

# Repair of Congenital Bilateral Upper Eyelid Colobomas with Mustard Flap in a 6<sup>th</sup> Week-Old Girl: Case Report

## Altı Haftalık Bir Kız Çocuğunda Mustard Flebi ile Doğumsal Bilateral Kapak Kolobomu Tamiri

Lale KÖZER BİLGİN, MD,<sup>a</sup>  
Barış YENİAD, MD<sup>a</sup>

<sup>a</sup>Department of Ophthalmology,  
İstanbul University  
İstanbul Faculty of Medicine, İstanbul

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Yazışma Adresi/Correspondence:  
Barış YENİAD, MD  
İstanbul University  
İstanbul Faculty of Medicine,  
Department of Ophthalmology, İstanbul,  
TÜRKİYE/TURKEY  
byeniad@yahoo.com

**ABSTRACT** A 7-day-old girl was referred to our clinic with congenital bilateral upper eyelid colobomas larger than the two-third of the entire eyelid and bilateral corneal perforations. Urgent penetrating keratoplasty were conducted in both eyes. Since the patient lacked adequate lower eyelid tissue, eyelids were reconstructed with temporary free tissue grafts. At the end of the 4<sup>th</sup> week, the free grafts necrosed but the globe maintained its integrity. At the 6<sup>th</sup> week, full-thickness lower eyelid rotational flaps were used for reconstruction. During the examinations the ophthalmic ultrasound revealed a normal fundus. The responses with flash visual evoked potential (VEP) were normal. The patient was referred to a center for low vision. In large eyelid colobomas, amblyopia may be unavoidable because of the compulsive treatment choices. However, in these severe cases the primary goal is to protect the eye and attain a self-sufficient level of sight.

**Key Words:** Coloboma; amblyopia; surgical flaps

**ÖZET** Yedi günlük kız çocuğu, kapakların üçte ikisini kaplayan bilateral üst kapak kolobomu ve korneal perforasyon ile kliniğimize başvurdu. Her iki göze acil penetran keratoplasti uygulandı. Aynı seansta her iki gözde yeterli alt kapak dokusu bulunmadığından serbest kapak greftleri ile kapak rekonstruksiyonu yapıldı. Dört hafta sonra kapak greftlerinde nekroz gelişti ancak glob bütünlüğünü korundu. Hasta 6 haftalık olduğunda tam kat rotasyonel flepler ile kapak tamiri yapıldı. Takipler süresince oftalmik ultrason ile normal bir fundus görünümü tespit edildi. Flaş görsel uyarılmış potansiyel (VEP) ile cevaplar normal olarak kaydedildi. Hasta az görenler için oluşturulan bir merkeze yönlendirildi. Geniş kapak kolobomlarında zorunlu tedavi seçenekleri nedeni ile ambliyopi kaçınılmaz olabilir. Ancak ciddi olgularda öncelikli amaç gözü korumak ve kendine yetecek bir görmenin sağlanması olmalıdır.

**Anahtar Kelimeler:** Kolobom; ambliyopi; cerrahi flep

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Congenital coloboma is a primary defect of the eyelid that develops through an embryological cleft. Inadequacy in the migration of the ectoderm or mesoderm flowing to the eyelid between the 5<sup>th</sup> and 7<sup>th</sup> weeks in the uterus causes a cleft to develop at the eyelid edge.<sup>1</sup> It typically appears at the medial one third of the upper eyelid, and the lateral one third of the lower eyelid. The main risks of the congenital eyelid coloboma is, the loss of the eye due to exposure keratitis and corneal perforation as a result of the corneal exposure. Bilateral colobomas are usually part of a syndrome and isolated bilateral eyelid colobomas are an infrequent entity.<sup>1</sup>

In this report, we discuss the treatment of a case with bilateral isolated coloboma larger than the two-third of the entire upper eyelid and bilateral corneal perforations.

## CASE REPORT

A 7-day-old girl was referred to our clinic with a large defect of both eyelids and corneal perforation. We examined bilateral upper eyelid coloboma larger than the two-third of the entire eyelid as well as the corneal perforation and ectopic lens (Figure 1). After the informed consent was obtained, the surgery was planned.

## SURGERY TECHNIQUE

Bilateral penetrating keratoplasty were conducted and two layers of continuous amniotic membrane that covered the whole ocular surface with continuous 8/0 Vicryl sutures were placed to create the upper fornix. Since the patient lacked adequate lower eyelid tissue due to very young age, and an opening of the eyelid was desirable for the purposes of monitoring the cornea following penetrating keratoplasty, a temporary upper eyelid was created with a free tissue graft from the thigh. At the end of the 4<sup>th</sup> week, the free grafts necrosed but the globe maintained its integrity (Figure 2).

During her 6<sup>th</sup> week of life, a full-thickness lower eyelid graft (Mustard flap) corresponding to the defective areas on both upper eyelids was performed and amniotic membrane was transplanted for reconstruction (Figure 3). In the 7<sup>th</sup> week of the operation, the upper and lower eyelids were created after the flaps were opened under general anesthesia. During the follow-up, ophthalmic ultrasound demonstrated no other pathologies of the vitreous and retina. The responses with flash VEP were normal.

The patient was referred to a center for low-vision. Now the patient is 15 months old, and observed to have developed a sense of light tracking, ability to discern coloured objects, and hand coordination. At the last biomicroscopic examination, the upper and lower eyelid contours for both eyes were normal, symblepharons were observed extending from both upper eyelids towards the cornea,

both corneas were opaque and vascularized (Figure 4). Despite the successful eyelid surgery, bilateral buphtalmos that caused inadequate eyelid closure were detected during the follow-up.

## DISCUSSION

Various theories are discussed for the formation of coloboma, the exact reason of which is unknown. Some intrauterine factors such as a decrease in placenta circulation, amniotic bands, inflammation, and an abnormal vascular system have been claimed to have an effect.<sup>2</sup>

In defects of the eyelid not covering the cornea or the conjunctiva, a wide range of ocular surface anomalies from simple conjunctival hyperaemia to corneal perforation may occur. The exposure of the cornea especially in upper eyelid coloboma is inevitable. Therefore, the major goal of coloboma treatment should be the protection of the cornea if possible, and ensure the globe integrity.

The size of the coloboma is an important factor for the treatment protocol. If the coloboma is involving less than the half of the entire eyelid and the cornea is healthy, surgical correction may be postponed until the second year of the patient with frequent examinations. More eyelid tissue may be available for reconstruction. If the surgical intervention is decided, various techniques may be performed. A coloboma of approximately one quarter of the eyelid margin can be closed directly. For a defect involving the one third of the eyelid, the semi-circular flap is commonly the treatment of choice.<sup>3-5</sup> If the defect involves up to one half of the eyelid margin, Cutlar-Beard procedure, full-thickness lower eyelid rotation flap or tarsomarginal grafts must be employed.<sup>5</sup> However, since full-thickness lower eyelid flaps cause severe amblyopia, especially in unilateral cases, it must be used very selectively. In a study of Seah et al, they concluded that congenital upper eyelid colobomas are a potential threat to vision and a significant cosmetic problem later in life. They also informed that eye-sharing techniques involve long periods of occlusion of the eye during the critical period of visual development so this amblyogenic factor should be remembered.<sup>6</sup>



**FIGURE 1:** Bilateral upper eyelid coloboma and corneal perforation before surgery



**FIGURE 2:** Necrosis of free tissue grafts in both eyelids.

In our case, because of the bilateral corneal perforation and lens luxation, we primarily performed penetrating keratoplasty to protect the eye. Once the integrity of the eye was achieved, both coloboma were closed with amniotic membrane and free tissue grafts. The patient was examined biomicroscopically every day and hospitalised for one and a half months. Since the skin grafts did not survive and no pathologies other than those expected developed during this interval, the defects on the upper eyelids were closed with bilateral lower eyelid full-thickness rotational flaps.

Large upper eyelid colobomas are severe anatomical defects that threaten the visual acuity. Multiple surgeries may be needed to get successful results. In our case bilateral spontaneous corneal perforations contributed to the eyelid colobomas. Therefore not only the eyelid surgery, also the re-

sults of the penetrating keratoplasties had an effect on the prognosis. The progressing buphtalmos was another factor that reduced the chance of success. Although we have plausible results for the eyelid surgery, the result for the visual acuity was limited. However, in these cases, the primary objective should be to protect the eye, and to initiate low-vision exercises to improve the visual acuity as possible. The patient must be monitored closely for visual development, and the ultimate goal should be ensuring that the patient attains a self-sufficient level of sight when the patient is older.



**FIGURE 3:** Full-thickness lower eyelid graft of the left eye.



**FIGURE 4:** The normal contour of upper eyelids of the 15 month-old patient. (Notice the symblepharon and vascularized corneas)

## REFERENCES

1. Collin JR. Congenital upper lid coloboma. *Aust N Z J Ophthalmol* 1986;14(4):313-7.
2. Mustardé JC. Congenital soft tissue deformities. In: Smith BC, Della Roca RC, Nesi FA, Lisman RD, eds. Vol 2. *Ophthalmic Plastic and Reconstructive Surgery*. 1<sup>st</sup> ed. St Louis: CV Mosby Co; 1987. p1238-61.
3. Tenzel RR, Stewart WB. Eyelid reconstruction by the semicircle flap technique. *Ophthalmology* 1978;85(11):1164-9.
4. Kidwell ED, Tenzel RR. Repair of congenital colobomas of the lids. *Arch Ophthalmol* 1979;97(10):1931-2.
5. Cutler NL, Beard C. A method for partial and total upper lid reconstruction. *Am J Ophthalmol* 1955;39(1):1-7.
6. Seah LL, Choo CT, Fong KS. Congenital upper lid colobomas: management and visual outcome. *Ophthal Plast Reconstr Surg* 2002;18(3):190-5.