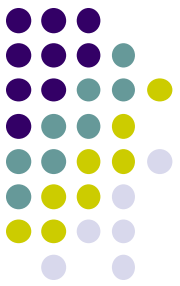


Corneal Dystrophies



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?

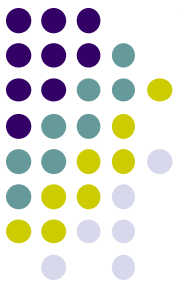
What are the four categories of corneal dystrophies?

?

?

Corneal Dystrophies

2



Epithelial and Subepithelial Dystrophies

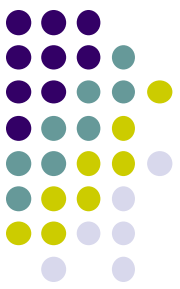
Epithelial-Stromal *TGFB* Dystrophies

What are the four categories of corneal dystrophies?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

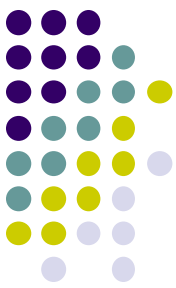
Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



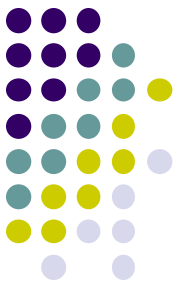
Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?
'Transforming growth factor beta induced'

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

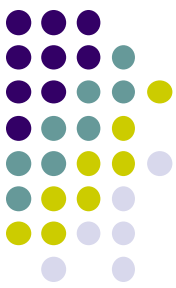
Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?
'Transforming **growth factor** beta induced'

To what does the term **growth factor** refer?

Endothelial Dystrophies

Corneal Dystrophies



6

Epithelial and Subepithelial Dystrophies

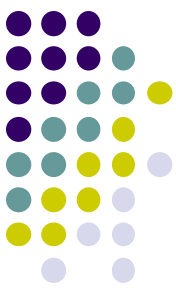
Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?
'Transforming **growth factor** beta induced'

To what does the term **growth factor** refer?
To any of a diverse group of protein (or steroid) molecules that promote cell growth

Endothelial Dystrophies

Corneal Dystrophies



7

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB β** Dystrophies

What does TGFB β stand for in this context?
'Transforming **growth factor** beta induced'

To what does the term **growth factor** refer?
To any of a diverse group of protein (or steroid) molecules that promote cell growth

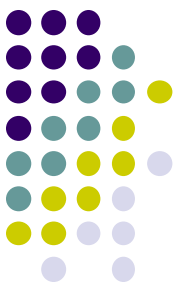
Which growth factor is likely most familiar to ophthalmologists?

two words

growth factor

Endothelial Dystrophies

Corneal Dystrophies



8

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB β** Dystrophies

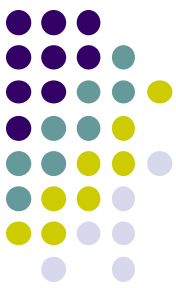
What does TGFB β stand for in this context?
'Transforming **growth factor** beta induced'

To what does the term **growth factor** refer?
To any of a diverse group of protein (or steroid) molecules that promote cell growth

Which growth factor is likely most familiar to ophthalmologists?
Vascular endothelial **growth factor**

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

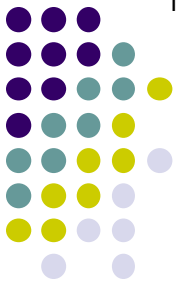
What does TGFB stand for in this context?

'Transforming growth factor beta induced'

*To what does the term **transforming growth factor** refer?*

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?

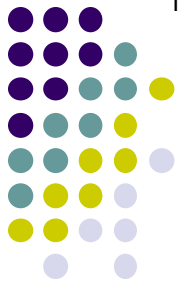
'Transforming growth factor beta induced'

*To what does the term **transforming growth factor** refer?*
A superfamily of related growth factors

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

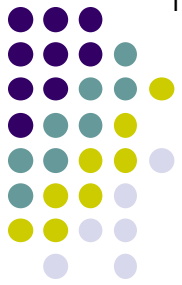
What does TGFB stand for in this context?

'Transforming growth factor beta' induced

*To what does the term transforming growth factor **beta** refer?*

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

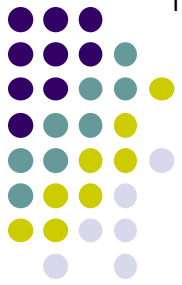
What does TGFB stand for in this context?

'Transforming growth factor beta induced'

*To what does the term transforming growth factor **beta** refer?
A subclass of transforming growth factors (the other subclass is, not surprisingly, transforming growth factor **alpha**)*

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

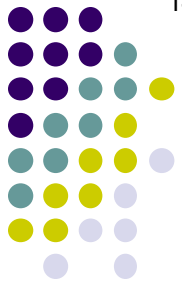
What does TGFB stand for in this context?

'Transforming growth factor beta induced'

*To what does the term transforming growth factor beta **induced** refer?*

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

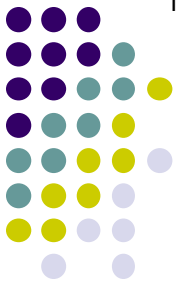
Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?

'Transforming growth factor beta induced'

*To what does the term transforming growth factor beta **induced** refer?*
It refers to a protein, the production of which is controlled by a product of the *transforming growth factor beta* family

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?

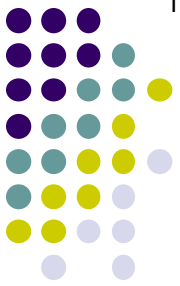
'Transforming growth factor beta induced'

*To what does the term transformant refer to?
It refers to a protein, the product of the transforming growth factor beta gene.*

a protein

What is the name of the protein involved in the TGFB1 dystrophies?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?

'Transforming growth factor beta induced'

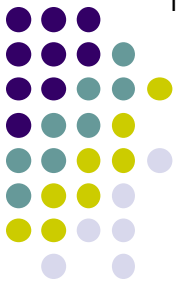
*To what does the term transform refer to?
It refers to a protein, the product of the transforming growth factor beta gene.*

a protein

What is the name of the protein involved in the TGFB1 dystrophies?

Keratoepithelin (the Cornea book refers in passing to the TGFB1 conditions as *keratoepithelin dystrophies*)

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?

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*To what does the term transform refer to?
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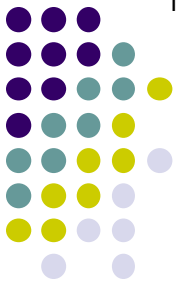
a protein

What is the name of the protein involved in the TGFB dystrophies?

Keratoepithelin (the Cornea book refers in passing to the TGFB conditions as *keratoepithelin dystrophies*)

What is the clinical hallmark of the keratoepithelin dystrophies?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?

'Transforming growth factor beta induced'

To what does the term refer to?
It refers to a protein, the product of the transforming growth factor beta gene.

a protein

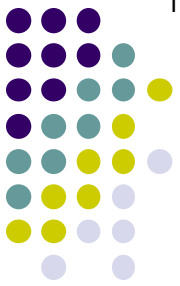
What is the name of the protein involved in the TGFB dystrophies?

Keratoepithelin (the Cornea book refers in passing to the TGFB conditions as *keratoepithelin dystrophies*)

What is the clinical hallmark of the keratoepithelin dystrophies?

Recurrent epithelial erosions

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB** Dystrophies

What does TGFB stand for in this context?

Transforming growth factor beta induced

To what does the term refer?
It refers to **a protein**, the product of the transforming growth factor beta gene.

What is the name of the protein involved in the TGFB dystrophies?

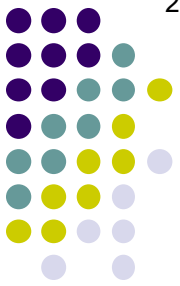
Keratoepithelin (the Cornea book refers in passing to the TGFB conditions as *keratoepithelin dystrophies*)

What is the clinical hallmark of the keratoepithelin dystrophies?

Recurrent epithelial erosions

The Cornea book considered this factoid important enough to make it one of three 'Highlights' for the Dystrophy chapter—take note of it!

Endothelial Dystrophies



Epithelial and Subepithelial Dystrophies

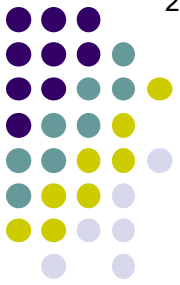
Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?
'Transforming growth factor beta induced'

What is TGFB1's chromosomal location?

Stromal Dystrophies

Endothelial Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

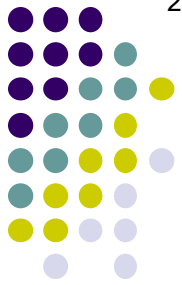
What does TGFB1 stand for in this context?
'Transforming growth factor beta induced'

What is TGFB1's chromosomal location?
5q31

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

What does TGFB1 stand for in this context?
'Transforming growth factor beta induced'

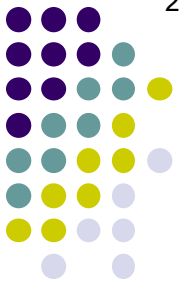
What is TGFB1's chromosomal location?
5q31

The TGFB1 gene was formerly known as what?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal **TGFB1** Dystrophies

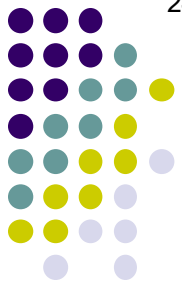
What does TGFB1 stand for in this context?
'Transforming growth factor beta induced'

What is TGFB1's chromosomal location?
5q31

The TGFB1 gene was formerly known as what?
BIGH3 (this factoid is important because you might encounter this name in the older literature)

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

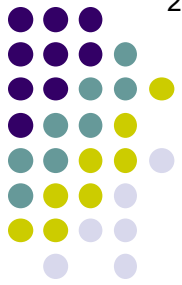
- 1) ?
- 2) ?
- 3) ?
- 4) ?
- 5) ?
- 6) ?

*What are the six
epithelial-stromal TGFBI
corneal dystrophies?*

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

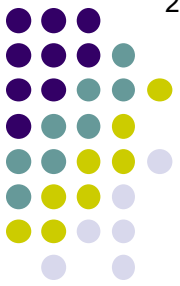
Epithelial-Stromal *TGFBI* Dystrophies

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

*What are the six
epithelial-stromal TGFBI
corneal dystrophies?*

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

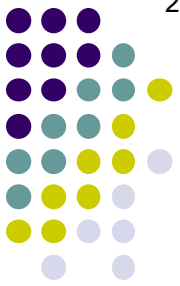
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- 5) Granular type 1
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How do you pronounce this?

How do you pronounce this?

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

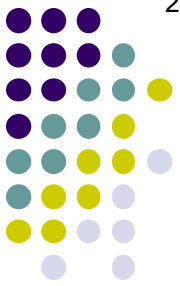
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- 4) Lattice, variant types (III, IIIA, IIIB, IV)
- 5) Granular type 1
- 6) Granular type 2

How do you pronounce this?
RICE BOO-klerz

How do you pronounce this?
TEAL BEN-key

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

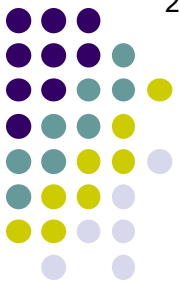
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- 3) Lattice, type 1

What is the histologic hallmark of Reis-Bücklers and Thiel-Behnke?
Disruption/fragmentation of Bowman's layer

4) Granular type 2 (Avellino dystrophy)

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

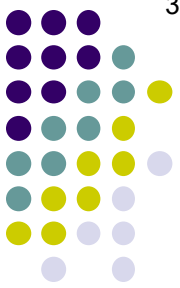
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Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

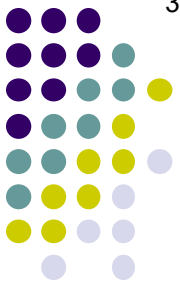
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The corneal-dystrophy section of the Cornea book underwent a major revision recently. In what 'category of corneal dystrophy' were Reis-Bücklers and Thiel-Behnke in previous editions?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

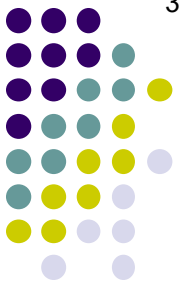
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The '**Corneal Dystrophies of Bowmans**'

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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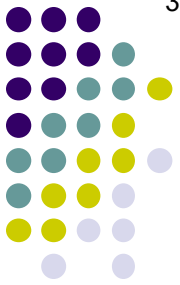
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Disruption/fragmentation of Bowman's layer

*Important aside: When you hear 'disruption/fragmentation of Bowman's layer,' a specific corneal **ectatic** disorder should come immediately to mind. Which one?*

The 'Corneal Dystrophies of Bowmans'

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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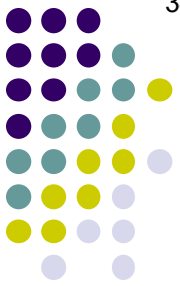
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Keratoconus

The 'Corneal Dystrophies of Bowmans'

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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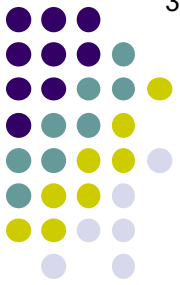
Disruption/fragmentation of Bowman's layer

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*Important aside: When you hear 'disruption/fragmentation of Bowman's layer,' a specific **corneal ectatic disorder** should come immediately to mind. Which one?*
Keratoconus

To finish off this aside: The Cornea book addresses four ectatic conditions. What are the other three?

- Keratoconus
- ?
- ?
- ?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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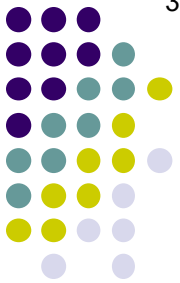
Disruption/fragmentation of Bowman's layer

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Keratoconus

To finish off this aside: The Cornea book addresses four ectatic conditions. What are the other three?

- Keratoconus
- Keratoglobus
- Pellucid marginal degeneration
- Iatrogenic (ie, post-keratorefractive surgery)



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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RBCD

TBCD

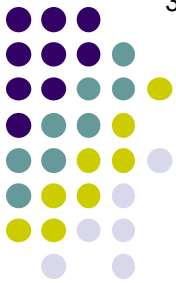
Age of onset

?

?

Stromal Dystrophies

Endothelial Dystrophies



Epithelial and Subepithelial Dystrophies

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RBCD

TBCD

Age of onset

Childhood

Childhood

Stromal Dystrophies

Endothelial Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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- 4) Lattice, type 2
- 5) Granular
- 6) Granular

RBCD

TBCD

	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	?	?
Stromal Dystrophies			
Endothelial Dystrophies			



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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RBCD

TBCD

	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFBI (BIGH3)	TGFBI (BIGH3)
Stromal Dystrophies			
Endothelial Dystrophies			

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

- | | |
|---|--------------|
| 1) Reis-Bücklers corneal dystrophy | |
| 2) Thiel-Behnke corneal dystrophy | |
| 3) Lattice, type 1 | |
| 4) Lattice, type 2 | Age of onset |
| 5) Granular | |
| 6) Granular | Genetics |

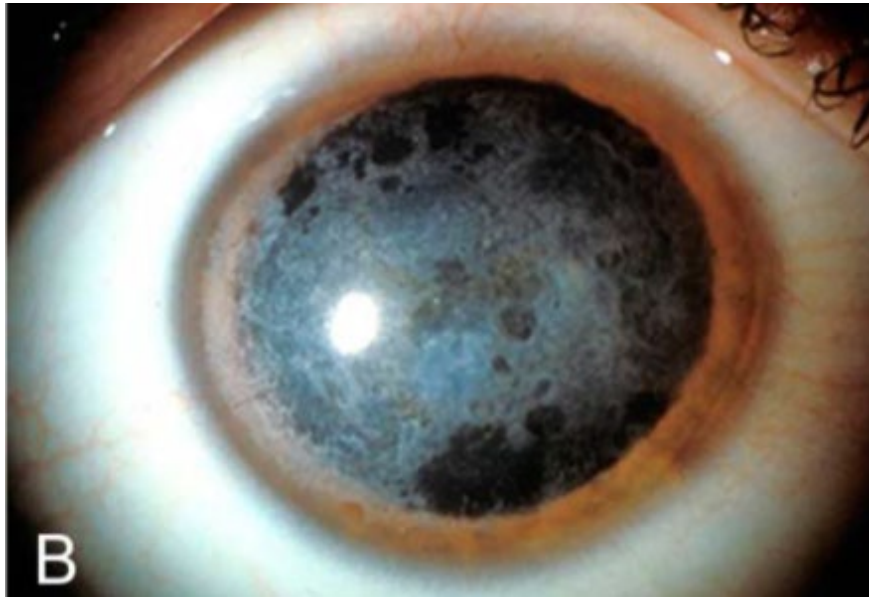
		RBCD	TBCD
Stromal Dystrophies	2) Thiel-Behnke corneal dystrophy		
	3) Lattice, type 1		
	4) Lattice, variant		
	5) Granular		
	6) Granular variant		
Endothelial Dystrophies	Age of onset	Childhood	Childhood
	Genetics	TGFBI (BIGH3)	TGFBI (BIGH3)
	SL appearance	?	?

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

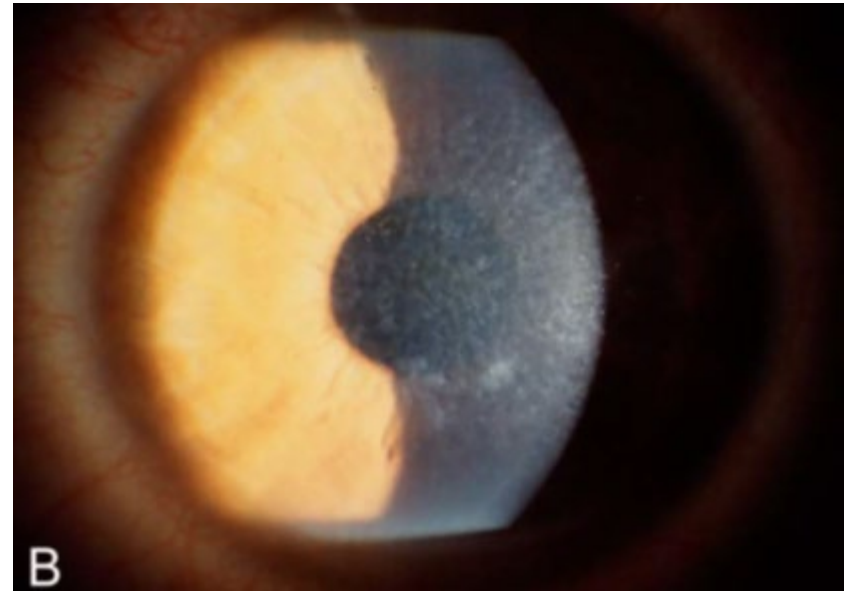
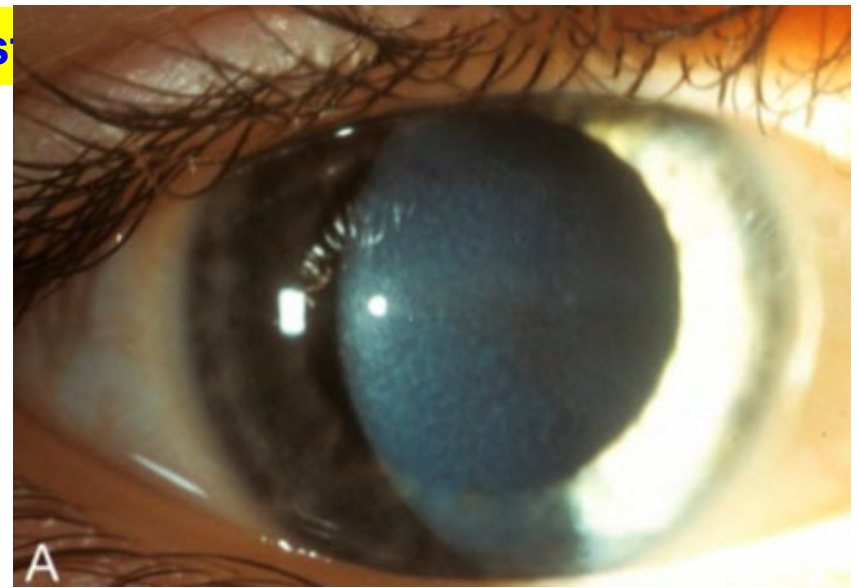
- | | |
|---|---------------------|
| 1) Reis-Bücklers corneal dystrophy | |
| 2) Thiel-Behnke corneal dystrophy | |
| 3) Lattice, type 1 | |
| 4) Lattice, type 2 | <i>Age of onset</i> |
| 5) Granular | |
| 6) Granular | <i>Genetics</i> |

		RBCD	TBCD
Stromal Dystrophies	2) Thiel-Behnke corneal dystrophy		
	3) Lattice, type 1		
	4) Lattice, variant		
	5) Granular		
Endothelial Dystrophies	6) Granular		



Reis-Bücklers corneal dystrophy:
Geographic opacities

Dys



Thiel-Benke corneal dystrophy:
Honeycomb appearance

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

- | | |
|---|---------------------|
| 1) Reis-Bücklers corneal dystrophy | |
| 2) Thiel-Behnke corneal dystrophy | |
| 3) Lattice, type 1 | |
| 4) Lattice, type 2 | <i>Age of onset</i> |
| 5) Granular | |
| 6) Granular | <i>Genetics</i> |

		RBCD	TBCD
Stromal Dystrophies	2) Thiel-Behnke corneal dystrophy		
	3) Lattice, type 1		
	4) Lattice, variant		
	5) Granular		
	6) Granular variant		
Endothelial Dystrophies	Age of onset	Childhood	Childhood
	Genetics	TGFBI (BIGH3)	TGFBI (BIGH3)
	SL appearance	'Geographic' opacification	'Honeycomb' opacification
	Painful?	?	?

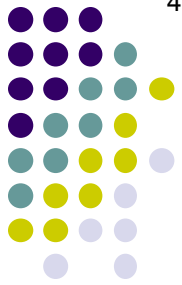
Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

- | | |
|---|---------------------|
| 1) Reis-Bücklers corneal dystrophy | |
| 2) Thiel-Behnke corneal dystrophy | |
| 3) Lattice, type 1 | |
| 4) Lattice, type 2 | <i>Age of onset</i> |
| 5) Granular | |
| 6) Granular | <i>Genetics</i> |

		RBCD	TBCD
Stromal Dystrophies	2) Thiel-Behnke corneal dystrophy		
	3) Lattice, type 1		
	4) Lattice, variant		
	5) Granular		
	6) Granular variant		
Endothelial Dystrophies	Age of onset	Childhood	Childhood
	Genetics	TGFBI (BIGH3)	TGFBI (BIGH3)
	SL appearance	'Geographic' opacification	'Honeycomb' opacification
	Painful?	Yes	Yes

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

1) **Reis-Bücklers** corneal dystrophy

2) **Thiel-Behnke** corneal dystrophy

3) Lattice, type 1

4) Lattice, type 2

5) Granular

6) Granular

RBCD

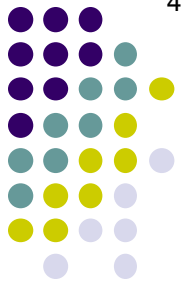
TBCD

Stromal Dystrophies

Endothelial Dystrophies

	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	?	?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

1) **Reis-Bücklers** corneal dystrophy

2) **Thiel-Behnke** corneal dystrophy

3) Lattice, type 1

4) Lattice, type 2

5) Granular

6) Granular

RBCD

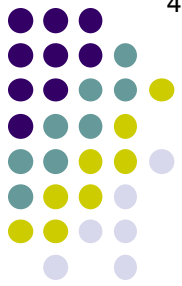
TBCD

Stromal Dystrophies

Endothelial Dystrophies

	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB1 (BIGH3)	TGFB1 (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

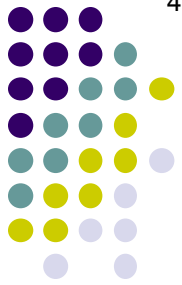
- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

Endothelial Dystrophies

		RBCD	TBCD
	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes
	<i>Stain/color</i>	?	

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

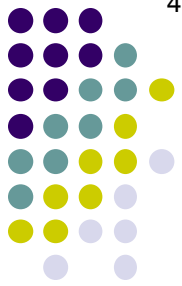
- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

Endothelial Dystrophies

		RBCD	TBCD
	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes
	<i>Stain/color</i>	Masson trichrome/Red	

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

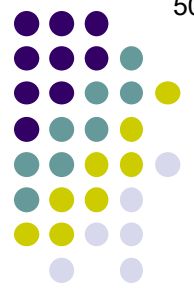
Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

Endothelial Dystrophies

		RBCD	TBCD
	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes
	<i>Stain/color</i>	Masson trichrome/Red	
	<i>Light microscopy appearance of Bowmans</i>	?	?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

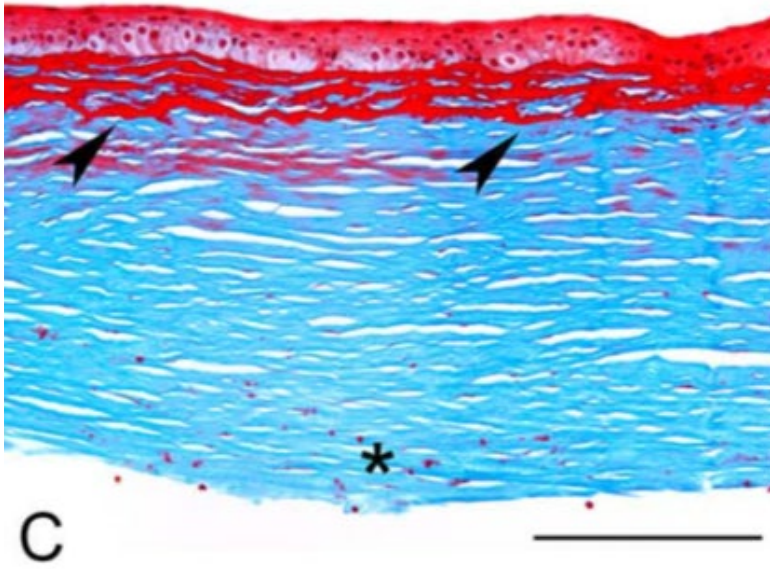
Endothelial Dystrophies

		RBCD	TBCD
	Age of onset	Childhood	Childhood
	Genetics	TGFB (BIGH3)	TGFB (BIGH3)
	SL appearance	‘Geographic’ opacification	‘Honeycomb’ opacification
	Painful?	Yes	Yes
	Affect vision?	Yes	Yes
	Stain/color	Masson trichrome/Red	
	Light microscopy appearance of Bowmans	‘Sheets’	‘Saw-toothed’

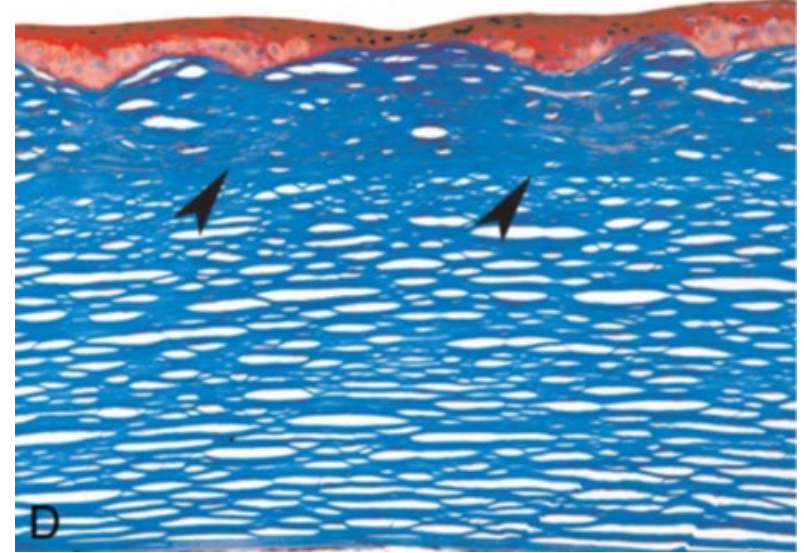
Reis-Bücklers: sheet-like layers

Corneal Dystrophies

Thiel-Behnke: Sawtooth pattern



Reis-Bücklers

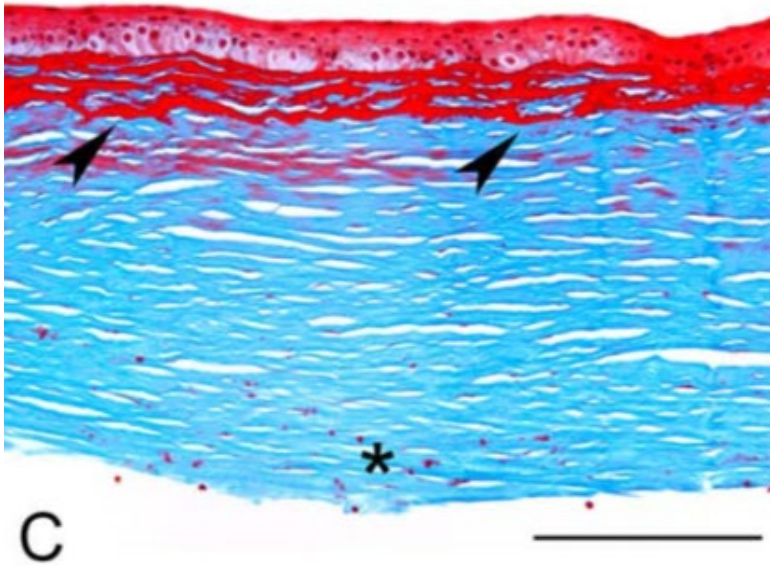


Thiel-Behnke

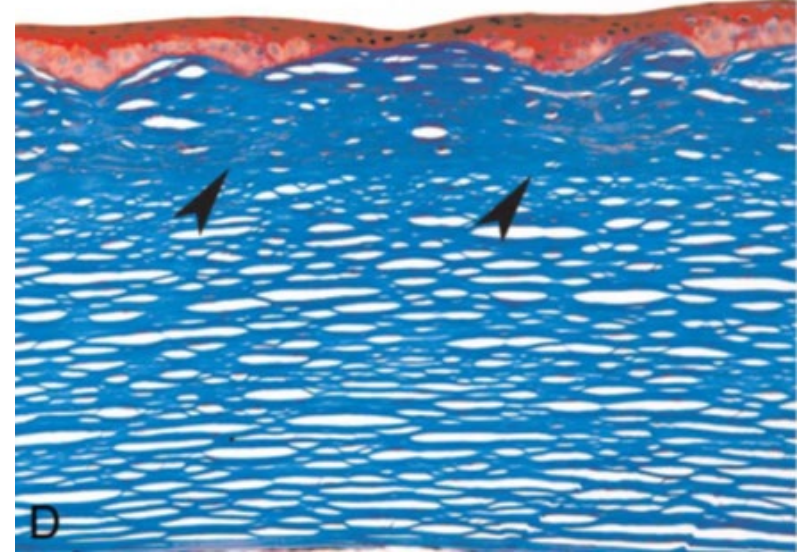
Reis-Bücklers: sheet-like layers

Corneal Dystrophies

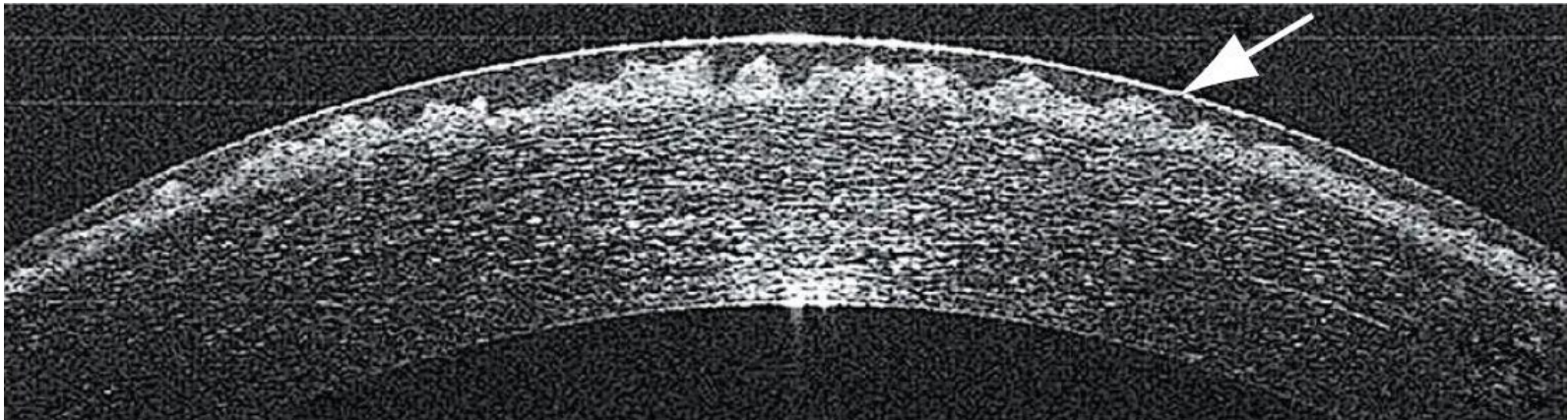
Thiel-Behnke: Sawtooth pattern



Reis-Bücklers

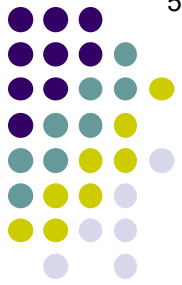


Thiel-Behnke



The sawtooth pattern in TBCD is also appreciable via anterior-segment OCT

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

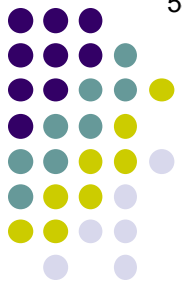
- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

Endothelial Dystrophies

		RBCD	TBCD
	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes
	<i>Stain/color</i>	Masson trichrome/Red	
	<i>Light microscopy appearance of Bowmans</i>	'Sheets'	'Saw-toothed'
	<i>Electron microscopy appearance</i>	?	?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, type 2
- 5) Granular
- 6) Granular

Stromal Dystrophies

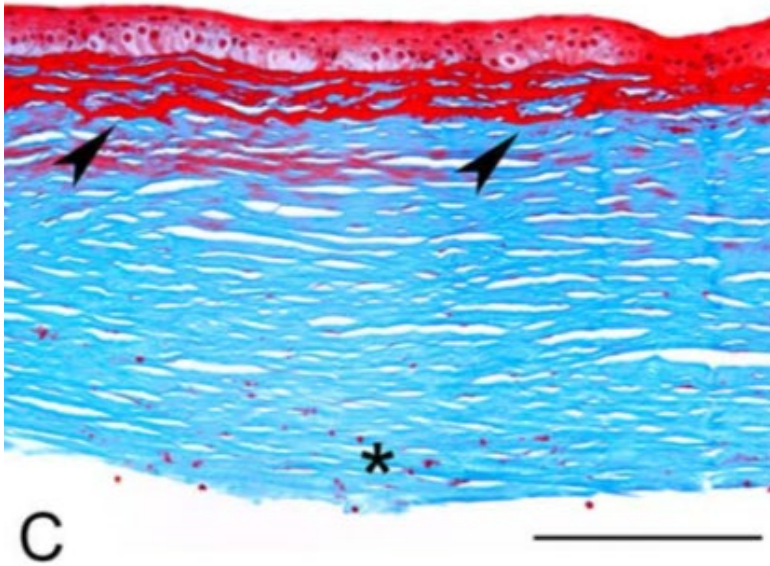
Endothelial Dystrophies

		RBCD	TBCD
	<i>Age of onset</i>	Childhood	Childhood
	<i>Genetics</i>	TGFB (BIGH3)	TGFB (BIGH3)
	<i>SL appearance</i>	'Geographic' opacification	'Honeycomb' opacification
	<i>Painful?</i>	Yes	Yes
	<i>Affect vision?</i>	Yes	Yes
	<i>Stain/color</i>	Masson trichrome/Red	
	<i>Light microscopy appearance of Bowmans</i>	'Sheets'	'Saw-toothed'
	<i>Electron microscopy appearance</i>	Rod-shaped fibers	Curly fibers

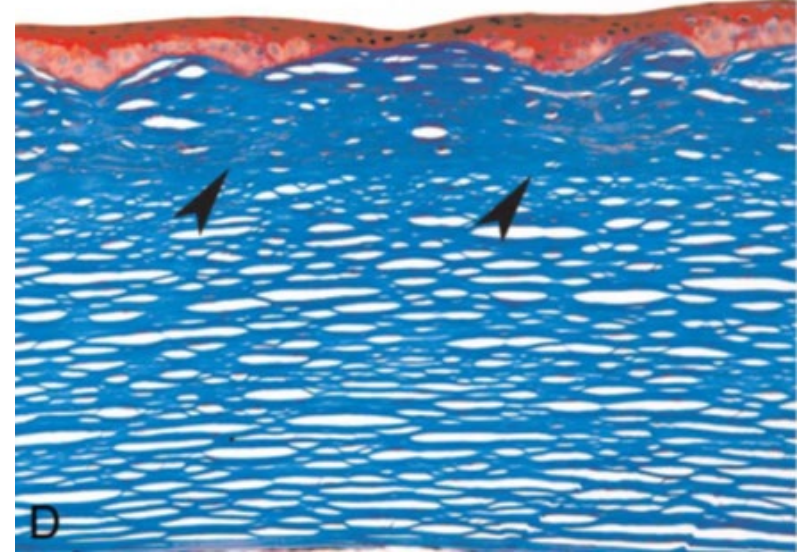
Reis-Bücklers: sheet-like layers

Corneal Dystrophies

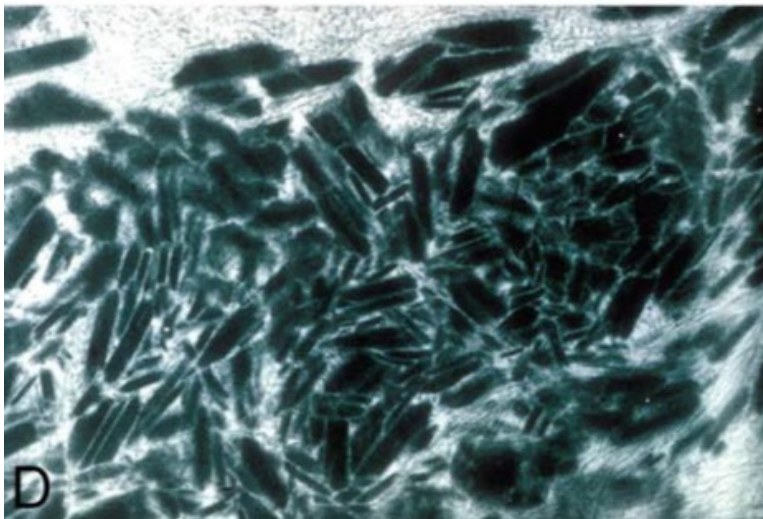
Thiel-Behnke: Sawtooth pattern



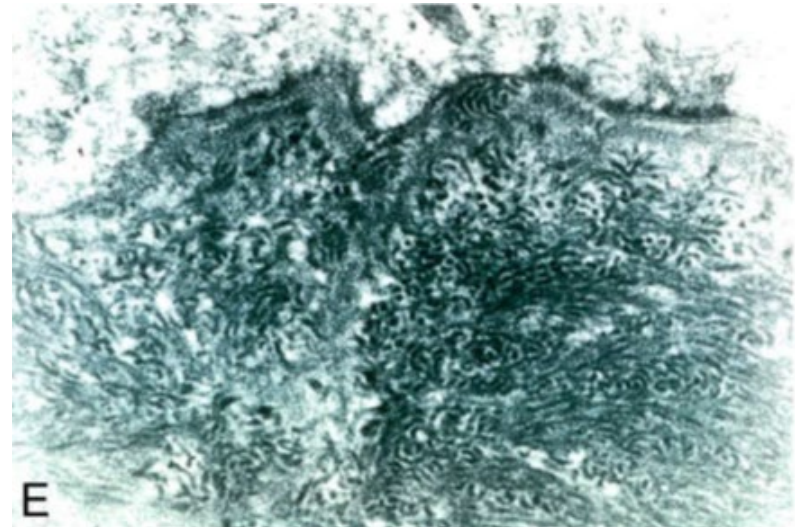
Reis-Bücklers



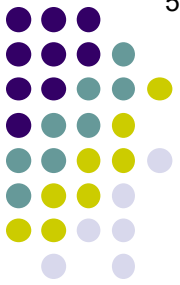
Thiel-Behnke



Reis-Bücklers: Rod-shaped fibers



Thiel-Behnke: Curly fibers



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

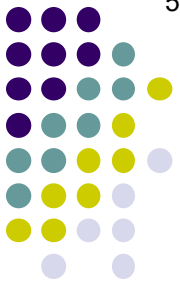
Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1

Can Reis-Bücklers and Thiel-Behnke be reliably differentiated from one another at the slit lamp?

Bowman's layer dystrophies: Which is which?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

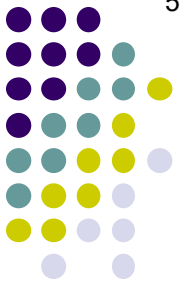
Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1

Can Reis-Bücklers and Thiel-Behnke be reliably differentiated from one another at the slit lamp?
The BCSC Cornea book says doing so is “difficult” (read: no, you can’t)

Bowman’s layer dystrophies: Which is which?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

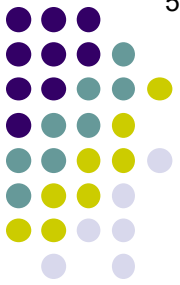
- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1

Can Reis-Bücklers and Thiel-Behnke be reliably differentiated from one another at the slit lamp?
The BCSC Cornea book says doing so is “difficult” (read: no, you can’t)

How can they be differentiated clinically?

Bowman’s layer dystrophies: Which is which?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) **Reis-Bücklers** corneal dystrophy
- 2) **Thiel-Behnke** corneal dystrophy
- 3) Lattice, type 1

Can Reis-Bücklers and Thiel-Behnke be reliably differentiated from one another at the slit lamp?
The BCSC Cornea book says doing so is “difficult” (read: no, you can’t)

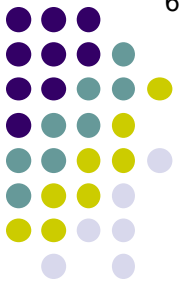
How can they be differentiated clinically?

Via anterior-segment OCT, and confocal microscopy

Bowman’s layer dystrophies: Which is which?

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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

Note: The *Cornea* book lumps together type 1 (aka 'classic lattice') and its variants

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

At what age does classic lattice type 1 begin to manifest?

Epithelial-Stromal *TGFB1* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

Epithelial-Stromal *TGFB1* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

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

Epithelial-Stromal *TGFB* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

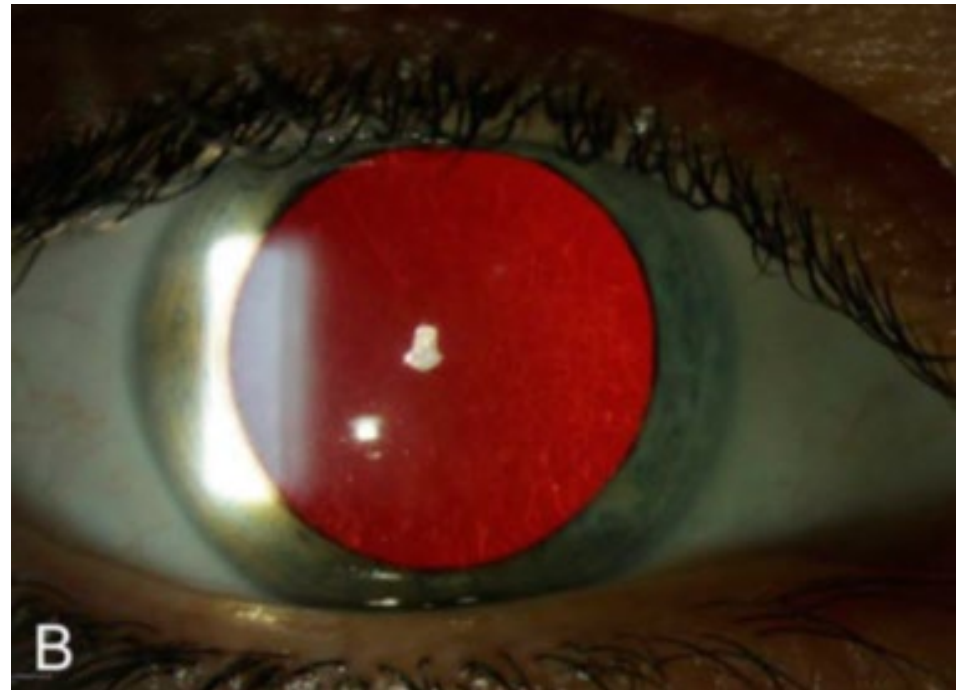
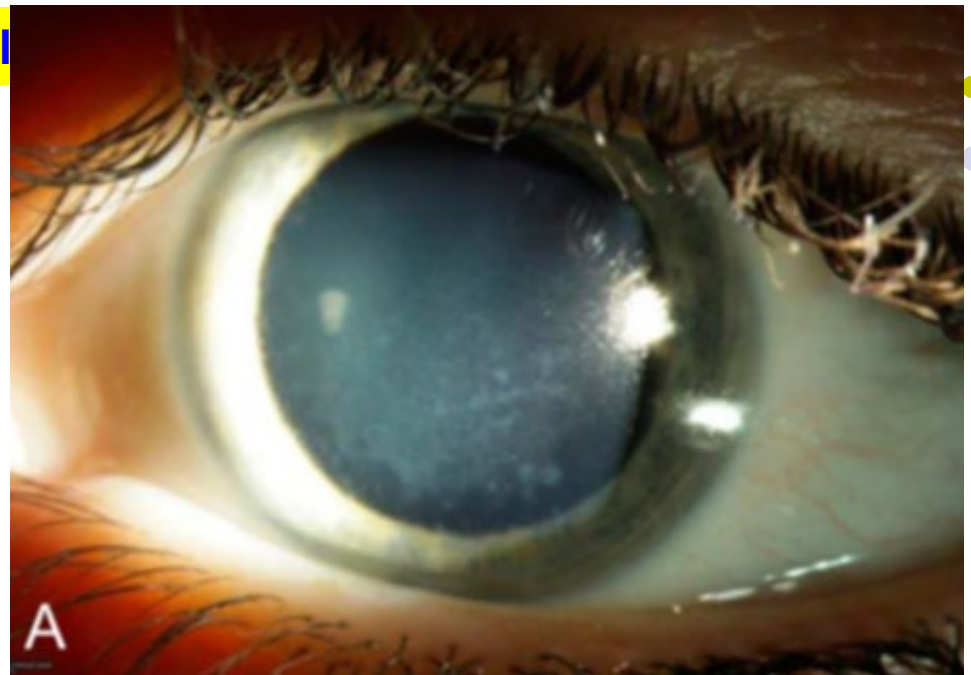
Stromal Dystrophies

Endothelial Dystrophies

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

How does it present? What is seen at the slit lamp?
Early in the disease, fleck-like central opacities and a diffuse stromal haze are present.

Lattice corneal dystrophy, type 1
(*aka* classic lattice). Direct (A) and
retroillumination (B) of early lattice
corneal dystrophy (LCD) with dots
and fine lattice lines.



Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

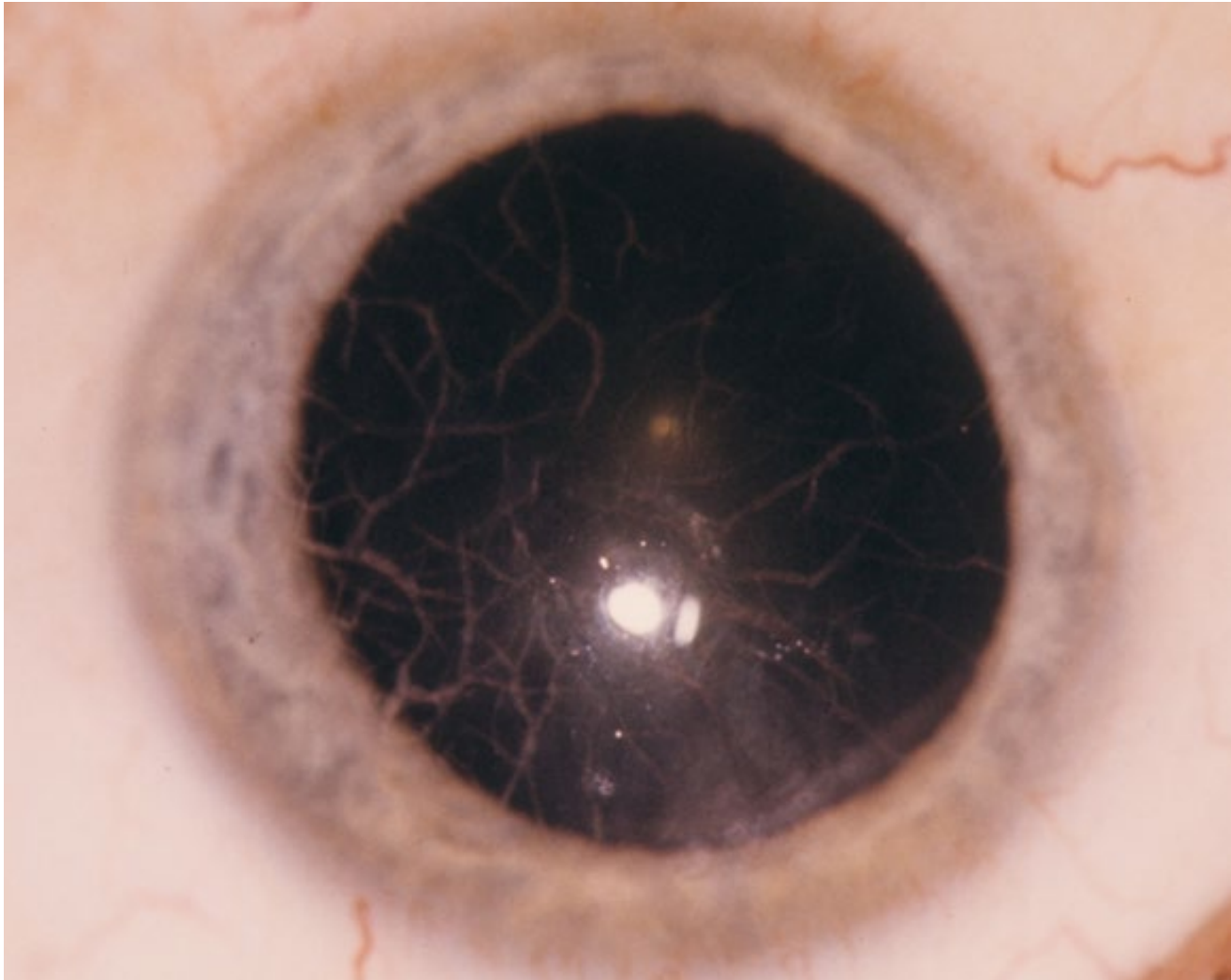
Stromal Dystrophies

Endothelial Dystrophies

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

How does it present? What is seen at the slit lamp?
Early in the disease, fleck-like central opacities and a diffuse stromal haze are present. Later, myriad crisscrossing refractile lines in the cornea predominate.

Corneal Dystrophies



Lattice corneal dystrophy, type 1 (*aka* classic lattice)



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

At what age does classic **lattice** type 1 begin to manifest?
Childhood to teens

Why is this condition called 'lattice'?

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

diffuse stromal haze are present. Later, myriad crisscrossing refractile lines in the cornea predominate.

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

At what age does classic **lattice** type 1 begin to manifest?
Childhood to teens

Why is this condition called 'lattice'?

Because the crisscrossing lines are reminiscent of a lattice structure (eg, like one might encounter in a garden)

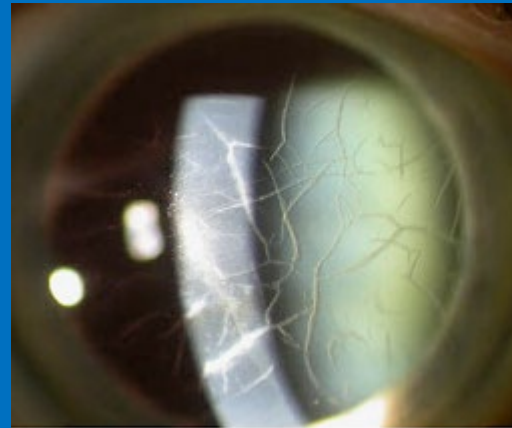
Epithelial-Stromal *TGFBI* Dystrophies

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

diffuse stromal haze are present. Later, myriads of crisscrossing refractile lines in the cornea predominate.

Stromal Dystrophies

Endothelial Dystrophies





Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

How does it present? What is seen at the slit lamp?
Early in the disease, fleck-like central opacities and a diffuse stromal haze are present. Later, myriad crisscrossing **refractile lines in the cornea** predominate.

What word is used to describe the appearance of the lines?

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) **Lattice, type 1**
- 4) **Lattice, variant types (III, IIIa)**
- 5) Granular type 1
- 6) Granular type 2

At what age does classic lattice type 1 begin to manifest?
Childhood to teens

How does it present? What is seen at the slit lamp?
Early in the disease, fleck-like central opacities and a diffuse stromal haze are present. Later, myriad crisscrossing **refractile lines in the cornea** predominate.

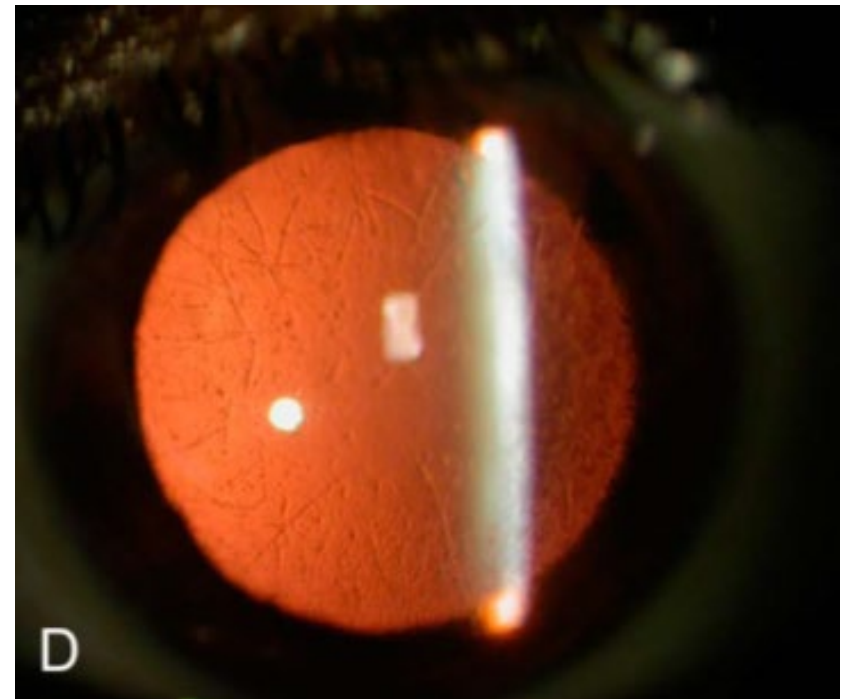
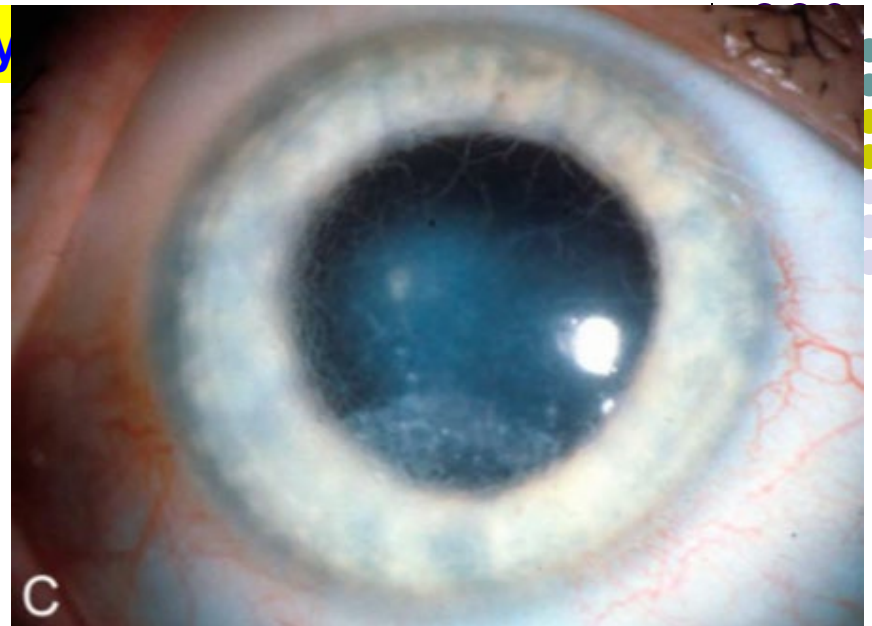
What word is used to describe the appearance of the lines?
'Glasslike'

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dy

Lattice corneal dystrophy, type 1
(*aka* classic lattice). C, Subepithelial groundglass haze of the central and inferior cornea, and diffuse lattice lines in advanced disease.
D, Dots and paracentral lattice lines in retroillumination.



Corneal Dystrophies



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Is it painful?

Corneal Dystrophies



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Corneal Dystrophies

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Surface irregularity and stromal haze often results in decreased vision



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Corneal Dystrophies



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What is the hallmark of lattice type 1 on light microscopy?
The presence of substance in the subepithelial space and anterior stroma

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Stromal Dystrophies

Endothelial Dystrophies

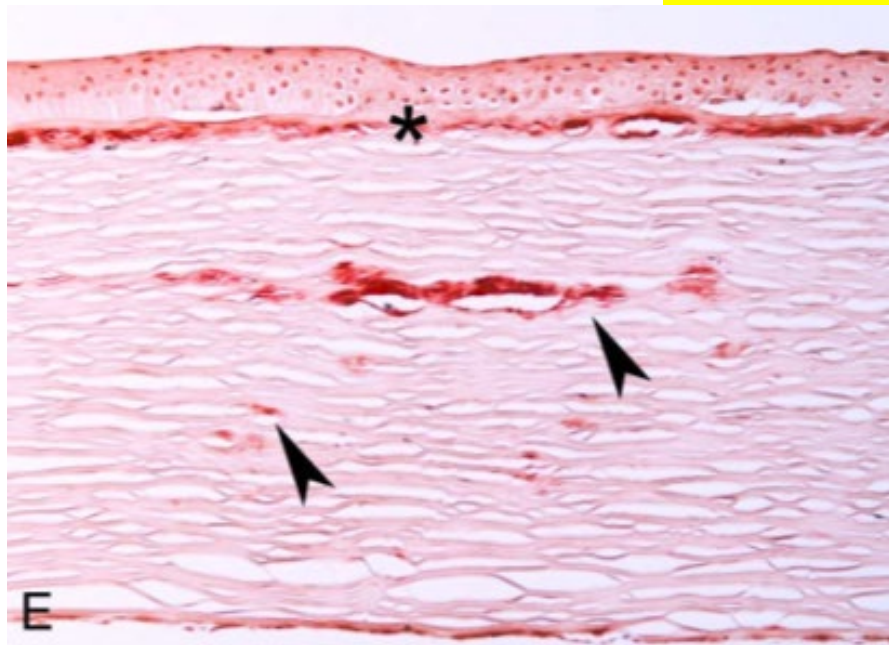
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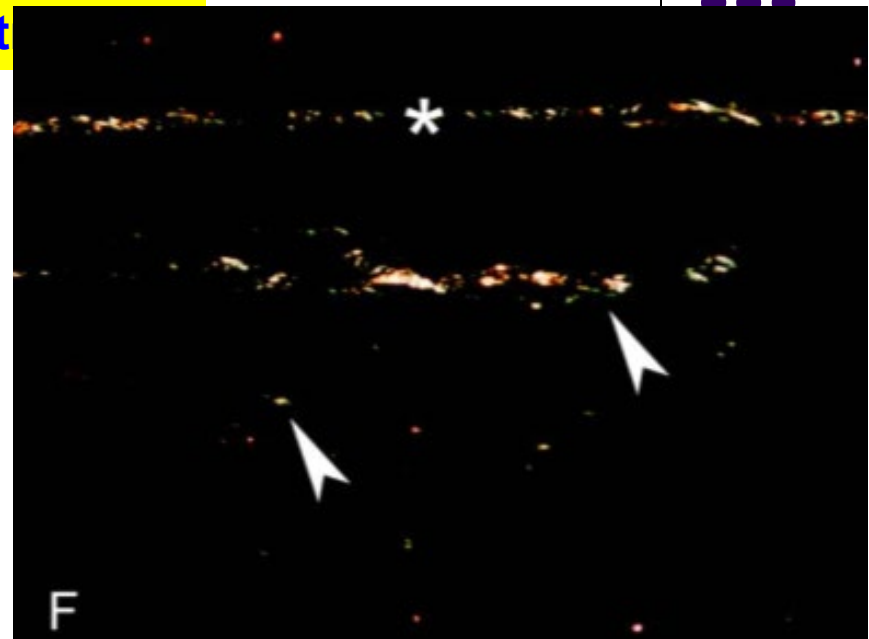
Is it painful?
Yes; pts get recurrent epithelial erosions

Does it affect vision?
Surface irregularity and stromal haze often results in decreased vision

What is the hallmark of lattice type 1 on light microscopy?
The presence of **amyloid** in the subepithelial space and anterior stroma

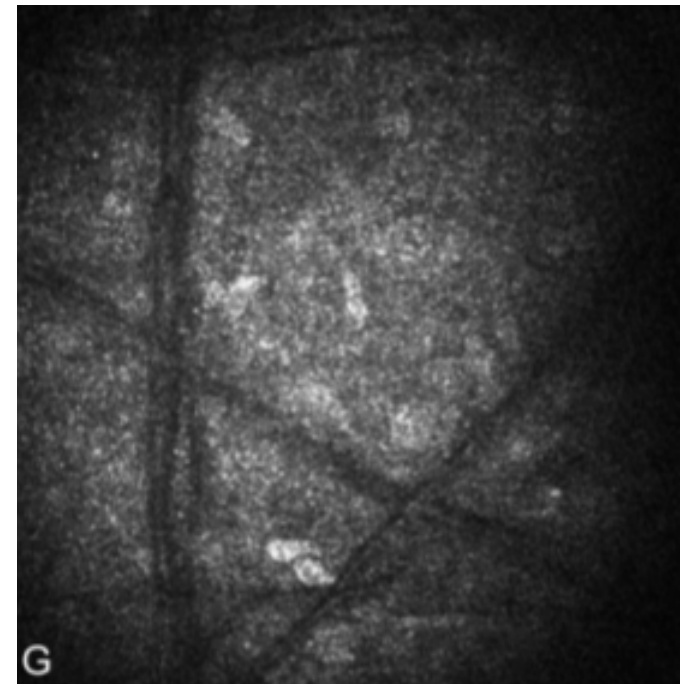


Dyst

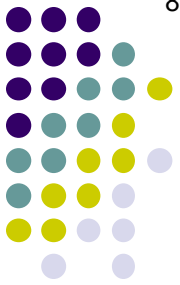


Lattice corneal dystrophy, type 1 (classic lattice).

E, Light microscopy: Congo red prominently stains a continuous layer of amyloid (asterisk) that underlies and partially destroys the Bowman layer and intrastromal amyloid deposits corresponding to lattice lines (arrowheads). F, This same section viewed with polarized light confirms deposits are birefringent and red-green dichroic, thus amyloid. G, In vivo confocal microscopy image shows filaments corresponding to lattice lines within the stroma.



Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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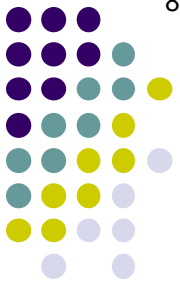
Lattice, gelsolin type

Why is lattice, gelsolin type not grouped with lattice?

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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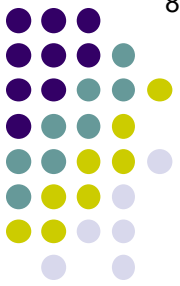
Lattice, gelsolin type

Stromal Dystrophies

Endothelial Dystrophies

Why is lattice, gelsolin type not grouped with lattice?
Because it is part of a **systemic syndrome**, it is no longer classified as a corneal dystrophy. (But given its corneal findings, we will touch briefly on it here.)

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB1* Dystrophies

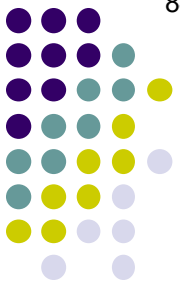
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Stromal Dys (aka... *Lattice, gelsolin type syndrome*)

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What is the name of the syndrome?

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

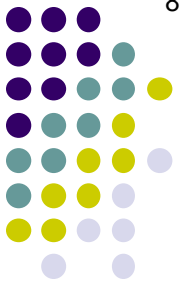
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Lattice, gelsolin type
 Stromal Dys (aka...Meretoja syndrome)

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Meretoja syndrome

Endothelial Dystrophies



Corneal Dystrophies

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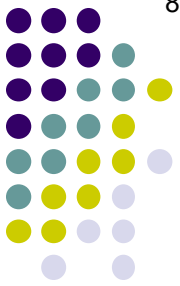
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Endothelial Dystrophies



Corneal Dystrophies

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Lattice, gelsolin type
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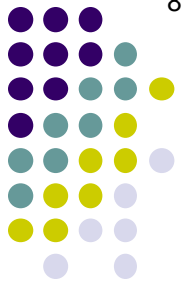
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What are the systemic findings in Meretoja syndrome?
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Endothelial Dystrophies

Corneal Dystrophies



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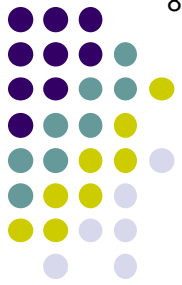
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Corneal Dystrophies



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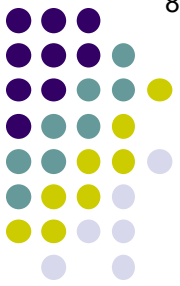
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'two words'

Endothelial Dystrophies

Corneal Dystrophies



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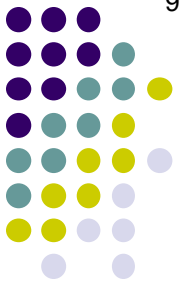
Endothelial Dystrophies

Corneal Dystrophies



Meretoja syndrome: '*Bloodhound facies*'

Corneal Dystrophies



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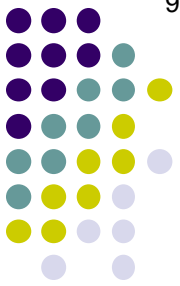
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Endothelial Dystrophies

What about the corneal findings?



Corneal Dystrophies

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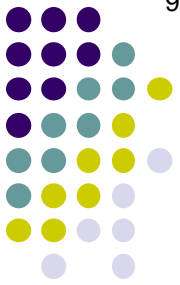
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Endothelial Dystrophies

What about the corneal findings?
 They are essentially identical to those of lattice type 1

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

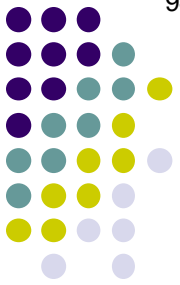
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Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

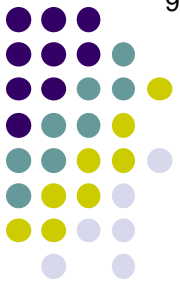
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Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?
Early childhood



Corneal Dystrophies

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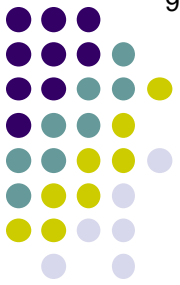
At what age does GCD1 begin to manifest?

Early childhood

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

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

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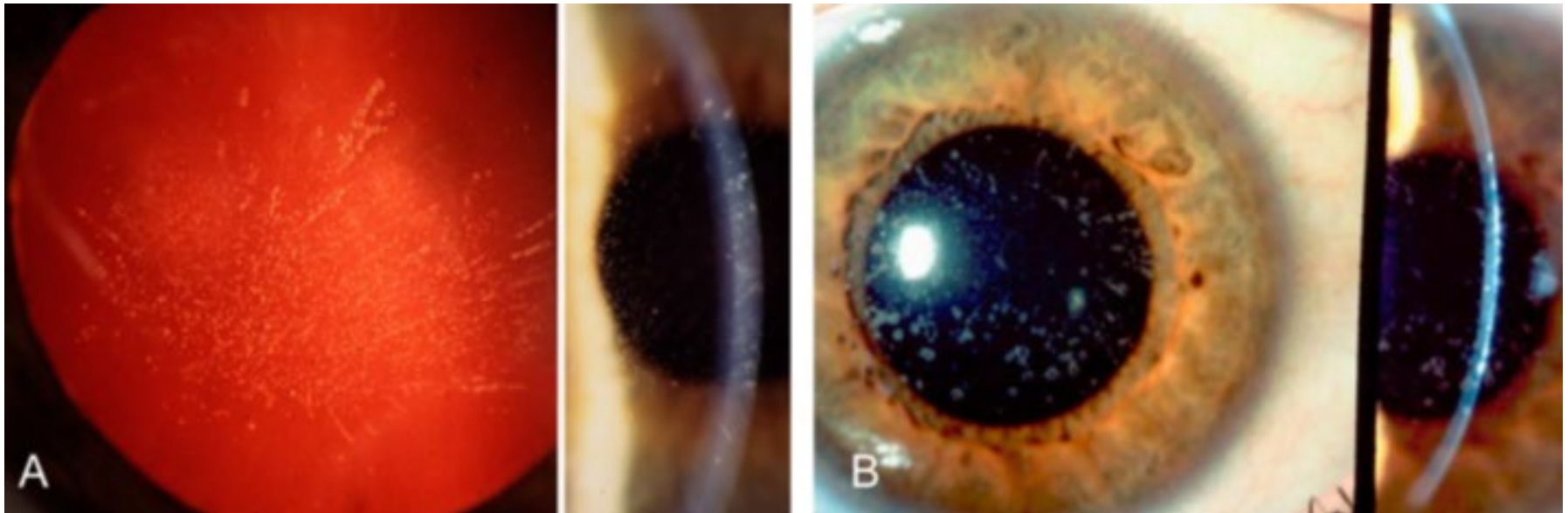
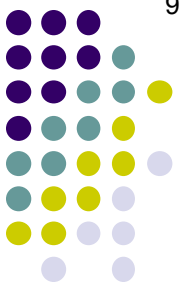
Early childhood

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

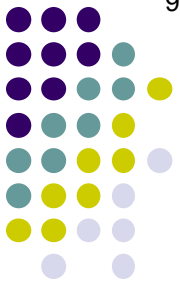
In early disease, tiny crumblike granules appear.

Stromal Dystrophies

Endothelial Dystrophies



Granular corneal dystrophy, type 1. A, In a child, early subepithelial verticillate-like opacities are evident by retro and direct illumination. B, With broad slit illumination, stromal deposits are both discrete and confluent, and are axially distributed in anterior stroma.



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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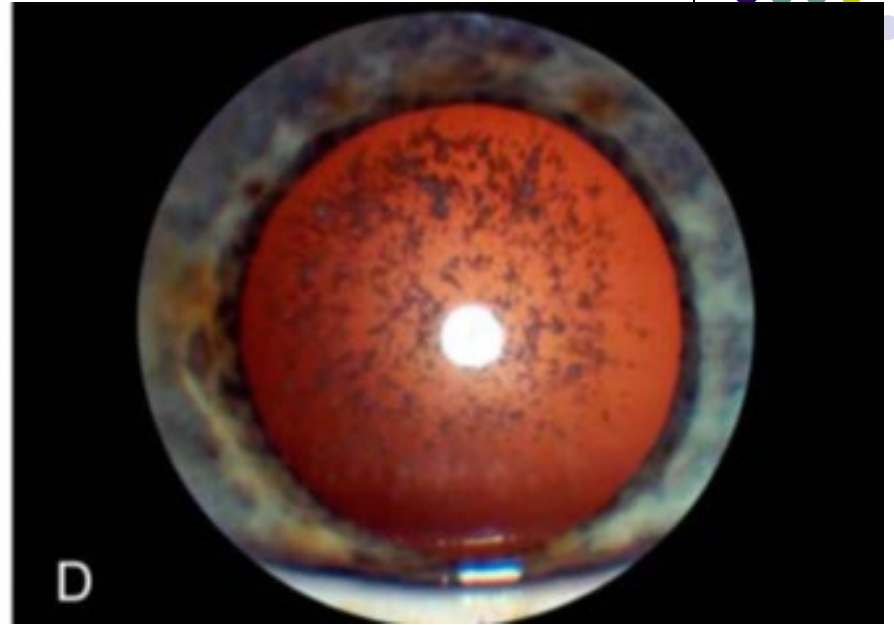
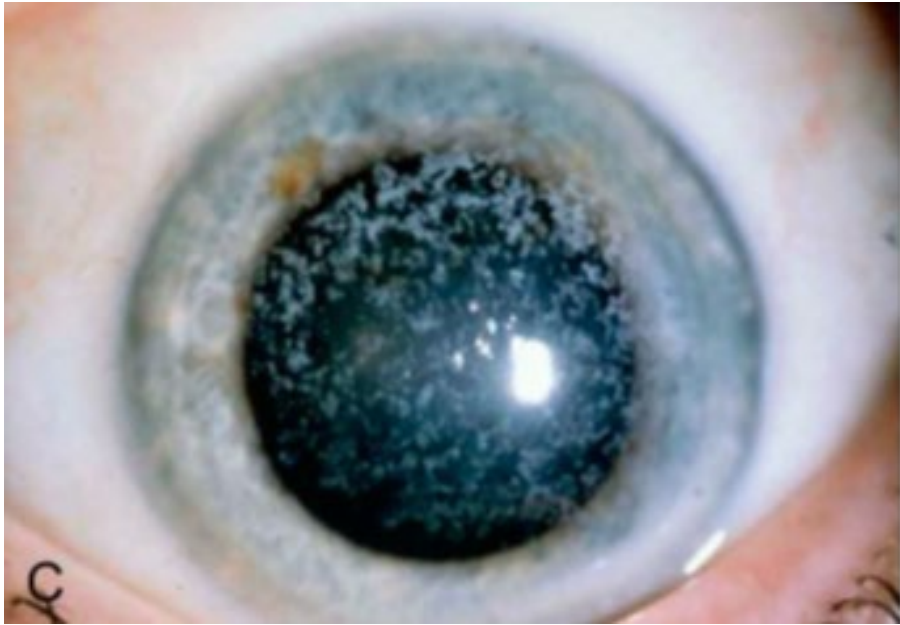
How does it present? What is seen at the slit lamp?

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

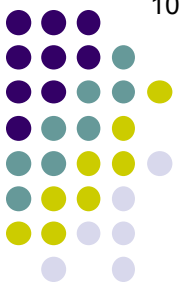
Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Granular corneal dystrophy, type 1. In an adult, more prominent diffuse granular opacities in the form of “snowfall” are apparent with direct (C) and retroillumination (D).



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

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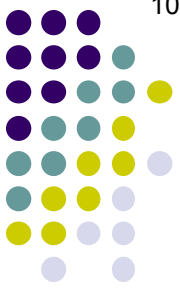
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Is it painful?

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (Lattice type 2)
- 5) **Granular type 1**
- 6) Granular type 2

At what age does GCD1 begin to manifest?

Early childhood

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

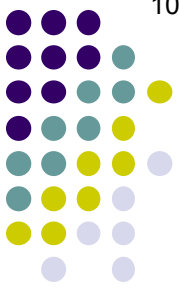
In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Stromal Dystrophies

Endothelial Dystrophies



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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- 3) Lattice, type 1
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- 5) **Granular type 1**
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Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

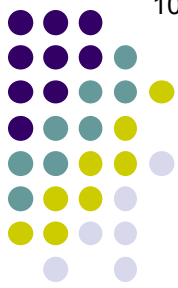
In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Does it affect vision?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal
- 2) Thiel-Behnke corneal
- 3) Lattice, type 1
- 4) Lattice, variant types (I)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

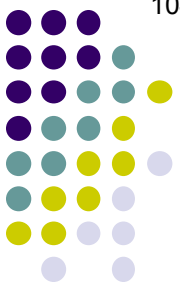
In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Does it affect vision?

Glare and stromal haze result in decreased vision



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFB* Dystrophies

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- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (Lattice type 2)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

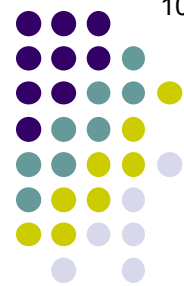
Is it painful?

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Does it affect vision?

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What is the histologic hallmark of GCD1 on light microscopy?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

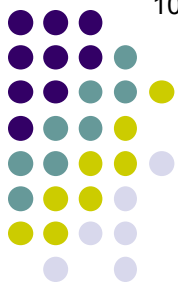
Does it affect vision?

Glare and stromal haze result in decreased vision

What is the histologic hallmark of GCD1 on light microscopy?

The presence of one word in the subepithelial space and anterior stroma (just like Reis-Bücklers). It stains bright color with two words (just like Reis-Bücklers).

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (type 2, 3, 4)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

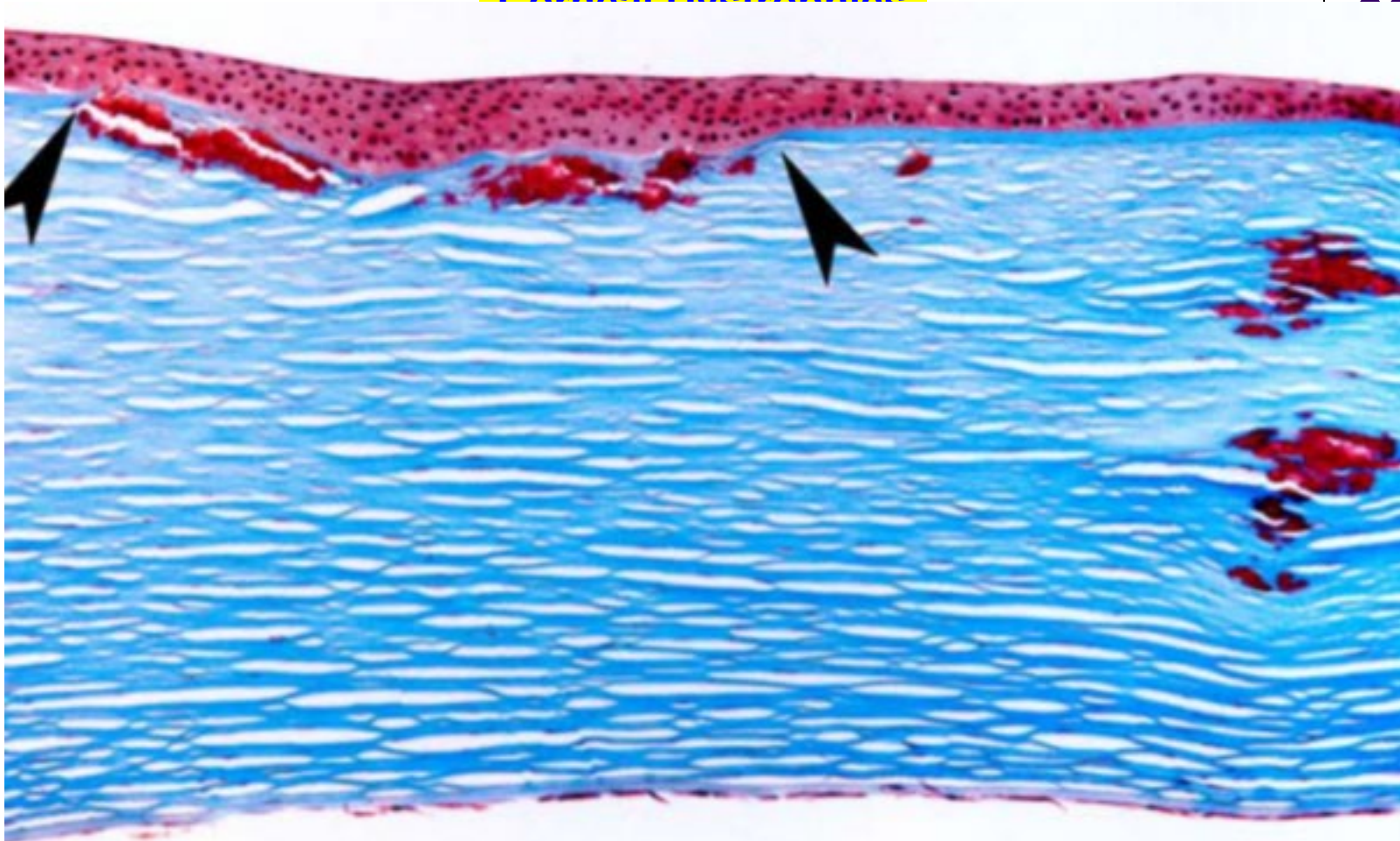
Does it affect vision?

Glare and stromal haze result in decreased vision

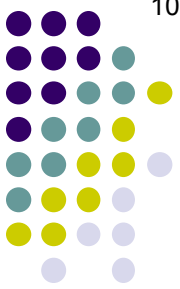
What is the histologic hallmark of GCD1 on light microscopy?

The presence of **hyaline** in the subepithelial space and anterior stroma (just like Reis-Bücklers). It stains bright **red** with **Masson trichrome** (just like Reis-Bücklers).

Corneal Dystrophies



Granular corneal dystrophy, type 1. Light microscopy—Masson trichrome highlights deposits of hyaline at various stromal layers and partial destruction of the Bowman layer (between arrowheads).



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (Lattice type 2)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Does it affect vision?

Glare and stromal haze result in decreased vision

What is the histologic hallmark of GCD1 on light microscopy?

The presence of **hyaline** in the subepithelial space and anterior stroma (just like Reis-Bücklers). It stains bright **red** with **Masson trichrome** (just like Reis-Bücklers).

What is the hallmark of GCD1 on electron microscopy?



Corneal Dystrophies

Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (Lattice type 2)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Does it affect vision?

Glare and stromal haze result in decreased vision

What is the histologic hallmark of GCD1 on light microscopy?

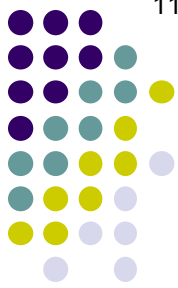
The presence of **hyaline** in the subepithelial space and anterior stroma (just like Reis-Bücklers). It stains bright **red** with **Masson trichrome** (just like Reis-Bücklers).

What is the hallmark of GCD1 on electron microscopy?

three words

(just like Reis-Bücklers)

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (1 and 2)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?

Early childhood

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

In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?

Yes; pts get recurrent epithelial erosions

Does it affect vision?

Glare and stromal haze result in decreased vision

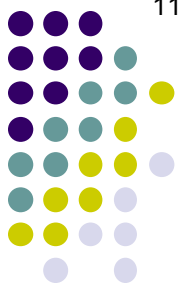
What is the histologic hallmark of GCD1 on light microscopy?

The presence of **hyaline** in the subepithelial space and anterior stroma (just like Reis-Bücklers). It stains bright **red** with **Masson trichrome** (just like Reis-Bücklers).

What is the hallmark of GCD1 on electron microscopy?

Rod-shaped fibers (just like Reis-Bücklers)

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers corneal dystrophy
- 2) Thiel-Behnke corneal dystrophy
- 3) Lattice, type 1
- 4) Lattice, variant types (Lattice A, B, C)
- 5) **Granular type 1**
- 6) Granular type 2

Stromal Dystrophies

Endothelial Dystrophies

At what age does GCD1 begin to manifest?
Early childhood

How does it present? What is seen at the slit lamp?
In early disease, tiny crumblike granules appear. As the disease progresses, the size and density of the granules increases. The granules never reach as far as the limbus.

Is it painful?
Yes; pts get recurrent epithelial erosions

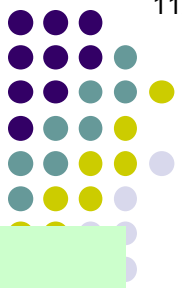
Does it affect vision?
Glare and stromal haze result in decreased vision

What is the histologic hallmark of GCD1 on light microscopy?
The presence of **hyaline** in the subepithelial space and anterior stroma (**just like Reis-Bücklers**). It stains bright red with **Masson trichrome** (**just like Reis-Bücklers**).

What is the hallmark of GCD1 on electron microscopy?
Rod-shaped fibers (**just like Reis-Bücklers**)



Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

By what other names is GCD2 known?

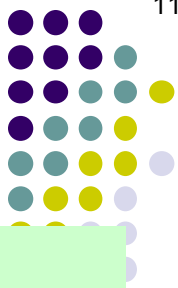
Epithelial-Stromal *TGFB1* Dystrop

- 1) Reis-Bücklers cornea
- 2) Thiel-Behnke corneal
- 3) Lattice, type 1
- 4) Lattice, variant types
- 5) Granular type 1
- 6) **Granular type 2**

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

By what other names is GCD2 known?

Combined granular-lattice dystrophy; Avellino dystrophy

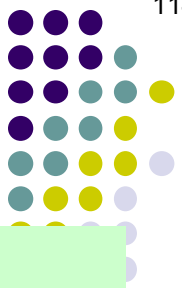
Epithelial-Stromal *TGFB1* Dystrophies

- 1) Reis-Bücklers cornea
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- 3) Lattice, type 1
- 4) Lattice, variant types
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- 6) **Granular type 2**

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

By what other names is GCD2 known?

Combined granular-lattice dystrophy; Avellino dystrophy

At what age does GCD2 begin to manifest?

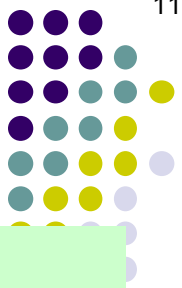
Epithelial-Stromal *TGFBI* Dystrophies

- 1) Reis-Bücklers cornea
- 2) Thiel-Behnke corneal
- 3) Lattice, type 1
- 4) Lattice, variant types
- 5) Granular type 1
- 6) **Granular type 2**

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

By what other names is GCD2 known?

Combined granular-lattice dystrophy; Avellino dystrophy

At what age does GCD2 begin to manifest?

Usually in the teen to early-adult years, can be younger

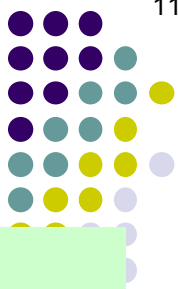
Epithelial-Stromal *TGFB* Dystrop

- 1) Reis-Bücklers cornea
- 2) Thiel-Behnke corneal
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- 4) Lattice, variant types
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- 6) **Granular type 2**

Stromal Dystrophies

Endothelial Dystrophies

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrop

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- 5) Granular type 1
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Stromal Dystrophies

Endothelial Dystrophies

By what other names is GCD2 known?

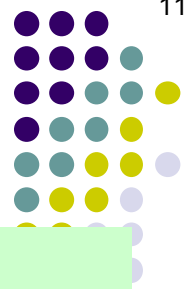
Combined granular-lattice dystrophy; Avellino dystrophy

At what age does GCD2 begin to manifest?

Usually in the teen to early-adult years, can be younger

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

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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By what other names is GCD2 known?

Combined granular-lattice dystrophy; Avellino dystrophy

At what age does GCD2 begin to manifest?

Usually in the teen to early-adult years, can be younger

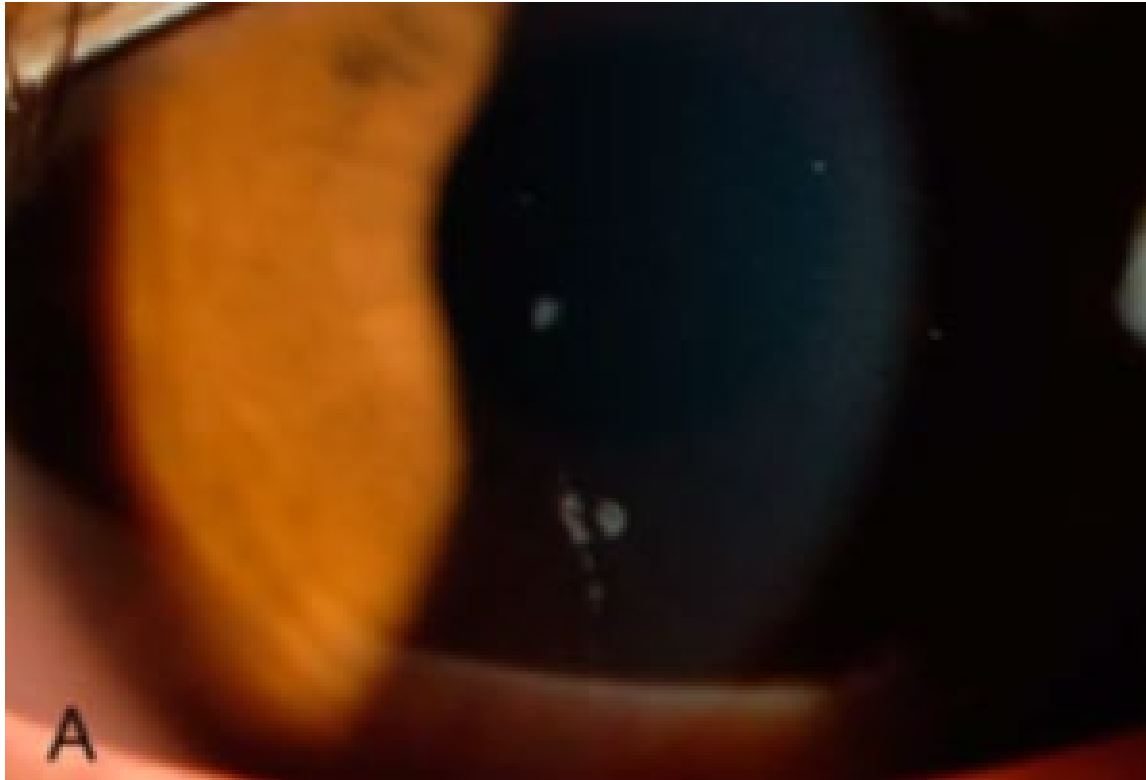
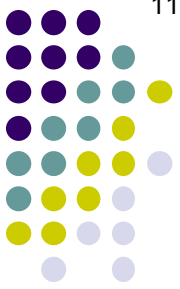
How does it present? What is seen at the slit lamp?

Basically, as a combination of GCD1 and lattice dystrophy:

The crumblike granules of GCD1, along with a version of the lattice lines seen in LCD

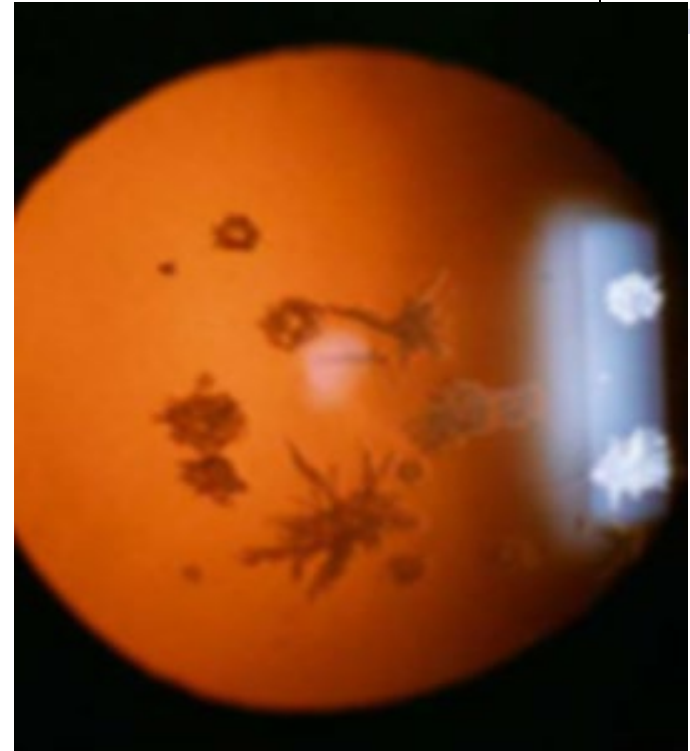
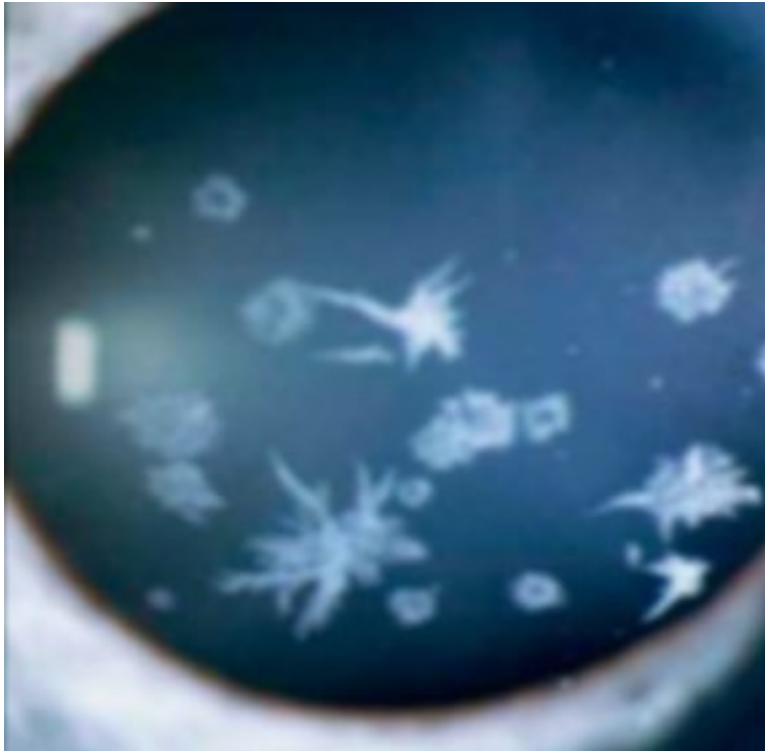
Stromal Dystrophies

Endothelial Dystrophies



Granular corneal dystrophy, type 2. 13-year-old with a few white dots.

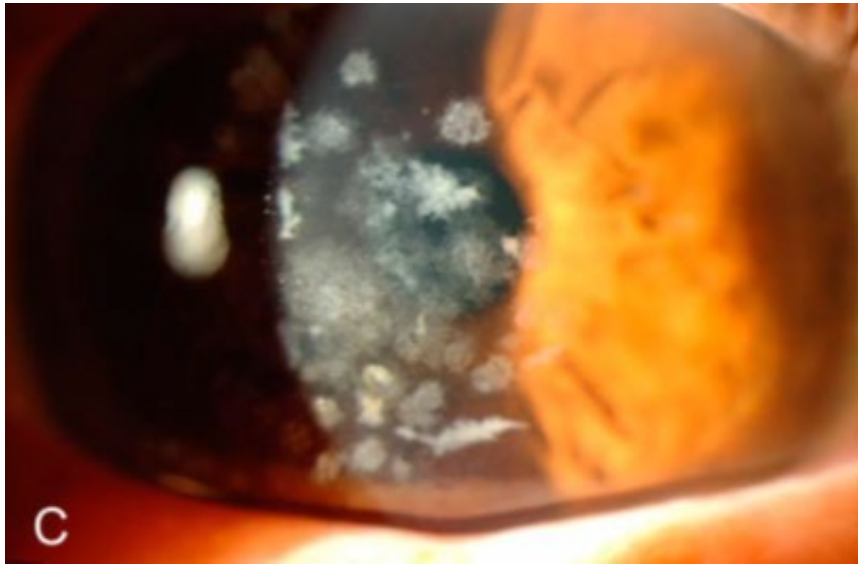
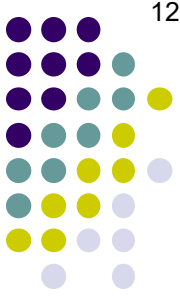
Corneal Dystrophies



Granular corneal dystrophy, type 2. Direct and retro-illumination views of an older pt demonstrate branching, star-shaped, spiny, and ring-like deposits.

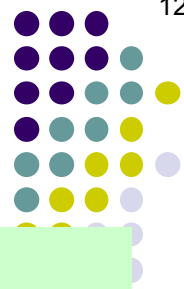
Corneal Dystrophies

120



Granular corneal dystrophy, type 2. C, pt with superficial translucent flattened breadcrumb opacity beneath the Bowman layer. Denser icicles and disc-like and ring-like opacities. D, Homozygote with denser and confluent opacities.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrop

- 1) Reis-Bücklers cornea
- 2) Thiel-Behnke corneal
- 3) Lattice, type 1
- 4) Lattice, variant types
- 5) Granular type 1
- 6) **Granular type 2**

Stromal Dystrophies

Endothelial Dystrophies

By what other names is GCD2 known?

Combined granular-lattice dystrophy; Avellino dystrophy

At what age does GCD2 begin to manifest?

Usually in the teen to early-adult years, can be younger

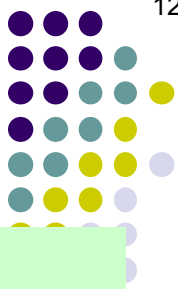
How does it present? What is seen at the slit lamp?

Basically, as a combination of GCD1 and lattice dystrophy:

The crumblike granules of GCD1, along with a version of the lattice lines seen in LCD

Is it painful?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Endothelial Dystrophies

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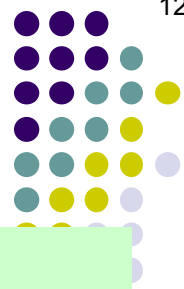
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Is it painful?

Yes; pts get recurrent epithelial erosions

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Stromal Dystrophies

Endothelial Dystrophies

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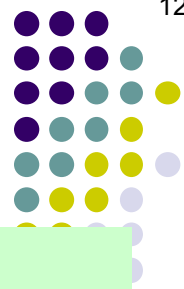
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Does it affect vision?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

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Stromal Dystrophies

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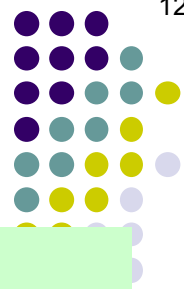
Is it painful?

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Does it affect vision?

Glare and stromal haze result in decreased vision

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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- 2) Thiel-Behnke corneal
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Is it painful?

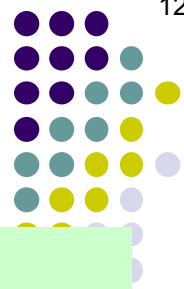
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What is the histologic hallmark of GCD1 on light microscopy?

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Stromal Dystrophies

Endothelial Dystrophies

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Basically, as a combination of GCD1 and lattice dystrophy:

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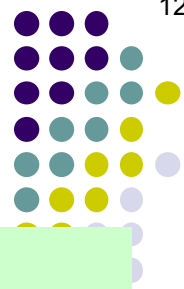
Does it affect vision?

Glare and stromal haze result in decreased vision

What is the histologic hallmark of GCD1 on light microscopy?

The presence of both and in the subepithelial space and anterior stroma.

Corneal Dystrophies



Epithelial and Subepithelial Dystrophies

Epithelial-Stromal *TGFBI* Dystrophies

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Stromal Dystrophies

Endothelial Dystrophies

By what other names is GCD2 known?

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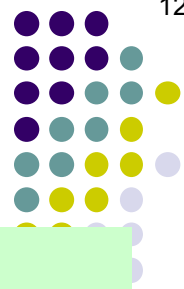
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Epithelial and Subepithelial Dystrophies

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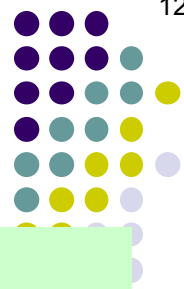
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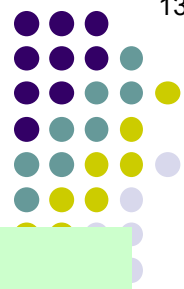
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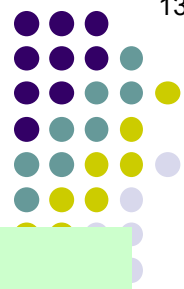
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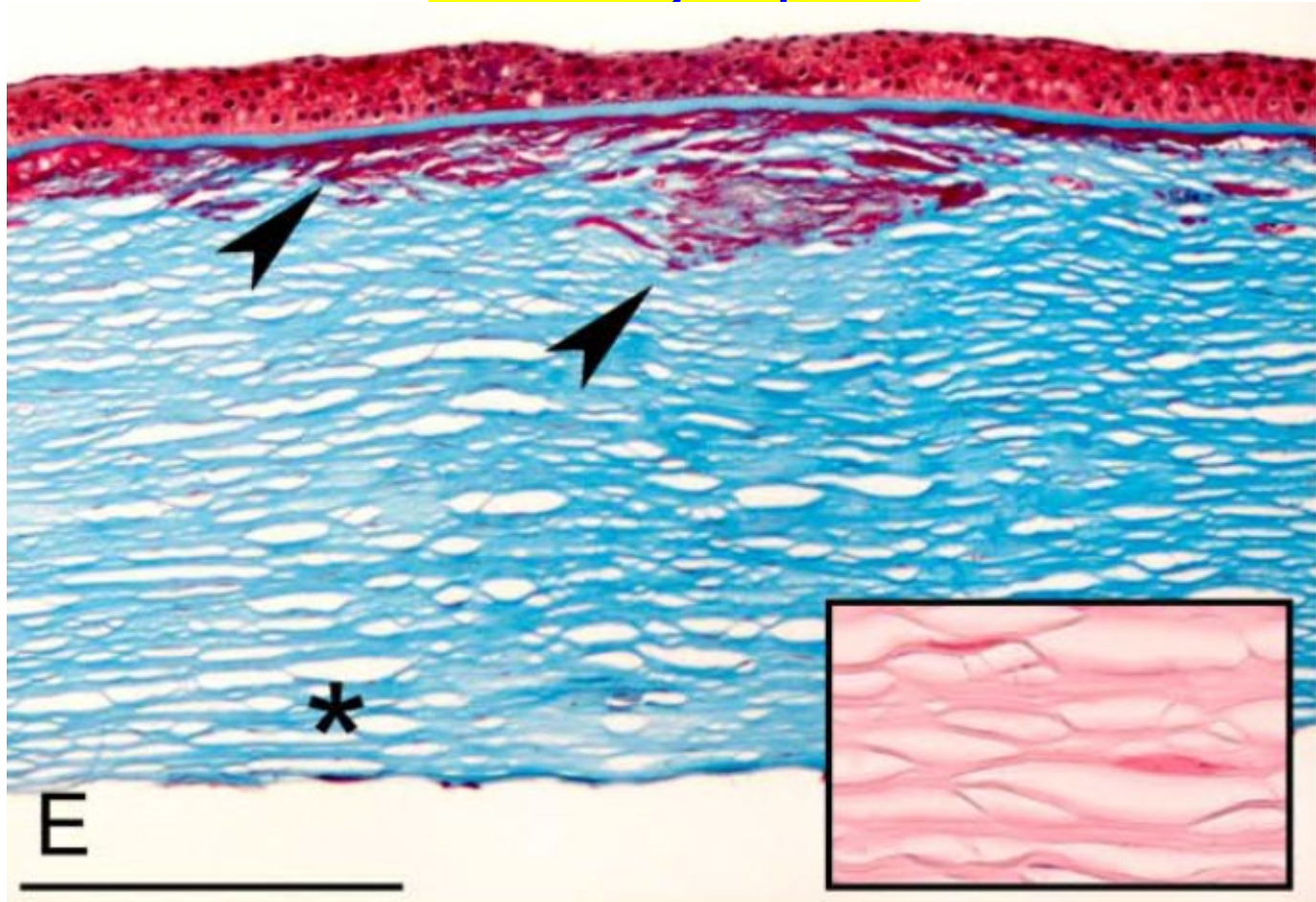
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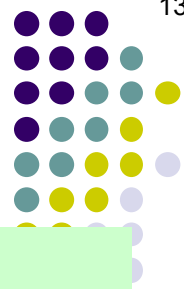
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Corneal Dystrophies



Granular corneal dystrophy, type 2. E, Light microscopy—Masson trichrome stains sub-Bowman and anterior stromal hyaline deposits red (arrowheads). Note that the deeper stromal layers do not have hyaline granules (asterisk). In the deep stroma, small amyloid deposits stain with Congo red (inset).

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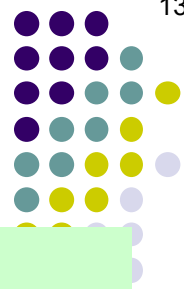
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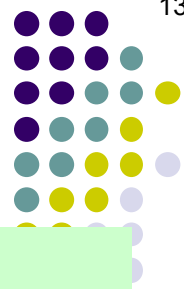
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three words

, albeit in reduced numbers

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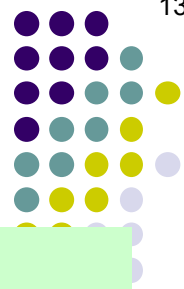
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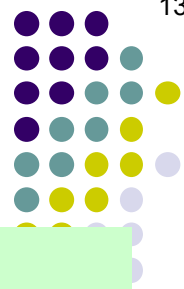
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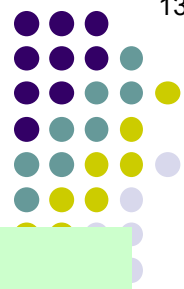
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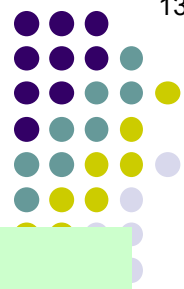
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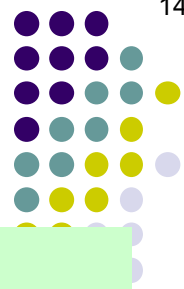
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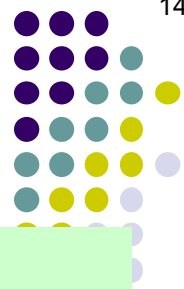
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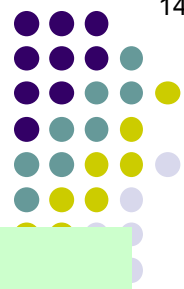
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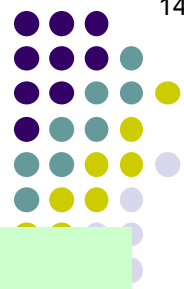
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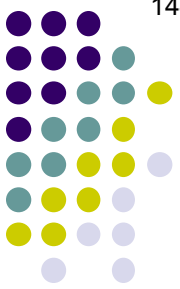
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- These two primarily involve Bowman's membrane
- There are two 'lattice' forms
- Two are 'granular' forms

Stromal Dy: As a way to help remember them, think of the *TGFB* dystrophies as consisting of three pairs of conditions:
 Two **Bowman's** dystrophies, two **lattice** dystrophies,
 and two **granular** dystrophies

Endothelial Dystrophies