What are the four categories of corneal dystrophies?
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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal $TGFBI$ Dystrophies

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

Endothelial Dystrophies

What are the six non-$TGFBI$ stromal dystrophies?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

What are the six non-TGFBI stromal dystrophies?
Stromal Dystrophies

1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
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Corneal Dystrophies

What is the inheritance pattern for macular dystrophy (MCD)?

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

Endothelial Dystrophies
Stromal Dystrophies

1) Macular corneal dystrophy
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Endothelial Dystrophies

Corneal Dystrophies

What is the inheritance pattern for macular dystrophy (MCD)?
AR

At what age does MCD begin to manifest?
Childhood (the corneas are clear at birth)

How does it present at the slit lamp?
It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of guttata), and extend limbus-to-limbus.

Is it painful?
Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
**Stromal Dystrophies**

1. **Macular corneal dystrophy**
2. Schnyder corneal dystrophy
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**Corneal Dystrophies**

*What is the inheritance pattern for macular dystrophy (MCD)??*  
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*How does it present at the slit lamp??*  
It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of guttata), and extend limbus-to-limbus.

*Is it painful??*  
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*What is the histologic hallmark of MCD on light microscopy??*  
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Endothelial Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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Corneal Dystrophies

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How does it present at the slit lamp?

Stromal Dystrophies
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4) Fleck corneal dystrophy
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6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
**Macular corneal dystrophy**

1) What is the inheritance pattern for macular dystrophy (MCD)?
   **AR**

2) At what age does MCD begin to manifest?
   Childhood (the corneas are clear at birth)

3) How does it present at the slit lamp?
   It starts with gray-white flecks in the anterior stroma. The cornea between lesions is clear vs hazy.

4) Does it affect vision?
   Yes, severe impairment occurs in the teens to 20s.

5) What is the histologic hallmark of MCD on light microscopy?
   Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
1) Macular corneal dystrophy
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**What is the inheritance pattern for macular dystrophy (MCD)?**
AR

**At what age does MCD begin to manifest?**
Childhood (the corneas are clear at birth)

**How does it present at the slit lamp?**
It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy.
Macular corneal dystrophy. Early stage with few central macular opacities.
Stromal Dystrophies

1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
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Endothelial Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Is it painful?

Epithelial erosions are rare, so generally no.

Does it affect vision?

Yes, severe impairment occurs in the teens to 20s.

What is the histologic hallmark of MCD on light microscopy?

Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
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**Corneal Dystrophies**

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Is it painful?
Epithelial erosions are rare, so generally no

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What is the histologic hallmark of MCD on light microscopy?

Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
Macular corneal dystrophy. Later, opacities are found limbus to limbus.
Macular corneal dystrophy. Later, opacities are found limbus to limbus. Note that the intervening spaces between lesions are hazy as well.
Stromal Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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When I hear ‘guttata,’ I think two words

Stromal Dystrophies

Endothelial Dystrophies
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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*When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’*

Is it painful?
Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.

When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’

No, corneal edema does not occur
Stromal Dystrophies

1) Macular corneal dystrophy
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Corneal Dystrophies

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When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think two diff words.

Does it affect vision?
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What is the histologic hallmark of MCD on light microscopy?
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Epithelial-Stromal TGFBI Dystrophies

When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think ‘corneal edema.’

Stromal Dystrophies

1) Macular corneal dystrophy
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Endothelial Dystrophies
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Is corneal edema a manifestation of MCD?

When I hear 'guttata,' I think 'Fuchs dystrophy.' And when I think Fuchs dystrophy, I think 'corneal edema.'
**Stromal Dystrophies**

1) **Macular corneal dystrophy**
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**What is the inheritance pattern for macular dystrophy (MCD)?**

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It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of guttata) and extend limbus-to-limbus.

*When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think ‘corneal edema.’ Is corneal edema a manifestation of MCD?*

No, corneal edema does not occur

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**Epithelial and Subepithelial Dystrophies**

**Epithelial-Stromal TGFBI Dystrophies**

**Stromal Dystrophies**

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**Endothelial Dystrophies**
Stromal Dystrophies

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Endothelial Dystrophies

Corneal Dystrophies

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When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think ‘corneal edema.’ Is corneal edema a manifestation of MCD?
No, corneal edema does not occur.

When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think ‘corneal edema.’ Is corneal edema a manifestation of MCD?
No, corneal edema does not occur.

Corneal thickness is impacted as well. Does MCD cause the cornea to thicken, or to thin?

Yes, severe impairment occurs in the teens to 20s.

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
Stromal Dystrophies

1) Macular corneal dystrophy
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Corneal thickness is impacted as well. Does MCD cause the cornea to thicken, or to thin?
To thin

Epithelial-Stromal TGFBI Dystrophies

When I hear ‘guttata,’ I think ‘Fuchs dystrophy.’ And when I think Fuchs dystrophy, I think ‘corneal edema.’ Is corneal edema a manifestation of MCD?
No, corneal edema does not occur

Epithelial and Subepithelial Dystrophies

What is the inheritance pattern for macular dystrophy (MCD)?
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At what age does MCD begin to manifest?
Children (the corneas are clear at birth)

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Is it painful?
Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
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Is it painful?

Epithelial-Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
1) Macular corneal dystrophy
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Endothelial Dystrophies
Corneal Dystrophies

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Childhood (the corneas are clear at birth)

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Is it painful?
Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.

1) Macular corneal dystrophy
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Stromal Dystrophies

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Corneal Dystrophies

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Is it painful?
Epithelial erosions are rare

Epithelial-Stromal TGFBI Dystrophies

Endothelial Dystrophies
Stromal Dystrophies

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Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFB1 Dystrophies

Corneal Dystrophies

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Is it painful?
Epithelial erosions are rare, so generally no

Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.
<table>
<thead>
<tr>
<th>Corneal Dystrophies</th>
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<tbody>
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### Stromal Dystrophies
1) **Macular corneal dystrophy**
2) Schnyder corneal dystrophy
3) Congenital stromal dystrophy
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### Endothelial Dystrophies
Corneal Dystrophies

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Childhood (the corneas are clear at birth)

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Is it painful?
Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs

Epithelial-Dystrophies

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Endothelial Dystrophies
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Endothelial Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Corneal Dystrophies

- What is the inheritance pattern for macular dystrophy (MCD)?
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  Epithelial erosions are rare, so generally no

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Stromal Dystrophies

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Endothelial Dystrophies
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**Corneal Dystrophies**

**What is the inheritance pattern for macular dystrophy (MCD)?**

AR

**At what age does MCD begin to manifest?**

Childhood (the corneas are clear at birth)

**How does it present at the slit lamp?**

It starts with gray-white flecks in the anterior stroma. The cornea *between* lesions is *hazy*. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of *guttata*), and extend *limbus-to-limbus*.

**Is it painful?**

Epithelial erosions are *rare*, so generally no

**Does it affect vision?**

Yes, severe impairment occurs *in the teens to 20s*
Corneal Dystrophies

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Abnormal mucopolysaccharides (aka glycosaminoglycans, GAGs) at all levels of the cornea; they stain with alcian blue.

Macular corneal dystrophy

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
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2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
**Corneal Dystrophies**

**What is the inheritance pattern for macular dystrophy (MCD)?**
AR

**At what age does MCD begin to manifest?**
Childhood (the corneas are clear at birth)

**How does it present at the slit lamp?**
It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of guttata), and extend limbus-to-limbus.

**Is it painful?**
Epithelial erosions are rare, so generally no

**Does it affect vision?**
Yes, severe impairment occurs in the teens to 20s

**What is the histologic hallmark of MCD on light microscopy?**
Abnormal

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**Stromal Dystrophies**

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What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka another long word) at all levels of the cornea

Endothelial Dystrophies
### Corneal Dystrophies

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**Macular corneal dystrophy.** Intracellular and extracellular accumulation of mucopolysaccharides (GAGs) at all levels of stroma and corneal endothelium. Subepithelial fibrous tissue also contains GAGs. Stain: Alcian blue
Stromal Dystrophies

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Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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Take note of this aka! The term mucopolysaccharide is considered outdated—glycosaminoglycan is the preferred nomenclature. This is reflected in recent editions of the Cornea book, which refer to MCD as a condition of defective glycosaminoglycan production, not mucopolysaccharide production. (The import of this shift in terminology will be made clear shortly.)

Endothelial Dystrophies

(No question—proceed when ready)
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**What is a glycosaminoglycan (GAG), and what is it doing in the cornea?**

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**Epithelial-Stromal Dystrophies**

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Epithelial-Stromal Dystrophies

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Endothelial Dystrophies

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Macular Corneal Dystrophy

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Three GAGs are present in the corneal stroma—what are they?
--?
--?
--?

Keratan sulfate

What’s wrong with it?
It isn’t properly sulfated

Glycosaminoglycans
Stromal Dystrophies

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--Chondroitin sulfate
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Good question. But before we address it, let’s back up a step…

The corneal stroma consists of three basic components. What are they?
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Which one ain’t right in MCD?

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What’s wrong with it?
It isn’t properly sulfated
What is the inheritance pattern for macular dystrophy (MCD)?
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At what age does MCD begin to manifest?
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Corneal Dystrophies

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Endothelial Dystrophies

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Good, but not 100% (there are subtypes of MCD that will not test positive)

Will the test be positive prior to the onset of corneal changes?
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**Stromal Dystrophies**

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Endothelial Dystrophies
Stromal Dystrophies

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Corneal Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Macular corneal dystrophy
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Corneal Dystrophies

What is the inheritance pattern for macular dystrophy (MCD)?
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How does it present at the slit lamp?
It starts with gray-white flecks in the anterior stroma. The cornea between lesions is hazy. The lesions spread to involve the full thickness of the stroma (and can even involve the endothelium in the form of guttata), and extend limbus-to-limbus.

The Cornea book stresses four characteristics that distinguish MCD from other stromal dystrophies:
--
--
--
--

What is the histologic hallmark of MCD on light microscopy?
Abnormal mucopolysaccharides (aka glycosaminoglycans) at all levels of the cornea; they stain with alcian blue.

Epithelial-Stromal TGFBI Dystrophies

Epithelial erosions are rare, so generally no

Does it affect vision?
Yes, severe impairment occurs in the teens to 20s

(No question yet—keep going)
Corneal Dystrophies

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--It’s inherited AR (most corneal dystrophies are AD)
--It involves the entire stroma (most pick a layer and stay there)
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Corneal Dystrophies

Endothelial Dystrophies

You’re probably familiar with the well-known mnemonic regarding the corneal dystrophies:

**Marilyn Monroe Always Gets Her Man in LA County**

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It’s now time to address this…

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Which helps us remember the name, abnormal material, and stain for each of the ‘Big 3’ stromal dystrophies:

**Marilyn**  macular dystrophy  
**Monroe**  mucopolysaccharide  
**Always**  Alcian blue  
**Gets**  granular dystrophy  
**Her**  hyaline  
**Man**  Masson trichrome  
**in**  lattice dystrophy  
**L**  amyloid  
**A**  Congo Red

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You’re probably familiar with the well-known mnemonic regarding the corneal dystrophies:

Marilyn Monroe Always Gets Her Man in LA County

Which helps us remember the name, abnormal material, and stain for each of the ‘Big 3’ stromal dystrophies:
Marilyn macular dystrophy
Monroe mucopolysaccharide
Always Alcian blue

But it’s not perfect:
The problem is readily apparent—the mnemonic only works if the abnormal material in MCD is called ‘mucopolysaccharide.’ So either modify the mnemonic to include GAGs (tweet your mods to me @EyeDentistAAO), or (gasp!) actually learn it.

A amyloid
County Congo Red

6) Pre-Descemet corneal dystrophy

(No question—keep going)
Stromal Dystrophies

1) Macular corneal dystrophy
2) **Schnyder corneal dystrophy**
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
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**What was the former name of this condition?**

Schnyder crystalline corneal dystrophy

**Why was the name changed?**

Only ~50% manifest corneal crystals

**What is the fundamental pathology in SCD?**

It is a localized disorder of lipid metabolism

**At what age does SCD begin to manifest?**

In the first year of life (but it often goes undiagnosed for many years)

**What is seen at the slit lamp?**

Early, in the disease, the cornea displays either a central opaque 'disc,' or central crystals. Later, arcus lipoides forms, and as the disease progresses, the cornea becomes more and more opaque.

**Is it painful?**

Generally no

**Does it affect vision?**

Yes--glare eventually becomes disabling

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

Phospholipids that stain with Oil red O

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Endothelial Dystrophies
Stromal Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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**Corneal Dystrophies**

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Epithelial and Subepithelial Dystrophies

**Epithelial-Stromal TGFBI Dystrophies**

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Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophies
Schnyder corneal dystrophy.
Early (<age 23 years):
Noncrystalline (A) and crystalline (B) forms.
Stromal Dystrophies

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Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFBI** Dystrophies

Stromal Dystrophies

Endothelial Dystrophies
Schnyder corneal dystrophy.
D, As dz progresses, arcus lipoides develops.
F, As the dz progresses further, midperipheral haze appears, and worsens throughout life (pt in [F] is 72)
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Phospholipids that stain with Oil red O

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**Phospholipids** that stain with **Oil red O**
Schnyder corneal dystrophy. Light microscopy—Oil Red O stains innumerable tiny lipid droplets red within the corneal stroma. Note also the spaces in the subepithelial and Bowman’s region.
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Endothelial Dystrophies

At what age does CSCD begin to manifest?

Birth (duh, it's congenital)

What is seen at the slit lamp?

Limbus-to-limbus, uniformly distributed haze.

On close inspection, innumerable white flaky opacities are present.

Is it progressive?

Generally no, or only modestly so

Is it painful?

No

Does it affect vision?

Yes, it results in significant visual loss

What is the histologic hallmark of CSCD on light microscopy?

Pronounced thickening of the corneal stroma with separation of corneal lamellae
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**Congenital stromal corneal dystrophy.** Diffuse clouding with flake-like opacities throughout the stroma in a 4-year old patient.
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Epithelial-Stromal TGFB1 Dystrophies

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What is the histologic hallmark of CSCD on light microscopy? Pronounced thickening of the corneal stroma with separation of corneal lamellae
**Congenital stromal corneal dystrophy.**
Light microscopy: the cornea is markedly thickened with stromal lamellae that are separated from each other in a regular manner.
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Endothelial Dystrophies

*The scenario of an infant with cloudy corneas should immediately bring to mind a mnemonic. Which one?*

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Endothelial Dystrophies
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The scenario of an infant with cloudy corneas should immediately bring to mind a mnemonic. Which one? STUMPED

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

Birth (duh, it’s congenital)

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Yes, it results in significant visual loss

What is the histologic hallmark of CSCD on light microscopy?
Pronounced thickening of the corneal stroma with separation of corneal lamellae

Stromal Dystrophies
1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
Stromal Dystrophies

1) Macular corneal dystrophy
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Endothelial Dystrophies

**Corneal Dystrophies**

Epithelial and Subepithelial Dystrophies

The scenario of an infant with cloudy corneas should immediately bring to mind a mnemonic. Which one? **STUMPED**

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

**Birth (duh, it’s congenital)**

*What is seen at the slit lamp?*
Limbus-to-limbus, uniformly distributed haze.

On close inspection, innumerable white flaky opacities are present.

*Is it progressive?*
Generally no, or only modestly so

*Is it painful?*
No

*Does it affect vision?*
Yes, it results in significant visual loss

*What is the histologic hallmark of CSCD on light microscopy?*
Pronounced thickening of the corneal stroma with separation of corneal lamellae
Corneal Dystrophies

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

- Sclerocornea; Stromal dystrophy (CHSD)
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED); Elevated IOP (ie, congenital glaucoma)
- Dermoid of the cornea

*Fill in the entities embedded in the mnemonic*
(Note: There are two Ss and two Es)
Next we will touch on distinguishing among CHSD, CHED and primary congenital glaucoma by highlighting key differences in their presentations.

- **Sclerocornea; Stromal dystrophy (CHSD)**
- **Trauma (endothelial; ie, from forceps)**
- **Ulcer**
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In CHSD, the cornea is modestly thickened by the presence of the material that causes the cloudiness.

*(No question—advance when ready)*
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The CCT is dramatically increased in CHED because of edema 2ndry to lack of adequate endothelial barrier and deturgescence function.

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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 is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?

Buphthalmos

What does buphthalmos translate to in English?

'Ox's eye'
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Congenital glaucoma: Increased corneal diameter
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'Buphthalmos' translates to 'Ox's eye'
Congenital glaucoma: Buphthalmos OD
Corneal Dystrophies

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Why is corneal diameter increased in congenital glaucoma? The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?

- Breaks in Descemet’s membrane (and its overlying endothelial layer)
- Horizontally
- Haab’s striae
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Increased
Horizontal Descemet’s breaks (*Haab’s striae*) in congenital glaucoma
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<td>Mildly increased</td>
<td>WNL</td>
<td>WNL</td>
<td>?</td>
</tr>
<tr>
<td>Primary congenital glaucoma</td>
<td>Variably increased (or WNL, or thin)</td>
<td>Increased</td>
<td>Duh</td>
<td>?</td>
</tr>
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</table>
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.

<table>
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<tr>
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<th>CCT</th>
<th>Corneal diameter</th>
<th>IOP</th>
<th>Tearing/Photophobia?</th>
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<tr>
<td>CHED</td>
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<td>WNL</td>
<td>WNL</td>
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1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

At what age does FCD begin to manifest?

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

Endothelial Dystrophies
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Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

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Endothelial Dystrophies

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*At what age does FCD begin to manifest?*

Very early—can even be congenital
Corneal Dystrophies

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1) Macular corneal dystrophy
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Epithelial-Stromal *TGFBI* Dystrophies

At what age does FCD begin to manifest?
Very early—can even be congenital

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

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At what age does FCD begin to manifest?
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How does it present? What is seen at the slit lamp?
Subtle light-gray discs in the stroma that have described as

Endothelial Dystrophies
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Epithelial-Stromal TGFBI Dystrophies

At what age does FCD begin to manifest?
Very early—can even be congenital

How does it present? What is seen at the slit lamp?
Subtle light-gray discs in the stroma that have described as 'dandruff-like.'
**Fleck corneal dystrophy.** Dandruff-like opacities seen in 2 different patients throughout the stroma using: (A) broad oblique illumination, and (B) at varying depths in the slit-lamp photograph.
Corneal Dystrophies

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Endothelial Dystrophies

At what age does FCD begin to manifest?
Very early—can even be congenital

How does it present? What is seen at the slit lamp?
Subtle light-gray discs in the stroma that have described as ‘dandruff-like.’ The intervening spaces are clear.
**Corneal Dystrophies**

**Epithelial and Subepithelial Dystrophies**

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Subtle light-gray discs in the stroma that have described as ‘dandruff-like.’ The intervening spaces are **clear**. The lesions are never found in non-stromal portions of the cornea.
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Subtle light-gray discs in the stroma that have described as ‘dandruff-like.’ The intervening spaces are clear. The lesions are never found in non-stromal portions of the cornea.

Is it painful?
Corneal Dystrophies

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Is it painful?
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Is it painful?
No

Does it affect vision?
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Is it painful?
No

Does it affect vision?
Usually not
First: *What sound-alike, more-familiar condition must you keep separate from PACD?*

1. Fleck corneal dystrophy
2. Posterior amorphous corneal dystrophy
3. Pre-Descemet corneal dystrophy

Endothelial Dystrophies
**First: What sound-alike, more-familiar condition must you keep separate from PACD?**

Posterior *poly*morphous corneal dystrophy. PPMD is an endothelial dystrophy, whereas PACD is a stromal (although it can affect the endothelium indirectly).

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5) *Posterior amorphous corneal dystrophy*

6) Pre-Descemet corneal dystrophy

---

Endothelial Dystrophies
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*Now then: At what age does PACD begin to manifest?*

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Now then: At what age does PACD begin to manifest? First decade. Can be present in infancy.

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First decade. Can be present in infancy.

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

---

1. Fleck corneal dystrophy
2. Pre-Descemet corneal dystrophy
3. Posterior amorphous corneal dystrophy
4. Pre-Descemet corneal dystrophy
**Corneal Dystrophies**

**First:** What sound-alike, more-familiar condition must you keep separate from PACD? Posterior polymeherphous corneal dystrophy. PPMD is an endothelial dystrophy, whereas PACD is a stromal (although it can affect the endothelium indirectly).

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1. Fleck corneal dystrophy
2. Posterior amorphous corneal dystrophy
3. Pre-Descemet corneal dystrophy

**Endothelial Dystrophies**
Posterior amorphous corneal dystrophy. Central deep stromal/pre-Descemet opacity with some degree of peripheral extension interrupted by few clear bands in the midperipheral cornea.
Corneal Dystrophies

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PACD is a dystrophy of the deep corneal stroma. Sheetlike opacities are present, and can be extensive. The deepest lesions can indent Descemet's and the endothelium. Further, the cornea tends to be both thinner and flatter than normal.
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*Is it painful?*

---

5) **Posterior amorphous corneal dystrophy**
6) Pre-Descemet corneal dystrophy
First: What sound-alike, more-familiar condition must you keep separate from PACD?
Posterior polymorphous corneal dystrophy. PPMD is an endothelial dystrophy, whereas PACD is a stromal (although it can affect the endothelium indirectly).

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PACD is a dystrophy of the deep corneal stroma. Sheetlike opacities are present, and can be extensive. The deepest lesions can indent Descemet’s and the endothelium. Further, the cornea tends to be both thinner and flatter than normal; as a result of the flatness, PACD pts are usually hyperopes.

Is it painful?
No

5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy
**Corneal Dystrophies**

**First**: What sound-alike, more-familiar condition must you keep separate from PACD? Posterior poly morphous corneal dystrophy. PPMD is an endothelial dystrophy, whereas PACD is a stromal (although it can affect the endothelium indirectly).

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Is it painful? No

Does it affect vision?

5) Poster-choral dystrophy
5) **Posterior amorphous corneal dystrophy**
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
**Corneal Dystrophies**

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**Is it painful?**
No

**Does it affect vision?**
Only mildly

---

4) Fleck corneal dystrophy
5) **Posterior amorphous corneal dystrophy**
6) Pre-Descemetary corneal dystrophy

**Endothelial Dystrophies**
**First:** What sound-alike, more-familiar condition must you keep separate from PACD?

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First decade. Can be present in infancy.

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

PACD is a dystrophy of the *deep* corneal stroma. Sheetlike opacities are present, and can be extensive. The deepest lesions can indent Descemet’s and the endothelium. Further, the cornea tends to be both *thinner* and *flatter* than normal; as a result of the flatness, PACD pts are usually *hyperopes*.

*Is it painful?*

No

*Does it affect vision?*

Only mildly

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

1. Fleck corneal dystrophy
2. Posterior amorphous corneal dystrophy
3. Pre-Descemet corneal dystrophy

**Endothelial Dystrophies**
**Corneal Dystrophies**

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PACD is a dystrophy of the **deep** corneal stroma. Sheetlike opacities are present, and can be extensive. The deepest lesions can indent Descemet’s and the endothelium. Further, the cornea tends to be both **thinner** and **flatter** than normal; as a result of the flatness, PACD pts are usually **hyperopes**.

**Is it painful?**
No

**Does it affect vision?**
Only mildly

**What is the histologic hallmark of PACD on light microscopy?**
Irregularities to the pre-Descemet’s deep stroma

---

**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**

TGFBI

**Dystrophies**

1) Reis-Bücklers corneal dystrophy
2) Thiel-Behnke corneal dystrophy
3) Lattice, type 1
4) Lattice, variant types (III, IIIA, I/IIIA, IV)
5) Granular type 1
6) Granular type 2

---

**Stromal Dystrophies**

1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

---

**Endothelial Dystrophies**
Posterior amorphous corneal dystrophy. Light microscopy—extracellular colloidal iron stains positive material (arrowheads) in the deep stroma
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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At what age does PDCD begin to manifest?

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Endothelial Dystrophies

At what age does PDCD begin to manifest?
Usually after age 30 years; rarely in childhood
Corneal Dystrophies

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At what age does PDCD begin to manifest?
Usually after age 30 years; rarely in childhood

What is seen at the slit lamp?
Fine punctate opacities just anterior to Descemet's

Endothelial Dystrophies
Pre-Descemet’s corneal dystrophy. A, With broadbeam illumination, punctate opacities anterior to Descemet membrane are apparent. B, Slit beam illumination of the same eye demonstrating punctate opacities anterior to Descemet membrane.
Corneal Dystrophies

1) Macular corneal dystrophy
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Stromal Dystrophies

At what age does PDCD begin to manifest?
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What is seen at the slit lamp?
Fine punctate opacities just anterior to Descemet’s

Is it painful?

Endothelial Dystrophies
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Is it painful?
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Usually after age 30 years; rarely in childhood

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Is it painful?
No

Does it affect vision?
**Stromal Dystrophies**

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Usually after age 30 years; rarely in childhood

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Fine punctate opacities just anterior to Descemet’s

**Is it painful?**
No

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No