ORIGINAL ARTICLE

Keratoconus in children in Tunisia: Epidemiological, clinical and therapeutic features

Kératocône chez l'enfant en Tunisie: Particularités épidémiologiques, cliniques et thérapeutiques

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Abstract

Aim: To report the clinical and therapeutic particularities of pediatric keratoconus (KC).

Methods: Retrospective study focusing on patients aged less than 18 years, presenting with KC and followed in a tertiary reference center in Sfax, Tunisia.

Results: Our study involved 38 eyes of 20 children. We found a mean age of 12.8 years, a family history of keratoconus for 25% of cases and an atopic background in 30% of children. The mean best-corrected visual acuity was 5.3/10, the mean myopia was -6.3D and the mean total astigmatism was -4.9D. The average maximum simulated keratometry (Kmax) was 54.6 D. The visual acuity decrease was correlated with the presence of corneal opacities, with all keratometric indices except Skewed Radial Axes (SRAX) at 3 and 5 mm, with all elevation parameters and those of pachymetry except the delocalization of the thinnest point. 71.1% of eyes were classified as clinical KC and 47% of eyes were classified as Amlser-Krumeich stage 4. 42.1% of eyes were fitted with glasses and 57.9% with contact lenses. Six patients (10 eyes) underwent cross-linking (CXL). We noted an evolution of the KC for 64.29% of eyes not treated with CXL and a stabilization of 80% of eyes treated with CXL. The difference in follow-up parameters between the two groups was significant.

Conclusion: Pediatric KC is a severe disease, often diagnosed at a late stage. CXL is a safe and effective way to stop the disease.

Key words: Keratoconus; Child; Corneal topography; Cross-linking

Résumé

Objectif: Rapporter les particularités cliniques et thérapeutiques du kératocône (KC) pédiatrique.

Méthodes: Etude rétrospective sur les patients de moins de 18 ans présentant un KC et suivis dans un centre de référence à Sfax en Tunisie.

Résultats: Notre étude a concerné 20 enfants (38 yeux). Nous avons trouvé un âge moyen de 12,8 ans, des antécédents familiaux de KC pour 25% des cas et un terrain atopique chez 30%. La meilleure acuité visuelle corrigée moyenne était 5,3/10, la myopie moyenne était de -6.3D et l'astigmatisme total moyen était de -4.9D. La moyenne de la kératométrie simulée maximale (Kmax) était de 54,6 D. La baisse de l'acuité visuelle était corrélée à la présence des opacités cornéennes, à tous les indices kératométriques sauf la SRAX (Skewed Radial Axes) à 3 et 5 mm, à tous les paramètres d'élévation et ceux de la pachymétrie sauf la délocalisation du point le plus fin. 71,1% des yeux étaient classés comme KC avéré et 47% des yeux étaient classés au stade 4 d'Amlser-Krumeich. 42,1% des yeux ont été équipés par des lunettes et 57.9% par des lentilles de contact. Six patients (10 yeux) ont eu un cross-linking (CXL). Nous avons noté une évolution du KC pour 64,29% des yeux non traités par CXL et une stabilisation de 80% des yeux traités par CXL. La différence des paramètres de suivi entre les deux groupes était significative.

Conclusion: Le KC pédiatrique est une maladie sévère, souvent diagnostiquée tardivement. Le CXL constitue un moyen sûr et efficace pour stopper la maladie.

Mots clés: Kératocône ; Enfant ; Topographie cornéenne ; Cross-linking

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Introduction

Keratoconus (KC) is an ectatic disease characterized by a deformation of the cornea that gradually becomes thinner. It is typically diagnosed in late adolescence and reaches its most advanced stage around 20-30 years of age and progresses until around age of 40 years, eventually stabilizing after (1). However, it is possible that it manifests in childhood in a more severe form, progressing more quickly and generally diagnosed at an advanced stage, affecting the child's quality of life, social development and schooling.(2). The diagnostic and therapeutic approaches for pediatric KC (PKC) are different from those for adults. The present study aimed to report the epidemiological, clinical and therapeutic particularities of PKC collected in a tertiary reference center in Sfax, Tunisia.

Methods

Patients selection and data analysis

We conducted a retrospective study, between August 2013 and February 2020, on patients aged under 18 years and presenting with KC followed in the Ophthalmology department of the Habib Bourguiba Hospital in Sfax. We excluded eyes presenting with acute KC, those which underwent keratoplasty (KP) or those having another ocular condition responsible for a decrease in visual acuity.

All available demographic and clinical data were collected including age, sex, functional signs, family history of KC and atopy, personal history of atopy and systemic pathology, ophthalmological history of allergic conjunctivitis, eye rubbing and other associated ocular pathology. Previous therapeutic modalities and duration of follow-up were also noted.

All patients underwent an ocular refraction specifying the value of myopia (S), total astigmatism (TA) and spherical equivalent (SE), and a complete ophthalmological examination. Best corrected visual acuity (BCVA) was measured using the Snellen scale, collected in decimal scale and converted to Log MAR. Slit lamp biomicroscopy included examination of the ocular adnexa, conjunctiva and cornea for signs of vernal keratoconjunctivitis (VKC) and documentation of signs of KC such as Munson's sign, thinning and corneal ectasia, Vogt's striae, scar lines, prominent corneal nerves, Fleischer's ring or deep opacities, and fundus examination after pupillary dilation. Corneal topography using ORBSCAN scanning-slit topography (Bausch and Lomb/Technolas perfect vision) was performed to confirm the diagnosis. The information collected in corneal topography was rendered in the form of a quad-map. The curvature map made it possible to classify the topographical pattern according to the Rabinowitz classification (3) and calculate the KISA index and the Skewed Radial Axes (SRAX). The anterior and posterior elevation maps gave the value of the maximum elevation relative to the reference sphere (BFS). The pachymetric map allowed the measurement of the central pachymetry (CP) and the thinnest point (TP) with its vertical (Y) and horizontal (X) coordinates and the delocalization of the TP. We also obtained numerical indices including maximum and minimum simulated keratometry (Kmax and Kmin), irregularity, average power, cylinder and axis of the two main meridians in the 3 and 5 mm zone. A swept source optical coherence tomography of the anterior segment (AS-OCT) by the DRI OCT Triton Swept Source machine was also performed. It allowed us to measure corneal pachymetry and visualize the different histological layers of the cornea.

KC was classified according to the clinical classification of Gatinel and Saad(4), according to the Amsler-Krumeich classification and according to the tomographic classification(5).

Management

Management included avoidance of eye rubbing and medical treatment of allergic conjunctivitis. Optical treatment was carried out by optical glasses for early forms and by contact lenses (CL) for advanced forms : soft toric CL, rigid gas permeable lenses (RGPL) or minisceral lenses (SL). Physical treatment with cross-linking (CXL) of corneal collagen was immediately indicated after eliminating the contraindications: corneal thickness < 400 µm, central opacity of the cornea and severe dry eye. (6). Surgical treatment with KP was indicated in cases of failure of all optical means, in cases of acute hydrops or in cases of stage 4 KC.

Regular follow-up of the patients was carried out every six months with a careful interview specifying the evolution of functional signs, an objective refraction, a measurement of the BCVA, a complete ophthalmological examination and an AS-OCT. Corneal topography was performed every year. It was requested at 6 months in the case of signs of progression of the KC or in the case of realization of a CXL. Disease progression was defined by an increase in Kmax ≥ 1 D, a decrease in cylinder ≥ 1 D at 6 months interval, decrease in BCVA or change in CL parameters at renewal.(7).

Statistical analysis

Statistical analysis was carried out with the Statistical Package for Social Sciences (SPSS) software (version 26.0). Quantitative variables were expressed as mean ± standard deviation or median. Qualitative variables were expressed in numbers and percentages (%), and compared using the chi-square test. For the comparison of means, we used the Student's t test or the Mann Whitney U test whenever the normality of the distributions was not respected or when the size of one of the groups was small. The connections between two quantitative variables were made by the Pearson rank correlation coefficient in the case of a Gaussian distribution and the Spearman correlation whenever the normality of the distributions was not respected. In all statistical tests, the significance level was set at 0.05.

Results

Thirty-eight eyes of 20 children were included. Eighteen children had bilateral KC, one patient had unilateral KC and one child had an eye operated on for KP. The average follow-up was 13.83 ± 8.45 months [6-28].

Patients characteristics

The mean age at diagnosis of our patients was 12.8 ±3.8 years. 20% were aged less than 10 years; 45% between 10 and 14 years old and 35% over 15 years old. The M/F sex ratio was 0.81. We found a familial atopic background in 6 patients (30%) and a family history of KC in 5 patients (20%) : 3 cases in siblings, 1 case in the father and 1 case in the cousin. Seven children (35%) had allergic conjunctivitis, two of whom (10%) presented with VKC. One patient had Turner syndrome (5%). The rest of the patients characteristics are summarized in Table 1.

Table 1. Patients Characterisitics

Parameter		Values {Mean (range) or No (%)}
Mean Age		12.8 ±3.8 years (6-18 years)
Sex		
	Male	9 (45%)
	Female	11 (55%)
Family history		
	Consanguineous marriage	8 (40%)
	Atopy	6 (30%)
	Keratoconus	5 (20%)
Personal history		
	Allergic conjunctivitis	7 (35%)
	Turner syndrome	1 (5%)
	Bilateral pseudophakia	1 (5%)
Discovery circumstances and symptoms		
	Fortuitous	14 (70%)
	Blurred vision	17 (85%)
	Eye rubbing	16 (80%)
	Decrease in visual acuity	15 (75%)
	Photophobia	3 (15%)
Optical correction		
	None	11 (55%)
	Eye glasses	7 (35%)
	Contact lenses	2 (10%)

Ophthalmological examination results

The mean BCVA was 0.368 ± 0.37 log MAR and half of the eyes (19 eyes; 50%) had a BCVA between 0.3 and 1 log MAR. The study of refraction was only possible in 32 eyes (84%) and it showed an average myopia of -6.3 ±5.8D and an average TA of -4.9 ±2.3 D (Table 2).

Slit lamp examination showed the presence of corneal protrusion in 27 eyes (71.1%), Munson's sign in 21 eyes (55.3%), Vogt's striae in 12 eyes (31. 6%), scar lines in three eyes (7.9%), Fleisher's ring in one eye (2.6%), and deep opacities in seven eyes (18.4%).

Table 2. Refraction Data

Refraction parameters	Mean ± standard deviation (D)	Extremes (D)
S	-6.3±5.8	[-17, +2.5]
C	-4.9±2.3	[-8.8, -1.5]
SE	-8.7±6.3	[-21.5, +1.5]
K1	48±5.5	[41 - 65]
K2	52.9±2.7	[42.8 - 74.5]
Kavg	50.3±6.3	[41.9 - 69.3]
CA	4.9±2.5	[1.5 – 12]

CA: corneal astigmatism; **C**: Cylinder; **D**: Diopter; **SE**: Spherical equivalent; **K1**: Minimal keratometry; **K2**: Maximum keratometry; **Kavg**: Average keratometry; **S**: Sphere

Topographic data

All patients underwent corneal topography. The majority of eyes (28 eyes; 73.7%) had a central (13 eyes; 34.2%) or paracentral cone (15 eyes; 39.5%). The most identified Rabinowitz pattern was the « G » pattern (9 eyes; 23.7%) followed by « H » (7 eyes; 18.4%) and « J » (6 eyes; 15.8%). Loss of enantiomorphism was noted in 26 eyes (68.4%).

We found a Kmax > 47.2 in 84.2%, a SRAX > 21° in 78.9% and KISA > 100% in 71.1%. The mean value was -5.4D for astigmatism; 54.6 D for Kmax and 49.3 D for Kmin. The rest of the means of the keratometric indices are summarized in Table 3. BCVA was inversely and significantly correlated with the different keratometric indices with the exception of SRAX (Table 3).

Table 3. Topographic keratometric, elevation and pachymetric parameters and their correlations with BCVA

Parameter		Mean ±standard deviation	Correlation coefficient with BCVA	p
Sim K	Ast (D)	-5.4±3.2	-0.707	<0.01
	Kmax (D)	54.6±8.4	-0.767	<0.01
	Kmin (D)	49.3±6.2	-0.668	<0.01
3mm area	Irregularity (D)	5.4±3.2	-0.483	0.002
	Average power (D)	50.3±5.5	-0.762	<0.01
	Average Ast (D)	4.5±2.4	-0.761	<0.01
	SRAX (Degree)	31.5±9.9	0.119	NS
5mm area	Irregularity (D)	6.8±4.3	-0.625	<0.01
	Average power (D)	47.2±3.7	-0.752	<0.01
	Average Ast (D)	2.13±1.1	-0.341	0.039
	SRAX (Degree)	40.3±7.1	-0.036	NS
Anterior elevation (µm)		49.6±17.2	-0.508	0.01
Posterior elevation (µm)		77.7±29.4	-0.511	0.01
CP (µm)		429.9±87.2	0.543	<0.01
TP (µm)		410.2±93	0.577	<0.01
CP-TP difference (µm)		20.8±13.5	0.05	NS
Y displacement of the TP: CP-TP (cm)		-0.52±0.76	-0.522	0.01

Ast: Astigmatism; **Kmax**: Maximum keratometry; Kmin: Minimum keratometry; **BCVA**: Best corrected visual acuity; **CP**: Central pachymetry; **Sim K**: Simulated keratometry; **SRAX**: Skewed radial axes; **TP**: finest pachymetry

We noted an elevation >40µm in more than half of the eyes, more frequent on the posterior surface than on the anterior surface (29 eyes (76.3%) and 25 eyes (65.8%) respectively). BCVA was inversely and significantly correlated with topographic elevation parameters (Table 3). The mean CP was 429.9±87.2 µm. Fifteen eyes (39.5%) had a CP > 450µm. The mean TP was 410.2±93 µm and 19 eyes (50%) had a TP between 200 and 400 µm. Thirty eyes (78.9%) had a difference of more than

10 µm between CP and TP and the average Y-shift of the TP relative to the CP was -0.52 ± 0.76 cm. BCVA was significantly correlated with the different pachymetric parameters with the exception of TP delocalization (Table 3). AS-OCT was performed in 18 eyes (47.4%). The average CP was 399.7±108.5 µm, significantly correlated with the topographic one (r=0.84; p<0.0001).

Classification

Twenty-seven eyes (71.1%) had proven KC according to the clinical classification of Gatinel and Saad, 18 eyes (47%) were in stage 4 according to the Amsler-Krumeich classification, and 8 eyes (21.1%) in stage 2 according to the tomographic classification (Table 4).

Table 4. Classification of keratoconus

Classification		Number of eyes (%)
Clinical of Gatinel and Saad	Proven KC	27 (71.1%)
	Suspect KC	5 (13.2%)
	Forme fruste KC	5 (13.2%)
Amsler-Krumeich	Stage 1	10 (26.3%)
	Stage 2	4 (10.5%)
	Stage 3	6 (15.8%)
	Stage 4	18 (47%)
Tomographic	Stage 1	3 (7.9%)
	Stage 2	8 (21.1%)
	Stage 3	2 (5.3%)
	Stage 4	4 (10.5%)
	stage 5b	1 (2.6%)

Therapeutic management

All our patients had optical correction. Sixteen eyes (42.1%) were equipped with glasses and 22 eyes (57.9%) with CL: RGPL (12 eyes; 31.6%); SL (8 eyes; 21%) and soft toric CL (2 eyes; 5.3%). The improvement in VA was significant for both groups and more significant for eyes equipped with CL (p=0.03) (Table 5).

Table 5. Average BCVA before and after correction and visual acuity gain

VA in Log MAR		Mean±standard deviation [extremes]	Gain (lines)	P
Correction by glasses	BCVA before	0.83 ±0.51[0.4 – 2]	4.9 ±0.32	**<0.0001**
	BCVA after	0.34 ± 0.48 [0 – 2]		**0.03**
Correction by CL	BCVA before	1.06 ± 0.35 [0.5 – 2]	8.4±0.22	**<0.0001**
	BCVA after	0.22 ± 0.14 [0 – 0.7]		

BCVA: Best corrected visual acuity; **CL**: contact lenses

Six patients (10 eyes; 26.3%) underwent CXL. Table 6 summarizes the refractive and topographical parameters of eyes with and without CXL.

The evolution for eyes without CXL was marked by a stabilization of the KC for 10 eyes (35.71%) and an evolution for 18 eyes (64.29%) including 4 eyes complicated by hydrops. For eyes with CXL, we noted stabilization of the KC in 4 patients (8 eyes) and progression in one patient (2 eyes). The latter had persistent eye rubbing, per-annual allergic conjunctivitis with a decentered cone in the corneal topography.

After 24 months of follow-up and comparing the two groups with and without CXL, we noticed a significant difference in the evolution of refractive parameters, BCVA, Kmax and also in terms of evolution and stabilization (Table 6). This difference was not significant for the evolution of TP. For eyes without CXL, we found a significant worsening of all parameters studied except Kmax. For eyes with CXL, we noted significant worsening of S, TA and SE (Table 6). A KP was indicated for 5 eyes.

Table 6. Evolution of refractive and topographical parameters of eyes with and without CXL

	Eyes without CXL (28 eyes)				Eyes with CXL (10 eyes)				Comparison of the differences of the two groups (p)
Parameter	**Initial value**	**Value at 24 months**	**Difference**	**p**	**Initial value**	**Value at 24 months**	**Difference**	**p**	
S(D)	-3.56 ± 4.51	-5 ±4.96	-1.44 ± 0.91	0.003	-6.17 ± 5.01	-6.85 ±5.42	-0.67 ±0.67	0.010	0.003
TA(D)	-5.16 ±2.37	-6.34 ±2.58	-1.19 ±0.99	0.01	-4.85 ±2.42	-5.57 ±2.95	-0.72 ±0.87	0.029	0.011
SE (D)	-6.28 ±6.27	-8.17 ±5.03	-1.89 ±0.99	0.001	-8.61 ±6.16	-9.63 ±6.78	-1.02 ±0.85	0.004	0.004
BCVA (log Mar)	0.34 ± 0.32	0.58 ±0.32	0.21 ±0.24	0.038	0.2 ± 0.19	0.235 ±0.17	0.04 ± 0.12	NS	0.035
Kmax (D)	54.16 ±4.89	58.238 ±7.01	4.08 ±5.82	NS	50.36 ±5.63	51.46 ±5.95	1.10 ±1.31	NS	0.027
TP (µm)	354 ±84.11	345.63 ±83.97	-8.37 ±9.35	0.039	470.2 ±46.71	457.8 ±48.13	-12.40 ±26.47	NS	NS
Evolution (number of eyes)		18 eyes (64.29%)				2 eyes (20%)			0.016
Stabilization (number of eyes)		10 eyes (35.71%)				8 eyes (80%)			0.016

TA: Total astigmatism; **CXL**: Cross-linking; **SE**: Spherical equivalent; **Kmax**: Maximum keratometry; **BCVA**: Best corrected visual acuity; **S**: Sphere; **TP**: thinnest pachymetry

Discussion

In our series, 20% of children had familial KC. Hashemi et al (8) after analyzing 29 articles, found that family history of the disease is the main risk factor for KC. PKC with family history was reported with an average rate of 10%, ranged from 5 to 27.9% (9). KC can be associated with other genetic diseases such as Down syndrome, Ehlers-Danlos syndrome, or Turner syndrome as in the case of our patient (7). Currently, the association between chronic, abnormal eye rubbing and the development of KC is well established (10) unlike the association with allergic conjunctivitis which is still debated. Indeed, some authors have suggested that atopy is not significantly associated with KC, but rather with eye rubbing (11) with the exception of VKC which is considered the most frequently associated ocular allergy with KC (12).

In our study, the average age of discovery was 12.8 years, comparable to that of S. Léoni-Mesplié et al (13) (13 years) and lower than those reported by Shahira Mahmoud et al (14) (14.4 years) and Naderan et al (15) (15 years old). The youngest child in our series was 6 years old, similar to that in the series of S. Léoni-Mesplié et al (13). In the literature, the youngest age reported was 4 years. This was a little girl with Down syndrome (16). Younger age of onset of KC tends to be associated with faster progression and more advanced disease stage (2). In fact, although children share most of the common signs and symptoms of adult KC such as blurred vision, distorted images, and double vision, their complaints are often mild. This is explained by the children's inability to express their visual discomfort and by the child's high accommodative power which will compensate for the anterior corneal aberrations induced by the KC (16).

Various classifications based on morphology, disease progression, ocular signs and keratoconus index systems have been proposed. The clinical classification is very uncommon, most probably linked to the different topographers used in the various centers. We found that ¾ of the patients had proven KC. Our results match those reported by Alyaa Saeed Ahmed et al (17) who found 12 proven KC among 16 diagnosed KC. Likewise, with the Amsler-Krumeich classification which is the most widespread classification, we found a significant rate of KC stage 4 (47.4%) exceeding those reported by El-khoury et al (16.7%), Naderan et al (22.8%), Léoni-Mesplié et al (27.8%) and Shahira Mahmoud et al (25%) (13–15, 18). For AS-OCT classification based on the modification of corneal layers, the use is still limited and to our best knowledge, this is the 1st study that classifies pediatric KC by AS-OCT. According to our results, PKC is most often diagnosed at an advanced stage. This can be explained by the delay in consultation and diagnosis of KC and also by the aggressiveness of PKC in our region. In fact, the current majority of authors agree on the fact that young age is associated with more severe forms of KC, with a more rapid progression of ectasia and with increased likelihood of corneal opacities and need for KP (19–23).This aggressiveness of the PKC could be linked to the dynamic environment of the young cornea which presents higher rates of collagen remodeling compared to adults and the phenomenon of ectasia is the result of exceeding the capacity of the reticulation process (24). Kotecha et al (25) suggested a negative correlation between the viscoelastic properties of the cornea and age. Similarly, Blackburn et al (22) found that the cornea becomes stiffer with age in an almost linear manner. On the other hand, the frequent coexistence of ocular pathologies such as atopy and VKC has been associated with faster progression and more frequent complications of PKC (21).

KC is responsible for irregular myopic astigmatism. The near VA is often preserved for a long time. The BCVA of our patients was 0.368±0.37 LogMar, similar to that reported by Léoni-Mesplié et al (0.31±0.05), Naderan et al (0.51±0.20), Padmanabhan et al (0.33±0.22) and Shahira Mahmoud et al (0.79 ± 0.63) (13–15, 26). Keratometric values vary between the different series. This variation could be linked to the difference in the anatomical and biomechanical characteristics of the corneas between different countries. In our study, the average Kmax was 54.6±8 D close to those reported by El khoury et al (18) (53.66 ± 11.12) and Shahira Mahmoud et al (14) (59.18 ± 10.38), with which we share several demographic and environmental similarities. Compared to adults, the Kmax was greater in children with a significant difference, once again demonstrating the severity of the PKC (19, 20).

Biomicroscopic examination of keratoconic corneas is often uninformative in the early stages of the disease, thus leading to a diagnostic delay, which is more significant in children. In more advanced stages, corneal opacities can be individualized and they are directly linked to the importance of the protrusion of the cone (27). In our series, they were objectified in 18.4% of cases reflecting the severity of PKC in our patients. Corneal elevation topography is currently the reference examination for the diagnosis and classification of KC, especially for the subclinical stages (28). It can also provide information on the prognosis of PKC. Indeed, we found that all keratometric indices except SRAX, all elevation parameters and those of pachymetry except TP delocalization are correlated with BCVA thus constituting prognostic factors for KC. Our results agree with those of Schmitt et al who found good correlations between Kmax values, average power in the central 3 mm area, maximum elevation amplitude on the anterior corneal elevation map and minimal corneal pachymetry with BCVA(29).

The biomechanical properties of the pediatric cornea, the inability of children to express their visual symptoms, the asymmetric nature of the disease and the significant accommodative power of the child, lead to that PKC is often diagnosed at an advanced stage. Therefore, it must be suspected in any child presenting with unexplained decrease in VA, recent amblyopia and appearance or evolution of an astigmatism. Screening must be done by corneal topography. There is no age limit for the latter and it must be carried out as soon as the child becomes able to place his chin on the device (30). The study of the epithelial profile using OCT allows even earlier detection of early KC by exploring the masking effect of the epithelium (31).

PKC management must be rapid. Its main objective is to stop progression, prevent visual deterioration and avoid the need for KP. Currently, there is no specific management protocol to PKC. The available therapeutic algorithms are similar to those for adults and depend on the stage of evolution of the KC (32). However, the care of children must be carried out in its entirety. It is imperative to stop all eye rubbing, carry out a pediatric assessment and carry out regular monitoring. The prescription of optical glasses is only possible at an early stage because of the rapid variation in astigmatism. CL remain the primary method of KC correction and are estimated to successfully treat 75% of cases while respecting ocular physiology (33–35). In children, they are indicated as soon as the diagnosis of PKC is made, regardless of age. In fact, it is known that the younger the child, the easier and faster the adaptation will be (36). This adaptation

must be bilateral even if the damage is mainly unilateral or asymmetric. In our series, 57.9% of children were equipped with CL, a rate close to those reported in the SFOALC and CLEK studies (75%) (37,38). RGPL were the main CL prescribed in our study, which is consistent with the literature (20,39,40). However, although they are generally better accepted by the child, their mobility is very often a source of discomfort, loss and abandonment of wearing. SL, without corneal contact and with scleral support, allow better tolerance and better centering of the lens, particularly in advanced cases. On the other hand, they are indicated in the treatment of surface pathologies, frequently associated with KC (34,41). Thus, they are increasingly used in the management of KC. In our series, SL were used in 36.4% of cases, a higher rate than that reported by Shneor et al (42) (4.2%).This is probably due to our dry and dusty climate, a source of intolerance to RGPL and the advanced KC stage of our patients. Surgical intervention is recommended in case of CL failure. Intracorneal rings (ICR) can be proposed only if the transparency of the cornea is preserved and the pachymetry is more than 400 µm at the implantation site (43). Although they appear safe and effective in children with poor VA, ICR are not common practice, especially in cases of associated VKC, because of the aggressive nature of the disease, the tendency to rub the eyes and the risk of migration and extrusion of the eyes, and the absence of nomograms adapted to PKC (43). KP is indicated in the case of failure of other therapeutic modalities and in case of irreversible opacities, preventing any visual recovery. Deep anterior lamellar KP is the technique of choice, penetrating KP is indicated in case of damage to the endothelial layer. The PKC represents 15 to 20% of all child KPs, but the number has decreased considerably with the advent of the CXL (44). The latter is considered the only treatment available to stop the progression of KC. However, no standards have been established so far. In our study, CXL was indicated from the outset, in accordance with the recommendations of the Global Consensus on Keratoconus and Ecstatic Diseases (45). However, Simantov et al (46) after 7 years of follow-up noted only 25% progression of KC in untreated eyes and suggested CXL only in case of progression of KC. In our study, we noted stabilization without CXL in 35.71% after 2 years of follow-up. Other authors have proposed an individualized approach and have considered the systematic performance of CXL in case of severe allergy or very advanced contralateral KC, while reducing the monitoring intervals (47). Several studies have shown the effectiveness of CXL on topographical stabilization and VA improvement (48). By comparing the two groups with and without CXL in our study, we have indeed highlighted the effectiveness of CXL on the evolution of refractive parameters, BCVA and Kmax. Compared to adults, the initial effectiveness of CXL is similar to that of adults in terms of improvement in visual and topographical results but long-term results are more variable (49). However, it should be kept in mind that CXL is not devoid of complications, even if they are rare such as infectious keratitis, corneal haze and temporary decrease in VA (50). Also, close monitoring after CXL in the pediatric group is necessary to detect the first signs of return of progression.

Conclusion

PKC has many unique clinical features. It is often associated with allergic conjunctivitis and frequent eye rubbing. Its association with a systemic pathology is also possible requiring a pediatric examination and multidisciplinary care. Screening for KC in the family must also be recommended. PKC at the time of diagnosis is more severe than that of adults with more frequent and rapid progression. Thus, early screening using corneal topography must be systematic for any significant and/or progressive astigmatism and for any unexplained decrease in VA in a child. CXL appears effective but the stabilization rate seems to be worse than in adults, suggesting a personalized approach.

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