Primary Congenital Glaucoma

- Evident at [insert age] or within [insert period of time]
Primary Congenital Glaucoma

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

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

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

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

...
Primary Congenital Glaucoma

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

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

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

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

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

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

- Evident at birth or within the 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

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

Clinical presentation

The classic triad of signs/symptoms:

1) Epiphora
2) Photosensitivity
3) Blepharospasm
Primary Congenital Glaucoma

Clinical presentation

The classic triad of signs/symptoms:
1) Epiphora
2) Photosensitivity
3) Blepharospasm
Primary Congenital Glaucoma

Congenital glaucoma: The triad

Epiphora

Photophobia/blepharospasm
Primary Congenital Glaucoma

- Clinical presentation
  - The classic triad of signs/symptoms:
    1) **Epiphora** (common misdiagnosis: Congenital
    2) **Photosensitivity**
    3) **Blepharospasm**
Primary Congenital Glaucoma

Clinical presentation

The classic triad of signs/symptoms:

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

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

**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
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

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?
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

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
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 > **mm**
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
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 #; age 7 years #
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
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:
  - **#** unanaesthetized
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
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
  - # even under anesthesia
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
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:
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
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:
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
Primary Congenital Glaucoma

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

- Natural history if untreated:
  - Progresses to **NLP**

*(NLP = No light perception)*
Primary Congenital Glaucoma

- Natural history if untreated:
  - Progresses to NLP
  - Cornea...

- bad sequelae 1
- and
- bad sequelae 2
Primary Congenital Glaucoma

Natural history if untreated:

- Progresses to NLP
- Cornea…
  - Opacifies and vascularizes
Primary Congenital Glaucoma

Congenital glaucoma: Corneal opacification, vascularization
Primary Congenital Glaucoma

- Natural history if untreated:
  - Progresses to NLP
  - Cornea…
    - Opacifies and vascularizes
    - Diameter: # (mm)
Primary Congenital Glaucoma

Natural history if untreated:
- Progresses to NLP
- Cornea...
  - Opacifies and vascularizes
  - Diameter ~17 mm
Primary Congenital Glaucoma

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

Translation: ox’s eye
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
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
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