General categories of ptosis etiology

Note: These categories are not unique to congenital ptosis; they apply to acquired ptosis as well

Name the categories first…
General categories of ptosis etiology

- Myogenic
- Neurogenic
- Aponeurotic
- Mechanical
- Traumatic

Name the categories first…

Congenital ptosis: Fill ‘er up
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**CNIII = Third cranial nerve (ie, the oculomotor nerve)**

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Which is the most common cause of congenital ptosis?

Localized myogenic dysgenesis (aka congenital fibrosis of the levator), by a mile.

In myogenic dysgenesis, what is the key finding on downgaze?

Lid lag.

The muscle is composed of fibrofatty tissue that can neither contract (causing ptosis) nor relax (causing lid lag). Frank lagophthalmos can be present.

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SR = Superior rectus
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Anisometropia and/or strabismus are the more-common causes

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**Aponeurotic**

Rare; associated with forceps injury

**Mechanical**

--Plexiform neuroma  
--Capillary hemangioma

**Traumatic**


**Congenital ptosis: Fill ‘er up**

What surgery is performed to correct congenital ptosis due to myogenic dysgenesis?

The specific procedure depends upon the amount of levator function present.

What determines the amount of levator function present?

It is proportional to the amount of normal muscle tissue present.

What are the specific guidelines relating levator function and choice of procedure?

- $\text{LF} > 10 \text{mm}$: Aponeurosis resection
- $6 \leq \text{LF} < 10$ : Aponeurosis + levator resection
- $\text{LF} < 6$ : Frontalis sling

What is lid lag?

The muscle is composed of fibrofatty tissue that can neither contract (causing ptosis) nor relax (causing lid lag). Frank lagophthalmos can be present.

What two nonlid findings are frequently associated with myogenic dysgenesis of the levator?

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**Traumatic**

Rare; associated with blunt trauma.

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**Lid lag**: The muscle is composed of fibrofatty tissue that can neither contract (causing ptosis) nor relax (causing lid lag). Frank lagophthalmos can be present.

**What two nonlid findings are frequently associated with myogenic dysgenesis of the levator?**
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- **Myogenic**
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- **Aponeurotic**
  - Rare; associated with forceps injury
- **Mechanical**
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#### In severe unilateral congenital ptosis secondary to myogenic dysgenesis, should you perform unilateral or bilateral slings?

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### Additional Notes

In myogenic dysgenesis, what is the key finding on downgaze?

Lid lag. The muscle is composed of fibrofatty tissue that can neither contract (causing ptosis) nor relax (causing lid lag). Frank lagophthalmos can be present.

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### Traumatic

- Rare; associated with forceps injury

## Congenital Ptosis: Fill ‘er up

### Which is the most common cause of congenital ptosis?

Localized myogenic dysgenesis (aka congenital fibrosis of the levator), by a mile

### In myogenic dysgenesis, what is the key finding on downgaze?

Lid lag

The muscle is composed of fibrofatty tissue that can neither contract (causing ptosis) nor relax (causing lid lag). Frank lagophthalmos can be present.

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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 necessitate destruction of normal tissue.
What ocular sign is present in congenital Horner’s that is not found in acquired Horner’s?

Heterochromia iridis—the iris on that side is lighter.

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

- Lighter hair on the ipsilateral side of the head
- A lighter-colored nipple on that side of the chest

What diagnosis must be considered as a cause of congenital Horner’s?

Neuroblastoma of the sympathetic chain

What imaging studies are needed to evaluate for neuroblastoma?

MRI of the head and neck

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

**What does synkinesis refer to?**

The involuntary movement of one body part in response to the voluntary movement of another

**Is the ptosis of MGJW unilateral, or bilateral?**

Unilateral

**What is the clinical hallmark of MGJW?**

The ptotic lid elevates in response to voluntary movements of the jaw

**Which jaw movements are involved?**

- Lateral displacement
- Protrusion
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It is while the infant is nursing (Mom may say the infant’s lid ‘twitches’ while nursing)
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

What does synkinesis refer to? The involuntary movement of one bodypart in response to the voluntary movement of another.

Is the ptosis of MGJW unilateral, or bilateral? Unilateral.

What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

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).

### General categories of ptosis etiology

<table>
<thead>
<tr>
<th>Category</th>
<th>Specific causes of congenital ptosis within each category</th>
</tr>
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Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)?
It is one of synkinesis

What does synkinesis refer to?
The involuntary movement of one body part in response to the voluntary movement of another

Is the ptosis of MGJW unilateral, or bilateral?
Unilateral

What is the clinical hallmark of MGJW?
The ptotic lid elevates in response to voluntary masticatory movements of the jaw

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

A
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

What does synkinesis refer to? The involuntary movement of one body part in response to the voluntary movement of another.

Is the ptosis of MGJW unilateral, or bilateral? Unilateral.

What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Can MGJW present such that the ptosis worsens with jaw movements? Yes, but this is distinctly uncommon.
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

What does synkinesis refer to?
The involuntary movement of one body part in response to the voluntary movement of another

Is the ptosis of MGJW unilateral or bilateral? Bilateral

What are the muscles of mastication?
- Medial (or internal) pterygoid
- Lateral (or external) pterygoid
- Masseter
- Temporalis

Which cranial nerve innervates them?
The trigeminal

What is the clinical hallmark of MGJW?
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Congenital ptosis: Fill ‘er up

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- Temporalis

What are the masticatory movements of the jaw?
- Lateral displacement
- Protrusion
- Wide opening
- Clenching

Which cranial nerve innervates them?
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Is the ptosis of MGJW unilateral, or bilateral?
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Which cranial nerve innervates them?
The trigeminal nerve.

What are the specific causes of congenital ptosis within each category?

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What are the muscles of mastication?
- Medial (or internal) pterygoid
- Lateral (or external) pterygoid
- Masseter
- Temporalis

Which cranial nerve innervates them?
The trigeminal (V)

**Congenital ptosis: Fill ‘er up**

The ptotic lid elevates in response to voluntary masticatory movements of the jaw
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The ptotic lid elevates in response to voluntary masticatory movements of the jaw

Which jaw movements are involved?
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- Medial (or internal) pterygoid
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- Temporalis

Which cranial nerve innervates them?

The trigeminal (V)

Which branch of the trigeminal?
- Mandibular (V3)

Which branch of the trigeminal innervates the muscles of mastication?
- Mandibular (V3)
Congenital ptosis: Fill ‘er up

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

What does synkinesis refer to? The involuntary movement of one body part in response to the voluntary movement of another.

What are the muscles of mastication? --Medial (or internal) pterygoid --Lateral (or external) pterygoid --Masseter --Temporalis

Which cranial nerve innervates them? The trigeminal (V)

Which branch of the trigeminal? The mandibular (V₃)

What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Are the movements of MGJW unilateral or bilateral? Bilateral.

Which movements are involved? --Lateral displacement --Protrusion --Wide opening --Clenching

Are the muscles of mastication innervated by the same cranial nerve? Yes, the trigeminal (V) innervates both the mandibular (V₃) and maxillary (V₂) branches. However, the spinal accessory nerve (CN XI) innervates the sternocleidomastoid and trapezius muscles, which are also involved in mastication.

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**Which jaw movements are involved?**

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**Is the ptosis of MGJW unilateral, or bilateral?**
Unilateral.

**What is the clinical hallmark of MGJW?**
The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

**Which jaw movements are involved?**
- Lateral displacement (contralateral to ptosis? ipsilateral to ptosis?)
- Protrusion
- Wide opening
- Clenching

**If lateral displacement is the movement that resolves the ptosis, is the direction of the displacement contralateral to the ptosis, or ipsilateral?**
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

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Is the ptosis of MGJW unilateral, or bilateral? Unilateral.

What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved? --Lateral displacement (contralateral to ptosis) --Protrusion --Wide opening --Clenching

If lateral displacement is the movement that resolves the ptosis, is the direction of the displacement contralateral to the ptosis, or ipsilateral? Contralateral.
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)?

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  - CNIII palsy
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  - Marcus Gunn jaw wink

- Aponeurotic

- Mechanical

- Traumatic; associated with forceps injury
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  - Capillary hemangioma

What do these movements have in common?

All are performed by the lateral pterygoid muscle.
Q/A

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

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Which jaw movements are involved? -- Lateral displacement (contralateral to ptosis) -- Protrusion -- Wide opening -- Clenching

(Note: Is aka the external pterygoid muscle)
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

What does synkinesis refer to? The involuntary movement of one body part in response to the voluntary movement of another.

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What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved? -- Lateral displacement (contralateral to ptosis) -- Protrusion -- Wide opening -- Clenching.

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

What does this imply about the pathophysiology of MGJW? That it usually involves an abnormal connection between the levator palpebrae muscle of the ptotic eye and the contralateral lateral pterygoid muscle.
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

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Which jaw movements are involved? --Lateral displacement (contralateral to ptosis) --Protrusion --Wide opening --Clenching

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

What does this imply about the pathophysiology of MGJW?
General categories of ptosis etiology

Specific causes of congenital ptosis within each category

<table>
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</tr>
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- Mechanical
- Traumatic
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Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)?

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What does synkinesis refer to?

The **involuntary** movement of one body part in response to the **voluntary** movement of another.

Is the ptosis of MGJW unilateral, or bilateral?

Unilateral

What is the clinical hallmark of MGJW?

The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved?

- Lateral displacement (contralateral to ptosis)
- Protrusion
- Wide opening
- Clenching

What does this imply about the pathophysiology of MGJW?

That it doesn’t **always** involve the lateral pterygoid muscle.
**Congenital ptosis: Fill ‘er up**

**General categories of ptosis etiology**

**Specific causes of congenital ptosis within each category**

- **Myogenic**
  - localized myogenic dysgenesis

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

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**What does synkinesis refer to?**

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

**Is the ptosis of MGJW unilateral, or bilateral?**

Unilateral

**What is the clinical hallmark of MGJW?**

The ptotic lid elevates in response to voluntary masticatory movements of the jaw

**Which jaw movements are involved?**

- Lateral displacement (contralateral to ptosis)
- Protrusion
- Wide opening

**--Clenching**

**What do these movements have in common?**

All are performed by the lateral pterygoid muscle

**What muscle is implicated when clenching is the triggering movement?**

The **medial pterygoid**
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

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What is the clinical hallmark of MGJW?
The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved?
--Lateral displacement (contralateral to ptosis)
--Protrusion
--Wide opening
--Clenching

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

What muscle is implicated when clenching is the triggering movement? The medial pterygoid.

(Note: Is aka the internal pterygoid)
Broadly speaking, what sort of disorder is Marcus Gunn jaw wink (MGJW)? It is one of synkinesis.

What does synkinesis refer to? The involuntary movement of one bodypart in response to the voluntary movement of another.

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What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved? -- Lateral displacement (contralateral to ptosis) -- Protrusion -- Wide opening -- Clenching.

Among pts with congenital ptosis, is MGJW a common, or uncommon finding? More common than you might think -- it is present in about 5% of congenital ptosis cases.
Q/A

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

What does synkinesis refer to? The involuntary movement of one body part in response to the voluntary movement of another.

Is the ptosis of MGJW unilateral, or bilateral? Unilateral.

Which jaw movements are involved? Lateral displacement (contralateral to ptosis), protrusion, wide opening, clenching.

Among pts with congenital ptosis, is MGJW a common, or uncommon finding? More common than you might think—it is present in about 5% of congenital ptosis cases.

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Which jaw movements are involved?--Lateral displacement (contralateral to ptosis)--Protrusion--Wide opening--Clenching

Is MGJW sporadic, or familial? The vast majority of cases are sporadic.

Specific causes of congenital ptosis within each category:

- Myogenic
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Q: "Congenital ptosis: Fill ‘er up"
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Which jaw movements are involved? --Lateral displacement (contralateral to ptosis) --Protrusion --Wide opening --Clenching

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What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw. Which jaw movements are involved? --Lateral displacement --Protrusion --Wide opening --Clenching. All are performed by the lateral pterygoid muscle.

In Marcus Gunn jaw-winking syndrome, CN5 (dys)innervates the levator. Thus, Marcus Gunn jaw wink is an example of a congenital cranial dysinnervation disorder. Another such disorder should readily come to mind—what is it?
A

**Congenital ptosis: Fill ‘er up**

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The ptotic lid elevates in response to voluntary masticatory movements of the jaw

**Which jaw movements are involved?**

- --Lateral displacement
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**Duane syndrome**
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Is the ptosis of MGJW unilateral, or bilateral? Unilateral.

What is the clinical hallmark of MGJW? The ptotic lid elevates in response to voluntary masticatory movements of the jaw.

Which jaw movements are involved? Lateral displacement, protrusion, wide opening, clenching (contralateral to ptosis).

All are performed by the lateral pterygoid muscle.

In Marcus Gunn jaw-winking syndrome, CN5 (dys)innervates the levator. Thus, Marcus Gunn jaw wink is an example of a congenital cranial dysinnervation disorder. Another such disorder should readily come to mind—what is it? Duane syndrome.

In Duane syndrome, which cranial nerve (dys)innervates what muscle?
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In Duane syndrome, which cranial nerve (dys)innervates what muscle? CN3 innervates the lateral rectus.
# General categories of ptosis etiology

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**What refractive problem is associated with virtually all forms of congenital ptosis?**

Astigmatism

**Does the astigmatism resolve after successful ptosis surgery?**

Generally no, so be sure to re-refract after surgery.
General categories of ptosis etiology | Specific causes of congenital ptosis within each category

<table>
<thead>
<tr>
<th>Myogenic</th>
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<tr>
<td></td>
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<td>Aponeurotic</td>
<td>Rare; associated with forceps injury</td>
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What refractive problem is associated with virtually all forms of congenital ptosis? Astigmatism
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--Horner’s  

--Marcus Gunn jaw wink |
| **Aponeurotic**| Rare; associated with forceps injury                       |
| **Mechanical** | --Plexiform neuroma  

--Capillary hemangioma |
| **Traumatic**  |                                                          |

**Congenital ptosis: Fill ‘er up**

What refractive problem is associated with virtually all forms of congenital ptosis?

Astigmatism

Does the astigmatism resolve after successful ptosis surgery?
**General categories of ptosis etiology**

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**Congenital ptosis: Fill ‘er up**

**What refractive problem is associated with virtually all forms of congenital ptosis?**
Astigmatism

**Does the astigmatism resolve after successful ptosis surgery?**
Generally **not**--so be sure to re-refract after surgery
### General categories of ptosis etiology

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#### Acquired ptosis: Fill ‘er up

Now let’s look at acquired ptosis
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<td></td>
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<td>CPEO <em>(chronic progressive external ophthalmoplegia)</em></td>
</tr>
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| **Myogenic**                         | MG  
Muscular dystrophy  
CPEO                                                   |
| **Neurogenic**                       | (2, maybe 3)                                           |
| Aponeurotic                          |                                                        |
| Mechanical                           |                                                        |
| Traumatic                            |                                                        |
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**Acquired ptosis: Fill ‘er up**

89
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### Specific causes of acquired ptosis within each category

- **Myogenic**: MG
- **Neurogenic**: Horner's CNIII palsy
- **Aponeurotic**
- **Mechanical**
- **Traumatic**

*(The BCSC books equivocate with respect to whether MG is a *myogenic* vs a *neurogenic* disorder)*
Acquired ptosis: Fill ‘er up

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Muscular dystrophy  
CPEO                                                  |
| Neurogenic                          | Horner's  
CNIII palsy                                            |
| Aponeurotic                         | (1)                                                  |
| Mechanical                          |                                                      |
| Traumatic                           |                                                      |
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| **Neurogenic** | Horner's  
CNIII palsy |
| **Aponeurotic** | Aponeurotic dehiscence |
| **Mechanical** |  |
| **Traumatic** |  |

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Muscular dystrophy  
CPEO                                             |
| **Neurogenic**    | Horner's  
CNIII palsy                                    |
| **Aponeurotic**   | Aponeurotic dehiscence                           |
| **Mechanical**    | Large chalazion  
Post-op edema  
BCC/SCC                                                  |
| **Traumatic**     |                                                        |

*Acquired ptosis: Fill ‘er up*

(basal-cell carcinoma/squamous-cell carcinoma)
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**Which is the most common cause of acquired ptosis?**

*Aponeurotic dehiscence, by a mile*
### General categories of ptosis etiology

| Myogenic     | MG  
Muscular dystrophy | CPEO |
|--------------|------|
| Neurogenic   | Horner's  
CNIII palsy |
| Aponeurotic  | Aponeurotic dehiscence |
| Mechanical   | CLarge chalazion  
Post-op edema  
BCC/SC |
| Traumatic    |                  |

### Specific causes of acquired ptosis within each category

Which is the most common cause of acquired ptosis? 
Aponeurotic dehiscence, by a mile

**Acquired ptosis: Fill ‘er up**
### General categories of ptosis etiology

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<th>Mechanical</th>
<th>Traumatic</th>
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<tr>
<td>Post-op edema, BCC/SCC</td>
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**What feared causes must be considered in acquired ptosis secondary to Horner’s?**

- Pancoast tumor
- Carotid dissection
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**What must you be sure to enquire about in any pt presenting with myasthenia?**

Be sure to ask about symptoms related to bulbar weakness (dysphagia, etc).
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### Aponeurotic
- Aponeurotic dehiscence

### Mechanical
- Large chalazion
- Post-op edema
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### Traumatic

**Acquired ptosis: Fill ‘er up**

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#### Acquired ptosis: Fill ‘er up

About 10% of MG pts harbor an occult neoplasm--what is it?

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  - MG
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A thymoma
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A thymoma

What about the reverse? That is, what percent of thymoma pts will have MG?

**Acquired ptosis: Fill ‘er up**
About 10% of MG pts harbor an occult neoplasm—what is it?  
A thymoma

What about the reverse? That is, what percent of thymoma pts will have MG?  
About half

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<tbody>
<tr>
<td>Myasthenia</td>
<td></td>
<td></td>
</tr>
<tr>
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**General categories of ptosis etiology**

Specific causes of **acquired** within each category

- **Myogenic**
  - MG
  - Muscular dystrophy
  - CPEO
- **Neurogenic**
  - Horner's CNIII palsy
  - Aponeurotic dehiscence
- **Mechanical**
  - Large chalazion
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**Q**

*About 10% of MG pts harbor an occult neoplasm--what is it?*
A thymoma

*What about the reverse? That is, what percent of thymoma pts will have MG?*
About half

*What is a thymoma, and where is it located?*

Is MG-associated thymoma usually malignant, or benign?
Benign (although it is malignant in a small number of cases)

How is thymoma diagnosed?
Radiographically, via CXR and/or CT

If an MG pt has a thymoma, what (if anything) should be done about it?
Thymectomy should be considered in select cases
About 10% of MG pts harbor an occult neoplasm--what is it?
A thymoma

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About half

What is a thymoma, and where is it located?
It is a neoplasm of the thymus, which is located in the anterior/superior mediastinum

MG
Muscular dystrophy
CPEO
Horner's
CNIII palsy
Aponeurotic dehiscence
Large chalazion
Post-op edema
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Acquired ptosis: Fill ‘er up
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What sort of organ is the thymus? What is its function?

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- CPEO
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‘Organ in which T cells mature’--how does this dovetail with the pathophysiology of MG?

MG
Muscular dystrophy
CPEO
Homer's CNIII palsy

Acquired ptosis: Fill ‘er up

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‘Organ in which T cells mature’–how does this dovetail with the pathophysiology of MG?
MG is a disease of autoantibodies, which are produced by B cells. However, B-cell autoantibody production in MG is prompted by a T-cell response to ACh-receptor antigens. Thus, MG is fundamentally a T-cell disease.
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**Acquired ptosis: Fill ‘er up**

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**Myogenic**
- MG
- Muscular dystrophy
- CPEO

**Neurogenic**
- Horner’s
- CNIII palsy
- Aponeurotic
- Aponeurotic dehiscence

**Mechanical**
- Large chalazion
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Is MG-associated thymoma usually malignant, or benign?
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How is thymoma diagnosed?
Radiographically, via CXR and/or CT

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Specific causes of acquired ptosis within each category

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- Thyroid
- Hyperthyroidism
- Other thyroid abnormalities
- Yes

About acquired ptosis:

- Fill ‘er up

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