

# Scleral Lenses for the Visual Rehabilitation of Keratoconus:

## A case report of a young dancer with red eyes

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### ABSTRACT

**Background:** Keratoconus is a progressive corneal disease that causes irregular astigmatism and results in blurred or distorted vision. In advanced cases the condition can result in corneal scarring that further limits vision. Medical management with scleral lenses is a viable treatment for vision rehabilitation. **Case Report:** A 24-year-old black male presented for a comprehensive eye exam and evaluation of his corneal RGP lenses. His examination revealed limited best visual acuity in spectacles, high irregular astigmatism on topography and apical thinning with striae of both corneas. He was diagnosed with bilateral keratoconus with minor contact lens induced corneal warpage and was treated with scleral lenses. Trial lens fitting was performed and the first pair of ordered lenses achieved excellent vision and comfort. He was followed for three months with no adverse ocular outcomes and no changes were needed to the lenses. **Discussion:** Keratoconus is caused by a combination of genetic and environment factors. Localized stromal thinning causes irregular astigmatism and higher and lower optical aberrations. As the disease progresses traditional spectacle and soft contact lens options can be ineffective in providing functional vision and specialty lens fitting can be indicated. Specialty lens options include corneal RGP lenses, custom soft lenses, piggyback systems, hybrid designs and scleral contact lenses. Intolerance to contact lenses or excessive corneal scarring may require a corneal transplant. Other surgical options include corneal cross-linking and intra-stromal corneal ring segments. Scleral contact lenses corrected our patient's problem of dislocation, provided improved comfort and significant improved his vision. Since he was successfully fit with good comfort and vision, no surgical interventions were required. **Conclusion:** Scleral lenses provide excellent vision correction and superior comfort compared to traditional contact lens options for the management of keratoconus. Scleral lenses should be considered before patients fail in other lens modalities since it may prevent or delay the need for surgical intervention. Fitting of the lenses can be simple and patients can easily adapt to the lenses.

**Keywords:** Keratoconus -- Scleral Contact Lenses --- Irregular Astigmatism --- Ectasia

# Sclearal Lenses for the Visual Rehabilitation of Keratoconus: A case report of a young dancer with red eyes

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## INTRODUCTION

**K**eratoconus is a bilateral corneal disease in which stromal thinning creates an irregular protrusion or “cone” shape of the cornea. The progressive irregular shape of the cornea results in blurred or distorted vision and can result in scarring of the tissue. The condition is typically bilateral but often presents asymmetrically. Prevalence is reported as high as 1 in 2000<sup>[1]</sup> and the decrease in vision caused can significantly affect quality of life as it is typically diagnosed during peak education, income-earning and child rearing years<sup>[2]</sup>.

The following case report summarizes the diagnosis and management of a 24-year-old male with keratoconus and his successful visual rehabilitation with scleral contact lenses.

## CASE REPORT

### *Initial Exam and CL fitting*

24-year-old black male SH presented to a private practice office on 4/4/2014 for a comprehensive exam and contact lens evaluation. SH reported good vision and comfort in his current rigid gas permeable (RGP) lenses but had noticed a slight decrease in comfort in the left eye for 3-4 weeks. His current lenses were about 2 years old and he felt he needed a new pair. He had previously diagnosed with Keratoconus in both eyes, which had been progressing for the past few years and he had no history of ocular surgeries. He was not taking any medications and had no known systemic medical conditions. The patient’s family ocular and systemic history was unknown. He denied tobacco use and reported social alcohol use.

Uncorrected visual acuity was noted as counting fingers at two feet with pinhole improvement to 20/30 in both right and left eyes. Pupils were equal, round and reactive with no afferent pupillary defect in either eye. Ocular alignment, motility and confrontation fields were normal in both eyes. Intraocular pressures were 16 and 17 mm Hg in the right and left eye respectively measured with non-contact tonometry at 4:10pm. Retinoscopy was performed and revealed a significant scissoring reflex in right and left eyes. Manifest refraction in the right eye was -10.25-4.00x180 to 20/30- and -12.00-2.00x130 to 20/40-2 in the left eye. The patient reported ghosting and distortion of the letters in both eyes with best spectacle correction.

Anterior segment examination revealed 1+ conjunctival injection of both eyes, inferior stromal thinning with striae but no scarring of both corneas, left eye slightly worse than right, and 1+ superficial punctate

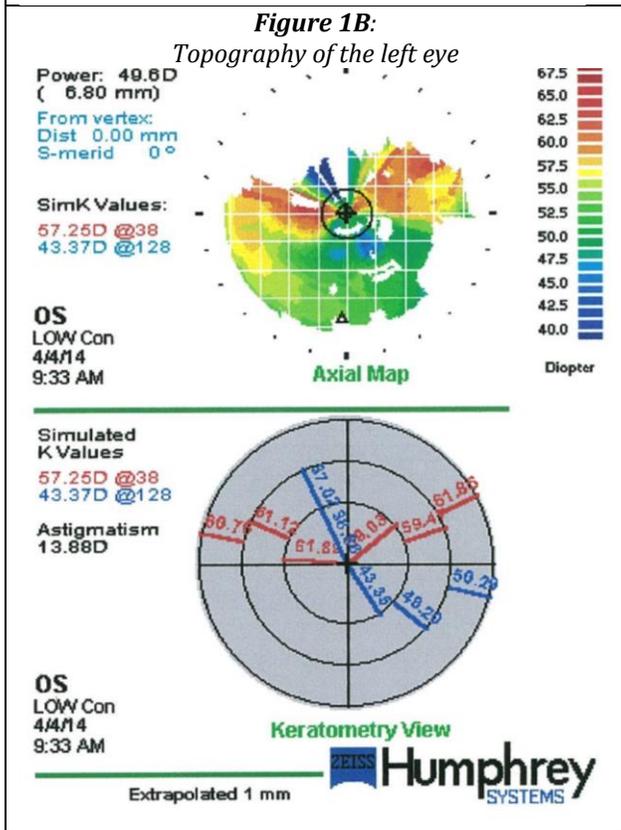
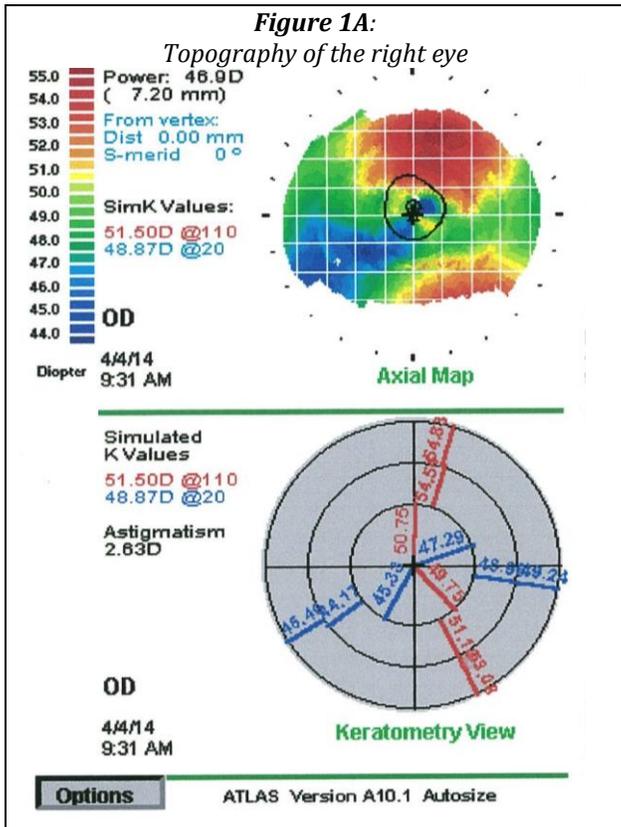
staining of both eyes. Lids and lashes were clean and clear, anterior chambers deep and quiet, irises flat and blue and the lenses were clear in both eyes.

A dilated fundus exam was performed after instilling 1 drop 0.5% proparacaine hydrochloride ophthalmic solution and 1 drop Paremyd® (1% hydroxyamphetamine hydrobromide and 0.25% tropicamide ophthalmic solution) in the right and left eye. Both eyes were found to have optic nerves of good color with cup to disc ratios of 0.4 by 0.4, maculae with good foveal reflexes, vessels of normal course and caliber, clear vitreous and flat and intact peripheries.

A corneal topography (*figure 1A and 1B*) was taken of each eye and showed high irregular astigmatism and steepening >50D in both eyes. Slight flattening pattern inferior was attributed to flat fitting RGP lenses.

The patient’s presenting corneal RGP lenses of unknown design measured as: right eye: BC: 6.96mm, Dia: 9.6, Pwr: -6.50D and left eye: BC 6.42, Dia: 9.6mm, Pwr: -7.50D. His vision was recorded as 20/40 with an over-refraction of -0.50DS to 20/30- in the right eye and 20/70 with an over-refraction of -0.50 to 20/50- with heavy fluctuations in the left eye. The right lens exhibited heavy touch over inferior cone with excessive edge lift and movement. The area of heavy touch correlated to the superficial punctate staining. His left lens had a large crack down the center of the lens and popped out during evaluation. Upon further questioning of the patient, he reported that the lenses were both very uncomfortable, popped out often and he had to remove them for his work as a professional dancer for fear or losing them. He also reported his eyes were always red and he was self conscious about them. SH was under the impression that it was the best that could be done for his condition and felt unable to function without them.

Differential diagnoses for SH were keratoconus, pellucid marginal degeneration, keratoglobus, posterior keratoconus, post surgical ectasias and contact lens included corneal warpage. His corneal topography did not demonstrate the common crab-claw pattern of pellucid margin degeneration or the overall bullous shape of keratoglobus. The topography of posterior keratoconus typically shows spherical or regular astigmatism on the front surface, which was not present for SH. A medical history absent of ocular surgeries ruled out post-surgical ectasias.



SH was diagnosed with keratoconus due to his apical steepening greater than 47D, inferior stromal thinning, limited BCVA with glasses, and scissoring reflex during retinoscopy. He was also determined to have minor contact lens induced corneal warpage from his flat fitting

RGP lenses. His condition was classified as moderate in the right eye and severe in the left eye.

Treatment recommended for the patient was medically necessary contact lens fitting for functional vision improvement. Correction with spectacle lenses was not practical due to advanced state of disease and poor vision through high, unstable manifest refraction. Since he was not contact lens intolerant, did not have heavy scarring and was happy with his overall level of vision through contact lenses, a corneal transplant was not indicated. Scleral lenses were recommended to solve issue of lenses popping out, improve comfort and possibly improve vision. Other lenses options discussed with patient were to refit corneal GP lenses, piggyback system, custom soft lenses or hybrid designs. Patient elected to proceed with scleral lens fitting.

Mini-scleral lenses were selected for this patient for easier handling compared to full scleral lenses; Atlantis® lenses were used. Initial trial lenses were selected based on manufacturer's recommendation of starting with lens labeled "C" with the standard edge design. Lens "C" was in the middle base curve of the trial lens set, allowing the fitter to move steeper or flatter depending of the assessment of initial lens. Since both eyes required the same starting trial lens, a steeper edge design of the same base curve was substituted for the left eye. The following lenses were filled with preservative free saline solution and a small amount of fluorescein dye and inserted in to the patent's eyes:

- OD: BC: 7.67mm, Dia: 15.0mm, Pwr: -3.00, Limbal Zone (LZ): Standard, Scleral Zone (SZ): Standard
- OS: BC: 7.67mm, Dia: 15.0mm, Pwr: -3.00, Limbal Zone (LZ): Standard, Scleral Zone (SZ): 1-Steep

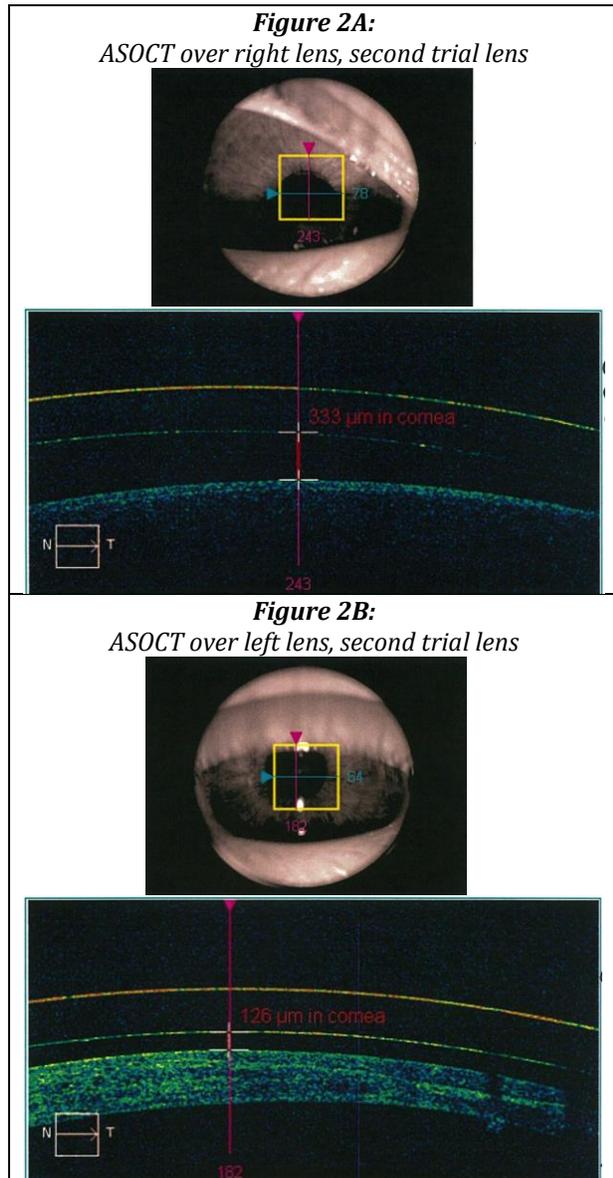
After 5 minutes of settling the lenses were evaluated and found to have no central clearance in either eye, thus indicating inadequate sagittal depth of the lens or too flat of a base curve.

The second trial lenses were selected with a steeper base curve and therefore more overall sagittal depth:

- OD: BC: 7.03mm, Dia: 15.0mm, Pwr: -3.00, LZ: Standard, SZ: Standard
- OS: BC: 7.03mm, Dia: 15.0mm, Pwr: -3.00, LZ: Standard, SZ: 1-Steep

Evaluation of both lenses revealed good centration and a diameter extending approximately 2mm beyond the limbus. The right lens showed an estimated 300µm of clearance over the highest corneal elevation (inferior cone), cleared over limbus without touch, and tight edges with mild compression and blanching of 360 degrees of the conjunctiva. The left lens showed an estimated 150µm of clearance over highest corneal elevation, cleared over the limbus without touch and tight edges with moderate compression and blanching of 360 degrees of the conjunctiva. Over refraction of the right eye was -1.00DS to

20/30- with no improvement with cylinder correction and -1.75DS to 20/20 in the left eye. 25 minutes after insertion anterior segment ocular coherence tomographies (ASOCT) (figure 2A and 2B) were performed over the lenses and the caliper function were used to measure the central clearance as 333 $\mu$ m in the right eye and 126 $\mu$ m in the left eye. With both lenses in, the patient reported incredible comfort and could hardly tell the lenses were even in. When shown the over-refraction over both eyes the patient reported vision was better than he could ever remember.



Since the lenses of both eyes were exhibiting excessive compression of the conjunctiva a flatter edge design was desired for the patient's lens order. The base curve was steepened to compensate for the sagittal depth loss to the flatter edge design and adjusted to achieve a target 200-400 $\mu$ m of central clearance. Power of the lens was calculated using the trial lens power, the over-refraction and compensation for base curve changes. The final lenses ordered were as follows:

OD: Atlantis® Scleral BC: 6.95mm, Dia: 15.0mm, Pwr: -6.50, LZ: Standard, SZ: 1- Flat  
OS: Atlantis® Scleral BC: 6.82mm, Dia: 15.0mm, Pwr: -8.25, LZ: Standard, SZ: 1-Flat

The patient was asked to return in 3 to 4 weeks for the contact lens dispensing visit with insertion and removal training.

#### Dispensing visit

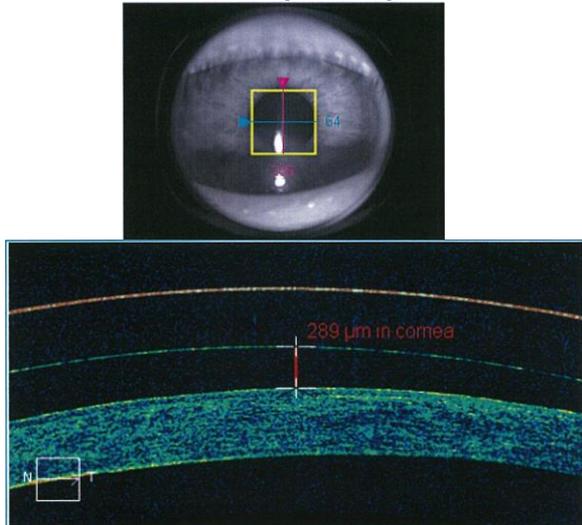
Patient SH presented to the office for a doctor directed dispensing visit on 05/01/2015. The ordered lenses were filled with preservative free saline solution and a small amount of fluorescein dye and inserted in to the patient's eyes. SH reported good initial comfort and vision. After settling five minutes, vision was measured as 20/20 in the right and the left eye and 20/15- with both eyes open. Over-refraction was performed and found to be plano in both eyes.

Evaluation of both lenses revealed good centration and a diameter extending approximately 2mm beyond the limbus. The right lens showed an estimated 300 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. The left lens showed an estimated 400 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. Both lenses rotated freely when manipulated and indicated that there was no touch on the cornea and that the edges were appropriate. AS OCTs (figure 3A and 3B) were performed over the lenses and the caliper function were used to measure the central clearance as 289 $\mu$ m in the right eye and 422 $\mu$ m in the left eye. Lens fit was deemed appropriate in both eyes and lenses were dispensed.

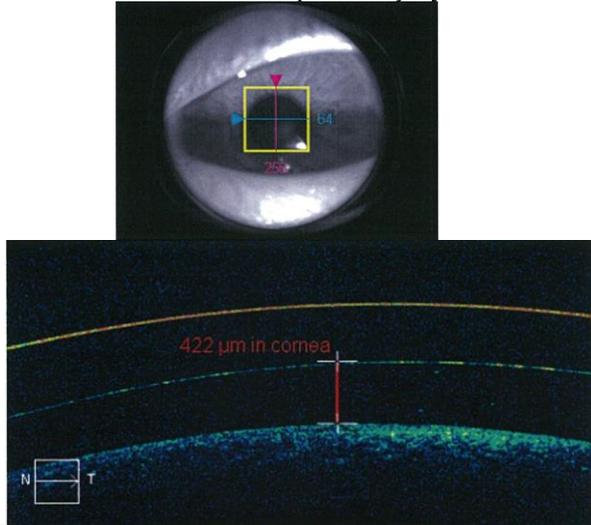
Anterior segment examination revealed inferior stromal thinning with striae but no scarring of both corneas, left eye slightly worse than right, and a 1+ injection of both eyes. Other findings were unremarkable.

Peroxide based cleaned solution and non-preserved saline solution for insertion was prescribed and a three-month supply of the Clear Care® and Unisol® solution was given to the patient. SH was also supplied with a large DMV plunger for inserting the lenses and a small DMV for removing them. He was educated on proper techniques for inserting and removing the lenses and grasped them very quickly. He was instructed to build up wearing time of the lenses, starting with only four hours the first day, adding two hours a day up to a max of twelve hours until he was seen for follow up in two weeks. The patient was extremely pleased with the comfort and the vision with the lenses and excited to leave with them.

**Figure 3A:**  
*ASOCT over dispensed right lens*



**Figure 3B:**  
*ASOCT over dispensed left eye*



manipulated. The lenses were then removed and fluorescein dye instilled into both eyes.

Anterior segment examination revealed inferior stromal thinning with striae but no scarring of both corneas, left eye slightly worse than right. There was no injection and no staining of the cornea or conjunctiva of either eye. Other findings were unremarkable. Lenses were determined to fitting well with no ocular health complications and no changes were required. The patient was instructed that he would wear lenses all day as needed but needed to remove the lenses each night for cleaning. He was asked to return in one month for follow up.

#### *One month contact lens follow up*

On 6/12/2014 the patient presented for his one month contact lens follow up. He was still extremely pleased with the comfort and vision of his lenses. He was excited to report that people had been commenting on how nice and bright his eyes were and that he no longer felt self conscious about them being red all the time. The lenses had not popped out once and been able to wear 16-18 hours without any discomfort. The lenses had been in for about six hours that day.

His vision was 20/20 in the right and left eyes and 20/15 with both eyes open. Over-refraction of the lenses was plano in each eye. Evaluation of both lenses revealed good centration and diameter. The right lens showed an estimated 220 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. The left lens showed an estimated 350 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. Both lenses rotated freely when manipulated.

Anterior segment examination revealed inferior stromal thinning with striae but no scarring of both corneas, left eye slightly worse than right. All other findings were unremarkable. No changes were needed to the lenses and the patient was instructed to continue lens wear and return in 3 months.

#### *Two week contact lens follow up*

SH presented to the office on 5/15/2014 for his two-week contact lens follow up. He reported excellent vision and comfort and had no difficulties with insertion and removal of the lenses. He was especially pleased that his eyes were no longer red and he was able to wear them all day long, even when he was performing. The lenses had been in for about eight hours.

His vision was 20/20 in the right and left eyes and 20/15 with both eyes together. Over-refraction of the lenses was plano in each eye. Evaluation of both lenses revealed good centration and diameter. The right lens showed an estimated 200 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. The left lens showed an estimated 300 $\mu$ m of central clearance, cleared over the limbus without touch, and good edge alignment without compression. Both lenses rotated freely when

#### *Three month contact lens follow up*

SH was evaluated on 9/24/2014 and his exam finding were almost identical to his to one month follow up visit and no changes to his lenses were needed. His contact lens prescription was finalized was instructed to return in nine months for a comprehensive exam and evaluation of his lenses.

### **DISCUSSION**

The cause of keratoconus is complex combination of genetic and environmental factors [3]. One preventable factor of excessive eye rubbing has been cited as a common contributor. Keratoconus onset at younger age is associated with faster progressing and more severe disease [4]. On a microscopic level, proteolytic enzymes degrade the stromal collagen and reduce the overall number of lamellae within a small region of the cornea [3]. This localized thinning allows the cornea to protrude forward creating an irregular anterior surface. As the

thinning progresses, it creates more irregular astigmatism and in advanced stages can cause scarring of the clear tissue. The irregular astigmatism causes progressive blurring and distortion of the vision and creates high and low order aberrations that are not well corrected with traditional spectacles and soft contact lenses.

As the disease progresses traditional spectacle or soft contact lenses no longer provide adequate functional vision and specialty lens options including corneal RGPs, custom soft lenses, hybrids and scleral contact lenses are appropriate. In severe cases of keratoconus where the patient has become intolerant to the sensation of the contact lenses or the corneal scarring prevents functional vision, a corneal transplant may be necessary [10]. Other surgical procedures including corneal cross-linking (CXL) to stiffen the cornea and intra-stromal corneal ring segments (INTACS) to flatten the protrusion and irregular astigmatism have also proven successful.

Corneal RGP lenses have long been the gold standard in correction of keratoconus and work by creating a smooth refractive surface and correcting the aberrations. With severe irregularities of the corneal lenses can decenter or dislocate often [5]. The corneal sensation of the lens can inhibit comfortable lens wear in certain patients and RGPs are associated with an increased risk of corneal scarring, especially with flat fitting lenses [4]. Custom soft lens designs have increased central thickness that partially corrects the irregularity and can provide improved comfort compared to corneal RGPs. The custom soft designs are limited in their ability to correct aberrations [6] and may have a lower quality of vision compared to RGPs. Hybrid lenses, with corneal RGP in the center with a soft skirt, and piggyback systems can improve comfort but they also increase the complexity and cost of lens wear and storage [5].

A well-fitted scleral lens does not touch on the cornea or limbus and bears its weight on the sclera, a tissue of relatively low sensitivity [7]. The size of the lens maximizes centration and allows it to tuck under the eyelids with minimal lid interaction and lens awareness compared to corneal RGPs. The fluid filled lenses provide good optical correction without the danger of increased corneal damage and may delay or prevent surgical involvement [8]. In our case, the scleral lenses corrected our patient's problem of dislocation, provided much improved comfort and significantly improve his best-corrected vision. They also resolved the superficial punctate staining and related conjunctival hyperemia caused by his flat fitting RGP lenses and potentially reduced his changes of corneal scarring. He was able to achieve longer comfortable wearing time, was able to use them for all his daily activities and felt less self-conscious about his eyes.

Scleral lenses, like any contact lens worn in the eye, carry a risk of microbial keratitis but it is not commonly reported in peer-reviewed literature [7]. The isolated cases reported are in severely compromised eyes and those with patients with poor hygiene. Fogging of the fluid inside the scleral lens chamber is a common

complication and typically correlates with ocular surface disease [7]. Improvement is noted with treatment of ocular surface disease and anecdotally reported to decrease after several months of successful wear. Conjunctivochalasis occurs with excessive limbal vault and results in loose conjunctiva being pulled towards the cornea under the lens [7]. Mild discomfort has been noted with conjunctivochalasis and it is typically resolved by refitting the lens with decreased limbal clearance. Based on theoretical models, oxygen supply to the cornea may be insufficient and result in some level of hypoxia induced corneal swelling [9]. According to scleral lens specialist in clinical practice, corneal edema is not often seen and active neovascularization may improve after switching to scleral lenses [7]. None of the above mentioned complications occurred with our keratoconus patient fit in scleral lenses but long-term follow up is necessary to monitor for corneal hypoxia.

Keratoconus patients who fail to achieve good comfort and vision with any contact lens modality or those with excessive scarring may require corneal surgery [10]. Penetrating keratoplasties (PKP) have a high success rate of 93-96% but many post operative patients still require contact lenses to best vision and the surgery carries a lifelong risk of rejection [5]. The relatively new procedure of deep anterior lamellar keratoplasty (DALK) is now being done in certain cases and carries a lower risk of rejection [8]. Corneal cross-linking (CXL) uses ultraviolet A radiation and vitamin B2 (riboflavin) to strengthen the corneal tissue [11]. It can create a small flattening effect that can improve vision but its primary effect is in slowing down or halting the progression of keratoconus [11]. Intra-stromal corneal ring segments (INTACS) are two semi-circular plastic rings inserted into the stroma that flatten the overall corneal curvature, however many patients still require contact lenses to achieve their best vision. Since our patient was able to achieve good vision and comfort with his lenses corneal transplants did not need to be explored. He could potentially benefit from CXL to halt the progression of his disease.

## CONCLUSION

Keratoconus is a good indication for use of scleral lenses as they correct irregular astigmatism and aberrations and provide superior comfort compared to traditional contact lens options. Scleral lenses should be considered before patients fail in other lens modalities since it may prevent or delay the need for surgical intervention.

This case report demonstrated that fitting of scleral lenses can be simple and that patients can easily adapt to this type of lens.

## CONFLICTS

Justine Siergey has been a paid educator for several scleral lens companies including: X-Cel Contacts (manufacturer of Atlantis®), Blanchard Contact Lens Inc. and TruForm Optics.

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