S Afr Optom 2013 **72**(1) 41-45

Keratoconus: a clinical exposé

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Received 28 October 2012; revised version accepted 26 February 2013

Introduction

Keratoconus is (or should be), clinically, a well known condition of the cornea. Cline *et al*[1] define keratoconus as: "a developmental or dystrophic deformity of the cornea in which it becomes cone-shaped due to a thinning and stretching of the tissue in its central area. It usually manifests itself during puberty, is usually bilateral and is more common in women than men". A clinical description of keratoconus might include the following: a non-inflammatory, bilateral, asymmetric progressive ectasia of the cornea associated with thinning, protrusion and distortion of the cornea[2-4]. Clinical signs of keratoconus include: Vogt's striae, Fleischer's ring, corneal thinning, corneal scarring, increased visibility of corneal nerves, characteristic topographical changes, protrusion and rupture/folding in the area of Descemet's membrane[5-7]. Keratoconus has a prevalence of between 50 and 230 people per 100 000 of the population depending on the criteria used to diagnose the condition[8].

Aspects of keratoconus that are perhaps not clear are the clinical characteristics of the condition that are common to a particular keratoconic population of patients. For instance: what is the prevalence of Vogt's striae in that clinic population? In how many patients is it not possible to get any reliable measurements of refractive state, keratometric or topographical data? How many patients present with corneal scarring? What are the locations of the cones? The answers to the above questions are not obvious. The aim of this investigation was to conduct a purely clinical appraisal of the prevalence of the characteristic signs and some symptoms that occur in a population of keratoconic patients attending a particular contact lens clinic in the Johannesburg area.

Method

It was decided to record a number of characteristic clinical findings that were noted during the initial routine clinical examination of keratoconic patients attending a contact lens clinic at the University of Johannesburg. Observations were recorded as and when I remembered to do so and/or when time allowed for the noting of the findings. In many instances the presence/absence of a particular sign was a clinical decision made during the slit lamp examination of each patient with no attempt being made to verify the decision by other means (for example: corneal thinning which could have been verified by Pentacam measurements, however, in many instances such measurements were not available).The clinical observations that were noted from each patient were the following: the location of the cone, whether scarring was present, increased nerve visibility, the presence/absence of Fleischer's ring or Vogt's striae, whether there was a history of rubbing of the eyes, history of atopy, family history of keratoconus, presence/absence of obvious (using a slit lamp) corneal thinning and whether refractive, keratometric and topographic measurements were possible. Both eyes of each patient were examined and if, for example, Vogt's striae were present in only one eye then the presence of Vogt's striae was recorded for that patient. Over a period of approximately 18 months the clinical observations for 25 keratoconic patients were recorded. What follows is a description of how decisions were made regarding the various aspects of keratoconus that were recorded in this study.

Cone location

McMahon[9] has stated that the location of the cone

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in a keratoconic cornea can be anywhere, with 10-20% of cones being above the horizontal meridian of the cornea. In this investigation the location of the cone was determined by assessing corneal topography obtained via a Pentacam topographer. In instances where measurements of the cornea could not be obtained with the Pentacam, flourescein patterns or the slit lamp were used to determine the location of the cone. Figure 1 shows a Pentacam image where the cone is located inferiorly for instance. Cone location was determined for both eyes of each patient.

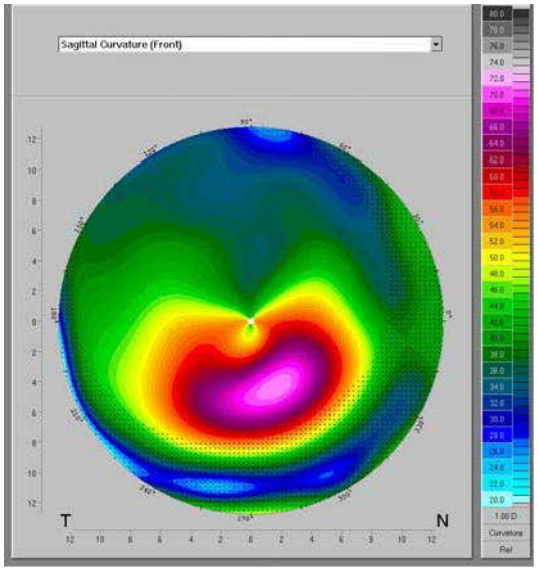

Figure 1. A Pentacam image is shown indicating an inferiorly located cone.

Alternatively Figure 2 shows a Pentacam image indicating a superiorly located cone and in Figure 3 a centrally located cone is indicated.

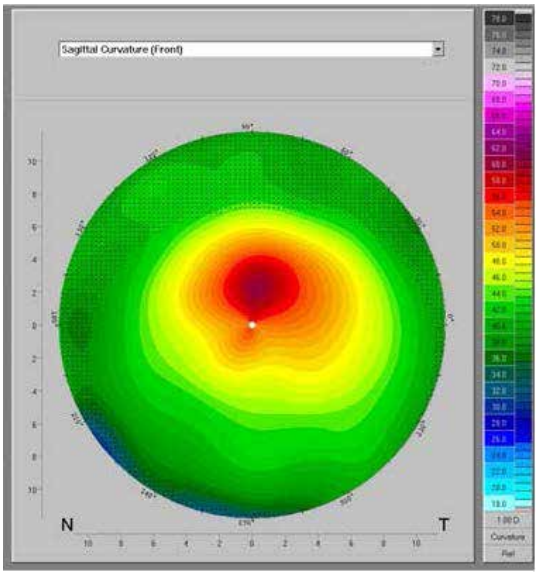

Figure 2. A Pentacam image showing a superiorly located cone.

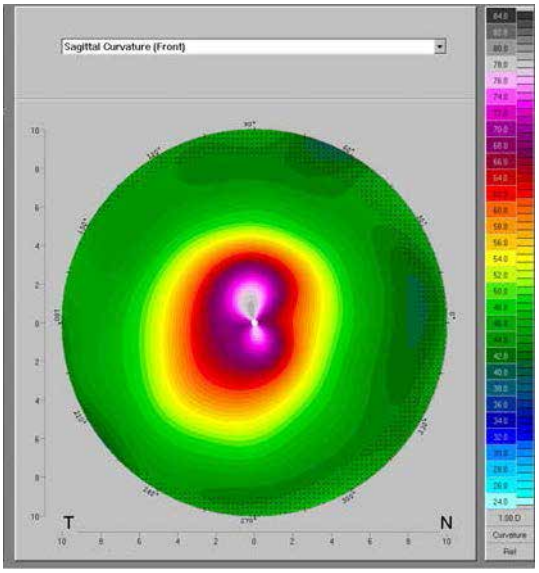

Figure 3. A central cone is suggested in this Pentacam image.

Scarring

The presence of corneal scarring was assessed during the routine slit lamp examination of the patient. Figures 4 and 5 show slit lamp photographs of examples of scarring that were observed in patients attending the clinic. Scarring was recorded even if only one eye was observed to have scarring.

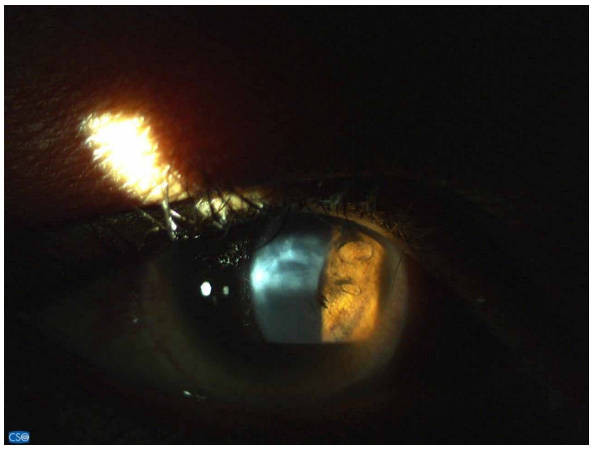

Figure 4. An example of scarring associated with keratoconus

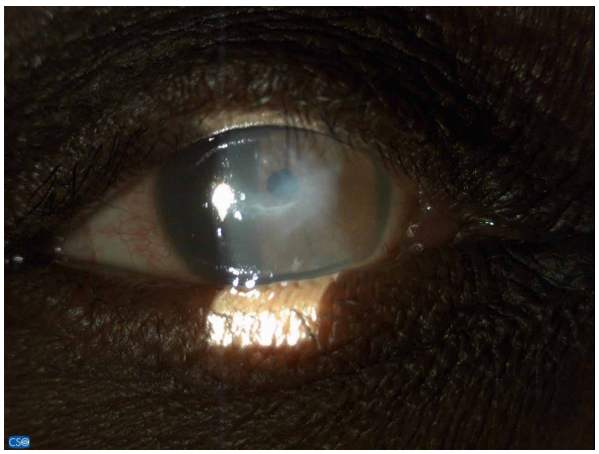

Figure 5. A second example of keratoconus-related scarring.

Nerve visibility, Fleischer's ring and Vogt's striae

Corneal nerves are more visible in patients presenting with keratoconus[10]. The assessment of this increased visibility, the presence of Fleischer's ring and/or Vogt's striae, in this study, was purely objective and was conducted during the routine examination of the patient. Figure 6 shows a well developed and obvious Fleischer's ring, however, this sign of keratoconus is not always so easy to observe. Vogt's striae are ubiquitous in keratoconic patients and two good examples of such are shown in Figure 7. If only one eye had increased nerve visibility, Fleischer's ring or Vogt's striae, then that patient was recorded as having that characteristic.

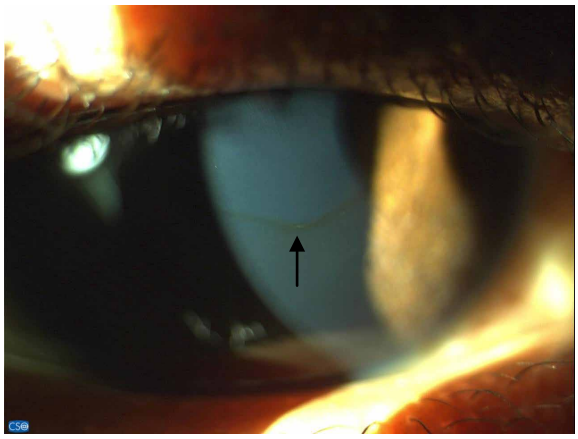

Figure 6. An obvious Fleischer's ring indicated by the arrow.

Rubbing, atopy and family history

Information relating to the presence of atopy and whether the patient had family members who were keratoconic or whether the patient had/does rub their eyes excessively was determined during the routine case history.

Corneal thinning

The slit lamp was used to determine if clinically obvious thinning of the cornea was present. Figure 8 shows a cornea where thinning is obvious. Thinning was recorded for a patient if only one eye was noted to have thinning.

Refractive measures

Retinoscopy, subjective refraction, keratometry and Pentacam topographical measurements were attempted in all patients. In many patients it was not possible to obtained useful measurements. In many instances keratometry was not possible due to severe distortion of the mires and an inability to align them. All too often accurate measurements were not possible as the scale was not extensive enough to measure the curvature of the cornea (measurements greater than 52 D). The Pentacam is designed to automatically trigger when the instrument has been aligned correctly. In numerous patients the cornea was too distorted for the Pentacam to trigger automatically. Manual triggering is possible but this usually resulted in inaccurate measurements. If refractive measurements were not possible in one eye then that patient was recorded as not being able to have refractive measurements determined.

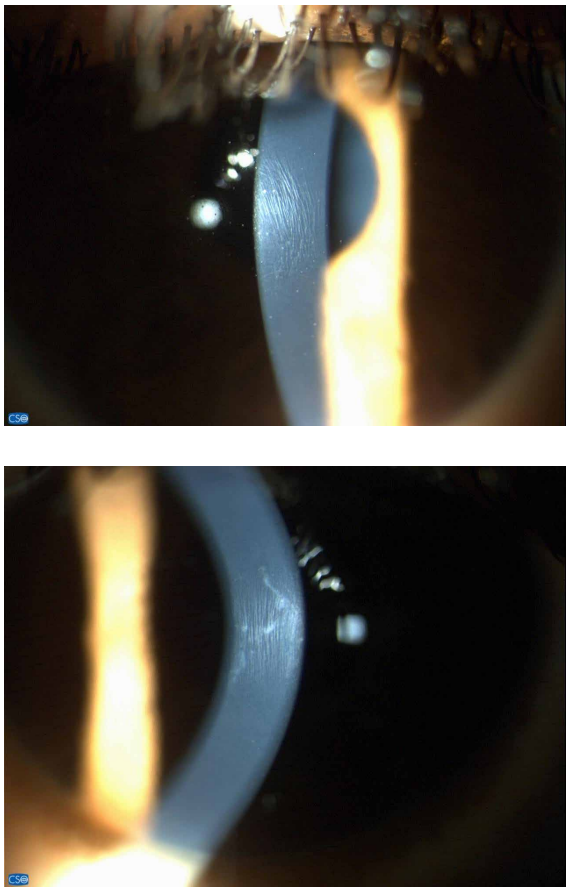

Figure 7. Examples of Vogt's striae.

Figure 8. An example of superior thinning observable using a slit lamp.

Table 1. Cone location for 49 eyes is presented. One subject only had one eye. In two patients the location of the cone could not be decided upon due to the distorted state of the corneas.

Location % (*n*)	**Inferior**	**Superior**	**Central**	**Temporal**	**Nasal**	**Unknown**
	20.4 (10)	14.3 (7)	53.1 (26)	4.1 (2)	0	8.2 (4)

Table 2. The number of patients presenting with the relevant characteristic is shown here. The total number of patients is 25. The following characteristics are noted: scarring, increased visibility of corneal nerves (NV), Fleischer's ring (FR), Vogt's striae (VS), rubbing, atopy, obvious corneal thinning and refractive measures (RM, patients who could not produce any refractive measures). Data are given as percentage and number of patients (*n*).

Scarring	NV	FR	VS	Rubbing	Atopy	Corenal Thinning	RM
56 (14)	28 (7)	68 (17)	48 (12)	76 (19)	48 (12)	52 (13)	56 (14)

Results

A total of 49 eyes were assessed for cone location (one patient only had one eye). Table 1 shows the data for the cone location of 49 eyes.

The results of other characteristics noted in this study are presented in Table 2. The number of patients presenting with each characteristic (even if only present in one eye) is recorded here (out of a total of 25 patients).

Each patient was asked whether they knew of a history of keratoconus in their family. Three patients out of the 25 in this study answered in the affirmative with the other 22 patients not knowing or stating that no family history of keratoconus was present.

Discussion

A clinical, mostly objective appraisal of characteristic signs and symptoms of keratoconus is presented in this article. It is very possible that a more rigorous methodology, with more stringent criteria being used to categorize observations would result in different findings. It is also possible that different results would be determined if another clinic population from another part of the country was involved in the investigation. However, the reason for this investigation was to present some information that might be applicable to clinicians, making clinical decisions, who are involved in the examination of keratoconic patients. There is a paucity of epidemiological information (as far as I know) relating to the presence of keratoconus in South Africa. It is not clear what percentage of the population has keratoconus, what the levels of severity of the disease are, what the presenting signs and symptoms of different aged patients might be and so on.

As McMahon[9] has stated, the cone of a keratoconic cornea can be found in any location on the cornea. In this study most keratoconic eyes had cones considered to be centrally located (53.1%). About 1 in 5 or 20.4% of cones were located in an inferior position (see Table 1). In a study conducted by Uçakhan *et al*[10] they found Fleischer's ring in 17% of eyes, prominent nerves in 9% of eyes, Vogt's striae in 13% and stromal scarring in 5% of eyes. These data were determined from a group of 24 keratoconic patients in Turkey. The results of the present study can be seen (and compared with Uçakhan *et al*[10]) in Table 2 (remembering that data shown in Table 2 is patient number and not number of eyes). An interesting clinical observation in the population of keratoconics being presented here was the severity of keratoconus when diagnosed in young children (as young as seven years old). These young patients are presenting with severe scarring, thinning and general progression of the keratoconus condition. This finding was also determined by Léoni-Mesplié *et al*[11] when investigating keratoconus in children in Bordeaux, France. Rubbing of the eyes has been implicated in the development of keratoconus[12]. In this study 76% of the 25 patients admitted to rubbing of the eyes, either presently or at some earlier stage.

No refractive information was obtainable in 56% of patients. Retinoscopy, subjective refraction, keratometry and Pentacam topography gave no useful information (other than a general indication of distortion of the cornea). In these instances initial trial lens fitting began at what was essentially a "guesstimate" of lens type and base-curve. Further investigations of keratoconus in South Africa are needed. We need to know how many people have the disease, what role genetics plays in the manifestation of the disease, how severe is the keratoconus when

patients present for care and so on so that eye care professionals have a better idea of the characteristics of keratoconus in the South African population.

In summary, information relating to 25 keratoconic patients, attending a contact lens clinic in the Johannesburg area, is presented. The prevalence of some characteristic signs and symptoms is given.

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