Pediatric Keratoconus in a Tertiary Referral Center: Incidence, Presentation, Risk Factors, and Treatment

Sylvain el-Khoury, MD; Youssef Abdelmassih, MD; Adib Hamade, MD; Elise Slim, MD; Carole G. Cherfan, MD; Elias Chelala, MD; Jamal Bleik, MD; Elias F. Jarade, MD

ABSTRACT

PURPOSE: To report the incidence, clinical presentation, risk factors, and treatment outcome of pediatric keratoconus in a tertiary referral eye hospital in Beirut, Lebanon.

METHODS: In this retrospective study, the authors evaluated all patients with keratoconus 14 years or younger newly diagnosed at the Beirut Eye Specialist Hospital, Beirut, Lebanon, between January 2010 and December 2014. The incidence of pediatric keratoconus among all pediatric patients and among patients with keratoconus of all ages was assessed. Patients with pediatric keratoconus were evaluated for keratoconus stage, initial presentation, uncorrected distance visual acuity, corrected distance visual acuity (CDVA), corneal topography, and pachymetry. Patients were classified according to different treatment regimen groups and different follow-up visits were evaluated.

RESULTS: During 5 years, 16,808 patients were examined, of whom 2,972 were 14 years or younger. A total of 541 patients were diagnosed as having keratoconus; of those, 16 were 14 years or younger at the time of diagnosis. Hence, the incidence of keratoconus was 0.53% among pediatric patients and 3.78% among adult patients (> 14 years). Initial presentation was during routine checkup (1 of 16) for allergic conjunctivitis (3 of 16), reduced vision (10 of 16), and corneal hydrops (mimicking keratitis) (2 of 16). Except for 2 patients lost to follow-up, all eyes received corneal cross-linking treatment and 16 eyes received additional intracorneal ring segment implantation.

CONCLUSIONS: The incidence of pediatric keratoconus indicates that increased awareness for keratoconus among children is needed, mainly in cases of family history of keratoconus, ocular allergy/pruritus, poor CDVA, corneal hydrops, and/or high astigmatism.

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Keratoconus is a progressive and noninflammatory corneal ectasia most frequently diagnosed after adolescence.[1] Studies reporting the prevalence of keratoconus have shown a wide range of results (between 0.3 per 100,000 in Russia[2] and 2,300 per 100,000 in Central India).[3] This difference is believed to be due to ethnic, genetic, and environmental factors and to the diagnostic criteria used.[3-8] In children, keratoconus is a rare disease. To our knowledge, detailed information concerning the prevalence or incidence of pediatric keratoconus has not been reported yet. Pediatric keratoconus seems to progress faster and to be more advanced at the time of diagnosis than keratoconus in adults.[9,10] Due to its advanced stage at diagnosis, pediatric keratoconus bears a higher risk of corneal scarring in comparison to keratoconus in adults, thus resulting in a greater need for penetrating keratoplasty.[9,11-14] Treatment of pediatric cases of keratoconus in comparison to the adult form demonstrates several distinctive issues, such as poor patient compliance, higher rates of intolerance to contact lens wear, and higher rates of corneal graft rejection.[15]

In an attempt to begin demographic quantification of pediatric keratoconus, we report in the current study its incidence over a 5-year period as diagnosed in a tertiary referral eye hospital in Beirut, Lebanon. A systematic analysis of outcomes and safety of the treatment was also conducted.

PATIENTS AND METHODS

Patient Selection and Data Analysis

We conducted a retrospective survey on patients examined at the Beirut Eye Specialist Hospital, a tertiary eye care center in Beirut, Lebanon. The data of all patients who pre-

From Beirut Eye Specialist Hospital, Beirut, Lebanon (SK, YA, AH, ES, CGC, EFJ); Saint-Joseph University, Faculty of Medicine, Beirut, Lebanon (SK, YA, ES, EC); the Department of Ophthalmology, Rizk Hospital, Beirut, Lebanon (JB); the School of Medical Sciences, Lebanese University, Beirut, Lebanon (AH, JB); and Mediclinic Dubai Mall, Dubai, United Arab Emirates (EFJ).

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Correspondence: Elias F. Jarade, MD, Beirut Eye Specialist Hospital, Al-Mathaf Square, P.O. Box 116-5311, Beirut, Lebanon. E-mail: ejarade@yahoo.com

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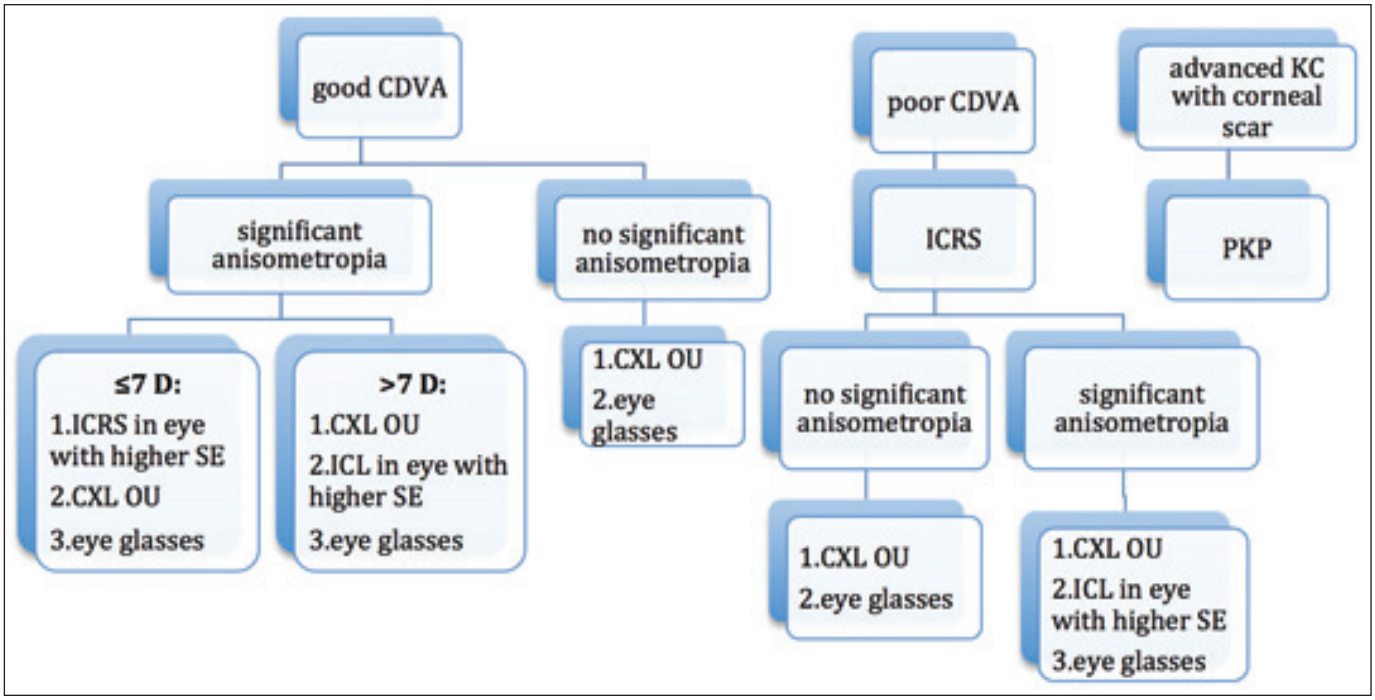

Figure 1. Dr. Jarade's nomogram for the treatment and visual rehabilitation of pediatric keratoconus. CDVA = corrected distance visual acuity; KC = keratoconus; ICRS = intracorneal ring segment; SE = spherical equivalence; CXL = corneal cross-linking; ICL = implantable collamer lens; PKP = penetrating keratoplasty; OU= oculus uterque

sented to the hospital between January 2010 and December 2014 were reviewed and categorized according to age (≤ 14 years and > 14 years) and presence of keratoconus. Institutional review board approval was obtained. The files of all patients with keratoconus 14 years or younger were retrieved and analyzed for relevant keratoconus data, such as stage, clearness of the cornea, uncorrected and corrected distance visual acuity (UDVA and CDVA, respectively), manifest refractive error, keratometric readings (K1 [flat], K2 [steep], and Kmax), and central corneal thickness. Diagnosis and classification of keratoconus were based on anterior and posterior corneal topography, which was performed using computed slit-scanning videokeratography (Pentacam HR; Oculus Optikgeräte, Wetzlar, Germany). Keratoconus stage was classified according to the Amsler–Krumeich criteria.[16] If applicable, procedures performed and relevant keratoconus data of the 6-month and 1-year follow-up were retrieved.

Statistical analysis was performed using SPSS for Windows software (version 22.0; SPSS, Inc., Chicago, IL). The Wilcoxon signed rank test for paired data was used to compare parameters. A two-tailed *P* value of .05 or less was considered statistically significant.

Patient Management

According to Dr. Jarade's protocol for the treatment of pediatric keratoconus (**Figure 1**), corneal collagen cross-linking (CXL) was recommended for all pediatric eyes with keratoconus regardless of whether progression was documented at the time of diagnosis. Intracorneal ring segment (ICRS) implantation was recommended to improve CDVA or to decrease anisometropia and occurred 4 weeks prior to CXL treatment. Implantable collamer lenses (ICLs) were indicated in the case of severe anisometropia, which could not be addressed by ICRS implantation alone, or in the case of a significant residual anisometropia (after ICRS implanation).

The treatment options, benefits, and risks were explained to the patients' parents preoperatively and written informed consent was obtained. All operative procedures were performed by the same experienced surgeon (EFJ) in the above-mentioned hospital.

The CXL procedure was performed in all patients under topical anesthesia and without sedation. Before entering the operating room, all children were psychologically well prepared in the presence of their parents and the surgical team. CXL was performed using standard epithelium-off CXL according to Wollensak et al.[17] In brief, during CXL the central 9-mm corneal epithelium was removed after topical anesthesia (proparacaine hydrochloride 0.5%), then soaked with 0.1% riboflavin–20% dextran solution for 10 minutes, followed by continuous instillation of the solution every 5 minutes for 30 minutes and administration of ultraviolet A light (UV-X illumination system, version 1000; IROC AG, Zu-

TABLE 1
Demographic Characteristics

Characteristic	No. of Patients (%)
Total	16,808
Adults (> 14 years)	13,836 (82.31%)
Pediatric (≤ 14 years)	2,972 (17.68%)
Keratoconus in all ages	541 (3.21%)
Pediatric keratoconus	16 (0.53%)[a]; (2.96%)[b]

[a]Percentage indicated in relation to number of pediatric patients.
[b]Percentage indicated in relation to number of patients with keratoconus of all ages.

rich, Switzerland) for 30 minutes. Postoperatively, soft contact lens was fitted until complete epithelialization of the cornea and all patients received topical antibiotics and steroids (gatifloxacin 0.3% [Zymar; Allergan Inc., Irvine, CA]) and tobramycin 0.3%–dexamethasone 0.1% (Alcon Laboratories, Inc., Fort Worth, TX) each six times daily for 1 week. After removal of the soft contact lens, loteprednol 0.5% eye drops (Bausch & Lomb, Inc., Rochester, NY) five times daily were initiated and slowly tapered over 5 weeks. Oral analgesic was prescribed whenever needed. The ICRS tunnels were created under topical anesthesia using a femtosecond laser (Intralase FS60; Abbott Medical Optics, Santa Ana, CA), except for 1 patient who was operated on under general anesthesia with manually dissected tunnels. In this patient, both eyes were operated on simultaneously. Depending on availability, Intacs SK (Addition Technology, Inc., Des Plaines, IL) or Keraring SI6 (Mediphacos Ltd., Minas Gerais, Brazil) ring segments were inserted. Incision site, ring thickness (0.25 to 0.45 mm), and number of segments (one or two) were largely dependent on the refractive errors to be treated by ICRS implantation. After ICRS implantation, patients were treated with topical antibiotics and steroids for 10 days.

RESULTS

Demographic Characteristics and Patient Presentation

In total, 2,972 patients aged 14 years or younger were examined at the Beirut Eye Specialist Hospital between January 2010 and December 2014, of whom 16 patients (30 eyes) were diagnosed as having keratoconus, resulting in a pediatric keratoconus incidence of 0.53% in children (**Table 1**). Pediatric keratoconus cases represented 2.96% of all keratoconus cases (541 patients) diagnosed at the hospital during the same period. Of the 16 pediatric keratoconus cases, 13 were boys and 3 were girls (male-to-female ratio = approximately 4:1). Mean age was 11.84 ± 2.14 years (range: 9 to 14 years). Three patients (21.4%) had a positive family history of keratoconus and 7 patients (43.8%) had a history of eye rubbing due to vernal keratoconjunctivitis, which was medically well controlled at the time of surgical treatment. Patient presentation and refractive variables are summarized in **Tables 2-3**, and **Table A** (available in the online version of this article).

TABLE 2
Characteristics of the Pediatric Population With Keratoconus

Characteristic	No. of Patients (%)
Age (y)	11.8 ± 2.0
Gender	
Male	13 (81%)
Female	3 (19%)
Initial clinical presentation	
Reduced visual acuity	10 (62.5%)
Allergic conjunctivitis	3 (18.75%)
Routine check-up	1 (6.25%)
Hydrops, mimicking keratitis	2 (12.5%)
Family history	
Yes	2 (12.5%)
No	14 (87.5%)
Allergy	
Yes	7 (43.8%)
No	9 (56.2%)
Stages and forms of keratoconus	
Stage I	13 (43.3%)
Stage II	7 (23.3%)
Stage III	1 (3.3%)
Stage IV	5[a] (16.7%)
Forme fruste keratoconus	4 (13.3%)
Treatment	
Lost to treatment and follow-up	4[b] (13.3%)
Only CXL	9 (30%)
Only CXL + ICRS	15 (50%)
CXL + ICRS + ICL	1 (3.3%)
PKP	1 (3.3%)

CXL = corneal cross-linking; ICRS = intracorneal ring segment implantation; ICL = implantable collamer lens implantation; PKP = perforating keratoplasty
[a]Of 5 keratoconus stage IV cases, two presented as corneal hydrops.
[b]Two patients were lost to treatment and follow-up.

Patient Management and Follow-up

Eyes were subcategorized in two large groups depending on CDVA and anisometropia and treated according to our treatment protocol for pediatric kerato-

TABLE 3
Refractive and Visual Acuity Parameters of Pediatric Patients Treated Only With CXL[a]

Parameter	Baseline	6-Month Follow-up After CXL	*P*
UDVA (logMAR)	0.43 ± 0.35	0.34 ± 0.40	.11
CDVA (logMAR)	0.07 ± 0.04	0.03 ± 0.04	.17
Sphere (D)	-2.03 ± 1.94	-1.64 ± 1.90	.03
Cylinder (D)	2.22 ± 2.10	1.78 ± 2.06	.06
Spherical equivalent (D)	-0.70 ± 1.42	-0.74 ± 1.28	.92
K flat (D)	41.90 ± 1.60	42.0 ± 1.40	.67
K steep (D)	44.20 ± 1.90	44.1 ± 2.10	.62
Kmax (D)	46.50 ± 4.20	45.3 ± 4.10	.18

CXL = corneal cross-linking; UDVA = uncorrected distance visual acuity; CDVA = corrected distance visual acuity; D = diopters; K = keratometry reading; Kmax = maximum keratometry
[a]*Parameters are shown before and 6 months after treatment.*

conus (**Figure 1**). Eyes with better CDVA received only CXL and eyes with worse CDVA or high anisometropia received ICRS implantation and CXL. Results are presented in detail below.

Two patients (4 eyes) were lost to follow-up. One eye presenting with severe hydrops was treated with 5% topical hypertonic saline, steroids, and artificial tears followed by perforating keratoplasty, but lost to follow-up after 6 months. One eye needed a secondary ICL insertion due to high residual myopia after ICRS-CXL.

CXL Without ICRS

CXL treatment alone was performed in 9 eyes of 7 patients. Patients were all boys and had a mean age of 12.8 ± 1.9 years and a relatively good CDVA of 0.07 ± 0.04 logMAR before treatment. Five eyes had stage I, 3 eyes had forme fruste, and 1 eye had stage II keratoconus. No progression of keratoconus was reported at the 6-month follow-up visit after CXL. Likewise, the 1-year follow up of 6 eyes showed stable results. UDVA, CDVA, sphere, cylinder, and keratometric readings showed no statistically significant changes at the 6-month follow-up visits compared to baseline. However, we noticed a trend toward a decrease in refractive error and a consecutive improvement in CDVA and UDVA (**Table 3**).

Combined ICRS and CXL

Sixteen eyes of 11 patients were treated with an ICRS implantation followed by CXL treatment 1 month later. Of those, 1 eye received a secondary ICL insertion. Mean CDVA was considerably lower than in the group treated only with CXL (0.42 ± 0.43 logMAR compared to 0.07 ± 0.04 logMAR). Four patients received bilateral ICRS implantation. Two patients were females and the mean age of all patients was 12.1 ± 1.9 years. Six eyes had stage I, 6 eyes had stage II, 1 eye had stage III, and 3 eyes had stage IV keratoconus. Of 11 patients, 1 was lost to follow-up and returned 4 years after treatment. This patient was not included in the statistical analysis.

Postoperative refractive and topographic results are summarized in **Table 4**. All variables showed a statistically significant improvement 6 months after combined treatment, resulting in 50% of eyes (7 of 14) reaching a UDVA of at least 20/40 (0.30 logMAR). Both UDVA and CDVA improved significantly from 0.95 ± 0.59 to 0.37 ± 0.26 logMAR ($P = .001$) and from 0.42 ± 0.43 to 0.15 ± 0.14 logMAR ($P = .004$), respectively. Sphere and spherical equivalent decreased significantly from -6.13 ± 4.28 to -3.86 ± 5.07 D ($P = .003$) and from -4.39 ± 4.12 to -2.67 ± 4.40 D ($P = .006$), respectively. Topography showed a statistically significant flattening of all keratometric readings, mostly in Kmax, which decreased from 55.60 ± 10.57 to 50.70 ± 5.66 D ($P = .009$).

The safety index (mean postoperative CDVA [decimal] / mean preoperative CDVA [decimal]) of the combined procedure at the 6-month follow-up was 1.84 and the efficacy index (mean postoperative UDVA [decimal] / mean preoperative CDVA [decimal]) was 1.12.

Combined ICRS and CXL Followed by ICL Implantation

In one eye, a high residual refractive error of -13.00 D sphere and 5.00 D cylinder persisted after the ICRS-CXL procedure, reaching a CDVA of 0.40 logMAR. After refraction stabilized, we opted for an ICL insertion, which was performed 3 years after the sequential treatment. Postoperative topography is not available

TABLE 4
Refractive and Visual Acuity Parameters of Pediatric Patients Treated With ICRS Insertion Combined With CXL

Parameter	Baseline	6-Month Follow-up After ICRS+CXL	*P*
UDVA (logMAR)	0.95 ± 0.59	0.37 ± 0.26	.001
CDVA (logMAR)	0.42 ± 0.43	0.15 ± 0.14	.004
Sphere (D)	-6.13 ± 4.28	-3.86 ± 5.07	.003
Cylinder (D)	3.46 ± 1.42	2.41 ± 1.56	.034
Spherical equivalent (D)	-4.39 ± 4.12	-2.67 ± 4.40	.006
K flat (D)	47.10 ± 6.67	43.8 ± 5.36	.001
K steep (D)	50.60 ± 6.25	47.70 ± 5.01	.001
Kmax (D)	55.60 ± 10.57	50.70 ± 5.66	.009

ICRS = intracorneal ring segment implantation; CXL = corneal cross-linking; UDVA = uncorrected distance visual acuity; CDVA = corrected distance visual acuity; D = diopters; K = keratometry reading; Kmax = maximum keratometry
[a]*Parameters are shown before and 6 months after treatment.*

yet; UDVA 2 months after ICL implantation was 0.47 logMAR.

Complications

No major complications, such as keratitis, endophthalmitis, ICRS rejection, or migration were recorded after treatment. All epithelial defects healed within 4 days following CXL.

DISCUSSION

Demographics

Between January 2010 and December 2014, 16 cases of keratoconus in children 14 years or younger were diagnosed in a tertiary referral eye center, resulting in an incidence of keratoconus of 0.53% among pediatric patients evaluated. In comparison, the incidence of keratoconus among patients older than 14 years was found to be 3.8% in the above time frame. These numbers are expected to be higher than those of the general population because they were measured out of a pool of ophthalmic patients examined in a tertiary referral center (selection bias). Further, we know that the prevalence of keratoconus is highly variable around the world, with indications that it is more prevalent in our region when compared to northern countries.[14,18,19]

Keratoconus has been reported to appear around puberty.[1] However, in some cases this disease has been found to start in the first 10 years of life.[20,21] Concerning the prevalence, a screening among school children undertaken in southwestern Nigeria reports a 0.08% prevalence of blindness due to keratoconus,[22] whereas a screening using topography performed in New Zealand on Maori/Polynesian and European adolescents (16.8 ± 1.05 and 16.2 ± 1.3 years, respectively) showed 19% of topographies reminiscent of emerging keratoconus in at least one eye.[23] Overall, we found a substantial percentage of pediatric keratoconus cases, representing 2.59% of all patients with keratoconus. Although this is a relatively small percentage, physicians should keep a high index of suspicion for keratoconus in children, especially because keratoconus in a younger age group is reported to be more severe than in adults[21] and rapidly progressive with a short interval between the onset of functional symptoms and the development of a severe form of keratoconus.[9,10] In our study, approximately 30% of patients (5 of 16) were diagnosed as having stage IV keratoconus at presentation, 2 of them having already developed a corneal hydrops. This is similar to the cases studied by Léoni-Mesplié et al., where 27.8% of children younger than 15 years had a stage IV keratoconus (compared to 7.8% of patients ≥ 27 years old[9]) and indicates the severity of pediatric keratoconus at time of presentation. Gender distribution also showed convergence with reported epidemiological data on keratoconus in children. Eighty-one percent of our patients were male, similar to the 75% male ratio in pediatric keratoconus (same age group) in the study by Léoni-Mesplié et al.[20]

Risk Factors and Presentation

In our clinic, a decrease in visual acuity was the most frequent complaint at initial presentation (62.5% of cases). Two patients (12.5%), 9- and 10-year-old boys, were referred to our center for a therapy-refractory keratitis. However, a careful study including topography unmasked the underlying cause as a corneal hydrops due to advanced keratoconus (for details on the first case refer to Slim et al.[24]). Three patients (18.8%) presented initially for ocular pruritus associated with allergic conjunctivitis. However, after careful examination and evaluation of case history, 7 patients

(43.8%) had an allergic conjunctivitis at the slit lamp or a history of eye rubbing. Two patients (12.5%) had a positive family history of keratoconus. The family screening performed subsequently revealed two new keratoconus cases in one family (not included in the study because the patients were older than 14 years).

In our pediatric population, the two major risk factors for pediatric keratoconus seem to be allergic conjunctivitis and a family history of keratoconus. In otherwise healthy children, atopy and subsequent eye rubbing have been reported as major factors associated with an increased incidence of keratoconus.[25-30] Therefore, every child with ocular allergies and/or eye rubbing should be considered as a keratoconus suspect to prevent misdiagnosis.[24,29,30] Concerning family history, studies where screening for keratoconus is done by corneal topography have shown that first-degree relatives of patients with keratoconus have an estimated 15 to 67 times higher prevalence of keratoconus than in the general population.[31] Hence, keratoconus should be further suspected in every child with a positive family history, an unexplained loss of vision (especially if associated with a high error of refraction), astigmatism that is not amenable to 20/20 correction, or a new onset of astigmatism. In the case of positive family history, corneal topography screening should be started as soon as the child is able to fixate and place his chin for topography. In the absence of suspicious findings, topography should be repeated yearly afterward. In the case of suspicious topographies, the examination should be repeated every 1 to 3 months (vs 6 to 12 months in adolescents and adults), due to the highly progressive nature of keratoconus at this age.

Treatment

Pediatric keratoconus treatment is time sensitive. The disease has a fast progression rate, with a more difficult visual rehabilitation mainly in the presence of anisometropia and with often-observed contact lens intolerance. Therefore, no time should be lost between diagnosis and treatment.

CXL was proven to be effective for halting the progression of the disease in children[32,33] and to have a good safety profile.[10,32-35] It also showed an effect on CDVA; however, this effect seems to be minimal, reported to be approximately one line.[34] In our study, 9 eyes that were treated only with CXL showed a stable situation with a minimal but statistically nonsignificant improvement in CDVA (0.07 ± 0.04 logMAR preoperatively versus 0.03 ± 0.04 logMAR postoperatively, $P = .17$), in UDVA (0.43 ± 0.35 logMAR preoperatively versus 0.34 ± 0.40 logMAR postoperatively, $P = .11$), and in any other refractive parameter studied, except for a statistically significant but small improvement in sphere (from -2.03 ± 1.94 to -1.64 ± 1.90 D, $P = .03$) 6 months after treatment. Note that for this treatment group, patients had been preoperatively selected to have a good CDVA.

However, for patients with poor CDVA and/or severe anisometropia, an improvement of CDVA and a decrease in anisometropia is needed. ICRS implantation was found to be safe and effective for improving CDVA in adult keratoconus[36] and similar results for Intacs SK (Addition Technology) implantation are reported in children and adults.[37] Apart from this, to the best of our knowledge, ICRS implantation in children is not yet reported. A 5-year follow-up after ICRS implantation (without CXL) in relatively young patients (25.75 ± 3.59 years) with progressive keratoconus showed no significant influence of the treatment in stopping the progression of the disease,[38] leading to the rationale of combining ICRS implantation and CXL. In the current study, a combined ICRS-CXL procedure was shown to be safe (safety index 1.84) and effective (efficacy index 1.12) in addressing the aforementioned difficulties of pediatric keratoconus. In 16 eyes treated with the combined procedure, CDVA significantly improved from 0.42 ± 0.43 logMAR preoperatively to 0.15 ± 0.14 logMAR 6 months postoperatively ($P = .004$). UDVA and spherical equivalent also showed a significant improvement from 0.95 ± 0.59 logMAR preoperatively to 0.37 ± 0.26 logMAR 6 months postoperatively ($P = .001$) and from -4.39 ± 4.12 to -2.67 ± 4.40 D ($P = .006$), respectively. A significant flattening of keratometric readings was also recorded.

In light of a lack of guidelines for the treatment of pediatric keratoconus (to the best of our knowledge, there is no standard protocol), we have set up a treatment nomogram that serves as a reference for the patients diagnosed in our clinic, including the patients analyzed in this article. **Figure 1** visualizes this treatment plan. It is based on the quality of CDVA and as the presence of significant anisometropia (one that might result in the patient developing amblyopia or intolerance to glasses) or corneal scar.

Due to the high rate of progression of keratoconus in children (the literature reports it to be significantly higher than in older age groups[9,12-14]), we suggest that CXL always be performed in pediatric keratoconus to stabilize the cornea, regardless of stage and evolution of the disease. This is in accordance with the keratoconus management algorithm suggested by Shetty et al., in which all demographic high-risk patients are advised to receive CXL treatment,[39] as well as with Dr. Jarade's protocol for the treatment of keratoconus or ectasia after surgery.[36] This approach is further supported

by our clinical observation of aggressive keratoconus behavior in young patients in the Middle East. Accordingly, in children with good CDVA and no significant anisometropia, CXL alone is performed. If a significant anisometropia is present, treatment depends on the refractive difference between eyes. If it surpasses 7.00 D, CXL is performed in both eyes and an ICL is inserted in the eye with the higher spherical equivalent value. ICL insertion should be done at least 6 months after CXL and only if the previous 2 months showed stable topography. If it is 7.00 D or less, an ICRS is inserted in the eye with the higher spherical equivalent and 1 month later CXL is performed in both eyes.

In pediatric keratoconus cases with poor CDVA, an ICRS is inserted in the respective eye and followed by CXL 1 month later for both eyes. If anisometropia persists and is not tolerated or has the risk of developing into amblyopia, an ICL is inserted in the eye with the higher spherical equivalent value. In cases of ICRS implantation, the number of ICRSs depends on the presence of significant anisometropia. If not present, corneal irregularities are corrected with one ICRS. In case of significant anisometropia and/or amblyopic risk, two ICRS are inserted. Penetrating keratoplasty should be reserved for advanced cases of keratoconus presenting with corneal scarring. We consider this to be the last option treatment because penatrating keratoplasty is fraught with many complications and a high incidence of graft failure.[11,15] Also, the success of the procedure is dependent on the dedication of the family in following a rigorous postoperative regimen. Postoperative glaucoma, strabismus, self-induced trauma, and immune rejection are difficult complications that may occur.

Controlling allergic disease and avoiding eye rubbing is of primary importance and should be assessed at every visit.

Overall, the presented treatment plan has led to good postoperative results; however, it remains a first attempt to unify treatment strategies and a consolidation of this plan needs more data and especially long-term results.

Although it is rarely reported, pediatric keratoconus is an existing condition and may present with different clinical manifestations. The prevalence of keratoconus in children is often underestimated and therefore an increased awareness is needed among ophthalmologists for this condition, especially if the child has a positive family history of keratoconus, a history of ophthalmic allergies and/or pruritus, poor CDVA, corneal hydrops, or high astigmatism. We recommend performing CXL in all cases of pediatric keratoconus regardless of evidence of progression. ICRS implantation proved to be safe and effective in enhancing CDVA and decreasing spherical equivalent. Phakic intraocular lenses are preserved to neutralize severe anisometropia and perforating keratoplasty should be kept as a last resort treatment in stage IV keratoconus because of its high risk of failure and complications. Concerning treatment, more data, especially long-term results, are needed to evaluate its risks and benefits.

AUTHOR CONTRIBUTIONS

Study concept and design (AH, EC, EFJ); data collection (SK, YA, AH, ES, JB); analysis and interpretation of data (SK, YA, CGC); writing the manuscript (SK, YA, AH, ES); critical revision of the manuscript (SK, YA, CGC, EC, JB, EFJ); administrative, technical, or material support (EFJ); supervision (EFJ)

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TABLE A

Relevant Variables of Patients With Pediatric Keratoconus at Time of Diagnosis

Variables	Minimum	Maximum	Mean ± SD
Age (y)	9	14	11.87 ± 2.03
UDVA (logMAR)	0	3.00	0.86 ± 0.73
CDVA (logMAR)	0	1.30	0.41 ± 0.65
Sphere (D)	-15.25	0.00	-4.58 ± 4.48
Cylinder (D)	0.00	7.00	3.12 ± 1.88
Spherical equivalent (D)	-12.63	1.37	-2.90 ± 4.05
K1 flat (D)	40.70	61.10	45.37 ± 5.53
K2 steep (D)	42.60	66.70	48.70 ± 6.38
Kmax (D)	43.20	86.00	53.66 ± 11.12
CCT (μm)	396	611	487 ± 54.9

SD = standard deviation; UDVA = uncorrected distance visual acuity; CDVA = corrected distance visual acuity; D = diopters; K = keratometry reading; Kmax = maximum keratometry; CCT = central corneal thickness