

A Retrospective Study of the  
Clinical Profile of Keratoconic Patients  
at Ferris State University Optometry Clinic.

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## ABSTRACT

We conducted a retrospective study of the records of 40 keratoconic patients presenting at the Ferris State University Optometry Clinic. The purpose of this study was to develop a clinical profile of the presentation of an average keratoconic patient. Parameters examined included the sex of the patient, the age of the patient at the time of diagnosis, initial keratometric readings, initial best visual acuity, initial corneal signs, laterality, history of eye rubbing, and family history. The progression of the disease was also studied by noting if the patients were fit with rigid gas permeable contact lenses, if surgery was indicated, and any corneal signs which developed during the time they were followed. A rate of change of the keratometric readings was also calculated in order to quantify the progressive steepening of the cornea. Lastly, data from the patients' most recent examination and corneal topography was studied to determine the most recent best visual acuity and the position of the cone itself.

## INTRODUCTION

Keratoconus is a noninflammatory ectasia of the cornea, in which the cornea assumes an irregular conical shape.(1-3) The disease is typically bilateral although asymmetrical involvement of the two eyes is not uncommon.(4) It is a progressive disease, resulting in thinning and distortion of the cornea.(5) This leads to a mild to marked impairment of vision secondary to advanced and irregular astigmatism or corneal scarring which is often present.(6)

The management of keratoconus depends on the severity of the disease. Patients with this disease can be divided into three groups on the basis of severity. In mild disease, patients can obtain good visual acuity with spectacle, soft contact lenses, or rigid gas permeable (RGP) contact lenses.(7) In moderately severe disease, there is more myopia and irregular astigmatism, without significant corneal scarring. Spectacle correction does not provide adequate visual acuity in this group because of the irregular astigmatism. RGP contact lenses are indicated for this degree of keratoconus because they can correct the irregular astigmatism and provide acceptable visual acuity without surgery.

In severe keratoconus, contact lens correction is not adequate because of discomfort or poor visual acuity. These problems may be secondary to severe corneal ectasia and steepening or corneal scarring, and penetrating keratoplasty is required to restore vision.(8) Corneal transplantation is necessary for 10 to 20% of patients with keratoconus.(9-10)

The onset of keratoconus is reported to occur at puberty with no significant difference between sexes. However, keratoconus has been diagnosed in patients as early as the first decade of life and as late as the fifth decade.(9) Keratoconus occurs in all races. The most valid prevalence figures are probably from a study from the Mayo Clinic, which cited a prevalence of 54.5 cases per 100,000 population in Olmstead County, Minnesota in 1982.(11)

The origin, pathogenesis and biochemistry of keratoconus are unknown.(4) Keratoconus appears to be a disease or condition where many causative factors result in similar clinical pictures.(5) Proposed causes of this disease include atopic disease (eye rubbing response), systemic conditions (connective tissue disease), heredity and rigid contact lens wear.(5) However, none of these associations have given rise to a uniform theory of the basis of this disease, genetic or otherwise.(4)

Investigation of the as yet unknown pathogenesis of keratoconus is another area in which accurate and objective diagnosis would be helpful. The purpose of this study is to aid the clinician in the diagnosis of keratoconus by developing a clinical profile of the presentation of an average keratoconus patient.

## **METHODS**

A retrospective study was performed by reviewing the corneal topographies and records of 40 keratoconic patients seen at the Ferris State University Optometry Clinic. Data was only collected for the patient's worst or most involved eye. Information gathered

included the patients age at the time of diagnosis, sex, initial keratometric readings, initial best visual acuity, any initial corneal signs present, laterality, history of eye rubbing, and any family history of the disease. We were also able to collect data on several patients regarding the progression of their condition. This data included things such as if the patient was fit with a RGP contact lens, if surgery was ever recommended, and the appearance of corneal signs during follow-up care. Data from the patients most recent examination and corneal topography was also collected. This included the final best visual acuity, the keratometric reading from the topography as well as the position of the cone itself. By comparing the patients final keratometric reading to the initial keratometric reading we were also able to calculate a rate of change in the keratometric readings of several patients.

## RESULTS

Due to the fact that this study was retrospective in nature, the data from some patient records was incomplete in one or more categories. In the event that a patient's data was incomplete in a certain category, that patient was not included in the calculations for that category. Therefore, the mean for several categories is based on less than 40 patients. When this is a factor it will be indicated along with the data.

First, the results from the patients' initial presentation at our clinic, or from the initial diagnosis if available, will be reviewed. The age of first diagnosis was known for 37 patients and the mean was found to be 29.2 years of age. The range of age when first diagnosed was 13 to 67 years of age. Our sample population had a significant prevalence of males, 31/40 or 77.5%, while only 9/40 or 22.5% were female. Initial keratometric readings were known for 35 patients and these readings ranged from 42.25 to 90.48 diopters. The mean initial keratometric reading was 54.33 diopters. Data regarding the initial best visual acuity (BVA) was available for 38 patients, and was measured with Snellen acuity charts. The range for the BVA was 20/20 to 20/200. One patient's

BVA was measured to be 6/400, but this piece of data was excluded from calculation of the mean because it was not representative of our sample and severely skewed the results. The mean BVA at initial presentation was found to be 20/36.1 , or approximately 20/40. Corneal signs were noted at the initial presentation of 26 out of the 40 (65%) patients in the study. The corneal signs included Fleisher's ring, epithelial staining, stromal thinning, corneal scars, positive Munson's sign, distorted keratometric mires and a scissors reflex with retinoscopy. The condition was found to be bilateral in 31 of the patients or 77.5%. A positive history of eye rubbing was only noted in 2 patients or 5%. A positive family history of keratoconus was known for only 3 patients or 7.5%.

Data regarding the progression of the disease was evaluated next. A significant number of patients were fit with RGP lenses at some point during their care, 31/40 or 77.5%. Surgery was indicated for 8/40 or 20% of the patients studied. Corneal signs became apparent in 29/40 or 72.5% of the patients. The rate of change of the keratometric readings was calculated for 8 of the 40 patients. The mean rate of change was found to be 0.248 diopters/month or 2.97 diopters/year. The range of this value was 0.06 to 0.67 diopters/month or 0.72 diopters/year to 8.04 diopters/year.

Finally, data from the patients' most recent exam and corneal topography were compiled. The most recent BVA was known for 38 of the 40 patients, and the mean was found to be 20/28.55 or approximately 20/30 Snellen acuity. The range of the most recent BVA was 20/20 to 20/80 Snellen acuity. The steepest keratometric reading from the most recent corneal topography of the most involved eye was recorded for the entire sample. The range was found to be 44.82 diopters to 90.48 diopters, with a mean value of 58.54 diopters. The location or position of the cone was measured from the steepest point on the corneal topography. In 65% of the patients, the right eye was the most involved or worst eye. The average position of the cone was calculated to be 1.57mm from

center inferior nasally, or more specifically 32.75 degrees nasally from the straight inferior position.

## DISCUSSION

In summary, the clinical profile of the average keratoconic patient is as follows. According to our study the patient is likely to be 29 years of age, male with keratometric readings of 54 diopters and best visual acuity of approximately 20/40 Snellen. Only 65% of the patients initially show corneal signs ranging from slit lamp examination findings to distorted keratometric mires to scissors reflex with retinoscopy. The condition is bilateral in 77.5% of patients with the right eye being most likely to be more involved. Only 5% of patients will give a positive history of eye rubbing, while only 7.5% of patients will have a known family history of the condition.

According to our results, the typical rate of progression is nearly 3 diopters of corneal steepening per year. The clinician can expect to eventually find corneal signs in 72.5% of keratoconic patients. He or she can also expect 77.5% of patients to eventually require RGP contact lenses and surgery to be indicated in 20% of patients with keratoconus.

On average, the position of the cone itself is likely to be inferior nasal approximately 1.57mm from center with a final keratometric reading of 58.54 diopters. The clinician can expect the average keratoconic patient to be correctable to approximately 20/30 in Snellen acuity. Our study seems to support the fact that most keratoconic patients can be managed with glasses or contact lenses, while surgery is only indicated in approximately 20% of patients.

Early diagnosis of keratoconus can be difficult. For a long time, it has been recognized that keratoconus can exist in the absence of corneal signs. Our study showed that 65% of patients could be diagnosed by slit lamp examination. We feel that for the

early detection of keratoconus, the study of paracentral corneal topography appears to be the most sensitive. Since corneal topography is not always available, several clinical techniques have been suggested to detect these early cases. These include retroillumination techniques such as the "Charleaux oil droplet" reflex and early scissors reflex with retinoscopy, pachometry, steepening of the keratometric readings in up gaze, and distortion of the mires on keratometry.

Patients should be informed of the progressive, yet unpredictable nature of keratoconus. Patients should also be informed of the hereditary nature of the disease as well. Onset of the disease at an early age (2nd to 3rd decade of life) generally indicates that the disease will progress to a more advanced stage. The prospect of treatment by keratoplasty increases with an earlier onset of disease.(5) Krachmer and co-workers tell their patients that the chances are less than 1 in 10 that a blood relative will have the disease.(11) Furthermore, the finding that only 20% of patients with keratoconus require keratoplasty when followed for 20 years after the diagnosis(9), coupled with the finding that 90% of keratoplasties for keratoconus result in a clear graft(11,14), suggests that there is only a 1 in 500 chance that a blood relative will have the disease, require keratoplasty, and end up with a cloudy graft.(4) Our study correlates quite well with these findings. We found that 1 in 13 patients with keratoconus, or 7.5%, have a family history of the disease and that keratoplasty was indicated in 20% of the patients in our study. These two factors coupled together with the surgical success rate of 90% for keratoconus keratoplasties suggests that there is only a 1 in 650 chance that a blood relative will end up with cloudy corneal graft. Keep in mind that this disease is often asymmetrical and the odds of profound bilateral visual impairment in a relative is further reduced.

We feel that our study has demonstrated the average clinical profile of the presentation of an average keratoconus patient and

the progressiveness of the disease. We hope our study and its results will aid the clinician in the early detection and diagnosis of keratoconus, so that these patients may be properly managed and fully informed of the disease.

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