# CLASSIFICATION, ASSESSMENT, AND MANAGEMENT OF CHILDHOOD PTOSIS

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## ARTICLES

CLASSIFICATION, ASSESSMENT, AND MANAGEMENT OF CHILDHOOD PTOSIS

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Ptosis, or more correctly blepharoptosis, denotes drooping of the upper eyelid. Ptosis may be caused by defective function of either the levator palpebrae superioris muscle supplied by the oculomotor nerve or the sympathetically innervated Muller's muscle. Ptosis in children comprises about 50% of all ptosis. [14]

## CLASSIFICATION

Childhood ptosis is usually congenital and is rarely associated with serious underlying disease. In practice a poorly developed levator muscle is by far the most common cause of congenital ptosis. Other causes of congenital ptosis include oculomotor palsy, Horner's syndrome, and congenital fibrosis of the extraocular muscles. [18] [24] [29] [34] Acquired ptosis in children requires thorough investigation, as it may sometimes be a harbinger of serious underlying disease. [15] [18] [24] [34] Ptosis has many classifications, which will be described further (Table 1) [14]

**TABLE 1** -- CLASSIFICATION OF CHILDHOOD PTOSIS

| Category | Examples |
|---|---|
| Myogenic | Simple congenital ptosis |
| | Congenital fibrosis syndrome |
| | Myasthenia gravis |
| Neurogenic | Oculomotor (III) palsy |

| | |
|---|---|
| | Horner's syndrome |
| Aponeurotic | Trauma |
| | Prolonged rigid contact lens wear |
| Mechanical | Eyelid tumours |
| | Capillary haemangioma |
| | Orbital metastases |
| | Trachoma, other conjunctival scarring |
| Specific Forms | Blepharophimosis syndrome |
| of Ptosis | Marcus Gunn jaw-winking phenomenon |
| Pseudo-optosis | Enophthalmos, microphthalmos |
| | Contralateral proptosis |
| | Hypotropia |

### *Myogenic Ptosis*

Simple congenital ptosis is said to be due to a dystrophic levator muscle, where muscle fibers are replaced by fatty degeneration. [5] However, a recent study has found no histolopathological difference between normal and so-called dystrophic levator muscles. [11] The muscle not only contracts poorly, creating the ptosis, but also relaxes incompletely, leading to lagophthalmos on down gaze. The levator function is variable in simple congenital ptosis, with most having poor lid excursion. Congenital fibrosis syndrome may result in ptosis, in addition to restricted ocular motility. [22] Myastheniagravis may rarely cause ptosis in children; it is usually accompanied by strabismus and ophthalmoplegia. [25]

### *Neurogenic Ptosis*

Oculomotor nerve palsy may be congenital and is commonly accompanied by aberrant regeneration. [18] [24] Most cases of acquired oculomotor palsy are traumatic or infective in origin, with only a small minority caused by neoplasm. [18] [24] Patients with congenital Horner's syndrome may demonstrate heterochromia, ipsilateral facial anihydrosis, and increased amplitude of accommodation. Some acquired cases of Horner's syndrome are due to underlying neoplasm, particularly neuroblastoma. [5] [34]

### *Aponeurotic Ptosis*

Aponeurotic ptosis is caused by damage to the aponeurosis connecting the levator muscle to the eyelid. It is uncommon in childhood and usually results from trauma to the eyelid such as forceps delivery or surgery. It may also occasionally result from rigid contact lens wear, perhaps due to repeated traction on the upper lid while inserting or removing the lenses. [20] [31]

### *Mechanical Ptosis*

Mechanical ptosis in children may be traumatic or secondary to tumor, particularly plexiform neurofibromas in neurofibromatosis. Other causes include capillary hemangiomas, metastases, trachoma, or other forms of conjunctival scarring.

### *Specific Forms of Ptosis*

Six percent of patients with congenital ptosis demonstrate jaw-winking (Marcus-Gunn) phenomenon, usually unilaterally, but occasionally bilaterally. The affected lid moves upwards when children chew or move their jaw to the opposite side. This

syndrome results from connection of the levator muscle to the motor division of the trigeminal. The ptosis may vary from small and barely noticeable to more severe forms. It may be associated with muscle weakness, such as paralysis of the superior rectus muscle. [27]

Blepharophimosis syndrome is an infrequent cause of childhood ptosis. This autosomal dominant disorder is characterized by telecanthus, flattening of the nasal bridge, epicanthus inversus, and severe ptosis. [8] [30] There are three subtypes of blepharophimosis. In Type I blepharophimosis, all four eyelids are phimotic, and the upper lids are ptotic. In Type II, there is telecanthus and ptosis. The epicanthic folds are absent. In Type III, there are no epicanthic folds, but telecanthus and upper lid ptosis are present. [6]

### *Pseudo-ptosis*

Pseudo-ptosis is an apparent ptosis that may have a variety of causes. Anophthalmia, micro-ophthalmia, or enophthalmos may all give an erroneous impression of ptosis. Conversely, lid retraction or proptosis of one eye may be misinterpreted as ptosis of the opposite eye. An eye that is hypotropic may also appear ptotic. If the opposite eye is occluded, the patient fixates with the previously hypotropic eye, and the eyelid returns to the normal position.

## ASSESSMENT

All children with ptosis must undergo a thorough ophthalmic examination with assessment of visual acuity for amblyopia, pupils, ocular motility, Bell's phenomenon, refraction, and fundus examination. Deprivation amblyopia occurs if the visual axis is occluded. [17] The occluded eye may become myopic, compared with the fellow eye, and astigmatism may also develop. [16] [19]

Evaluation of the ptosis must include the following important eyelid measurements:

- Palpebral aperture height (normal value approximately 9 mm).
- Distance from the upper lid margin to the corneal light reflex (normally value approximately 4 mm to 5 mm).
- Levator function (the total excursion of the eyelid from extreme downgaze to upgaze while immobilizing the frontalis muscle, normal range 12 mm to 17 mm).
- The position and depth of the upper lid crease. The upper lid crease is frequently shallow or absent in congenital ptosis.
- Bellsphenomenon. Upward rotation of the globe on eyelid closure is an important ocular protective mechanism. When it is poor or absent, there is increased likelihood of corneal exposure following surgical repair of ptosis.

## SURGICAL CORRECTION OF CHILDHOOD PTOSIS

Ideally, correction achieves symmetry of both lids, normal contour of the lid margins, absence of lagophthalmos, no corneal exposure, and uncovering of both pupils. A number of operations are commonly used for treating ptosis (Table 2) . Where levator function is poor, a frontalis suspension (sling) is the procedure of choice. [9] [10] In this operation, the eyelids are suspended from the brow muscles, and the levator ceases to have a role in eyelid movement. [9] Levator muscle resection or advancement is the preferred treatment where levator function is moderate to good. [4] Some cases of mild ptosis with good levator function can be corrected by tarsal conjunctival resection (Fasanella-Servat) or similar procedures. [13] [28] The carbon dioxide laser is also effective for some forms of levator surgery. [2]

**TABLE 2** -- CHOICE OF OPERATION

| Degree of Ptosis (mm) | Levator Function (mm) | Operation |
|---|---|---|

| | | |
|---|---|---|
| 1–3 | 10–15 | Fasanella-Servat<br>Gavarais<br>Aponeurotic tuck |
| 3–5 | 6–10 | Levator resection |
| 5–7 | <5 | Frontalis suspension |

As with all childhood ophthalmic disorders, glasses and or occlusion therapy for amblyopia should be prescribed as appropriate.

### *Poor Levator Function*

Where levator function is poor (<6 mm), frontalis suspension (sling) is the most appropriate operation. [9] [10] This procedure is suitable for children with simple congenital ptosis who have poor levator function or following failed levator resection. This operation is alsothe procedure of choice for ptosis due to congenital fibrosis syndrome, severe jaw-winking syndrome, blepharophimosis syndrome, oculomotor palsy, or trauma. Numerous materials have been used to create the sling, including silicone rods, mersilene mesh, assorted suture materials, and autogenous and banked fascia lata. [7] [21] [32] A number of series of ptosis correction with stored fascia lata have demonstrated a recurrence rate of up to 28%. [12] [26] Autogenous fascia lata is now the material of choice for children over 4 years of age, where the thigh is sufficiently developed to harvest the fascia. [9] for children under 4 years, the fascia lata is not sufficiently developed, and autogenous fascia lata or synthetic materials such as silicone rods must be employed. [7] [9]

### *Technique for Fascia Lata Frontalis Sling Procedure*

The lateral aspect of the thigh provides the site for fascia removal. Using the Crawford (Storz, Canada) separator and stripper, a large strip of fascia can be removed through a small incision just above the knee joint (Figs. 1 (Figure Not Available) and 2) (Figure Not Available) . Several techniques for performing the brow suspension have been described. Crawford popularized a technique employing three to four small horizontal skin/muscle incisions on the upper lid, approximately 2 mm above the lid margin. [9] Two further incisions are made in the skin of the forehead or brow. The temporal incision is made just temporal to a vertical line through the most temporal eyelid incision. The nasal brow incision is similarly placed just nasal to a vertical line from the nasal most eyelid incision. A further central brow incision is made above and parallel to these incisions, and is located mid-way between them. Wright's fascia needle is used to thread the fascia from the lid margin, under the skin and through the brow incisions (Figs. 3 (Figure Not Available) and 4) (Figure Not Available) . The eyelid is elevated so that it is just lifted off the globe. This is typically 2 mm to 3 mm more than desired, as the eyelid will drop somewhat post-operatively with the return of orbicularis muscle tone. [9] [19] Fascia lata harvesting from the thigh results in mild discomfort lasting less than a week, the only long-term consequence being the scar. 35

**Figure 1.** (Figure Not Available) Fascia separator and forceps (*A*). Crawford (Storz, Canada) fascia stripper (*B*). *(Courtesy of Storz, Inc., Canada: with permission)*

**Figure 2.** (Figure Not Available) *A,* Attention is first directed to the lateral thigh. An imaginary line connecting the head of the fibula and the anterior superior iliac spine lies in the direction of the fibers of the fascia lata. A 5-cm mark is made along this line starting 5−8 cm above the tibial plateau to indicate the proposed skin incision. *B,* A scapel incision is made through skin, subcutaneous tissue and fat, exposing the fascia. Metzenbaum scissors bluntly separate the fascia and fat for several centimeters above the wound.*C,* Parallel longitudinal incisions are made 8−10 mm apart and joined inferiorly to form a tongue of fascia. *D,* The fascia is introduced into a stripper (the Crawford stripper is illustrated), which is advanced while applying countertraction with a hemostat on the fascial tongue. An 8−10 cm strip is desirable. The cutting blade is used to sever the fascia. A second strip can be similarly removed if needed. *(From Beyer-Machule CK, von Noorden GK: In Heilman K, Paton D [eds]: Atlas of Ophthalmic Surgery, vol 1. New York, Thieme-Stratton, 1985, p 144−147; with permission.)*

**Figure 3.** (Figure Not Available) Similarly, the fascia is threaded along the tarsal surface and out at the central lid incision. From here, it is passed to the nasal brow incision forming a triangle. *(From Beyer-Machule CK, von Noorden GK: In Heilman K, Paton D [eds]: Atlas of Ophthalmic Surgery, vol 1. New York, Thieme-Stratton, 1985, p 144−147; with permission.)*

**Figure 4.** (Figure Not Available) *A,* The same procedure is carried out laterally forming two triangles of fascia. The strip ends are tied together in square knots at the apex of each triangle. The knots are reinforced with 5-0 chromic catgut sutures and allowed to retract. *B,* One end of each of the strips is now tunneled subcutaneously

to the upper forehead incision. A hemostat is attached to each strip. Traction is applied to each hemostat until the lid contour is correct. A third hemostat then grasps the two strips at skin level, and the first hemostats are removed. *(From Beyer-Machule CK, von Noorden GK: In Heilman K, Paton D [eds]: Atlas of Ophthalmic Surgery, vol 1. New York, Thieme-Stratton, 1985, p 144−147; with permission.)*

Other sling procedures, including the use of orbicularis oculi and orbital septum have been described for very severe congenital ptosis. [1]

### *Moderate Levator Function*

Levator resection is the operation of choice when there is considerable ptosis (3 mm to 5 mm) but reasonable levator function (6 mm to 10 mm). The amount of levator muscle resection depends on the degree of ptosis. [4]

### *Good Levator Function*

Mild ptosis (1 mm to 3 mm) but good levator function (10 mm to 15 mm) can be treated using one of several procedures. Tarsal conjunctival resection (Fasanella-Servat procedure), Muller's muscle-conjunctival resection (Gavarais procedure), or aponeurotic repair may all give excellent results. [13] [28]

### *Treatment of Specific Forms of Ptosis*

The jaw-winking syndrome does not require treatment if the ptosis is mild. If the ptosis is marked, levator resection is the treatment of choice. However, if the jaw winking is severe, a 5 mm to 10 mm segment of the levator is removed to eliminate the wink, this creates a complete ptosis. One month later, a bilateral frontalis suspension procedure is performed to elevate the lid and create symmetry with the opposite side. [27]

The treatment of blepharophimosis depends on the subtype. [1] [6] Type I (all four eyelids phimotic with upper lid ptosis) is treated by rearranging the epicanthic folds to shift the medial canthal structures medially and correct the ptosis. The ptosis in Type II (telecanthus and ptosis, absent epicanthic folds) is treated by the frontalis sling procedure. In Type III (epicanthic folds but telecanthus and upper lid ptosis), the treatment is staged with skin grafting, craniofacial surgery, and the frontalis sling. [6]

### *Treatment of Other Forms of Ptosis*

Ptosis resulting from oculomotor palsy must be approached with caution because of the high risk of corneal exposure in an eye that may be hypotropic with poor elevation and poor Bell's phenomenon. [23] Where indicated, the ptosis is treated by frontalis suspension or levator resection depending on levator function. Ptosis secondary to Horners syndrome is usually mild and can be corrected by tarsal conjunctival resection. [13] Ptosis in ocular fibrosis syndromes can be treated by frontalis suspension. [22]

## COMPLICATIONS OF PTOSIS SURGERY

Undercorrection is the most common post-operative problem (Box 1) . With levator resection undercorrection may result from inadequate resection of the muscle or from performing the procedure where levator function is very poor. Recurrence of the ptosis may occur, particularly following a frontalis suspension operation where stored fascia lata has been employed. [26] Reaction to the sling material may also occur, particularly with stored fascia lata. [8]

**Box 1. Complications of Ptosis Surgery**

Undercorrection (or recurrence)

Overcorrection

Lagophthalmos

Ectropion

Entropion

Conjunctival prolapse

Hematoma

Reaction to sling material

Protrusion of sling material

Over-correction is uncommon following repair of congenital ptosis. All patients will have some exacerbation of their lagophthalmos on down gaze, and some develop mild corneal exposure problems. Exposure usually occurs during sleep, particularly if the Bell's phenomenon is poor. [7] Most cases of corneal exposure in children resolve quickly with the aid of lubricant ointment before going to sleep. [9] Corneal exposure problems are most likely to occur following repair of ptosis secondary to congenital fibrosis syndrome or oculomotor palsy. [22] [23] Overcorrection following ptosis surgery may improve with time or with massage of the eyelid. If over-correction persists, a number of surgical procedures are available to lower the upper lid. These procedures include levator tenotomy, recession, or lengthening using a tendon expander. [3]

Upper lid entropion is a rare complication. This may occur if the fascia lata is placed too deeply in the eyelid. Alternatively, if the fascia lata is placed too anteriorally, ectropion may result. Placement of sutures too inferiorally on the tarsal plate in a levator resection may also lead to upper lid ectropion.

Uneven lid contour may result from uneven placement of the tarsal sutures in an anterior levator resection (carried-out through the skin). During tarsal conjunctival resection, excessive removal of tarsal plate may result in the center of the eyelid being raised too high. [3]

Post-operative edema can cause the conjunctiva to prolapse between the eyelids or even become partially strangulated. If prolapse occurs, a firm pad and pressure bandage may help the edema to resolve. If this fails, it may be necessary to place sutures through the conjunctiva of the upper fornix and bring them out through the skin where they are tied on the surface.

## SUMMARY

Ptosis in childhood may both impair normal visual development and be cosmetically disfiguring. Ptosis may sometimes be a component part of a more extensive disorder involving the extraocular muscles, facial structures, or nervous system. Each patient must receive a thorough ocular examination, as well as careful assessment of the ptosis itself. Optimum outcome following surgical repair depends on the correct choice of operation for the specific type of ptosis and degree of levator muscle function.

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