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

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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What are the six non-TGFB1 stromal dystrophies?

Endothelial Dystrophies
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Endothelial Dystrophies

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

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Endothelial Dystrophies
Macular corneal dystrophy. Early stage with few central macular opacities.
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Macular corneal dystrophy. More diffuse opacities and haze involving the entire stroma.
**Epithelial and Subepithelial Dystrophies**

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The presence of **mucopolysaccharides** (aka **glycosaminoglycans**, or **GAGs**) at all levels of the cornea, that stains with **Alcian Blue**.
**Macular corneal dystrophy.** Light microscopy—intracellular and extracellular accumulation of mucopolysaccharides (GAGs) at all levels of stroma and corneal endothelium. Subepithelial fibrous tissue also contains GAGs.
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Endothelial Dystrophies

Epithelial and Subepithelial Dystrophies

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

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## Endothelial Dystrophies

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## Epithelial and Subepithelial Dystrophies

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## Epithelial-Stromal TGFBI Dystrophies

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## Stromal Dystrophies

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

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### 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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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
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 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
**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.
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

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
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
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Endothelial Dystrophies

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

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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Endothelial Dystrophies

At what age does CSCD begin to manifest?
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What is seen at the slit lamp?
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Epithelial-Stromal TGFBI Dystrophies

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6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

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**Corneal 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.
Congenital stromal corneal dystrophy. Diffuse clouding with flake-like opacities throughout the stroma in a 4-year old patient.
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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5) Posterior amorphous corneal dystrophy
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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?

No
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

Endothelial Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

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Is it progressive?
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Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

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Birth (duh, it’s congenital)

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

**Is it painful?**
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

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Endothelial Dystrophies

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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
**Corneal Dystrophies**

**Epithelial and Subepithelial Dystrophies**

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

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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

**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**
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
1) Macular corneal dystrophy
2) Schnyder 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?
Yes, it results in significant visual loss
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
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Endothelial Dystrophies

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**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?**
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFβI 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
**Congenital stromal corneal dystrophy.**
Light microscopy: the cornea is markedly thickened with stromal lamellae that are separated from each other in a regular manner.
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 and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

At what age does FCD begin to manifest?

Stromal Dystrophies

Epithelial and Subepithelial Dystrophies

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 and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

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

Stromal Dystrophies

Endothelial Dystrophies
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

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

Epithelial-Stromal TGFBI Dystrophies

Endothelial Dystrophies
Corneal 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 and Subepithelial Dystrophies

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.

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

Stromal 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.
**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

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

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?

No

Usually not
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

Is it painful?
No
Stromal Dystrophies

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

Epithelial and Subepithelial 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?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
1) Macular corneal dystrophy
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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?
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
First: *What sound-alike, more-familiar condition must you keep separate from PACD?*

- Fleck corneal dystrophy
- **Posterior amorphous corneal dystrophy**
- 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).

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?*

5) Posterior amorphous corneal dystrophy

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies
**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).

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

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

**Endothelial Dystrophies**
**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).

*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?*

1) Fleck corneal dystrophy
2) **Posterior amorphous corneal dystrophy**
3) Pre-Descemet corneal dystrophy

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**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?
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.

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?*

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.
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.
**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?
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?
Irregularities to the pre-Descemet’s deep stroma

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

**Endothelial Dystrophies**
**Corneal 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? 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.
Posterior amorphous corneal dystrophy. Slit beam demonstrates decreased corneal thickness and posterior stromal lamellar opacification.
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? 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?

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?
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
### 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

### Corneal 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?

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?**

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?
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

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

Endothelial Dystrophies
Corneal 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? 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) Lisch epithelial corneal dystrophy
3) Lattice, type 1
4) Lattice, variant types (III, IIIA, I/IIIA, IV)
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?*

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?*

Irregularities to the pre-Descemet's deep stroma

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5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy
Posterior amorphous corneal dystrophy. Light microscopy—extracellular colloidal iron stains positive material (arrowheads) in the posterior stroma.
**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**

*At what age does PDCD begin to manifest?*
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

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

Epithelial-Stromal TGFBI Dystrophies

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
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?
Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies
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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.
Stromal Dystrophies

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**Is it painful?**
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