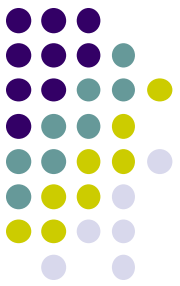


Corneal Dystrophies



?

?

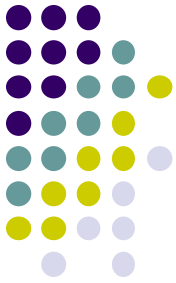
What are the four categories of corneal dystrophies?

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Corneal Dystrophies

2



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

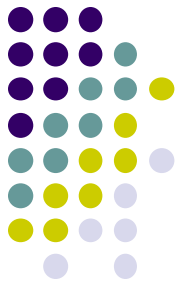
What are the four categories of corneal dystrophies?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies

3



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

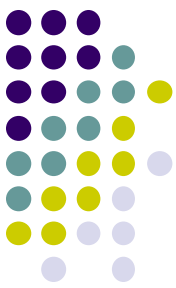
What are the six non-TGFB stromal dystrophies?

Stromal Dystrophies

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

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

What are the six non-TGFB stromal dystrophies?

Stromal Dystrophies

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

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dys

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

Epithelial-Stromal *TGFB* Dystro

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dys

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Epithelial-Stromal *TGFB* Dystro

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dys

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At what age does MCD begin to manifest?

Epithelial-Stromal *TGFB* Dystro

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

Epithelial-Stromal *TGFBI* Dystrophies

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dys

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How does it present at the slit lamp?

Epithelial-Stromal *TGFB* Dystro

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Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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

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

clear vs
hazy

Epithelial-Stromal TGFB1 Dystro

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

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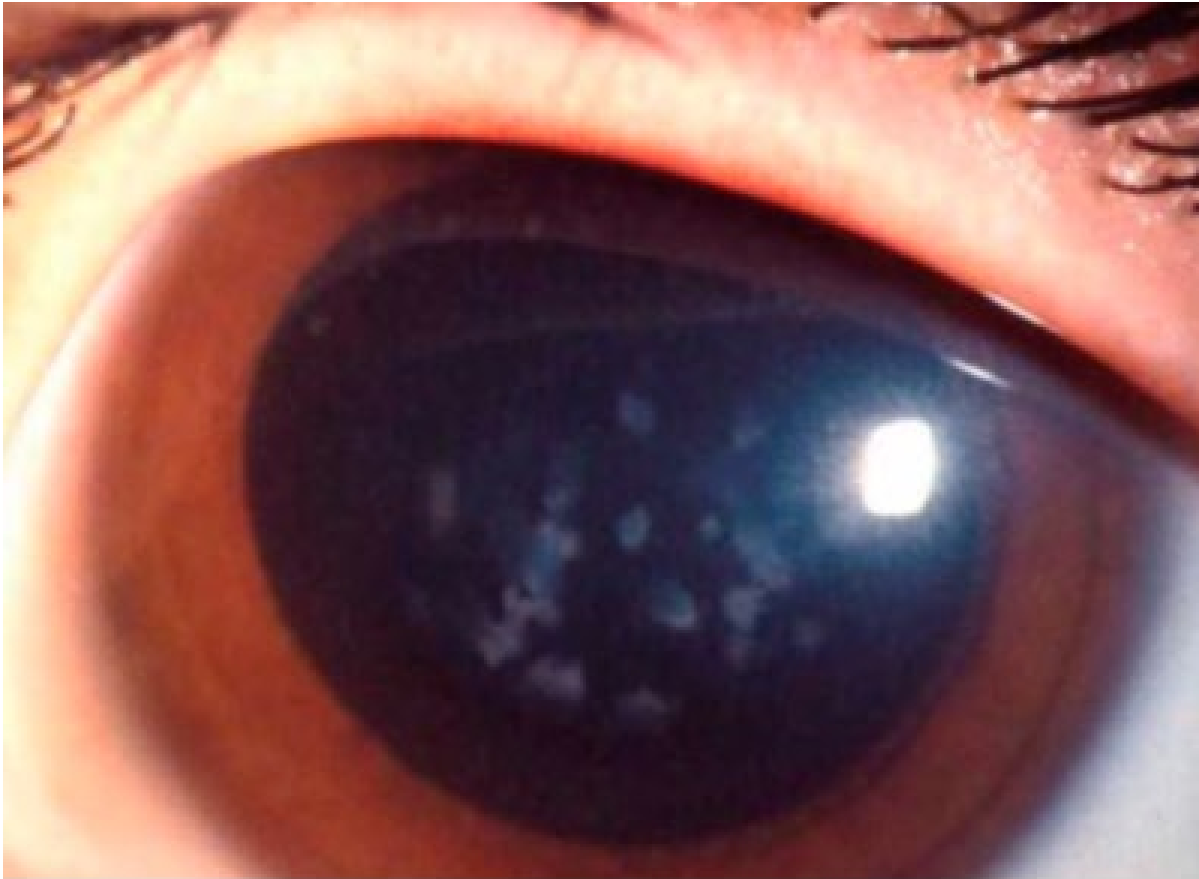
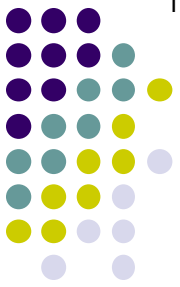
Epithelial-Stromal TGFB1 Dystro

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Endothelial Dystrophies

Corneal Dystrophies



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



Corneal Dystrophies


Epithelial and Subepithelial Dys

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Epithelial-Stromal TGFBI Dystro

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Epithelial-Stromal TGFBI Dystro

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where –to-where

Epithelial-Stromal TGFBI Dystro

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Endothelial Dystrophies



Corneal Dystrophies

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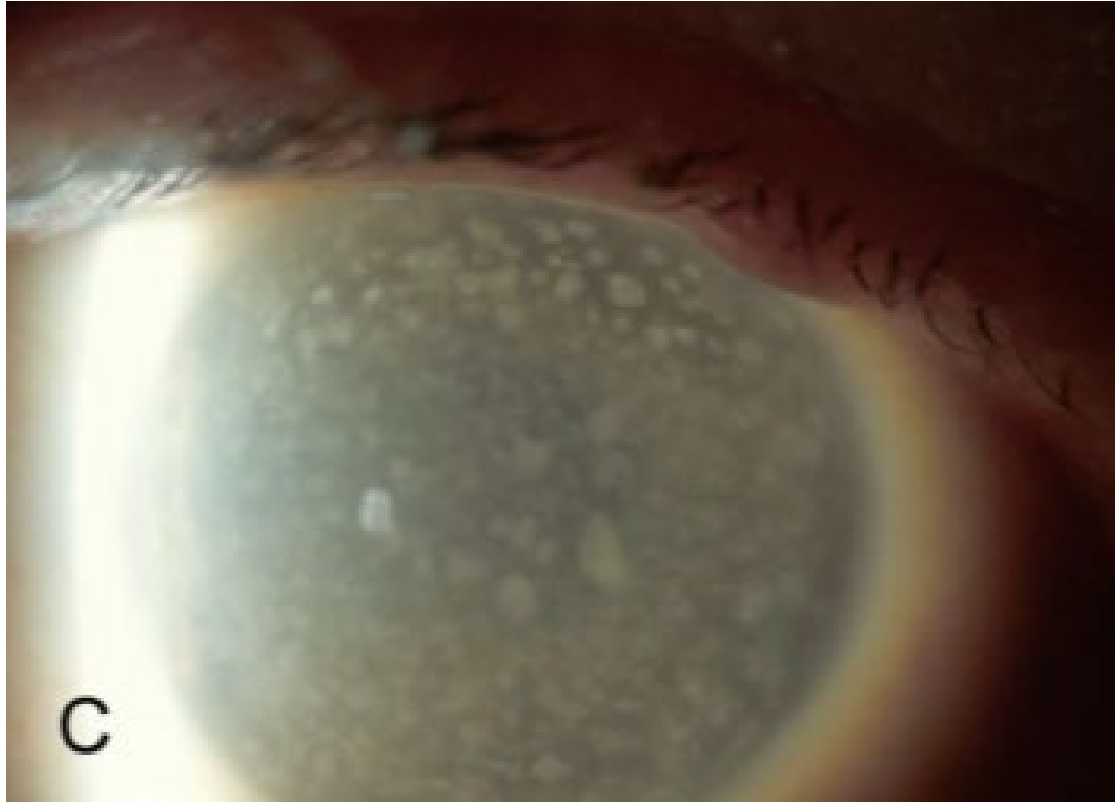
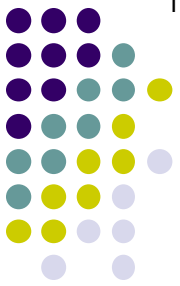
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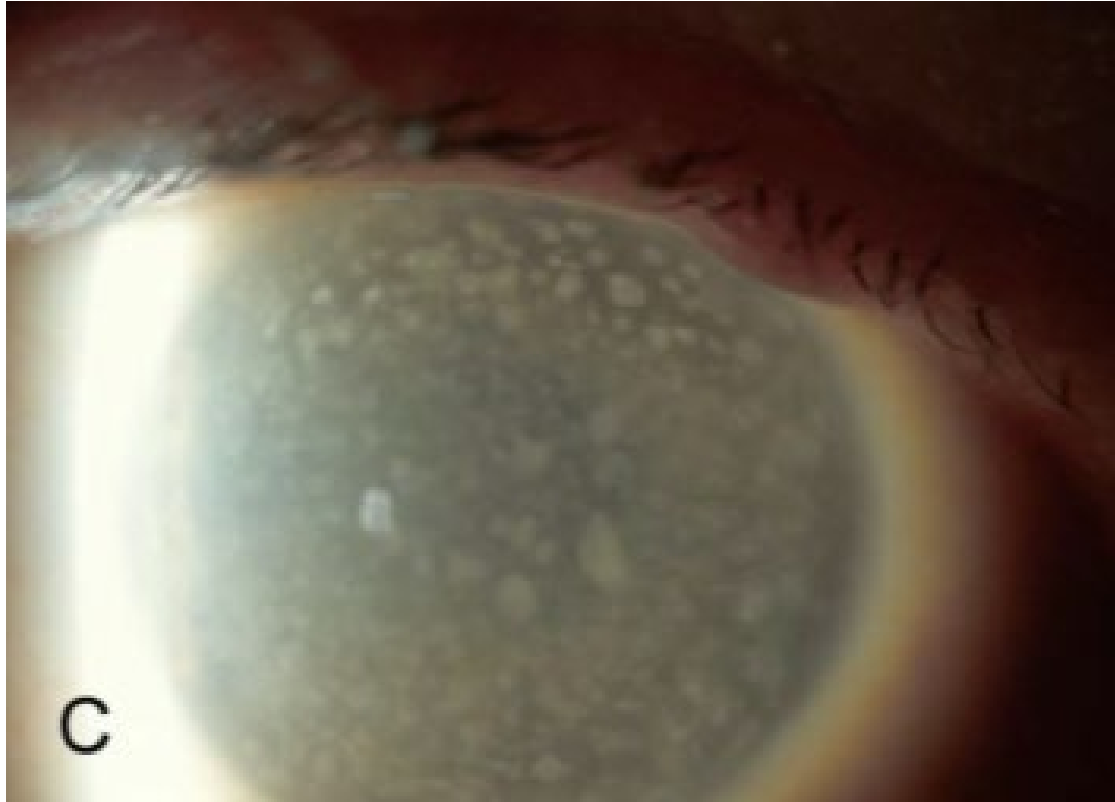
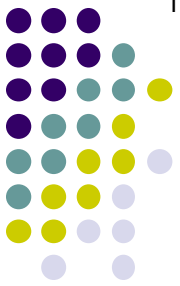
Endothelial Dystrophies

Corneal Dystrophies



Macular corneal dystrophy. Later, opacities are found limbus to limbus.

Corneal Dystrophies



Macular corneal dystrophy. Later, opacities are found limbus to limbus.
Note that the intervening spaces between lesions are hazy as well.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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

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

How does it present at the slit lamp?

Epithelial-Stromal TGFBI Dystrophy

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

When I hear 'guttata,' I think

two words

Stromal Dystrophies

- 1) **Macular corneal dystrophy**
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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophies

When I hear 'guttata,' I think 'Fuchs dystrophy.'

Stromal Dystrophies

- 1) **Macular corneal dystrophy**
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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophies

When I hear 'guttata,' I think 'Fuchs dystrophy.' And when I think Fuchs dystrophy, I think

two diff words

Stromal Dystrophies

- 1) **Macular corneal dystrophy**
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Endothelial Dystrophies



Corneal Dystrophies

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Epithelial-Stromal TGFBI Dystrophies

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

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophies

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

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophy

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

No, corneal edema does not occur

Stromal Dystrophies

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Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophy

When I hear 'guttata,' I think 'Fuchs dystrophy.' And when I think Fuchs dystrophy, I think 'corneal edema.' Is cornea No, corneal edema does not occur *Corneal thickness is impacted as well. Does MCD cause the cornea to thicken, or to thin?*

Stromal Dystrophies

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Corneal Dystrophies

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To thin

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Is it painful?

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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Is it painful?

Epithelial erosions are

rare vs
common

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Is it painful?

Epithelial erosions are rare

Epithelial-Stromal TGFBI Dystrophies

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Endothelial Dystrophies



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Is it painful?

Epithelial erosions are rare, so generally no

Epithelial-Stromal TGFBI Dystrophies

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Is it painful?

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Does it affect vision?

Epithelial-Stromal TGFBI Dystrophies

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Is it painful?

Epithelial erosions are rare, so generally no

Does it affect vision?

Yes, modest v severe impairment occurs

Stromal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

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Is it painful?

Epithelial erosions are rare , **so generally no**

Does it affect vision?

Yes, severe impairment occurs

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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Is it painful?

Epithelial erosions are rare, so generally no

Does it affect vision?

Yes, severe impairment occurs in the

age range

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

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Is it painful?

Epithelial erosions are rare, so generally no

Does it affect vision?

Yes, severe impairment occurs in the teens to 20s

Epithelial-Stromal TGFBI Dystrophies

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Is it painful?

Epithelial erosions are rare, so generally no

Does it affect vision?

Yes, severe impairment occurs in the teens to 20s

Epithelial-Stromal TGFBI Dystrophies

Stromal Dystrophies

1) **Macular corneal dystrophy**

2) Schnyder corneal dystrophy

3) Congenital stromal dystrophy

4) Fleck corneal dystrophy

5) Posterior amorphous dystrophy

6) Pre-Descemet corneal dystrophy

What is the histologic hallmark of MCD on light microscopy?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

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

How does it present at the slit lamp?

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Abnormal

one long word

Endothelial Dystrophies



Corneal Dystrophies

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Endothelial Dystrophies



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

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

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What is the histologic hallmark of MCD on light microscopy?
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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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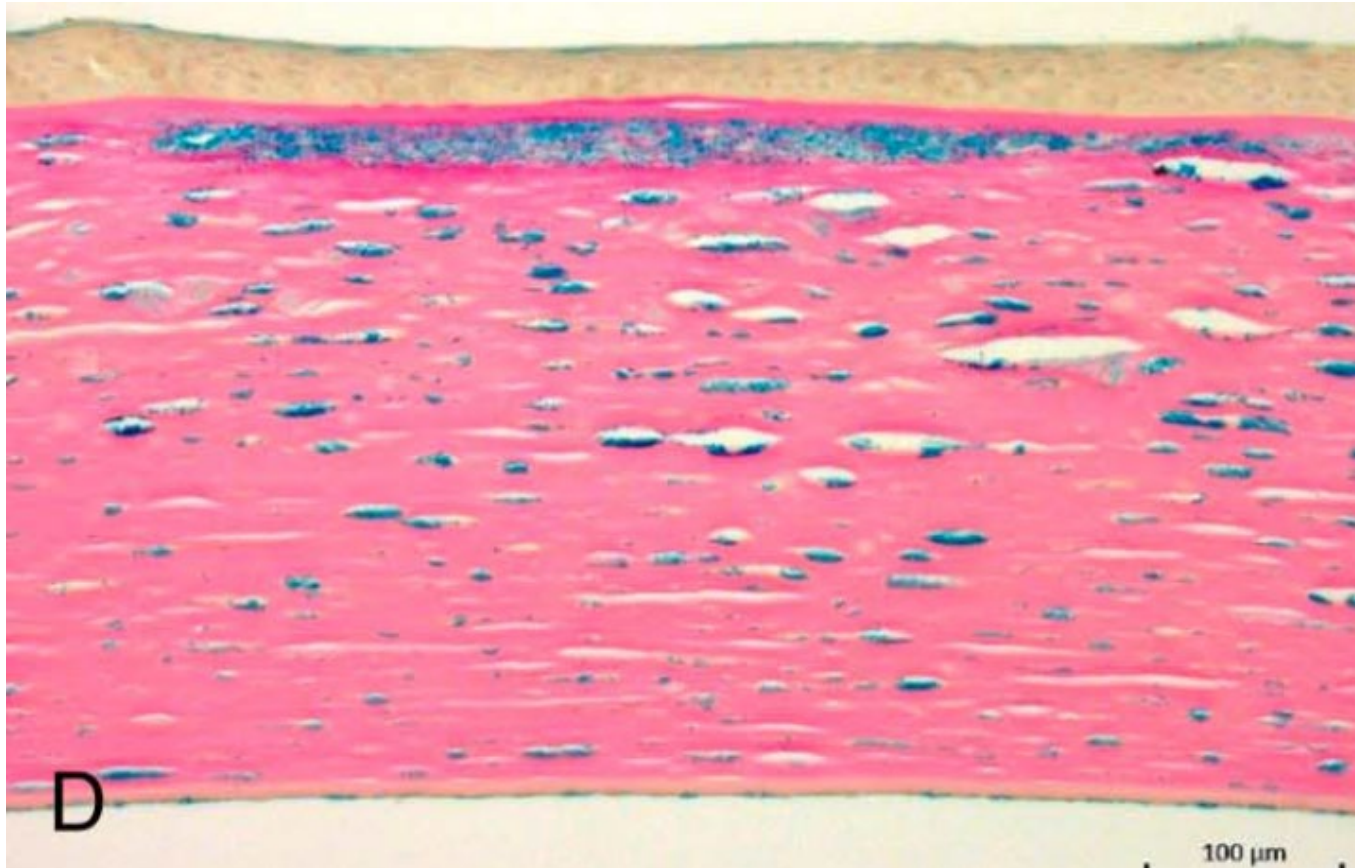
5) Posterior amorphous dystrophy

6) Pre-Descemet corneal dystrophy

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

Endothelial Dystrophies

Corneal Dystrophies



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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal TGFBI Dystrophy

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

Stromal Dystrophies

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Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?

Abnormal accumulation of mucopolysaccharides at all levels of the cornea; they stain with alcian blue

mucopolysaccharides (aka glycosaminoglycans)

Endothelial Dystrophies

(No question—proceed when ready)

Corneal Dystrophies



Epithelial and Subepithelial Dys

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

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

What is a glycosaminoglycan (GAG), and what is it doing in the cornea?

Epithelial-St

Stromal Dys

- 1)
- 2)
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- 6) Pre-Descemet corneal dystrophy

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

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dys

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Good question. But before we address it, let's back up a step...

The corneal stroma consists of three basic components. What are they?

--?

--?

--?

Stromal Dys

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at all levels of the cornea; they stain with alcian blue.

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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The corneal stroma consists of three basic components. What are they?

--? fibers organized into

--?

--?

Stromal Dys

1)

2)

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4) Fleck corneal dystro

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6) Pre-Descemet corneal dystrophy

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Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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--Collagen fibers organized into lamellae;

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Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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Epithelial-St

What is a glycosaminoglycan (GAG), and what is it doing in the cornea?
Good question. But before we address it, let's back up a step...

The corneal stroma consists of three basic components. What are they?

--Collagen fibers organized into lamellae;

--two words, the gooey material that fills the space between lamellae; and
--?

Stromal Dys

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Abnormal mucopolysaccharides (aka **glycosaminoglycans**)
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Endothelial Dystrophies

Corneal Dystrophies



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Endothelial Dystrophies

Corneal Dystrophies



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- , the cells that make the collagen and ground substance

Stromal Dys

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Abnormal mucopolysaccharides (aka **glycosaminoglycans**)
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Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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Corneal Dystrophies



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- Keratocytes, the cells that make the collagen and ground substance

Ground substance is composed of not GAGs, strictly speaking and water.

Stromal Dys

- 1)
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Endothelial Dystrophies

Corneal Dystrophies



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Ground substance is composed of proteoglycans and water.

Stromal Dys

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Endothelial Dystrophies

Corneal Dystrophies



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Ground substance is composed of proteoglycans and water. At last, the payoff:
A proteoglycan is simply a protein with a Ta da! **attached to it.**

Stromal Dys

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A proteoglycan is simply a protein with a glycosaminoglycan attached to it.

Stromal Dys

- 1) So, if the proteoglycans contain abnormal GAGs, the cornea ain't gonna be right,
- 2) and this is the case in MCD.

3) Congenital stromal

4) Fleck corneal dystro

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

- ?
- ?
- ?

e right,

glycosaminoglycans

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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

- Keratan sulfate
- Chondroitin sulfate
- Dermatan sulfate

glycosaminoglycans

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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

- Keratan sulfate?**
- Chondroitin sulfate?**
- Dermatan sulfate?**

Which one ain't right in MCD?

glycosaminoglycans

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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- Keratan sulfate!**
- Chondroitin sulfate
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glycosaminoglycans

Which one ain't right in MCD?
Keratan sulfate

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dys

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- Keratan sulfate!**
- Chondroitin sulfate
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glycosaminoglycans

Endothelial Dystrophies

Which one ain't right in MCD?
Keratan sulfate

What's wrong with it?

Corneal Dystrophies



Epithelial and Subepithelial Dys

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- Keratan sulfate!**
- Chondroitin sulfate
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glycosaminoglycans

Endothelial Dystrophies

Which one ain't right in MCD?
Keratan sulfate

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

Corneal Dystrophies



Epithelial and Subepithelial Dys

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

At what age does MCD begin to manifest?
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A blood test to diagnose MCD is available. What is evaluated?

Epithelial-St

What is a gly
Good question

The corneal
--Collagen fi
--Ground sub
--Keratocytes

Ground subs
A proteoglyc

Stromal Dys

- 1) So, if the proteoglycan
- 2) and this is the case in
- 3) Congenital stromal
- 4) Fleck corneal dystro
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- 6) Pre-Descemet cornea

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

- Keratan sulfate!**
- Chondroitin sulfate
- Dermatan sulfate

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Epithelial-Stromal *TGFB* Dystrophy

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

--
--
--
--

to 20s

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

Endothelial Dystrophies

(No question yet—keep going)



Corneal Dystrophies

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- It involves the entire stroma (most pick a layer and stay there)

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Endothelial Dystrophies

(No question—keep going)



Corneal Dystrophies

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- It can involve the endothelium (others don't)

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to 20s

Endothelial Dystrophies

(No question—keep going)



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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Epithelial-Stromal *TGFB* Dystrophy

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

- It's inherited AR (most corneal dystrophies are AD)
- It involves the entire stroma (most pick a layer and stay there)
- It can involve the endothelium (others don't)
- It's limbus-to-limbus (others tend to spare the corneal periphery)

to 20s

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(No question—keep going)

Corneal Dystrophies



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Epithelial-Stromal TGFBI Dystro

Take note of this aka! The term *mucopolysaccharide* is considered outdated—*glycosaminoglycan* is preferred in recent editions of the C. of defective glycosaminoglycan production, **not** mucopolysaccharide production. (The import of this shift in terminology will be made clear shortly.)

It's now time to address this...

Stromal Dystrophies

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Yes, severe impairment occurs in the teens to 20s

What is the histologic hallmark of MCD on light microscopy?

Abnormal accumulation of mucopolysaccharides at all levels of the cornea; they stain with alcian blue

mucopolysaccharides (aka glycosaminoglycans)

Endothelial Dystrophies

(No question—keep going)



Corneal Dystrophies

Epithelial and Subepithelial Dys

What is the inheritance pattern for macular dystrophy (MCD)?
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At what age does MCD begin to manifest?

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

Marilyn Monroe Always Gets Her Man in LA County

b) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

(No question—keep going)

Corneal Dystrophies



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

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

b) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

(No question—keep going)

Corneal Dystrophies



Epithelial and Subepithelial Dys

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Marilyn macular dystrophy

Monroe mucopolysaccharide

Always Alcian blue

The problem is readily apparent—the *mnemonic only works if the abnormal material in MCD is called 'mucopolysaccharide.'* So either modify the mnemonic to include GAGs (tweet your mods to me @EyeDentistAAO), or (gasp!) actually learn it.

A amyloid

County Congo Red

b) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

(No question—keep going)



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

What was the former name of this condition?

Epithelial-Stromal *TGFB* Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

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

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

What was the former name of this condition?

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Why was the name changed?

Only ~50% manifest corneal crystals

Epithelial-Stromal *TGFB* Dystrophies

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Endothelial Dystrophies



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Endothelial Dystrophies



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Early in the disease, the cornea displays either a central opaque 'disc,' or central crystals

Epithelial-Stromal *TGFB* Dystrophies

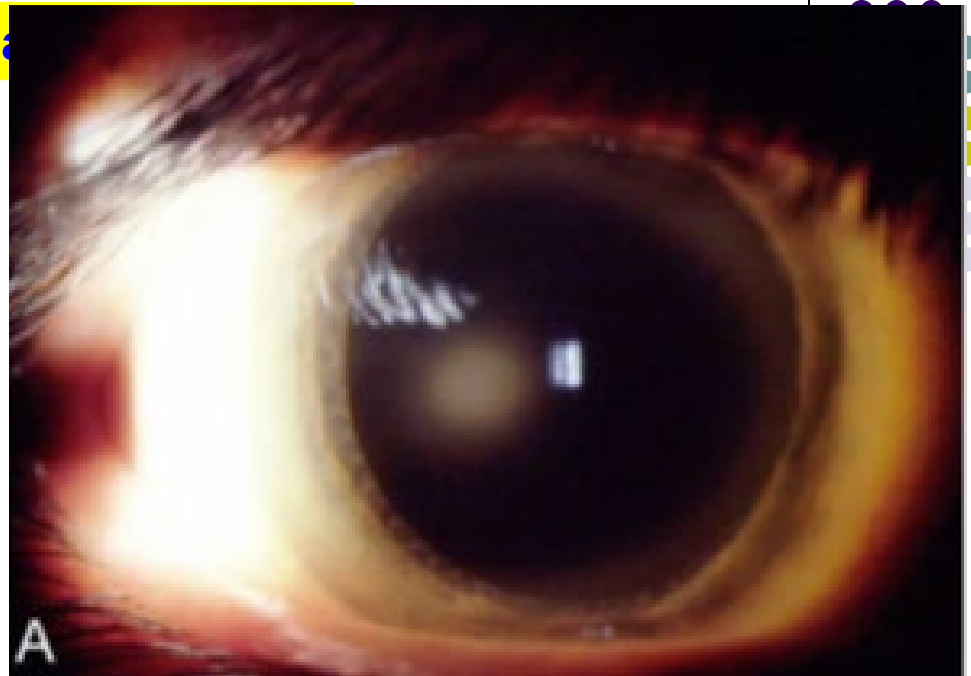
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Endothelial Dystrophies

Cornea

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





Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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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.

Epithelial-Stromal *TGFB* Dystrophies

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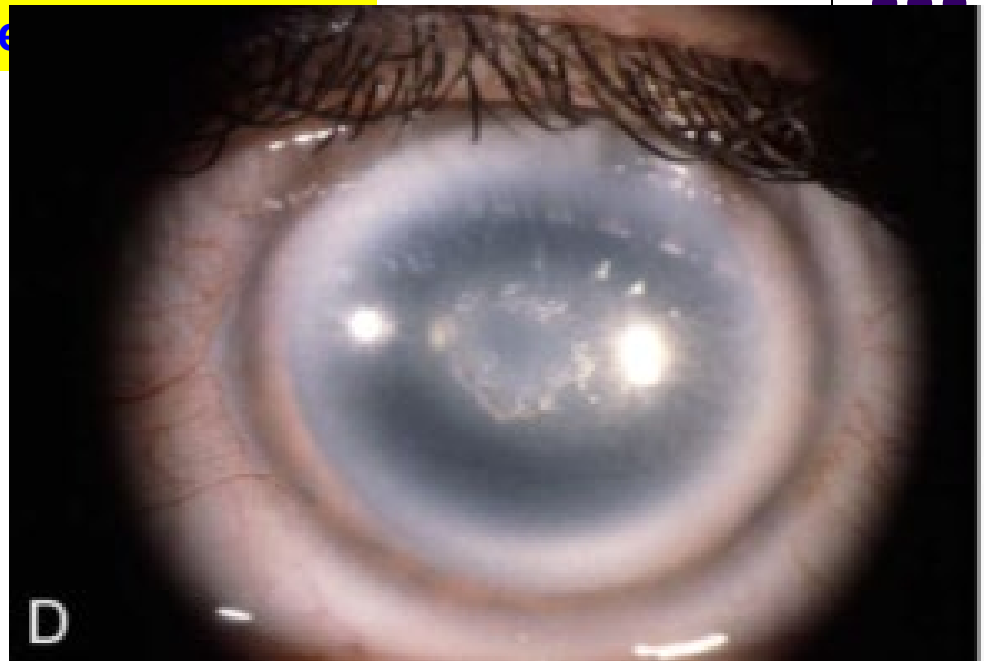
Endothelial Dystrophies

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

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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?

Epithelial-Stromal *TGFB* 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?

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one word

that stain with

three words

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What is the histologic hallmark of SCD on light microscopy?

Phospholipids that stain with Oil red O

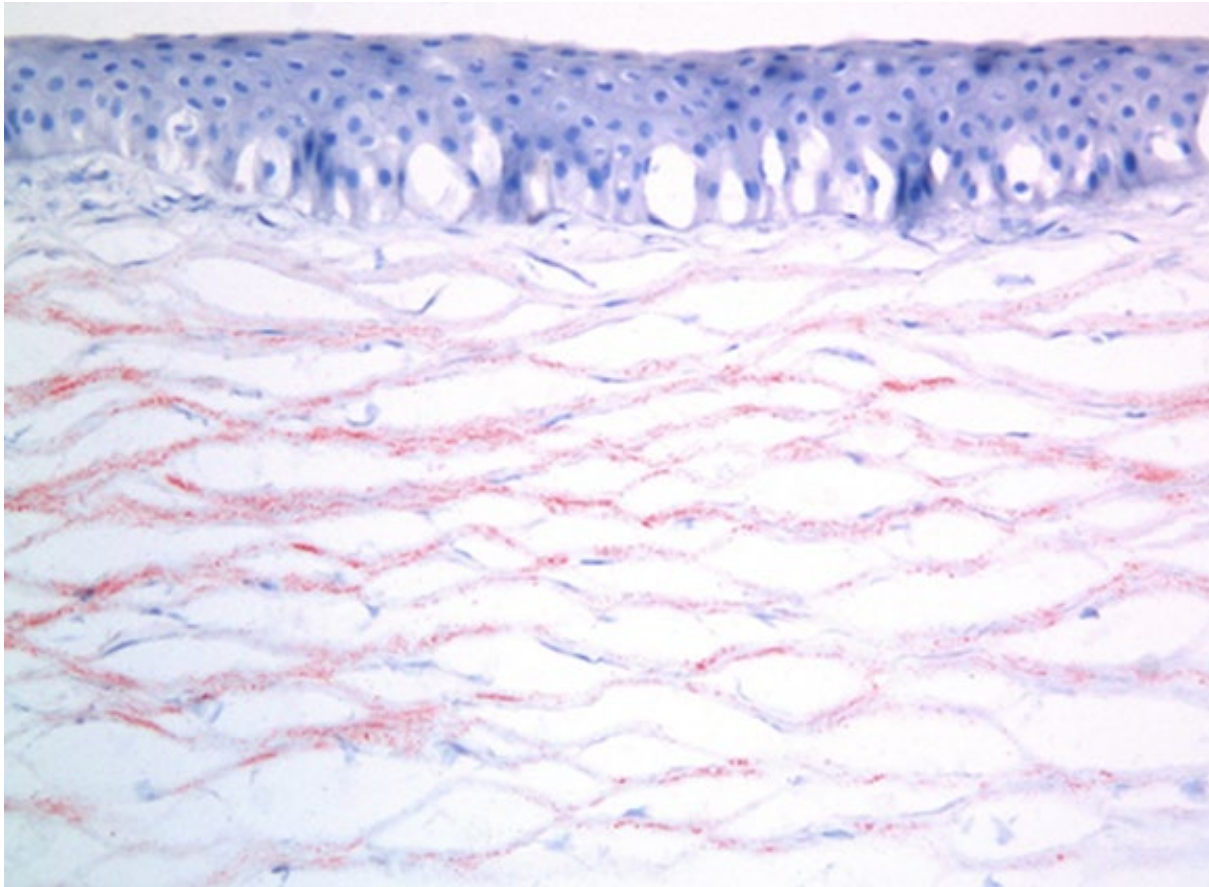
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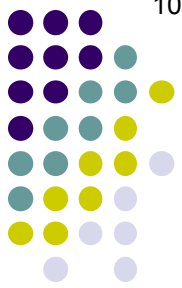
Endothelial Dystrophies

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 *TGFB* Dystrophies

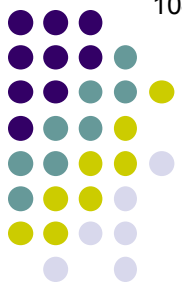
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Endothelial Dystrophies

At what age does CSCD begin to manifest?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

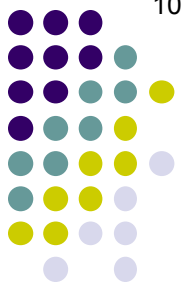
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Endothelial Dystrophies

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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

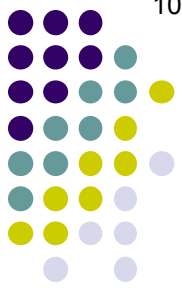
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Endothelial Dystrophies

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

What is seen at the slit lamp?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

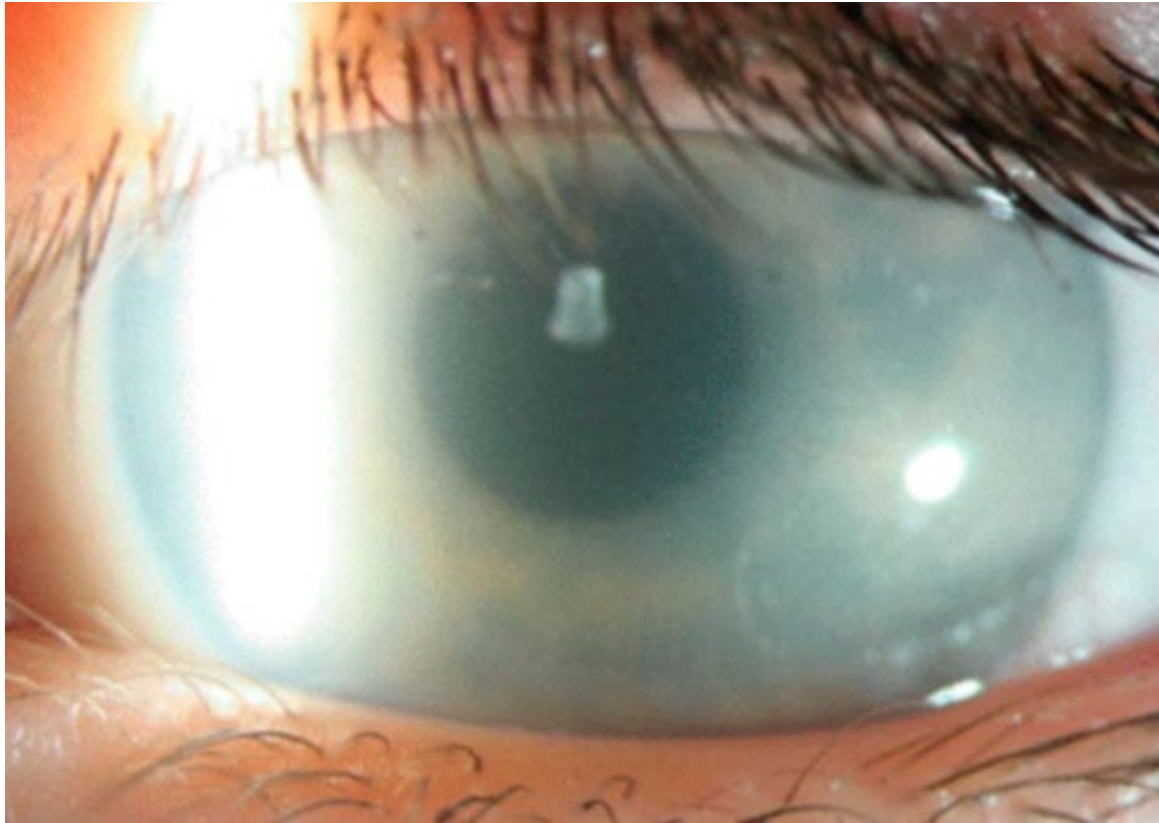
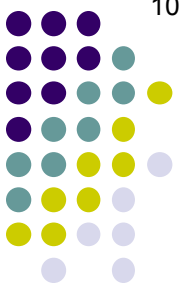
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Endothelial Dystrophies

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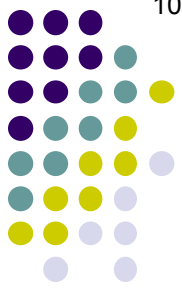
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Limbus-to-limbus, uniformly distributed haze.
On close inspection, innumerable white flaky opacities are present.

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 *TGFB* Dystrophies

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Endothelial Dystrophies

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Is it progressive?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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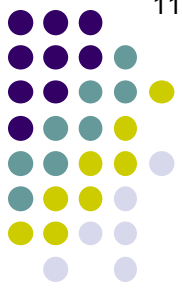
Endothelial Dystrophies

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Is it progressive?
Generally no, or only modestly so

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies

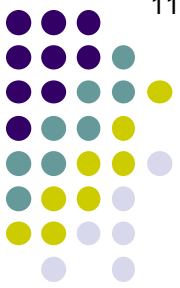
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Is it painful?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies

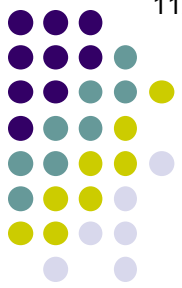
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Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Endothelial Dystrophies

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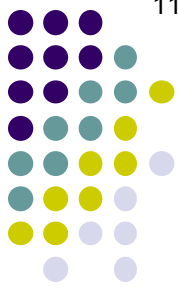
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Does it affect vision?

Corneal Dystrophies



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Endothelial Dystrophies

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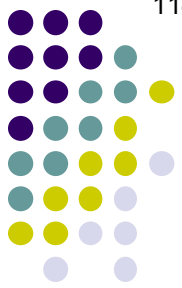
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Is it painful?
No

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

Corneal Dystrophies



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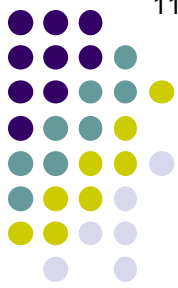
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What is the histologic hallmark of CSCD on light microscopy?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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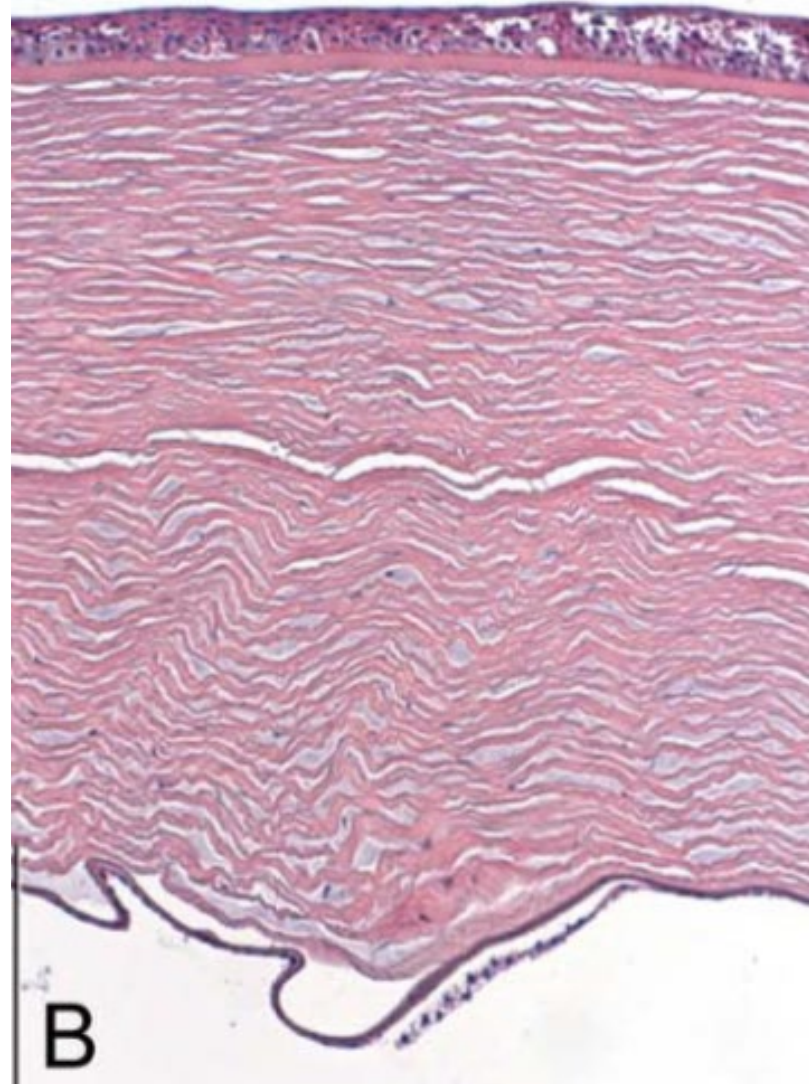
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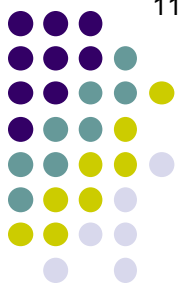
What is the histologic hallmark of CSCD on light microscopy?
Pronounced thickening of the corneal stroma with separation of corneal lamellae

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

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

Birth (duh, it's congenital)

What is seen at the slit lamp?

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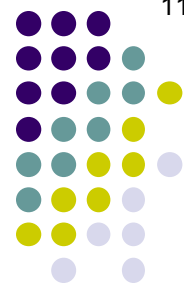
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Endothelial Dystrophies



Corneal Dystrophies

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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

STUMPED

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

Birth (duh, it's congenital)

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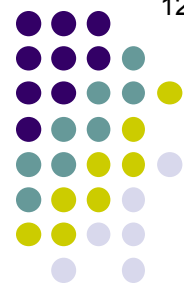
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Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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

STUMPED

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

Coming in hot...

Birth (duh, it's congenital)

What is seen at the slit lamp?

Limbus-to-limbus, uniformly distributed haze.

What are the clinical hallmarks of congenital corneal dystrophy?
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Is it progressive?

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Is it painful?

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Does it affect vision?

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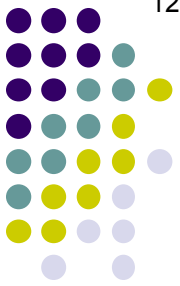
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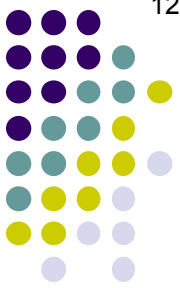
Endothelial Dystrophies



- S
- T
- U
- M
- P
- E
- D

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

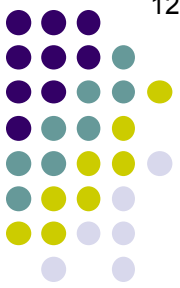
Corneal Dystrophies



- **S**clerocornea; **S**tromal dystrophy (CHSD)
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**ndothelial dystrophy (CHED); **E**levated IOP
(ie, two words)
- **D**ermoid of the cornea

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

Corneal Dystrophies



- **S**clerocornea; **S**tromal dystrophy (CHSD)
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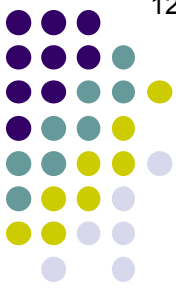
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(Note: There are two Ss and two Es)*

Corneal Dystrophies



- Sclerocornea; **Stromal dystrophy (CHSD)**
- Trauma (endothelial; ie, from forceps)
- Ulcer
- *Next we will touch on distinguishing among CHSD, CHED and primary congenital glaucoma by highlighting key differences in their presentations*
- Peters anomaly
- **Endothelial dystrophy (CHED); Elevated IOP**
(ie, congenital glaucoma)
- Dermoid of the cornea

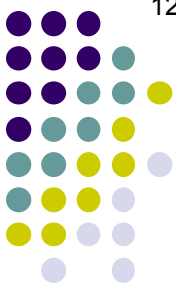
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	?			
CHSD	?			
Primary congenital glaucoma	?			

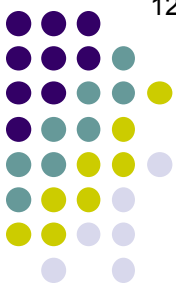
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased			
CHSD	Mildly increased			
Primary congenital glaucoma	Variably increased (or WNL, or thin)			

Corneal Dystrophies

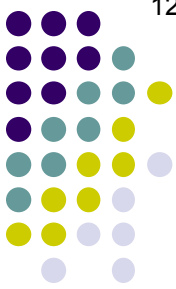


CCT, corneal diameter, IOP and the presence/absence of tearing & photophobia are key to differentiating among CHED, CHSD, and primary congenital glaucoma. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased			
CHSD	Mildly increased	In CHSD, the cornea is modestly thickened by the presence of the material that causes the cloudiness		
Primary congenital glaucoma	Variably increased (or WNL, or thin)			

(No question—advance when ready)

Corneal Dystrophies

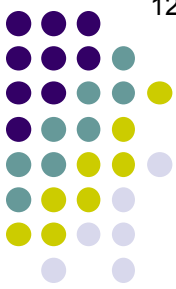


CCT, corneal diameter, IOP and the presence/absence of tearing & photophobia are key to differentiating among CHED, CHSD, and primary congenital glaucoma. Fill in the blanks below.

	CCT	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	Markedly increased	The CCT is dramatically increased in CHED because of edema 2ndry to lack of adequate endothelial barrier and deturgescence function		
CHSD	Mildly increased			
Primary congenital glaucoma	Variably increased (or WNL, or thin)			

(No question—advance when ready)

Corneal Dystrophies

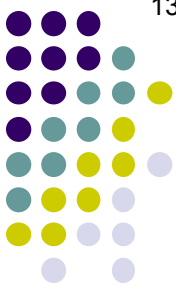


CCT, corneal diameter, IOP and the presence/absence of tearing & photophobia are key to differentiating among CHED, CHSD, and primary congenital glaucoma. Fill in the blanks below.

	CCT	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	Markedly increased			
CHSD	Mildly increased			
Primary congenital glaucoma	Variably increased (or WNL, or thin)	In congenital glaucoma, corneal thickness depends upon 1) whether the endo is healthy and 2) how high the IOP is		

(No question—advance when ready)

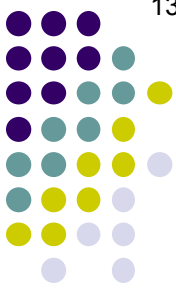
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	?		
CHSD	Mildly increased	?		
Primary congenital glaucoma	Variably increased (or WNL, or thin)	?		

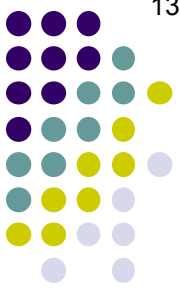
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	WNL		
CHSD	Mildly increased	WNL		
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased		

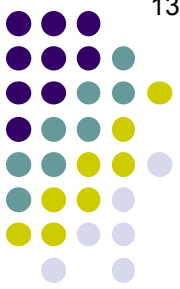
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

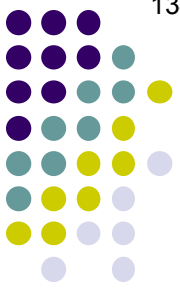
	Corneal Diameter	Intraocular Pressure (IOP)	Tearing & Photophobia
CHED	<p>Why is corneal diameter increased in congenital glaucoma?</p>		
CHSD			
Primary congenital glaucoma			
	increased (or WNL, or thin)	Increased	

Corneal Dystrophies



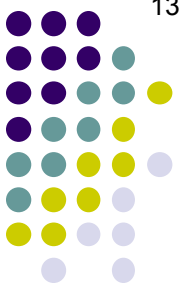
CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

		Corneal Diameter	Intraocular Pressure (IOP)	Tearing & Photophobia
	Why is corneal diameter increased in congenital glaucoma? Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall			
CHED				
CHSD				
Primary congenital glaucoma	Corneal Diameter increased (or WNL, or thin)	Increased		



Congenital glaucoma: Increased corneal diameter

Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	IOP	Tearing & Photophobia
	<p><i>Why is corneal diameter increased in congenital glaucoma?</i> Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall</p>		
CHED	<p><i>What is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?</i></p>		
CHSD			
Primary congenital glaucoma			
	increased (or WNL, or thin)	Increased	

Corneal Dystrophies



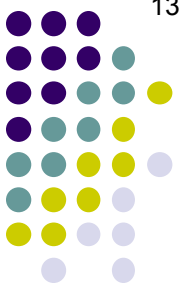
CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	IOP	Tearing/Photophobia
	<p><i>Why is corneal diameter increased in congenital glaucoma?</i> Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall</p>		
CHED	<p><i>What is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?</i> Buphthalmos</p>		
CHSD			
Primary congenital glaucoma			
	increased (or WNL, or thin)	Increased	



Congenital glaucoma: Buphthalmos OD

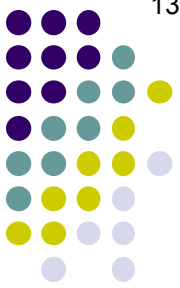
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	Intraocular Pressure (IOP)	Tearing & Photophobia
	<p><i>Why is corneal diameter increased in congenital glaucoma?</i> Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall</p>		
CHED	<p><i>What is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?</i> Buphthalmos</p>		
CHSD	<p><i>What does buphthalmos translate to in English?</i></p>		
Primary congenital glaucoma	increased (or WNL, or thin)	Increased	

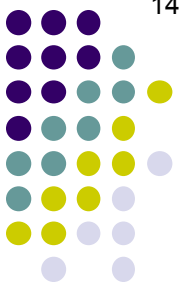
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	Intraocular Pressure (IOP)	Tearing & Photophobia
	<p><i>Why is corneal diameter increased in congenital glaucoma?</i> Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall</p>		
CHED	<p><i>What is the formal term for eye enlargement secondary to elevated IOP in congenital glaucoma?</i> Buphthalmos</p>		
CHSD	<p><i>What does buphthalmos translate to in English?</i> 'Ox's eye'</p>		
Primary congenital glaucoma	increased (or WNL, or thin)	Increased	

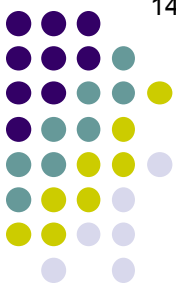
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	IOP	Tearing/Photophobia
	Why is corneal diameter increased in congenital glaucoma?		
CHED	The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?		
CHSD			
Primary congenital glaucoma			
	increased (or WNL, or thin)	Increased	

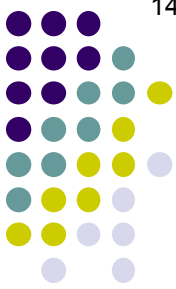
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	IOP	Tearing/Photophobia
	Why is corneal diameter increased in congenital glaucoma?		
CHED	The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort? Breaks in Descemet's membrane (and its overlying endothelial layer)		
CHSD			
Primary congenital glaucoma			
	increased (or WNL, or thin)	Increased	

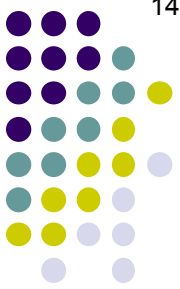
Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

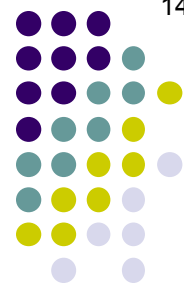
		Why is corneal diameter increased in congenital glaucoma?		
		The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?		
CHED		Breaks in Descemet's membrane (and its overlying endothelial layer)		
		How (ie, in what direction) are these breaks oriented?		
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	Increased		

Corneal Dystrophies



CCT, corneal diameter, IOP and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

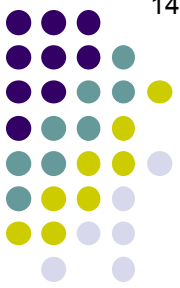
	Corneal Diameter	IOP	Photophobia	Tearing
	<p><i>Why is corneal diameter increased in congenital glaucoma?</i></p> <p><i>The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?</i></p> <p><i>Breaks in Descemet's membrane (and its overlying endothelial layer)</i></p> <p><i>How (ie, in what direction) are these breaks oriented?</i></p> <p><i>Horizontally</i></p>			
CHED				
CHSD				
Primary congenital glaucoma	increased (or WNL, or thin)	Increased		



Corneal Dystrophies

CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	Corneal Diameter	IOP	Photophobia	Tearing
	Why is corneal diameter increased in congenital glaucoma?			
CHED	The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?			
	Breaks in Descemet's membrane (and its overlying endothelial layer)			
CHSD	How (ie, in what direction) are these breaks oriented?			
	Horizontally			
Primary congenital glaucoma	What is the eponymous name for these breaks?			
	increased (or WNL, or thin)	Increased		

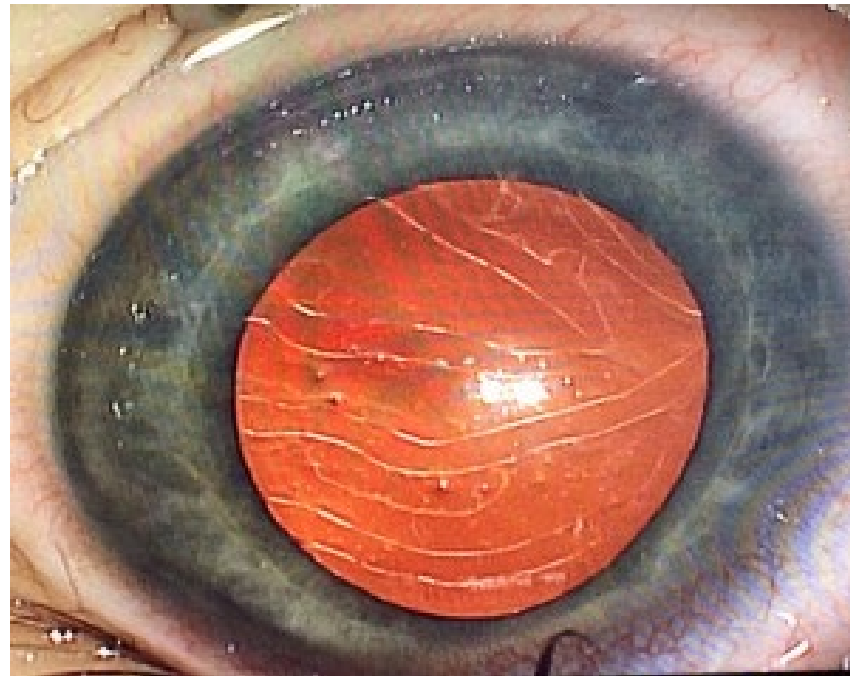
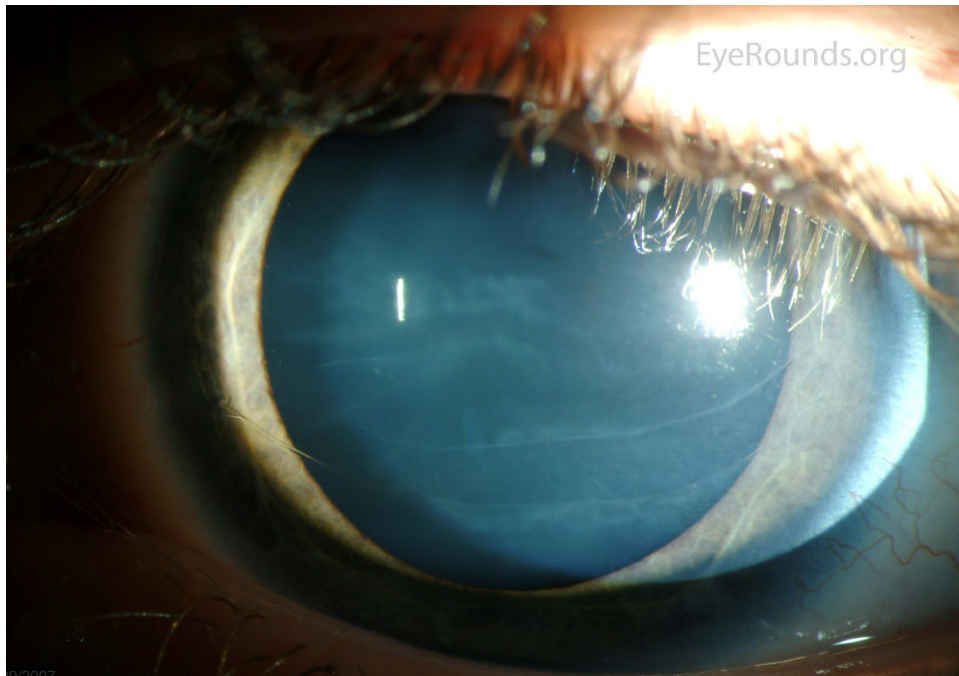


Corneal Dystrophies

CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

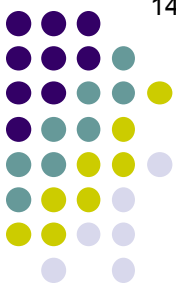
		Why is corneal diameter increased in congenital glaucoma?		
		The high IOP in congenital glaucoma causes further mechanical damage to the cornea—what sort?		
CHED		Breaks in Descemet's membrane (and its overlying endothelial layer)		
		How (ie, in what direction) are these breaks oriented?		
CHSD		Horizontally		
		What is the eponymous name for these breaks?		
Primary congenital glaucoma		Haab's striae		
	increased (or WNL, or thin)	Increased		

Corneal Dystrophies



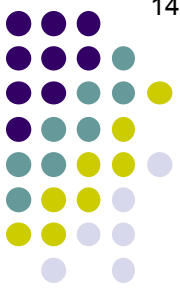
Horizontal Descemet's breaks (*Haab's striae*) in congenital glaucoma

Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	WNL	?	
CHSD	Mildly increased	WNL	?	
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased	?	

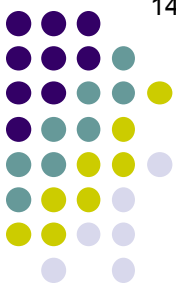


Corneal Dystrophies

CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	WNL	WNL	
CHSD	Mildly increased	WNL	WNL	
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased	Duh	

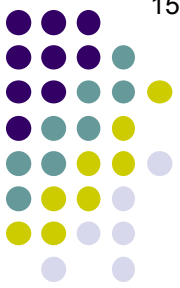
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	WNL	WNL	?
CHSD	Mildly increased	WNL	WNL	?
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased	Duh	?

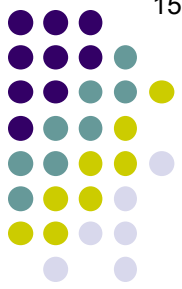
Corneal Dystrophies



CCT, *corneal diameter*, *IOP* and the *presence/absence of tearing & photophobia* are key to differentiating among *CHED*, *CHSD*, and *primary congenital glaucoma*. Fill in the blanks below.

	<i>CCT</i>	<i>Corneal diameter</i>	<i>IOP</i>	<i>Tearing/ Photophobia?</i>
CHED	Markedly increased	WNL	WNL	No
CHSD	Mildly increased	WNL	WNL	No
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased	Duh	Yes

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

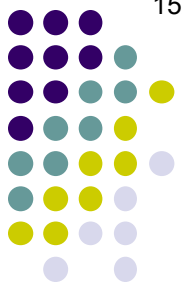
Stromal Dystrophies

- 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 *TGFB* Dystrophies

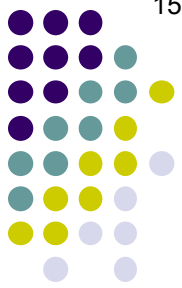
Stromal Dystrophies

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

Endothelial Dystrophies

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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

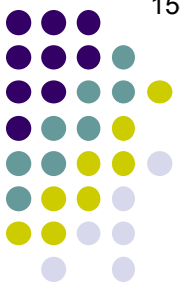
- 1) Macular corneal dystrophy
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- 6) Pre-Descemet corneal dystrophy

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 *TGFB* 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?

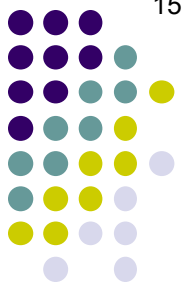
Very early—can even be congenital

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 *TGFB* Dystrophies

Stromal Dystrophies

- 1) Macular corneal dystrophy
- 2) Schnyder corneal dystrophy
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Endothelial Dystrophies

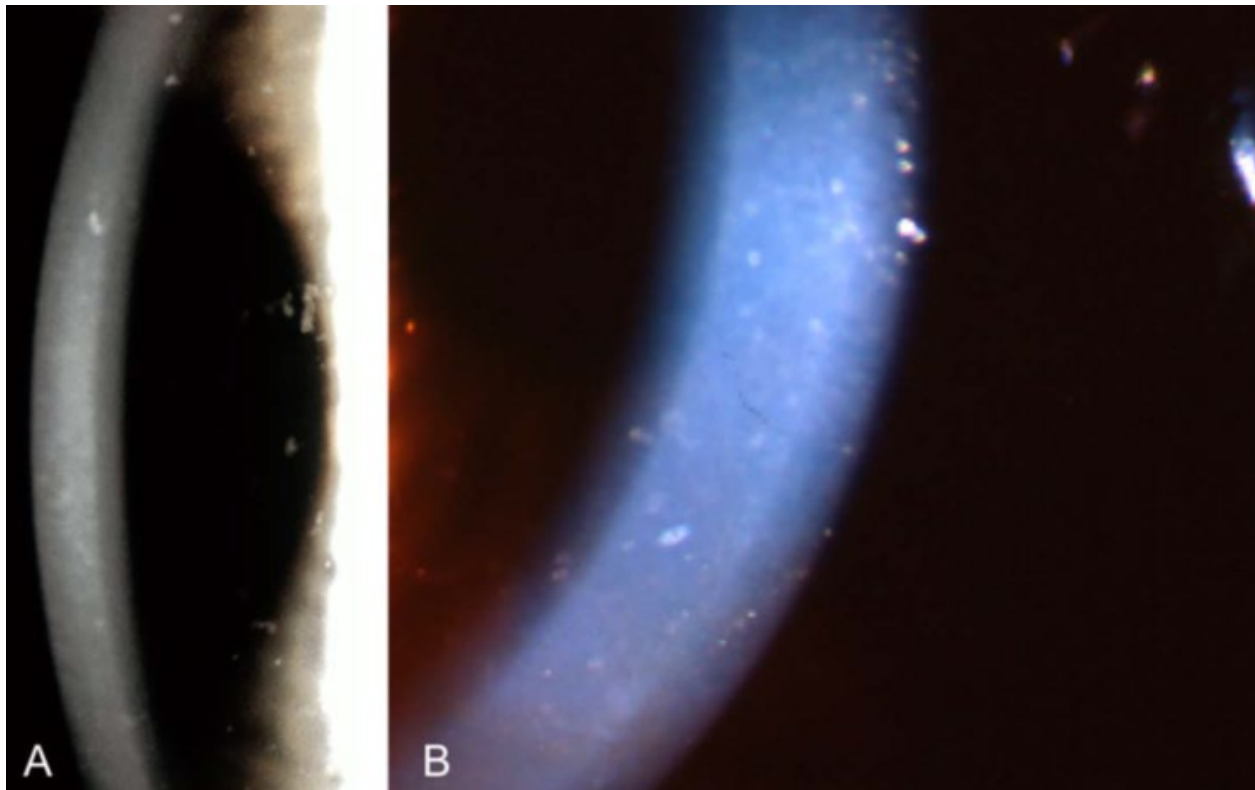
At what age does FCD begin to manifest?

Very early—can even be congenital

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

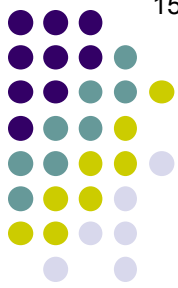
Subtle light-gray discs in the stroma that have described as '**dandruff-like**.'

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 *TGFB* Dystrophies

Stromal Dystrophies

- 1) Macular corneal dystrophy
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- 3) Congenital stromal corneal dystrophy
- 4) **Fleck corneal dystrophy**
- 5) Posterior amorphous corneal dystrophy
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Endothelial Dystrophies

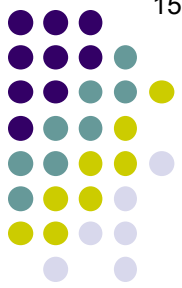
At what age does FCD begin to manifest?

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 hazy v clear.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFI* Dystrophies

Stromal Dystrophies

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

Endothelial Dystrophies

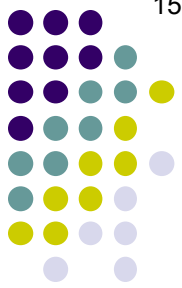
At what age does FCD begin to manifest?

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 *TGFB* Dystrophies

Stromal Dystrophies

- 1) Macular corneal dystrophy
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Endothelial Dystrophies

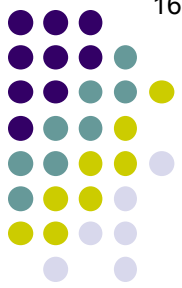
At what age does FCD begin to manifest?

Very early—can even be congenital

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

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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

- 1) Macular corneal dystrophy
- 2) Schnyder corneal dystrophy
- 3) Congenital stromal corneal dystrophy
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Endothelial Dystrophies

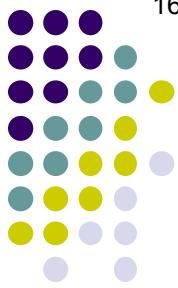
At what age does FCD begin to manifest?

Very early—can even be congenital

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

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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFI* 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?

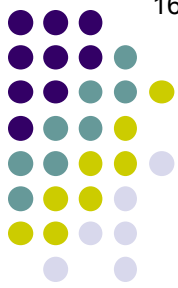
Very early—can even be congenital

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 *TGFB* 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

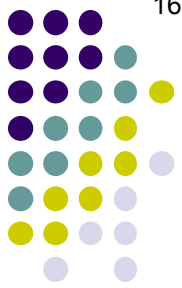
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Is it painful?

No

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* 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?

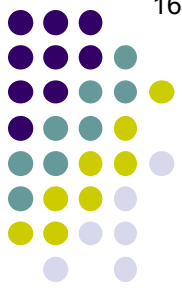
Subtle light-gray discs in the stroma that have described as '**dandruff-like**.' The intervening spaces are **clear**. The lesions are **never** found in non-stromal portions of the cornea.

Is it painful?

No

Does it affect vision?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFI* 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) Pre-Descemet 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?*

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

First decade. Can be present in infancy.

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies

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

First decade. Can be present in infancy.

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

4) **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) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies

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First decade. Can be present in infancy.

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

PACD is a dystrophy of the **deep** corneal stroma.

4) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies

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

First decade. Can be present in infancy.

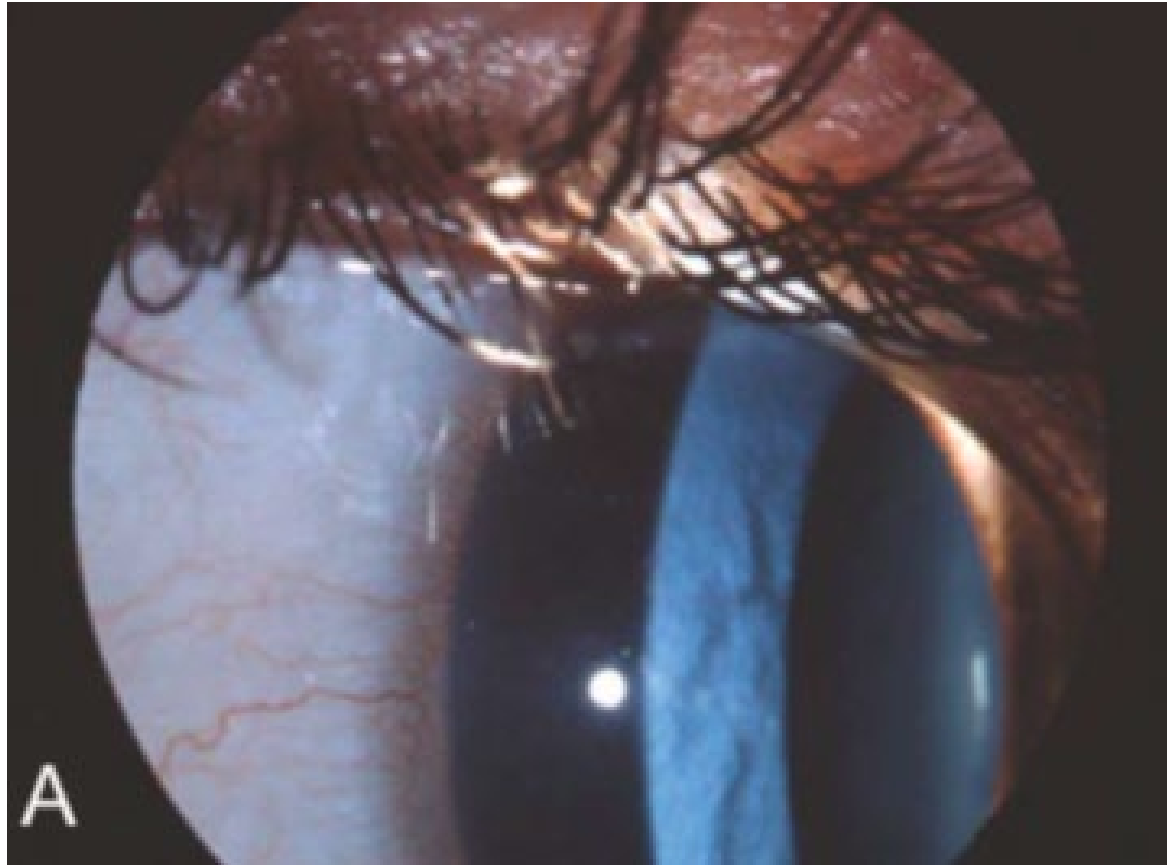
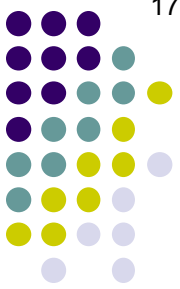
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PACD is a dystrophy of the **deep** corneal stroma. Sheetlike opacities are present, and can be extensive. The deepest lesions can indent Descemet's and the endothelium.

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies



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

Corneal Dystrophies

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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

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies

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Endothelial Dystrophies

Corneal Dystrophies



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Endothelial Dystrophies

Corneal Dystrophies

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

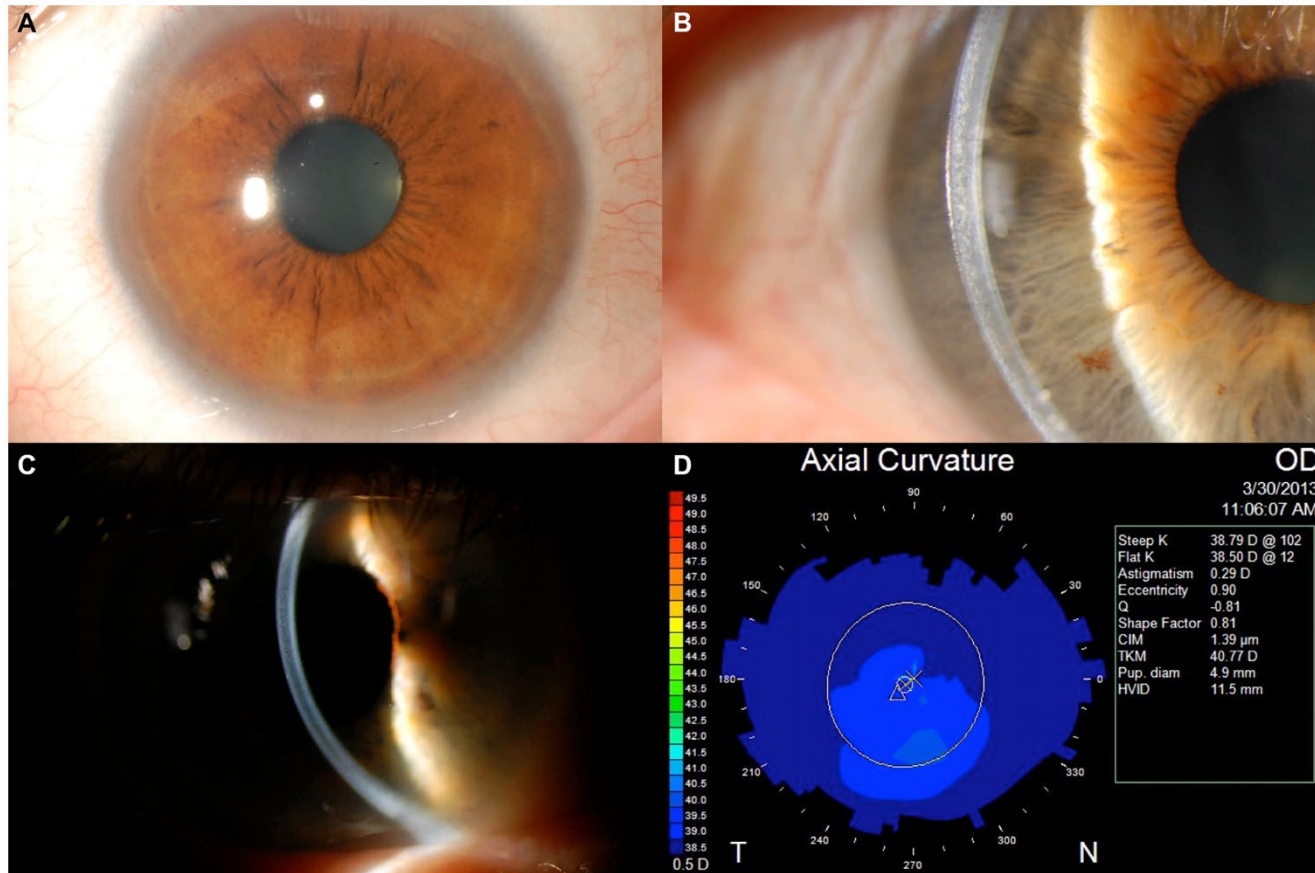
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5) **Posterior amorphous corneal dystrophy**

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

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Is it painful?

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

Endothelial Dystrophies

Corneal Dystrophies



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No

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Is it painful?

No

Does it affect vision?

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Endothelial Dystrophies

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Is it painful?

No

Does it affect vision?

Only mildly

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Endothelial Dystrophies

Corneal Dystrophies

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What is the histologic hallmark of PACD on light microscopy?

5) **Posterior amorphous corneal dystrophy**

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Endothelial Dystrophies

Corneal Dystrophies

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What is the histologic hallmark of PACD on light microscopy?

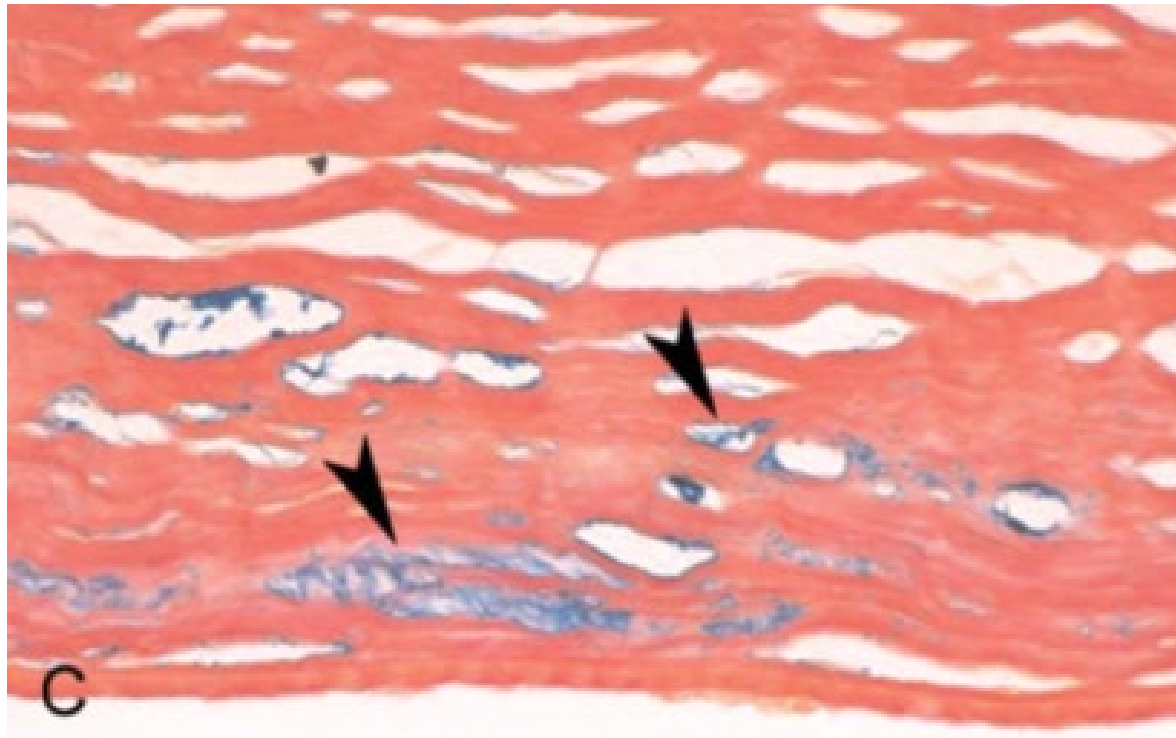
Irregularities to the pre-Descemet's deep stroma

5) **Posterior amorphous corneal dystrophy**

6) Pre-Descemet corneal dystrophy

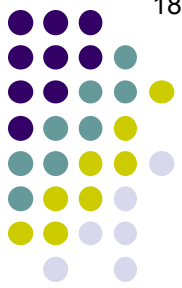
Endothelial Dystrophies

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 *TGFB* Dystrophies

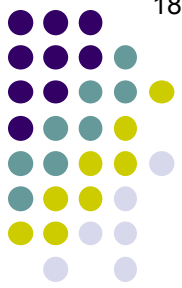
Stromal Dystrophies

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Endothelial Dystrophies

At what age does PDCD begin to manifest?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

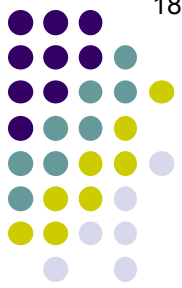
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- 6) **Pre-Descemet corneal dystrophy**

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

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

Stromal Dystrophies

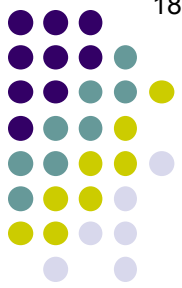
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Endothelial Dystrophies

Corneal Dystrophies



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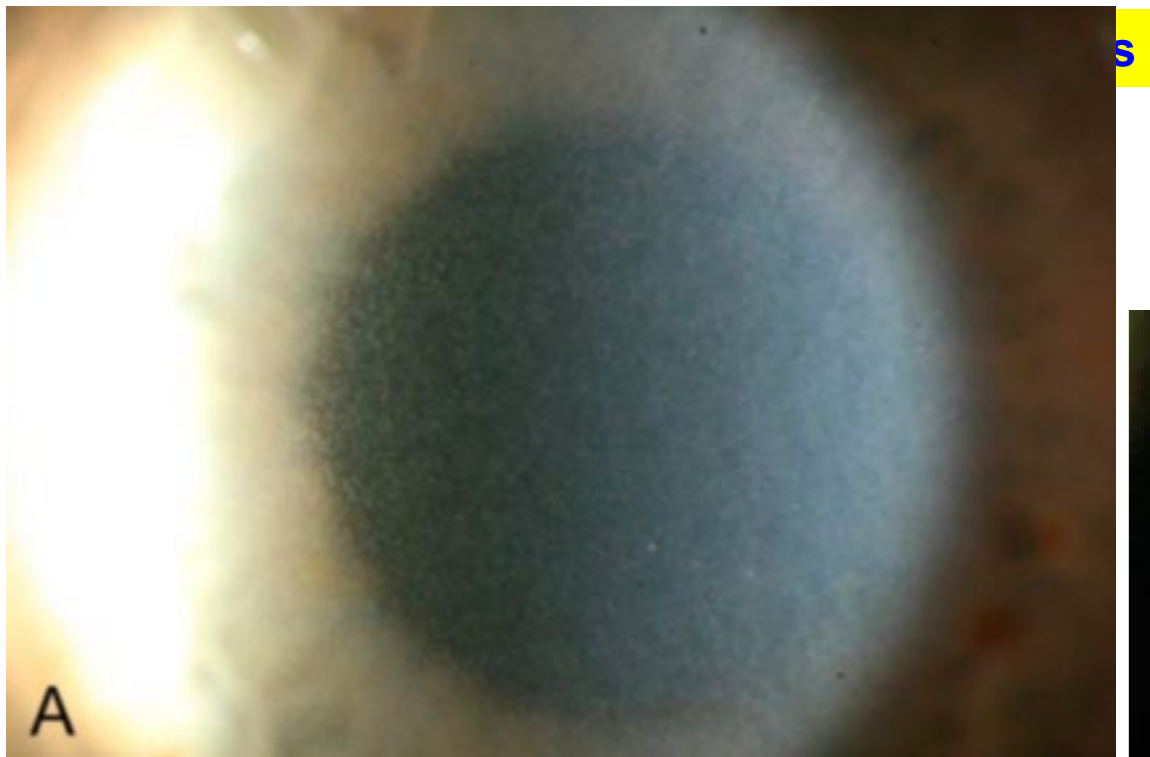
At what age does PDCD begin to manifest?

Usually after age 30 years; rarely in childhood

What is seen at the slit lamp?

Fine punctate opacities just anterior to Descemet's

Endothelial Dystrophies



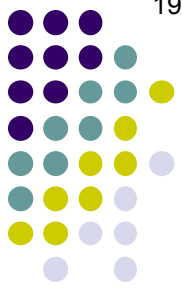
Pre-Descemet's corneal dystrophy.

A, With broadbeam illumination, punctate opacities anterior to Descemet membrane are apparent.

B, Slit beam illumination of the same eye demonstrating punctate opacities anterior to Descemet membrane.



Corneal Dystrophies



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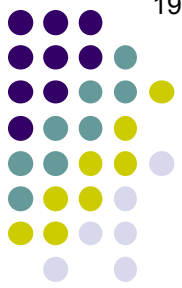
What is seen at the slit lamp?

Fine punctate opacities just anterior to Descemet's

Is it painful?

Endothelial Dystrophies

Corneal Dystrophies



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Endothelial Dystrophies

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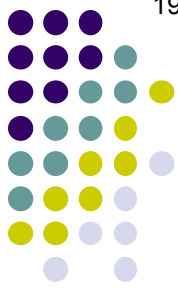
What is seen at the slit lamp?

Fine punctate opacities just anterior to Descemet's

Is it painful?

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Corneal Dystrophies



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Epithelial-Stromal *TGFI* Dystrophies

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Endothelial Dystrophies

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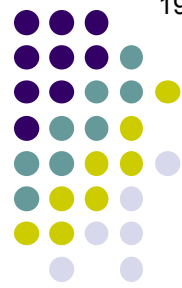
Fine punctate opacities just anterior to Descemet's

Is it painful?

No

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Corneal Dystrophies



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