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Original research article

A Questionnaire Based Clinical Study to Assess the Sequence of Events Leading to Diagnosis of Keratoconus and its Impact on Quality of Life

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Abstract

Aim: To evaluate the sequence of events leading to diagnosis of keratoconus and its impact on quality of life.

Methods: This survey-based study was done the Department of Ophthalmology, Nalanda Medical College and Hospital, Patna, Bihar, India for 10 months, after taking the approval of the protocol review committee and institutional ethics committee. The study included patients over the age of 13 years who were diagnosed with subclinical or clinical KCN for the first time at our tertiary eye center.

Results: The present study included 200 eyes of 100 patients diagnosed with clinical or subclinical KCN. The mean age of patients at the time of diagnosis was 21.82 ± 5.89 years. Despite a drop in BCVA, 50(50%) patients reported never having visited an ophthalmologist before presenting to our tertiary eye care center. 40 (40%) patients were advised a screening test (corneal topography) to rule out KCN before presenting to our tertiary care center. 16 (40%) of these patients did not get it done. The time interval between the last screening test and KCN diagnosis at the tertiary center was 19 ± 5.9 months. The mean pachymetry at the thinnest point was 63.78 ± 60.8 Microns. Distribution of pachymetry at the thinnest location .The distribution of inter‑asymmetry score was <3 in 45 (27%), 3 in 15 (15%), and 4–5 in 58 (58%) patients. It was found that 68 (68%) had never noticed a difference in vision in the two eyes, whereas 32(32%) were aware of some difference.

Conclusion: Keratoconus is a disease of the young and severely affects their quality of life. Improving awareness of the general public, ensuring timely referral by optometrists, and keeping a high index of suspicion for KCN is emphasized.

Keywords: Quality of life, keratoconus

Introduction

Keratoconus (KC) is a corneal disorder characterized by corneal thinning, vision deterioration and irregular astigmatism that usually starts in the early teens.[1] The prevalence of KC was reported to be 1.38 per 1000 in a recent review.[2] KC etiology is currently unclear now, and the genetic and environmental factors have been reported to play important roles.[3-6] Epidemiological investigation of KC is helpful to further understand this condition. Most people in developing countries rely on optometrists to get their refraction checked and may not find it necessary to visit an ophthalmologist for an issue that they consider as minor, including a change in the refractive error. Moreover, many eye care centers lack facilities for corneal topography, which is the primary diagnostic tool for early KCN detection. Inability to refract to a BCVA of 20/20, presence of irregular/oblique astigmatism, scissoring reflex on retinoscopy, and high keratometry values of auto refractor should arise suspicion to screen a patient for KCN. Awareness among the public about this disease pathology is limited, unlike other common eye pathologies.[7] With the introduction of corneal cross linking (CXL), we can halt the progression of KCN and have reduced the need for corneal transplantation.[8] Still, we continue to see patients in cornea clinics with KCN related reduced quality of life either due to reduced BCVA or dependence on rigid contact lenses.[9] Many undiagnosed cases are seen to present with acute hydrops.[10] It is indeed unfortunate that most of these consequences could have been prevented with timely intervention.

Material and methods

This survey‐based study was done the Department of Ophthalmology, Nalanda Medical College and Hospital,Patna, Bihar, India for 10 months. after taking the approval of the protocol review committee and institutional ethics committee.

The study included patients over the age of 13 years who were diagnosed with subclinical or clinical KCN for the first time at our tertiary eye center. The diagnosis of KCN was performed via corneal topography, refraction, and clinical examination. Demographic data, BCVA, and pentacam records of every subject were retrieved. Staging of KCN was done according to the modified Amsler Krumeich staging classification system, and the inter‐eye asymmetry score was assessed according to a scoring system. They were provided with a proforma and the National Eye Institute Visual Function Questionnaire (NEI‐VFQ‐25) questionnaire and were asked to answer the given set of questions at the time of diagnosis. The sequence of events leading to the diagnosis of KCN was thoroughly investigated.

Results

The present study included 200 eyes of 100 patients diagnosed with clinical or subclinical KCN at a tertiary eye care center in India. The mean age of patients at the time of diagnosis was 21.82 ± 5.89 years. The youngest patient was 13 years old and the oldest was 35 years old. There were 65(65%) males and 35 females (35%). Out of 100patients of KCN, 2 (2%) were younger than 14 years of age and were classified as pediatric KCN. Two patients (2%) presented with acute hydrops as the initial presentation at the time of diagnosis. These two male patients were 20 and 18 years old respectively. Systemic illness was present in 5(5%) patients, 1 had bronchial asthma, 1 had allergic dermatitis, and 2 had allergic rhinitis.

History of vernal keratoconjunctivitis (VKC) was present in 34(34%) patients, with 23 (23%) patients being symptomatic for more than one year. 56 (56%) patients were not aware of the need to avoid eye rubbing, whereas 16(16%) and 28(28%) patients remembered being advised

against eye rubbing by an optometrist and ophthalmologist, respectively. 62 (62%) patients gave a history of sleeping more often on the side with worse KCN. 72(72%) used spectacles for vision correction as compared to 28(28%) using contact lenses At the time when diagnosis of KCN was made, 68(68%) patients were not aware of a disease called "keratoconus." 6 (6%) and 22 (22%) patients were hinted about the possibility of this disease by their optometrist or ophthalmologist respectively. 4(4%) were aware of the condition due to the presence of a similar condition in a family member or acquaintance. Out of 52 patients who were aware of the condition, 48 (92.3%) were aware of the possible consequences of progression in this condition. 45 (45%) patients preferred visiting an optometrist for their complaints. The distribution of the preferred primary point of contact for patients.

Despite a drop in BCVA, 50(50%) patients reported never having visited an ophthalmologist before presenting to our tertiary eye care center. The factors associated with not consulting an ophthalmologist are summarized in Table 1. 40 (40%) patients were advised a screening test (corneal topography) to rule out KCN before presenting to our tertiary care center. 16 (40%) of these patients did not get it done. Factors attributed to not getting a screening test done are summarized in Table 2. 13 (13%) patients underwent a screening test for KCN in the form of corneal topography. However, none of them were diagnosed as KCN at that time. The time interval between the last screening test and KCN diagnosis at the tertiary center was 19 ± 5.9 months.

First visit to a tertiary eye care system where diagnosis of KCN was made

32 (32%) patients were referred by an optometrist or previous practitioner. The reasons for visiting our tertiary eye care center when the first diagnosis of KCN was made were as follows: referred by previous practitioner [10 (10%)], referred by optometrist [24 (24%)], "not satisfied with glasses prescribed elsewhere" [28(28%)], first consultation for reduced vision[35 (35%)], and consult for some other complaint [2 (2%)].

Visual acuity and refractive error

Corrected distance visual acuity and refractive variables are summarized in Table 3.

Table 1: Factors associated with not consulting an ophthalmologist

Factors associated with not consulting an ophthalmologist	Number of patients (Percentage patients
No ophthalmologist in the vicinity	3 (6%)
Faith in local practitioner/optician	6 (12%)
Considered it a minor problem	39 (78%)
Cost factor	2 (4%)

Table 2: Factors for not undergoing screening corneal topography

Factors for not undergoing screening corneal topography	Number of patients (Percentage patients)
Found the test to be unnecessary/ considered the disease a minor problem	16 (40%)
High cost	11 (27.5%)
Non‑availability of the machine required for test in the concerned center	10 (25%)
Lack of time/too busy	3 (7.5%)

Corneal tomography

A diagnosis of clinical KCN was made based on clinical features, refraction, and corneal tomography. Subclinical KCN was diagnosed based on corneal tomography suggestive of KCN.

The mean pachymetry at the thinnest point was 63.78 ± 60.8 Microns. Distribution of pachymetry at the thinnest location is shown in Fig. 3.

The distribution of inter‐asymmetry score was <3 in 45 (27%), 3 in 15 (15%), and 4–5 in 58 (58%) patients. It was found that 68 (68%) had never noticed a difference in vision in the two eyes, whereas 32(32%) were aware of some difference.

The distribution of stage of KCN (modified Amsler Krumeich staging) in the worse eye at the time of diagnosis was as follows: Stage 1 in 24(24%), 2 in 48 (48%), 3 in 4 (4%), and 4 in 23(23%) patients. 1(1%) patients presented with acute hydrops at the time of diagnosis.

After diagnosing KCN, they were offered options for visual rehabilitation. Ninety‐six (58%) selected spectacles, 16(16%) selected rigid contact lenses, and 26(26%) selected scleral contact lenses.

Quality of life

- NEI‐VFQ‐25

All 100 patients completed the NEI‐VFQ‐25 questionnaire and were included in the analysis. The scores are summarized in Table 4. Further, we asked the patients about the effect of reduced vision on the career they wish to pursue, and 28 (28%) patients self‐reported that they felt that their choice of career was now compromised because of poor vision attributed to the diagnosis of KCN. We studied the correlation of vision‐targeted composite score with various variables. It showed no relation with age of diagnosis (r value: −0.005) but showed a significant negative correlation with grade of KCN (r value: −0.481) and positive correlation (r value: 0.544) with LogMAR vision at presentation.

Table 3: Visual Acuity and Refractive variables

Parameter	Mean±standard deviation (range)
CDVA (logMAR)	0.28±0.24 (0‐1.6)
Spherical Equivalent (D)	2.72±1.95 (0.25‐11.5)
Cylinder (D)	2.90±1.45 (0.5‐7)
Axis	106.81±49.96 (10‐180)
K mean (D)	49.26±4.75 (40.6‐66.1)
K max (D)	55.19±7.35 (42.3‐87.6)

Table 4: National Eye Institute Visual Function Questionnaire (NEI‐VFQ‐25) scores

	General Health	General Vision	Ocular Pain	Near Activities	Distance Activities	Social Function
Mean± SD	72.91±28.3	56.8±19.2	81±21.7	83.82±14.7	79.8±17.7	89.12±15.7
Range	0‐100	20‐100	20‐100	25‐100	25‐100	50‐100

	Mental Health	Role difficulties	Dependency	Driving	Color vision	Peripheral vision
Mean± SD	**63.9±18.5**	**82.9±21.6**	**93.1±14.5**	**84.3±11.6**	**97.6±8.6**	**95.1±6.6**
Range	**18.75-93.75**	**25-100**	**33.3-100**	**58.3-100**	**75-100**	**75-100**

Discussion

KCN is primarily a disease of early adulthood, beginning typically at about the age of puberty, and usually progresses over the next 10 − 20 years. The mean age of patients at the first visit to an ophthalmologist, as also the age at which diagnosis of KCN was made, was 20.82 ± 5.89 years in our study. It affects both genders; however, gender predisposition is unclear, with some studies reporting equal prevalence between genders;[11,12] while other investigators have found a greater prevalence in males,[13,14] as is also supported by our study. The mean cylindrical refractive error at presentation was 2.90 ± 1.45 D. It is imperative to point out that KCN can present with low cylindrical power, and a high index of suspicion is necessary to diagnose this condition. Correlation of history of VKC to KCN goes in coherence with other studies,[15,16] but the tragic part highlighted is the lack of awareness among patients about the relation between eye rubbing and KCN, with the majority of patients (56%) not knowing about this correlation. Ignorance among patients and lack of regulations necessitating regular follow-up with an ophthalmologist might be the reason, which needs to be worked on.

In our study, 62 (62%) patients gave a history of sleeping more often on the side with worse KCN. Similarly, a recent study highlighted that in KCN patients, the most affected eye correlated with the preferential side on which patients were used to sleeping.[17] This likely association can be explained by compression forces on the eye, which results in the release of inflammatory mediators, which further result in keratocyte apoptosis, contributing to stromal thinning.

The natural course of KCN is progressive, and the disease can only be halted at the stage at which it is diagnosed.[14] In our study, 13(13%) patients, despite undergoing a screening test for KCN (corneal topography), were not diagnosed as KCN at that time. The time interval between the last screening test and KCN diagnosis at our tertiary center was 19 ± 5.9 months. Therefore, a higher suspicion and regular examination will ensure that patients are diagnosed at an early stage of KCN. In our study, we found that at the time when the diagnosis of KCN was made for the first time, as many as 23 (23%) patients were at stage four of KCN. Furthermore, in two patients, acute hydrops was the presenting feature of KCN. Literature supports that most of the cases of acute hydrops are seen in the second or the third decade with preponderance for the male gender.10 In our study too, the two patients were males and were 19 and 20 years old, respectively. Both these patients, despite being spectacle users for 5 and 6 years, respectively, had never visited an ophthalmologist and were never screened for KCN. Reduced vision at the time of diagnosis is also highlighted in the study. This indicates that a substantial loss of visual acuity had already occurred in the patients by the time a confirmatory diagnosis of KCN was made. Furthermore, 12(12%) of these patients had a thinnest pachymetry of <400 □, making CXL a challenge.[8]

We investigated the sequence of events related to the diagnosis of KCN in these patients. The preferred primary point of contact was an optometrist in the majority of patients (45%), indicating that the role of optometrists needs to be emphasized to ensure early diagnosis of KCN. Despite BCVA getting worse than 6/6, patients did not prefer to consult an ophthalmologist/ tertiary eye care center; 50% of patients had never visited an ophthalmologist for their complaints. It is alarming to note that 68(68%) of these patients never visited an

ophthalmologist as they considered it to be a minor issue. Most of the patients (68%) were unaware of the disease entity and were never screened (68%), suggesting the need to improve awareness among patients and healthcare professionals.

KCN is known to be a bilateral asymmetric condition.[18,19] The inter‐eye asymmetry score in our study was 4 − 5 in 57 (57%) patients. Further, upon assessing whether patients had ever noticed any difference in visual acuity in the two eyes prior to being diagnosed with KCN, it was found that 68 (68%) had never noticed a difference in vision in the two eyes. Children may not notice a difference in vision in the two eyes, necessitating routine ophthalmological examination.

Similar studies on KCN reporting to tertiary care centers suggest that KCN in India presents at a younger age than in the Western population and progresses more rapidly.[20] This emphasizes the need to build up our reach of tertiary care facilities in the developing world. We follow a protocol of screening patients with corneal topography when either of the following criteria is met: inability to refract to 20/20 with high cylindrical power against the rule/oblique astigmatism, high keratometry value, and progressive increase in cylindrical power or keratometry value. Using this protocol, we have been able to pick up subclinical KCN at a relatively early stage. Various centers can come together to create a protocol to screen patients for KCN so that these cases can be picked up early in the course of the disease. We found that poor quality of life scores were associated with worse grade of KCN and BCVA at the time when the diagnosis was made. This is in coherence with previous studies.[21] Patients with the disease, unlike other ocular pathologies, belonged to a lower age group and hence reduction in quality of life seems more important and impactful.

Conclusion

Keratoconus is a disease of the young and severely affects their quality of life. Improving awareness of the general public, ensuring timely referral by optometrists, and keeping a high index of suspicion for KCN is emphasized.

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