ORIGINAL ARTICLE

Cosmetic Outcome of Y-V Medial Canthoplasty in Blepharophimosis Syndrome

Ibrar Hussain and Tajamul Khan

ABSTRACT

Objective: To determine the cosmetic outcome of Y-V medial canthoplasty with medial canthal tendon shortening in Blepharophimosis syndrome.

Study Design: Quasi experimental study.

Place and Duration of Study: Department of Ophthalmology, Khyber Teaching Hospital, Peshawar, Pakistan, from June 2005 to April 2008.

Methodology: Thirteen patients of Blepharophimosis syndrome aged 4 years and above were included in the study. Oculoplastic assessment including measurement of horizontal length of palpebral fissure, ptosis and intercanthal distance were done. All patients underwent Y-V medial canthoplasty with medial canthal tendon shortening by a single surgeon. Same measurements were repeated one week postoperatively and then 6 weeks after surgery and results compared with pre-operative findings.

Results: Twenty six eyes of 13 patients were included in the study. Ten (77%) patients were female and 3 (23%) were male. Mean age of all patients was 13.54 ± 6.99 years with a range from 4 to 28 years. Mean horizontal length of palpebral fissure of all 26 eyes was 22.88 mm before surgery and increased to 26.77 mm after surgery. Vertical pre-operative and postoperative height of palpebral fissure was 5.69 mm and 5.34 mm respectively. Mean pre-operative intercanthal distance of 37.46 mm was reduced to 32.08 mm postoperatively. Epicanthus inversus which was present in all (100%) eyes before surgery was successfully treated in 22 (84.6%) eyes after surgery.

Conclusion: Y-V medial canthoplasty with medial canthal shortening in Blepharophimosis syndrome has excellent cosmetic outcome.

Key words: *Blepharophimosis syndrome. Y-V medial canthoplasty. Cosmetic outcome medial canthal shortening.*

INTRODUCTION

Blepharophimosis syndrome (BPES) is a rare autosomal dominant disorder characterized by bilaterally shortened horizontal palpebral fissure (Blepharophimosis), severe ptosis, a vertical fold arising from the lower lid and inserts medially in the upper lid (epicanthus inversus) and an increased intercanthal distance (telecanthus, Figure 1).[1]

It was first described by Dimitry in 1921.[2] The mutation causing this disorder is found in FOX L2 gene, a forkhead transcription factor, located in 3q23.[3,4] Although many patients with this syndrome have an affected parent (usually father), a conspicuous number of sporadic cases have also been reported.[5] Sporadic cases account for about one half of the diagnosed cases.[6] Chaudry *et al*. reported this condition affecting four members of a Pakistani family across three generations.[7]

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Surgical treatment of the condition is needed not only for cosmetic purpose but also to prevent amblyopia which is present in about half of the cases.[8,9] Surgery is recommended before going to school (usually at the age of 4 years) except when there is risk of amblyopia when early intervention is needed. Most of oculoplastic surgeons prefer a two-stage surgery. In the first stage, horizontal dimensions of palpebral fissure is lengthened by medial canthoplasty combined with medial palpebral ligament shortening and in the second stage ptosis is corrected. Known procedures for first stage surgery are Mustarde's double-Z plasty and Y-V medial canthoplasty. Parameters for good cosmetic outcome of these procedures are increased in horizontal length of palpebral fissure, disappearance of epicanthus inversus and reduction in telecanthus. Y-V medial canthoplasty is a relatively easier procedure to learn and perform as compared to Mustarde's double-Z plasty.

The purpose of this study was to determine the cosmetic outcome of Y-V medial canthoplasty with medial canthal tendon shortening (first stage) in patients with Blepharophimosis syndrome.

METHODOLOGY

This quasi experimental study was performed at the Department of Ophthalmology, Khyber Teaching Hospital, Peshawar, Pakistan, from June 2005 to April 2008.

Twenty six eyes of 13 patients of BPES were included in the study. Patients below the age of 4 years were excluded. Those patients, who had already undergone ptosis or squint surgery were also excluded from the study.

Detailed history and ocular examination was performed on each patient. Special attention was given to oculoplastic assessment including measurement of horizontal length of palpebral fissure, ptosis and telecanthus.These measurements were taken in mm using simple ruler. All patients underwent Y-V medial canthoplasty with medial canthal tendon shortening by single surgeon (IH). Same measurements were repeated one week postoperatively and then 6 weeks after surgery. From thereafter, the patients were evaluated as and when they visited the outpatient department.

The data was evaluated and analysed using Statistical Package for Social Sciences (SPSS) version 14. For nominal variables like gender and postoperative scarring, frequencies were taken and for numerical variables like horizontal and vertical length of palpebral fissure and amount of telecanthus, means with standard deviation were considered. Paired sample t-test was used to find out the correlation between pre-operative and postoperative measurement of above mentioned numerical variables and p-value calculated. P-value < 0.01 was considered as significant.

RESULTS

Twenty six eyes of 13 patients were included in the study. Ten (76.9%) were female and 3 (23.1%) were male. The mean age of the patients was 13.54 ± 6.99 years ranging from 4 to 28 years. The duration of follow-up ranged from 6 to 30 months.

The mean pre-operative horizontal length of palpebral fissure (HPFL) was 22.88 ± 2.63 mm. Postoperatively this figure increased to 26.77 ± 2.86 mm (p < 0.001). Mean pre-operative vertical height of palpebral fissure was 5.69 ± 2.08 mm. Postoperatively this figure changed to 5.34 ± 2.14 mm (p = 0.55). Mean pre-operative distance between right and left medial canthus i.e., intercanthal distance (ICD) was 37.46 ± 5.82 mm and it reduced to 32.08 ± 5.04 mm after surgery (p = 0.001).

Hence, pre-operative ICD/HPFL ratio of 1.66 ± 0.35 decreased to 1.22 ± 0.25 after surgery (p = 0.001, Table I and Figure 2).

Epicanthus inversus was totally corrected in 22 (84.6%) eyes while it was still present in 4 (15.4%) eyes. Till the last follow-up (minimum 6 months), moderate scarring in the medial canthal area was present in 16 (61.5%) eyes and minimum scarring in 10 (38.5%) eyes. Post-operatively caruncle became visible in all 26 (100%) eyes.

Table I: Postoperative changes in horizontal palpebral fissure length, vertical height of palpebral fissure and intercanthal distance and their significance.

Variable	Pre-operative	Postoperative	p-value
Horizontal palpebral fissure length (HPFL)	22.88±2.63 mm	26.77±2.86 mm	< 0.001
Vertical height of the palpebral fissure	5.69±2.08 mm	5.34±2.14 mm	0.552
Intercanthal distance (ICD)	37.46±5.82 mm	32.08±5.04 mm	0.001
ICD/HPFL ratio	1.66±0.35	1.22±0.25	< 0.001

Figure 1: Blepharophimosis syndrome showing Blepharophimosis, ptosis, telecanthus and epicanthus inversus.

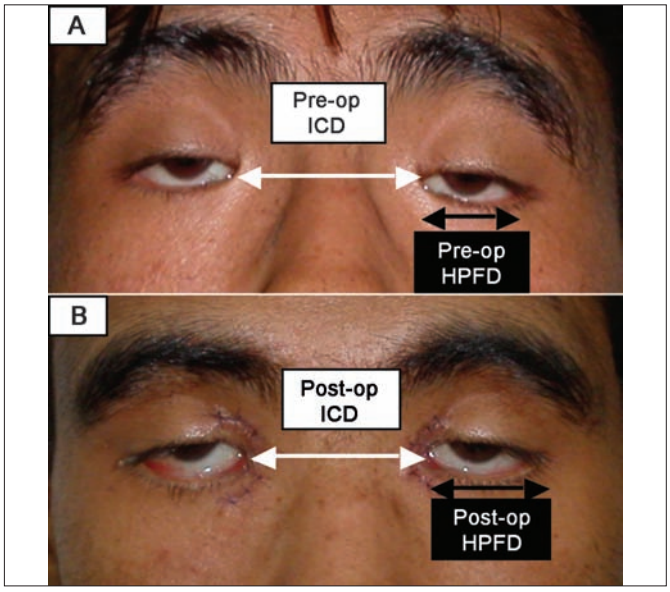

Figure 2: (A) Shows pre-operative horizontal palpebral fissure distance (HPFD) and intercanthal distance (ICD). **(B)** Shows changes in HPFD and ICD after surgery.

DISCUSSION

Blepharophimosis syndrome is a complex condition and its management requires input of several specialists including paediatric ophthalmologist, orthoptist and oculoplastic surgeon. Various methods of treatment to improve cosmetic appearance have been advocated.[10,11] In the present study, Y-V medial canthoplasty with medial canthal shortening was adopted as first

stage surgery to deal with Blepharophimosis, epicanthus inversus and telecanthus.

Timing of eyelid surgery is controversial. It involves weighing the balance of early surgery to prevent amblyopia and late surgery to allow for more reliable ptosis measurement. The unoperated cases presenting in adulthood with bilateral ptosis were bilaterally amblyopic whereas those children who underwent surgery early did better.[9] Surgery is hampered by dysplastic structure of eyelids.[12] The surgical management traditionally involves a medial canthoplasty for correction of blepharophimosis, epicanthus inversus and telecanthus at 3 – 5 years of age (first stage) followed a year later by ptosis correction (second stage).[13] Some surgeons insist on this two stage surgery,[14,15] while few others recommend one stage procedure.[16-18] In this series, results of first stage surgery were evaluated and the age range for this was 4 – 28 years. Patients who presented before the age of 4 years were not considered for first stage surgery. Backingsale and colleagues recommended that patients with severe ptosis have their ptosis corrected before 3 years of age and other patients should undergo surgery before 5 years of age.[12]

For medial canthoplasty, most of the surgeons perform Mustarde's double-Z technique while others adopt Y-V technique. In present study, latter technique was adopted and horizontal palpebral fissure length (HPFL) increased from 22.88 ± 2.63 mm to 26.77 ± 2.86 mm ($p < 0.001$) while intercanthal distance (ICD) reduced from 37.46 ± 5.82 mm to 32.08 ± 5.04 mm ($p < 0.001$) and hence ICD to HPFL ratio of 1.66±0.35 reduced to 1.22 ± 0.25 ($p < 0.001$). Li and colleagues from China,[19] who also adopted multistage surgery technique, reported that pre-operative ICD to HPFL ratio of 2.04 ± 0.25 reduced to 1.25 ± 0.13 ($p < 0.001$). In a study by Taylor and colleagues, pre-operative intercanthal distance to horizontal palpebral fissure length ratio (ICD/HPFL) was 1.26 – 1.60 (non BPES = 1.0). Median reduction post-surgery was 26% ($p = 0.001$).[20] In a study by Wei-Qing and colleagues, who adopted one-stage correction of Blepharophimosis syndrome and performed double-Z technique for medial canthoplasty, HPFL increased from 20 mm to 27 mm and ICD reduced from 40 mm to 34 mm.[21]

In this study, there was also some modification in shortening the medial canthal ligament i.e., instead of anchoring the cut end of ligament to periosteum of anterior lacrimal crest, both medial canthal ligaments were fixed together under deep fascia of dorsum nasi. In another study by Sebastia and colleagues from Brazil, who adopted the same technique as by Wei-Qing, it was reported that mean HPFL increased from 20.90 ± 2.14 mm to 26.36 ± 1.4 mm, mean ICD reduced from 42.45 ± 2.19 to 32.07 ± 1.96 mm and the ICD/HPFL ratio reduced from 2.04 ± 0.14 to 1.23 ± 0.09 ($p < 0.001$).[22] Ren and colleagues in their study proved that HPFL increased from 13 – 22 mm to 24 – 32 mm and ICD reduced from 33 – 44 mm to 29 - 34 mm.[23] Epicanthal fold was eliminated in 84.6% patients in the present series while in the above mentioned studies,[4,8] it was corrected in 100% of cases and caruncle was unmasked. Till the last follow-up (minimum 6 months) in the present study, moderate scarring in medial canthal area was present in 61.5% cases and mild scarring in 38.5% cases. In one of the above mentioned studies,[20] 50% of the cases had mild scarring in the medial canthal region.

Comparison of the results of the present series with the above mentioned studies show that results are comparable with international studies in tackling Blepharophimosis and telecanthus, while slightly lacking in tackling epicanthal fold and medial canthal scarring.

CONCLUSION

Y-V medial canthoplasty with medial canthal shortening in Blepharophimosis syndrome has excellent cosmetic outcome.

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