

Bilateral congenital eyelids eversion: case series and review of literature

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ABSTRACT

Congenital Upper Eyelid Eversion (CUEE) is a rare and benign congenital condition characterized by everted upper eyelids with prominent chemosis. We report two cases of bilateral congenital upper eyelid ectropion; an ophthalmic examination of one of the cases showed asymmetry in the extent of upper eyelid eversion and chemosis. The further paediatric evaluation showed no revealing in the two cases. For the two cases, treatment of the eyelids eversion was conservative, combining topical steroids, antibiotics, and lubricants. Chemosis was reduced progressively. One of the cases reported by the authors had an asymmetric severity and variation in time to respond to conservative treatment, which makes it peculiar to other reported cases of congenital upper eyelid ectropion. In this patient, the upper eyelid with severe chemosis and eversion took extended time. Congenital upper eyelid ectropion is common among black infants, and most infants show excellent anatomic and functional results with conservative treatment if managed timely and promptly.

Key words: Congenital ectropion, Upper eyelid eversions, Asymmetric presentation, Black infants, Conservative management

INTRODUCTION

Congenital Upper Eyelid Eversion (CUEE) is a rare and benign congenital disorder that usually presents at birth and is described as a complete eversion of the upper eyelid associated with swelling, conjunctival prolapse, and severe conjunctival chemosis¹. In addition, CUEE is associated with blepharophimosis syndrome, colloidal skin disease, and Down syndrome. Moreover, a higher preponderance of the disease is seen in black infants²⁻⁴.

We report two rare cases of bilateral upper eyelid eversion presented to hospital of which one had an asymmetric clinical feature at presentation. Our case report aims to describe a previously unreported asymmetric feature of upper eyelid ectropion. It also highlights the treatment response to conservative management.

CASE REPORTS

Case 1

A 4-hour-old male neonate, the first child to a 26-year-old mother, was referred to our centre from a private clinic where he was delivered on account of protrusions of red mass of tissue from both eyes since birth. Both the pregnancy and delivery were uneventful. The mother had an ante natal follow-up at the same clinic. The baby was born at full term via vaginal delivery without using any instruments. Ophthalmic examination revealed complete eversion of both upper eyelids with severe conjunctival chemosis and hyperemia (Figure 1). The eyeballs were

examined by carefully opening the eyelids with a cotton-tipped applicator under topical anaesthesia. Both globes were normal size, and the anterior segment evaluation was unremarkable in both eyes. A further detailed systemic examination was unremarkable.



Figure 1: A 4-hour-old child born with complete bilateral eversion of the upper eyelids with severe conjunctival chemosis and hyperemia

Manual repositioning of the eyelids was not successful. Moist dressings with hypertonic saline, a topical antibiotic-corticosteroid ophthalmic ointment, Terra-cortril ointment® (10 mg/mL Oxytetracycline, 5 mg/mL Hydrocortisone Acetate and 10000 u/mL Polymyxin B Sulphate) three times daily and lubricants with Tear Naturale® (dextran 701 mg / ml and hypromellose mg / ml) applied hourly for lubrication, infection prophylaxis, and chemosis reduction as part of a conservative treatment under close supervision. The eyes were patched between

drops. After four weeks of conservative treatment, the upper eyelid's chemosis had sufficiently subsided, the lid naturally flipped (Figure 2), and there was no ocular complication.



Figure 2: Four weeks after the presentation, the same patient had complete resolution of bilateral upper eyelid eversion and chemosis.

Case 2

A 2-day-old female neonate presented to the emergency ophthalmology clinic with a chief complaint of outward eversion of both eyelids since birth, with the left eyelid intermittently reverting back by itself. She was the fourth child of a 25-year-old woman. Pregnancy was uneventful with regular antenatal follow-ups. The child was full term and was born by normal vaginal delivery, with no instrumentation.

Ophthalmic examination revealed complete of the right upper eyelid and partial eversion of the left upper eyelid with prominent conjunctival hyperemia and chemosis ((Figure 3A).



Figure 3A: A two-day-old infant with fully bilateral everted upper eyelids and chemosis, worse on the right upper eyelid

The eyeballs were examined by carefully opening the eyelids using a cotton tip applicator under topical anaesthesia. Both globes were normal-sized, and anterior segment evaluation was unremarkable in both eyes. Further detailed systemic examination was unremarkable.

Manual repositioning of the eyelids was not successful. Similar management to the first patient was given. After three days, the left upper eyelid's chemosis had sufficiently subsided, the lid naturally flipped, and the right upper eyelid chemosis markedly reduced (Figure 3B).



Figure 3B: After three days of topical therapy, resulting in resolution of eyelid edema in the left eye and naturally flipped left upper eyelid and no longer taped.

The same conservative treatment with eye patches was continued to the right eye, which did not improve, until day 18, when the right upper lid began to realign naturally (Figure 4). Two months later, both eyelids were normal, and there was no ocular complication.

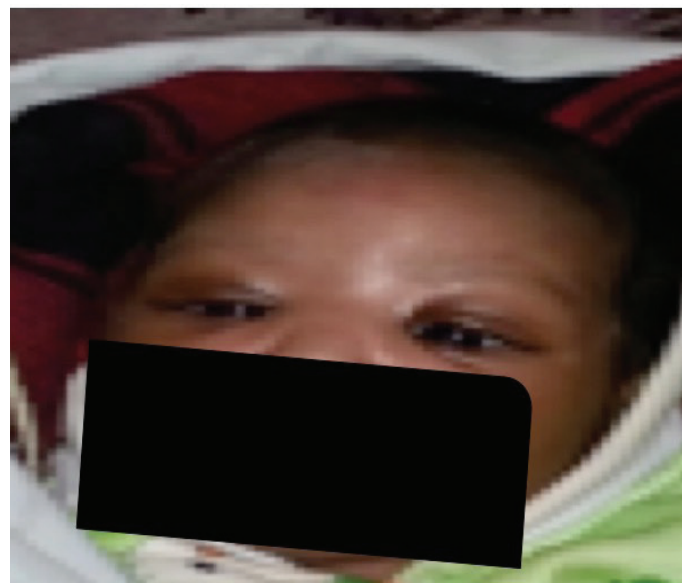


Figure 4: The same patient 5 with complete resolution of bilateral upper eyelid eversion and chemosis

DISCUSSION

Congenital Upper Eyelid Eversion (CUEE) is a rare abnormality, and Adams initially documented it as a case of “double congenital ectropion” in 1896¹. Congenital Eyelid Imbrication Syndrome (CEIS) and Congenital Floppy Eyelid Syndrome (CFES) are important differential diagnoses for CUEE. CEIS is characterized by upper eyelids overriding the lower eyelids, usually seen in patients with floppy eyelid syndrome⁵. Floppy eyelid syndrome is frequently associated with Down syndrome⁶. In the present cases, no overriding of the upper lid over the lower eyelid and no features of Down syndrome were noted on systemic evaluation.

Bentsi-Ebchill from Ghana reported the greatest number of CUEE cases, 14 in total⁷. The exact aetiology of CUEE is unknown. The absence of a tarsal plate, vertical

shortening of the anterior lamella, levator disinsertion, and overlapping of the lower eyelid edge under the upper lid are some of the hypothesized aetiologies for CUEE^{7,8}. In addition, the orbicularis spasm may serve as a sphincter once it has everted, creating a cycle of conjunctival strangulation and edema due to venous stasis⁴. However, a histopathologic investigation of a deceased 9-day-old neonate with CUEE failed to reveal anatomic lid anomalies to explain the illness⁹.

Rarely, the disorder may be linked to collodion skin disease or Down syndrome^{2,10-12}. Otherwise, as seen in Table 1, most cases reported in healthy children were in Africans^{3,4}. Birth trauma has been suggested as a potential mechanism for CUEE, even if the actual aetiology is unknown^{4,13}. Similar to our case, the majority of the reported cases have been following both normal vaginal delivery^{7,14-17}.

Table 1: Clinical presentation of reported cases of congenital upper eyelid ectropion

Features	Sayadi <i>et al</i> ¹³	Ibraheem <i>et al</i> ¹⁷	Bentsi-Enchill ⁷	Adeoti <i>et al</i> ¹⁴	Present
No. of cases	1	1	14	3	2
Age at presentation	4 h	6 days	1h-1 case 6h- 1 case 14h- 1 case 1day-6 cases 2 days-2 case 3 days- 3 case	4 h 3days 4 days	4 h 2 days
Sex	Male	Female	10-male 4-Female	3 Male	1 Male 2 Female
Ethnicity	White	Black	Black	Black	Black
Method of delivery	Vaginal	Vaginal	13 vaginal 1 caesarean	Vaginal-all	Vaginal
Laterality of eversion	Bilateral	Bilateral RE>LE	Unilateral RE-3 Unilateral LE-3 Bilateral -6 Bilateral LE>RE-1 Bilateral RE>LE-1	Bilateral-2 Bilateral LE>RE	Bilateral Bilateral RE>LE-1
Country	Tunisia	Nigeria	Ghana	Nigeria	Ethiopia
Systemic diseases	Umbilical hernia	None	None	None	None
Treatment	Topical Lubrication, Antibiotics and Application Hypertonic Saline	Topical lubrication, antibiotics and application hypertonic saline	Lid suture Subconjunctival hyaluronidase injection Antibiotic treatment	Topical lubrication, antibiotics and application hypertonic saline	Topical lubrication, antibiotics and application hypertonic saline
Recovery	21 days	5 Days	1-2 days	8 th day-1case 10 th day-2 cases	2 weeks

Like in our cases, the disorder is frequently present from birth^{1,17-20}; however, there have been recorded cases of later presentation¹⁰. It is clinically defined by the protrusion of the edematous upper lid conjunctival fornix leading to upper eyelid eversion^{1,18,19}.

Although unilateral involvement has been noted^{7,15-19}, most publications describe bilateral involvement of the upper lids. Bilateral asymmetric presentation is rare and these cases were seen in black Africans like our cases (Table 1).

The role of surgical interventions and injections was only limited in studies published earlier⁷ or in cases of

CUEE with Down syndrome¹². Conservative management with local antibiotic ointment, hypertonic saline, lubricants, and moist pressure with manual repositioning of the eyelids has been reported to resolve CUEE in a few days to weeks^{2,16,17,19,20-22}. In the current cases, similar to the above reports, the patient responded to conservative management. The left upper eyelid, which had partial eversion, responded in 2 days, whereas the right upper eyelid, which had total eversion, took three weeks to respond fully.

CONCLUSIONS

CUEE is a rare eyelid disorder of uncertain cause. As seen in our cases, black infants with CUEE have no other associated systemic disorders. Black infants with CUEE rarely present with asymmetric CUEE, and most infants with CUEE usually show excellent anatomic and functional results with conservative treatment. These case series may expand the current recognition and understanding of this rare entity.

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