



What is the 'proper' term for eyelid ptosis?





What is the 'proper' term for eyelid ptosis?

Blepharoptosis





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Blepharoptosis

What is the formal definition of ptosis?





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What is the formal definition of ptosis? Inferodisplacement of the upper lid





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What is pseudoptosis?





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What is **pseudo**ptosis? Apparent inferodisplacement of the lid secondary to a non-lid condition





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What is **pseudo**ptosis? Apparent inferodisplacement of the lid secondary to a non-lid condition

The Peds book lists two causes of pseudoptosis that might mimic congenital ptosis—what are they?



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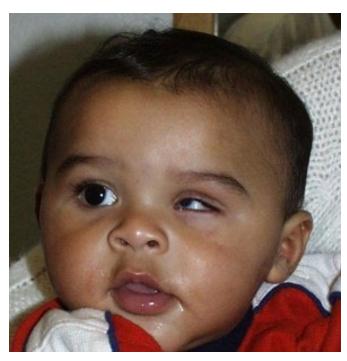
What is pseudoptosis?

Apparent inferodisplacement of the lid secondary to a non-lid condition

The Peds book lists two causes of pseudoptosis that might mimic congenital ptosis—what are they?

Microphthalmia and hypotropia. Make sure you're not missing one of these before locking in on a dx of congenital ptosis!





Microphthalmia



Hypotropia





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What are the two very general forms of ptosis?





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What are the two very general forms of ptosis? Congenital and acquired

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Lid-related observations in the ptosis patient	

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Lid-related observations in the ptosis patient
MRD1
MRD2
Vertical fissure height
Upper lid crease position
Levator function
Presence of lagophthalmos



What does MRD stand for in this context?

What are the two very general forms of ptosis?
Congenital and acquired

Ated observations in e ptosis patient

MRD1

MRD2

rtical fissure height

er lid crease position

Levator function

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What does MRD stand for in this context? Margin-reflex distance

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To what does the word reflex refer?

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What does MRD stand for in this context? Margin-reflex distance

To what does the word reflex refer? The corneal light reflex

What are these specific measurements?
MRD1 is...

MRD2 is...

What are the two very general forms of ptosis? Congenital and acquired

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MRD1

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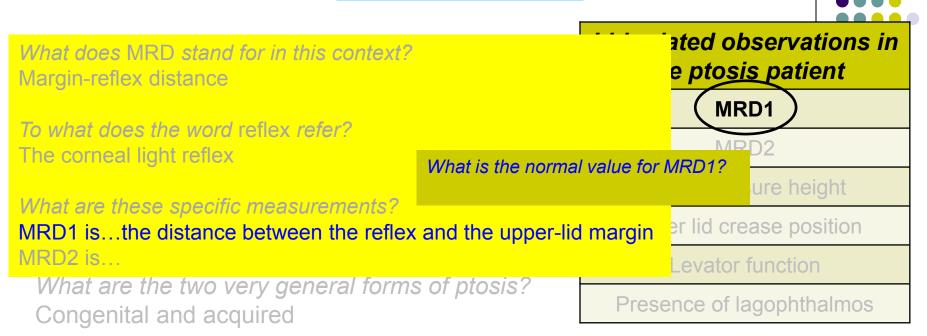
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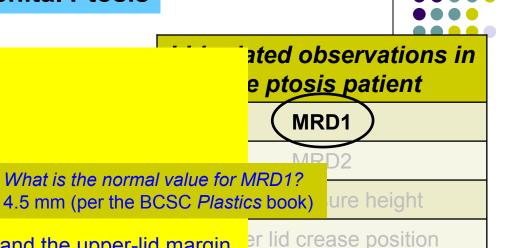
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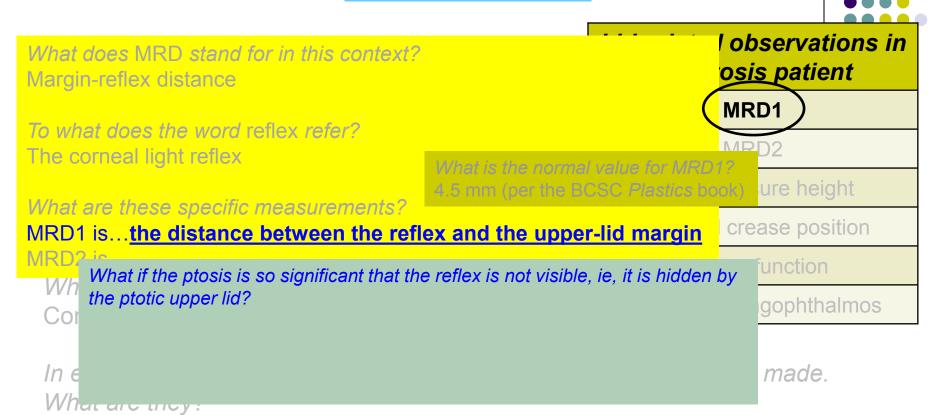
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crease position

What are these specific measurements?

MRD1 is...the distance between the reflex and the upper-lid margin

MRD24

What if the ptosis is so significant that the reflex is not visible, ie, it is hidden by the ptotic upper lid?

Then the MRD1 is defined as the distance the upper lid has to be **elevated** in order for the reflex to be seen, and is assigned a **negative** value. So, eg, if the upper-lid margin has to be elevated 2 mm in order for the reflex to be seen, MRD1 is -2 mm.

What are tricy:

function

osis patient

MRD1

MRD2

gophthalmos

made

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MRD1 is...the distance between the reflex and the upper-lid margin MRD2 is...the distance between the reflex and the **lower**-lid margin

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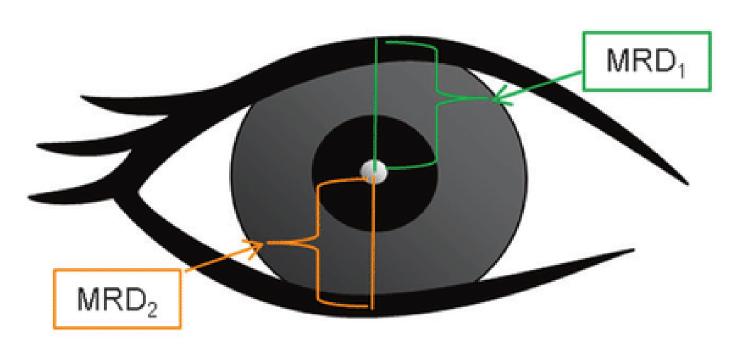
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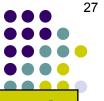
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How is vertical fissure height quantified?

What a Conger

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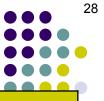
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What is the relationship between vertical fissure height and MRD1/MRD2? Assuming all have been measured correctly, vertical fissure height will be

In evaluequal to MRD1 + MRD2

What are they?

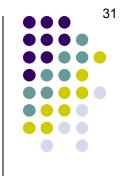
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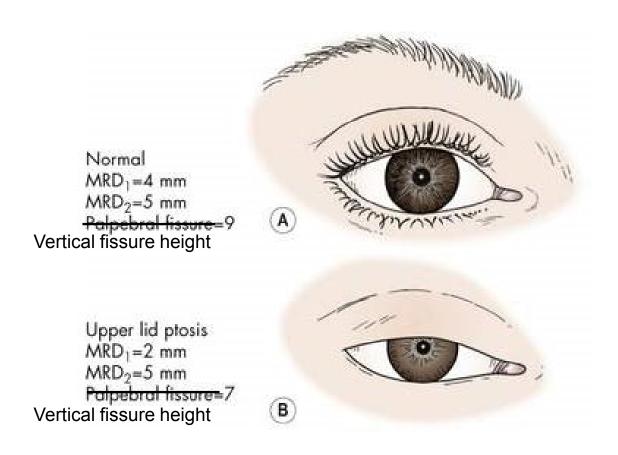
MRD1

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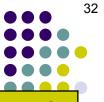
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MRD and vertical fissure height



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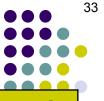
Upper lid crease pisition

Levator function

Congen

What ar Anatomically speaking, what structures create the upper lid crease?

In evalu What ar



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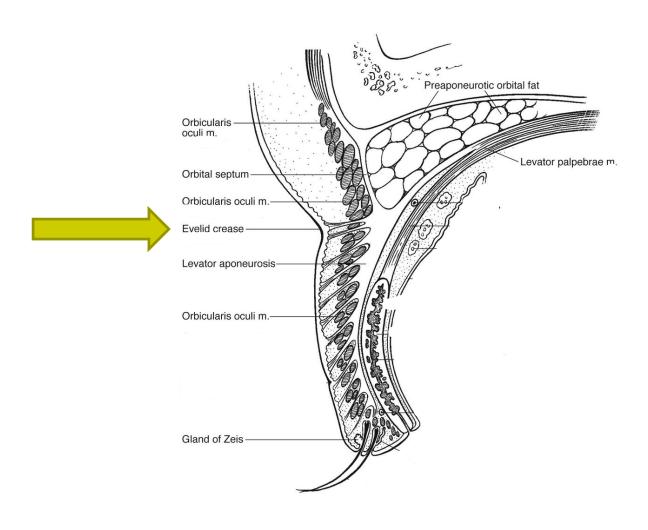
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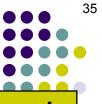
Congen As the aponeurosis of the levator muscle approaches the tarsal plate, it splits into anterior and posterior portions. The anterior portion consists of fine tendrils of In evalu aponeurotic material, some of which will attach to the skin overlying the superior What ar margin of the tarsus. These attachments draw the skin inward, thereby producing the eyelid **crease**. This indrawing of skin causes the skin, muscle and fat superior to the crease to 'overhang;' this overhang is called the eyelid **fold**.

> (In case you were wondering: The *posterior* portion of the aponeurosis inserts on the tarsal plate.)



Eyelid crease





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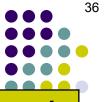
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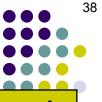




Lid crease measurement

What

Congenital Ptosis



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In eva. What are typical values for this measure?

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Well, the *Plastics* book only gives values for Caucasians...

Lid-related observations in the ptosis patient

MRD1

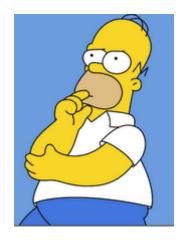
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Lid-related observations in

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those values are #### for males, for females.

the ptosis patient MRD1

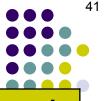
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What

In eva What are typical values for this measure? Well, the *Plastics* book only gives values for Caucasians... those values are 8-9 mm for males, 9-11 mm for females. The book goes on to say the crease "is typically lower or obscured in the Asian eyelid, with or without ptosis."

Lid-related observations in the ptosis patient

MRD1

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Vertical fissure height

Upper lid crease position

Levator function

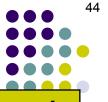
of lagophthalmos





Lid crease is often poorly defined or absent in congenital ptosis





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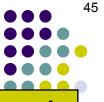
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In evalu when the pt is in down- and upgaze

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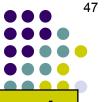
Levator function

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Levator function measurement



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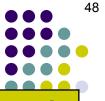
Levator function

Presence of lagophthalmos

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What important qualifier is placed on measuring levator function?





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What important qualifier is placed on measuring levator function?

The measurement must be performed in way that prevents the muscle from assisting lid elevation



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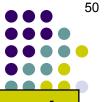
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What are the two Congenital and a

In evaluating a pt

What are they?

What is lagophthalmos?

Lid-related observations in the ptosis patient

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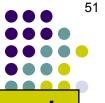
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What is lagophthalmos? Failure of the lids to close completely

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Lagophthalmos OD

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What are the two What is lagophthalmos?
Congenital and a Failure of the lids to close completely

In evaluating a pt What are they?

In evaluating a pt Hol up--why wouldn't a ptotic lid close completely?

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What is lagophthalmos? Failure of the lids to close completely

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In evaluating a pt Hol up--why wouldn't a ptotic lid close completely? We'll get to that shortly

Lid-related observations in the ptosis patient

MRD1

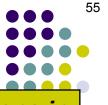
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Of these six, which measurement does the Plastics book single out as being the most useful/important?

What are the two very general forms of ptosis? Congenital and acquired

Lid-related observations in the ptosis patient MRD1? MRD2? Vertical fissure height? Upper lid crease position? Levator function? Presence of lagophthalmos?

In evaluating a ptosis pt, six observations/measurements should be made. What are they?

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MRD1

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Other things you gotta check
?
?
?
?
?
?



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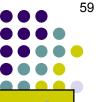
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In evaluating a ptosis pt, six non-lid observations/measurements should be made. What are they?

Other things you gotta check Pupils Corneal sensitivity Bell's phenomenon Visual acuity Refractive error EOMs



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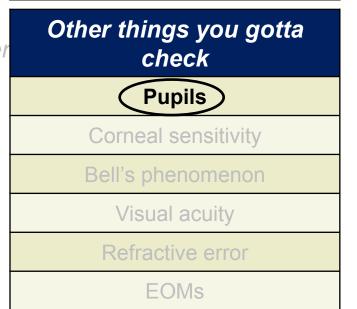
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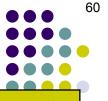
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In the context of a ptosis eval, what pupil finding would be particularly noteworthy?

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MRD2
Vertical fissure height
Upper lid crease position
Levator function
Presence of lagophthalmos





What is the 'proper' term for eyelid ptosis?

Blepharoptosis

What is the formal definition of ptosis? Inferodisplacement of the upper lid

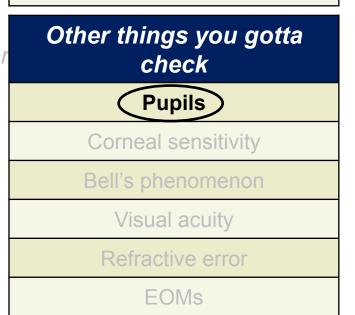
What are the two very general forms of ptosis?

Congenital and acquired

In the context of a ptosis eval, what pupil finding would be particularly noteworthy?

Anisocoria

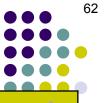
Lid-related observations in the ptosis patient
MRD1
MRD2
Vertical fissure height
Upper lid crease position
Levator function
Presence of lagophthalmos







Ptosis with anisocoria



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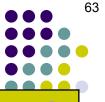
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Refractive error

EOMs



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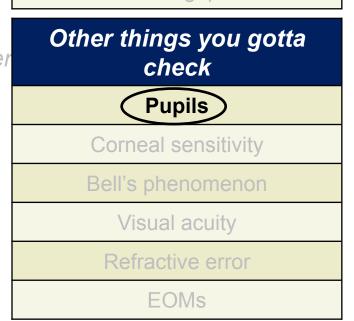
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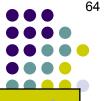
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Horner syndrome

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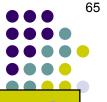
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MRD2
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EOMs

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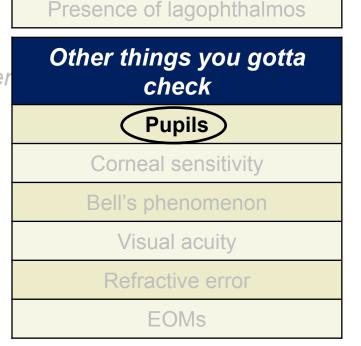
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CN3 palsy

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Levator function



66

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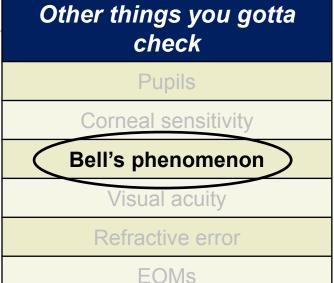
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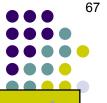
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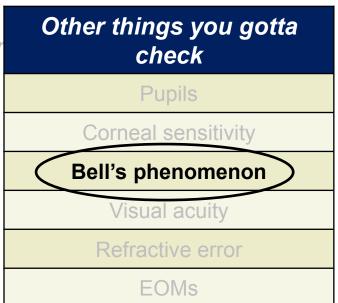
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la avaluation a atonia at air ann lid

What is Bell's phenomenon?

A reflex in which unsuccessful lid closure cause the globe to roll up and laterally

Lid-related observations in the ptosis patient MRD1 MRD2 Vertical fissure height Upper lid crease position Levator function Presence of lagophthalmos







Bell's phenomenon

69

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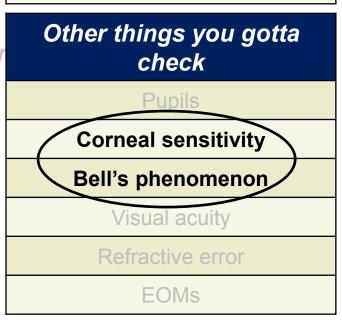
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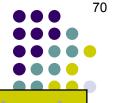
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Why must corneal sensitivity and Bell's phenomenon be assessed?

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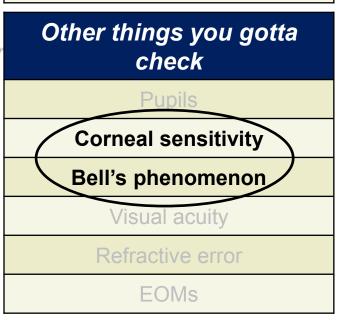
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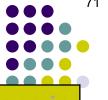
In evaluating a ptosis pt, six observations/measurer

Why must corneal sensitivity and Bell's phenomenon be assessed?

Ptosis often necessitates lid-elevation surgery, which may leave the ocular surface exposed. Thus, it is of paramount importance that the surgeon be aware of the status of the eye regarding corneal sensitivity and Bell's phenomenon, as these factors play a vital role in surface health.

Lid-related observations in the ptosis patient MRD1 MRD2 Vertical fissure height Upper lid crease position Levator function Presence of lagophthalmos





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What makes VA of particular concern in ptosis?

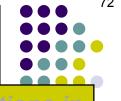
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MRD1 MRD2 Vertical fissure height Upper lid crease position Levator function

Presence of lagophthalmos

Other things you gotta check Pupils Corneal sensitivity Bell's phenomenon Visual acuity Refractive error **EOMs**





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Vertical fissure height

Upper lid crease position

Levator function

Presence of lagophthalmos

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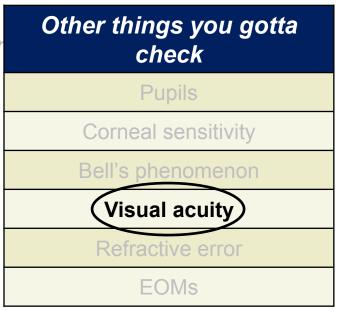
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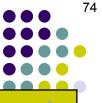
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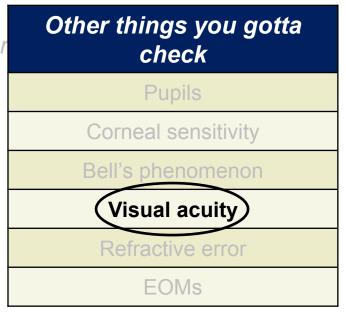
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What are they?

This is why assessing refractive error is an important component of a congenital ptosis workup

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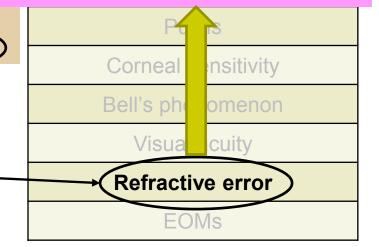
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MRD1

MRD2

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Upper lid crease position

Levator function

80

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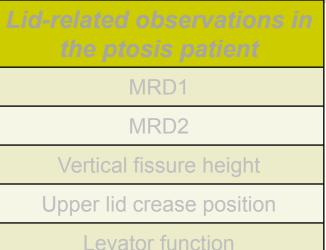
Congenital and ac Astigmatism

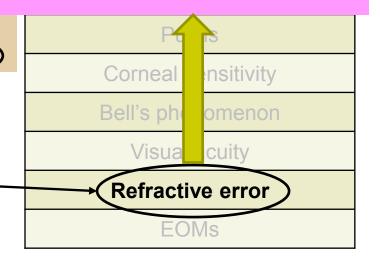
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Does the astigmatism resolve after successful ptosis surgery?

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Does the astigmatism resolve after successful ptosis surgery? Obviously, if the ptotic lid col Generally **not**--so be sure to re-refract after surgery

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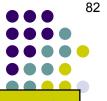
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What two EOM issues are of particular concern when evaluating congenital ptosis?

Lid-related observations in the ptosis patient

MRD1

MRD2

Vertical fissure height

Upper lid crease position

Levator function

Presence of lagophthalmos

Other things you gotta check

Pupils

Corneal sensitivity

Refractive error

(EOMs

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--Ruling out a palsy

--Assessing for the presence of

three words

(aka

three diff words

Refractive error

EOMs

MRD1

MRD2

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Upper lid crease position

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Presence of lagophthalmos

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--Ruling out a CN3 palsy

--Assessing for the presence of monocular elevation deficiency (aka double elevator palsy)

Lid-related observations in the ptosis patient

MRD1

MRD2

Vertical fissure height

Upper lid crease position

Levator function

Presence of lagophthalmos

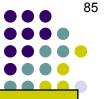
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In evalua Briefly, what is monocular elevation deficiency? What are

In evalua

- --Ruling out a CN3 palsy
- --Assessing for the presence monocular elevation deficiency aka double elevator palsy)

MRD1

MRD2

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Levator function

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Other things you gotta

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Refractive error

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Other things you gotta

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MRD2

Vertical fissure height

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Levator function

Presence of lagophthalmos

eck

ensitivity

In evalua Briefly, what is monocular elevation deficiency?

A form of vertical strabismus in which the hypotropic eye demonstrates deficient elevation in all fields of gaze

In evalua

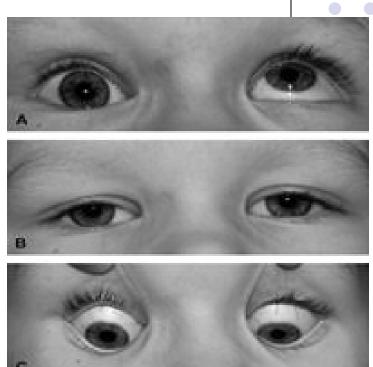
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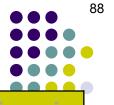
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Double elevator palsy



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In evalua What is the cause?

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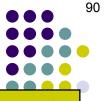
It can be restrictive, innervational, or a combo of both

--Assessing for the presence monocular elevation deficiency aka double elevator palsy)

Refractive error

EOMs

--Ruling out a CN3 palsy



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Ptosis etiology can be classified into six categories. What are they?

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Q

Congenital Ptosis



General categories ptosis etiology	of		
?			
?			
?		lote: These categories are not unique to cong tosis—they apply to acquired ptosis as well	genital
?			
?			
?			

Name the categories first...

A

Congenital Ptosis



General categories ptosis etiology	of		
Myogenic			
Neurogenic			
Aponeurotic		lote: These categories are not unique to cong tosis—they apply to acquired ptosis as well	genital
Mechanical			
Syndromic			
Traumatic			

Name the categories first...





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	? start here
Neurogenic	
Aponeurotic	
Mechanical	
Syndromic	
Traumatic	





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	
Aponeurotic	
Mechanical	
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Traumatic	





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General categories of ptosis etiology	Specific causes of congenital ptosis within each category
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Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	
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General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	Rare; associated with forceps injury
Mechanical	
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General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	Rare; associated with forceps injury
Mechanical	
Syndromic	
Traumatic	(So an argument could be made that it actually belongs here)





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	Rare; associated with forceps injury
Mechanical	? ?
Syndromic	
Traumatic	





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Which is the most common cause of congenital ptosis?





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Which is the most common cause of congenital ptosis?

Congenital myogenic ptosis (aka congenital fibrosis of the levator), by a mile





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
In three words, what is the e	etiology of congenital myogenic ptosis?
Traumatic	





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In three words, what is the etiology of congenital myogenic ptosis? Levator muscle dysgenesis

Traumatic





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What does that mean?





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What does that mean?

The levator fails to develop properly, with some or all of its muscle fibers replaced by fibrous and adipose tissue





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How is it inherited?





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What does that mean?

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How is it inherited?

This is not addressed in either the *Peds* or *Plastics* book, but both mention that it can be familial (so be sure to inquire re family hx)





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In congenital myogenic ptosis, what important finding manifests in downgaze?

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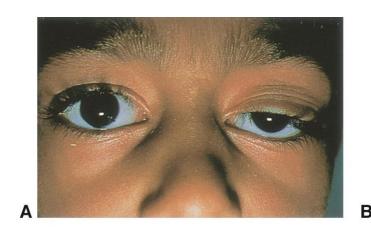
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Congenital myogenic ptosis: Lid lag





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In severe cases, ie, those in which fibrofatty tissue constitute nearly all of the substance of the levator, this inability to relax can lead to what related finding?





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In severe cases, ie, those in which fibrofatty tissue constitute nearly all of the substance of the levator, this inability to relax can lead to what related finding? **Lagophthalmos**

What is lagophthalmos?
Failure of the lids to close completely

Note that we have at last 'gotten to this shortly'

Hol up--why wouldn't a ptotic lid close completely?
We'll get to that shortly





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With regard to correction of congenital myogenic ptosis: At around what age should surgery be performed?

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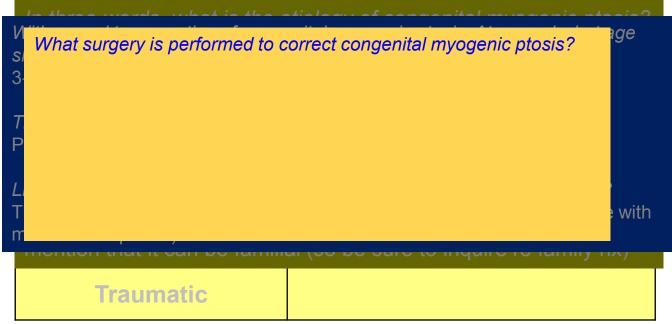
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Likewise, there is an indication for intervening in toddlerhood—what is it? The adoption by the child of a significant chin-up position (it may interfere with motor development)





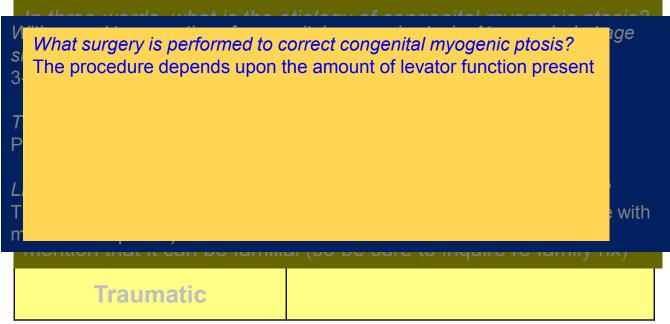
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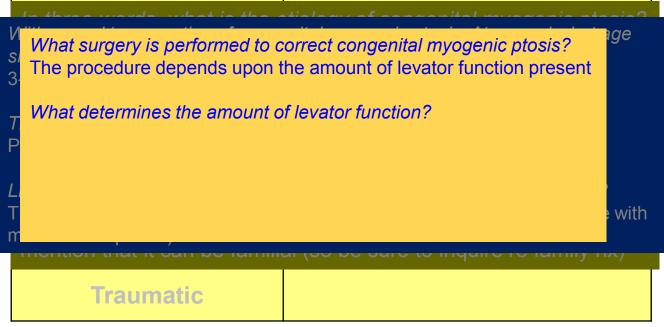
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Traumatic





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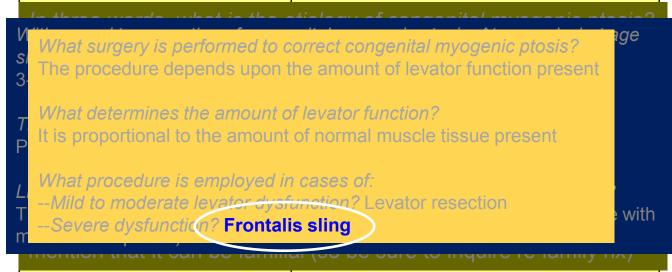
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What is the underlying premise of the frontalis sling procedure?





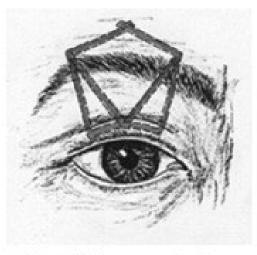
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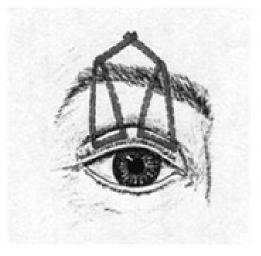
What is the underlying premise of the frontalis sling procedure?

When levator contraction is inadequate to clear the visual axis, people naturally engage the frontalis muscle to assist. The *frontalis sling procedure* directly connects the frontalis muscle to the lid margin, greatly increasing the effectiveness of frontalis contraction for elevating the lid.

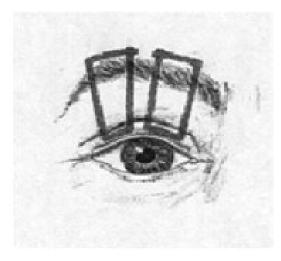




Collin & Kemp technique



Crawford technique



Double rhomboid technique

Frontalis sling*





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In severe unilateral congenital ptosis secondary to myogenic dysgenesis, should you perform unilateral, or bilateral slings?

Tough call. A unilateral sling spares the normally functioning lid, but leaves the patient with a markedly asymmetric appearance; bilateral slings yield a more symmetric appearance, but involves the destruction of normal tissue.

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What ocular sign is present in congenital Horner's that is not found in acquired Horner's?

Syndromic	Blepharophimosis syndrome
Traumatic	



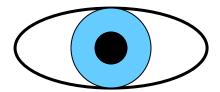


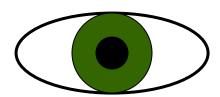
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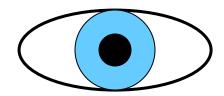
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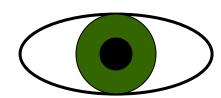
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Which iris is lighter—the one on the ptotic side, or the other?

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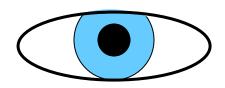
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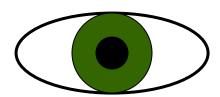
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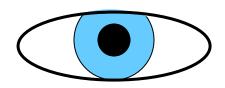


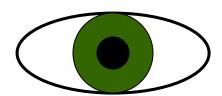
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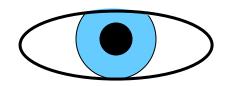


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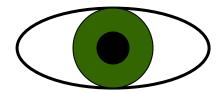
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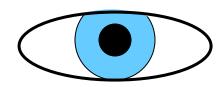
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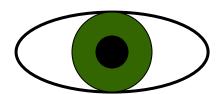
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- --The nipple on the ipsilateral side of the body is

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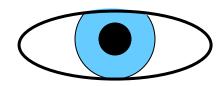
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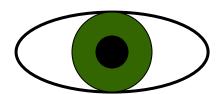
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What nonocular signs are present that aren't found in acquired Horner's?

- -- The hair on the ipsilateral side of the head is lighter
- -- The nipple on the ipsilateral side of the body is lighter

Syndromic	Blepharophimosis syndrome
Traumatic	









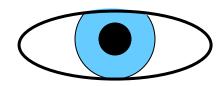
General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	CN3 palsy Horner's Marcus Gupp jaw wink

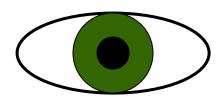
What ocular Another hair-related finding: er's that is not found in

Curly hair on the ipsilateral side of the head is viess ecurly

The ptotic side

Syndromic	Blepharophimosis syndrome
Traumatic	





ed Horner's?



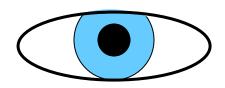


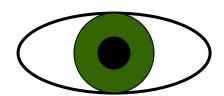
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What ocular Another hair-related finding: er's that is not found in

Curly hair on the ipsilateral side of the head is less curly

Syndromic	Blepharophimosis syndrome
Traumatic	







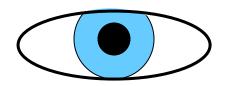


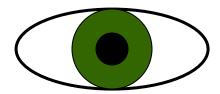
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Syndromic	Blepharophimosis syndrome
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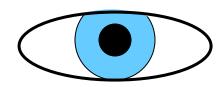
malignancy

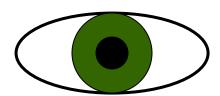
of the

two words

vvriich ins is lighter—the one on the ptotic side, or the other?
The ptotic side

Syndromic	Blepharophimosis syndrome
Traumatic	







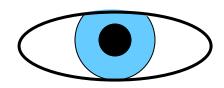


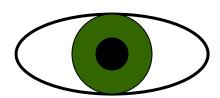
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What diagnosis must be considered as a cause of congenital Horner's? Neuroblastoma of the sympathetic chain

Syndromic	Blepharophimosis syndrome
Traumatic	









Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)?

ecific causes of congenital osis within each category

Congenital myogenic ptosis

CN3 palsy Horner's

Marcus Gunn jaw wink

e; associated with forceps injury

Plexiform neurofibroma Capillary hemangioma





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MGJW







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What is the classic story regarding when parents first note their infant has MGJW?

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What is the classic story regarding when parents first note their infant has MGJW? It is while the infant is nursing (Mom may say the infant's lid 'twitches' while nursing)

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Can MGJW present such that the ptosis worsens with jaw movements?





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Can MGJW present such that the ptosis worsens with jaw movements? Yes, but this is distinctly uncommon





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masticatory movements o

What are the muscles of mastication?

__

--Temporalis

Which cranial nerve innervates them? The trigeminal (V)

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eurofibroma emangioma

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Unilateral

Congenital Ptosis



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Which branch of the trigeminal?





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Q

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Which jaw movements are involved?

- -- Lateral displacement (contralateral to ptosis? ipsilateral to ptosis?)
- --Protrusion --Wide ope If lateral displacement is the movement that resolves the ptosis, is the
- direction of the displacement contralateral to the ptosis, or ipsilateral?





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What do these movements have in common?

All are performed by the _____ muscle





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Horner's Marcus Gunn jaw wink

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osis within each category

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- -- Lateral (or external) pterygoid
- --Masseter
- --Temporalis

Which cranial nerve innervates them? The trigeminal (V)

Which jaw movements are

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What do these movements have in common? All are performed by the lateral pterygoid muscle





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- eurofibroma -- Lateral (or external) pterygoid emangioma

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More common than you might think--it is present in about 6 of congenital ptosis cases

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The **involuntary** movement of one bodypart in response to the **voluntary** movement of another

ecific causes of congenital osis within each category

Congenital myogenic ptosis

CN3 palsy Horner's

Marcus Gunn jaw wink

Is MGJW sporadic, or familial?

Among pts with congenital ptosis, is Notes and the More common than you might think--it

--iviediai (or internai) pterygoid

--Lateral (or external) pterygoid

eurofibroma emangioma

What is the clinical hallmark -- Masseter

The ptotic lid What does this imply about the pathophysiology of MGJW?

masticatory r That it usually involves an abnormal connection between the levator palpebrae muscle of the ptotic eye and the contralateral lateral pterygoid muscle

Which jaw movements are I.....

-- Lateral displacement (contralateral to ptosis)

- --Protrusion
- --Wide opening

--Clenching <

What do these movements have in mmon?

All are performed by the lateral pterygoid muscle

What does this imply about the pathophysiology of MGJW? That it doesn't always involve the lateral pterygoid muscle

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What is the clinical ha muscle o

What doe The ptotic lid masticatory

Which jaw movements

- --Lateral displaceme
- --Protrusion
- --Wide opening

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205

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'Mask-like'

Which jaw movements

--Lateral displaceme

--Protrusion

--Wide opening

--Clenching <

on? oid muscle



210

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Which jaw movement: 'Mask-like'

--Lateral displaceme

--Protrusion

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--Clenching •

Are vertical eye movements affected as well?

on? oid muscle



211

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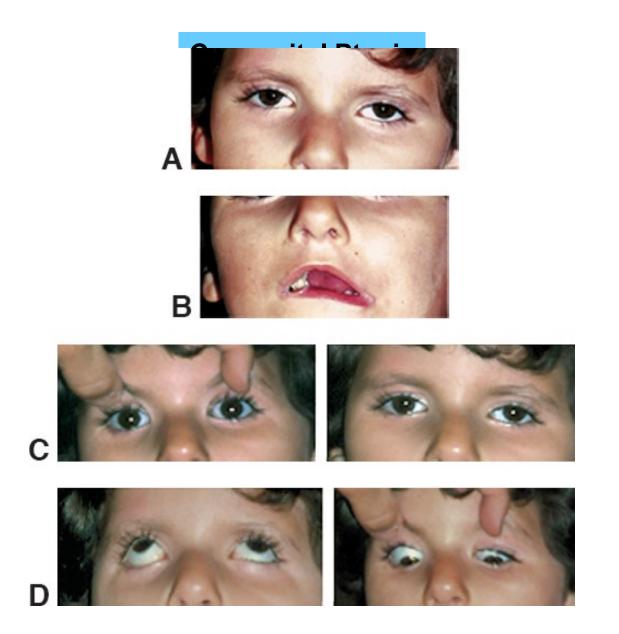
--Protrusion

--Wide opening

--Clenching <

Are vertical eye movements affected as well? No, they are intact

non? oid muscle



212

Mobiüs syndrome. **A**, orthotropia in primary. **B**, inability to smile. **C**, bilaterally absent adduction and severely limited abduction. **D**, Vertical movements are not affected.



--Clenching <

Congenital Ptosis

213

Broadly speaking, what sort of disorder is ecific causes of congenital Marcus Gunn jaw wink (MGJW)? osis within each category It is one of synkinesis congenital myogenic ptosis What does synkinesis refer to? CN3 palsy The **involuntary** movement of one bodypart in respo anothe In a nutshell, what findings are typical in Mobiüs syndrome? Another conge What tops the DDx for a Mobiüs-like presentation in a newborn? Mobiüs syndrome What is the clinical ha V What doe The ptotic lid masticatory Which jaw movements --Lateral displaceme non? -- Protrusion oid muscle --Wide opening



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Nah—the condition resolves spontaneously and rapidly

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General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis
Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	Rare; associated with forceps injury
Mechanical	Plexiform neurofibroma Capillary hemangioma

What two specific cell types are implicated in plexiform neurofibroma formation?





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Are plexiform neurofibromas associated with a systemic condition?





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Neurogenic	CN3 palsy Horner's Marcus Gunn jaw wink
Aponeurotic	Rare; associated with forceps injury
Mechanical	Plexiform neurofibroma Capillary hemangioma

What two specific cell types are implicated in plexiform neurofibroma formation? Schwann cells and fibroblasts

These nonencapsulated, diffusely infiltrative tumors have a distinct consistency when palpated. What icky term is used to describe it?

'Bag of worms'

Are plexiform neurofibromas associated with a systemic condition? Yes—they are strongly associated with





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What proportion of plexiform neurofibroma pts have NF1?







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tl;dr Not all NF1 pts get plexiform neuofibromas, but everyone with plexiform neurofibromas has NF1

The word, what sort of condition is NF1?

A phakomatosis





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Their predilection for the temporal aspect of the upper lid produces a specific configuration of the involved lid. What is it?

Yes—they are strongly associated with NF1

A phakomatosis





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tl;dr Not all NF1 pts get plexiform r with plexiform neurofibromas has Their predilection for the temporal aspect of the upper lid produces a specific configuration of the involved lid. What is it?

The tumor gives rise to an 'S-shaped lid'

Yes—they are strongly associated with NF1

A phakomatosis



S-shaped lid deformity



Plexiform neurofibroma: S-shaped lid





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In addition to producing ptosis, plexiform neurofibromas of the upper lid are a strong risk factor for what potentially blinding ocular condition?

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Glaucoma

tinct consistency

How does a plexiform neurofibroma cause glaucoma?

Their predilection for the temporal aspect of the upper lid produces olived lid. What is it?

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are a **strong risk factor for** what potentially blinding ocular condition?

Glaucoma

tinct consistency

Their predilection for the temporal aspect of the upper lid produces

How does a plexiform neurofibroma cause glaucoma?

It doesn't (so far as we know); rather, there is simply a strong association between them and the development of ipsilateral glaucoma

rolved lid. What is it?

aped lid'

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General categories of	Specific causes of congenital
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Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

Mechanical	Plexiform neurofibroma Capillary hemangioma
Syndromic	Blepharophimosis syndrome
Traumatic	





General categories of | Specific causes of congenital

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General categories of | Specific causes of congenital

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Is there a gender predilection?

Mechanical	Plexiform neurofibroma Capillary hemangioma
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General categories of | Specific causes of congenital

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Is there a gender predilection?

Yes, it is more common in MUE

Mechanical	Plexiform neurofibroma Capillary hemangioma
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General categories of | Specific causes of congenital

Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

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Mechanical	Plexiform neurofibroma Capillary hemangioma
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General categories of | Specific causes of congenital

Among the orbital vascular lesions, where does the capillary hemangioma rank in terms of prevalence?

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What proportion of orbital capillary hemangiomas are congenital?

Mechanical	Plexiform neurofibroma Capillary hemangioma
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General categories of | Specific causes of congenital

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Mechanical	Plexiform neurofibroma Capillary hemangioma
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Mechanical

Plexiform neurofibroma Capillary hemangioma

How does capillary hemangioma present?





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General categories of | Specific causes of congenital

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Mechanical

Plexiform neurofibroma Capillary hemangioma

How does capillary hemangioma present?

As a **bluish** or **strawberry-like** lesion of the periorbital skin



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Capillary hemangioma





General categories of | Specific causes of congenital

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How does capillary hemangioma present?

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What determines whether it appears bluish vs strawberry-like?





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Among the orbital vascular hemangioma rank in terms It is the most common	lesions, where does the capillary of prevalence?	
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Mechanical

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- --Superficial lesions can be treated with topical timolol, or laser

About 1/3

Mechanical

Plexiform neurofibroma

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About 1/3

Mechanical

Plexiform neurofibroma

Capillary hemangioma

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Mechanical

Plexiform neurofibroma Capillary hemangioma

Disclaimer: These treatment guidelines were taken from the latest (2018-19) version of the BCSC *Peds* book. It should be noted that the *Orbit* book indicates that **steroids** (systemic or local) are the preferred first-line treatment for capillary hemangiomas. In contrast, the *Peds* book doesn't even mention them. Which book is right? I'm going with the *Peds* version. But as always, caveat emptor.

It appears **bluish** if it's...**deep** It appears strawberry-like if it's...superficial





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

--Ptosis

--

_

and, of course,

--

	Ouplinary Herriangionna
Syndromic	Blepharophimosis syndrome
Traumatic	





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

- --Ptosis
- --Telecanthus
- --Epicanthus inversus and, of course,
- --Blepharophimosis

Syndromic	Blepharophimosis syndrome
Traumatic	





Blepharophimosis syndrome





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- --Ptosis
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Blepharophimosis syndrome has two other names. What are they?

-

-

	Capillary Herriangionna
Syndromic	Blepharophimosis syndrome
Traumatic	





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of

blepharophimosis syndrome aka congenital eyelid syndrome, and

--Ptosis

blepharophimosis-ptosis-epicanthus inversus syndrome

- --Telecanthus
- --Epicanthus inversus and, of course,
- --Blepharophimosis

Blepharophimosis syndrome has two other names. What are they?

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

Syndromic	Blepharophimosis syndrome
Traumatic	





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What does it mean to say a pt has blepharophimosis?

Syndromic	Blepharophimosis syndrome
Traumatic	





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What does it mean to say a pt has blepharophimosis?

It means her/his palpebral fissures are abnormally in both vertical and horizontal extent

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Syndromic Blepharophimosis syndrome
Traumatic





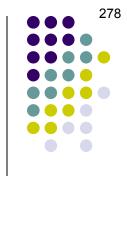
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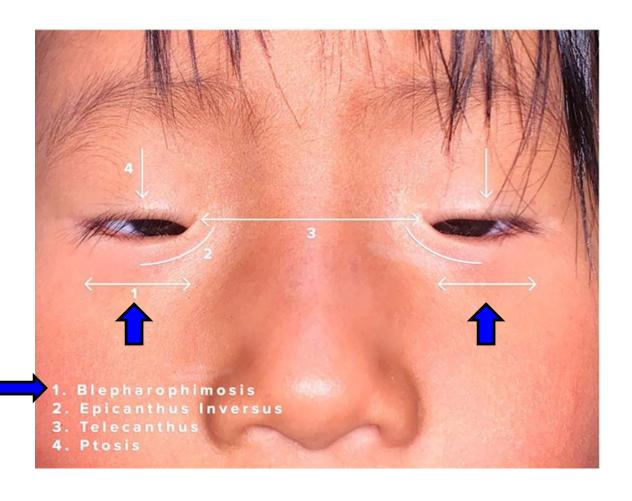
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- --Ptosis
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- --Epicanthus inversus and, of course,
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What does it mean to say a pt has blepharophimosis? It means her/his palpebral fissures are abnormally short in both vertical and horizontal extent

Syndromic	Blepharophimosis syndrome
Traumatic	





Blepharophimosis syndrome: Blepharophimosis





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

--Ptosis

What is telecanthus?

- --Telecanthus
- --Epicanthus inversas and, of course,
- --Blepharophimosis

	Ouplinary normangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





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What is telecanthus?

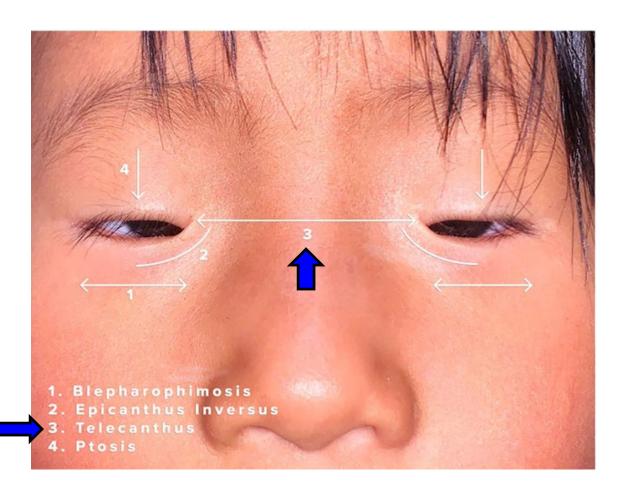
--Telecanthus

An abnormally increased distance between the medial canthi

- --Epicanthus inversal and, of course,
- --Blepharophimosis

	Ouplinary normangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





Blepharophimosis syndrome: Telecanthus





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Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

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What does the term epicanthus refer to in this context?

iomangioma





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What is epicanthus inversus?





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What is epicanthus inversus? It is one type of epicanthus





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What is epicanthus inversus? It is one type of epicanthus

How many types are there?





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

- --Ptosis
- --Telecanthus
- -- Epicanthus inversus

What does the term epicanthus refer to in this context? An epicanthus is a fold of skin extending above and below her remains an income and the properties and the properties and the properties are remains an income and the properties are remains an income and the properties and the properties and the properties and the properties are remains an income and the premains are remains an income and the properties are remains an in the medial canthal region

What is epicanthus inversus? It is one type of epicanthus

How many types are there? Four





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What are the other three?





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How many types are there? Four

What are the other three? Coming in hot...

nosis syndrome





What are the four types of epicanthus?

- --Epicanthus
- -- Epicanthus inversus mnemonic forthcoming...
- -- Epicanthus
- -- Epicanthus

-- relecantnus

-- Epicanthus inversus

and, of course,

	Capillary Hornarigionia
Syndromic	Blepharophimosis syndrome
Traumatic	



What are the four types of epicanthus?

- -- Epicanthus **t** (start here)
- -- Epicanthus inversus mnemonic forthcoming... TIPS

- --Epicanthus p
- --Epicanthus s

-- relecantnus

-- Epicanthus inversus

and, of course,

	Capillary Herriangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	



What are the four types of epicanthus?

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

-- relecantitus

-- Epicanthus inversus

and, of course,

	Capillary Hernangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





What are the four types of epicanthus?

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

-- relecantitus

-- Epicanthus inversus

and, of course,

	Ouplinary Herriangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





What are the four types of epicanthus?

- -- Epicanthus tarsalis
- -- Epicanthus *inversus*
- --Epicanthus *palpebralis*
- -- Epicanthus supraciliaris

-- relecantnus

-- Epicanthus inversus

and, of course,

	Capillary Herriangionia
Syndromic	Blepharophimosis syndrome
Traumatic	



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

- --Epicanthus *tarsalis:* Primarily lid
- --Epicanthus inversus:
- --Epicanthus *palpebralis*:
- -- Epicanthus *supraciliaris*:

-- relecantitus

-- Epicanthus inversus

and, of course,

	Capillary Hernangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





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

- -- Epicanthus tarsalis: Primarily upper lid
- -- Epicanthus inversus:
- --Epicanthus *palpebralis*:
- -- Epicanthus *supraciliaris*:

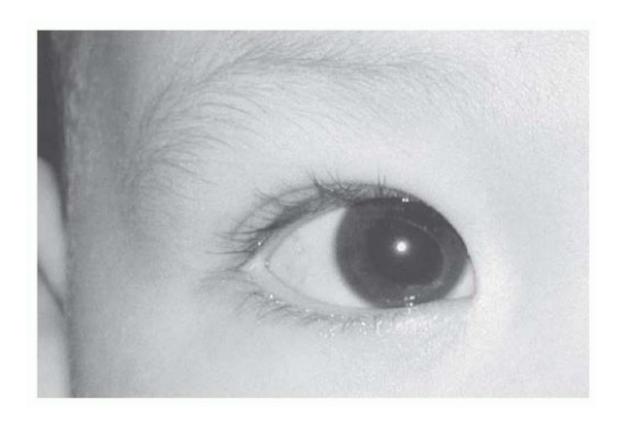
-- relecantitus

-- Epicanthus inversus

and, of course,

Syndromic	Blepharophimosis syndrome
Traumatic	





Epicanthus tarsalis



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

- -- Epicanthus tarsalis: Primarily upper lid
- --Epicanthus inversus: Primarily lid
- --Epicanthus *palpebralis*:
- -- Epicanthus *supraciliaris*:

-- relecantitus

-- Epicanthus inversus

and, of course,

	Ouplinary Hornarigionna
Syndromic	Blepharophimosis syndrome
Traumatic	





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- --Epicanthus *palpebralis*:
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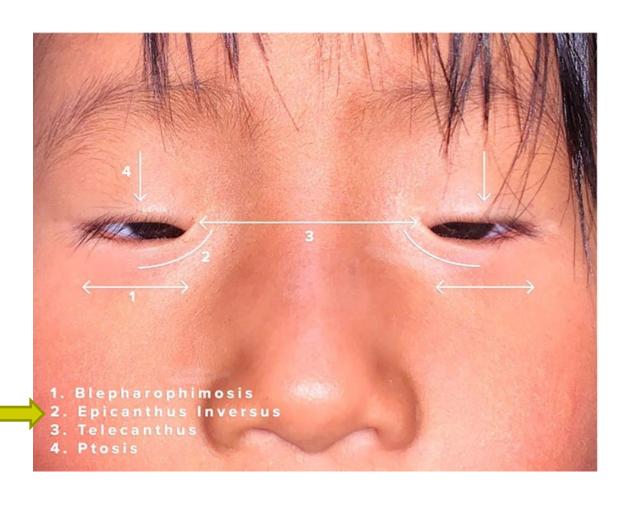
-- rejecantinus

-- Epicanthus inversus

and, of course,

Syndromic	Blepharophimosis syndrome
Traumatic	





Blepharophimosis syndrome: 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*: and equally
- -- Epicanthus *supraciliaris*:

-- relecantitus

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and, of course,

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Syndromic	Blepharophimosis syndrome
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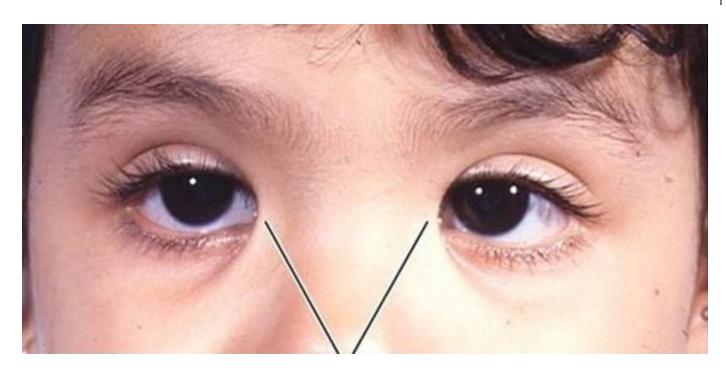
-- relecantinus

-- Epicanthus inversus

and, of course,

	Oapmary normangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





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 equally
- --Epicanthus supraciliaris: From to lid

-- relecantinus
--**Epicanthus inversus**and, of course,
--Blepharophimosis

	Oapillary Herriangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	



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: From brow to lower lid

-- relecantinus

-- Epicanthus inversus

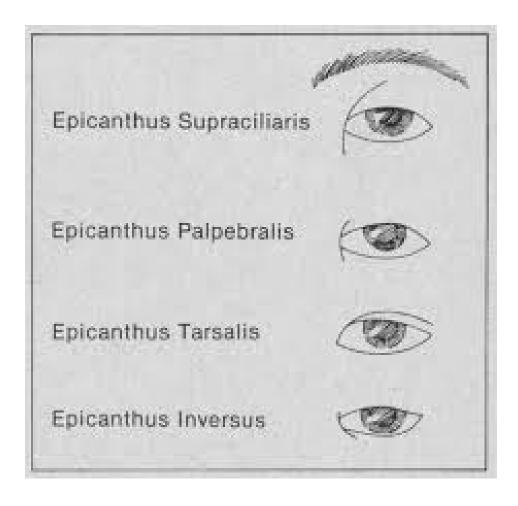
and, of course,

	Capillary Hernangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





Epicanthus supraciliaris



Epicanthal folds overview







General categories of ptosis etiology	Specific causes of congenital ptosis within each category
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Other than ptosis, what are the three other defining ophthalmic features of

blepharop Is the ptosis purely structural (ie, 2ndry to blepharophimosis),

- --Ptosis or is there a problem with the levator?
- --Telecantl
- --Epicanthus inversus and, of course,
- --Blepharophimosis

	Ouplinary normangiorna
Syndromic	Blepharophimosis syndrome
Traumatic	





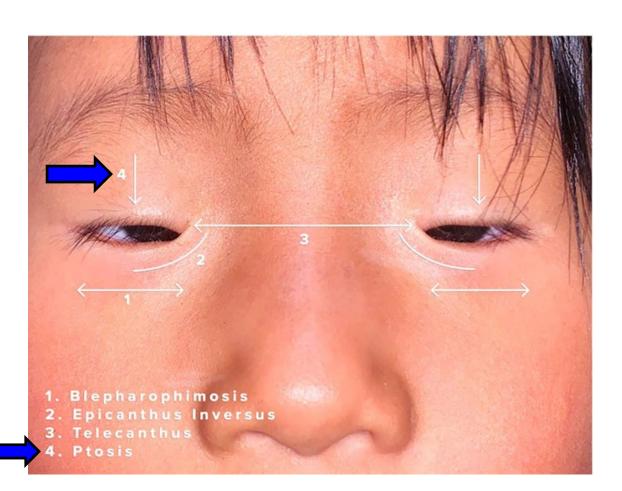
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Syndromic	Blepharophimosis syndrome
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Blepharophimosis syndrome: Ptosis







General categories of ptosis etiology	Specific causes of congenital ptosis within each category
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and, of co What does this imply regarding surgical repair?

--Blephan

	Ouplifully Hernangiornal
Syndromic	Blepharophimosis syndrome
Traumatic	





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Syndromic	Blepharophimosis syndrome
Traumatic	





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Syndromic	Blepharophimosis syndrome
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- --Ptosis
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Syndromic	Blepharophimosis syndrome
Traumatic	





General categories of ptosis etiology	Specific causes of congenital ptosis within each category
Myogenic	Congenital myogenic ptosis

Other than ptosis, what are the three other defining ophthalmic features of blepharophimosis syndrome?

--Hypoplastic

- --Ptosis
- --Telecanthus
- --Epicanthus inversus and, of course,
- --Blepharophimosis

	Hypop	lastic	two words		
	one word	ears			

Syndromic	Blepharophimosis syndrome
Traumatic	





General categories of ptosis etiology	Specific causes of congenital ptosis within each category		
Myogenic	Congenital myogenic ptosis		

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- --Ptosis
- --Telecanthus
- -- Epicanthus inversus and, of course,
- --Blepharophimosis

 Н١	/no	olas	stic	nasal	hr	ida	e
 шу	יטטי	via	Suc	Hasai	וט	ıuy	

-- 'Lop' ears

	Capillary Herriangionia
Syndromic	Blepharophimosis syndrome
Traumatic	





Hypoplastic nasal bridge (note also the chin-up positioning)



Lop ears

Blepharophimosis syndrome