Scleral Lenses Reduce the Need for Corneal Transplants in Severe Keratoconus

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• PURPOSE: To investigate the success and failure rates of scleral lens correction in severe keratoconus.

• DESIGN: Retrospective case series.

• METHODS: Study population comprised patients with keratoconus who attended the keratoconus clinic at the Antwerp University Hospital, Belgium, between January 1, 2010, and December 31, 2014. The included eyes had maximal keratometry values ≥ 70 diopters, as measured using the sagittal curvature map that was obtained by Scheimpflug tomography. The exclusion criteria included amblyopia, mental retardation, and concomitant ocular disease that limited their visual potential.

• RESULTS: Scleral lens fitting was proposed for the 75 eyes included in the study. Eight eyes underwent transplant surgery because of insufficient visual acuity with the lenses, lens intolerance, and issues with handling the lenses. Twelve eyes did not have lenses fitted because of good visual acuity in the other eye or a contraindication for lens wear. Three eyes were successfully fitted with corneal or hybrid lens wear. Scleral lenses were prescribed in 51 of 75 eyes. The mean gain in visual acuity (lens vs spectacle-corrected visual acuity) was 0.54 ± 0.18 (decimal fraction, Snellen eye chart). Seven eyes were lost to follow-up, 4 eyes abandoned wearing the scleral lens because of an inability to handle the lenses, and 40 eyes wore the lenses at their last follow-up visit, with a mean follow-up interval of 30.15 ± 12.83 months.

• CONCLUSIONS: Forty of the 51 eyes with severe keratoconus that would otherwise have undergone transplant surgery were successfully treated with long-term scleral lens wear. In this way, the indication for keratoplasty was more than halved in our keratoconus population. (Am J Ophthalmol 2018;185:43–47. © 2018 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license [http://creativecommons.org/licenses/by-nc-nd/4.0/].)

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For more than 50 years, the traditional treatment paradigm for mild stages of keratoconus has consisted of glasses, with rigid corneal contact lenses being used to treat moderate forms and penetrating keratoplasty for more advanced cases. The indication for surgery was typically intolerance to contact lenses or a limitation of the best lens-corrected visual acuity.

New surgical techniques have led to major shifts in the traditional treatment paradigm. Corneal collagen cross-linking (CXL) is currently routinely applied to stabilize progressive ectatic disease. In Europe, this technique has been available for more than 10 years. The effectiveness of CXL in reducing the need for corneal transplants for keratoconus was described by Godefrooij and associates in a Dutch study.1 Refractive surgery has also become part of keratoconus management in the form of intracorneal ring segments, phakic lens implants, and laser surgery. These forms of non–transplant surgery for keratoconus are indicated in mild and moderate keratoconus but not in advanced stages of the disease with corneal scarring.2 In such severe cases, corneal transplantation appears to be the only option. Many authors currently promote the choice of deep anterior lamellar keratoplasty (DALK) over penetrating keratoplasty (PK) as a first-line corneal transplant procedure. Gomes and a group of Global Delphi Panel of Keratoconus panelists concluded that contact lens intolerance was the leading indication for DALK surgery, while corneal scarring was the most important indication for proceeding to PK.3 Conclusive evidence demonstrating the superiority of DALK over PK, however, is still lacking, and visual and refractive results remain equally variable and unpredictable.4

Contact lens technology has experienced major advances as well, particularly over the last 5 years, with an emphasis on scleral lenses that rest on the conjunctiva and vault over both the cornea and limbus.5 Scleral lenses made of gas-permeable materials were introduced more than 25 years ago, but their use was limited to a handful of highly specialized centers around the world owing to the complexity of fitting these lenses.5–8 The advantages of scleral lenses compared to corneal lenses in the correction of irregular corneas are numerous: scleral lenses provide a more stable and better-centered fit; they do not touch the cornea, so scar formation is not exacerbated; and they are more comfortable, thereby improving quality of life for patients.9–12 Scleral lenses can be adapted to fit almost any degree of corneal ectasia by changing the vault of the optic zone. Deloss
and associates published that even eyes with advanced corneal ectasia can be successfully fitted with scleral lenses (PROSE).\textsuperscript{11,12} These authors concluded that the visual outcome for stage 4 ectasia was better and more rapid with lens correction compared to keratoplasty. From our in-practice experience, we can confirm that scleral lenses take the nonsurgical management of severe keratoconus cases to a new level. Specifically, without scleral lenses, many more patients require transplant surgery. To more comprehensively investigate this clinical finding, we selected a subgroup of patients with extremely advanced keratoconus. From our keratoconus population, a cohort of eyes was identified with maximal keratometry (K max) values of $\geq 70$ diopters (D) on Scheimpflug tomography. The success or failure of scleral lens correction in this group of eyes is described in this manuscript.

### METHODS

**STUDY DESIGN:** This study was designed as a retrospective case series of patients attending a dedicated keratoconus clinic in a university hospital setting over a 5-year period (January 1, 2010 through December 31, 2014). Ethical approval for the study and data accumulation was obtained from the institutional ethics committee.

**PATIENT SELECTION:** All patients with keratoconus were identified using the electronic agenda system (Ultra-Genda, Destelbergen, Belgium). The patients were diagnosed with keratoconus on using a slit-lamp examination and Scheimpflug tomography (Pentacam; Oculus Optikgerate GmbH, Wetzlar, Germany). The patients with clinical evidence of keratoconus and/or consistent tomographic evidence of a localized area of steepening in the anterior curvature, lower pachymetry, and increased posterior elevation were diagnosed with keratoconus. Based on the Pentacam database, all eyes with K max $\geq 70$ D on the sagittal curvature map were included in our study group. A trial fitting with contact lenses was offered to all patients regardless of the presence and degree of corneal scarring.

**CONTACT LENS FITTING:** Contact lens fitting was performed by specialist optometrists in collaboration with an ophthalmologist. The lens design used was the mini-MISA or the larger MISA scleral lens, both of which were provided by Microlens (Arnhem, The Netherlands). The lenses are made of highly gas-permeable materials and have diameters ranging from 16.5 to 17.5 mm for the mini-MISA lens and 18.0 mm or more for the MISA lens. The clearance of the lens over the cornea can vary by patient, but the lens is refitted if it touches the apex of the cornea at any of the follow-up visits.

### RESULTS

A TOTAL OF 846 PATIENTS DIAGNOSED WITH KERATOCONUS attended the clinic from January 1, 2010, to December 31, 2014. There was a predominance of male subjects in this population, 545 (64.42%), vs 301 (35.58%) female subjects, and the mean age of the population was 36.71 ± 14.72 years.

Of the total keratoconus cohort, 86 of the 1692 eyes (5.1%) had a K max value $\geq 70$ D. Eleven eyes were subsequently excluded from the study for the following reasons: 2 were amblyopic; 6 were in patients with Down syndrome or other forms of mental retardation; 2 eyes in a single patient showed retinal dystrophy, limiting visual potential; and 1 eye was diagnosed with cataract. Seventy-five eyes from 63 patients were ultimately included. There were 53 male and 10 female patients, with a mean age of 36.70 ± 14.73 years. Thirty-three were left eyes and 42 were right eyes.

K max varied from 70.00 to 130.0 D, with a mean value of 81.70 ± 11.05 D. Pachymetry at the thinnest point ranged from 186 to 478 μm, with a mean of 360.33 ± 67.44 μm. Slit-lamp examinations showed macroscopic deformation and scarring in 24 eyes (K max 89.65 ± 13.82 D), Vogt striae and scarring in 6 (K max 82.99 ± 7.11 D), scarring only in 12 eyes (K max 80.52 ± 8.31 D), Vogt striae only in 15 (K max 78.25 ± 5.71 D), and clear corneas in 18 eyes (K max 74.34 ± 3.76 D).

Of the 75 eyes that were included, 9 could not be fitted successfully with corrective contact lenses. One eye developed hydrops, while the other 8 cases underwent transplant surgery (4 DALK: K max 84.97 ± 9.41 D; 4 PK: K max 90.23 ± 10.68 D). All eyes that underwent keratoplasty displayed obvious corneal scarring. The reason for surgery was inadequate improvement of vision in 5 eyes, intolerance to scleral lens wear in 2 eyes, and inability to manipulate the scleral lens in 1 eye.

Another 15 of the remaining cohort of 66 eyes did not have scleral lenses fitted. A hybrid lens (Synergeyes, Carlsbad, California, USA) was used in 1 eye: a visual acuity of 0.8 was achieved. Two eyes were comfortable in corneal lens wear and were refitted with corneal contact lenses (Rose K; Menicon, Nagoya, Japan). These eyes exhibited a mean visual acuity 0.65. With respect to the final 12 eyes in the cohort, the patients chose to undergo neither contact lens correction nor corneal transplantation. Nine patients preferred not to have their vision corrected in their worse eye, given that the better eye had a best-corrected visual acuity (BCVA) that was satisfactory for the patient (mean best spectacle-corrected visual acuity [BSCVA] 0.9 decimal fraction, Snellen eye chart). For 1 patient, further contact lens wear was contraindicated for both eyes owing to a severe allergy; fortunately, the patient had an acceptable visual acuity with glasses. One patient experienced a great
deal of anxiety and did not wish to use any type of contact lens.

Scleral lenses were ultimately prescribed for 51 eyes: 45 mini scleral lenses (K max 79.36 \pm 7.38 D) and 6 full scleral lenses (K max 87.75 \pm 12.44 D). In 16 eyes, scleral lenses were the first choice for the lens trial because of the advanced nature of the cone. For the remaining 35 eyes, the patients had experienced the typical problems of intolerance and instability of corneal lens fitting, which can be expected for such high degrees of corneal steepening. The mean BSCVA for this group of 51 eyes was 0.13 \pm 0.14, and BCVA with the scleral lenses was 0.66 \pm 0.18. The mean gain in visual acuity (scleral lens vs spectacle-corrected acuity) amounted to 0.54 \pm 0.18 (decimal fraction, Snellen eye chart).

Of the 51 eyes, 7 were lost to follow-up. For 4 of the eyes, the patients abandoned scleral lens wear because of difficulties in handling the lens. For 40 eyes, the patients were still wearing the lenses at the last follow-up visit, with a mean follow-up interval of 30.15 \pm 12.83 months. All of the eyes were followed for at least 6 months.

### DISCUSSION

**THE KERATOCONUS CLINIC OF THE DEPARTMENT OF OPHTHALMOLOGY AT ANWERP UNIVERSITY HOSPITAL** is a tertiary referral center that provides service on both a regional and national level. Based on an estimated prevalence of keratoconus in the general population of 54 per 100,000, the number of patients in Belgium can be calculated at approximately 5940. The 846 individual patients who visited the keratoconus clinic over a 5-year period therefore represent approximately 14% of the Belgian keratoconus population. A well-established team of optometrists and ophthalmologists closely collaborate to offer all specialty lens options, thereby providing patients with optimal visual acuity and maximizing patient comfort. Penetrating and anterior lamellar keratoplasty are offered to patients when specialty lens correction cannot achieve the double goal of satisfactory visual acuity and a comfortable wearing time of at least 10–12 hours per day.

In view of the technological advances in refractive and transplant surgery, several reviews have been published that have rewritten the treatment paradigm for keratoconus. McGhee and associates described a comprehensive management paradigm for moderate-to-severe keratoconus with poor BCVA. For the treatment of mild-to-moderate cases, contact lenses are classified as a conservative route of treatment and are briefly mentioned. Parker and associates reviewed the treatment options for advanced keratoconus, although contact lenses are only discussed in the context of cases where lens wear is necessary to supplement transplant or keratorefractive surgery. With currently available lens designs, especially scleral lenses, we have demonstrated that there is no technical limit for which keratoconus cases can successfully be treated with lenses. This study indicates that even eyes with very severe scarring and deformation can achieve functional visual acuities. Only a trial fitting is required to allow the patient to judge whether a satisfactory result is obtained. Unfortunately, there is no clinically adequate classification system for grading keratoconus. To ensure that only the most advanced cases of keratoconus were included in this study, we arbitrarily defined a cut-off value for K max as \( \geq 70 \) D. The severity of ectatic disease in the study group is highlighted by the low pachymetry values (mean value of 360.33 \pm 67.44 \mu m) and the presence of scarring (42 of 75 eyes).

Scleral lenses were prescribed in 51 out of a total of 75 eyes in the study cohort. For 40 eyes, scleral lenses were proven to be an effective optical correction. This conclusion can be drawn because these patients continued wearing the lens for a mean follow-up interval of 30 months. Of note, 9 patients felt no need for correction of their worst eye, given that their better eye had very good uncorrected or spectacle-corrected visual acuity (mean value 0.9 decimal fraction). These eyes illustrate to an extreme degree the asymmetric nature of keratoconus. None of these 9 patients were visually disturbed by this enormous interocular difference; and from the patient’s perspective, there was no need for correction of the worse eye.

Before scleral lenses became widely available, the 40 corneal contact lens–intolerant eyes would have undergone transplant surgery; instead, these patients are now successfully wearing scleral lenses. The interest in the literature for new specialty lenses has increased, but more awareness must be generated. Even in the most recent reviews on modern treatment paradigms for keratoconus, scleral lenses are looked upon as unpractical and costly. It has been suggested that tear exchange is reduced when compared to corneal lenses and that the absence of long-term studies on the physiological aspects of scleral lens wear warrant caution. As a consequence, patients are not always offered the choice for scleral or other specialty lenses by their ophthalmologists. Lens tolerance is difficult to assess objectively and depends on the skill and diligence of the prescriber, the disposition of the patient, and the type of lenses available for use. It is clear that the manner in which the choice between lenses and keratoplasty is
presented to the patient plays a major role in the decision process of the ophthalmologist and patient. In an editorial entitled “Keratoconus: Why and when do we turn to surgical therapy?” Mannis eloquently notes why patients should not lightly make the choice for keratoplasty: “In truth, the patient who opts for a corneal transplant trades one set of problems for another. One gives up contact lens intolerance and/or poor visual acuity or poor qualitative vision for better post-PK acuity. With this, however, comes the risk of complications, such as high corneal astigmatism, anisometropia, graft rejection, infection, cataract, glaucoma, and ocular surface disease, all of which may complicate the course of a superlatively performed graft.”20

Anterior lamellar keratoplasty has solved some of these issues but, specifically with respect to vision and refraction, DALK does not score better than penetrating keratoplasty.4 Patients should be fully informed regarding the risk of requiring additional refractive surgery and/or contact lens correction to correct for postkeratoplasty astigmatism, anisometropia. A major advantage of scleral lens wear vs keratoplasty is its total reversibility. If the patient does not find that lens wear provides good visual acuity and high quality of life, lens wear can be abandoned and surgery performed.

In the study group of 75 eyes with K max ≥ 70 D, 8 eyes could not be successfully fitted with scleral lenses (inadequate improvement of vision: n = 5; intolerance: n = 2; inability to manipulate the scleral lens n = 1); these patients subsequently underwent transplant surgery. Of the total cohort of 846 patients who visited the keratoconus clinic over the 5-year study period, 28 eyes of 25 patients underwent lamellar or penetrating keratoplasty. Of these 28 eyes, 8 had a K max on Pentacam analysis of ≥ 70 D, and 20 eyes had a K max < 70 D. If the 40 eyes with proven, long-term successful scleral lens wear had been included in the transplantation group, the incidence of transplantation in our keratoconus population would have more than doubled to approximately 4%. Currently, the rate of corneal transplantation in our population is 28 of 1692 eyes (1.65%) over a 5-year period.

We acknowledge that our treatment strategy for keratoconus is strongly centered on specialty contact lenses. Other centers may take a different view on surgery and present it, and accurately so, as a solution that can provide good visual acuity all day without the hassle and risks associated with contact lens wear. Therefore, our results may not be representative for all practices. However, despite satisfactory visual outcome measures obtained after keratoplasty, vision-related quality of life in patients with keratoconus too often remains impaired.21 We inform patients extensively regarding all therapeutic options so that they are assured that the therapeutic approach will be tailored to their individual physical and psychological needs and to their specific social and professional conditions.

In conclusion, the use of specialty lenses, specifically scleral lenses, has led to a decreased need for corneal transplants in severe keratoconus as defined by K max ≥ 70 D.

REFERENCES