

## CASE REPORT

# Corneal Collagen Cross-linking in a Prepubescent 10-Year-Old Girl with Aggressive Keratoconus

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## ABSTRACT

Keratoconus (KC) is a corneal ectatic disease that results in bilateral and asymmetrical corneal distortion, altered refractive powers and reduced vision. In 20 to 25% of cases, corneal transplantation may be required, although a number of conservative procedures are available. We report a case of an aggressive stage II keratoconus in a prepubescent 10-year-old girl, successfully treated with corneal collagen cross-linking (CXL) with epithelium removal.

**Keywords:** Cornea, Corneal collagen cross-linking, Keratoconus, Pediatric.

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## INTRODUCTION

Keratoconus (KC) is a corneal ectatic disease characterized by the gradual loss of thickness and change into a more conical shape of the corneal stroma. This leads to irregular astigmatism and subsequent decrease in visual acuity<sup>1</sup> while, in 20 to 25% of cases, corneal transplantation may be required. The clinical onset is generally bilateral and occurs at a young age, usually in the second decade of life.

Conservative surgical techniques, aimed at avoiding the need for a corneal transplantation, include asymmetric radial keratotomy (ARK),<sup>2</sup> mini asymmetric radial keratotomy (MARK),<sup>3</sup> circular keratotomy (CK)<sup>4</sup> and intrastromal corneal ring segments (ICRS).<sup>5</sup>

Another original technique recently developed is corneal collagen cross-linking (CXL), a procedure that directly addresses the intrinsic structural weakness of the cornea by reinforcing the corneal stroma, thus stopping keratoconus progression.<sup>6</sup> Corneal collagen cross-

linking consists in the removal of corneal epithelium, followed by the local application of a riboflavin solution and by subsequent UV-A irradiation, a technique firstly developed by surgeons at the Dresden Clinic, Germany.

We have successfully performed CXL since 2005, reporting both a mean refractive improvement of 1, 2 D and a stabilization of the disease following the intervention,<sup>7</sup> alone or combined with other refractive procedures, including MARK, photorefractive keratectomy (PRK) and phakic intraocular lens (PIOL) implantation. This case report, however, focuses on the outcomes of a single CXL treatment in a prepubescent 10-year-old girl with an aggressive stage II keratoconus.

## CASE REPORT

In May 2014, a 10-year-old girl presented with a bilateral keratoconus diagnosed 45 days before, ocular history included 6 years of vernal keratoconjunctivitis. The first diagnosis of hyperopia and astigmatism has been advised 4 years before by her optician.

The patient underwent a thorough clinical examination prior to the intervention, which included measurement of uncorrected (UCVA) and best corrected visual acuity (BCVA), computerized corneal topography, axial biometry, pachymetry, endothelial cell count, keratometry, Scheimpflug camera corneal analysis and slit lamp examination.

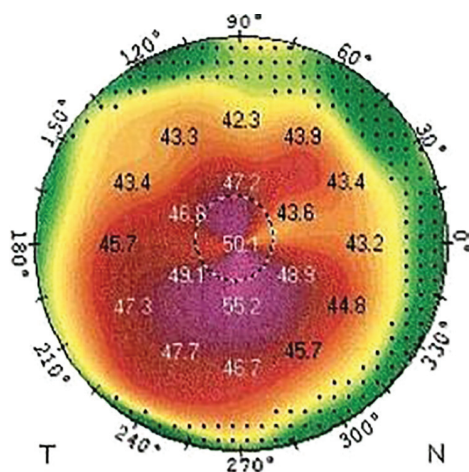
Corrected distance visual acuity (CDVA) was 20/25 in the left eye with +0.50/-1.50 × 160 and 20/32 in the right eye with -2.50 × 5. Central keratometry (K) values were 45 × 48 Dioptres (D) in the left eye and 45.5 × 52.5 D in the right eye. K<sub>max</sub> was 55.2 D in the right eye, showing a dramatic worsening compared to prior examinations, when K<sub>max</sub> values in the left eye and the right eye were comparable. Central pachymetry (Pentacam-Oculus Optikgerate GmbH, Wetzlar) was 534 μm in the right eye and 561 μm in the left eye. Thinnest pachymetry was 473 μm in the right eye (Fig. 1).

Given the rapid progression of the disease, we performed the CXL treatment in the right eye according to the Dresden protocol (epi-off), with benoxinate chloride 0.4% drops as a topical anesthesia. Removal of corneal epithelium was performed using an ophthalmic scalpel (MicroFeather; Feather Safety Razor Co, Ltd, Osaka,

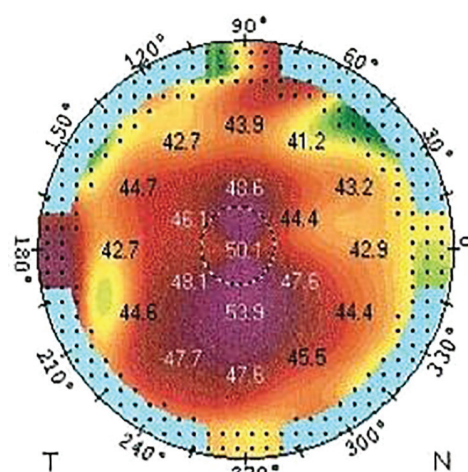
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**Fig. 1:** Corneal topography of the right eye before the intervention,  $K_{max}$  value was 55.2 D



**Fig. 2:** Corneal topography of the right eye 6 months after CXL,  $K_{max}$  and astigmatism improved

Japan) and a surgical microscope. Eyelids were kept open using surgical forceps during the procedure.

Riboflavin eye drops (B2 vitamin) were applied on the corneal stroma 30 minutes before UV-A application 10 times at 3 minute intervals, then riboflavin was applied six times at 5 minute intervals during UV-A application, with a total UV-A exposure time of 30 minutes. The UV-A source used was the UV-X illumination system version 1000, from IROC AG, Switzerland.

After treatment, a contact lens was placed and topical antibiotics and non-steroidal anti-inflammatory drops were prescribed. Clinical examination was performed on the following day. The contact lens was removed 3 days following the treatment and corneal epithelial healing was checked.

Three months after CXL, K values were  $45.75 \times 52$  D and  $K_{max}$  was 55 D. Six months after CXL, CDVA was 20/32, K values were  $45.5 \times 52$  and  $K_{max}$  decreased to 53.9 D (Fig. 2 and Table 1).

The patient's left eye has been regularly monitored with thorough clinical examinations, which showed a stable condition and no need for CXL.

## DISCUSSION

Corneal collagen cross-linking with epithelium removal is effective in reducing the progression of keratoconus, along with a painless and safe protocol for adult patients. For this reason, CXL has been recently applied to children as well.

Ophthalmic case studies in pediatric and adolescent patients with keratoconus have shown that, following CXL, KC is stabilized with a significant visual and topo-

graphic improvement. Due to an increasing awareness of the disease, a number of studies demonstrate that pediatric keratoconus is now often diagnosed and treated with success. While the number of children and adolescents affected by keratoconus varies depending on the kind of clinical trial, two studies show that it ranges between 29<sup>8</sup> and 38%<sup>9</sup> of all treated keratoconus patients. This parasurgical treatment is safe and there is no visually significant risk,<sup>10-12</sup> although one study suggests that the halting of the disease might not be as long lasting as in adults.<sup>12</sup> It is worth pointing out, however, that we never had to repeat the treatment in our 10-year experience with CXL, even when dealing with pediatric keratoconus.

Pediatric keratoconus shows a higher incidence rate and a more rapid progression compared to adult keratoconus,<sup>12</sup> notwithstanding the fact that the disease is most frequently diagnosed after adolescence. The progressive visual impairment influences the social life as well as the educational development of affected children. An early diagnosis of keratoconus, by adoption of adequate screening procedures, is therefore fundamental.

In our case, the diagnosis of keratoconus was early and its treatment with 'epi-off' CXL was timely. Manifest refraction remained stable during follow-up, as the K2 value, while  $K_{max}$  value improved. In our last examination, 6 months after CXL, the apex of the cone had decreased by 1.3 D and there were no complications, thus showing a stabilization of keratoconus as well as a visual improvement, despite the aggressiveness of the disease. These data let us hope for new improvements in the near future.

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**Table 1:** Pre- and post-CXL values, 6 months after the intervention

	IHA	IHD	ISV	IVA	KI	CKI	$K_{max}$
Pre-CXL	44.7	0.054	0.061	0.061	1.19	1.05	55.2
Post-CXL	44.8	0.051	0.061	0.047	1.18	1.05	53.9

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