Failure of lid differentiation

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

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- Lid coloboma
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Failure of lid differentiation

**Cryptophthalmos**

What is the classic appearance of cryptophthalmos?

- Ankyloblepharon
- Blepharophimosis
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

An expanse of uninterrupted skin from the forehead region to the cheek

Of sorts, but it is virtually always profoundly malformed

Can relatively normal lids be surgically constructed?

Not easily. The problem is, not only are the lids undifferentiated, but their architectural elements—tarsal plates, orbicularis muscle, meibomian glands, etc—are absent.
Failure of lid differentiation

What is the classic appearance of cryptophthalmos?
An expanse of uninterrupted skin from the forehead region to the cheek

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos

- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Cryptophthalmos
What is the classic appearance of cryptophthalmos?
An expanse of uninterrupted skin from the forehead region to the cheek

Is there an eye under the skin?

- Ankyloblepharon
- Blepharophimosis
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

- Ankyloblepharon
- Blepharophimosis
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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

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- Epicanthus (epicanthal folds)
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- Congenital ptosis
**Failure of lid differentiation**

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- Blepharophimosis
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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
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Failure of lid differentiation

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- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Upper-lid coloboma in Goldenhar
Failure of lid differentiation

**Associated with Goldenhar**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)

**Lid coloboma**

*Lid colobomas associated with Goldenhar—are they found in the upper, or the lower lid?*

Depends on who you ask. The BCSC Cornea book says the upper, whereas the Plastics book indicates the lower. EyeWiki says upper > lower. The Peds book doesn’t address the issue. Caveat emptor.
Failure of lid differentiation

Associated with Goldenhar

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- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- **Lid coloboma**
- Congenital ptosis

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Congenital lid abnormalities: Matching

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- Dystopia canthorum
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- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- **Lid coloboma**
- Congenital ptosis

What other ocular structures are commonly colobomatous?
(in general—not in Goldenhar)

Lid colobomas associated with Goldenhar—are they found in the upper, or the lower lid?
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**Congenital lid abnormalities: Matching**

*What other ocular structures are commonly colobomatous?*
- The iris
- The choroid
- The optic nerve head

*Lid colobomas associated with Goldenhar—are they found in the upper, or the lower lid?*
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- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- *Lid coloboma*
- Congenital ptosis
What other ocular structures are commonly colobomatous?
-- The iris
-- The choroid
-- The optic nerve head

What is the pathological process that accounts for all of these coloboma types?

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Failure of the embryonic fissure of the optic vesicle to close

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**Are lid colobomas associated with these others?**

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- The choroid
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- **Blepharophimosis**
- **Cryptophthalmos**
- **Congenital ectropion**
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- **Lid coloboma**
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- The iris
- The choroid
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**What is the pathological process that accounts for all of these coloboma types?**
Failure of the embryonic fissure of the optic vesicle to close

**Are lid colobomas associated with these others?**
No

**Why are lid colobomas not associated with the other types?**
Because the eyelids do not derive embryologically from the optic vesicle.

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Q

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What is the noneponymous name of Goldenhar syndrome?

Mnemonic: Goldenhar

Answer starts with ‘O’
A

Failure of lid differentiation

Associated with Goldenhar

Mnemonic: Goldenhar

What is the noneponymous name of Goldenhar syndrome?
Oculo-Auriculo-Vertebral (OAV) syndrome

Answer starts with ‘O’

Goldenhar

OAV syndrome

Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

What other adnexal/surface finding is common?
Limbal dermoids

What strabismus syndrome is associated with Goldenhar?
Duane syndrome

What nonocular findings are usually present?
Ear abnormalities (pre-auricular appendages; aural fistulae)

Emiefacial microsomia (maxillary/mandibular hypoplasia)

Are Goldenhar pts cognitively impaired?
Retardation is present in a minority (~10%)
Q

Congenital lid abnormalities: Matching

- Failure of lid differentiation
- Associated with Goldenhar
- Ankyloblepharon
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- Epicanthus (epicanthal folds)
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What is the non-eponymous name of Goldenhar syndrome?
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Mnemonic: Goldenhar

Goldenhar
OAV syndrome

Starts with ‘L’
Failure of lid differentiation
Associated with Goldenhar

Congenital lid abnormalities: Matching
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OAV syndrome
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Goldenhar

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Mnemonic: **Goldenhar**

- Goldenhar
- OAV syndrome
- Limbal dermoids
- Duane syndrome
- E
- N
- H
- A
- R

- Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

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Duane syndrome

What nonocular findings are usually present?

- E
- H
- N
- O
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Mnemonic: Goldenhar

Goldenhar
OAV syndrome
Limbal dermoids
Duane syndrome
Nothing starts with 'N'

- Congenital ptosis
A

Congenital lid abnormalities: Matching

- Failure of lid differentiation
- Associated with Goldenhar

Goldenhar

Mnemonic: Goldenhar

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- Hemifacial microsomia
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---

**Mnemonic: Goldenhar**

- Goldenhar
- OAV syndrome
- Limbal dermoids
- Duane syndrome
- Ear abnormalities
- Nothing starts w/ ‘N’
- Hemifacial microsomia
- Also, ‘A’
- R

---

- Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

**Congenital lid abnormalities: Matching**

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Mnemonic: Goldenhar

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- Limbal dermoids
- Duane syndrome
- Ear abnormalities
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- Nothing starts w/ ‘N’
- Also, ‘A’
- Retardation in ~10%

- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins

Congenital lid abnormalities: Matching

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- Epicanthus (epicanthal folds)
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- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Ankyloblepharon
An infant has several fine strands of tissue connecting the upper and lower lid margins. Is this ankyloblepharon?
Q/A

- Failure of lid differentiation
- Associated with Goldenhar
- **Fusion of all or part of the upper/lower margins**

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

An infant has several fine strands of tissue connecting the upper and lower lid margins. Is this ankyloblepharon? Yes, this is a variant called **ankyloblepharon**.
An infant has several fine strands of tissue connecting the upper and lower lid margins. Is this ankyloblepharon?
Yes, this is a variant called **ankyloblepharon filiform adnatum**.
Congenital lid abnormalities: Matching

Ankyloblepharon filiform adnatum
An infant has several fine strands of tissue connecting the upper and lower lid margins. Is this ankyloblepharon?
Yes, this is a variant called ankyloblepharon filiform adnatum.

How is this managed?
An infant has several fine strands of tissue connecting the upper and lower lid margins. Is this ankyloblepharon? Yes, this is a variant called **ankyloblepharon filiform adnatum**.

**How is this managed?**
By cutting the strands under local anesthesia in the office.
Congenital lid abnormalities: Matching

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- Lid coloboma
- Congenital ptosis

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion

not this one, duh
Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
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- Epicanthus (epicanthal folds)
- Lid coloboma
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- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
Congenital lid abnormalities: Matching

Congenital tarsal kink. (A) This child presented with a swollen and sore left eye with blepharospasm. (B) On eversion of the lid the horizontal kink in the tarsus can be seen. It runs the whole length of the tarsal plate, which is bent to 90°.
Q

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
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- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
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- Congenital ptosis
A

Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
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Congenital lid abnormalities: Matching

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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Epiblepharon
The ‘redundant lid tissue’—of what is it composed?

- A band of excess pre-tarsal obicularis muscle and overlying skin
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
**Redundant lid tissue** causes lashes to abut ocular surface

The ‘redundant lid tissue’—of what is it composed?
A band of excess pre-tarsal obicularis muscle and overlying skin

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- **Epiblepharon**
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- Congenital tarsal kink
- Congenital epicanthus (epicanthal folds)
- Lid coloboma
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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

Does epiblepharon usually involve the upper lid, or the lower?
Typically it involves only the nasal portion.
A

**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

Does epiblepharon usually involve the upper lid, or the lower?
The lower (epiblepharon rarely if ever occurs in the upper lid)
Q

Congenital lid abnormalities: Matching

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Does epiblepharon usually involve the upper lid, or the lower? The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
Q/A

**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

**Does epiblepharon usually involve the upper lid, or the lower?**
The lower (epiblepharon rarely if ever occurs in the upper lid)

**Does it typically involve the entirety of the lower lid, or only a portion?**
Typically it involves only the nasal portion

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- *Epiblepharon*
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Congenital ptosis
- Lid coloboma
- Epicanthus (epicanthal folds)
Failure of lid differentiation

Associated with Goldenhar

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- **Epiblepharon**
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Congenital lid hemifissure (epicanthal folds)
- Lid coloboma
- Congenital ptosis

**Does epiblepharon usually involve the upper lid, or the lower?**
The lower (epiblepharon rarely if ever occurs in the upper lid)

**Does it typically involve the entirety of the lower lid, or only a portion?**
Typically it involves only the nasal portion
Congenital lid abnormalities: Matching

Epiblepharon. Note primarily nasal involvement
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)?
Asian (#1) and Native-American

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Typically it involves only the nasal portion
A

**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
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- Ankyloblepharon
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- **Epiblepharon**
- Dystopia canthorum
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**Questions**

Does epiblepharon usually involve the upper lid, or the lower?
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Does it typically involve the entirety of the lower lid, or only a portion?
Typically it involves only the nasal portion
Epiblepharon in pt of Asian heritage
Two other common lid disorders also involves apposition of the lashes to the cornea. What are they?

- Epiblepharon
  - Posterior rather than anterior
  - Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)? Asian (#1) and Native-American

- Congenital ptosis

- Congenital tarsal kink

- Lid coloboma

- Epicanthus (epicanthal folds)

- Congenital entropion

- Euryblepharon

- Congenital entropion

- Bichirn

- Epiblepharon

- Ankyloblepharon

- Blepharophimosis

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- Congenital ectropion

- Epiblepharon

- Dystopia canthorum

- Epicanthus (epicanthal folds)

- Lid coloboma

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Associated with Goldenhar

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Ankyloblepharon

Blepharophimosis

Cryptophthalmos

Congenital ectropion

Epiblepharon

Dystopia canthorum

Congenital entropion

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Epicanthus (epicanthal folds)

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Congenital ptosis

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Two other common lid disorders also involves apposition of the lashes to the cornea. What are they? Entropion and trichiasis

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Two other common lid disorders also involves apposition of the lashes to the cornea. What are they? Entropion and trichiasis

In a few words, define…

Entropion: An inward rotation of the eyelid margin

Trichiasis: An inward rotation of eyelashes

So is epiblepharon a form of entropion, or trichiasis?

Neither. Note that, by definition, entropion involves an inward turning of the lid margin, and trichiasis an inward turning of the lashes. But in epiblepharon, both the lid margin and the lashes are positioned normally; it's the excess skin and muscle that pushes the lashes against the cornea.
Two other common lid disorders also involve apposition of the lashes to the cornea. What are they? Entropion and trichiasis.

In a few words, define…

Entropion:

Trichiasis:

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

Epiblepharon

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The lower (epiblepharon rarely if ever occurs in the upper lid)

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Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis

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In a few words, define…

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Two other common lid disorders also involves apposition of the lashes to the cornea. What are they? Entropion and trichiasis.

In a few words, define...
Entropion: An inward rotation of the eyelid margin
Trichiasis: A turning inward of eyelashes

Epiblepharon
- Definition: Redundant lid tissue causes lashes to abut ocular surface
- Does epiblepharon usually involve the upper lid, or the lower? The lower (epiblepharon rarely if ever occurs in the upper lid)
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Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)? Asian (#1) and Native-American

Congenital entropion

But to answer the question:

- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

Entropion and trichiasis:

- Two other common lid disorders also involves apposition of the lashes to the cornea. What are they? Entropion and trichiasis
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Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

Epiblepharon
Repetitive, often coarse, excess of skin and muscle in the lateral canthus (epicanthal folds)

Does epiblepharon usually involve the upper lid, or the lower?
The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
Typically it involves only the nasal portion

Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)?
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Other common lid disorders involves apposition of the lashes to the cornea. What are they?
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Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

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Fusion of all or part of the upper/lower margins

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Two other common lid disorders also involve apposition of the lashes to the cornea. What are they? Entropion and trichiasis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

Two other common lid disorders also involves apposition of the lashes to the cornea. What are they?

- Entropion
- Trichiasis

To summarize (this is worth re-reading until you grock it):

In **entropion**, the abnormally-positioned lid margin causes the normally-directed lashes to abut the ocular surface;

In **trichiasis**, abnormally-directed lashes growing from the normally-positioned lid margin abut the ocular surface; and

In **epiblepharon**, an overriding band of tissue causes the normally-directed lashes growing from the normally-positioned lid margin to abut the ocular surface.

Does epiblepharon usually involve the upper lid, or the lower?
The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
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Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)?
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To summarize (this is worth re-reading until you grock it):

In **entropion**, the abnormally-positioned lid margin causes the normally-directed lashes to abut the ocular surface;

In **trichiasis**, abnormally-directed lashes growing from the normally-positioned lid margin abut the ocular surface; and

In **epiblepharon**, an overriding band of tissue causes the normally-directed lashes growing from the normally-positioned lid margin to abut the ocular surface.
Two other common lid disorders also involve apposition of the lashes to the cornea. What are they?
Entropy and trichiasis

In a few words, define…
Entropy: An inward rotation of the eyelid margin
Trichiasis: An inward rotation of eyelashes

You evaluate an infant with epiblepharon and extensive lash-cornea touch. Should this be managed surgically or medically, and why?

Redundant lid tissue causes lashes to abut ocular surface

Does epiblepharon usually involve the upper lid, or the lower?
The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
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Epiblepharon

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Typically it involves only the nasal portion

Epiblepharon

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Two other common lid disorders also involve apposition of the lashes to the cornea. What are they?

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Redundant lid tissue causes lashes to abut ocular surface

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Two other common lid disorders also involve apposition of the lashes to the cornea. What are they? Entropion and trichiasis.

In a few words, define…
Entropion: An inward rotation of the eyelid margin
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You evaluate an infant with epiblepharon and extensive lash-cornea touch. Should this be managed surgically or medically, and why?
Medically. Infant's eyelashes are very soft, and typically do not cause significant corneal damage.
Epiblepharon usually resolves spontaneously as the facial planes mature. If it has not resolved by the time coarse, mature lashes have developed—and the cornea is suffering—surgical intervention should be considered.

Redundant lid tissue causes lashes to abut ocular surface

Does epiblepharon usually involve the upper lid, or the lower? The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion? Typically it involves only the nasal portion

Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)? Asian (#1) and Native-American

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Epiblepharon

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The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion? Typically it involves only the nasal portion

Congenital tarsal kink

Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)? Asian (#1) and Native-American
**Congenital lid abnormalities: Matching**

**Epiblepharon.** In this child the lower lid lashes have turned in from birth, but the cornea has remained undamaged.
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching
- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
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- Lid coloboma
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Q

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- **In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?**
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

**In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?**

- The temporal portion of the lower lid is displaced inferiorly

Does euryblepharon require surgical intervention?

- It does if it results in exposure keratitis
Failure of lid differentiation

- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins

Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpapalpebral fissure

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
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- Euryblepharon
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- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?
The temporal portion

Redundant lid tissue causes lashes to abut ocular surface

Abnormal widening of interpapalpebral fissure

Does euryblepharon require surgical intervention?
Yes, if it results in exposure keratitis
Congenital lid abnormalities: Matching

Euryblepharon
**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis

**In euryblepharon**, which portion (ie, medial, central, temporal) of the fissure is widened?

The temporal portion

With regard to which portion of the lid is involved... Recall regarding which portion of the lid is involved in epiblepharon. What was the answer?

Redundant lid tissue causes lashes to abut ocular surface

Does **epiblepharon** usually involve the upper lid, or the lower?

The lower (epiblepharon rarely occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?

Typically it involves only the **nasal** portion

- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
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In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?
The temporal portion

Does euryblepharon usually involve the upper lid, or the lower?
The lower (euryblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
Typically it involves only the nasal portion

With regard to which portion of the lid is involved…Recall regarding which portion of the lid is involved in epiblepharon. What was the answer?
The nasal portion

So, take note: Euryblepharon and epiblepharon involve opposite ends of the lid. I point this out because, if you can remember which portion is involved for either condition, along with the fact that they’re the opposite of one another, you will know which portion is involved for both conditions.

Ankyloblepharon
Blepharophimosis

In congenital ptosis, which eye is affected?

Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma

Does epiblepharon usually involve the upper lid, or the lower?
The lower (epiblepharon rarely if ever occurs in the upper lid)

Does it typically involve the entirety of the lower lid, or only a portion?
Typically it involves only the nasal portion
**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Ankyloblepharon
- Blepharophimosis

In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?
The temporal portion

What is responsible for this widening, ie, is it the upper, or the lower lid that is displaced?

Redundant lid tissue causes lashes to abut ocular surface

Abnormal widening of interpaplebral fissure

Euryblepharon

Congenital entropion

Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis

Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened? The temporal portion
- What is responsible for this widening, ie, is it the upper, or the lower lid that is displaced? The temporal portion of the lower lid is displaced inferiorly

- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

Does euryblepharon require surgical intervention? It does if it results in exposure keratitis
In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?
The temporal portion

What is responsible for this widening, ie, is it the upper, or the lower lid that is displaced?
The temporal portion of the lower lid is displaced inferiorly

Does euryblepharon require surgical intervention?

Abnormal widening of interpalpebral fissure

Euryblepharon

Congenital tarsal kink

Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis

Ankyloblepharon

Blepharophimosis

Congenital lid abnormalities: Matching

Redundant lid tissue causes lashes to abut ocular surface
Failure of lid differentiation

- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins

Type of congenital entropion

- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- In euryblepharon, which portion (ie, medial, central, temporal) of the fissure is widened?
  - The temporal portion
- What is responsible for this widening, ie, is it the upper, or the lower lid that is displaced?
  - The temporal portion of the lower lid is displaced inferiorly
- Does euryblepharon require surgical intervention?
  - It does if it results in exposure keratitis

- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
Abnormal widening of interpalpebral fissure

What is dystopia canthorum?

Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis
**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut the ocular surface
- Abnormal widening of the interpalpebral fissure
- What is dystopia canthorum?
  Lateral displacement of the canthi (ie, telecanthus) **PLUS** laterally displaced lacrimal puncta

---

**What is dystopia canthorum?**
Lateral displacement of the canthi (ie, telecanthus) **PLUS** laterally displaced lacrimal puncta

---

**Failure of lid differentiation**

**Associated with Goldenhar**

**Fusion of all or part of the upper/lower margins**

**Type of congenital entropion**

**Redundant lid tissue causes lashes to abut the ocular surface**

**Abnormal widening of the interpalpebral fissure**

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**Dystopia canthorum**

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**Epicanthus (epicanthal folds)**

**Lid coloboma**

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**Congenital ptosis**

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Congenital lid abnormalities: Matching

Dystopia canthorum. Note the telecanthus, and laterally displaced lacrimal puncta
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
Abnormal widening of interpalpebral fissure

What is dystopia canthorum?
Lateral displacement of the canthi (ie, telecanthus) PLUS laterally displaced lacrimal puncta

How on earth are you supposed to recognize that the puncta are ‘too lateral’?

Q
Ankyloblepharon

Blepharophimosis

Cryptophthalmos

Congenital ectropion

Epiblepharon

Dystopia canthorum

Congenital entropion

Euryblepharon

Congenital tarsal kink

Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis

Failure of lid differentiation

Associated with Goldenhar

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

Abnormal widening of interpalpebral fissure

What is dystopia canthorum?
Lateral displacement of the canthi (ie, telecanthus) PLUS laterally displaced lacrimal puncta

How on earth are you supposed to recognize that the puncta are ‘too lateral’?
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced. (Next time you examine a pt at the slit-lamp, take note of whether such a line crosses their cornea [it won’t].)
**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure

**What is dystopia canthorum?**
Lateral displacement of the canthi (i.e., telecanthus) PLUS laterally displaced lacrimal puncta

**How on earth are you supposed to recognize that the puncta are ‘too lateral’?**
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced. (Next time you examine a pt at the slit-lamp, take note of this.)

**What is the difference between telecanthus and hypertelorism?**
- Telecanthus refers to an abnormally increased distance between the medial canthi.
- Hypertelorism refers to an abnormally increased distance between the medial orbital walls which manifests as an increased interpupillary distance.
**Q/A**

**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure

What is dystopia canthorum?
Lateral displacement of the canthi (i.e., telecanthus) PLUS laterally displaced lacrimal puncta

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**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls.

**What is dystopia canthorum?**
Lateral displacement of the canthi (i.e., telecanthus) PLUS laterally displaced lacrimal puncta.

**How on earth are you supposed to recognize that the puncta are ‘too lateral’?**
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced.

**What is the difference between telecanthus and hypertelorism?**
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Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
Abnormal widening of interpalpebral fissure

What is dystopia canthorum?
Lateral displacement of the canthi (i.e., telecanthus) PLUS laterally displaced lacrimal puncta

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**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls.

A

**Congenital lid abnormalities: Matching**

- Dystopia canthorum
- Epiblepharon
- Congenital ectropion
- Lid coloboma
- Congenital ptosis
- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure
**Q**

**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpupillary fissure

**Dystopia canthorum**

What is dystopia canthorum? Lateral displacement of the canthi (i.e., **telecanthus**) PLUS laterally displaced lacrimal puncta.

How on earth are you supposed to recognize that the puncta are ‘too lateral’?
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced. (Next time you examine a pt at the slit-lamp, take note of this.)

What is the difference between telecanthus and hypertelorism? **Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls.

Which manifests as an increased interpupillary distance?
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure

**What is dystopia canthorum?**
Lateral displacement of the canthi (ie, telecanthus) PLUS laterally displaced lacrimal puncta

**How on earth are you supposed to recognize that the puncta are ‘too lateral’?**
Draw an imaginary vertical line from the upper to the lower puncta. If this line crosses the cornea, the puncta are displaced. (Next time you examine a pt at the slit-lamp, take note of this.)

**What is the difference between telecanthus and hypertelorism?**
**Telecanthus** refers to an abnormally increased distance between the medial canthi, whereas **hypertelorism** refers to an abnormally increased distance between the medial orbital walls.

**Which manifests as an increased interpupillary distance?**
**Hypertelorism**
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Failure of lid differentiation
Associated with Goldenhar

Say... Why does this person with dystopia canthorum also have heterochromia iridis?

Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching

Say... Why does this person with dystopia canthorum also have heterochromia iridis?

Dystopia canthorum

What non-ophthalmic finding is classic for Waardenburg syndrome?
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

Say... Why does this person with dystopia canthorum also have heterochromia iridis?

Dystopia canthorum

What non-ophthalmic finding is classic for Waardenburg syndrome?
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Q/A

- Failure of lid differentiation
- Associated with Goldenhar

**Say... Why does this person with dystopia canthorum also have heterochromia iridis?**

Because dystopia canthorum and heterochromia iridis, along with synophrys, are the three classic ophthalmic hallmarks of Waardenburg syndrome.

What non-ophthalmic finding is classic for Waardenburg syndrome?
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

- Abnormal widening of interpalpebral fissure
Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

Failure of lid differentiation
- Associated with Goldenhar

Say... Why does this person with dystopia canthorum also have heterochromia iridis? Because dystopia canthorum and heterochromia iridis, along with synophrys, are the three classic ophthalmic hallmarks of Waardenberg syndrome.

Abnormal widening of interpalpebral fissure

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Congenital lid abnormalities: Matching

Waardenburg syndrome: Heterochromia iridis, dystopia canthorum, and mild synophrys

(What the heck is synophrys?)
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Failure of lid differentiation
Associated with Goldenhar
Lid coloboma

Say... Why does this person with dystopia canthorum also have heterochromia iridis?
Because dystopia canthorum and heterochromia iridis, along with synophrys, are the three classic ophthalmic hallmarks of Waardenberg syndrome.

What non-ophthalmic finding is classic for Waardenberg syndrome?
The presence of a white forelock (ie, an isolated streak of white hair in the forehead region).

What is synophrys?
The formal medical term for a unibrow.
Failure of lid differentiation
Associated with Goldenhar

Say... Why does this person with dystopia canthorum also have heterochromia iridis?
Because dystopia canthorum is associated with heterochromia iridis along with synophrys and synophrys are hallmarks of Waardenburg syndrome

What the heck is synophrys?
The formal medical term for a unibrow

Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Waardenburg syndrome: Synophrys
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

Failure of lid differentiation
Associated with Goldenhar

Say…Why does this person with dystopia canthorum also have heterochromia iridis?
Because dystopia canthorum and heterochromia iridis, along with synophrys, are the three classic ophthalmic hallmarks of Waardenburg syndrome

What non-ophthalmic cosmetic finding is classic for Waardenburg syndrome?

Abnormal widening of interpalpebral fissure

Q
Congenital lid abnormalities: Matching

Dystopia canthorum
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

- Failure of lid differentiation
- Associated with Goldenhar

Say... Why does this person with dystopia canthorum also have heterochromia iridis? Because dystopia canthorum and heterochromia iridis, along with synophrys, are the three classic ophthalmic hallmarks of *Waardenberg* syndrome.

What non-ophthalmic cosmetic finding is classic for *Waardenburg* syndrome? The presence of a white forelock (ie, an isolated streak of white hair in the forehead region)

- Abnormal widening of interpalpebral fissure

**Dystopia canthorum**
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Waardenburg syndrome: White forelock
Congenital lid abnormalities: Matching

Note that Waardenburg syndrome has forms that do not involve heterochromia.
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

What does the term *epicanthus* refer to in this context?

*Epicanthus (epicanthal folds)*
**Congenital lid abnormalities: Matching**

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

**What does the term epicanthus refer to in this context?**

An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- *Epicanthus (epicanthal folds)*
- Lid coloboma
- Congenital ptosis
What are the four types of epicanthus?

--Epicanthus
--Epicanthus
--Epicanthus
--Epicanthus

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

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An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus--more on this shortly.

Epiblpharon

Dystopia canthorum

Congenital entropion

Euryblepharon

Congenital tarsal kink

Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis
What are the four types of epicanthus?
--Epicanthus t (start here)
--Epicanthus i
--Epicanthus p
--Epicanthus s

Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

What does the term epicanthus refer to in this context?
An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.

Epicanthus (epicanthal folds)

Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Lid coloboma
Congenital ptosis
### Q/A

**What are the four types of epicanthus?**

- **Epicanthus *tarsalis***
- **Epicanthus *i*** (next)
- **Epicanthus *p***
- **Epicanthus *s***

### Congenital lid abnormalities: Matching

- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

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An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.

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**Epicanthus (epicanthal folds)**
What are the four types of epicanthus?

-- Epicanthus *tarsalis*
-- Epicanthus *inversus*
-- Epicanthus *p* (next)
-- Epicanthus *s*

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

What does the term epicanthus refer to in this context?

An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.
What are the four types of epicanthus?

-- Epicanthus *tarsalis*
-- Epicanthus *inversus*
-- Epicanthus *palpebralis*
-- Epicanthus *s* (next)

- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

What does the term epicanthus refer to in this context? An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus--more on this shortly.

- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- **Epicanthus (epicanthal folds)**
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

What are the four types of epicanthus?
--Epicanthus tarsalis
--Epicanthus inversus
--Epicanthus palpebralis
--Epicanthus supraciliaris

What does the term epicanthus refer to in this context?
An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus--more on this shortly.

What are the four types of epicanthus?
--Epicanthus tarsalis
--Epicanthus inversus
--Epicanthus palpebralis
--Epicanthus supraciliaris

Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink

Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis
## What are the four types of epicanthus? What’s involved for each?

- **Epicanthus tarsalis**: Primarily upper lid
- **Epicanthus inversus**: Primarily lower lid
- **Epicanthus palpebralis**: Upper and lower equally
- **Epicanthus supraciliaris**: Brow to lower lid

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### What does the term epicanthus refer to in this context?

An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.

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### Congenital lid abnormalities: Matching

- **Epicanthus (epicanthal folds)**
- **Epiblepharon**
- **Dystopia canthorum**
- **Congenital entropion**
- **Euryblepharon**
- **Congenital tarsal kink**
- **Lid coloboma**
- **Congenital ptosis**

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- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

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What are the four types of epicanthus? What’s involved for each?
--Epicanthus *tarsalis*: Primarily upper lid
--Epicanthus *inversus*
--Epicanthus *palpebralis*
--Epicanthus *supraciliaris*

Fusion of all or part of the upper/lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface

What does the term epicanthus refer to in this context? An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus--more on this shortly.

**Epicanthus (epicanthal folds)**
Congenital lid abnormalities: Matching

Epicanthus tarsalis
What are the four types of epicanthus? What’s involved for each?

--Epicanthus *tarsalis*: Primarily upper lid

--Epicanthus *inversus*: (next)

--Epicanthus *palpebralis*

--Epicanthus *supraciliaris*

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

What does the term epicanthus refer to in this context? An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.
**Q/A**

**Congenital lid abnormalities: Matching**

**What are the four types of epicanthus? What’s involved for each?**
- Epicanthus *tarsalis*: Primarily upper lid
- Epicanthus *inversus*: Primarily lower lid
- Epicanthus *palpebralis*
- Epicanthus *supraciliaris*

**Fusion of all or part of the upper/lower margins**

**Type of congenital entropion**

**Redundant lid tissue causes lashes to abut ocular surface**

**What does the term epicanthus refer to in this context?**

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**Epiblepharon**

**Dystopia canthorum**

**Congenital entropion**

**Euryblepharon**

**Congenital tarsal kink**

**Epicanthus (epicanthal folds)**

**Lid coloboma**

**Congenital ptosis**
Congenital lid abnormalities: Matching

Epicanthus inversus
What are the four types of epicanthus? What’s involved for each?

- Epicanthus *tarsalis*: Primarily upper lid
- Epicanthus *inversus*: Primarily lower lid
- Epicanthus *palpebralis*: (next)
- Epicanthus *supraciliaris*

What does the term epicanthus refer to in this context? An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.
Q/A

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- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface

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- Epiblepharon
- Dystopia canthorum
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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

Epicanthus palpebralis
What are the four types of epicanthus? What’s involved for each?

- **Epicanthus tarsalis**: Primarily upper lid
- **Epicanthus inversus**: Primarily lower lid
- **Epicanthus palpebralis**: Upper and lower lids equally
- **Epicanthus supraciliaris**: (next)

What does the term epicanthus refer to in this context?

An *epicanthus* is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.

Fusion of all or part of the upper/lower margins

Type of congenital entropion

Redundant lid tissue causes lashes to abut ocular surface

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Congenital lid abnormalities: Matching

Epicanthus supraciliaris
Epicanthal folds overview
Congenital lid abnormalities: Matching

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- Is considered a normal variant when found in a child of East Asian descent?
- Can produce pseudostrabismus?
- Is associated with ptosis?
- Resolves without surgical intervention?

Epicanthus (epicanthal folds)

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- Can produce pseudostrabismus? All of them
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What is pseudostrabismus?
A false impression (on the part of an examiner) regarding the presence of strabismus.

Are pts with prominent epicanthal folds more likely to be (mis)diagnosed with esotropia, or exotropia?
Esotropia

What is it about the appearance of these pts that might lead one to incorrectly conclude the pt is ET?
The epicanthal folds cover some of the normally-visible sclera, thereby giving the impression the eyes are turned in.

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- **Epiblepharon**
- **Dystopia canthorum**
- **Congenital entropion**
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- **Congenital tarsal kink**
- **Epicanthus (epicanthal folds)**
- **Lid coloboma**
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Fusion of all or part of the upper/lower margins

Type of congenital entropion

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### Congenital lid abnormalities: Matching

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Congenital lid abnormalities: Matching

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Fusion of all or part of the upper and lower margins
Type of congenital entropion
Redundant lid tissue causes lashes to abut ocular surface
Abnormal widening of interpalpebral fissure

Why the hedging about whether epicanthus inversus can resolve without surgery?
--- Per the Peds book: “Because the epicanthus [inversus…] may improve with age, repair of the defects is often delayed.”
--- Per the Orbit book: “Most forms of epicanthus resolve with normal growth…Epicanthus inversus, however, rarely respond to facial growth.”

So caveat emptor, peeps.

What does the term epicanthus refer to in this context?
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**What are the four types of epicanthus?**

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---

**Why the hedging about whether epicanthus inversus can resolve without surgery?**

Because the BCSC books are somewhat in conflict on the subject (at least, the most recent editions in my possession are).

---

**An epicanthus is a fold of skin extending above and below the medial canthal region. How far the fold extends in each direction is a function of the type of epicanthus—more on this shortly.**

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**What does the term epicanthus refer to in this context?**

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**Why might epicanthus inversus be a reason for pseudostrabismus?**

Because the epicanthus inversus may improve with age, repair of the defects is often delayed.

---

**Can epicanthus inversus be associated with ptosis?**

Most forms of epicanthus resolve with normal growth...Epicanthus inversus, however, rarely respond to facial growth.

---

So caveat emptor, peeps.
Speaking of epicanthus inversus… It is exclusively syndromic. With which syndrome is it most closely associated?

- Blepharophimosis syndrome

What are the three other defining ophthalmic features of blepharophimosis syndrome?

- Epicanthus inversus
- Telecanthus
- Ptosis

and, of course,

- Blepharophimosis

Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
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Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
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What does it mean to say a pt has blepharophimosis?

It means her/his palpebral fissures are abnormally short in both vertical and horizontal extent.
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Blepharophimosis syndrome has two other names. What are they?
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Blepharophimosis syndrome has two other names. What are they? -- Congenital eyelid syndrome -- Blepharophimosis-ptosis-epicanthus inversus syndrome (BPES)

- Ankyloblepharon
- Cryptophthalmos
- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut corneal surface
- Abnormal widening of interpalpebral fissure
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
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Speaking of epicanthus inversus... It is exclusively syndromic. With which syndrome is it most closely associated?
Blepharophimosis syndrome

What are the three other defining ophthalmic features of blepharophimosis syndrome?
-- Epicanthus inversus
-- Telecanthus
-- Ptosis

And, of course...

Is the ptosis purely structural (i.e., secondary to blepharophimosis), or is there a problem with the levator?

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Epiblepharon
- Congenital ectropion
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
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Is the ptosis purely structural (ie, 2ndry to blepharophimosis), or is there a problem with the levator?

In fact levator function is usually very poor

Congenital lid abnormalities: Matching

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Are there any ophthalmic manifestations beyond these?
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
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Q/A

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Are there any ophthalmic manifestations beyond these?
Two of note:
- Hypertelorism
- Ectropion
Congenital lid abnormalities: Matching

- Failure of lid differentiation
  - Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
  - Redundant lid tissue causes lashes to abut ocular surface
  - Abnormal widening of interpupillary fissure
  - Poor levator function

Ankyloblepharon
Blepharophimosis
Cryptophthalmos

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Two of note: **Hypertelorism** and **ectropion**

- Congenital tarsal kink
- Epicanthus (epicanthal folds)
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**Type of congenital entropion**

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**Congenital lid abnormalities: Matching**

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- Telecanthus
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and, of course,

- Blepharophimosis syndrome

- Hypertelorism
- Ectropion

What are the two main non-ophthalmic features of blepharophimosis syndrome?
Congenital lid abnormalities: Matching

- Failure of lid differentiation
- Associated with Goldenhar
- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
- Redundant lid tissue causes lashes to abut ocular surface
- Abnormal widening of interpalpebral fissure
- Poor levator function
- Hypertelorism
- Ectropion
- Widening of interpupillary fissure
- Ankyloblepharon
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- Telecanthus
- Ptosis

and, of course,

- Blepharophimosis

What are the two main non-ophthalmic features of blepharophimosis syndrome?

- Hypoplastic ears
- ‘Lop’ ears

Hypoplastic nasal bridge

Ectropion

Congenital tarsal kink

Epicanthal (epicanthal folds)

Lid coloboma

Congenital ptosis
Congenital lid abnormalities: Matching

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A

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- Hypertelorism
- Ectropion
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- Lid coloboma
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- Epicanthus (epicanthal folds)
Congenital lid abnormalities: Matching

Hypoplastic nasal bridge

Lop ears

Blepharophimosis syndrome