Original Article

Reconstruction of Congenital Lid defects

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Purpose: To study the cosmetic results in congenital lid defect repairs.

Material and Methods: Retrospective case series review of ten patients operated during the last five years (2006 to 2011). All patients with congenital lid defects were included. These included patients with colobomas of upper or lower lid and abortive cryptophthalmos. Reconstruction was done using rotation flaps or lid sharing procedures by a single surgeon.

Results: Four patients underwent direct closure, four patients underwent Tenzel flap, one underwent composite graft and one underwent Cutler Beard flap. There were three patients with abortive cryptophthalmos who also underwent mucous membrane graft or conjunctival advancement. Three patients had good, four patients had satisfactory and three patients had poor results post-operatively. Three patients had dehiscence of wound after surgery which required further reconstruction. Exposure keratopathy due to delayed presentation was seen in three patients while cornea was scarred in three cases of abortive cryptophthalmos. Mild upper lid notch was seen in three patients and mild upper lid symblephron was seen in one patient.

Conclusion: Early surgery with adequate sized advancement flap for closure is the key to good cosmetic result.

Congenital coloboma is partial or full thickness lid defect[1] which is unilateral or bilateral.[2,3] The upper lid colobomas are more common and are present at the junction of middle and medial third of lid. The lower lid coloboma is present at the lateral third of lid. In abortive crytophthalmos, the lids are replaced by a layer of skin which is fused with microphthalmos. These lid defects are associated with systemic conditions like Goldenhar syndrome,[4] Treacher Collins syndrome and Fraser syndrome. The exact cause is not known but genetic and environmental factors are involved. Any delay or interference in the union of mesodermal sheets with the frontonasal processes or maxillary processes leads to lid defects.

The main complications resulting from lid defects are exposure keratitis, corneal opacity and visual deprivation leading to amblyopia.[5] The ideal treatment of lid defect is early surgical reconstruction to restore the anatomical structure of eyelid.

MATERIAL AND METHODS

A retrospective review of a series of 10 patients with upper and lower lid defects was conducted at Bahawal Victoria Hospital, Bahawalpur and Mayo Hospital Lahore from 2006 to 2011. The age of the patients ranged from 5 days to 2 years with a mean of 6 months. Out of these 10 patients 6 were male and 4 were female (Table 1). There were 8 (80 %) cases with defects of upper lid and 2 (20%) with defects of lower lid. In the series of upper lid defect patients, 3 cases had abortive cryptophthalmos. The extent of the defect varied from less than one third to more than half of the horizontal lid dimension. Only patients with congenital lid defects were included. Family history for any congenital lid defect was taken with details of pregnancy and delivery. Thorough ocular and systemic examination was done. Possible ocular associations like coloboma of iris and choroid, symblepharon, exposure keratopathy (Fig. 1), corneal opacity, nystagmus, trichiasis, dermolipoma and lacrimal abnormality were noted. Pre and post

operative photographs were taken. Reconstruction was done by using direct closure (Fig. 2), lid sharing procedure, Tenzel rotational flap and Cutler Beard technique (Fig. 3) by a single surgeon (Table 2). The patients were followed up at weekly interval for 1 month and then at every month for 6 months. The mean follow up was 6 months (2 months to 1 year). Assessment of the eyelid stability and corneal status was noted.

Table 1: Gender Distribution

Gender	No. of Patients n (%)
Male	6 (60)
Female	4 (40)

Table 2: Procedures performed for reconstruction of lid coloboma

Procedure Performed	No. of Lids n (%)
Direct closure	2 (20)
Direct closure and Lateral cantholysis	2 (20)
Tenzel semicircular flap	4 (40)
Mustarde cheek rotation flap	0
Cutler - Beard Technique	1 (10)
Hughes Procedure	0
Composite graft	1 (10)

RESULTS

Reconstruction was done by using direct closure, lid sharing procedure, Tenzel rotational flap and Cutler Beard technique by a single surgeon (Table 2). The success of post operative results was defined as "Good" if it was cosmetically acceptable with no complication, "Satisfactory" if it was adequate cosmetically with minor complications and "Poor" if it was inadequate cosmetically. 3 (30%) patients had good while 4 (40%) patients had satisfactory and 3 (30%) patients had poor post operative results. 3 (30%) patients had wound dehiscence after surgery which required further reconstruction. Exposure keratopathy (Fig. 1) was seen in 3 (30%) patients while corneal scarring occurred in 3 (30%) of cases of abortive crytophthalmos due to delayed presentation for surgery. These patients were referred to corneal surgeon for corneal graft. Other complications included mild upper lid notch in 3 (30%) cases and mild upper lid symblepharon in 1 (10%) case (Table 3).

Table 3: Postoperative Complications

Complications	No. of Lids n (%)
Exposure keratitis	3 (30)
Corneal scarring	3 (30)
Bulking of skin	0
Orbital hemorrhage	0
Marginal entropion	0
Marginal ectropion	0
Huge scar mark	0
Ischemia necrosis	0
Symblepharon	1 (10)
Punctual occlusion	0
Wound dehiscence	3 (30)
Lateral tissue sag	0
Unstable upper lid	0
Shallow depression	0
Skin hair irritation	0
Lid notching	3 (30)

DISCUSSION

Anatomically eyelid is divided into two lamellae. The anterior lamella consists of skin and orbicularis oculi muscle while posterior lamella consists of tarsus and conjunctiva. Both these lamellae should be replaced in the repair of lid defects.

For lid reconstruction, the defects are classified into small, medium and large according to the size of defect[6]. Small defects are less than one third of horizontal dimension of the lid margin, are closed directly[7]. Medium defect are one third to half of the horizontal lid dimension. These are repaired as direct closure with lateral cantholysis, Tenzel semicircular

flap[8] or Mustarde cheek rotation flap.[9] Large defects are more than half of the horizontal dimension of the lid. These defects are repaired with lid sharing procedure like Cutler Beard technique,[10] Hughes procedure[11,12] or composite graft.[13] In composite graft, the full thickness pentagonal graft is taken either from a donor eye or from contralateral eyelid when the defect involves the only seeing eye of the patient. The aim of eyelid reconstructive surgery is to reconstruct anatomically and cosmetically better eyelid with protection of globe and restoration of good vision as early as possible.

The main vision threatening complication resulting from the congenital lid defect is keratopathy. Aggressive treatment of exposure keratopathy by the referring pediatrician is essential to prevent permanent corneal scarring. We found that poor lubrication of the cornea had led to permanent corneal scarring in 3 (30%) of our cases. Early repair is required to combat this complication. These patients were referred to corneal surgeon. Proper alignment of the anatomical structures is a key factor for good results. The data shows good / satisfactory results (70%) in our series. The lid is well vascularized and tissue survives reasonably well. If the lid margins are not precisely aligned, complications like distortion, trichiasis and notching of the lid occur. Therefore adequate sized flap should be made to prevent these complications. Lateral cantholysis of lower lid without reestablish-ment of lateral support may cause lower lid laxity and ectropion.

Fig. 1: Exposure Keratopathy due to coloboma.

Fig. 2: Director Closure for Bilateral upper lid coloboma reconstruction with follow-up pictures for 5 years.

Fig. 3: Cutler Beard Technique for large upper lid defect.

In direct closure, wound is closed along the anatomical lines. This causes minimum tension on the wound. The lower lid is more lax so it can be pulled maximally to close the defect. But in upper lid excessive pull can lead to ptosis. After lid sharing procedures, if the flap is cut near the newly formed lid margin, the lid margin can roll inward[14]. Lid sharing procedure is also a major cause of visual deprivation amblyopia because of prolong period of closure of lids especially in very young patients. The marginal artery is situated 3 - 4 mm from lid margin. During bridging flap, inadequate blood supply can lead to flap necrosis.[15] This can be avoided if the horizontal incision to form flap is made about 3 - 4 mm from lid margin to avoid injury to vascular arcade. The vertical height of the pedicle of graft must be at least 4 - 6 mm. The upper lid defect repair can damage the lacrimal system leading to epiphora. The graft should be of proper size to cover the gap, otherwise the contraction will lead to scar formation. Proper homeostasis of bed and cauterization of bleeding vessel prevents the flap necrosis and hematoma formation.[16] Oedema of lids is common due to interruption of lymphatic drainage. It resolves after several days. Cold ice packs are indicated. The flaps should be broad. Broad flaps retract less than narrow flaps. The retraction of narrow flap leads to distortion of constructed lid. Sometimes altered pigmentation occurs in the scars. It usually improves with time. In Tenzel semicircular flap, the line of incision from the lateral canthus should not be horizontal but a continuation of the lid to be constructed. A shallow depression will form in the lateral part of lid if the incision is horizontal.

Visual outcome after penetrating keratoplasty in children is significantly worse with congenital corneal opacity due to low scleral rigidity, forward displacement of lens iris diaphragm cataract formation and post surgical anterior segment inflammation leading to corneal graft rejection.[17]

CONCLUSION

Early surgery with adequate sized advancement flap for closure is the key to good cosmetic result. Early surgical treatment prevents pre-operative complications and produces acceptable cosmetic results. There is a high rate of wound dehiscence in cases with tight closure. Lid sharing procedure should be reserved for cases with wound dehiscence.

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