What are the four categories of corneal dystrophies?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal \textit{TGFBI} Dystrophies

Stromal Dystrophies

Endothelial Dystrophies

What are the four categories of corneal dystrophies?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

Endothelial Dystrophies

*What are the six epithelial/subepithelial 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 *TGFB1* Dystrophies

Stromal Dystrophies

Endothelial 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

These two are not discussed in detail in the BCSC Cornea book, and thus will not be in this slide-set either.

Epithelial-Stromal TGFB1 Dystrophies

Stromal Dystrophies

Endothelial 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 are three other common names for EBMD?
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Epithelial and Subepithelial Dystrophies

1) **Epithelial basement membrane dystrophy**
2) Meesmann epithelial corneal dystrophy
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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
Epithelial and Subepithelial Dystrophies

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

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

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**What is the slit-lamp appearance of EBMD?**
Epithelial and Subepithelial Dystrophies

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What are three other common names for EBMD?
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What is the slit-lamp appearance of EBMD?
It can present in any combo of the following:
--Thin, curvy, hairlike lines that often occur in groups that follow the same highly curved course; these are called **fingerprint lines**. These represent abnormal basement membrane material.
--Thicker lines that follow a more jagged course; these are called **map lines**. These also result from abnormal basement membrane.
--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.
Epithelial and Subepithelial Dystrophies

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

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

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Epithelial basement membrane dystrophy. Map lines
**Epithelial and Subepithelial Dystrophies**

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Corneal Dystrophies

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Epithelial basement membrane dystrophy. Dots
Corneal Dystrophies

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

Epithelial and Subepithelial Dystrophies

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--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.
Epithelial basement membrane dystrophy. Typical appearance
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies
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What do pts complain of?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies
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--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 three words, then their abb.
Epithelial and Subepithelial Dystrophies

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

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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.
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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

1) Epithelial basement membrane dystrophy
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What are the major histologic abnormalities in EBMD?

--?
--?
--?

--Cogan’s microcystic dystrophy
--Anterior basement membrane dystrophy

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**What are the major histologic abnormalities in EBMD?**

--The epithelial cells are...

--Cogan’s microcystic dystrophy
--Anterior basement membrane dystrophy

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Epithelial and Subepithelial Dystrophies

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**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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**Corneal Dystrophies**

**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
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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

--Cogan’s microcystic dystrophy
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It can present in any combo of the following:
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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
--Anterior basement membrane dystrophy

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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.
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.
EBMD: Intraepithelial fibrillar material
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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What are three other common names for EBMD?
--Map-dot-fingerprint dystrophy
--Cogan’s microcystic dystrophy
--Anterior basement membrane dystrophy

How is EBMD inherited?
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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--Anterior basement membrane dystrophy

What is the slit-lamp appearance of EBMD?
It can present in any combo of the following:
--Thin, curvy, hairlike lines that often occur in groups that follow the same highly curved course; these are called **fingerprint lines**. These represent abnormal basement membrane material.
--Thicker lines that follow a more jagged course; these are called **map lines**. These also result from abnormal basement membrane.
--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.

What are the ‘6 F’s’ of EBMD?
--**Female**
--**Fifties and older** (usually)
--**Fifteen percent of the population are afflicted**
--**Fifty percent of pts who suffer REE have it**
--**Fibrillar material accumulates under the basement membrane**
Epithelial and Subepithelial Dystrophies

1) Epithelial basement membrane dystrophy
2) Meesmann epithelial corneal dystrophy
3) Lisch epithelial corneal dystrophy
4) Gelatinous droplike corneal dystrophy

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?

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

Are these microcysts evenly distributed?

Fairly so, with a modest tendency to be most numerous in the interpalpebral zone

Are the cysts visually disabling?

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

Electron microscopy indicates the presence of granular and/or filamentary debris within epithelial cells, which has to date eluded precise identification. By what name is this debris known?

It is called 'peculiar substance'
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Meesmann corneal dystrophy appearing as multiple dull-gray opacities
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A, Meesmann corneal dystrophy, appearing as tiny bubblelike blebs with indirect slit-lamp illumination. B, Blebs are also well seen against the red reflex
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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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Very rarely
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When symptomatic, what intervention is adequate in most cases?
Soft contact lens usage
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*At what age does LECD begin to manifest?*
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That sounds just like Meesman's. How do you distinguish between the two?
Unlike Meesman's, in which cysts are found everywhere on the cornea, in LECD the cysts are concentrated in discrete band- or feather-shaped lesions.

Are the cysts visually disabling?
Only if they happen to block the visual axis.

Are they painful?
No.

Do recurrent epithelial erosions occur in this condition?
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Lisch epithelial corneal dystrophy: Feather-shaped lesions
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In one word (not ‘dystrophy,’ duh), what sort of condition is GDCD?
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.
Gelatinous drop-like corneal dystrophy.
B, Congo Red stain. D, Polarized light reveals apple-green birefringence
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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

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--In a **band keratopathy**-like fashion
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Late in the process, the entire cornea becomes opaque--the so-called ‘**kumquat cornea**’

Another finding is that of **epithelial hyperpermeability**--the normal tight junctions between epi cells are absent. This means the lesions will stain with fluorescein even in the absence of an epi defect.

Is it visually disabling and/or painful?
It is both, eventually

How is it managed?
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Epithelial and Subepithelial Dystrophies

1) Epithelial basement membrane dystrophy
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A, Band keratopathy type. B, Mulberry type.
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**Gelatinous drop-like corneal dystrophy.** Advanced disease: Kumquat-like diffuse stromal opacity.
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**Does GDCD recur after surgery?**
Almost inevitably

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