

Clinical Evaluation of Ptosis

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History

All ptosis patients presenting with either unilateral droopy eyelid or bilateral droopy eyelids need a thorough examination that includes a medical history, a family history, a history of drug or allergic reactions, and a review of systems

Family photographs can help determine onset or variability of the ptosis. Providing photographs also gives the surgeon a chance to examine the other family members. A patient with a strong family history of congenital ptosis may not need an extensive workup.

- In severe cases where surgery is needed, historical emphasis should be placed on any anticoagulant use or bleeding disorder to avoid potential complications during surgery. The surgeon should also inquire about a family history of malignant hyperthermia and cardiac disorders. Patients with ptosis and Kearns-Sayre syndrome or chronic progressive external ophthalmoplegia may also have a cardiac conduction disorder.
- A history of fluctuating ptosis with strabismus may indicate myasthenia gravis.
- A careful medical history regarding cancer should be obtained. Metastatic or primary orbital tumors can result in malpositioning of the eyelid.
- A history of trauma with orbital wall fractures can result in pseudoptosis with enophthalmos. Also, third cranial nerve palsy from trauma may result in ptosis.

- A history of drug or allergic reactions may be helpful. Allergic reactions can result in eyelid edema and droopy eyelid.
- A history of difference in the size of the pupil may be helpful in diagnosing Horner syndrome. Patients with Horner syndrome will have ptosis and miosis on the same side. Cervical or apical thoracic tumors can cause damage to the sympathetic chain and result in this condition. Neuroblastoma, which is one of the most common childhood cancers, should be ruled out.
- A history of dry eyes, intermittent epiphora, or chronic conjunctivitis can indicate a dry eye disorder or corneal surface disease.



Role of Old Family Photographs: This lady in the top most photograph had come with complaints of ptosis in the right eye for the last 2 years. After proper clinical evaluation our diagnosis was a mild form of congenital ptosis. The patients guardians were pretty convinced that it had been present for only a few years, so we asked for her old album photographs. If you note carefully it is quite obvious in the bottom-left photograph, as well as in her childhood photo (at the bottom-right), which the parents had tried to mask by applying lots of eye liner.

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Physical Examination

All ptosis patients presenting with either unilateral droopy eyelid or bilateral droopy eyelids need a thorough physical evaluation.

- Visual acuity: Careful vision testing is performed using an age-appropriate method. In the past, ptosis alone was not considered to produce amblyopia, and other associated factors (eg, anisometropia, strabismus) were always thought to be the cause. However, recent studies have documented that amblyopia is possible with an isolated ptosis. This problem should be searched for and treated as necessary.
- Orthoptic evaluation: Look for associated problems (eg, double elevator palsy) or other more common forms of strabismus. If indicated, muscle surgery can be performed at the same time as ptosis surgery.



Ptosis with Nv III palsy

- Visual field: Obtain visual field tests in patients who are able to cooperate in order to document peripheral and superior visual field restriction.
- Slit lamp examination: Include slit lamp examination, intraocular pressure measurement, and fundus examination in the preoperative evaluation.
- Refraction: A cycloplegic refraction is indicated in all children with ptosis, since a significant number of them have anisometropia primarily due to astigmatism on the ptotic side. Correct any significant refractive error.
- Tear function testing: In adults, obtain a measure of basal tear secretion by performing a Schirmer test of the anesthetized eye. In addition, evaluate the corneal tear film for evidence of abnormal debris or tear breakup.

- Corneal sensitivity should be tested if possible. This may be a difficult test in young pediatric patients.
- An exophthalmometer can be used to assess relative proptosis or enophthalmos of each eye. In pseudoptosis, a proptosis of the contralateral eye gives the false impression that the normal upper eyelid is droopy.
- Pupil: Pupillary reaction, direct, consensual and accommodative reflex should be checked in all cases. The pupillary size and the iris color differences between the eyes should be examined for Horner syndrome.
- The patient should be examined for Bell's phenomenon. Bell's phenomenon is a normal defence reflex present in about 75% of the population, resulting in upward and outward turning of the globes when blinking.

It becomes noticeable only when the orbicularis muscle becomes weak as in, for example, facial palsy.

It is, however, present behind forcibly closed lids in most healthy people and should be assessed prior to ptosis surgery.

The patient closes both eyes tightly as the examiner holds the upper and lower lids apart. If the globe elevates during the forced lid closure, a normal Bell phenomenon is present. This evaluation can help the surgeon to determine the risk of exposure keratopathy following the eyelid surgery.



Bell's phenomenon Assessment

Absent Bells is a contraindication to Ptosis surgery, as the post operative lagophthalmos may give rise to corneal exposure with resultant exposure keratopathy.

Absent Bells Phenomenon: Bells is almost absent or very weak in CPEO. Surgical intervention for ptosis in this type of situation may cause exposure keratopathy.



Bell's Phenomenon Absent

- Fluctuating Ptosis: Fluctuation in the amount of Ptosis at different times of the day is highly suggestive of Myasthenia gravis



Fluctuating Ptosis: This boy's parents complained of variable and fluctuating ptosis. We asked the parents to take multiple photographs at different times of the day. When we observed the photographs carefully, the ptosis was changing and varying in amount among the two eyes at different times of the day. It is almost pathognomic of myasthenia gravis.

Ptosis measurements

Simple observation:

- Begin with simple inspection of the patient. Observe for any associated head posture, tilt or chin elevation. Longstanding head posture can give rise to torticollis. The parents need to be counsel led to opt for early surgical intervention.



Head Tilt: This boy has a ptosis of the right eye, Note the head tilt that has developed.



Chin Elevation: Bilateral congenital ptosis in this child has resulted in a compensatory chin elevated posture. The boy lifts his chin to keep his visual axis clear in primary position.

- Observe the lid level relative to the globe and to the other lid. The presence or absence of a lid fold or lid crease gives a significant clue to the degree of levator function. An absent lid crease is often accompanied by poor levator function. If a lid crease is present but is higher than normal (a normal lid crease is 8-10 mm from the lid margin measured above the pupil) and if a deeper upper lid sulcus is found on that side, note these as signs of a levator aponeurosis disinsertion. When the patient is asked to look up, both the sulcus and the lid crease may move superiorly slightly before the lid moves. This is caused by a delay in levator action due to the attenuation or stretching of the normal aponeurotic attachments to the tarsus. Measure and record the position of both upper lid creases.



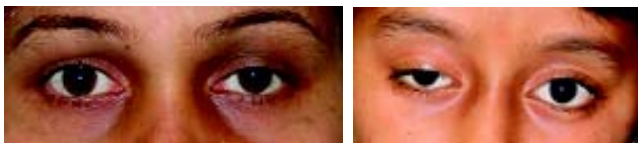
Elevated Superior Lid Crease: Right sided senile ptosis. The superior lid crease is elevated to almost the superior orbital rim. Compare the level of the lid crease with the normal left sided non ptotic eye. The lid in the non ptotic eye is at a slightly higher level than what should be normal, this is due to the over action of the frontalis (the folds on the forehead) to lift the ptotic eye, resulting in compensatory elevation of the contralateral eye.



Absence of Lid Crease: This child has marked ptosis, in the left eye covering the papillary axis. There is almost absence of presence of the superior lid crease, a feature very characteristic of Congenital Ptosis.

Upper lid height:

- This is a measure of the amount of ptosis in the primary position with the patient's brows relaxed. The upper limbus can be used as a baseline from which to estimate the amount of ptosis. The upper lid normally crosses the cornea approximately 0.5-2 mm below the upper limbus, and under normal situations, it maintains this approximate relation in upward and downward gaze. The cornea is about 11 mm in height; therefore, in a patient with 3.5 mm of ptosis, the lid margin splits the visual axis, assuming the measurement from the visual axis to the upper limbus is 5.5 mm (2 mm below the upper limbus normal position + 3.5 mm ptosis = 5.5 mm).



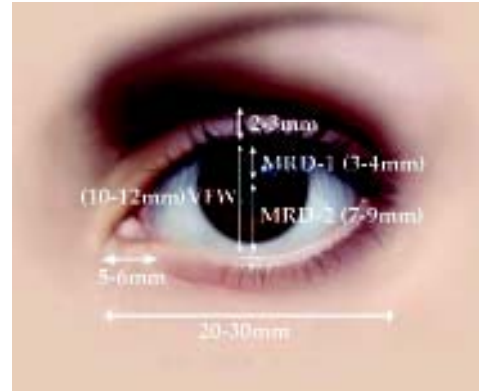
Mild Ptosis

Moderate Ptosis



Severe Ptosis

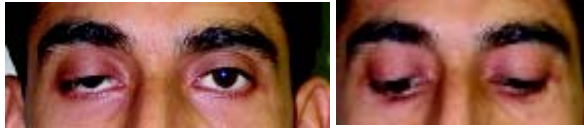
- Employing the corneal light reflex and its distance to the lid margin, keeping the above relationships in mind, also can provide a relatively precise estimation. For instance, if the lid margin appears to be approximately 1.5 mm above the corneal light reflex, 2-3 mm of ptosis is present. Ptosis is considered mild if 1-2 mm, moderate if 2-3 mm, and severe if 4 mm or more.



Average measurements of superficial anatomy

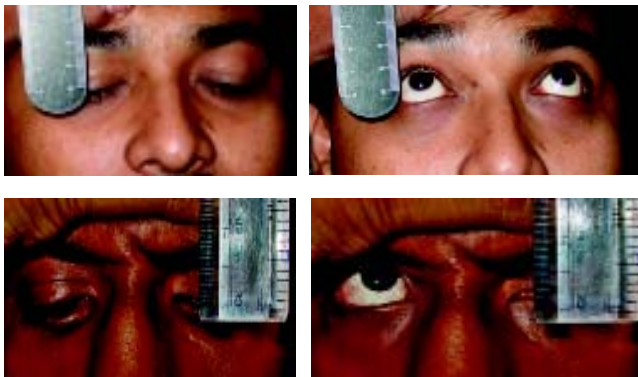
Levator function:

- Ptosis can be subdivided on the basis of levator function into:
 - Severe (LPS action <5mm),
 - Moderate (LPS action 6-10mm),
 - Mild (LPS action >10mm).
- Evaluation of the levator function is extremely important and begins with evaluation of the lid crease, as noted above. Presence of a crease suggests that some levator function exists even if the lid moves poorly. If the examiner everts the lid and it does not flip back to its normal position when the patient is asked to look up (known as Iliff sign), poor levator action is certain. With the exception of patients of Asian ancestry, in whom the lid crease may not be present, an absent lid crease usually indicates absent levator function.
- In patients with abnormal levator function, the amplitude of action of the muscle is diminished and the lid fails to elevate and depress normally, producing a lid lag in downgaze. The involved lid may be higher than the uninvolved one in downgaze. This lid lag is exaggerated by ptosis surgery and hence must be discussed in advance with the patient or parents.



Lid Lag : This gentleman has a congenital ptosis right side as is evident by absence of lid crease on primary position. When he is asked to look downwards, the LPS fails to depress normally, producing a lid lag in downgaze. The involved lid seems to be higher than the uninvolved one in downgaze

- In a patient with ptosis due to a dehiscence of the levator aponeurosis, the levator muscle is normal, its function generally is quite good, and the amount of ptosis remains the same in both upgaze and downgaze. This simple relationship allows the surgeon to readily differentiate true congenital ptosis from early-acquired ptosis, congenital ptosis associated with birth injury, and other forms of ptosis in which the levator muscle is normal. Although aponeurotic defects have been reported in congenital ptosis, these are not typical.
- The levator function is determined by holding the brow immobile, placing a millimeter ruler over the lid in the plane of the pupil, and measuring the levator excursion from extreme downgaze to extreme upgaze. Levator function may be classified as poor levator function (<5 mm), fair levator function (6-9 mm), and good levator function (>10 mm). Generally, in patients with congenital ptosis, mild ptosis usually is accompanied by good levator function, and moderate-to-marked ptosis is associated with fair-to-poor levator function. The measurements of the degree of ptosis and degree of levator function require the cooperation of the patient; usually a child must be aged 2-3 years before this cooperation can be obtained.



LPS (Levator Palpabrae Superioris) function Test

Additional observations:

- The lid contour, lashes, and skin are also evaluated. The presence of jaw winking (Marcus Gunn phenomenon) can be assessed by asking the child to move the jaw from side to side or to chew or by allowing the infant to nurse under observation. In adults, redundant skin and ptosis of the brow may mask a true ptosis or produce ptosis due to mechanical factors. Test orbicularis oculi muscle function, corneal sensation, and Bell phenomenon. Palpation of the lids and orbits is important because it may reveal a mass not otherwise appreciable as the cause of acquired ptosis.

Marcuc-Gunn Jaw-winking Phenomenon: Marcuc-Gunn Jawwinking in the left side following opening of mouth



- Careful external examination along with palpation of the eyelids and the orbital rim should be performed. A lid mass can cause extra weight in the lid, resulting in ptosis. Plexiform neuromas, lymphoma, or leukemia can result in an eyelid mass. Rhabdomyosarcoma may present with a mass that is palpable through the lid.

Laboratory Studies

- If myasthenia gravis is suspected, check serum acetylcholine receptor antibody levels.

Imaging Studies

- The following are indications to perform neuroimaging studies (eg, MRI, CT) of the orbit and brain:
 - History not consistent and onset not clear
 - Other neurologic findings along with ptosis
 - Orbital wall fracture suspected with history of trauma
 - Visible or palpable lid mass
 - Orbital tumors (eg, lymphoma, leukemia, rhabdomyosarcoma) suspected

- New onset of Horner syndrome with or without other neurologic findings
- New onset third cranial nerve palsy with or without other neurologic findings
- Globe displacement with either enophthalmos or proptosis
- In patients with acute ptosis or if any suspicion of an orbital process, sulcus filling, or exophthalmos exists.

Other Tests (MH)

- If myasthenia gravis is suspected, the following tests are recommended:
- Single fiber electromyography (EMG)
- Tensilon test
- Ice test
- If a mitochondrial disorder is suspected, an ECG is recommended.

Procedures (MH)

- If a mitochondrial disorder is suspected, a muscle biopsy should be performed.

Eyelid Measurements

Test	Measurement	Normal Value
VFW	palpebral fissure vertical	10 mm
VFW (down gaze)	palpebral fissure vertical in downgaze	2-4 mm
MRD1	light reflex to upper lid margin	4 mm
MRD2	light reflex to lower lid margin	>5 mm
LPS Action	upper lid margin from down gaze to upgaze	12-18 mm
MCD	on down gaze lid margin to crease	4-5 mm
lag	lagophthalmos	0 mm

Eyelid Measurements

The **vertical fissure width** is the distance between the upper and lower eyelid in vertical alignment with the center of the pupil.

The **marginal reflex distance-1** (MRD-1) is the distance between the center of the pupillary light reflex and the upper eyelid margin with the eye in primary gaze. A measurement of greater than 2.5 mm is considered normal.

The **marginal reflex distance-2** (MRD-2) is the distance between the center of the pupillary light reflex and the lower eyelid margin with the eye in primary gaze.

The **marginal crease distance**(MCD) is the distance from the upper eyelid margin to the lid crease on down gaze.

In white women, a central measurement of 10-11 mm is considered normal, and in white men, 8-10 mm is considered normal.

The **Levator Palpabrae Superioris** (LPS) Action. is measured by measuring the excursion of lid from down to up gaze after immobilizing the frontalis with firm pressure by thumb on the brow.

About the Author

Dr Arnab Biswas, MS DO, FRCS, consultant Oculoplastic and Orbital Surgeon at Alo Eye Care & B. B. Eye Foundation & Kolkata, underwent the International Council of Ophthalmology Fellowship in Oculoplasty & Orbital diseases from University of Limoges, France. He is the author of a comprehensive and elaborative "Atlas of Oculoplastic and Orbital Surgery".