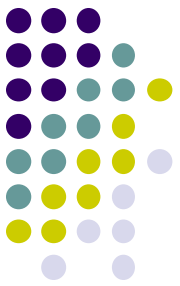


Corneal Dystrophies



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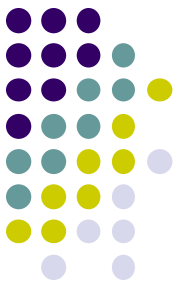
What are the four categories of corneal dystrophies?

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Corneal Dystrophies

2



Epithelial and Subepithelial Dystrophies

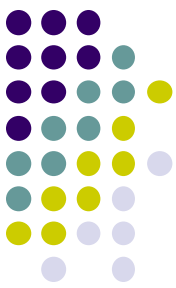
Epithelial-Stromal *TGFB1* Dystrophies

What are the four categories of corneal dystrophies?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) ?
- 2) ?
- 3) ?
- 4) ?
- 5) ?
- 6) ?

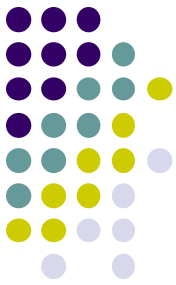
Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

*What are the six
epithelial/subepithelial
corneal dystrophies?*

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) Epithelial basement membrane dystrophy
- 2) Meesmann epithelial corneal dystrophy
- 3) Lisch epithelial corneal dystrophy
- 4) Gelatinous droplike corneal dystrophy
- 5) Epithelial recurrent erosion dystrophies
- 6) Subepithelial mucinous corneal dystrophy

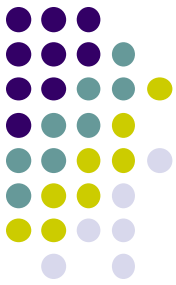
Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

What are the six epithelial/subepithelial corneal dystrophies?

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) Epithelial basement membrane dystrophy
- 2) Meesmann epithelial corneal dystrophy
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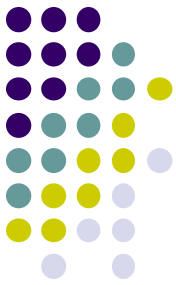
These two are not discussed in detail in the BCSC *Cornea* book, and thus will not be in this slide-set either

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



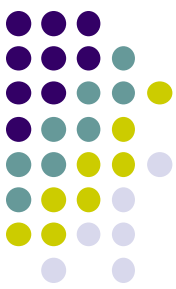
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- 5) Epithelial recurrent erosion dystrophies

What are three other common names for EBMD?

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Corneal Dystrophies



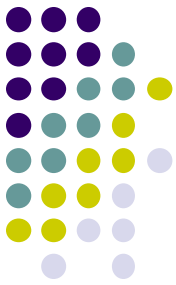
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- Cogan's microcystic dystrophy
- Anterior basement membrane dystrophy

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

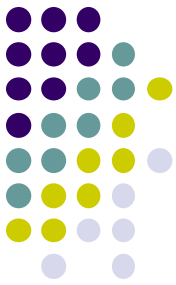
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What is the slit-lamp appearance of EBMD?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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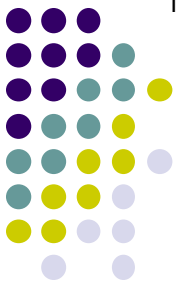
- Thin, curvy, hairlike lines that often occur in groups that follow the same highly curved course; these are called two words. These represent abnormal basement membrane material.

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Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
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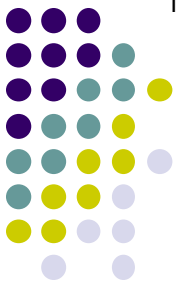
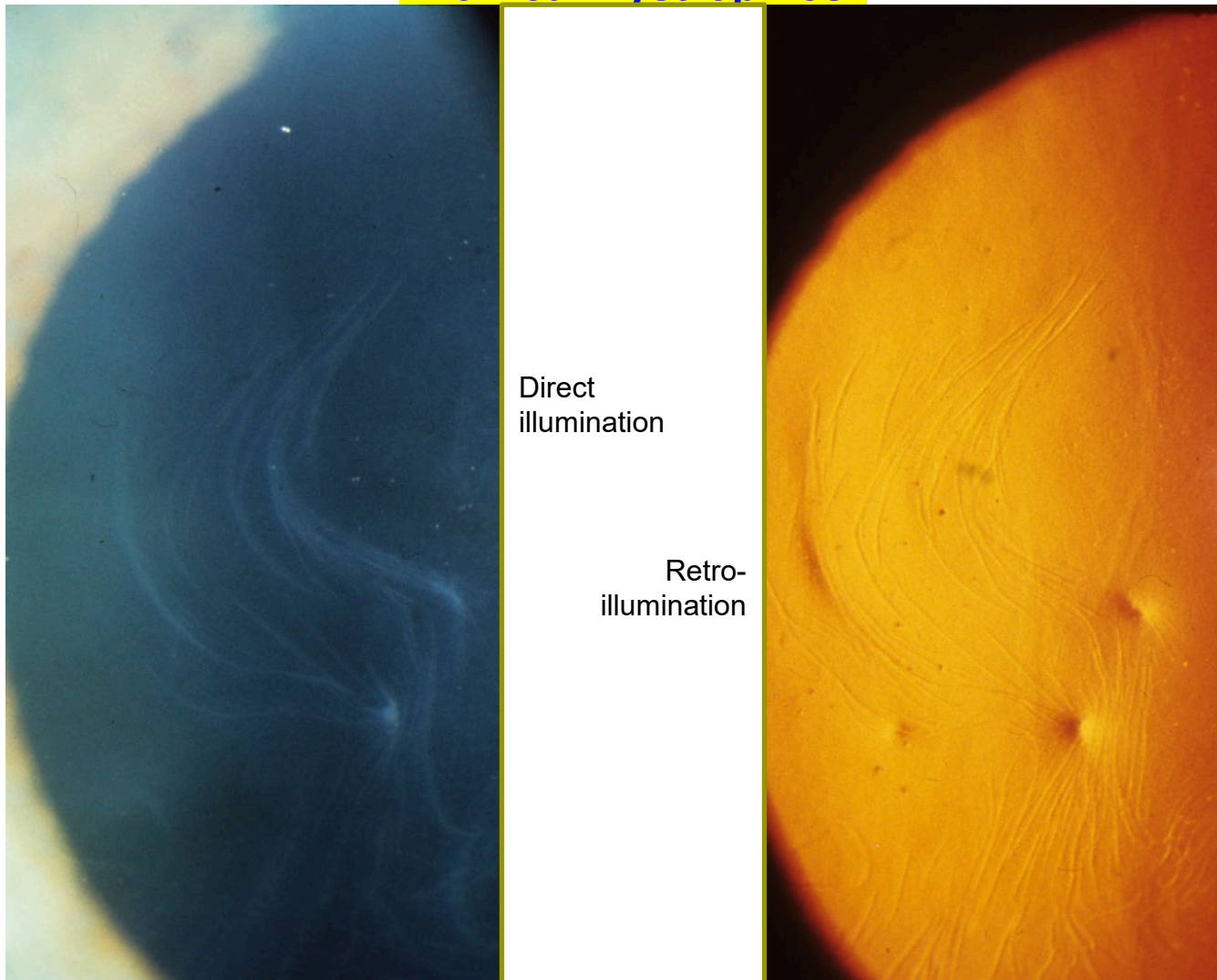
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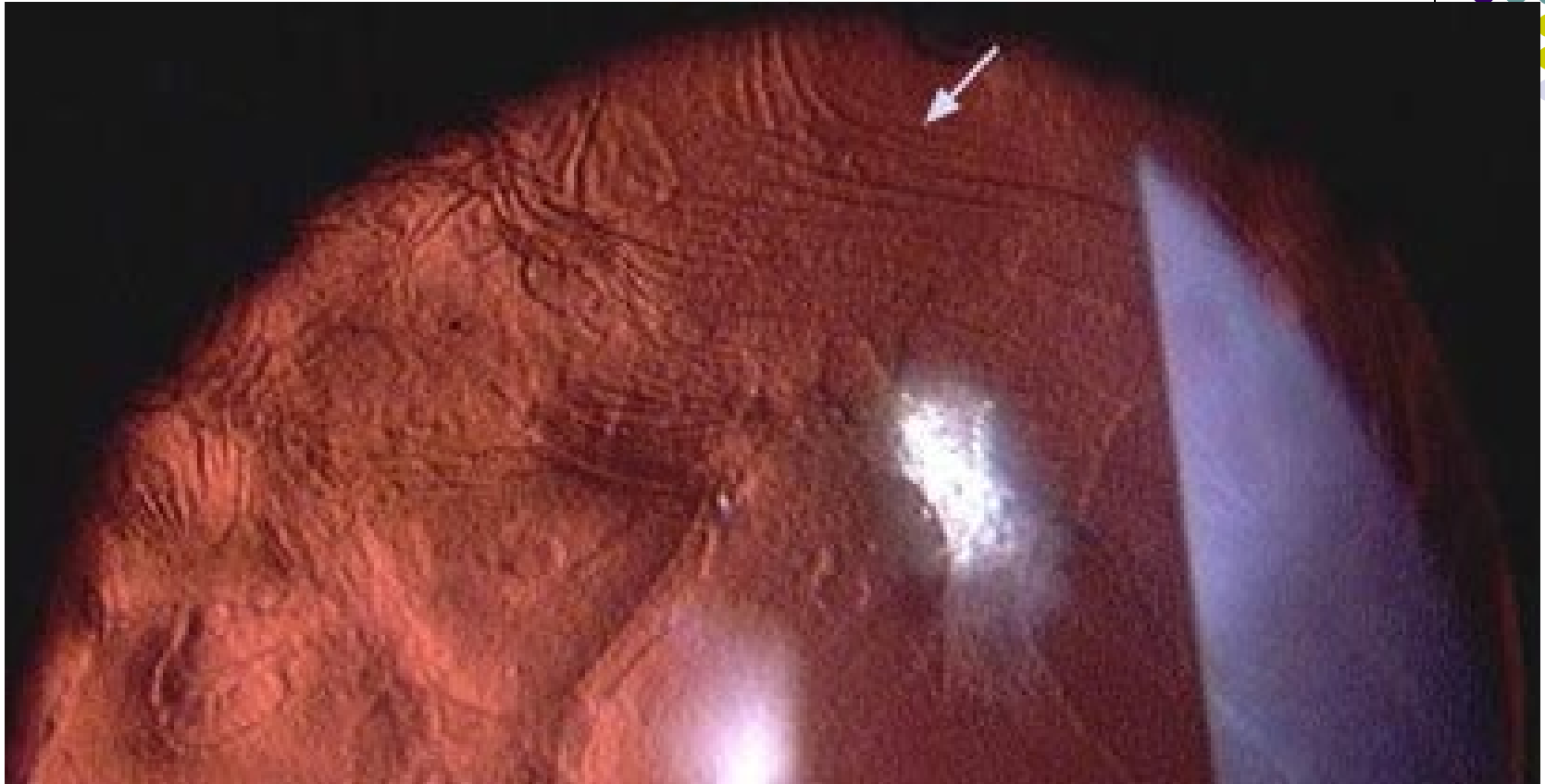
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Corneal Dystrophies



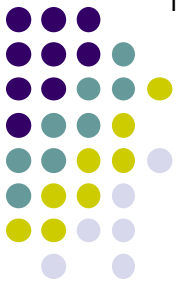
Epithelial basement membrane dystrophy. Fingerprint lines in direct and retro-illumination

Corneal Dystrophies



Epithelial basement membrane dystrophy. Fingerprint lines

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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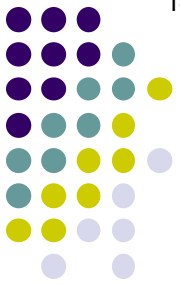
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- Thicker lines that follow a more jagged course; these are called two words. These also result from abnormal basement membrane.

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Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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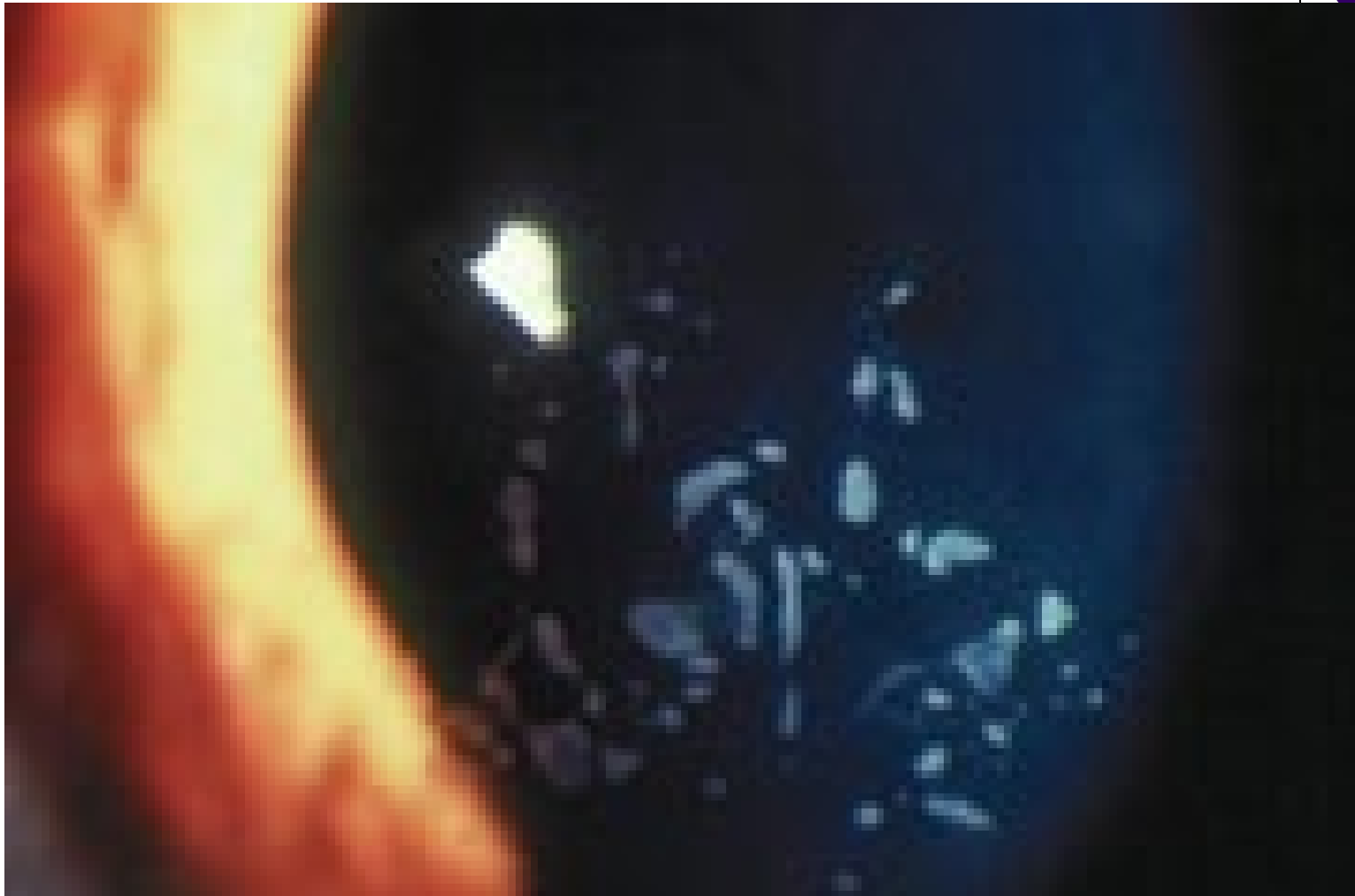
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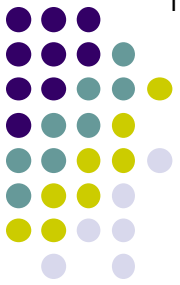
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Corneal Dystrophies



Epithelial basement membrane dystrophy. Map lines

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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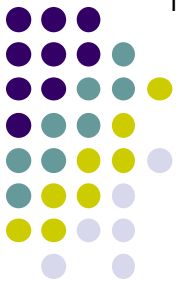
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- Multiple gray two/words . These consist of degenerated epithelial cell debris in the intraepithelial space.
-

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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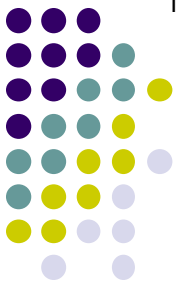
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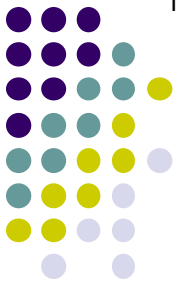
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-

Corneal Dystrophies



Epithelial basement membrane dystrophy. Dots

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
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What are three other common names for EBMD?

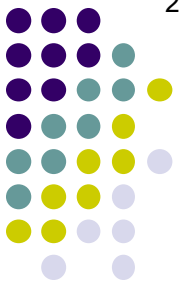
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- The so-called two words , which is seen in retroillumination to have a pebbled-glass appearance.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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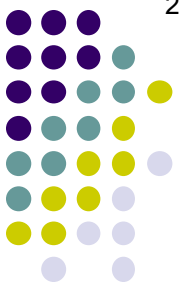
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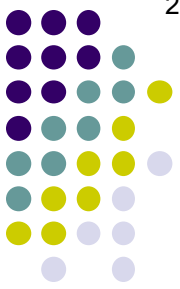
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- Multiple gray **dots/cysts** . These consist of degenerated epithelial cell debris in the intraepithelial space.
- The so-called **bleb pattern** , which is seen in retroillumination to have a pebbled-glass appearance.



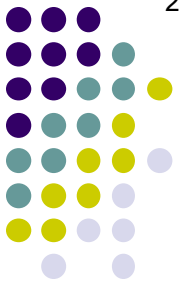
Epithelial basement membrane dystrophy. D, Multiple crowded blebs only visible in retroillumination.

Corneal Dystrophies



Epithelial basement membrane dystrophy. Typical appearance

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
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- Map-dot-fingerprint dystrophy
- Cogan's microcystic dystrophy
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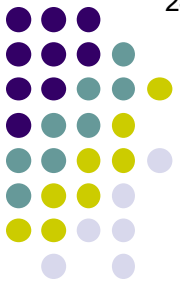
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What do pts complain of?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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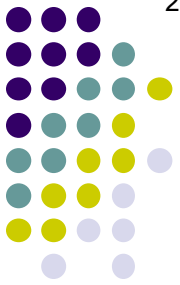
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What do pts complain of?

Most cases present with symptoms related to

three words, then their abb.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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- Map-dot-fingerprint dystrophy
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What is the slit-lamp appearance of EBMD?

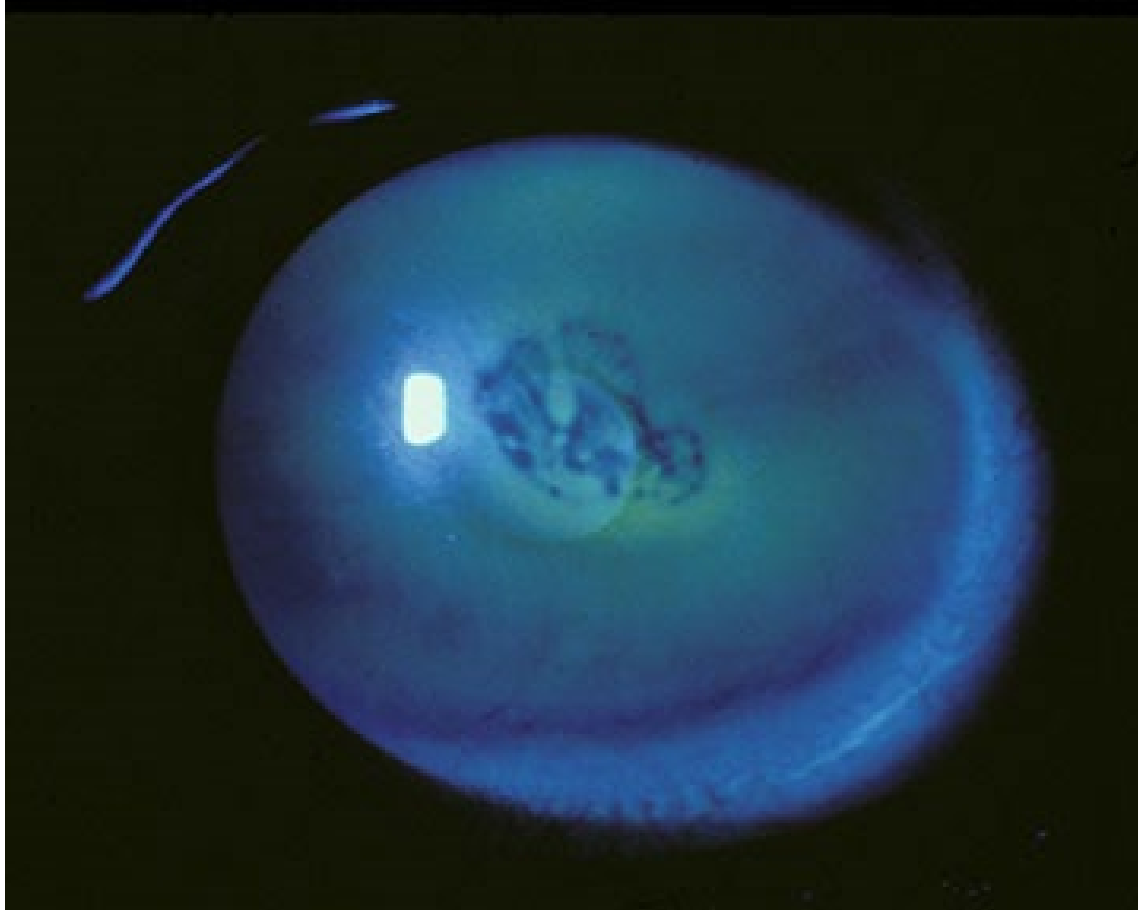
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What do pts complain of?

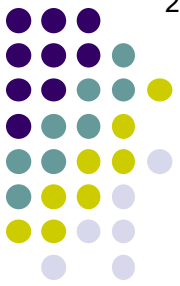
Most cases present with symptoms related to **recurrent epithelial erosions (REE)**

Corneal Dystrophies



REE: Small areas of epithelial loss with adjacent areas of rapid tear film breakup associated with poorly adherent corneal epithelium in the axial cornea

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
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- Map-dot-fingerprint dystrophy
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- Anterior basement membrane dystrophy

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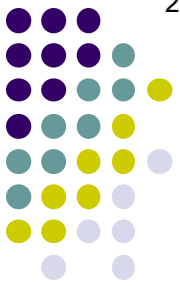
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- Multiple gray **dots/cysts**. These consist of degenerated epithelial cell debris in the intraepithelial space.
- The so-called **bleb pattern**, which is seen in retroillumination to have a pebbled-glass appearance.

What do pts complain of?

Most cases present with symptoms related to **recurrent epithelial erosions (REE)**. Additionally, if the epithelium in the visual axis gets heaped up and distorted enough, it can cause two words with resulting decreased vision.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
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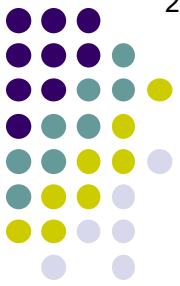
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Most cases present with symptoms related to **recurrent epithelial erosions (REE)**. Additionally, if the epithelium in the visual axis gets heaped up and distorted enough, it can cause **irregular astigmatism** with resulting decreased vision.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
- 2) Meesmann epithelial corneal dystrophy
- 3) Lisch epithelial corneal dystrophy

What are the major histologic abnormalities in EBMD?

--?

--?

--?

--Cogan's microcystic dystrophy

--Anterior basement membrane dystrophy

What is the slit-lamp appearance of EBMD?

It can present in any combo of the following:

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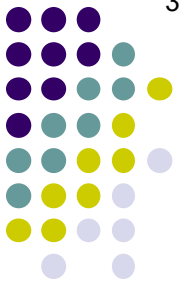
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Most cases present with symptoms related to **recurrent epithelial erosions** (REE). Additionally, if the epithelium in the visual axis gets heaped up and distorted enough, it can cause **irregular astigmatism** with resulting decreased vision

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
- 2) Meesmann epithelial corneal dystrophy
- 3) Lisch epithelial corneal dystrophy

What are the major histologic abnormalities in EBMD?

--The epithelial cells are...

--?

--?

--Cogan's microcystic dystrophy

--Anterior basement membrane dystrophy

What is the slit-lamp appearance of EBMD?

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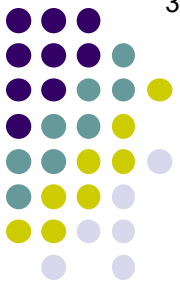
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Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) **Epithelial basement membrane dystrophy**
- 2) Meesmann epithelial corneal dystrophy
- 3) Lisch epithelial corneal dystrophy

What are the major histologic abnormalities in EBMD?

--The epithelial cells are...disordered, contain microcysts, and lack normal adhesion structures

--?

--?

--Cogan's microcystic dystrophy

--Anterior basement membrane dystrophy

What is the slit-lamp appearance of EBMD?

It can present in any combo of the following:

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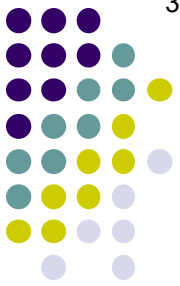
--Multiple gray **dots/cysts**. These consist of degenerated epithelial cell debris in the intraepithelial space.

--The so-called **bleb pattern**, which is seen in retroillumination to have a pebbled-glass appearance.

What do pts complain of?

Most cases present with symptoms related to **recurrent epithelial erosions** (REE). Additionally, if the epithelium in the visual axis gets heaped up and distorted enough, it can cause **irregular astigmatism** with resulting decreased vision

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

1) **Epithelial basement membrane dystrophy**

2) Meesmann epithelial corneal dystrophy

3) Lisch epithelial corneal dystrophy

What are the major histologic abnormalities in EBMD?

--The epithelial cells are...disordered, contain microcysts, and lack normal adhesion structures

--The basement membrane is...

--?

--Cogan's microcystic dystrophy

--Anterior basement membrane dystrophy

What is the slit-lamp appearance of EBMD?

It can present in any combo of the following:

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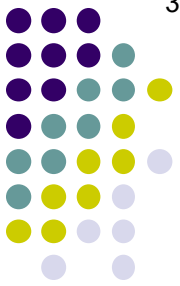
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What are the major histologic abnormalities in EBMD?

- The epithelial cells are...disordered, contain microcysts, and lack normal adhesion structures
- The basement membrane is...thickened, irregular, and may extend into the epithelium above
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What is the slit-lamp appearance of EBMD?

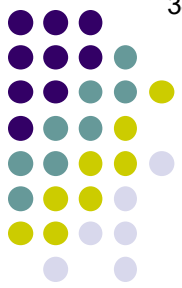
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What are the major histologic abnormalities in EBMD?

- The epithelial cells are...disordered, contain microcysts, and lack normal adhesion structures
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- A material accumulates between the BM and the underlying Bowman's layer

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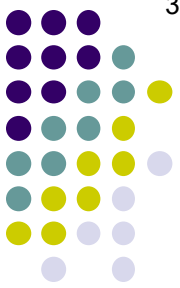
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What are the major histologic abnormalities in EBMD?

- The epithelial cells are...disordered, contain microcysts, and lack normal adhesion structures
- The basement membrane is...thickened, irregular, and may extend into the epithelium above
- A fibrillar material accumulates between the BM and the underlying Bowman's layer

--Cogan's microcystic dystrophy

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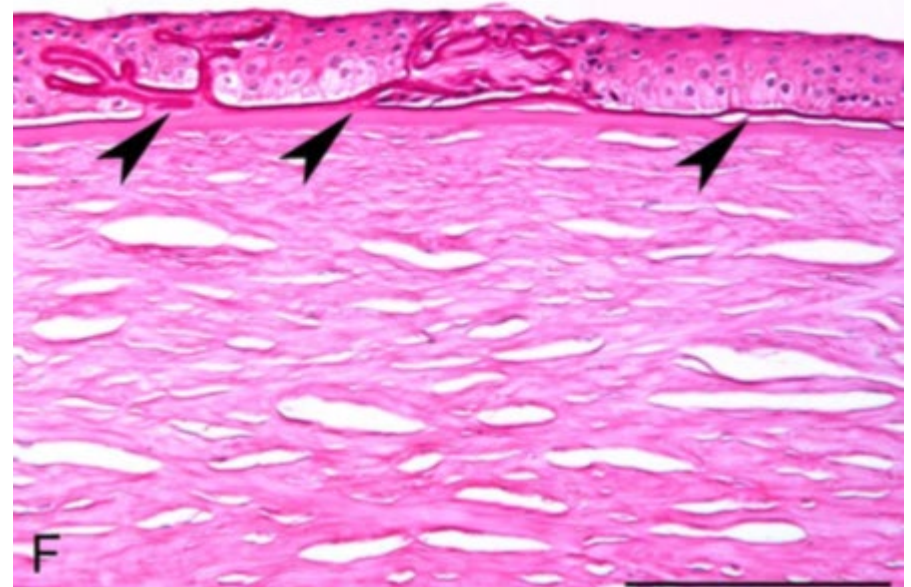
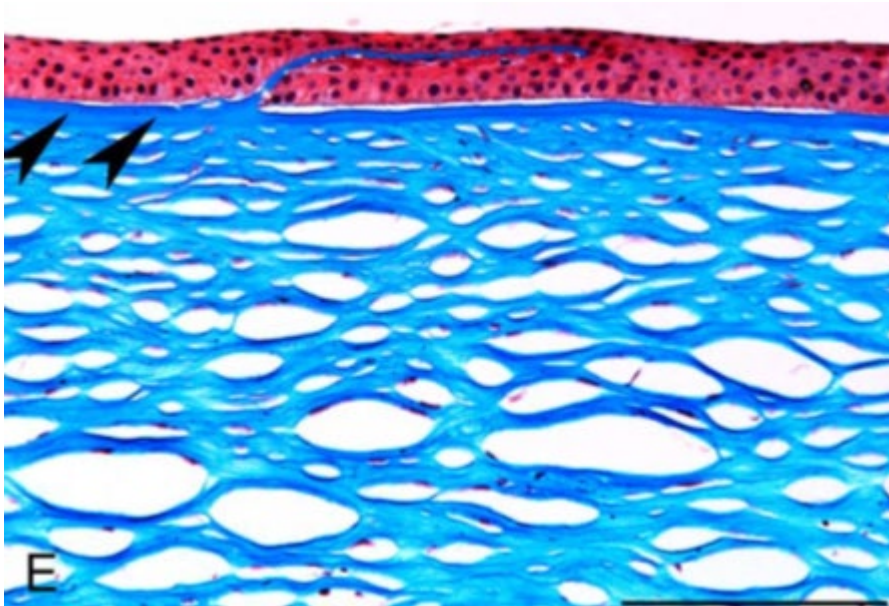
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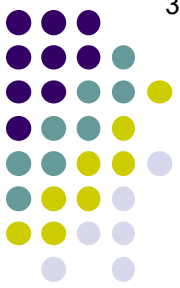
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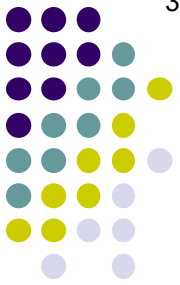
Epithelial basement membrane dystrophy. Light microscopy shows excessive basement membrane material (arrowheads) intervening between distorted epithelium and the intact Bowman layer to form redundant sheets corresponding to maps (E) and fingerprint.

Corneal Dystrophies



EBMD: Intraepithelial fibrillar material

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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- 3) Lisch epithelial corneal dystrophy
- 4) Gelatinous droplike corneal dystrophy
- 5) Epithelial recurrent erosion dystrophies

What are three other common names for EBMD?

- Map-dot-fingerprint dystrophy
- Cogan's microcystic dystrophy
- Anterior basement membrane dystrophy

How is EBMD inherited?

What is the inheritance pattern?

It can be

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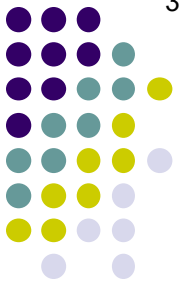
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Corneal Dystrophies



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How is EBMD inherited?

What is the inheritance pattern? As of the 2019 *Cornea* book, no inheritance pattern has been reliably identified. (In fact, the book allows that EBMD may not be a dystrophy at all, but rather a **corneal degeneration**.)

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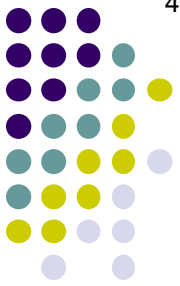
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Corneal Dystrophies



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What are the '6 F's' of EBMD?

- F
- F
- F
- F
- F

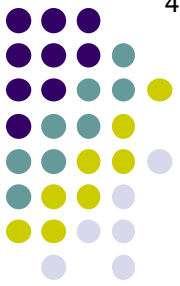
F

What do pts complain of?

Most cases present with recurrent epithelial in the visual field resulting decreased vis

Additionally, if the cornea is irregular, it can cause **irregular astigmatism** with

Corneal Dystrophies



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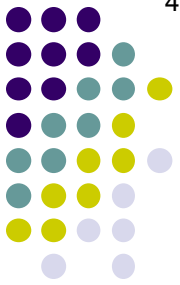
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- The so-called **bleb** appearance.

What do pts complain of?

- Most cases present with decreased vision. Additionally, if the dystrophy is severe, it can cause **regular astigmatism** with resulting decreased vision.
- F**emale
- F**ifties and older (usually)
- F**ive to **F**ifteen percent of the population are afflicted
- F**ifty percent of pts who suffer REE have it
- F**ibrillar material accumulates under the basement membrane

Corneal Dystrophies

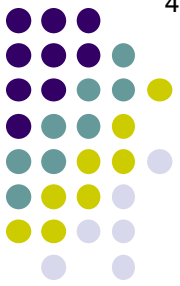


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What is the former name for this condition?

Corneal Dystrophies



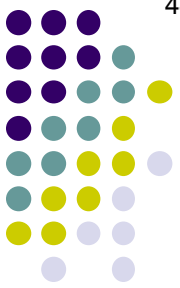
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Juvenile hereditary epithelial dystrophy

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

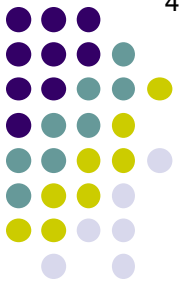
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At what age does it begin to manifest?

Corneal Dystrophies



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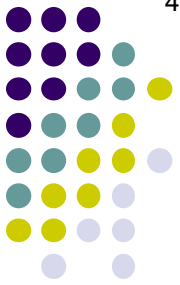
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Very young--usually before the child's second birthday (hence the word *juvenile* in the old name)

Corneal Dystrophies



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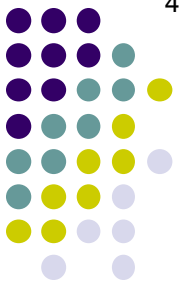
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Corneal Dystrophies



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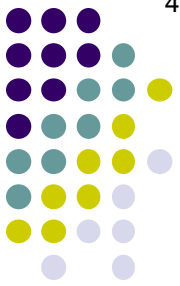
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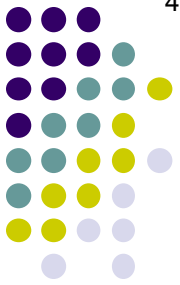
Direct illumination reveals multiple dull-gray opacities; retroillumination reveals innumerable tiny epithelial microcysts extending limbus-to-limbus

Corneal Dystrophies



Meesmann corneal dystrophy appearing as multiple dull-gray opacities

Corneal Dystrophies



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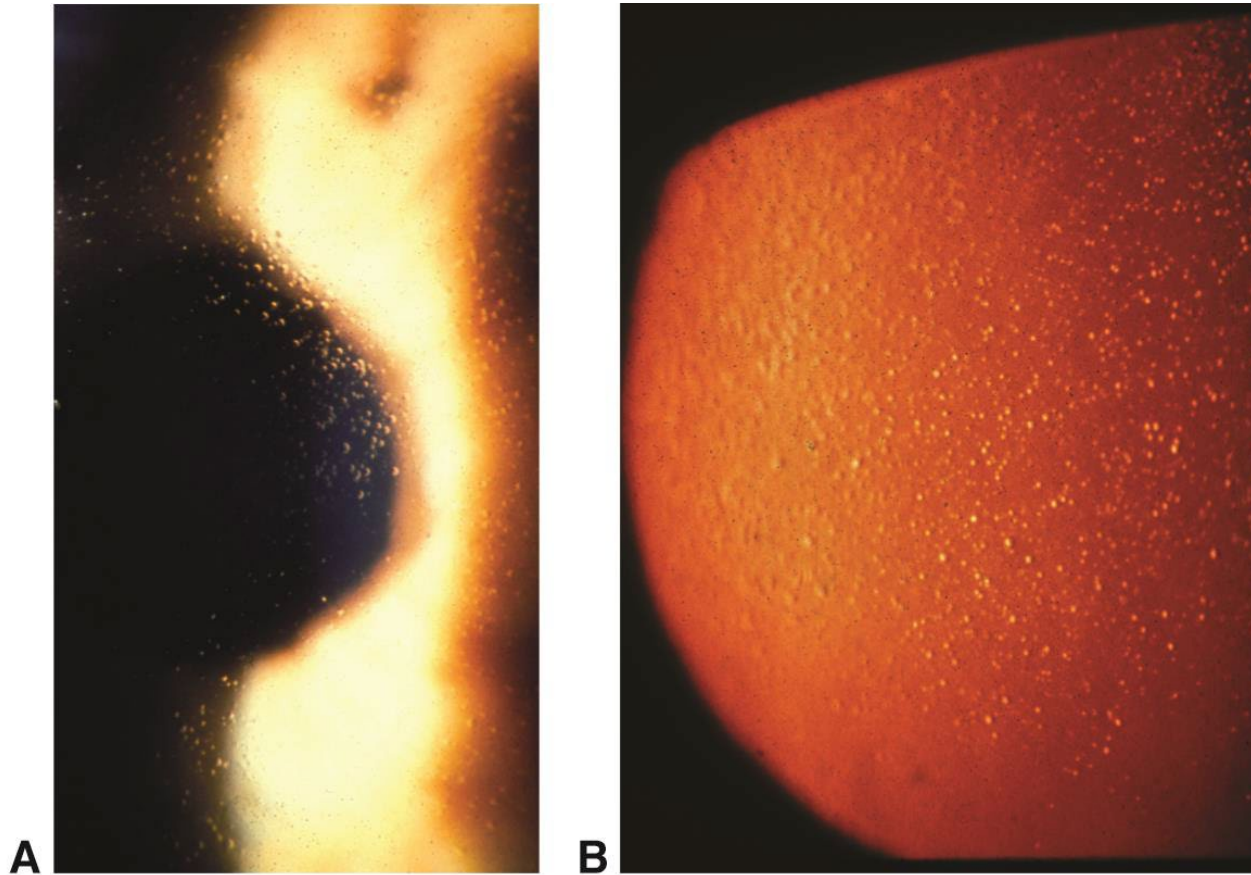
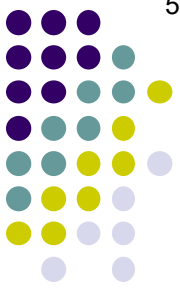
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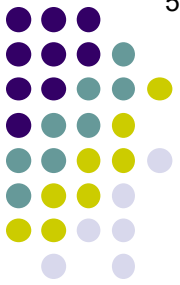
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Corneal Dystrophies



A, Meesmann corneal dystrophy, appearing as tiny bubblelike blebs with indirect slit-lamp illumination. B, Blebs are also well seen against the red reflex

Corneal Dystrophies



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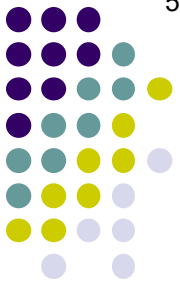
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Are these microcysts evenly distributed?

Corneal Dystrophies



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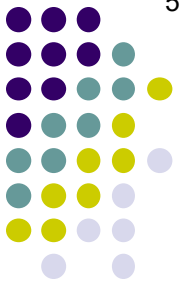
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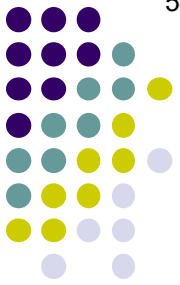
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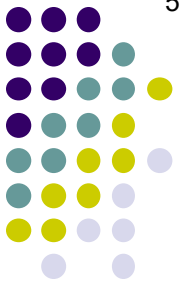
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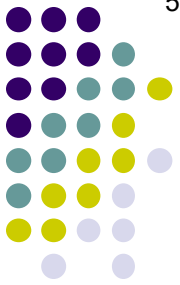
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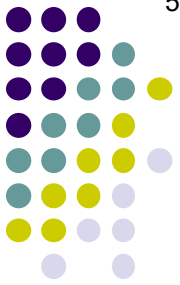
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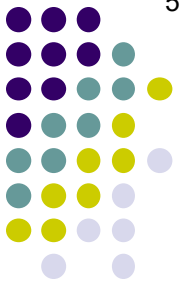
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Do recurrent epithelial erosions occur in this condition?

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No, vision is only mildly affected

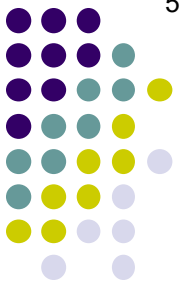
Are they painful?

A mild foreign-body sensation may occur, but it is not a particularly painful condition. Unless...

Do recurrent epithelial erosions occur in this condition?

Yes, and these episodes **are** quite painful

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) Epithelial basement membrane dystrophy
- 2) **Meesmann epithelial corneal dystrophy**
- 3) Lisch epithelial corneal dystrophy
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What is the former name for this condition?

Juvenile hereditary epithelial dystrophy

At what age does it begin to manifest?

Very young--usually before the child's second birthday (hence the word *juvenile* in the old name)

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

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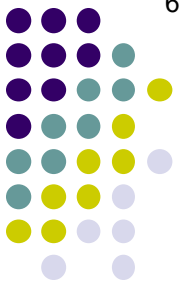
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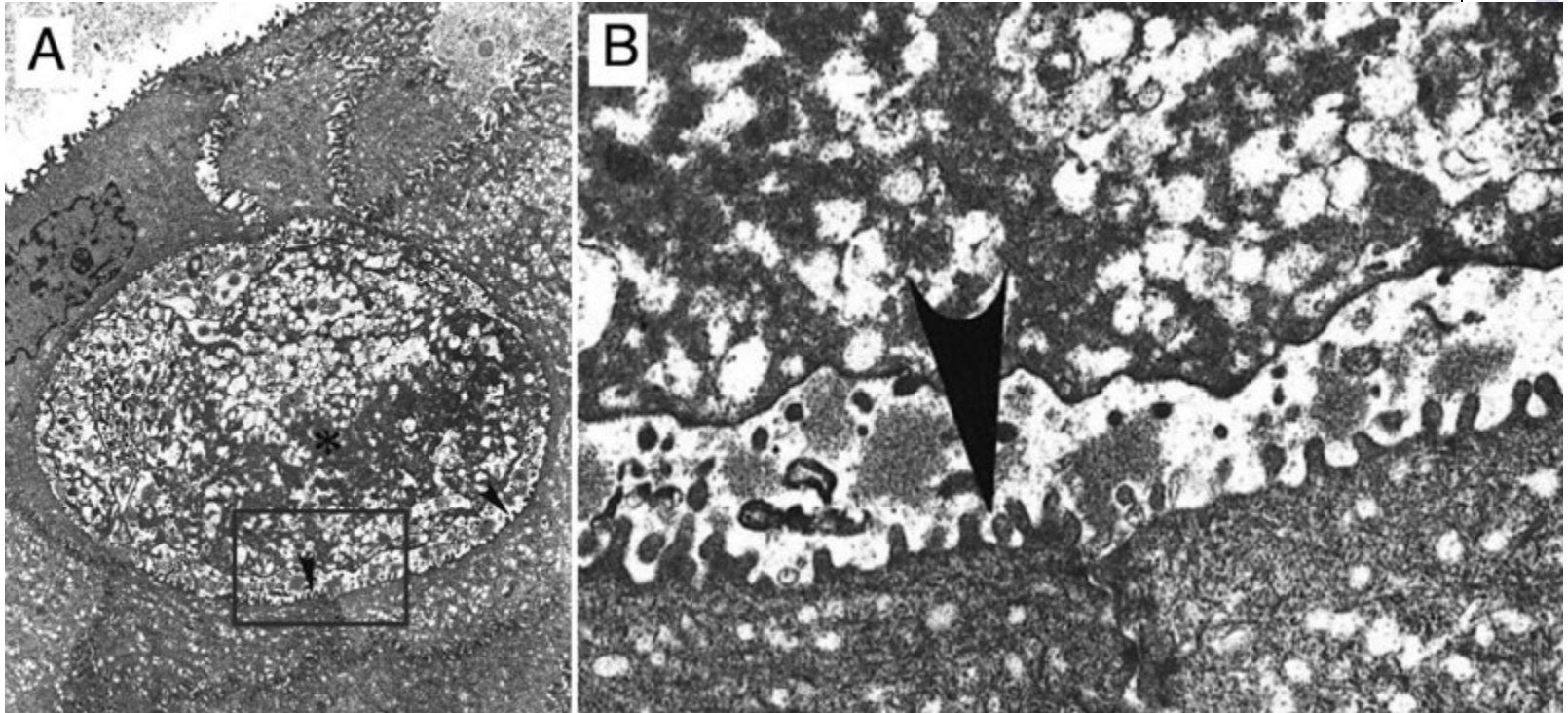
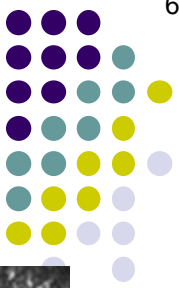
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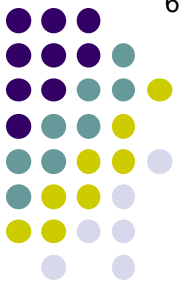
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It is called '**peculiar substance**'



A: Electron micrograph of corneal epithelium depicting an intraepithelial cyst containing a "peculiar" electron-dense substance (asterisk) intermixed with small vacuoles and electron-dense filamentous material.
B: Higher magnification of A.

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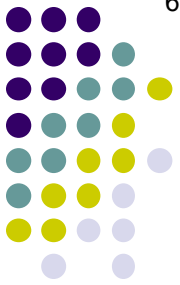
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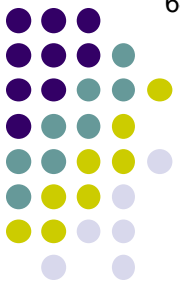
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When symptomatic, what intervention is adequate in most cases?

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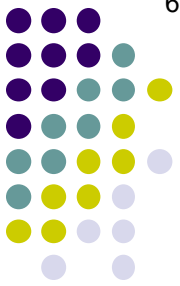
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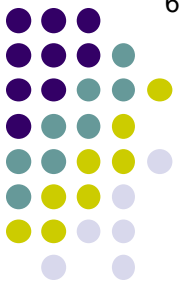
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Corneal Dystrophies

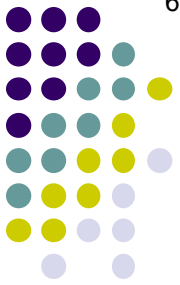


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At what age does LECD begin to manifest?

Corneal Dystrophies



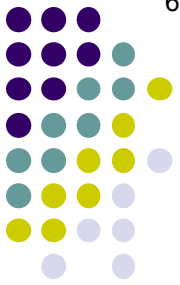
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Corneal Dystrophies



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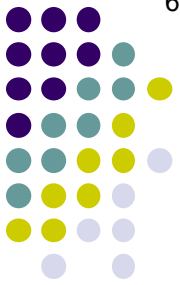
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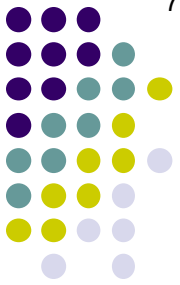
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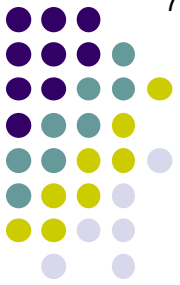
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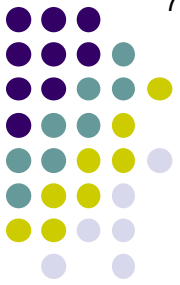
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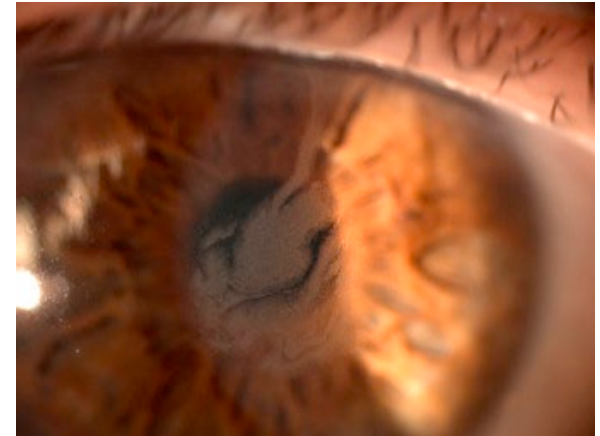
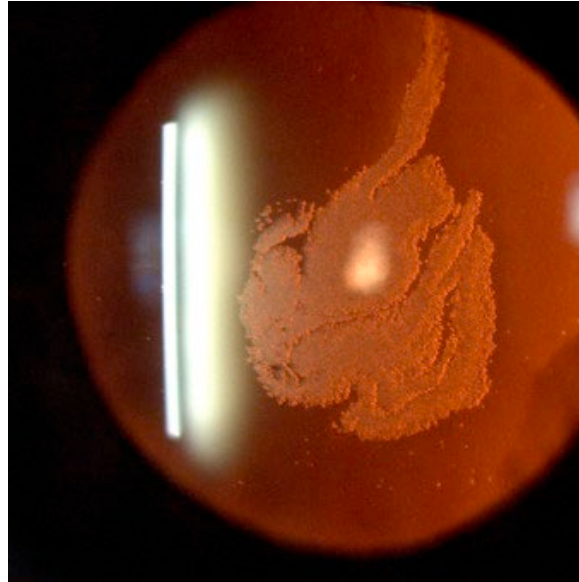
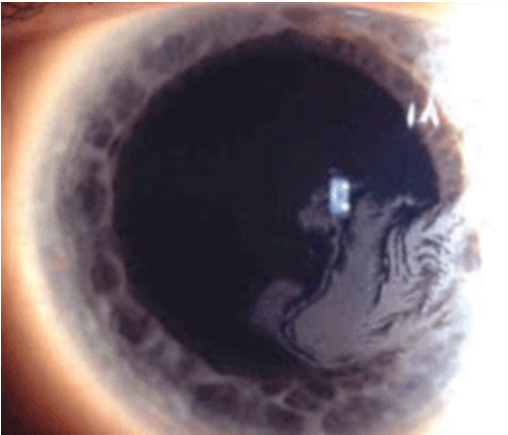
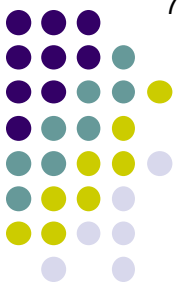
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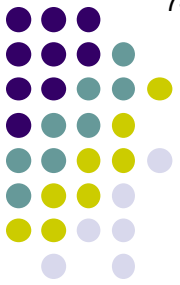
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Corneal Dystrophies



Lisch epithelial corneal dystrophy: Feather-shaped lesions

Corneal Dystrophies



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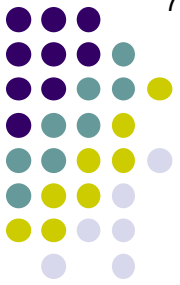
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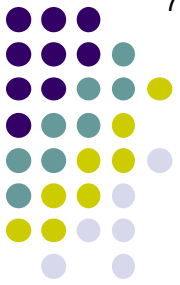
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Only if they happen to block the visual axis

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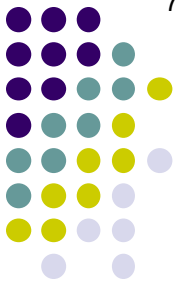
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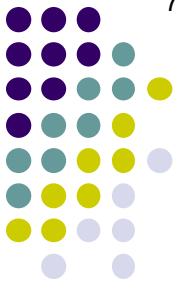
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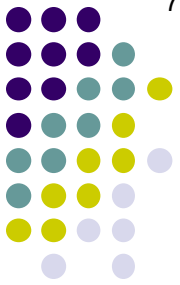
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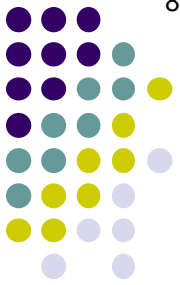
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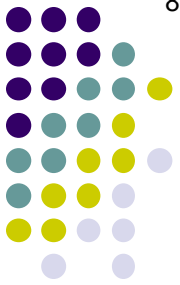


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In one word (not 'dystrophy,' duh), what sort of condition is GD CD?

Corneal Dystrophies

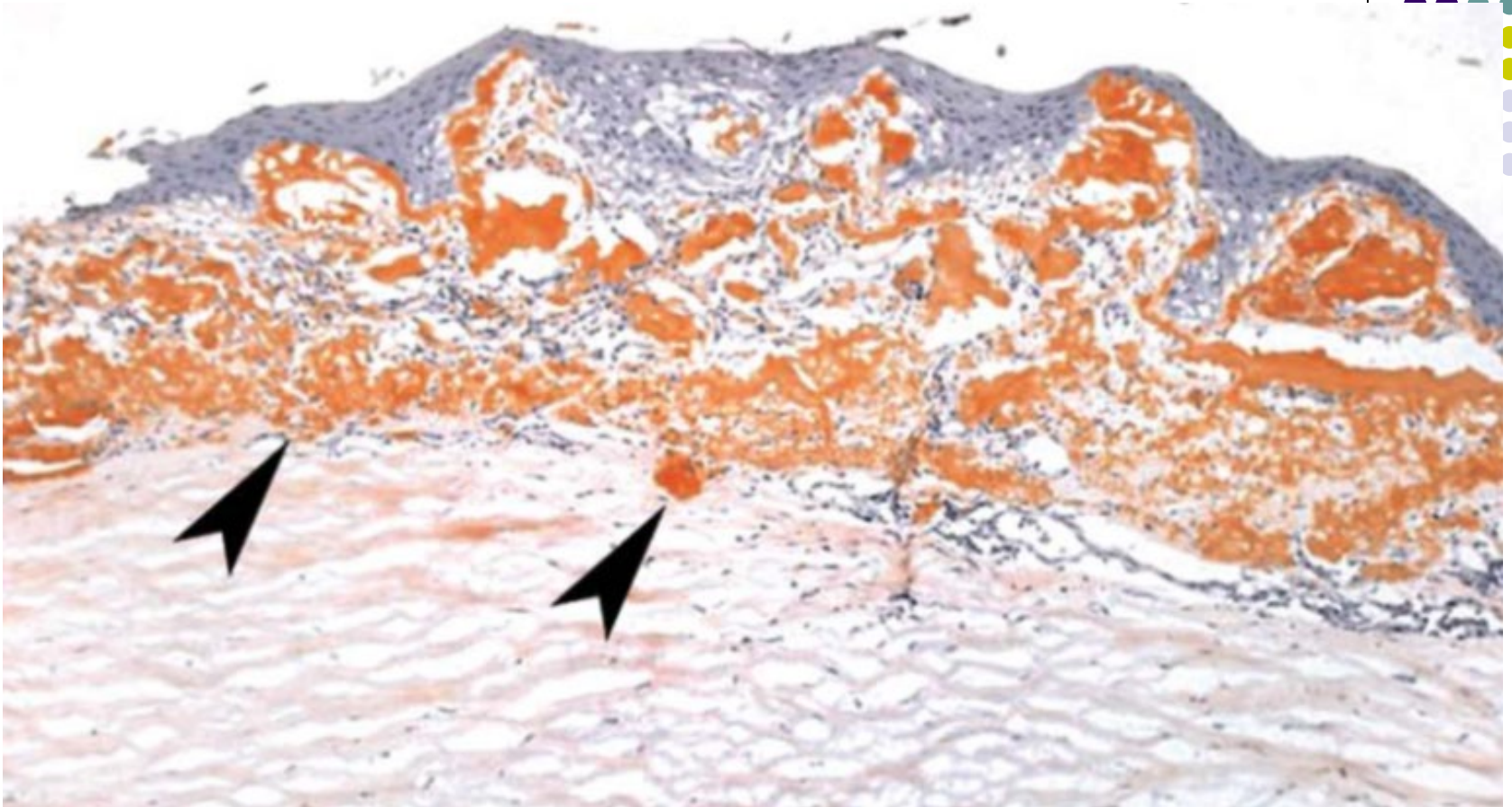


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An **amyloidosis**. Amyloid is deposited in the subepithelial space, and within the corneal stroma.



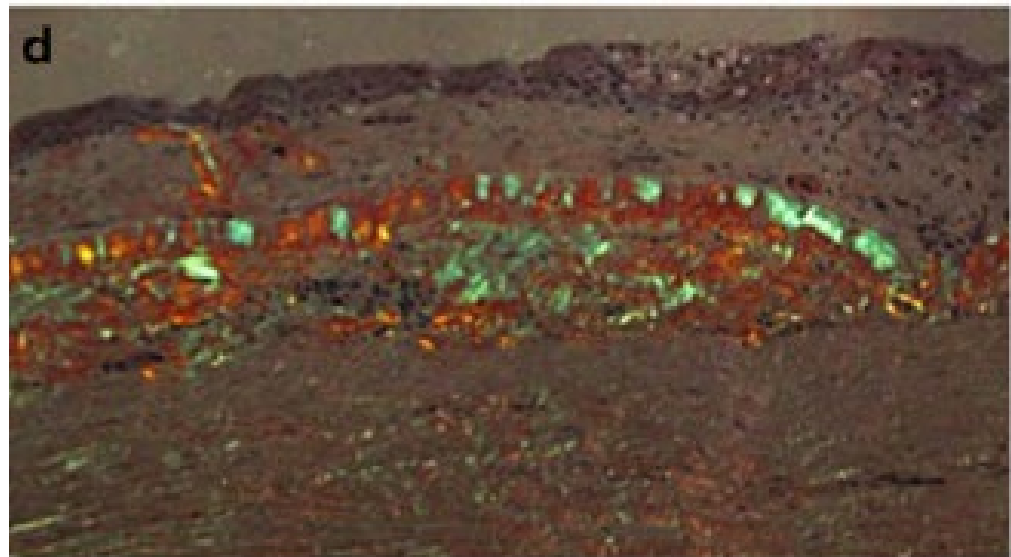
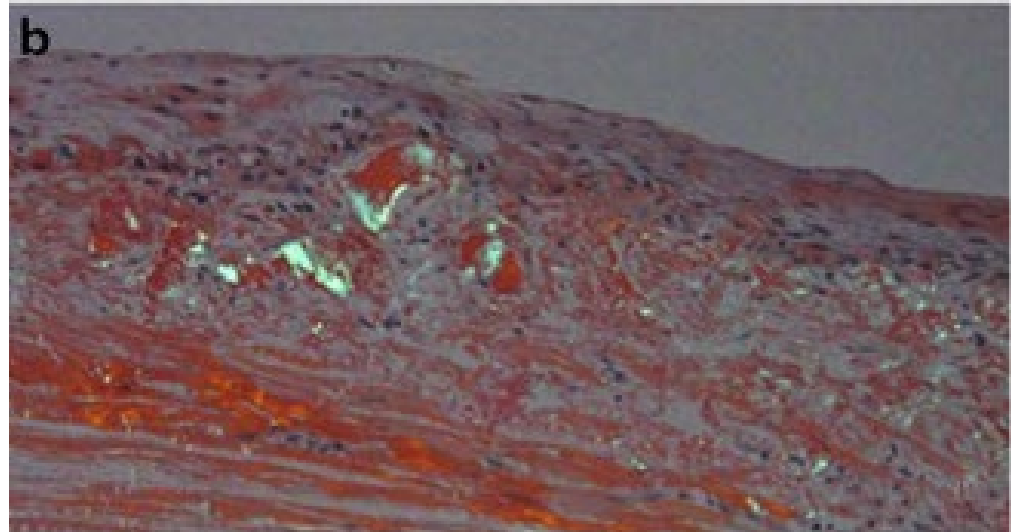
Gelatinous drop-like corneal dystrophy. Light microscopy: massive amyloid in a subepithelial lesion (arrowheads) extending to the midstromal cornea.

Corneal Dystrophies

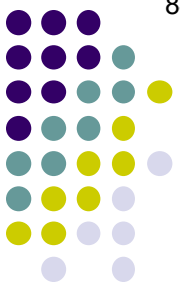


Gelatinous drop-like corneal dystrophy.

B, Congo Red stain. D, Polarized light reveals apple-green birefringence



Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

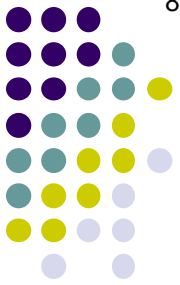
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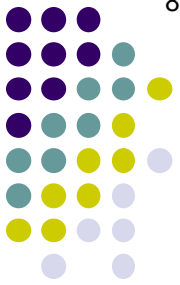
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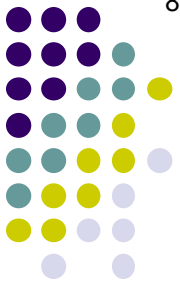
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How does it present? What is seen at the slit lamp?

Corneal Dystrophies



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An **amyloidosis**. Amyloid is deposited in the subepithelial space, and within the corneal stroma.

At what age does it begin to manifest?

Young--childhood to teens

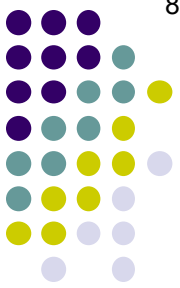
How does it present? What is seen at the slit lamp?

Early in the disease process it presents in one of two ways:

--In a two words like fashion

--As groups of nodules (aka two different words)

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

- 1) Epithelial basement membrane dystrophy
- 2) Meesmann epithelial corneal dystrophy
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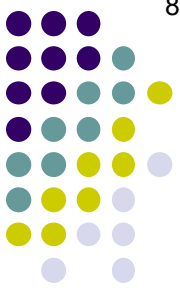
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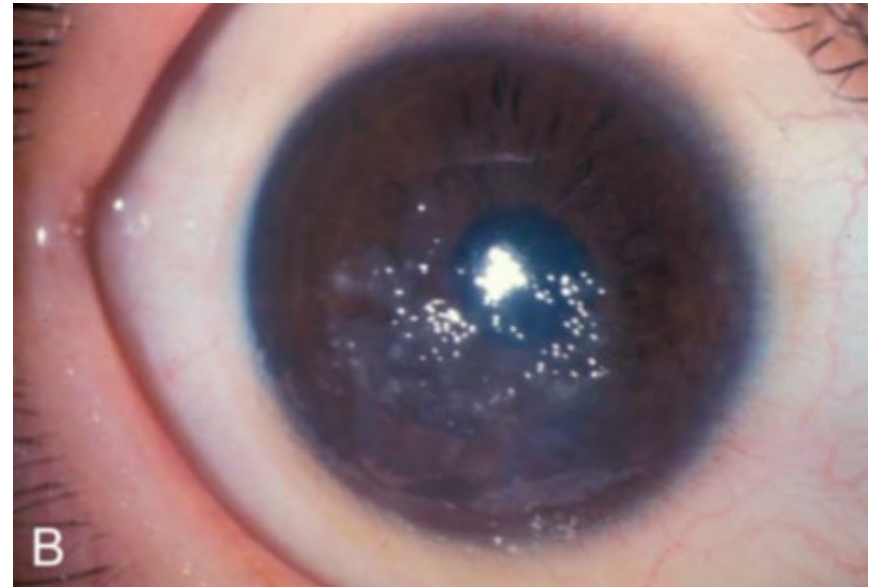
--In a **band keratopathy** -like fashion

--As groups of nodules (aka '**mulberry cornea**')

Corneal Dystrophies



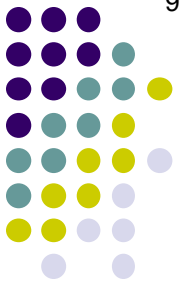
Initial presentation: **'Band-like'**



Initial presentation: **'Mulberry cornea'**

Gelatinous drop-like corneal dystrophy.
A, Band keratopathy type. B, Mulberry type.

Corneal Dystrophies



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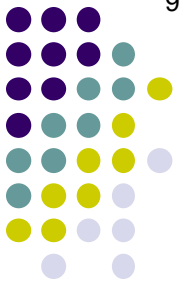
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Late in the process, the entire cornea becomes opaque--the so-called

two words

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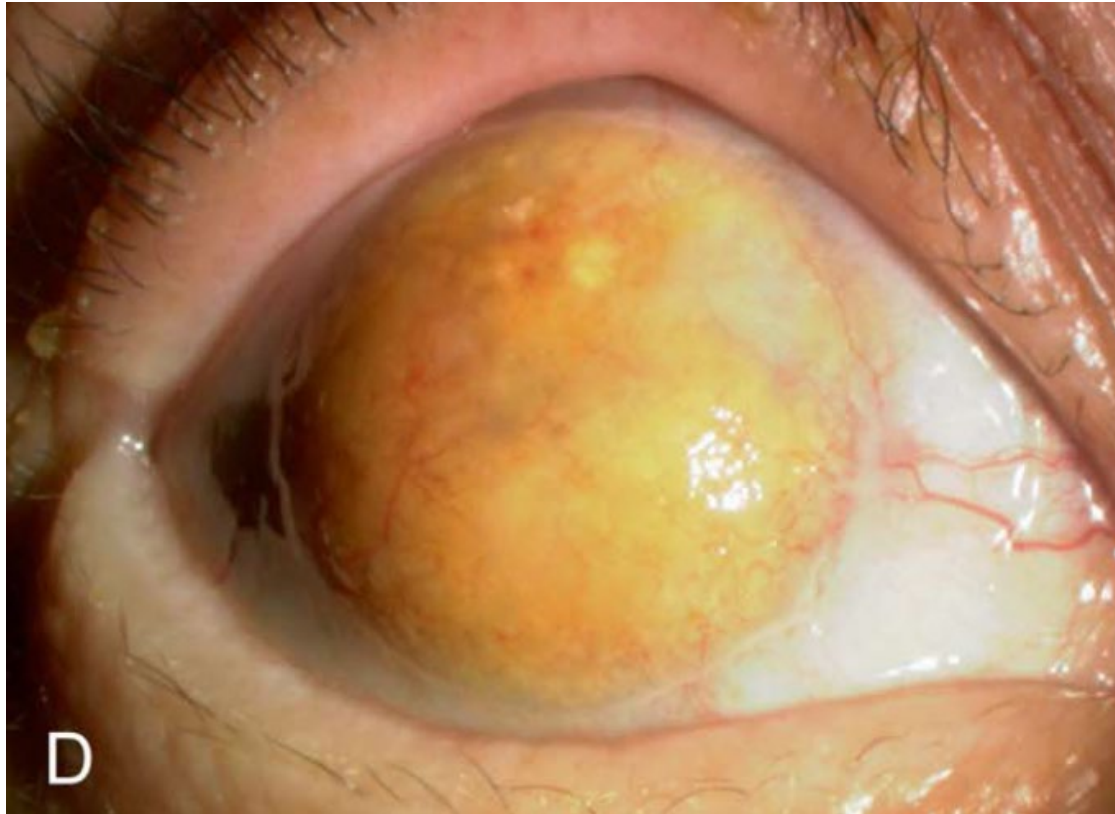
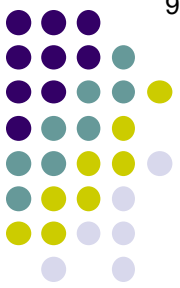
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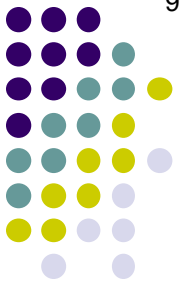
--As groups of nodules (aka '**mulberry cornea**')

Late in the process, the entire cornea becomes opaque--the so-called '**kumquat cornea**'



Gelatinous drop-like corneal dystrophy. Advanced disease: Kumquat-like diffuse stromal opacity.

Corneal Dystrophies



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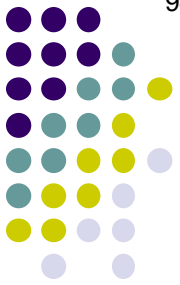
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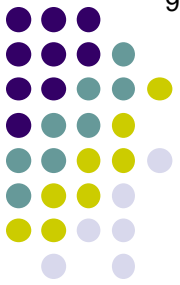
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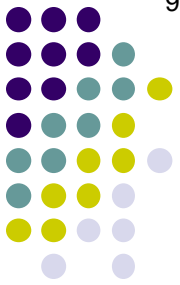
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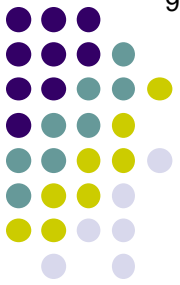
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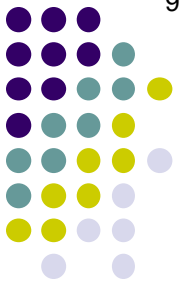
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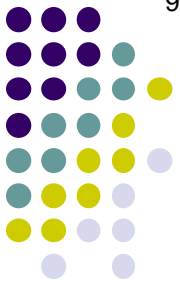
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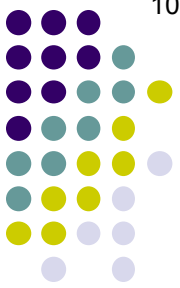
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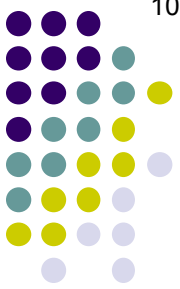
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Almost inevitably

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