## **Review Article**

## Management of Blepharoptosis update - A Review

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#### Abstract

Blepharoptosis, or ptosis of the eyelid, refers to drooping of the upper eyelid. The condition is cosmetically noticeable even in the earliest stages .It usually results from a congenital or acquired abnormality of the muscles that elevate the eyelid. Ptosis may be the presenting sign or symptom of a serious neurologic disease. Regardless of the etiology, when ptosis obstructs vision, it is disabling. The appropriate management requires recognition of the underlying cause. we discuss the clinical approaches to the management of ptosis .This review article highlights the various aspects of ptosis evaluation and management.

# Keywords: Blepharoptosis, levator muscle, Muller's muscle, Fasanella-Servat procedure, Frontalis sling surgery.

#### Introduction

Blepharoptosis is defined as drooping or inferior displacement of upper evelid when the eye is in the primary position. It is a common clinical sign that may affect individuals of all ages rang. it might be severe in that the pupils are completely covered causing visual disturbances. Apart from the cosmetic concern, patients with ptosis suffer from problems in the quality of vision. Although the superior visual field is most often involved, central vision can also be affected. Many patients with ptosis report difficulty with reading because the ptosis worsens in downgaze. Ptosis has also been shown to decrease the overall amount of light reaching the macula and therefore, can reduce visual acuity, especially at night.<sup>1</sup> In children, ptosis is of a major concern as it can cause amblyopia. Ptosis can be an early sign of a life-threatening disease. Early diagnosis and proper evaluation are the important factors aiding treatment of this condition which can improve the quality of life of the people affected.

#### **Anatomical considerations**

Normally, the upper eyelid is positioned 1 to 2 mm below the upper corneal limbus, and the lower eyelid is placed at the sclerocorneal junction. The muscles concerned with elevation of upper eyelid are levator palpebrae superioris (LPS), Muller's muscle, and frontalis.<sup>2</sup> LPS is the major muscle involved which is innervated by the oculomotor nerve (third cranial nerve). The levator originates from the lesser wing of the sphenoid bone. As it traverses the orbit, it broadens and becomes a fibrous aponeurosis that inserts on the anterior aspect of the tarsal plate. The upper eyelid skin crease is formed by attachments of the aponeurosis to the orbicularis muscle and skin.<sup>3</sup> Muller>s muscle is a smooth muscle that arises from the undersurface of the levator and inserts into the superior tarsus. The Muller's muscle contributes 1-2 mm of eyelid elevation. Muller's is under sympathetic control and when gets fatigued or dysfunctional, leads to mild ptosis, for example, Horner's syndrome. The frontalis muscle lift the brows, and it is innervated by 7<sup>th</sup> cranial nerve.

#### Classification

Ptosis can be classified as congenital and acquired. According to the etiology, it can be further classified as myogenic, aponeurotic, neurogenic, neuromuscular, mechanical, traumatic, and pseudoptosis.

#### Myogenic ptosis

Myogenic ptosis occurs when the muscles elevating the lid are dysfunctional, mainly the levator. Most commonly seen as congenital ptosis where there is LPS dysgenesis. This can occur either as autosomal dominant or as sporadic. Congenital myogenic ptosis is usually unilateral. LPS function may be severely affected in these cases, and they present with a lid lag on down gaze as the LPS fails to relax.<sup>4</sup> Some of them will have associated ipsilateral superior rectus muscle weakness, and some others may have associated craniofacial syndromes such as blepharophimosis syndrome and Marcus Gunn jawwinking syndrome.<sup>4</sup>

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Blepharophimosis-ptosis-epicanthus inversus syndrome (BPES) is an autosomal dominant condition characterized by severe bilateral congenital ptosis associated with telecanthus, epicanthus inversus, hypoplasia of the superior orbital rims, horizontal shortening of the eyelids, ear deformities, hypertelorism, and hypoplasia of the nasal bridge.

Marcus Gunn Jaw winking is caused by a miscommunication between the third cranial nerve that innervates the levator and the fifth cranial nerve that innervates the muscles of mastication. The result is a unilateral ptosis that improves when the patient opens the mouth or moves the lower jaw in a contralateral direction.

The causes of myogenic ptosis in adults include oculopharyngeal dystrophy, chronic progressive external ophthalmoplegia (CPEO), myotonic dystrophy, and infiltrative ptosis.<sup>5</sup>

Oculopharyngeal dystrophy patients will have ptosis and associated difficulty in swallowing due to the weakness of oropharyngeal muscles.<sup>5</sup> CPEO presents with bilaterally symmetric ptosis with ophthalmoplegia without any diplopia and may have associated retinal pigmentary abnormalities and cardiac conduction defects. Myotonic dystrophy has an autosomal dominant inheritance and the classical features are ptosis, orbicularis weakness, and extraocular muscle weakness and may also be associated with cardiac conduction defects. Myogenic ptosis can also be caused by infiltrative processes such as amyloidosis which can affect levator as well as extraocular muscles and biopsy of the muscles confirms the diagnosis. <sup>5</sup>

Myasthenia gravis is a neuromuscular junction disorder with autoantibodies against acetylcholine receptor causing destruction of them. Ocular myasthenia presents with unilateral or bilateral ptosis with the involvement of extraocular muscles, levator muscle, orbicularis muscle, and without any systemic features. The ptosis usually worsens with fatigue and improves with the ice test and edrophonium test. Other signs include ptosis that worsens in the evening, paradoxical eyelid retraction , and Cogan's lid twitch, which is eyelid retraction that occurs during upgaze after sustained downgaze.<sup>4</sup>

#### **Aponeurotic ptosis**

The most common etiology of blepharoptosis is aponeurotic. The levator aponeurosis transmits levator force to the eyelid. Thus any disruption in its anatomy or function can lead to ptosis. It is due to stretching, attenuation, dehiscence or detachment of LPS aponeurosis from its tarsal attachments. 3,4 Usually, bilaterally symmetric ptosis, but can be unilateral and asymmetric also. They will have normal LPS function, normal ocular movements, normal pupils, high lid crease and deep upper lid sulcus. Stretching of aponeurosis commonly occurs in patients with constant contact lens use and chronic eye rubbing as in ocular allergy/Down's syndrome.<sup>4,6</sup> Many patients after cataract surgery can develop aponeurotic ptosis.<sup>5</sup> It is thought to be due to trauma to the LPS and superior rectus complex which has strong intermuscular fascial connections to the levator muscle and can be disrupted either by postcataract eyelid swelling or by the eyelid speculum used to separate the eyelids at the time of surgery.

## **Neurogenic ptosis**

Third nerve palsy and Horner's syndrome are the important causes of neurogenic ptosis. Third nerve palsy can result from vascular/inflammatory/neurotoxic or compressive etiology. It presents with ptosis, extraocular muscle involvement sparing the lateral rectus and superior oblique, down and out eyeball and with or without pupillary involvement(Mydriasis).<sup>4</sup>

Horner's syndrome is due to damage to the sympathetic supply to the eye from tumors, aneurysms, or inflammations. It results in ptosis, anhidrosis, miosis, apparent enophthalmos, and decrease pigmentation of iris on the involved side. The ptosis in this case is usually mild with good LPS function. <sup>4,5</sup>

## **Traumatic ptosis**

Traumatic ptosis may not deserve its own category. Trauma can damage LPS muscle/aponeurosis/neural input which results in myogenic/aponeurotic/neurogenic ptosis.

# **Mechanical ptosis**

The causes of mechanical ptosis include excessive skin overhanging the eyelid and mass or scar that weigh down the upper lid. It may be caused by congenital abnormality, such as plexiform neurofibroma or hemangioma or by acquired such as a large chalazion or skin carcinoma. The correction of ptosis in this regard is by correcting the underlying cause first, and the residual ptosis can be corrected by levator advancement in patients with good LPS function

## **Pseudoptosis**

Conditions which mimic ptosis include enophthalmos, anophthalmos, dermatochalasis, hypotropia, contralateral lid retraction, contralateral proptosis, facial nerve palsy, superior sulcus deformity, and brow ptosis. Proper examination should be done to differentiate these conditions from that of true ptosis.<sup>3</sup>

#### **Clinical management**

#### History

A proper history is always an important tool for diagnosis. Apart from the usual ophthalmic and medical history, the points that should be emphasized in a case of ptosis are: Management of Blepharoptosis update - A Review

- Is one or are both eyes affected?
- Are there associated symptoms, specifically pain, malaise, visual disturbance, diplopia, dysphagia or muscle weakness elsewhere?
- What was the speed of onset, the duration and extent of progression of the ptosis?
- Does the ptosis fluctuate?
- Are there any obvious relieving and exacerbating factors?
- Does the patient have any comorbidities?
- In particular, are there vascular risk factors, a history of injury to the head, neck or chest, a history of HIV or other cause of immunosuppression, features of the metabolic syndrome, cancer or ocular disease?
- Are there any systemic features of giant cell arteritis?
- Is there a history of trauma, ophthalmic surgery or rubbing of the eyelid?
- Does the patient wear contact lenses?
- Has the patient had a blepharoplasty in the past?
- Is the patient taking any medications, regular or new?
- Is there a family history of ptosis or of other muscle weakness?

#### Examination

The examination of a patient with ptosis should be proper and should aim at confirmation of the diagnosis and decision of treatment. The proper examination of ptosis involves elicitation of signs, lid measurements, and ancillary tests.

#### Signs

- Examination starts from the moment a patient enters the room head posture and face turn if any should be noted. Chin-up position is the most commonly encountered posture
- Frontalis overaction: patient may compensate the ptosis by lifting eyebrows with the frontalis muscle
- External examination including the palpation of eyelids and orbital rim should be done
- Features suggestive of BPES: Telecanthus, epicanthus inversus, hypoplasia of the superior orbital rims, horizontal shortening of the eyelids, ear deformities, hypertelorism, and hypoplasia of the nasal bridge<sup>5</sup>
- Cigarette paper appearance of lid skin due to recurrent edema occurs in blepharochalasis

- Proptosis or enophthalmos should be ruled out which may contribute to pseudoptosis. Exophthalmometry with Hertel's exophthalmometer is relevant in this regard
- Strabismus if present should be evaluated. Cover/uncover test should be done in all cases of ptosis. It should be noted that hypertropia can mimic ptosis
- The presence of lagophthalmos should be assessed-ptosis surgery can cause worsening of the same
- Bell's phenomenon: Grading should be done. Good bell's is always needed for the health of cornea after surgery
- Lid retraction in other eye should be checked for which can make the impression of ptosis in the affected eye
- Lid position in downgaze: Lid lag in downgaze (higher position of upper eyelid in downgaze) in the absence of trauma/surgery is suggestive of dysgenesis of levator muscle (commonly in congenital ptosis) since the dysgenetic muscle is not able to relax properly<sup>7</sup>
- Synkinesis: The variation of the amount of ptosis with jaw movements is seen in Marcus Gunn jaw-winking ptosis, and variation in ptosis with ocular movements is noted in aberrant regeneration of the oculomotor nerve or the facial nerve, and some types of Duane's retraction syndrome
- Extraocular movements affected in CPEO, myasthenia as well as in the third nerve palsy
- Best corrected visual acuity and cycloplegic refraction should be done especially in children to asses amblyopia and visual problems
- Anterior segment examination with special emphasis on pupils: pupils affected in Horner's, third nerve palsy etc.
- Posterior segment examination: Example, abnormal retinal pigmentation seen in Kearn-Sayre syndrome
- Cogan's lid twitch: Elicited by having the patient look in downgaze, followed by upgaze. As the affected eye saccades up, the upper lid overshoots. Seen in myasthenia<sup>7</sup>
- Herring's law : Hering's law of equal innervation states that the reciprocal eye muscle of each eye is innervated equally. As such, manual elevation of the more ptotic eyelid decreases the muscle strength required to keep the lid elevated, and so the contralateral LPS relaxes and causes ptosis in the other eye.

## Measurements

- Margin-reflex distance 1 (MRD1): The distance between the central corneal light reflex and upper eyelid margin with eyes in primary position. Normal MRD 1 is 4–5 mm
- Margin-reflex distance 2 (MRD2): The distance between the central corneal light reflex and lower eyelid margin with eyes in primary position
- Palpebral fissure height (PFH): It is the distance between the upper and lower eyelid margins at the axis of the pupil. The sum of the MRD1 and the MRD2 should equal the vertical PFH 6,7
- Levator function: Berke's method estimated by measuring the upper eyelid excursion, from downgaze to upgaze with frontalis muscle function negated and with the positioned in the frontal or Frankfort plane. The amount of lid elevation is recorded in millimeters (mm) of levator function. The classification of levator function:

Poor: 0–4 mm lid elevation Fair: 5–11 mm lid elevation Good: 12–14 mm lid elevation Normal: >15 mm lid elevation.

- Iliff test: It is used to assess levator function in infants. Upper eyelid of the child is everted as the child looks down. If the levator action is good, lid reverts on its own.
- Margin crease distance: Upper eyelid crease position is the distance from the upper eyelid crease to the eyelid margin. It is normally 7–8 mm in males and 9–10 mm in females. High skin crease suggests aponeurotic defect. The depth of skin crease is a guide to determine the levator function in young children
- Margin limbal distance (MLD) : Putterman's method-measurement of the distance between the middle of upper eyelid margin to the 6 o'clock limbus in extreme upgaze. Normal is about 9 mm
- Pretarsal show: It is an important aspect of finding out the symmetry of eyelids. It is the distance between the lid margin and the skin fold with the eyes in primary position.

#### Tests

• Fatigue test: MRD1 should be measured first. Then the patient should be asked to look up for 2 min after which the MRD 1 is to be measured again. Worsening of ptosis is seen in myopathies, myasthenia as well as senile aponeurotic ptosis

- Ice test: Glove containing ice pack is applied on the closed ptotic eye for 2 min. If the lid elevates by 2 mm or more, it is suggestive of myasthenia
- Tensilon test: In cases of suspected myasthenia, 2 mg of edrophonium is injected slowly in 15– 30 s. The needle is left in situ, and the remaining 8 mg is injected slowly if no adverse reaction is observed within 1 min. If myasthenia is the cause, ptosis improves after the injection 2
- Phenylephrine test: Sympathomimetic agents, such as phenylephrine or apraclonidine, can be instilled under the eyelid to test the function of Muller>s muscle
- Schirmer's test: To evaluate tear function
- Tear breakup time in individuals suspected of having dry eyes due to the potential risk of incomplete eyelid closure and exposure keratopathy following surgical correction
- Corneal sensitivity should be tested in all cases.

## Treatment

In children, when there is risk of amblyopia, immediate surgery is needed; otherwise, if there is no risk of amblyopia, we can wait up to 3–5 years of age when we can get more accurate measurements.

#### Fasanella-Servat procedure

Upper border of the tarsus is excised with lower part of Muller's muscle and the overlying conjunctiva. Indications are mild congenital or acquired ptosis with good levator function, Horner's syndrome and minor contour adjustment after any ptosis surgery.<sup>3</sup>

#### Procedure

- A stab incision is to be made in the lid crease in the most lateral part
- Evert the lid and clamp over the upper attached border of the tarsus and lower part of Muller's muscle with overlying conjunctiva
- Pass one needle of double-armed 6-0 catgut suture through the everted lid medial to lateral just below the clamp as a continuous suture 8
- Remove the clamp and cut along the crush marks using scissors
- Use the other arm of the absorbable suture to approximate the cut edges of the wound
- Pass both arms of the suture through the conjunctival wound in the lateral part of the lid and through the stab incision in the skin, tie them without tension and bury the knot under the skin

- Bandage Contact lens is placed for 1 week.
- Complications include corneal abrasion, foreign body sensation, and skin crease lowering.<sup>9</sup>

#### **Muller's muscle-conjunctival resection**

This involves the excision of Muller's muscle and overlying conjunctiva with reattachment of resected edges. This procedure can achieve a maximal elevation of  $2-3 \text{ mm.}^{10}$ 

Indication-Mild ptosis with good levator function (Figure-1 ) a and (Figure-1 ) b. Usually done in Horner's syndrome and mild congenital ptosis.<sup>11</sup>



Figure 1: (a) Mild aponeurotic ptosis - Left eye (b) Postoperative picture following Muller's muscle conjunctival resection - Left eye

#### Levator advancement

A levator advancement procedure can be employed in all cases of aponeurotic ptosis. For patients with aponeurosis dehiscence or disinsertion and good levator function, surgery can be done by anterior/posterior approach. For adults, local anesthesia is recommended to be able to adjust lid height with the patient's cooperation.



In the aponeurosis surgery by anterior approach, the aponeurosis is approached through a skin incision, it is advanced and sutured to the tarsus, or the aponeurotic defect is repaired directly.<sup>6</sup> Then, the lid contour is assessed with the patient sitting up, once the eyelid height and contour have been verified and symmetry is created, the sutures are trimmed when the desired height is achieved. Excess skin is excised, and skin crease is formed with sutures. This technique is more used when

there is aponeurotic weakness with excess skin to be excised<sup>12</sup> (Figure-2) a and (Figure-2) b.

Figure 2: (a) Aponeurotic ptosis –Left eye (b) Postoperative picture following levator advancement surgery- Left eye

Complications include lid level too high or low, contour abnormalities, asymmetrical skin crease and upper lid show.<sup>1</sup>

#### Levator resection

It can be done in congenital as well acquired ptosis. Indications: Levator function  $> 4 \text{ mm.}^{14}$ 

In the conventional approach, a 20 mm sized horizontal incision is made on the skin, orbicularis dissected; orbital septum opened and under the pad of fat, levator aponeurosis is identified. Then the muscle is resected at the indented height and subsequently advanced up to the middle of the tarsal plate and secured with sutures. Lid crease forming sutures are made. The amount of resection needed is preoperatively determined by the amount of ptosis and the level of LPS function by Burke's method (Table-1). The lid margin is placed at the superior corneal limbus when levator muscle function is poor to fair, and a postoperative fall of several millimeters is expected. With fair to good function, the lid is placed 2-3 mm below the limbus and no postoperative fall is expected. Such intraoperative adjustment is a useful addition to the quantitative estimation made preoperatively. However, when epinephrine is present in the local, there will be some Muller's muscle stimulation so that a postoperative fall of 1-2 mm is typical. In adult patients, adjustable sutures can be put, which allows postoperative manipulation<sup>15</sup> (Figure-3) a , (Figure-3)b and (Figure-3)c.



Figure 3 : (a) Aponeurotic ptosis - Both eye L > R (b) Levator resection - Left eye

#### (c) Levator resection - Both eye

Table 1: The amount of levator muscle to be resected according to the grade of LPS function(Burke's method) and the severity of ptosis Journal of Z H Sikder Women's Medical college

Ptosis (mm)	Levator muscle	Levator muscle
	function(mm)	reection(mm)
Mild (<2	>10 mm	Small (10-13 mm)
mm)		
Moderate (3	>8 mm	Moderate (14-18 mm)
mm)	< 8 mm	Large (18-22 mm)
Severe (>4	< 5 mm	Maximum ( > 23
mm)		mm )

# Whitnall Sling

The Whitnall sling procedure involves maximal levator resection, up to Whitnall's ligament, combined with suturing of the superior tarsal edge to Whitnall's ligament with either absorbable or non-absorbable sutures. It can be used in cases of moderate to severe ptosis with poor to fair levator function.<sup>16,17</sup> A superior tarsectomy can be performed as well, with an estimated 1 to 1.5 mm of additional lid elevation expected from a 5-mm resection.<sup>3</sup> This is often recommended as cases of isolated Whitnall sling have been reported to have a high incidence of late undercorrection.<sup>16</sup> However, patients with maximal or supermaximal levator resection commonly have significant lagophthalmos in the immediate postoperative period and must be counseled on frequent lubrication.<sup>17</sup>

# **Frontalis sling**

The eyelid is suspended directly from the frontalis muscle so that movement of the eyebrow is efficienctly transmitted to the eyelid. This eyelid elevation by frontalis is enhanced by connecting frontalis and eyebrow to the upper eyelid using a subcutaneous sling. This surgery is used in patients with ptosis having poor LPS function and good frontalis action. Used primarily in the congenital ptosis with poor levator function, neurogenic ptosis (Figure-4) a, (Figure-4) b, (Figure-4) c, myogenic ptosis and blepharophimosis syndrome.<sup>3,5</sup> Skin incisions are placed at the tarsus and eyebrow in a fox pentagon pattern to insert the sling material in the suborbicularis plane. The material is placed anterior to the orbital septal plane to raise the eyelid towards the eyebrow. A "hood" can also be formed under the pretarsal and preseptal skin, delaying eyelid lowering in downgaze.



(a)



(b)



(c)

(d)



Figure 4: (a) Left eye- congenital ptosis, (b) Postoperative ptosis correction by prolene

(c) Right eye- congenital ptosis (d) S/P frontalis sling surgery by fascia lata.

Sling materials can be autogenous (fascia lata, temporalis fascia)<sup>18</sup> or nonautogenous. Nonautogenous can be integratable (Mersilene mesh, Gortex) and nonintegratable (prolene, silicone).<sup>19</sup> The patient should be at least 3-year-old for sufficient leg size for removing an appropriate fascia. Autogenous fascia lata is the best sling material, and the disadvantage is that it has to be harvested. Hence, it is not used in very young children (<3 years) and very old or debilitated persons. There is less risk of infection, absorption or rupture as compared to the foreign materials.

Whenever possible, all the ptosis surgeries should be performed under local anesthesia to allow for the precise adjustment of lid height and contour. In cooperative children,local anesthesia can be used in children as early as 12 years of age.20

# **Modified Tarsal Resection**

An alternative for patients with fair levator function is a modified tarsal resection with graded levator advancement, as described by Sarit Patel, MD, and colleagues.<sup>21</sup> From an anterior approach, the levator aponeurosis is exposed and disinserted from the tarsus, then separated from Müller muscle. The anterior face of the tarsus is exposed. A predetermined amount of tarsus and Müller (in a 1:2 ratio) to be excised is marked, depending on the amount of ptosis, then tarsus, Müller muscle and conjunctiva is resected en bloc. The inferior edge of Müller muscle and conjunctiva is then advanced to the new superior tarsal border with absorbable sutures. Levator aponeurosis is then advanced to the anterior face of the tarsus and adjusted to set the upper lid margin at the superior corneal limbus.All patients had mild lagophthalmos in the immediate postoperative period only , with good results in 25 of 30 patients(30 of 36 lid).<sup>21</sup>

## **Complications of Ptosis Surgery**

Complications of the surgery include under correction, overcorrection, corneal exposure, infection, sling granuloma (Figure-5)a, conjunctival prolapse (Figure-5) b and notching of the eyelid.22



Figure 5: (a) Sling granuloma on forehead (b) Conjunctival prolapse on left eye

#### Conclusion

Ptosis repair is a challenging oculoplastic surgical procedure that requires correct diagnosis, thoughtful planning, experience and good surgical technique. The clinical examination is easily understood once the underlying is known. To avoid unnecessary investigations, it is useful to try to determine whether the cause is likely to be structural, myogenic, neurogenic, neuromuscular or central. A directed history and examination often allows the clinician to localise the cause of the ptosis to a particular anatomical area, if not to reach a specific diagnosis. This in turn permits appropriate investigations and treatment to be instituted.

The management of ptosis is very much dependent on the underlying cause but oculoplastic surgery should be considered for patients with static lesions. Proper evaluation is necessary in deciding the correct management of ptosis. The choice of treatment is mainly dependent on the amount of ptosis and level of LPS function. The accurate measurement and a meticulous surgery will always give you good results and a happy patient.

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