

A Cause of Corneal Collagen Cross Linking Failure: Keratoconus Associated with the Floppy Eyelid Syndrome

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Abstract

Floppy eyelid syndrome FES is a condition strongly associated with keratoconus. The characteristic findings are an upper lid that may be readily everted, tarsal laxity, and diffuse papillary conjunctival changes. Floppy eyelid syndrome could be a risk factor for corneal collagen cross-linking (CXL) failure. We report a case of patient with floppy eyelid syndrome who had CXL for keratoconus in both eyes, an increase in the Keratometric (K) values indicating keratoconus progression (CXL failure) was seen in the eye with the more prominent floppy eyelid syndrome.

Keywords: Keratoconus; Floppy eyelid syndrome; Corneal collagen cross-linking; Keratoconus progression

Introduction

Floppy eyelid syndrome FES is a condition strongly associated with keratoconus. The characteristic findings are an upper lid that may be readily everted, tarsal laxity, and diffuse papillary conjunctival changes. The cause of floppy eyelid syndrome is believed to be a mechanical disorder due to the eversion of the lids while sleeping. The cause of keratoconus remains uncertain. There are strong proponents to a mechanical etiology for this disease [1,2].

Aim of the study: Patient with floppy eyelid syndrome that had CXL for keratoconus in both eyes, an increase in the Keratometric (K) values indicating keratoconus progression (CXL failure) was seen in the eye with the more prominent floppy eyelid syndrome.

Case Presentation

We report the case of a patient with binocular FES associated with keratoconus (Figure 1 and Figure 2) in which after uneventful bilateral corneal cross-linking (CXL) treatment. No intra-operative or early postoperative complications were found.

At 6 months; Topographic examination after CXL revealed a continuous increase in the keratometric values in the left eye in which the floppy eyelid syndrome was more prominent, with K values of 51.00/52.1 D, indicating keratoconus progression (CXL failure) (Figure 3).

There was no sign of keratoconus progression in the right eye during the follow-up period (Figure 4).

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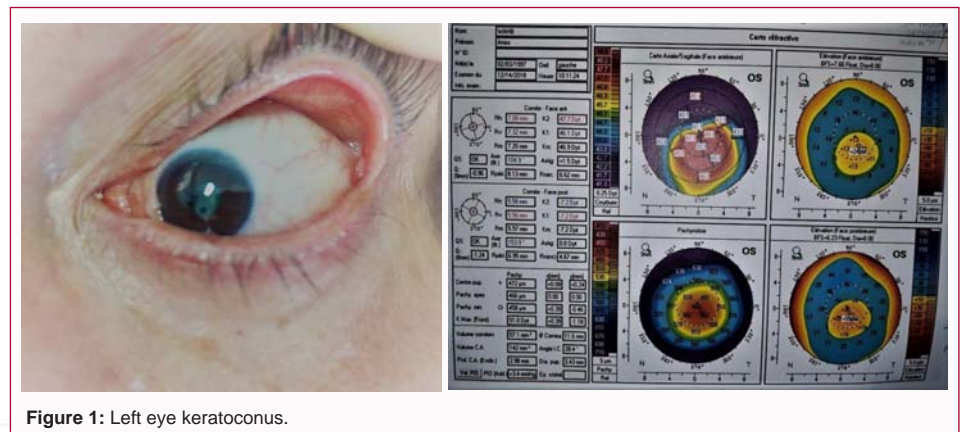


Figure 1: Left eye keratoconus.

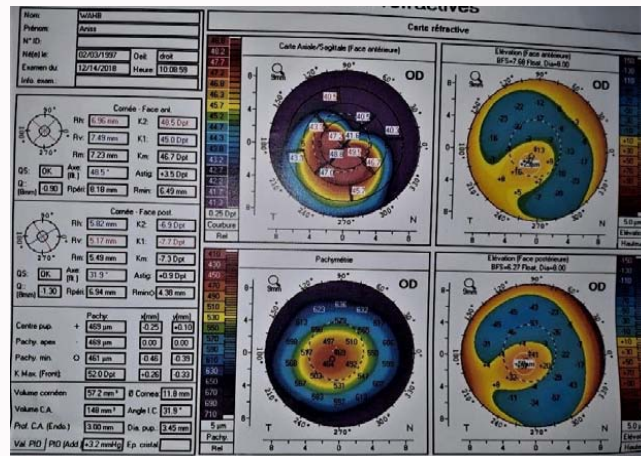


Figure 2: Right eye keratoconus.

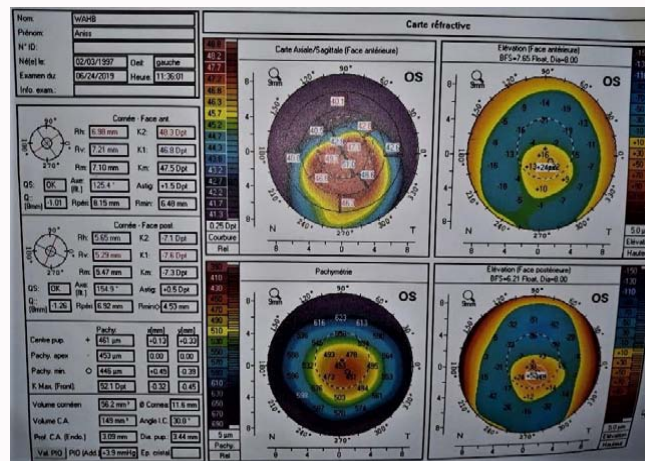


Figure 3: 6 Months after CXL, Left eye keratoconus progression.

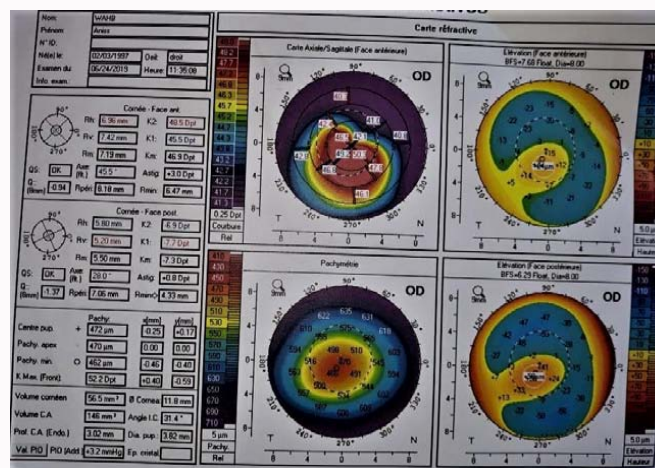


Figure 4: 6 Months after CXL, Right eye: Stabilized keratoconus.

We suggested that possibly the flaccid eyelids could be easily everted during sleeping prone on the affected side and allow mechanical contact with bed sheets, causing irritation of the cornea and conjunctiva, finally leading to keratoconus progression even in a cross-linked (stiffened) cornea. Interestingly, the fellow eye (with the less prominent FES) was stable during every follow-up, indicating

that the CXL failure was probably due to a biomechanical effect rather than a possible systemic alteration.

Discussion

Since the initial description of the floppy eyelid syndrome in 1981, several hypotheses have been made for its pathogenesis. Culbertson

and Ostler, who first pointed out this condition, postulated that spontaneous nocturnal eversion of the upper eyelid would cause events, such as mechanical insult to the conjunctiva, subsequent papillary conjunctivitis and conjunctival keratinization leading to the loss of tarsal elasticity. They made this hypothesis based on the observation that the ocular signs and symptoms are generally more frequent and/or more severe in the eye corresponding to the side on which the patient prefers to sleep. They also suggested an X-chromosome-linked inheritance pattern or hormone influence associated to the problem, since they only found it in men [3].

The floppy eyelid syndrome has been associated with a variety of ocular and systemic conditions, such as keratoconus and obstructive sleep apnea syndrome. It is a unilateral or bilateral condition characterized by flaccid eyelids that easily evert during sleeping prone on the affected side and allow mechanical contact with bed sheets, causing irritation of the cornea and conjunctiva. Several studies have shown the association of keratoconus with floppy eyelid syndrome [1].

Corneal collagen cross-linking (CXL) is a minimally invasive surgical procedure for the treatment of keratoconus. Several studies have reported stabilization and halting of keratoconus progression after CXL treatment. However, further progression of keratoconus during the first year after CXL has been reported. Floppy eyelid syndrome could be a risk factor for CXL failure [4,5].

The cause of floppy eyelid syndrome is believed to be a mechanical disorder due to the eversion of the lids while sleeping. The cause of

keratoconus remains uncertain. There are strong proponents to a mechanical etiology for this disease.

Conclusion

Independent of the nature of decreased corneal biomechanics in patients with FES, primary or secondary, we would like to highlight that these patients should be followed closely with corneal topography, especially during puberty, because of possible progression of ectasia.

References

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