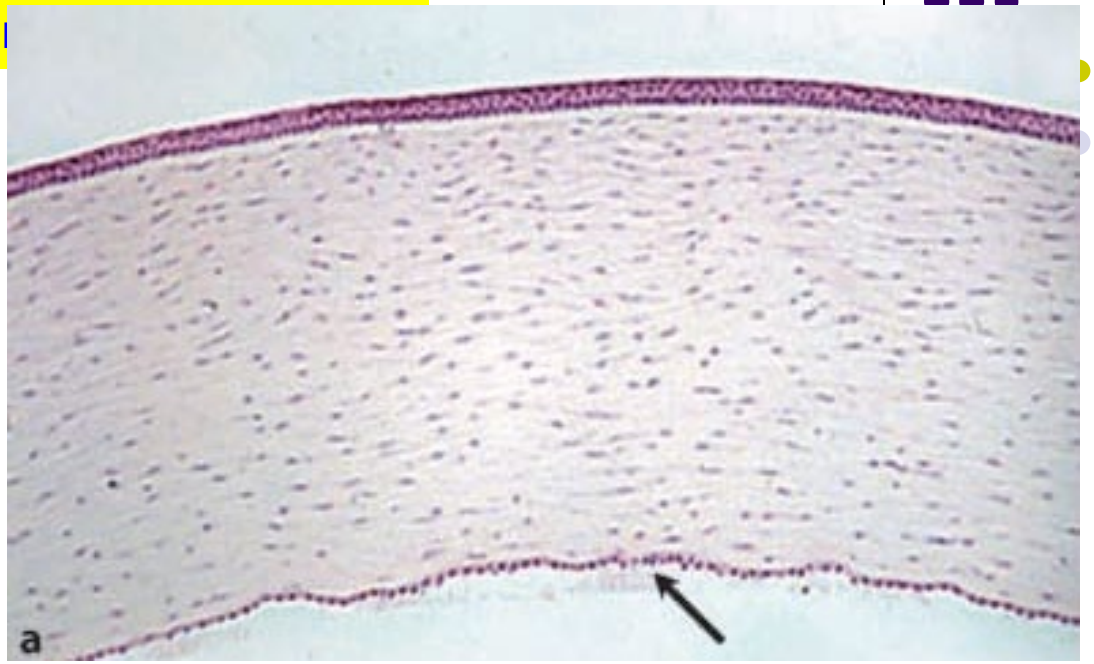
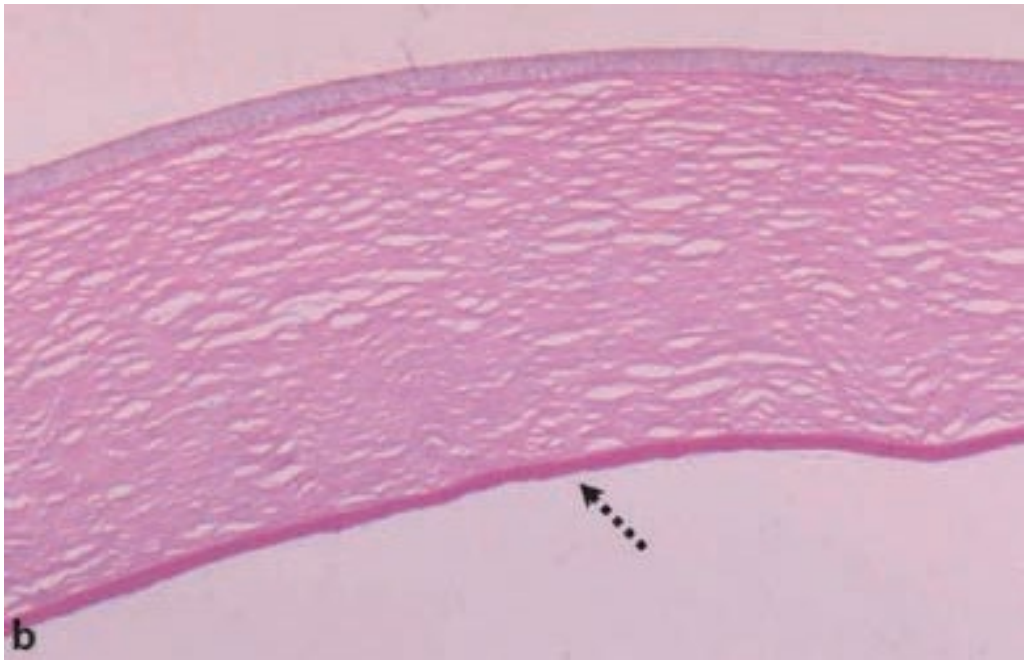
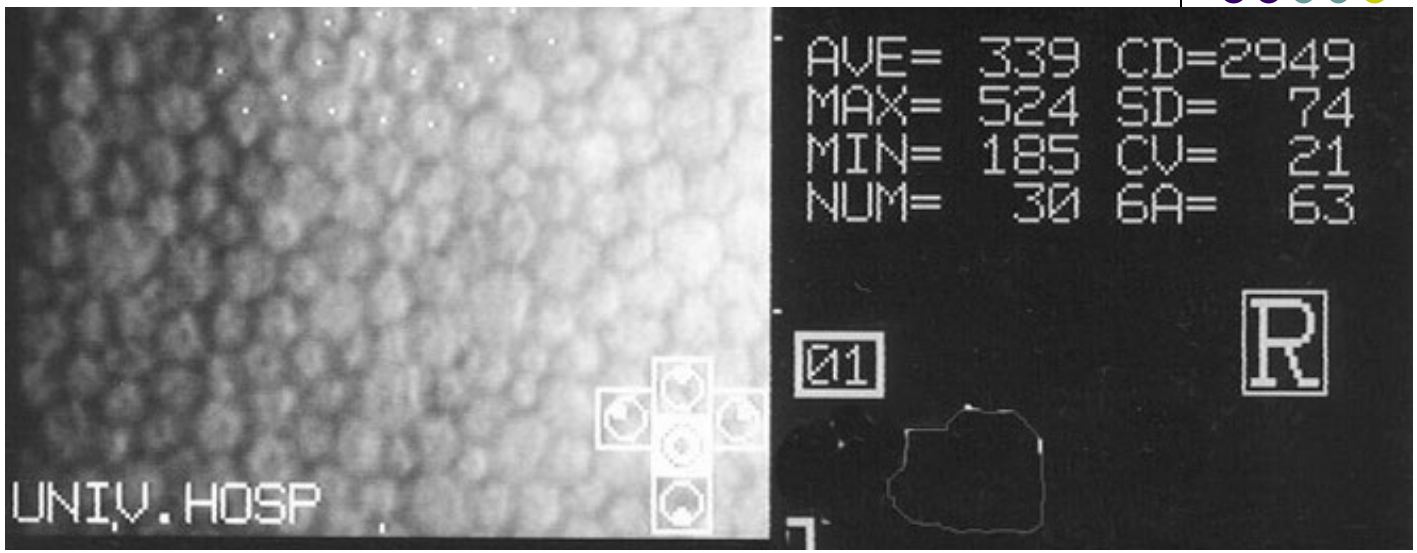


Fuchs endothelial corneal dystrophy. Light microscopy section of FED cornea. Note the markedly thickened Descemet's membrane and the absence of endothelial cell nuclei on the posterior surface (*dashed arrow*).



Corneal Dystrophies

Specular microscopic image, **normal cornea**. Note the plethora of polygonal cells of uniform size, and the absence of empty spaces. The cell density is 2949/mm^2 (nl 2-3K, avg ~2400)



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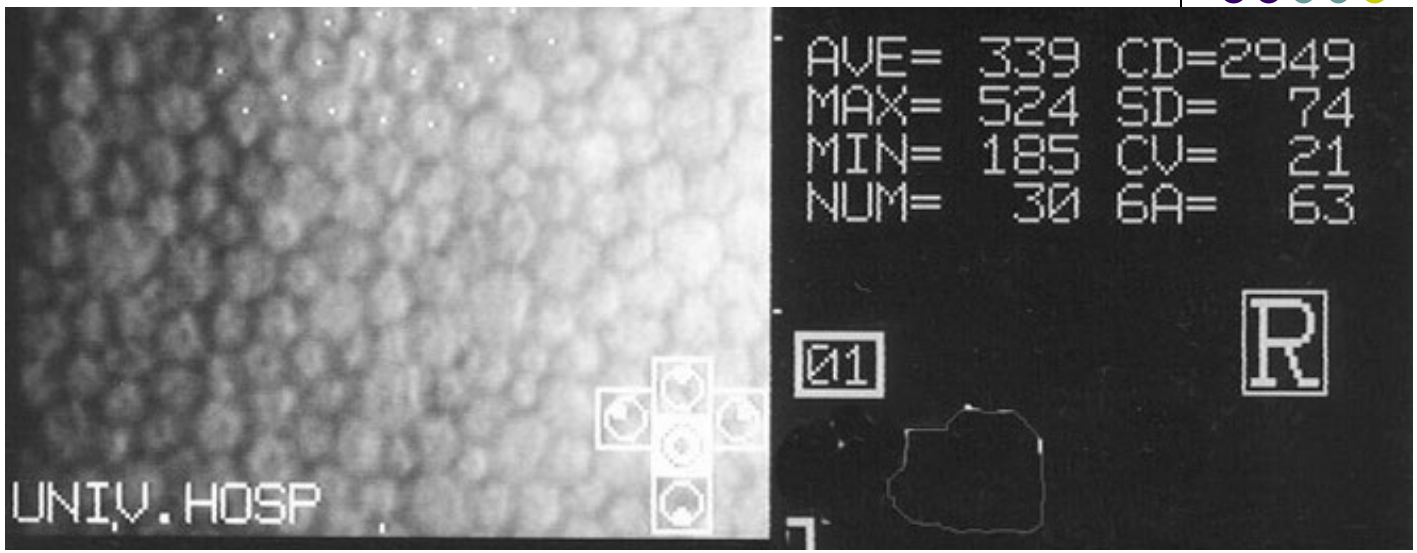

Specular microscopic image, **Fuchs**. Note the polymegathism and polymorphism, and the empty spaces (= guttata). The cell density is 1763/mm^2.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does PPMD begin to manifest?

Endothelial Dystrophies

1) Fuchs endothelial corneal dystrophy
2) **Posterior polymorphous corneal dystrophy**
3) Congenital hereditary endothelial dystrophy

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does PPMD begin to manifest?
Usually teens or 20s; rarely in childhood

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How does it present? What is seen at the slit lamp?

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does PPMD begin to manifest?
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How does it present? What is seen at the slit lamp?
Endothelial-surface abnormalities, including:
- two words, either isolated or in groups
--Linear gray two-words opacities
--Diffuse 'word' opacities that often have an overlying haze in the posterior stroma

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does PPMD begin to manifest?
Usually teens or 20s; rarely in childhood

How does it present? What is seen at the slit lamp?
Endothelial-surface abnormalities, including:
--**Vesicular lesions** , either isolated or in groups
--Linear gray **band-shaped** opacities
--Diffuse '**geographic**' opacities that often have an overlying haze in the posterior stroma

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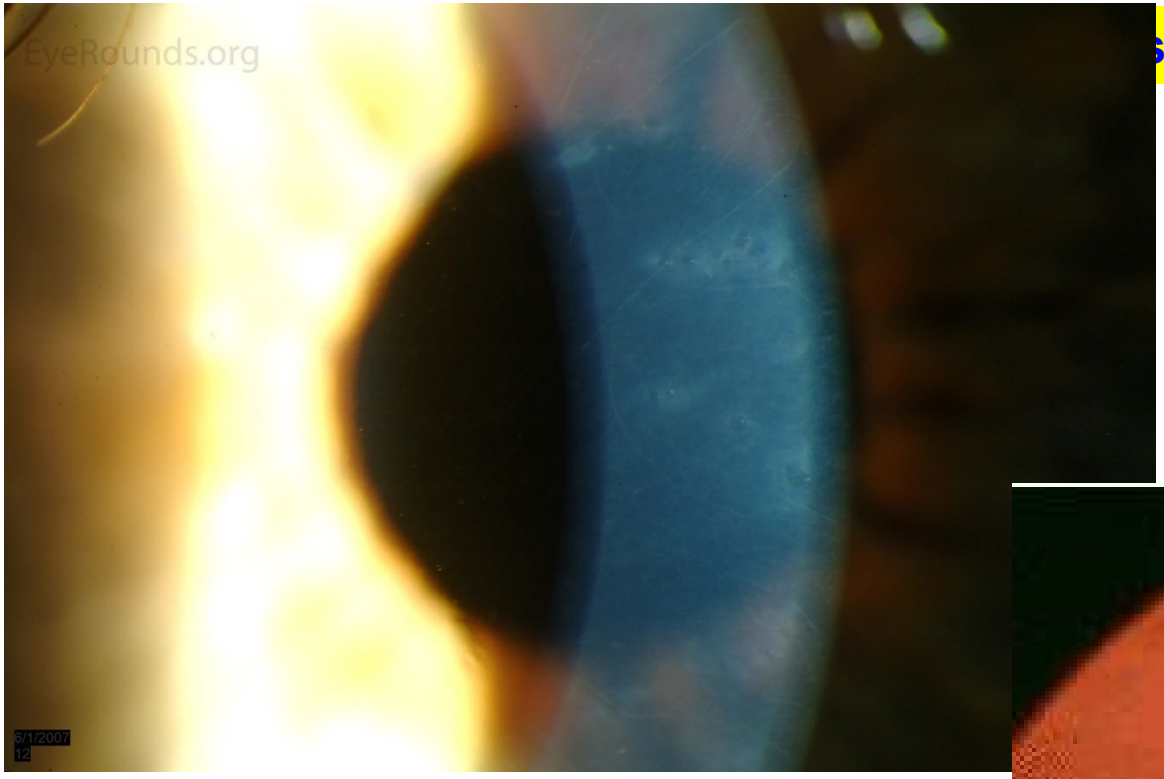
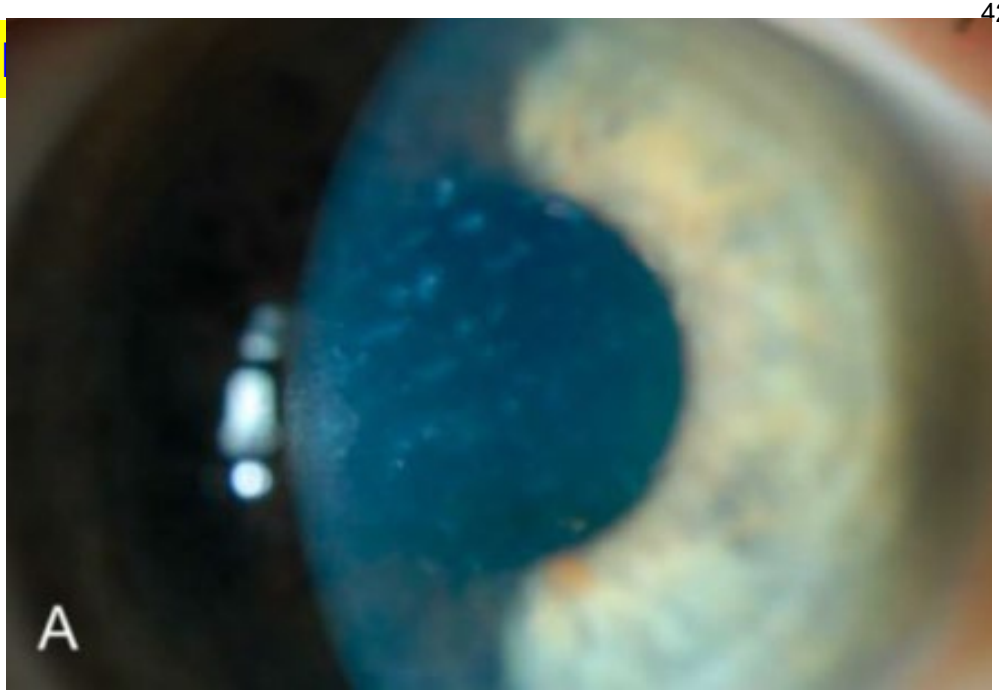
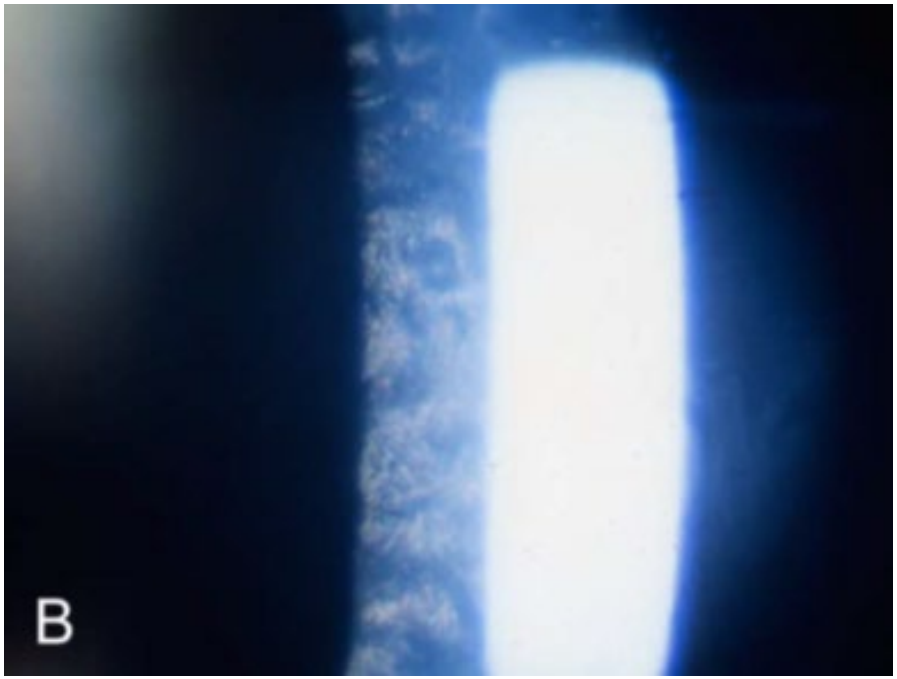


Posterior polymorphous corneal dystrophy:
Vesicular lesions

Corneal



Posterior polymorphous corneal dystrophy
A: Endothelial plaque-like lesions
B: Irregular crater-like figures on Descemet membrane viewed with specular reflection



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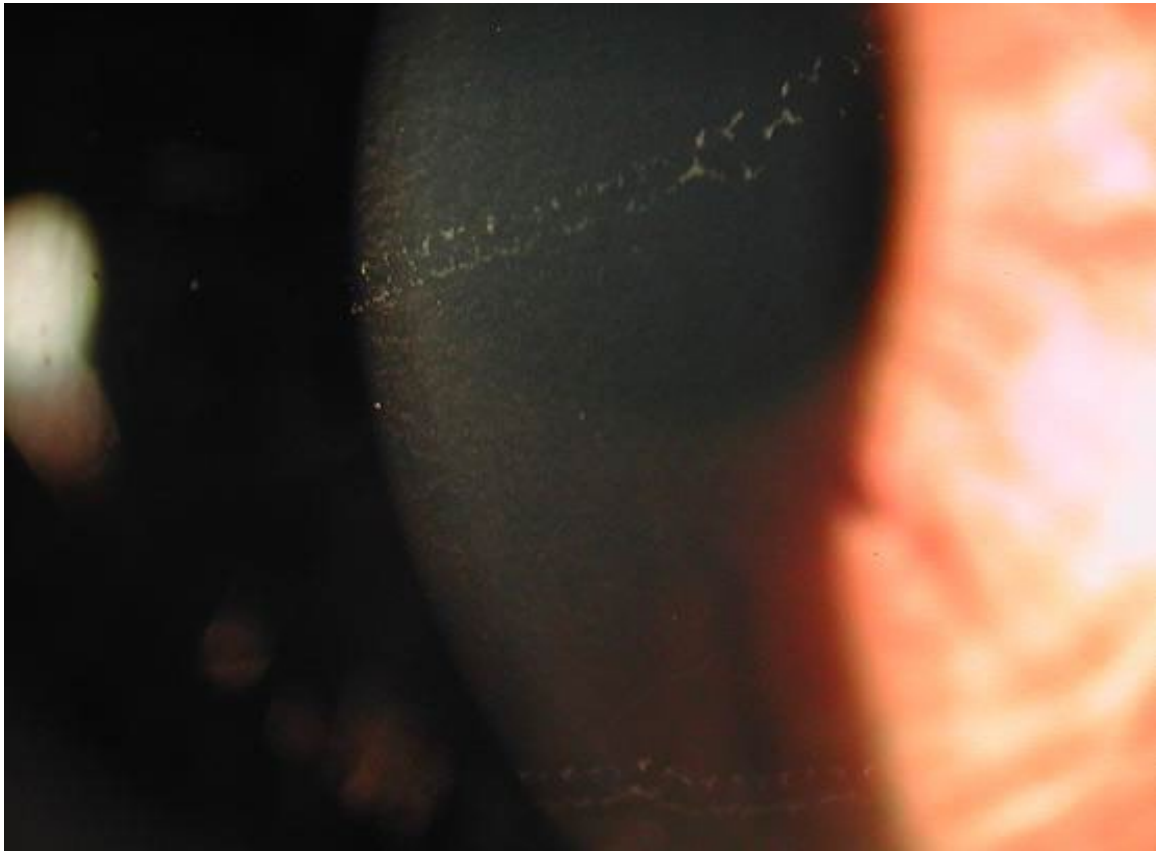
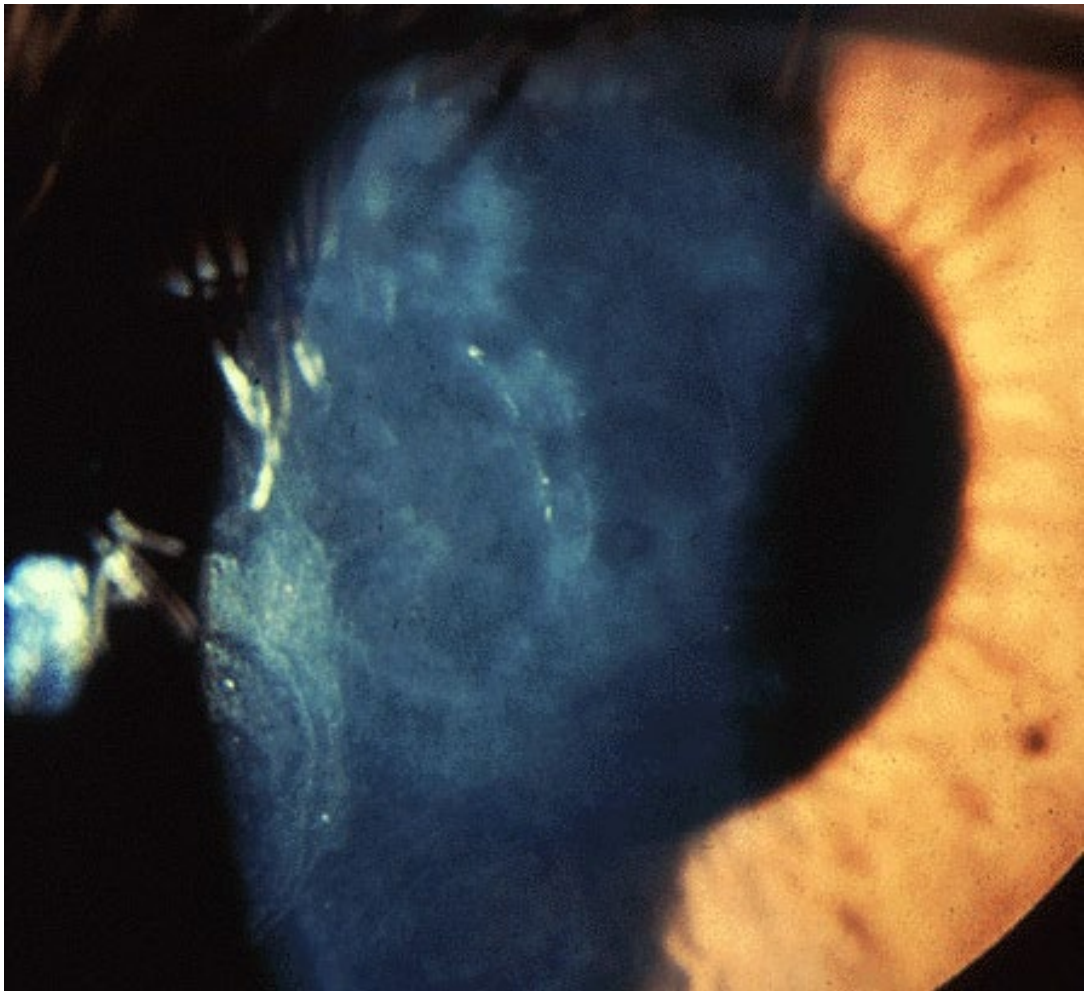
Corneal Dystrophies



Posterior polymorphous corneal dystrophy: *snail* or *railroad tracks*

Corneal Dystrophies



Posterior polymorphous corneal dystrophy. The clinical appearance of PPMD is highly variable.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does PPMD begin to manifest?
Usually teens or 20s; rarely in childhood

How does it present? What is seen at the slit lamp?
Endothelial-surface abnormalities, including:
--**Vesicular lesions** , either isolated or in groups
--Linear gray **band-shaped** opacities, often with edges described as '**snail** or **railroad** tracks'
--Diffuse '**geographic**' opacities that often have an overlying haze in the posterior stroma

Which lesion is most common in PPMD?

Endothelial Dystrophies

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Are the vesicles actually vesicular; ie, are they fluid-filled?

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No, they contain a collagenous material

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And now, a sidebar seemingly from out of left field: *What are* Haab striae*?*

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What do Haab striae look like?
Parallel horizontal lines on the posterior surface of the cornea

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Because unlike Haab striae, which taper and meet at their ends, the parallel lines in PPMD do **not** meet at their terminuses

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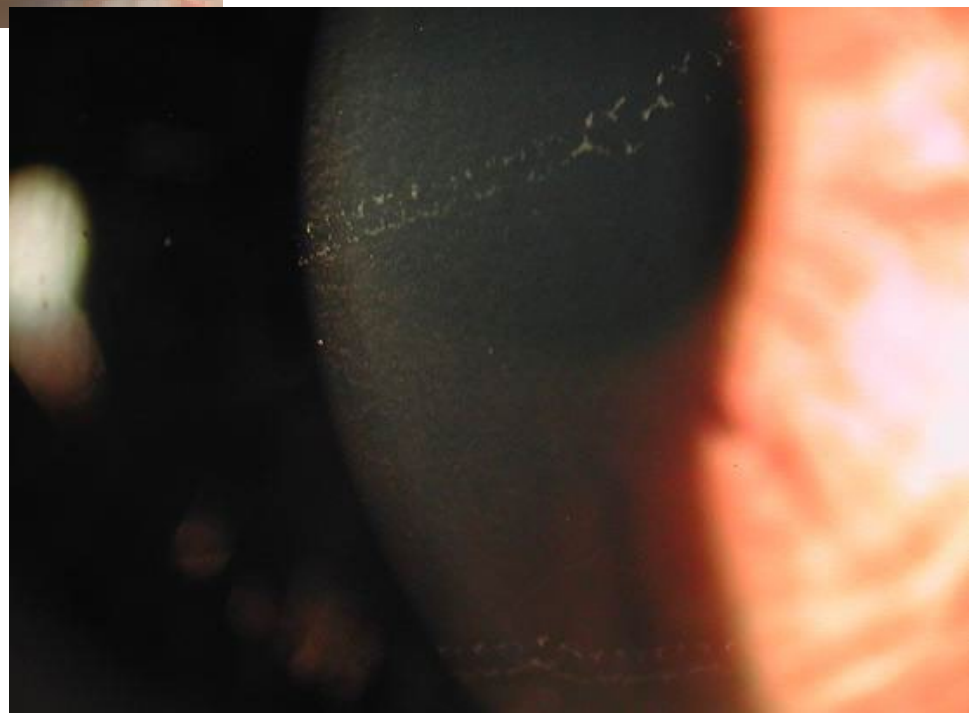
Haab's striae (note that the lines taper and meet)

Snail tracks in PPMD
(the lines don't meet)



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In the majority of pts, PPMD is a stable, painless and visually insignificant condition.

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The presence of multiple cell layers on the endothelial surface

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In addition to being multilayered, what else is abnormal about the endothelial cells in PPMD?

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Is it painful? Does it affect vision?

In addition to being multilayered, what else is abnormal about the endothelial cells in PPMD?
In PPMD, the endothelial cells 'behave' like epithelial cells and/or fibroblasts; ie, they **proliferate**, they **form multiple layers**, they **migrate**

What is the histologic hallmark of PPMD on light microscopy?
The presence of **multiple cell layers on the endothelial surface**

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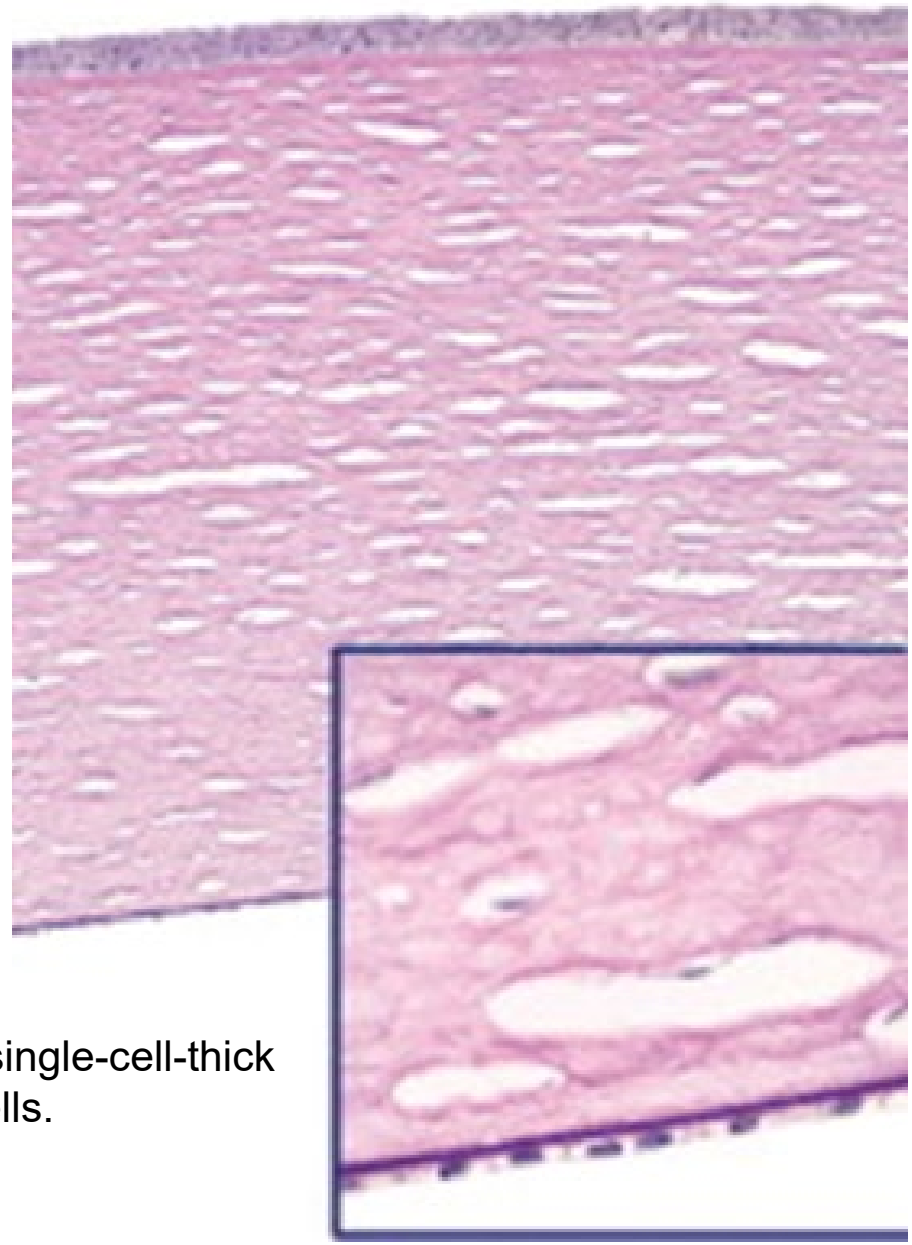
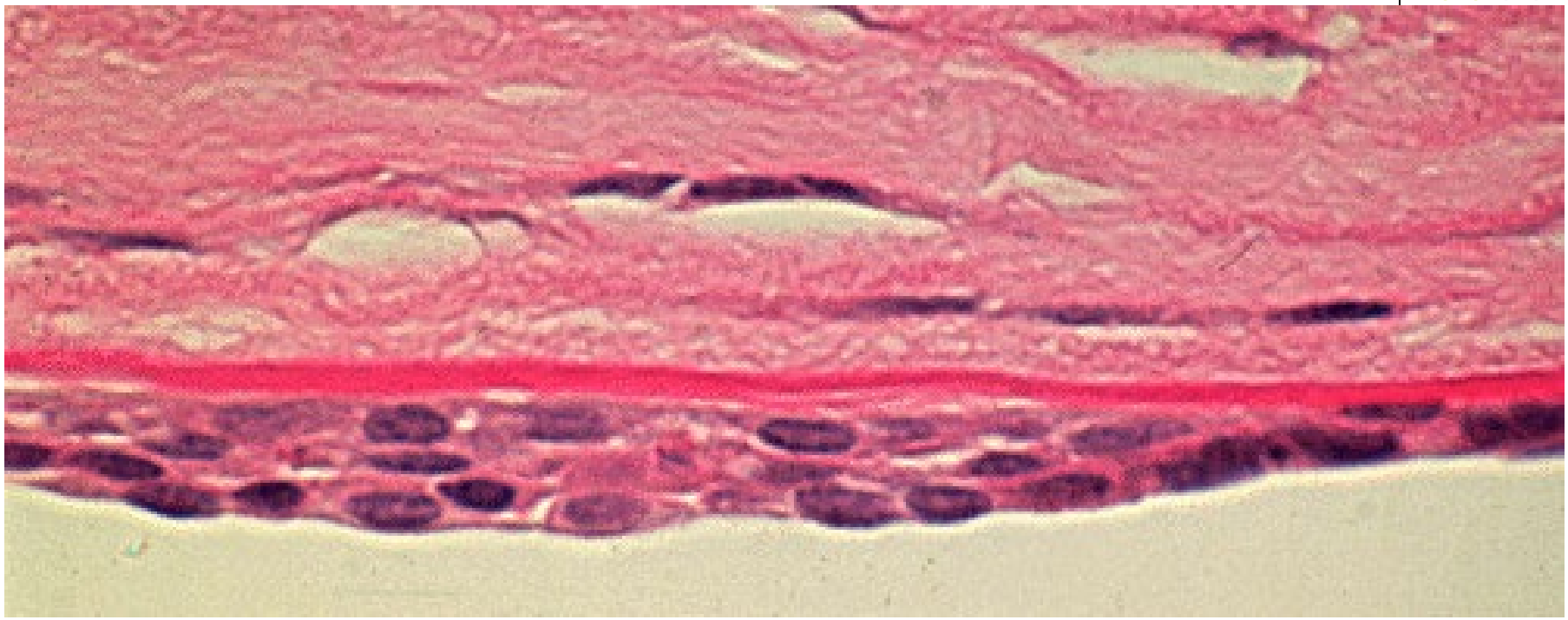
Corneal Dystrophies



Normal cornea. Note the single-cell-thick nature of the endothelial cells.

Corneal Dystrophies



Posterior polymorphous corneal dystrophy. Instead of being lined by cells with the attributes of corneal endothelium, the posterior cornea is covered by cells with **epithelial-** or **fibroblast-like** features.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

What condition manifests findings that render it easily confusable with PPMD?

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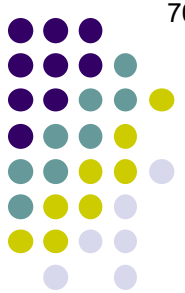

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--Unlike PPMD, ICE is always [inheritance]

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Speaking of ICE…PPMD can involve several ICE-like lesions that are not found in other corneal dystrophies. What are they?
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--Peripheral anterior synechiae
--Glaucoma
--Iris changes (eg, corectopia)

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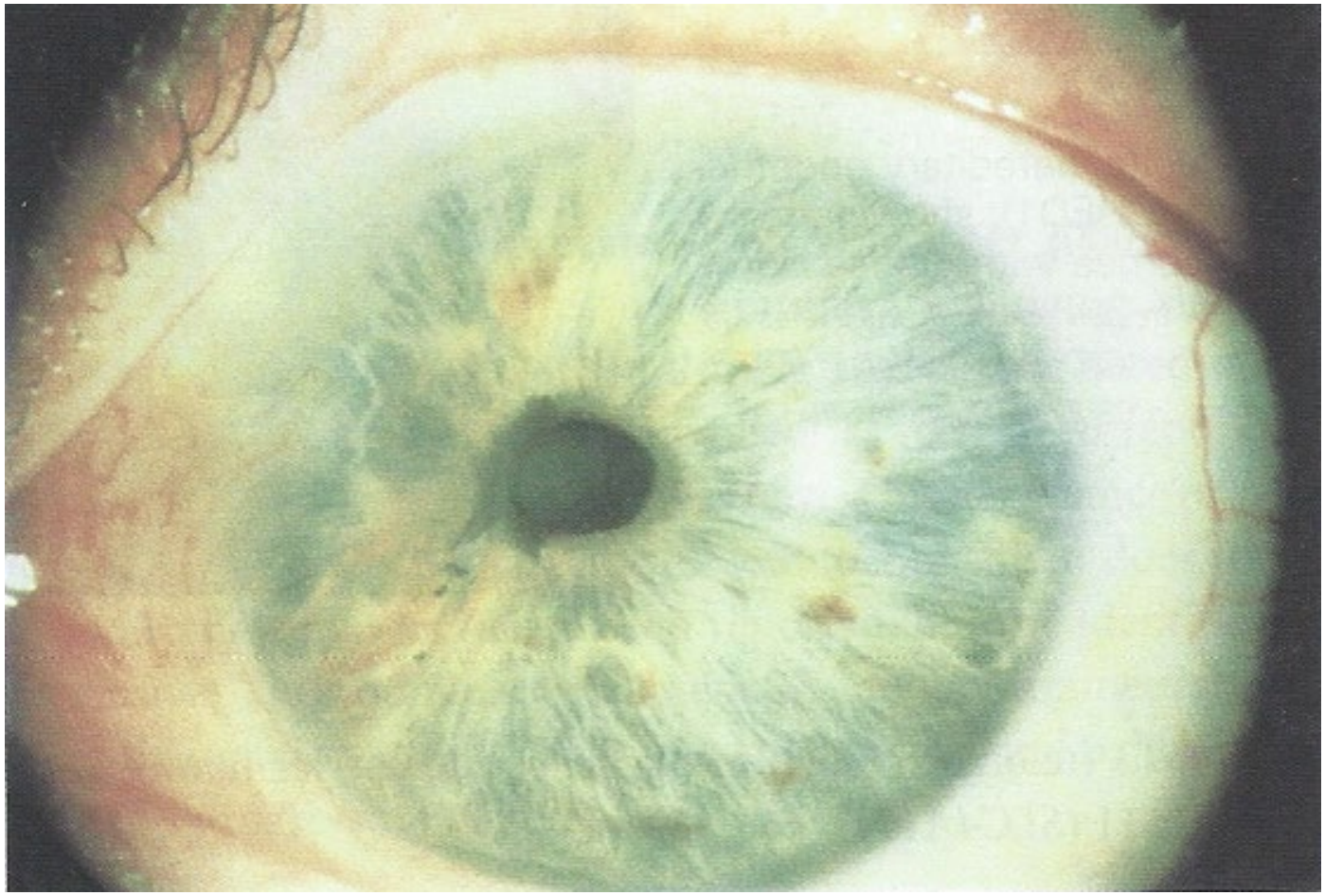
Corneal Dystrophies



Posterior polymorphous corneal dystrophy. Iridocorneal adhesion and corectopia. This presentation of PPMD could easily be mistaken for ICE.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

How is CHED inherited?

Endothelial Dystrophies

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Unlike most dystrophies, it usually isn't

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The corneas are diffusely cloudy (described as two words or one word)

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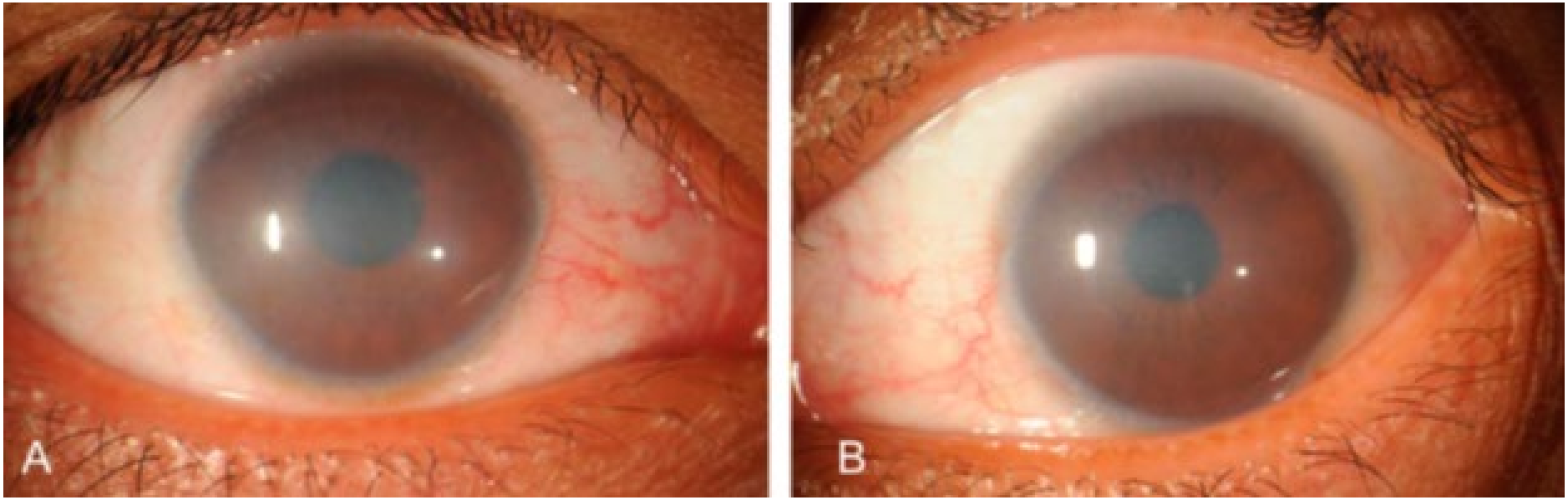

Congenital hereditary endothelial dystrophy. Right and left corneas of a 30-year-old woman with CHED.

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Is it progressive?
Unlike most dystrophies, it usually isn't

What is seen at the slit lamp?
The corneas are diffusely cloudy (desc

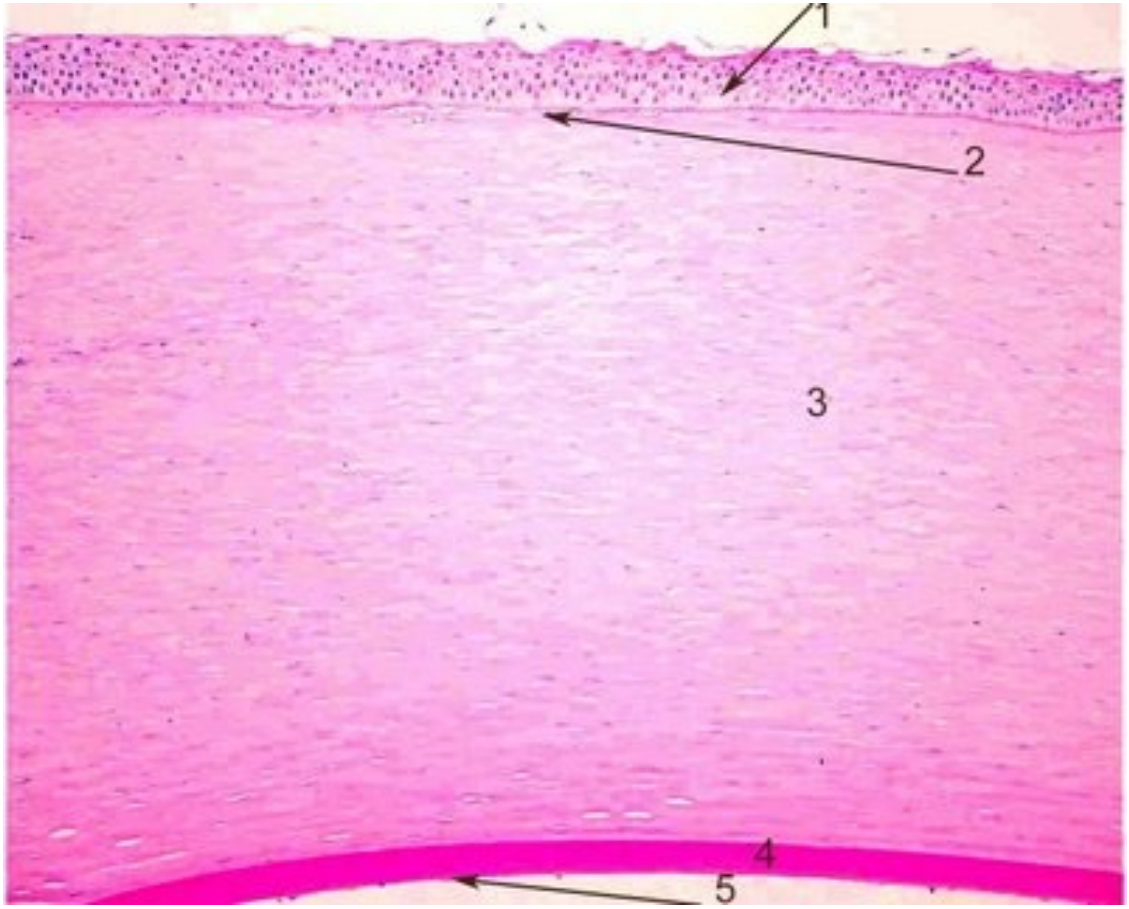
How thickened?
Like, 2-3 times normal

thickened

Endothelial Dystrophies

1) Fuchs endothelial corneal dystrophy
2) Posterior polymorphous corneal dystrophy
3) **Congenital hereditary endothelial dystrophy**

Corneal Dystrophies



Congenital hereditary endothelial dystrophy. Get a load of how thicc this cornea is!

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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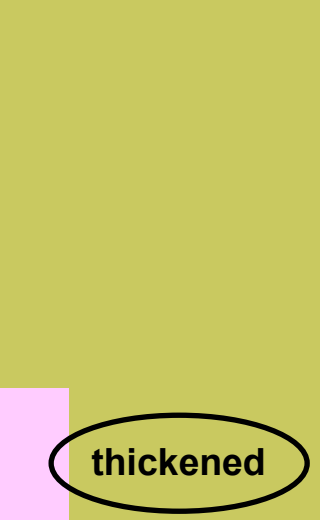

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Descemet's is thinned vs thickened

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Is it painful?
No

What is the histologic hallmark of CHED on light microscopy?
Descemet's is thickened, and the number of endothelial cells is substantially more *vs* less than normal

Endothelial Dystrophies

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Descemet's is thickened , and the number of endothelial cells is substantially less than normal

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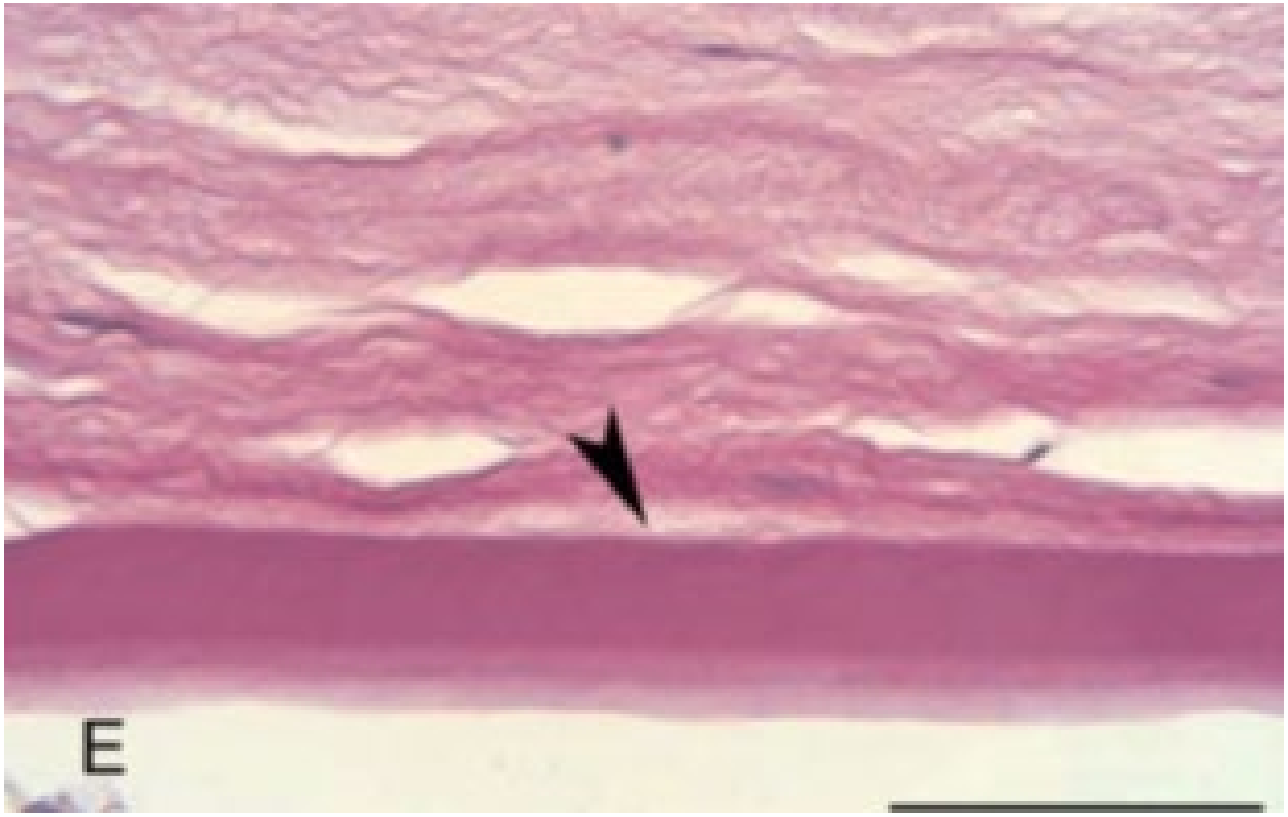

Congenital hereditary endothelial dystrophy. Thickened Descemet membrane with no visible endothelial cells.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Is it painful?
No

The few endothelial cells that are present—are they normal?

What is the histologic hallmark of CHED on light microscopy?
Descemet's is thickened, and the number of endothelial cells is substantially less than normal

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Is it painful?
No

The few endothelial cells that are present—are they normal?
No, most if not all are atrophic

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Descemet's is thickened, and the number of endothelial cells is substantially less than normal

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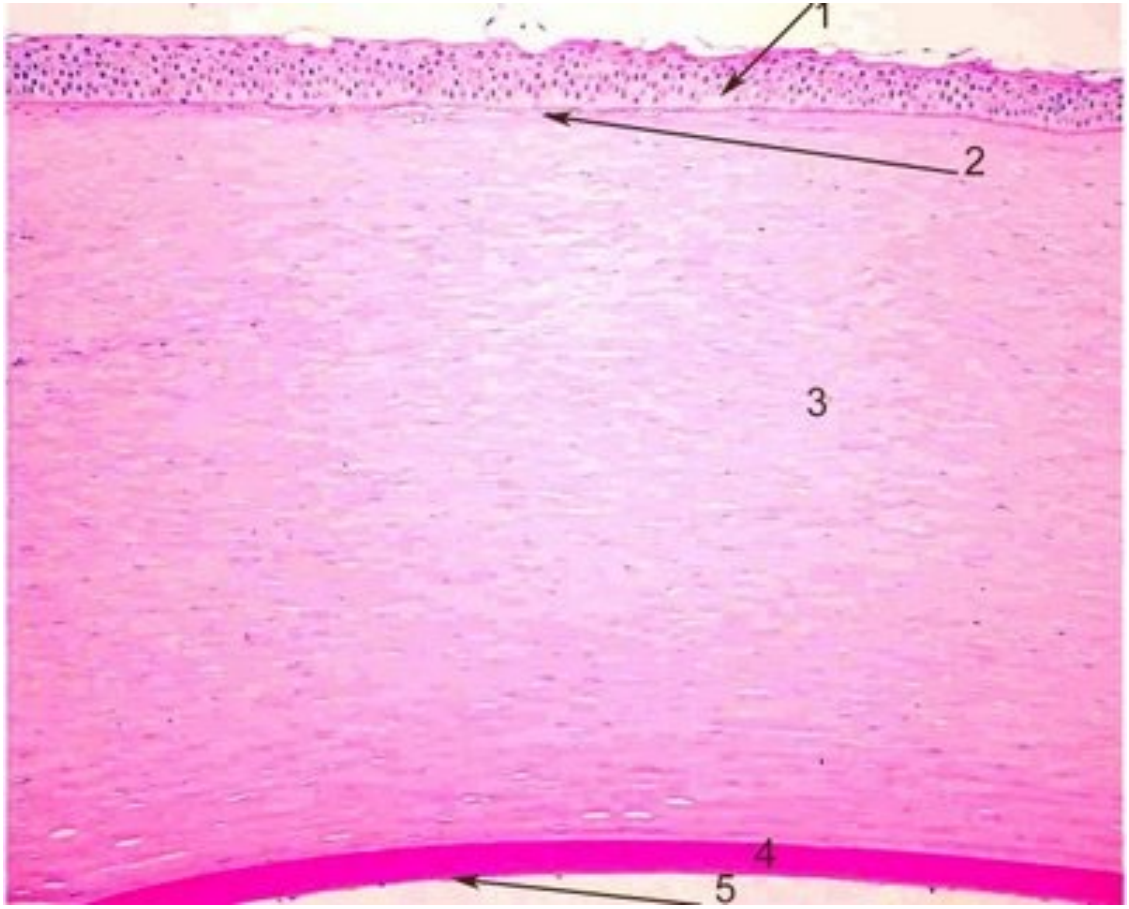

Congenital hereditary endothelial dystrophy. Same pic shown previously. This time, take note of the thickened Descemet's (4) and the paucity of endothelial cells (5)

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

How is CHED inherited?
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We have now 'gotten to' the reason CHED corneas are thickened:
The paucity of functioning endothelial cells

thickened

Is vision affected?
Yes, significantly enough that a sensor

Why is the cornea thickened in CHED?
We'll get to that shortly

Is it painful?
No

What is the histologic hallmark of CHED on light micr copy?
Descemet's is thickened, and the number of endothelial cells is substantially less than normal

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We have now 'gotten to' the reason CHED corneas are thickened:
The paucity of functioning endothelial cells

How does a lack of endothelial cells lead to a thicker cornea?

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Is it painful?
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The paucity of functioning endothelial cells

How does a lack of endothelial cells lead to a thicker cornea?
Recall that one of the main functions of the endothelium is maintaining a proper level of corneal [redacted].

thickened

Is vision affected?
Yes, significantly enough that a sensor

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We'll get to that shortly

Is it painful?
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The paucity of functioning endothelial cells

How does a lack of endothelial cells lead to a thicker cornea?
Recall that one of the main functions of the endothelium is maintaining a proper level of corneal hydration . The endo accomplishes this by 1) acting as a barrier preventing aqueous ingress, and 2) actively [redacted] the cornea via Na+/K+/ATPase pumps.

thickened

Why is the cornea thickened in CHED?
We'll get to that shortly

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What is the average endothelial count (cells/mm^2) of the human cornea?

We have now
The paucity o

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Recall that one c
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Indeed it does—it decreases with advancing age, from about 3400 at age 15 to about 2400 at age 85

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How low does the endo count have to get for corneal edema to set in?

We have now
The paucity o

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thickened

Why is the cornea thickened in CHED?
We'll get to that shortly

Is vision affected?
Yes, significantly enough that a sensor

Is it painful?
No

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thickened

What is the average endothelial count (cells/mm^2) of the human cornea?
Estimates vary among the *BCSC* books; for adults, 3000 is a reasonable value

'For adults' implies that endo count varies with age—does it?
Indeed it does—it decreases with advancing age, from about 3400 at age 15 to about 2400 at age 85

How low does the endo count have to get for corneal edema to set in?
Around 500 or so

Why is the cornea thickened in CHED?
We'll get to that shortly

Endothelial Dystrophies

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cornea via Na+/K+/ATPase pumps. When the endo count is too low to accomplish these functions, the cornea swells, which in turn thickens it.

What is the average endothelial count (cells/mm^2) of the human cornea?
Estimates vary among the *BCSC* books; for adults, 3000 is a reasonable value

'For adults' implies that endo count varies with age—does it?
Indeed it does—it decreases with advancing age, from about 3400 at age 15

Can CHED be treated?

a to set in?

thickened

Why is the cornea thickened in CHED?
We'll get to that shortly

Is vision affected?
Yes, significantly enough that a sensor

Is it painful?
No

What is the histologic hallmark of CHED on light micr copy?
Descemet's is thickened, and the number of endothelial cells is substantially less than normal

Endothelial Dystrophies

1) Fuchs endothelial corneal dystrophy
2) Posterior polymorphous corneal dystrophy
3) **Congenital hereditary endothelial dystrophy**

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

How is CHED inher
AR

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B

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Th

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The paucity o

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Recall that c
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cornea via Na+/K+/ATPase pumps. When the endo count is too low to accomplish these functions, the cornea swells, which in turn thickens it.

a to set in?

Can CHED be treated?
Severe cases go to transplantation—either full thickness (PK) or endothelial (eg, DSAEK)

thickened

Is vision affected?
Yes, significantly enough that a sensor

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We'll get to that shortly

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

How is CHED inherited?
AR

At what age does it manifest?
Birth

Is it progressive?
Unlike most dystrophies, it usually is

What is seen at the slit lamp?
The **corneas are diffusely cloudy** ed

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What is the histologic hallmark of CHED on light microscopy?
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The scenario of an infant with cloudy corneas should immediately bring to mind a mnemonic. Which one?

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What are the elements in the STUMPED mnemonic for cloudy corneas in an infant?

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Coming in hot…

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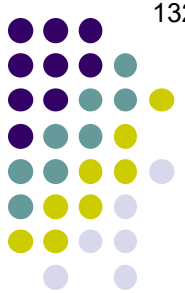
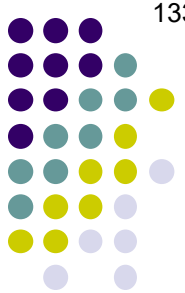
Corneal Dystrophies



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

Fill in the entities embedded in the mnemonic
(Note: There are two *Ss* and two *Es*)

Corneal Dystrophies



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

(ie, two words)

Fill in the entities embedded in the mnemonic
(Note: There are two *Ss* and two *Es*)

Corneal Dystrophies

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- **E**ndothelial dystrophy (CHED); **E**levated IOP
  (ie, congenital glaucoma)
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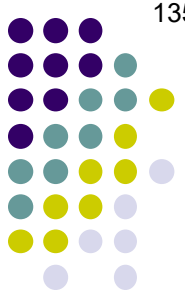
Corneal Dystrophies



- **S**clerocornea; **S**tromal dystrophy (CHSD)
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- *Next we will touch on distinguishing among CHSD, CHED and primary congenital glaucoma by highlighting key differences in their presentations*
- **P**eters anomaly
- **E**ndothelial dystrophy (CHED); **E**levated IOP (ie, congenital glaucoma)
- **D**ermoid of the cornea

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	*?*			
CHSD	*?*			
Primary congenital glaucoma	*?*			

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased			
CHSD	**Mildly** increased			
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)			

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased	As just discussed, the CCT is dramatically increased in CHED because of edema 2ndry to lack of adequate endothelial barrier and deturgescence function		
CHSD	**Mildly** increased			
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)			

(No question—advance when ready)

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

	CCT	***Corneal diameter***	***IOP***	***Tearing/ Photophobia?***
CHED	**Markedly** increased			
CHSD	**Mildly** increased	In CH***S***D, the cornea is modestly thickened by the presence of the material that causes the cloudiness		
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)			

(No question—advance when ready)

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased			
CHSD	**Mildly** increased			
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	In congenital glaucoma, corneal thickness depends upon 1) whether the endo is healthy and 2) how high the IOP is		

(No question—advance when ready)

Corneal Dystrophies

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	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased	WNL		
CHSD	**Mildly** increased	WNL		
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	Increased		

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

CHED				
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	**Increased**		

Why is corneal diameter increased in congenital glaucoma?

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

CHED				
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	**Increased**		

Why is corneal diameter increased in congenital glaucoma?
Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall

Corneal Dystrophies

Congenital glaucoma: Increased corneal diameter

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

CHED				
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	**Increased**		

Why is corneal diameter increased in congenital glaucoma?
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What is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

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Buphthalmos

Corneal Dystrophies

Congenital glaucoma: Buphthalmos OD

Corneal Dystrophies

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CHED				
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	**Increased**		

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Buphthalmos

What does buphthalmos *translate to in English?*

Corneal Dystrophies

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'Ox's eye'

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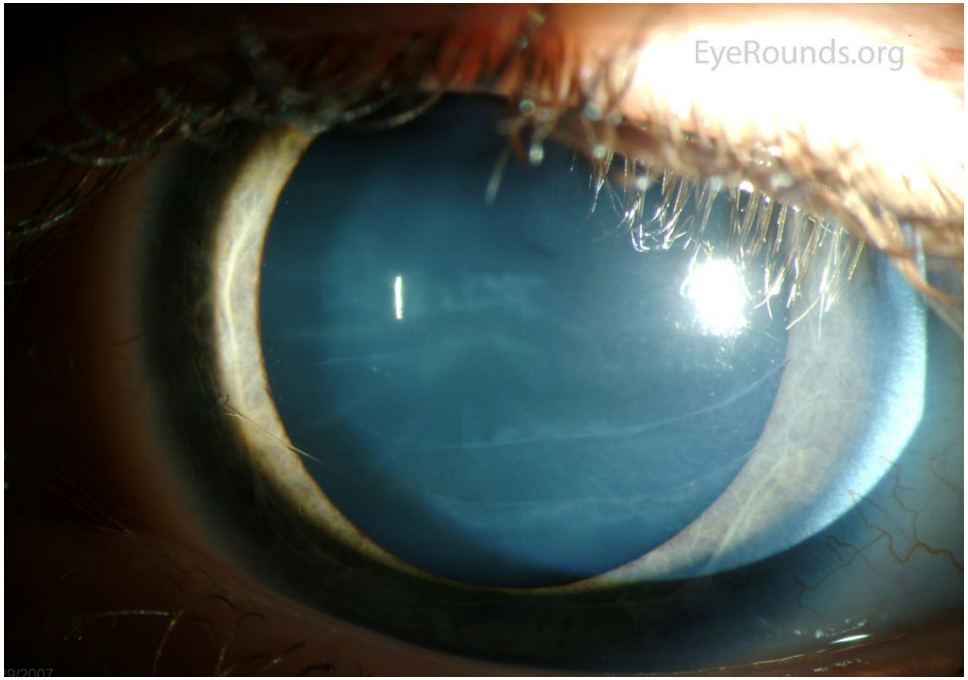
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	**Increased**		

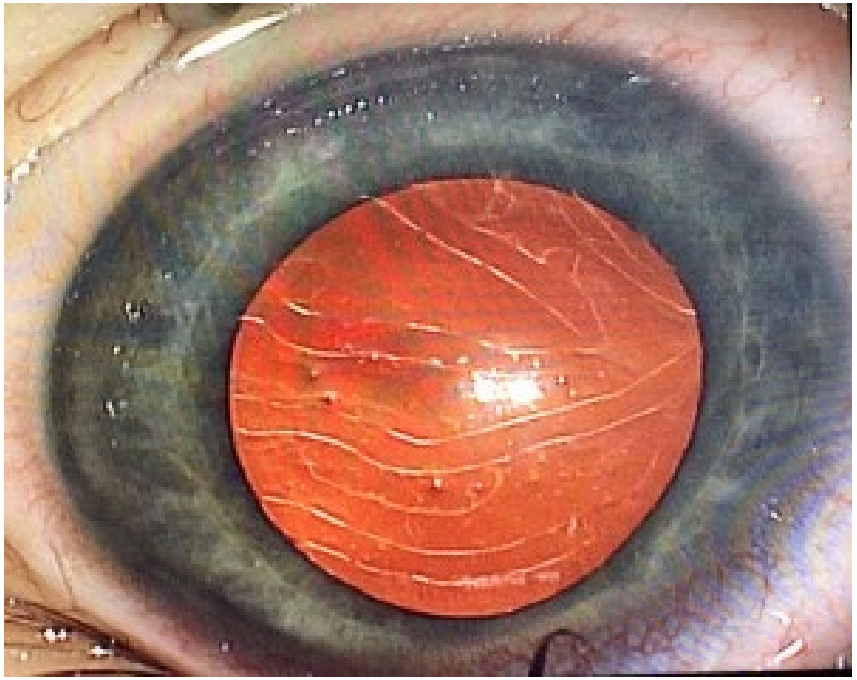
Why is corneal diameter increased in congenital glaucoma?
Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall

What does buphthalmos *translate to in English?*
'Ox's eye'

And as mentioned earlier in the slide-set, the high IOP in congenital glaucoma causes *Haab's striae*—horizontal breaks in Descemet's and the overlying endothelium

Corneal Dystrophies





Horizontal Descemet's breaks (*Haab's striae*) in congenital glaucoma

Corneal Dystrophies

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	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased	WNL	***?***	
CHSD	**Mildly** increased	WNL	***?***	
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	Increased	***?***	

Corneal Dystrophies

CCT, *corneal diameter, IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD,* and *primary congenital glaucoma*. Fill in the blanks below.

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CHED	**Markedly** increased	WNL	WNL	
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Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	Increased	Duh	

Corneal Dystrophies

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Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	Increased	Duh	?

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	CCT	*Corneal diameter*	*IOP*	*Tearing/ Photophobia?*
CHED	**Markedly** increased	WNL	WNL	No
CHSD	**Mildly** increased	WNL	WNL	No
Primary congenital glaucoma	**Variably** increased (or WNL, or thin)	Increased	Duh	Yes