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

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

Congenital lid abnormalities: Matching

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

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? 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

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

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

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

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis

Failure of lid differentiation

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?
Of sorts, but it is virtually always profoundly malformed

- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
**Failure of lid differentiation** ➔ **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?*
Of sorts, but it is virtually always profoundly malformed

*Can relatively normal lids be surgically constructed?*
Failure of lid differentiation

Cryptophthalmos

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

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Of sorts, but it is virtually always profoundly malformed

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Failure of lid differentiation
Associated with Goldenhar

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Failure of lid differentiation
Associated with Goldenhar

Congenital lid abnormalities: Matching

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- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

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

Lid coloboma
A

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion

- Failure of lid differentiation
- **Associated with Goldenhar**

---

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

Lower

---

- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

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

Colobomas of the upper lid—is that a thing?
Failure of lid differentiation

Associated with Goldenhar

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

Lower

Colobomas of the upper lid—is that a thing?
Yes
Failure of lid differentiation

**Associated with Goldenhar**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion

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

- Lower

*Colobomas of the upper lid—is that a thing?*

- Yes

*There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?*

- Epicanthus (epicanthal fold)

- **Lid coloboma**

- Congenital ptosis
A

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion

- Failure of lid differentiation
- **Associated with Goldenhar**

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

Colobomas of the upper lid—is that a thing?
Yes

There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?
Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar)

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

Associated with Goldenhar

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion

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?

Lower

Colobomas of the upper lid—is that a thing?

Yes

There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?

Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar)

- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

Congenital lid abnormalities: Matching

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

\textit{What other ocular structures are commonly colobomatous?}

- The iris
- The choroid
- The optic nerve head

\textit{Lid colobomas associated with Goldenhar—are they found in the upper, or the lower lid?}

Lower

\textit{Colobomas of the upper lid—is that a thing?}

Yes

\textit{There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?}

Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar)
Lid colobomas associated with Goldenhar—are they found in the upper, or the lower lid?
Lower

Colobomas of the upper lid—is that a thing?
Yes

There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?
Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar).
A

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- 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?**

Failure of the embryonic fissure of the optic vesicle to close

---

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

Lower

---

**Colobomas of the upper lid—is that a thing?**

Yes

---

**There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?**

Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar).
Congenital ptosis

Lid coloboma

Goldenhar syndrome (Frontonasal defect)

Failure of lid differentiation

Associated with Goldenhar

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

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

Lower colobomas of the upper lid—are they found in the upper, or the lower lid?

Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar).

There is a general rule of thumb that characterizes upper-vs lower-lid colobomas—what is it?

Yes

Colobomas of the upper lid—"is that a thing?"

Lower

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

Are lid colobomas associated with these others?

- The iris
- The choroid
- The optic nerve head

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

Failure of the embryonic fissure of the optic vesicle to close

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

Failure of the embryonic fissure of the optic vesicle to close
Failure of lid differentiation
Associated with Goldenhar

Congenital lid abnormalities: Matching

- Ankyloblepharon
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- 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?
Failure of the embryonic fissure of the optic vesicle to close

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

Colobomas of the upper lid—is that a thing?
Yes

There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it?
Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar)
What other ocular structures are commonly colobomatous?
- The iris
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- The optic nerve head

Are lid colobomas associated with these others?
No

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

Colobomas of the upper lid—is that a thing?
Yes

There is a general rule of thumb that characterizes upper- vs lower-lid colobomas—what is it? Colobomas of the upper lid typically are isolated findings, whereas colobomas of the lower lid are usually associated with other craniofacial abnormalities or syndromes (most commonly Goldenhar)

Why are lid colobomas not associated with the other types?
Because the eyelids do not derive embryologically from the optic vesicle
Failure of lid differentiation
Associated with Goldenhar

Congenital lid abnormalities: Matching

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

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- The iris
- The choroid
- The optic nerve head

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Congenital lid abnormalities: Matching

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

In addition to lid colobomas, what other adnexal/surface finding is common?
 Limbal dermoids

What strabismus syndrome is associated with Goldenhar?
D uane syndrome

What nonocular findings are usually present?
- Ear abnormalities (pre-auricular appendages; aural fistulae)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

Are Goldenhar pts cognitively impaired?
Retardation is present in a minority (~10%)
Failure of lid differentiation

Associated with Goldenhar

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

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

Mnemonic: Goldenhar

Answer starts with ‘O’

Goldenhar

OAV syndrome

Goldenhar

LDENH

emifacial microsomia

AR

retardation is present in a minority (~10%)
Failure of lid differentiation

Associated with

Goldenhar

Mnemonic: Goldenhar

OAV syndrome

Goldenhar

In addition to lid colobomas, what other adnexal/surface finding is common?

Lid coloboma

What is the nonpneymous name of Goldenhar syndrome?

Oculo-Auriculo-Vertebral (OAV) syndrome

In addition to lid colobomas, what other adnexal/surface finding is common?

Lid coloboma

What nonocular findings are usually present?

Ear abnormalities (pre-auricular appendages; aural fistulae)

Hemifacial microsomia (maxillary/mandibular hypoplasia)

Are Goldenhar pts cognitively impaired?

Retardation is present in a minority (~10%)

Ankyloblepharon

Blepharophimosis

Cryptophthalmos

Congenital ectropion

Epiblepharon

Dystopia canthorum

Congenital entropion

Euryblepharon

Congenital tarsal kink

Epicanthus (epicanthal folds)

Lid coloboma

Congenital ptosis
**Goldenhar Syndrome**

- Failure of lid differentiation
- Associated with Goldenhar

### 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

**What is the non-eponymous name of Goldenhar syndrome?**

Oculo-Auriculo-Vertebral (OAV) syndrome

**In addition to lid colobomas, what other adnexal/surface finding is common?**

Limbal dermoids

**Mnemonic: Goldenhar**

- **Goldenhar**
- OAV syndrome
- Limbal dermoids
- Denials
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

**Are Goldenhar pts cognitively impaired?**

Retardation is present in a minority (~10%)
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

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

In addition to lid colobomas, what other adnexal/surface finding is common?
Limbal dermoids

What strabismus syndrome is associated with Goldenhar?
Duane syndrome

Nonocular findings associated with Goldenhar:
- Ear abnormalities (pre-auricular appendages; aural fistulae)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)
- Retardation is present in a minority (~10%)
<table>
<thead>
<tr>
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<th>Goldenhar</th>
</tr>
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<tbody>
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<td></td>
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<td>Duane syndrome</td>
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Mnemonic: Goldenhar

| Goldenhar | OAV syndrome | Limbal dermoids | Duane syndrome | En | Hemifacial microsomia | Maxillary/mandibular hypoplasia | Retardation is present in a minority (~10%) |
**Congenital lid abnormalities: Matching**

- 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
- Congenital ptosis

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

In addition to lid colobomas, what other adnexal/surface finding is common?
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What strabismus syndrome is associated with Goldenhar?
**Duane syndrome**

What nonocular findings are usually present?
--

**Mnemonic: Goldenhar**

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

- Congenital ptosis
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What is the nonpneymous name of Goldenhar syndrome?
- Oculo-Auriculo-Vertebral (OAV) syndrome

In addition to lid colobomas, 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)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

Mnemonic: Goldenhar
- OAV syndrome
- Limbal dermoids
- Duane syndrome
- Ear abnormalities
- Hemifacial microsomia
- Narasimhan syndrome
- Congenital ptosis
What is the noneponymous name of Goldenhar syndrome?
Oculo-Auriculo-Vertebral (OAV) syndrome

In addition to lid colobomas, what other adnexal/surface finding is common?
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What strabismus syndrome is associated with Goldenhar?
Duane syndrome

What nonocular findings are usually present?
--Ear abnormalities (pre-auricular appendages; aural fistulae)
--Hemifacial microsomia (maxillary/mandibular hypoplasia)

Are Goldenhar pts cognitively impaired?

Mnemonic: Goldenhar

Goldenhar
OAV syndrome
Limbal dermoids
Duane syndrome
Ear abnormalities

Hemifacial microsomia

AR

Congenital ptosis
Failure of lid differentiation

Associated with Goldenhar

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

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Are Goldenhar pts cognitively impaired?
Retardation is present in a minority (~10%)
Failure of lid differentiation
Associated with Goldenhar

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

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Are Goldenhar pts cognitively impaired?
Retardation is present in a minority (~10%)
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins

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

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

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

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.
Q/A

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
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- Congenital tarsal kink
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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.
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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

- 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**

*Not this one, duh*
A

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

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

- 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

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

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- **Epiblepharon**
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Concanthal canthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
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? (Epiblepharon rarely if ever occurs in the upper lid)

Typically it involves only the nasal portion
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)
Congenital lid abnormalities: Matching

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

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

Temporal v medial v nasal
Failure of lid differentiation
Associated with Goldenhar
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Congenital lid abnormalities: Matching

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

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

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- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- **Epiblepharon**
- Epicanthus (epicanthal folds)
- Epiblepharon
- Dystopia canthorum
- Congenital tarsal kink
- Lid coloboma
- Congenital ptosis
A

Congenital lid abnormalities: Matching

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

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

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

- Fusion of all or part of the upper/lower margins
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Epiblepharon

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

- Entropion
- 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.
Failure of lid differentiation
Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
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In a few words, define…
Entropion:
Trichiasis:

Fusion of all or part of the upper/lower margins

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Entropion:
Trichiasis:

Congenital tarsal kink
Congenital ptosis

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Congenital ptosis

Epicanthus (epicanthal folds)
Lid coloboma
Congenital tarsal kink
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
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Fusion of all or part of the upper/lower margins
Type of congenital entropion
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- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
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- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Failure of lid differentiation

- Associated with Goldenhar
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Congenital lid abnormalities: Matching
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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.

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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Children of what two ethnic heritages are most likely to present with epiblepharon (and which is #1)?
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You evaluate an infant with epiblepharon and extensive lash-cornea touch. Should this be managed surgically or medically, and why?

Epiblepharon

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In a few words, define...
Entropion: 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?
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

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

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

Entropion: An inward rotation of the eyelid margin
Trichiasis: An inward rotation of eyelashes
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

- Ankyloblepharon
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Congenital lid abnormalities: Matching

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Abnormal widening of interpalpebral fissure

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- Blepharophimosis

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

- Euryblepharon

Redundant lid tissue causes lashes to abut ocular surface

Abnormal widening of interpalpebral fissure

- Congenital entropion
- 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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Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Ankyloblepharon
- Euryblepharon
- Congenital entropion
- 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

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

Does euryblepharon require surgical intervention?
It does if it results in exposure keratitis
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

- Ankyloblepharon
- Blepharophimosis
- **Euryblepharon**
- **Epiblepharon**
- **Dystopia canthorum**
- Epicanthus (epicanthal folds)
- **Congenital ptosis**
Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- 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

In **eurylepharon**, 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 this question 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.

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
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 interpalpebral fissure.

Euryblepharon.

Ankyloblepharon.

Blepharophimosis.

Congenital entropion.

Epiblepharon.

Dystopia canthorum.

Congenital epicanthus.

Lid coloboma.

Congenital ptosis.

Congenital tarsal kink.
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

Abnormal widening of interpalpebral fissure

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis
- Ankyloblepharon
- Blepharophimosis

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

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
- 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?

- 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

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

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
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- Congenital ptosis

What is dystopia canthorum?

+ 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].)
A

**Congenital lid abnormalities: Matching**

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

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

- Congenital ectropion
- Epiblepharon
- Dystopia canthorum
- Lid coloboma
- Congenital ptosis
Ankyloblepharon
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Cryptophthalmos
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Q

Congenital lid abnormalities: Matching

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Associated with Goldenhar
Fusion of all or part of the upper/lower margins
Type of congenital entropion
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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’?
Ankyloblepharon
Blepharophimosis
Cryptophthalmos
Congenital ectropion
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Epicanthus (epicanthal folds)
Lid coloboma
Congenital ptosis

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- Blepharophimosis
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- Epiblepharon
- Dystopia canthorum
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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
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- 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...

**What is the difference between telecanthus and hypertelorism?**
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
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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.

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

What is dystopia canthorum?
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

**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...)

*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
- 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

Blepharophimosis

Cryptophthalmos

Congenital ectropion

Epiblepharon

Dystopia canthorum

- Congenital entropion
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis

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...)

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

Abnormal widening of interpalpebral fissure

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

Dystopia canthorum

Congenital lid abnormalities: Matching

Due to the abnormal widening of the interpalpebral fissure, the eyes may appear convergent, leading to heterochromia iridis. Additionally, the lateral displacement of the lids can cause epicanthus (epicanthal folds), which further contributes to the appearance of heterochromia iridis.

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 (i.e., an isolated streak of white hair in the forehead region).
Failure of lid differentiation

Associated with Goldenhar

Failure of synthesis

Abnormal widening of interpalpebral fissure

Conjugate lid abnormalities: Matching

- 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) eponym
A

**Congenital lid abnormalities: Matching**

- 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

- Congenital ectropion
- Epiblepharon
- **Dystopia canthorum**
- Congenital entropion
- Euryblepharon
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
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, heterochromia iridis, and synophrys are all associated with Waardenberg syndrome.

What is synophrys?
Abnormal widening of interpalpebral fissure

Synophrys
Dystopia canthorum

Congenital lid abnormalities: Matching

White forelock (isolated streak of white hair in the forehead region)

What non-ophthalmic finding is classic for Waardenberg syndrome?
**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 ocular surface
- Abnormal widening of interpalpebral fissure

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

Because dystopia canthorum and heterochromia iridis along with synophrys often are diagnostic features of Waardenberg syndrome.

**Synophrys**
- The formal medical term for a unibrow

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

What is synophrys?
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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?
- 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 (i.e., an isolated streak of white hair in the forehead region)

- Abnormal widening of interpalpebral fissure

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

- Ankyloblepharon
- Blepharophimosis
- Cryptophthalmos
- Congenital entropion
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
- Dystopia canthorum
- Congenital lid abnormalities: Matching
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?

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

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 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
--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

Epiblepharon
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**: mnemonic forthcoming… **TIPS**
- **Epicanthus p**
- **Epicanthus s**

### 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.

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

**TIPS**

- **T**: Primarily upper lid
- **I**: Primarily lower lid
- **P**: Upper and lower equally
- **S**: From brow to lower lid

**Mnemonic forthcoming**…
What are the four types of epicanthus?
--Epicanthus *tarsalis*
--Epicanthus *i* (next)
--Epicanthus *p*
--Epicanthus *s*

- Fusion of all or part of the upper/lower margins
- Type of congenital entropion
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Epicanthus *tarsalis*: Primarily upper lid
Epicanthus *i*: Primarily lower lid
Epicanthus *p*: Upper and lower equally
Epicanthus *s*: From brow to lower lid

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

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Epicanthus (epicanthal folds)
Epiblepharon
Dystopia canthorum
Congenital entropion
Euryblepharon
Congenital tarsal kink
Lid coloboma
Congenital ptosis
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
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Epiblepharon
Dystopia canthorum
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**Epicanthus (epicanthal folds)**
Lid coloboma
Congenital ptosis
What are the four types of epicanthus? What's involved for each?
--Epicanthus *tarsalis*: (start here)
--Epicanthus *inversus*
--Epicanthus *palpebralis*
--Epicanthus *supraciliaris*

- Fusion of all or part of the upper/lower margins
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- *Epicanthus (epicanthal folds)*
- Lid coloboma
- Congenital ptosis

**Q**

**Congenital lid abnormalities: Matching**
**Congenital lid abnormalities: Matching**

**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**

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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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**Epicanthus (epicanthal folds)**

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- Dystopia canthorum
- Congenital entropion
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What are the four types of epicanthus? What’s involved for each?

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- **Epicanthus inversus**: Primarily lower lid
- **Epicanthus palpebralis**: (next)
- **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.

Epiblepharon

Dystopia canthorum

Congenital entropion

Euryblepharon

Congenital tarsal kink

**Epicanthus (epicanthal folds)**

Lid coloboma

Congenital ptosis
Q/A

**Congenital lid abnormalities: Matching**

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

--Epicanthus *tarsalis*: Primarily upper lid
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- Is associated with ptosis?
- Resolves without surgical intervention?

**Epicanthus (epicanthal folds)**
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- Associated with Goldenhar
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A false impression (on the part of an examiner) re the presence of strabismus.

Is considered a normal variant when found in a child of East Asian descent?

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- All of them

Is associated with ptosis?

- All of them

Resolves without surgical intervention?

- All (with the possible exception of inversus)

What is pseudostrabismus?

A false impression (on the part of an examiner) re 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.

- Euryblepharon
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- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
What do the terms entropion, ectropion, and ptosis refer to in this context? Are these specific congenital conditions? If so, what are the clinical features?
Failure of lid differentiation
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Congenital lid abnormalities: Matching
- Ankyloblepharon
- Blepharophimosis
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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).

- 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.

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**Congenital lid abnormalities: Congenital ptosis**

- **Epicanthus (epicanthal folds)**
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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, 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
- Cryptophthalmos
- Anophthalmia
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What does it mean to say a pt has 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.

Congenital lid abnormalities: Matching

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Blepharophimosis syndrome has two other names. What are they?
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- Ankyloblepharon
- Blepharophimosis syndrome
- 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 ocular surface
- Abnormal widening of interpalpebral fissure

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

Blepharophimosis syndrome has two other names. What are they?

-- Congenital eyelid syndrome

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

1. Ankyloblepharon
2. Blepharophimosis syndrome
3. Cryptophthalmos
4. Blepharophimosis syndrome
5. Ankyloblepharon
6. Epicanthus (epicanthal folds)
7. Lid coloboma
8. Congenital ptosis

Q/A
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 syndrome

Blepharophimosis syndrome has two other names. What are they?

- Congenital eyelid syndrome
- Blepharophimosis-ptosis-epicanthus inversus syndrome (BPES)

Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis syndrome
- 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 ocular surface
- Abnormal widening of interpalpebral fissure
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis syndrome

- 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

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

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

And, of course, Blepharophimosis

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

Is the ptosis purely structural (i.e., secondary to blepharophimosis), or is there a problem with the levator? In fact, levator function is usually very poor.
Congenital lid abnormalities: Matching

- Ankyloblepharon
- Blepharophimosis syndrome
- Cryptophthalmos

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

Are there any ophthalmic manifestations beyond these?

- Hypertelorism
- Ectropion
- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Q/A

Congenital lid abnormalities: Matching

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

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

Are there any ophthalmic manifestations beyond these? Two of note:

Hypertelorism

Ectropion
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

A

Congenital lid abnormalities: Matching

- Ankyloblepharon syndrome
- Blepharophimosis syndrome

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

Are there any ophthalmic manifestations beyond these? Two of note: Hypertelorism and ectropion

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

**Congenital lid abnormalities: Matching**

- Ankyloblepharon syndrome
- Blepharophimosis syndrome

- 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

**Speaking of epicanthus inversus…** It is exclusively syndromic. With which syndrome is it most closely associated?

**A**

Blepharophimosis syndrome

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

- Epicanthus inversus
- Telecanthus
- Ptosis

And, of course, Blepharophimosis syndrome

- Hypertelorism
- Ectropion

- Congenital tarsal kink
- Epicanthus (epicanthal folds)
- Lid coloboma
- Congenital ptosis
Q/A

**Congenital lid abnormalities: Matching**

- Ankyloblepharon syndrome
- Blepharophimosis syndrome

**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

**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

**Hypertelorism**
- Ectropion
- Poor levator function

**Ectropion**
- Hypoplastic nasal bridge
- ‘Lop’ ears

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

**Congenital lid abnormalities: Matching**

- Ankyloblepharon
- **Blepharophimosis syndrome**

- 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 distance

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

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

-- Hypoplastic nasal bridge
-- 'Lop' ears

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