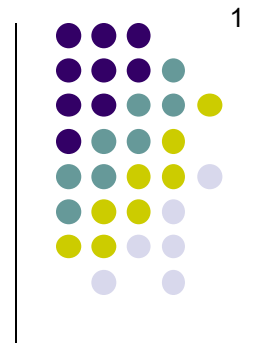


# Corneal Dystrophies



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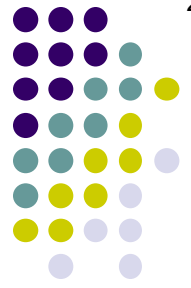
?

*What are the four categories of corneal dystrophies?*

?

?

# 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

*What are the three endothelial dystrophies?*

Stromal Dystrophies

**Endothelial Dystrophies**

- 1) ?
- 2) ?
- 3) ?

# Corneal Dystrophies

## Epithelial and Subepithelial Dystrophies

## Epithelial-Stromal *TGFBI* Dystrophies

## Stromal 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.)

## Endothelial Dystrophies

- 1) **Fuchs endothelial corneal dystrophy**
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# 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.)

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

## Endothelial Dystrophies

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# 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.)

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

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

## Endothelial Dystrophies

- 1) **Fuchs endothelial corneal dystrophy**
- 2) Posterior polymorphous corneal dystrophy
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# Corneal Dystrophies



## Epithelial and Subepithelial Dystrophies

*At what age does FECD begin to manifest?*

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## Endothelial Dystrophies

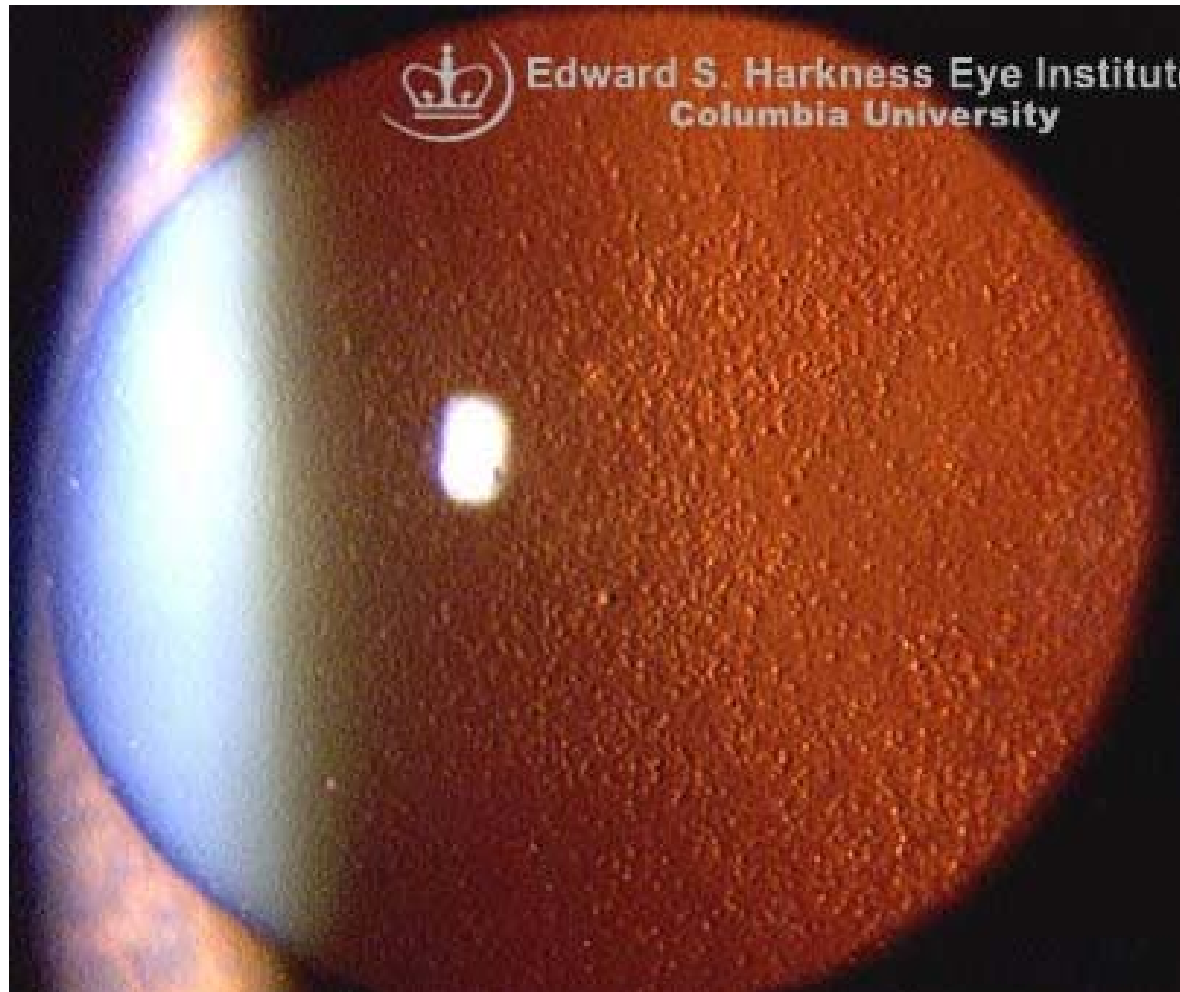
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## Corneal Dystrophies



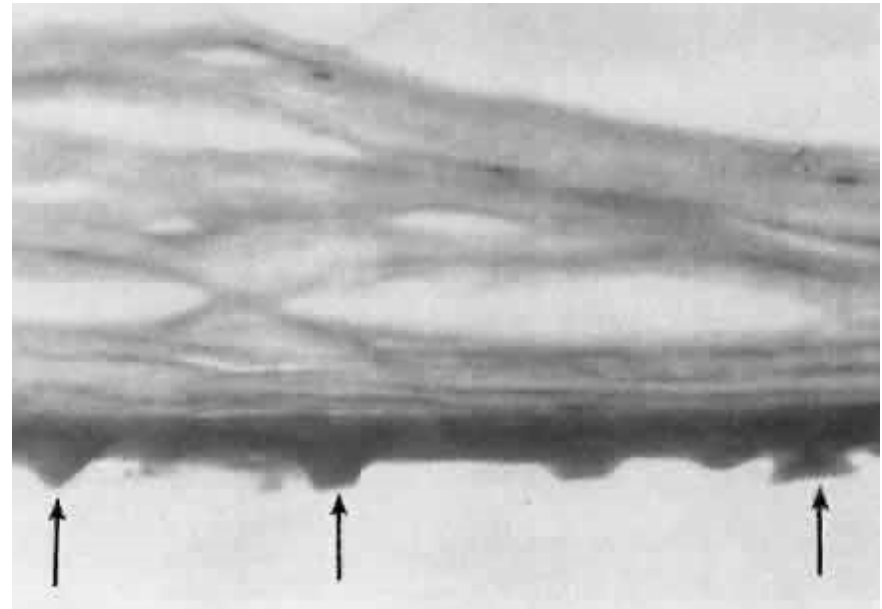
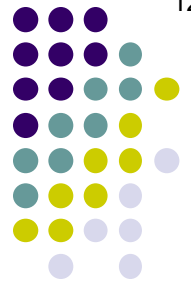
**Fuchs endothelial corneal dystrophy. Cornea guttata, slit-lamp view**

## 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: ).

### Endothelial Dystrophies

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## Corneal Dystrophies

### Epithelial and Subepithelial Dystrophies

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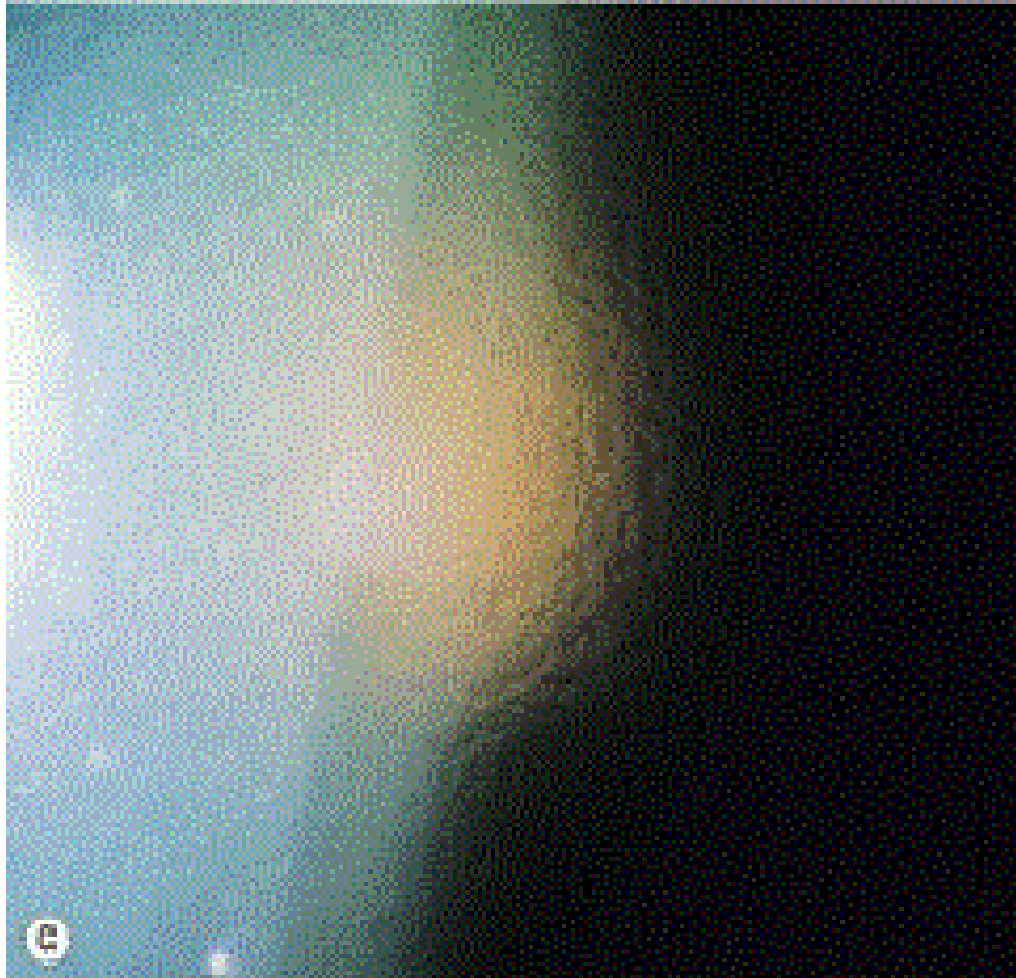
*How does it present? What is seen at the slit lamp?*

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## Corneal Dystrophies



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





## Corneal Dystrophies

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### Endothelial Dystrophies

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## Corneal Dystrophies

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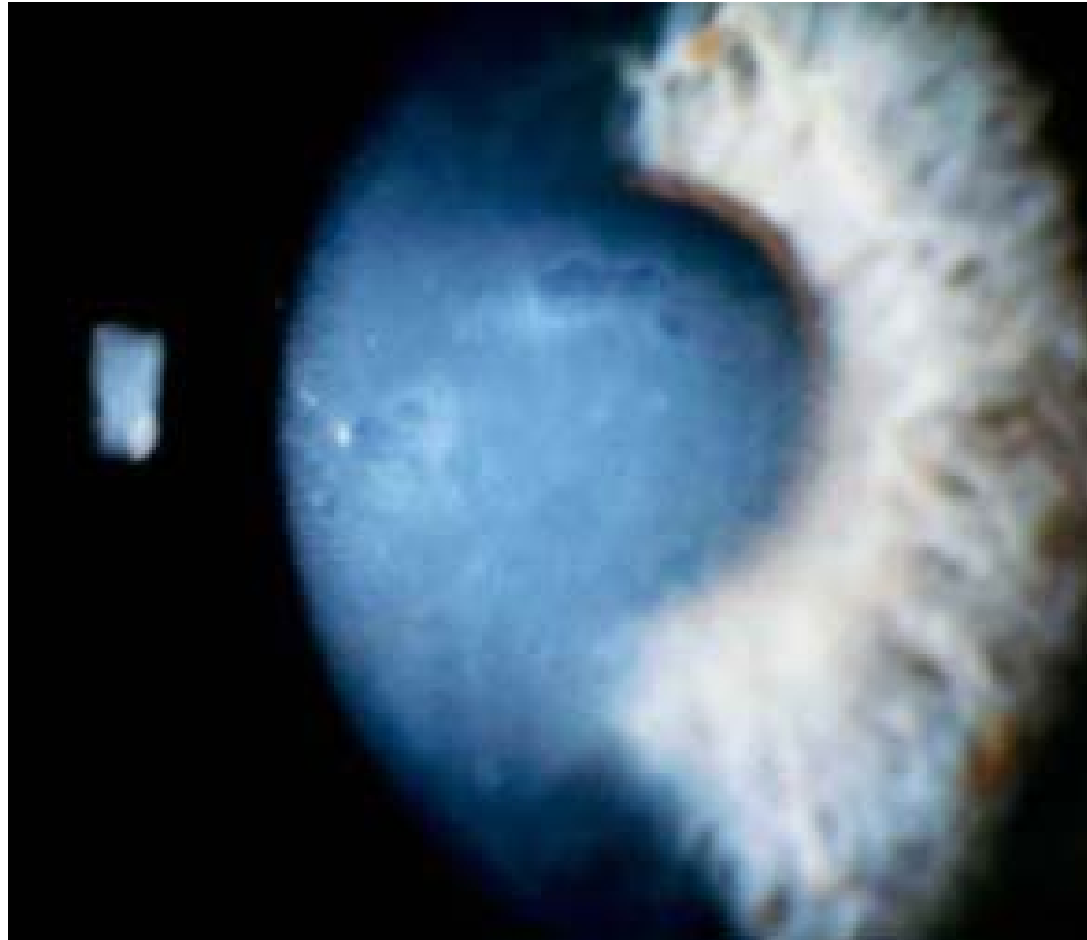
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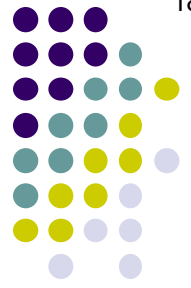
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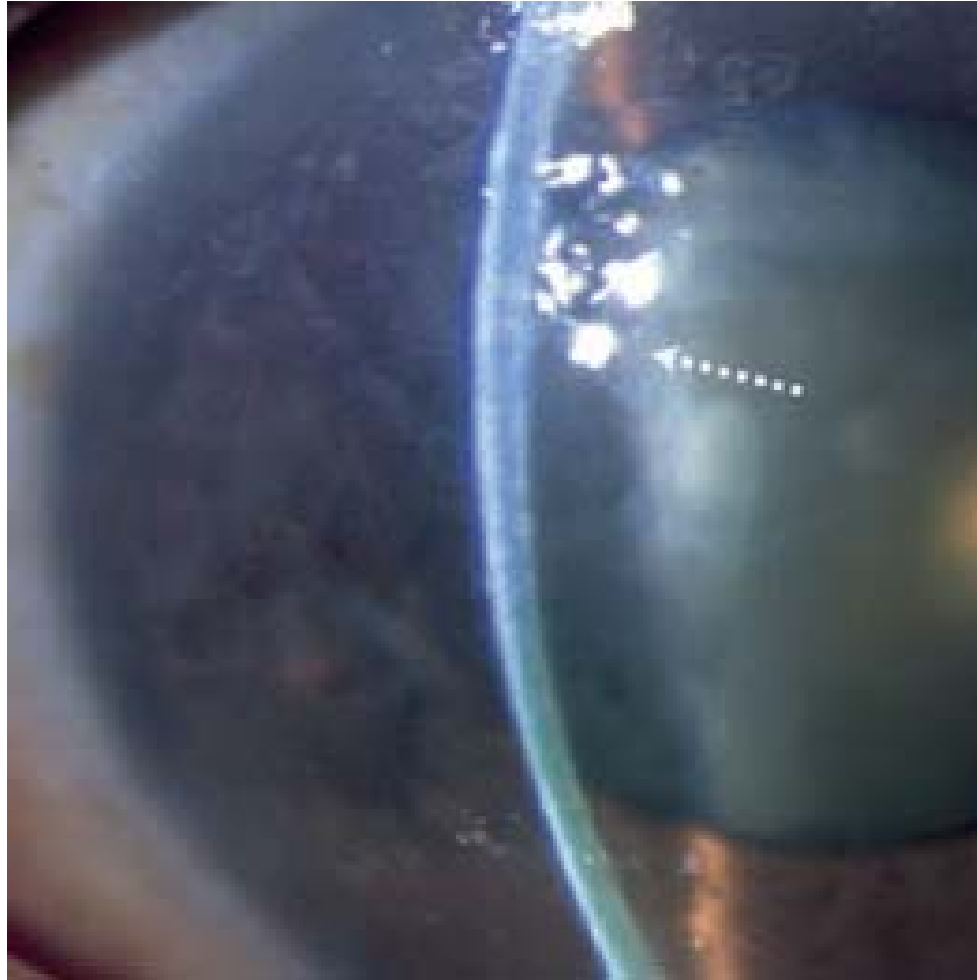
## Corneal Dystrophies



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



## Corneal Dystrophies



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





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*Is it painful?*

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*Is it painful?*

If/when epithelial bullae rupture, yes

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*Is it painful?*

If/when epithelial bullae rupture, yes

*Does it affect vision?*

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*Is it painful?*

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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## Corneal Dystrophies

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*Is it painful?*

If/when epithelial bullae rupture, yes

*Does it affect vision?*

Stromal/epithelial edema causes decreased vision, which is **usually worse in the morning**

*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

### Endothelial Dystrophies

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## Corneal Dystrophies

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*Is it painful?*

If/when epithelial bullae rupture, yes

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*What is the histologic hallmark of FECD on confocal microscopy?*

1)

2)

### Endothelial Dystrophies

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## Corneal Dystrophies

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If/when epithelial bullae rupture, yes

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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  ) and an increase in the cell-to-cell variability in size (known as  )

2) The presence of guttata

### Endothelial Dystrophies

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## Corneal Dystrophies

### Epithelial and Subepithelial Dystrophies

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If/when epithelial bullae rupture, yes

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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 **polymegathism**) and an increase in the cell-to-cell variability in size (known as **pleomorphism**)

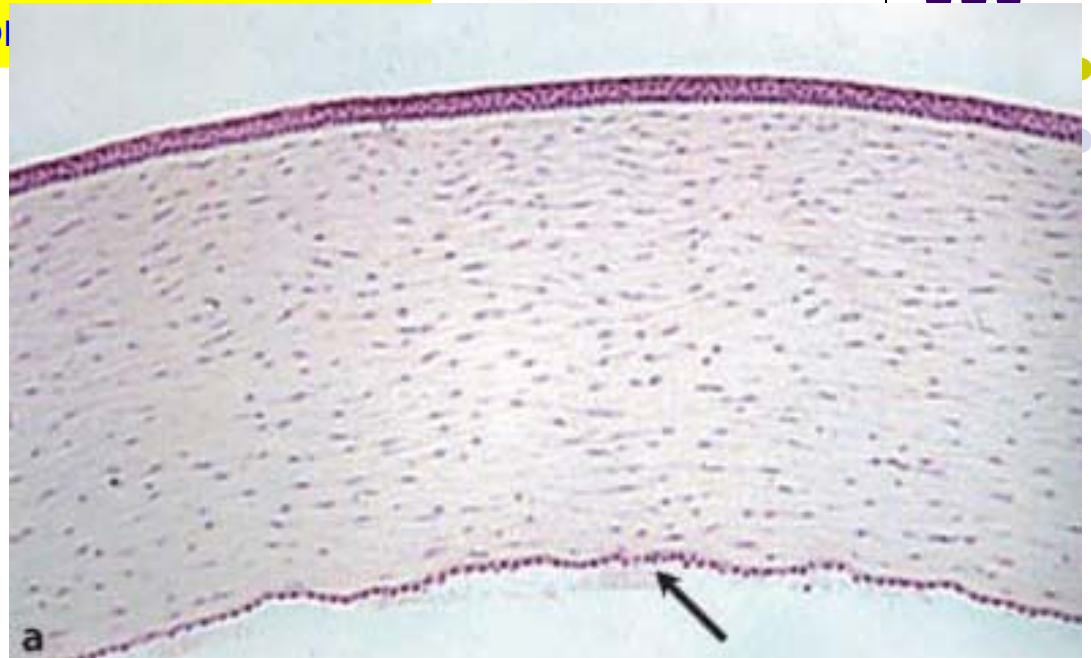
2) The presence of guttata

### Endothelial Dystrophies

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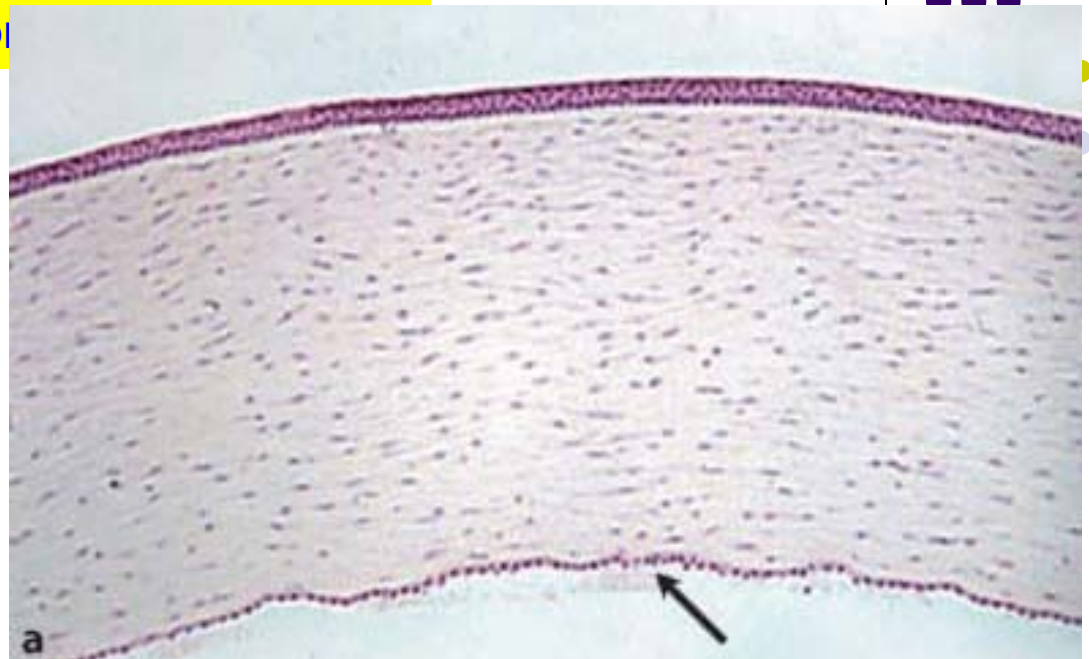
Cor

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

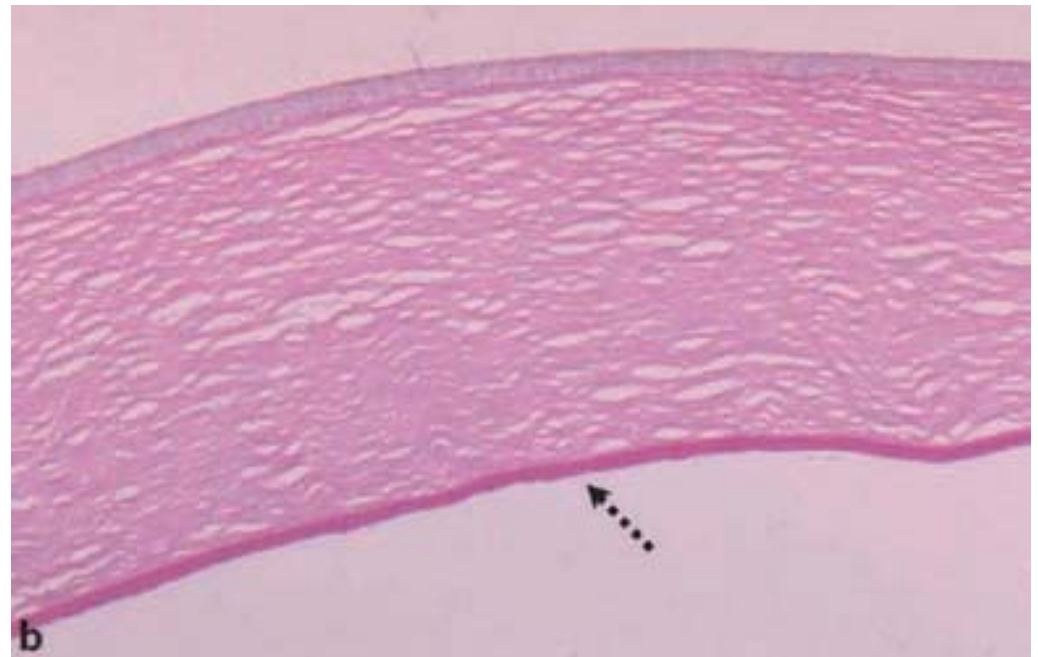


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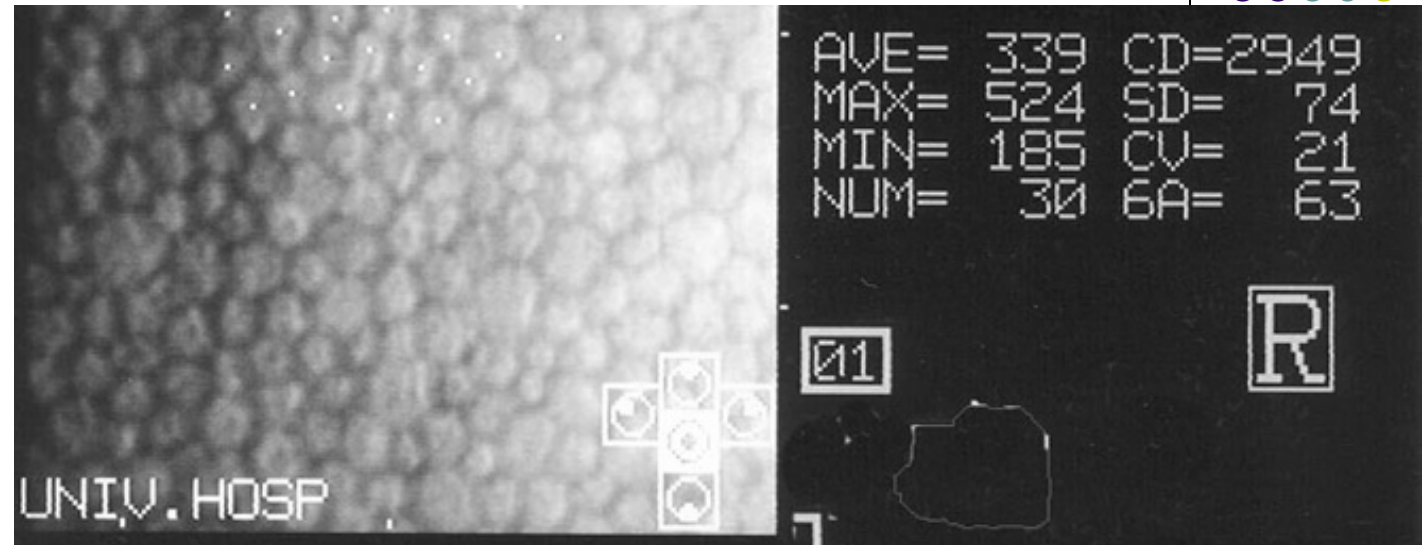
**Fuchs endothelial corneal dystrophy.** Light microscopy section of FED cornea. Note the markedly thickened Descemet's membrane and the absence of endothelial cell nuclei on the posterior surface (*dashed arrow*).





## Corneal Dystrophies

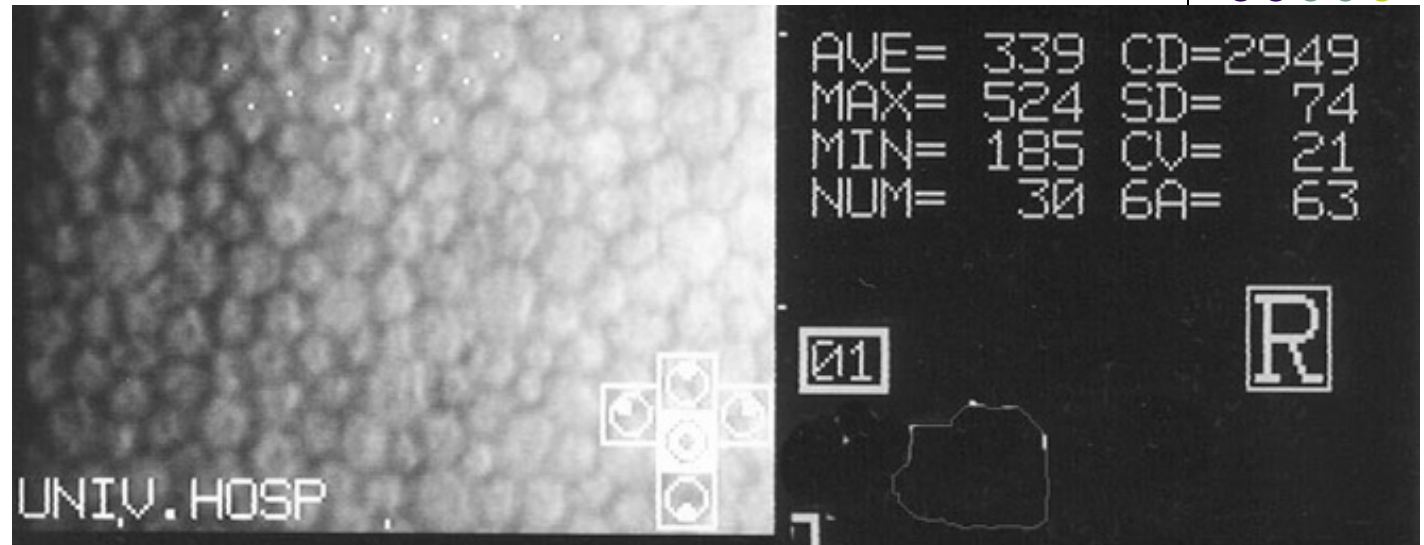
Specular microscopic image, **normal cornea**. Note the plethora of polygonal cells of uniform size, and the absence of empty spaces. The cell density is 2949/mm<sup>2</sup> (nl 2-3K, avg ~2400)



## 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<sup>2</sup> (nl 2-3K, avg ~2400)



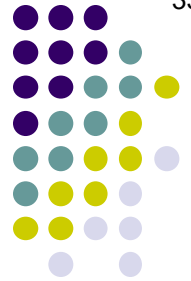
Specular microscopic image, **Fuchs**. Note the polymegathism and polymorphism, and the empty spaces (= guttata). The cell density is 1763/mm<sup>2</sup>.





# 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

## Endothelial Dystrophies

- 1) Fuchs endothelial corneal dystrophy
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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?*

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# Corneal Dystrophies

## Epithelial and Subepithelial Dystrophies

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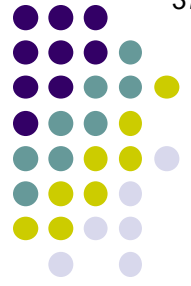
*How does it present? What is seen at the slit lamp?*

Endothelial-surface abnormalities, including:

- , either isolated or in groups
- Linear gray  opacities, often with edges described as  or  tracks'
- Diffuse  opacities that often have an overlying haze in the posterior stroma

## Endothelial Dystrophies

- 1) Fuchs endothelial corneal dystrophy
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# Corneal Dystrophies

## Epithelial and Subepithelial Dystrophies

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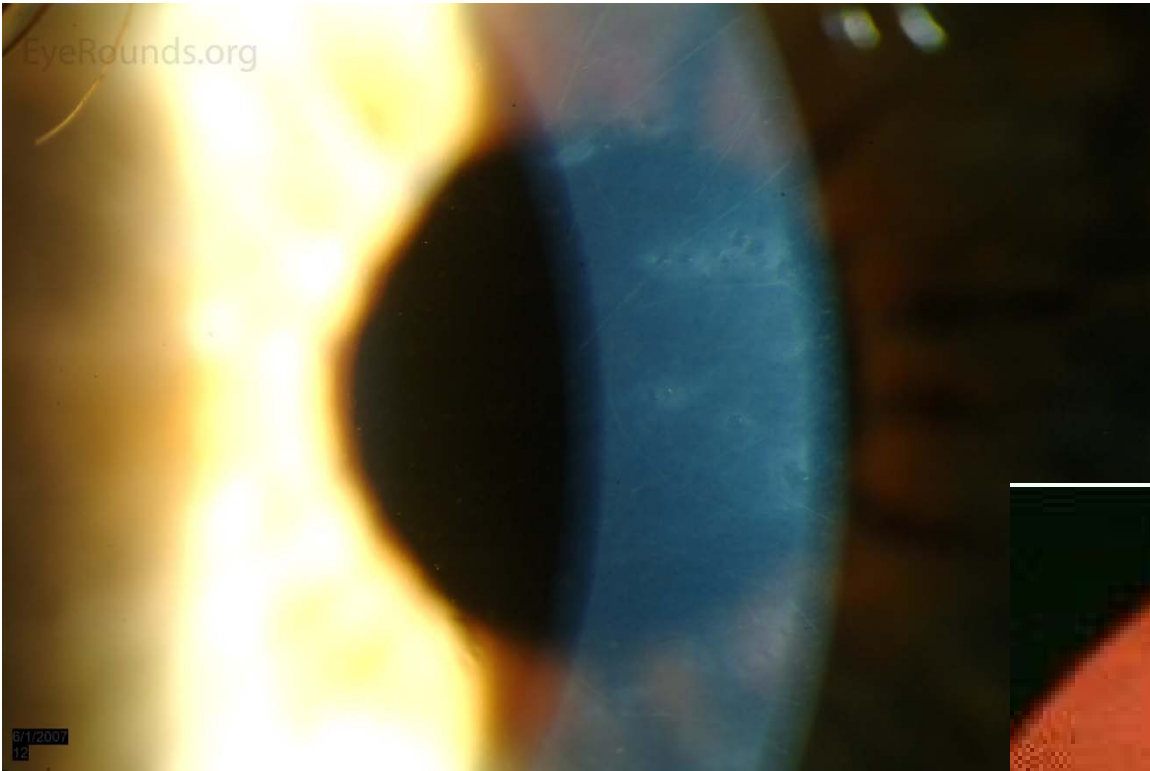
*How does it present? What is seen at the slit lamp?*

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

## Endothelial Dystrophies

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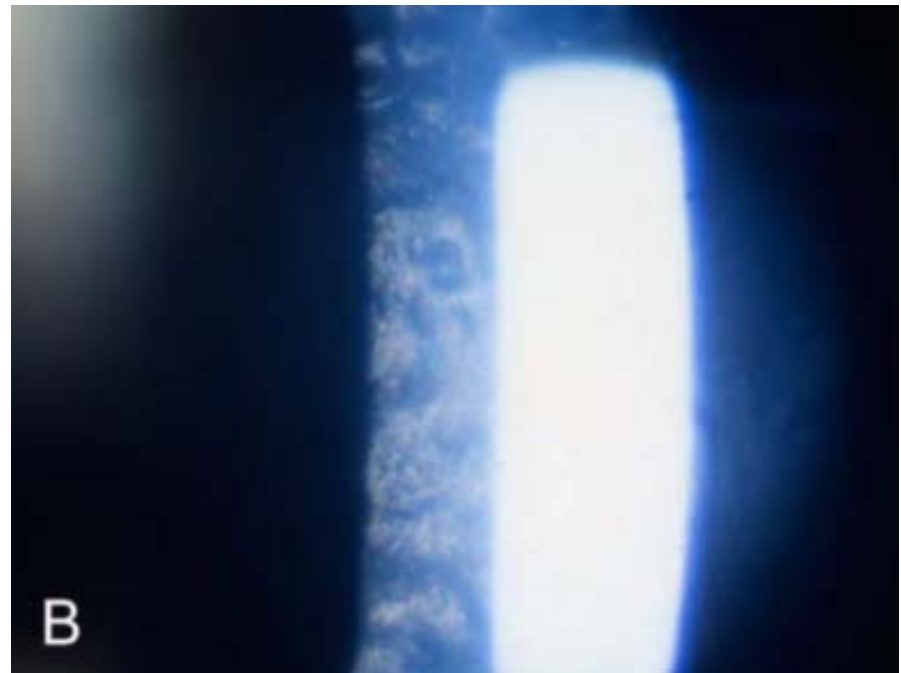
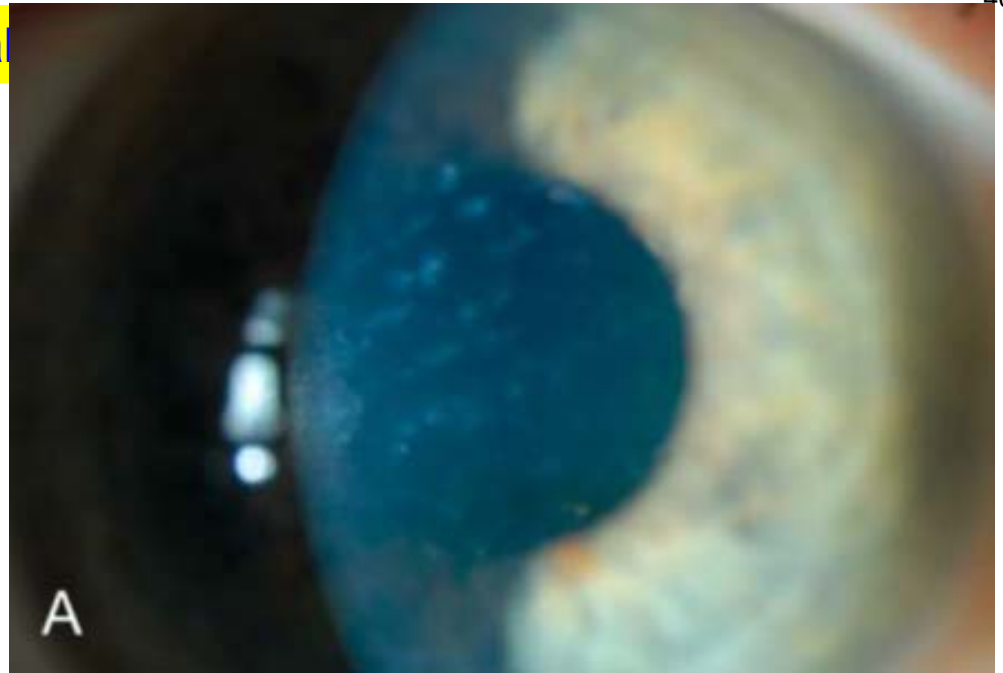
**Posterior polymorphous corneal dystrophy:**  
Vesicular lesions



## Corneal Dystrophies

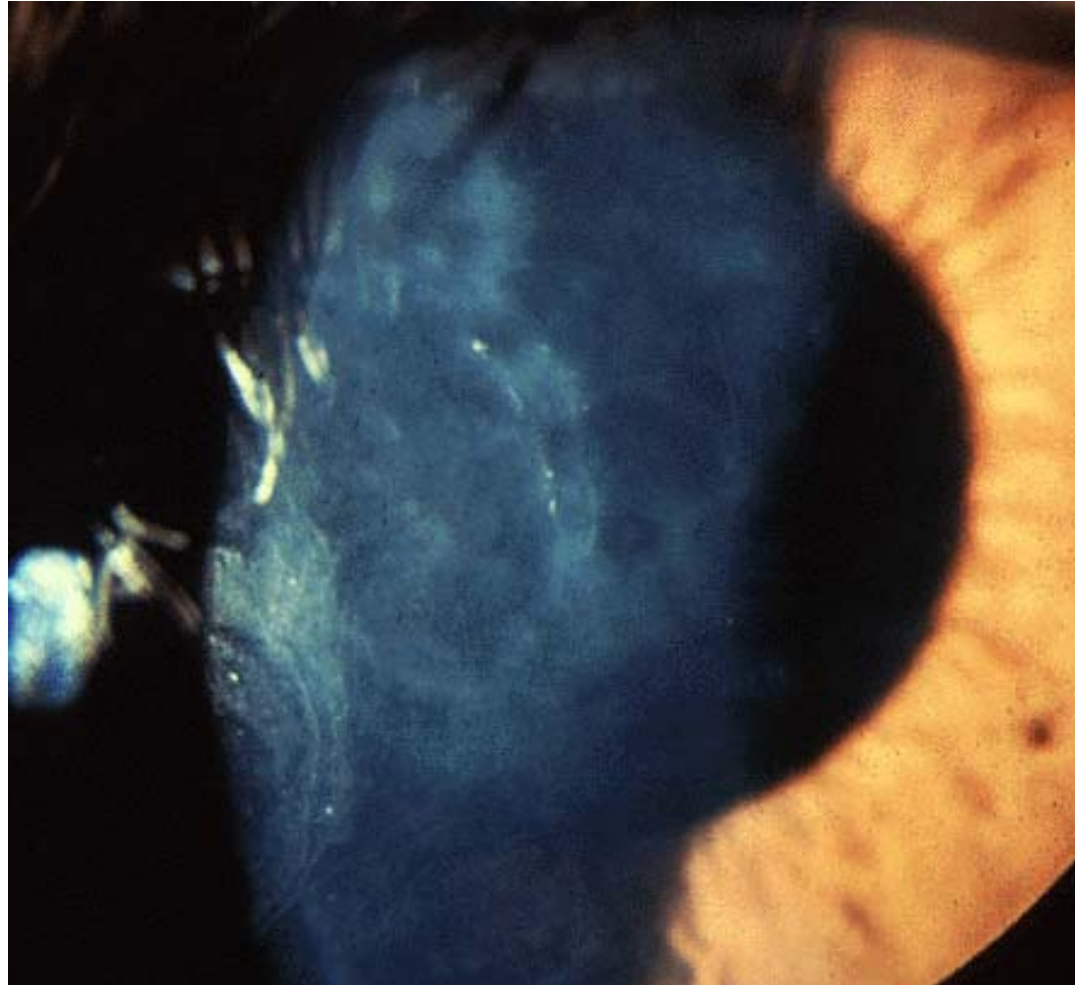


**Posterior polymorphous corneal dystrophy: *snail* or *railroad tracks***

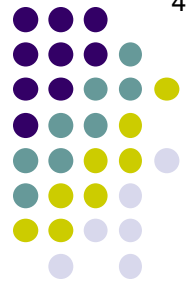
**Cornea****Posterior polymorphous corneal dystrophy****A:** Endothelial plaque-like lesions**B:** Irregular crater-like figures on Descemet membrane viewed with specular reflection

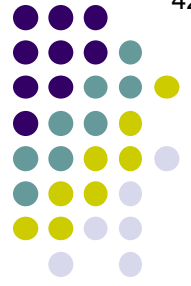


## Corneal Dystrophies



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





# Corneal Dystrophies

## Epithelial and Subepithelial Dystrophies

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

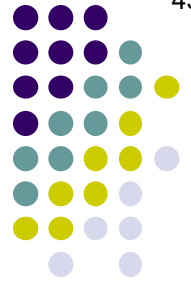
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*Which lesion is most common in PPMD?*

## Endothelial Dystrophies

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# Corneal Dystrophies

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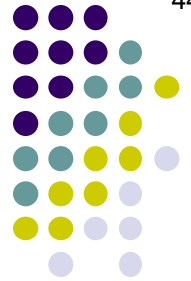
--Diffuse '**geographic**' opacities that often have an overlying haze in the posterior stroma

*Which lesion is most common in PPMD?*

Vesicular lesions--these are found in essentially all PPMD pts

## Endothelial Dystrophies

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# Corneal Dystrophies

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--**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?*

Vesicular lesions--these are found in essentially all PPMD pts

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

## 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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No, they contain a collagenous material

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## Corneal Dystrophies

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

At w  
Usu

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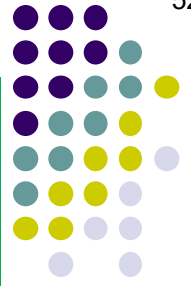
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Because unlike Haab striae, which taper and meet at their ends, the parallel lines in PPMD do **not** meet at their terminuses

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### Endothelial Dystrophies

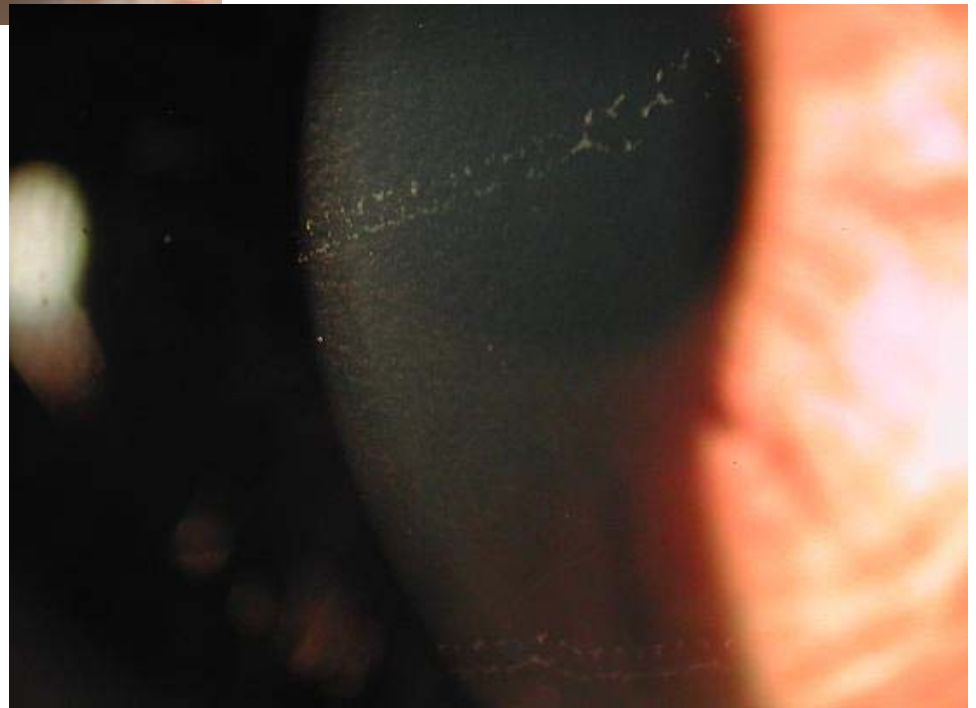
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phies

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

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





# 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:

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*Is it painful? Does it affect vision?*

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In the majority of pts, PPMD is a stable, painless and visually insignificant condition. However, severe cases can be associated with glaucoma, stromal edema, and significant vision loss.

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*What is the histologic hallmark of PPMD on light microscopy?*

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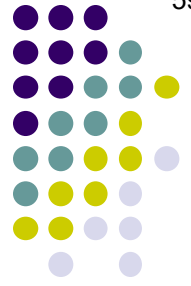
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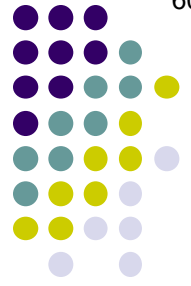
*In addition to being multilayered, what else is abnormal about the endothelial cells in PPMD?*

*What is the histologic hallmark of PPMD on light microscopy?*

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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**

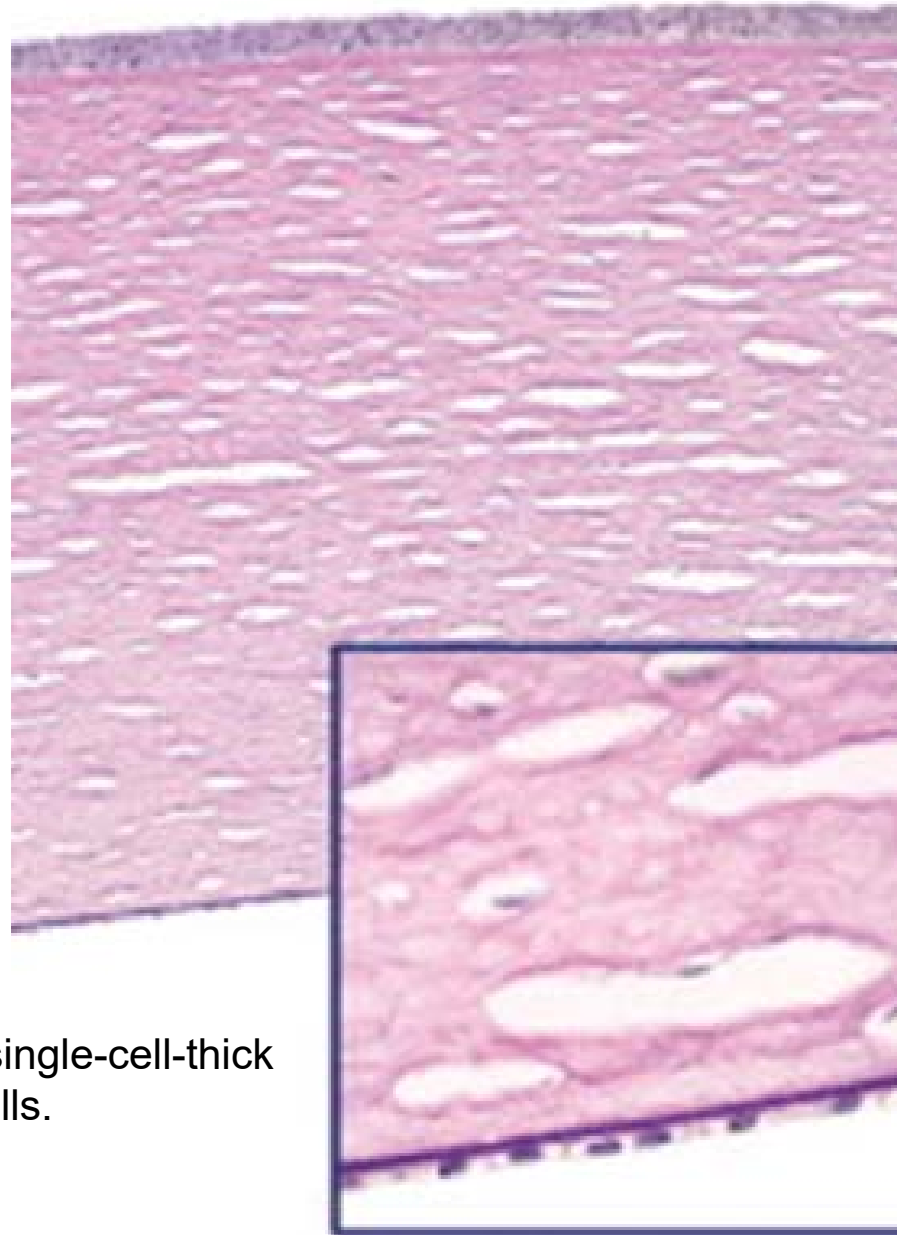
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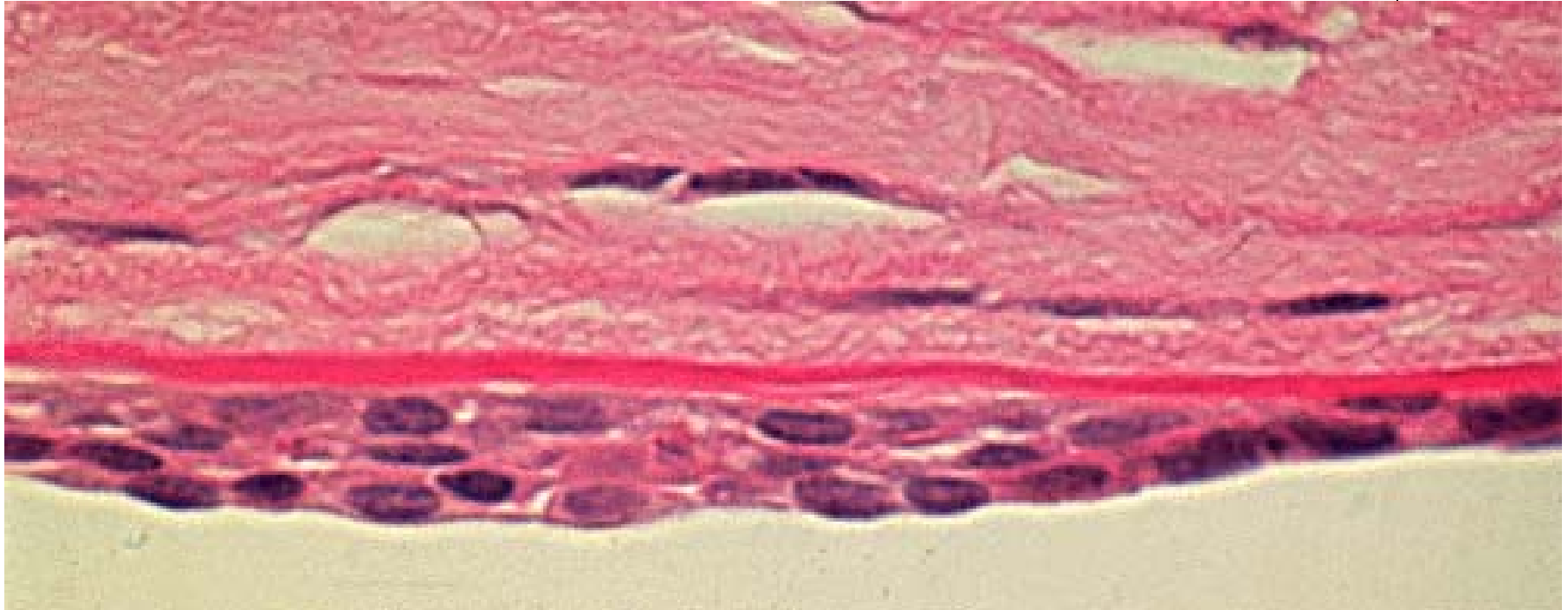
## Corneal Dystrophies



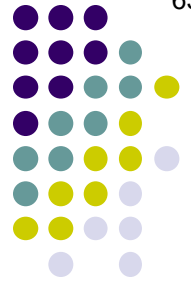
**Normal cornea.** Note the single-cell-thick nature of the endothelial cells.



## 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

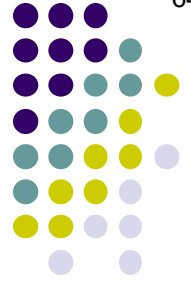
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# Corneal Dystrophies

## Epithelial and Subepithelial Dystrophies



## Epithelial-Stromal *TGFBI* Dystrophies

*What condition manifests findings that render it easily confusable with PPMD?*  
Iridocorneal endothelial (ICE) syndrome

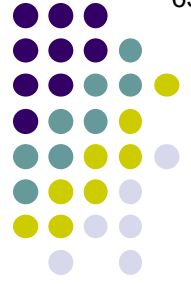
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# Corneal Dystrophies

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## Epithelial-Stromal *TGFBI* Dystrophies

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*How can you tell if a pt has PPMD vs ICE?*

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# Corneal Dystrophies

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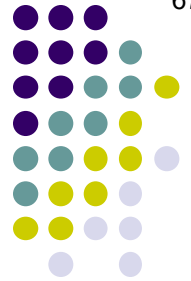
--Unlike PPMD, ICE is always inheritance

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--Unlike PPMD, ICE is always **sporadic**

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--Unlike PPMD, ICE is always **laterality**

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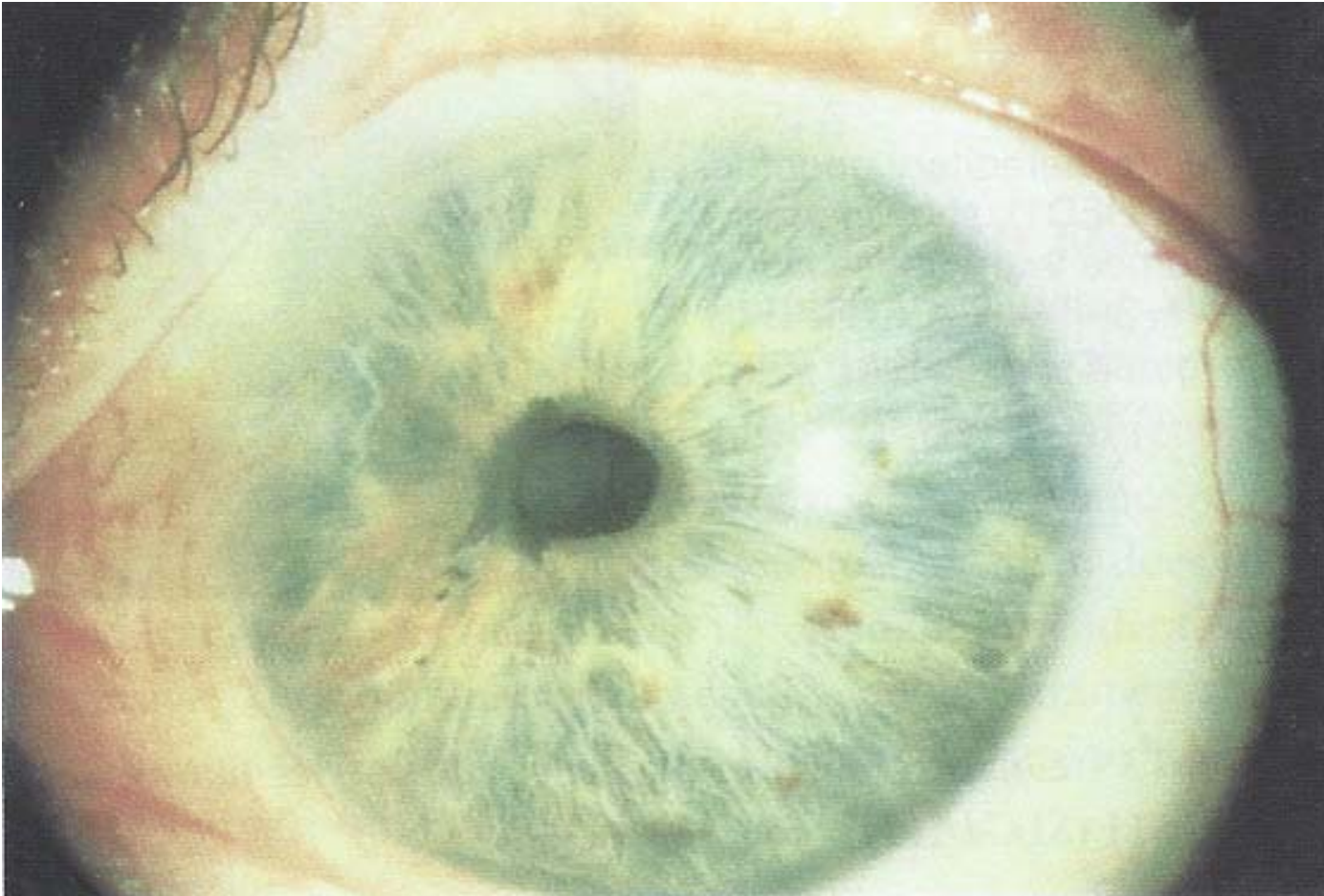
--Glaucoma

--Iris changes (eg, corectopia)

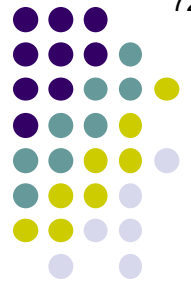
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## Corneal Dystrophies



**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?*

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*Is it progressive?*

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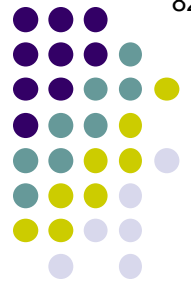
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The cornea is diffusely cloudy (described as 'ground glass' or 'milky'), and thickened

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## Corneal Dystrophies



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

# Corneal Dystrophies

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*How thickened?*

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*How thickened?*

Like, 2-3 times normal

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*Is vision affected?*

Yes, significantly enough to result in nystagmus is common

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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 isn't

*What is seen at the slit lamp?*

The cornea is diffusely cloudy (described as 'ground glass' or 'milky'), and thickened

*Is vision affected?*

Yes, significantly enough to result in nystagmus is common

*Is it painful?*

## Endothelial Dystrophies

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

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*What is the histologic hallmark of CHED2 on light microscopy?*

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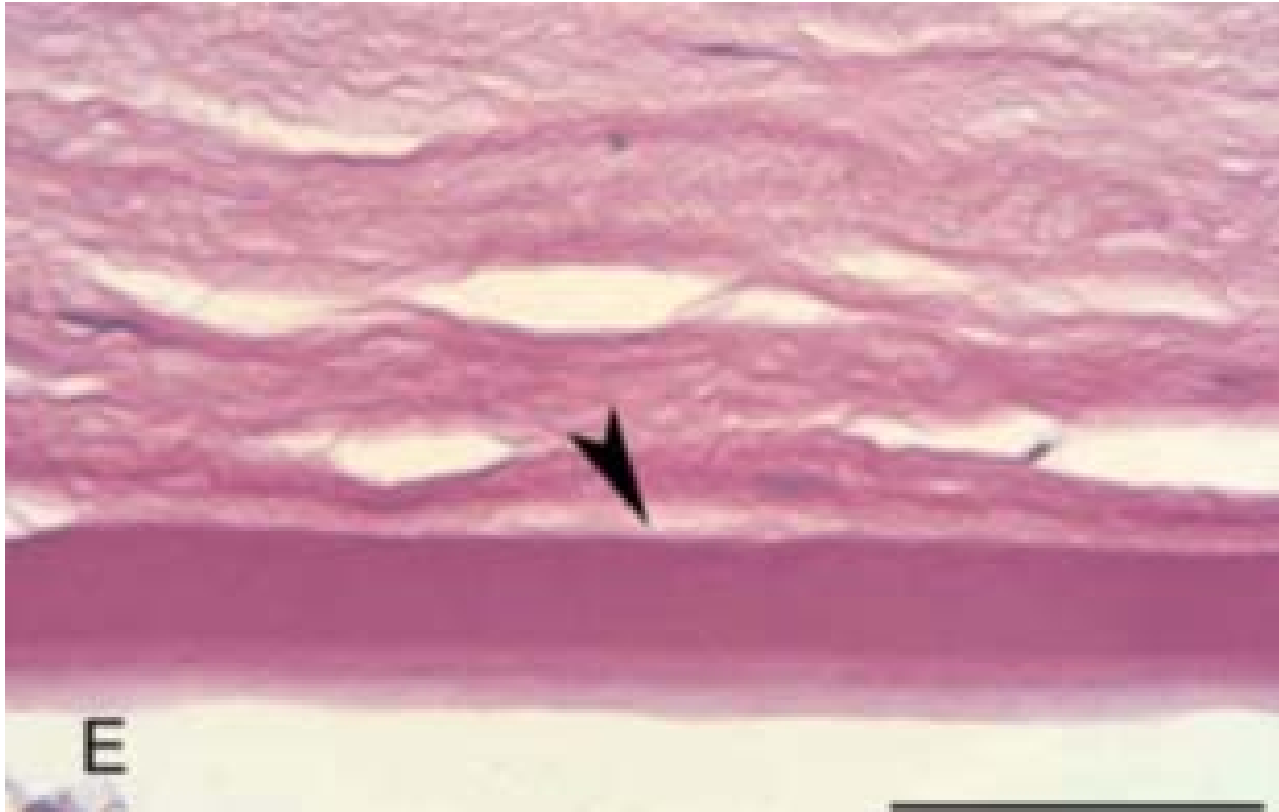
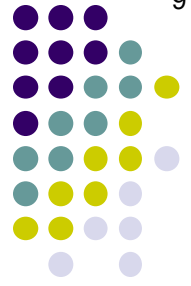
*What is the histologic hallmark of CHED2 on light microscopy?*

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

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## Corneal Dystrophies



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