

PUB: 7/2014

www.ijrhs.com
ISSN (o):2321-7251



Congenital total eversion of upper eyelids in a newborn with Down's syndrome- Case Report

Nagesha C. K¹, Ashish K. Deb², Chanchal Poddar³

Dept of Ophthalmology. 1- Assistant professor, Sri Devaraj Urs Medical College, Tamak, Kolar, Karnataka. 2- Associate professor, Silchar Medical college and Hospital, Silchar. 3- Postgraduate, Silchar Medical college and Hospital, Silchar.

Submission Date: 07-12-2013, Acceptance Date: 10-01-2013, Publication Date: 31-01-2014

How to cite this article:

acouver/ICMJE Style

KNC, Deb AK, Poddar C. Congenital total eversion of upper eyelids in a newborn with Down's syndrome- Case Report. *Int J Res Health Sci* [Internet]. 2014 Jan31;2(1):000-00. Available from <http://www.ijrhs.com/issues.php?val=Volume2&iss=Issue1>

Harvard style

K,N.C, Deb, A.K., Poddar, C. (2014) Congenital total eversion of upper eyelids in a newborn with Down's syndrome- Case Report. *Int J Res Health Sci*. [Online] 2(1). p. 000-00 Available from: <http://www.ijrhs.com/issues.php?val=Volume2&iss=Issue1>

Corresponding Author:

Nagesha C. K, Dept. Of Ophthalmology, Assistant professor, Sri Devaraj Urs Medical College, Tamak, Kolar, Karnataka.
Email: drnageshck_2006@yahoo.com

Abstract:

Congenital eversion of the upper eyelids in Down's syndrome is a rare condition associated with serious socio-psychological consequences. We report congenital eversion of both upper eyelids in neonate with Down's syndrome and secondary conjunctival chemosis and prolapse managed with 5% hypertonic normal saline, lubricants and antibiotics. Complete eye opening achieved after 5 weeks from the presentation and condition is totally resolved. Pediatrician's assessment of the child is very important to rule out associated congenital anomalies. Non-invasive management of congenital eyelid eversion was found to be effective with no need for surgical management

Key words: Congenital ectropion; Congenital eyelid eversion; Conjunctival chemosis; Down's syndrome; Subconjunctival hyaluronidase.

Introduction

Congenital eyelid eversion is a rare entity which usually presents at birth and more commonly involves the upper eyelids. It was first reported by Adam's [1] in 1896, who used the term 'Double congenital ectropion'. Sellar [2] in 1992, reviewed the literature and found 51 reported cases. By 2006 four [3-6] more cases were added to the pool,

additional three [7] cases were added later on till 2010. We report a case of congenital total eversion of both upper eyelids in a newborn with Down's syndrome.

Case History

A 6 day old female neonate was seen, with the parents complaining of fleshy protrusion of the lids and inability to open both eyes since birth.

The mother was a 24yr-old-gravida 2 para 1. The full term infant weighing 1.985kg was born after prolonged labour of more than 20hrs but without any instrumentation. Ocular examination revealed eversion of both upper eyelids and severe conjunctival chemosis with prolapse of forniceal conjunctiva. (Figure 1) The lid could not be repositioned to the normal position, even with pressure. Cornea was clear with brisk pupillary reactions. Pediatric assesment revealed that the child had peripheral stigmata of Down's syndrome like Single transverse palmar crease and short 5th digit with clinodactyly and, Joint hyperflexibility.

A diagnosis of bilateral congenital eversion with Down's syndrome was made. The child was put on 5% hypertonic saline 2hrly, moxifloxacin 0.5% eye drops 6 times a day, carboxymethyl cellulose 6 times a day and Tobramycin eye ointment 2 times a day and padding of the eyelids with 5% hypertonic saline-soaked gauze dressing. Patient was treated on outpatient basis and advised a follow up weekly.

At 2nd week follow-up, condition improved and child could open her left eye partially with regression of prolapsed conjunctiva. In the right eye condition persisted but was of less severity. By the end of 4th week, there was complete resolution in the left eye but incomplete resolution in the right eye (Figure 2). At the end of 5th week, child achieved normal eyelids with complete eye opening in both the eyes. (Figure 3). Dilated fundoscopy with 0.5% tropicamide drops disclosed normal fundus.

Discussion

Congenital eyelid eversion is reported to be very rare. The condition is typically bilateral, but unilateral [7] cases have been described. The incidence appears higher in black infants [8], infants with trisomy 21[2] and infants born with collodion skin disease [9].

Several factors have been implicated in its pathophysiology including orbicularis oculi hypotonia, birth trauma, vertical shortening of anterior lamella or vertical elongation of posterior lamella of the eyelids with failure of the orbital septum to fuse with the levator aponeurosis, absence of an effective lateral canthal ligament and lateral elongation of the eyelids [4].

Venous stasis during delivery also caused marked chemosis and prolapse of the conjunctiva, leading to eversion of the eyelids [10]. The conjunctival chemosis protects the cornea from exposure and hence, corneal complications are rare.

In this report, prolonged labour could be attributed as a possible factor for causation of congenital eversion, as similary reported in cases by the previous authors [11].

Congenital eyelid eversion can be treated conservatively by topical lubrication, antibiotics and hypertonic saline [6] or surgically by lid sutures, subconjunctival hyaluronidase, and eye padding [12]. Our case was managed by topical antibiotics, lubricants, hypertonic saline and padding where improvement was observed within 1 week and complete resolution occurred in 5 weeks.

Although both minor and major surgical procedures have been advocated, their efficacy must be questioned because conservative measures appear very successful as shown in this case.

Conclusion

Congenital eyelid eversion in Down's syndrome is very rare and a pediatrician's assessment of the child is very important, further to ascertain patient's health, and to rule out associated congenital anomalies such as Down's syndrome. Despite the alarming presentation of congenital eyelid eversion, its benign course justifies a conservative approach in anticipation of an excellent result.

Source of Funding: Nil

Source of Conflict: Nil

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Figure 1: Total eversion of both upperlids with conjunctival chemosis and prolapse.



Figure 2: Resolution of eversion in left eye by 4weeks.

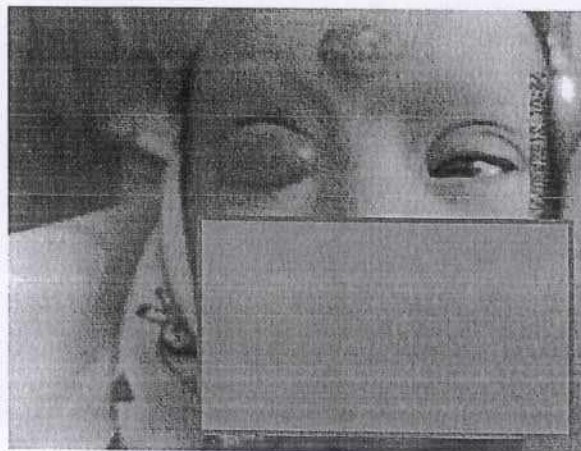


Figure 3: Complete resolution in both eyes by 5 weeks.



