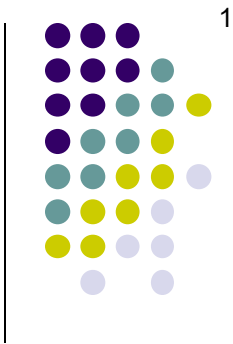


Corneal Dystrophies



?

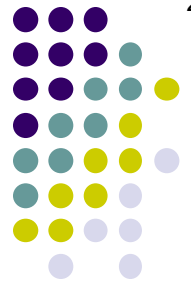
?

What are the four categories of corneal dystrophies?

?

?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

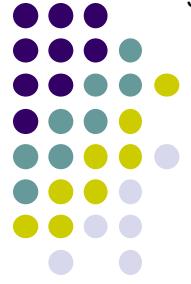
What are the four categories of corneal dystrophies?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies



Epithelial-Stromal *TGFBI* Dystrophies

What are the six non-TGFBI stromal dystrophies?

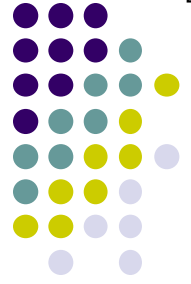
Stromal Dystrophies

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

Endothelial Dystrophies

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies



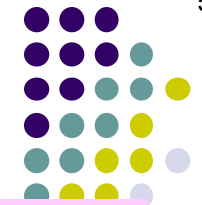
Epithelial-Stromal *TGFBI* Dystrophies

What are the six non-TGFBI stromal 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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

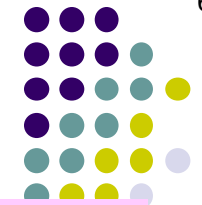
At what age does MCD begin to manifest?

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

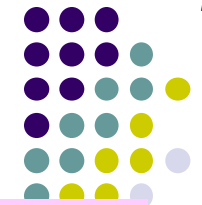
*At what age does MCD begin to manifest?
Childhood (the corneas are clear at birth)*

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

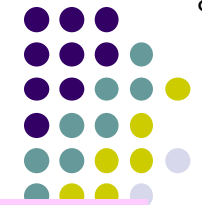
How does it present at the slit lamp?

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

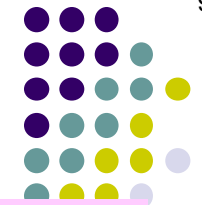
Stromal Dystrophies

- 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 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 .



Corneal Dystrophies

Epithelial and Subepithelial 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**.

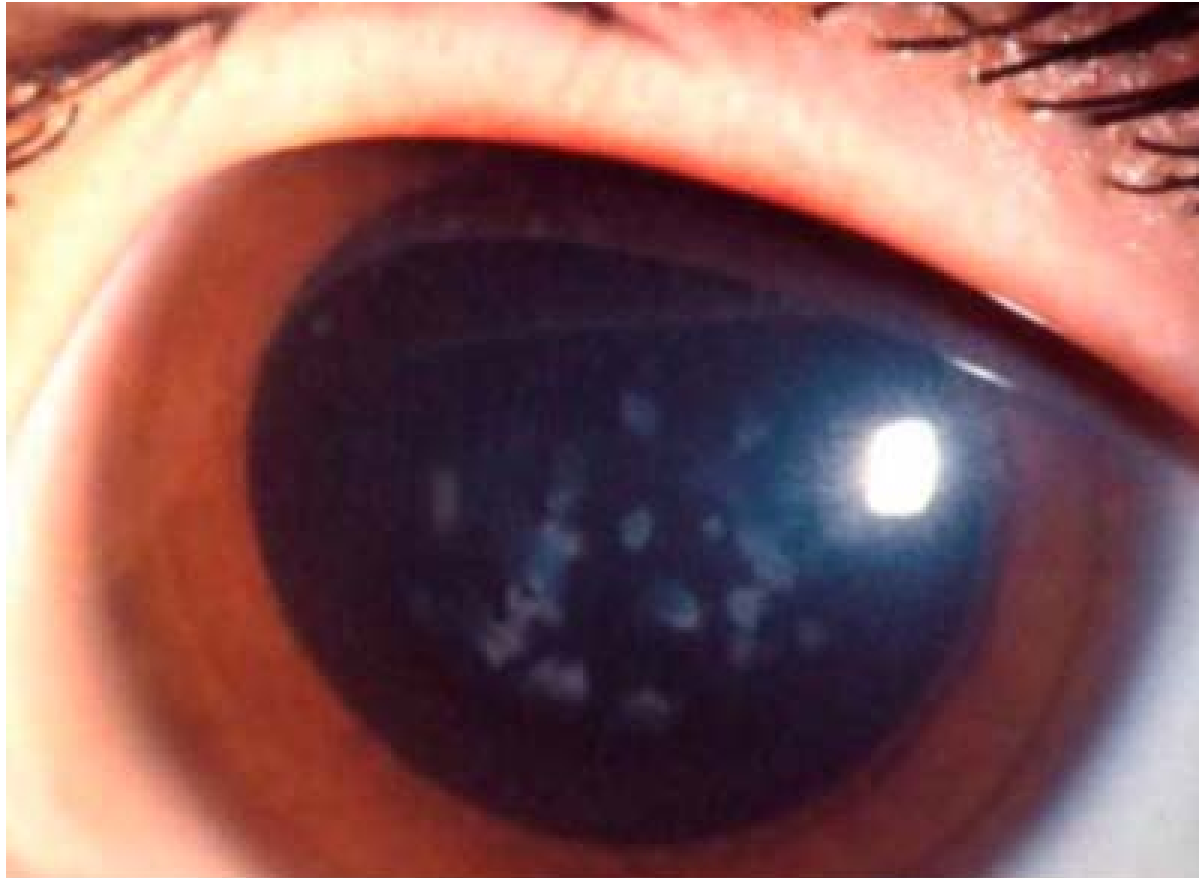
Epithelial-Stromal *TGFBI* Dystrophies

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

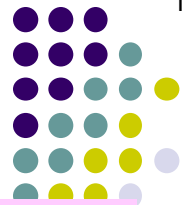
Endothelial Dystrophies

Corneal Dystrophies



Macular corneal dystrophy. Early stage with few central macular opacities.





Corneal Dystrophies

Epithelial and Subepithelial 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.

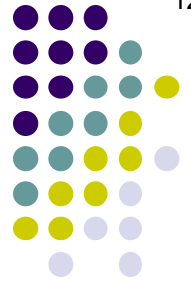
Epithelial-Stromal TGFBI Dystrophies

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

Corneal Dystrophies



Macular corneal dystrophy. More diffuse opacities and haze involving the entire 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 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?

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 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

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 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?

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 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

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 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?

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 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 one long word (aka another long word or its abb.) at all levels of the cornea, that stains with two words

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 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.

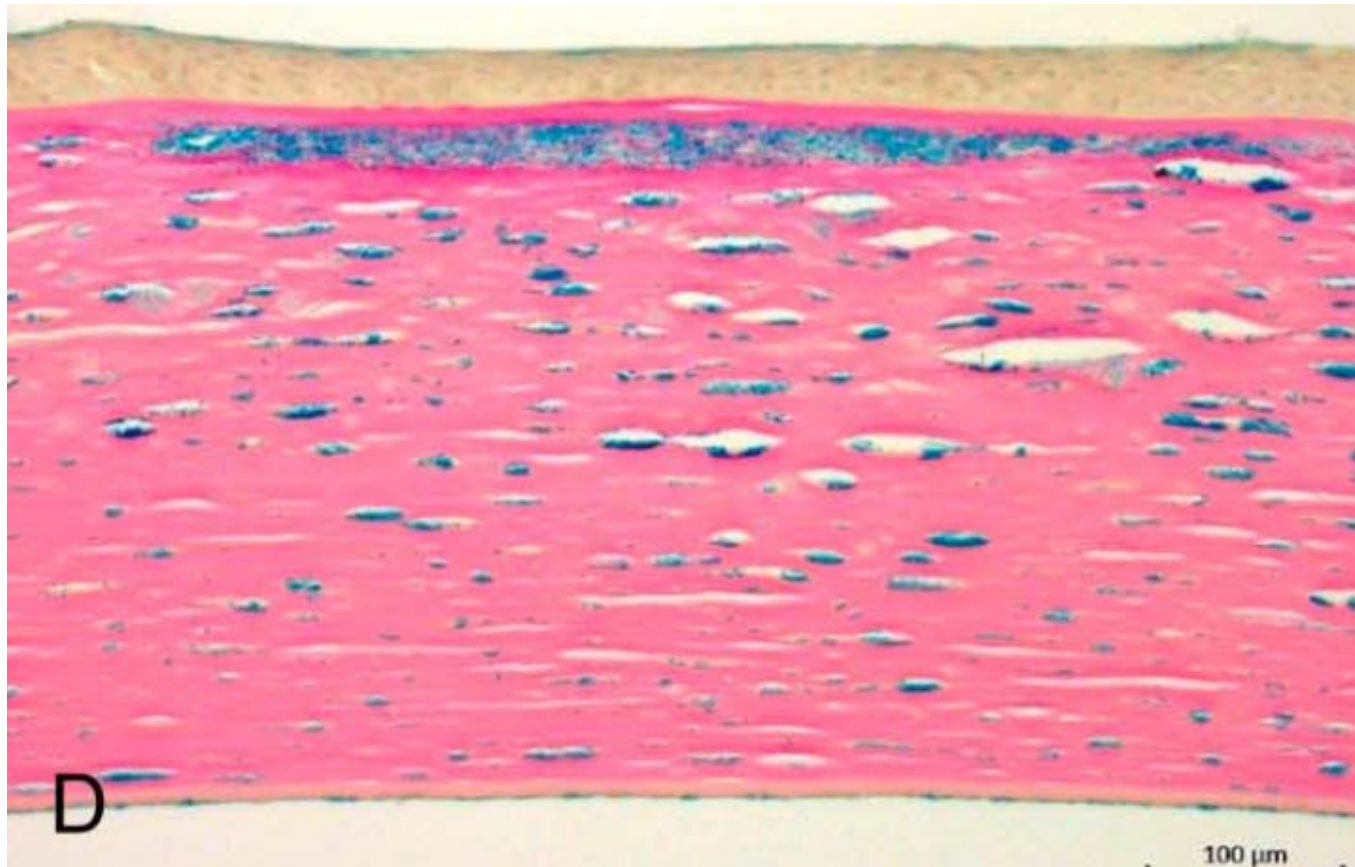
Is it painful?
Pts can get recurrent epithelial erosions, but generally do so at a much lower rate than is seen with other dystrophies

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

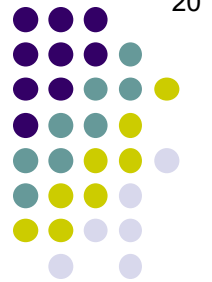
What is the histologic hallmark of MCD on light microscopy?

The presence of **mucopolysaccharides** (aka **glycosaminoglycans**, or **GAGs**) at all levels of the cornea, that stains with Alcian Blue.

Corneal Dystrophies



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.





Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

What was the former name of this condition?

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

What was the former name of this condition?
Schnyder **crystalline** corneal dystrophy

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

What was the former name of this condition?
Schnyder **crystalline** corneal dystrophy

Why was the name changed?

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



Corneal 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

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



Corneal 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?

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



Corneal 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

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



Corneal 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?

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



Corneal 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

Epithelial-Stromal *TGFBI* Dystrophies

At what age does SCD begin to manifest?
 In the first year of life (but it often goes undiagnosed for many years)

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

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

Epithelial-Stromal *TGFBI* Dystrophies

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?

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

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

Epithelial-Stromal *TGFBI* Dystrophies

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

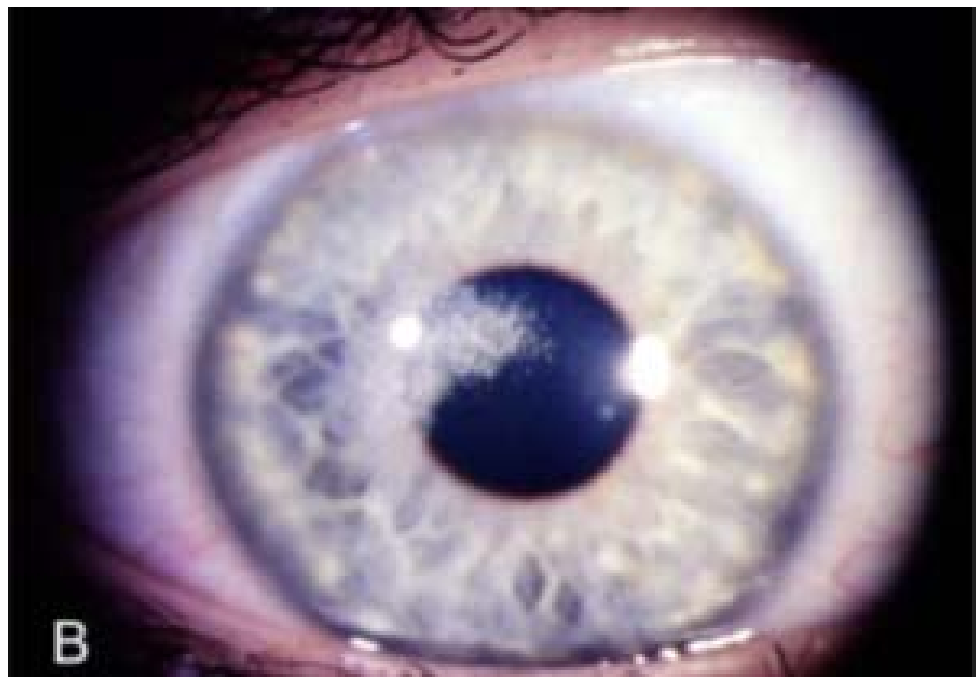
Stromal Dystrophies

- 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

Cornea

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





Corneal 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

Epithelial-Stromal *TGFBI* Dystrophies

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.

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

Corne**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)





Corneal 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?

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



Corneal 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

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



Corneal 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

Epithelial-Stromal *TGFBI* Dystrophies

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.

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

Is it painful?
Generally no

Does it affect vision?

Endothelial Dystrophies



Corneal 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

Epithelial-Stromal *TGFBI* Dystrophies

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.

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

Is it painful?
Generally no

Does it affect vision?
Yes--glare eventually becomes disabling

Endothelial Dystrophies



Corneal 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

Epithelial-Stromal *TGFBI* Dystrophies

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.

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

Is it painful?
Generally no

Does it affect vision?
Yes--glare eventually becomes disabling

Endothelial Dystrophies

What is the histologic hallmark of SCD on light microscopy?



Corneal 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?

one word

that stain with

three words

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



Corneal 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

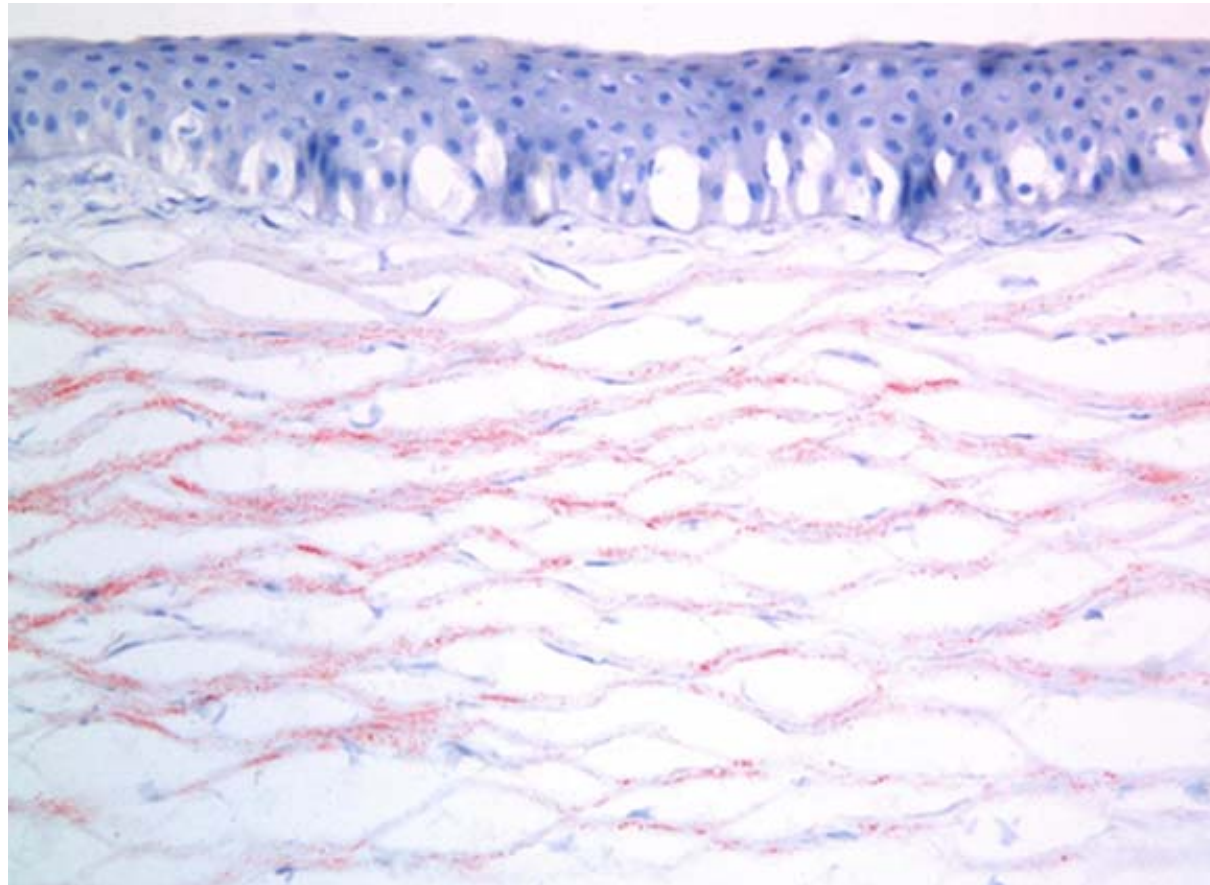
Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

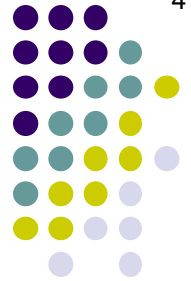
- 1) Macular corneal dystrophy
- 2) **Schnyder corneal dystrophy**
- 3) Congenital stromal corneal dystrophy
- 4) Fleck corneal dystrophy
- 5) Posterior amorphous corneal dystrophy
- 6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies



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?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

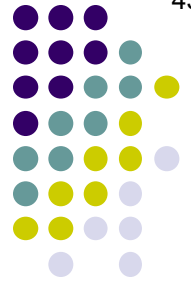
Epithelial-Stromal *TGFBI* Dystrophies

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)*





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?

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

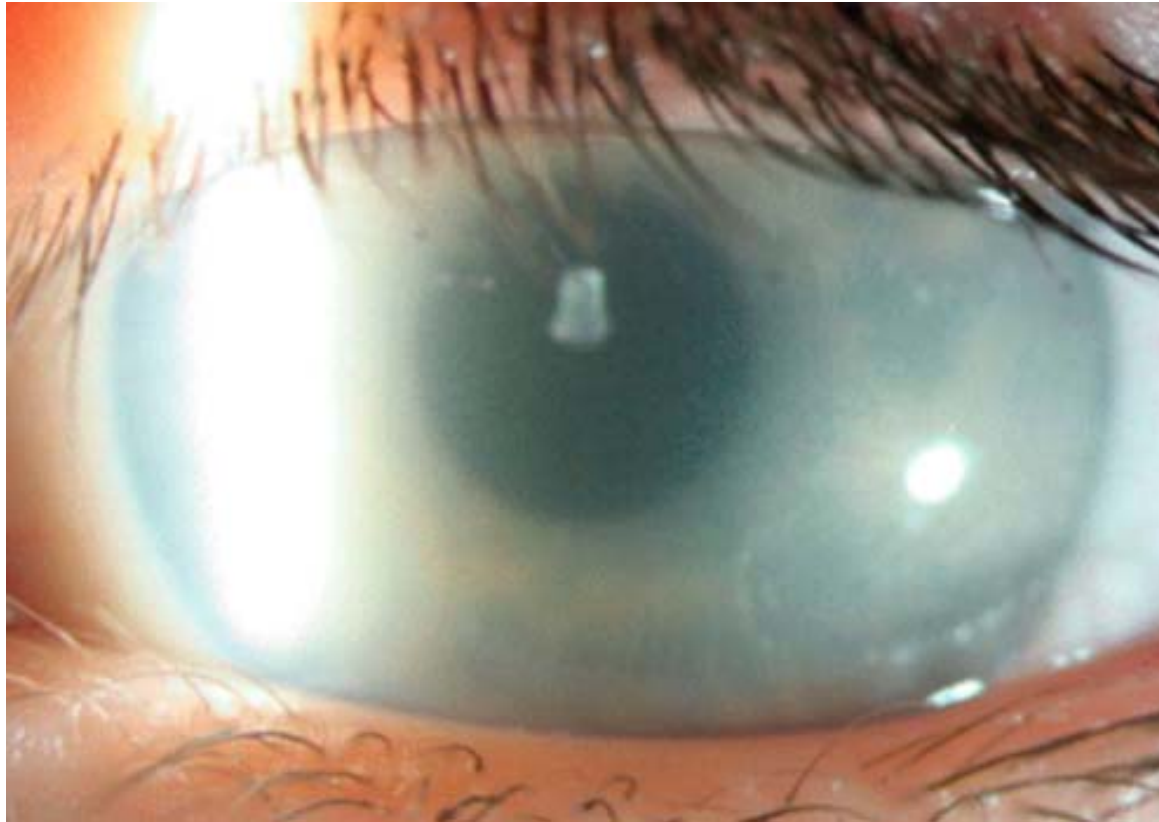
- 1) Macular corneal dystrophy
- 2) Schnyder corneal dystrophy
- 3) **Congenital stromal corneal dystrophy**
- 4) Fleck corneal dystrophy
- 5) Posterior amorphous corneal dystrophy
- 6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

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

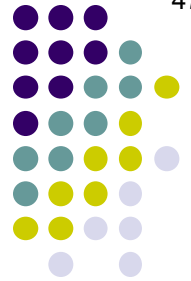
What is seen at the slit lamp?
Limbus-to-limbus, uniformly distributed haze.
On close inspection, innumerable white flaky opacities are present.

Corneal Dystrophies



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

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?



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

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

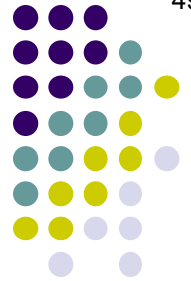
Endothelial Dystrophies

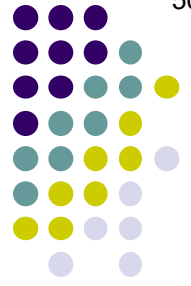
At what age does CSCD begin to manifest?
Birth (duh, it's congenital)

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?





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

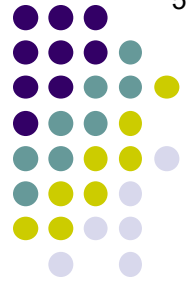
Endothelial Dystrophies

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



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?



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

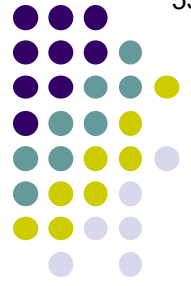
At what age does CSCD begin to manifest?
Birth (duh, it's congenital)

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



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?



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

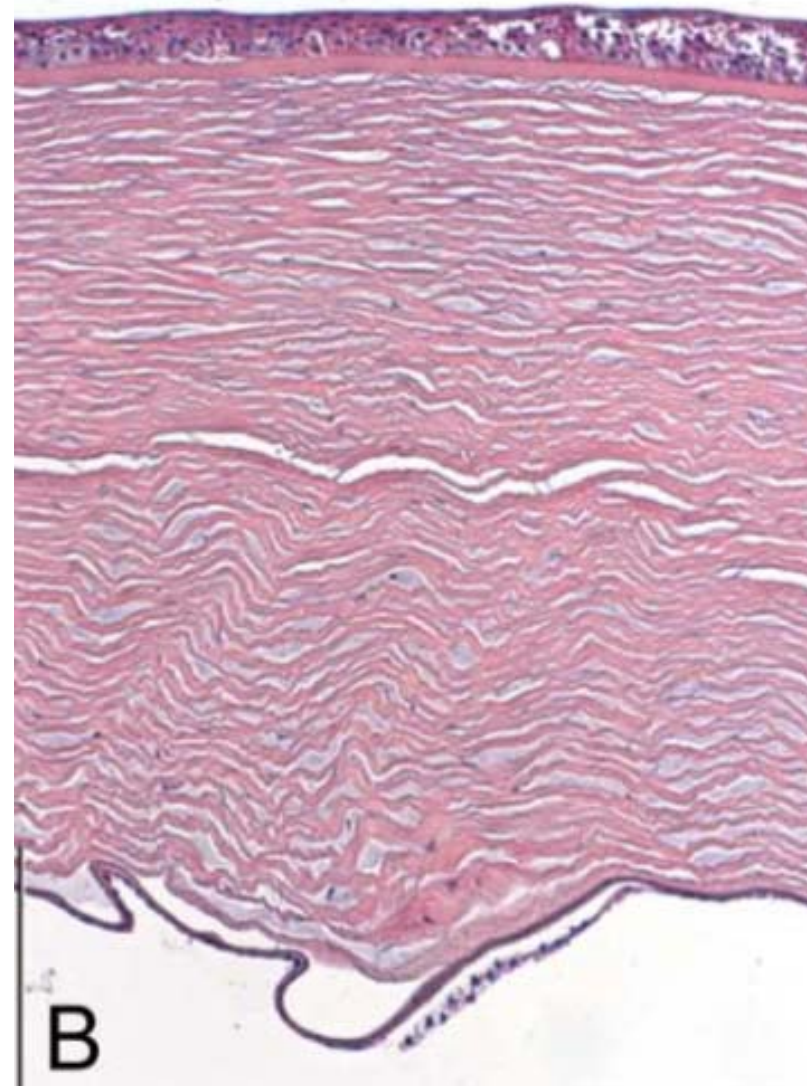
Is it painful?
No

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

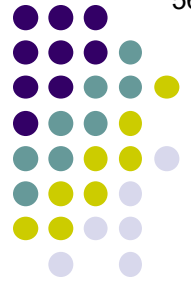
What is the histologic hallmark of CSCD on light microscopy?
Pronounced thickening of the corneal stroma with separation of corneal lamellae

Corneal Dystrophies

Congenital stromal corneal dystrophy.
Light microscopy: the cornea is markedly thickened with stromal lamellae that are separated from each other in a regular manner.



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 FCD begin to manifest?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* 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

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

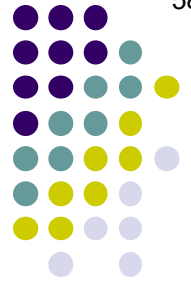
Stromal Dystrophies

- 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 FCD begin to manifest?
Very early—can even be congenital*

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



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies



Epithelial-Stromal *TGFBI* Dystrophies

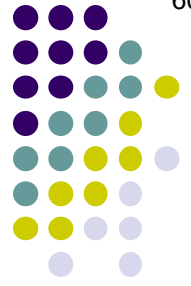
Stromal Dystrophies

- 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 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 so embarrassing



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

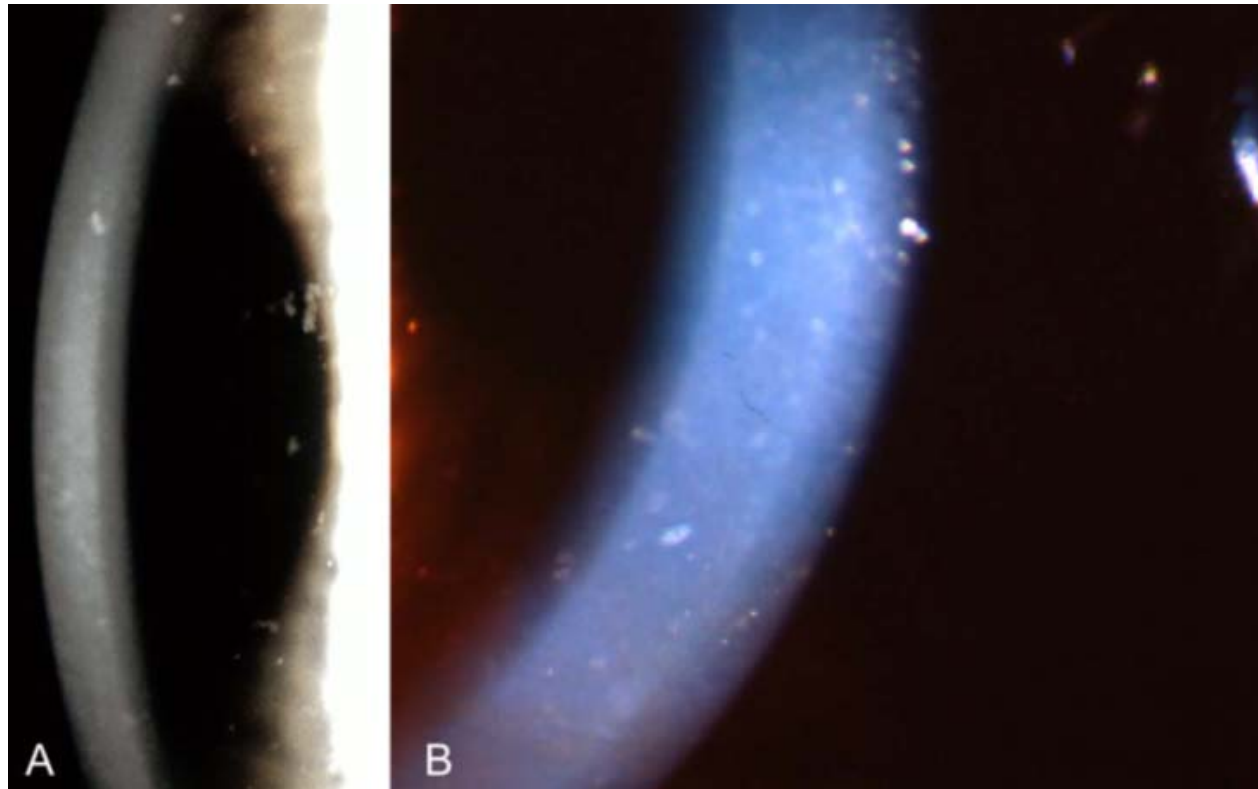
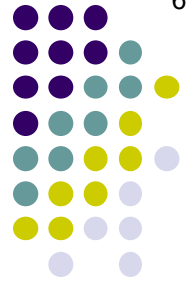
- 1) Macular corneal dystrophy
- 2) Schnyder corneal dystrophy
- 3) Congenital stromal corneal dystrophy
- 4) **Fleck corneal dystrophy**
- 5) Posterior amorphous corneal dystrophy
- 6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

At what age does 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.**'

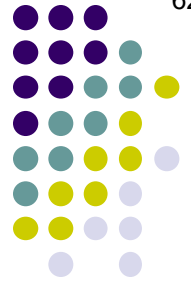
Corneal Dystrophies



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



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 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 hazy v
clear.

Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

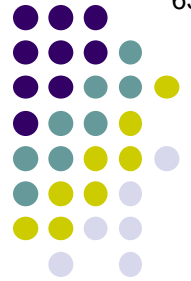
Stromal Dystrophies

- 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 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



Epithelial-Stromal *TGFBI* Dystrophies

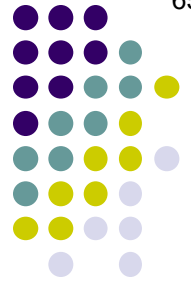
Stromal Dystrophies

- 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 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 v
always found in non-stromal portions of the cornea.



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

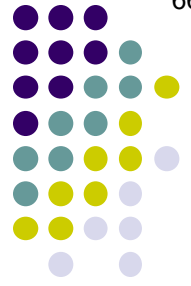
Stromal Dystrophies

- 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 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

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

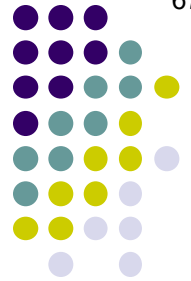
- 1) Macular corneal dystrophy
- 2) Schnyder corneal dystrophy
- 3) Congenital stromal corneal dystrophy
- 4) **Fleck corneal dystrophy**
- 5) Posterior amorphous corneal dystrophy
- 6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

At what age does 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?



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 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



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 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
- 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?
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?*

- 4) Fuchs 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).

- 4) Fick 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?

- 4) Fick 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.

- 4) Fick 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?

- 4) Fick 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 v
shallow corneal stroma.

4) Fick 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.

4) Fick 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.

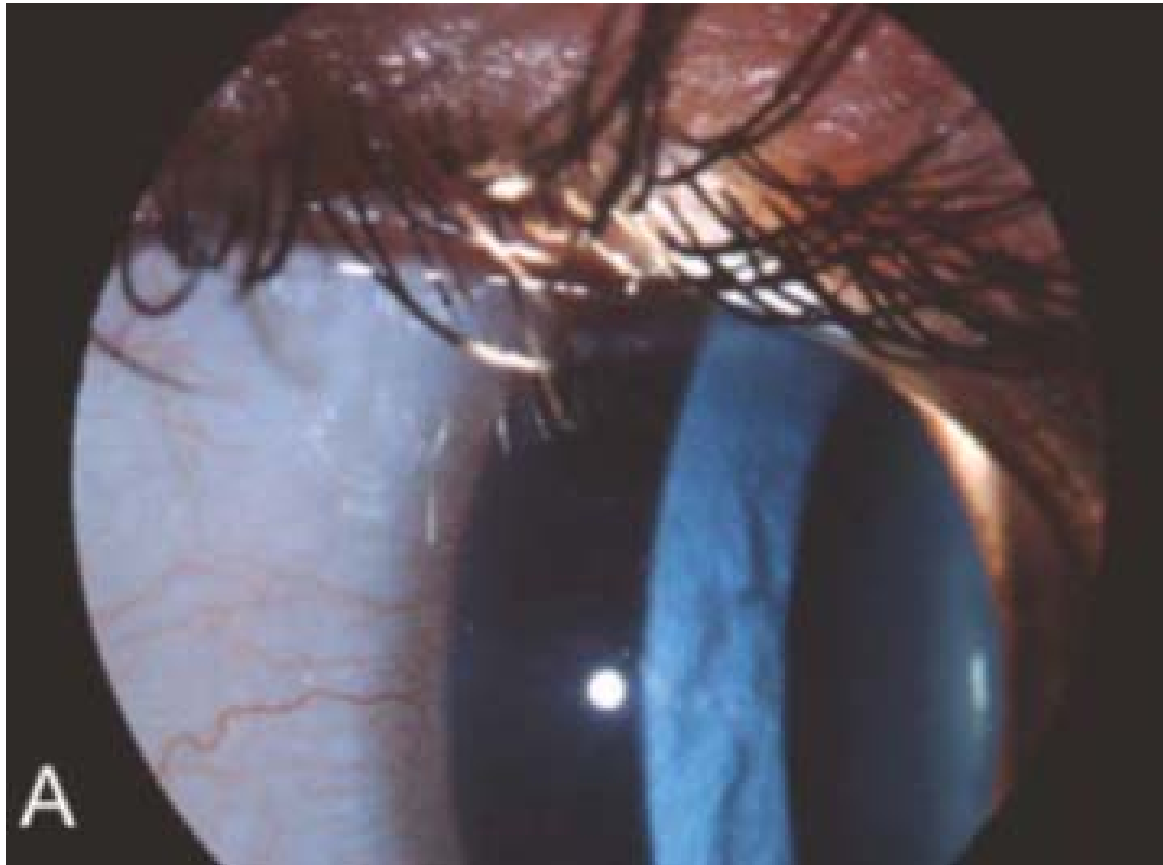
4) Fick corneal dystrophy

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal 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



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 vs
thicker

and

flatter v
steeper

than normal

1) Fick 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

4) Fick 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 refractive status.

4) Fick 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**.

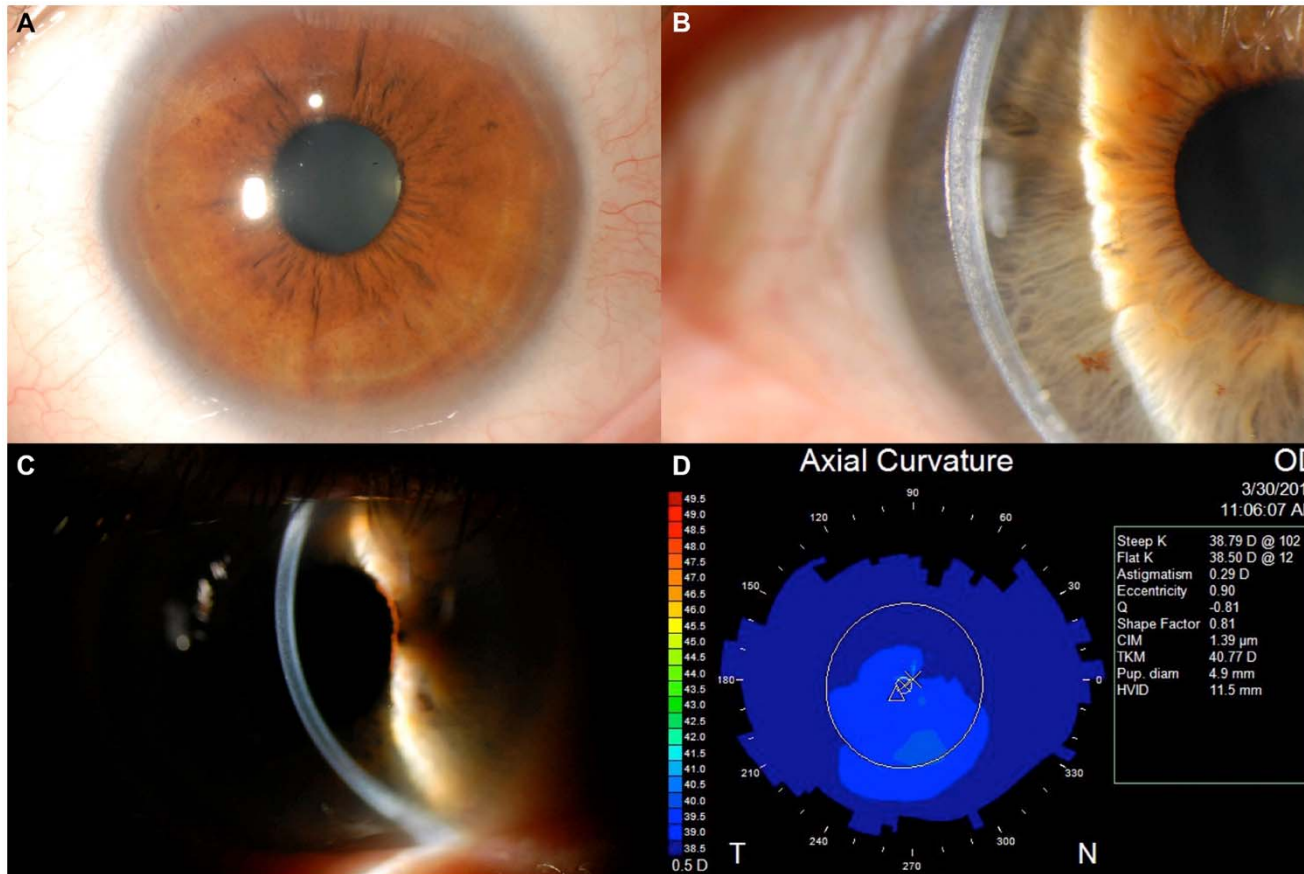
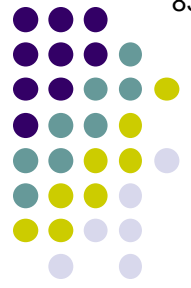
4) Fick corneal dystrophy

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies



Posterior amorphous corneal dystrophy. **A.** Slit lamp photomicrograph demonstrating peripheral corneal opacification. **B–C.** Slit lamp photomicrograph of central and peripheral corneal opacification **D.** Corneal topographic imaging demonstrates significant flattening of the corneal curvature (steep K 39D)

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?

4) Fick 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

1) Fick 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?

1) Fuchs 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

1) Fick 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) Fuchs corneal dystrophy
- 2) Keratic precipitates
- 3) Pre-Desemet corneal dystrophy
- 4) Posterior polymorphous corneal dystrophy
- 5) **Posterior amorphous corneal dystrophy**
- 6) Pre-Desemet 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

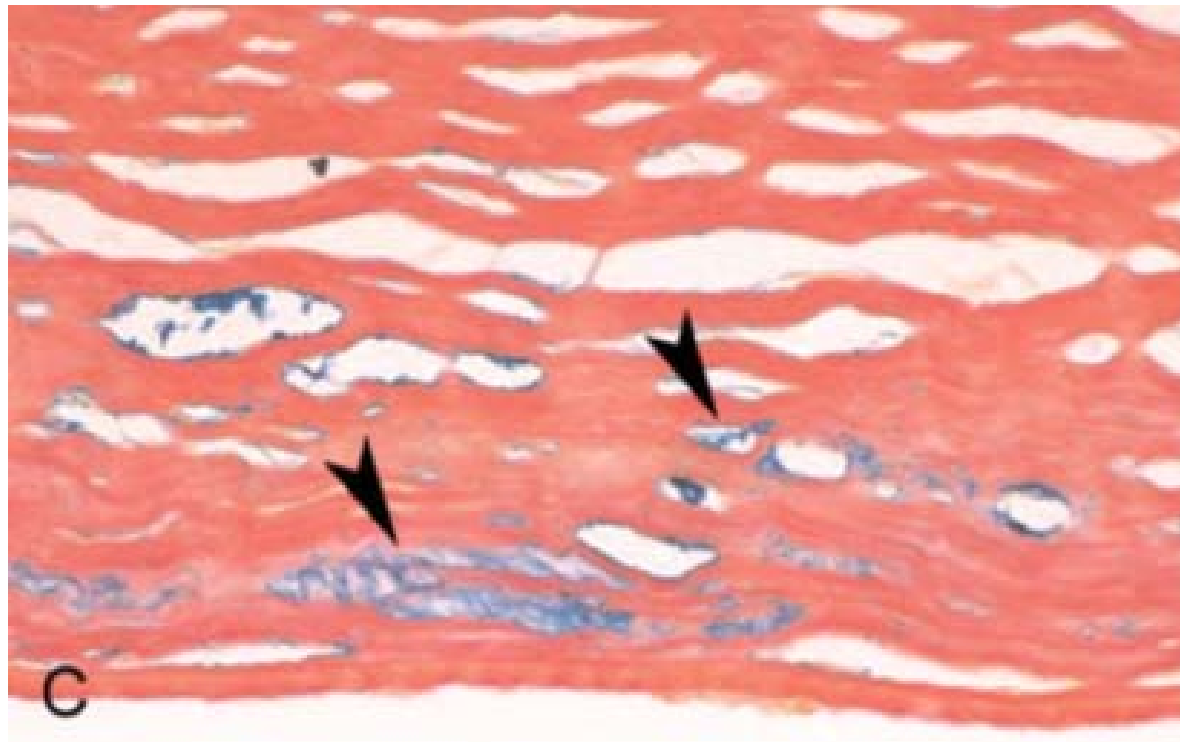
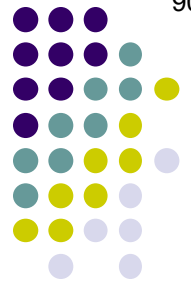
What is the histologic hallmark of PACD on light microscopy?

Irregularities to the pre-Descemet's deep stroma

- 1) Pre-Descemet corneal dystrophy
- 2) Posterior polymorphous corneal dystrophy
- 3) Granular corneal dystrophy
- 4) Lattice corneal dystrophy
- 5) **Posterior amorphous corneal dystrophy**
- 6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal 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

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

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?
Usually after age 30 years; rarely in childhood



Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

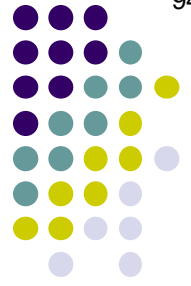
Stromal Dystrophies

- 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?
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

- 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**

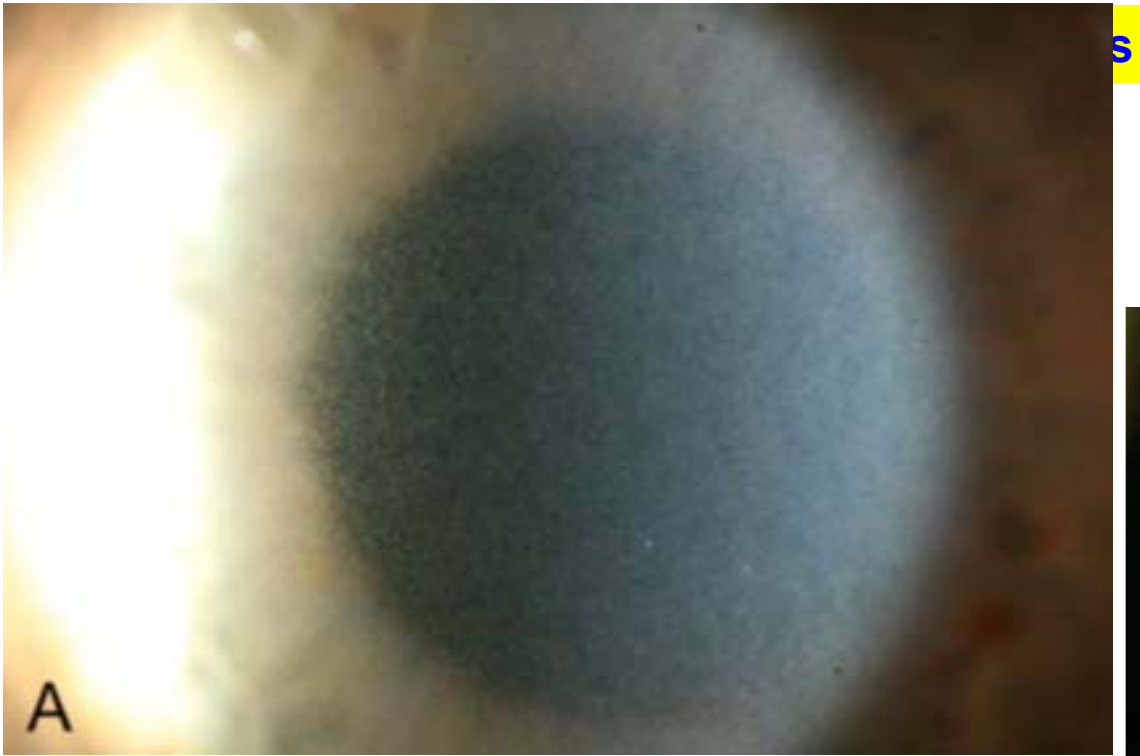
Endothelial Dystrophies

At what age does PDCD begin to manifest?

Usually after age 30 years; rarely in childhood

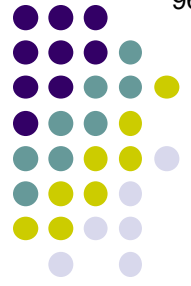
What is seen at the slit lamp?

Fine punctate opacities just anterior to Descemet's



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

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?

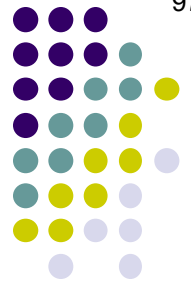
Usually after age 30 years; rarely in childhood

What is seen at the slit lamp?

Fine punctate opacities just anterior to Descemet's

Is it painful?

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?
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



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?

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

Does it affect vision?



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?

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

Does it affect vision?

No