Brief Communication

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Single-triangle technique for congenital ptosis repair with a frontalis sling in blepharophimosis patients

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Abstract:

The purpose of this study was to describe the single-triangle technique for congenital ptosis repair with a frontalis sling in blepharophimosis patients. The single-triangle technique was used in 40 eyes of 20 patients of blepharophimosis syndrome. The center point of the lid is marked. The desired base length is calculated depending on the available horizontal fissure width. Two marks are inked 2 mm above the lid margin, equidistant from the central mark. A single brow mark is placed in such a way that it is directly above the center point of the lid. These are now joined to complete the triangle. In blepharophimosis patients, the mean preoperative margin reflex distance (MRD₁) was 1.0 ± 1.1 mm which increased to 4.1 ± 1.6 mm after surgery. The MRD₁ increased by 3.1 ± 1.7 mm. Cosmetic outcome was graded with a score of 0, 1, or 2 to indicate poor, good, and excellent results, respectively. Out of the 40 eyes that were operated, 33 eyes had a score of 2, 5 eyes had a score of 1, and 2 eyes were scored 0. The single-triangle technique has several advantages over both the Fox pentagon technique and modified Crawford technique in severe blepharophimosis patients. It is not only a much simpler procedure to perform but also has a better control over the curvature of the lid without any central focal notching, thus providing better cosmesis and esthetic results.

Keywords:

Blepharophimosis, Fox pentagon, modified Crawford, single triangle

Introduction

The frontalis sling surgery (FSS) is a commonly used surgical procedure for the treatment of severe ptosis with poor levator palpebrae superioris (LPS) action. Several materials have been tried and several techniques have been described for passing the sling.

The different techniques described for passing the sling are single pentagon (Fox pentagon),^[1] double pentagon, double triangle (Crawford procedure),^[2] single rhomboid (Friedenwald-Guyton procedure),^[3] double rhomboid (Iliff procedure),^[4] and double trapezoid

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(Wright procedure). The Fox pentagon and modified Crawford technique are the two commonly used methods of passing the sling.^[5]

However, in cases like severe Blepharophimosis syndrome (BPES), these two techniques are not very effective and the lid contour may not be satisfactory. In many patients with severe blepharophimosis, despite medial canthal surgeries, the horizontal fissure width (HFW) achieved may not be more than 20–22 mm.^[6] In these cases, the two commonly used techniques, i.e., Fox pentagon and modified Crawford, do not provide good contour to the lids due to disturbed dynamics of lids.

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Submission: 01-09-2020 Accepted: 19-02-2021 Published: 17-04-2021 We used a single-triangle technique in 40 eyes of 20 patients of severe blepharophimosis and studied the lid contour achieved.

Materials and Methods

Written and informed consent was taken from all patients. This study was approved by the Institutional Ethics Committee and Institutional Review Board (2019/01-78). All procedures performed in our study involving human participants were in accordance with the 1964 Helsinki Declaration and its later amendments.

Inclusion criteria

Twenty patients of blepharophimosis were evaluated to be included in this cohort study.

Preoperative assessment for ptosis surgery included margin reflex distance (MRD₁ and MRD₂), vertical fissure height (VFH), HFW, levator function, and frontalis action. All measurements were done after blocking the action of the frontalis for accurate pre- and postoperative comparison. For blepharophimosis assessment, the interpupillary distance and inter-medial-canthal distance were recorded. Intact corneal sensations and a good Bell's phenomenon were confirmed. Marcus-Gunn jaw-winking syndrome, congenital third nerve palsy, and congenital fibrosis of the extraocular muscles (CFOEM) were ruled out by examination of extraocular muscle movements.

Thus, all blepharophimosis patients included in this cohort study had bilateral severe congenital ptosis with poor levitation function with good Bell's reflex. The patients with epicanthal folds underwent Mustarde's double Z-plasty as a primary treatment plan for the epicanthal folds as well as horizontal fissure lengthening while the patients without any epicanthal folds underwent Spaeth's V-Y plasty for additional horizontal fissure length. All the patients were included in this cohort study for correction of ptosis 8 weeks after their primary surgery.

The single-triangle technique was used in 40 eyes of 20 patients, i.e., all patients were operated bilaterally. The important preoperative clinical findings in cohort patients are presented in Table 1.

Rationale of technique

The tension along the lateral arms in both the Fox pentagon and the modified Crawford does not uniformly distribute along the lid in cases of severe blepharophimosis. In such cases, the primary desired end-result generally remains uncovering the pupillary area which can be effectively achieved by the single-triangle method.

Markings

Single triangle [Figure 1]

The center point of the lid is marked. The desired base length is calculated depending on the available HFW. Two marks are inked 2 mm above the lid margin,

Table 1: Demographic and clinical profile of blepharophimosis epicanthus-inversus syndrome patients undergoing single-triangle technique for ptosis correction

Age sex	Epicanthal fold	Primary surgery	HFW (mm)		MRD1 (mm)			MRD2 (mm)		
			Right	Left	Right Left		.eft	Right	Left	
					Preoperative	Postoperative	Preoperative	Postoperative		
3/male	None	V-Y	19	19	1	4	1	4	5	5
6/male	Inversus	Double-Z	20	20	2	5	2	5	5	5
7/male	Inversus	Double-Z	18	18	0	3	0	3	5	5
18/female	Palpebralis	Double-Z	19	19	1	4	1	4	5	5
16/female	Palpebralis	Double-Z	17	17	0	4	0	4	5	5
5/male	None	V-Y	22	22	2	5	2	5	6	6
8/female	None	V-Y	17	17	1	4	1	4	5	5
4/male	Inversus	Double-Z	18	18	1	4	1	4	5	5
5/female	Palpebralis	Double-Z	21	21	1	5	1	5	5	5
9/female	None	V-Y	18	18	2	3	2	3	5	5
15/male	Inversus	Double-Z	21	21	0	5	0	5	5	5
24/male	Palpebralis	Double-Z	22	22	1	5	1	5	5	5
17/female	Inversus	Double-Z	18	18	0	4	0	4	5	5
21/male	None	V-Y	19	19	2	3	2	3	5	5
13/male	None	V-Y	22	22	1	3	1	3	5	5
10/male	Palpebralis	Double-Z	17	17	1	4	1	4	5	5
9/female	Inversus	Double-Z	20	20	1	5	1	5	6	6
5/female	None	V-Y	21	21	2	4	2	4	5	5
4/female	Inversus	Double-Z	18	18	0	4	0	4	5	5
11/male	Palpebralis	Double-Z	19	19	1	4	1	4	5	5

HFW=Horizontal fissure width, MRD=Margin reflex distance

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equidistant from the central mark. The distance between these two points is equal to desired calculated base length of the triangle. A single brow mark is placed in such a way that it is directly above the center point of the lid. These are now joined to complete the triangle.

Surgical steps

An incision is made at the brow mark described above and a pocket is made within the frontalis muscle $(3 \text{ mm} \times 5 \text{ mm})$ superior to the incision. Lid incisions are given corresponding to the lid marks described above. The silicon rod (aurosling ptosis sling model – T9052; needle length 6.3 cm, needle diameter 920 um, silicon rod length 40 cm, silicon sleeve length 7 mm; ©Aurolabs India) is passed in a triangular fashion from the brow incision through the two lid incisions and back. The lid contour, curvature, and height dictate the amount of silicon rod tightening and adjustment required. The goal end-point for lid height adjustment is to leave the upper lid just at the margin of the superior limbus. The two ends of the silicon rod are inserted into the sleeve and a 6-0 polypropylene suture is tied around it before burying the rod, sleeve, and the suture in the frontalis pocket created above. This brow incision is finally sutured with a single 6-0 polypropylene suture.

Results

Demographic data

The youngest blepharophimosis patient that we operated was 3 years old and the oldest patient was 24 years old. Overall, the cohort study included 11 males and 9 females (male: female ratio = 1.2:1).

Out of the total 20 patients (40 eyes), 13 patients had epicanthal folds, of which 7 patients had epicanthus inversus and six patients had epicanthus palpebralis. These 13 patients underwent Mustarde's double



Figure 1: Schematic diagram to illustrate the single-triangle technique of passing in the frontalis sling in blepharophimosis patients

Z-plasty as their primary surgery whereas the remaining 7 patients underwent V-Y plasty for correction of their blepharophimosis.

Assessment of vertical fissure height

The postoperative VFH was objectively assessed by comparing the pre- and postoperative MRD measurements. The functional success was evaluated by measuring the difference between the pre- and postoperative MRD₁ [Table 1 and Figures 2, 3].

In our cohort study patients, the mean preoperative MRD_1 was 1.0 ± 1.1 mm which increased to 4.1 ± 1.6 mm after surgery. The MRD_1 increased by 3.1 ± 1.7 mm.

Assessment of lid contour

The lid contour was objectively assessed by two independent observers (another oculoplasty surgeon from a different institute). The objective assessment was based on clinical photographs of the seated patients taken under uniform space and lighting conditions. Cosmetic outcome was graded with a score of 0, 1, or 2 to indicate poor, good, and excellent results, respectively.

Out of the 40 eyes that were operated, 33 eyes had a score of 2, 5 eyes had a score of 1, and 2 eyes were scored 0.

Complications and follow-up

The patients were followed up for a period of 12 months after the sling surgery.

Lagophthalmos was reported in 18 eyes of 9 patients. The lagophthalmos at 1 week postoperative was 1.5 ± 0.6 mm which decreased to 0.6 ± 0.4 mm at the 12-month postoperative mark.

Complications included foreign body granuloma in 2 eyes (5%), undercorrection in 6 eyes (15%), and sling



Figure 2: Representative summary of Case 1. (a) Preoperative clinical photograph showing the blepharophimosis, epicanthus inversus, and ptosis. (b) Postoperative clinical photograph after V-Y plasty and single-triangle technique for Ptosis repair



Figure 3: Representative summary of Case 2. (a) Preoperative clinical photograph showing the blepharophimosis, epicanthus inversus, and ptosis. (b) Postoperative clinical photograph after first surgery, i.e., V-Y plasty. (c) Postoperative clinical photograph after single-triangle technique for ptosis repair

extrusion in 1 eye (2.5%). Silicon rod readjustment was performed in cases of undercorrection. No patient in either group developed exposure keratopathy, overcorrection, or recurrence of ptosis in the 6 months of follow-up.

Discussion

Blepharophimosis syndrome (BPES), by definition, includes blepharophimosis, telecanthus, epicanthus inversus, and severe ptosis even though all patients may have varying degrees of some components.^[6] Hence, the surgical management is directed toward solving each of these anomalies individually or together in a staged or a single-procedure approach. Timing of the surgery is decided primarily on the basis of the severity of ptosis, i.e., eyelid position because of its potential to lead to sensory amblyopia and secondarily on the cosmetic aspect.[7-10] The blepharophimosis is corrected with Mustarde's double Z-plasty or Spaeth's V-Y plasty (depending on the presence or absence of epicanthal folds). The ptosis is corrected preferably with frontalis sling surgeries as most blepharophimosis patients have severe ptosis with poor LPS action.

Review of literature yielded not only very few studies in blepharophimosis patients but also did these studies not have a large sample size. Wang *et al.* in 2020 carried out a modified Fox pentagon technique using a polytetrafluoroethylene sling in frontalis suspension to treat blepharophimosis patients and achieved good cosmetic results.^[6] Bhattacharjee *et al.* in 2013 carried 53 out a study in 11 patients to report the functional and cosmetic outcome of a single-stage surgical procedure for the correction of classical blepharophimosis syndrome wherein they performed a Mustarde's double-Z plasty for the epicanthus inversus, transnasal wiring for the telecanthus, and frontalis sling suspension with fascia lata by the modified Crawford technique for correction of ptosis. They reported good and stable cosmetic correction and functional results with shortened treatment time.^[7] Wu *et al.* in 2008 carried out a similar study in 24 patients wherein they employed the double-rhomboid technique in 19 patients and used modified Mauriello's technique of maximal levator resection in 5 patients with acceptable results both in function and cosmesis.^[8] Taylor *et al.* conducted a retrospective review in 2007 in 14 blepharophimosis patients wherein they performed the Crawford technique for frontalis sling suspension about 9–12 months after the Mustarde's double-Z plasty and reported similar effective results.^[9]

Fox pentagon and modified Crawford are the two most commonly preferred techniques for passing the sling in ptosis patients. Fox pentagon utilizes a base length of 12 mm¹ while the base length of each triangle in the modified Crawford double-triangle method is 10-12 mm². Even though these techniques are very effective in achieving the desired lid curvature and height in the normal population, these are unfortunately cumbersome in blepharophimosis patients and lack both surgical and patient satisfaction. First, in the normal population, the HFW ranges from 28 to 30 mm, whereas in blepharophimosis patients, the HFW may be grossly shortened to as low as 14 mm in severely affected cases.^[10] Even the Mustarde's double Z-plasty or Spaeth's V-Y plasty may yield us a HFW of 18-22 mm in such severe scenarios at best. Moreover, in such cases, it becomes difficult to accommodate the base of the Fox pentagon or the modified Crawford for efficient pupillary uncovering. Second, if we attempt to shorten the base length in both these techniques, even though we may be able to uncover the pupil, it would distort the lid curvature such that the upper lid would either flatten out at its highest point (as with the Fox pentagon) or focal notch in an unesthetic manner (as with the modified Crawford) and the desired lid curvature may not be obtained. Finally, as a result of the excessive tension along the silicon sling, especially with a small base length in the Fox pentagon or modified Crawford, in an effort to uncover the pupil and yet maintain esthetic lid contour, the direction of this tension along the silicon rods is not distributed evenly and thus disturbs the dynamics of the techniques resulting in unacceptable lagophthalmos.

Now, the single-triangle technique described by us is not novel in entirety. Only two documented studies have been found in the literature. In 2006, Seider *et al.*^[11] reported a cohort study with a similar technique in adult myogenic blepharoptosis using Tutoplast, a commercially available processed fascia lata allograft (Tutogen Medical and IOP Inc., Costa Mesa, California, USA) costing \$425 per $0.3 \text{ cm} \times 12 \text{ cm}$ sling. Besides the cost, there are other deterrents to the use of this technique. First, their brow pocket necessitated an incision made to the level of the periosteum. Furthermore, an ellipse of redundant lid skin at the lid crease needed to be incised removed so that tarsal plate could be dissected. Third, the Tutoplast was required to be inserted with a Wright's needle suborbicularis and sutured to the upper tarsus with three partial-thickness 6.0 Prolene (Ethicon, Inc., Somerville, New Jersey, USA) sutures. The second cohort study of 15 cases reported in 1985 by Betharia SM. et al.[12] employed a 15-mm curved cutting needle carrying 4-0 Supramid suture as the sling material and utilized a base length 5 mm as this was probably the maximum possible width of the bite that could be taken with the curved cutting needle.

The advantages of the single-triangle technique described by us are worth mentioning. Since we defined the technique using silicon rod as the sling material (aurosling ptosis sling model - T9052; needle length 6.3 cm, needle diameter 920um, silicon rod length 40 cm, silicon sleeve length 7 mm; ©Aurolabs India), we by-and-large circumvented all the suture-related complications (infection, extrusion, and granuloma) that could be encountered with using 4-0 Supramid. Second, the silicon rod can be autoclaved and the total length of 40 cm suffices two patients, thus making it extremely cost-effective in a developing country like India. The third most important advantage is that our technique requires practically no dissection other than a simple frontalis pocket to bury the silicon rod with its sleeve. Fourth, our single-triangle method utilizes a base length of 8-10 mm in blepharophimosis patients depending on the available HFW. Since the base length is short, the lid contour obtained is esthetically much better. Furthermore, since the tension along the silicon rod is directed to the central point above, it effectively elevates the central part of the lid and uncovers the pupil. This calculated base length is neither too short nor too long. Compared to other techniques of passing the sling (i.e., Fox or modified Crawford), the base length may be termed "short" but compared to the available HFW in blepharophimosis patients, the base length is just "adequate." We recommend utilizing a base length 50% available HFW in patients of blepharophimosis while doing the single-triangle technique, such that:

Base length = $\frac{1}{2}$ of available HFW

10 mm for HFW of 20 mm [Table 2].

Even though silicone rod is an effective material in frontalis suspension for blepharophimosis patients, ironically, there is but one inherent disadvantage of the procedure owing to sling material, i.e., silicon.

Table 2: Base length of frontalis sling surgery by						
single-triangle method depends on horizontal fissure						
width in blepharophimosis epicanthus-inversus						
syndrome patients						

HFW (mm)	Base length of triangle (mm)	Number of patients (number of eyes)
<18	8	Three patients (6 eyes)
≥18 but<20	9	Nine patients (18 eyes)
≥20	10	Eight patients (16 eyes)

HFW=Horizontal fissure width

Complications reported after FSS using silicone rod are undercorrection of ptosis, overcorrection of ptosis, lagophthalmos, corneal exposure, recurrence of ptosis due to slippage of sling, and sling site granuloma.^[13-16] The reported incidence of sling site granulomas varies from 7% in fascia lata-based FSS^[13] to 17% with silicone rod-based FSS.^[5,17] Higher rates of complications are associated with nylon monofilament and PTFE.^[18-20]

It may not be possible to achieve an ideal desired MRD_1 of 5 mm in all blepharophimosis cases. In such patients, in fact, an MRD_1 of 3–4 mm is sufficient enough to uncover the pupil and provide an acceptable cosmetic result. More importantly, the MRD_1 and lid contour should be symmetrical in both eyes.

Conclusion

The single-triangle technique has several advantages over both the Fox pentagon technique and the modified Crawford technique in severe blepharophimosis patients. It is not only a much simpler procedure to perform but also has a better control over the curvature of the lid without any central focal notching thus providing better cosmesis and better esthetic results.

Informed consent

Informed consent was obtained from all individual participants included in the study.

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Conflicts of interest

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

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