Paediatric ptosis

BY PARTH SHAH, ANGELOS SINAPIS AND MANOJ PARULEKAR

Manoj Parulekar and colleagues provide a comprehensive overview of the diagnosis, assessment and management of childhood ptosis.

Blepharoptosis (commonly referred to as ptosis – Greek, πτῶσις, 'to fall') is a condition where the upper eyelid is in an abnormally low position in primary gaze.

The prevalence of childhood ptosis is estimated at 1:10,000 [1]. The aetiopathogenesis and management are quite distinct from adult onset ptosis. The effects of paediatric ptosis are:

- Amblyopia: deprivation or refractive (anisometropia, astigmatism)
- Cosmetic disfigurement
- Altered head posture.

Clinical assessment

The clinical assessment of paediatric ptosis focuses on severity and functional impact (visual development) and identification of the underlying cause. These factors will guide decision-making for management.

History

A directed history from the parents includes:

- Time course: when the ptosis was first noticed, and change over time. Early post-natal photos are helpful for comparison
- Birth trauma
- Variation in lid position when feeding (Marcus-Gunn jaw-winking syndrome) or variability through the day
- Family history of congenital ptosis
- Maternal myasthenia gravis
- Is the eye open or shut when the child is asleep?
- Is the eye more than 50% closed, i.e. lid covering the pupil – if so, what proportion of waking hours is the pupil covered?

Clinical examination

- Visual function: age-appropriate uni-ocular visual acuity measurements are obtained to assess for amblyopia. In younger children, objection to occlusion and forced preferential looking tests are used
- Observation
  - Unilateral or bilateral involvement
  - Abnormal (chin-up) head posture (Figure 1)
  - Features of blepharophimosis syndrome
  - Signs of birth trauma
  - Brow over action
  - Periocular fullness: suggestive of mechanical cause.
- Assessment of the severity of ptosis when the child is fully awake:
  - Hirschberg corneal light reflex is of critical importance in young infants to assess whether or not the visual axis is clear.
  - Estimate of palpebral aperture and MRD1 measurements.
  - Presence and depth of upper eyelid crease: a very useful surrogate for levator function.
  - Estimate of levator function: can be challenging to perform reliably in young infants (Figure 2).
- Jaw-winking: observe for changes in lid height when child sucks on a pacifier or bottle (Figure 3).
- Pupil assessment for anisocoria in third nerve palsy or Horner syndrome and relative afferent pupil defect for orbital lesions.
- Ocular motility assessment: specifically looking for any limitation of upgaze or hypotropia in primary to suggest superior rectus weakness in monocular elevation deficit (Figure 4), generalised motility limitation in chronic progressive external ophthalmoplegia.
- Cycloplegic refraction: particularly looking for astigmatism and anisometropia in cases of unilateral ptosis.
- Dilated fundus examination.
- Palpate and lift up / evert upper eyelid for lacrimal / sub-tarsal / superior orbital masses.
- Assessment of corneal protective mechanisms in older children: corneal sensation, Bell's reflex and tear film.

Table 1: Causes of paediatric ptosis.

True ptosis		Pseudoptosis
Congenital	**Acquired**	
• Isolated • Dysinnervational ptosis • Syndromic ptosis: blepharophimosis syndrome • Cranio-facial anomalies, e.g. Treacher Collins syndrome, Crouzon syndrome	• Mechanical • Traumatic • Myogenic • Neurogenic	• Contralateral eyelid retraction or proptosis • Hypotropia, hypoglobus • Microphthalmos, enophthalmos • Duane retraction syndrome • Monocular elevation deficit • Aberrant re-innervation of facial nerve

Differential diagnoses

True ptosis must be differentiated from the various causes of pseudo-ptosis. The various aetiologies are listed in Table 1.

Isolated simple congenital ptosis occurs in approximately 1:842 births [1]. Levator palpebrae superioris muscle fibres are poorly developed (dystrophic) and replaced with fibrous tissue and fat. Seventy percent of cases are unilateral. Ptosis is present from birth, and non-progressive. The lid crease is poorly formed and there is limited upper eyelid movement in both upgaze (due to poor muscle function) and downgaze due to the dystrophic changes in the muscle (inelastic levator).

Mechanical causes of ptosis include:

- Inflammation (cellulitis)
- Orbital or eyelid mass lesions (chalazion, capillary haemangioma, dermoid cyst, neurofibroma)
- Blepharophimosis syndrome (BPES) is associated with ptosis, telecanthus and epicanthus inversus (has mechanical as well as aponeurotic elements) (Figure 5).

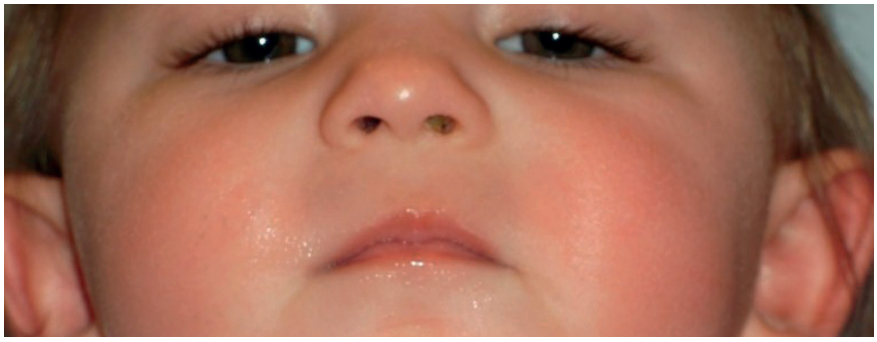

Figure 1: Chin up head posture to clear the lid from the visual axis.

Figure 2: Assessing the levator function by measuring the excursion of the lid between upgaze and downgaze while stabilising the brow (to inactivate the frontalis).

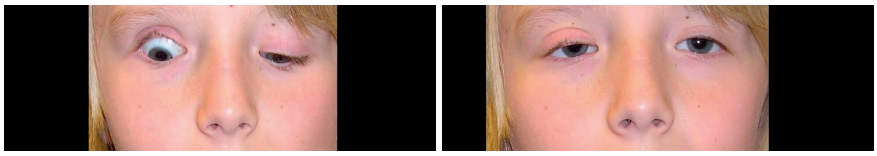

Figure 3: Marcus Gunn Jaw-winking phenomenon.

Figure 4: Right ptosis following levator resection showing limitation of upgaze (monocular elevation deficiency).

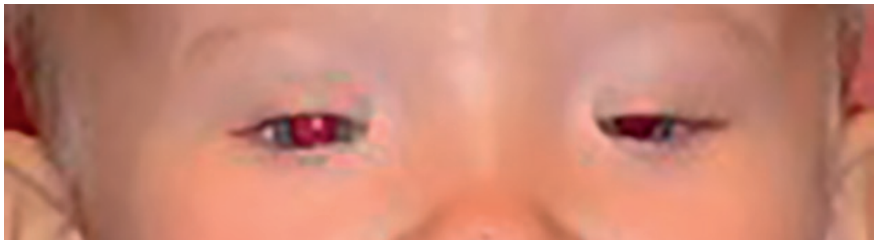

Figure 5: Blepharophimosis syndrome.

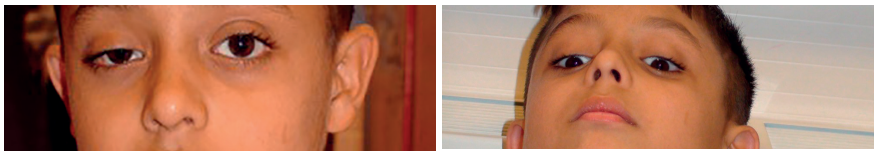

Figure 6: Downgaze lagophthalmos in congenital ptosis.

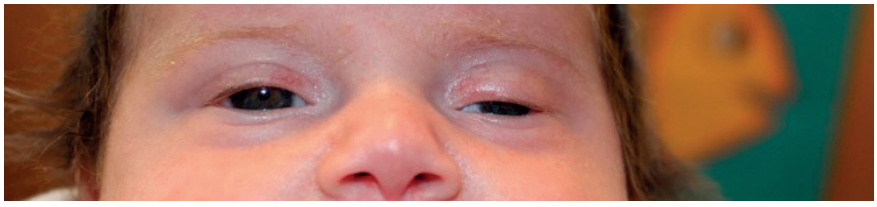

Figure 7: Amblyogenic ptosis.

Aponeurotic ptosis can be secondary to birth trauma, particularly with forceps-assisted delivery, or secondary to a developmental anomaly. Levator function is generally good and the lid crease may be normal or high if the aponeurosis is disinserted.

Myogenic causes include congenital fibrosis of the extraocular muscles (CFEOM) and chronic progressive external ophthalmoplegia (CPEO).

Neurogenic causes include congenital third nerve paresis, Horner syndrome or congenital cranial dys-innervational disorders such as Marcus-Gunn jaw-winking (Figure 3). There are usually associated pupil or ocular motility disturbances that suggest the diagnosis. Congenital and Acquired (autoimmune) Myasthenia are rare but an important cause with systemic implications.

Management

Although the definitive treatment of most forms of ptosis is surgical, medical management is indicated for some secondary causes, e.g. myasthenia. Supportive management of amblyopia and refractive errors is critical to ensure normal visual development.

Regular cycloplegic refraction is necessary and glasses are prescribed for significant anisometropia, commonly due to astigmatism. Amblyopia is treated where required.

Mechanical causes of ptosis require treatment of the underlying cause. In all other cases, surgical ptosis repair may be indicated either for visual function or to restore normal appearance. Surgical decision-making revolves around timing and choice of technique.

There are several considerations when formulating the management strategy:

1. Distinction between congenital and acquired ptosis

Congenital ptosis is characterised by downgaze lagophthalmos (Figure 6) and also some degree of blink and nocturnal (incomplete closure when asleep) lagophthalmos because of the inelastic dystrophic levator. This is an important consideration because levator surgery (which involves shortening an already inelastic levator muscle) as well as brow suspension surgery will increase lagophthalmos. This can be quite noticeable, particularly in unilateral cases. The postoperative lagophthalmos improves over several months, and will necessitate the use of ocular lubricants to reduce the risk of corneal exposure during this period.

2. Is the ptosis visually significant?

There may be risk of amblyopia with unilateral as well as bilateral ptosis. If the lid margin crosses / bisects the pupil for a major part of the waking hours, there is a risk of amblyopia. The presence of a chin-up acquired head posture (AHP) is reassuring, and indicates the child is making an effort to clear the lid from the visual axis. Marked chin up AHP, or lack of any AHP in the presence of severe ptosis is an indication for early surgery.

3. The timing of surgery

Early surgery is indicated if the visual axis (pupil) is occluded due to risk of deprivation

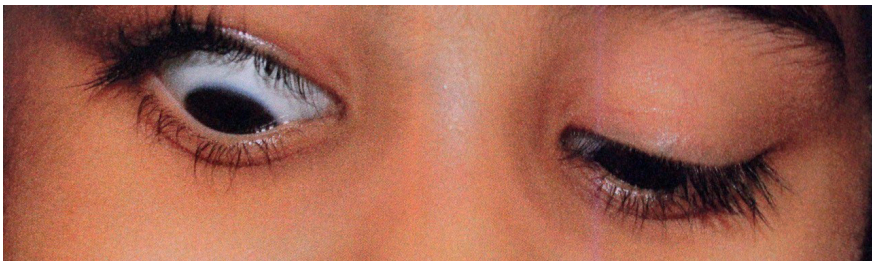

Figure 8: Postoperative downgaze lagophthalmos.

Figure 9: Pre and postoperative brow suspension with silicone.

amblyopia [2,3] (Figure 7). If the pupil is not occluded, surgery is carried out electively, usually deferred until at least three to four years of age when levator function can be accurately measured[4]. During this time the child's visual development is monitored. The most common timing of surgery is before the child commences primary school at around five years of age, when the psychosocial implications of the ptosis will become significant.

4. Aim of surgery

The aim of surgery is symmetry in primary position, and good cosmesis (good lid height and contour, and a well-formed skin crease). The expectation of postoperative nocturnal and down gaze lagophthalmos) must be emphasised to parents and the older child. This is particularly relevant with unilateral cases, where asymmetry in down-gaze is expected because of the induced lagophthalmos (Figure 8).

5. Choice of procedure

All paediatric ptosis surgery is done under general anaesthesia. The choice of surgery depends largely on the function of the levator palpebrae superioris muscle, and the laterality, i.e. unilateral or bilateral (symmetric or asymmetric).

There are two types of procedures for paediatric ptosis:

a. Utilising the levator muscle – the levator muscle is resected / advanced by the anterior or posterior approach.
b. Frontalis / brow suspension. It is common for children with ptosis to over-work their forehead muscles to compensate for the droopy lid, and this mechanism is used in the brow suspension operation.

The brow suspension operation compensates for a weak levator by creating a sling from artificial or natural (patients own tissues) materials to connect the forehead muscle and the eyelid. This sling suspends the eyelid at a higher level, providing some static lift when the forehead muscles are relaxed, and also provides a dynamic lift by lifting the eyelid even higher when the child works the brow muscles, transmitting the pull to the eyelid through the sling (Figure 9).

Various natural (autologous or banked fascia lata) [5] or artificial materials (silicone, polyfilament cable-type nylon (Supramid Extra: S. Jackson, Alexandria, VA, USA), Mersilene Mesh (Ethicon, Somerville, NJ, USA) and expanded poly-tetra-fluoro-ethylene (Gore-Tex: W.L. Gore & Associates, Newark, DE, USA)) [6, 7] can be used.

There are two methods of placement of the preferred material with the frontalis sling procedure: the Fox (pentagon) technique [5,8] or Crawford (double triangle) technique [5,9]. Most surgeons use the Fox (pentagon) technique with artificial materials, and either Fox or Crawford technique with fascia lata.

As a general rule, levator surgery is used in mild to moderate ptosis with good levator function.

Frontalis (brow) suspension is used in severe ptosis with poor levator function.

Advantages and disadvantages of the techniques

Levator resection uses the existing muscle, and is a more physiological procedure, providing a static lift. It results in a better skin crease and contour. As it is not reliant on the brow muscles, it works well in unilateral as well as bilateral cases. The lagophthalmos and asymmetry is less marked than brow suspension, particularly in unilateral cases.

However, predictability and longevity of this operation is unpredictable, especially when levator function is poor. It is not suitable if there is abnormal innervation, e.g. jaw-winking.

The amount of resection is inversely proportional to the levator function: the less the levator function the more the muscle needs to be resected. There are two formulas to calculate the amount of levator resection, one based on the levator function [10] and the other based on the Margin Reflex Distance [11] – see Table 2.

The levator resection operation is usually done via an anterior approach, through a skin crease incision. This approach enables not only the intraoperative correction of lid height but also the reformation of the skin crease which is important for symmetry and cosmesis.

Posterior approach ptosis correction (Muller's muscle-conjunctival resection [12] or white line advancement [13]) can be performed for mild ptosis with good levator function. This approach is preferred when the risk of overcorrection is significant or when there is risk of exposure, such as in cases of deficient upgaze or poor Bell's phenomenon [4].

Table 2: Estimating the amount of resection for simple congenital ptosis.

Levator function technique (Berke, 1959)	
Levator function	**Intraoperative lid height**
2-3mm (poor function)	at upper limbus
4-5mm (poor function)	1-2mm overlap
6-7mm (fair function)	2mm overlap
8-9mm (good function)	3-4mm overlap
10-11mm (good function)	5mm overlap
MRD technique (Beard, 1981)	
Preoperative MRD1	**Amount of resection**
3-4mm (mild ptosis)	10-13mm
2-3mm (moderate ptosis)	14-17mm
1-2mm (marked ptosis)	18-22mm
0-1mm (severe ptosis)	>23mm

Brow suspension is minimally invasive, works well in cases of poor levator function, and is suitable even if there is abnormal innervation, e.g. jaw-winking. It is easier to achieve a symmetrical result in bilateral cases with a brow suspension. However, the skin crease is less well formed, and the excess skin fold might necessitate additional surgery at the same time or at a later date.

The brow suspension has a static component resulting from the degree of tightening of the material at the time of surgery, and a dynamic component, which relies on elevation of the brow muscles. The lagophthalmos is more marked than with levator surgery, and asymmetry greater in unilateral cases.

The challenge of unilateral ptosis

As a general rule, unilateral cases do well with levator resection if there is reasonable muscle function. Unilateral ptosis with poor levator function is more challenging.

In unilateral cases children often do not over-work their brow muscles, as the drive to do this comes from the dominant eye. This is in contrast to bilateral cases, where the child usually demonstrates significant brow muscle over-action to lift both eyelids. Unilateral cases therefore need a tighter sling (static component) to achieve the same effect compared to bilateral cases because brow muscle over action (dynamic component) cannot contribute nearly as much. If the material is tightened excessively to create a greater static lift, there will be more marked lagophthalmos (blink, nocturnal and downgaze). If the surgeon creates a moderate static lift and relies on the dynamic component, there can be a partial ptosis when the brow muscles relax.

Planned sequential surgery in unilateral cases with poor levator function

For severe unilateral ptosis, it may be advantageous to perform a levator shortening operation initially, to provide a good skin crease and achieve a moderate static lift [14]. A subsequent brow lift can augment the effect of the first operation, and need not be very tight, thus minimising the lagophthalmos, and asymmetry in down gaze.

Recurrent ptosis

Recurrence can occur following childhood ptosis surgery, often several years later. This is due to gradual stretching of the resected weak levator muscle, or loss of tension in the material used for brow suspension.

Recurrences are more common if the child has a severe ptosis at presentation. Approximately 12% of operations fail over the first five years, and need further surgery [15,16,17].

"Congenital ptosis is characterised by downgaze lagophthalmos, and this is compounded by any form of ptosis surgery."

In some cases, a second operation might be required to address skin crease or contour issues. Special forms of ptosis such as blepharophimosis need multiple procedures performed in a planned sequence.

Recurrences can be corrected with either repeat levator resection, or brow suspension, often using a different material.

Management of unusual types of congenital ptosis

The management of ptosis associated with Marcus-Gunn jaw winking syndrome can be challenging. The jaw wink tends to improve by the end of the first decade. Observation during infancy and early childhood, rather than surgical intervention, is therefore recommended. The ptosis can be corrected in most cases with silicone brow suspension of the affected side. This corrects the ptosis without exacerbating the lid retraction associated with jaw-winking.

Marked residual jaw-wink in the older child is rare. Complete levator ablation followed by brow suspension is an option is such cases. However, the degree of asymmetry and lagophthalmos induced by this surgical approach is often unacceptable. Some authors advocate bilateral surgery for symmetry [18], however, this requires surgery on the unaffected side and is rarely accepted by parents. Unilateral and sometimes bilateral silicone brow suspension can be a reasonable compromise in such cases.

BPES syndrome

Surgical correction of BPES involves correction of the telecanthus and epicanthus inversus with medial canthoplasties, and correction of the ptosis with brow suspension. In mild cases, silicone may be used, but fascia lata is the preferred material for moderate to severe cases. Depending on the severity, the two operations may be performed simultaneously (in mild cases) or sequentially (in severe cases).

Postoperative care and long-term follow-up

Topical lubricant gel or ointment is prescribed four to six times daily, and at night until the blink and nocturnal lagophthalmos recovers. Treatment is continued until only the sclera is visible when the child is asleep, generally two to three months post-op. It is important to follow up children with congenital ptosis even after successful surgery in view of potential amblyopia, refractive error and astigmatism, which need to be monitored and treated appropriately. This is particularly important for unilateral cases.

Complications of paediatric ptosis surgery

The main risk after paediatric ptosis surgery is undercorrection or recurrence. Parents are counselled preoperatively of the need for subsequent operations, which may include early revision surgery. Overcorrection occurs less often. Corneal exposure is rarely problematic as children have a robust Bell's reflex and their corneas adapt rapidly to exposure. It is, however, important to use ocular lubricants in all cases. The risk of exposure is greatest with myopathies, e.g. CPEO, where the Bell's phenomenon is impaired, and there may be orbicularis weakness.

Eyelid asymmetry and poor lid contour or lid fold anomaly can occur with paediatric ptosis surgery. Skin incisions heal remarkably well in children and postoperative scarring is usually minimal. Infection is uncommon but can occur with silicone sling procedures. If the artificial material is exposed or extrudes, it must be removed and replaced after an interval of weeks to months with a similar or other material.

TAKE HOME MESSAGE

- Paediatric ptosis can be associated with amblyopia, cosmetic disfigurement and altered head posture.
- True ptosis must be differentiated from the various causes of pseudo-ptosis.
- Surgical decision-making revolves around timing and choice of technique.
- Early surgery is indicated if the visual axis (pupil) is occluded due to risk of deprivation amblyopia.
- As a general rule, levator surgery is used in mild to moderate ptosis with good levator function. Frontalis (brow) suspension is used in severe ptosis with poor levator function.

Summary

Ptosis can occur in children due to a variety of underlying causes. Management is complicated by the developing visual system. The timing and approach of surgical intervention is guided by severity of ptosis and amblyogenic potential, parental concerns, and social considerations such as teasing at school.

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