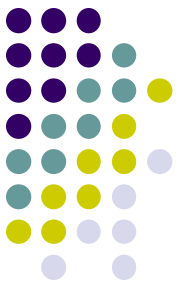


Q

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

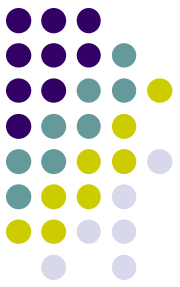
- Evident at or within



A

Primary Congenital Glaucoma

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



Q

Primary Congenital Glaucoma

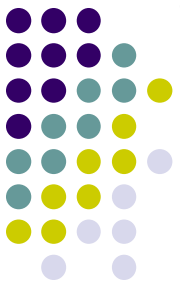
- **Primary Congenital Glaucoma**
 - Evident at **birth** or within **first few years**
 - 1/ live births



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...

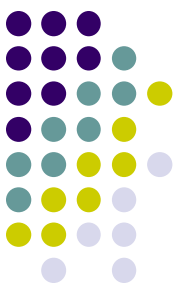
...

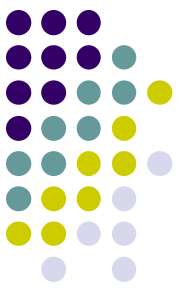
unilateral
v bilateral

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

...

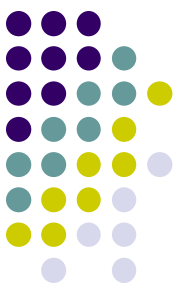
male vs
female

A

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

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

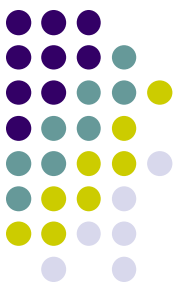


Q

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

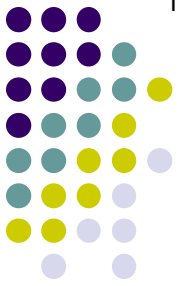


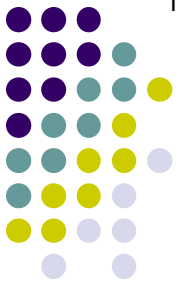
A

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**





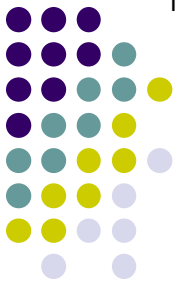
Q

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?

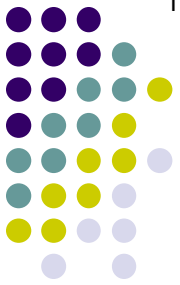
yes/
no



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**

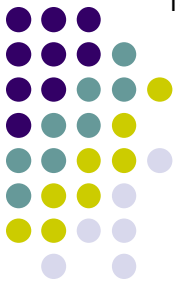


Q

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**



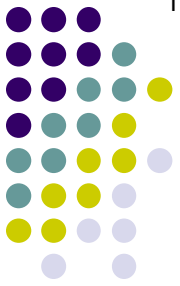
Q

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:**

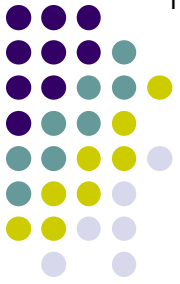
1)

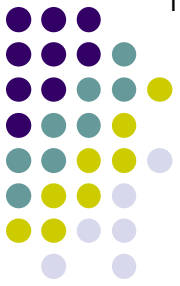


2)



3)





- **Primary Congenital Glaucoma**

- Clinical presentation

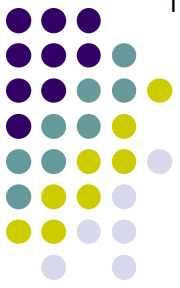
- **The classic triad of signs/symptoms:**

- 1) **Epiphora**

- 2) **Photosensitivity**

- 3) **Blepharospasm**

Primary Congenital Glaucoma

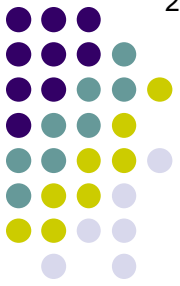


Epiphora



Photophobia/blepharospasm

Congenital glaucoma: The triad



- **Primary Congenital Glaucoma**

- Clinical presentation

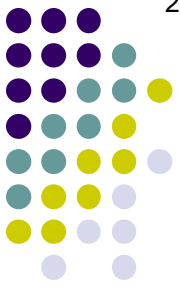
- **The classic triad of signs/symptoms:**

- 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 **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasmia**

Q

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

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

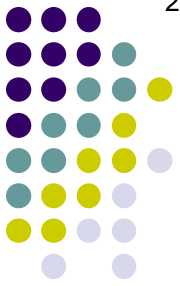
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

- Cornea:

- Cloudiness secondary to

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Clinical presentation

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

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

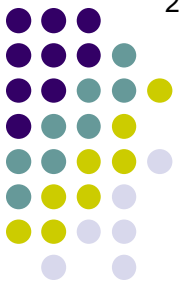
- 2) **Photosensitivity**

- 3) **Blepharospasm**

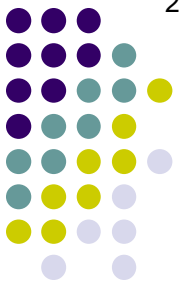
- Cornea:

- Cloudiness secondary to **edema**

Primary Congenital Glaucoma



Congenital glaucoma: Corneal cloudiness



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

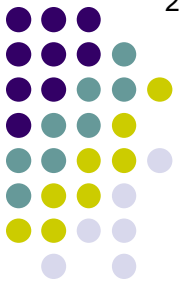
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

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

A

Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

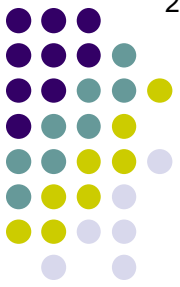
- Clinical presentation

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

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

- Cornea:

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



Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

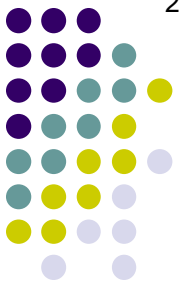
● The classic triad of signs/symptoms:

- 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:

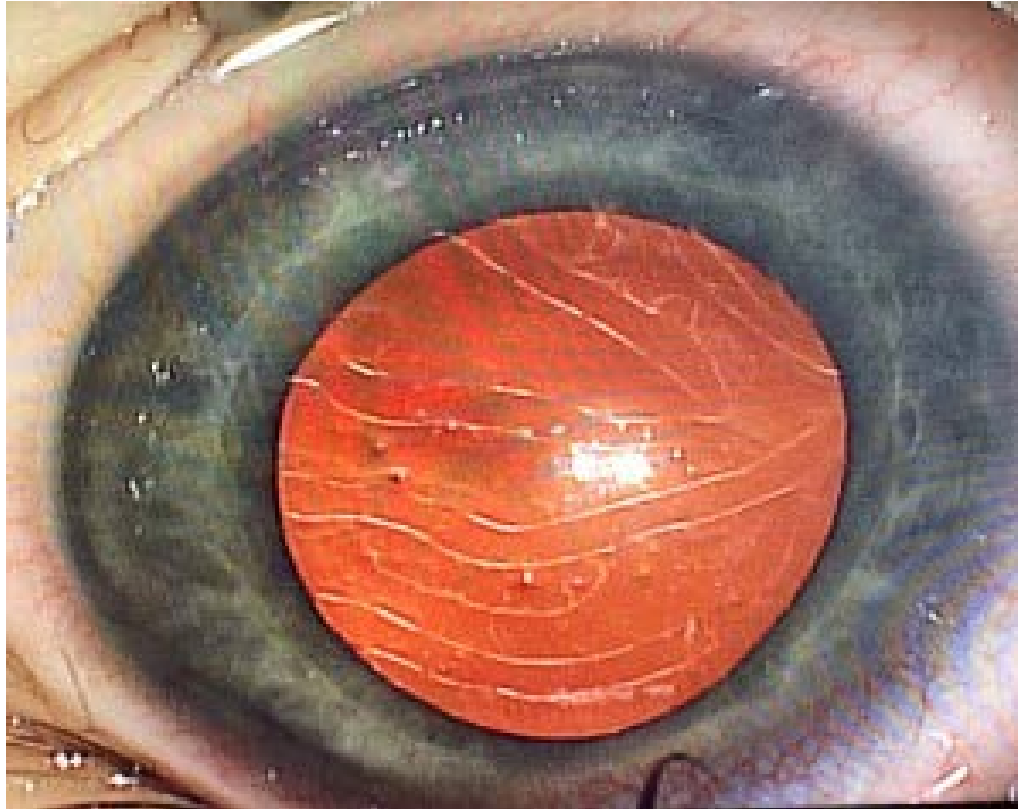
- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● Cornea:

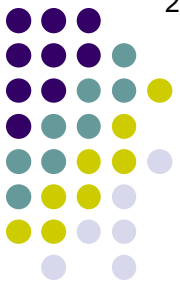
- Cloudiness secondary to
- Haab's striae

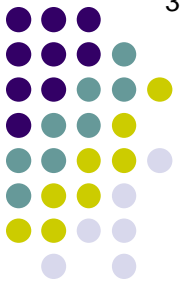
*Haab striae represent what sort of pathology, ie, which corneal structure is damaged, and in what way?
Tears in Descemet's secondary to corneal stretching*

Primary Congenital Glaucoma



Congenital glaucoma: Haab striae





Q

Primary Congenital Glaucoma

● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

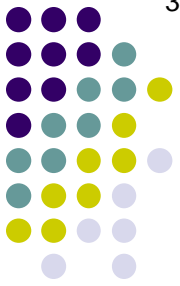
- 1) Epiphora (common misdiagnosis: Congenital lacrimal sac obstruction)
- 2) Photosensitivity
- 3) Blepharospasm

● 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

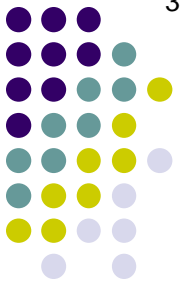
● Cornea:

- 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

● Clinical presentation

● The classic triad of signs/symptoms:

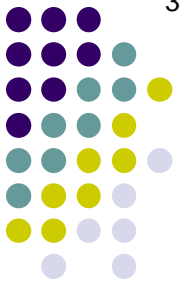
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

- Cloudiness secondary to **edema**
- **Haab's** striae
- Horizontal diameter >

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
- Horizontal diameter > **11.5 mm**



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**
- **Haab's** striae
- 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:

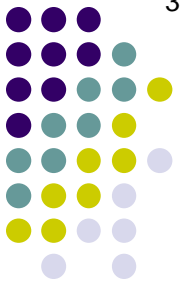
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

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

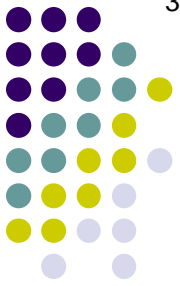
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrimonal sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

- Cloudiness secondary to **edema**
- **Haab's** striae
- Horizontal diameter > **11.5 mm**

● IOP:

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



● Primary Congenital Glaucoma

● Clinical presentation

● The classic triad of signs/symptoms:

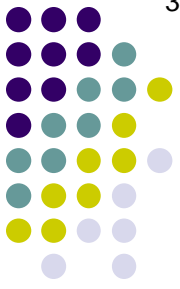
- 1) **Epiphora** (common misdiagnosis: Congenital **lacrima sac obstruction**)
- 2) **Photosensitivity**
- 3) **Blepharospasm**

● Cornea:

- Cloudiness secondary to **edema**
- **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

● 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
- Horizontal diameter > **11.5 mm**

● IOP:

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



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
 - 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



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

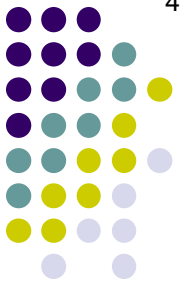
● Cornea:

- Cloudiness secondary to edema
- 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:



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

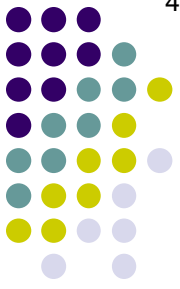
● Cornea:

- Cloudiness secondary to edema
- 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



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:

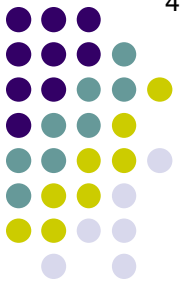
- Cloudiness secondary to edema
- Haab's striae
- Horizontal diameter > 11.5 mm

● 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:



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
- 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

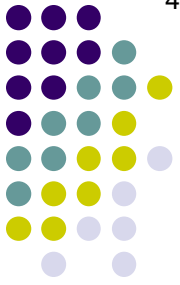
General anesthetic(s) that raise IOP:
Ketamine

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

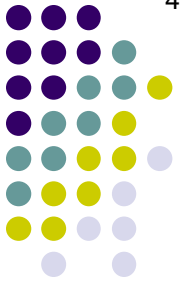
Primary Congenital Glaucoma

- **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...

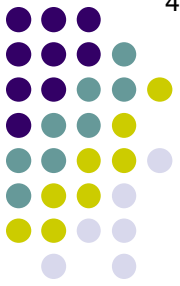
- bad sequelae 1 and bad sequelae 2

A

Primary Congenital Glaucoma

- **Primary Congenital Glaucoma**

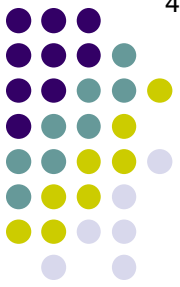
- Natural history if untreated:
 - Progresses to **NLP**
 - Cornea...
 - **Opacifies** and **vascularizes**

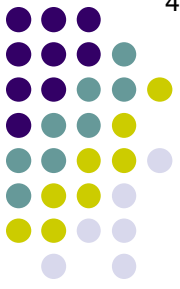


Primary Congenital Glaucoma



Congenital glaucoma: Corneal opacification, vascularization





Q

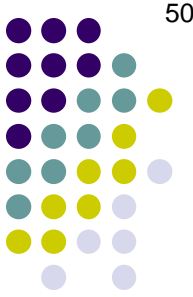
Primary Congenital Glaucoma

● Primary Congenital Glaucoma

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

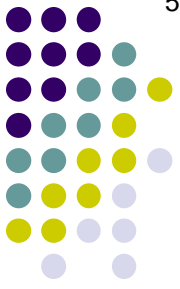
A

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...

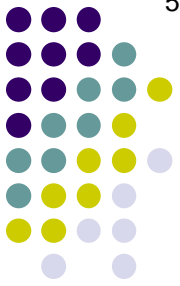
 - Opacifies and vascularizes

 - diameter ~17 mm

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

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Primary Congenital Glaucoma



- **Primary Congenital Glaucoma**

- Natural history if untreated:

- Progresses to **NLP**

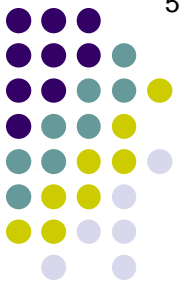
- Cornea...

- **Opacifies** and **vascularizes**

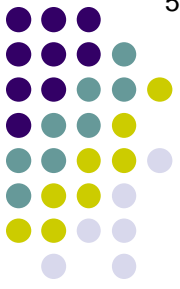
- diameter **~17 mm**

- Pseudoproptosis 2° to **buphthalmos** (enlargement of globe secondary to chronically elevated IOP)

Primary Congenital Glaucoma



Congenital glaucoma: Pseudoproptosis 2ndry to buphthalmos

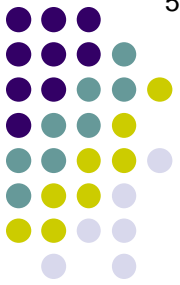


Q

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



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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