ORIGINAL ARTICLE

Prevalence and clinical profile of keratoconus in patients presenting at a provincial hospital in KwaZulu, Natal. South Africa: A case study

NONKULULEKO M. GCABASHE, VANESSA RAQUEL MOODLEY and REKHA HANSRAJ

Discipline of Optometry, School of Health Science, University of KwaZulu-Natal, Durban, South Africa

DOI: 10.4081/jphia.2023.2356

Abstract. Keratoconus (KC) is a progressive, asymmetrical 1 2 corneal disease, characterized by stromal thinning that leads 3 to distortion, causing vision loss. The visual loss is secondary 4 to corneal scarring, irregular astigmatism, and myopia. The prevalence of KC has been reported to differ in different parts 5 of the world. The study aimed to determine the prevalence and 6 7 profile of patients with KC presenting to a provincial hospital in KwaZulu-Natal, South Africa. A retrospective study design 8 9 was used to review 412 clinical records of patients attending the 10 McCord Provincial Eye Hospital (MPEH) during a five-year 11 period (2016-2020). Data on age, race, refraction, clinical 12 profile, treatment plan, and diagnosis were ascertained. The prevalence of KC in MPEH was found to be 13.7% with a 13 14 mean age of 24.7±7.94 years. Black African and females had a higher frequency of KC compared to males and other ethnic 15 groups. Most of the patients presented with a severe stage of 16 17 KC and referral was the most common management. Central 18 corneal thinning and Munson's sign were the most prevalent 19 clinical signs. There was no statistically significant difference 20 between the worse and better eye when comparing the clinical signs. The prevalence and clinical profile of patients with KC 21 22 in this study was similar to that reported by previous studies 23 and more in Blacks and females. Population based epidemiological studies are needed to determine the prevalence of KC 24 25 in South Africa to enable early clinical interventions. 26

27 Introduction

28

29 Keratoconus (KC) is a progressive, non-inflammatory 30 thinning of the cornea associated with myopia and irregular

Correspondence to: Nonkululeko M. Gcabashe, Discipline of Optometry, School of Health Science, University of KwaZulu-Natal, Durban, South Africa E-mail: thwala@ukzn.ac.za

Key words: keratoconus, vernal keratoconjunctivitis, prevalence, central corneal thinning, corneal scarring

astigmatism. Patients experience a decrease in the quality of 31 vision due to monocular diplopia, halos or ghost images (1). 32 It is the most common form of corneal dystrophy, which 33 may lead to severe visual impairment if left untreated (2). In 34 the absence of a single identifiable cause, ongoing research 35 into the aetiology of KC has revealed complex interactions 36 between genetic, environmental and hormonal factors (3,4). 37 Risk factors for KC already identified include demographics, 38 ethnicity, genetics and the environment (5). Ecological factors 39 associated with KC include, among others, eye rubbing, 40 atopy and exposure to ultraviolet radiation (5). While KC has 41 been noted in populations worldwide, it is more frequently 42 reported in certain ethnic groups such as South Asians, East 43 Mediterranean's and North Africans (6,7). 44

The incidence and prevalence of KC vary worldwide; 45 with its diagnosis being made using a variety of methods 46 such as retinoscopy, slit-lamp biomicroscopy, pachymetry, 47 keratometry and video keratography, topography and tomog-48 raphy (8,9). The prevalence of KC was reported to be 54.5 per 49 100 000 population in the United States of America, while the 50 prevalence of KC in a hospital-based study in Denmark in 2007 51 reported an estimated prevalence of 86 patients per 100 000 52 residents with an annual incidence of 1.3 per 100 000 (10,11). 53 However, a similar study, conducted in the same hospital in 54 Denmark in 2019, reported a prevalence of 44 per 100 000 55 and an increase in the incidence rate of 203 folds during the 56 last 10-15 years (11,12). A study done in Jerusalem reported a 57 prevalence of 2.34% and a significantly higher prevalence in 58 men (4.91%, CI 2.6-7.3) than women (1.07%, CI 0.3-1.9) (13). 59 An African hospital-based study found a prevalence of 10.6% 60 by clinical diagnosis, 14.6% by keratometry and 30.9% by 61 topography in patients with allergic conjunctivitis attending 62 Kenyatta National Hospital (14). 63

The onset of KC is usually at puberty and progresses 64 until 40-50 years of age. However, due to the self-limiting 65 nature of the disease, this progression may stop at any stage 66 between mild to severe KC (13-15). Although the disease 67 is bilateral, it presents asymmetrically in line with studies 68 reporting that the disease starts unilaterally, with delayed 69 70 onset in the other eye (16,17). Ocular signs and symptoms vary with the severity of KC (18). Vinciguerra et al (2016) 71 noted that abnormal biomechanical changes are observed 72

at an early stage before tomographic changes and conical 1 2 signs (19). The early signs of KC may go unnoticed, with 3 symptoms varying from increased sensitivity to light and 4 glare to distorted vision (19). In moderate cases, Fleischer's 5 ring is seen around the base of the cone together with Vogt's 6 striae which are vertical, fine, whitish lines in the deep/poste-7 rior stroma (20). In advanced stages, Munson's sign, Rizzuti's 8 sign and corneal hydrops and or corneal scarring, which 9 develop as a result of a split in the Descemet's membrane, 10 may be observed (20,21).

The severity of KC has been classified by previous studies 11 12 using either the Amsler-Krumeich (AKC), ABCD, Keratoconus 13 Severity Score (KSS) or Collaborative Longitudinal Evaluation 14 of Keratoconus (CLEK) classification systems (22-25). AKC is 15 based on the mean corneal power, astigmatism, transparency and thinnest corneal thickness while the CLEK classification 16 17 is founded on the change in visual acuities, mean keratometry, 18 slit-lamp biomicroscopic signs, presence or absence of corneal scarring, and visual-related quality of life (22). The ABCD 19 20 keratoconus staging system incorporates anterior and posterior 21 curvature centered on the thinnest point of the cornea, thinnest 22 pachymetry values and distance visual acuity (24).

23 The management of KC is dependent on the severity of 24 the condition, with the first treatment option for reduced 25 visual acuity (VA) generally being spectacles. For mild or 26 moderate KC, soft-toric and custom soft spherical or toric 27 contact lenses can be used. Moderate to severe KC requires the use of corneal and scleral rigid gas permeable (RGP) 28 29 lenses (26). Contact lenses often provide better vision than 30 spectacles by masking higher-order aberrations due to 31 irregular astigmatism (15). When vision can no longer be 32 corrected with optical corrections such as spectacles and 33 contact lenses, corneal surgery is then recommended (27). 34 The surgical management of KC includes intrastromal rings, 35 corneal intrastromal corneal ring segments (INTACS) and 36 keratoplasty, amongst others. Corneal cross-linking (CXL) 37 is a minimally invasive surgical procedure used to stabilize 38 the progression of KC (28). Studies have shown that CXL 39 is effective in halting the progression of keratoconus over a 40 couple of years (28,29).

41 Keratoconus is a significant cause of severe visual impair-42 ment (VI) worldwide, if undiagnosed and left untreated. 43 Despite this observation, it has received little attention in 44 terms of public health efforts to address the health care needs 45 of affected persons, particularly in Africa, including South 46 Africa. The estimation of prevalence is important in estab-47 lishing strategies and programs geared towards the prevention of VI caused by an ocular condition that can be treated (30). 48 Even though a few studies on KC have been conducted on the 49 50 African continent, no previous study has been conducted in 51 a hospital in South Africa, nor at the site of the current study 52 to determine prevalence of KC (14,31,32). Anecdotal evidence 53 suggests that there has been an increase in the number of refer-54 rals of patients with KC to the MPEH, currently dedicated as 55 the only public eye hospital in the province of KZN, South Africa. Therefore, this study aimed at identifying the preva-56 57 lence and clinical profile of patients with KC presenting at 58 MPEH to better understand this corneal ectasia towards an 59 improvement in the diagnosis and management of presenting 60 KC patients.

Materials and methods

This quantitative study involved a retrospective review of 63 clinical records that were randomly selected to obtain infor-64 mation on the prevalence, demographic and clinical profiles, 65 and management of patients with KC at MPEH. The study 66 67 commenced after ethical clearance and relevant gatekeeper permissions were obtained from the Biomedical Research 68 Ethics Committee (BE332/19), Department of Health in KZN, 69 and MPEH, respectively. As MPEH is the only public eye 70 hospital situated in the eThekwini district that offers eye care 71 services at a provincial level, patients seen at this hospital are 72 73 referred from other health care providers. A random sampling method was used to determine the minimum sample size 74 of 391. Data on the demographic, clinical presentation and 75 management strategy for keratoconic patients seen at MPEH 76 over five years were extracted and analyzed using descriptive 77 and inferential statistics with the Statistical Package for Social 78 Sciences version 25. 79 80

Results

83 Demographics. A random sample of 412 patient clinical records from 2016 to 2020 were reviewed, however, 70 of the 84 clinical records did not have the diagnosis for the respective 85 patients recorded and were excluded. The prevalence of KC 86 was therefore calculated based on the remaining 342 clinical 87 records that had a diagnosis recorded. Of these, 47 were 88 noted to have KC indicating the facility prevalence of KC of 89 13.7%. The mean age of patients with KC was 24.7±7.94 years 90 91 (median age of 24.0 years) with a higher percentage (63.8%)being female. There were more Black African patients with 92 93 KC (66%) compared to Indians (29.8%) with no Caucasians or patients of mixed race, and a further 4.2% for whom race 94 count not be classified. 95

97 Clinical data. The most common ocular disease and medical condition noted in patients with KC was vernal keratoconjunc-98 tivitis (VKC) (53%) and sinusitis (6.4%), respectively. Marfans 99 syndrome was noted for 2.1% of keratoconic patients with no 100 other underlying systemic condition recorded. While refrac-101 tive status was not noted in 53.2% of the record cards for the 102 keratoconic patients, based on those cards in which refractive 103 findings and best-corrected visual acuity (VA) were recorded 104 (n=22), 19.1% had no VI, 12.8% had mild VI, 12.8% had 105 moderate VI and 2.1% severe VI. 106 107

Keratoconus profile. All of the keratoconic patients had 108 bilateral KC and the most common clinical sign that was 109 noted was apical scarring (n=12) and central corneal thinning 110 (413 \pm 80.1 μ m) (n=11). Fig. 1 illustrates the clinical signs noted 111 in the keratoconic patients. These values are derived from only 112 42 of the 47 record cards for KC patients, as in the remaining 113 five cards, there was no recording of the absence or presence 114 of the clinical signs. 115

Table I outlines a comparison of the frequency of clinical 116 signs being present in the better eye and the worse eye of the 117 keratoconic patients. Only hydrops was found to be present in 118 a significant number of better eyes than worse eyes (P=0.007). 119 Furthermore, no significant difference was noted in the visual 120

81

82

1 Table I. Frequency of clinical signs present in the better and 2 worse eye.

| | Better eye (%) | Worse eye (%) |
|-------------------------|-------------------|------------------|
| Munson's Sign (n=38) | 21.05 | 21.05 |
| Vogt's Striae (n=38) | 10.5 | 10.5 |
| Fleischer's Ring (n=38) | 7.89 | 7.89 |
| Hydrops (n=38) | 10.5 | 10.5 |
| Corneal Thinning (n=38) | 21.05 | 21.05 |
| Apical Scarring (n=36) | 22.2 | 27.78 |

acuities of the better eye in patients that presented with clinicalsigns.

Table II shows the mean and standard deviation for 18 corneal curvature (K), central corneal thickness (CCT), and 19 20 the nearest equivalent sphere (NES RX) of the left and right 21 eyes of patients that have KC. The NES range for right and left 22 eves was -18.75 DS to +0.75DS, with corneal curvature range 23 of 40.25 to 73.30 D and that for corneal thickness of 217 to 24 516 μ m. The better eves had mean visual acuity (in decimal 25 notation) of 0.449±0.253 respectively while the worse eyes had mean visual acuity mean of 0.160±0.158 with a range of 0.01 26 27 to 0.63.

The severity of KC was graded using the AKC. As shown 28 29 in Fig. 2, most of the keratoconic patients (35.5%) had Stage 3 30 KC, followed by Stage 1 (25.8%), Stage 2 (22.6%), and lastly 31 Stage 4 (16.1%). In terms of the CLEK, most patients had 32 severe KC (66.7%), followed by moderate (30.3%), and then mild (3%). Chi-squared analysis was run to determine if there 33 34 was any association between the clinical signs and stages of 35 KC as per CLEK and AKC and there was no significant relationship noted. 36

The most common treatment plans used for patients with KC at MPEH, amongst other management options as shown in Fig. 3, was referral only (36.2%) followed by monitoring the condition only (14.9%).

42 Discussion

43

41

14

15

The prevalence of KC was found to be 13.7% (n=47) which is 44 45 similar to the prevalence of KC using clinical diagnoses (10.6%) and keratometry (14.6%) reported in Kenyatta hospital, Kenya 46 in patients with allergic conjunctivitis (14). Although both 47 studies were conducted in Africa and were hospital-based, the 48 participants in the study by Mugho (2016) were considerably 49 50 younger (mean of 14.9 years vs. 24.7 years in the current study) 51 with all participants diagnosed with allergic conjunctivitis (14). 52 Lower prevalence has been reported elsewhere. Nielsen et al in 53 2007, found a prevalence of 86 per 100 000 in Denmark with 54 more recent studies revealing the prevalence of KC in middle 55 eastern countries including Iran, Saudi Arabia, and Israel to vary from 3.18 to 4.91% (11,33-35). While Netto et al (2018) 56 57 also calculated the prevalence of KC in Saudi Arabia to be 58 4.79% using a retrospective chart review, it was based on a 59 pediatric sample with a mean age of 16.8 ± 4.2 years (35). The 60 study by Shneor et al (2014) determined the prevalence of KC in Israel by analyzing the video keratographic indices and 61 images to confirm if the students with a mean age of 25.08±8.83 62 had KC (34). Millodot et al (2011), in a cross-sectional study, 63 found that the prevalence amongst the 981 volunteers (mean 64 age 24.4 years) was higher (2.3%) in Jerusalem as compared 65 to Western Countries such as the Republic of Macedonia with 66 a mean age of 26.81 years ±1.25 (6.8:100 000) and Norway 67 (192.1 per 100 000) (13,36,37). The comparatively higher 68 prevalence of KC reported by African studies could be because 69 many of those studies were hospital-based while the European 70 studies were mainly population-based and may also be associ-71 ated with environmental factors and genetics. Many studies 72 73 have also suggested links between allergy, atopy, or asthma and KC (13,38). A study by Lucas and Burdon (2020) stated 74 that genetics play a major role in some individuals showing a 75 pattern of autosomal inheritance, with eye rubbing also impli-76 cated (39). Furthermore, the higher prevalence of KC found in 77 the current study could be related to the capacity at the study 78 site; as MPEH serves as a referral hospital in KwaZulu-Natal 79 due to the facility having all the necessary equipment for accu-80 rate and early diagnosis of KC. This may result in a greater 81 likelihood of referrals from lower-level facilities and increased 82 attendance by patients with KC at MPEH. Variations in preva-83 lence of KC previously reported in studies worldwide may 84 thus be attributed to differences in research designs, such as 85 86 population-based vs. hospital-based, differences in methods used to investigate and diagnose the condition, demographics 87 of the participants, as well as the utilization of varying KC 88 classification criteria. 89

Keratoconus starts as a unilateral corneal ectasia 90 91 however, it progresses to the fellow eye within the first five to six years after the onset (3). In the present study, all the 92 keratoconic participants presented with bilateral KC which 93 was similar to the 98.3% of bilateral keratoconics reported 94 by Rashid et al (2016) in a study in Africa (40). In addition, 95 a clinic-based study conducted by Rupnarain et al (2020), 96 in KZN, South Africa also found that most (71.3%) of 97 their participants had bilateral KC (31). Outside of Africa, 98 Naderan et al (2015) reported bilateral KC in all their partici-99 pants, in Iran. Other studies, in similar low-to-middle-income 100 countries, have also shown that bilateral KC prevalence ranges 101 between 56 to 93% (41,42). A possible reason for a predomi- 102 nance of bilateral KC could be that perhaps in the absence 103 of KC advocacy programmes and limited resources, patients 104 only seek medical help when vision in both eyes is affected 105 and they can no longer function adequately. Early signs of KC 106 are not easily visible without the use of equipment such as 107 tomographers and topographers. Many of the referral centers 108 may not have the necessary diagnostic equipment and, in the 109 current study the instruments used to diagnose KC was not 110 documented and no images (tomography/topography) were 111 attached to the participants' records. It is likely therefore 112 that some early keratoconic participants, who may also have 113 had unilateral KC, might have been misdiagnosed or KC not 114 identified. 115

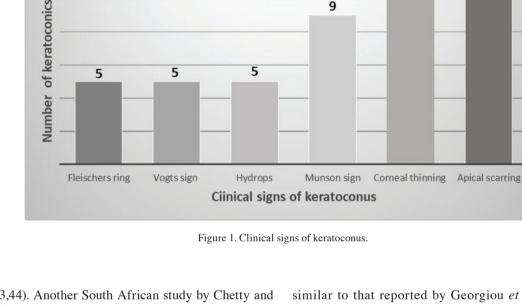
In the current study, there were twice as many female 116 (66.7%) than male (33.3%) keratoconics. Several studies 117 revealed that, on average, females generally frequent primary 118 health care facilities than males which could be the reason why 119 higher number of females than males having been referred to 120

| | Laterality | Minimum | Maximum | Mean | Std. Deviation |
|-----------------------------|------------|---------|---------|--------|----------------|
| | Right eye | -17.00 | +0.75 | -5.63 | 5.01 |
| | Left eye | -18.75 | +1.00 | -5.87 | 5.15 |
| | Right eye | 40.75 | 72.50 | 55.97 | 7.79 |
| | Left eye | 40.25 | 73.30 | 54.99 | 9.36 |
| Corneal thickness (μm) | Right eye | 217 | 513.00 | 414.17 | 75.03 |
| | Left eye | 242 | 516.00 | 397.23 | 72.28 |
| | - | | | | |
| | | | | | |

9

Table II. Corneal radius power (K), corneal thickness (CCT) and refractive error (NES).





this site (43,44). Another South African study by Chetty and 38 Rubin (2019) reported similar results of a higher prevalence of KC in females (53%) (32). Valdez-Garcia et al (2014), in a 39 40 study amongst Mexican adolescents, found a higher prevalence in females (66.7%) compared to males (33.3%), which was the 41 42 same ratio as noted in the current study (45). In contrast, other 43 studies have reported a higher prevalence of KC in males than in females (1,4). The study conducted by Godefrooji et al (2017) 44 45 reported 72% of their keratoconic sample being male compared to female, with Millodot et al (2011) reporting a 46 slightly higher prevalence of 4.91% in men compared to in 47 women (1.07%) (4,13). Although KC has been mentioned to 48 49 affect both genders, studies have reported different prevalence 50 when comparing the genders. The differences in prevalence 51 of KC between genders may be attributed to demographics, biological and anatomical factors unique to different regions, 52 53 as well as physiological factors such as pregnancy and the 54 menstrual cycle which could also lead to variability, and 55 thus further investigation into gender predilection may be warranted (46). 56

57 Ethnicity has been noted as an important risk factor 58 in KC. A study by Pearson et al (2000), found a higher prevalence of KC in Asians of 229 per 100 000 compared 59 60 to 57 per 100 000 in the Caucasian population, which is similar to that reported by Georgiou et al (2004) (47,48). 97 Both studies concluded that the high prevalence could be 98 due to ethnic differences (47,49). Of the total number of 99 keratoconus identified in this study, there were only Black 100 Africans and Indians with a higher percentage being in Black 101 Africans (66%). The vulnerability of the Black race to KC 102 was also noted by Tuft et al (1994) during an investigation 103 of the prognostic factors for the progression of KC in a study 104 sample which constituted 79.0% White, 15.7% Asian, and 105 5.3% Black subjects (49). Their study revealed that racial 106 grouping had a significant effect on the time from diagnosis 107 to requiring penetrating keratoplasty (PK) and that Black 108 patients progressed to PK at a greater rate than either Asians 109 or Whites (P<0.001) (49). The findings of the current study 110 are similar to that reported by Chetty and Rubin (2019) in 111 their university-based clinic study conducted in another 112 province in South Africa (Johannesburg). The majority (74%) 113 of the keratoconics were Black Africans when compared 114 to other ethnic groups such as Indians (12%), Caucasians 115 (9%), and Mixed race (5%) (32). The high number of Black 116 patients with KC in these two studies could be attributed to 117 the fact that the majority of South Africans are Black (81%) 118 compared to other races and thus their predominance at the 119 chosen sites. It was also noted in the current study that the 120

12

11

61

73

74 75

76

77 78

79

80

81

82

83

84

85

86

87 88

89

90

91

92

93

94

95 96

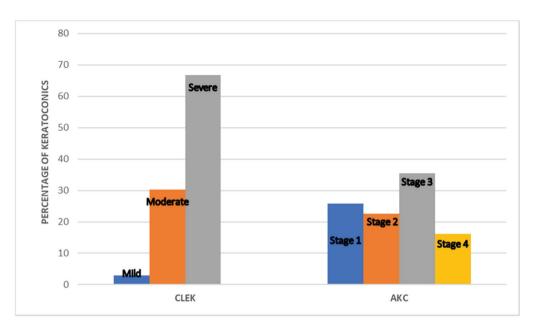


Figure 2. Comparison of stage of keratoconus based on the Amsler-Krumeich (AKC) and CLEK classification systems.

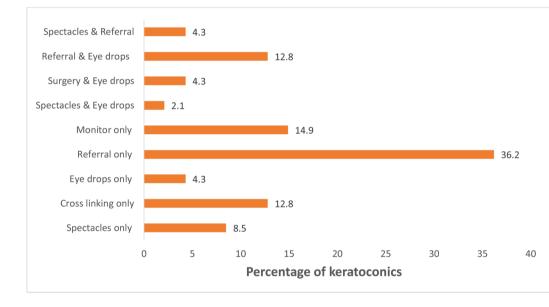


Figure 3. Treatment options considered for patients with KC at MPEH.

clinical records, even though randomly selected, were mainly of Black (72.8%) participants followed by Indians (14.6%), Mixed race (3.3%) and Whites (1.7%). Moreover, both the study by Chetty and Rubin (2019) and the current study were conducted at public health care facilities. The 2018 general household survey (GHS) in South Africa highlighted that only 9.9% of Black African households had access to medical insurance thereby limiting access to private health care by this racial group and promoting the seeking of health care in the public sector which is where MPEH currently lies. Similarly, the survey noted that 72.9% of the White popu-lation (72.9%) had access to health care privately, through health insurance, and thus they do not frequent public health facilities; which may account for the low percentage of that racial group attending the site of the current study (50).

The mean age of the patients with KC in this study 106 was 24.7 ± 7.94 years, which is similar to that reported by 107 Weed *et al* (2008) of a mean age of 25.05 ± 8.97 years in a 108 keratoconic population in Scotland (3). The mean age of the 109 current study is, however higher than that found in other 110 African countries namely Kenya and Sudan (40,51). Two 111 clinic-based studies in South Africa reported that the mean 112 age of patients that visit their KC clinic were 25.2 ± 9.9 years 113 and 26.1 ± 7.5 years, respectively, which is also similar to the 114 mean age noted in the current study. Studies in Asia have also 115 reported similar age profiles whereby the mean age of the 116 patients was 21.46 ± 6.17 years in Malaysia and 29.5 ± 9.40 years 117 in Singapore (43,44). The age profile of keratoconics generally 118 being in their twenties could be related to the finding that even 119 though KC tends to progress until the third or fourth decade of 120

life, patients with KC appear to only present for medical care 1 in hospitals or clinics in their second decade of life (39). In the 2 3 current study, the keratoconic patients were categorized into 4 four age groups with the finding of the majority (76.3%) being 5 in the 19-34 year age group. A study by Millodot et al (2016) 6 reported that the onset of KC in their participants was at the 7 age of 18.4 ± 3.8 years (52). In the current study the onset of KC was not investigated, however, this aspect may be complicated 8 9 by keratoconics possibly presenting to another health facility 10 before subsequent referral to MPEH.

Most of the keratoconic patients in the current study were 11 12 classified with Stage 3 (35.3%), or severe KC (66.7%) based 13 on the Amsler-Krumeich and CLEK classifications, respec-14 tively. Previous studies have reported a high percentage of patients presenting with severe KC based on the CLEK clas-15 sification to specialized evecare clinics or hospitals (52,53). 16 Serdarogullari et al (2013) using Pentacam-derived param-17 18 eters reported that the mean corneal curvature of patients with KC was 57.7±9.0 D, which is similar to this finding in 19 20 the current study (52). Mahadevan et al (2009) revealed that 21 most KC patients presenting to a tertiary eye care hospital 22 had advanced KC with corneal curvatures of greater than 23 52D (53). Similarly, a university clinic-based study in Durban, 24 SA found the mean corneal curvature for patients with KC 25 to be 54.16±7.65D. A possible reason why the patients may 26 be seeking medical intervention in the advanced or in the 27 later stages of KC also noted in the current study, could be that spectacles no longer provide adequate functional vision 28 29 at that stage. The avoidance or the delay in seeking medical 30 help by patients could also be due to factors such as the 31 fear of the diagnosis, denial of the diagnosis and financial 32 constraints (54,55). Access to cost-effective treatment may 33 also be a factor as highlighted in a study conducted within the 34 same district by Maake and Moodley (2018) (56). The authors 35 stated that financial constraints was one of the reasons that the patients did not seek medical care. 36

37 Keratoconus causes thinning and protrusion of the cornea and the current study revealed Munson's sign and 38 39 central corneal thinning, with a mean corneal thickness of 40 $413\pm80.1 \ \mu\text{m}$, to be the most noted clinical signs. Munson's 41 sign was mostly reported in keratoconics in the severe stage 42 (33.3%) and Stage 3 (45.5%), while the central corneal 43 thinning was more frequently noted in moderate stage 2 (66.7%) and Stage 3 (45.5%) according to the CLEK and 44 45 Amsler Krumeich classification, respectively. This finding further highlights the late-stage presentation of the disease 46 47 by patients, which potentially enhances the treatment challenges and costs to the facility. Similar percentages 48 49 of keratoconics with corneal thinning were noted in 50 Stages 1, 2 and 4 of KC when applying the Amsler-Krumeich 51 classification. A hospital-based study conducted in Turkey 52 by Serdarogullari et al (2013) also reported that the patients 53 with KC showed clinical signs such as stromal thinning, 54 bulging of the cornea, Fleischer ring, and Vogt's striae (57). 55 Similarly, Salooti and Amir (2001) found that 35.5% of 56 patients presented with central or paracentral thinning, 57 40% with Fleisher rings, and 31.4% with Vogt's striae (58). 58 Elmassry et al (2021) focused on corneal endothelial cell 59 changes in the different stages of KC as classified using 60 Amsler-Krumeich classification, and found no significant endothelial changes in Stages 1 and 2 (59). However, Stage 361showed significant changes, often exhibiting polymegathism62and pleomorphism. Therefore, KC does cause physiological63changes in the different layers of corneal tissue which subse-64quently manifest as clinical signs depending on the severity65of the disease.66

67 The management strategy for keratoconics presenting to MPEH was most often a referral to the local univer-68 sity-based clinic followed by cross-linking, with contact 69 lenses not offered as a treatment modality. Previous hospital 70 71 and clinic-based studies have reported a different hierarchy of treatment options used for patients with KC, in order of 72 highest to lowest frequency, being contact lenses, spectacles 73 and surgery (4,46,56). A hospital-based study in London by 74 Lim and Vogt (2002) reported that 78.5% of the patients 75 were treated with binocular contact lenses, 18.5% with 76 monocular contact lenses and only 3% had received no 77 78 intervention (59). A similar trend was observed in a study in Sudan, Africa which reported that 60.8% of patients were 79 treated with contact lenses while 24.5% were treated with 80 spectacles (41). Contact lenses were a treatment of choice 81 because it allows the best possible vision for the patients 82 requiring optical correction and somehow delays the need 83 for surgery. The reason for most patients being referred in the 84 current study could be related to the facility, despite having 85 the equipment for a correct diagnosis of KC, not having the 86 necessary equipment for the fitting of contact lenses and 87 supplies, such as fitting sets. Furthermore, those patients that 88 are still in the early stages of KC are treated by crosslinking 89 to delay the progression of KC, which will have positive 90 91 long-term benefits to the patient and reduce treatment costs at the health care facility. The Health Professions Council 92 of South Africa (HPCSA) has a guideline for the minimum 93 tests to be conducted for any person that presents for a basic 94 eye examination. A study by Gcabashe et al (2022) revealed 95 that none of the eye care facilities in their study, including 96 MPEH had contact lens fitting sets, contact lenses, suction 97 holders among other instruments needed for management of 98 . Thus, a possible negative impact on the quality of life in the 99 keratoconic patients represented in the current study may be 100 expected, as many did not receive the required care when 101 they were first diagnosed and were subsequently not treated 102 at the public sector facility to which they were referred (60). 103 The deficiency in care of keratoconic patients at the facility 104 should be addressed by the relevant stakeholders to provide 105 patients with the expected minimum standards of eye 106 health care. 107

The current study has provided epidemiological data on the 108 prevalence of keratoconus and a clinical profile of keratoconic 109 patients presenting to a public eye hospital in KZN which has 110 not been determined by any previous study. This information 111 is key to effective strategies for the delivery of equitable and 112 adequate eye care for the diagnosis and management of KC 113 in the public sector in South Africa. The study however is not 114 without limitations. The information presented following this 115 retrospective study may not be complete as the instruments or 116 clinical tests used to detect KC were not always documented 117 in the clinical records. There was also no standardization 118 of record keeping amongst the large number of eye care 119 personnel employed at any one time. Furthermore, the clinical 120

61 62

68

69

70

71

72

73

74 75

76

77

78

records did not have any imaging data (tomography or topog-1 2 raphy) from which more data regarding KC could have been retrieved. As this was a hospital-based study, the prevalence 3 determined cannot be assumed to represent the larger popula-4 5 tion in KwaZulu-Natal. Moreover, screening for the optometry 6 department is done by a team of primary health caregivers 7 that does not include an optometrist hence leading to possible 8 referrals of keratoconics. Furthermore, as the current study 0 was cross-sectional, the age of onset of keratoconus cannot be 10 commented on.

11

12 Conclusions

13

14 The World Health Organization (WHO) in its strategic plan 15 for 2009-2013 emphasized the need for population-based data on the frequency of VI. One of the causes of VI is 16 uncorrected refractive error which may, in turn, be related 17 to ocular disease including KC. It is therefore important that 18 countries have access to clinical data that can inform policies 19 20 and strategies for the management of the ocular disease. The 21 prevalence of KC in the study was similar to that reported 22 by previous studies. Many patients that presented at the 23 facility in the current study were at the severe stage 4 of 24 KC highlighting the need for screening programs at lower 25 levels of care, efficiency in diagnostic protocols and patient 26 education programmes on KC. The screening team at hospi-27 tals, which should include optometrists, should be trained to identify patients at risk, or those with early signs of KC. 28 29 Vernal keratoconjunctivitis and sinusitis were the most noted 30 ocular disease and medical conditions present in the partici-31 pants. As previous studies highlight the strong link with these conditions, it is important that all patients that present 32 with VKC be screened for KC. The prevalence of KC in the 33 34 current study can be utilized to guide proper implementation 35 of appropriate diagnosis and management strategies and also assist in early diagnosis. This will help to reduce the likeli-36 37 hood of the affected patients becoming visually impaired 38 due to KC.

40 Acknowledgments

41

39

The authors thank Dr. Partson Tinarwo and Dr. FikileNkwanyana for assistance with statistical analysis.

45 Funding

46

44

The University of KwaZulu-Natal (UKZN) College of HealthSciences funded the fieldwork.

49

50 Ethical approval and consent to participate

51

The study commenced after ethical clearance and relevant
gatekeeper permissions were obtained from the Biomedical
Research Ethics Committee (BE332/19), Department of
Health in KZN, and MPEH, respectively.

57 Availability of data and material

- 56 57
- 58
- New and original data were collected, analyzed, and availableupon request from the corresponding author, NMG.

Contributions

NMG, VRM, RH conceptualized the project and designed the63methodology; NG, drafted the initial manuscript and wrote64the manuscript; VRM, RH supervised the project, guided and65reviewed drafts up to the final article. All the authors approved66the final version to be published.67

Conflict of interest

The authors declare no potential conflict of interest.

Accepted: 16, Deceber 2022; submitted: 18, October 2022.

References

- 1. Kummelil MK, Hemamalini MS, Bhagali R, Sargod K, Nagappa S, Shetty R and Shetty BK: Toric implantable collamer lens for keratoconus. Indian J Ophthalmol 61: 456-460, 2013.
- 2. Arne JL and Fournié P: Keratoconus, the most common corneal dystrophy. Can keratoplasty be avoided? Bull Acad Natl Med 195: 113-129, 2011 (In French).
- Weed KH, MacEwen CJ, Giles T, Low J and McGhee CN: The Dundee University Scottish keratoconus study: Demographics, corneal signs, associated diseases, and eye rubbing. Eye (Lond) 22: 534-541, 2008.
- Godefrooji DA, de Wit GA, Uiterwaal CS, Imhof SM and Wisse RP: Age-specific incidence and prevalence of keratoconus: A nationwide registration study. Am J Ophthalmol 175: 169-172, 2017.
- 5. Sharma N, Rao K, Maharana PK and Vajpayee RB: Ocular allergy and keratoconus. Indian J Ophthalmol 61: 407-409, 2013.
- 6. Kok YO, Tan GF and Loon SC: Review: Keratoconus in Asia. Cornea 31: 581-593, 2012.
- 7. Gillan WDH: Keratoconus: A clinical exposé. S Afr Optom 72: 9 41-45, 2013.
- Rabinowitz YS: Keratoconus. Surv Ophthalmol 42: 297-319, 1998.
- Maeda N, Klyce SD, Smolek MK and Thompson HW: Automated keratoconus screening with corneal topography analysis. Invest Ophthalmol Vis Sci 35: 2749-2757, 1994.
- Kennedy RH, Bourne WM and Dyer JA: A 48-year clinical and epidemiologic study of keratoconus. Am J Ophthalmol 101: 267-273, 1986.
- Nielsen K, Hjortdal J, Nohr EA and Ehlers N: Incidence and prevalence of keratoconus in Denmark. Acta Ophthalmol Scand 85: 890-892, 2007.
 Di L, Di L
- Bak-Nielsen S, Ramlau-Hansen CH, Ivarsen A, Plana-Ripoll O and Hjortdal J: Incidence and prevalence of keratoconus in Denmark. Acta Ophthalmol 97: 752-755, 2019.
- Millodot M, Shneor E, Albou S, Atlani E and Gordon-Shaag A: 103
 Prevalence and associated factors of keratoconus in Jerusalem: A Cross-sectional Study. Ophthalmic Epidemiol 18: 91-97, 104
 2011. 105
- Mugho SN, Ilako D and Nyenze EM: Prevalence of keratoconus in patients with allergic conjunctivitis attending Kenyatta National Hospital eye clinic. JOECSA 21: 18-23, 2016.
- Espandar L and Meyer J: Keratoconus: Keratoconus: Overview 108 and update on treatment. Middle East Afr J Ophthalmol 17: 109 15-20, 2010.
- Holland DR, Maeda N, Hannush SB, Riveroll LH, Green MT, ¹¹⁰ Klyce SD and Wilson SE: Unilateral keratoconus. Incidence 111 and quantitative topographic analysis. Ophthalmology 104: 112 1409-1413, 1997.
- 17. Lim X, Rabinowitz YS, Rasheed K and Yang H: Longitudinal ¹¹³ study of the normal eyes in unilateral keratoconus patients. ¹¹⁴ Ophthalmology 111: 440-446, 2004. ¹¹⁵
- Duncan JK, Belin MW and Borgstrom M: Assessing progression of keratoconus: Novel tomographic determinants. Eye Vis (Lond) 3: 6, 2016.
- Vinciguerra R, Ambrósio R Jr, Elsheikh A, Roberts CJ, Lopes B, 118 Morenghi E, Azzolini C and Vinciguerra P: Detection of keratoconus with a new biomechanical index. J Refract Surg 32: 119 803-810, 2016. 120

20. Romero-Jiménez M, Santodomingo-Rubido J and Wolffsohn JS: Keratoconus: A review. Contact Lens and Anterior Eye 33: 157-166, 2010.

8

1

2

49

50

51

52

53 54

55

56

57

58

59

60

- 3 21. Barsam A, Petrushkin H, Brennan N, Bunce C, Xing W, Foot B
 and Tuft S: Acute corneal hydrops in keratoconus: A national prospective study of incidence and management. Eye (Lond) 29: 469-474, 2015.
- 6 22. Amsler M: Le kératocône fruste au Javal. Ophthalmologica 96:
 77-83, 1938.
- 23. Mcmahon TT, Szczotka-Flynn L, Barr J, Anderson R, Slaughter M, Lass J and Iyengar S: A new method for grading the severity of keratoconus. Cornea 25: 794-800, 2006.
- 10 24. Belin M and Duncan J: Keratoconus: The ABCD grading system. Int J Kerat Ect Cor Dis 4: 85-93, 2015.
- 11 11
 25. Wagner H, Barr JT and Zadnik K: Collaborative longitudinal evaluation of keratoconus (CLEK) Study: Methods and findings to date. Cont Lens Anterior Eye 30: 223-232, 2007.
- 26. Zhang XH and Li X: Effect of rigid gas permeable contact lens on keratoconus progression: A review. Int J Ophthalmol 13: 1124-1131, 2020.
- 27. Saraç Ö, Kars ME, Temel B and Çağıl N: Clinical evaluations of different types of contact lenses in keratoconus management. Cont Lens Anterior Eye 42: 482-286, 2019.
- 28. Hoyer A, Raiskup-Wolf F, Sporl E and Pillunat LE: Collagen cross-linking with riboflavin and UVA light in keratoconus-results from Dresden. Ophthalmologe 106: 133-140, 2009 (In German).
- 20 29. York NJ and Tinley C: Corneal donations in South Africa:
 21 A 15-year review. S Afr Med J 107: 697-701, 2017.
- 30. Bezabih L, Abebe TW and Fite RO: Prevalence and factors associated with childhood visual impairment in Ethiopia. Clin Ophthalmol 11: 1941-1948, 2017.
- Rupnarain S, Madlala N, Memela N, Ngcobo S, Shabalala N, Simjee N, Gcabashe N and Rampersad N: Clinical characteristics of keratoconus patients at the University of KwaZulu-Natal eye clinic. African Vision and Eye Health 9: 7, 2020.
- 27 32. Chetty E and Rubin A: Preliminary demographics for patients with keratoconus attending a university-based clinic in Johannesburg, South Africa. South Africa. Afr Vision Eye Health: 78, 2019.
- 30 33. Hashemi H, Khabazkhoob M, Yazdani N, Ostadimoghaddam H,
 31 Norouzirad R, Amanzadeh K, Miraftab M, Derakhshan A and
 32 Yekta A: The prevalence of keratoconus in a young population in
 Mashhad, Iran. Ophthalmic Physiol Opt 34: 519-527, 2014.
- 34. Shneor E, Millodot M, Gordon-Shaag A, Essa M, Anton M, Barbara R and Barbara A: Prevalence of Keratoconus among Young Arab Students in Israel. Int J Keratoconus Ectatic Corneal Dis 3: 9-14, 2014.
- 36
 35. Netto TEA, Al-Otaibi WM, Hafezi NL, Kling S, Al-Farhan HM, Randleman JB and Hafezi F: Prevalence of keratoconus in paediatric patients in Riyadh, Saudi Arabia. Br J Ophthalmol 102: 1436-1441, 2018.
- 39 36. Ljubic A: Keratoconus and its prevalence in Macedonia.
 Macedonian J Med Sci 2: 58, 2009.
- 37. Kristianslund O, Hagem AM, Thorsrud A and Drolsum L: Prevalence and incidence of keratoconus in Norway: A nationwide register study. Acta Ophthalmol 99: 694-699, 2021.
- 38. Naderan M, Shoar S, Rezagholizadeh F, Zolfaghari M and Naderan M: Characteristics and associations of keratoconus patients. Cont Lens Anterior Eye 38: 199-1205, 2015.
- 45 39. Lucas SEM and Burdon KP: Genetic and environmental risk factors for keratoconus. Annu Rev Vis Sci 6: 25-46, 2020.
- 40. Rashid ZA, Millodot M and Evans KS: Characteristics of keratoconic patients attending a specialist contact lens clinic in Kenya. Middle East Afr J Ophthalmol 23: 283-287, 2016.

- 41. Khor WB, Wei RH, Lim L, Chan CM and Tan DTH: Keratoconus in Asians: Demographics, clinical characteristics and visual function in a hospital-based population. Clin Exp Ophthalmol 39: 299-307, 2011.
- 42. Mohd-Ali B, Abdu M, Das S and Mohidin N: Ethnicity related to keratoconus: A study with clinical implications. Int Med J 18: 237-240, 2011.
- Kapur N, Hunt I, Lunt M, McBeth J, Creed F and Macfarlane G: Primary care consultation predictors in men and women: A cohort study. Br J Gen Pract 55: 108-113, 2005.
 68
- 44. Bertakis KD, Azari R, Helms LJ, Callahan EJ and Robbins JA: Gender differences in the utilization of health care services. J Fam Pract 49: 147-152, 2000.
 68
 69
 69
 70
- Valdez-García JE, Sepúlveda R, Salazar-Martínez JJ and Lozano-Ramírez JF: Prevalence of keratoconus in an adolescent population. Revista Mexicana de Oftalmología 88: 95-98, 2014.
- 46. Ertan A and Muftuoglu O: Keratoconus clinical findings according to different age and gender groups. Cornea 27: 1109-1113, 2008.
- 48. Georgiou T, Funnel CL, Cassels-Brown A and O'Conor R: ¹⁷ Influence of ethnic origin on the incidence of keratoconus and associated atopic disease in Asian and white patients. Eye (Lond) 18: 379-383, 2004.
 40. Tyff SL, Maedalaw LC, Creegew WM, Daviage CB, and ⁸⁰
- 49. Tuft SJ, Moodaley LC, Gregory WM, Davison CR and Buckley RJ: Prognostic factors for the progression of keratoconus. Ophthalmology 101: 439-447, 1994.
- 50. Mhlanga D and Garidzirai R: The influence of racial differences in the demand for healthcare in South Africa: A case of public healthcare. Int J Environ Res Public Health 17: 5043, 2020.
 84
- 51. Abdu M, Binnawi KH, Elmadina AM and Hassan R: 85 Clinical profile of keratoconus patients in Sudan. Sudanese J Ophthalmol 8: 20-25, 2016.
- 52. Serdarogullari H, Tetikoglu M, Karahan H, Altin F and Elcioglu M: Prevalence of keratoconus and subclinical keratoconus in subjects with astigmatism using pentacam derived parameters. J Ophthalmic Vis Res 8: 213-219, 2013.
- 53. Mahadevan R, Arumugam AO, Arunachalam V and 90 Kumaresan B: Keratoconus-a review from a Tertiary Eye-care 91 center. J Optom 2: 166-172, 2009.
 92
- 54. Larkey LK, Hecht ML, Miller K and Alatorre C: Hispanic cultural norms for health-seeking behaviors in the face of symptoms. Health Educ Behav 28: 65-80, 2001.
- 55. Persoskie A, Ferrer RA and Klein WM: Association of cancer Worry and perceived risk with doctor avoidance: An analysis of information avoidance in a nationally representative US sample. J Behav Med 37: 977-987, 2014.
- 56. Maake ME and Moodley VR: An evaluation of the public sector optometric service provided within the health districts in KwaZulu-Natal. South Africa. Afr Vision Eye Health 77: 407, 2018.
- 57. Elmassry A, Osman A, Sabry M, Elmassry M, Katkat M, 100 Hatata MY and El-Kateb M: Corneal endothelial cells changes in different stages of Keratoconus: A multi-Centre clinical study. BMC Ophthalmol 21: 143, 2021.
- 58. Salooti Ř and Amir J: Characteristics of keratoconus patients in 103 Shiraz. BINA Journa 7: 19-24, 2001.
 59. Lim N and Voot II: Characteristics and functional actions of 104
- 59. Lim N and Vogt U: Characteristics and functional outcomes of 104 130 patients with keratoconus attending a specialist contact lens 105 clinic. Eye (Lond) 16: 54-59, 2002.
 106
- Gcabashe N, Moodley VR and Hansraj R: Keratoconus management at public sector facilities in KwaZulu-Natal, South Africa: Practitioner perspectives. Afr Vision Eye Health 81: a698, 2022.

- 110 111
- 112
- 113
- 114
- 117
- 115
- 116
- 117
- 118
 - 119 120