

# Keratoconus and its Association with Other Ocular Diseases

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**Purpose:** To determine the association of keratoconus with other ocular diseases.

**Material and Methods:** A study on Keratoconus (KC) was conducted in the Department of Ophthalmology, Gomal Medical College, DHQ Teaching Hospital, Dera Ismail Khan (NWFP) from July 2003 to June 2006.

Fifty cases of Keratoconus were studied. Each patient underwent a complete ocular examination including visual acuity, refraction, slitlamp biomicroscopy, keratometry, applanation tonometry, direct and indirect ophthalmoscopy.

**Results:** The majority of patients 41 (82%) presented between 13 and 28 years of age. Male to female ratio was 7:1. Forty one (82%) patients had bilateral keratoconus and nine (18%) had unilateral keratoconus. The most common ocular association was vernal catarrh (64%). Penetrating keratoplasty was suggested for 49 eyes (53.85%), and spectacles correction alone or spectacles with contact lens correction in 42 eyes (46.15%) in order to improve the visual acuity of the patients to acceptable level.

**Conclusion:** The commonest ocular association was vernal catarrh (64%). Keratoconus is a problem of young people and 53.85% of eyes needed corneal graft. Because of non availability of donor tissue material, educating the people in our society through all the possible media to donate their eyes is required.

**K**eratoconus (KC) is a progressive, non inflammatory<sup>1</sup>, bilateral (but usually asymmetrical) disease of the cornea, characterized by paraxial stromal thinning that leads to corneal surface distortion. Painless visual loss<sup>2</sup> occurs primarily from irregular astigmatism, myopia and secondarily from corneal scarring.

All layers of the cornea are believed to be affected by KC, although the most notable features are the thinning of the corneal stroma, breaks in bowman layer, and the deposition of iron in the basal epithelial cells, forming the Fleischer ring. Breaks in and folds

close to the descemet's membrane result in acute hydrops and Vogt's striae respectively.

Although KC may be associated with a number of ocular and systemic disorders, its exact pathogenesis is not clear<sup>3</sup>. Commonly recognized ocular associations include vernal keratoconjunctivitis, retinitis pigmentosa and Leber's congenital amaurosis.

Particular risk factors include atopic history, especially ocular allergies, rigid contact lens wear and vigorous eye rubbing<sup>4</sup>.

Early in its course, optical correction is achieved through spectacles. As the irregular astigmatism

progresses, rigid contact lens can be quite successful in restoring vision. When patient's visual acuity can not be significantly improved with contact lenses or contact lenses are not tolerated well, then surgery in the form of penetrating keratoplasty is indicated.

## **MATERIAL AND METHODS**

We included 50 cases of KC at the department of ophthalmology, Gomal Medical College, D.H.Q Teaching Hospital Dera Ismail Khan from July 2003 to June 2006.

The details of history and clinical examination of the patients were recorded on a specially designed performa. Each patient underwent a complete ocular examination. Ocular examination included the following:

### **Visual Acuity**

Both uncorrected and best corrected visual acuity with glasses was checked with standard Snellen's chart.

### **Refraction**

Retinoscopy, autorefractometry and trial and error methods were performed to achieve best corrected visual acuity with glasses.

### **Biomicroscopy**

Slitlamp biomicroscopy was performed to examine the anterior segment in detail with special attention to detect.

- ❖ Corneal Thinning.
- ❖ Vogt's striae.
- ❖ Fleischer's ring (with cobalt blue filter).
- ❖ Stromal scarring.
- ❖ Rupture in Descemet's membrane.
- ❖ Acute corneal hydrops.
- ❖ Vernal conjunctivitis.

### **Keratotomy**

It was performed to study the general shape of the cornea.

### **Keratometry**

Keratometry with von Helmholtz keratometer was performed to measure the radius of curvature of the central cornea and dioptric refracting power of the cornea.

### **Corneal Sensation**

Corneal sensation at the apex of the cone was checked with cotton wick.

### **Munson's Sign**

Munson's sign was checked by asking the patient to look down, to see the angulation of the lower lid.

### **Rizzuti Illumination Test**

Penlight was thrown from the temporal side anterior to the iris plane. In KC, the ectatic cornea focuses the light sharply inside the nasal limbus.

### **Intraocular Pressure**

Before checking the intraocular pressure both eyes were anaesthetized by putting drops of proparacaine hydrochloride (Alcaine) twice, one minute apart. Sterile fluorescein paper strips were used to colour the lachrymal fluid. Intraocular pressure was measured with hand-held Perkin's applanation tonometer. The graduation value of the prism in the axis of minus cylinder was set at the mark of the prism holder (equivalent of red 43° mark of Goldmann applanation tonometer).

### **Direct Ophthalmoscopy**

Before ophthalmoscopy pupils were dilated by putting drops of 10% phenylephrine (Isonefrine) and 1% Tropicamide (Mydriacyl) in the conjunctival sac. Direct ophthalmoscopy was performed to see the dark area with in the illuminated field.

### **Indirect Ophthalmoscopy**

Fundus examination was difficult with direct ophthalmoscope due to higher astigmatism, for detailed fundus examination indirect ophthalmoscopy was performed.

## **RESULTS**

The majority of patients, i.e. 41 (82%), presented between 13 and 28 years of age. Their age ranged from 9 to 50 years. Mean age was 22.3 years (Figure 1). There were 44 males (88%) and 6 females (12%). Male to female ratio was 7:1.

Forty one patients (82%) had bilateral KC and nine patients (18%) had unilateral KC. So the total number of eyes involved were 91. Out of 41 bilateral cases 36(87.8%) were males and 5(12.2%) were females, while out of 09 unilateral cases 8(88.89%) were male and 1(11.11%) was female (Table 1).

Uncorrected visual acuity was hand movement (H.M) to 6/60 in 60 eyes (65.93%), 6/60 - 6/24 in 21 eyes (23.07%), 6/18 in 5 eyes (5.50%), 6/12 in 2 eyes (2.20%) and 6/9 in 3 eyes (3.30%) (Figure 2).

Best corrected visual acuity with glasses was H.M. to 6/60 in 33 eyes (36.26%), 6/60-6/24 in 24 eyes (26.37%), 6/18 in 9 eyes (9.89%), 6/12 in 8 eyes (8.79%), 6/9 in 9 eyes (9.89%) and 6/6 in 8 eyes (8.79%) (Figure 2).

The duration of KC at the time of presentation ranged from 3 months to 30 years. Mean duration was 7.9 years (Table 2).

35 patients (70%) gave positive history of excessive eye rubbing, while 12 patients (24%) gave no such history, and 3 patients (6%) gave history of occasional eye rubbing.

Munson's sign was present in 72 eyes (79.12%) and was not detectable in the remainder of eyes.

Rizzuti's sign was present in 82 eyes (90.10%), while in the rest of eyes it was absent. Fleischer's ring was seen in 27 eyes (29.67%), out of which in 7 eyes it was complete and in 20 eyes the ring was incomplete. Vogt's striae were present in 51 eyes (56.04%), and were absent in 34 eyes (37.36%), while in 6 eyes (6.59%) Vogt's striae were not detectable due to acute corneal hydrops. Corneal sensation at the apex of cone was normal in 67 eyes (73.63%), while it was decreased in 24 eyes (26.37%). Stromal scarring was present in 60 eyes (65.93%) and was absent in 31 eyes (34.07%).

**Table 1:** Laterality, Sex distribution and total number of eyes involved in KC patients

	No. of Patients	No. of Eyes	Males	Females
Bilateral KC	41	82	36	5
Unilateral KC	9	9	8	1
Total	50	91	44	6

**Table 2:** Duration of disease at the time of presentation

Duration in Years	No of patients n (%)
0-5	28 (56)
6-10	8 (16)
11-20	12 (24)
21-30	2 (4)
Total	50 (100)

Intraocular pressure ranged from 2 to 15 mm Hg in 89 eyes. Mean intraocular pressure was 7.6 mm Hg. In 2 eyes intraocular pressure was not recordable. Five patients (10%) presented with acute corneal hydrops.

At the time of presentation their ages ranged from 7-25 years, mean age was 12.2 years. One patient at the age of 12 years developed bilateral acute corneal hydrops, first left, followed by right eye after 40 days. She had bilateral aphakia, operated for steroid induced cataract three years back. One other patient at the age of 7 years developed right acute corneal hydrops, which resolved after some weeks, but he developed left acute corneal hydrops after 7 months following resolution of the right hydrops.

We found oval cones in 54 eyes (59.34%) and round cones in 37 eyes (40.66%). In 23 patients (46%) there was family history of vernal catarrh and KC. In three families, two brothers in each were suffering from vernal catarrh and KC. In the rest of 27 patients (54%) there was no family history of either KC or any other associated disorder (Table 3).

Most of the patients in our series belonged to poor and lower middle class i.e. 40 patients (80%). Eight patients (16%) belonged to upper middle class and 2 patients (4%) belonged to upper class.

**Ocular Associations** (Table 4)

**Table 3:** Family history of KC patients

Disease	No of patients n (%)
Nil	27 (54)
Vernal Catarrh	20 (40)
Vernal Catarrh + KC	3 (6)
Total	50 (100)

**Table 4:** Ocular association of patients with KC

Disease	No of patients n (%)
Vernal conjunctivitis	32 (64)
Steroid induced cataract	6 (12)
Pre senile mature Cataract	2 (4)
Blepharoptosis	3 (6)
Strabismus	3 (6)
Arcus Juvenilis	2 (2)
Steroid induced glaucoma	1 (1)
Epicanthus	1 (1)

Total	50 (100)
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### **Vernal Catarrh**

Thirty two patients (64%) were suffering from vernal catarrh in our series.

### **Cataract**

Eight patients (16%) had cataract, of which 6 patients had bilateral steroid induced posterior subcapsular lenticular opacities and 2 patients had bilateral pre-senile mature cataract.

### **Blepharoptosis**

Three patients (6%) had bilateral mild ptosis due to vernal catarrh and or topical steroids.

### **Strabismus**

Three patients (6%) had strabismus, 2 had convergent and 1 had divergent strabismus.

### **Arcus Juvenilis**

Two patients (4%) had arcus juvenilis.

### **Glaucoma**

Bilateral steroid induced glaucoma was present in 1 patient (2%), for which he had bilateral trabeculectomy.

### **Epicanthus**

We noted bilateral epicanthus in 1 patient (2%).

## **DISCUSSION**

KC has generated considerable interest in a large number of ophthalmologists since it was recognized as a pathological entity in the middle of the eighteenth century. About a century later KC was differentiated from other corneal ectatic disorders by Nottingham<sup>5</sup>, who defined its essential clinical features. Since then a considerable amount of work has been published on the subject, most addressing either the clinical or the etiological aspects of the disorder.

In our series 82% of patients presented between 13 and 28 years of age. KC is thus a problem of young people and should be suspected in any adolescent with progressive myopic astigmatism<sup>6</sup>.

A female predominance has been observed in most studies, however the ratio varies<sup>1</sup>. In our study males were affected more than females with a ratio of 7:1. This high male: female ratio is probably because of high number of vernal catarrh patients, which is predominantly a disease of male children. Our male dominant society, religious veil and dependence of females on males might also play some role so they are

unable to seek medical advice in time for most of minor and some of the major problems.

We detected 18% of patients with unilateral KC, which closely resembles the figure 14.3% reported by Amsler<sup>7</sup>.

In our study 70% of patient with KC gave positive history of excessive eye rubbing. Several reports have implicated eye rubbing as an important etiological factor in the development of KC<sup>4,5,8-11</sup>. The reported prevalence ranges from 66% to 73%<sup>10-12</sup>. Eye rubbing may be the etiological link between conical cornea and associated systemic and ocular diseases. Itching, ocular irritation and eye rubbing are common features of vernal catarrh and atopic disease. We found 32 patients (64%) associated with vernal catarrh. Perhaps the chronic ocular massage is responsible for the increased occurrence of KC in these patients. Gritz and McDonnell<sup>13</sup> also suggest such an association, but a direct cause and effect relationship has not been proven.

Waardenburg et al<sup>14</sup> cites seven cases of KC involving blood relatives and thus suspects a recessive genetic factor. The early development of KC and the consanguinity of the parents support the hypothesis that this is likely to be an autosomal recessive disorder<sup>15</sup>. Our results support this hypothesis.

Mild forms of KC are treated with spectacles or contact lenses. Contact lens wear becomes necessary when cone progression increases the degree of irregular astigmatism. Although contact lenses have been speculated to cause KC<sup>16</sup>, they are also essential for the optical treatment of this disorder, and their use can not be avoided.

In South Africa 0.92% of all the blind were due to KC<sup>17</sup>. Blindness due to KC is treatable. Surgery for KC is indicated when the patient is unable to obtain clear, comfortable vision with contact lenses. KC patients have one of the highest success rates of any group of patients undergoing penetrating keratoplasty<sup>18</sup>. We advised penetrating keratoplasty in 49 eyes (53.85%).

Penetrating keratoplasty in this part of the world is a difficult option because of non availability of donor tissue material.

The answer naturally is to tap a national source. The major obstacle for tissue donation is the religious faith that by enucleating the eye, you are desecrating the human body which is a sacred gift from God. This thinking is based on total misconception. The eyes of the deceased individual are enucleated after due permission from the deceased when he was alive, or

from the next of kin after his death and with the noble idea of transplanting it to a needy fellow human being. This point needs to be properly explained to all the relevant religious quarters by ophthalmologists and other members of eye bank society, a society dedicated to the promotion of corneal grafts in Pakistan. The newspapers and the electronic media may be involved to play their due role in this campaign.

## CONCLUSIONS

After studying 50 cases of KC we arrived at the following conclusions:-

1. The commonest ocular association is vernal catarrh (64%).
2. KC is a problem of young people, majority of patients present between 13 and 28 years of age.
3. Males are affected more commonly than females
4. 53.85% of eyes needed corneal graft. Indeed the commonest indication for keratoplasty nowadays is KC at some centers.
5. Educating the people in our society through all the possible media to donate their eyes may be of great help in improving the grim future of KC patients.

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### Editor's Note:

As the endothelium of most of the keratoconus patients is healthy, we should utilize it for better, lasting results and laser complications etc etc by recommending lamellar keratoplasty rather than penetrating keratoplasty requiring training our young corneal surgeons for this techniques.