Primary Congenital Glaucoma

- Evident at a young age or within a 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...
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
- 2/3 are male
- Diagnosed by age 6 months
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
Primary Congenital Glaucoma

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

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

Clinical presentation

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

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

Start here (there are two S's)
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- T
- U
- M
- P
- E
- D

next
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
  - (Tears in Descemet’s membrane works too)
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)

next
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer

next
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- P
- E
- D

next
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- E next (there are two E's)
- D
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Elevated IOP; Endothelial dystrophy (CHED)
- D (Edema works too)
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Elevated IOP; Endothelial dystrophy (CHED)

finally
Primary Congenital Glaucoma

- Sclerocornea; congenital hereditary Stromal dystrophy
- Trauma (endothelial; ie, from forceps)
- Ulcer
- Metabolic disorders
- Peters anomaly
- Elevated IOP; Endothelial dystrophy (CHED)
- Dermoid 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
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 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
    - 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 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)
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

*Note: NLP = No Light Perception*
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