

Congenital bilateral upper lid eversion

Faten Almajed, Hamad M. Alsulaiman¹, Nada AlMadhi², Silvana A. Schellini³, Gorka Sesma⁴

Access this article online

Quick Response Code:



Website:

www.saudijophthalmol.org

DOI:

10.4103/1319-4534.322620

Abstract:

We report two healthy Saudi newborns with congenital bilateral upper lid eversion evolving with good outcome using conservative treatment. The current literature including epidemiology, clinical characteristics, possible etiologic factors, and treatment was reviewed.

Keywords:

Congenital ectropion, conjunctival chemosis, upper lid eversion

INTRODUCTION

Congenital bilateral upper lid eversion (CBULE) was first described by Adams in 1896, who referred to the condition as double congenital ectropion.^[1] It was also referred to as bilateral congenital eublepharon.^[2] CBULE is a rare benign condition. Until the year 1992, only 50 cases were reported in a literature review.^[3] This disease has been associated with collodion skin disease^[4] and is very prominent in Down syndrome patients.^[5-9]

The vast majority of reported cases in healthy children were described in Africans^[10-19] or Afro-Americans^[20-23] except one Arabian patient,^[24] two Indian patients,^[25,26] and one Turkish patient.^[27] This entity is likely only encountered by few ophthalmologists throughout their career, making it important to report the clinical characteristics, feasible etiologies, and treatment options for CBULE to raise awareness and a correct diagnosis.

We report the first two cases of healthy Saudi newborns with CBULE presenting to King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia. This report was conducted in compliance with the moral, ethical, and scientific principles governing clinical research as set out in the Declaration of Helsinki and was approved by the scientific committee of our hospital.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

CASE REPORTS

Case 1

A 2-day-old Saudi female newborn from a postterm normal spontaneous vaginal delivery was referred to our hospital due to severe bilateral upper lid ectropion, conjunctival chemosis, and yellow discharge since birth. The neonate was otherwise healthy with normal birth weight from a healthy mother with insignificant maternal pregnancy history. On examination, there was a bilateral total upper eyelid eversion with chemotic exposed upper palpebral conjunctiva and yellow discharge overlying the palpebral conjunctiva. The protruded chemotic conjunctiva was occluding the palpebral fissure which had to be retracted to examine the eyes. However, the remainder of the ophthalmic examination was unremarkable, bilaterally. The patient was diagnosed with CBULE. A conjunctival swab for culture was negative. A conservative treatment with close observation was started using isotonic natural tears (Carboxy Methyl Cellulose Na 0.5%, Jamjoom Pharma, Jeddah, Saudi Arabia) every 2 h around the clock and artificial tears gel (Carbomer 0.2%, Fabrik GmbH, Berlin, Germany) twice daily with topical prophylactic antibiotic drops (Moxifloxacin 0.5%, Alcon Laboratories, Fort Worth < TX, USA) four times daily. The eyes were patched between drops. A marked regression of the condition was observed after 48 h, and the child remained in good condition subsequently [Figure 1].

How to cite this article: Almajed F, Alsulaiman HM, AlMadhi N, Schellini SA, Sesma G. Congenital bilateral upper lid eversion. Saudi J Ophthalmol 2020;34:324-7.

Department of Ophthalmology, Imam Abdulrahman Bin Faisal University Dammam, ¹Division of Oculoplastic and Reconstructive Surgery, King Khaled Eye Specialist Hospital, ²Residency Program, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia, ⁴Pediatric Ophthalmology, Department - King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia, ³Department of Ophthalmology, Medical School, Sao Paulo State University, Botucatu, Sao Paulo State, Brazil

Address for correspondence:

Dr. Hamad M. Alsulaiman, Division of Oculoplastic and Reconstructive Surgery, King Khaled Eye Specialist Hospital, Al-Oruba Street, PO Box 7191, Riyadh 11462, Saudi Arabia. E-mail: dr.halsulaiman@gmail.com

Submitted: 07-May-2020

Accepted: 18-Jun-2020

Published: 29-Jul-2021

Case 2

A 3-day-old male Saudi newborn from a postterm delivery by cesarean section was referred to our hospital because of bilateral upper eyelids eversion with intense chemosis occluding the eyelid palpebral fissure since birth. The neonate was otherwise healthy from a healthy mother with insignificant maternal pregnancy history. The neonate was delivered through cesarean section. The ophthalmic examination was very similar to Case 1 [Figure 2]. The patient was diagnosed with CBULE. A similar management to the first patient was given and the condition markedly regressed within 48 h.

DISCUSSION

In this paper we present the first two cases of CBLUE in healthy Saudi neonates. In 1955, Mazhar^[24] described the first Arabian case reinforcing the rare occurrence of CBULE in healthy children in the Arabian Peninsula. There was a previous report of a Saudi child with Down syndrome with an unusual presentation.^[7] In our case, one patient was female and the other male. However, others have reported a male preponderance for CBULE.^[10,16,23]

The newborns were presented to our eye hospital at 2 and 3 days after birth without a suspected diagnosis. Most of the children affected present soon after delivery, probably because of its rarity, obscure nature, and pronounced appearance.

Our two cases were postterm. However, the role of multiparity as well as postmaturity on the occurrence of CBULE has been questioned and remains ambiguous,^[28] with cases occurring from the first pregnancy^[17,22] and preterm infants.^[23] Parity of the mother and maturity of the baby may be coincidental findings rather than predisposing factors.^[13]

Our cases were delivered by two different methods. One was delivered via normal vaginal delivery and the other by a cesarean section. It is unlikely that delivery method can be a predisposing factor, as similar cases have been documented following both normal vaginal delivery^[10,13-16,18,20,22,24,25] and

cesarean section.^[10,17,21,29] Mechanical eversion of the eyelids during passage through the birth canal has been suggested as a cause of CBULE.^[15] However, no definitive evidence has been presented of an association between CBULE and the duration of labor or mode of delivery.^[13] There were no gestational issues in our cases, but the condition could occur in cases of intrauterine inflammation.^[13]

The disorder is clinically characterized by prolapse of the edematous upper lid conjunctival fornix leading to upper eyelid eversion that is usually present since birth, but later presentation has been documented.^[16] Most publications report bilateral involvement of the upper lids, but unilateral involvement has been observed.^[10,13]

Apart from the pronounced appearance (such as bilateral prolapse), the eyes were normal, with the need to retract the edematous conjunctiva to perform an ophthalmic examination. The conjunctival chemosis appears to protect the cornea from exposure, and hence, corneal complications are rare,^[8,17,25] except in cases of Down syndrome.^[7]

The origin of CBULE is still speculative. As CBULE is more common in Africans,^[10-19,29] theories have been postulated on differences in anatomic aspects of the lids as the superior height of the tarsal plate or vertical elongation of the posterior lamellae facilitating eyelid eversion and conjunctival exposure, resulting in chemosis. However, this hypothesis needs confirmation. A previous report of a detailed histological examination of the eyelids of a patient with total lid ectropion reported no intrinsic defect in the eyelids.^[30] Other proposed etiologies include overlapping of the lower eyelid margin under the upper lid as a predisposing factor,^[10] absence of a tarsal plate,^[31] vertical shortening of the anterior lamella and levator disinsertion,^[3] and laxity causing the eyelids to evert,^[32] while others have proposed the opposite.^[12] Other possible anatomical causes can be an absence of an effective lateral canthal ligament, lateral elongation of the eyelid, or hypotonia of the orbicularis.^[5,13,15,25,33] Once everted, it has been postulated that orbicularis spasm may act as a sphincter, setting up a vicious cycle of conjunctival strangulation and edema secondary to venous stasis.^[21]

Differential diagnosis of CBULE includes congenital lid imbrication syndrome which is an extremely rare, transient, self-limiting eyelid malposition disorder and presents with bilateral upper eyelids overriding the lower eyelids when the child is sleeping, concurring with bilateral medial and lateral canthal tendon laxity and tarsal conjunctival hyperemia.^[34-36] Another differential diagnosis is floppy eyelid syndrome which can rarely occur in children.^[36]



Figure 1: (a) Left, the pronounced appearance of bilateral upper lid eversion and conjunctival chemosis overlying yellowish mucoïd discharge and occluding the lid fissure. (b) Right, the same child after 2 days of clinical treatment with total regression of the condition



Figure 2: (a) At presentation with severe bilateral upper eyelids eversion with bilateral chemosis worse on the left side. (b) 2 days after presentation with resolution of the eyelid eversion and persistence of upper eyelid swelling and redness in the right side with reduction in the amount of chemosis in the left side. (c) 5 days after the presentation with complete resolution of bilateral of upper eyelid eversion and chemosis

Conservative management to prevent conjunctival desiccation with patching of the eyelids, lubricants, antibiotics, and hypertonic saline has been used successfully.^[6,13,18,37] The use of hypertonic saline 5% has been postulated to reduce chemosis by osmosis of fluid from the edematous conjunctiva through the semipermeable subconjunctival membrane,^[18] but good outcomes are achieved in many cases using normal saline. It is unclear whether the hypertonic solution is better than the isotonic saline. In addition, hypertonic solution may induce irritation.

Success has been reported by manual repositioning of the eyelid,^[20] maintaining the repositioned lids in place with lid taping, pressure bandages, suturing of the lids, or tarsorrhaphy. However, it has been documented that lid manipulation itself can lead to autonomic effects such as respiratory arrest in neonates.^[22] Hence, cardiac monitoring should be performed during the procedure. This recommendation reinforces the importance of a more conservative approach to managing CBULE.

Success has also been reported with puncture of the liquid localized under the everted conjunctiva.^[14] In addition, there are reports of surgical options including compression sutures,^[15] temporary tarsorrhaphy,^[11,12] and a modified entropion repair technique to form the superior fornix and evert the lids.^[33] A comparison between eyelid sutures or the addition of hyaluronidase injection showed that the addition of hyaluronidase resulted in faster permanent reversion of the condition.^[10]

In most infants, the disease course of CBULE is benign. However, infants with Down syndrome have a more complicated course, probably due to the mechanism of eversion that is related to the vertical shortening of the anterior lamella or vertical elongation of the posterior lamella with failure of the orbital septum to fuse with the levator aponeurosis in addition to the eyelid laxity. Hence, skin grafting of the upper eyelids may be necessary.^[7,8]

In conclusion, this article aims to raise awareness among health-care practitioners and specifically ophthalmologists and pediatricians regarding the sight-threatening presentation of CBULE, a condition with a benign course in normal newborns that is usually successfully managed conservatively.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Adams AL. A case of double congenital ectropion. *Med Fortnightly* 1896;9:137-8.
- Sally Suliman CM. A case of bilateral congenital eublepharon or ectropion. *West Lond Med J* 2010;2:37-41.
- Sellar PW, Bryars JH, Archer DB. Late presentation of congenital ectropion of the eyelids in a child with Down syndrome: A case report and review of the literature. *J Pediatr Ophthalmol Strabismus* 1992;29:64-67.
- Chakraborti C, Tripathi P, Bandopadhyay G, Mazumder DB. Congenital bilateral ectropion in lamellar ichthyosis. *Oman J Ophthalmol* 2011;4:35-6.
- Gilbert HD, Smith RE, Barlow MH, Mohr D. Congenital upper eyelid eversion and Down's syndrome. *Am J Ophthalmol* 1973;75:469-72.
- Stern EN, Campbell CH, Faulkner HW. Conservative management of congenital eversion of the eyelids. *Am J Ophthalmol* 1973;75:319-20.
- Al-Hussain H, Al-Rajhi AA, Al-Qahtani S, Meyer D. Congenital upper eyelid eversion complicated by corneal perforation. *Br J Ophthalmol* 2005;89:771.
- Krishnappa NC, Deb AK, Poddar C. Congenital total eversion of upper eyelids in a newborn with Down's syndrome. *Oman J Ophthalmol* 2014;7:98-9.
- Corredor-Osorio R, Tovilla-Pomar JL, Tovilla-Canales JL. Congenital upper eyelids ectropion in Down's syndrome. *GMS Ophthalmol Cases* 2017;7:Doc03.
- Bentsi-Enchill KO. Congenital total eversion of the upper eyelids. *Br J Ophthalmol* 1981;65:209-13.
- Ibechukwu BI, Iya CK. Early surgical management of two cases of bilateral congenital prolapse of the superior conjunctival fornices with lid eversion. *West Afr J Med* 1991;10:433-8.
- Dawodu OA. Total eversion of the upper eyelids in a newborn. *Niger Postgrad Med J* 2001;8:145-7.
- Adeoti CO, Ashaye AO, Isawumi MA, Raji RA. Non-surgical management of congenital eversion of the eyelids. *J Ophthalmic Vis Res* 2010;5:188-92.
- Monebenimp F, Kagmeni G, Chelo D, Bilong Y, Moukouri E. Congenital bilateral eversion of the eyelids: Management of a case according to the conservative approach at the University Hospital of Yaounde, Cameroon. *Pan Afr Med J* 2012;11:34.
- Fasina O. Management of bilateral congenital upper eyelid eversion with severe chemosis. *J Ophthalmic Vis Res* 2013;8:175-8.
- Ibraheem WA. Bilateral congenital upper eyelid eversion: The clinical course and outcome of conservative management. *Pan Afr Med J* 2014;17:215.
- Awoyesuku EA, Pedro-Egbe CN, Sibeudu OA. Congenital upper lid eversion and severe chemosis in a new born. *Niger J Clin Pract* 2014;17:248-50.
- Dohvoma VA, Nchifor A, Ngwanou AN, Attha E, Ngounou F, Bella AL, *et al.* Conservative management in congenital bilateral upper eyelid eversion. *Case Rep Ophthalmol Med* 2015;2015.
- Dzidzinyo K, Ayena KD, Amedome KM, Vonor K, Maneh N, Nagbe Y, *et al.* Ectropion congénital bilatéral avec important chémosis. *J Fr Ophtalmol* 2018;41:e319-20.
- Lu LW, Bansal RK, Katzman B. Primary congenital eversion of the upper lids. *J Pediatr Ophthalmol Strabismus* 1979;16:149-51.
- Raab EL, Saphir RL. Congenital eyelid eversion. *Ophthalmic Surg* 1990;21:736.
- Watts MT, Dapling RB. Congenital eversion of the upper eyelid: A case report. *Ophthalmic Plast Reconstr Surg* 1995;11:293-5.
- Kirkpatrick A, Ledlow D, Dixon E, Philips JB. Congenital bilateral eyelid eversion and chemosis: A case study. *Neonatal Netw* 2018;37:137-40.
- Mazhar M. Congenital eversion of upper eyelids. *Br J Ophthalmol* 1955;39:702.
- Maheshwari R, Maheshwari S. Congenital eversion of upper eyelids: Case report and management. *Indian J Ophthalmol* 2006;54:203-4.
- Devika P, Bhagavan V, Rao DD, Sudhir HK, Kotian VB. Case report of bilateral congenital upper eyelid eversion in a newborn. *RRJMHS* 2014;3:22-4.
- Cingü AK, Sahin A, Yüksel H, Ozkök A, Arı S, Çaça I, *et al.* Congenital

- bilateral upper eyelid eversion: Report of a case. *Semin Ophthalmol* 2014;29:36-8.
28. Kronish JW, Lingua R. Pressure patch treatment for congenital upper eyelid eversion. *Arch Ophthalmol* 1991;109:767-8.
 29. Ren L, Liu J. Congenital total ectropion of upper lid. *Vestn Oftalmol* 1999;115:32-4.
 30. Young RJ. Congenital ectropion of the upper lids. *Arch Dis Child* 1954;29:97-100.
 31. Pico G. Congenital ectropion and distichiasis; etiologic and hereditary factors: A report of cases and review of the literature. *Am J Ophthalmol* 1959;47:363-87.
 32. Johnson CC, McGowan BL. Persistent congenital ectropion of all four eyelids with megaloblepharon. *Am J Ophthalmol* 1969;67:252-6.
 33. Loeffler M, Hornblase A. Surgical management of congenital upper-eyelid eversion. *Ophthalmic Surg* 1990;21:435-7.
 34. Rumelt S, Kassif Y, Rehany U. Congenital eyelid imbrication syndrome. *Am J Ophthalmol* 2004;138:499-501.
 35. De Silva DJ, Fielder AR, Ramkissoo YD. Congenital eyelid imbrication syndrome. *Eye* 2006;20:1103-4.
 36. Chandravanshi SL, Rathore MK, Tirkey ER. Congenital combined eyelid imbrication and floppy eyelid syndrome: Case report and review of literature. *Indian J Ophthalmol* 2013;61:593-6.
 37. deLuise V. Primary congenital eversion of the eyelids. *Ann Ophthalmol* 1984;16:796-7.