

Spectrum approach to congenital ectropion

Abordagem espectral do ectrópio congênito

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ABSTRACT

In the literature, there is a confusing classification among congenital floppy eyelid, eyelid eversion and ectropion. They are described as eyelid malposition with laxity and out-turning of the eyelids in newborns, usually associated with conjunctival prolapse and chemosis. Although the underlying pathophysiology of these rare conditions is obscure, they share anatomic characteristics. Thus, instead of a plethora of denominations, a spectrum approach should join these entities. In this paper, the authors present a case series of four patients that illustrates distinctive presentations of this condition and advocate that it should be considered as variants of a spectrum of congenital ectropion. Mild cases, when promptly treated, can benefit from clinical treatment. On the other hand, severe and delayed cases will need surgical correction as in the case of acquired ectropion.

RESUMO

Na literatura, existe uma classificação confusa entre *floppy eyelid* congênita, eversão palpebral e ectrópio congênito. Essas afecções são similarmente descritas como pálpebras frouxas e evertidas em recém-nascidos e geralmente associadas a prolapso de conjuntiva e quemose. Embora a fisiopatologia dessas raras afecções seja incerta, elas apresentam íntimas características anatômicas em comum. Assim, ao invés dessa nomenclatura variada, seria interessante incluí-las em um espectro de uma só doença. Neste artigo, apresenta-se uma série de quatro casos que ilustram diferentes apresentações dessa afecção e propõe-se que todas devam ser consideradas variações do espectro de ectrópio congênito. Casos leves são beneficiados quando tratados precocemente. Por outro lado, casos mais graves ou que são tratados tardiamente necessitarão de procedimento cirúrgico semelhante ao ectrópio adquirido.

INTRODUCTION

Congenital eyelid ectropion is a rare condition defined as a complete out-turning of the eyelids usually associated with conjunctival prolapse and chemosis in newborns. In its first description, this condition was called “double congenital ectropion”.^(1,2) It can occur isolated or in association with other disorders, such as infections, inflammation, trauma through the birth canal, or associated with systemic diseases, such as lamellar ichthyosis, collodion skin disease, and Down’s syndrome.⁽³⁾ There is a racial predilection for black infants, usually both eyes are affected, but unilateral cases were documented.⁽⁴⁾

There is a confusing classification of this condition. Many authors term it as congenital eyelid eversion and others as congenital ectropion.⁽⁵⁾ Further, other authors describe a similar condition with intermittent eyelid eversion as congenital floppy eyelid.⁽⁶⁾

Instead of this plethora of denominations, spectrum approach may join conditions that were previously considered separately. Herein we report a case series of four patients that illustrates distinctive presentations of this condition. We also discuss the treatment options and advocate they are not different entities but different degrees of the spectrum of congenital ectropion.

CASE REPORTS

Written informed consent from the parents/guardians of individuals and institutional review board were obtained and approved by Ethics Committee under CAAE:# 36234520.9.0000.5083.

Case 1

A Down’s syndrome Caucasian newborn presented left upper eyelid complete eversion and a mild margin eversion at right upper eyelid (Figure 1A). We performed manual inverting of the eyelid and patching for 3 days. Eyelid position was normalized with no eversion recurrence (Figure 1B).

Case 2

A 7-day-old male Caucasian newborn with no systemic disease was seen with clean and non-discharging eyes with bilateral complete eversion of the upper eyelids and marked hyperemic conjunctival chemosis prolapsed (Figure 2A). We tried conservative management at first, but after 3 days, there was no improvement of eversion. The lids showed moderate laxity and vertical posterior lamellae elongation (Figure 2B). A bilateral partial temporary tarsorrhaphy 7-0 nylon mattress suture was performed. At the sixth postoperative day, sutures were removed, and eyelids were in proper anatomic position, without chemosis and complete eye opening (Figure 2C).

Case 3

A Down’s syndrome black newborn with congenital unilateral eyelid eversion and prolapsed chemosis (Figure 3A). He was submitted to surgical treatment under general anesthesia. We performed skin graft associated with eyelid tightening. Despite margin suture dehiscence, the patient had acceptable functional result (Figure 3B).

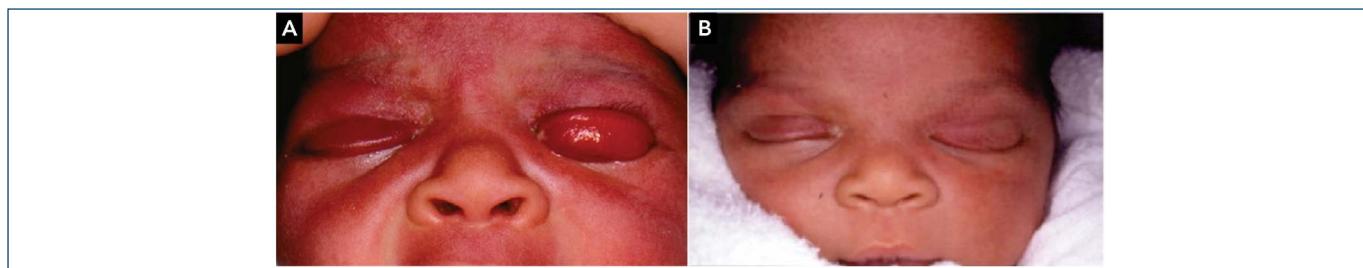


Figure 1. (A) Bilateral eversion; (B) Spontaneous resolution. grafting.

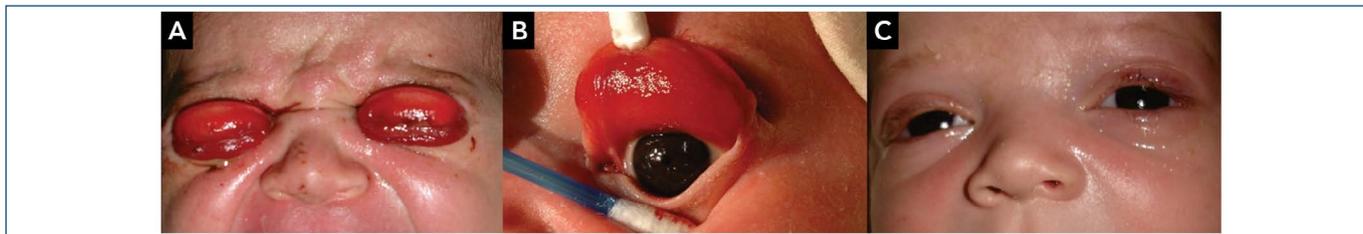


Figure 2. (A) Eyelid eversion with hiperemic conjunctival chemosis prolapsed; (B) moderate eyelid laxity and vertical posterior lamellae elongation; (C) 6 days after tarsorrhaphy.



Figure 3. (A) Unilateral eyelid eversion; (B) post-operative with released sutures.



Figure 4. (A) Superior and inferior ectropion; (B) After skin.

Case 4

A 4-year-old Down's syndrome black child with upper and lower eyelid ectropion (Figure 4A) presenting tearing and red eye since birth. The eyelids showed horizontal laxity and vertical skin deficiency. Skin grafts were used combined to lateral ligament reinforcement (Figure 4B).

DISCUSSION

Congenital eyelid ectropion is a rare condition, and since it was first described in 1896 by Adams, no more than one hundred cases have been reported.⁽²⁾ Most commonly, it involves upper eyelids and is bilateral. Unilateral cases can occur, and all were related to black infants with lid laxity. Bentsi-Enchill reported six cases of unilateral ectropion among the total of 14 cases, suggesting that unilateral occurrence may not be unusual.⁽⁷⁾ Lower eyelid is also rare and occurs in more severe cases with vertical deficiency of skin.^(5,8) Main associations include difficult labor, male gender, Down syndrome,⁽⁹⁾ collodion skin disease and lamellar ichthyosis.⁽³⁾ Grandmultiparity and post-maturity have been questioned in Nigeria.⁽¹⁰⁾ Inheritance has been also suggested.⁽⁴⁾

Some articles refer these cases as eyelid eversion and others as congenital ectropion.^(1,4,5,7,9) Others, also use the term congenital floppy eyelid in cases in which the eyelids have a tendency for intermittent spontaneous eversion on crying and yawning.⁽⁶⁾ Although some authors stress eyelid eversion as a different entity,⁽²⁾ we believe a spectrum approach should join these entities. The word "ectropion" derives from the Greek and means "out-turning",

so it is reasonable to propose all types of eyelid eversion under this term.

The complete pathophysiology of congenital eyelid eversion or ectropion is uncertain; however, they share anatomic characteristics. We had three cases of Down syndrome. According to Krishnappa, these patients usually have a more complicated course, probably due to the mechanism of eversion that is related to the vertical shortening of 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. In two of these three cases, we performed skin grafts and eyelid shortening. The cases present variable laxity of medial and lateral canthal tendons and of the attachment between the anterior and posterior lamellae. Delivery-related trauma may trigger the eyelid out-turning and, once everted, orbicularis spasm may act as a sphincter that leads to a vicious cycle of conjunctival strangulation and edema secondary to venous stasis.⁽³⁾ More severe cases are associated with vertical insufficiency of the anterior lamella.

When recognized early and treated promptly, this pattern can be broken in mild cases like our first patient, with topical anti-inflammatory and antibiotic lubricants, moist dressings, eyelid taping, manual lid inverting, and pressure patching. However, manual eyelid inversion must be done cautiously, as it can cause cardiac and respiratory arrest by oculocardiac reflex.⁽⁸⁾ If clinical treatment is unsuccessful, minor procedure such as tarsorrhaphy can be tried, as in the second

case. Severe cases (cases 3 and 4) are managed as adult ectropion: horizontal lid shortening, skin grafting, and lateral tendon reinforcement. Our case 4 example also showed that the problem can last for years when not treated.

We herein have reported four cases that illustrate variants of this pathology: mild and temporary, and severe, and permanent. Instead of a plethora of denominations, spectrum approach may join conditions that were previously considered separately. Mild cases, when treated early, can benefit from clinical treatment only. Otherwise, severe and delayed cases will need surgical correction, such as in the case of acquired ectropion.

The spectrum approach to congenital ectropion may improve recognition and understanding of this rare entity among healthcare professionals, particularly those who are involved in obstetric and neonatal care.

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