

Referral patterns, treatment management and visual outcome in keratoconus

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Abstract

Purpose/Method To determine the patterns of referral, the hospital-based management and the subjective degree of visual success achieved by keratoconics attending the Corneal Service at Sunderland Eye Infirmary, England, a specifically designed anonymous questionnaire was mailed to 160 keratoconus subjects.

Results The following responses were obtained from 97 (61%) respondents: mean age of first referral (22.88 ± 8.36 years); gender (male:female ratio 3:1); source of initial referral (optometrists 90%); visual symptoms experienced before referral (46% complained of blurred/distorted vision); and the type of visual correction before referral (70% wore a refractive correction: 72% spectacles, 28% contact lenses). Initial post-referral treatment included bilateral contact lens fitting (65%), monocular contact lens correction (26%), penetrating keratoplasty (8%) and no intervention (1%). Two-thirds of subjects were presently wearing contact lenses (67%), with the mean period of successful lens wear being 8.26 ± 7.92 years, and 31% had undergone penetrating keratoplasty (PK) since initial referral. A subjective evaluation of visual acuity with contact lenses was made by respondents using a visual analogue scale. The mean value was 86.86 ± 15.25 (range 50–100 mm), with 50 mm representing moderate visual acuity. Of those who underwent PK the mean subjective evaluation of the change in post-operative vision compared with pre-operatively was 82.41 ± 26.57 (range 1–100 mm), with 50 mm representing no visual change. The majority of subjects therefore noted a moderate to marked visual improvement following PK.

Conclusion In this fairly young group of patients, predominantly male, the majority wore a refractive correction for approximately 10 years before referral to the Hospital Eye Service (HES). Most referrals were initiated by optometrists. On initial HES assessment the treatment for two-thirds was bilateral contact lens fitting; only a quarter were fitted monocularly. However, by the time of this

study only a tenth retained monocular contact lens correction, with the mean length of time from diagnosis to bilateral contact lens wearing being approximately 5½ years. The progressive nature of this disease is further evidenced by the large number of contact lens sets required by patients (approximately 5) and by the number of patients ($n = 22$) requiring PK, the mean length of time from diagnosis to corneal graft being approximately 8½ years.

Key words Contact lenses, Keratoconus, Penetrating keratoplasty, Visual acuity

The management of keratoconus usually includes refractive error correction, initially with spectacles, thereafter by contact lenses; eventually surgical therapy such as penetrating keratoplasty (PK) may be required.¹ Keratoconus has consequences that are not only medical and clinical but also social, such as employment restrictions which can mean individuals are prevented from following their chosen career or have to discontinue it.² Kennedy *et al.*³ found over a period of 48 years an average annual incidence of 2 cases per 100 000 and a prevalence rate of 54.5 per 100 000 population. With the advent of computerised videokeratography (CVK) it is possible to detect subclinical keratoconus which may or may not progress to overt keratoconus.⁴ Therefore the relative incidence may apparently increase with the availability of CVK analysis and consequently increase demands on medical services. In the United Kingdom (UK) keratoconic patients generally have their contact lenses provided within the National Health Service as part of their long-term treatment.

A specifically designed questionnaire was mailed to 160 individuals who were attending Sunderland Eye Infirmary, Sunderland (UK) with a diagnosis of keratoconus, in order to ascertain their biometric data, the management of their keratoconus and the degree of visual success experienced.

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Subjects and methods

To establish the pattern of contact lens wear, visual rehabilitation after PK and the subjective assessment of keratoconus management, a questionnaire was designed and forwarded to 160 patients with diagnosed keratoconus who were attending the corneal service at Sunderland Eye Infirmary.

The questionnaire consisted of 31 questions. Twenty single-response questions elicited information on age, gender, occupation, age of first referral to the HES, source of initial referral, current type of contact lens, time difference between monocular and binocular fitting, whether presenting contact lens required refitting, number of contact lens sets fitted since referral, mean age of present contact lenses, maximum contact lens survival, number of days per week contact lenses worn, number of hours per day contact lenses worn, whether presently a spectacle wearer, age at PK, length of time contact lenses worn prior to PK, length of time off work after PK, contact lens comfort pre- versus post-PK, and PK visual expectations. Seven multiple-response questions concerned presenting visual problems, type of visual correction before referral, initial treatment after referral, type of contact lenses worn since referral, current contact lens problems, type of optical correction worn after PK and medical history. There were also four questions with visual analogue scales.

Visual analogue scales provide a simple, reliable technique for measuring subjective experience.⁵ They typically consist of a 100 mm line anchored at both ends with words descriptive of the maximal and minimal extremes of the dimension being measured.⁵ In this study visual analogue scales (Fig. 1), anchored with adjectival descriptions, were used in which the respondent placed a vertical mark on a 100 mm horizontal line for subjective evaluation of four questions relating to: vision, visual acuity with spectacles, visual acuity with contact lenses, and the difference in visual acuity attained after PK. On three of these analogue scales, 0 represented 'cannot see anything at all', 50 mm represented 'moderate vision' and 100 mm represented 'can see everything I need to'. For the final question, 0 represented 'very much worse' than before the operation, 50 mm 'no difference' and 100 mm 'very much better' than before PK. Responses were calculated by accurately measuring, in millimetres (mm), the position of the patient's mark on each scale.

Results

Ninety-seven (97) questionnaires were returned, giving a response rate of 61% (97/160). The responses are analysed below.

Cannot see anything at all Moderate Can see everything I need to

Fig. 1. Visual analogue scale.

Age, gender, occupation

The mean age of the patients at first referral to hospital eye services (HES) was 22.88 ± 8.36 years (range 8–60 years); the mean age on receiving the questionnaire was 31.1 ± 10.3 years (range 11–64 years). The male:female ratio was 3:1. The predominant occupation was clerical in nature (21%); 19% were classified as manual workers, 14% were students, 10% were unemployed, 9% were teachers, 7% were engineers and 20% had miscellaneous occupations.

Pre-referral features and method of referral to HES

Hospital referral was mainly initiated by optometrists (90%), and the visual problems experienced by the patients when initially referred are shown in Table 1. The most common complaint was blurred/distorted vision (46%). Only 30% did not wear a visual correction prior to referral. Of the 70% wearing a refractive correction 72% (49/68) wore spectacles and 28% (19/68) wore contact lenses. The mean period wearing a refractive correction was 8.15 ± 8.04 years (range 0.5–36 years) for spectacles and 11.68 ± 6.23 years (range 1–25 years) for contact lenses. Of the 19 contact lens wearers 42% (8/19) wore rigid gas permeable (RGP) lenses, 37% (7/19) wore hard (PMMA) lenses and 21% (4/19) wore soft contact lenses.

Initial hospital treatment

Of the respondents who wore contact lenses prior to referral, 94% were refitted with a different contact lens design after referral to the hospital and 93.3% replied that this refit solved their visual problem. The most frequently cited initial treatment was contact lens fitting to both eyes (63/97, 65%). Monocular correction was the initial treatment in 26% of subjects (25/97), of whom 64% subsequently had the fellow eye fitted (16/25), with a lag period of 68.1 ± 58 months (range 1–156 months). Only 8% (8/97) of subjects were listed for PK at the first HES assessment and 1% were reviewed without alteration to management.

Type of contact lens worn

Of 194 eyes (97 subjects), 121 eyes (62%) were presently wearing contact lenses, and in total 65 of the 97 patients (67%) were current contact lens wearers, i.e. 9 patients wore only one contact lens. Four subjects had discontinued contact lens wear, 3 of whom were presently utilising spectacles with good subjective visual acuity (100, 100 and 92 mm on the visual analogue scale).

Table 1. Presenting visual problems

Visual problem	No.	%
Blurred vision	75	47
Poor VA with spectacles	42	26
Sensitivity to light	28	17
Frequent refractive changes	17	10

Table 2. Current or former contact lens type

Contact lens type	Right eye	Left eye
Hard	9 (11%)	7 (8.4%)
Rigid gas permeable	69 (85%)	69 (83.1%)
Softperm	3 (4%)	6 (7.3%)
Soft	0	1 (1.2%)
Total	81	83

However, one respondent reported using no form of visual correction with poor subjective vision (32 mm on the visual analogue scale). The types of contact lenses the subjects presently or formerly (if discontinued) wore are shown in Table 2. The reason for refitting in 32% of patients (31/97) was discomfort in 42%, whereas 39% were refitted on advice of a practitioner and 19% due to poor visual acuity.

Mean age of present contact lens and the number of sets worn

The mean age of the contact lens presently worn was 11.64 ± 12.22 months (range 0–72 months) for right eyes ($n = 65$), and 12.30 ± 13.22 months (range 1–60 months) for left eyes ($n = 64$). The approximate number of lens sets patients had worn ranged from 1 to 36, with a mean of 5.17 ± 5.50 ($n = 65$), whilst 11 patients noted there were too many to remember! The mean time period of contact lens survival was 21.4 ± 17.01 months, range 4–84 months ($n = 65$). The number of years of successful contact lens wear was 8.26 ± 7.92 years (range 0.5–32 years). Subjects rated each contact-lens-associated symptom they experienced as either a major or minor problem. Major problems related to vision, discomfort, redness and the contact lens falling out were noted in 20% of eyes. Minor problems were reported by 82% of eyes.

Contact lens wearing schedule and visual acuity

Only 20% of subjects wore their contact lenses for less than 8 h a day (56% wore contact lenses for over 12 h) and 62% of subjects wore their contact lenses 7 days a week (8% for 3 days or less). Using visual analogue scales, the mean subjective evaluation of the respondents' vision without correction was 30.32 ± 24.79 , range 0–95 mm ($n = 93$); mean visual acuity with spectacles was 52.73 ± 31.37 , range 0–100 mm ($n = 41$); and the mean visual acuity with contact lenses was 86.86 ± 15.25 , range 50–100 mm ($n = 69$). For all three scales 0 represented 'cannot see anything at all', moderate vision was represented by 50 mm, and 100 mm represented 'can see everything I need to'.

Penetrating keratoplasty

Thirty patients (31%) underwent a PK; 37 procedures were carried out (i.e. 7 patients had bilateral grafts). Sixteen (43%) of these subjects currently wore spectacles, 14 (38%) required no visual correction and 7 (19%) wore

post-graft contact lenses. Of those wearing contact lenses 4 subjects found the resumption of contact lens wearing easier and 3 noted no difference in comfort. The mean age at which PK was performed was 30.41 ± 9.59 years (range 16–60 years) and the mean length of time taken off work due to the operation was 6.47 ± 3.48 weeks (range 4–13 weeks). The mean length of time from diagnosis to PK was 8.65 ± 8.59 years (range 0–24 years). The mean survival of the grafts was 3.85 ± 5.17 years (range 0.5–25 years). The main reasons for patients undergoing PK were contact lens discomfort (67.5%), the contact lens falling out (19%) and poor contact lens visual acuity (VA) (13.5%).

The result of the operation was better than expected for 19 (51.4%) eyes, for 6 (16.2%) was as expected, and 10 (27%) eyes results were worse than patient expectations. Two eyes (5.4%) still had sutures *in situ* and therefore the final visual result was not known (as some subjects had bilateral PKs, eyes are quoted not individuals). Using a visual analogue scale the mean subjective evaluation of the visual improvement post-operatively was 82.41 ± 26.57 mm (range 1–100 mm), where 50 mm represented no difference. The majority of subjects therefore noted a moderate to marked visual improvement following PK.

Skin disorders, hay fever, asthma and food allergies

Subjects were asked whether they had a history of acne, eczema, psoriasis, hay fever symptoms, asthma or allergies. In total 34% recorded a skin disorder, 9% (9/97) had a history of acne, 19.6% (19/97) a history of eczema and 5% (5/97) exhibited psoriasis. Two patients reported a combination of skin conditions: eczema and acne; and eczema and psoriasis. Hay fever symptoms were reported by 36%, 21% suffered from asthma, and 4% reported food allergies. Nine patients suffered a combination of asthma and hay fever and one patient suffered a combination of asthma and food allergy.

Eye rubbing and inheritance

Eye rubbing has been reported as a possible causative factor in keratoconus development. In this group 15% reported that they 'eye rubbed' a great deal, 33% a fair amount, 40% eye rubbed sometimes and only 11% never rubbed their eyes. With regard to inheritance 5% reported a family member exhibiting keratoconus (aunt, $n = 1$; brother/sister, $n = 2$; cousin, $n = 2$).

Discussion

One of the diagnostic signs of keratoconus, even before biomicroscopically visible signs such as Vogt's striae, is an irregular retinoscopy reflex (scissors) and/or distorted keratometry mires. It is therefore not surprising that in this study optometrists were the main source of initial referral (90%) or that the most common visual complaint was that of blurred or distorted vision (46%). Keratoconus usually develops during teenage years, with

a mean age of onset of 16 years of age.⁶ In the present study the mean age of diagnosis by the HES was somewhat older, at 22.88 years, but with a large standard deviation of 8.36 years. The youngest patient was 8 years old, although keratoconus has previously been reported in children as young as 6 years old.⁷ The oldest patient at primary diagnosis was 60 years old; however, keratoconus rarely develops beyond 30 years of age.⁸ There are varying views on the significance of gender. Initially a greater preponderance of female sufferers was reported;⁹ however, recent investigations report a higher incidence in males.¹⁰ In the present study there was a pronounced male preponderance by a factor of 3:1.

As initial treatment in this study 65% of subjects had bilateral contact lenses fitted and 26% had monocular contact lens correction. In a large retrospective review of 295 keratoconics,¹¹ 33% wore unilateral contact lenses; however, Rabinowitz *et al.*⁴ state that keratoconus is rarely unilateral and even in cases with no clinical signs in the contralateral eye there is a suspiciously high positive inferior/superior steepening on CVK. Of those who were initially fitted monocularly with a contact lens in this study, 36% were later fitted with a contralateral contact lens, the lag time to fit the fellow eye being approximately 5.5 years. Therefore at the time of the present study only 16 patients were uni-ocularly corrected by a contact lens. A similar lag time frame has been reported elsewhere.¹² The number of lens sets fitted per patient, approximately 5, was not unsurprising as the active state of keratoconus progression is usually about 5–7 years in duration,^{12–14} after which there may be a period of stability. The mean age of the patients' contact lenses at the time of the study was approximately 1 year (11.64 months for the right eye and 12.30 months for the left) and the mean contact lens survival time was approximately 2 years (21.4 months). During the active progression stage, usually 1 to 2 years from diagnosis, contact lenses may have to be altered several times.

Visual analogue scales have been used as a method for assessing subjective responses for over 70 years.¹⁵ Some difficulties can be encountered with such scales; however, they are thought to be no weaker than other comparable psychological measures and have advantages such as graduation of response and ease of use.⁵ Visual analogue scales were used in this study to subjectively evaluate the subject's vision or VA. Although, not surprisingly, the respondents subjectively rated their level of VA higher with the contact lens (86.86 mm) than with the spectacles (52.73 mm), where 50 mm represented 'moderate' visual acuity and 100 mm represented 'can see everything I need to', 44% of subjects were still able to maintain a 'moderate' level of VA. Although most patients with significant keratoconus tend to wear a contact lens correction (80% of subjects wore their contact lenses for more than 8 h per day; 64% for 7 days a week), spectacles even with their inherent optical limitations should not be overlooked in keratoconus, especially as the subjective rating of the subjects' unaided vision was poor (30.32 mm). Following PK the majority of subjects noted a moderate to marked

visual improvement (the mean subjective evaluation of the visual improvement post-operatively was 82.41 mm).

As the questionnaire was entirely anonymous to encourage unbiased patient response, unfortunately no differences between the responders and non-responders could be identified in relation to age, gender, quality of VA or mode of treatment, and therefore we are unable to conclude that non-responders would have produced the same data as responders. However, the questionnaire was only sent to 'active' hospital files, i.e. patients who had been seen within the previous 3 years, and therefore could reasonably be considered a representative sample of keratoconics attending the HES.

The aetiology and pathogenesis of keratoconus is unknown but may be associated with a variety of factors such as eye rubbing, atopic disease, connective tissue disease (Ehlers–Danlos syndrome, Reiger's syndrome, Marfan's syndrome), tapetoretinal degeneration, Down's syndrome and other genetic factors.^{16–18} In this study 15% of subjects admitted to rubbing their eyes a great deal and 33% a fair amount; only 11% reported that they never rubbed their eyes. Karseras and Rubin¹⁹ reported that eye rubbing was a dominant aetiological factor in two-thirds of patients. Whilst there are various genetic theories for the inheritance of keratoconus,²⁰ there is support within the literature^{21–33} for the hypothesis that in certain cases there is an autosomal dominant mode of inheritance. In our study 5% reported a family member being diagnosed with keratoconus, similar to the 6% reported by Rabinowitz *et al.*²⁴ In total 34% of respondents in our study exhibited a skin disorder and 21% suffered from asthma. In a study²⁵ of 182 keratoconics, atopy was identified in 35% in comparison with 12% in a matched control group. Gasset *et al.*²⁶ also noted 35.7% of keratoconic patients suffered from asthma.

A relationship between the wearing of rigid contact lenses and the development of keratoconus^{27–29} has also been postulated. In this study only 15 of 97 (15%) wore rigid contact lenses prior to their referral to the HES; however, it is difficult to elicit a convincing cause–effect relationship, as by its very nature, irregular corneal shape may lead to the wearing of rigid contact lenses.³⁰ The number of patients wearing hard contact lenses may appear high (7/19) in comparison with RGP (8/19); however, the subjective reporting of whether PMMA or RGP lenses were worn may be prone to error. In this study, even amongst the successful contact lens wearers, 82% experienced minor problems with poor VA, discomfort, hyperaemia and the contact lens falling out. However, 80% of eyes were corrected by contact lenses for more than 8 hours a day and approximately 64% wore the lenses 7 days a week. Discomfort was the major reason for contact lens refitting (42%), with 19% being refitted due to poor VA. Most of the patients who wore contact lenses prior to initial referral (94%) were refitted and 93.3% of those recorded that this refit did solve their presenting visual or discomfort problem.

In this study, for 63% of patients the present management of their keratoconus was contact lens wear;

however, 31% had undergone PK. Other studies³¹⁻³⁷ report from 10% to 31% of subjects undergoing PK. The need for contact lens wear after graft surgery has been reported to be as high as 60%;³⁸ however, in the present study only 19% required contact lenses post-operatively to achieve adequate VA. Included in the 31% who underwent PK were 8 patients who were listed for surgery at initial HES presentation; for the remainder, the mean length of time from diagnosis to PK was 46.2 months. Smiddy *et al.*³⁸ reported that 31% of subjects with keratoconus required PK after a mean period of 38.4 months of contact lens wear. The mean age at the time of surgery in this study was approximately 30 years, with the average patient taking 6 weeks' absence from work. Only 4 patients discontinued contact lens wear without proceeding to surgery, and of these only one was not able to receive satisfactory VA utilising spectacles.

The primary reason for patients undergoing PK in the present study was significant discomfort with contact lens wear (67.5%), poor VA with the contact lens (13.5%) and problems with contact lens falling out (19%). In another series³⁹ the main reason for PK was poor VA (43%) with contact lens intolerance second (32%). The corneal graft survival rate from a representative series⁴⁰ was 91% at 1 year, 72% at 5 years and 69% at 7 years; another⁴¹ noted 89% survival at the 1 year stage. The mean survival of the grafts was 3.85 ± 5.17 years in this study. It is possible for keratoconus to re-occur following PK,⁴² or for regrafting to be necessary due to corneal graft failure. As yet, in this group of 37 eyes, there has been no re-occurrence; however, one graft (2.7%) did decompensate. Keratoconus is now the most common indication for PK⁴³ in most series of corneal transplantation.

Conclusion

This study examined keratoconus treatment and management within the HES from a patient's perspective. The vast majority of subjects were referred by optometrists, they were predominantly male, and had a mean age at diagnosis of 23 years. Most subjects were initially fitted with bilateral contact lenses (65%) and this enabled the majority to achieve a good quality of corrected VA. At the time of the study 90% of subjects exhibited features of bilateral keratoconus and 5% provided a family history of the disease. Approximately one-third of subjects proceeded to PK at a mean time of 8½ years from diagnosis, with only 19% requiring contact lens correction after PK. The preferred initial management of keratoconus remains the fitting of appropriate contact lenses; however, with approximately 5 contact lens sets per subject over a mean treatment time of 8 years, this entails a significant continuing commitment for the HES.

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