

Case report

Corneal cross-linking in a 10-year-old child with stage III keratoconus

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Abstract

Background: Keratoconus is a corneal dystrophy characterized by progressive thinning, conical shape of the cornea and irregular astigmatism. It is particularly insidious when it occurs in very young patients. **Case:** We report the case of a 10-year-old child with an aggressive stage III keratoconus, who was suggested to undergo a Penetrating Keratoplasty. We performed Corneal Collagen Cross-linking with epithelium removal instead, which was successful in arresting the rapid progression of keratoconus. **Observations:** Eighteen months later, Kmax had decreased by 1.3 D, astigmatism by 0.8 D, thinnest pachimetry had improved by 69 μm and CDVA was 20/32. **Conclusion:** This case confirms that Corneal Collagen Cross-linking is a safe and effective procedure for the treatment of keratoconus even in its aggressive forms. We encourage ophthalmic surgeons to favor conservative treatments when dealing with very young patients.

Keywords: keratoconus, corneal cross-linking, pediatric keratoconus,

Introduction

Keratoconus (KC) is a bilateral, non-inflammatory ectatic corneal disease, characterized by progressive thinning, due to changes in the collagen structure and subsequent stromal weakening, which results in a conical shape of the corneal tissue. This pathology has been studied since the 18th century. However, it was adequately described and distinguished from other degenerative corneal disorders only in 1854 by British physician John Nottingham (Grzybowski, 2013). This corneal dystrophy leads to irregular astigmatism and distorted and decreased vision, with an onset that is generally bilateral and occurs at a young age, usually during puberty, and progresses until the third decade of life, more rarely until the fourth.

Keratoconus affects approximately 1 in 2,000 in the general population – despite

rates vary greatly in different geographic regions – while invasive surgery such as Penetrating Keratoplasty (PKP) is performed in approximately 20% of cases. In the last 25 years, however, a number of conservative procedures has been developed, with the aim of preserving the original cornea and avoiding corneal transplantation. Corneal Collagen Cross-linking (CXL), the most recent among these, was first developed in 1998 by ophthalmic surgeons in Dresda, Germany (Wollensak et al, 2003). It's a non-invasive procedure that addresses the structural weakness of the cornea by reinforcing the corneal stroma with new bonds between its layers, thus halting the progression of keratoconus. Other conservative procedures, aimed at improving vision in stage I and II keratoconus, include Mini Asymmetric Radial Keratotomy (MARK) (Abbondanza et al, 2016), Circular Keratotomy (CK) (Krumeich and Kezirian, 2009) and Intrastromal Corneal Ring Segments (ICRS) (Colin et al, 2000), which can be combined with CXL, in some

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specific cases, in order to obtain both corneal reinforcement and vision correction.

We report the case of a 10-year-old child with an aggressive stage III keratoconus, who was suggested to undergo a PKP, a condition that was successfully treated with Corneal Collagen Cross-linking with epithelium removal instead. Such cases are rarely described in literature and could provide important insights for the conservative treatment of very young pediatric patients with severe KC. The patient was informed about the risks of the treatment and informed consent was obtained

Case presentation

A prepubescent 10-year-old girl presented with a bilateral keratoconus in our clinic, diagnosed less than 2 months earlier by another practice, lamenting poor and decreasing vision. Her ocular history included 4 years of hyperopia and astigmatism and 6 years of vernal keratoconjunctivitis. Corrected distance visual acuity (CDVA) was 20/32 in the right eye and 20/25 in the left eye, however, in a few months the Kmax of her right eye worsened suddenly, reaching 58.3 D (fig. 1), whereas the Kmax of the two eyes was previously comparable. Central pachimetry (Oculus Pentacam HR – Oculus Optikgerate GmbH, Wetzlar, Germany) was 534 μ m in the right eye, thinnest pachimetry was 473 μ m and astigmatism was 8.7 D.

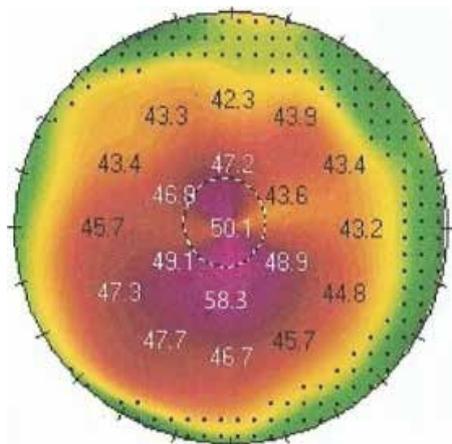


Figure 1: pre-CXL corneal topography, Kmax was 58.3 D

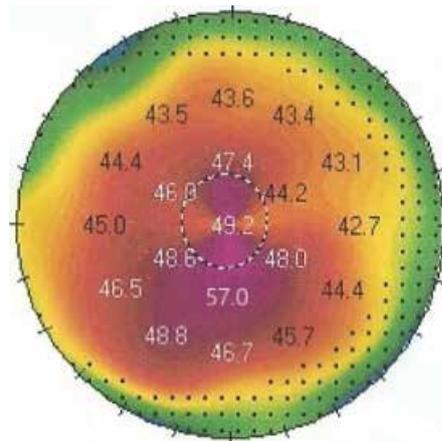


Figure 2: post-CXL corneal topography, Kmax was 57 D

Given the rapid progression of keratoconus, we decided to perform Corneal Collagen Cross-linking with epithelium removal (“epi-off”) in her right eye, according to the standard Dresden Protocol, with benoxinate chloride 0.4% drops as a topical anesthesia. A written informed consent was required as a part of our pre-operative routine. The patient’s eyelids were kept open with the aid of surgical forceps during the procedure, while her corneal epithelium was removed with an ophthalmic scalpel (Micro Feather; Feather Safety Razor Co., Ltd., Osaka, Japan). Riboflavin eye drops (B2 vitamin) were applied on the corneal stroma 30 minutes before the procedure, 10 times at 3 minute intervals; riboflavin was then applied 6 times at 5 minute intervals during UV-A irradiation. The UV-A source used was the UV-X illumination system, version 1000, from IROC AG, Switzerland, while total UV-A exposure time was 30 minutes. Immediately after CXL, we placed a bandage contact lens and prescribed topical antibiotics and non-steroidal anti-inflammatory drops.

One and a half years later, a new clinical examination assessed the level of efficacy of our intervention, deemed at avoiding a corneal transplantation in a 10-year-old child. 18 months after CXL was performed in her right eye, Kmax decreased from 58.3 D to 57 D (fig. 2), Kmed from 48.7 D to 48.4 D,

astigmatism decreased from 8.7 D to 7.9 D, central pachimetry increased from 534 μm to 546 μm and, more importantly, thinnest pachimetry further increased from 473 μm to 542 μm . Accordingly with the halting of the disease and with these significant changes, the child's CDVA was restored to 20/32. Moreover, the change in relevant topographic and tomographic parameters confirmed these

improvements: the Index of Surface Variance (ISV) decreased to 60 (-7), the Index of Vertical Asymmetry (IVA) was 0.54 (-0.07), Keratoconus Index (KI) was 1.16 (-0.03) and the Center Keratoconus Index (CKI) was stable at 1.04 (table 1). Her left eye, on the other hand, was regularly checked but showed no sign of KC progression and, therefore, no need to be treated.

Table 1: pre- and post-CXL values, 18 months after the intervention

	Kmax	Kmed	Astigmatism	Central Pachimetry	Thinnest Pachimetry	CDVA	ISV	IVA	KI	CKI
pre-CXL	58.3	48.7	8.7	534	473	20/32	67	0.61	1.19	1.04
post-CXL	57	48.4	7.9	546	542	20/32	60	0.54	1.16	1.04
Difference	-1.3	-0.3	-0.8	+12	+69	/	-7	-0.07	-0.03	0

Discussion

Today, Corneal Collagen Cross-linking is widely regarded as a safe and effective treatment for keratoconus. Given the success with adults and young adults, CXL is increasingly used for the treatment of adolescents and children as well. This is mainly due to the fact that it increases the strength of the corneal stroma and reduces corneal ectasia, thus providing a small vision improvement and the possibility of being repeated, in case of further progression of keratoconus (Hanna et al, 2015). There is sufficient literature regarding CXL in adolescent patients, but only a few reports have been made on very young pre-teen patients (Sabti et al, 2015). Other clinical studies, examining the efficacy of CXL in mostly adolescent patients, show that this procedure is both safe and effective, a condition that was confirmed in case reports on pre-teen patients as well (Agora et al, 2012; Vinciguerra et al, 2012). A new study, moreover, was able to quantify a complication rate of 6% when using CXL for the treatment of pediatric patients (Steinwender et al, 2016).

There is an ongoing debate regarding the validity of Transepithelial Corneal Cross-linking, a technique that gives less discomfort to the patient but is also generally regarded as less effective (Wollensak and Iomdina, 2009),

something that has also been confirmed in the treatment of pediatric patients (Buzzonetti and Petrocelli, 2012). Our experience with CXL, which started in 2005, brought us to the same conclusion and allowed us to assess that Corneal Collagen Cross-linking, with epithelium removal, successfully halts KC progression and results in an average reduction in corneal curvature of 1.2 D, an improvement that may avoid the need for further surgery (Abbondanza et al, 2009).

Conclusion

In this case, CXL with epithelium removal was successful in arresting a stage III keratoconus and avoiding the need for a corneal transplantation in a 10-year-old child. It produced a corneal flattening of 1.3 D, along with a reduction of astigmatism and an improvement in pachimetry and other topographic and tomographic parameters. It is important to remind that keratoconus can be insidious, especially when it develops monolaterally and before puberty. In such cases, it may negatively influence a child's social life and educational development, thus the importance of stressing that a timely diagnosis of keratoconus is fundamental in order to address it at an early stage. We encourage fellow ophthalmic surgeons to use

conservative treatments whenever possible (Rademaker, 2014), especially when dealing with very young patients with aggressive keratoconus.

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