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Surgical treatment of unilateral severe simple congenital ptosis

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Abstract:

Unilateral congenital ptosis with poor levator function of ≤ 4 mm continues to be a difficult challenge for the oculoplastic surgeon. Surgical correction can be accomplished with unilateral frontalis suspension, maximal levator resection, or bilateral frontalis suspension with or without levator muscle excision of the normal eyelid. Bilateral frontalis suspension was proposed by Beard and Callahan to overcome the challenge of postoperative asymmetry, allowing symmetrical lagophthalmos on downgaze, postoperatively. However, most surgeons and patients prefer unilateral correction on the abnormal eyelid either with a frontalis suspension or maximal levator resection. Frontalis suspension may be performed through the various surgical techniques using different autogenous or exogenous materials. Autogenous fascia lata is considered the material of choice with low recurrence rates but carries the drawbacks of the difficulty of harvesting and postoperative morbidity from the second surgical site. Recent reports have suggested that maximal levator resection provides improved cosmesis, a more natural contour, and avoids brow scars. Although both treatments have shown to have similar success rates, there is much debate about what the most favorable method for treating severe unilateral ptosis, including the rationale, advantages and disadvantages of each technique.

Keywords:

Congenital ptosis, frontalis suspension, maximal levator resection, poor levator function

Introduction

ongenital ptosis is most frequently

caused by myogenic dystrophy of the levator muscle. It may also be associated with blepharophimosis syndrome, Marcus-Gunn jaw-winking syndrome, and other systemic causes of muscle weakness.^[1] It has been estimated that levator function is poor in 71.8% of congenital ptosis and unilateral in 64.7%–75.0%.^[2,3] Patients with congenital ptosis are more predisposed to developing amblyopia, especially in unilateral cases, usually due to convergent strabismus, high astigmatism, or anisometropia.^[4-6] The correction of congenital ptosis, therefore, is crucial to avoid visual disturbance as well as for reasons of cosmesis. Levator function and severity of ptosis determine the choice

of surgical treatment of congenital ptosis.^[7] Levator function is categorized into three groups depending on the amount of lid excursion: good (excursion >8 mm), fair (5–7 mm), and poor (0-4 mm).^[8,9]

Congenital ptosis with poor levator function is one of the most challenging procedures for oculoplastic surgeons. In particular, the optimal surgical correction for unilateral severe congenital ptosis remains highly controversial. Several surgical procedures have been demonstrated to treat severe unilateral congenital ptosis with variable outcomes and complications. Two main surgical options are frontalis suspension and maximal levator resection. Although both methods have been reported to have satisfactory surgical success for unilateral congenital ptosis the debate on which is the optimal treatment with lower complication rates still exists. For the treatment of congenital ptosis, levator function, severity of ptosis,

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laterality, associated diseases (i.e., blepharophimosis, Marcus-Gunn jaw-winking syndrome), and surgeon's preference should all be factors to be considered.

Surgical Considerations

Frontalis suspension surgery versus levator surgery

The ideal surgical choice for unilateral congenital ptosis with poor levator function remains divisive. Frontalis suspension surgery has been considered as the most successful surgical option for correcting congenital ptosis with poor levator function of $\leq 4 \text{ mm.}^{[10-14]}$ This technique was described by Wright,^[15] and further developed with the use of autogenous fascia lata as the standard suspension material by Crawford.^[11] It allows the frontalis muscle to elevate the upper eyelid upward by creating a connection between the frontalis muscle and the upper tarsal plate. Various sling materials and surgical techniques have been used for frontalis suspension. Autologous fascia lata has been reported to be the ideal material for frontalis suspension with comparatively lower rates of complications such as infection and granuloma formation.[11] However, it requires the harvesting of fascia lata from the child's leg and therefore leads to potential donor-site morbidities.[16] Alternative substances are available as off-the-shelf solutions such as preserved fascia lata, silicone rods, and e-polytetrafluoroethylene. Many articles have shown that frontalis suspension operation may cause hypertrophic scars in the forehead region, inflammation, extrusion, infection of the suspensory material, and late failure.^[14,17]

Levator resection is conventionally carried out for patients who have fair-to-good levator function of more than 4 mm, whereas frontalis suspension is reserved for those with poor levator function of <4 mm.^[1,5,14,18,19] Some oculoplastic surgeons have advocated maximal levator resection as the first choice of surgery even in ptosis patients with poor levator function.^[20-28] However, there is always concern that levator resection in patients with poor levator function would result in undercorrection, even with maximal levator dissection and resection.^[29,30] This would naturally result in some surgeons preferring frontalis suspension in cases of severe ptosis. However, a study of maximal levator resection in unilateral congenital ptosis with poor levator function found no statistically difference in outcomes between two groups of patients, one with preoperative levator function of 0-2.0 mm and the other 2.5-4.0 mm.^[28] Another study similarly showed that unilateral congenital ptosis with <2 mm of levator function achieved satisfactory results in 81.8% with maximal levator resection.^[22]

Spontaneous brow elevation is crucial for obtaining excellent lid height after frontalis suspension surgery.^[31] In cases of amblyopia or operation on the nondominant

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eye, failure to recruit the ipsilateral frontalis muscle, can occur after unilateral brow suspension due to fixation preference of the fellow normal eye.^[32] Since patients with unilateral congenital ptosis are susceptible to amblyopia, undercorrection is not uncommon after unilateral frontalis suspension surgery. For this reason, bilateral frontalis suspension or unilateral levator surgery can be recommended to overcome the above problem.^[28]

Levator resection surgery has been shown to result in marked the improvement of postoperative levator muscle function of 2.9–5.4 mm, which induces a favorable effect on the surgical success, especially for patients with a poor levator function.^[25,33] Natural elevation movement of the eyelid can be produced in a superior-posterior direction following levator resection surgery.^[34] It allows normal physiologic motion of the upper eyelids as opposed to the enforced use of sling materials in frontalis suspension surgery.

Frontalis Suspension Surgery

A general surgical technique for treating unilateral congenital ptosis with poor levator function of ≤ 4 mm is frontalis suspension surgery. As the levator aponeurosis is inherently weak, another motor vector is necessary to raise the eyelid. The frontalis muscle provides both functions of suspension and elevation.^[35] The important goal of the surgery is to raise the eyelid by creating a direct connection between the frontalis muscle and the tarsal plate of the upper lid. This allows an elevating force to be transmitted to the eyelid during contraction of the frontalis muscle, in addition to a suspensory force in primary gaze. Frontalis suspension surgery is generally considered a technically simple and effective procedure to elevate the eyelid.

Laterality: Unilateral versus bilateral surgery

Frontalis suspension can be accomplished with either bilateral or unilateral surgery. Some authors recommend bilateral frontalis suspension procedure for the treatment of unilateral poor-function congenital ptosis.[36-38] Advocates of the bilateral approach are concerned about postoperative asymmetry during eyelid closure, blinking, and down gaze. Beard advocated excision of both the affected and normal levator muscles to create bilateral symmetrical ptosis followed by bilateral frontalis suspension with fascia lata.^[36] Some levator function is believed to remain in the normal eyelid even after being severed due to unseparated attachments such as the suspensory ligament in the superior conjunctival fornix, resulted in the elevation of the upper eyelid.^[12] Callahan recommended bilateral frontalis suspension with preservation of the normal levator muscle.^[37] However, bilateral surgery would mean putting the normal eye at risk of postoperative complications such as lagophthalmos, exposure keratopathy, entropion, and eyelash ptosis.^[38] Most oculoplastic surgeons, patients, and guardians would naturally prefer to operate only on the abnormal side when treating unilateral congenital ptosis, without needing to destroy normal structures compared to bilateral surgery. Patients with unilateral surgery unconsciously adjust relatively well to unilateral lagophthalmos and lid lag, both becoming less apparent by voluntary orbicularis contraction with time.^[12] Unilateral frontalis suspension^[18,32,35] and maximal levator resection^[20-22,26-28,34] have been recommended as approaches that intervene only on the deficient side.

Sling materials for frontalis suspension

Autogenous fascia lata has been regarded as the best material for frontalis suspension. Histopathologically, implanted autogenous fascia lata has been shown to remain with markedly increased vascularization and incorporation into adjacent tissues even after 42 years.^[39,40] However, it is difficult to obtain the fascia lata and requires two surgical sites. The donor site at the thigh bears the morbidity risks associated with harvesting fascia lata such as early leg pain, limping, and scar formation.^[16] At the eyelid surgical site, contracture of the implanted fascia lata may result in eyelash inversion, entropion, and tarsal deformity.^[41,42]

Other various autogenous and exogenous suspensory materials, including banked fascia lata,^[43,44] silicone rod,^[10,45,46] mersilene mesh,^[47,48] nylon polyfilament cable-type suture (Supramid sutures),^[42,49,50] prolene sutures,^[51] and polytetrafluoroethylene (Gore-Tex)^[42,52] have been used, each with diverse rates of success and complications. The latter includes recurrence of ptosis or complications related to allogenic materials such as infection, exposure of graft, immune rejection, and granuloma formation.^[11,17,43,51] For all sling materials, the inability of patients to elevate their brow reliably can lead to unsatisfactory results.^[21,53]

Levator Surgery

Maximal levator resection versus Whitnall's sling Maximal levator resection is a synonym for super-maximal levator resection, supramaximal levator resection, and total levator aponeurosis resection.^[20,24-26,34] Super-maximal levator resection was first introduced for severe unilateral congenital blepharoptosis in 1984.^[20] Epstein and Putterman showed cosmetically acceptable results in 6 of 8 cases (75%) with super-maximum levator resection. It was compared to bilateral frontalis suspension with autogenous fascia lata with excision of the normal levator aponeurosis, which achieved good results in 4 of 8 cases (50%).^[20] Many surgeons have since performed maximal levator resection for severe congenital ptosis with satisfactory outcomes rates of 28.6% to 100% [Table 1]. ^[20-27,34,54-56] For unilateral ptosis cases, reported success rates have ranged from 63.6% to 100% [Figure 1].^[20,22,24,26,56]

Whitnall's sling differs from maximal levator resection in that the upper eyelid is attached to the Whitnall's ligament after maximal resection of the aponeurosis in the former, while fixation sutures may be placed as high above the Whitnall's ligament as necessary in the latter to obtain satisfactory intraoperative eyelid level.^[54] Opponents of the Whitnall's sling may argue that Whitnall's ligament of patients with ptosis may be atrophic or dehiscent, and hence may provide weak and insufficient support for severe congenital ptosis.^[58]

Some articles have reported that superior tarsectomy can augment maximal levator resection or Whitnall's sling for treatment of severe congenital ptosis with poor levator function.^[34,54,55] The Whitnall's sling procedure in conjunction with an excision of 4-5 mm of superior tarsus can achieve a lid height within 1 mm of the fellow eyelid in 68% of severe ptosis cases.^[55] Superior tarsectomy augments the eyelid height of the maximal levator resection by increasing the resection of the upper eyelid tissue.^[34] The excision of the tarsal plate, however, can cause an unstable eyelid position with undesirable complications including ectropion and eyelid instability, particularly with maximal levator resection. The removal of normal tarsus also makes surgery even more difficult in cases requiring reoperation.^[26,34] A large retrospective and a prospective study, in fact, showed maximal levator resection without tarsectomy to have a significantly higher success rate in severe unilateral congenital ptosis patients than that with tarsectomy.^[27,28]

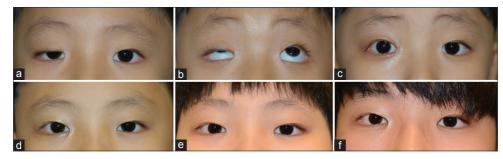


Figure 1: Representative case of good surgical outcome after maximal levator resection (a and b) a 5-year-old patient with severe unilateral congenital ptosis with 3.0 mm levator function (c) 1 month after maximal levator resection (d) 2-year postmaximal levator resection (e) 5 years after operation (f) 7-year after maximal levator resection

Author	Surgical technique	Follow up	Laterality	Number of cases	Levator function	Success rate (%)
Epstein and Putterman ^[20]	Super-maximal levator resection	NA	Unilateral	8	0-4	6/8 (75)
Mauriello <i>et al</i> . ^[21]	Maximal levator resection	18 months	24 unilateral, 4 bilateral	32	0-2	28/32 (87.5)
Anderson <i>et al.</i> ^[54]	Whitnall's sling±superior tarsectomy	>1 year	59 unilateral, 5 bilateral	69	1-5	49/69 (71)
Holds et al.[55]	Whitnall's sling with superior tarsectomy	3-24 months	Unilateral	25	1-7	17/25 (68)
Press and Hübner ^[22]	Maximal levator resection	NA	Unilateral	44	0-2	36/44 (81.8)
Pak <i>et al</i> . ^[34]	Super-maximum levator resection with superior tarsectomy	NA	1 unilateral, 7 bilateral	8	3-4.5	4/6 (66.7)
	Super-maximum levator resection only	NA	1 unilateral, 9 bilateral	10	1.5-4.0	2/7 (28.6)
Park et al. ^[23]	Levator resection	27 months	35 unilateral, 15 bilateral	65	2-4	35/65 (53.9)
Al-Mujaini and Wali ^[24]	Total levator aponeurosis resection	2-24 months	Unilateral	7	1-5	7/7 (100)
Kasaei <i>et al</i> . ^[56]	Levator resection with tarsal resection	2-12 months	Unilateral	17	1-5	13/17 (76.4)
Decock et al.[25]	Supramaximal levator resection	>1 year	Unilateral	11	0-4	7/11 (63.6)
Cruz et al.[26]	Supramaximal levator resection	5-85 months	Unilateral	35	6.6	32/35 (91.4)
Mete <i>et al</i> . ^[27]	Maximal levator resection	10-36 months	17 unilateral, 6 bilateral	29	0-4	16/23 (69.6)
Lee <i>et al</i> . ^[28]	Maximal levator resection	40.9 months	210 unilateral, 33 bilateral asymmetric	243	0-4	226/243 (93.0)
Chen <i>et al.</i> ^[57]	Levator resection with suspensory ligament of the superior fornix suspension	12-18 months	10 unilateral, 15 bilateral	40	0-4	Unilateral - 8/10 (80) Bilateral - 14/15 (93.3

Table 1: Surgical outcomes of maximal levator resection and Whitnall's sling for congenital ptosis

NA = Not available

The medial and lateral horns of the levator aponeurosis are cut during maximal levator resection according to the desired amount of levator complex to be resected.^[17,22,25,26,34,57] Some have emphasized that complete dissection of the lateral and medial horns is the most important surgical step in maximal levator resection.^[22] Recently, however, some surgeons have preferred to preserve the horns for providing vertical support to the eyelid and allow better eyelid contour.^[27,28] Transection of the medial and lateral horns can result in the destruction of the normal structures of the lacrimal gland and reflected tendon of the superior oblique muscle.^[59] It has been proposed that complete dissection of the levator aponeurosis and Muller's muscle from the tarsal plate and conjunctiva to above the level of Whitnall's ligament until the desired intraoperative lid height is obtained is the crucial step to obtaining a good surgical outcome, rather than transection of the levator horns per se.[27,28] In a series of 243 patients with unilateral severe congenital ptosis who underwent maximal levator resection, the levator complex was sufficiently dissected superiorly without severing the medial and lateral horns of the levator aponeurosis. Anchoring sutures from the tarsal plate were fixed at the level of or above the Whitnall's ligament with the intraoperative height of the upper eyelid adjusted to the level of the superior

limbus in patients under general anesthesia and 1 mm higher than the contralateral eyelid in patients with local anesthesia. Ninety-three percent (226/243 eyelids) had satisfactory results after maximal levator resection without the need to transect of the levator horns.

Complications of Severe Ptosis Correction

After maximal levator resection or frontalis suspension operation, postoperative lid lag on down-gaze and lagophthalmos are inevitable. Unnatural eyelid movement and sluggish or incomplete blinking are almost always present although eyelid symmetry is generally obtained in primary gaze with favorable surgical outcomes. Mild to moderate exposure keratopathy can develop in all patients after correction of severe congenital ptosis, whatever the procedure. The cornea can be protected by voluntary contraction of orbicularis muscle and keratopathy is usually prevented with lubricants or lid taping. Nonetheless, significant corneal problems can be encountered in the postoperative period.^[11,60,61] Furthermore, postoperative lagophthalmos, lid lag on down-gaze, and sluggish blinking should be fully informed to the patient and parents preoperatively. If necessary, this can be improved with the release of the levator complex and skin incision, as well as with the recovery of the pretarsal orbicularis muscle with time after the operation. $^{\scriptscriptstyle [55]}$

Entropion or eyelash inversion occurs in 5.4%–11.9% of cases after correction of severe congenital ptosis, which is associated with increased posterior lamella vertical tension.^[28,62,63] The imbalance between a shortened posterior lamella and redundant anterior lamella leads to overhanging of the eyelid and ciliocorneal touch.^[28] It can be avoided by excising a segment of skin and orbicularis muscle above the crease and eyelash-rotating sutures (skin of the inferior flap-tarsal plate– skin of the superior flap).^[28,61]

Other common complications of frontalis suspension surgery or maximal levator resection include undercorrection, overcorrection, poor eyelid contour, ectropion, deformity of the crease, and conjunctival prolapse.^[61] The use of nonautogenous material in frontalis suspension surgery can lead to potential foreign body tissue reaction, granuloma formation, hypertrophic scar, extrusion, and infection.^[64]

Conclusions

Surgical treatment of unilateral congenital ptosis with poor levator function is still one of the most challenging tasks for oculoplastic surgeons since complications such as undercorrection, lid asymmetry, and poor cosmetic results are common postoperative complications. Frontalis suspension surgery has been favored in the treatment of unilateral severe congenital ptosis in the past decades. However, maximal levator resection also provides favorable results with more physiological and natural eyelid movement and possibly fewer complications. Challenges pertaining to the best option for the management of unilateral congenital ptosis with poor levator function still remain, and the debate regarding the ideal surgical procedure may persist for some time to come.

Ethical approval

The study was conducted in accordance with the Declaration of Helsinki and was approved by the local ethics committee of the institute. Informed written consent was obtained from all patients prior to their enrollment in this study.

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Nil.

Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

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