

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

What is the purpose of the mnemonic, ie, what does it help to remember?



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

What is the purpose of the mnemonic, ie, what does it help to remember?
The DDx for a cloudy cornea in an infant



S

Start here

- 1
- U
- M
- P
- E
- D



- Sclerocornea
- T

Next

- U
- M
- P
- E
- D



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- (Tears in Descemet's membrane works too)
- M
- P
- E
- D



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- U
- M
- P
- E
- D





- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- M
- P
- E
- D





- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- F
- E
- D





- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- **E**
- D





- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)

(CHED = congenital hereditary endothelial dystrophy)

(*Edema* works too)



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- D



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



How does sclerocornea present?

- Sclerocornea
- Trauma (endot
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea

How does sclerocornea present?

The name says it all—the cornea looks like sclera

14

- Sclerocornea
- Trauma (endot
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea





Sclerocornea



How does sclerocornea present?

The name says it all—the cornea looks like sclera?

- Sclerocornea
 - Is it the case that corneal tissue has been literally replaced by scleral tissue?
- Trauma (e
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



How does sclerocornea present?

The name says it all—the cornea looks like sclera?

- Scierocorpes

 Is it the case that corneal tissue has been literally replaced by scieral tissue?
- Trauma (e
- Ulcer
- Depends on who you ask, unfortunately. The *Cornea* book asserts that such corneas have undergone "scleralization." However, the *Peds* book states that the cornea merely 'resembles' sclera, pointedly noting that the term 'sclerocornea' is "descriptive" and "does not suggest causation."
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



How does sclerocornea present?

The name says it all—the cornea looks like sclera?

Sclerocorpea

Trauma

Ulcer

Is it the case that corneal tissue has been literally replaced by scleral tissue? Depends on who you ask, unfortunately. The *Cornea* book asserts that such corneas have undergone "scleralization." However, the *Peds* book states that the cornea merely 'resembles' sclera, pointedly noting that the term 'sclerocornea' is "descriptive" and "does not suggest causation." In fact, the *Peds* book goes so far as to suggest the term should be avoided entirely. Caveat emptor.

- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea

19

- Sclerocornea
- Trauma (endot
- Ulcer
- Metabolic disorders
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- Dermoid of the cornea

How does sclerocornea present?
The name says it all—the cornea looks like sclera

Does it present unilaterally, or bilaterally?



- Sclerocornea
- Trauma (endot
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea

How does sclerocornea present?
The name says it all—the cornea looks like sclera

Does it present unilaterally, or bilaterally?

It is bilateral in the vast majority of cases (>90%)

Q

The STUMPED Mnemonic



- Sclerocornea
- Trauma (endot)
- Ulcer

How does sclerocornea present?
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Another congenital corneal abnormality is strongly associated with sclerocornea. What is it?

- Metabolic disorders
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22

- Sclerocornea
- Trauma (endot
- Ulcer
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Cornea plana



- Sclerocornea
- Trauma (endot
- Ulcer

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Does it present unilaterally, or bilaterally?
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Another congenital corneal abnormality is strongly associated with sclerocornea. What is it?

- Metabolic disorders
 - What is the defining feature of cornea plana?
- Peters
- Endot
- Dermo



- Sclerocornea
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Cornea plana

Metabolic disorders

What is the defining feature of cornea plana? The cornea is markedly flatter than normal

- Peters
- Endot
- Dermo





Cornea plana



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- Metabolic disorders
- What is the defining feature of cornea plana?
 The cornea is markedly flatter than normal

What is the average central corneal power in (non-plana) adults?

- Endot
- Dermo



- Sclerocornea
- Trauma (endot
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- How flat does the central cornea have to be to qualify as cornea plana?



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- Trauma (endot
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 There's no set cutoff, but suffice to say plana corneas are **much** flatter than 43D



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- Trauma (endot
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 There's no set cutoff, but suffice to say plana corneas are **much** flatter than 43D

OK, what would be a typical power of a plana cornea?

A

The STUMPED Mnemonic



- Sclerocornea
- Trauma (endot
- Ulcer

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 There's no set cutoff, but suffice to say plana corneas are **much** flatter than 43D

OK, what would be a typical power of a plana cornea? Values in the 30-35D range are common



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Does this sort of birth trauma tend to be unilateral, or bilateral?

- Demoio of the comea



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Does this sort of birth trauma tend to be unilateral, or bilateral?

 It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)





Corneal haze 2ndry to birth trauma



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Does this sort of birth trauma tend to be unilateral, or bilateral?

 It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)
 - At what point post-partum does the traumatized cornea become cloudy?

A



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 It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)
 - At what point post-partum does the traumatized cornea become cloudy?
- Usually within a day or two

Q



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- How does endothelial damage lead to a cloudy cornea?
- Dennoid of the confea



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 Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy
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Do these traumatic Descemet breaks tend to run vertically, or horizontally?



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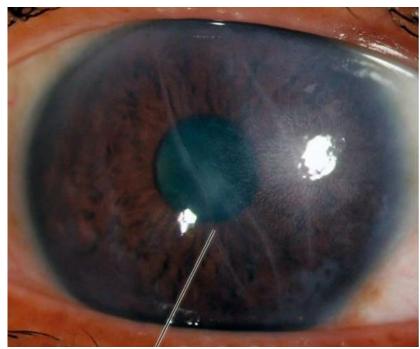
Do these traumatic Descemet breaks tend to run vertically, or horizontally?

Vertically









Vertical Descemet's breaks after birth trauma

Q



- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Does this sort of birth trauma tend to be unilateral, or bilateral?

 It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)
 - Another congenital condition is associated with Descemet's breaks—what is it?

zed cornea become cloudy?

- How does endothelial damage lead to a cloudy cornea?
- Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy

Do these traumatic Descemet breaks tend to run vertically, or horizontally?

Vertically



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 Congenital glaucoma

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- How does endothelial damage lead to a cloudy cornea?
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- Do these traumatic Descemet breaks tend to run vertically, or horizontally?

 Vertically



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zed cornea become cloudy?

Do these traumatic Descemet breaks tend to run vertically, or horizontally?

Vertically



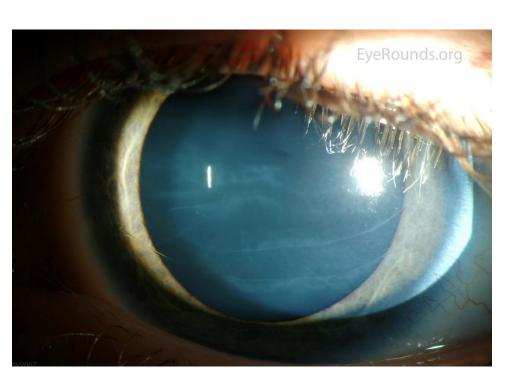
- Sclerocornea
- Do these traumatic Descemet breaks tend to run vertically, or horizontally?
- Horizontally teral, or bilateral?
 - It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)
 - Another congenital condition is associated with Descemet's breaks—what is it?
 - Congenital glaucoma
 - How does endothelial damage lead to a cloudy cornea?
 - Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy

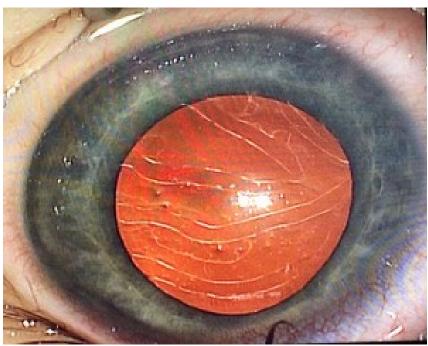
zed cornea become cloudy?

Do these traumatic Descemet breaks tend to run vertically, or horizontally?

Vertically







Horizontal Descemet's breaks in congenital glaucoma

Q

The STUMPED Mnemonic



What is the eponymous name for the Descemet's breaks associated with congenital glaucoma?

- Scierocornea
- Do these traumatic Descemet breaks tend to from forceps run vertically, or horizontally?
- Horizontally

teral, or bilateral?

It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)

Another congenital condition is associated with Descemet's breaks—what is it?

zed cornea become cloudy?

Congenital glaucoma

How does endothelial damage lead to a cloudy cornea?

Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy

Do these traumatic Descemet breaks tend to run vertically, or horizontally?
Vertically

48

What is the eponymous name for the Descemet's breaks associated with congenital glaucoma? Haab's striae



- Scierocornea
- Do these traumatic Descemet breaks tend to from forceps run vertically, or horizontally?
- Horizontally

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It is almost always unilateral (would take seriously bad luck to injure both corneas simultaneously)

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zed cornea become cloudy?

Congenital glaucoma

How does endothelial damage lead to a cloudy cornea?

Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy

Do these traumatic Descemet breaks tend to run vertically, or horizontally?
Vertically

Q

The STUMPED Mnemonic

With what syndrome is corneal dermoid associated?



Dermoid of the cornea



With what syndrome is corneal dermoid associated? Goldenhar syndrome



Mnemonic: Goldenhar

Goldenhar

E N

Н

Α

R

Dermoid of the cornea

What is the incidence of Goldenhar? About 1/4000 live births



Dermoid of the cornea

Goldenhar

O

D E

N

Н

A

Goldenhar syndrome

What is the incidence of Goldenhar?
About 1/4000 live births

What is its inheritance pattern?



Dermoid of the cornea

Goldenhar

C

D

E N

Н

A

What is the incidence of Goldenhar?
About 1/4000 live births

What is its inheritance pattern? It is sporadic



Dermoid of the cornea

Goldenhar

O

D

E N

Н

A

What is the incidence of Goldenhar? About 1/4000 live births

What is its inheritance pattern? It is sporadic

Is there a sex predilection?



Goldenhar

O

D

E N

Н

A

R

Dermoid of the cornea

What is the incidence of Goldenhar? About 1/4000 live births

What is its inheritance pattern? It is sporadic

Is there a sex predilection?

Dermoid of the cornea

Yes, MyF are #x as likely to be affected



Goldenhar

D

N

E

Н

Α

What is the incidence of Goldenhar?
About 1/4000 live births

What is its inheritance pattern? It is sporadic

Is there a sex predilection?
Yes, males are twice as likely to be affected



Goldenhar

C

D

E N

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R

Dermoid of the cornea



With what syndrome is corneal dermoid associated? **Goldenhar syndrome**

What is Goldenhar's noneponymous name?



Answer starts with an 'O'

Dermoid of the cornea

Goldenhar

E N Н

Α



With what syndrome is corneal dermoid associated?

Goldenhar syndrome

What is Goldenhar's noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

Told ya

Dermoid of the cornea



Goldenhar
OAV syndrome

D E

> N H

A R

Q

The STUMPED Mnemonic

With what syndrome is corneal dermoid associated?

Goldenhar syndrome

What is Goldenhar's noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

-

--

One answer starts with an 'L'

The other with a 'D'

Dermoid of the cornea



Goldenhar OAV syndrome

D

E N H

A R



With what syndrome is corneal dermoid associated? Goldenhar syndrome

What is Goldenhar's noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

- --Lid coloboma
- -- Duane syndrome

One answer starts with an 'L'

The other with a 'D'

Dermoid of the cornea



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

E

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A





Goldenhar syndrome: Lid coloboma

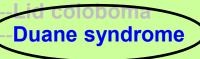


With what syndrome is corneal dermoid associated?

Goldenhar syndrome

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Briefly, what is Duane syndrome?

Dermoid of the cornea



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

E

N

Н

A



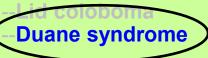
With what syndrome is corneal dermoid associated?

Goldenhar syndrome

What is Goldenhar's noneponymous name?

Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?



Briefly, what is Duane syndrome?

A motility disorder with the following key findings:

- --At least some limitation of horizontal movement
- --Attempted adduction causes the globe to retract, and may cause it to up- or downshoot

Dermoid of the cornea



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

E

N

H

A



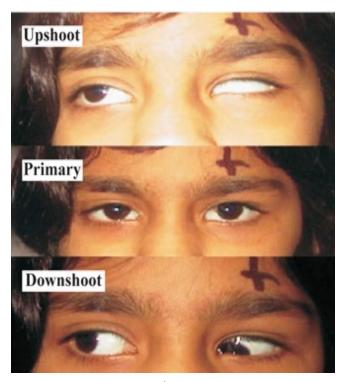




Horizontal movement limitation



Globe retraction



Upshoot/downshoot

Duane syndrome

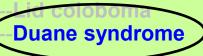


With what syndrome is corneal dermoid associated?

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Briefly, what is Duane syndrome?

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- --At least some limitation of horizontal movement
- --Attempted adduction causes the globe to retract, and may cause it to up- or downshoot

What is the cause?

Dermoid of the cornea

66

Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

E

N

Н

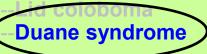
A



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Briefly, what is Duane syndrome?

A motility disorder with the following key findings:

- --At least some limitation of horizontal movement
- --Attempted adduction causes the globe to retract, and may cause it to up- or downshoot

What is the cause?

The nucleus for cranial nerve VI is missing, and the lateral rectus is innervated by cranial nerve III

Dermoid of the cornea



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

E

N

Н

A



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What nonocular findings are usually present?

-

--

One answer starts with an 'E'

Dermoid of the

The other with a 'H'



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

Ε

Nothing starts w/ 'N'

Н

Α



With what syndrome is corneal dermoid associated? Goldenhar syndrome

What is Goldenhar's noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

- --Lid coloboma
- -- Duane syndrome

What **nonocular** findings are usually present?

- --Ear abnormalities (pre-auricular appendages; aural fistulae)
- --Hemifacial microsomia (maxillary/mandibular hypoplasia)

One answer starts with an 'E'

Dermoid of the

The other with a 'H'



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

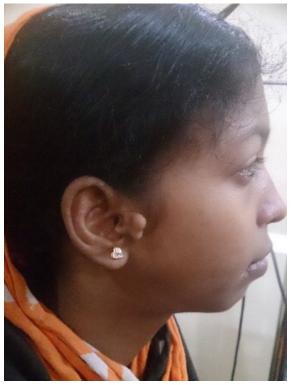
Ear abnormalities

Nothing starts w/ 'N'

Hemifacial microsomia











70

Hemifacial microsomia

Goldenhar syndrome

Q

The STUMPED Mnemonic

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Goldenhar syndrome

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Where are epibulbar dermoids commonly located in Goldenhar?

Dermoid of the cornea

Etc.



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

Ear abnormalities

Nothing starts w/ 'N'

Hemifacial microsomia

A



With what syndrome is corneal dermoid associated?

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Where are epibulbar dermoids commonly located in Goldenhar?

At the limbus

Dermoid of the cornea

Etc.



Goldenhar

OAV syndrome

Lid coloboma

Duane syndrome

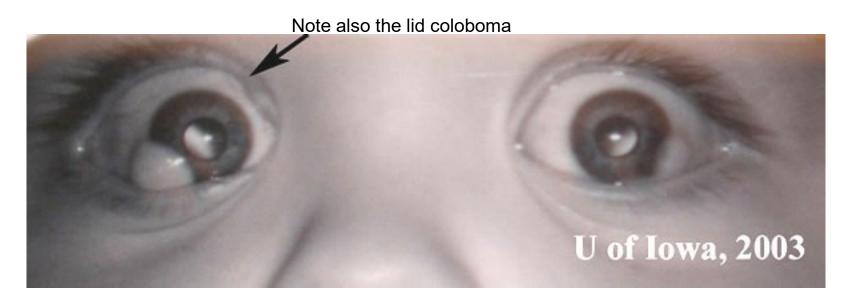
Ear abnormalities

Nothing starts w/ 'N'

Hemifacial microsomia

At the limbus





Goldenhar syndrome: Limbal (epibulbar) dermoids OU



With what syndrome is corneal dermoid associated? Goldenhar syndrome

What is Goldenhar's noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

- --Lid coloboma
- -- Duane syndrome

What nonocular findings are usually present?

- --Ear abnormalities (pre-auricular appendages; aural fistulae)
- --Hemifacial microsomia (maxillary/mandibular hypoplasia)

Where are epibulbar dermoids commonly located in Goldenhar?

At the limbus

Are Goldenhar pts cognitively impaired?

Dermoid of the cornea



Goldenhar
OAV syndrome

Lid coloboma

Duane syndrome

Ear abnormalities

Nothing starts w/ 'N'

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Are Goldenhar pts cognitively impaired?

A minority (~10%) have mental retardation

Dermoid of the cornea



OAV syndrome
Lid coloboma
Duane syndrome
Ear abnormalities
Nothing starts w/ 'N'
Hemifacial microsomia
At the limbus

Retardation in ~10%

Goldenhar



The Peds book mentions one specific sort of metabolic disorder—what is it?

- Scle
- Trau
- Ulce
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



The Peds book mentions one specific sort of metabolic disorder—what is it? Mucopolysaccharidosis (MPS)

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The CTHMDED Masmania

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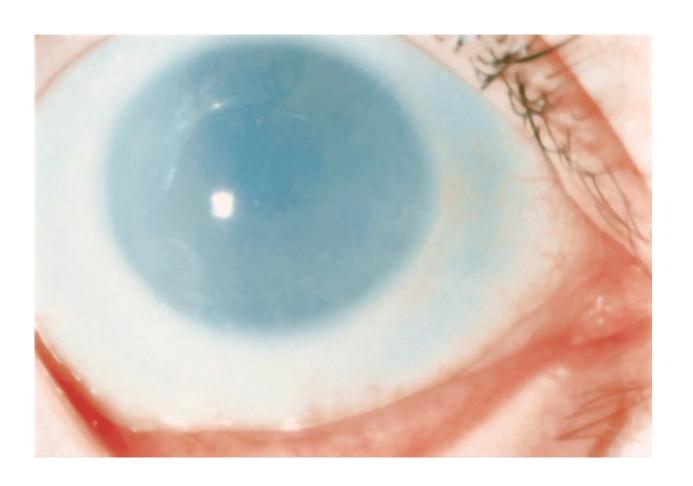
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Hurler, Scheie, and Morquio syndromes

Ulce

- Metabolic disorders
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MPS (Hurler syndrome)

The CTIIMDED Masmania



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The STIIMDED Masmania



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In a nutshell, what is a mucopolysaccharidosis?

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- Do these syndromes present with cloudy corneas at birth?
- No, significant cloudiness takes 6 weeks to 24 months or so to develop, depending upon the syndrome
- Hurler, Scheie, and Morquio syndromes
- Ulce
- Metabolic disorders
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The CTIIMDED Masses



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How is the cloudy MPS cornea managed?

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- Hurler, Scheie, and Morquio syndromes
- How is the cloudy MPS cornea managed?
 Via transplantation—either PK, or DALK
- Metabolic disorders
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The Peds book mentions three What do these stand for?

Trau Hurler, Scheie, and Morqu

PK: DALK:

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Trau Hurler, Scheie, and Morqu

What do these stand for? PK: Penetrating keratoplasty

DALK: Deep anterior lamellar keratoplasty

How is the cloudy MPS comed managed Via transplantation—either PK, or DALK

- Metabolic disorders
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In three words, what sort of condition is Peters anomaly? It is a classic exemplar of an

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- Endothelial dystrophy (CHED)
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In three words, what sort of condition is Peters anomaly? It is a classic exemplar of an anterior segment dysgenesis

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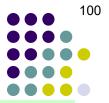


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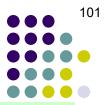
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In one word, what sort of condition is it?

It is a which means it arises secondary to the failure of two-words cells to migrate and/or differentiate properly

- Peters anomaly
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In three words, what sort of condition is Peters anomaly?
It is a classic exemplar of an anterior segment dysgenesis

In one word, what sort of condition is it?
It is a neurocristopathy, which means it arises secondary to the failure of neural-crest cells to migrate and/or differentiate properly

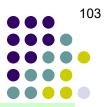
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How does it present?

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In three words, what sort of condition is Peters anomaly? It is a classic exemplar of an anterior segment dysgenesis

How does it present?

As a corneal opacity at birth (duh, it's in the STUMPED mnemonic). The opacity ranges in severity from a faint haze to an opaque, elevated and vascularized mess.

- Peters anomaly
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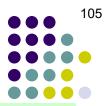
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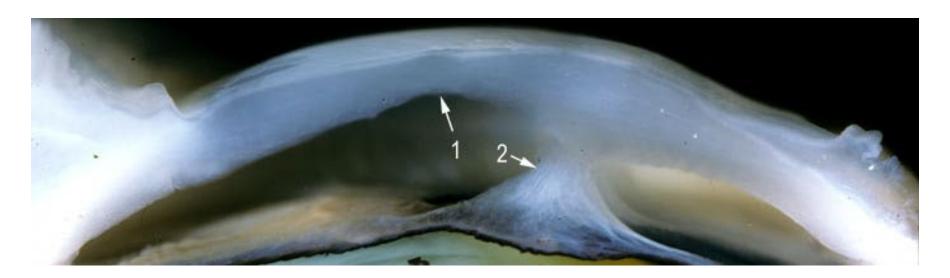
There is a defect in the posterior central cornea, including the absence of Descemet's and subjacent endothelium. Adhesions extending from the iris to the posterior corneal defect are often present.

- Peters anomaly
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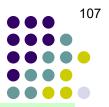


1. Defect of the posterior central cornea, including the absence of Descemet's and subjacent endothelium

2. Adhesions extending from the iris to the posterior corneal defect







In three words, what sort of condition is Peters anomaly? It is a classic exemplar of an anterior segment dysgenesis

How does it present?

As a corneal opacity at birth (duh, it's in the STUMPED mnemonic). The opacity ranges in severity from a faint haze to an opaque, elevated and vascularized mess.

What specific abnormalities are commonly present?

There is a defect in the posterior central cornea, including the absence of Descemet's and subjacent endothelium. Adhesions extending from the iris to the posterior corneal defect are often present. The lens may be small, cataractous and misshapen, and may be adherent to the defect in the posterior cornea.

- Peters anomaly
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Peters anomaly: Small, cataractous, misshapen lens



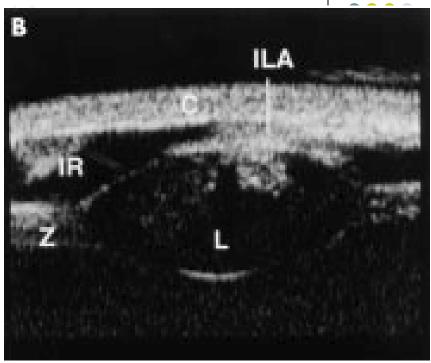
This image shows complete corneal opacification thought clinically to be sclerocornea







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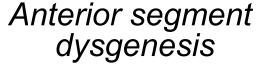


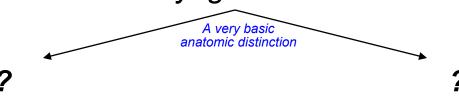
UBM of the same eye shows keratolenticular adhesion (ILA), aniridia with an iris stump (IR), and a small lens, revealing the correct diagnosis as Peters anomaly. This case demonstrates the importance of a complete anterior segment exam with UBM to make an accurate diagnosis.

Q

The STUMPED Mnemonic



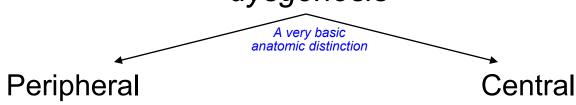


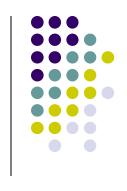


Let's take a moment to overview the anterior segment dysgeneses. We divvy them up into two broad categories based on a very basic anatomic consideration—what is that consideration?



Anterior segment dysgenesis

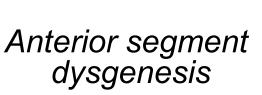


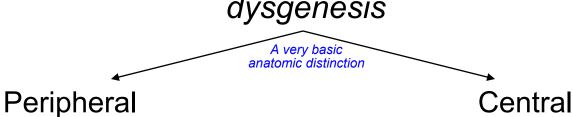


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Q

The STUMPED Mnemonic



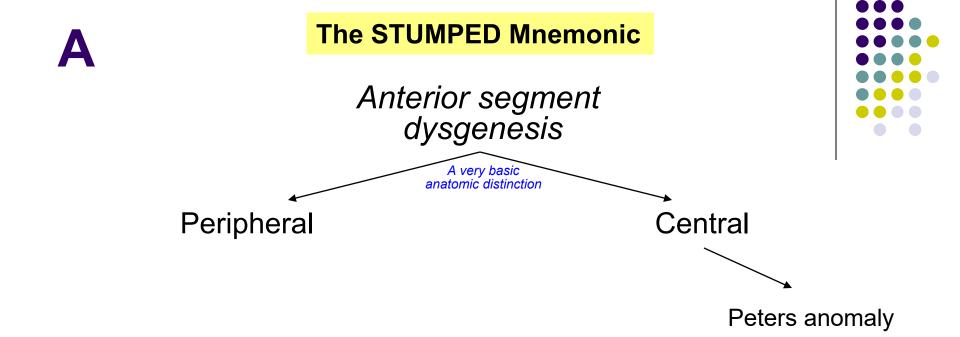




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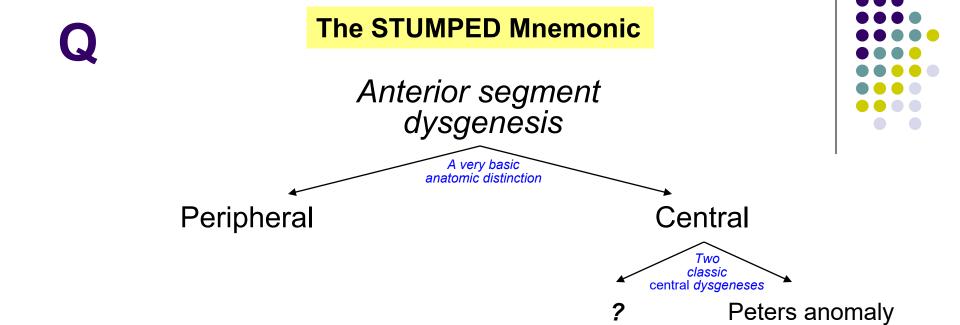
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To which category does Peters anomaly belong?



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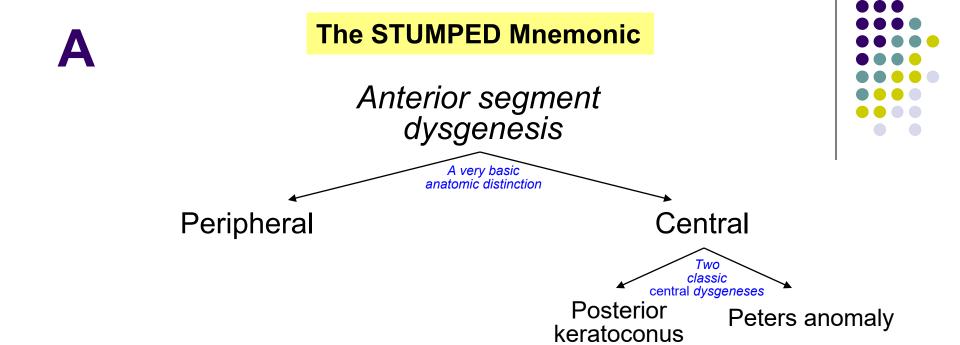
Peters is a **central** form of anterior segment dysgenesis



To which category does Peters anomaly belong?

Peters is a **central** form of anterior segment dysgenesis

What is the other classic form of central anterior segment dysgenesis?

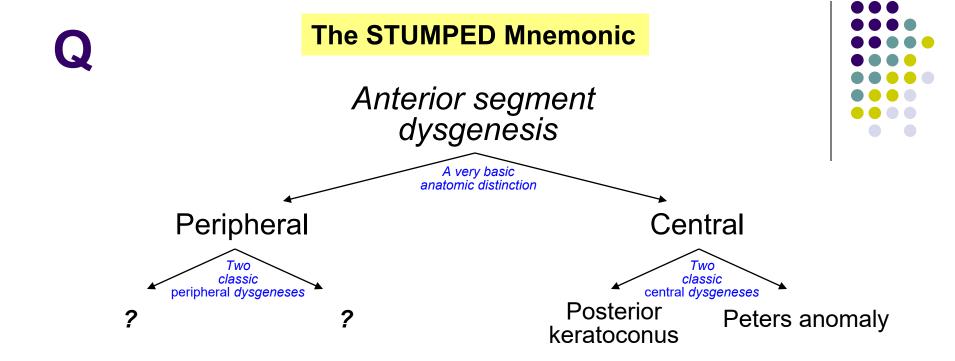


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Posterior keratoconus (Note: This is nothing like the anterior form you're familiar with)



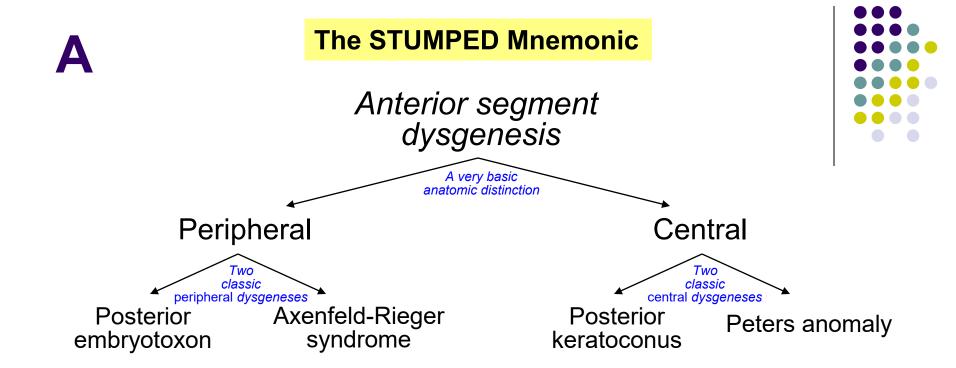
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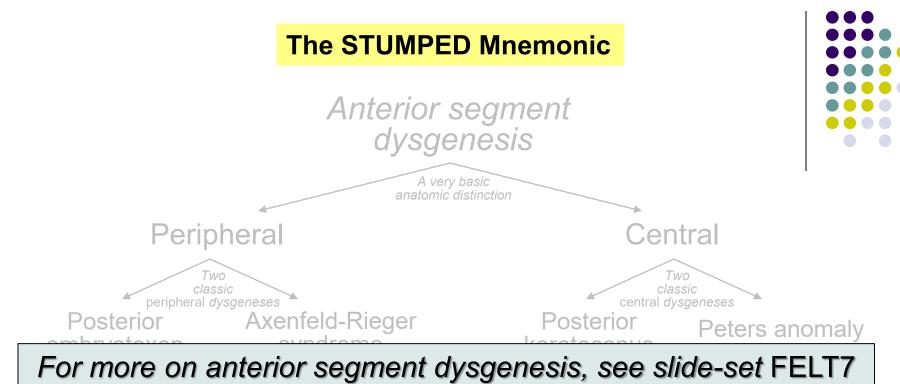
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Posterior embryotoxon and Axenfeld-Rieger syndrome



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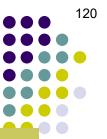
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The Cornea book discusses three endothelial dystrophies. What are the other two? --CHED
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- Endothelial dystrophy (CHED)
- Dermoid of the cornea



The Cornea book discusses three endothelial dystrophies. What are the other two? --CHED

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- --Posterior polymorphous corneal dystrophy

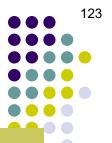
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--CHED
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--Posterior polymorphous corneal dystrophy

In CHED, what is seen at the slit lamp?

The corneas are diffusely cloudy (described as

two words

or one word

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In CHED, what is seen at the slit lamp?

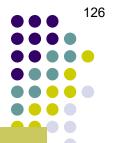
The corneas are diffusely cloudy (described as 'ground glass' or 'milky')

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125

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 - N e x t

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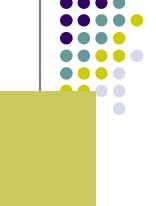
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In CHED, what is seen at the slit lamp?

The corneas are diffusely cloudy (described as 'ground glass' or 'milky'), and thickened

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127

- The Cornea book discusses three endothelial dystrophies. What are the other two? --CHED --Fuchs endothelial corneal dystrophy --Posterior polymorphous corneal dystrophy In CHED, what is seen at the slit lamp? How thickened? thickened The corneas are diffusely cloudy (desc
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128

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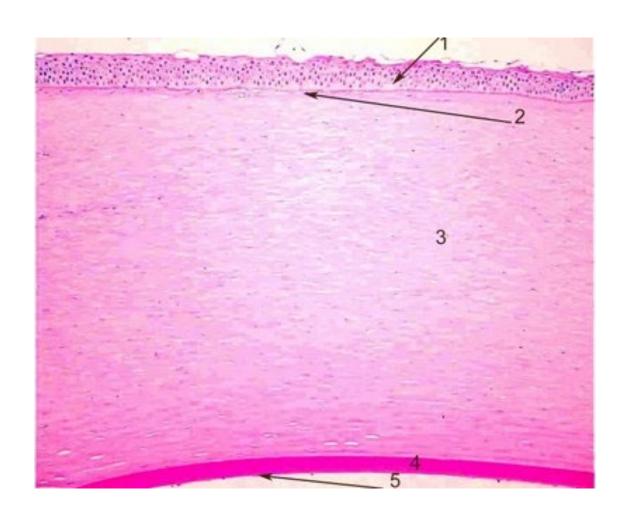
In CHED, what is seen at the slit lamp? How thickened?

The corneas are diffusely cloudy (desc Like, 2-3 times normal

thickened

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Congenital hereditary endothelial dystrophy. Get a load of how thicc this cornea is!

130

thickened

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Why is the cornea thickened in CHED?

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Why is the cornea thickened in CHED? We'll get to that shortly

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- Is vision affected?

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- Dermoid of the cornea



The Cornea book discusses three endothelial dystrophies. What are the other two? --CHED

- --Fuchs endothelial corneal dystrophy
- --Posterior polymorphous corneal dystrophy

In CHED, what is seen at the slit lamp?

The corneas are diffusely cloudy (described as 'ground glass' or 'milky'), and thickened

Is vision affected?

Yes, significantly enough that a sensory may be present

Endothelial dystrophy (CHED)

Dermoid of the cornea



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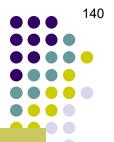
 Descemet's is thinned vs thickened
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



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- What is the histologic hallmark of CHED on light microscopy?
 Descemet's is thickened, and the number of endothelial cells is substantially than normal
- Endothelial dystrophy (CHED)
- Dermoid of the cornea





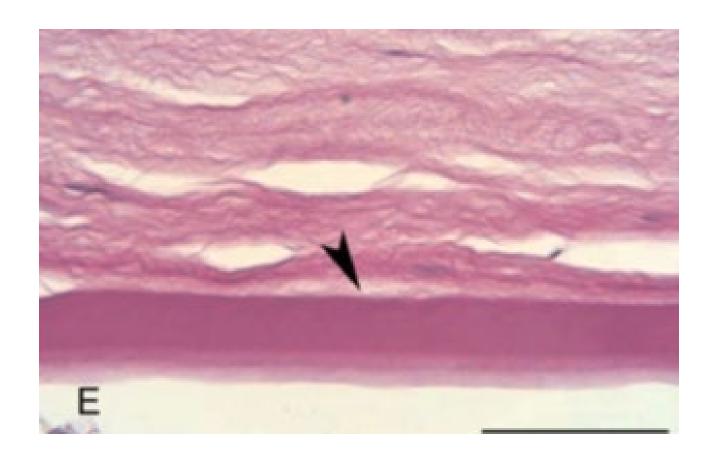
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Congenital hereditary endothelial dystrophy. Thickened Descemet membrane with no visible endothelial cells.



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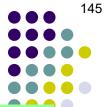
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The few endothelial cells that are present—are they normal? No, most if not all are atrophic

- What is the histologic hallmark of CHED on light microscopy?

 Descemet's is thickened, and the number of endothelial cells is substantially less than normal
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We have now 'gotten to' the reason CHED corneas are thickened: The Cornea book di The paucity of functioning endothelial cells --CHED --Fuchs endothelial --Posterior polymork In CHED, what is se The corneas are diff Is vision affected? Yes, significantly enough that a sensor We'll get to that shortly Is it painful? No What is the histologic hallmark of CHED on light micros ppy? Descemet's is thickened, and the number of endothelial cells is substantially less than normal

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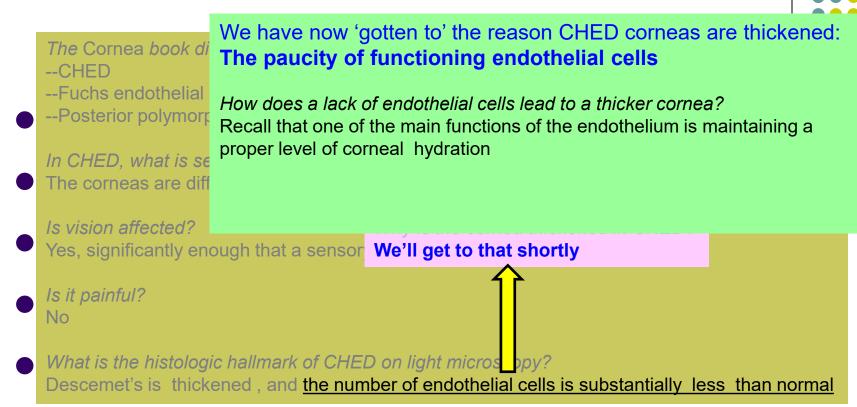
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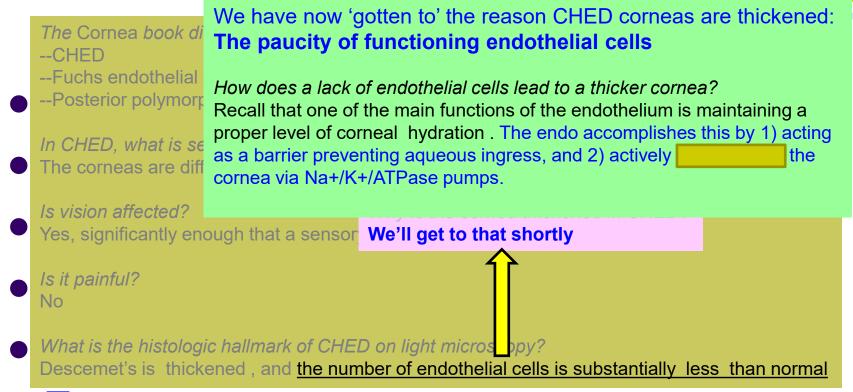




- Endothelial dystrophy (CHED)
- Dermoid of the cornea

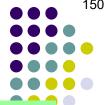
QA

The STUMPED Mnemonic



- Endothelial dystrophy (CHED)
- Dermoid of the cornea





The Cornea book di --CHED

We have now 'gotten to' the reason CHED corneas are thickened: The paucity of functioning endothelial cells

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How does a lack of endothelial cells lead to a thicker cornea? Recall that one of the main functions of the endothelium is maintaining a proper level of corneal hydration. The endo accomplishes this by 1) acting as a barrier preventing aqueous ingress, and 2) actively dehydrating the cornea via Na+/K+/ATPase pumps.

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Is vision affected?

Yes, significantly enough that a sensor We'll get to that shortly

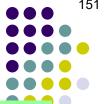
Is it painful? No

What is the histologic hallmark of CHED on light micros ppy?

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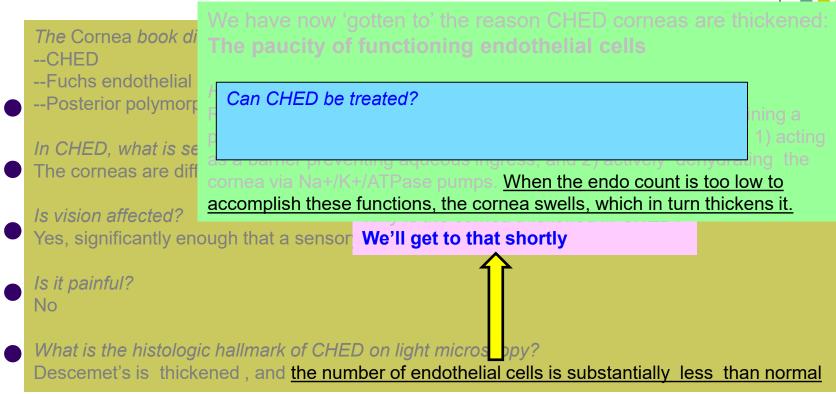
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How does a lack of endothelial cells lead to a thicker cornea? Recall that one of the main functions of the endothelium is maintaining a proper level of corneal hydration. The endo accomplishes this by 1) acting as a barrier preventing aqueous ingress, and 2) actively dehydrating the cornea via Na+/K+/ATPase pumps. When the endo count is too low to accomplish these functions, the cornea swells, which in turn thickens it.

Yes, significantly enough that a sensor We'll get to that shortly

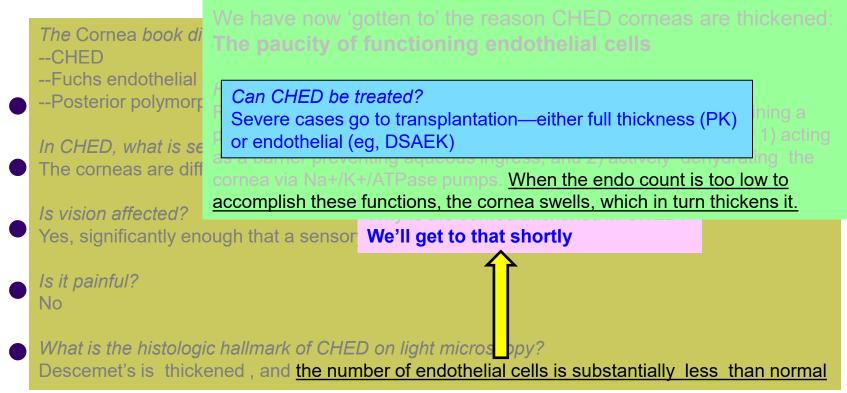
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- Endothelial dystrophy (CHED)
- Dermoid of the cornea





- Endothelial dystrophy (CHED)
- Dermoid of the cornea



Sclerocornea;

- not the first letter in this disorder
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders

The mnemonic should be modified to include these as well

- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea



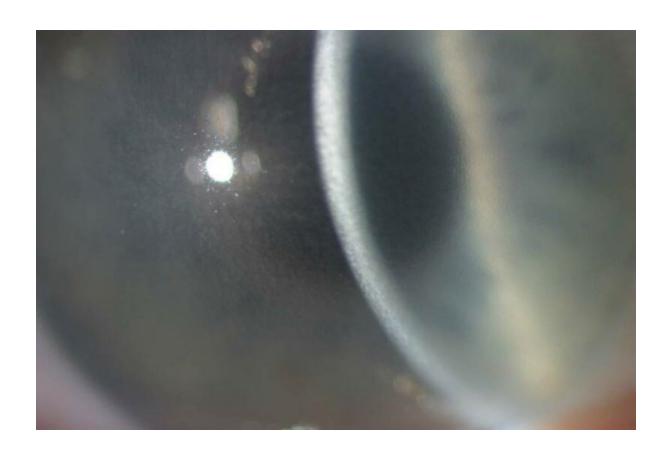


- Sclerocornea; congenital hereditary Stromal dystrophy (CHSD)
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders

The mnemonic should be modified to include these as well

- Peters anomaly
- Endothelial dystrophy (CHED)
- Dermoid of the cornea







- Sclerocornea; congenital hereditary Stromal dystrophy (CHSD)
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED);
- Dermoid of the cornea

The mnemonic should be modified to include these as well





- Sclerocornea; congenital hereditary Stromal dystrophy (CHSD)
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Endothelial dystrophy (CHED); Elevated IOP (congenital glaucoma)
- Dermoid of the cornea

The mnemonic should be modified to include these as well



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



	ССТ	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	?			
CHSD	?			
Primary congenital glaucoma	?			



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



	ССТ	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	warkediy	CHED because	e of edema 2n	s dramatically increased in dry to lack of adequate gescence function
CHSD	Mildly increased			
Primary congenital glaucoma	Variably increased (or WNL, or thin)			



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



	ССТ	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		

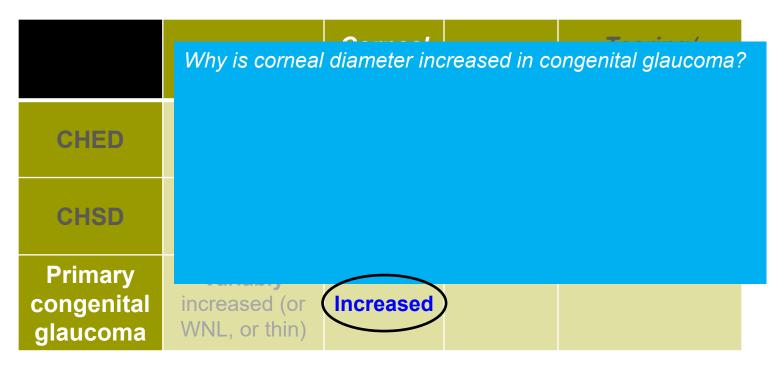


	ССТ	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	Markedly increased	?		
CHSD	Mildly increased	?		
Primary congenital glaucoma	Variably increased (or WNL, or thin)	?		

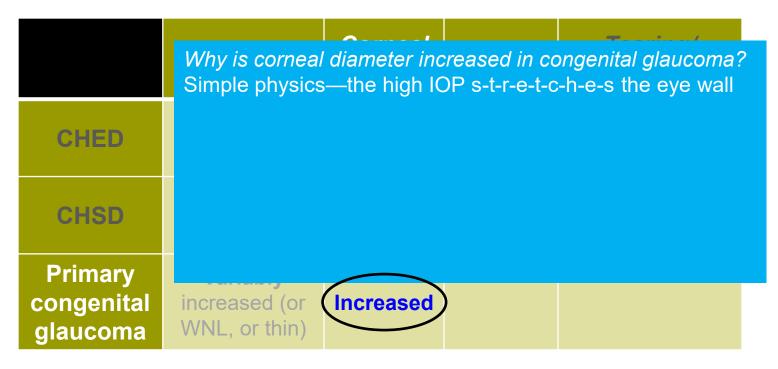


	ССТ	Corneal diameter	IOP	Tearing/ Photophobia?
CHED	Markedly increased	WNL		
CHSD	Mildly increased	WNL		
Primary congenital glaucoma	Variably increased (or WNL, or thin)	Increased		

167



168

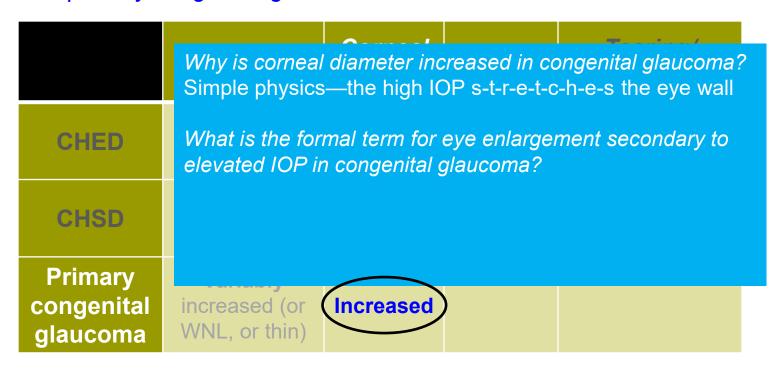




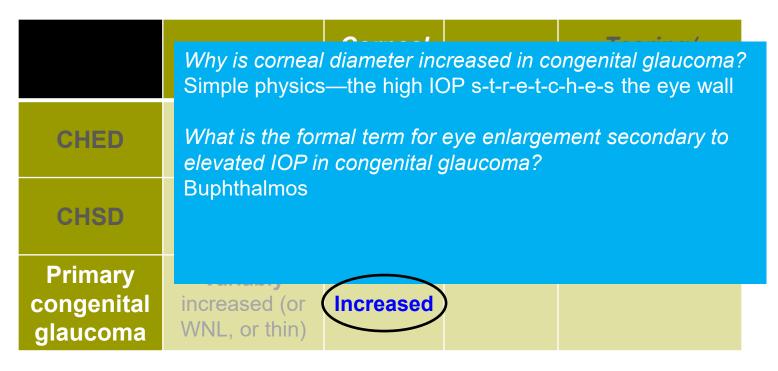


Congenital glaucoma: Increased corneal diameter

170





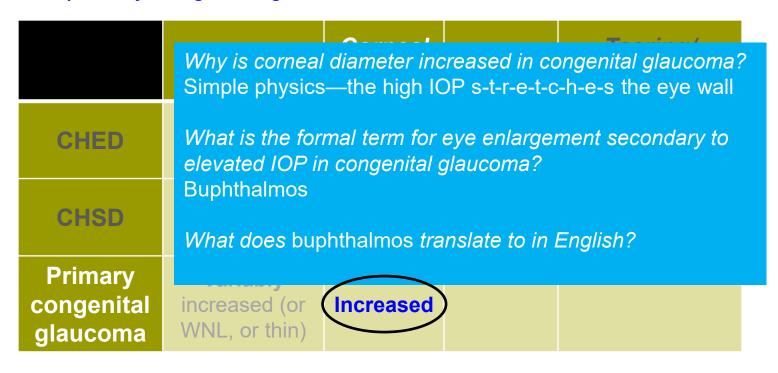




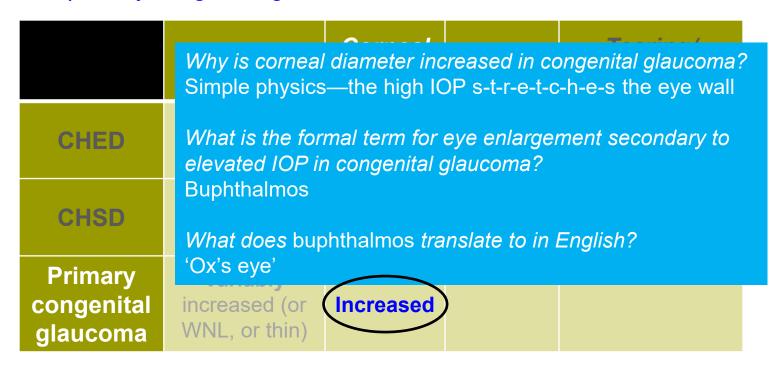


Congenital glaucoma: Buphthalmos OD











& 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? Simple physics—the high IOP s-t-r-e-t-c-h-e-s the eye wall

And as mentioned earlier in the slide-set, the high IOP in congenital glaucoma causes *Haab's striae*—horizontal breaks in Descemet's and the overlying endothelium

CHSD

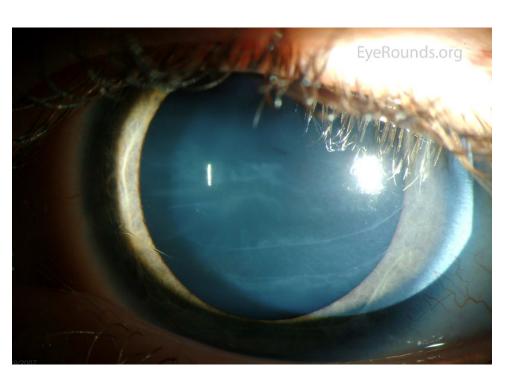
Primary congenital glaucoma

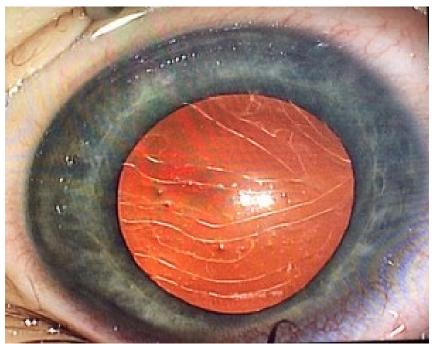
What does buphthalmos translate to in English? 'Ox's eve'

increased (or WNL, or thin)

Increased







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



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



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



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



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