Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**ndothelial dystrophy (CHED) *(Congenital hereditary endothelial dystrophy)*
- **D**ermoid of the cornea
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

How does sclerocornea present?

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

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

Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

How does sclerocornea present?
As bilateral (in % of cases) opacified corneas
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

- **Sclerocornea**
- **Trauma (endothelial; from forceps, etc)**
- **Ulcer**
- **Metabolic disorders**
- **Peters anomaly**
- **Endothelial dystrophy (CHED)**
- **Dermoid of the cornea**
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **S**clerocornea
  - How does sclerocornea present?
    - As bilateral (in 90% of cases) **opacified corneas**
  - In sclerocornea, is it the case that normal corneal tissue has literally been replaced by scleral tissue?

- **P**eters anomaly
- **E**ndothelial dystrophy (CHED)
- **D**ermoid of the cornea
Sclerocornea

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

In sclerocornea, is it the case that normal corneal tissue has literally been replaced by scleral tissue?
This is unclear. The BCSC Cornea book asserts that such corneas have undergone “scleralization.”
However, the Peds book states that the cornea merely ‘resembles’ sclera, and pointedly notes that the term sclerocornea is “descriptive,” and “does not suggest causation.” In fact, the Peds book suggests the term sclerocornea should be avoided entirely!

So what should you do? You should caveat emptor.

Peters anomaly
Endothelial dystrophy (CHED)
Dermoid of the cornea

Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

**Sclerocornea**

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

**How does sclerocornea present?**
As bilateral (in 90% of cases) opacified corneas

**Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?**
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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

**How does sclerocornea present?**
As bilateral (in 90% of cases) opacified corneas

**Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?**
Cornea plana
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

How does sclerocornea present?  
As bilateral (in 90% of cases) opacified corneas

Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?  
Cornea plana

What is the defining feature of cornea plana?

Values in the 30-35D range are common
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?

**Cornea plana**

What is the defining feature of cornea plana?
The cornea is markedly flatter than normal
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?

**Cornea plana**

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?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **Sclerocornea**
  - How does sclerocornea present?
    - As bilateral (in 90% of cases) opacified corneas
  - Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?
    - Cornea plana

- **Trauma (endothelial)**
  - Endothelial dystrophy (CHED)
  - Dermoid of the cornea

- **Ulcer**

- **Ulcus**

- **Metabolic disorders**

- **Peters anomaly**

- **Endothelial dystrophy (CHED)**

**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?**
- About 43D
Sclerocornea

- How does sclerocornea present?
  - As bilateral (in 90% of cases) opacified corneas

- Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?
  - Cornea plana

- 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?
  - About 43D

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

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?

Cornea plana

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?
About 43D

How flat does the central cornea have to be to qualify as cornea plana?
Technically, less than 43D, but don’t get it twisted--most corneas within shouting distance of 43D are not ‘plana.’ Plana corneas are much flatter.

Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **Sclerocornea**
  - How does sclerocornea present?
    - As bilateral (in 90% of cases) opacified corneas
  - Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?
    - Cornea plana

- **Trauma (endothelial; from forceps, etc)**

- **Ulcer**

- **Metabolic disorders**

- **Peters anomaly**

- **Endothelial dystrophy (CHED)**

- **Dermoid of the cornea**

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?
- About 43D

How flat does the central cornea have to be to qualify as cornea plana?
- Technically, less than 43D, but don’t get it twisted--most corneas within shouting distance of 43D are not ‘plana.’ Plana corneas are much flatter.

OK, what is the typical power of a plana cornea?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **Sclerocornea**
  - How does sclerocornea present?
    - As bilateral (in 90% of cases) opacified corneas
  - Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?
    - Cornea plana

- **Trauma (endothelial)**

- **Ulcer**

- **Metabolic disorders**

- **Peters anomaly**

- **Endothelial dystrophy (CHED)**

- **Dermoid of the cornea**

---

How does sclerocornea present?
As bilateral (in 90% of cases) opacified corneas

Other than being opaque, what corneal abnormality is found in most cases of sclerocornea?
Cornea plana

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?
About 43D

How flat does the central cornea have to be to qualify as cornea plana?
Technically, less than 43D, but don’t get it twisted--most corneas within shouting distance of 43D are not ‘plana.’ Plana corneas are **much** flatter.

OK, what is the typical power of a plana cornea?
Values in the 30-35D range are common
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- Sclerocornea
- Trauma (endothelial; ie, from forceps)
- Dermoid of the cornea

Does this sort of birth trauma tend to be unilateral, or bilateral?
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **S**clerocornea
- **T**rauma (endothelial; ie, from forceps)
  - *Does this sort of birth trauma tend to be unilateral, or bilateral?*
  - It is almost always unilateral (would take some seriously bad luck to injure both corneas simultaneously)
- **D**ermoid of the cornea
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **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 some seriously bad luck to injure both corneas simultaneously)

  *At what point post-partum does the traumatized cornea become cloudy?*

- **Dermoid of the cornea**
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- 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 some 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
- Dermoid of the cornea
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **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 some 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
  - *How does endothelial damage lead to a cloudy cornea?*

- **Dermoid of the cornea**
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **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 some 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
  - **How does endothelial damage lead to a cloudy cornea?**
    - Breaks in Descemet's/endothelium allow the cornea to become edematous, and thus hazy
- **Dermoid of the cornea**
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- 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 some 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

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

  What is the eponymous name for these trauma-induced Descemet breaks?
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 some 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

How does endothelial damage lead to a cloudy cornea?

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

What is the eponymous name for these trauma-induced Descemet breaks? Vogt striae
Sclerocornea

Trauma (endothelial; ie, from forceps)

*Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?*

**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 some seriously bad luck to injure both corneas simultaneously)

At what point post-partum does the traumatized cornea become cloudy?

Another congenital condition is associated with Descemet’s breaks--what is it?

How does endothelial damage lead to a cloudy cornea?

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

What is the eponymous name for these trauma-induced Descemet breaks?

Vogt striae
Sclerocornea

Trauma (endothelial; ie, from forceps)

Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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

At what point post-partum does the traumatized cornea become cloudy?

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

What is the eponymous name for these trauma-induced Descemet breaks?

Vogt striae
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- 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 some seriously bad luck to injure both corneas simultaneously)

**What is the eponymous name for these glaucoma-induced Descemet breaks?**

**At what point post-partum does the traumatized cornea become cloudy?**

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

**What is the eponymous name for these trauma-induced Descemet breaks?**
Vogt striae
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

- **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 some seriously bad luck to injure both corneas simultaneously).

  *What is the eponymous name for these glaucoma-induced Descemet breaks?*
  
  Haab striae

  *At what point post-partum does the traumatized cornea become cloudy?*

  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.

  *What is the eponymous name for these trauma-induced Descemet breaks?*

  Vogt striae
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 some 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

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

What is the eponymous name for these trauma-induced Descemet breaks?
Vogt striae

Haab striae

Let’s briefly review Vogt and Haab striae…

What is the eponymous name for these glaucoma-induced Descemet breaks?
Haab striae

Another congenital condition is associated with Descemet’s breaks--what is it?
Congenital glaucoma
For each statement, identify whether it applies to **Haab striae vs Vogt striae**

- **Associated with congenital glaucoma:**

  - Vogt
  - Haab

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: **Haab**
- Associated with birth trauma: **Vogt**

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to Haab striae vs Vogt striae

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt
- Usually oriented vertically:

Vogt agina
Haab igh IOP

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt
- Usually oriented vertically: Vogt

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt
- Usually oriented vertically: Vogt
- Usually oriented horizontally:

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: **Haab**
- Associated with birth trauma: **Vogt**
- Usually oriented vertically: **Vogt**
- Usually oriented horizontally: **Haab**

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is? First of several ‘hint questions’ forthcoming

As the questions above are answered, check down to here to see the mnemonics bloom
Q/A

For each statement, identify whether it applies to *Haab striae* vs *Vogt striae*

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal type of dz…

First of several ‘hint questions’ forthcoming

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is? It is a corneal dystrophy …

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to Haab striae vs Vogt striae

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal layer …

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- **Associated with congenital glaucoma:** Haab
- **Associated with birth trauma:** Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is? It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …

As the questions above are answered, check down to here to see the mnemonics bloom
Associated with congenital glaucoma: Haab
Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …
Its histologic hallmark is the finding of three words on the endothelial surface…

As the questions above are answered, check down to here to see the mnemonics bloom.
For each statement, identify whether it applies to Haab striae vs Vogt striae

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?
It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …
Its histologic hallmark is the finding of multiple cell layers on the endothelial surface …

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to Haab striae vs Vogt striae

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition—completely unrelated—that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …
Its histologic hallmark is the finding of multiple cell layers on the endothelial surface …
The bands are sometimes referred to as ‘two words’ or ‘two words, same second word’.

Vogt agina vertical

Haab high IOP horizontal

As the questions above are answered, check down to here to see the mnemonics bloom.
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …
Its histologic hallmark is the finding of multiple cell layers on the endothelial surface …
The bands are sometimes referred to as ‘snail tracks’ or ‘railroad tracks’

As the questions above are answered, check down to here to see the mnemonics bloom.

Last chance--answer next!
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition—completely unrelated—that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy …
Specifically, it is a dystrophy of the corneal endothelium …
Its histologic hallmark is the finding of multiple cell layers on the endothelial surface …
The bands are sometimes referred to as ‘snail tracks’ or ‘railroad tracks’
Posterior polymorphous membrane dystrophy (PPMD)

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands:

**Posterior polymorphous membrane dystrophy (PPMD)**

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?

-- Haab striae are present at birth, whereas PPMD changes don't appear until teens/early adulthood
-- Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

Finally, the configuration of the lines differ in a subtle but crucial way:
-- Haab striae taper and meet at their ends, whereas
-- The parallel lines in PPMD do not meet at their ends

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy…

Specifically, it is a dystrophy of the corneal endothelium…

Its histologic hallmark is the finding of multiple cell layers on the endothelial surface…

The bands are sometimes referred to as “snail tracks” or “railroad tracks.”

**Posterior polymorphous membrane dystrophy (PPMD)**

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?

--- Haab striae are present at birth, whereas PPMD changes don’t appear until teens/early adulthood

--- Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

Finally, the configuration of the lines differ in a subtle but crucial way:

--- Haab striae taper and meet at their ends, whereas

--- The parallel lines in PPMD do not meet at their ends

As the questions above are answered, check down to here to see the mnemonics bloom.
Associated with congenital glaucoma: Haab
Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy…

Specifically, it is a dystrophy of the corneal endothelium…

Its histologic hallmark is the finding of multiple cell layers on the endothelial surface…

The bands are sometimes referred to as ‘snail tracks’ or ‘railroad tracks’…

Posterior polymorphous membrane dystrophy (PPMD)

Haab striae are present at birth, whereas PPMD changes don’t appear until teens/early adulthood.

Finally, the configuration of the lines differ in a subtle but crucial way:

-- Haab striae taper and meet at their ends, whereas
-- The parallel lines in PPMD do not meet at their ends.
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands:

**Posterior polymorphous membrane dystrophy (PPMD)**

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?
--Haab striae are present at birth, whereas PPMD changes don’t appear until teens/early adulthood
--Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

Finally, the configuration of the lines differ in a subtle but crucial way:
--Haab striae taper and meet at their ends, whereas
--The parallel lines in PPMD do not meet at their ends

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to **Haab striae vs Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that can be confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy... Specifically, it is a dystrophy of the corneal endothelium... Its histologic hallmark is the finding of multiple cell layers on the endothelial surface... The bands are sometimes referred to as 'snail tracks' or 'railroad tracks'.

Posterior polymorphous membrane dystrophy (PPMD)

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?

-- Haab striae are present at birth, whereas PPMD changes don't appear until teens/early adulthood
-- Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

As the questions above are answered, check down to here to see the mnemonics bloom
For each statement, identify whether it applies to
Haab striae vs Vogt striae

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands. Do you know what that condition is?

It is a corneal dystrophy…
Specifically, it is a dystrophy of the corneal endothelium…
The bands are sometimes referred to as "snail tracks" or "railroad tracks." It is called Posterior polymorphous membrane dystrophy (PPMD).

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?
--Haab striae are present at birth, whereas PPMD changes don't appear until teens/early adulthood
--Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

(But note: PPMD can present very asymmetrically)

As the questions above are answered, check down to here to see the mnemonics bloom.
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- **Associated with congenital glaucoma**: Haab
- **Associated with birth trauma**: Vogt

There is another condition—completely unrelated—that presents with narrow, horizontally oriented bands.

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?

---

- Haab striae are present at birth, whereas PPMD changes don’t appear until teens/early adulthood.
- Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral.

Finally, the configuration of the lines differ in a subtle but crucial way:

- The bands are sometimes referred to as “snail tracks” or “railroad tracks.”

**Posterior polymorphous membrane dystrophy (PPMD)**

As the questions above are answered, check down to here to see the mnemonics bloom.
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands that are easily confused with Haab striae. Do you know what that condition is?

It is a corneal dystrophy...

Specifically, it is a dystrophy of the corneal endothelium...

Its histologic hallmark is the finding of multiple cell layers on the endothelial surface...

The bands are sometimes referred to as ‘snail tracks’ or ‘railroad tracks’...

**Posterior polymorphous membrane dystrophy (PPMD)**

---

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?

--Haab striae are present at birth, whereas PPMD changes don’t appear until teens/early adulthood
--Haab striae are essentially always unilateral, whereas PPMD changes are always bilateral

Finally, the configuration of the lines differ in a subtle but crucial way:

--Haab striae taper and meet at their ends, whereas
--The parallel lines in PPMD do not meet at their ends

As the questions above are answered, check down to here to see the mnemonics bloom.
For each statement, identify whether it applies to **Haab striae** vs **Vogt striae**

- Associated with congenital glaucoma: Haab
- Associated with birth trauma: Vogt

There is another condition--completely unrelated--that presents with narrow, horizontal bands. Do you know what that condition is? It is a corneal dystrophy. Specifically, it is a dystrophy of the corneal endothelium. Its histologic hallmark is the finding of multiple cell layers on the endothelial surface. The bands are sometimes referred to as “snail tracks” or “railroad tracks.”

**Posterior polymorphous membrane dystrophy (PPMD)**

How can the snail/railroad tracks of PPMD be distinguished from Haab striae?
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As the questions above are answered, check down to here to see the mnemonics bloom.
With what syndrome is corneal dermoid associated?

- Dermoid of the cornea
With what syndrome is corneal dermoid associated?

**Goldenhar syndrome**

### Mnemonic: Goldenhar

- **G**oldenhar
- **O**ptics
- **L**id colobomas
- **D**uane syndrome
- **E**ar abnormalities
- **N**othing starts w/ 'N'
- **H**emifacial microsomia
- **A**t the limbus
- **R**tardation in 5-15%

- **Dermoid of the cornea**
What is the incidence of Goldenhar?

What is the incidence of Goldenhar?

About 1/4000 live births

What is the inheritance pattern?

It is sporadic

Is there a sex predilection?

Yes, males are twice as likely to be affected
What is the incidence of Goldenhar?
About 1/4000 live births

- Dermoid of the cornea

Goldenhar syndrome

- Goldenhar syndrome
  - Lid colobomas
  - Duane syndrome
  - Ear abnormalities
  - Hemifacial microsomia

A minority (5-15%) have mental retardation

What nonocular findings are usually present?
- Hemifacial microsomia (maxillary/mandibular hypoplasia)
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With what syndrome is corneal dermoid associated?

Goldenhar syndrome

Oculo-Auriculo-Vertebral (OAV) syndrome

Where specifically are dermoids commonly located in Goldenhar?

At the limbus

What other ocular/periocular abnormalities are common in Goldenhar?

Upper lid colobomas
Duane syndrome

Are they cognitively impaired?

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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? Yes, M v F are # x as likely to be affected

Goldenhar syndrome

- Dermoid of the cornea

- Upper lid colobomas
- Duane syndrome

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

- Nothing starts w/ 'N'

- About 1/4000 live births
- It is sporadic
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Goldenhar

G O L D E N H A R
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Goldenhar

Goldenhar

Goldenhar
With what syndrome is corneal dermoid associated? Goldenhar syndrome

What is the ‘full’ name of the dermoid in question? one word epibulbar dermoid

● Dermoid of the cornea
With what syndrome is corneal dermoid associated?
Goldenhar syndrome

What is the 'full' name of the dermoid in question?
Epibulbar dermoid

Note: There is another legit answer, so if you came up with that one, no worries (we'll identify it shortly)

- Dermoid of the cornea
With what syndrome is corneal dermoid associated? **Goldenhar syndrome**

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

Where specifically are dermoids commonly located in Goldenhar? At the limbus

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What nonocular findings are usually present? Hemifacial microsomia (maxillary/mandibular hypoplasia), Ear abnormalities (pre-auricular appendages; aural fistulae)

Answer starts with an ‘O’
With what syndrome is corneal dermoid associated?
Goldenhar syndrome

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

- Dermoid of the cornea

Goldenhar
OAV syndrome

Told ya
With what syndrome is corneal dermoid associated?
Goldenhar syndrome

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

What is the classic vertebral finding?
Hemivertebrae, aka butterfly vertebrae

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What are dermoids commonly associated with? Goldenhar syndrome

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What other ocular/periocular abnormalities are common in Goldenhar? Upper lid colobomas, Duane syndrome

Are they cognitively impaired? A minority (5-15%) have mental retardation

What nonocular findings are usually present? Hemifacial microsomia, ear abnormalities

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What is the non-epponymous name for Alagille syndrome? Arterohepatic dysplasia

Under what circumstance is an ophthalmologist likely to encounter an Alagille pt? An infant with jaundice will present to the eye service as a 'rule out Alagille syndrome' consult

What eye finding is the ophthalmologist looking for in order to rule in/out Alagille syndrome? The presence of a posterior embryotoxon on the infant's corneas

What is a posterior embryotoxon? An anteriorly displaced and thickened Schwalbe's line

Dermoid of the cornea
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<th>Answer</th>
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What is a posterior embryotoxon?
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Dermoid of the cornea
**Q**

*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?*

---

*Now the ‘L’ and the ‘D’ have changed colors…*

---

**Dermoid of the cornea**

---

One answer starts with an ‘L’

The other with a ‘D’
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What is Goldenhar’s noneponymous name? **Oculo-Auriculo-Vertebral (OAV) syndrome**

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--Lid coloboma  
--Duane syndrome

*Dermoid of the cornea*
With what syndrome is corneal dermoid associated?
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What is Goldenhar’s noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?
--Lid coloboma
--Duane syndrome

Does the coloboma tend to be in the upper lid, or the lower?

Dermoid of the cornea
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

Does the coloboma tend to be in the upper lid, or the lower?
Depends on who you ask. The BCSC Cornea book says the upper, whereas the Plastics book indicates the lower. (The Peds book doesn’t address this issue.) Caveat emptor.

Dermoid of the cornea
With what syndrome is corneal dermoid associated?  
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Where specifically are dermoids commonly located in Goldenhar?  
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Are they cognitively impaired?  
A minority (5-15%) have mental retardation

One answer starts with an ‘E’  
The other with a ‘H’
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One answer starts with an ‘E’ 
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What nonocular findings are usually present?  
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Which side of the face is more likely to be affected?  

### Dermoid of the cornea
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--Ear abnormalities (pre-auricular appendages; aural fistulae)
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Which side of the face is more likely to be affected?
The right

● Dermoid of the cornea
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Why?

● Dermoid of the cornea

Goldenhar  
OAV syndrome  
Lid coloboma  
Duane syndrome  
Ear abnormalities  
Nothing starts w/ ‘N’  
Hemifacial microsomia  
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

What nonocular findings are usually present?  
--Ear abnormalities (pre-auricular appendages; aural fistulae)  
--Hemifacial microsomia (maxillary/mandibular hypoplasia)

Which side of the face is more likely to be affected?  
The right

Why?  
I have no idea

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Where are epibulbar dermoids commonly located in Goldenhar?

- Dermoid of the cornea

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At the limbus

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Where are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Several slides ago I acknowledged that epibulbar dermoids had another legit name. At long last—what is it?
**Limbal** dermoids

● Dermoid of the cornea

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At the limbus

Are Goldenhar pts cognitively impaired?  
A minority (5-15%) have mental retardation

---

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Retardation in ~10%
With what syndrome is **corneal dermoid** associated?
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What is the relationship between corneal/limbal dermoids and **dermoid cysts**?

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There is none; they are distinct and separate entities

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- **Dermoid of the cornea**

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Where are dermolipomas typically located?

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Where are dermolipomas typically located?
The temporal fornix

Dermoid of the cornea

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Where are dermolipomas typically located? The temporal fornix.

With what similar-appearing but etiologically-distinct condition are dermolipomas often confused? Prolapsed orbital fat.

Dermoid of the cornea

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Where are dermolipomas typically located? The temporal fornix

With what similar-appearing but etiologically-distinct condition are dermolipomas often confused? Prolapsed orbital fat

How can the two be differentiated at the slit lamp? --

- -

Dermoid of the cornea
A/Q

With what syndrome is corneal dermoid associated?
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Where are dermolipomas typically located?
The temporal fornix

With what similar-appearing but etiologically-distinct condition are dermolipomas often confused?
Prolapsed orbital fat

How can the two be differentiated at the slit lamp?
--By color (prolapsed fat is yellow, whereas a dermolipoma is pinkish-white)
--By texture (prolapsed fat is squishy, whereas a dermolipoma is firm)

Dermoid of the cornea

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

What is the relationship between corneal/limbal dermoids and *dermoid cysts*?
There is none; they are distinct and separate entities

*Is there a relationship between corneal/limbal dermoids and *lipodermoids* (aka *dermolipomas*)?*
Yes. The relationship is that, like dermoids, lipodermoids are associated with Goldenhar syndrome.

Where are *dermolipomas* typically located?
The temporal fornix

What about on MRI/CT—how can they be differentiated?

- By **color** (prolapsed fat is *yellow*, whereas a dermolipoma is *pinkish-white*)
- By **texture** (prolapsed fat is squishy, whereas a dermolipoma is firm)

---

**Dermoid of the cornea**

- At the limbus
- Retardation in ~10%

Nothing starts w/ ‘N’

**Hemifacial microsomia**

* Goldenhar syndrome
* Oculo-Auriculo-Vertebral (OAV) syndrome
  - Upper lid colobomas
  - Duane syndrome
- Ear abnormalities (pre-auricular appendages; aural fistulae)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

- Dermoid of the cornea
  - Peters anomaly
  - Endothelial dystrophy
  - Trauma (endothelial, from forceps, etc)
  - Ulcer

- Goldenhar mnemonic for infantile cloudy cornea

- Dermoid cysts

- Lipodermoids (aka dermolipomas)

- Prolapsed orbital fat

- Protrusio bulbi

- Phthisis bulbi

- Proptosis

- Ptosis

- Premacular dermoid

- Proximal retinal detachment

- Retinal dialysis

- Retinal traction

- Retinal detachment

- Rubeosis iridis

- Rhabdophthalmos

- Rheumatogenous retinal detachment

- Degenerative keratitis

- Keratomalacia

- Keratocanthoma

- Keratoconus
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Where are dermolipomas typically located?
The temporal fornix

What about on MRI/CT--how can they be differentiated?
On imaging, prolapsed orbital fat will be seen to be continuous with intraconal fat, whereas a dermolipoma will not.

How can the two be differentiated at the slit lamp?
--By color (prolapsed fat is yellow, whereas a dermolipoma is pinkish-white)
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Dermoid of the cornea

● At the limbus

Retardation in ~10%
QA

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

Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?
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- Sclerocornea;
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- Endothelial dystrophy (CHED)
- Dermoid of the cornea
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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

*The mnemonic should be modified to include these as well*
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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- Sclerocornea; congenital hereditary Stromal dystrophy (CHSD)
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- Peters anomaly
- Endothelial dystrophy (CHED); Elevated IOP (infantile glaucoma)
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Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

<table>
<thead>
<tr>
<th>Condition</th>
<th>CCT</th>
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</tr>
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<tbody>
<tr>
<td>CHED</td>
<td></td>
<td></td>
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*CCT, corneal diameter and IOP are key to differentiating among CHED, CHSD and infantile glaucoma. Fill in the blanks with the terms increased, decreased, and WNL.*

*(CCT = Central corneal thickness)*
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

CCT, corneal diameter and IOP are key to differentiating among CHED, CHSD and infantile glaucoma. Fill in the blanks with the terms increased, decreased, and WNL.

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Sclerocornea: congenital hereditary Stromal dystrophy (CHSD)

Trauma (endothelial; from forceps, etc)

Ulcer

Metabolic disorders

Peters anomaly

Endothelial dystrophy (CHED); elevated IOP (infantile glaucoma)

Dermoid of the cornea

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**CCT, corneal diameter** and **IOP** are key to differentiating among **CHED**, **CHSD** and **infantile glaucoma**. Fill in the blanks with the terms **increased**, **decreased**, and **WNL**.

But the OHTS* demonstrated that glaucoma is associated with **decreased** CCT, not increased. Why is CCT increased in infantile glaucoma?

---

*OHTS = Ocular Hypertension Treatment Study*
Everyone knows the STUMPED mnemonic for infantile cloudy cornea—or do you?

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But the OHTS* demonstrated that glaucoma is associated with decreased CCT, not increased. Why is CCT increased in infantile glaucoma?
The high pressure leads to endothelial-cell dysfunction with subsequent corneal edema.

*OHTS = Ocular Hypertension Treatment Study