Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal $TGFBI$ Dystrophies

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

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

Endothelial 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
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5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

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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 that are similar in appearance to those of GCD1; however, unlike GCD1, the spaces between lesions are hazy. The lesions quickly spread to involve the full thickness of the corneal stroma, and can involve Descemet’s and the endothelium (in the form of guttata) as well.

Is it painful?
Pts can get recurrent epithelial erosions, but generally do so at a much lower rate than is seen with other dystrophies.

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

What is the histologic hallmark of MCD on light microscopy?
The presence of mucopolysaccharides (aka glycosaminoglycans, or GAGs) at all levels of the cornea, that stains with Alcian Blue.
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies

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**Epithelial-Stromal TGFBI Dystrophies**
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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**Corneal Dystrophies**

Epithelial and Subepithelial Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal \textit{TGFB1} Dystrophies

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Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Endothelial Dystrophies
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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**Corneal Dystrophies**

**Epithelial and Subepithelial Dystrophies**

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**Endothelial Dystrophies**

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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 corneal 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
Stromal Dystrophies

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**Endothelial Dystrophies**

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### Epithelial and Subepithelial Dystrophies

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Yes--glare eventually becomes disabling

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Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

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**Phospholipids** that stain with Oil red O
Corneal Dystrophies

Epithelial and Subepithelial 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

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

Epithelial-Stromal *TGFBI* Dystrophies
Corneal Dystrophies

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Endothelial Dystrophies

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Endothelial Dystrophies

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Endothelial Dystrophies

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\textbf{Does it affect vision?}
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\textbf{What is the histologic hallmark of CSCD on light microscopy?}
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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1) Macular corneal dystrophy
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3) **Congenital stromal corneal dystrophy**
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Endothelial Dystrophies

At what age does CSCD begin to manifest?
Birth (duh, it’s congenital)

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Does it affect vision?
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Pronounced thickening of the corneal stroma with separation of corneal lamellae
Corneal Dystrophies

Epithelial and Subepithelial 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

At what age does FCD begin to manifest?

Epithelial-Stromal TGFBI Dystrophies
Epithelial and Subepithelial Dystrophies

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

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
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

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?
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

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 'something-like' The intervening spaces are clear. The lesions are never found in non-stromal portions of the cornea.
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

Epithelial-Stromal TGFBI Dystrophies

Corneal 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. The lesions are never found in non-stromal portions of the cornea.
Corneal Dystrophies

Epithelial and Subepithelial 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
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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.’ The intervening spaces are clear. The lesions are never found in non-stromal portions of the cornea.

Is it painful?

Endothelial 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

Epithelial-Stromal \textit{TGFB1} Dystrophies

Corneal 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 ‘\textit{dandruff-like}.’ The intervening spaces are clear. The lesions are never found in non-stromal portions of the cornea.

Is it painful?
No

Endothelial Dystrophies
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Stromal Dystrophies

1) Macular corneal dystrophy
2) Schnyder corneal dystrophy
3) Congenital stromal corneal dystrophy
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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. The lesions are never found in non-stromal portions of the cornea.

Is it painful? No

Does it affect vision?
**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**

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**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. The lesions are never found in non-stromal portions of the cornea.

**Is it painful?**
No

**Does it affect vision?**
Usually not
Corneal Dystrophies

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
Corneal Dystrophies

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

4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
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).

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

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5) Posterior amorphous corneal dystrophy

6) Pre-Descemet corneal dystrophy
**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).

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

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

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

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First: *What sound-alike, more-familiar condition must you keep separate from PACD?*
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*Now then: At what age does PACD begin to manifest?*
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.

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5) **Posterior amorphous corneal dystrophy**
6) Pre-Descemet corneal dystrophy
**First:** *What sound-alike, more-familiar condition must you keep separate from PACD?*

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Now then: *At what age does PACD begin to manifest?*
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*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?
Irregularities to the pre-Descemet's deep stroma

Endothelial Dystrophies

5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy
**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).

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

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.
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**Is it painful?**
No.
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*Is it painful?*
No
**Corneal Dystrophies**

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*Is it painful?*
No

*Does it affect vision?*

---

1) Fleck corneal 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

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5) Posterior amorphous corneal dystrophy

6) Pre-Descemet corneal dystrophy

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Endothelial Dystrophies
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*Is it painful?*
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*What is the histologic hallmark of PACD on light microscopy?*
1) Fleck corneal dystrophy  
5) **Posterior amorphous corneal dystrophy**  
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**Endothelial Dystrophies**
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**Is it painful?**
No

**Does it affect vision?**
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**What is the histologic hallmark of PACD on light microscopy?**

Irregularities to the pre-Descemet's deep stroma

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5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

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**Endothelial Dystrophies**
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
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Endothelial Dystrophies

At what age does PDCD begin to manifest?

Corneal Dystrophies

At what age does PDCD begin to manifest?

Corneal Dystrophies

At what age does PDCD begin to manifest?

Corneal Dystrophies

At what age does PDCD begin to manifest?
Stromal Dystrophies
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3) Congenital stromal corneal dystrophy
4) Fleck corneal dystrophy
5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

Epithelial and Subepithelial Dystrophies

Endothelial Dystrophies
Stromal Dystrophies

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

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

What is seen at the slit lamp?

Endothelial 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

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

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Endothelial Dystrophies
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Stromal 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

Is it painful?

Endothelial Dystrophies
Stromal Dystrophies

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5) Posterior amorphous corneal dystrophy
6) Pre-Descemet corneal dystrophy

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Usually after age 30 years; rarely in childhood

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

Is it painful?
No

Epithelial and Subepithelial Dystrophies

Corneal Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Endothelial Dystrophies
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies

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Fine punctate opacities just anterior to Descemet's

Is it painful?
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Does it affect vision?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

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Fine punctate opacities just anterior to Descemet’s

Is it painful?
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Does it affect vision?
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Endothelial Dystrophies