

Grand Rounds

A 5-day-old-newborn with a large right upper eyelid coloboma

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History

A 5-day-old girl presented to the Cleveland Clinic Cole Eye Institute Oculoplastics service for evaluation of right eye exposure. She was born at full term by spontaneous vaginal delivery after an uncomplicated pregnancy. Family history was unremarkable for any known heritable conditions or ocular disease.

Examination

On initial examination, the patient was found to have a right preauricular skin appendage, visual acuity of light perception in both eyes, a small right eye temporal limbal dermoid, and a right upper eyelid coloboma that involved approximately 70%–80% of the upper eyelid. There was near total exposure of the right corneal surface, with trace inferior punctate epithelial erosions but no epithelial defect. Anterior and posterior segment examinations were normal. Aggressive surface lubrication was initiated, and the patient was followed closely. At 2 weeks' follow-up the patient developed a small right inferior corneal ulcer. Topical antibiotics were initiated, and the patient was scheduled for surgical repair (Figure 1).

Ancillary Testing

Routine blood work, newborn metabolic screening, chromosomal microarray analysis, and hearing screen were unremarkable. An echocardiogram revealed a moderate secundum atrial septal defect with mild dilation of the right ventricle. X-ray of the chest and abdomen revealed thoracic vertebral and left rib segmentation anomalies.



Figure 1. Large upper eyelid coloboma in neonatal patient with inferior corneal ulcer and ointment laden corneal surface.

Treatment

Under general endotracheal anesthesia, local anesthetic solution was injected into the lateral canthus and tissue surrounding the eyelid coloboma. This consisted of 1 ml of lidocaine 1% with 1:100,000 dilution of epinephrine and sodium bicarbonate 8.4% in a 1:10 ratio with 50 units per 10 ml of hyaluronidase. The coloboma borders were then squared, and direct approximation of the edges was attempted; however, correction via this method caused significant tension, resulting in ptosis. The internal cantholysis technique was then used to recruit additional eyelid for surgical repair (Figure 2). The lateral eyelid was held with digital tension to expose the temporal aspect of the conjunctiva. A horizontal 2 mm transconjunctival incision was made using electro-

Published September 28, 2017.

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doi:10.5693/djo.03.2017.08.001

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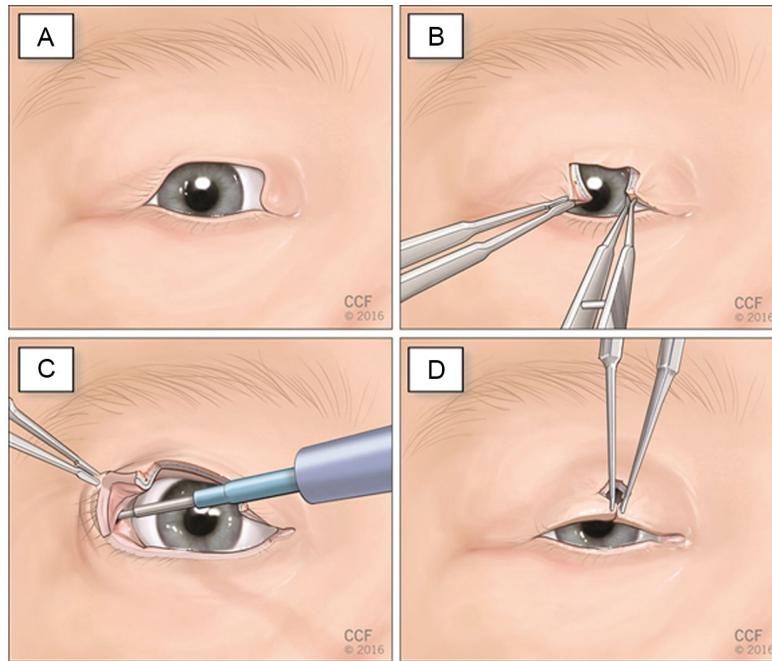


Figure 2. A, Large upper eyelid coloboma B, Edges of the defect are freshened with #15 blade and an initial attempt at direct approximation failed to close the defect. C, The conjunctiva is incised approximately 3 mm superior to the tarsus in the right upper eyelid lateral fornix, and the superior crux of the lateral canthal tendon is lysed with a monopolar cautery instrument. D, The wound edges are precisely anastomosed without undue tension.

cautery approximately 3 mm superior to the tarsus, just lateral to the lateral canthal tendon. A Jaeger lid plate was placed adjacent to the conjunctival incision, and the electrocautery device was used to strum and identify the superior crux of the lateral canthal tendon. These attachments were severed using the electrocautery unit. The lateral eyelid could then be reapproximated.

The tarsus was closed with two lamellar 5-0 absorbable braided polyglactin 910 sutures. The orbicularis was closed with a single 5-0 polyglactin 910 suture. A small amount of redundant cutaneous tissue was excised using a Burrow's triangle along the relaxed skin tension lines inferolaterally. The eyelid margin was repaired with a vertical mattress 7-0 silk suture, which was then run superiorly to close the skin. The silk suture was removed 2 weeks after surgery. Antibiotic ointment was placed on the surgical wounds.

Differential Diagnosis

In this patient the differential diagnosis included Goldenhar syndrome, CHARGE syndrome, and isolated eyelid coloboma. There were several options for how to repair upper eyelid colobomas. Direct closure, which generally yields excellent functional and cosmetic out-

comes, was the most simple and straightforward surgical option.¹ However, application of this technique is greatly limited by defect size and lid laxity, because undue tension could easily result in notch formation and trichiasis.² For colobomas larger than 1/3 of the horizontal lid margin, additional tissue must be mobilized in order to avoid these complications. External canthotomy and cantholysis and semicircular or rotational flaps are often used for defects involving up to 50% of the lid margin, whereas the Cutler-Beard technique is generally reserved for defects of larger size.³ Because two-stage techniques and prolonged occlusion of the pupil at this age can cause significant amblyopia, one-stage techniques, such as sliding tarsoconjunctival/myocutaneous flaps,⁴ skin grafts,⁵ or large lateral myocutaneous flaps³ have been employed for large defects. However, these procedures carry significant risks for wound morbidity, prolonged healing time, and visible scarring. Often, the recruited or grafted tissue is not lash bearing. This compromises both the cosmetic and functional result, because granulation can result in a keratinized lid margin, with subsequent mechanical keratopathy.² Although not specifically reported before to repair upper eyelid colobomas, internal cantholysis with subsequent direct closure is a one-step procedure that has been employed successfully for closure of full-thickness defects of up to

25 mm in adults following excision of cutaneous lid malignancies.⁶

Diagnosis and Discussion

Eyelid colobomas are rare congenital malformations caused by failure of the mesodermal lid folds to fuse during embryogenesis. The resultant defect is often triangular in shape and located at the junction of the medial and middle 1/3 of the upper eyelid, although the shape, size, and location are all highly variable.¹ Colobomas are most commonly idiopathic and can present at birth as an isolated finding, but they may also be accompanied by systemic features, as in Goldenhar or Fraser syndrome.⁷ Given the constellation of our patient's signs and symptoms a presumptive diagnosis of Goldenhar syndrome was made.

Correction of large upper eyelid defects in the neonatal period presents unique challenges, because unilateral lid-sharing procedures can result in significant amblyopia. When possible, surgery may be delayed until the age of 3 or 4 years, when more lid tissue is available and the risk of induced amblyopia is decreased.³ However, especially in larger defects, as in this case, surgery should be performed promptly to lessen or prevent exposure keratopathy. Historically, sliding tarsoconjunctival/myocutaneous flaps,⁴ skin grafts,⁵ or large lateral myocutaneous flaps³ have been used with varying success.

In this case, we used internal cantholysis followed by direct closure to repair the large right upper eyelid coloboma. The silk sutures were removed 3 weeks after surgery. A video taken at this time demonstrates excellent eyelid movement and contour (Video 1). At 9 months' follow-up, there was a favorable eyelid contour, mild ptosis that did not obstruct the visual axis, and very slight lagophthalmos, without noticeable notch formation (Figure 3). There was persistence of superomedial periorbital hollowing, because the severe coloboma also involved the superomedial fatty tissues in addition to the full thickness of the eyelid. Artificial tear ointment and drops were no longer necessary.

This technique produced significant lid mobility and avoided complications associated with other procedures commonly used for closure of large wounds, such as external scar formation and prolonged occlusion of vision. Although we did not encounter lateral canthal dystopia or lid buckling in this patient, such complications tend to be temporary and improve spontaneously in the early postoperative period.⁶ In the event that the amount of recruited tissue is initially inadequate, the



Video 1. Patient demonstrating appropriate upper eyelid contour and blink.

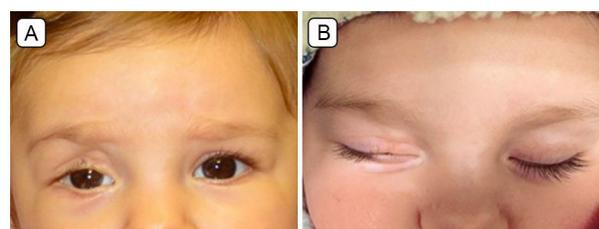


Figure 3. A, Clinical photograph showing appropriate eyelid contour and mild ptosis 9 months postoperatively. B, Trace lagophthalmos without noticeable notch formation.

procedure can easily be converted intraoperatively into a semicircular flap to allow for wound closure. The technique requires minimal equipment, requires no supplementary sutures, and produces excellent cosmetic and functional results, which is especially important in cases with potential for good visual outcome. Internal cantholysis is easily learned and is a powerful technique that should be considered as an option for repairing larger full-thickness lid defects in both adults and children.

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