

Single stage surgery for Blepharophimosis syndrome

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Purpose: The purpose of this study was to report the functional and cosmetic outcome of single stage surgical procedure for correction of the classic components of Blepharophimosis syndrome. **Materials and Methods:** We report a retrospective case file review of 11 patients with Blepharophimosis syndrome operated between July 2004 and April 2008. Each patient had undergone the correction of epicanthus inversus, telecanthus, palpebral phimosis, and bilateral ptosis as a single-stage surgical procedure. Patients were examined and photographed before and after surgery. The mean follow-up was 3 years (range 2-6 years). **Results:** A total of 11 patients (8 males, 3 females) with a mean age of 9 years (range 6-22 years) were reviewed. The surgical outcome was assessed both functionally and cosmetically. The mean preoperative visual acuity was 0.729 ± 0.316 SD and the mean postoperative visual acuity was 0.856 ± 0.277 SD ($P < 0.0428$). There was a statistically significant decrease of astigmatism following ptosis correction ($P < 0.05$), improvement of telecanthus ($P < 0.0001$) in terms of IICD (inner intercanthal distance), and HPFL (horizontal palpebral fissure length) ($P = 0.019$) along with improvement of the superior visual field. The mean preoperative and postoperative IICD was 3 ± 0.33 SD and 2.418 ± 0.189 SD, respectively. There was also a significant postoperative improvement of ptosis ($P < 0.01$), as measured by IPFH (vertical interpalpebral fissure height). All the patients had a stable functional and cosmetic result after a mean follow-up period of 3 years. **Conclusion:** Single-stage surgical correction of the classic anomalies of Blepharophimosis syndrome provides stable and successful long-term results.

Key words: Blepharophimosis, cosmetic, functional, single stage

Congenital Blepharophimosis (BPS) is an oculofacial maldevelopmental syndrome comprising the four classic anomalies of epicanthus inversus, telecanthus, palpebral phimosis, and bilateral ptosis. Moreover, it is also accompanied by poorly developed nasal bridge and hypoplasia of the superior orbital rim. The condition may occur either as an autosomal dominant trait (types 1 and 2), or sporadically.^[1,2] Out of the two clinical subtypes type 1 is associated with premature ovarian failure.^[3] However both types 1 and 2 of BPS are linked to 3q23, and mutation in the FOXL2 gene.^[4] These patients are usually at an increased risk of developing refractive error, strabismus, and amblyopia affecting one or both eyes. Systemic abnormalities like mental retardation, gonadal atrophy with infertility, and lacrimal abnormalities may coexist with BPS.

So far, most of the reported surgical techniques for BPS involve multistage procedures. The treatment includes multiple surgical procedures done between 3 and 9 months interval which require repeated hospitalization and prolonged follow-up. There are a few reports wherein the single-stage surgical procedure had been performed for correcting this syndrome. But most surgeons who advocated one-stage surgical treatment had addressed single-stage correction of epicanthus inversus along with ptosis only and had ignored the correction order of other two steps which may affect the adjustment of vertical and horizontal lengths of eyelids.^[5,6]

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Since 2004, the authors have adopted correction of the classic tetrad of oculofacial anomalies of BPS as a single-stage surgery thereby addressing both the horizontal and the vertical components of BPS. Herein, the authors present the functional and cosmetic outcome of a series of 11 patients of BPS who underwent single-stage surgical correction.

Materials and Methods

This is a retrospective study of 11 patients with Blepharophimosis syndrome who underwent surgical correction of the four classic clinical anomalies in the same sitting as a one-stage procedure. The study was approved by the Institutional Ethics Committee. The pertinent information was gathered by examining the medical records of the patients. Consent for use of these data as well as publication of photographs of the patients was obtained as a routine procedure. The study is a case file review of patients operated for correction of BPS as a single-stage procedure between July 2004 and April 2008. Exclusion criteria included patients of BPS who had undergone correction of only one or two clinical features and not the four classic tetrads as either a single-stage or multistage surgery. Moreover patients with severe ptosis with vertical interpalpebral fissure height (IPFH) of < 2 mm were excluded from the present study (severity of ptosis as classified by Freuh).^[7] Patients' demographics, clinical findings, and functional parameters like refractive status, best corrected visual acuity (BCVA), corneal topography (for documentation of astigmatism), and superior visual field analysis were assessed preoperatively and 3 months postoperatively. Amblyopia was defined as BCVA of 20/40 or less in the affected eye or two lines of difference in Snellen's chart.^[8,9] Cosmetic analysis included measurement of telecanthus by IICD (inner intercanthal distance), ptosis by IPFH (vertical interpalpebral

fissure height), and palpebral phimosis by horizontal palpebral fissure length (HPFL), measured preoperatively and 3 months postoperatively customarily by a rule. Patients having preoperative amblyopia were started with amblyopia therapy a week following surgery and analyzed 3 months postoperatively. Moreover steps of the surgical procedure, complications during surgery, length of follow-up, and complications during follow-up period were also recorded. Data pertaining to functional and cosmetic recovery were tabulated, summarized, and statistically analyzed by Friedman repeated measures of ANOVA.

The surgical procedure was performed in supine position under general anesthesia. Local infiltrative anesthesia consisting of 4 ml of 2% lidocaine with epinephrine 1:100,000 in a 50:50 mixture with 0.5% bupivacaine was injected into the medial canthal region bilaterally and to the root of the nose. A single surgeon performed all the cases by using a consistent technique.

An initial correction of the cosmetic anomalies started with a lateral canthotomy and canthoplasty for correction of palpebral phimosis and to enlarge eyelid fissure, which was followed by correction of epicanthus inversus by the Mustarde's double Z-plasty and telecanthus correction by bilateral shortening and transnasal wiring of medial palpebral ligaments. Finally, attention was directed in correction of the vertical component wherein bilateral ptosis has been corrected by brow suspension, frontalis sling surgery (Crawford double-triangle technique) using autologous fascia lata to elevate the upper eyelid.

In Mustarde's double Z-plasty the flaps were fashioned at an angle of 60° to the line joining the present medial canthus and the site of the proposed medial canthus [Figures 1 and 2]. After fashioning the flaps, the underlying subcutaneous tissue and muscle were excised and the medial canthal tendon was exposed. The periosteum and the medial canthal tendon attachment were dissected from the anterior lacrimal crest and the medial wall of the lacrimal fossa was exposed. In the case of very prominent anterior lacrimal crest, excess bony tissues were removed with the bone punch [Figure 3]. The same surgical procedure was performed bilaterally and a bone drill was then used to make an opening just anterior to the posterior lacrimal crest and drilled across the nose from one side to emerge in a similar position on the other side. A loop of stainless steel wire was passed through the opening to which the medial canthal tendons on either side were sutured along with the subcutaneous tissues, and pulled medially toward the nose. The stainless steel wire was fixed and tightened after titration of midline shift of medial canthus and subcutaneous tissue [Figure 4]. The subcutaneous tissues were sutured with 6-0 polyglactin sutures and skin flaps transposed and sutured with 6-0 silk sutures. Finally, the bilateral ptosis was corrected by frontalis brow suspension with autologous fascia lata harvested from the thigh [Figure 5].

The skin sutures were removed a week later postoperatively. Patients were followed up at 1 week, 1 month, 3 months, 6 months, and annually thereafter.

Results

The records of 11 patients of BPS (eight males, three females) operated between July 2004 and April 2008 were reviewed

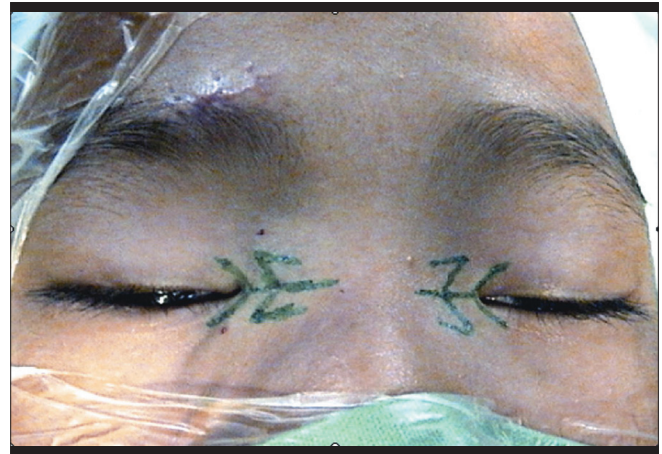


Figure 1: Patient of Blepharophimosis syndrome planned for correction of epicanthal fold



Figure 2: Correction of epicanthus inversus by Mustarde's double Z plasty



Figure 3: Photograph of prominent and laterally displaced anterior lacrimal crest seen in Blepharophimosis syndrome wherein the excess bony tissues removed with bone punch

retrospectively. The mean age was 9 years with a range of 6--22 years. All patients had type 2 Blepharophimosis syndrome and had no positive family history. The mean follow-up was 3 years with a range of 2--6 years. The outcome of the surgery was assessed both functionally and cosmetically, measured preoperatively and 3 months postoperatively.

The functional parameters documented were refractive status, best corrected visual acuity (BCVA), corneal astigmatism

by topography, and superior visual field analysis [Table 1]. Cosmetic analysis included measurement of IICD, IPFH, and HPFL [Table 2].

Out of the 11 patients, three patients were emmetropes, six had asymmetrical mixed astigmatism, one patient had symmetrical myopic astigmatism of -3.50 D at 180° bilaterally and one had anisometropic hyperopic refractive status. The mean preoperative visual acuity was 0.729±0.316 SD

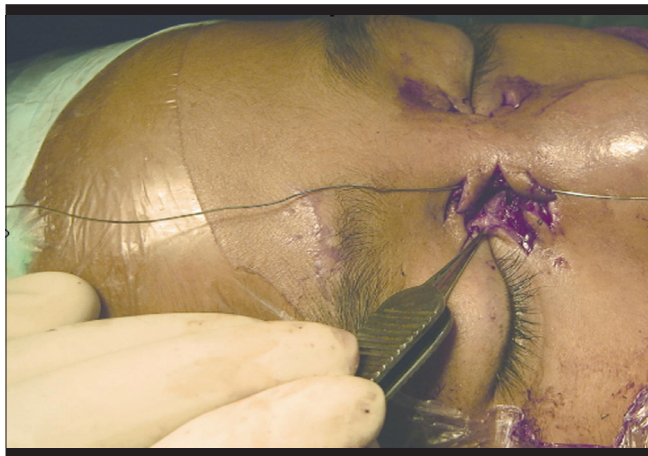


Figure 4: Photograph of a loop of stainless steel wire seen passed through the nose to which the medial canthal tendons on either side along with the subcutaneous tissues were sutured



Figure 5: Bilateral ptosis corrected by frontalis brow suspension with fascia lata harvested from the thigh

Table 1: Preoperative and 3 months postoperative record of best corrected visual acuity (BCVA), refractive status, visual field analysis

		Preoperative				Postoperative					
BCVA		RE		VFD		BCVA		RE		VFD	
OD	OS	OD	OS	OD	OS	OD	OS	OD	OS	OD	OS
20/20	20/60	+2.00@-1.25 x 180°	+3.00@-1.00 x 180°	SFD	SFD	20/20	20/20	+1.25@-0.50 x 180°	+1.00@ -1.50 x 172°	Imp	Imp
20/20	20/20	±	±	-	-	20/20	20/20	+0.50	+1.25@ -1.00 x 160°	Un	Un
20/20	20/20	+1.50 @- 2.00 x 170°	+2.00@ -1.50 x 180°	SFD	SFD	20/20	20/20	UN	+0.50 @-0.50 x 170°	Imp	Imp
20/20	20/20	±	±	-	-	20/20	20/20	±	±	Un	Un
20/40	20/100	+2.50 @-2.25 x 180°	+5.00@ -3.50 x 175°	SFD	SFD	20/20	20/60	+1.25 @-1.00 x 170°	+3.50 @-1.50 x 180°	Imp	Imp
20/40	20/40	+0.50 @-1.00 x 180°	+0.75 @ -2.50 x 170°	-	-	20/20	20/20	+0.50 @-0.50 x 170°	+0.75 @-1.50 x 180°	Un	Un
20/40	20/60	+0.50 @-2.00 x 176°	+1.00@-4.00x 180°	SFD	SFD	20/20	20/20	-0.50 x 180°	+0.50 @-1.00 x 180°	Rd	Rd
20/80	20/80	+2.00@ -2.50x 170°	+3.00 @-2.50 x 180°	SFD	SFD	20/60	20/60	+1.00 @- 1.00 x 180°	+1.00@- 1.00 x 180°	Imp	Imp
20/20	20/20	+0.50@- 3.50 x 180°	+0.75 @-3.50 x 180°	-	-	20/20	20/20	- 1.00 x 180°	-1.00 x 180°	Un	Un
20/20	20/20	±	±	-	-	20/20	20/20	±	±	Un	un
20/20	20/80	+2.50	+4.50	-	-	20/20	20/80	Un	Un	Un	Un

OD: Right eye, OS: Left eye, BCVA: Best corrected visual acuity, RE: Refractive error (in Diopters), VFD: Visual field defect, @: Combined with, SFD: Superior field defect; Rd: Reduced, Imp: Improved; Un: Unchanged.

Table 2: Preoperative and 3 months postoperative record of horizontal palpebral fissure length

Preoperative					Postoperative				
HPFL (mm)		IPFH (mm)		IICD (cm)	HPFL (mm)		IPFH (mm)		IICD (cm)
OD	OS	OD	OS		OD	OS	OD	OS	
15	15	4	4	2.8	25	25	8	9	2.2
15	15	4	4	2.9	25	25	9	9	2.5
20	19	6	6	3.0	27	28	10	10	2.5
16	16	6	6	3.1	26	26	10	10	2.6
16	16	6	6	3.5	26	26	10	10	2.4
14	14	5	5	2.8	25	25	9	10	2.4
15	15	5	5	3.2	25	25	8	8	2.8
16	16	5	5	3.5	25	26	10	10	2.5
18	19	6	6	2.9	26	26	9	9	2.2
20	18	6	6	2.7	27	28	10	10	2.3
21	21	5	5	2.6	26	26	10	10	2.2

(HPFL), interpalpebral fissure height (IPFH), and inner intercanthal distance (IICD)

and the mean postoperative visual acuity was 0.856 ± 0.277 SD ($P < 0.0313$). Though there was a significant decrease of astigmatism following surgical correction of ptosis ($P < 0.05$), the mean refractive error in terms of spherical equivalent was $+0.757 \pm 2.017$ preoperatively and -0.181 ± 0.246 postoperatively ($P = 0.05$).

Four patients having amblyopia had visual improvement by two Snellen lines by third postoperative month following amblyopia treatment which was started a week following surgery. Five patients had shown an impairment of superior visual field preoperatively and on postoperative examination, there was an improvement of visual field in four patients. However, in one patient there was progressive constriction of visual field during the follow-up period of 4 years due to associated fundus changes of retinitis pigmentosa.

In the setting of cosmesis, success was defined as reduction of telecanthus (IICD), ptosis (IPFH), and palpebral phimosis (HPFL) measured preoperatively and 3 months postoperatively. Cosmetic correction of epicanthus inversus was observed by appearance of postoperative prominent caruncles. Although there was cosmetically acceptable correction of all the four classic anomalies postoperatively, telecanthus correction showed a statistically significant improvement ($P < 0.0001$) with a mean preoperative IICD of 3.00 ± 0.33 SD (cm) and postoperative IICD of 2.418 ± 0.189 SD (cm) [Figures 6, 7a,b, 8, and 9]. There has been a significant improvement of ptosis in terms of IPFH. The mean preoperative IPFH was 5.27 mm and postoperative being 9.5 mm ($P < 0.01$). There has been adequate correction of palpebral phimosis with a preoperative mean HPFL of 16.8 mm and postoperative HPFL of 25.85 mm with a difference of 9.05 ($P = 0.0019$).

Two patients had subcutaneous hemorrhage over the nasal bridge in the immediate postoperative period that settled spontaneously within 2 weeks and did not require any treatment. As the levator palpebralis muscle action was

less than 4 mm in all the 11 patients, the ptosis was corrected by bilateral frontalis brow suspension with autologous fascia lata harvested from thigh. That the study did not encounter any rejection or sling related complications was attributed to the use of autologous tissue as sling material. Though all patients showed consistent and acceptable cosmetic correction postoperatively, one patient showed a high superior sulcus deformity on one side 1 month postoperatively as documented in the clinical records [Figure 10]. Overall stable and good long-term results in terms of functional and cosmetic correction was achieved and maintained in all the patients through a mean follow-up period of 3 years, follow-up ranging from 2 years to 6 years.

Discussion

Congenital Blepharophimosis syndrome consists of a constellation of periocular anomalies and is generally recommended for multistage surgery with a belief that planned multiple stage surgery results in better functional and cosmetic recovery. However, there have been some reports of single-stage surgical procedures being done to correct the anomalies of BPS.^[10,11] These reported studies mainly focused on the correction of epicanthal fold and bilateral ptosis. Keracaoglan *et al.* have reported one-stage repair of BPS wherein medial canthoplasty, facial suspension, and widening of the bridge of the nose had been done with the use of the bone graft taken from the iliac crest.^[12] But they had not addressed the correction of telecanthus and palpebral phimosis. Transnasal wiring of the medial canthal ligaments done to correct the telecanthus is considered as a complex manipulation.^[10] However, transnasal wiring gives the best correction of telecanthus in BPS as the patients have prominent anterior lacrimal crest and laterally displaced medial palpebral ligaments of variable degree. Simple refixation of shortened medial canthal tendon results in under correction of telecanthus.^[13] Bilateral transnasal wiring anchors the medial canthal tendons on both sides and pulls each other across the nose and redirects the laterally displaced medial palpebral ligament medially toward the nose, resulting in better correction.^[13] Moreover, this also provides a parallax of decreased width of the nasal bridge. In the present series, in all 11 patients the authors had performed lateral canthoplasty, medial canthoplasty by Mustarde's double-Z technique, transnasal wiring with stainless steel wire, and bilateral brow suspension with fascia lata. In this study, the authors found significant correction of telecanthus postoperatively ($P < 0.0001$). As these patients had very poor levator function, bilateral facial suspension was done when the child's leg was developed enough to obtain the autogenous fascia lata. The authors had achieved good esthetic correction in all the 11 patients. Besides, there was definite improvement of functional parameters in terms of BCVA ($P = 0.0313$), decrease in corneal astigmatism ($P = 0.05$) along with improvement in amblyopia, and improvement of superior visual field. Wu *et al.* reported 31% association of amblyopia in Blepharophimosis syndrome.^[14] Beckengsal *et al.* reported that in their series of 28 patients, 64% had amblyopia with coexistent strabismus, and 24% had amblyopia without strabismus. Moreover, they found 40% of these patients had significant astigmatism.^[15] In the present series 36.36% of patients had amblyopia and had shown visual improvement by 2 Snellen lines by third postoperative month following amblyopia treatment, started



Figure 6: Photograph of Blepharophimosis syndrome, preoperative, case 4



Figure 7: (a and b) Photograph of Blepharophimosis syndrome, postoperative, case 4



Figure 8: Photograph of Blepharophimosis syndrome, preoperative, case 8



Figure 9: Photograph of Blepharophimosis syndrome, postoperative, case 8



Figure 10: Photograph of high superior sulcus deformity seen following ptosis correction in 1 month postoperative of case 2

a week following surgery. However, one patient did not show any improvement in terms of visual acuity and visual field following the surgical procedure due to associated retinitis pigmentosa. Similar association between retinitis pigmentosa and BPS has also been reported by Vedantham *et al.*, and has mentioned that this association may be due to chromosomal anomaly probably in the region of chromosome14.^[16]

Decreased visual acuity in BPS is attributable to either strabismic amblyopia due to loss of eye parallelism or due to isoametropic amblyopia due to refractive defects.

In the present study none had strabismic amblyopia; however 36.36 % had isoametropic amblyopia mainly attributed to high order astigmatism.

Improvement of visual acuity following Frontalis sling surgery for ptosis correction has also been reported by Gustavo Rodríguez *et al.*^[17] Their results showed a visual acuity improvement in 100% of patients, to varying degrees.

Prior to surgery, 72% had visual acuity of 0.1–0.5. Six months postsurgery, with visual rehabilitation, 90.9% exhibited visual acuity of >0.5.

The functional analysis of the results of Tables 1 and 2 were done 3 months postoperatively keeping in the mind the natural wound healing process which is completed by approximately 3 months.

Natural history of wound healing and scar formation undergoes the phases of inflammation (ingrowth of blood vessels and extracellular material 5-6 weeks), proliferation (incision line softens, bleaches, and fades 6-7 weeks) and remodeling (maximum relaxation in 3 months approximately).^[18]

This study is an attempt to correct all the four classic signs of BPS in a single-stage procedure. Therefore in the present study, the results of Tables 1 and 2 are interrelated in a way that IICD, HPFL, and IPFH affect the postoperative results of astigmatism, visual acuity, and visual field.

Bilateral ptosis correction by the Crawford's double triangle technique using autogenous fascia lata effectively enhanced the postoperative IPFH in patients with BPS. The mean preoperative IPFH was 5.27 and postoperative IPFH was 9.5 ($P < 0.01$). The significant result in the present study might be because of inclusion of patients above 6 years with maturity of the musculoskeletal system and exclusion of patients with severe ptosis with IPFH < 2 mm. In this study IPFH measurement alone was taken as a measure of ptosis and had not taken MRD (margin reflex distance) into consideration as it is affected by the chin position. Taylor *et al.* had also reported on their objective analysis of surgery on patients with BPS that MRD measurement for ptosis in pediatric population is often inaccurate as chin-up posturing often results in overestimation of MRD1 and under estimation of MRD 2 with a resultant under estimation of ptosis surgery results.^[19]

Wu *et al.*, in their study on one-stage correction of BPS, focused on severity of syndrome and the surgical outcome based on IPFH and the IICD to HPFL ratio as per the normative database in the Chinese population.^[14] In their study the pre- and postoperative IICD difference was 6.65 and HPFL difference was 7.03 mm. In the present study there has been a similar improvement of telecanthus with a pre- and postoperative difference of 6.00 mm ($P < 0.001$). Moreover there has been adequate correction of palpebral phimosis with a preoperative mean HPFL of 16.8 mm and postoperative HPFL of 25.85 mm ($P = 0.0019$) having a difference of 9.05 mm. In the study by Wu *et al.*, the one-stage corrective procedure on BPS provided acceptable results both in functional and cosmesis. However in their study 30.43% needed a repeat operation which was attributed to a preoperative IPFH < 2 mm (severe ptosis).^[14]

In the present study repeat operation was not performed in any of the 11 patients. This may be attributed to the musculoskeletal maturity of the patients being above the age of 6 years and the exclusion of patients with severe ptosis (IPFH < 2 mm).

Nakajima *et al.* had reported many systemic associations of BPS including heart disease.^[10] The associated systemic problems may not permit multiple exposures to general anesthesia in multiple stage surgeries for BPS. Some authors

mentioned that in consideration of emotional and physical stress imposed in a child by staged operation, one-stage operation is the most desirable method.^[10]

Patients with BPS seen by the authors 7 years prior to the study period were treated with conventional multistage procedure. At that time the authors experienced very high percentage of follow-up losses following the first surgical procedure and refusal for the next stage of surgery either due to emotional factors or refusal to multiple exposures to general anesthesia. This made the authors aware and as such the single-stage surgical technique has been done to address all the shortcomings of the multistage surgical procedure. The authors found that the single-stage procedure has several advantages over the multistage procedure such as need for only one surgical option, decreased hospitalization and recovery time, more cost-effective, and potentially less anxiety for the patients. Moreover, single-stage surgery seems to be a better option when follow-up losses are a factor worth mentioning in the case of multistage surgery for BPS.

The authors conclude that the single-stage surgical procedure for Blepharophimosis Syndrome offers good and stable cosmetic correction and functional results with shortened treatment time. Though this is the first study report of single-stage surgical outcome of BPS in the Indian population having an adequately long follow-up duration of 6 years with all clinical measures performed using the standard protocol, it is subject to methodological limitation due to the small sample size. The other limitation of the study was the noncomparative retrospective design. Thus further studies with larger sample size may shed new light to many of the unresolved queries regarding the single-stage surgery as a line of treatment in patients with BPS to achieve the best possible outcome.

References

1. Stromme P, Sandboe F. Blepharophimosis-ptosis-epicanthus inversus syndrome (BPES). *Acta Ophthalmol Scand* 1996;74:45-7.
2. Maw M, Kar B, Biswas J, Biswas P, Nancarrow D, Bridges R, *et al.* Linkage of blepharophimosis syndrome in a large Indian pedigree to chromosome 7p. *Hum Mol Genet* 1996;5:2049-54.
3. Zlotogora J, Sagi M, Cohen T. The Blepharophimosis, ptosis, and epicanthus inversus syndrome: Delineation of two types. *Am J Hum Genet* 1983;35:1020-7.
4. De Baere E, Dixon MJ, Small KW, Jabs EW, Leroy BP, Devriendt K, *et al.* Spectrum of FOXL2 genemutations in blepharophimosis-ptosis-epicanthus inversus (BPES) families demonstrates a genotype-phenotype correlation. *Hum Mol Genet* 2001;10:1591-600.
5. Zhang HM, Sun GC, Liu Z. Treatment of congenital eyelid syndrome (CES). *Chin J Plast Burn Surg* 1997;13:175-8.
6. Huang WQ, Qiao Q, Zhao R, Wang XJ, Fang XQ. Surgical strategy for congenital blepharophimosis syndrome. *Chin Med J* 2007;120:1413-5.
7. Freuh BR. The mechanistic classification of ptosis. *Ophthalmology* 1980;87:1019-21.
8. Beaconsfield M, Walker JW, Collin JR. Visual development in the blepharophimosis syndrome. *Br J Ophthalmol* 1991;75:746-8.
9. Anderson RL, Baumgartner SA. Amblyopia in ptosis. *Arch Ophthalmol* 1980;98:1068-9.
10. Nakajima T, Yoshimura Y, Onishi K, Sakakibara A. One stage repair of blepharophimosis. *Plast Reconstr Surg* 1991;87:24-31.
11. Dang H, Zhao GC. One-stage surgical reconstruction of congenital

- blepharophimosis syndrome. *Chin J Plast Surg Burn (Chin)* 1993;9:353-4.
12. Karacaoglan N, Sahin U, Ercan U, Bozdogan N. One-stage repair of blepharophimosis: A new method. *Plast Reconstr Surg* 1994;93:1406-9.
 13. Tyers AG, Collin JR. *Colour atlas of ophthalmic plastic surgery*. 3rd ed., Vol. 18. Butterworth- Heinemann: Elsevier Science Ltd; 2008. p. 435.
 14. Wu SY, Ma L, Tsai YJ, Kuo JZ. One-stage correction for blepharophimosis syndrome. *Eye (Lond)* 2008;22:380-8.
 15. Beckingsale PS, Sullivan TJ, Wong VA, Oley C. Blepharophimosis: A recommendation for early surgery in patients with severe ptosis. *Clin Exp Ophthalmol* 2003;31:138-42.
 16. Vedantham V, Jethani J, Agarwal A, Vijayalakshmi P. Retinitis Pigmentosa, associated with blepharophimosis, blue dot cataract and primary inferior oblique overaction: A New Syndrome Complex. *Indian J Ophthalmol* 2007;55:150-1.
 17. Salinas GR, Centelles IA, Rondón IR, Sánchez Tde J, Salas SV, Oduardo MD. Improved visual acuity after frontalis sling surgery for simple congenital ptosis. *MEDICC Rev* 2011;13:23-8.
 18. William PD, Chen, Khan JA, McCord CD Jr. *Colour Atlas of Cosmetic and Oculofacial surgery*. 1st ed., Vol. 3. Butterworth Heinemann: Elsevier Science Ltd; 2004. p. 20.
 19. Taylor A, Stike PW, Tyers AG. Blepharophimosis-ptosis-epicanthus inversus syndrome: Objective analysis of surgical outcome in patients from a single unit. *Clin Exp Ophthalmol* 2007;35:262-9.

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