

Repair of Bilateral Upper Eyelid Colobomas in Infants

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Eyelid colobomas are congenital abnormalities that result from derailed or incomplete cell migration in embryogenesis. The physical result is a discontinuity in the eyelid margin. These malformations may occur in the upper or lower eyelids and may affect a variable portion of the eyelid margin.

Jacques Guillemeau first described eyelid colobomas in 1585. John Mustardé later observed that upper eyelid colobomas may occur 1) as an isolated condition, 2) with facial abnormalities or 3) as a component of a larger systemic syndrome. Facial defects that may be associated with eyelid colobomas include the loss of eyebrow hair, a less prominent supraorbital margin, the absence of an upper eyelid fornix, and a bifid nose.¹ Among the syndromes that may include eyelid colobomas are Goldenhar, Treacher Collins, Delleman, Fraser and nasopalpebral lipoma coloboma syndrome.

Visual Consequences

Normally, eyelid tissue lubricates the corneal surface and protects it from the external environment. Lack of protective eyelid tissue predisposes the cornea to opacification and visual loss, including blindness. These devastating potential consequences underscore the importance of promptly restoring normal eyelid function in these patients.

The timing of surgical eyelid correction depends on the size and location of the eyelid margin defect. Patients with smaller defects may be



SLIDING HUGHES PROCEDURE. Preoperative presentation of a 2-month-old infant with bilateral medial upper eyelid colobomas (1). Creation of the lateral tarsoconjunctival flap (2). Attachment of the tarsoconjunctival flap to the medial canthal tendon (3). Seven-week postoperative surgical result with skin graft (4).

able to delay surgery for years with aggressive topical lubrication. However, earlier intervention is often suggested for patients with large defects and significant corneal exposure. Patients with bilateral colobomas may require very early intervention because both corneas are at risk of visual loss from exposure.

Preoperative Planning

The initial evaluation of an upper eyelid coloboma consists of measuring the size of the eyelid margin defect and comparing it with the overall length of

the horizontal palpebral fissure. The surgical reconstruction technique is determined by the size of the defect.

Small defects. If the defect in the upper eyelid involves less than one-third of the margin, it may be closed with direct tissue apposition. This surgery may require a lateral canthotomy and/or superior cantholysis to rotate or advance adjacent tissue in order to prevent excessive tension on the wound.

Moderately sized defects. Larger defects, however, often require more creative and technically difficult surgical techniques. A Tenzel semicircular

rotational flap may be used for defects involving approximately one-third of the eyelid margin.² In this technique, the lateral segment of the eyelid is advanced by incising the lateral canthal tendon and creating a semicircular flap from skin below the lateral portion of the eyebrow and canthus. The mobilized lateral eyelid covers the medial defect.

Large defects. If the defect is larger than one-half of the upper eyelid, other surgical procedures should be used. A free tarsoconjunctival graft from the contralateral upper eyelid may be used in unilateral cases when there is adequate tarsal tissue.

The Cutler-Beard procedure is another option frequently used for large defects. In this technique, an incision is made below the lower eyelid tarsus, and a full-thickness lower eyelid flap is moved into the defect of the upper eyelid by advancing the flap behind the remaining lower eyelid margin. This procedure results in temporary occlusion of the palpebral fissure. Techniques that require long-term occlusion of an eye during the critical early stage of development of the visual sensory pathway may produce debilitating occlusion amblyopia.

The surgical treatment described for the following patient demonstrates an effective way to manage bilateral upper eyelid colobomas without placing an infant at risk for occlusion amblyopia.

Our Patient

A 2-month-old girl with bilateral upper eyelid colobomas presented to the oculoplastic surgery clinic (Fig. 1). The patient was born after a 31-week gestation through spontaneous vaginal delivery. No cardiac or genetic defects had been detected in earlier examinations.

On physical examination, the patient had full-thickness defects involving more than half of the medial margin in both eyelids. The upper medial skin was attached directly to the bulbar conjunctiva approximately 2 to 3 mm above the superior limbus, a condition known as symblepharon.

The patient also had a bifid nose. Her eyes were orthotropic, both her pupils were 2.5 mm, and there was no relative afferent pupillary defect. The corneas appeared clear, and the rest of the slit-lamp examination was unremarkable. A dilated retinal examination revealed a cup-to-disc ratio in both eyes of 0.1, a normal retina and no evidence of retinal coloboma. She had been given artificial tear ointment every four hours and artificial tear drops every two hours since her birth.

Treatment Decision

This patient presented a surgical challenge for several reasons. The symblepharon had created small, superior fornices bilaterally and resulted in a small surgical space within which to operate. At the onset of the case, the symblepharon was managed by injecting the superior palpebral conjunctiva with anesthetic to create as much space in the superior medial fornix as possible. It was decided to leave her superior medial fornices short at the conclusion of the case, with the understanding that additional surgical intervention may be required in the future, should this condition present a problem for the patient.

Since the patient was an infant, any eyelid-sharing technique from the lower eyelid (e.g., Cutler-Beard procedure) could increase the risk of occlusion amblyopia. A free tarsoconjunctival graft from the contralateral upper eyelid also was not an option because the lesion was bilateral. Because the existing lateral eyelid had a substantial tarsal remnant to work with, a modified Hughes procedure was chosen as the method of reconstruction.

Surgical Technique for the Hughes Procedure

The original Hughes procedure was devised to correct a 50 to 100 percent lower eyelid defect.³ This procedure consists of reconstructing a defect in the lower eyelid by advancing a flap of conjunctiva and tarsus from the upper eyelid to replace the lower eyelid defect. To make this tarsoconjunctival flap, Müller's muscle is dissected from

the tarsus and conjunctiva of the upper eyelid. The flap is then sutured into the defect in the lower eyelid and a free skin graft or skin-muscle advancement flap is placed over the tarsus in the lower eyelid. The bridge holding the eyelids closed (and obstructing vision) is opened one to six weeks postoperatively.

Lateral Modified Hughes Procedure

A variation of the Hughes procedure may be performed in a patient with bilateral medial upper eyelid defects. This procedure, sometimes referred to as a "sliding Hughes" procedure, creates a tarsoconjunctival flap from the ipsilateral eyelid and rotates it horizontally (medially in this case) to bridge the eyelid defect created by the coloboma.⁴ In this procedure, the upper eyelid is everted and a horizontal incision is made 4 mm to 6 mm below the superior border of the tarsus, extending from the coloboma edge for a distance equal to the amount of horizontal dimension needed to cover the medial defect.⁵ At the end of the incision, a second incision, perpendicular to the first, is made in the direction of the superior fornix.⁵ After delicate dissection of the tarsus and conjunctival tissue from the underlying Müller's muscle, the tarsoconjunctival flap is created (Fig. 2). It is important when creating the tarsoconjunctival flap not to damage Müller's muscle or the vascular supply to the flap.

The flap is then sutured medially to the medial canthal tendon (Fig. 3), and laterally to the remaining tarsus in the lateral upper eyelid, using interrupted 6-0 polyglactin sutures. Prior to suturing the flap laterally to the coloboma edge, the surgeon must trim the medial edge of the "free" eyelid tissue. This aspect of the procedure, commonly referred to as "freshening the edge" of the tissue, removes necrotic tissue and allows the surgeon to suture the eyelid defect closed, tarsus-to-tarsus. This helps prevent any eyelid contour abnormalities in the future. After attaching the tarsoconjunctival flap both medially and laterally, the eyelid is prepared for the placement of a skin graft.

Once one eyelid flap is sutured into place and prepared for the skin graft, the contralateral eyelid can be done in a similar fashion.

Forming a Free Skin Graft

The final step involves harvesting a skin graft to replace the anterior lamellar defect of each eyelid and to cover the exposed tarsus of the tarsoconjunctival flap. It is important to choose donor skin that resembles the naturally occurring skin of the eyelid. This skin must be thin, smooth and hairless. Common sites include the retroauricular and supraclavicular areas or the contralateral eyelids.

Our patient. In this infant, a free skin graft was obtained from a retroauricular location on one side. The free skin graft was divided to provide enough skin for the anterior lamellar defects remaining on both sides, and then sewn into place using interrupted, absorbable sutures. It is advisable to use absorbable sutures in infants because removal of sutures in this age group can be very challenging. A central horizontal mattress 7-0 chromic gut suture was also placed to fix the graft to the recipient bed below. When suturing down the graft, it is important to make sure that the new conjunctival surface of the upper eyelid margin is lower than the skin. This will prevent any corneal abrasions. In addition, the free skin graft should be slightly larger than the defect it is intended to cover. This will account for skin contraction in the future.

Postoperative Follow-Up

Infants with upper eyelid colobomas require close follow-up during the postoperative period. Treating the surgical wounds with antibiotic ointment two times a day is recommended for one week. Frequent eye examinations to assess eyelid stability and function and overall eye development are recommended. Our patient returned to the clinic seven weeks postoperatively with continuous upper eyelids bilaterally (Fig. 4), having had no complications in the interim.

The case described above illustrates

the challenges involved in surgically correcting infants with bilateral upper eyelid colobomas that involve more than 50 percent of both eyelids. Proper planning of the surgery preoperatively is critical for a functionally appropriate and cosmetically pleasing surgical outcome.

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