The Management of Dry Eye Related Corneal Distortion via Scleral Shell

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Candidate #251 Case 1
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ABSTRACT

Background: Dry eye disease (DED) is an insidious problem for patients of all ages. Although treatable, it is as yet incurable. According to the TFOS DEWS II report (published in the *Ocular Surface Journal*, July 2017), Dry Eye is defined as, "*a multifactorial disease of the ocular surface characterized by a loss of homeostasis of the tear film, and accompanied by ocular symptoms, in which tear film instability and hyperosmolarity, ocular surface inflammation and damage, and neurosensory abnormalities play etiological roles.*"
In mild cases, DED causes discomfort which may be constant or infrequent. As the condition progresses, through the spectrum of mild to moderate to severe, patients may report pain and fluctuating or blurred vision. The cornea may become distorted and compromised to the point of scarring. The spectrum of treatment may range from standard techniques; i.e. patient education and lifestyle modification, hot compress and lid hygiene techniques; to secondary treatments. These include artificial tears, Meibomian gland expression, punctal occlusion, in-office eyelid therapy (e.g IPL, LLLT, MiboFlo, LipoFlo, RF, etc.) and topical steroids and antibiotic-steroid combinations. When these methods do not stem the cascade of dry eye sequelae, tertiary treatments like autologous serum, bandage contact lenses and Scleral lenses may become a viable methods available to Optometry to treat advanced DED[13]. **Epidemiology[6]:** Based on data from the National Health and Wellness Survey, 6.8 percent of the United States adult population (approximately 16.4 million people) have been diagnosed with DED. The prevalence increased with age (2.7 percent in those 18 to 34 years old versus 18.6 percent in those ≥75 years old) and was higher in women than men (8.8 versus 4.5 percent). Prevalence (see Table 1) was not affected by education or location of residence.[7.]

Table 1: Dry Eye Disease Risk Factor Categorization[8]		
Consistent	**Probable**	**Inconclusive**
aging	diabetes	Hispanic ethnicity
female sex	rosacea	menopause
Asian race	viral infection	acne
meibomian gland dysfunction	thyroid disease	sarcoidosis
connective tissue disease	psychiatric conditions	smoking
Sjögren syndrome	pterygium	alcohol
androgen deficiency	low fatty acid intake	pregnancy
computer use	refractive surgery	demodex infestation
contact lens wear	allergic conjunctivitis	botulinum toxin injection
use of medications, such a, antihistamines, antidepressants, anxiolytics, and isotretinoin	medications (for example, anti-cholinergics, diuretics,	multivitamins
estrogen replacement therapy		oral contraceptives
hematopoietic stem cell transplantation		
environmental conditions, such as pollution, low humidity, and sick building syndrome		

Case Report: A 27-year-old Latino male with a history of juvenile leukemia and DM, presented with long standing ocular dryness, right eye > left. Vision was reduced in the right eye and normal in his left with only slight improvement with pinhole. Key entrance findings were a central corneal opacity RE and a distorted corneal topography. General exam was followed up with a dry eye workup and ultimately, the patient's RE was fit with a scleral lens. At five months the patient has reported clearer vision and a more comfortable RE. **Discussion:** Over the past 40 years, the Dry Eye treatment paradigm has steadily evolved. For many years the best advice doctors could give patients was the use of artificial tears (a stand-alone treatment many doctors strongly rely on even today). Then came the idea of disrupting the lacrimal drainage system either with silicone punctal plugs or surgical cautery. This philosophy subscribed to the thinking that dry eye was mostly related to decrease tear production and by keeping more tears in our patients' eyes we could alleviate their symptoms. Often the final result was "plugging up a swamp" of poor quality tears. Around the start of this millennium, the oil producing meibomian glands were given the attention they deserved and treatments revolved along the lines of applying heat first to the outside of the eyelids and eventually in combination with physical manipulation and life-style/environmental modifications. At the same time, the inflammatory nature of DED was also recognized and two topical prescription therapies began to be employed in its amelioration: corticosteroids, and topical cyclosporine A (CsA) (with lifitegrast ultimately joining this list). Oral essential fatty acid supplementation and tetracycline-class antibiotics began to be commonly prescribed as well[12]. As we continue forward to our present, the devices to diagnose, measure and treat DED have multiplied. As previously mentioned in this case study, our current treatment methodology now has 4-step process (see Table 2). In Step 3 the use of properly fit, Scleral lens designs allow for a constant neutral saline reservoir to bathe the compromised cornea throughout all hours of wear. Additionally, the scleral shell protects the cornea from exogenous insult related to foreign matter and gives the lids a smooth surface to blink over.
Conclusion: This case of a severe dry eye and corneal distortion was handled successfully by the use of a Scleral lens. [1]
Keywords: Dry eye disease / Scleral Contact Lenses / Corneal topography

Table 2[13]
Staged management & treatment recommendations for dry eye disease

Step 1:

- Education regarding the condition, its management, treatment and prognosis
- Modification of local environment
- Education regarding potential dietary modifications (including oral essential fatty acid supplementation)
- Identification and potential modification/elimination of offending systemic and topical medications
- Ocular lubricants of various types (if MGD is present, then consider lipid-containing supplements)
- Lid hygiene and warm compresses of various types

Step 2:
If above options are inadequate consider:

- Non-preserved ocular lubricants to minimize preservative-induced toxicity
- Tea tree oil treatment for Demodex (if present)
- Tear conservation
    - Punctal occlusion
    - Moisture chamber spectacles/goggles
- Overnight treatments (such as ointment or moisture chamber devices)
- In-office, physical heating and expression of the meibomian glands (including device-assisted therapies, such as LipiFlow)
- In-office intense pulsed light therapy for MGD
- Prescription drugs to manage DED
    -
    - Topical antibiotic or antibiotic/steroid combination applied to the lid margins for anterior blepharitis (if present)
    - Topical corticosteroid (limited-duration)
    - Topical secretagogues
    - Topical non-glucocorticoid immunomodulatory drugs (such as cyclosporine)
    - Topical LFA-1 antagonist drugs (such as lifitegrast)
    - Oral macrolide or tetracycline antibiotics

Step 3:
If above options are inadequate consider:

- Oral secretagogues
- Autologous/allogeneic serum eye drops
- Therapeutic contact lens options

- Soft bandage lenses
- Rigid scleral lenses

Step 4:
If above options are inadequate consider:

- Topical corticosteroid for longer duration
- Amniotic membrane grafts
- Surgical punctal occlusion
- Other surgical approaches (eg tarsorrhaphy, salivary gland transplantation)

INTRODUCTION

Systemic therapies used for a wide range of cancer types are associated with distinct adverse effects, several of which may affect the eye. Ocular side effects from systemically administered chemotherapy can be severe, disabling, and irreversible. These agents may cause dry eye, meibomian gland dysfunction and nonhealing corneal epithelium.[2]

DED is the most frequently encountered sequelae of diabetes mellitus (DM). Although the pathogenesis of DM related dry eye is unknown (and correcting metabolic imbalance can assist in lessening symptomology), its prevalence has increased in recent years.[3, 10] Studies have linked antineoplastics to dry eye disease as an iatrogenic causative factor. [9,] Out of 77 bone marrow transplanted patients surviving at least 1 year, 43% developed ocular manifestations of DED. Irradiation may contribute to the development of dry eye, since all patients with hematological malignancies, in contrast to aplastic anemia patients, had undergone total body irradiation. [11]

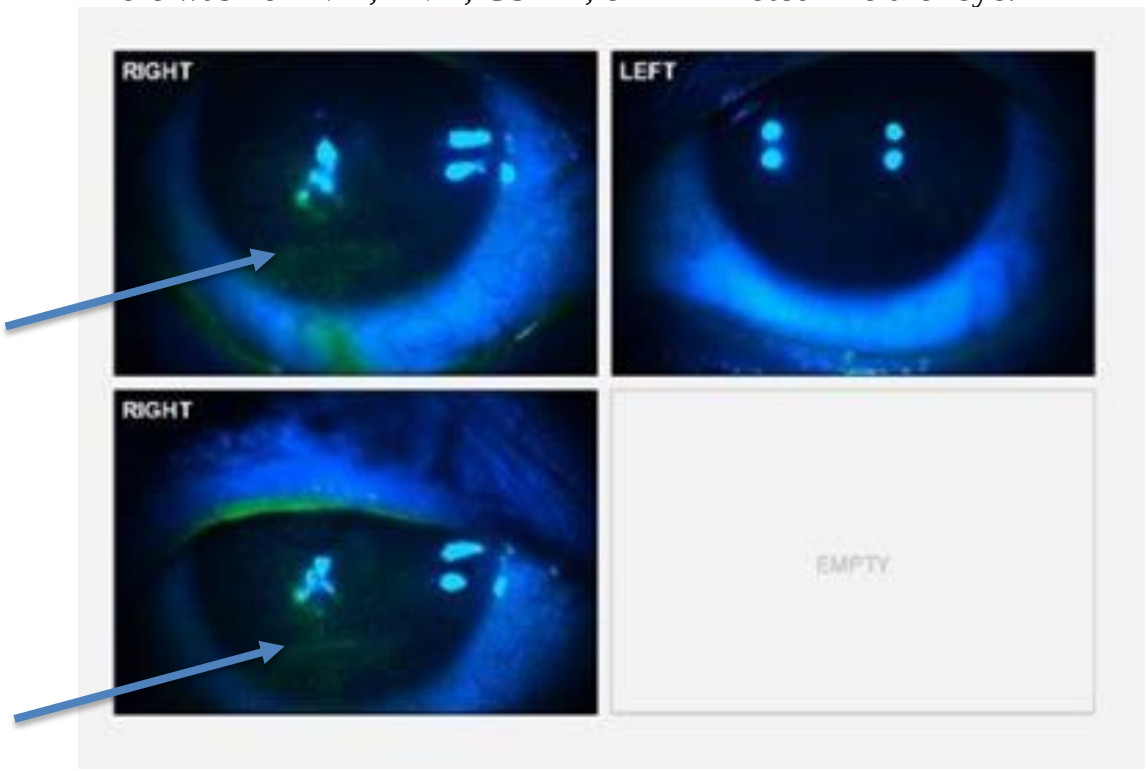
The case report which follows describes the initial encounter with a young man suffering from poor vision related to his previous and ongoing health history and the methods used to ameliorate his condition.

CASE REPORT

Chief Complaint: MR, a 27 year old, Latino male, presented to our office **4/19/19**. He reported no allergies and a non-contributory family history. He denied alcohol or tobacco use. Current medications include Aspart Insulin and Gemfibrozil. The patient was diagnosed with Acute Lymphoid Leukemia in 2005. His therapy included chemotherapy, radiation, and bone marrow transplant. He has been in cancer remission since. Type II Diabetes Mellitus was first diagnosed in 2018 and reported as being under good control.

Chief complaints consisted of dry eye discomfort and general blurred vision beginning shortly after treatment of patient's leukemia, the overall symptoms being significantly worse for the Right eye than the Left. Patient reported RUL cautery in 2008 and RLL punctal occlusion along with Bruder-type mask and artificial tear in-home therapy, with no relief of symptoms; this factor along with the patient's condition of his cornea led to the decision to use a scleral lens for tertiary treatment. Other symptoms included: glare, photophobia, eye fatigue, burning, dryness, epiphora, soreness, redness, and sandy or gritty feeling to the right eye.

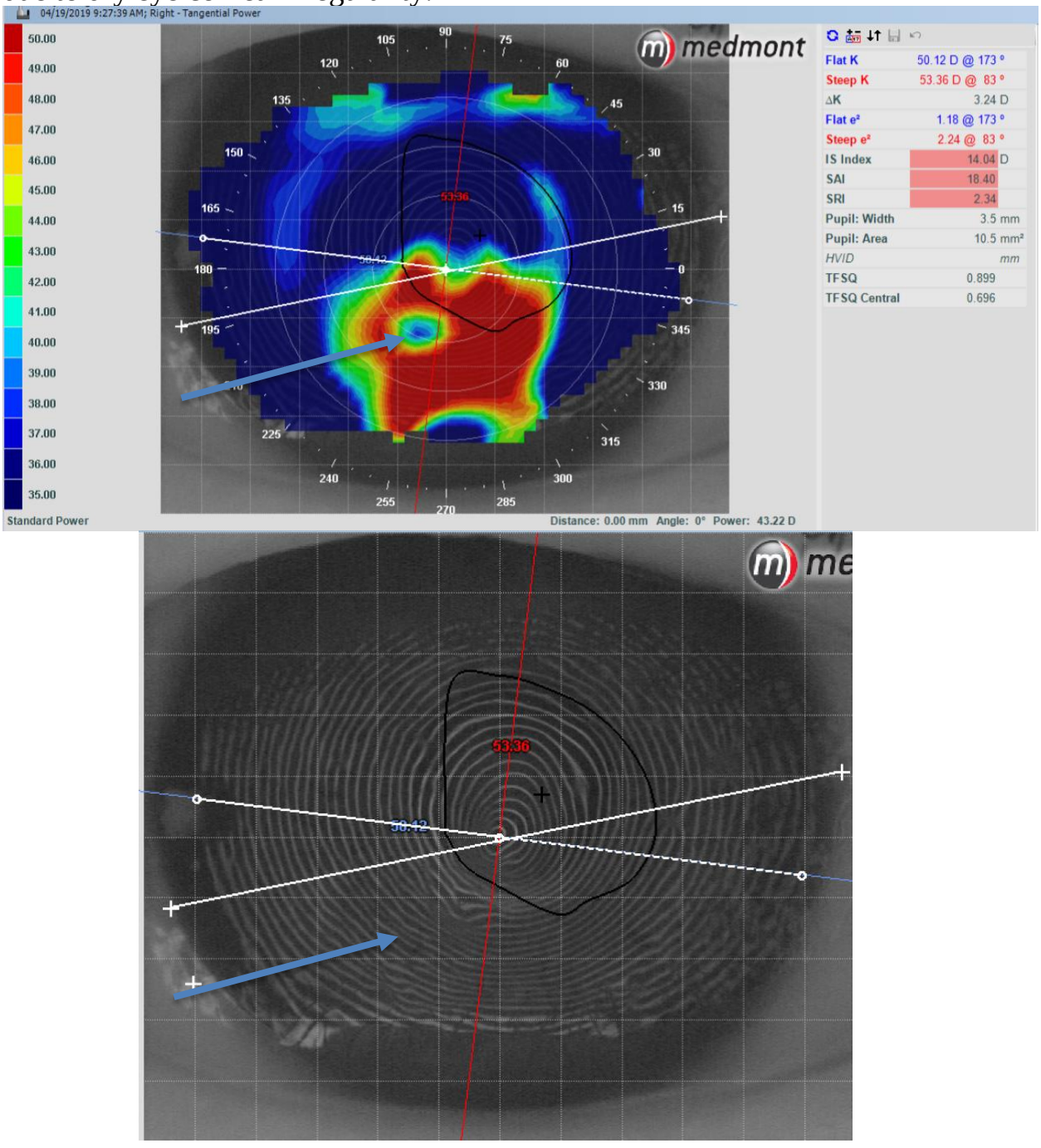
Initial Exam Findings: MR's unaided distance acuity was RE 20/70 (improved to 20/60-2 with pinhole), LE 20/20 and 20/15 OU. Near point VA was RE 20/200, LE 20/20, and OU 20/20. His eyes were orthophoric distance and near with full range of motion, normal color vision, and NPC to nose. His stereopsis was decreased to 2/6 owing to the poor acuity in his RE.
His autorefraction and autokeratometry was as follows: RE -1.75-6.00x96 / 43.37@88 x 54.25@176 with 176 micron elevation difference and LE +.25-.50x19 / 41.75@03 x 42.37@93 with 16 micron elevation difference.
Manifest refraction was: RE -.50-1.25x50 w/20/70 PHNI and LE 0.00 w/20/20.
IOP was RE 15mm Hg and LE 13mmHg
Slit lamp exam demonstrated RE epithelial staining and a pericentral corneal opacity approximately 1mm in size from Bowman's membrane to the anterior stroma, 3+ stringy mucous, lid swelling, 4+ bulbar and conjunctival injection, and trace anterior lenticular opacities. LE only showed 2+ bulbar conjunctival injection and OU the anterior chambers were deep and clear. Pupils were equally round and reactive to light. There was no APD present.
Internal eye health exam showed normal nerves, maculae, and blood vessels. There was no NVD, NVE, CSME, or BDR noted in either eye.



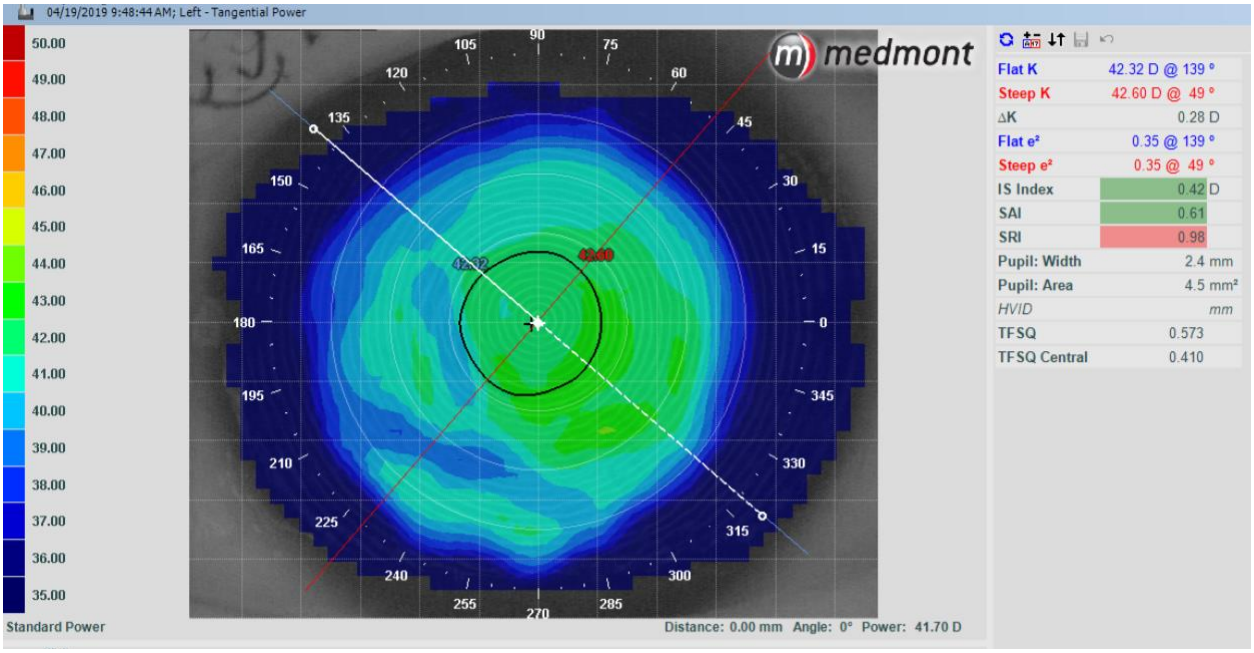
The above photos demonstrate significant elevated RE corneal disruption and staining in the inferior midperiphery. The LE is clear of any such defects.

Topographical Analysis:

MR's right eye demonstrated significant distortion which also gave the eye the appearance of being much steeper than the LE with keratoconic-like elevation due to dry eye corneal irregularity.



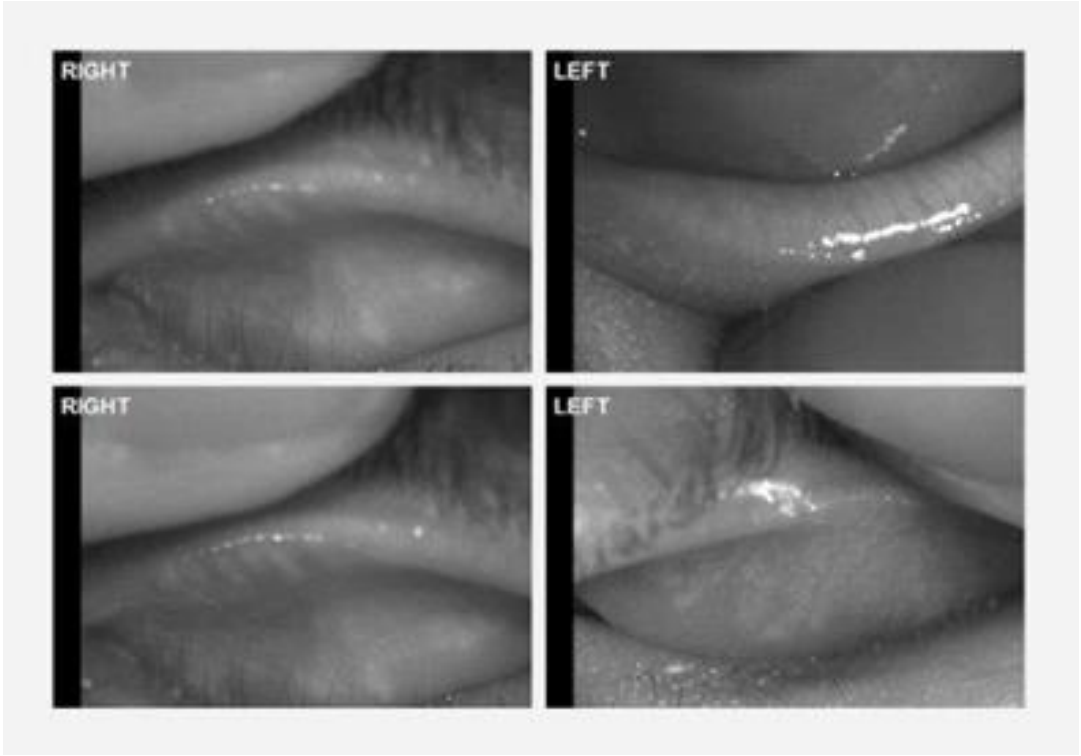
RE topography and mire pattern (arrows indicate area of greatest dry eye related cornea insult)

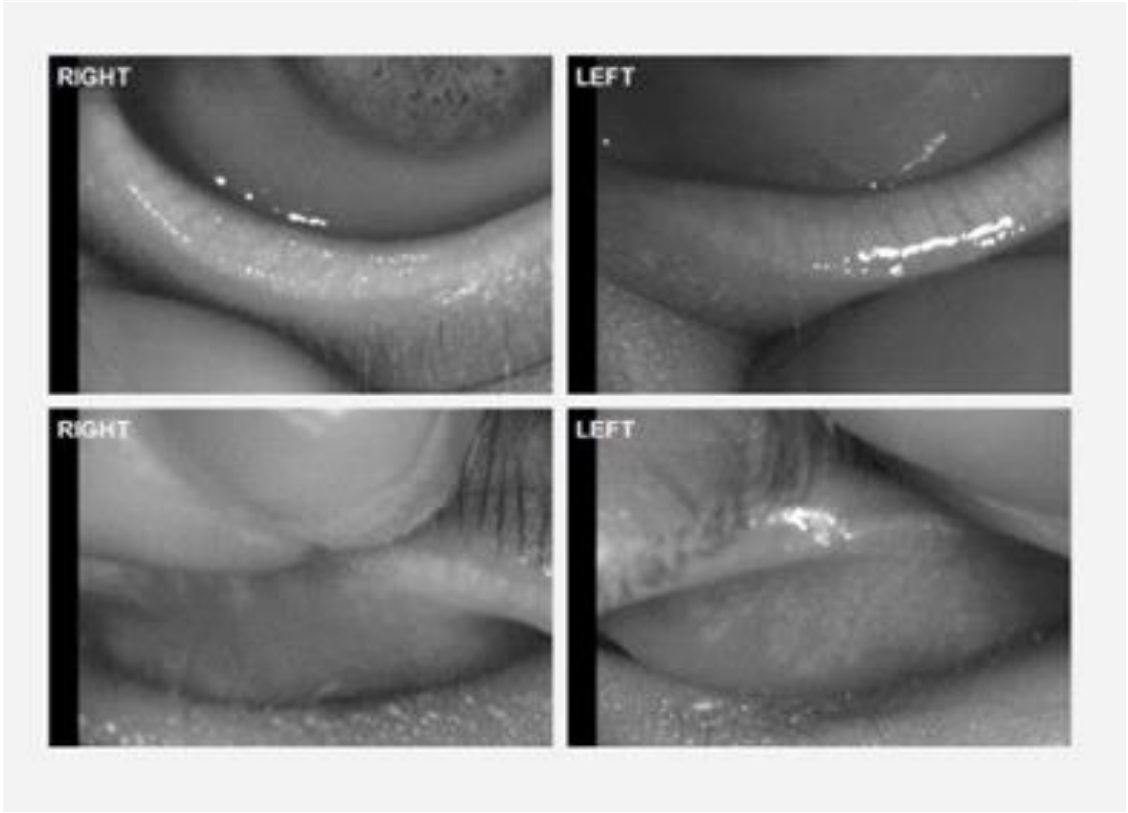


LE topography

Dry Eye Evaluation: this eval was performed on **4/26/19**. MR's Right cornea had 4+ epithelial staining with a 3 second TBUT. The Left cornea demonstrated no staining but also had a reduced TBUT of 2 seconds. His zone quick results showed possible reflex tearing as his RE was 15mm and his LE was 3mm for the 15 second test. Inflammadry yielded positive results OU. Meibography demonstrated Grade 2 gland atrophy for his Right upper and lower lids, and Grade 1 Left upper lid and Grade 0 Left lower lid. He also showed OU inferior and Superior lid wiper epitheliopathy and 1+ positive nasal aspect LE lissamine green staining of the bulbar conjunctiva. The RE had 4+ bulbar and palpebral conjunctival injection and the LE had only 2+ bulbar conjunctival injection. On gross evaluation, the RE had a distinct shrunken appearance as compared with the left.

Diagnosis and Management: MR was diagnosed with aqueous deficient dry eye and Meibomian gland dysfunction OU. Treatment has consisted with Restasis 1gtt OU bid and Systane prn for the naked RE. Again, it should be noted that DES signs and symptoms for the LE were considerably less than for the RE. He was recommended to use Thermolon hot compresses qhs. As previous dry eye therapies were less than adequate for the amelioration of the patient's chief complaint and symptoms (i.e. RE discomfort and blur) it was decided to proceed with a Scleral lens fit for the RE. [4,5]





Grade 2 meibomian gland atrophy RUL / RLL. Grade 1 meibomian gland atrophy LUL Grade 0 meibomian gland atrophy LLL

Initial Scleral Lens Evaluation 5/3/19:
Trial Lens Selection: Given the general steepness as demonstrated by topography and the extreme elevation difference (176 microns) combined with a relatively small cornea (HVID = 11.27mm) and narrow vertical fissure it was decided to select the smallest OAD available in the Acculens Maxim fitting set. Sizes in this set range from 16.0 to 18.2mm with sagittal clearance matched for each trial lens to each OAD.

Trial Lens Fitting: TL 1: Acculens Maxim 7 / BC 6.49 / SAG 4.89 / OAD 16.0 OZ 9.0 / -12.00 This is a "stock" design with a spherical haptic.
As per our SOP, my assistant evaluated the lens briefly to rule out decentration and air bubbles via visual examination. After our standard protocol of 1 hour of wearing time it was noted to move excessively and >700 microns of central clearance was observed.
Please Note: All clearance measurements in this case were made via slit lamp estimation. Proper Limbal clearance and Haptic-Scleral interaction were likewise made using slit lamp observation for this entire case. These observations will take the form of denoting whether or not there was observable limbal or scleral blanching and the location it occurred. We are defining blanching as inside of the landing haptic and not at the lens edge; otherwise referred to as "compression". This is specifically different from "impingement" which is located at the edge of the haptic.

TL 2: Acculens Maxim 7 / BC 7.03 / SAG 4.64 / OAD 16.0 / OZ 9.0 / -6.00 This trial lens was chosen to maintain the OAD but decrease the central sag. This lens was made with 5 limbal curves and 2 haptic curves. Each limbal curve is tangential and flatter as we traverse from the OZ to the haptic enabling us to attempt an average vault of 200 microns (see Table 3). After 1 hour of wearing time it was noted that the lens exhibited no movement, there was no limbal or scleral blanching, and central clearance was 350 microns, superior clearance was 200 microns; and inferior, nasal, and temporal clearance was 100 microns. The over refraction was 0.00 with 20/20-1 vision.

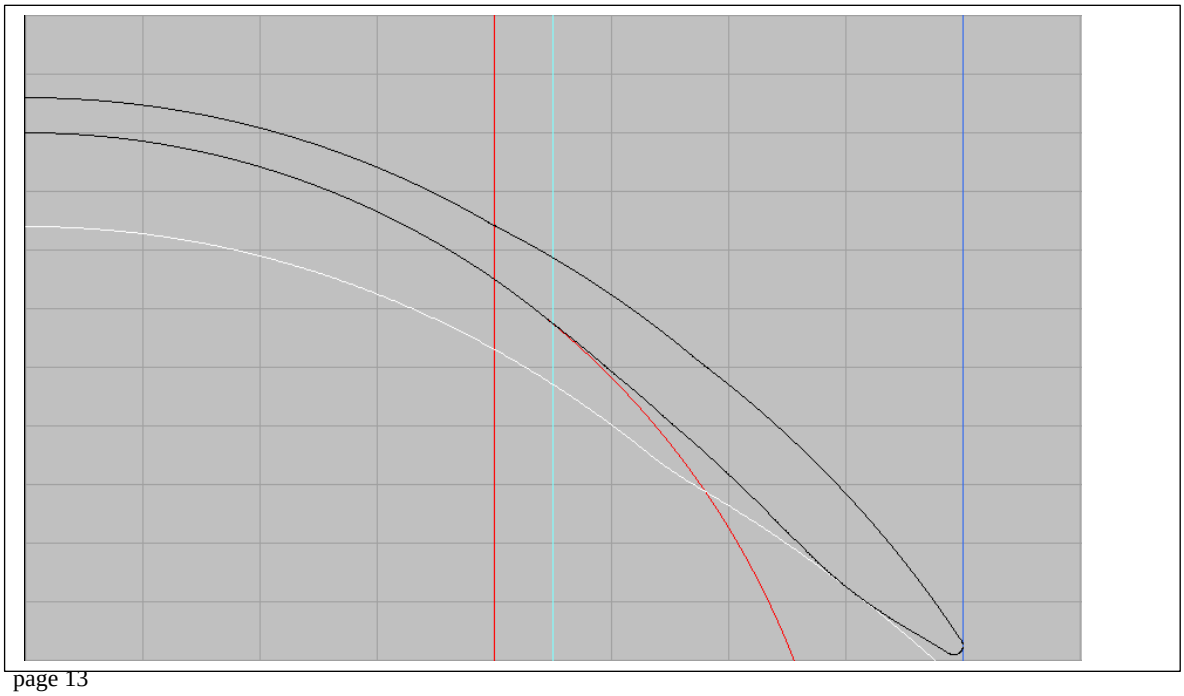
Lens Dispensing: Trial lens 2 was ordered and dispensed as **MR Lens #1** on **5/13/19**. Our SOP for scleral lens filling is single dose ampules of 0.9% inhalation saline. Additionally, we dispense all scleral lenses with Sauflon/Refine H2O2 disinfection systems. After 1 hour of wearing time, there was no limbal or scleral blanching. Clearances were observed as follows: 400

microns central, 300 microns superior and inferior, and 100 microns nasal and temporal, and 200 microns over the inferior mid-peripheral corneal irregularity previously noted. MR saw 20/30 with only +.25 over-refraction which improved acuity to 20/25-1. Follow up was set at 1 week with a standard wearing time buildup of: Day 1= 6 hours, Day 2 = 8 hours, Day 3 = 10 hours, Day 4 = 12 hours and Day 5+ = 14-16 hours maximum wearing time.

Table 3: MR Lens #1

BC Radius	7.03
Sag	4.64
OAD	16.0
OZ	9.0
Power	-6.00
CT	.25
Limbal Curve (1) Radius / Width	7.50 / .5
Limbal Curve (2) Radius / Width	8.04 / .5
Limbal Curve (3) Radius / Width	8.65 / .5
Limbal Curve (4) Radius / Width	8.88 / .45
Limbal Curve (5) Radius / Width	9.38 / .5
Haptic Curve (1) Radius / Width	13.50 / .5
Haptic Curve (2) Radius / Width	15.00 / .55

CAD profile MR Lens #1



Follow up 5/31/19 with 6.5 hours wearing time. MR was seeing 20/25 with 0.00 over-refraction. He reported that although he was "seeing better, (my) eyes seem more red." Clearances were observed as follows: 350 microns central, 100 microns inferior, 200 microns superior, 100 microns temporal, 50 microns nasal and 150 over the inferior mid-peripheral corneal irregularity previously noted. There was slight temporal scleral blanching inside of the haptic edge. Using a "measure twice-cut once" philosophy it was decided to recheck all findings prior to redesign so another appointment was set.

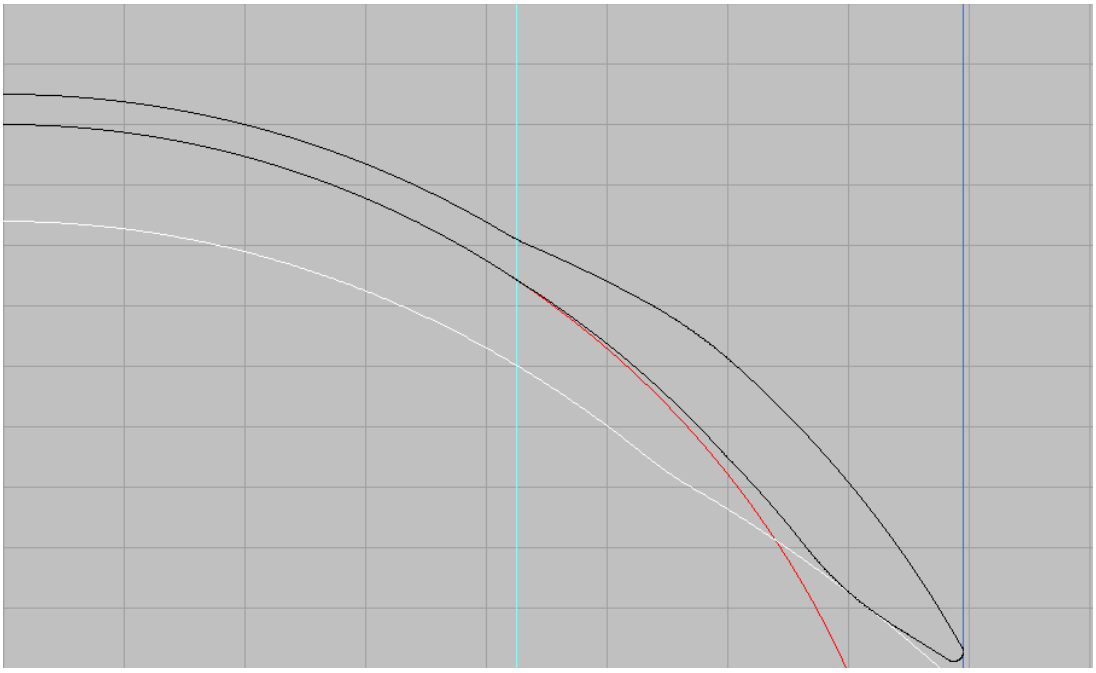
Follow up 6/5/19: MR came in for follow up again with 6.5 hours wearing time. He reported that his vision was "slightly blurry today". His vision was 20/30-2 but felt that a -.50 over-refraction improved his quality of vision. Clearances were observed as follows: 300 microns central, 100 microns inferior, 100 microns superior, 50 microns temporal, 50 microns nasal with touch over the inferior mid-peripheral corneal irregularity previously noted. Although there was no limbal or scleral blanching, there was 2+ conjunctival injection under the haptic with 3+ bulbar conjunctival injection. It was decided to re-design MR's lens as a more prolate design to loosen the haptic. In this context, we wanted to flatten the lens centrally (which in turn allowed for a decrease in optical power via a more minus tear lens to improve VA) and go to 2 limbal curves which were flatter and wider to achieve a faster rate of peripheral lift (see Table 4).

Follow up 8/23/19: MR came into the office to have MR Lens #2 dispensed and evaluated. After 45min of wearing time, MR's vision was 20/25. There was 500 microns of central clearance, 400 microns inferior, 300 microns superior, 200 microns temporal and 100 microns nasal. There was no limbal or scleral blanching and no excessive bulbar conjunctival injection. The patient reported Lens #2 "felt better like it was less tight". Follow up was set for 2 weeks.

Table 4: MR Lens #2

BC Radius	7.67
Sag	4.63
OAD	15.9 (finished diameter)
OZ	8.5
Power	-2.50 (includes 6/5/19 over refraction)
CT	.25
Limbal Curve (1) Radius / Width	8.04 / 1.75
Limbal Curve (2) Radius / Width	8.44 / .90
Haptic Curve (1) Radius / Width	13.00 / .5
Haptic Curve (2) Radius / Width	14.50 / .55

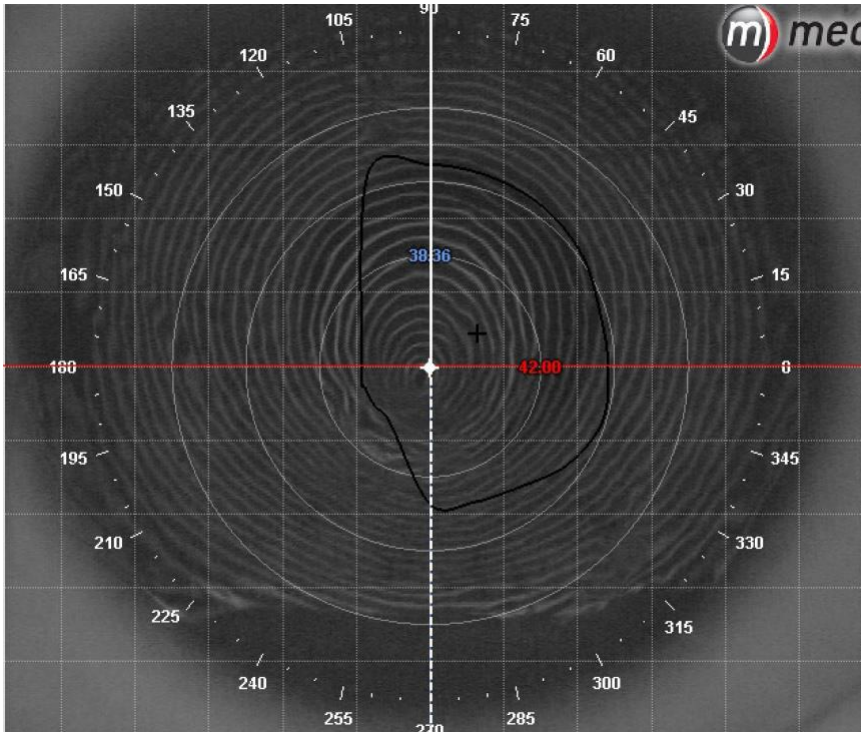
CAD profile MR Lens #2



Follow up 9/6/19: MR came in for a follow up visit. He reported that the new lens "feels better, not as red, and vision is good". His vision was 20/30-1 after 7 hours of wearing time. Lens clearances were 100 microns central, 250 microns inferior, 100 microns superior, 250 microns temporal and 100 microns nasal. There was no limbal or scleral blanching and only a trace amount of bulbar conjunctival injection. Follow up was set for 4 months.

Follow up 1/29/20: we saw MR at the end of 7 hours of wearing time. He reported that his vision was good, he was seeing 20/50; and the "lens felt good except for a little dryness by the end of the day". Clearances were 200 microns central, 100 microns inferior and superior, 50 microns temporal and 100 microns nasal. Once again, there was only a trace amount of bulbar conjunctival injection and no limbal or scleral blanching. It was decided to deep clean his lens with progent at this visit and set follow up at the time of his regular yearly comprehensive exam.

Follow up 1/28/21: MR came into our office for a yearly progress appointment. He reported that scleral lens wear "really helps and (my) vision is more clear". With the exception of some mask related dryness and a decrease of light sensitivity and redness, MR also reported a resolution of all pre-fit symptoms. His RE visual acuity at 12.5 hours wearing time was 20/30+1 with a -.50 over-refraction which only improved VA by one letter. There was no limbal blanching, but we observed trace scleral blanching from 7-11:00 along with 3+ bulbar conjunctival injection (this was at 4+ pre-fit). Clearances were 250 microns central, 150 microns inferior, 100 microns superior, 100 microns temporal and 100 microns nasal. Limbal clearances were estimated to be 100 microns inferior, 50 microns superior, 50 microns temproal and 50 microns nasal. We performed a progent deep cleaning and he was pre-appointed for a 6 month return visit. Pictured below is the topography mire pattern for MR's RE cornea from this visit. It demonstrates slightly less corneal distortion than in the orignal, pre-fit map via the use of a scleral contact lens.



RE topography and mire pattern (20 months post-fit)

Appendix 1: Commonly accepted abbreviations

DED dry eye disease
TFOS DEWS tear film ocular surface dry eye workshop
IPL intense pulsed light therapy
LLET low level laser therapy
RF radio frequency
DM diabetes mellitus
RUL right upper lid
RLL right lower lid
RE right eye
LE left eye
OU ocular unitas or both eyes
VA visual acuity
NPC near point of convergence
PHNI pin hole no improvement
mmHg millimeters of mercury
APD afferent pupillary defect
NVD neovascularization of the optic disc
NVE neovascularization everywhere
CSME clinically significant macular edema
BDR background diabetic retinopathy
1gtt OU bid 1 drop both eyes twice daily
TL trial lens
HVID horizontal visible iris diameter
OAD overall diameter
BC base curve
SAG sagittal height
OZ optic zone
SOP standard operating procedure
H2O2 hydrogen peroxide

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