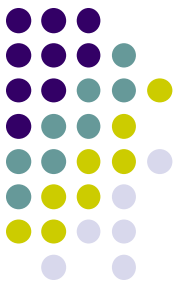


Q

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

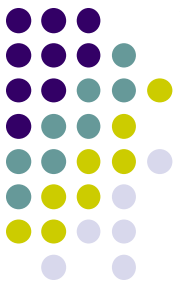
- Evident at or within



A

Primary Congenital Glaucoma

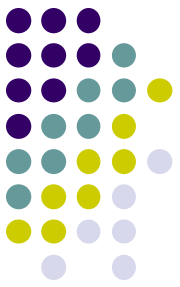
- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**



Q

Primary Congenital Glaucoma

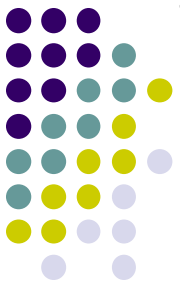
- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - 1/ live births



A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - **1/10K** live births





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...

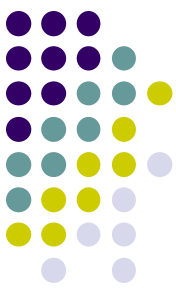
...

unilateral
v bilateral

A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - 1/**10K** live births
 - 2/3 are...
...**bilateral**



Q

Primary Congenital Glaucoma

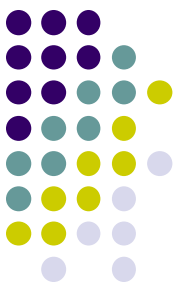
- **Primary Congenital Glaucoma**

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...

...**bilateral**

...

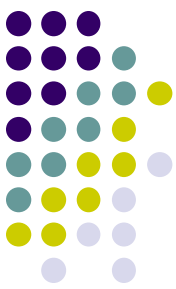
male vs
female



A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - 1/**10K** live births
 - 2/3 are...
 - ...**bilateral**
 - ...**male**

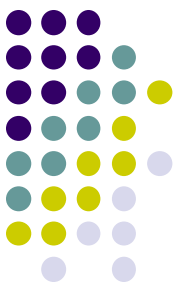


Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age

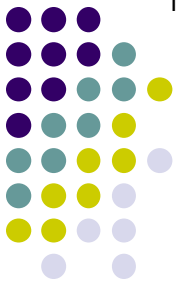


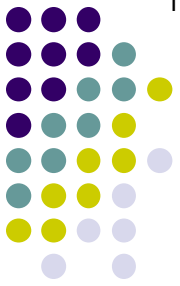
A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age **6 months**





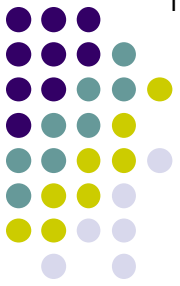
Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age **6 months**
- Associated genetically with POAG?

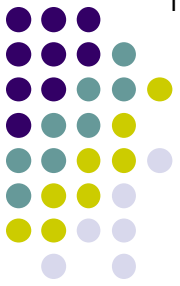
yes/
no



A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - 1/**10K** live births
 - 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age **6 months**
 - Associated genetically with POAG? **No**

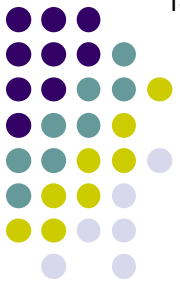


Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age **6 months**
- Associated genetically with POAG? **No**
- Is secondary to **structure** dysplasia in the **absence** of other

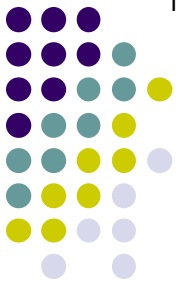


A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at **birth** or within **first few years**
- 1/**10K** live births
- 2/3 are...
 - ...**bilateral**
 - ...**male**
 - ...diagnosed by age **6 months**
- Associated genetically with POAG? **No**
- Is secondary to **angle** dysplasia in the **absence** of other **ocular or systemic abnormalities**



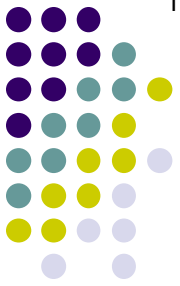
Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at birth or within first few years
- 1/10K live births
- 2/3 are...
 - ...bilateral
 - ...male
 - ...diagnosed by age 6 months
- Associated genetically with POAG? No *association with*
- Is secondary to angle dysplasia in the ~~absence~~ of other ocular or systemic abnormalities

What if other ocular and/or systemic abnormalities are present?



A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Evident at birth or within first few years
- 1/10K live births
- 2/3 are...
 - ...bilateral
 - ...male
 - ...diagnosed by age 6 months
- Associated genetically with POAG? No *association with*
- Is secondary to angle dysplasia in the ~~absence~~ of other ocular or systemic abnormalities

*What if other ocular and/or systemic abnormalities are present?
Would be **secondary** congenital glaucoma (or confusingly,
primary **developmental** glaucoma)*

Q

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

- **The classic triad of signs/symptoms:**

1)

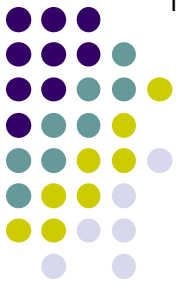


2)



3)





- **Primary Congenital Glaucoma**

- Clinical presentation

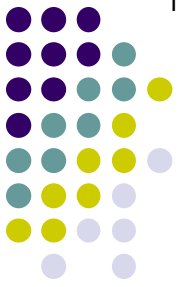
- **The classic triad of signs/symptoms:**

- 1) **Epiphora**

- 2) **Photophobia**

- 3) **Blepharospasm**

Primary Congenital Glaucoma

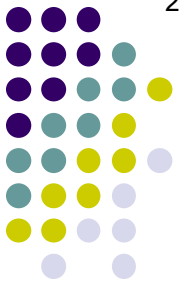


Epiphora



Photophobia/blepharospasm

Congenital glaucoma: The triad



- **Primary Congenital Glaucoma**

- Clinical presentation

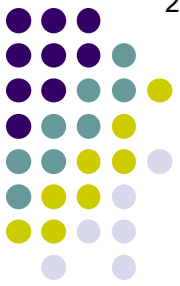
- **The classic triad of signs/symptoms:**

- 1) **Epiphora** (common misdiagnosis: Congenital
- 2) **Photosensitivity**
- 3) **Blepharospasm**

three words

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

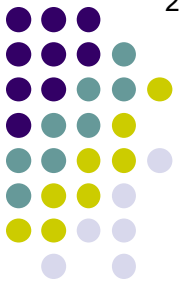
- Clinical presentation

- **The classic triad of signs/symptoms:**

- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

Q

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

- **The classic triad of signs/symptoms:**

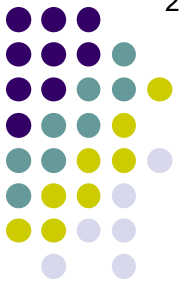
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

- Cornea:

- Cloudiness secondary to

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

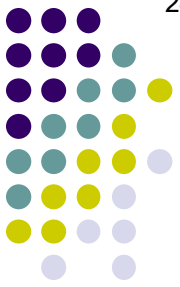
- **The classic triad of signs/symptoms:**

- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

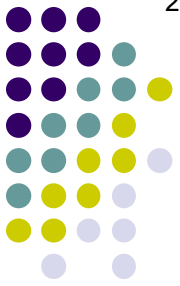
- Cornea:

- Cloudiness secondary to **edema**

Primary Congenital Glaucoma



Congenital glaucoma: Corneal cloudiness



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

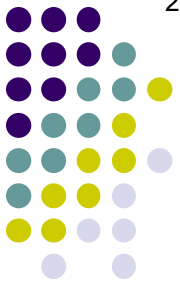
● The classic triad of signs/symptoms:

- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

- Cloudiness

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



A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

- Clinical presentation

- **The classic triad of signs/symptoms:**

- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimal sac obstruction**)

- 2) **Photosensitivity**

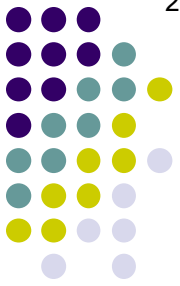
- 3) **Blepharospas**

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

STUMPED

- **Cornea:**

- **Cloudiness**



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

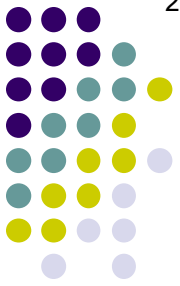
● Cornea:

- Cloudiness

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

STUMPED

Why?



A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

- Cloudiness

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

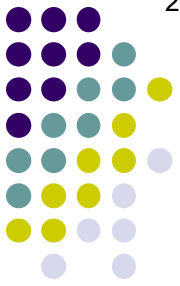
STUMPED

Why?

Coming in hot...

Q

Primary Congenital Glaucoma

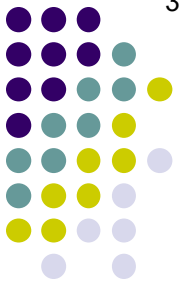


- S
- T
- U
- M
- P
- E
- D

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

A

Primary Congenital Glaucoma

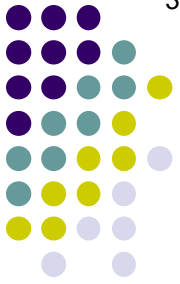


- S
- T
- U
- M
- P
- E
- D

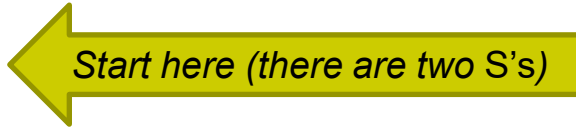
What is the purpose of the STUMPED mnemonic, ie, what does it help to remember?
The DDx for a cloudy cornea in an infant

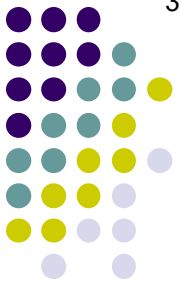
Q

Primary Congenital Glaucoma



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



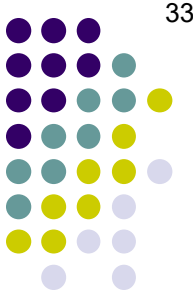


- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**
- **U**
- **M**
- **P**
- **E**
- **D**



A

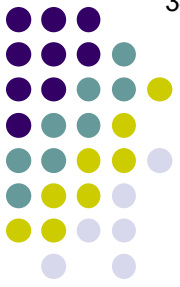
Primary Congenital Glaucoma



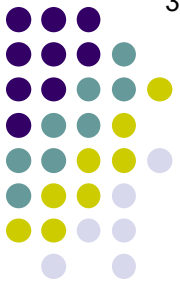
- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U** (*Tears in Descemet's membrane* works too)
- **M**
- **P**
- **E**
- **D**


Q

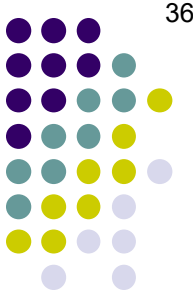
Primary Congenital Glaucoma



- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U** ← *next*
- **M**
- **P**
- **E**
- **D**

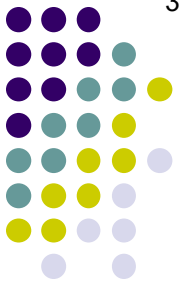


- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M** 
- **P**
- **E**
- **D**



- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**
- **E**
- **D**

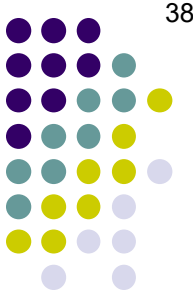




- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E** ← *next (there are two E's)*
- **D**

A

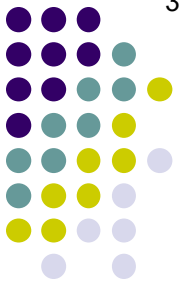
Primary Congenital Glaucoma




- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**levated IOP; **E**ndothelial dystrophy (CHED)
- **D** (*Edema* works too)

Q

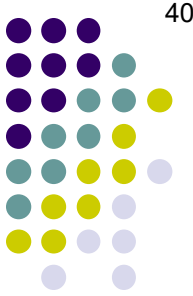
Primary Congenital Glaucoma



- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**levated IOP; **E**ndothelial dystrophy (CHED)
- **D** ← 

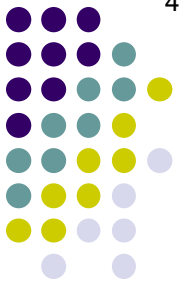
A

Primary Congenital Glaucoma

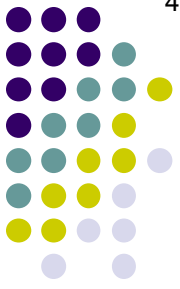


- **S**clerocornea; congenital hereditary **S**tromal dystrophy
- **T**rauma (endothelial; ie, from forceps)
- **U**lcer
- **M**etabolic disorders
- **P**eters anomaly
- **E**levated IOP; **E**ndothelial dystrophy (CHED)
- **D**ermoid of the cornea

Primary Congenital Glaucoma



- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- *For more on the STUMPED mnemonic, see slide-set K9)*
- Peters anomaly
- Elevated IOP; Endothelial dystrophy (CHED)
- Dermoid of the cornea



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

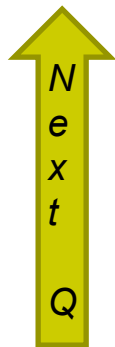
● Clinical presentation

● The classic triad of signs/symptoms:

- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

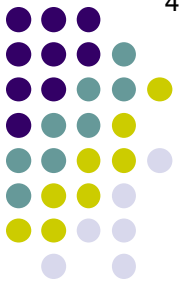
● Cornea:

- Cloudiness secondary to **edema**
- **eponym** striae



A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

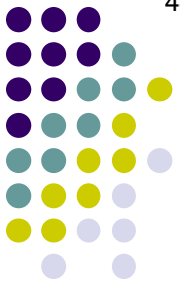
- Clinical presentation

- **The classic triad of signs/symptoms:**

- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

- Cornea:

- Cloudiness secondary to **edema**
- **Haab's** striae



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

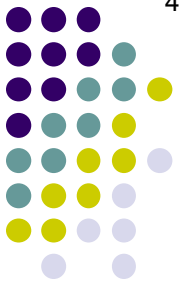
● The classic triad of signs/symptoms:

- 1) **Epiphora** (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

- Cloudiness secondary to
- **Haab's striae**

Haab striae represent what sort of pathology, ie, which corneal structure is damaged, and in what way?



A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

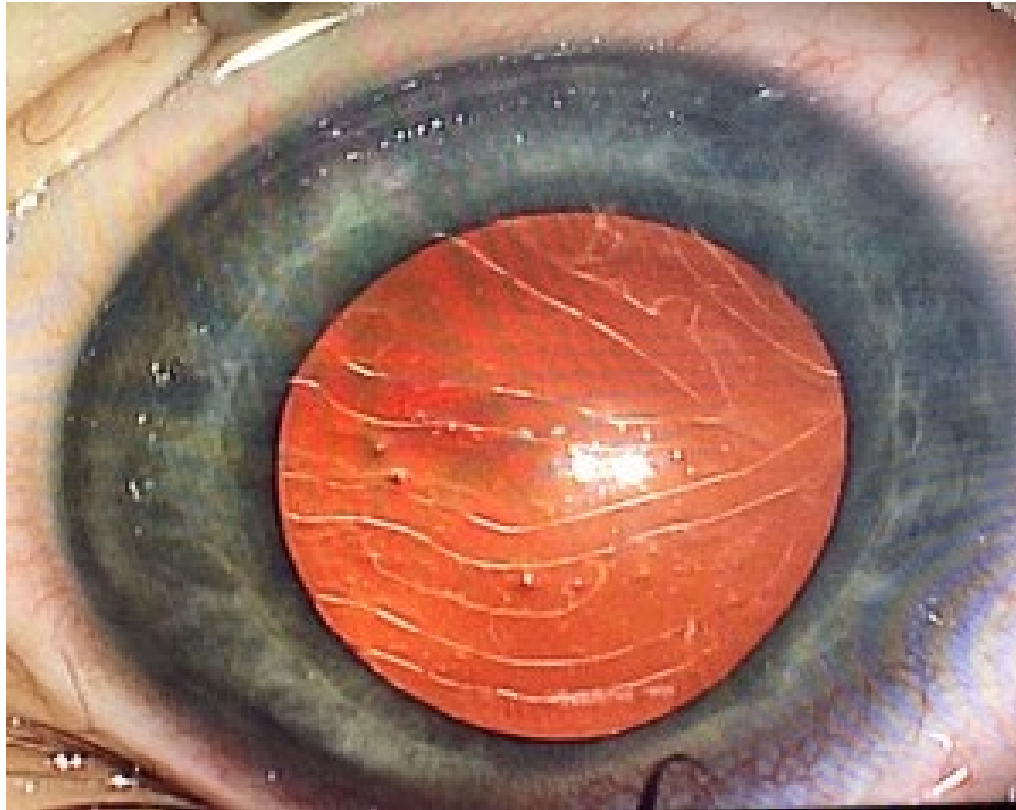
- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

- Cloudiness secondary to
- Haab's striae

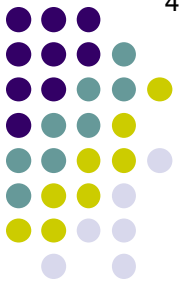
*Haab striae represent what sort of pathology, ie, which corneal structure is damaged, and in what way?
Tears in Descemet's secondary to corneal stretching*

Primary Congenital Glaucoma



Congenital glaucoma: Haab striae





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

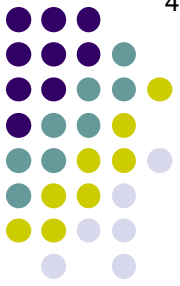
- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

- Cloudiness secondary to
- **Haab's striae**

*Haab striae represent what sort of pathology, ie, which corneal structure is damaged, and in what way?
Tears in Descemet's secondary to corneal stretching*

What is the mechanism?



A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

- Cloudiness secondary to
- **Haab's striae**

*Haab striae represent what sort of pathology, ie, which corneal structure is damaged, and in what way?
Tears in Descemet's secondary to corneal stretching*

What is the mechanism?

Mechanical deformation caused by the elevated IOP



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

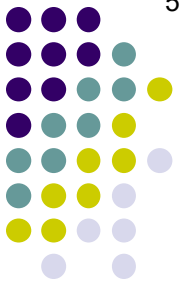
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● Cornea:

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- **Haab's** striae
- Horizontal diameter >



● Primary Congenital Glaucoma

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- Horizontal diameter > **11.5 mm**



Q

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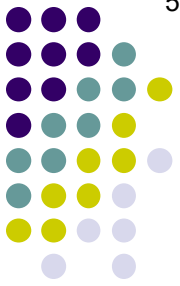
- Cloudiness secondary to **edema**
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- Horizontal diameter > **11.5 mm**

● IOP:

- **Normal IOP:** Infancy ; age 7 years

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

- **The classic triad of signs/symptoms:**

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- 2) **Photosensitivity**
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- Cornea:

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- Horizontal diameter > **11.5 mm**

- IOP:

- **Normal IOP:** Infancy **10-12** ; age 7 years **~14**



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

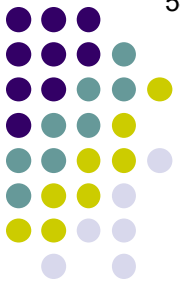
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- **Haab's** striae
- Horizontal diameter > **11.5 mm**

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- In congenital glaucoma:
 - **#** unanaesthetized



● Primary Congenital Glaucoma

● Clinical presentation

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- **Haab's** striae
- Horizontal diameter > **11.5 mm**

● IOP:

- **Normal** IOP: Infancy **10-12** ; age 7 years **~14**
- In congenital glaucoma:
 - **30-40** unanaesthetized



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

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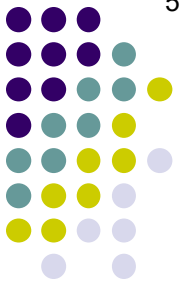
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 - **30-40** unanaesthetized
 - **#** even under anesthesia



A

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- 2) Photosensitivity
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● Cornea:

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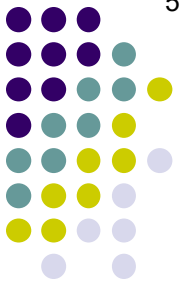
● IOP:

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 - >20 even under anesthesia

General anesthetic(s) that raise IOP:

A

Primary Congenital Glaucoma



- Primary Congenital Glaucoma

- Clinical presentation

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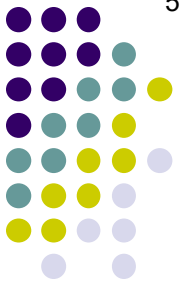
- Cornea:

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- IOP:

- Normal IOP: Infancy 10-12 ; age 7 years ~14
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*General anesthetic(s) that raise IOP:
Ketamine*



Q

Primary Congenital Glaucoma

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● Cornea:

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- **Haab's** striae
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● IOP:

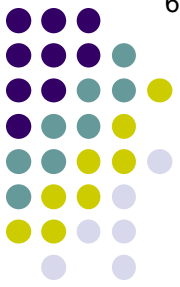
- **Normal IOP:** Infancy **10-12** ; age 7 years **~14**
- In congenital glaucoma:
 - **30-40** unanaesthetized
 - **>20 even under anesthesia**

General anesthetic(s) that raise IOP:
Ketamine

General anesthetic(s) that lower IOP:

A

Primary Congenital Glaucoma



- Primary Congenital Glaucoma

- Clinical presentation

- The classic triad of signs/symptoms:

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- 3) Blepharospasm

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- Haab's striae
- Horizontal diameter > 11.5 mm

- IOP:

- Normal IOP: Infancy 10-12 ; age 7 years ~14
- In congenital glaucoma:
 - 30-40 unanaesthetized
 - >20 even under anesthesia

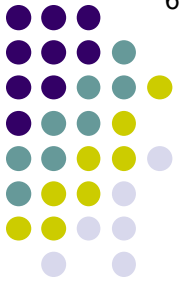
General anesthetic(s) that raise IOP:
Ketamine

General anesthetic(s) that lower IOP:
Pretty much everything else

Q

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**
 - Natural history if untreated:
 - Progresses to visual acuity



A

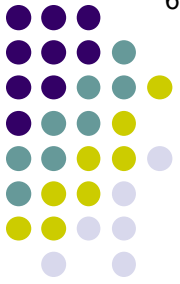
Primary Congenital Glaucoma

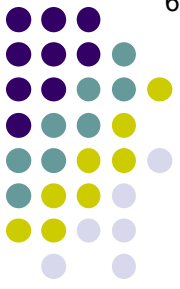
- **Primary Congenital Glaucoma**

- Natural history if untreated:

- Progresses to **NLP**

(NLP = No light perception)





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Natural history if untreated:

- Progresses to NLP

- Cornea...

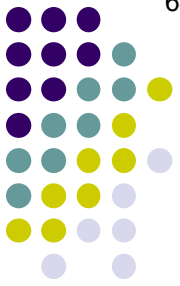
- bad sequelae 1 and bad sequelae 2

A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

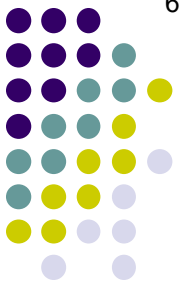
- Natural history if untreated:
 - Progresses to **NLP**
 - Cornea...
 - **Opacifies** and **vascularizes**



Primary Congenital Glaucoma



Congenital glaucoma: Corneal opacification, vascularization





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

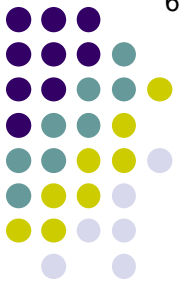
- Natural history if untreated:
 - Progresses to NLP
 - Cornea...
 - Opacifies and vascularizes
 - diameter # (mm)

A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Natural history if untreated:
 - Progresses to NLP
 - Cornea...
 - Opacifies and vascularizes
 - diameter ~17 mm





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Natural history if untreated:

- Progresses to NLP

- Cornea...

 - Opacifies and vascularizes

 - diameter ~17 mm

- Pseudoproptosis 2° to translation: ox's eye (enlargement of globe secondary to chronically elevated IOP)

A

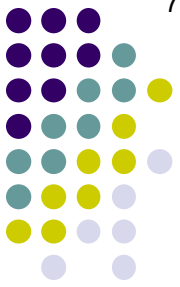
Primary Congenital Glaucoma



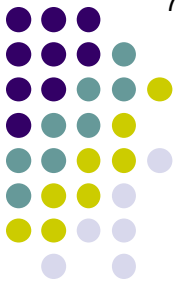
● Primary Congenital Glaucoma

- Natural history if untreated:
 - Progresses to NLP
 - Cornea...
 - Opacifies and vascularizes
 - diameter ~17 mm
 - Pseudoproptosis 2° to buphthalmos (enlargement of globe secondary to chronically elevated IOP)

Primary Congenital Glaucoma



Congenital glaucoma: Pseudoproptosis 2ndry to buphthalmos



Q

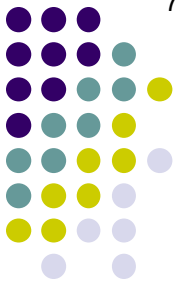
Primary Congenital Glaucoma

● Primary Congenital Glaucoma

- Natural history if untreated:
 - Progresses to NLP
 - Cornea...
 - Opacifies and vascularizes
 - diameter ~17 mm
 - Pseudoproptosis 2° to buphthalmos (enlargement of globe secondary to chronically elevated IOP)
 - Amblyopia common vs uncommon

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Natural history if untreated:

- Progresses to **NLP**
- Cornea...
 - **Opacifies** and **vascularizes**
 - diameter **~17 mm**
- Pseudoproptosis 2° to **buphthalmos** (enlargement of globe secondary to chronically elevated ↑ IOP)
- Amblyopia **common**