Congenital Primary Upper Eyelid Entropion

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ABSTRACT

A case of primary congenital upper eyelid entropion (PCUEE) is reviewed. The author proposes a congenital shortness of the levator muscle with anomalous insertion onto the tarsal plate as the etiology, and a new simplified surgical correction is described. The classification of PCUEE as a syndrome with multiple congenital anomalies is supported, and the need for a complete systemic evaluation is stressed.

Congenital primary upper eyelid entropion is an exceedingly rare condition. In 1969, Hiles and Wilder reviewed the 13 known cases in the ophthalmic literature, and presented a new case. They suggested the possibility that congenital upper eyelid entropion was part of a syndrome involving multiple systemic anomalies.

Surgical correction of the entropion is usually necessitated by corneal erosion secondary to trichiasis,2,3 and a multifarious assortment of surgical procedures has been described.4-8

The following is a case report of primary congenital upper eyelid entropion which corroborates the categorization as a syndrome, suggests a pathophysiologic abnormality of the eyelid as the etiology, and describes a new, simplified technique of surgical repair.

Case Report

The patient was the first-born male of a nonconsanguineous 21-year-old couple. Pregnancy was complicated by oligohydramnios at six months gestation, and C-section was required at 42 weeks gestation for fetal distress. The patient had an Apgar of 3, and required assisted ventilation and admittance to the Intensive Care Nursery. Birthweight was 2220 g (10% ile), length 50 cm (5% ile).

Neonatal exam revealed a dysmorphic facies with antimongoldoid slanting of the palpebral fissures, low-set ears which appeared dysplastic with underdeveloped pinnae, micrognathia, and long thin feet with rocker bottoms.

A normal 46XY karyotype was obtained. Complete biochemical analysis of blood and urine was normal, renal ultrasound and chest x-ray were unremarkable.

A CT scan of the central nervous system revealed agenesis of the corpus callosum (Figures 1 & 2). Cardiac evaluation revealed a mild ventrículo-septal defect and cardiomegaly.

Initial ophthalmic exam at age four days revealed a chemical conjunctivitis secondary to silver nitrate drops, normal appearing 'lids, clear corneas and lenses, and a normal fundus. Cycloplegic refraction was +1.00 sphere O.U., and ocular motility was normal.

At four weeks of age, a superficial central corneal ulcer was present right eye and Staphylococcus aureus was cultured. The upper eyelids appeared normal when closed, but on opening the lids, a marked entropion with trichiasis was present bilaterally. Bell's phenomenon could not be demonstrated. An intensive course of Neosporin ointment cured the ulcer, but superficial corneal erosions continued bilaterally.

At age seven weeks, the patient was taken to surgery for plastic repair (Figures 3-6). Forced duction of the upper lids revealed tight upper lids which were unable to be stretched inferiorly. The external surface of the upper lids were normally elastic.

A 5-mm ellipse of skin and orbicularis muscle was excised starting 3 mm above the upper lid margin, from the medial to the lateral ends of each upper eyelid. The skin adjacent to the excised ellipse was undermined for 3 mm (Figures 7 & 8). Hemostasis was achieved with light cautery.

A 6-0 Vycryl suture was placed through the inferior skin edge, advanced superiorly on the tarsal plate with two tarsal bites, and brought out through the superior skin edge, as depicted in Figure 9. Two similar sutures were equidistantly spaced nasally and temporally, respectively. Tying the sutures nicely everted the lid margin, producing a minimal ectropion (Figure 10). The identical procedure was per-
FIGURE 1: CT scan of brain revealing enlarged third ventricle elevated above the level of the lateral ventricles with configuration of the frontal horns of the lateral ventricles typical of agenesis of the corpus callosum.

FIGURE 2: Same as Figure 1, different section.

FIGURE 3: Frontal view, showing absence of upper eyelid fold, bilateral corneal leukomas, and marked inturning of cilia.

FIGURE 4: Close-up O.D.

FIGURE 5: Close-up O.S. Note normal lower lid.

FIGURE 6: O.D., showing severe cilia positioning against cornea.
formed upon the opposite lid. Histopathology of the surgical specimen was reported as normal eyelid skin, with no evidence of orbicularis hypertrophy.

Rapid resolution of corneal haze and ulcers occurred over a two week interval (Figures 11-14). It was necessary to maintain lubricating ointments bilaterally to prevent xerotic keratitis. Schirmer tests revealed reduced tear production bilaterally.

The patient demonstrated developmental delay, and at age eight months developed focal tonic-clonic seizures, controlled with Phenobarbital. Orthopedic consultation revealed bilateral mild knee flexion contractures and bilateral

**FIGURE 7:** Preoperative state, depicting excessive length of external half of upper lid, relative shortness of internal lid, and resultant entropion.

**FIGURE 8:** Surgical excision of skin/orbicularis crescent to reduce external lid length.

**FIGURE 9:** Sutures are placed to elevate inferior skin flap on tarsus and pull down superior skin flap, creating a skin fold and evening lid margins.
FIGURE 10: Postoperative lid position, with elimination of entropion. Lid fold will form at level of skin closure.

FIGURE 12: Side view O.D., showing normal cilia position.

FIGURE 13: Close-up O.D., showing upper lid fold, clear cornea, and eversion of cilia away from cornea.

FIGURE 14: Side view O.S., showing normal positioning of lid margin.

FIGURE 11: One week post-op, showing eversion of upper lid margins, clearing of corneas, and new lid folds.
eral calcaneal valgus feet. Repeat CT scan indicated evidence of cerebral atrophy, and EEG showed progressive deterioration.

Ophthalmic exam at 18 months of age revealed clear corneas, good fixing and following movements, normal optokinetic nystagmus, and orthophoria. The anterior segment and fundus exam was benign. The absence of Bell’s phenomenon persisted.

At age 20 months, the patient suffered a seizure with cardiac arrest, and despite emergency resuscitation efforts, expired.

Discussion

Multifarious etiologies have been proposed for congenital primary upper eyelid entropion. An abnormality of the tarsal plate has been documented in several case reports, but other case reports have described normal tarsal plates with spasm and/or hypertrophy of the marginal portion of the orbicularis muscle. Lack of an upper lid fold without ptosis has also been commonly observed.

Clinically, the pathophysioloogy of primary congenital upper eyelid entropion has an iatrogenic model in the secondary entropion following large levator resection, as described by Beard. The tightness of the forced duction test suggests that the levator muscle may be fibrotic or shorter than normal in length. In addition, the absence of an upper eyelid fold suggests the absence of tendinous fibers extending from the levator to the pretarsal skin and/or anterior portion of the tarsal plate. This would effectively subdivide the lid functionally into two distinct halves. The tight internal half would result in a traction on the lid margin. The loose external half would tend to accentuate the entropion of the margin, since the orbicularis muscle would tend to invert the margin, and the excess loose skin would gravitationally increase the net force.

As soon as the entropion is severe enough to produce trichiasis, a component of spastic entropion is likely to occur, secondary to blepharospasm, causing the pseudohypertrophy of the orbicularis muscle. If this occurs in utero, the soft fetal tarsus is likely to be deformed, producing the tarsal kink. The kink of the tarsal plate in turn seems to shorten the height of the tarsal plate.

The author's method of surgical correction of congenital entropion is similar to the procedure recommended by Beard for upper eyelid entropion following external levator resection.

This particular case corroborates Hiles and Wilder's suggestion that primary congenital upper eyelid entropion is a part of a syndrome with multiple systemic anomalies. The agenesis of the corpus callosum has not been previously described. Pediatricians, geneticists, and ophthalmologists must be aware that a complete systemic evaluation is mandatory in such cases. The developmental delay and abnormal skull findings described previously suggest central nervous system abnormalities may be a part of the syndrome.

The simple surgical procedure described would probably be successful in most cases, assuming that pathophysiologic mechanism is correct.

References