Primary Congenital Glaucoma
- Evident at birth or within a period of time
A

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

- 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
  - male
Primary Congenital Glaucoma

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

POAG = Primary open-angle 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
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...
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
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?
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

• Cornea:
  • Cloudiness secondary to edema
  • Haab's striae
  • Horizontal diameter >
Primary Congenital Glaucoma

Clinical presentation

The classic triad of signs/symptoms:
1) Epiphora (common misdiagnosis: Congenital lacrimal 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 edema
- Haab's striae
- Horizontal diameter esia
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

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

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
- 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:** 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
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 poor 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

- 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
Primary Congenital Glaucoma

Natural history if untreated:

- Progresses to NLP
- Cornea…
  - Opacifies and vascularizes
  - diameter ~17 mm
- Pseudoproptosis 2° to ox’s eye (enlargement of globe secondary to chronically elevated IOP)
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

- 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
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