Case Report: Clinical findings of bilateral Keratoconus with custom scleral lens management

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Abstract:

This case report details the management of complicated bilateral keratoconus in a young patient. Keratoconus is a multifactorial condition with differing rates of progression between each patient. Additionally, there are several different treatment options including surgical intervention and long-term scleral lens wear, the option discussed in this case report. Scleral lenses are the current go to contact lens option for patients with moderate to severe cones and scarring as discussed in this case report. For severely irregular cases EyePrint Prosthetic scleral lenses have been shown to improve vision the greatest for these patients. It is important for clinicians to know the available options for more complex patients that go beyond the standard scleral lens.

Key words: Keratoconus, Scleral lens, Eyeprint prosthetic

Introduction

Keratoconus is a bilateral yet asymmetric ectatic corneal degeneration that typically onsets between late teens to early 20's with progression usually into the patient's fourth decade. Keratoconus affects both genders as well as all ethnicities.[1] The disease process includes steepening of the cornea with associated inferior thinning. This causes an increase in irregular astigmatism and increase in higher order aberrations leading to an overall decrease in visual acuity. Complications such as scarring, and hydrops can cause further reduction in visual acuity. Scleral lenses have been found to improve Keratoconic patient's visual acuity more than rigid gas permeable lenses or glasses.[2] With an increase in technology, scleral lens choices now allow patients to have a fully customized lens. An example of this new technology is utilizing EyePrint Prosthetic impression molded lenses.

Case Report:

Initial visit:

A 26 year old African American male presented to the Contact Lens Institute of Nevada on 06/07/2022 complaining of blurry vision during both the day and night, worse in his left eye with an increase in fogging of his current habitual left scleral lens fit by his primary care optometrist. The patient also complains of constant redness, irritation and dryness of the left eye while wearing his current lens. Patient wears his scleral lenses for 12-14 hours a day. Patient presents out of his habitual lens in his left eye for 13 hours.

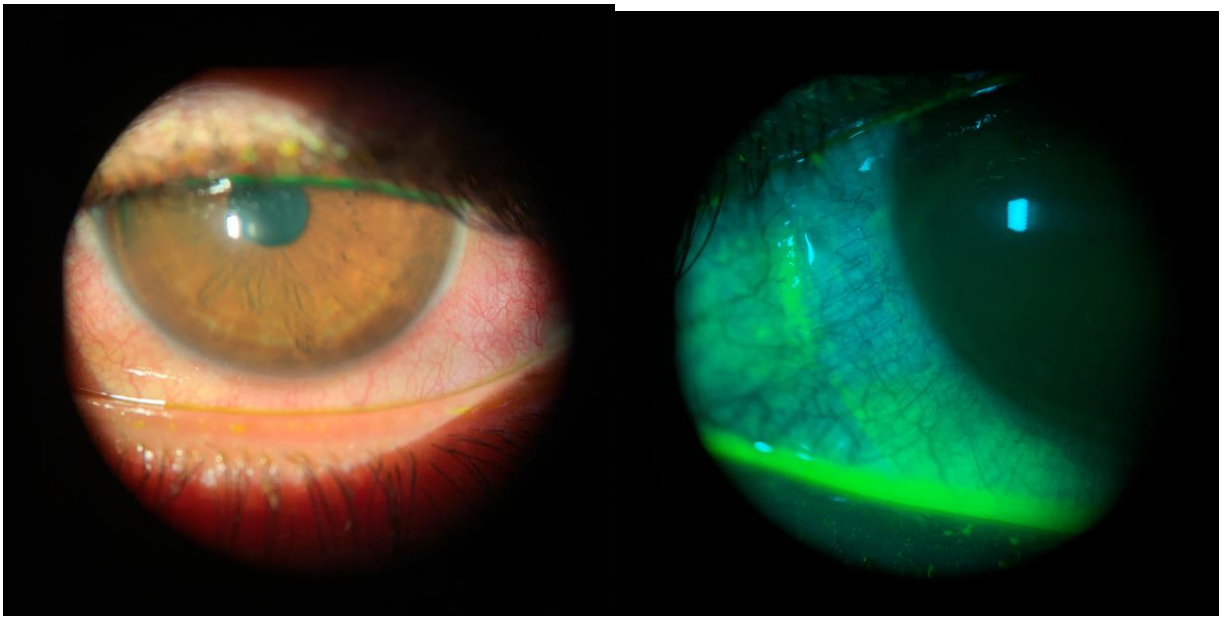
History: The patient was diagnosed with Keratoconus at the age of 18 years old and was initially seen at this clinic 05/19/2021, before my rotation at the clinic. At this time, the patient presented for similar issues as discussed above, but mostly in the right eye originally. Surgical history of corneal crosslinking done in 2019 to both eyes. Additional ocular history of corneal scarring in the right eye secondary to severe keratoconus. Patient was fit into a Eyeprint Pro prosthetic in his right eye in 2021 with higher order aberration correction and has been successful with the fit and vision. Patient's last eye exam at the clinic was 12/06/2021 for adjustments to his right scleral lens. Patient receives comprehensive eyecare (glasses needs and dilation) with his primary care optometrist. Family ocular history includes Keratoconus in his Maternal grandmother. Negative medical and social history. Negative family medical history. Negative medication and allergy history. Patient is currently in the police academy.

Examination:

Prior to the appointment, the patient was instructed to present not wearing his lens for at least 48 hours. This is to allow the conjunctiva to return to its unaltered state. Scleral lenses can often leave compression marks within the conjunctiva and then skew the data that is used to create the custom scleral lens. Due to the patient's work schedule, he was unable to be out of the lens for that long and presented out of the lens for 13 hours.

The visual acuity obtained for the left eye (uncorrected) was hand motion. Right eye (corrected) 20/25-

Slit lamp examination of the right eye (wearing his habitual EyePrint Prosthetic lens) shows severe keratoconus with scarring of the central cornea with superior pannus extending into midperiphery and 2mm neovascularization inferiorly. Slit lamp examination of the left eye reveals moderate keratoconus and inferior thinning on optic section examination. The bulbar conjunctiva of the left eye reveals overall inflammation with 3+ injection due to habitual scleral lens compression, worse nasally. Sodium Fluorescein dye was instilled OS which confirmed scleral lens edge compression 360, worse nasally.



Photos 1a and 1b. 1a shows the 3+ bulbar injection and irritation that the patient complained of. 1b shows the conjunctival staining nasally caused by scleral lens compression. The patient had been out of his left habitual scleral lens for 13 hours before the photos were taken.

Following slit lamp examination, Medmont corneal topography was obtained. Keratoconus' primary mode of diagnosis is via topography analysis where inferior corneal steepening can be detected.[1] After this scan, 5 drops of Flures (sodium fluorescein and proparacaine) were instilled to gather central, inferior, and superior scleral data using the sMap. The sMap is a corneal-scleral topographer that images 1 million points to create a highly detailed map of the cornea and scleral. These points are used to create a completely custom scleral lens. This is often used at the Contact Lens Institute of Nevada because the typical patient's cornea and scleral are too irregular. This can be due to advanced disease or post-surgical results, but the corneal complexity prevents them from being fit in a foundational scleral lens. The sMap delineates specific irregular areas that will require microvaults or additional areas of clearance (commonly required in patients with INTACs).

After imaging was obtained, EyePrint Pro impression of the left eye was taken. This involves 1 drop of tetracaine in the left eye, with a piece of Tegaderm tape placed to the lower lid before injecting an eye safe mold covering the entire sclera while the patient looks forward in primary gaze. The impression was left on for 60 seconds before removal. Patient tolerated the procedure well. This impression will be used to create a completely custom scleral lens molded around all ocular irregularities.

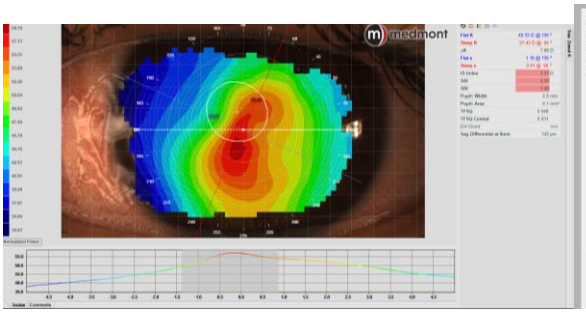


Image 2a: Medmont Topography.

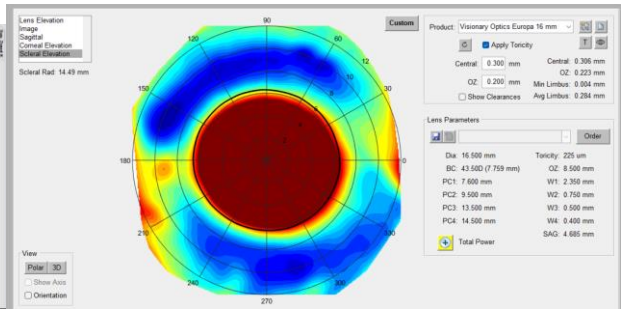


Image 2b: sMap topography

After sMap scleral imaging and EyePrint impression mold were obtained a diagnostic lens fit was preformed utilizing Europa Toric scleral lens diagnostic kit. Based on the sagittal depth and toricity recommended from the sMap topography, a lens was chosen that should allow adequate microns of central clearance.

The sMap suggested 4685 microns of sag and 225 microns of toricity. To ensure central clearance, a diagnostic lens should always have greater than the amount of sag suggested by the sMap. The closest lens in the fit set that included both parameters was: Europa T3, 50 D BC, 16mm overall diameter, 5050 microns of sag, -4.00D of power, 200 microns of toricity.

The diagnostic lens was placed on the left eye and anterior segment OCT (optical coherence tomography) imaging was preformed over top to allow fitting information to be gathered when ordering the patient's custom lens. Then a subjective over refraction was completed revealing –5.25 DS with an improvement to 20/25- visual acuity. Fit assessment via slit lamp examination and OCT imaging revealed 700 microns of clearance, nasal, temporal, and inferior edge compression with an overall small lens diameter compared to the patient's sclera.

All topography, fit assessment, anterior segment OCT images, over-refraction data and EyePrint Pro impression molding of the left eye were then sent to the EyePrint lab for custom scleral lens creation.

Differential diagnosis:

Due to the nature of this specialty clinic all patients present with their diagnoses and the clinic provides tailored treatment options. But based on this patient's chief complaint of increasing blurry vision at both distance and near differentials include:

Keratoconus

Dry Eye

Refractive error

Chronic retinal disease

Chronic/worsening corneal dystrophy

Optic neuropathy

Cataract

Amblyopia

The patient does complain of mild dry eye secondary to extended wear of scleral lenses, patient uses artificial tears as well as Nutrifil solution in his scleral lenses to combat any contributions of dry eye towards his visual acuity. Due to the slit lamp findings of inferior thinning of the cornea, dry eye was ruled out as the major cause of the patient's chief complaint. Dry Eye could serve as a secondary diagnosis.

The patient does have an unstable refractive error, but this is secondary to corneal changes caused by Keratoconus versus the unstable refractive error alone causing blurred vision at distance and near.

Chronic retinal disease includes retinitis pigmentosa, diabetic retinopathy, and macular degeneration. Retinal Wellness scan with RNFL analysis on the Maestro 2 OCT was performed on patient examination to rule out any retinal/macular/optic nerve contributions to the patient's reduction in vision. Retinal scan of both left and right eye revealed no signs of retinal, optic nerve, or macular disease. This data can be used to rule out Optic Neuropathy as well.

Chronic and worsening corneal dystrophies encompass EBMD, Macular/Granular/Lattice stromal dystrophies, and Fuch's endothelial dystrophy. There were no signs of epithelial disruptions, stromal deposits, or signs of gutatta or corneal edema on slit lamp examination, ruling out corneal dystrophies as primary diagnosis.

Cataracts were ruled out after slit lamp examination of the lens revealed clear lenses OU.

Amblyopia was ruled out due to negative history, ability to correct down to 20/20 in the past and no tropia on cover test. The patient cannot correct down now as an adult to 20/20 secondary to advancement of Keratoconus. A negative history includes: No history of patching, strabismus surgery, glasses at an early age or eye turn.

Follow up #1 for OS: 07/26/2022

Due to the patient only being out of his habitual scleral lens for 13 hours, the original EyePrint Pro molding obtained on 06/07/2022 was unable to be used due to the scleral shape being altered by his habitual scleral lens. EyePrint Pro recommends a minimum of 24 hours out of habitual lenses before obtaining molds to entire accuracy for the true scleral shape. During this follow up a secondary Eyeprint pro mold was obtained.

Patient presented at follow up out of his habitual left scleral lens for greater than 48 hours which will allow for optimal data collection.

VA obtained again for the left eye (uncorrected) was hand motion.

Slit lamp examination of the left eye utilizing NaFl revealed 3 vertical track like epithelial defects in the inferior nasal cornea (image 3A). There was also a small superficial defect at 11'oclock on the cornea (image 3B). These defects are caused by the patient's left habitual scleral lens fitting improperly and causing localized friction against the cornea due to areas of low clearance.

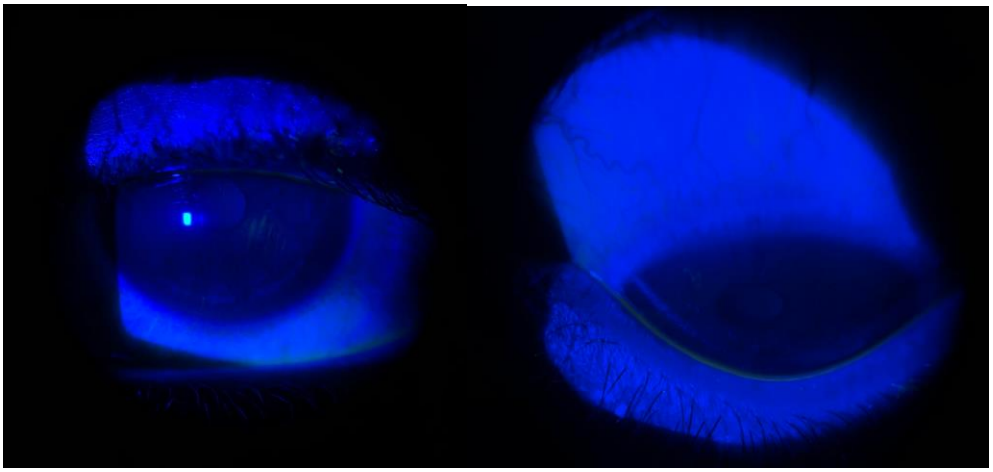
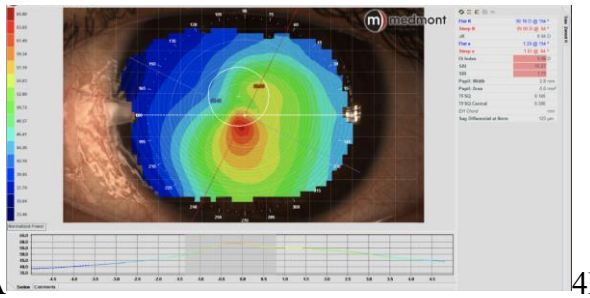
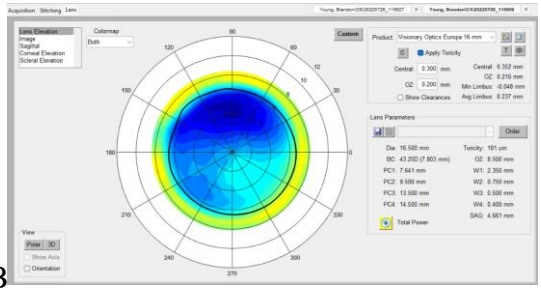


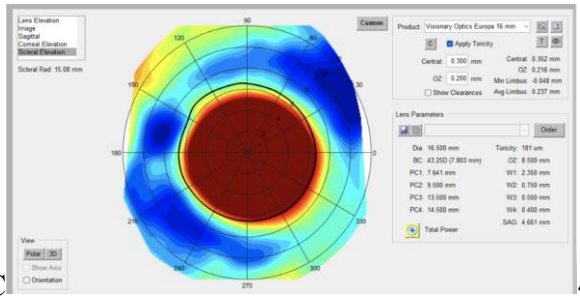
Image 3A. Shows 3 vertical track like epithelial defects near pupillary margin inferior nasal.

Image 3B. Shows small superficial epithelial defect at 11'oclock

Like at the original appointment: Medmont topography of the left eye was obtained followed by sMAP data. 5 drops of Flures were instilled in the left eye and data from upgaze, downgaze and center gaze was gathered. A lens elevation and scleral elevation map were generated. Following the sMAP, the Eyeprint impression molding was obtained. 1 drop of Tetracaine was instilled in the left eye before placing Tegaderm tape on the lower eyelid for control. The patient then looked straight forward while the molding was placed inside the eye. The mold set for 60 seconds before removal. At this follow up, 2 moldings of the patient's left eye were obtained for additional safety precaution.

4A 

4B 

4C 

4D 

Image 4a is the Medmont Topography. Image 4b is the lens elevation map by sMap. Image 4c is the scleral elevation map by sMap. Image 4d courtesy of Dr. Woo, is an example of the lens impression molding obtained with EyePrint (not the patient's molding in photo)

The topography, sMap elevation and lens maps, anterior segment photos and Eyeprint impression moldings of the left eye were then sent to the lab for custom scleral lens creation.

The patient will return to the clinic in 3-4 weeks depending on the length of the time needed to create the lens and have it shipped to the clinic. At this follow up, the lens will be dispensed to the patient and then fit, vision and comfort will be analyzed. The patient will then leave with the lens to trial for 1-2 weeks. After this trial period, the patient will return for re-assessment of fit, vision and comfort. If all 3 elements are satisfactory the lens can be finalized for 1 year of wear for the patient. The patient will then continue co-management with his primary care optometrist for dilated fundus examination and glasses needs.

Discussion:

The mechanism behind the development and progression of Keratoconus is still under research. Keratoconus is a multifactorial disease with risks including: history of atopy (dermatitis, allergies, asthma, eczema), other diseases that increase frequency of eye rubbing (floppy eyelid syndrome, ROP), connective tissue diseases and family history.[1] The pathophysiology of Keratoconus involves degeneration of proteoglycans within the microfibrils of stromal collagen, creating a weak cornea subject to protrusion and associated thinning creating the pathognomonic "cone". Studies have also found there is a change in oxidative enzyme function allowing more oxidative stress to damage the cornea.[1] Overall, the development and progression of Keratoconus has many factors that impact the patient's potential for visual acuity and complicate the management process.

The usual standard of care for Keratoconus before considering surgical intervention or in conjunction with a surgical treatment begins with spectacles, soft contacts, piggyback systems, rigid gas permeable lenses alone, hybrid lenses and then scleral lenses.[3] There are also different options within the scleral lens family, starting with a standard scleral (Jupiter, Zen Lens, Europa), a more custom scleral like Latitude, and finally a fully custom option EyePrint Prosthetics. Latitude custom scleral lenses also utilize sMap technology but unlike EyePrint there is no impression molding taken to contribute to the customization parameters. Additionally at this clinic, scleral lenses can incorporate higher order aberration correction utilizing Ovitz technology. Some studies have shown that long term scleral lens usage has decreased the need for surgical intervention.[4] The standard of care for surgical options in managing Keratoconus include: Intrastromal Corneal Ring Segments called Intacs, Corneal Crosslinking, and Keratoplasty including Penetrating (PK) and Deep Anterior Lamellar (DALK). Keratoplasties are typically reserved for patients with advanced Keratoconus that has caused scarring, preventing visual improvement with other modalities, including scleral lenses. The DALK has increased in popularity due to a decrease in frequency of graft rejection and preservation of corneal endothelial cells.[3] When it comes to pursuing a surgical option to manage keratoconus, many factors should be considered as no two patients are exactly alike. Primarily, stage of progression should be considered. Mild cases can be successful with spectacles, contact lens options and corneal cross linking. If the patient presents with severe scarring and history of hydrops perforation, penetrating keratoplasty is more likely to improve vision than a contact lens option. When looking at surgical options complications including: graft rejection, refractive error change leading to anisometropia, infection, cataracts, glaucoma and worsening ocular surface disease (dry eye) should be considered.[4] Other factors to consider include patient lifestyle and visual demands, as well as previous success in other modalities. For example, if a patient has had multiple lens fit failures in differing lens types, surgery intervention is likely the only option to improve vision. In this patient's case, custom scleral lenses were used in conjunction with corneal cross linking as management of his keratoconus. Due to the patient's severe presenting signs and symptoms as well as previous difficulty with his habitual standard scleral lens; a more custom option was required. Additionally, the patient's previous success with the EyePrint Pro molding to provide vision and comfort to his right eye allowed this treatment option to be easily considered.

Conclusion:

This case illustrates the complex nature of Keratoconus itself as well as managing it. All patient presentations, symptoms, history, and prognosis differ, preventing just one treatment from being the clear answer. Each patient should be treated individually while factoring in their lifestyle and previous treatment history. Scleral lenses have become one of the forefront of treatment options for patients with Keratoconus allowing there to be one more step before considering keratoplasties. As exemplified by this case, custom scleral lenses allow patients with advanced keratoconus to achieve significant improvements in vision. Additionally, this case shows that collecting all the information exactly as directed is critical for ensuring the custom lens has the best and most accurate fit for the patient and that this process takes time and patience.

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