

Abnormalities of Eyelid Position and Function

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ABSTRACT

Evaluation of the eyelids is an important part of the neuro-ophthalmic examination. Abnormal eyelid position and function can be caused by disorders involving the third cranial nerve, the oculosympathetic pathway, and the seventh cranial nerve, as well as supranuclear pathways, or as a result of neuromuscular diseases. To avoid unwarranted neurological investigations, it is also important for the clinician to recognize non-neurological eyelid abnormalities (such as ptosis from levator dehiscence or eyelid edema).

Keywords: Eyelids, blepharospasm, ptosis

EYELID ANATOMY

The opposing arches of the upper and lower eyelids frame the ocular fissure. The eyelids are anchored at the medial canthus and lateral orbital tubercle by the canthal tendons. The eyelid *protractor* (e.g., muscle that closes the eye) is the orbicularis oculi: a broad, flat, circumferential muscle innervated by the facial nerve (CN VII) (Fig. 1). The levator palpebrae superioris muscle is the primary *retractor* (raises the eyelid and opens the eye) of the upper eyelid and is innervated by the oculomotor nerve (CN III). This muscle has its origin at the orbital apex and extends anteriorly adjacent and superior to the

superior rectus muscle. The distal portion fans out as the *levator aponeurosis*, which inserts onto the anterior surface of the tarsus and into the upper eyelid skin to form the *eyelid crease*.¹ *Muller's muscle*, a sympathetically innervated muscle, provides additional limited eyelid elevation. It has a short course in the upper eyelid arising from the underside of the levator palpebrae superioris muscle and inserting into the superior border of the tarsus in the upper eyelid (Fig. 2).

The lower eyelid organization is similar to the upper eyelid. The primary retractor of the lower eyelid is the *capsulopalpebral fascia*. This fascial slip extends from the inferior rectus muscle to insert along the infe-

Objectives

On completion of this article the reader will be able to list the differential diagnosis of ptosis and other eyelid abnormalities, including those caused by disorders of the third cranial nerve, the oculosympathetic pathway, the seventh cranial nerve, and the supranuclear pathways as well as the neuromuscular disorders. The reader will also be able to recognize non-neurologic causes of ptosis, and know when to avoid unnecessary neurological tests.

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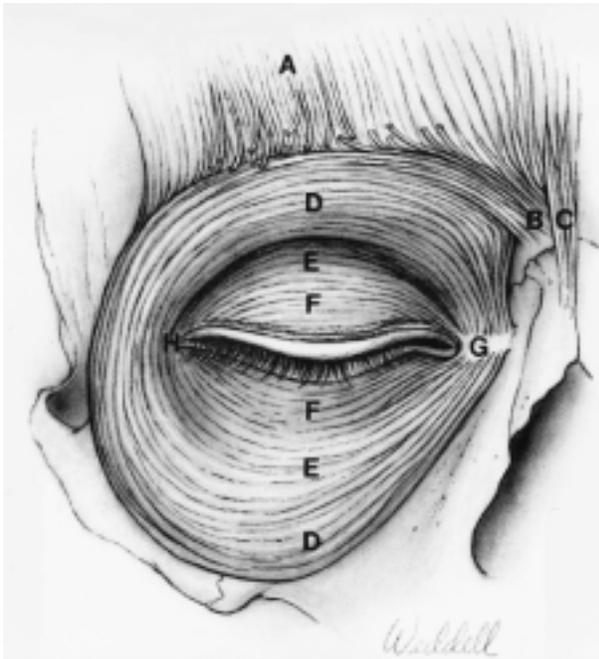


Figure 1. Orbicularis muscle and adjacent musculature. The frontalis muscle (A), superciliary corrugator muscle (B), and procerus muscle (C), are identified. The orbicularis muscle is divided into orbital (D), preseptal (E), and pretarsal (F) portions, and is anchored by the medial (G) and lateral (H) canthal tendons. (Courtesy of Beard C. Ptosis. 3rd ed. St. Louis: The C.V. Mosby Company; 1981, Figure 2.2.)

rior border of the tarsus in the lower eyelid and into the eyelid skin creating the lower eyelid crease. The *inferior tarsal* muscle in the lower lid is sympathetically innervated, and is analogous to Muller's muscle in the upper lid (Fig. 2).

The eyelid can be divided anatomically into an *anterior lamella*, comprised of skin and the orbicularis oculi muscle; *middle lamella*, containing the retractor mechanism of the eyelid and the orbital septum; and the *posterior lamella*, comprised of tarsus and conjunctiva. The orbital septum arises from the periosteum at the orbital rim (arcus marginalis) and compartmentalizes periocular structures into *preseptal* and *postseptal* (orbital) anatomic regions (Fig. 2).

Eyelid anatomy may vary according to race and gender. The upper eyelid crease in women is often higher (a greater distance from the lid margin) than in men.²

The oriental eyelid has a low or absent upper eyelid crease resulting from the absence of fusion of the orbital septum and the levator aponeurosis. Orbital fat (preaponeurotic fat pad) extends inferiorly, anterior to the tarsus. The lower eyelid may also have absent lid crease or a crease close to the origin of the lashes.³

EXAMINATION TECHNIQUES

Table 1 summarizes the important components of examining the eyelids, with more detailed explanations in the text below.

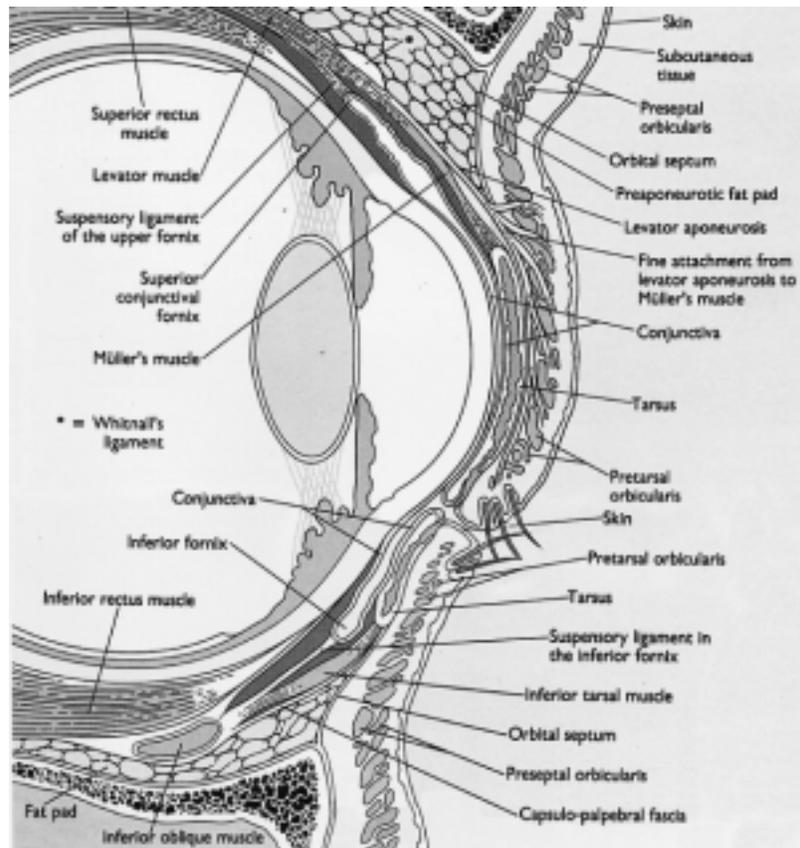


Figure 2. Eyelid anatomy, saggital view. (Courtesy of Tenzel RR. Surgery of acquired lid malpositions. In: Jaffe NS, ed. Atlas of Ophthalmic Surgery. New York: Gower Medical Publishing; 1990, Figures 6.2 and 6.9.)

Table 1. Evaluation of Eyelid Position and Function

<i>Exam Component</i>	<i>Method of Measurement</i>	<i>Examples of Abnormalities</i>
Observe eyelid position	Record as mm of ptosis (or retraction) from the normal position of the lid, or the margin-to-light reflex (MLR), which is the distance of the upper lid from a central cornea light reflex (see Figure 3)	A Horner's syndrome typically causes only 1–2 mm of ptosis; the degree of ptosis may be highly variable in myasthenia
Evaluate levator function	Measure upper eyelid excursion in mm from extreme down-gaze to extreme up-gaze, with the brow fixed by the examiner's hand to prevent any contribution from the frontalis musculature (see Figure 4)	Levator function is normal in levator dehiscence, but usually poor in congenital ptosis, myasthenia, or third cranial nerve palsy
Look for lid lag	Present when there is failure of the upper lid to follow the downward excursion of the eyeball	Common in congenital ptosis, also confirms true lid retraction
Measure lid crease	Distance from the lid margin to the major upper eyelid crease, formed by a portion of the levator insertion	Absent crease or increased distance suggests levator dehiscence
Evaluate orbicularis function; look for lagophthalmos	An inability to fully close the eye suggests weak orbicularis oculi, (or restriction of retractors)	Orbicularis weakness from facial palsies can result in corneal ulceration and blindness
Observe ocular motility	Concurrent motility disorders may suggest a third-nerve palsy, orbital Graves' disease, or myasthenia gravis	
Observe pupils	Anisocoria: ptosis with a small pupil suggests an oculosympathetic paresis (Horner's syndrome), whereas ptosis with a large pupil may occur with third cranial nerve palsies	

RECORDING EYELID POSITION IN PRIMARY POSITION

The normal position of the upper eyelid in primary gaze is about 1 to 2 mm inferior to the superior limbus (the junction of the white sclera and clear cornea). The position of an eyelid can be recorded as the distance in millimeters from this idealized position (*millimeters of ptosis* or retraction). Alternatively, the distance in millimeters from the center of the corneal light reflex and the lid margin, the so-called *margin-light reflex (MLR) distance* can be used also as a measure of eyelid position

(Fig. 3). The lower eyelid normally rests at the inferior limbus. Like the upper lid, the lower lid can be retracted, or even ptotic (*rising* to narrow the fissure).^{4,5}

ASYMMETRIC FISSURE HEIGHT

The distance between the upper and lower eyelid at its greatest vertical height is the *palpebral fissure height*. This measurement has the advantage of being more easily measured than millimeters of ptosis or MLR, but provides less specific information as it does not specify the

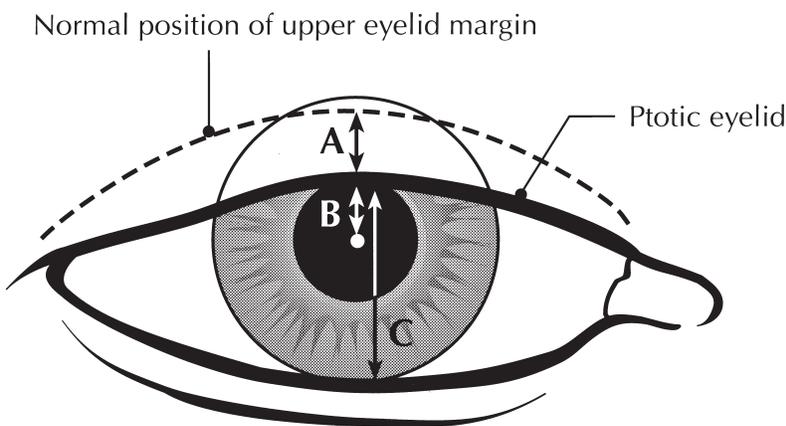


Figure 3. Measuring eyelid position. (A) Millimeters from ideal position. The upper eyelid normally rests about 2 mm below the superior limbus. Measuring from this ideal position, millimeters of ptosis or retraction can be determined. (B) Margin to light reflex distance (MLR). A positive number represents an eyelid above the light reflex, and a negative number below the light reflex. (C) Palpebral fissure height. This measurement is the easiest to perform, but does not identify the actual position of the eyelids on the globe.

Table 2. Causes of Ocular Fissure Abnormalities

Observation	Possible Causes
Ocular fissure too narrow	Ptosis (retractor weakness) Orbicularis overaction ("pseudoptosis")
Ocular fissure too wide	Eyelid retraction (retractor overactivity) Orbicularis weakness

actual position of the eyelid relative to the globe (Fig. 3). The size of the ocular fissure is the net result of forces opening the eye (eyelid retractors), and those closing the eye (eyelid protractors). Thus, an ocular fissure that is abnormally narrow may be the result of a retractor weakness (ptosis), or protractor hyperactivity (orbicularis spasm). A palpebral fissure that is abnormally wide can result from retractor hyperactivity (such as lid retraction in Graves' disease) or protractor weakness (acute facial palsy) (Table 2).

It is not always obvious which eye is abnormal when there is an asymmetry of the palpebral fissures. Does the eye with the smaller fissure have eyelid ptosis, or does the opposite eye have eyelid retraction? Furthermore, ptosis on one side may cause a contralateral lid retraction because the effort generated to lift the ptotic lid is shared bilaterally. This form of lid retraction promptly reverses when the examiner gently lifts the ptotic lid.

MEASURING EYELID FUNCTION

The *function* of the levator palpebrae superioris muscle is determined by the measurement of eyelid excursion from the extremes of down-gaze to up-gaze with the brow immobilized (Fig. 4).

Lagophthalmos is the inability of the lids to close completely. Weakness of the protractor (orbicularis oculi) or restriction of the retractor (levator palpebrae superioris) may cause lagophthalmos. Cranial nerve VII paralysis with weakening of the protractor muscles typically causes a profound lagophthalmos. Poor compliance of the levator muscle such as seen in congenital ptosis, Graves' disease, trauma, or following surgical resection of the levator palpebrae superioris muscle, are other potential causes.⁵

A patient who displays *lid lag* will have a higher resting position of the eyelid in relation to the eye in down-gaze when compared to primary gaze (the eyelid "lags" behind the eye in down-gaze). Lid lag is often seen in Graves' disease, and is the result of the decreased elasticity of the levator muscle secondary to muscle fibrosis or infiltration. Lid lag is also a characteristic of congenital ptosis, but not typical of most forms of acquired ptosis.

OTHER OBSERVATIONS

A high, asymmetric, or absent *lid crease* is a sign of the loss of attachment of the levator aponeurosis to the tarsus and skin (levator dehiscence), or may occur when levator function is poor. Ptosis and a *motility distur-*



Figure 4. Measuring eyelid function. A ruler is placed perpendicular to the central upper eyelid margin, with the thumb or hand placed to immobilize the brow. The full excursion of the eyelid margin from extreme downgaze to extreme upgaze is measured. Less than 10 mm is abnormal.

bance may be observed in oculomotor paresis, myasthenia gravis, chronic progressive external ophthalmoplegia, or Duane's syndrome. Congenital ptosis may be associated with superior rectus dysfunction or Marcus Gunn jaw winking phenomena. A large *pupil* on the side of the ptosis may indicate a CN III palsy or post-traumatic ptosis associated with iris sphincter injury; and a miotic pupil on the side of the ptosis may indicate an ocular sympathetic paresis. A mass in the lacrimal gland may give an "S-shaped" deformity to the eyelid. Inspection of the conjunctival fornices and gross palpation of the orbit for orbital masses should be performed on all patients who present with ptosis.⁶

Protractor function should also be assessed. An incomplete blink or subtle flattening of the nasolabial groove may indicate CN VII weakness. Orbicularis strength can be estimated by having the patient close their eyes tight while the examiner attempts to manually open them. Raising the brows to wrinkle the forehead,

puckering the lips, and smiling to show teeth further test facial motor function.⁵

CAUSES OF ABNORMAL EYELID POSITION AND FUNCTION

PTOSIS

Ptosis, or more precisely *blepharoptosis*, describes an abnormally low resting position of the upper eyelid because of levator insufficiency. The many causes of ptosis are outlined in Table 3, and described in more detail below.^{7,8}

Neuropathic

OCULOMOTOR PALSY. The oculomotor nuclear complex consists of several subnuclei clustered together in the dorsal mesencephalon. Most of the third cranial nerve subnuclei are paired, parasagittal motor nuclei. However, innervation to the right and left levator muscles originates in a single midline subnucleus—the central caudal nucleus. Thus, ptosis occurring from lesions affecting the third nerve nuclear complex will be bilateral. Although most lesions in this area usually affect other third nerve functions, isolated bilateral ptosis can occur.⁹

Fibers serving the levator muscle course with the other components of the oculomotor nerve to the orbit, travelling with the superior division of the oculomotor nerve (with superior rectus fibers) to innervate the levator palpebrae muscle. Ptosis occurring from lesions affecting the oculomotor nerve (such as a posterior communicating aneurysm) is invariably associated with other signs of third nerve dysfunction.

OCULOSYMPATHETIC PARESIS (HORNER'S SYNDROME). The oculosympathetic pathway takes a cir-

cuitous, multisynaptic route from the brain to the orbit, eventually innervating Muller's muscle and the pupillary dilator muscle in the iris. Thus, lesions of the sympathetic pathway produce ptosis and relative miosis. However, given the limited action of Muller's muscle, the ptosis never exceeds 2 mm. The lower eyelid may also be affected, resting at a higher position on the globe than the unaffected eye ("upside-down ptosis").

SUPRANUCLEAR PTOSIS. Rarely, unilateral or bilateral ptosis can be the result of supranuclear lesions. Extensive lesions involving the nondominant (usually right) hemisphere can cause bilateral, often asymmetric cortical ptosis, invariably associated with other neurological dysfunction. Unilateral cortical ptosis is rare and poorly understood, but is reported to occur with focal hemispheric lesions ranging from the frontal to the temporal lobes.

Lesions involving the limbic (extrapyramidal) pathways do not typically affect the resting position of the eyelid, but may cause *apraxia of eyelid opening*.¹⁰ This transient inability to open the eyelids is the result of inhibition of the levator (not orbicularis activation), and can be a component of progressive supranuclear palsy, Parkinson's disease, and other disorders.⁵

SYNKINETIC SYNDROMES. A number of congenital synkinetic syndromes can cause transient ptosis by inhibition of the levator muscle, as outlined in Table 4.

Neuromuscular

OCULAR MYASTHENIA. Myasthenia gravis is a disorder of the neuromuscular junction that commonly causes ptosis and diplopia. The disease may affect the ocular muscles only (ocular myasthenia), or may cause systemic weakness. Clinical characteristics include variable weakness and worsening of symptoms with fatigue. Symptoms may be minimal or absent on arising from sleep, and are usually worse late in the day.

Ptosis is present in many cases. The classic history of an *alternating ptosis*, in which a ptosis "switches sides" should be considered myasthenia until proven otherwise. The ptosis can often be seen to worsen during the examination, especially with prolonged up-gaze (Fig. 5). *Cogan's lid twitch sign* is often present, but is not entirely specific for myasthenia. This finding is evoked by a rapid saccade from down-gaze to primary position, and consists of an initial overshoot of the upper eyelid, with slow downward return. Orbicularis oculi and other facial muscle weakness is common.

In some cases, variability is only evident between examinations, underscoring the importance of careful and accurate measurement of eyelid position and ocular motility on each encounter. Examinations later in the day, when the patient is more fatigued, are usually more productive than early morning visits. Because myasthenia can produce virtually any motility pattern and/or ptosis, it should be included in the differential diagnosis of every patient with ptosis and ocular motility disorders.⁵

The short-acting anticholinesterase drug *edrophonium chloride* (Tensilon®, ICN Pharmaceuticals, Inc., Costa Mesa, CA) is used in a diagnostic test for myasthenia gravis—the "Tensilon test." The agent only lasts a

Table 3. Differential Diagnosis of Acquired Ptosis

Neuropathic
Oculomotor nucleus/fascicles/nerve palsy
Oculosympathetic paresis
Supranuclear ptosis
Neuromuscular junction
Myasthenia Gravis
Botulism
Myogenic
Myotonic dystrophy
CPEO
Aponeurotic (levator dehiscence)
Involitional ptosis ("senile" ptosis)
Contact lens wear
Postedema
Trauma
Mechanical
Dermatochalasis
Eyelid tumors
"Pseudoptosis"
Orbicularis activation (blepharospasm, etc.)
Globe retraction
Duane's retraction syndrome
Enophthalmos

Table 4. Synkinetic Syndromes Affecting the Palpebral Fissure

Observation	Mechanism	Disorder	Precipitating Movement	Cause
Palpebral fissure narrows	Inhibition of levator	Paradoxical levator inhibition	With adduction or abduction	Congenital oculomotor miswiring
		Inverse Marcus Gunn phenomenon	Movement of jaw to opposite side (external pterygoid)	Trigeminal-oculomotor synkinesis
	Activation of orbicularis	Aberrant regeneration of the facial nerve	Facial movement	Aberrant facial motors fibers innervating orbicularis muscle
Palpebral fissure widens	Activation of levator	Paradoxical levator excitation	With adduction or abduction	Congenital oculomotor miswiring
		Marcus Gunn phenomenon	Movement of jaw to opposite side (external pterygoid)	Trigeminal-oculomotor synkinesis

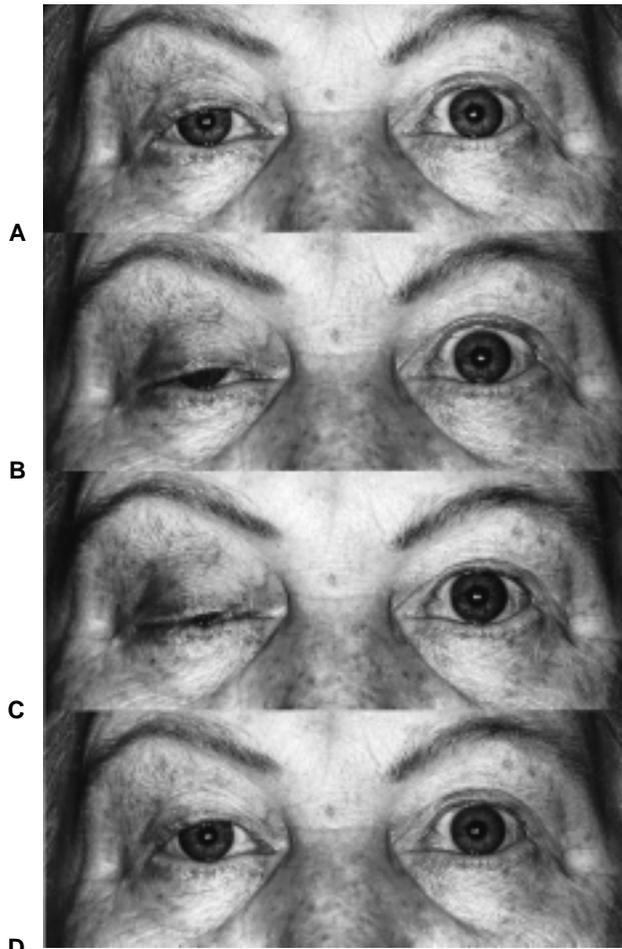


Figure 5. Variable ptosis with ocular myasthenia. A 64-year-old woman presented with ptosis of the right upper lid. (A) Ptosis at the start of the examination. (B) After sustained up-gaze, the amount of ptosis has increased. (C) With additional fatigue, the ptosis is nearly complete. (D) After resting with her eyes closed for 5 min, the amount of ptosis is much less.

matter of minutes, so the motility disturbance or ptosis needs to be significant enough to allow confident appraisal of improvement over just a few minutes of observation. Reversal of ptosis is easily observed, but motility patterns may be more difficult to assess, especially if subtle.

BOTULISM. Botulism is a neurological disorder caused by ingestion of food contaminated with *Clostridium botulinum*. This bacterium elaborates botulinum toxin, a powerful neurotoxin that blocks the release of acetylcholine at the neuromuscular junction and destroys nerve endings. Ocular signs include ptosis, ophthalmoparesis, and dilated pupils. Systemic symptoms include dizziness, headache, dysphagia, and weakness. Surprisingly, only a third of patients with ingestion of botulinum toxin have gastrointestinal symptoms.⁵

CPEO AND MUSCULAR DYSTROPHIES. Chronic progressive external ophthalmoplegia (CPEO) is clinical designation describing symmetric, slowly progressive bilateral ptosis and limitation of eye movements. This clinical entity can occur as the result of a number of different disorders, most of which are heritable, and many that are associated with systemic signs and symptoms.

Mitochondrial myopathies are a group of systemic disorders that cause muscular weakness and CPEO as a result of mitochondrial dysfunction. In addition to ptosis and ophthalmoplegia, mitochondrial myopathies frequently demonstrate weakness of facial and systemic muscles, cardiac conduction abnormalities, pigmentary retinopathies, and other systemic signs. Kearns-Sayre syndrome is a type of mitochondrial myopathy presenting as CPEO that is recognized as a distinct syndrome, often diagnosed before age 20. It is caused by a *mitochondrial* chromosomal defect, and is associated with cardiac conduction defects (including heart block and sudden death).

Myotonic dystrophy is a systemic myopathy that is a rare cause of bilateral ptosis and limited ocular motility. Additional ocular signs include a characteristic "polychromatic" cataract and macular and retinal

pigmentary changes. Electromyography is diagnostic, demonstrating characteristic myotonic discharges (“dive-bomber” discharges). Visible (and palpable) wasting of the temporalis muscles is characteristic.⁵

Oculopharyngeal dystrophy is an heritable (autosomal dominant) condition affecting patients of French-Canadian ancestry in their fifth and sixth decade, causing progressive ptosis, limited ocular motility, and weakness of the bulbar musculature with dysphagia.

Congenital Ptosis

Congenital ptosis refers to a developmental abnormality of the levator palpebrae superioris muscle, which results in ptosis. Ptosis may be unilateral or bilateral and may occur sporadically or may be a component of an inherited syndrome (e.g., the autosomal dominant blepharophimosis syndrome).

Congenital ptosis may be mild (2 mm), moderate (3 mm), or severe (>4 mm). The more severe the ptosis, the less striated muscle is present to lift the eyelid, and the poorer the levator function. A severe congenital ptosis is commonly associated with fair to poor levator function (eyelid excursion of 6 mm or less), and a poor eyelid crease.⁸

In congenital ptosis the levator palpebrae superioris muscle contains more fibroblasts than muscle fibers, resulting in poor contraction and relaxation. Therefore, lid lag in down-gaze is a characteristic feature of congenital ptosis, not present in most acquired forms of ptosis.⁷ The presence of lid lag also explains why children with either unilateral or bilateral congenital ptosis develop a head position (with chin up) to maintain unobstructed vision.

Involitional Ptosis

Involitional ptosis (also called “senile” ptosis) is the result of age-related changes in the levator aponeurosis or levator muscle, and is the most common type of ptosis encountered in clinical practice.⁸

Aging and other factors can cause the levator aponeurosis to become stretched, thinned, or detached from its insertion at the tarsus and skin of the upper lid. This results in ptosis, but with normal levator function (excursion). The upper eyelid crease is typically elevated or absent, reflecting loss of the levator insertion.⁷ Other factors such as contact lens wear,¹¹ recurrent eyelid edema, topical steroid use, and trauma are other common causes of levator dehiscence that may occur in younger patients^{12,13} (Fig. 6).

Surgery is generally successful in correcting involitional ptosis, and is often used when the eyelid blocks the superior visual field or for cosmetic reasons.

Other Causes

Blunt orbital trauma can cause a profound ptosis, but will often spontaneously resolve. Periocular trauma that results in a horizontal laceration of the eyelid fre-

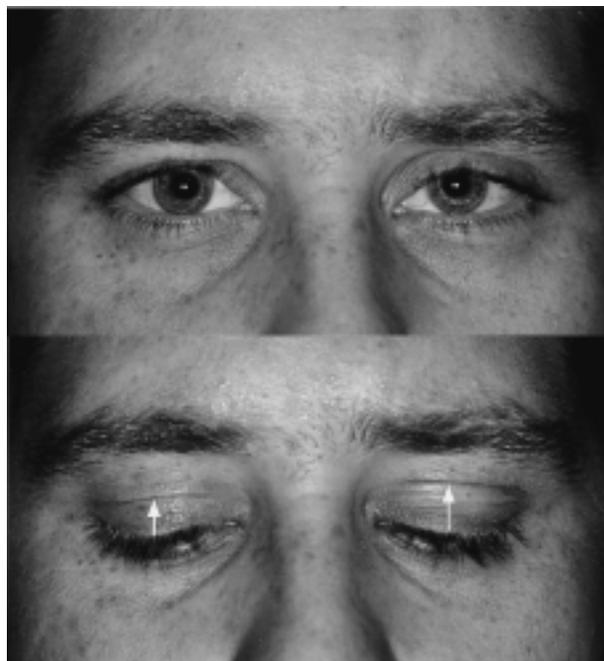


Figure 6. Elevated lid crease in aponeurosis dehiscence. A 27-year-old man was referred to evaluate a possible neurological cause for unilateral ptosis. There was no associated motility disturbance (to suggest a third-nerve paresis), anisocoria (to suggest a Horner's syndrome), or variability (suggesting myasthenia). The patient was a contact lens wearer for many years. The presence of an elevated eyelid crease (pictured) with normal levator function confirmed levator dehiscence as the cause of ptosis. (Courtesy of Martin TJ, Corbett JJ. Requisites in Neuro-ophthalmology. St. Louis: Mosby; 2000, Figure 7.8.)

quently involves the levator aponeurosis, and may require a surgeon experienced in eyelid repair.

Periocular tumors cause ptosis by a mechanical means or by affecting levator muscle function. A plexiform neurofibroma, for example, may mechanically limit lid excursion by its sheer bulk or may infiltrate the levator muscle, reducing its function. Contour abnormalities are often seen with tumors extrinsic to the levator muscle and limit eyelid excursion in sectorial fashion.

Pseudoptosis

Patients with marked dermatochalsis may have prolapse of the skin of the upper lid over the eyelid margin, simulating a ptosis (Fig. 7).

Globe retraction such as observed in Duane's syndrome or traumatic enophthalmos may be misinterpreted as ptosis. In these cases, the palpebral fissure is narrowed due to retraction of the globe into the orbit.

Patients who feign ptosis (functional or nonorganic ptosis) will demonstrate an increase in orbicularis tone affecting the upper and lower lids that cannot be sustained indefinitely.

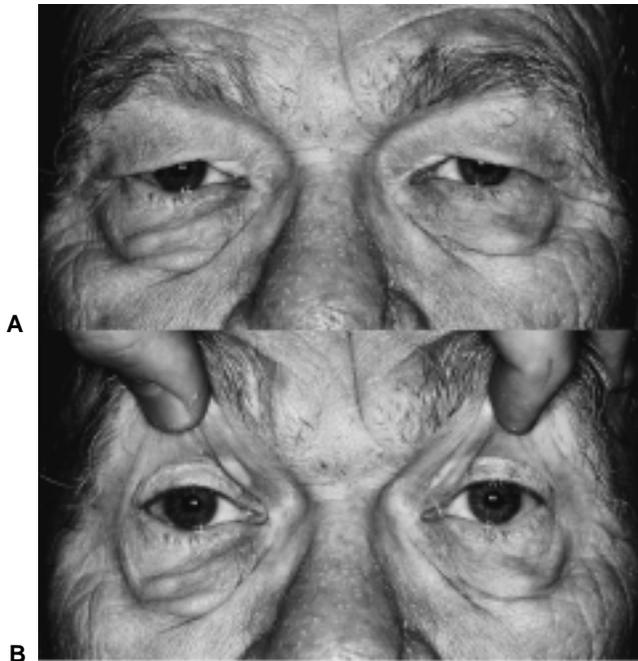


Figure 7. Pseudoptosis from dermatochalasis. (A) This 62-year-old man appears to have narrowed ocular fissures. However, this appearance is the result of loose skin from the upper eyelids prolapsing over the eyelid margin. (B) When the skin of the upper eyelids is gently retracted, the normal position of the upper eyelid margin can be appreciated.

ORBICULARIS OVERACTION: FACIAL NERVE HYPERACTIVITY

Lesions involving the facial nerve or its supranuclear pathways can produce irritative states causing overactivity, including blepharospasm, hemifacial spasm, and other abnormal activation.^{14,15} Orbicularis activation narrows the ocular fissure, but can usually be distinguished from ptosis as increased orbicularis tone in the upper and lower lids is usually clinically evident.

Blepharospasm

Benign essential blepharospasm (BEB) is an idiopathic condition in which there is intermittent spasm of the orbicularis oculi bilaterally, thought to be related to dysfunction of the basal ganglia and extrapyramidal (limbic) system. These unwanted spasms of eye closure can severely disable patients, making activities such as driving, ambulating, and reading difficult or impossible. This condition is most common in patients in the fourth through sixth decades.¹⁶ When blepharospasm is associated with other facial movements, such as grimacing or torti-retrocollis, the condition is called *Meige syndrome*.¹⁷

Secondary blepharospasm can be caused by ocular surface disease or intraocular inflammation, and rarely from meningeal irritation associated with intracranial lesions. Blepharospasm can be a manifestation of tardive dyskinesia, resulting from neuroleptic drugs, and can oc-

cur in extrapyramidal disorders such as Parkinson's disease, Huntington's chorea, and basal ganglia infarction.

Local injection of botulinum toxin (Botox®, Allergan, Inc., Irvine, CA) into the orbicularis oculi to weaken the muscle is an effective treatment for blepharospasm. Repeat injections are necessary, as the effect lasts only several months at best.^{18,19} Surgical procedures to weaken the facial nerve or to remove the orbicularis oculi muscle are complex and fraught with complications.

Hemifacial Spasm

Hemifacial spasm is intermittent spasm involving the upper and lower face on one side. Facial nerve function is otherwise normal. This condition is most likely caused by irritation and intermittent compression of the facial nerve by the anterior inferior cerebellar artery or other branches of the basilar artery in the subarachnoid space.²⁰ Patients with typical, isolated, long-standing hemifacial spasm may not require neuroimaging. However, patients with hemifacial spasm who also have facial weakness must have neuroimaging (preferably magnetic resonance imaging with GAD), as this combination of signs is suggestive of a compressive lesion such as a cerebellopontine angle tumor.²¹

Hemifacial spasm can usually be distinguished from facial tics, as tics can be suppressed voluntarily for a period of time and typically begin in childhood. Focal epilepsy involving the face is rare and is typically followed by postictal facial paralysis (Todd's paralysis).

Medical treatment of hemifacial spasm has included baclofen, carbamazepine (Tegretol®, Ciba-Geigy Corp., Summit, NJ), clonazepam, and more recently Neurontin, but these medications are rarely helpful. As with blepharospasm, botulinum toxin injection is often effective.²² Neurosurgical exploration of the root of the facial nerve and placement of a sponge between the facial nerve and impinging vascular structures has been successful in many cases, but the surgical risks of suboccipital craniotomy (including stroke and deafness) must be considered.^{21,23}

Facial Myokymia

Unlike the intermittent spasms of hemifacial spasm, facial myokymia is a continuous, unilateral, undulating movement that often appears to ripple across the face. This disorder may be isolated to the orbicularis oculi muscles initially, but over time involves all of the unilateral facial muscles, and is occasionally bilateral.²⁴ In advanced cases, the myokymia progresses to a continuous tonic facial contracture, associated with facial paralysis. This is called *spastic-paretic facial contracture*.

Facial myokymia is generally caused by lesions immediately rostral to or involving the facial nucleus and its fascicles within the pons. Common causes include multiple sclerosis in adults, and pontine glioma in children or young adults. Facial myokymia has also been associated with extra-medullary brain stem compression, brain stem infarction and infection, Guillain-Barré syn-

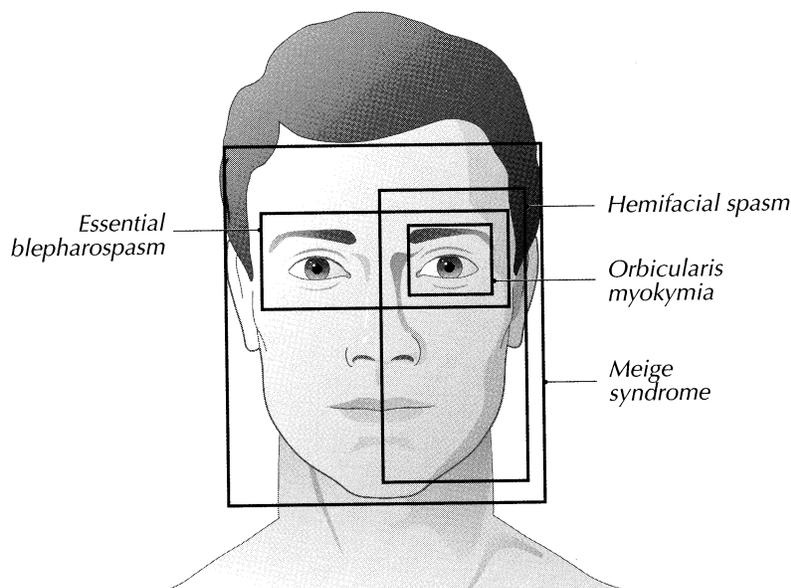


Figure 8. Facial involvement in hyperactivity states. Regions of the face commonly involved in facial hyperactivity states are shown. (Courtesy of Martin TJ, Corbett JJ. *Requisites in Neuro-ophthalmology*, St. Louis: Mosby; 2000, Figure 12.7.)

drome, toxins, anoxia, and obstructive hydrocephalus. Neuroimaging is absolutely required in all patients with facial myokymia. In most cases, the underlying cause cannot be definitively removed. Carbamazepine (Tegretol®), baclofen, or clonazepam may offer limited symptomatic relief.^{5,24}

Benign Orbicularis Myokymia

Benign orbicularis myokymia is a common complaint in otherwise normal individuals, consisting of an irritating unilateral twitching movement of the lower (or less often, upper) eyelids. The twitching is rapid and episodic, lasting hours or even days at a time, and does not involve any other portions of the face. Associated factors include stress, fatigue, as well as caffeine and nicotine intake. This benign disorder can be distinguished from hemifacial spasm and blepharospasm by the lack of involvement of any other portion of the face or opposite eye, and the absence of any other neurological or neuro-ophthalmic findings (Fig. 8).

Following Facial Nerve Palsy

Aberrant regeneration of the facial nerve frequently occurs following facial nerve injury. Facial nerve fibers that originally were directed to the lower face may be misdirected to the orbicularis, resulting in a narrowing of the fissure with movements of the lips or mouth (Table 4).

Generalized increased facial tone commonly occurs following recovery from Bell's palsy, resulting in a narrowing of the ocular fissure on the affected side.

EYELID RETRACTION

Eyelid retraction refers to an abnormally high position of the upper lid, giving the appearance of “staring.”

Retraction of the lower lid may also contribute to this appearance. Lid retraction may give the false impression of exophthalmos, and patients may present with a complaint that their “eye has gotten bigger.” Causes of eyelid retraction are outlined in Table 5, and described in more detail below.

Orbital Graves' Disease

Orbital Graves' disease is an autoimmune disorder characterized by enlargement of the extra-ocular muscles and an increase in orbital fat volume, causing proptosis, diplopia, and ocular congestion (Fig. 9). Eyelid retraction is a conspicuous sign, and occurs independently of the proptosis by an unknown mechanism.

Eyelid retraction can occur from other disorders,²⁵ but is often specific for Graves' disease when accompanied by proptosis and restrictive orbitopathy. Not uncommonly, lid retraction may be the first and only visible sign of orbital Graves' disease. Lid lag, or failure of the eyelid to follow with the globe in down-gaze, is usually present with lid retraction.

The combination of proptosis, lid retraction, and lid lag can produce an exposure keratopathy, causing blurred vision and pain. The severity of ocular surface disease dictates the intensity of treatment, and may include artificial tears, ointment, moisture shields, taping

Table 5. Differential Diagnosis of Eyelid Retraction

Graves' disease (unilateral or bilateral)
Midbrain disorders (bilateral, but may be asymmetric)
Aberrant regeneration of the oculomotor nerve
Pseudo-retraction
from contralateral ptosis
from ipsilateral facial palsy (acute)
Levator restriction from trauma or surgery



Figure 9. Eyelid retraction in Graves disease. This 45-year-old woman with orbital Graves disease has bilateral lid retraction and proptosis.

the eye closed at night, tarsorrhaphy, or levator recession surgery.

About 5% of patients with Graves' disease also have myasthenia gravis. The coexistence of multiple autoimmune disorders is not uncommon, and suggests an underlying fundamental immune derangement. Therefore, patients with orbital Graves' disease can actually present with ptosis, rather than lid retraction.

The orbit and thyroid gland are end-organs affected by a poorly understood underlying systemic autoimmune disorder. Therefore, thyroid dysfunction is not invariably present with Graves' orbitopathy and normal thyroid function does not rule out this entity.

Supranuclear Disorders

Lesions involving the dorsal midbrain can cause symmetric bilateral eyelid retraction (called *Collier's sign*), as part of a constellation of signs called *dorsal midbrain syndrome* or *Parinaud's syndrome*. Other commonly associated findings include convergence-retraction nystagmus, pupillary light-near dissociation, vertical gaze dysfunction, and convergence abnormalities.⁵ This form of lid retraction can be distinguished from the lid retraction in Graves' disease, as the eyelids follow the globe normally in down-gaze (no lid lag). If a dorsal midbrain lesion also affects the oculomotor fascicles on one side, the patient can present with lid retraction on one side, and ptosis on the other (called "plus-minus" syndrome).²⁶

Aberrant Regeneration of the Oculomotor Nerve

Aberrant regeneration of the third cranial nerve often involves the eyelid. Not uncommonly, eyelid *retraction* will occur with attempted adduction and/or depression of the globe, as fibers originally innervating the medial or inferior rectus now innervate the levator muscle. This important finding will be missed unless the examiner specifically looks for eyelid elevation in the infe-

rior positions of gaze (without holding the eyelids). Other synkinetic syndromes are discussed in Table 4.

Mechanical

Mechanical factors that restrict the normal compliance of the levator palpebrae or other eyelid components typically cause lid lag, but may cause lid retraction when severe. Examples include metastatic schirrous carcinoma of the breast and scarring from lacerations, burns, or orbital infections.

Pseudo-retraction from Contralateral Ptosis

The levator palpebrae muscles in the right and left eyes are *yoked muscles*, operating with equal innervation according to Hering's law. The extra effort generated to lift a partially ptotic lid on one side can give the appearance of eyelid retraction on the opposite side. This pseudo-retraction will disappear when the ptotic eyelid is manually elevated.

ORBICULARIS WEAKNESS

Unilateral orbicularis weakness may cause an asymmetry of the ocular fissures in primary position at rest, but is usually manifest clinically as an incomplete blink or poor eye closure.

Facial Nerve Palsies

BELL'S PALSY. Bell's palsy is the most common facial neuropathy. This condition has always been described as an idiopathic facial paralysis in the past, but recent evidence suggests a viral (herpes simplex) etiology.²⁷⁻²⁹

Facial weakness in Bell's palsy generally develops quite rapidly. Pain (usually retroauricular) is common

and may antedate the onset of paralysis by hours or days. Additional signs and symptoms include facial numbness, numbness of the tongue, decreased tearing, altered taste, and dysacusis. Orbicularis weakness is often profound, and the resultant exposure keratopathy can have serious consequences.

Greater than 80% of patients have a “satisfactory” recovery. Improvement usually begins within 3 weeks of onset. By 3 to 4 months, the degree of recovery is usually complete, and improvement is not expected beyond this point. The treatment recommendations for Bell’s palsy continue to evolve, and currently include the use of antiviral agents.¹⁴

OTHER CAUSES OF FACIAL NERVE PALSY. Infectious diseases that can cause facial nerve palsy include Herpes Zoster oticus, Lyme disease, human immunodeficiency virus (HIV). Guillian-Barré syndrome (acute inflammatory demyelinating polyradiculopathy, or AIDP), sarcoidosis, head trauma, and tumors (cerebellopontine angle tumors, parotid tumors) commonly involve the facial nerve.²⁷

Möbius’ syndrome is a congenital disorder of nuclear agenesis that consists of bilateral facial palsies, gaze palsies, and multiple other potential cranial neuropathies.

As in Bell’s palsy, treatment of the potentially devastating complications of orbicularis weakness—exposure keratopathy and its sequelle—must be addressed.

Myopathic Diseases and Myasthenia Gravis

Disorders that cause myopathic facial weakness (such as myotonic dystrophy, CPEO, or other muscular dystrophies) and myasthenia gravis can also cause weakness of eyelid closure. Interestingly, most of these same disorders are characterized by ptosis from weakness of eyelid retractors. Orbicularis weakness may only be evident when the patient is asked to tightly close their eyes. Patients with myasthenia may be able to initiate tight closure of the eyelids, but with fatigue weakness of the orbicularis muscle becomes evident (the so-called “peek sign”).³⁰

EXPOSURE KERATOPATHY FROM ORBICULARIS WEAKNESS

Preventing the potentially serious sequelae of exposure keratopathy is vital in caring for patients with poor eye closure. Exposure keratopathy leads to epithelial breakdown with corneal erosion, and the potential for corneal ulceration, corneal perforation, or endophthalmitis.

The degree of *orbicularis weakness* is the primary determinant of corneal exposure risk. Another important factor is the presence and effectiveness of the *Bell’s phenomenon*. This reflexive upper rotation of the eyes on eye closure may protect the cornea even when the eye cannot be completely closed. The presence of relative *corneal anesthesia* adds considerable risk for the development of keratopathy in the setting of orbicularis weakness. Specifically evaluating these three factors:

the degree of orbicularis weakness, the Bell’s phenomenon, and corneal sensation, along with the anticipated recovery time, determine how aggressive the treatment must be to protect the eye. All patients at risk for exposure keratopathy should be started on topical artificial tears at regular intervals throughout the day and ointment at night. Prompt referral to an ophthalmologist is required in most cases, especially those patients with profound orbicularis weakness or those with an anticipated chronic course. Tarsorrhaphy, injection of the levator palpebrae muscle with botulinum toxin, placement of gold weights in the upper eyelid, or other therapies may be necessary.

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