

Reversal of inverse Bell's reflex after sling removal and scar lysis in an 11-year-old patient: a case report

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Abstract

We report a case of inverse Bell's reflex of the right eye in an 11-year-old boy who underwent frontalis suspension ptosis correction at the age of 1 year and developed scarring secondary to exposure and infection of the sling implant 5 months later. At the age of 11 years, the patient had an episode of right eye infective keratitis secondary to corneal exposure and was first noted to have developed the inverse Bell's reflex. The patient underwent removal of sling, scar lysis, and limited full thickness blepharotomy. At postoperative 11 months, the Bell's reflex was completely normal. This suggests that eyelid scar tissue may contribute to the development of inverse Bell's reflex and that scar lysis, despite delayed, may resolve the problem.

Key words: Blinking; Eyelid diseases; Keratitis; Reflex

Introduction

Bell's reflex, in which the globe turned upward during eyelid closure, is an important mechanism to protect the cornea, particularly in patients with congenital ptosis who require ptosis correction surgery, because lid lag and lagophthalmos are common postoperative complications that may lead to

exposure keratopathy and visual debilitation. Inverse Bell's reflex, in which the globe turn downward, is a variation in a small proportion of patients.

Case presentation

In November 2006, a 32-day-old male infant presented to Hong Kong Eye Hospital with right eye congenital ptosis. On presentation, the visual axis was partially obscured. The patient was noted to have a chin-up head posture. He developed right eye amblyopia at 8 months old. Despite good compliance to left eye patching, the visual acuity was 6/12 for the right eye and 6/9.5 for the left eye based on the Cardiff acuity test. At the age of 1 year, the patient underwent frontalis sling operation for the right eye using the double trapezoid technique with a silicone rod. The visual acuity of the right eye improved to 6/9.5. Five months later, the patient had exposure and infection of the sling implant, which was immediately treated with wound revision, drainage, and intralesional antibiotic injection. Nonetheless, scarring developed on the upper lid and resulted in lid notching, upper lid retraction, and lagophthalmos. The palpebral fissure height was 10 mm for the right eye and 8 mm for the left eye. The margin to reflex distance was 4 mm for the right eye and 2 to 3 mm for the left eye. A revision surgery for eyelid retraction and exposure keratopathy was proposed but was declined by the parents. The patient was thus managed conservatively with topical lubricants.

At the age of 6 years, the patient underwent bilateral lateral rectus recession with upward transposition for intermittent

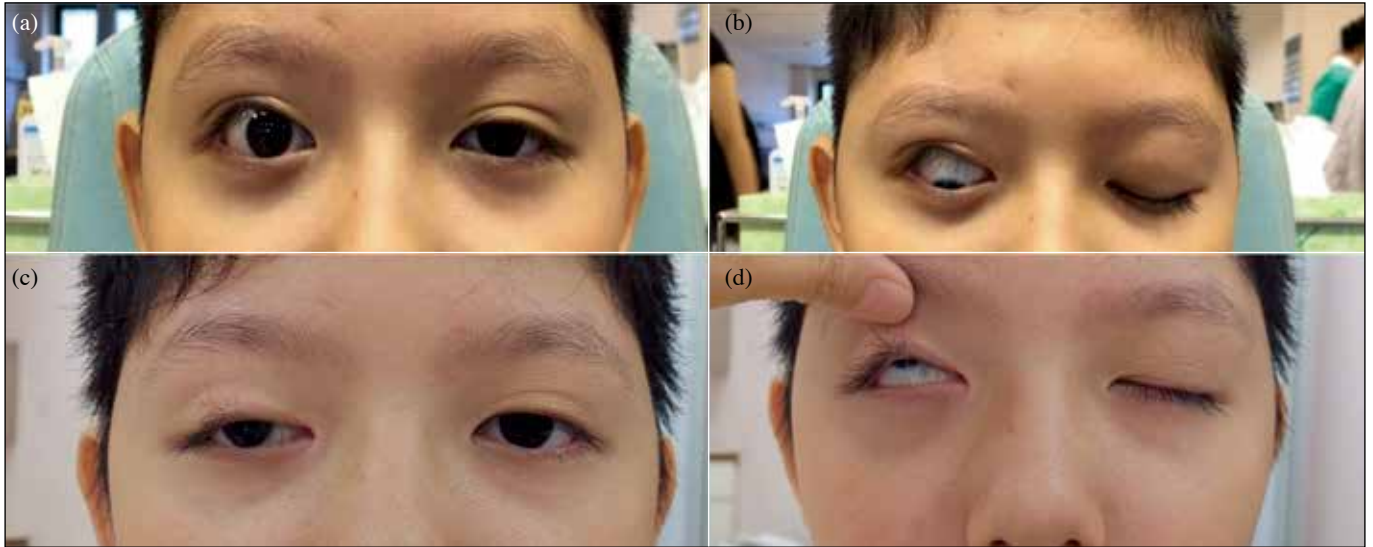


Figure. Clinical photographs showing (a) right eye upper lid retraction and (b) lagophthalmos and eyeball rolling down during eye closing, consistent with the inverse Bell's reflex; (c) correction of right eye upper lid retraction and (d) reversal of inverse Bell's reflex 41 months after sling removal and scar lysis.

exotropia; recovery was uneventful. At the age of 11 years, the patient had an episode of right eye infective keratitis secondary to corneal exposure. On examination, he was first noted to have developed inverse Bell's reflex (**Figure**). The inverse Bell's reflex aggravated the exposure keratopathy and contributed to the episode of keratitis, which was resolved after intensive topical antibiotic eyedrops and copious lubricants application.

One month later, the patient underwent removal of sling, scar lysis, and limited full thickness blepharotomy. Reverse frost suture was placed. Intra-operatively, right upper lid extensive middle lamellar scarring and shortening of right upper lid skin were noted. Scar above tarsus was lysed, the posterior and middle lamellar were completely freed. Subcutaneous adhesions were released as much as possible. Postoperatively, the upper lid retraction was corrected, and the palpebral fissure height was 5 mm for the right eye and 6 mm for the left eye. Residual lagophthalmos was mild. The inverse Bell's reflex normalized. At 4 months, there were occasional occurrences of inverse Bell's reflex. At 11 months, the Bell's reflex was completely normal. The cornea condition significantly improved with no further episodes of exposure keratopathy. At 41 months, the visual acuity was 0.8 for both eyes, and the cosmetic outcome was satisfactory, with no limitation in the extraocular movement (**Figure**).

Discussion

Assessing the Bell's reflex is essential before any ptosis correction surgery. In the normal Bell's reflex, the eyes turn upward during eyelid closure to protect the cornea from exposure. The inverse Bell's reflex, in which the eye turns downward, can be observed in 2% of the population and

also in patients with Bell's palsy.¹

Development of inverse Bell's reflex has been reported after repeat levator resection surgery for congenital ptosis and after a frontalis sling surgery, with the inverse Bell's reflex spontaneously resolved few weeks later.²⁻⁷ Development of inverse Bell's reflex after levator surgery for ptosis correction is rare. The exact mechanism is unknown; possible causes include intra-operative tissue trauma and postoperative soft tissue edema or injury to the nervous system causing aberrant connections.^{7,8} Our patient underwent the initial operation at the age of 1 year, which is much younger than that of other patients reported.³⁻⁷ The inverse Bell's reflex was first noted 10 years after the initial surgery, probably owing to its difficulty to detect at young age with poor patient cooperation for clinical examination. Spontaneous recovery did not occur in our patient, which is different from other cases reported.²⁻⁷ The inverse Bell's reflex resolved after scar lysis. This suggests that the presence of scarring may be a cause of the inverse Bell's reflex and that removal of scar tissue may reverse the inverse Bell's reflex. Although development of inverse Bell's reflex is rare, it is important to assess the Bell's reflex pre- and post-operatively for patients with congenital ptosis, particularly in those with coexisting lagophthalmos who are at higher risk of cornea complications. Copious lubricants and close monitoring are useful to prevent exposure keratopathy until normalization of the Bell's reflex. Prolonged placing of a frost suture to prevent exposure keratopathy is recommended.⁴

Contributors

All authors designed the study, acquired the data, analysed the data, drafted the manuscript, and critically revised the manuscript for important intellectual content. All authors

had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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Data availability

All data generated or analysed during the present study are available from the corresponding author on reasonable request.

Ethics approval

The patient was treated in accordance with the tenets of the Declaration of Helsinki. The patient/parents provided written informed consent for all treatments and procedures and for publication.

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