Ptosis and Lid Lag

Ptosis is a term applied to drooping of the eyelid. It can be unilateral or bilateral, complete or incomplete, acquired or congenital. Lid lag means delay in moving the eyelid as the eye moves downwards. It is a common finding in thyroid disease when it is known as Graefe’s sign.

Epidemiology

There is no race or gender predilection. Acquired ptosis usually develops later in life. Congenital ptosis usually presents at birth but is sometimes detected within the first year of life.

Presentation

Ptosis

History

Adults usually notice a change in their appearance and may complain that they always look sleepy and tired. They may also note reduced visual fields. A history should cover the date of onset, any previous trauma or eye surgery, relieving or aggravating surgery and any relevant family or medical history. In children with congenital ptosis, there is a history of unilateral or bilateral drooping eyelid, the onset of which may be elicited by looking at family photographs if available. On examining such photographs, other relatives with ptosis may be identified, in which case a familial cause is highly likely. The history should also enquire about:

- Medication and allergy reactions - eyelid oedema can mimic ptosis.
- Fluctuating ptosis - may indicate myasthenia gravis.
- History of cancer or symptoms indicative of cancer - metastases or primary orbital tumours can cause malposition of the eyelid.
- A history of trauma - a fractured orbit can result in pseudoptosis with enophthalmos. A third nerve palsy may also result in ptosis.
- Headaches - these can result from overactivity of the frontalis muscle as the patient tries to raise the eyelid.

Examination

A limited examination can be conducted in primary care but various measurements can be taken with the eye in primary gaze (defined as the position of the eyes when fixating straight ahead on an object at infinity) including:

- The palpebral fissure (PF) - the distance between the upper and lower eyelid in vertical alignment with the centre of the pupil.
- The marginal reflex distance 1 (MRD-1) - the distance between the centre of the pupillary light reflex and the upper eyelid margin with the eye in primary gaze.
- MRD-2 - the distance between the centre of the pupillary light reflex and the lower eyelid margin with the eye in primary gaze.
- Levator function - the distance the eyelid travels from downgaze to upgaze while the frontalis muscle is held inactive at the brow.
- The margin fold distance (MFD) - the distance from the upper eyelid margin to the fold of skin.

Other features to look out for on examination

These include:

- Abnormalities of tear production.
- Lower eyelid laxity or scleral show.
- Lagophthalmos (difficulty in complete closure of the eyelid over the eyeball).
- Anterior displacement of the globe within the orbit.
- Pseudoptosis, which can result from a number of conditions that alter the appearance of the eyeball (see ‘Differential diagnosis’, below).
- Eyelid retraction - requires exclusion of thyroid orbitopathy.

Lid lag

This is not normally noticed by the patient, but is discovered when a patient with thyroid disease has an eye examination. The feature is elicited by asking the patient to follow with their eyes an object moving slowly from their upper to lower field of vision and vice versa. In lid lag, the upper eyelid lags behind the upper edge of the iris as the eye moves downward. A similar phenomenon is seen with the lower edge, when the eye moves upwards. If the object is moved too quickly, the diagnosis may be missed.

Differential diagnosis
**Differential diagnosis of congenital ptosis**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Diagnosis</th>
<th>Associated Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic progressive external ophthalmoplegia (a disorder characterised by slowly progressive paralysis of the extraocular muscles)</td>
<td>Myasthenia gravis</td>
<td>Acquired myogenic ptosis</td>
</tr>
<tr>
<td>Blepharochalasis (relaxation of the skin of the eyelid, due to atrophy of the intercellular tissue)</td>
<td>Aponeurotic ptosis (the result of a failure of the distal end of the aponeurosis to attach in the usual position)</td>
<td>Neurogenic ptosis</td>
</tr>
<tr>
<td>Mechanical ptosis</td>
<td>Traumatic ptosis</td>
<td>Pseudoptosis - usually caused by alteration in the appearance of the orbit, such as microphthalmos, enophthalmos or orbital fracture</td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>Acquired blepharoptosis (see 'Adult ptosis', below)</td>
<td>Kearns-Sayre syndrome - this triad includes onset in persons younger than 20 years, chronic, progressive, external ophthalmoplegia, pigmentary degeneration of the retina and cardiac problems</td>
</tr>
</tbody>
</table>

There is a long list of conditions which enter the differential diagnosis of acquired ptosis. The main ones are as follows:[4]

<table>
<thead>
<tr>
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<th>Associated Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anophthalmos</td>
<td>Bell's palsy</td>
<td>Benign essential blepharospasm</td>
</tr>
<tr>
<td>Orbital cellulitis</td>
<td>Preseptal cellulitis</td>
<td>Chalazion</td>
</tr>
<tr>
<td>Chronic progressive external ophthalmoplegia</td>
<td>Giant papillary conjunctivitis</td>
<td>Atopic dermatitis</td>
</tr>
<tr>
<td>Duane's syndrome (dysfunction of the eye muscles, normally evident by the age of ten)</td>
<td>Exophthalmos</td>
<td>Capillary haemangioma</td>
</tr>
<tr>
<td>Horner's syndrome</td>
<td>Eyelid laceration</td>
<td>Lyme disease</td>
</tr>
<tr>
<td>Marcus Gunn jaw-winking syndrome</td>
<td>Multiple sclerosis</td>
<td>Myasthenia gravis</td>
</tr>
<tr>
<td>Neuro-ophthalmic history</td>
<td>Neurofibromatosis type 1</td>
<td>Oculomotor nerve palsy</td>
</tr>
<tr>
<td>Apex orbital fracture</td>
<td>Fracture of the floor of the orbit</td>
<td>Undiagnosed congenital ptosis</td>
</tr>
<tr>
<td>Thyroid ophthalmopathy</td>
<td>Uveitis</td>
<td>Cerebrovascular accident (CVA)</td>
</tr>
</tbody>
</table>

**Investigations**[5]

**Congenital ptosis**

Test to exclude myasthenia gravis, such as serum acetylcholine receptor antibody level, edrophonium chloride (Tensilon®) test and electromyography (EMG) should be checked if this condition is suspected.

MRI or CT scanning of the orbit and brain may be helpful in certain circumstances - for example:[1]

- The diagnosis is not evident from the history and examination.
- A history of trauma suggests orbital fracture.
- Other neurological symptoms are suggestive of neurological disease.
- There is a visible or palpable lid mass.
- There is a suspicion of orbital tumours.
- There is new-onset Horner’s syndrome or third cranial nerve palsy, with or without other neurological findings.
- Enophthalmos or proptosis is present with globe displacement.
Other investigations
Depending on the presentation, other relevant tests may be needed, such as mitochondrial assays to exclude mitochondrial disorders of the muscle.

Adult ptosis
- Tests for myasthenia gravis as above.
- ECG, EMG, mitochondrial studies and electoretinogram may be needed to exclude chronic progressive external ophthalmoplegia.
- Consider TFTs for patients with suspected thyroid ophthalmopathy.

Lid lag
TFTs should be performed. CT scan of the orbit may be required in cases of diagnostic difficulty.

Associated diseases

Congenital ptosis
This is often idiopatic but may be associated with:
- Poor visual acuity or strabismus.
- Birth trauma.
- Periorbital tumours.
- Neuroma or neurofibroma.

Neuropathic:
- Horner’s syndrome.
- 3rd nerve lesion.
- Papilloedema.
- Marcus Gunn jaw-winking syndrome - the eye winks when the jaw moves; also known as trigemino-oculomotor synkinesis and thought to be an aberrant connection between the motor branches of the trigeminal nerve and fibres of the superior division of the oculomotor nerve.

Acquired
- Horner’s syndrome - paralysis of the sympathetic nerves, causing unilateral ptosis with ipsilateral constriction of the pupil and sweating.
- 3rd nerve lesion - usually complete ptosis, with ophthalmoplegia with a dilated, unreactive pupil.
- Plexiform neurona - this is associated with neurofibroma type 1.
- Myopathy, myasthenia gravis - the ptosis may be unilateral or bilateral.
- Levator dehiscence (aponeurotic ptosis) - this is due to a dysfunction of the superior rectus and levator muscles. It may be iatrogenic (e.g., damage during suturing), in which case the condition is unilateral, or degenerative due to senility, which is usually bilateral.
- Corneal abrasion.
- Foreign body.

Lid lag
The majority of lid lag is seen in thyroid ophthalmopathy (also known as Graves’ ophthalmopathy). It is seen in 25-50% of adults with Grave’s disease. Other common associated conditions are Graves’ disease in children, hyperthyrotoxicosis and anxiety (all these are reflections of increased adrenergic tone).

Management

Congenital ptosis
- Mild cases may require no initial treatment. The patient should, however, be monitored every 3-4 months for signs which suggest the development of amblyopia (poor or blurry vision, usually in one eye, also called ‘lazy eye’), strabismus, or abnormal head posture.
- Surgical treatment should be considered for patients who start to develop complications. The type of operation depends on the underlying diagnosis and the degree of function of the levator muscle. Options include levator muscle suspension, suspension of the frontalis muscle (tightening of the brow muscles) and, rarely, in congenital cases, the Fasanella-Servat procedure, in which a block of tissue is removed from the underside of the lid. Other options include:
  - Whitnall’s sling - use of Whitnall’s ligament to form a sling.
  - Müller’s procedure (a technique which involves the restiting of Müller’s muscle, an involuntary, sympathetically innervated muscle that originates below the levator aponeurosis just distal to the Whitnall’s ligament).

Adult ptosis
- Underlying disease should be treated (e.g., myasthenia gravis).
Patients who do not wish to undergo surgery may prefer the lid being taped, using Micropore® tape. They should also use lubricants when employing this method, as the blinking mechanism will be less efficient. Alternatively, some patients may wish to use spectacles with a lid attachment which holds up the eyelid.

Surgical options include frontalis slings, levator resection, correction of any abnormality of the levator muscle and the Fasanella-Servat procedure.\[^9\]

Lid lag
This usually does not cause symptoms requiring treatment and may resolve when the patient becomes euthyroid. In severe cases of thyroid eye disease with exophthalmos, lubricants may be required to prevent or treat exposure keratitis.\[^10\]

Complications

**Congenital ptosis**
Complications may include amblyopia, strabismus and abnormal posture of the head (the chin-up position).

**Adult ptosis**
Reduced field of vision and headaches due to tension of the frontalis muscle can occur. Anxiety about appearance may result in social isolation and cause significant psychological complications.

**Lid lag**
There are no significant complications.
Prognosis

**Congenital ptosis**

Surgical correction usually produces excellent results and amblyopia can be avoided or treated successfully if detected early enough.

**Adult ptosis**

- The treatment of myasthenia gravis results in the correction of ptosis in about 70% of patients.
- Excellent results are usually achieved when operative procedures are performed by experienced surgeons.

Further reading & references

- Cohen AE et al; Adult Ptosis., Medscape, Mar 2013
- Sudhaikar P; Upper Eyelid Ptosis Revisited, American Journal of Clinical Medicine,6(3): 2009.
- Hsiao C; Update on Thyroid Eye Disease, Review of Ophthalmology, 2009.

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