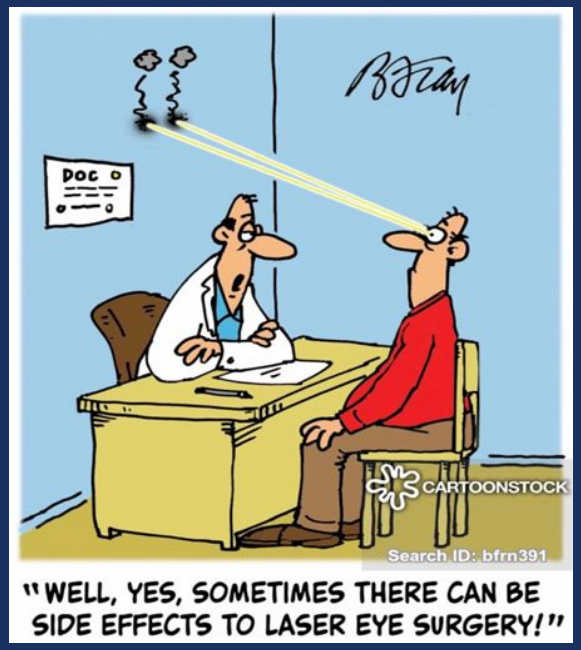
# CONTACT LENS FITTING FOR POST-SURGICAL COMPLICATIONS

MARCUS NOYES, FAAO, FSLS & BRYAN WILLIAMS, OD, FAAO, FSLS

# BACKGROUND

- Texas Tech University
  - Class of 2014

- University of Houston College of Optometry
  - Class of 2018

- UAB School of Optometry
  - Cornea & Contact Lens Resident 2018-2019

# FINANCIAL DISCLOSURES

- None

UNI University of Northern Iowa

THE OHIO STATE UNIVERSITY
WEXNER MEDICAL CENTER

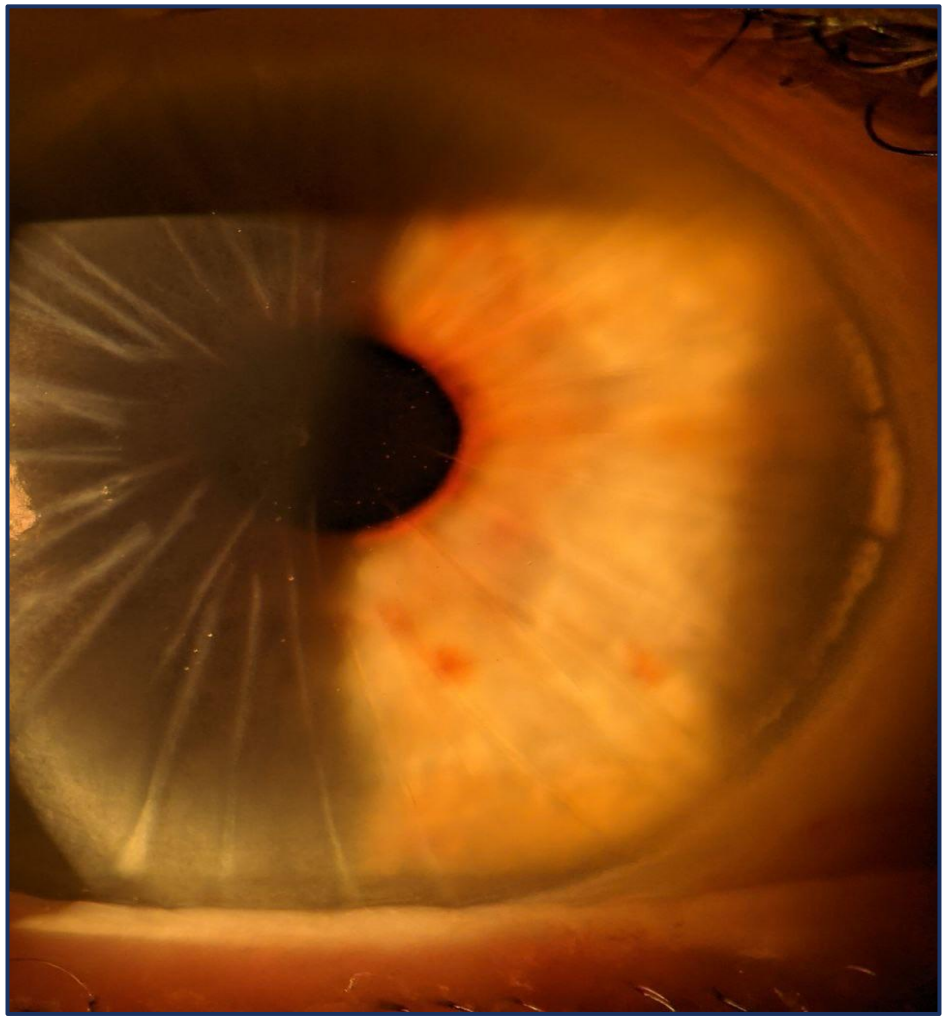
# FINANCIAL DISCLOSURES

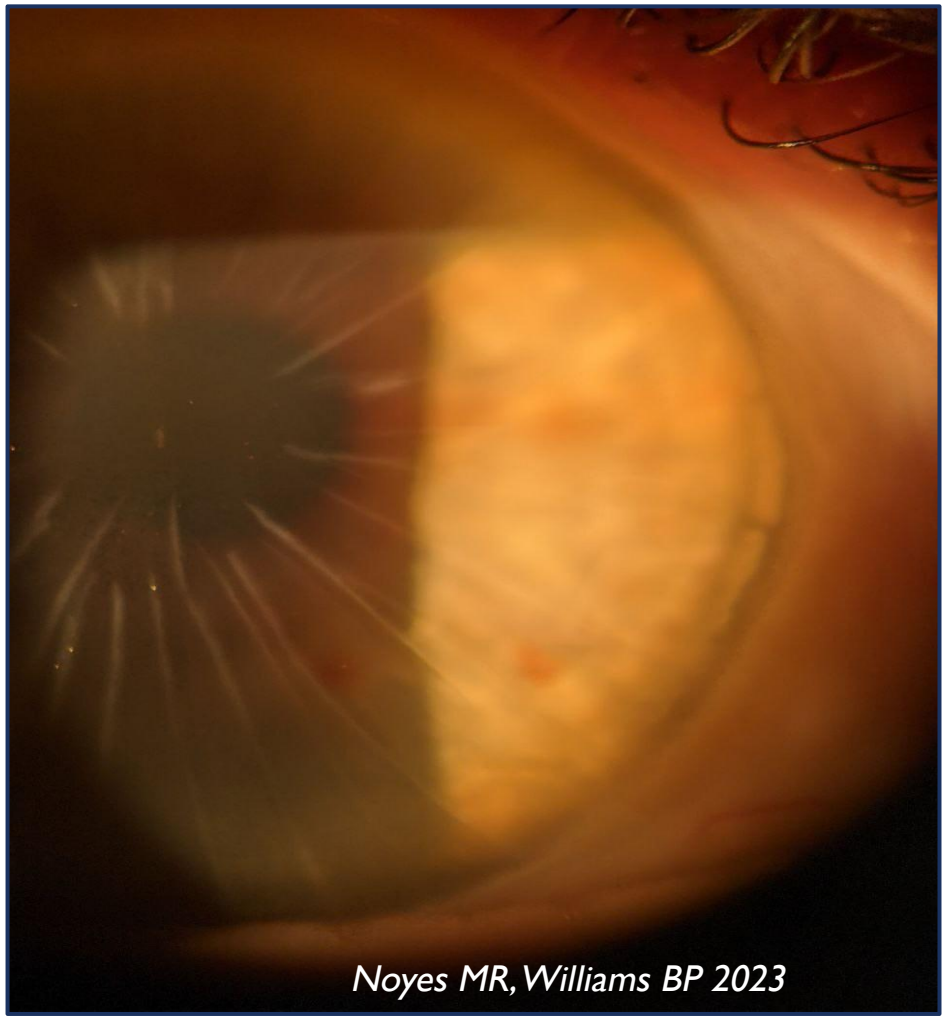
- None

# LECTURE OBJECTIVES

- **1**. Attendees will learn clinical pearls about managing patients that have undergone radial keratotomy surgery.
- **2**. Attendees will learn clinical pearls about managing patients that suffer from exposure keratitis.
- **3**. Attendees will learn clinical pearls about managing patients that have undergone corneal surgery to repair a corneal laceration.
- **4**. Attendees will learn ways to trouble-shoot obstacles that present themselves when trying to complete a complex scleral contact lens fitting.
- **5**. Sometimes you can do everything right, and things still go wrong (Case #5 specifically)
- **6**. Remember, you are treating the patient! Not the lens! If a "perfect fit" doesn't optimize the patient's health... Is it even worth it?

# CASE 1 – POST-RADIAL KERATOTOMY





# CASE HISTORY

- **CHIEF COMPLAINT**
  - Blurred vision at distance and near
  - Referred by cornea specialist for scleral contact lens evaluation
- **DEMOGRAPHICS**
  - 58yo WF
- **OCULAR HISTORY**
  - Bilateral RK surgery in 1991
  - Dry Eye Syndrome
  - Bilateral early cataracts
- **OCULAR MEDICATIONS**
  - None

# CASE HISTORY

- **MEDICAL HISTORY**
  - Thyroid disease
  - Sjogren's Syndrome
  - Hypertension
  - Elevated Cholesterol
  - Arthritis
  - Migraines
  - History of COVID related complications (hospitalized in 8/2021 as a result)
  - Seizure disorder

- **COMANAGEMENT**
  - Currently being followed by a neuro-optometrist as well as a cornea specialist

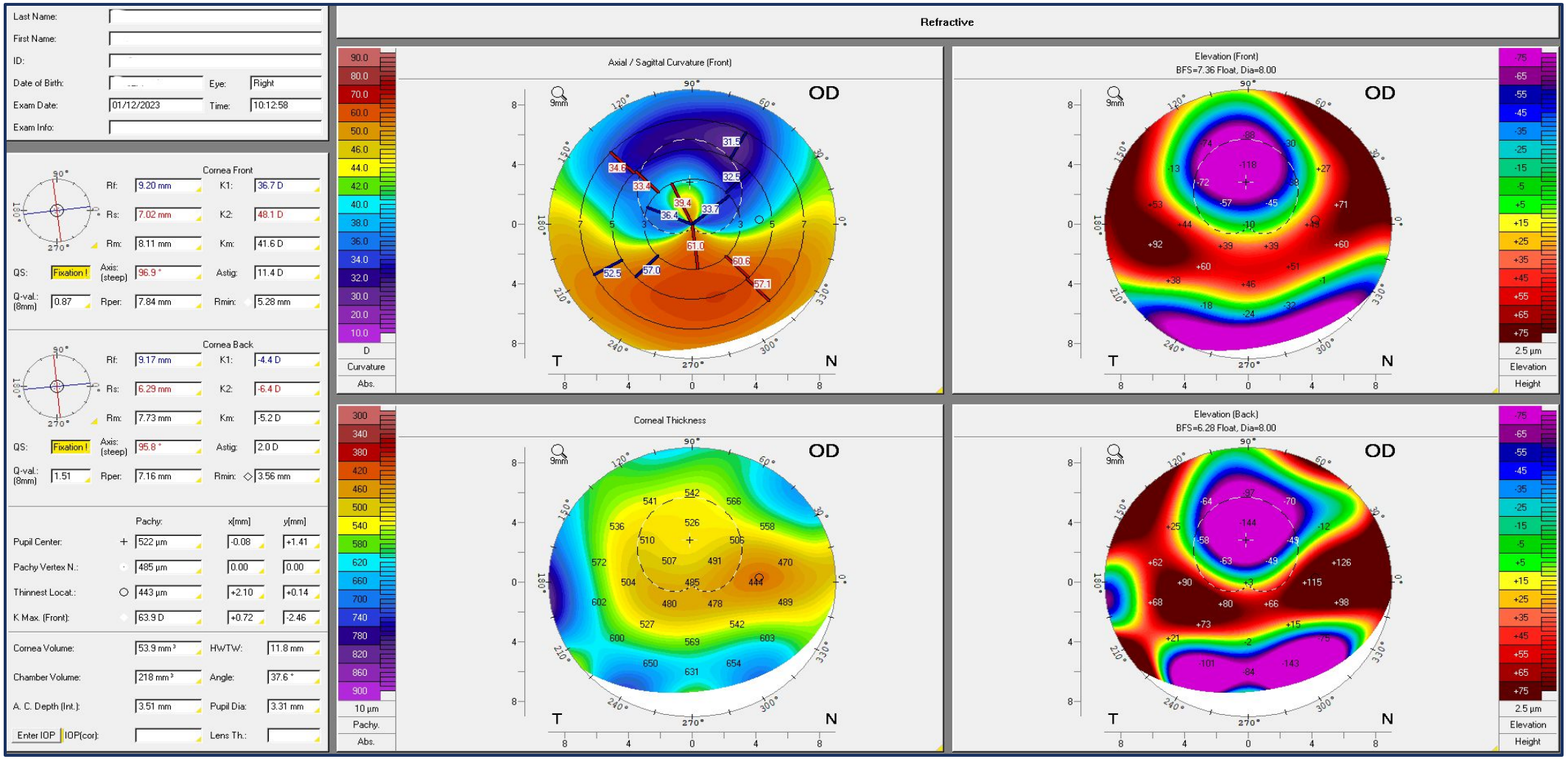
# EXAM FINDINGS

- **ACUITIES**
  - OD with glasses: 20/40--
  - OS with glasses: 20/50--
- **HABITUAL SPEC RX**
  - OD: +3.25-1.50x126
  - OS: +6.25-1.50x161
- **KERATOMETRY**
  - OD Kmax: 63.9D
  - OS Kmax: 68.5D
- **PACHYMETRY**
  - OD minimum: 443um
  - OS minimum: 407um
- **MANIFEST SPEC RX**
  - OD: +3.25-1.50x126
  - OS: +6.25-1.50x161
- **High Exophoria at distance and near**

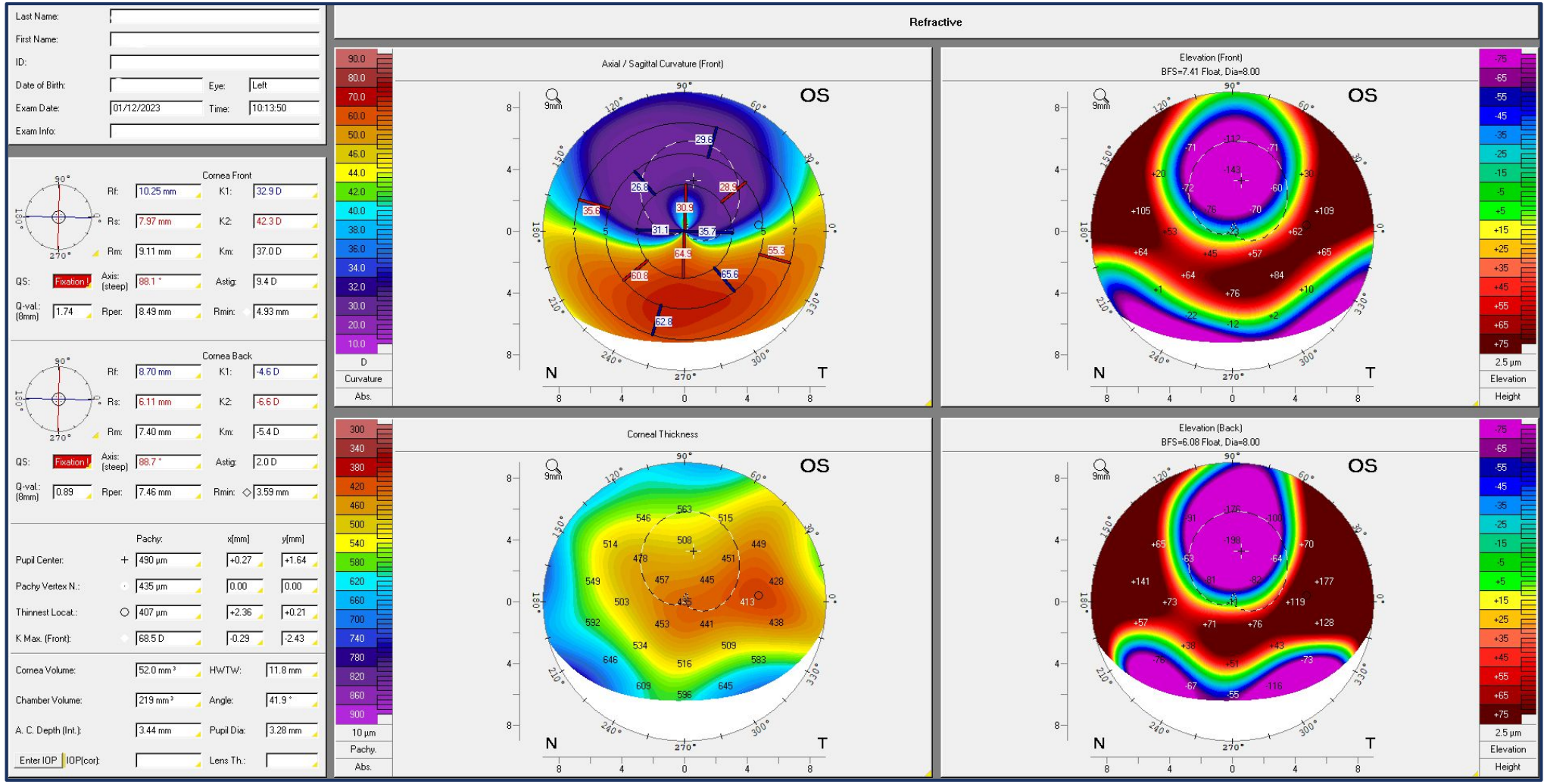
# EXAM FINDINGS

- **SLIT LAMP**
  - OU: 1+ MGD
  - OU: 16 cut RK
  - OU: 1-2+ NS
- **IOP/FUNDUS EXAM**
  - OU: WNL

# PENTACAM SCANS



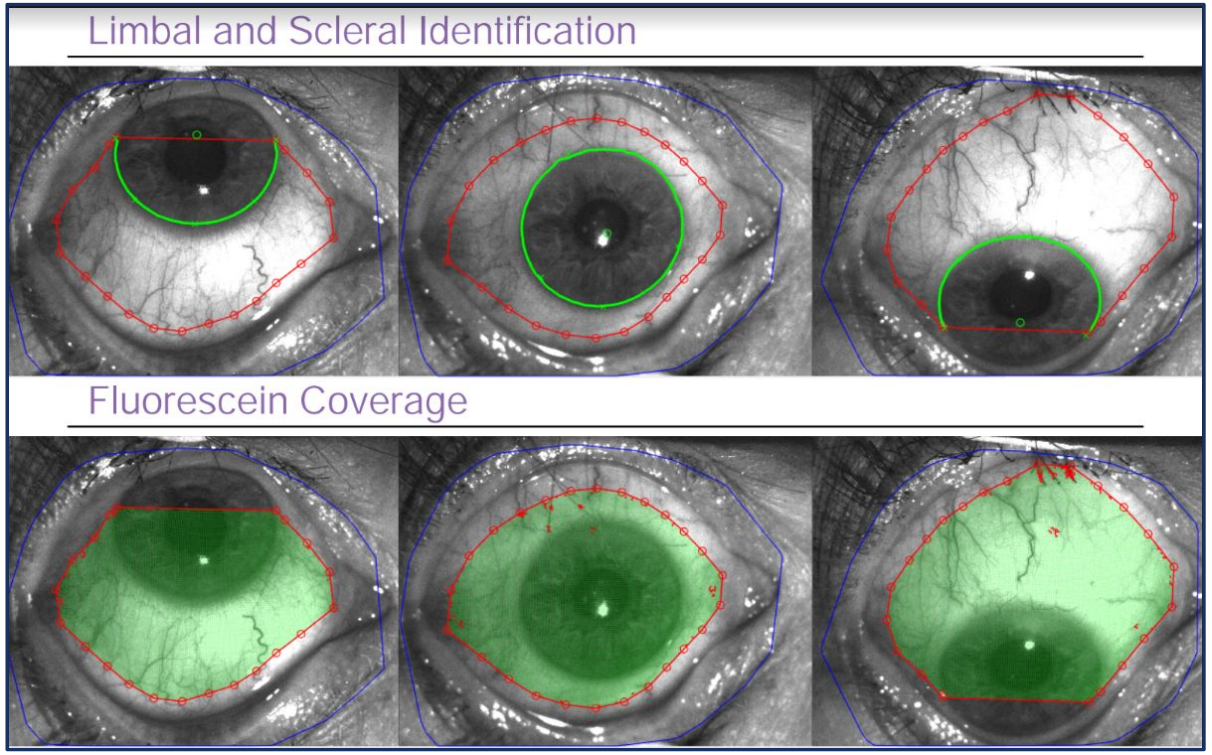
# PENTACAM SCANS

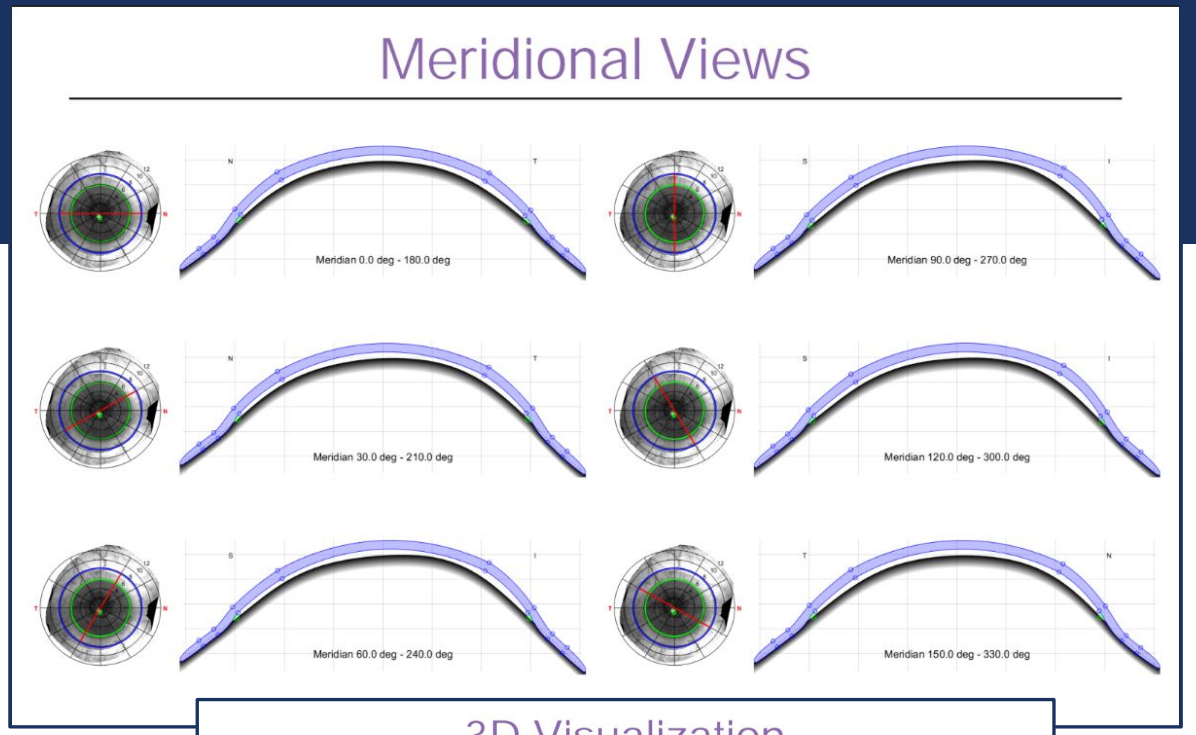


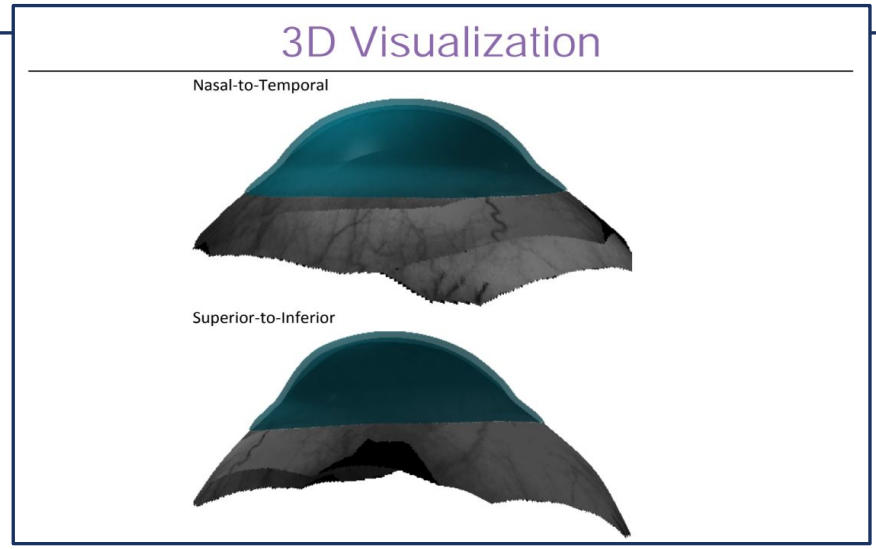
# CONTACT LENSES

- **1st TRIAL LENS**
  - OD - Europa
    - BC: 7.5D
    - Diameter: 16.0mm
    - PWR: -1.50
    - SAG: 4560
  - OS – Europa
    - BC: 7.34
    - Diameter: 16.0
    - PWR: -2.00
    - SAG: 4660
- **FIT**
  - **OD:** Adequate central and limbal clearance, well centered, stable
    - **ORx:** -8.00, 20/40-
  - **OS:** Adequate central and limbal clearance, well centered, stable
    - **ORx:** -10.00, 20/40-
- **PLAN**
  - Order lenses based off SMap3D scan OU
  - Return for A&R training when lenses arrive

# CONTACT LENS – OD

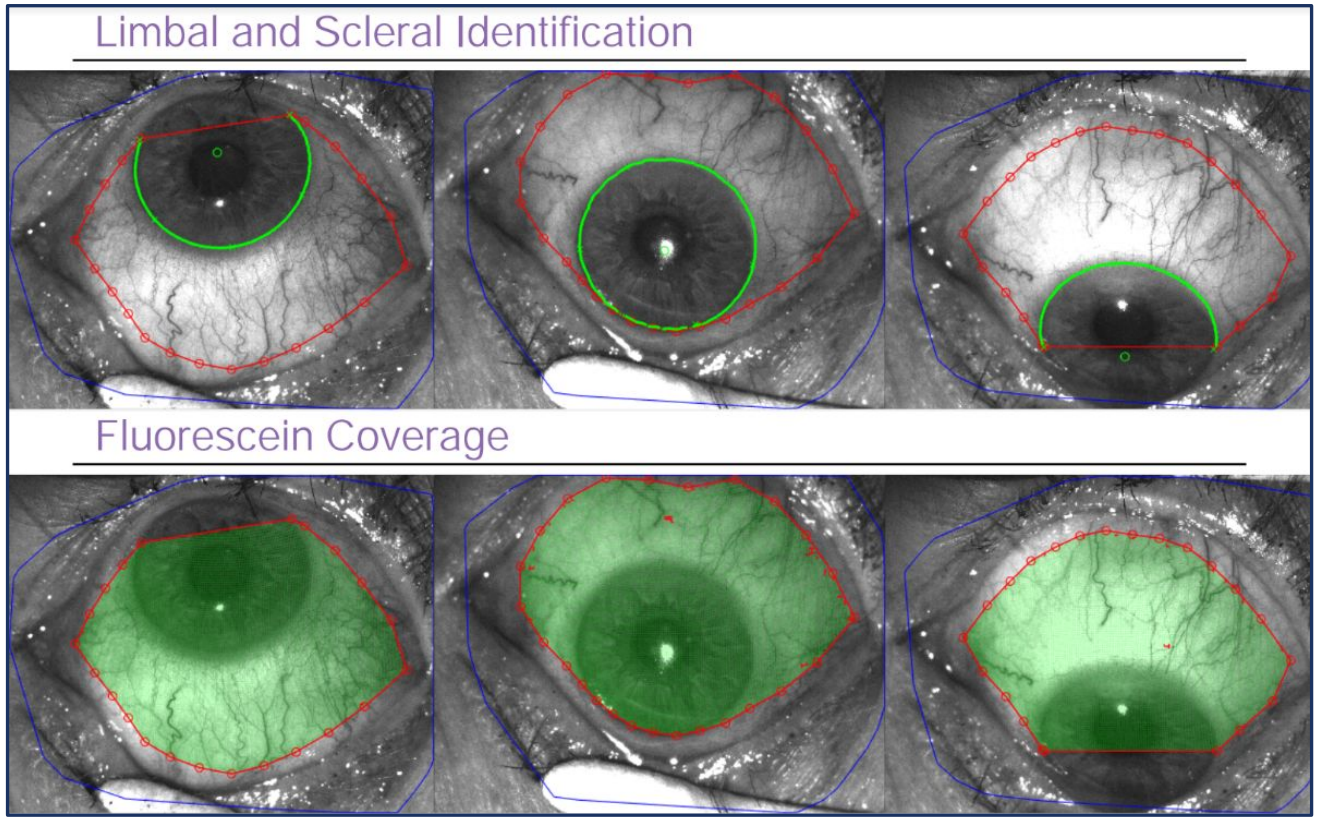


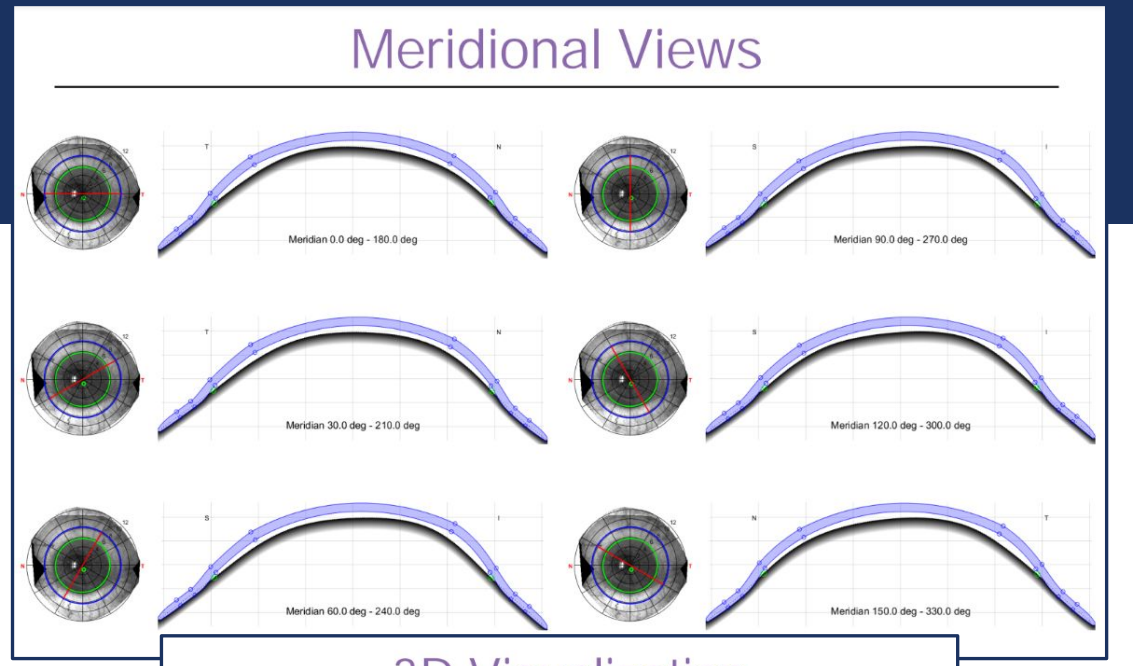



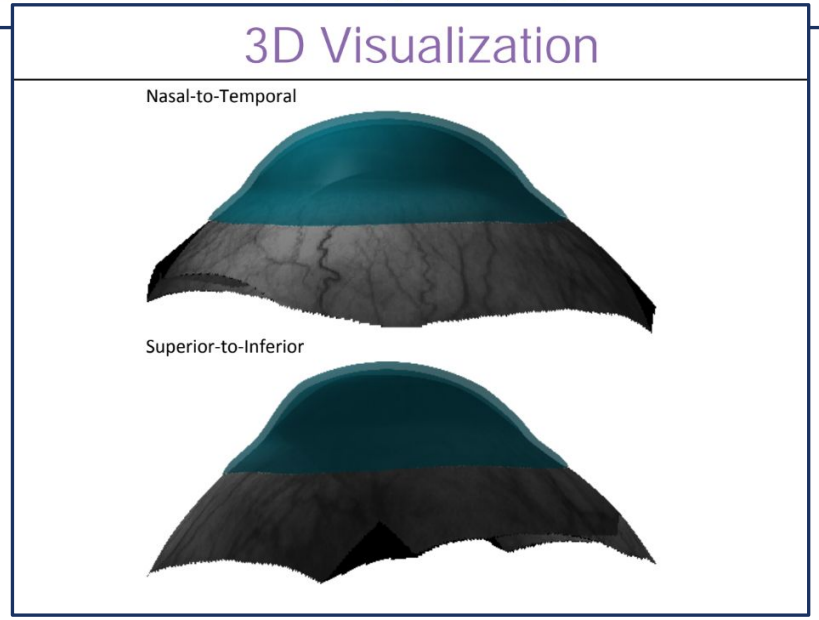




Diameter: 16.50mm, SAG: 5093, BC: 8.71, Rx: -2.50D, Freeform PCs

# CONTACT LENS – OD




Diameter: 16.50mm, SAG: 4991, BC: 8.99, Rx: -2.41D, Freeform PCs

# CONTACT LENSES – DISPENSE APPT 1/23/23

- **First Latitude Lens Fit OD**
  - Between 275-300um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/30-

- **First Latitude Lens Fit OS**
  - Between 275-300um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: +1.00, 20/25

- **A&R training Completed successfully**
- **Both CLs dispensed**
- **No lens changes made at this time**
- **Plan to RTC in 1-2 weeks for CL F/U**

# CONTACT LENSES – FOLLOW UP APPT 2/23/23

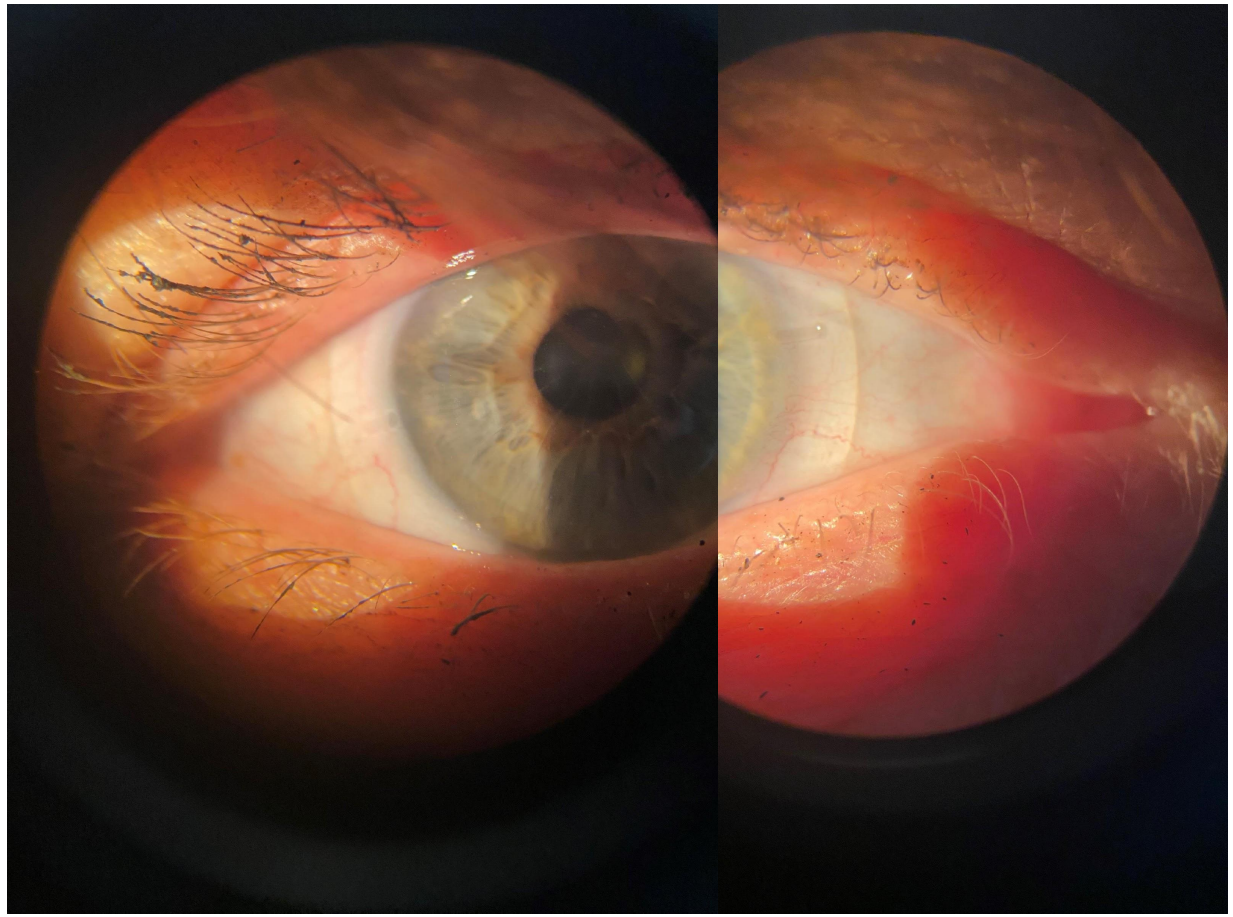
- **Patient reports she is happy with vision and comfort in both contact lenses**

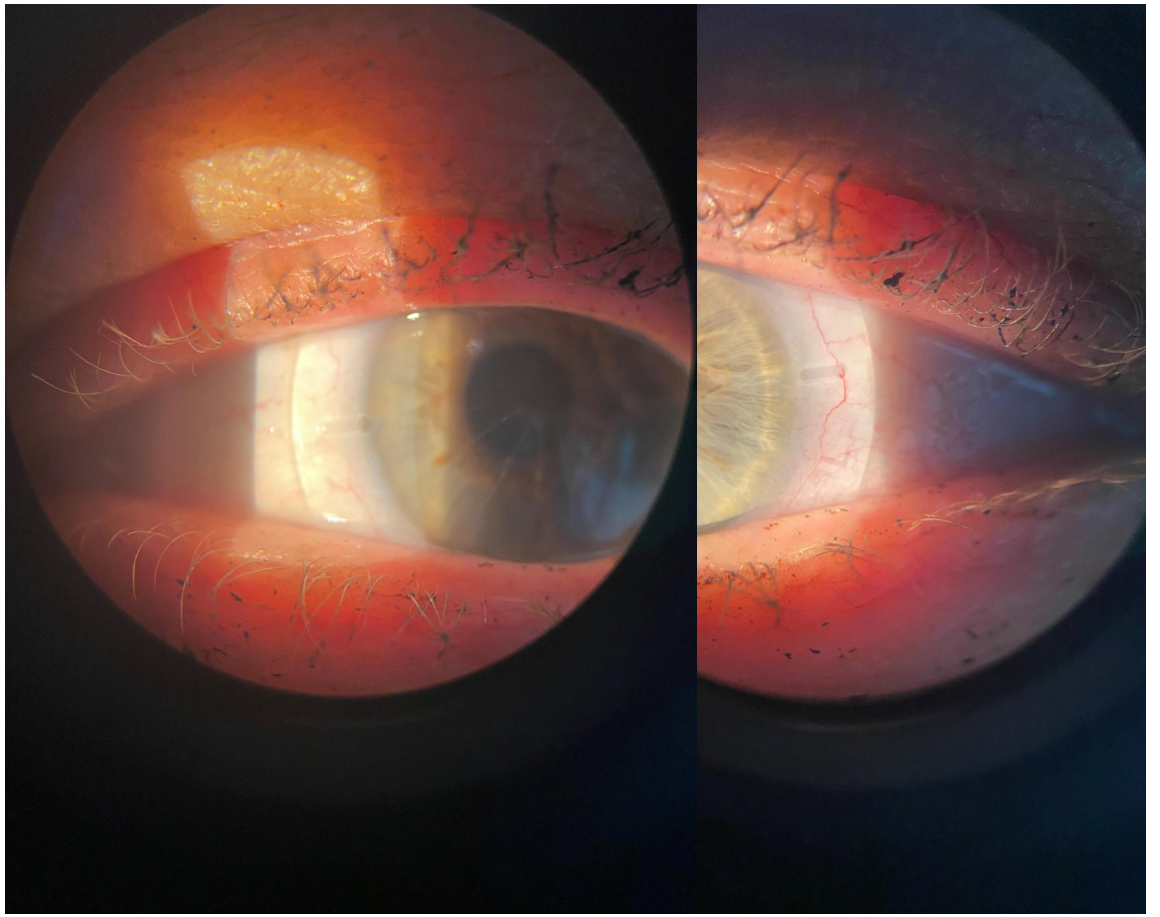
- **First Latitude Lens Fit OD** (4 hours wear time)
  - Between 225-240um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/40

- **First Latitude Lens Fit OS** (4 hours wear time)
  - Between 175-200um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: +1.25, 20/25--

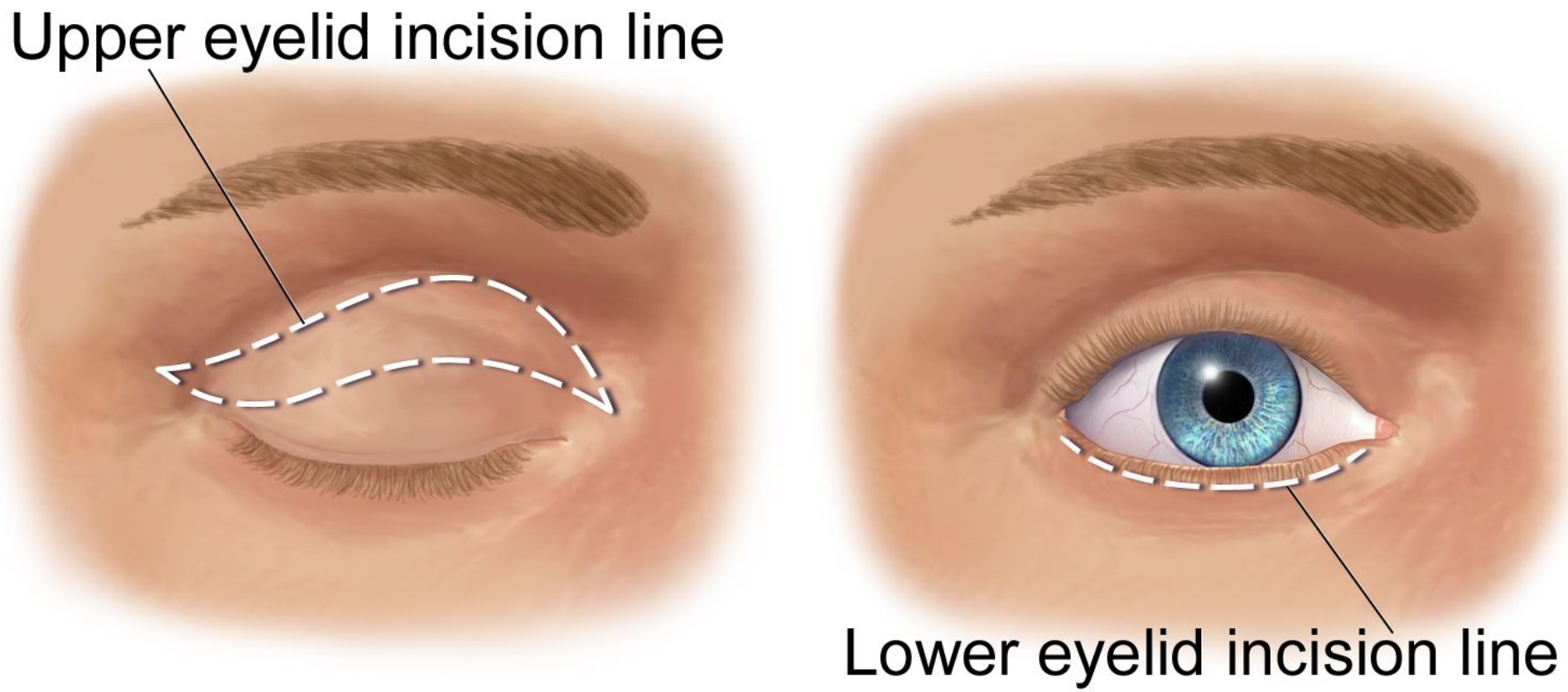
- **Good fit and vision with contact lenses**
- **Order ORx in left lens and ship to patient**
- **Plan to RTC in 2 months for CL F/U**

# CONTACT LENSES – FOLLOW UP APPT 2/23/23





# CASE 2 – LAGOPHTHALMOS AND EXPOSURE KERATITIS FOLLOWING BLEPHAROPLASTY SURGERY




Blepharoplasty

# CASE HISTORY

- **CHIEF COMPLAINT**
  - Severe dryness in both eyes
  - Referred by primary optometrist for scleral contact lens evaluation
- **DEMOGRAPHICS**
  - 71yo WF
- **OCULAR HISTORY**
  - Blepharoplasty on both upper eyelids
  - Lagophthalmos OU
  - Dry Eye Syndrome
  - Bilateral moderate cataracts
- **OCULAR MEDICATIONS**
  - Latisse 0.03%
  - RegenerEyes qid OU
- **MEDICAL HISTORY**
  - Thyroid disease
  - Rheumatoid Arthritis
  - Hypertension
- **SYSTEMIC MEDICATIONS**
  - Allopurinol
  - Estrogen
  - Fosamax
- **ALLERGIES**
  - Seasonal

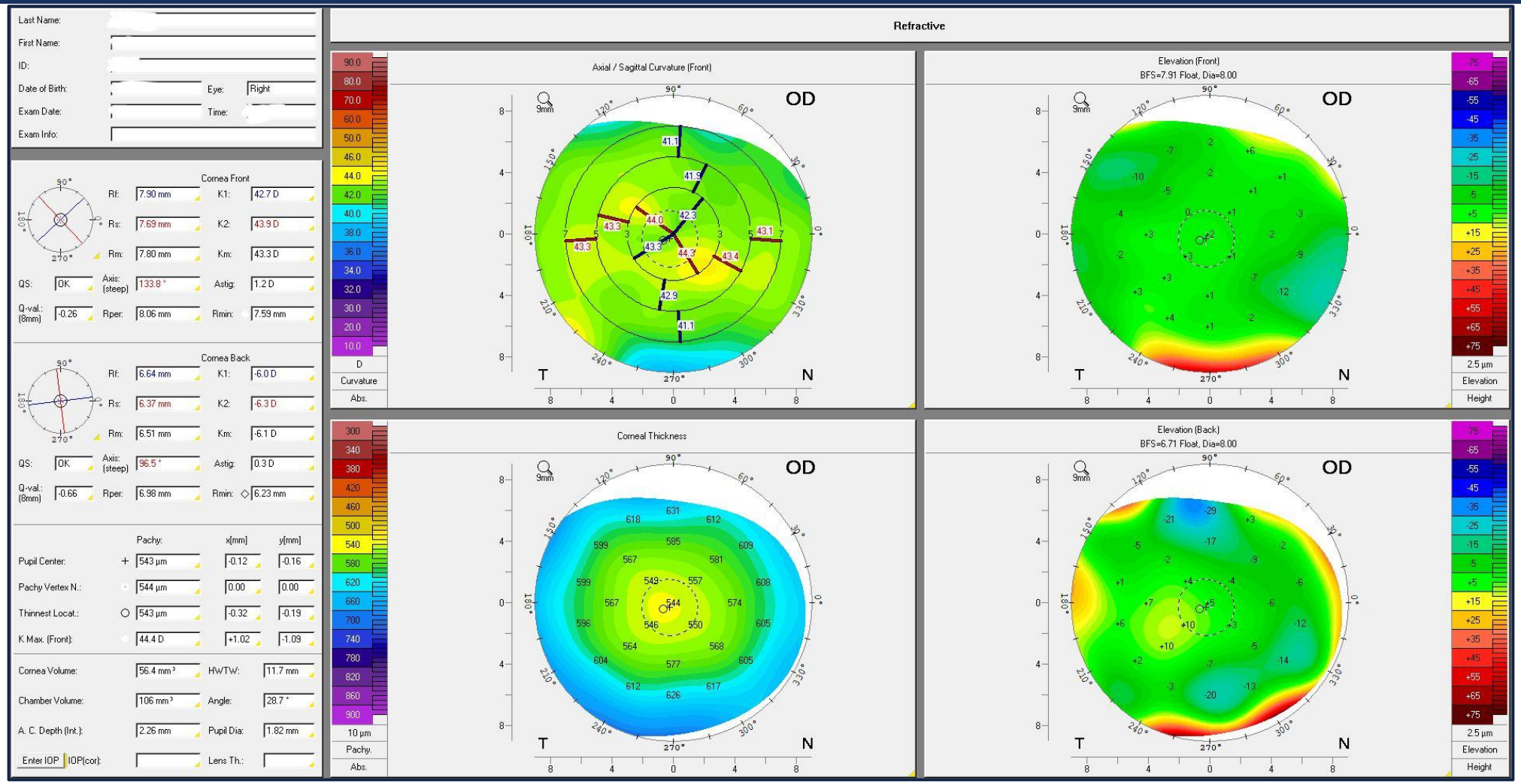
# EXAM FINDINGS

- **ACUITIES**
  - OD unaided: 20/30
  - OS unaided: 20/30
- **KERATOMETRY**
  - OD Kmax: 63.9D
  - OS Kmax: 68.5D
- **PACHYMETRY**
  - OD minimum: 443um
  - OS minimum: 407um
- **MANIFEST SPEC RX**
  - OD: +0.25-0.75x055, 20/30
  - OS: Plano-0.75x110, 20/30
- **IOP**
  - 13 OU with Goldmann

# EXAM FINDINGS

- **SLIT LAMP**
  - OU: 3+ MGD, heavy makeup debris on lashes and eyelid margin
  - OU: incomplete blinks covering ~2/3 of cornea
  - OU: scarring inferiorly, RTBUT, @+ SPK inferiorly
  - OU: 2+ NS
- **FUNDUS EXAM**
  - OU: WNL

# PENTACAM SCANS



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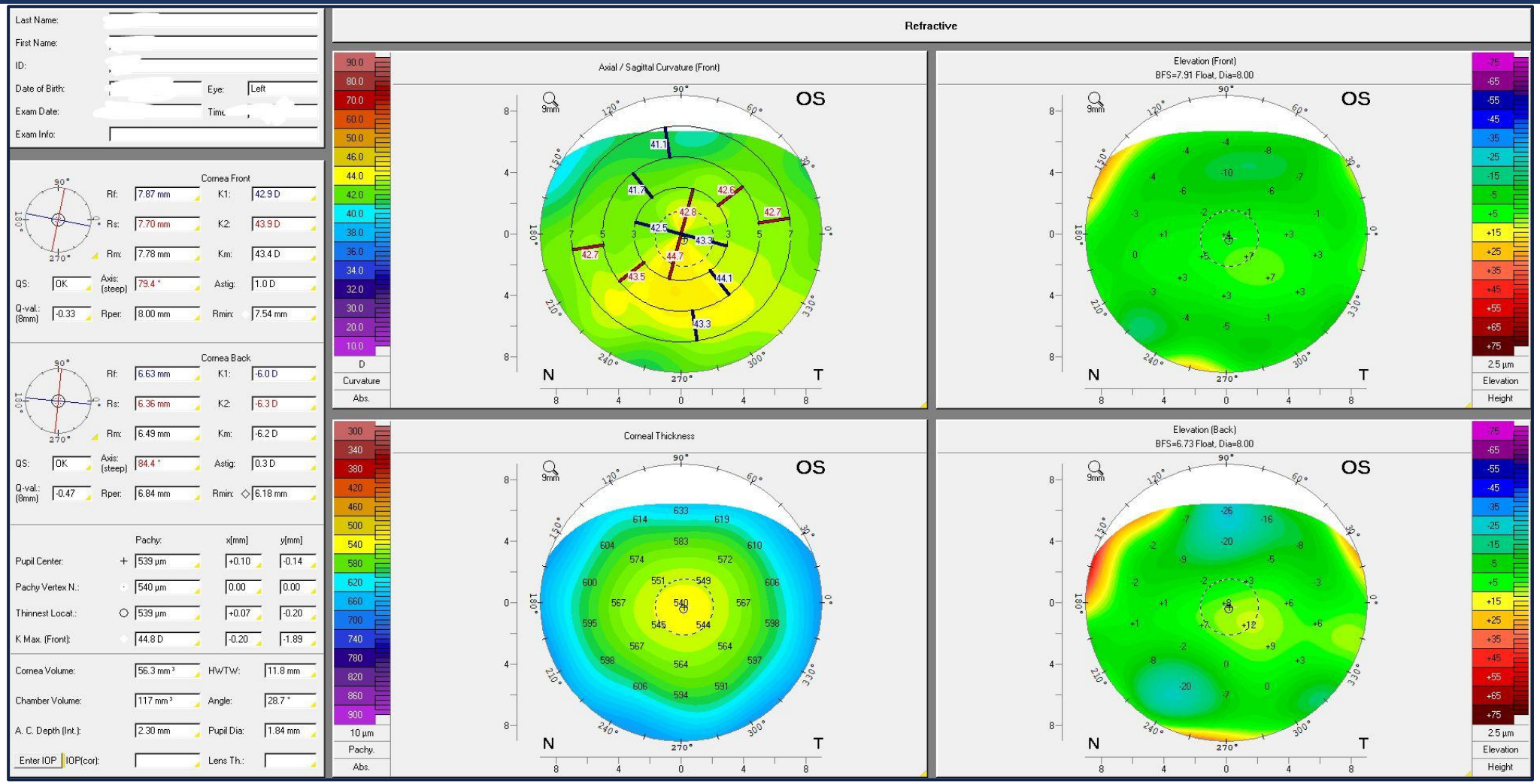
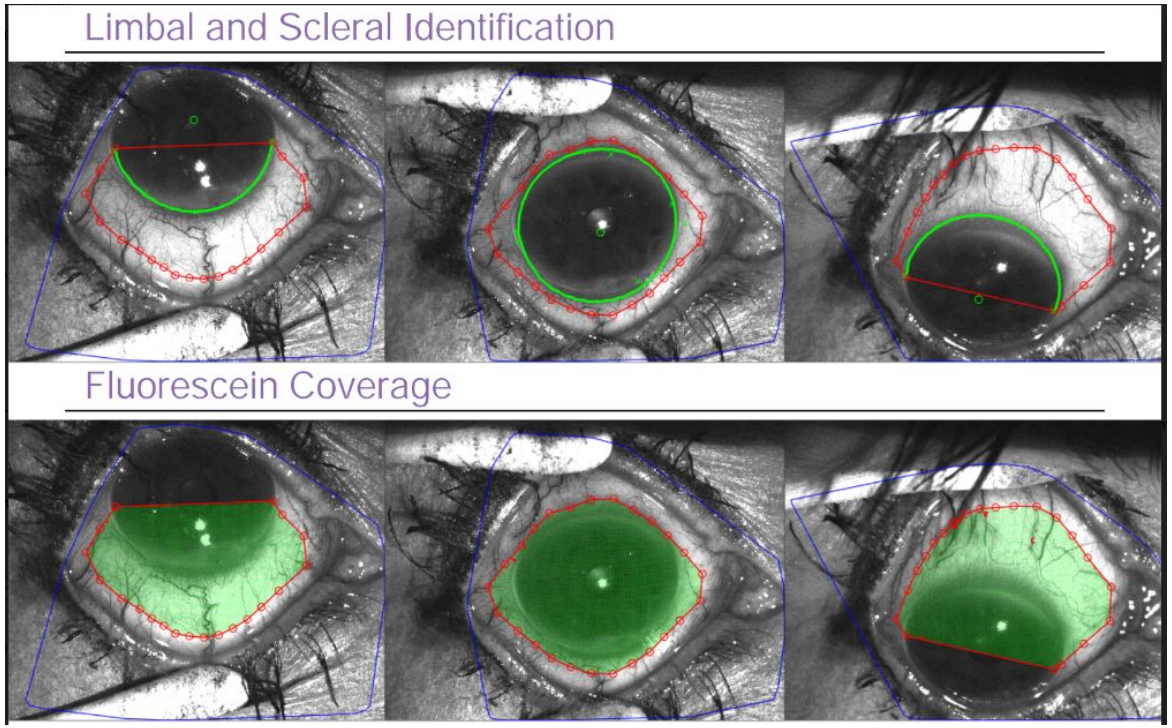
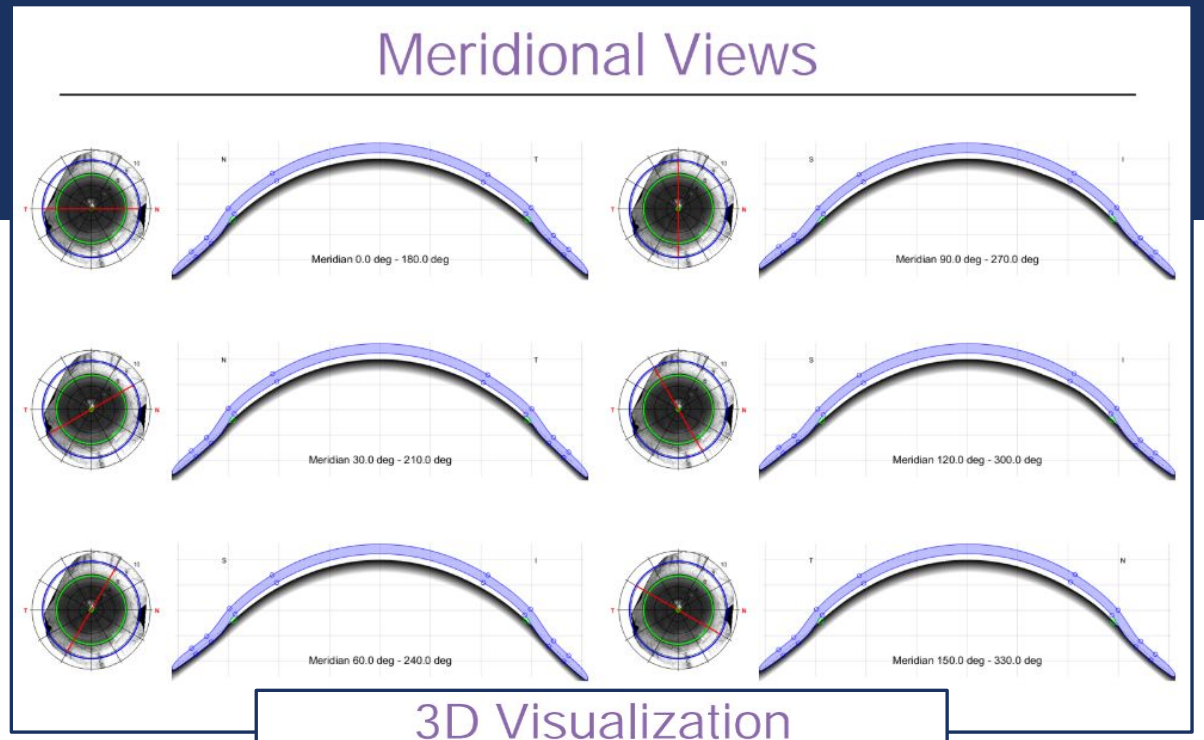


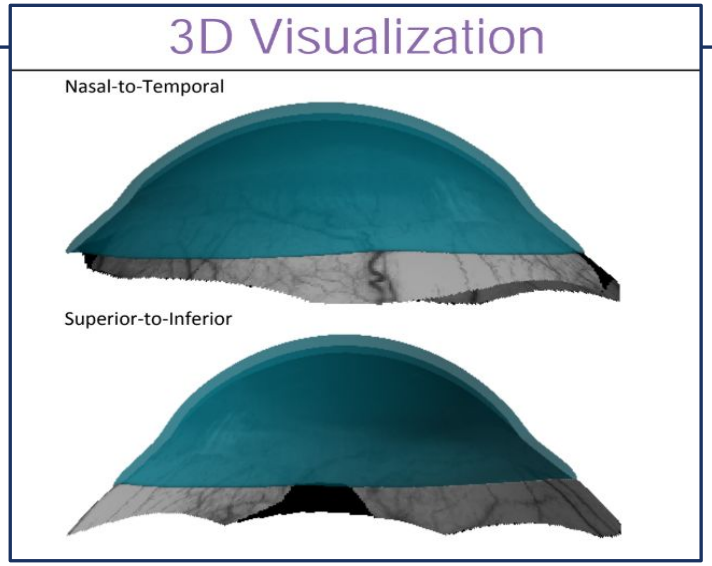
# CONTACT LENSES

- **1st TRIAL LENS**
  - OD - Europa
    - BC: 7.67D
    - Diameter: 16.0mm
    - PWR: -1.00
    - SAG: 4470
  - OS – Europa
    - BC: 7.85
    - Diameter: 16.0
    - PWR: -0.50
    - SAG: 4390
- **FIT**
  - **OD:** 450um central clearance, adequate limbal clearance, well centered, stable
    - **ORx:** -0.25, 20/20
  - **OS:** 150um central clearance, adequate limbal clearance, well centered, stable
    - **ORx:** -0.25, 20/20
- **PLAN**
  - Order lenses based off SMap3D scan OU
    - Aim for monovision, OD distance/OS near
  - Return for A&R training when lenses arrive

# CONTACT LENS – OD

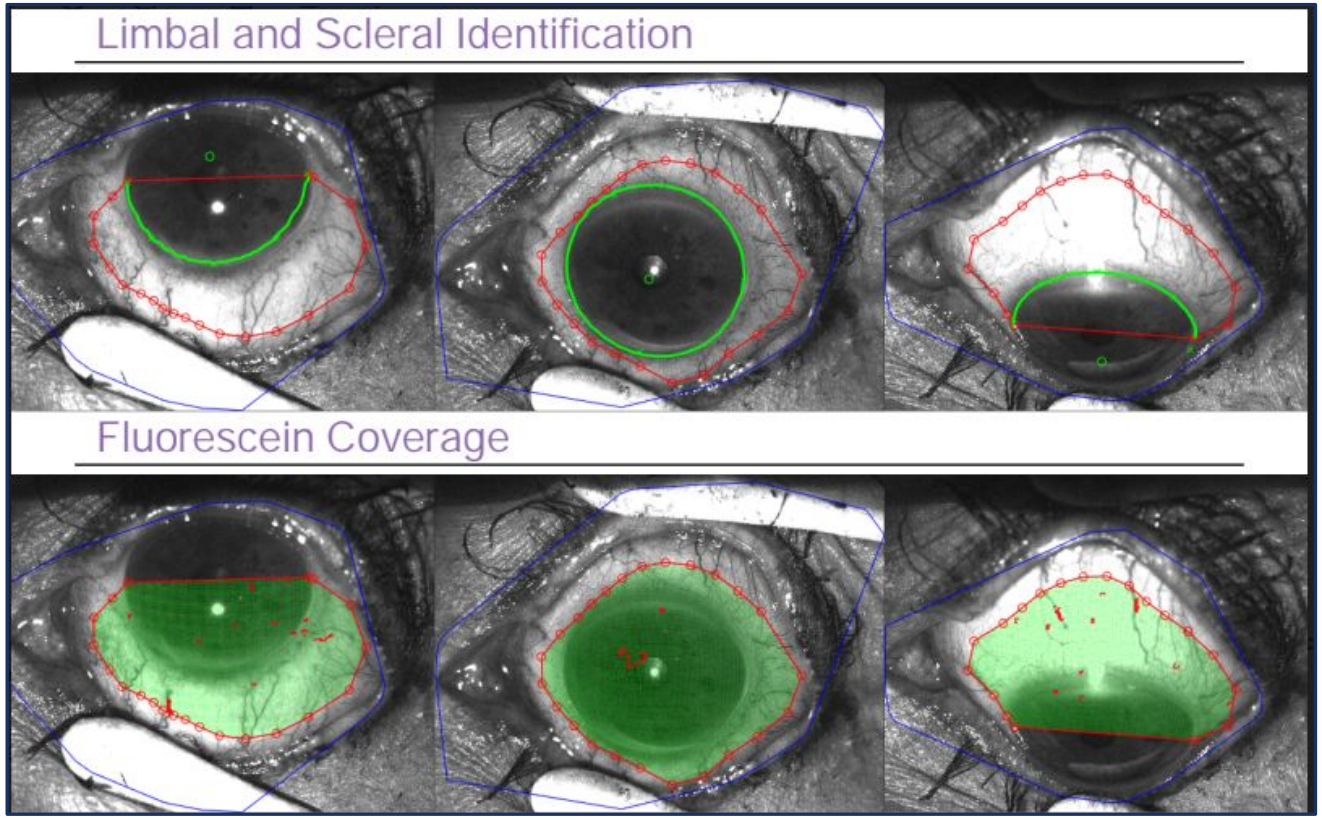


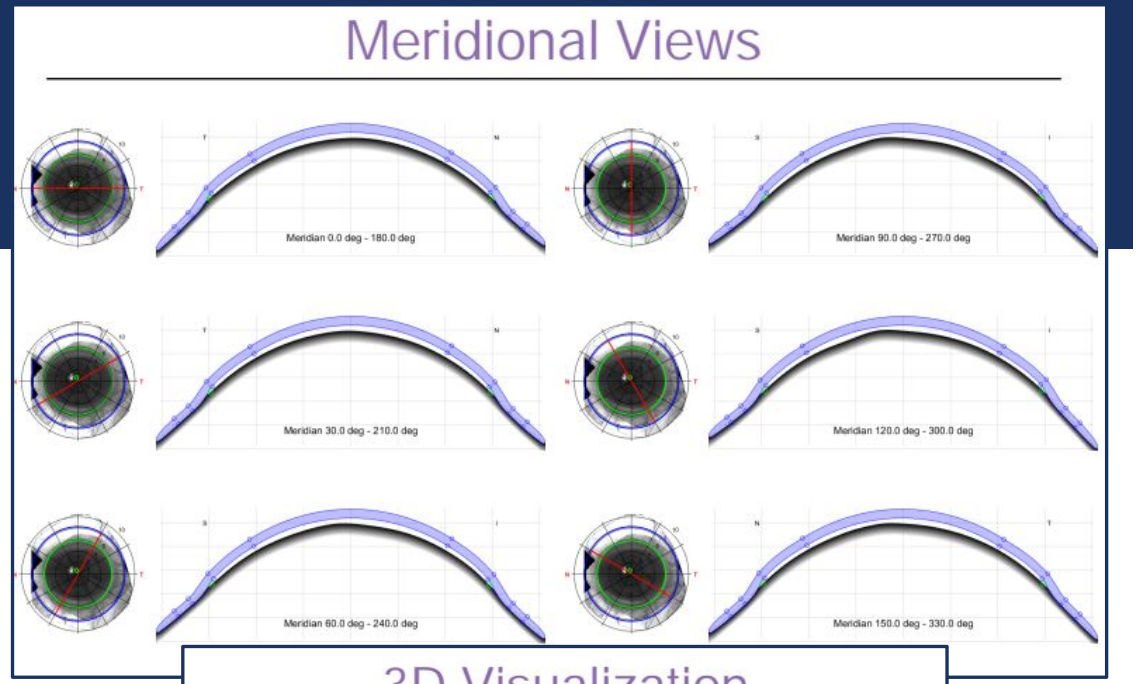



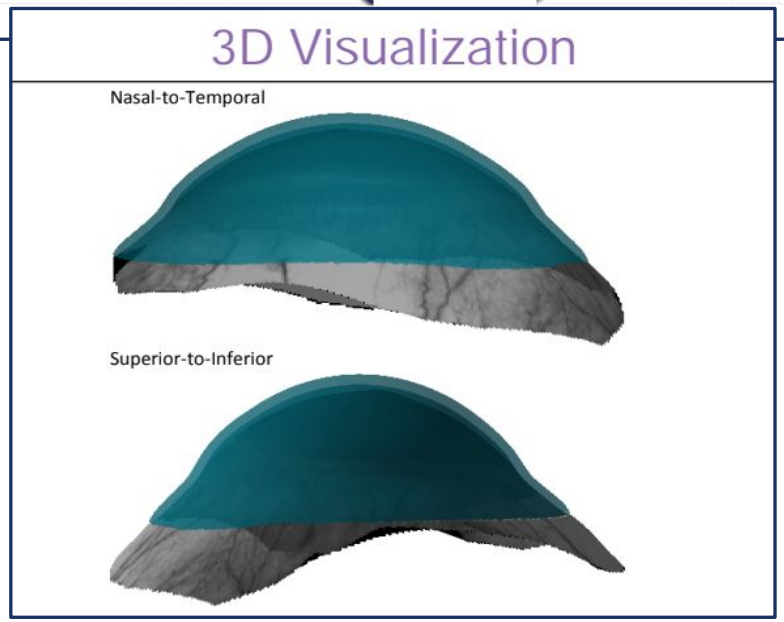




Diameter: 16.50mm, SAG: 5068, BC: 7.836, Rx: -0.32D, Freeform PCs

# CONTACT LENS – OD




Diameter: 16.50mm, SAG: 5267, BC: 7.642, Rx: -0.66D, Freeform PCs

# CONTACT LENSES – DISPENSE APPT 2/07/23

- **First Latitude Lens Fit OD**
  - 350um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/20

- **First Latitude Lens Fit OS**
  - 450um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/25 near; -1.25, 20/20- distance

- **A&R training completed but patient greatly struggled with application, instructed to use PFAT gel to apply lenses**
- **Both CLs dispensed**
- **No lens changes made at this time**
- **Plan to RTC in 1-2 weeks for CL F/U**

# CONTACT LENSES – PHONE CALL 2/12/23

- **Patient called reporting they had not yet successfully been able to apply the lenses on their own at home**
- **Voiced that she thought she would be more successful if the CLs were smaller**
- **Consulted with Visionary Optics and designed a 15.0mm diameter CLs OU**
- **Plan to RTC for repeat CL dispense appt and review of A&R training when new CLs arrive**

- **NEW CL PARAMETERS**
  - OD - Diameter: 15.0mm, SAG: 4344, BC: 7.84, Rx: -0.32D, Freeform PCs
  - OS - Diameter: 15.0mm, SAG: 4475, BC: 7.64, Rx: -0.66D, Freeform PCs

# CONTACT LENSES – DISPENSE APPT #2 2/21/23

- **Patient reports she is happy with vision and comfort in both contact lenses**

- **First Latitude Lens Fit OD**
  - Between 380um central clearance
  - Thin but acceptable limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: -0.25, 20/20

- **First Latitude Lens Fit OS**
  - 333um central clearance
  - Thin but acceptable limbal clearance
  - 1+ nasal and temporal edge lift, no blanching
  - ORx: 20/40 near; -1.00, 20/25-- distance

- **Good fit and vision with contact lenses**
- **Continue with PFAT gel to apply CLs**
- **Dispensed one bottle of diluted 15% solution of 0.5% proparacaine in AT bottle to aid patient in application**
  - **Patient instructed to only use a max of twice in one day, discontinue once she became more comfortable with application**
- **Plan to RTC in 2-4 weeks for CL F/U**

# CONTACT LENSES – DISPENSE APPT #2 2/21/23

# CASE 3 – TRAUMATIC CORNEAL LACERATION

# CASE HISTORY

- **CHIEF COMPLAINT**
  - Blurred vision at all distances and light sensitivity in his right eye
  - Referred by cornea specialist for scleral contact lens evaluation
- **DEMOGRAPHICS**
  - 56yo HM
- **OCULAR HISTORY**
  - Full thickness corneal laceration in right eye (10/2022)
  - Laceration repair surgery in 10/2022
- **OCULAR MEDICATIONS**
  - ATs qid OD
- **MEDICAL HISTORY**
  - Hypertension
- **MEDICATIONS**
  - None
- **ALLERGIES**
  - None
- **COMANAGEMENT**
  - Currently being followed by a cornea specialist

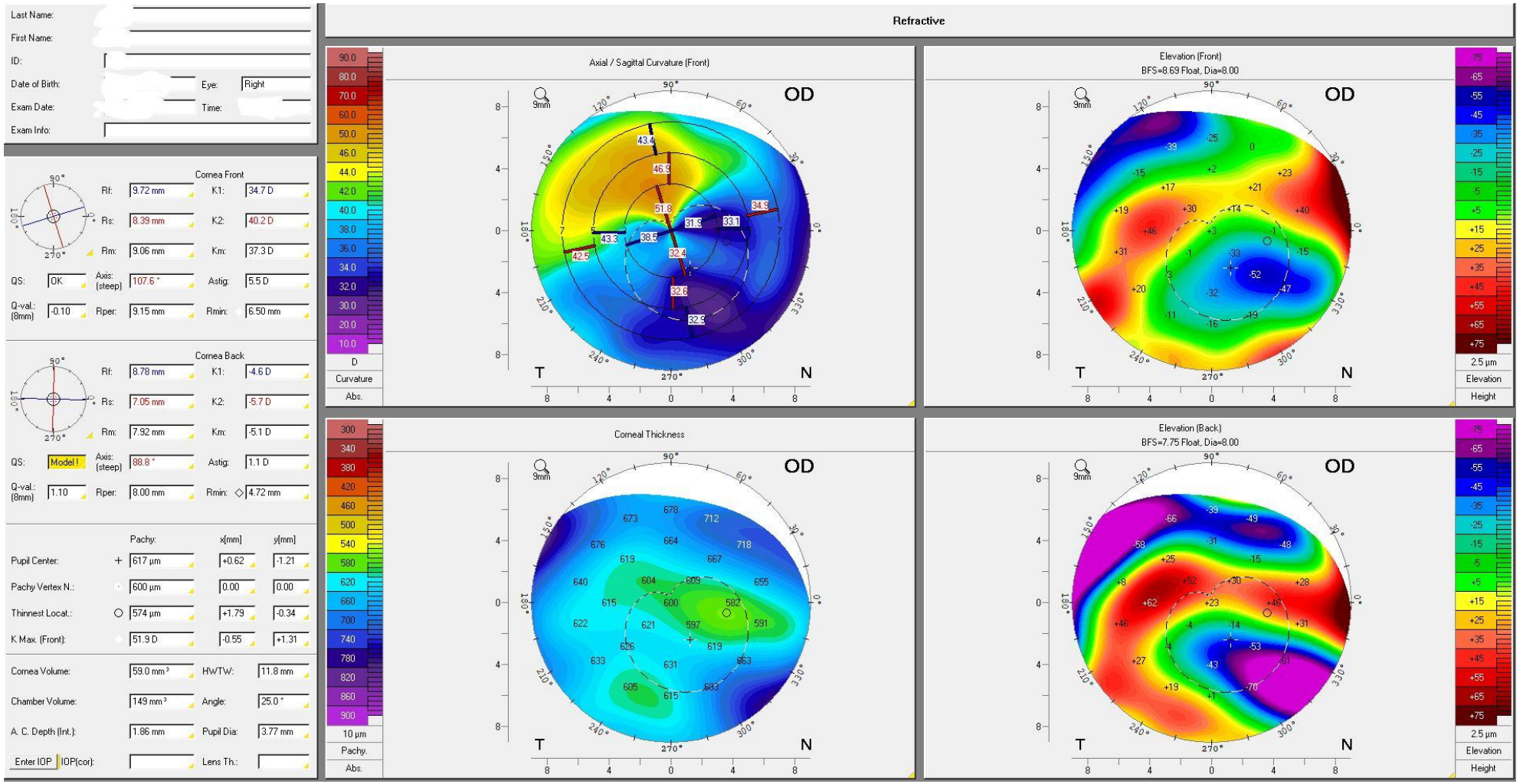
# EXAM FINDINGS

- **ACUITIES**
  - OD unaided: 20/500, PHNI
  - OS unaided: 20/30-
- **KERATOMETRY**
  - OD Kmax: 51.9D
  - OS Kmax: 43.6D
- **PACHYMETRY**
  - OD minimum: 443um
  - OS minimum: 407um
- **MANIFEST SPEC RX**
  - OD: +0.75, 20/500 (Balance lens)
  - OS: +0.75, 20/20

# EXAM FINDINGS

- **SLIT LAMP**
  - OU: 2+ MGD
  - OD: full thickness linear corneal laceration from limbus @ 4 o'clock extending superior temporally through visual axis (12 intact nylon sutures)
  - OD: Irregular pupil with nasal iris defect
  - OD: 2+ bulbar injection, large nasal pinguecula
- **IOP/FUNDUS EXAM**
  - OU: WNL

# PENTACAM SCANS

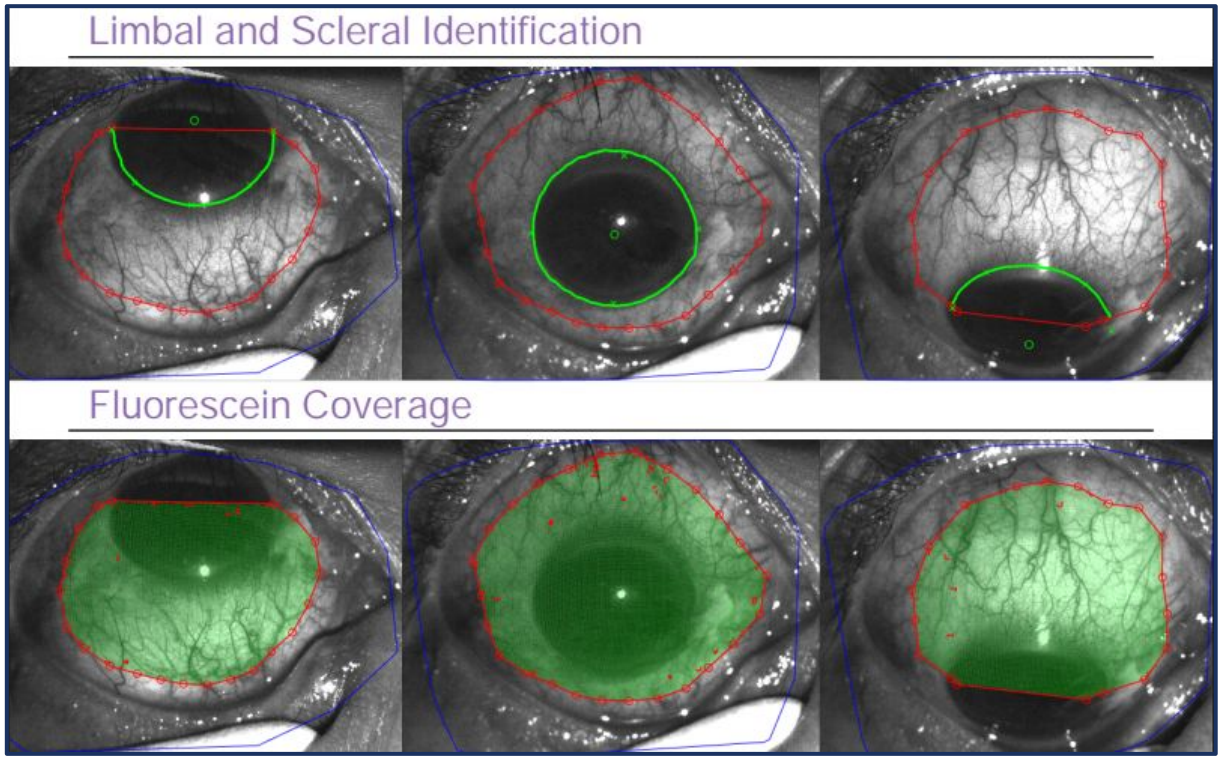


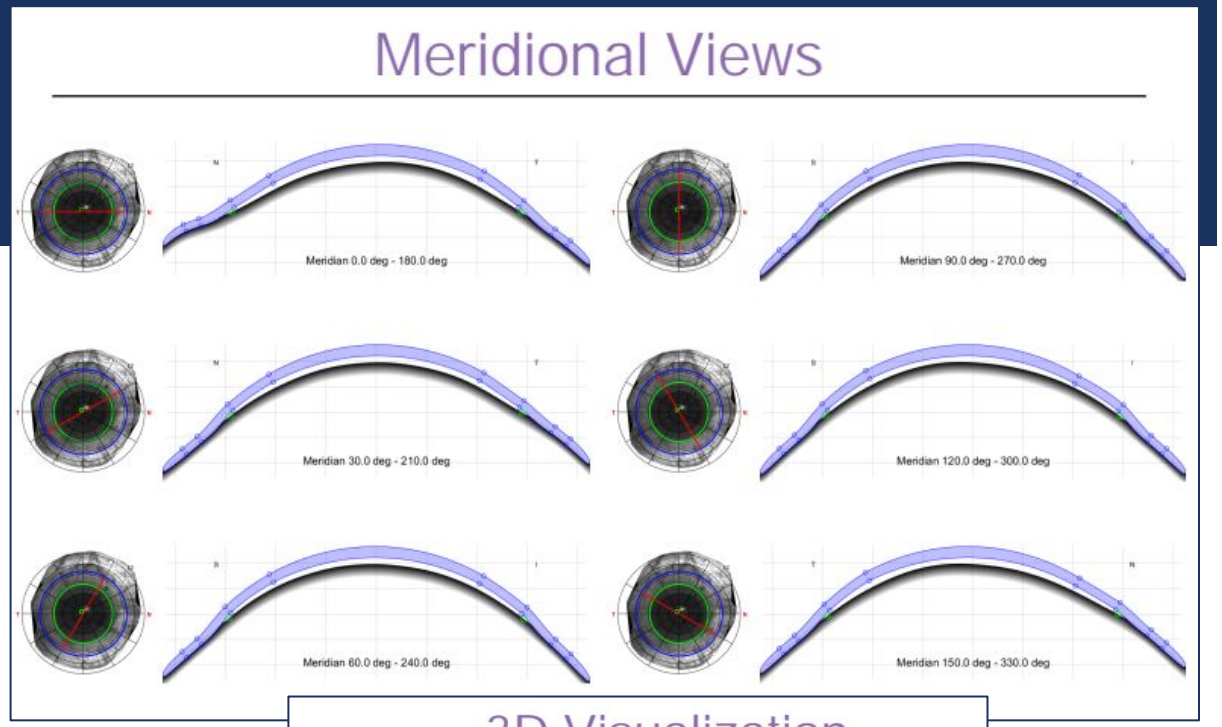
# CONTACT LENSES

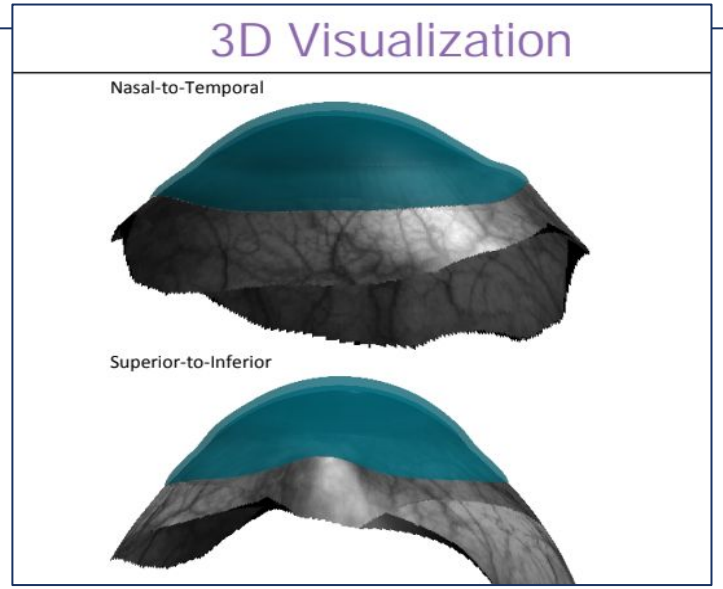
- **1st TRIAL LENS**
  - OD - Europa
    - BC: 8.04D
    - Diameter: 16.5mm
    - PWR: Plano
    - SAG: 4201

- **FIT**
  - **OD:** Very temporally decentered due to nasal pinguecula and elevation, adequate central clearance, stable enough to obtain ORx
    - **ORx:** –1.50, 20/50
- **PLAN**
  - Order lenses based off SMap3D scan OD
  - Return for A&R training when lenses arrive

# CONTACT LENS – OD




Diameter: 17.00mm, SAG: 4968, BC: 8.74, Rx: +1.89D, Freeform PCs

# CONTACT LENS – OD

Diameter: 16.50mm, SAG: 4991, BC: 8.99, Rx: -2.41D, Freeform PCs

# CONTACT LENSES – DISPENSE APPT 12/21/22

- **First Latitude Lens Fit OD**
  - 350um central clearance after application
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: +1.00, 20/40

- **A&R training Completed successfully**
- **CL dispensed**
- **No lens changes made at this time**
- **Plan to RTC in 2 weeks for CL F/U**

# CONTACT LENSES – FOLLOW UP APPT 1/20/23

- **Patient reports he is happy with comfort and vision in contact lens but is still very photophobic**

- **First Latitude Lens Fit OD (**1 hour of wear time**)**
  - 200um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/30

- **Good fit and vision with contact lenses**
- **Fit tinted spectacle lens to wear full time over CL**
- **Plan to RTC in 1 months for CL F/U**

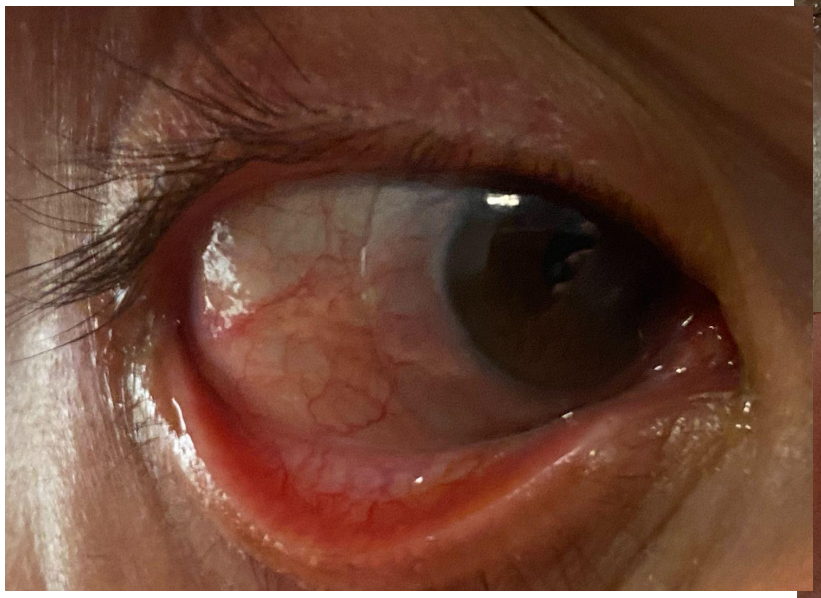
# CONTACT LENSES – FOLLOW UP APPT 3/13/23

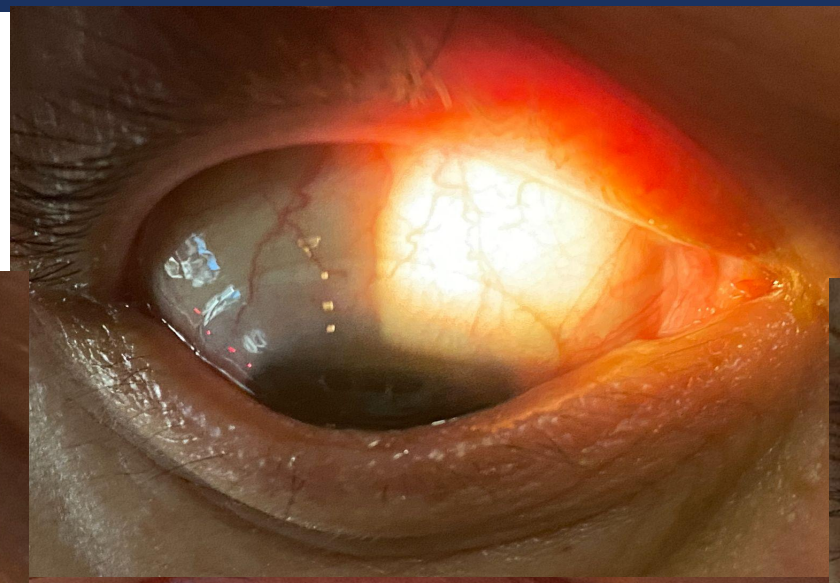
- **Patient reports he is happy with comfort and vision in contact lens**

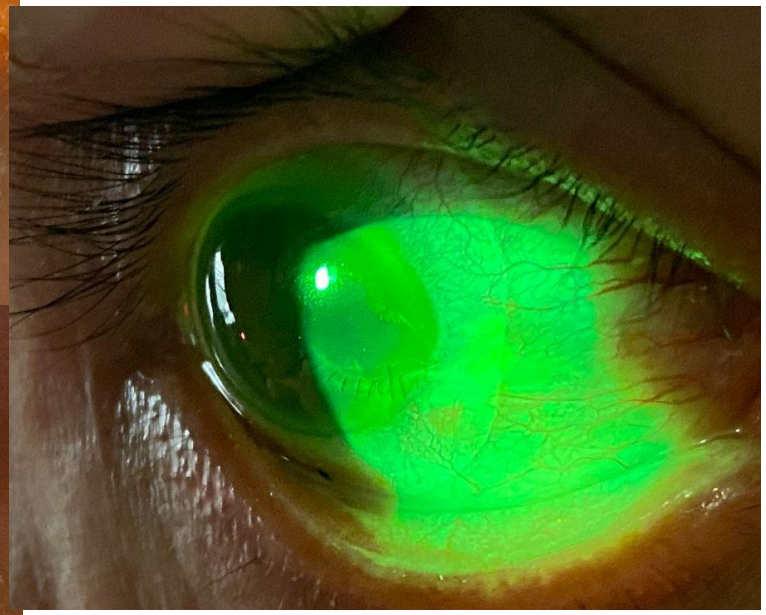
- **First Latitude Lens Fit OD** (3 hour of wear time)
  - 175um central clearance
  - Good limbal clearance
  - Smooth landing zone, no edge lift or blanching
  - ORx: Plano, 20/30

- **Good fit and vision with contact lens**
- **Continue tinted spectacle lens to wear full time over CL**
- **Plan to RTC in 6 months for CL F/U**

# CONTACT LENSES – FOLLOW UP APPT 3/13/23



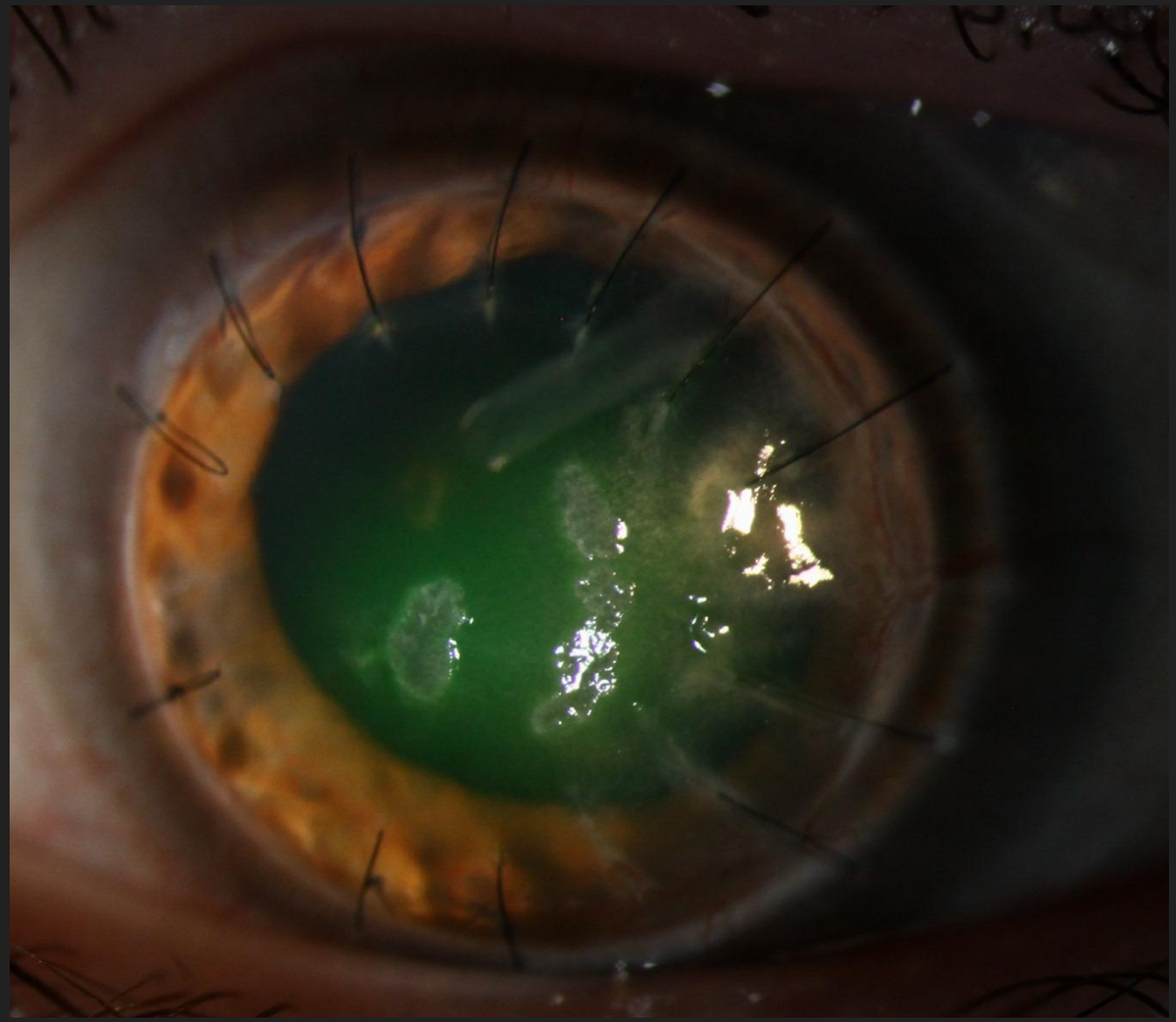




# TAKE IT AWAY MARCUS

# Case 1

# PK’s, Tubes, and Acanthamoeba/Mycotic Superinfection



# Case History

- Background: 20 year-old Caucasian female referred from cornea service for scleral lens evaluation OU
- Chief Complaint: Decreased visual acuity and dryness OU
- Past Medical History: (+) Type 2 diabetes mellitus
- Ocular History: (+) acanthamoeba OU, (+) central fungal ulcer OU, (+) penetrating keratoplasty OU, (+) amblyopia OS, (+) accommodative esotropia OS, (+) secondary angle closure glaucoma, (+) ahmed tube valve OS, (+) baerveldt tube OS

# Case History

- Systemic Medications: doxycycline 100mg bid, others non-contributory
- Ocular Medications: artificial tears prn, brimonidine, dorzolamide-timolol, moxifloxacin, muro, prednisolone acetate, autologous serum drops
- Prior Failed therapies: artificial tears, hydrating ung, bandage soft lens, antibiotic drops

# Exam Findings

Visual Acuity:

OD: 20/70sc→20/40 PH

OS: 20/60sc → 20/50 PH

EOMs: Normal OU

Pupils: Irregular and nonreactive OD, normal OS

IOP: 10mmHg OD, 19mmHg OS

Fundus: 0.1 C/D OU

CVF: Normal OU

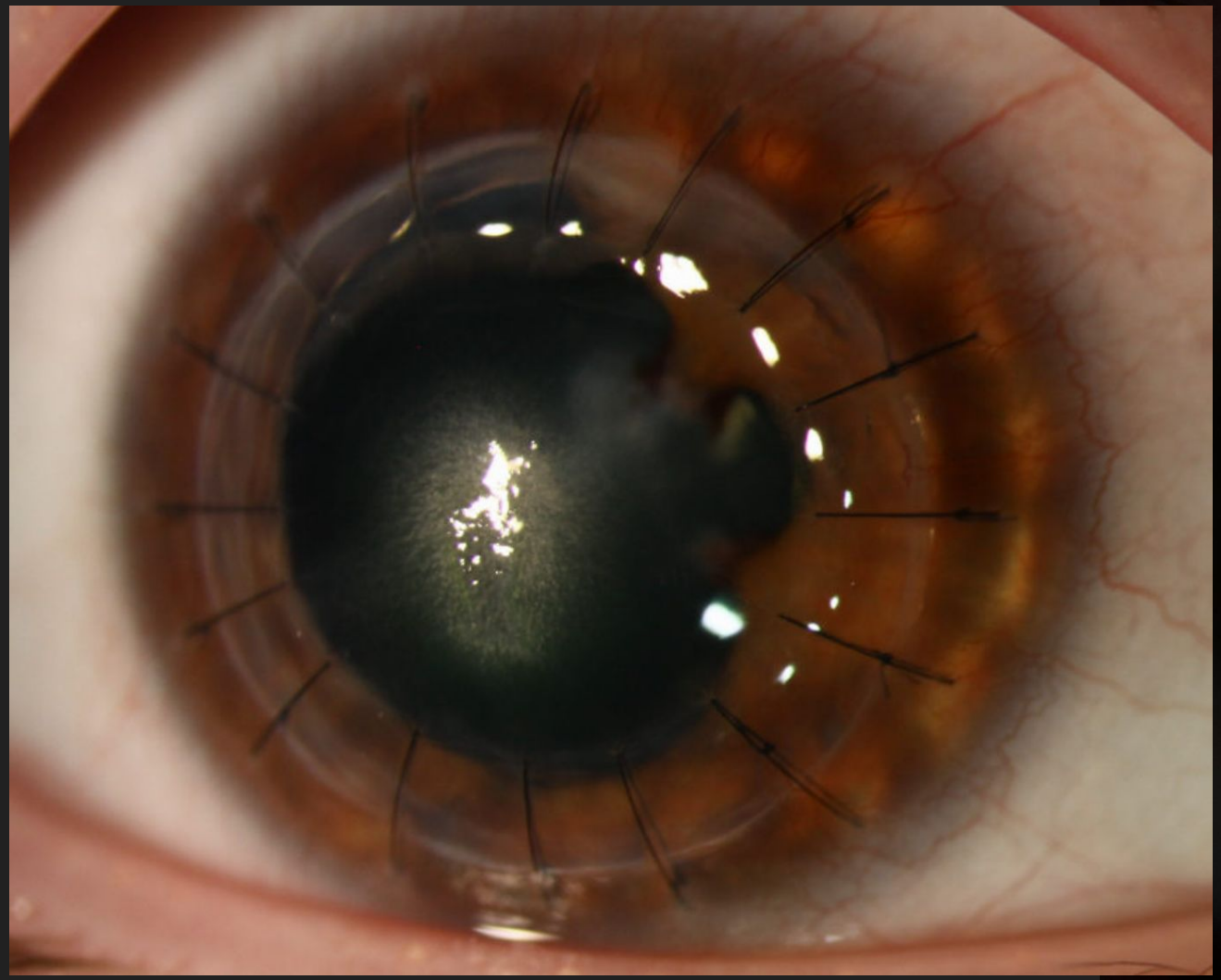
# Slit lamp examination OD

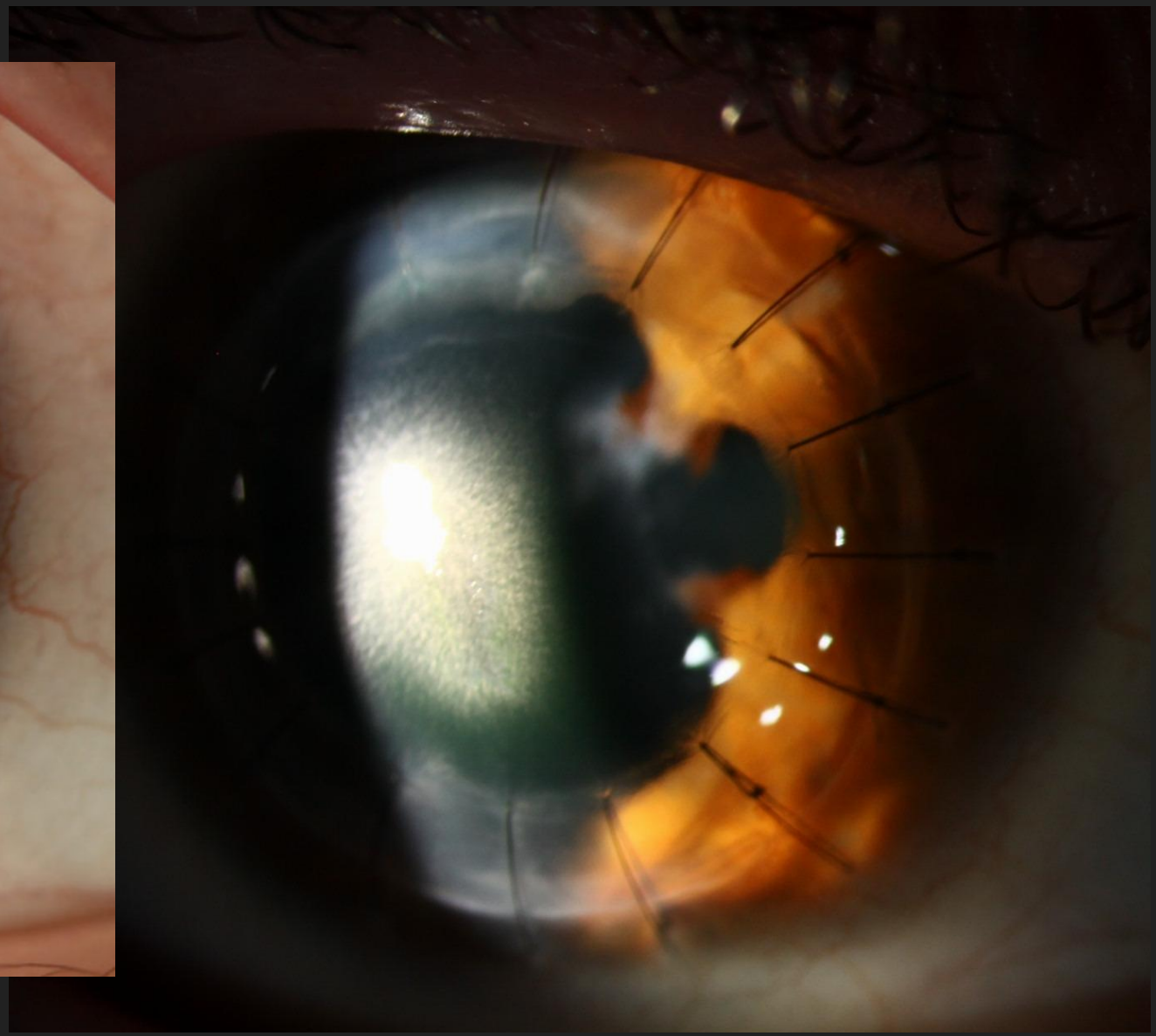
| | |
|---|---|
| **Eyelids/Adnexa** | Normal |
| **Conjunctiva** | Clear and quiet |
| **Cornea** | **PKP with peripheral NV** approaching GHJ at 3 and 5 o'clock, **central superficial haze, epi intact** |
| **Anterior Chamber** | Deep and quiet |
| **Iris** | Posterior synechiae at 2 and 4 o'clock |
| **Lens** | Anterior haze near posterior synechiae |
| **Vitreous** | Normal |

# Slit lamp examination OS

| | |
|---|---|
| **Eyelids/Adnexa** | Normal |
| **Conjunctiva** | **Superior and inferior tube shunts** without erosion |
| **Cornea** | **PKP with clumps of irregular epithelium, multifocal dendritiform pattern (stable).** No focal defect or infiltrate. Vessels at 0300 and 0500 marching barely inside graft |
| **Anterior Chamber** | Deep and quiet, tubes in good position |
| **Iris** | Normal |
| **Lens** | Normal |
| **Vitreous** | Tubes in sulcus |

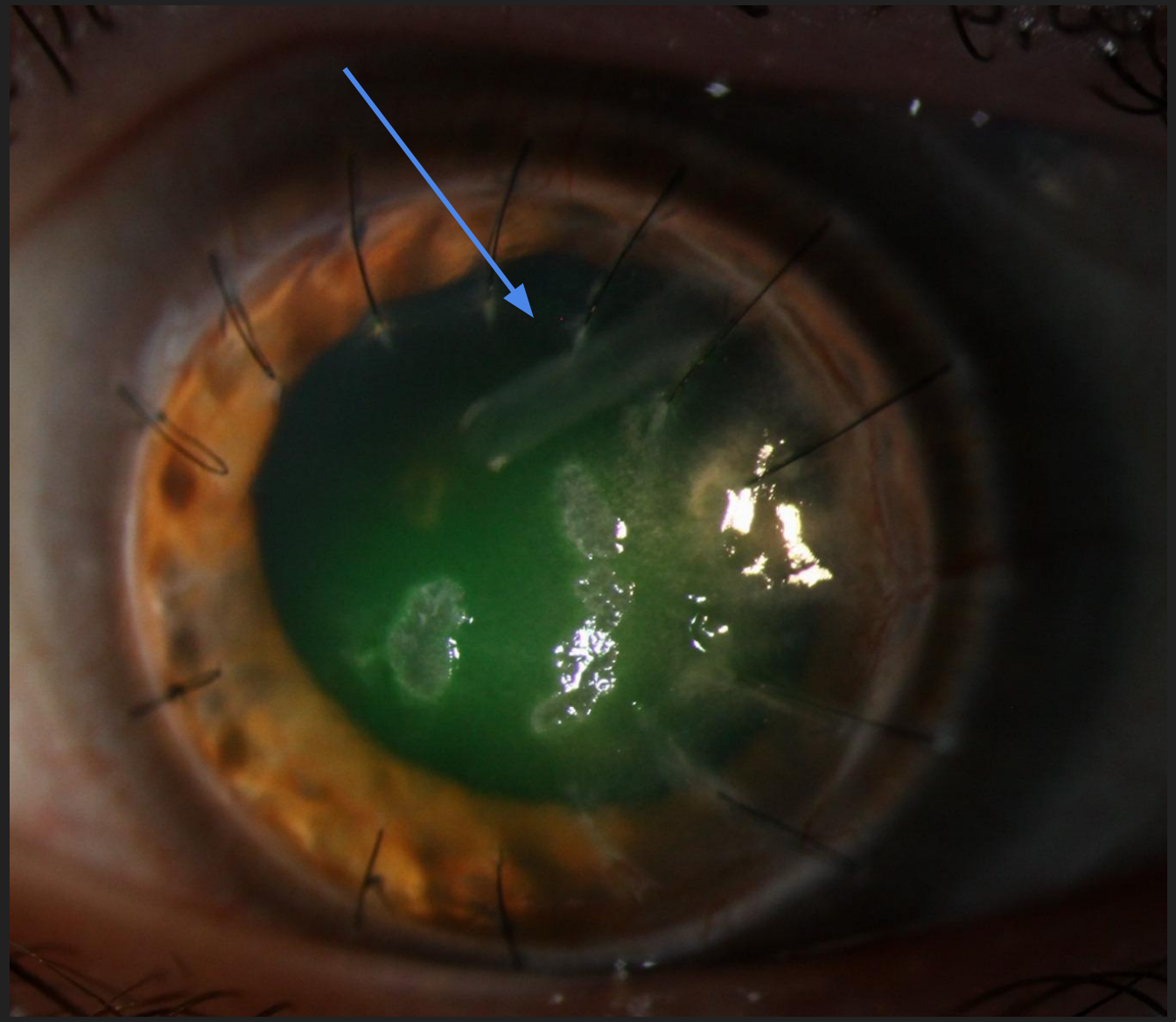
# Slit Lamp Photos OD





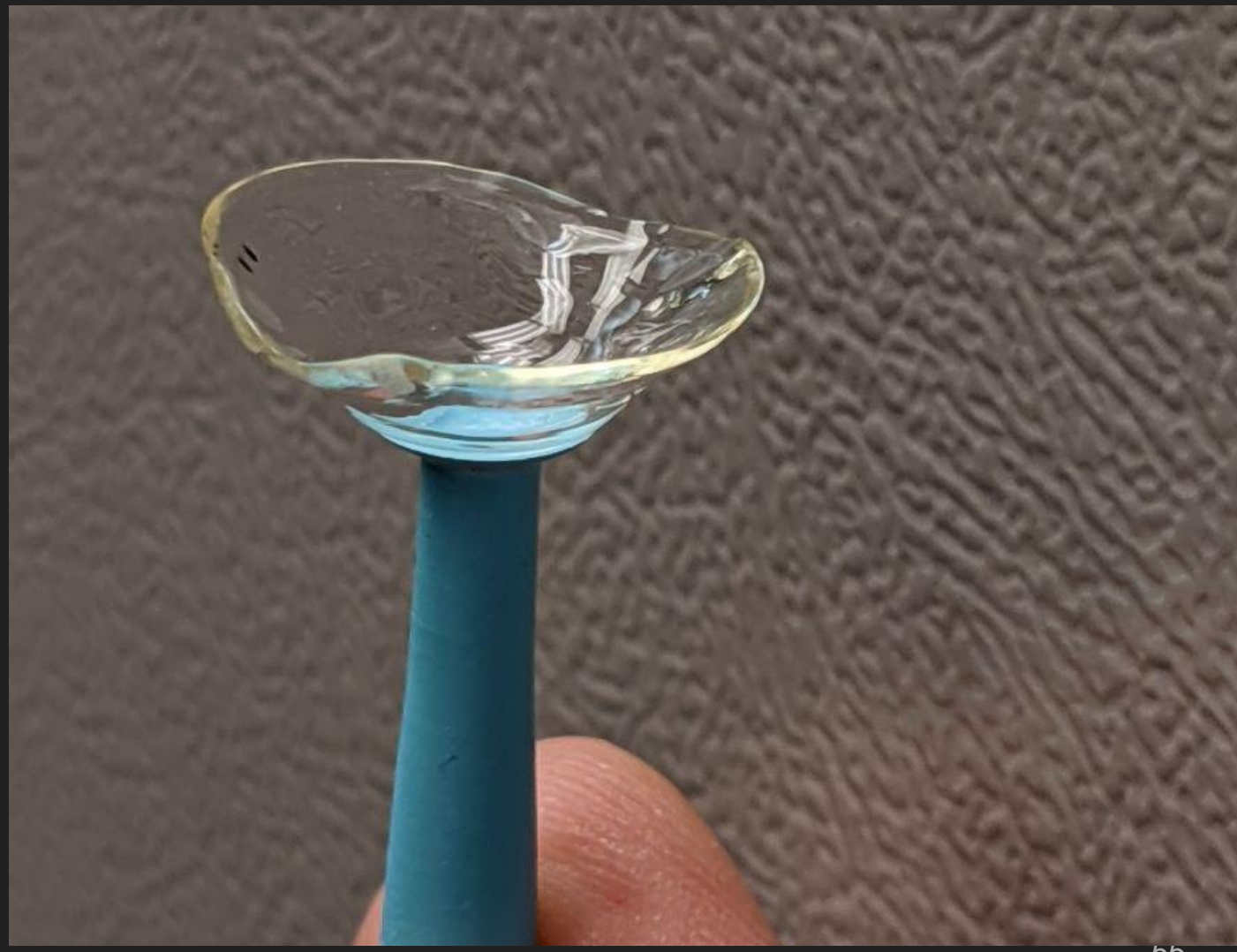
# Slit Lamp Photos OS

- Large diameter PKP
- **Tube** (just one in this photo)
- Strange epi after mycotic infection

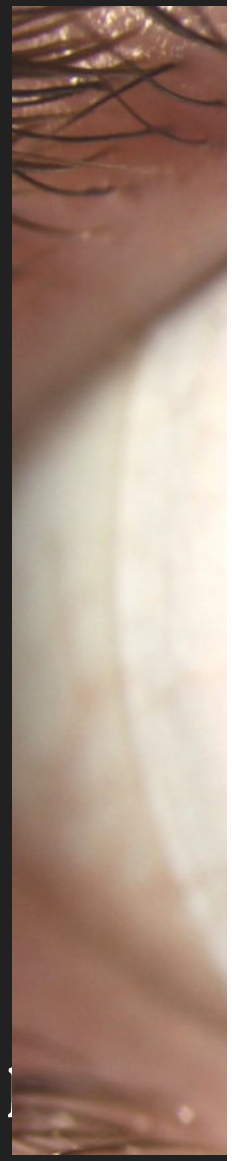


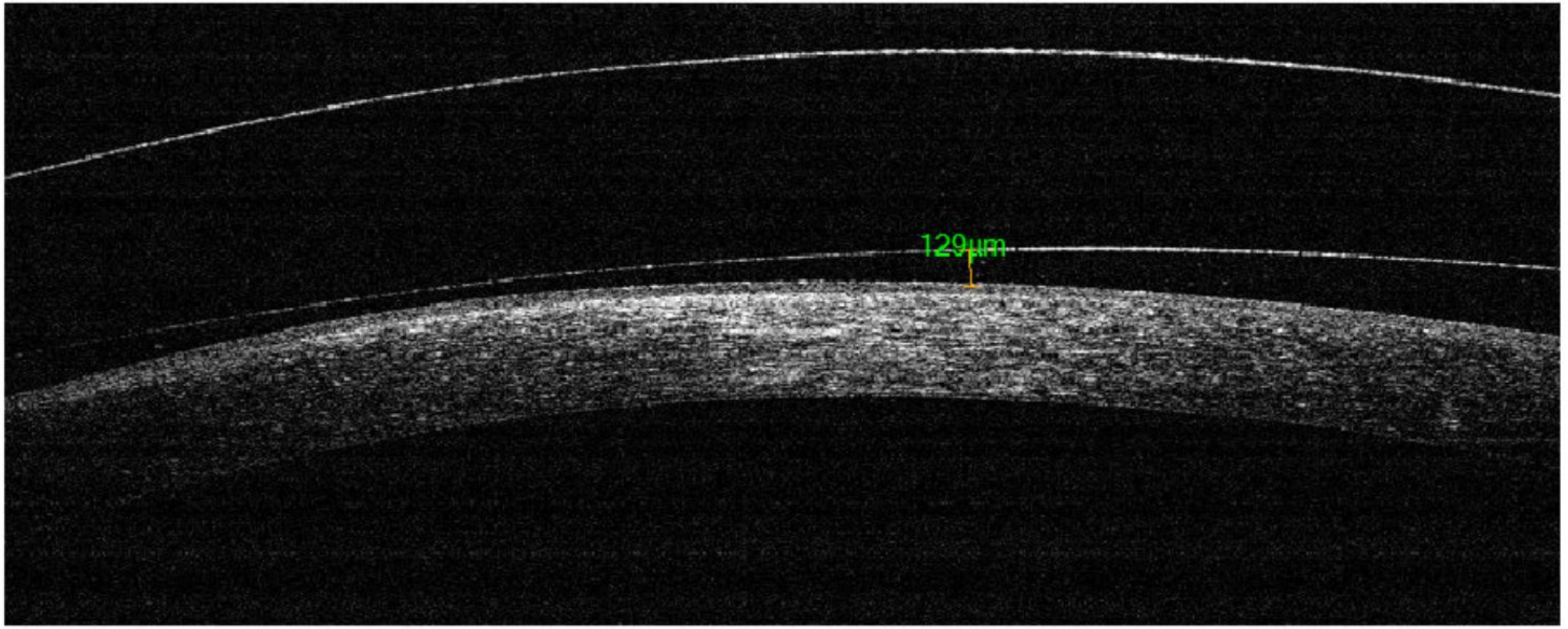
# The Scleral Fit Process

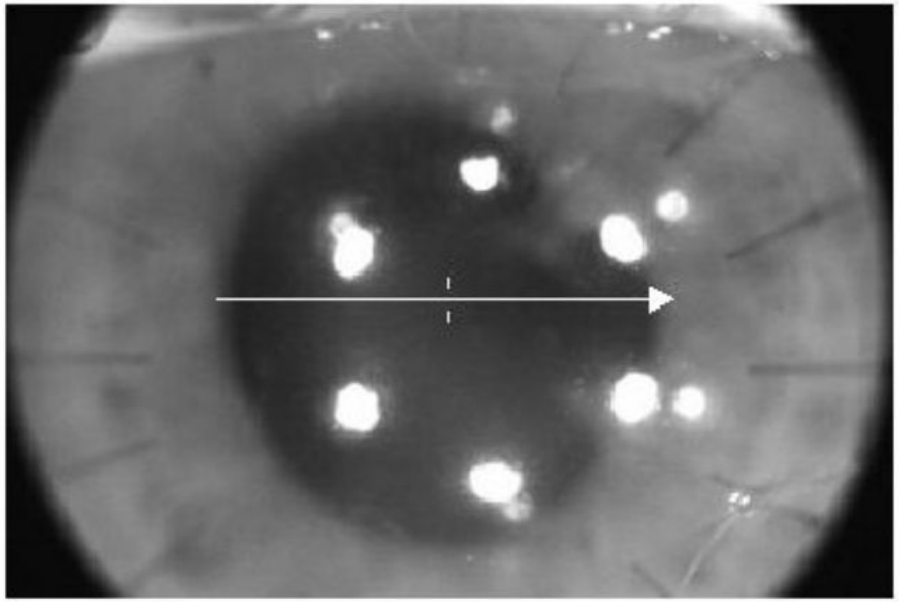
- Over-refracted with RGP’s
- Fit with an impression-based design



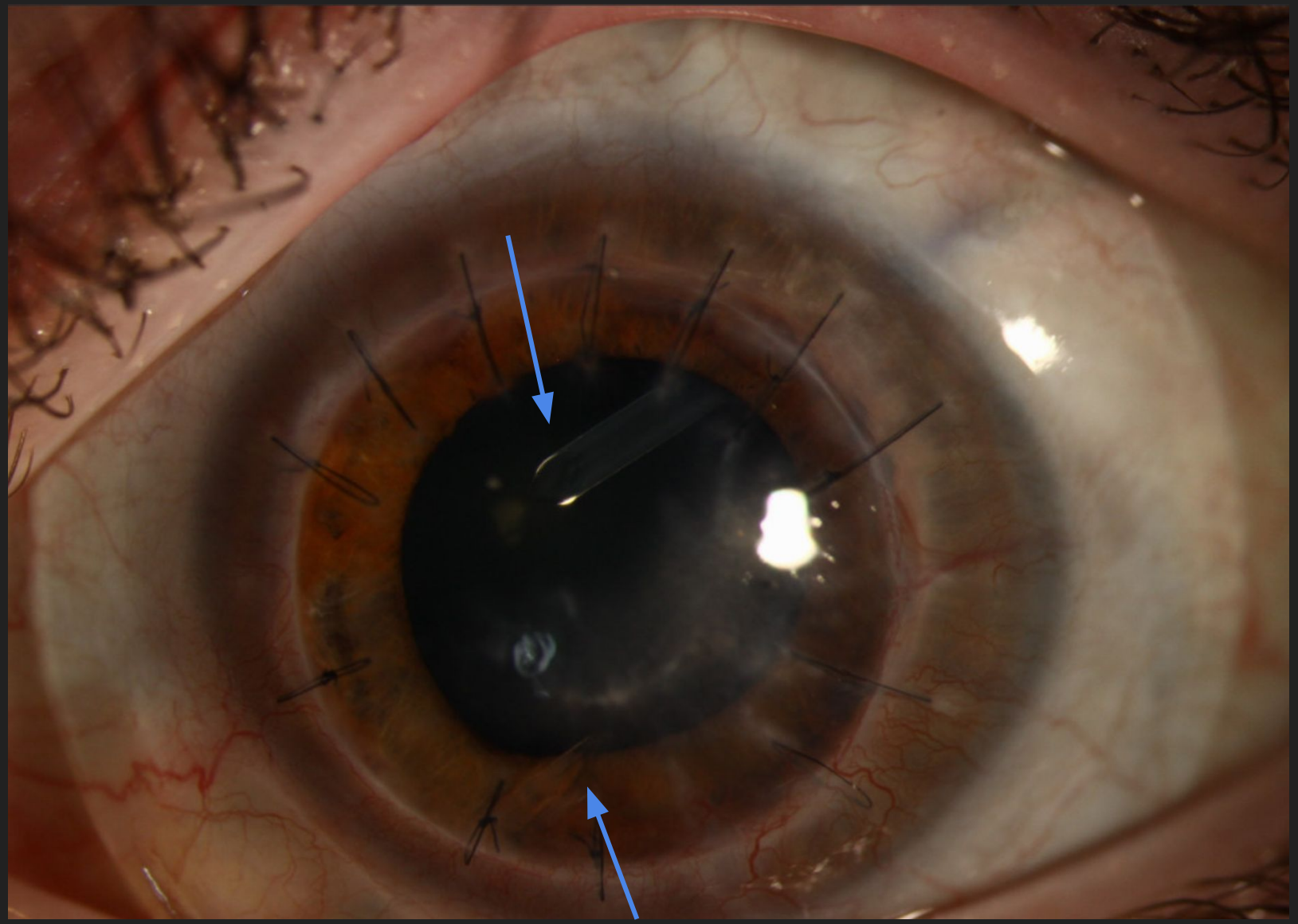
# The



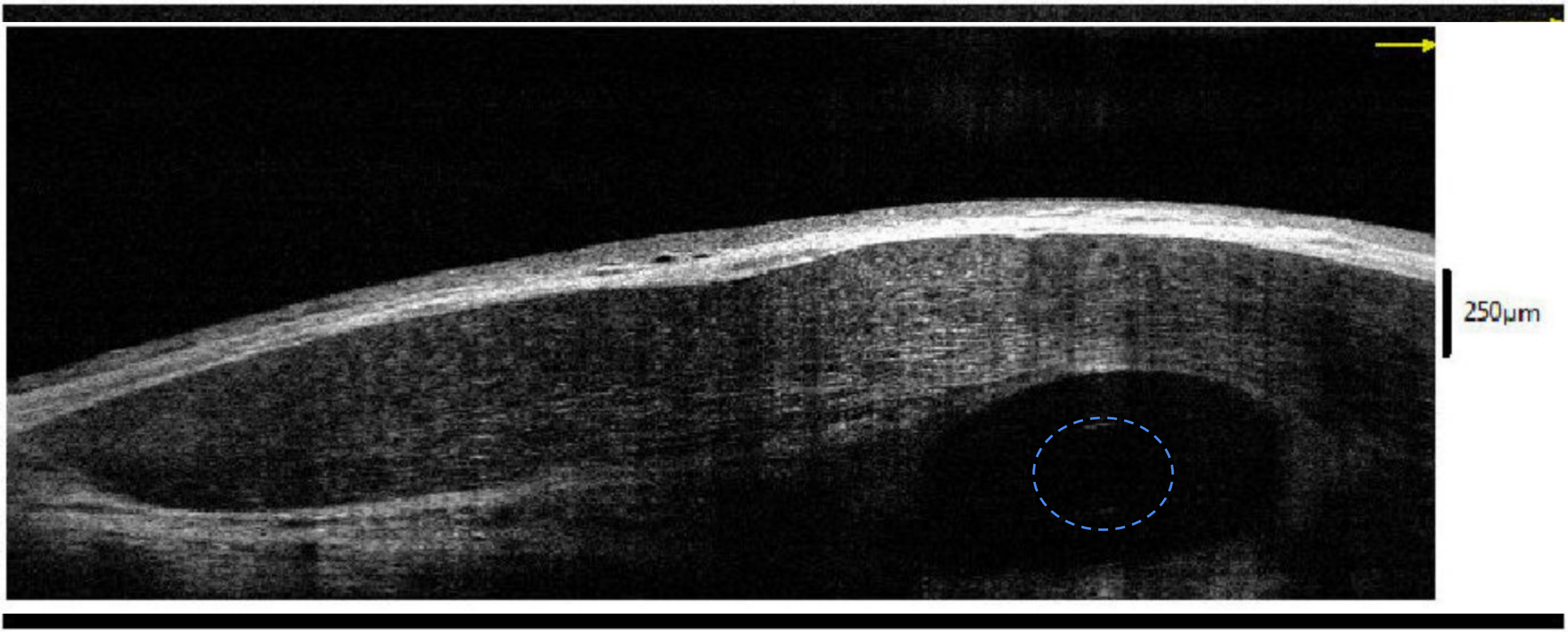


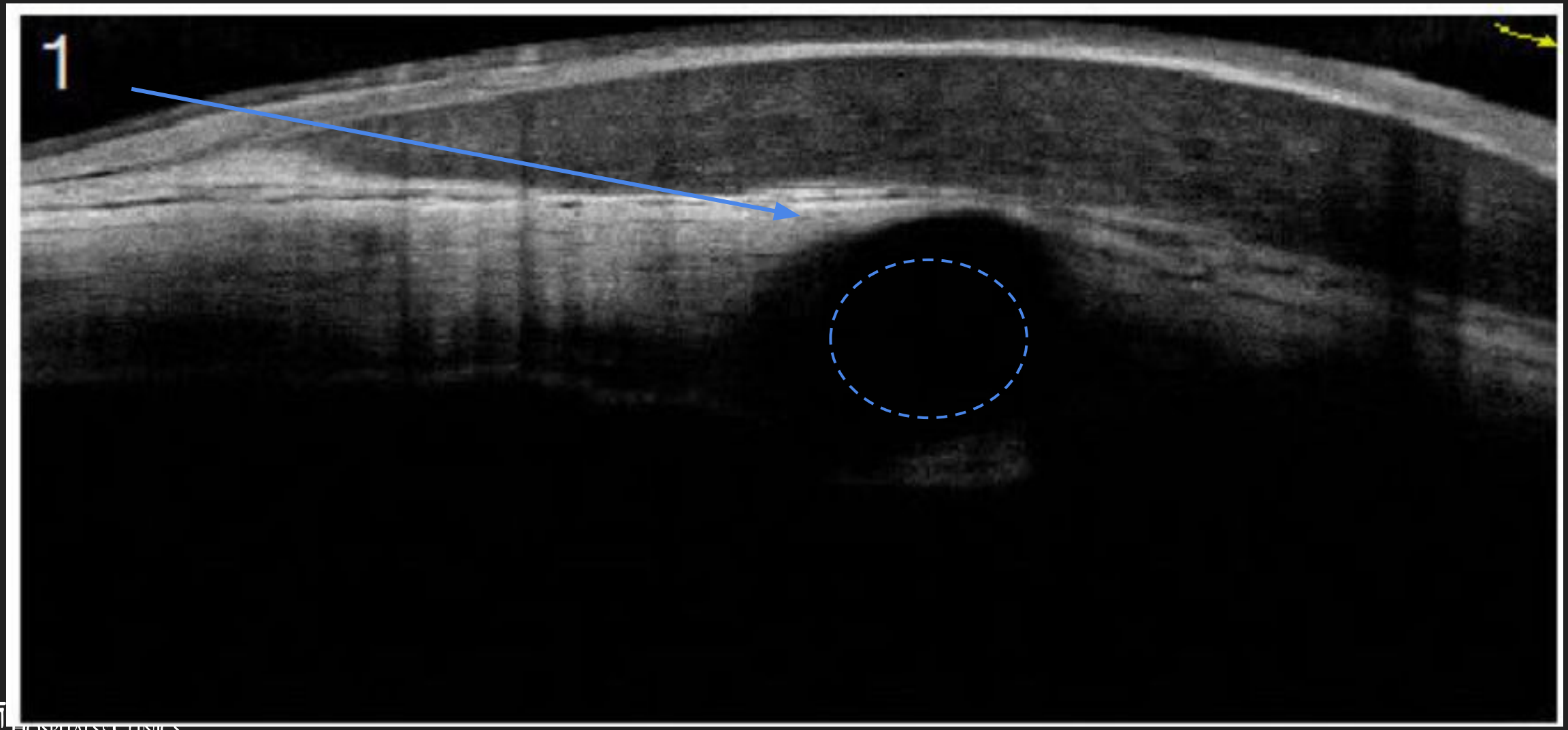
# The Scleral Fit OS



# The Scleral Fit OS



# Superior Tube Erosion OS…… 3 months later



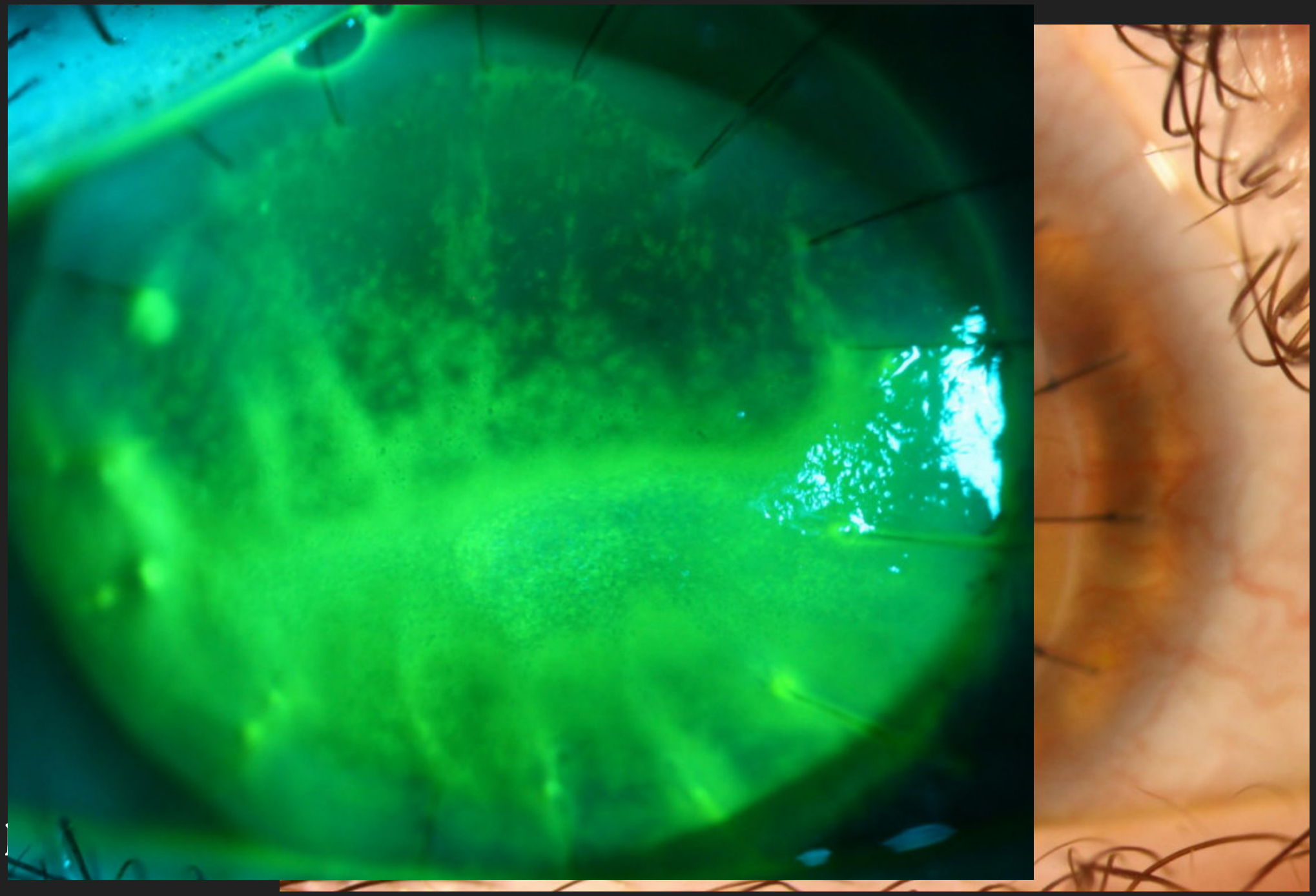
# Final Lens Parameters

| Brand | Overall Diameter | Base Curve | Power | CT | Material | Add-ons |
|---|---|---|---|---|---|---|
| EyeFit PRO (EyePrint Prosthetics) | 17.8mm | 9.954mm | +11.38 DS | 0.68mm | Optimum Infinite | Hydrapeg |
| EyePrint PRO (EyePrint Prosthetics) | 17.8mm | 7.586mm | -2.75 DS | 0.35mm | Optimum Infinite | Hydrapeg, extra vault over tubes |

**BCVA 20/25 OD, 20/30 OS!**

# Success! …right?

- Successful wear for 1.5 years
- Grafts start rejecting (after receiving COVID and flu vaccines)
- Re-PKP OD… and the graft gets microbial keratitis and new HSV coinfection
- And the left eye starts failing too
- Must stay out of sclerals… OD 20/150 OS 20/600



# Why a Scleral Lens?

- Avoid dessication of the cornea/limbus
- Improved patient comfort
- Can help with tube erosion

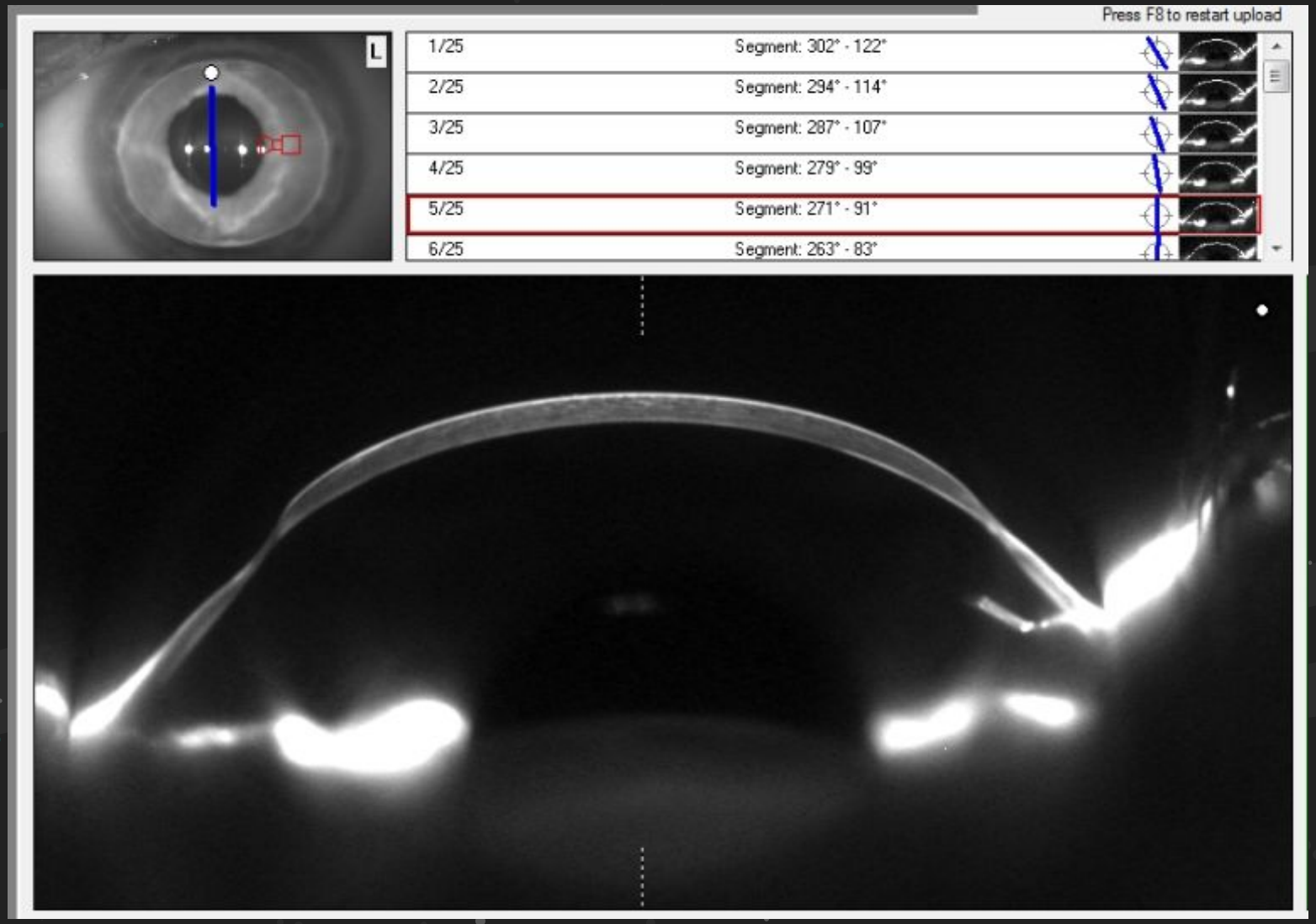
# Risk and Alternatives?

- Risks
  - Without impression-based fitting, very difficult to have proper fit over tube
  - Fragile eye; patient must be properly trained
- Alternatives?
  - RGP and…. That’s it

# Lesson Learned

- Over-vault tubes and blebs– need plenty of room to “settle-in”!
- Sclerals can re-model conjunctival tissue to prevent erosion
- Rejection/graft failure can occur independently of scleral fit!
    - Can be easy to get in an “adjusting cycle”

# Case 2: Fitting the Ectatic Graft



# Case History

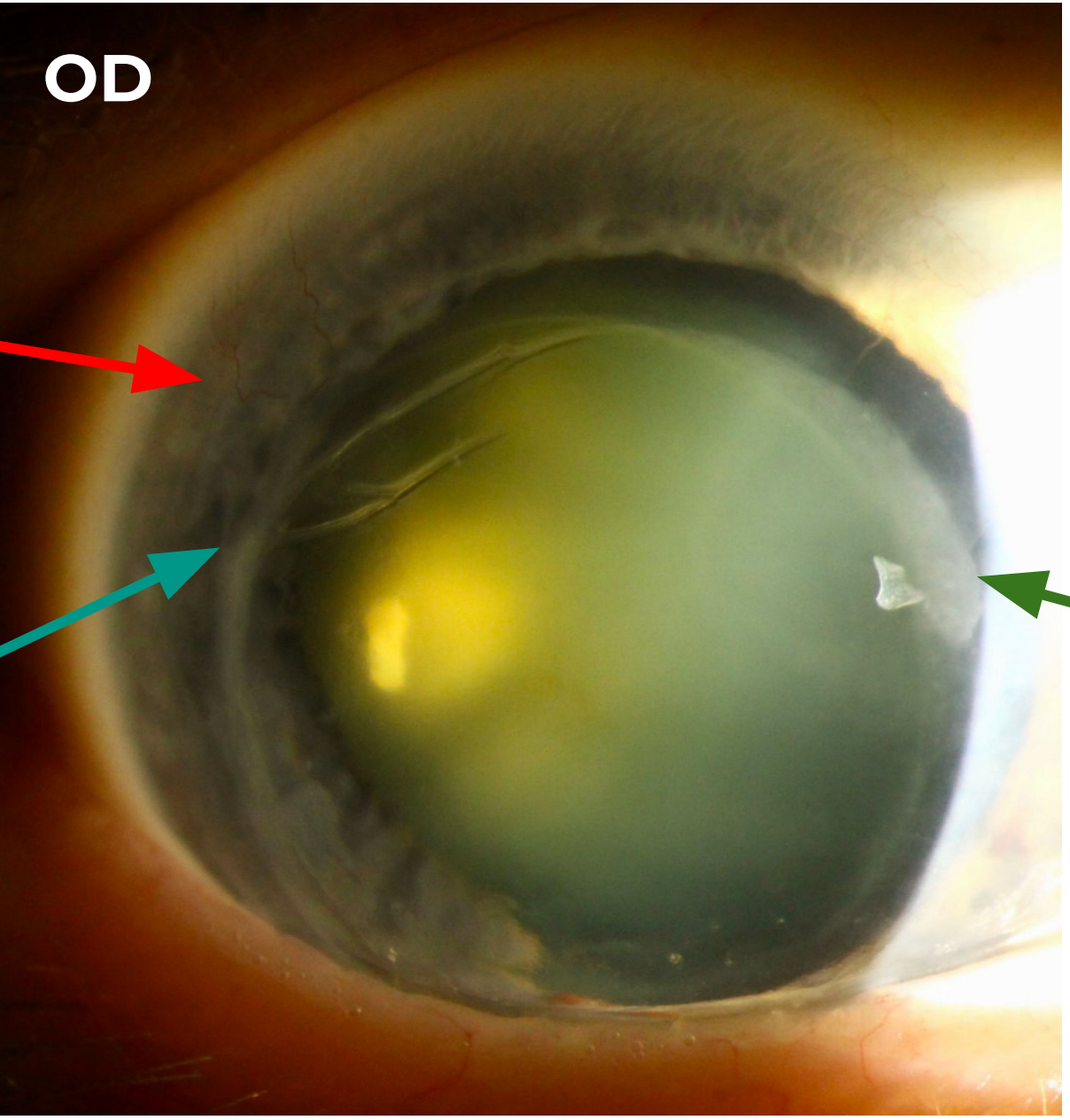
- Background: 65 year-old white male with keratoconus s/p PK OU referred from the cornea service for specialty lens evaluation
- Chief Complaint: Doesn’t want surgery
- Ocular History: (+) s/p PK “in the 1970’s” OU. (+) nuclear sclerotic cataracts, (+) keratoconus

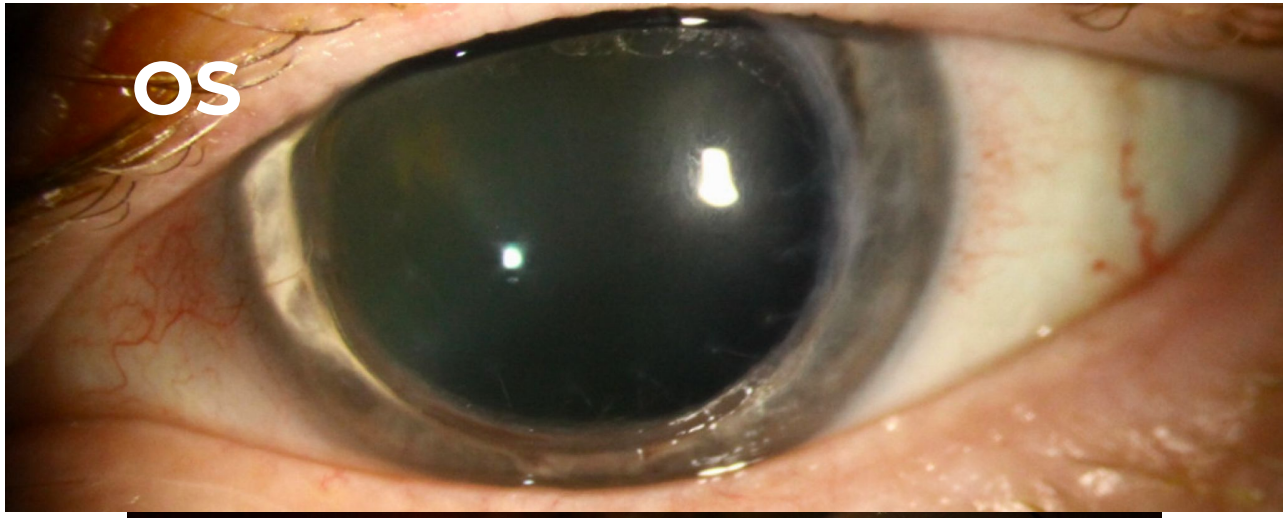
# Exam Findings

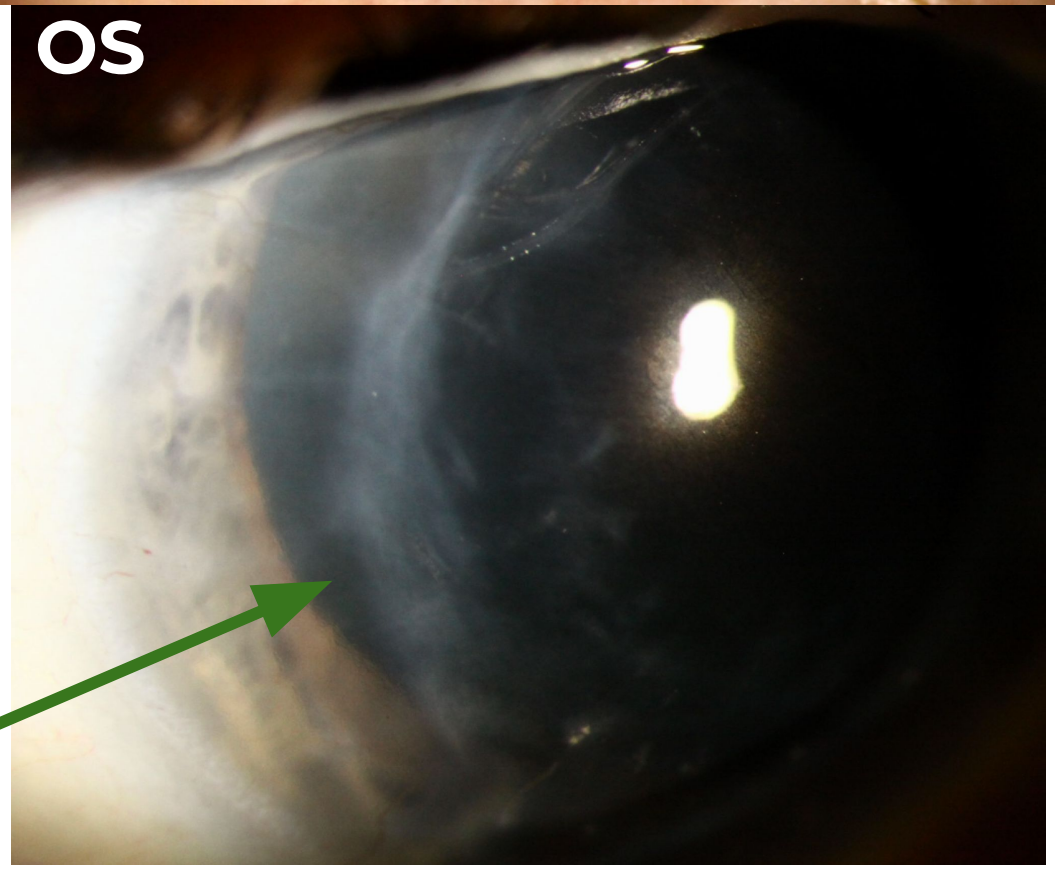
- Visual Acuity:
  - **OD: 20/60ish**
  - **OS: 20/300**
  - (with spectacles)
- **Pachymetry:**
  **660um OD, 735um OS**
- **Cell count: 140/mm2 (unreliable)**
  **OD, 844/mm2 OS**
- IOP: 15mmHg OD, 18mmHg OS
- Pupils: normal OU
- Fundus: normal OU
- CVF: Normal OU

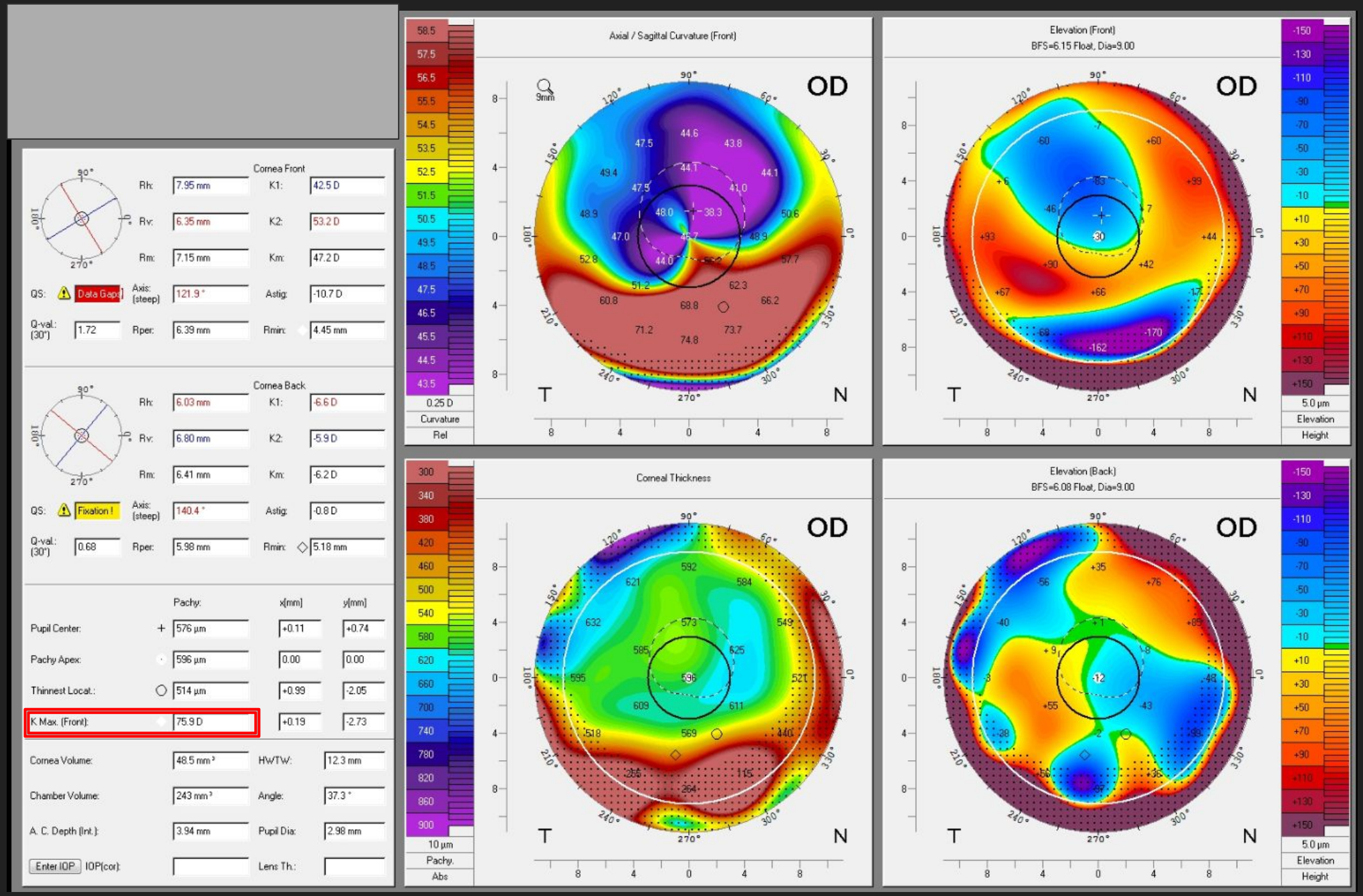
# Slit Lamp Examination OD

| | |
|---|---|
| **Eyelids/Adnexa** | Floppy eyelid, meibomian gland dysfunction, incomplete blink |
| **Conjunctiva** | Normal |
| **Cornea** | **PKP graft with ectasia, inferior cornea (host and ~1.5 mm of donor button) are thin and ectatic. Mild stromal haze and peripheral suture scars; trace punctate epithelial erosions, irregular surface OD; *Same with (+) keratoglobus OS*** |
| **Anterior Chamber** | Deep & Quiet |
| **Iris** | Normal |
| **Lens** | **Grade 3+ Nuclear sclerotic cataracts OU** |
| **Vitreous** | Normal |


OD


OS


OS

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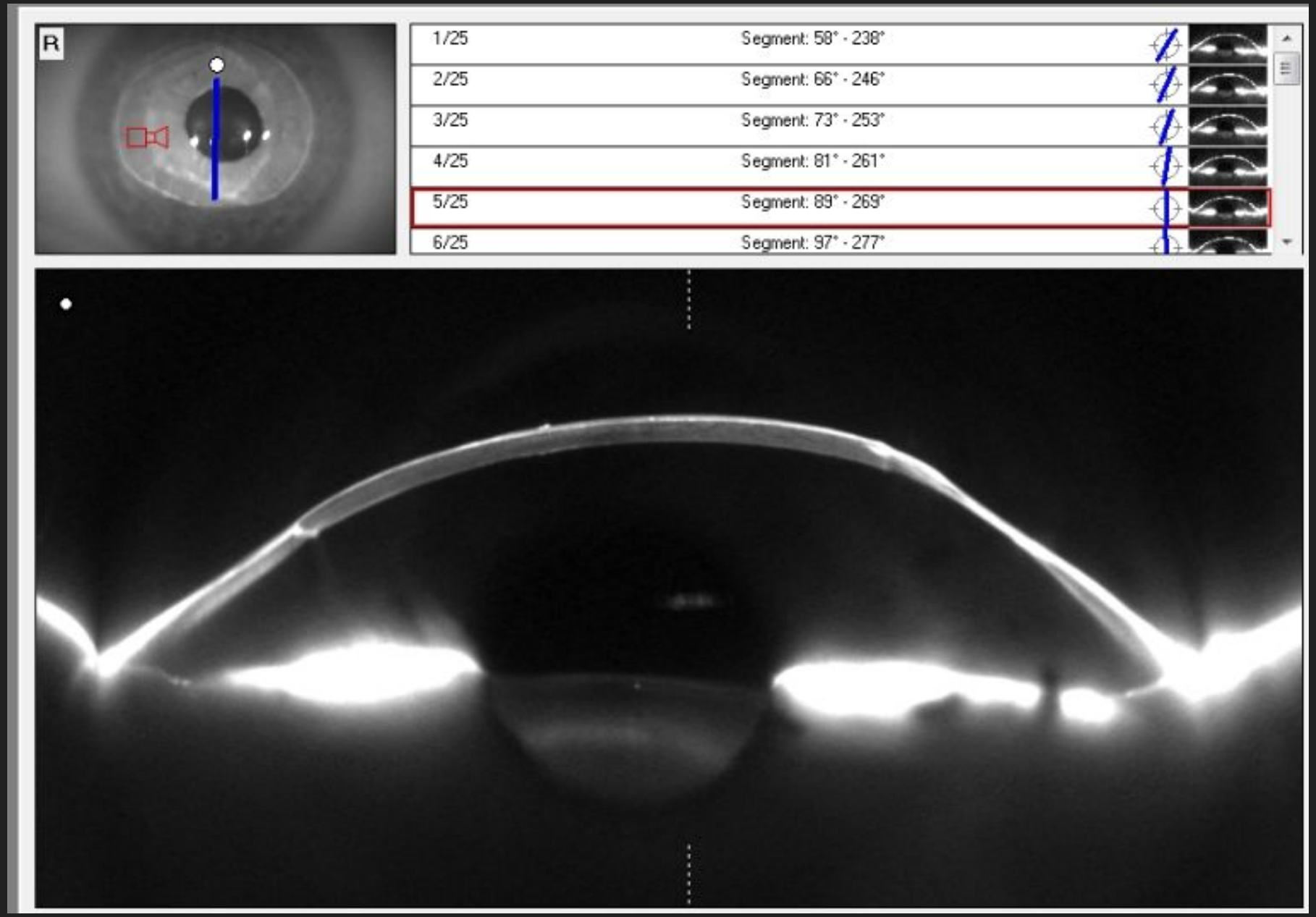
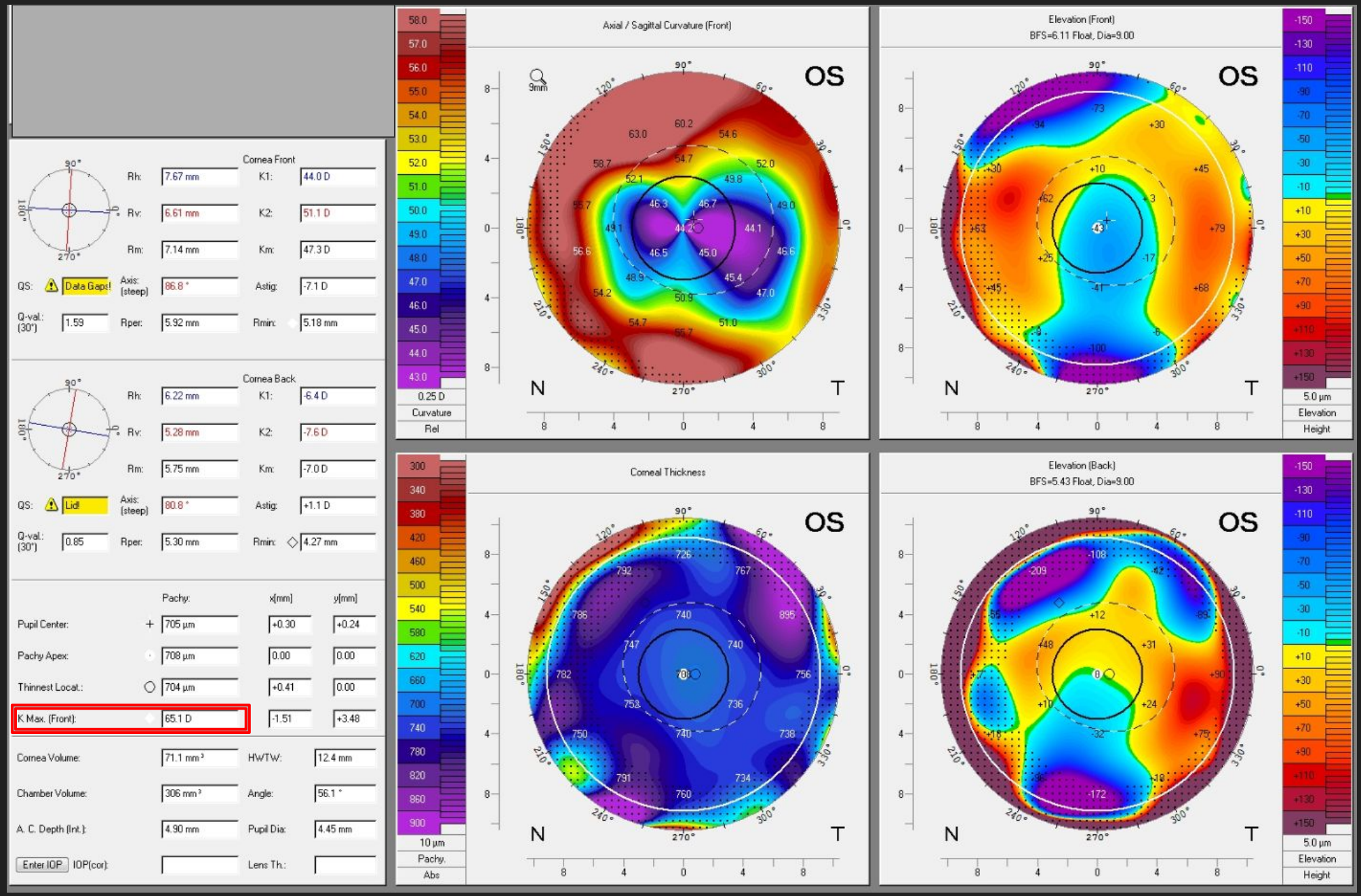
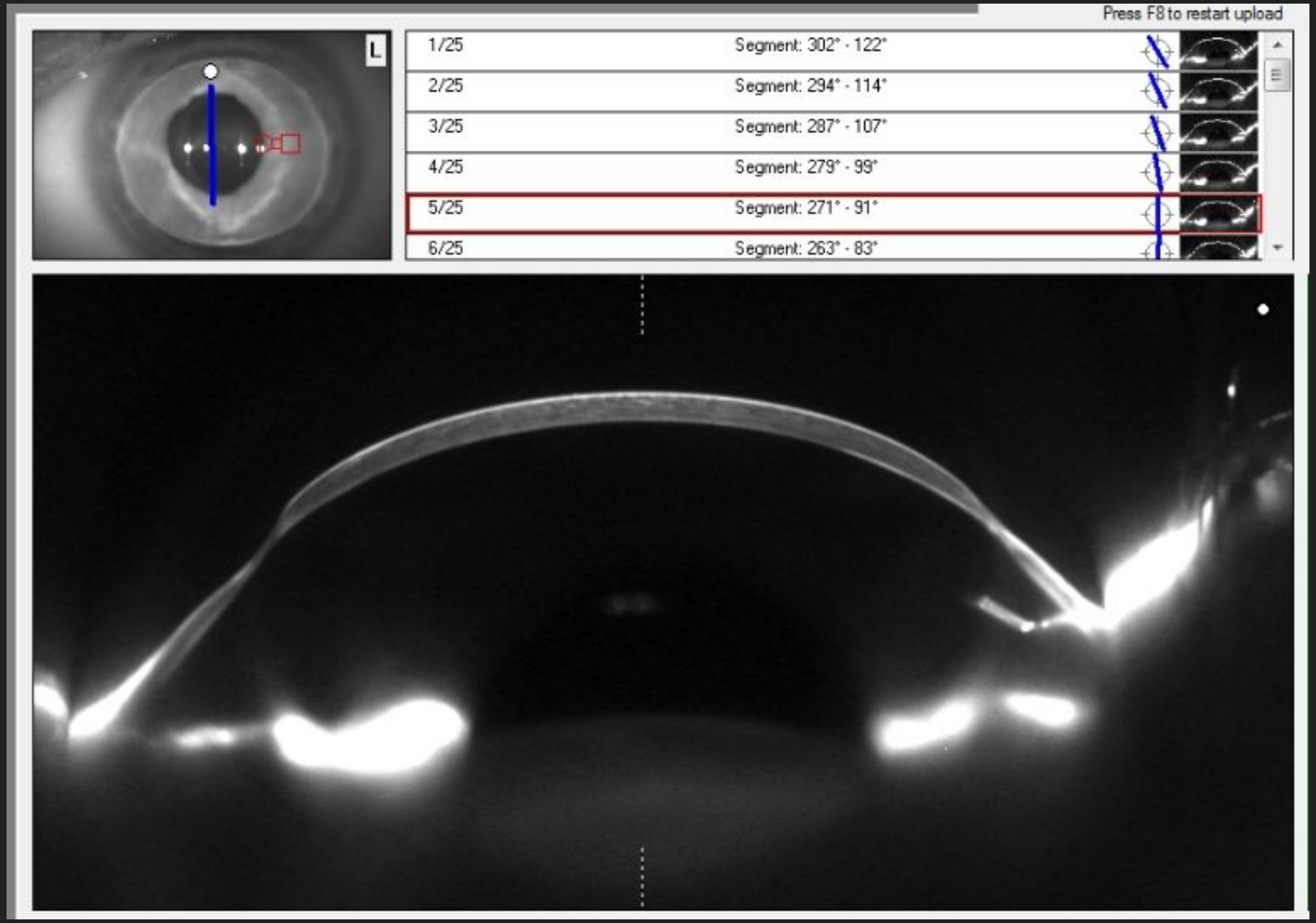

R
1/25 Segment: 58° - 238°
2/25 Segment: 66° - 246°
3/25 Segment: 73° - 253°
4/25 Segment: 81° - 261°
5/25 Segment: 89° - 269°
6/25 Segment: 97° - 277°


Cornea Front
Rh: 7.67 mm
K1: 44.0 D
Rv: 6.61 mm
K2: 51.1 D
Rm: 7.14 mm
Km: 47.3 D
QS: Data Gaps!
Axis: (steep) 86.8 °
Astig: -7.1 D
Q-val.: (30°) 1.59
Rper: 5.92 mm
Rmin: 5.18 mm
Cornea Back
Rh: 6.22 mm
K1: -6.4 D
Rv: 5.28 mm
K2: -7.6 D
Rm: 5.75 mm
Km: -7.0 D
QS: Lid!
Axis: (steep) 80.8 °
Astig: +1.1 D
Q-val.: (30°) 0.85
Rper: 5.30 mm
Rmin: 4.27 mm
Pachy:
x[mm]
y[mm]
Pupil Center: 705 µm +0.30 +0.24
Pachy Apex: 708 µm 0.00 0.00
Thinnest Locat.: 704 µm +0.41 0.00
K Max. (Front): 65.1 D -1.51 +3.48
Cornea Volume: 71.1 mm³
HWTW: 12.4 mm
Chamber Volume: 306 mm³
Angle: 56.1 °
A. C. Depth (Int.): 4.90 mm
Pupil Dia: 4.45 mm
Enter IOP
IOP(cor):
Lens Th.:
Axial / Sagittal Curvature (Front)
OS
N
T
0.25 D
Curvature
Rel
Elevation (Front)
BFS=6.11 Float, Dia=9.00
OS
N
T
5.0 µm
Elevation
Height
Corneal Thickness
OS
N
T
10 µm
Pachy.
Abs
Elevation (Back)
BFS=5.43 Float, Dia=9.00
OS
N
T
5.0 µm
Elevation
Height

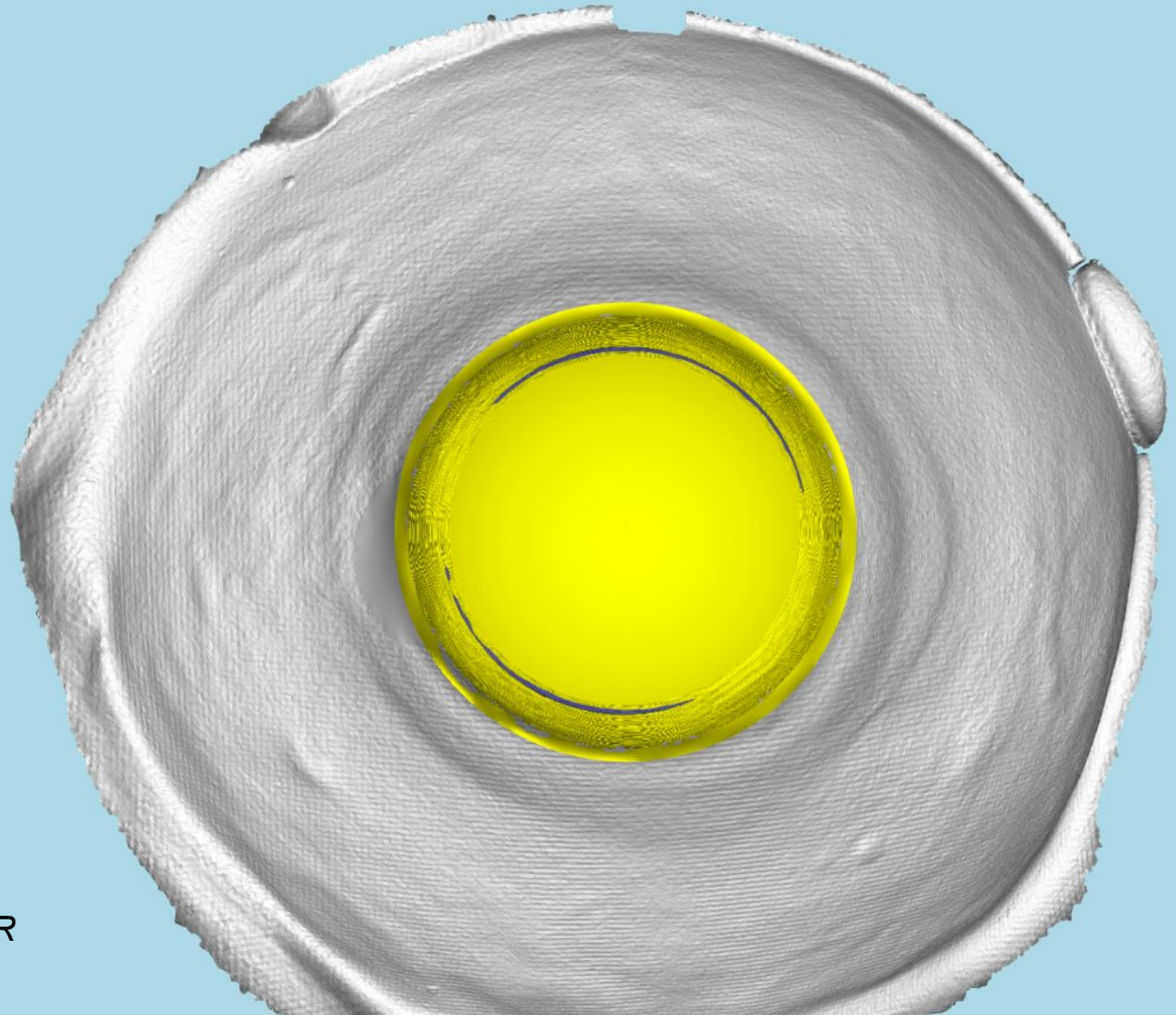

Press F8 to restart upload
L
1/25 Segment: 302° - 122°
2/25 Segment: 294° - 114°
3/25 Segment: 287° - 107°
4/25 Segment: 279° - 99°
5/25 Segment: 271° - 91°
6/25 Segment: 263° - 83°

# Exam Findings

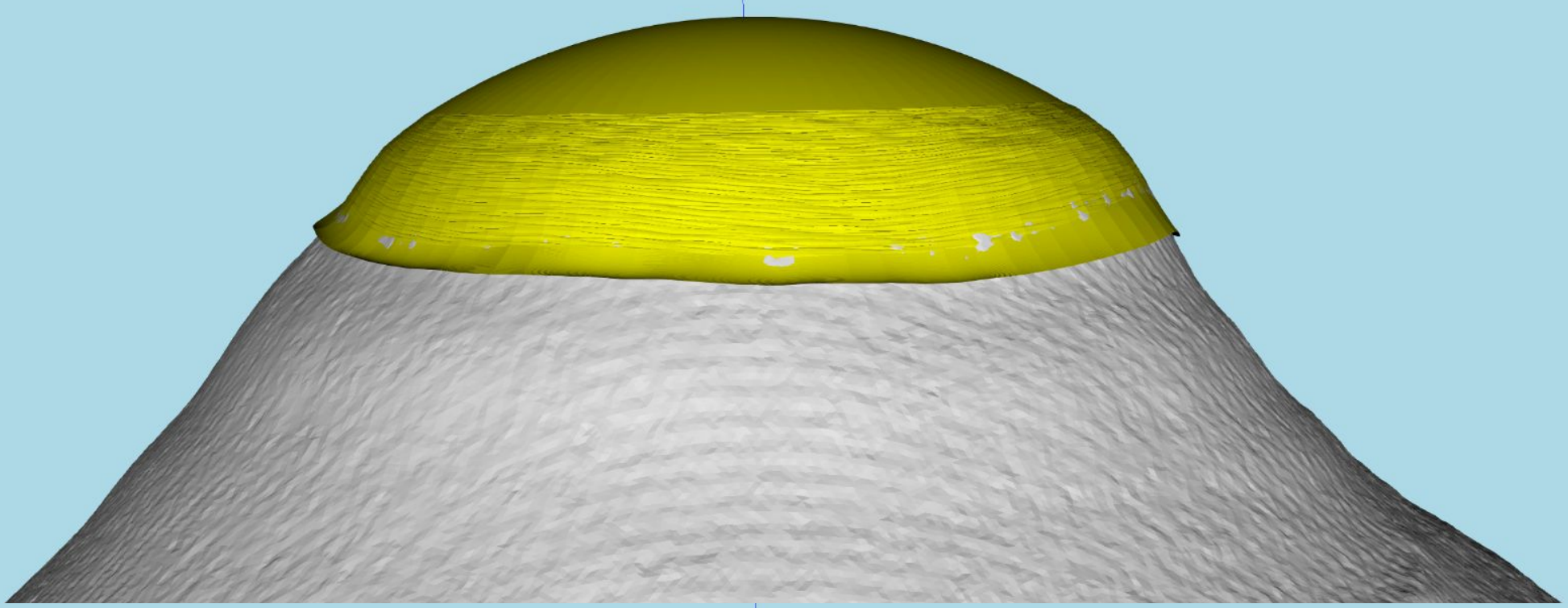
- RGP ORx OD: 20/40ish
- Scleral ORx OS: 20/25ish

But not a candidate for sclerals due to low Endo Cell Count and overall instability of graft.....

# EyePrint Gas Permeable (EPGP)



*Slater DJ, Williams BP, Noyes MR*



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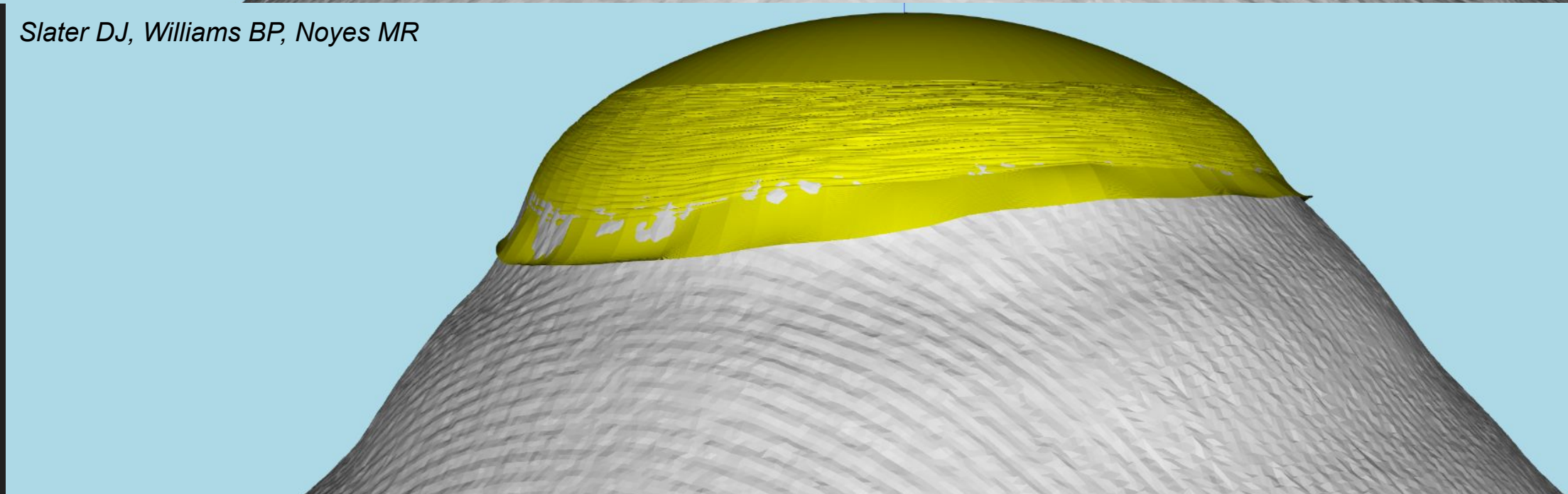
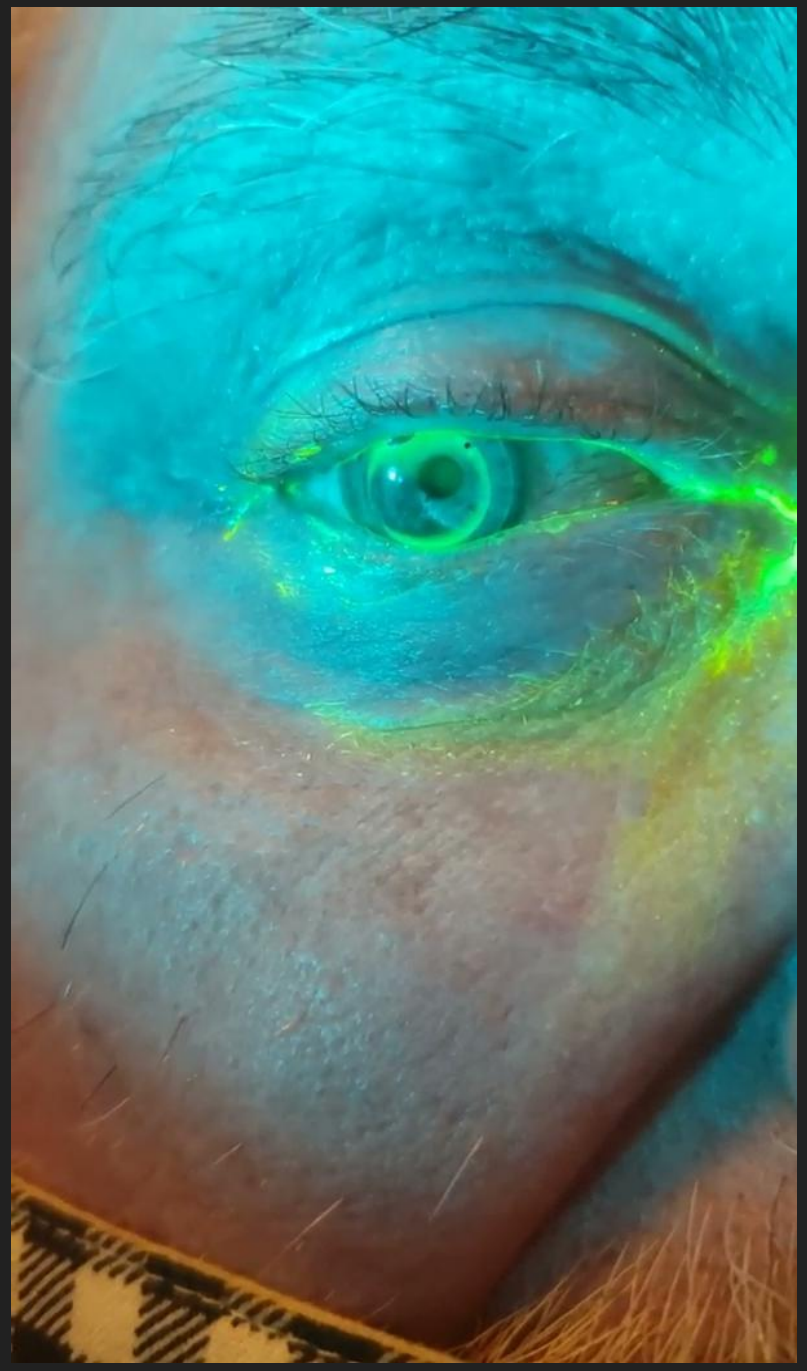




← OD

OS →

# Final Lens Parameters

| Brand | Overall Diameter | Base Curve | Power | CT | Material | Additional Modifications |
|---|---|---|---|---|---|---|
| EyePrint GP | 10.0mm | 7.104mm | -7.00 DS | 0.18mm | Optimum Infinite | HydraPeg |
| EyePrint GP | 9.0mm | 6.323mm | -13.75 DS | 0.18mm | Optimum Infinite | HydraPeg |

BCVA: **20/25 OD, 20/20 OS**

# Case 2 Key Points

- Novel technology such as impression-based GP's allow for a much wider variety of corneas to be fit (compared to that of standard GP's)

- It never hurts to try!

# Congenital Ectodermal Dysplasia secondary to Charl

# **Congenital Ectodermal Dysplasia** secondary to Charlie M Syndrome

**Ectodermal dysplasia is a rare hereditary disorder with a characteristic physiognomy**. It is a genetic disorder affecting the development or function of the teeth, hair, nails and sweat glands. Depending on the particular syndrome ectodermal dysplasia can also affect the skin, the lens or retina of the **eye**, parts of the inner ear, the development of **fingers** and toes, the **nerves** and other parts of the body.


- Deshmukh S, Prashanth S. Ectodermal dysplasia: a genetic review. Int J Clin Pediatr Dent. 2012 Sep;5(3):197-202. doi: 10.5005/jp-journals-10005-1165. Epub 2012 Dec 5. PMID: 25206167; PMCID: PMC4155886.

# Congenital Ectodermal Dysplasia secondary to **Charlie M Syndrome**

**Table 2.** Oromandibular Limb Hypogenesis Snydromes (OLHS): Chicarilli classification[14].

| | Type I; Micrognathia (mandibular) with | Type II; Microglossia with | Type III; Dysgnathia with | Type IV; Miscellaneous |
|---|---|---|---|---|
| A | Pierre Robin syndrome | Hypoglossia | Glossopalatine ankylosis | Möbius syndrome |
| B | Hanhart syndrome | Hypoglossia-hypodacytly | Glossopalatine ankylosis-hypodactyly | Charlie M syndrome |
| C | | | | (Amniotic band syndrome) |

**Table 3.** Overview and phenotypical description of type V (Hall)/type IV (Chicarilli) OLHS syndromes without amniotic band syndrome (c.r.: case reports, AD: autosomal dominant, AR: autosomal recessive, XR: X-linked recessive).

| | Inheritance | Orofacial | Skeletal | Other |
|---|---|---|---|---|
| Charlie M | Sporadic, prevalence unknown (<5 c.r.) | - facial asymmetry<br>- hypertelorism, telecanthus<br>- short philtrum<br>- micrognathia<br>- microstomia<br>- aglossia, hypoglossia<br>- absent teeth<br>- cleft palate<br>- gingival fibromatosis, glossopalatine ankylosis | - ectromelia<br>- etrodactyly, oligodactyly | / |

Oromandibular and Limb Hypogenesis Syndrome: Charlie M subvariant (Type IV B)

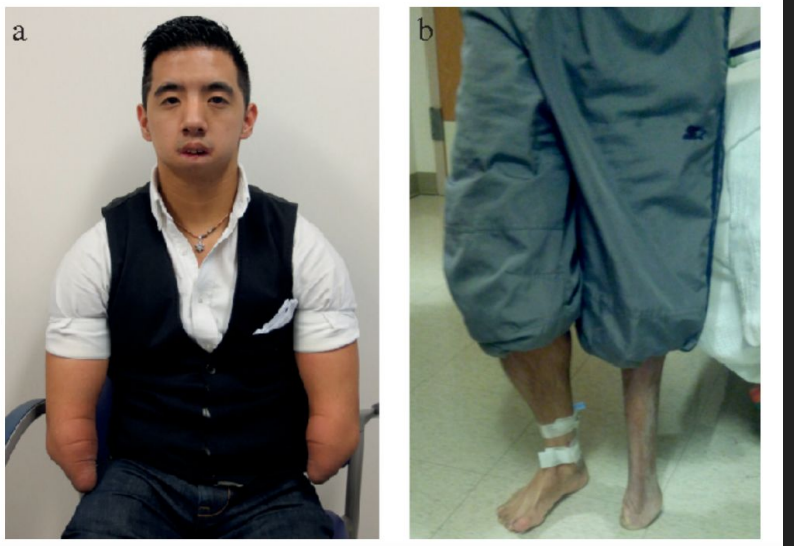



Fig. 2. a (left), b (right): Transverse deficiencies of the upper extremity below the elbow (left) and an absent foot below the upper ankle of the left lower extremity (right).

Jung, Ole, et al. "A patient with Charlie M Syndrome: Differential diagnosis of Oromandibular Limb Hypogenesis Syndromes." Biomedical Papers of the Medical Faculty of Palacky University in Olomouc 160.2 (2016).

# Case History

- Background: 35 year-old Caucasian male referred from oculoplastics for scleral lens evaluation OD
- Chief Complaint: Decreased visual acuity, pain, and dryness OD
- Past Medical History: (+) Congenital ectodermal dysplasia, (+) Charlie M Syndrome, (+) hypodactyly (2 digits per hand)
- Ocular History: (+) limbal stem cell deficiency, (+) symblepharon OD, (+) complete conjunctivalization OS

# Case History

- Systemic Medications: Not contributory
- Ocular Medications: artificial tears prn, cyclosporine 0.05%
- Prior Failed therapies: artificial tears, hydrating ung, bandage soft lens, antibiotic drops

# Exam Findings

Visual Acuity:

- OD: 20/200sc→NIPH
- OS: HM → NIPH

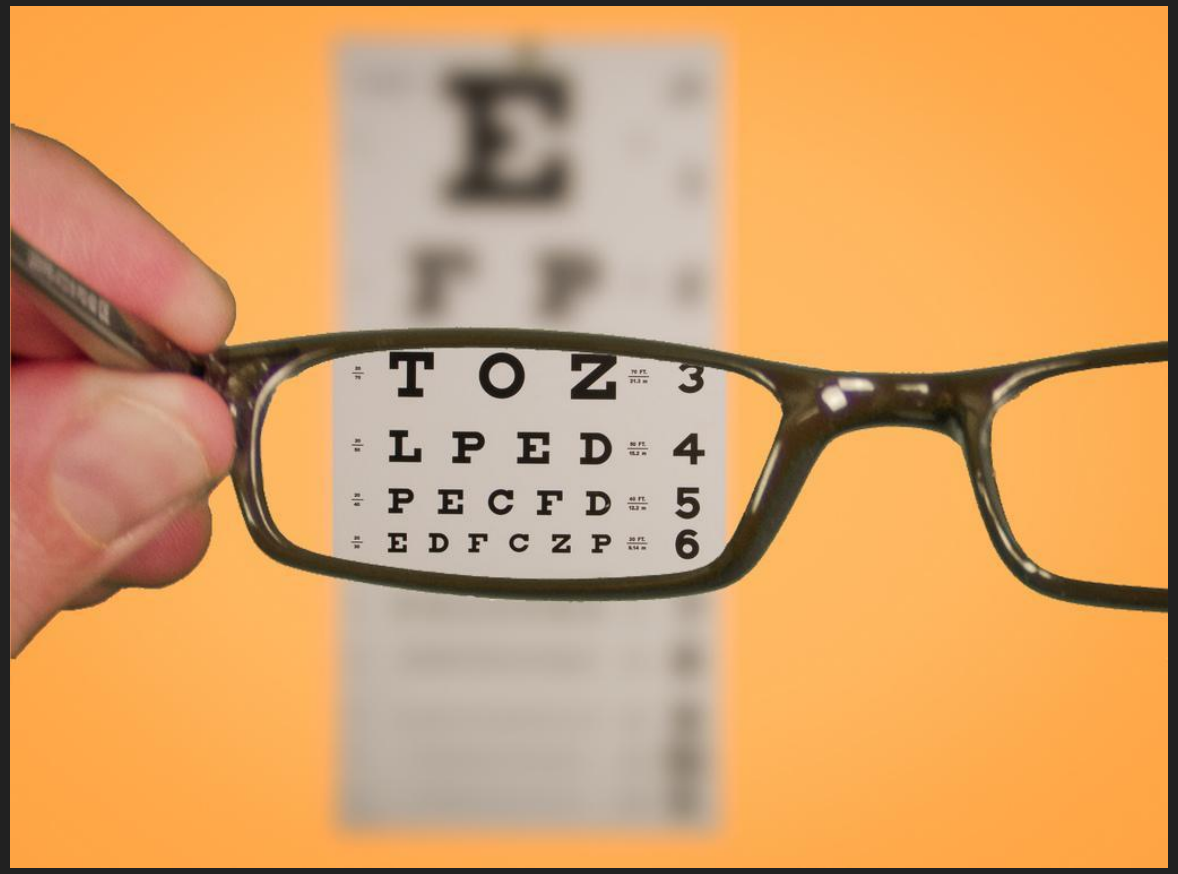
Pupils: Normal OD

EOMs: Normal OD

IOP: 17mmHg OD, STP OS

Fundus: 0.2 C/D OD, No view OS

CVF: Normal OD



# Slit lamp examination OD

| | |
|---|---|
| **Eyelids/Adnexa** | Thickened lids with trichiasis |
| **Conjunctiva** | **Inferior and nasal symblepharon** formation with **fusion of lid margin to bulbar conjunctiva** just inferior to the limbus, 2 lashes from meibomian glands on central upper lid, 3 misdirected lashes at nasal canthus |
| **Cornea** | Moderate central and nasal **haze impinging on visual axis**, **deep stromal neovascularization** nasally from 2 to 5 o'clock |
| **Anterior Chamber** | Deep and quiet |
| **Iris** | Normal |
| **Lens** | Lens |
| **Vitreous** | Normal |

# Slit lamp examination OS

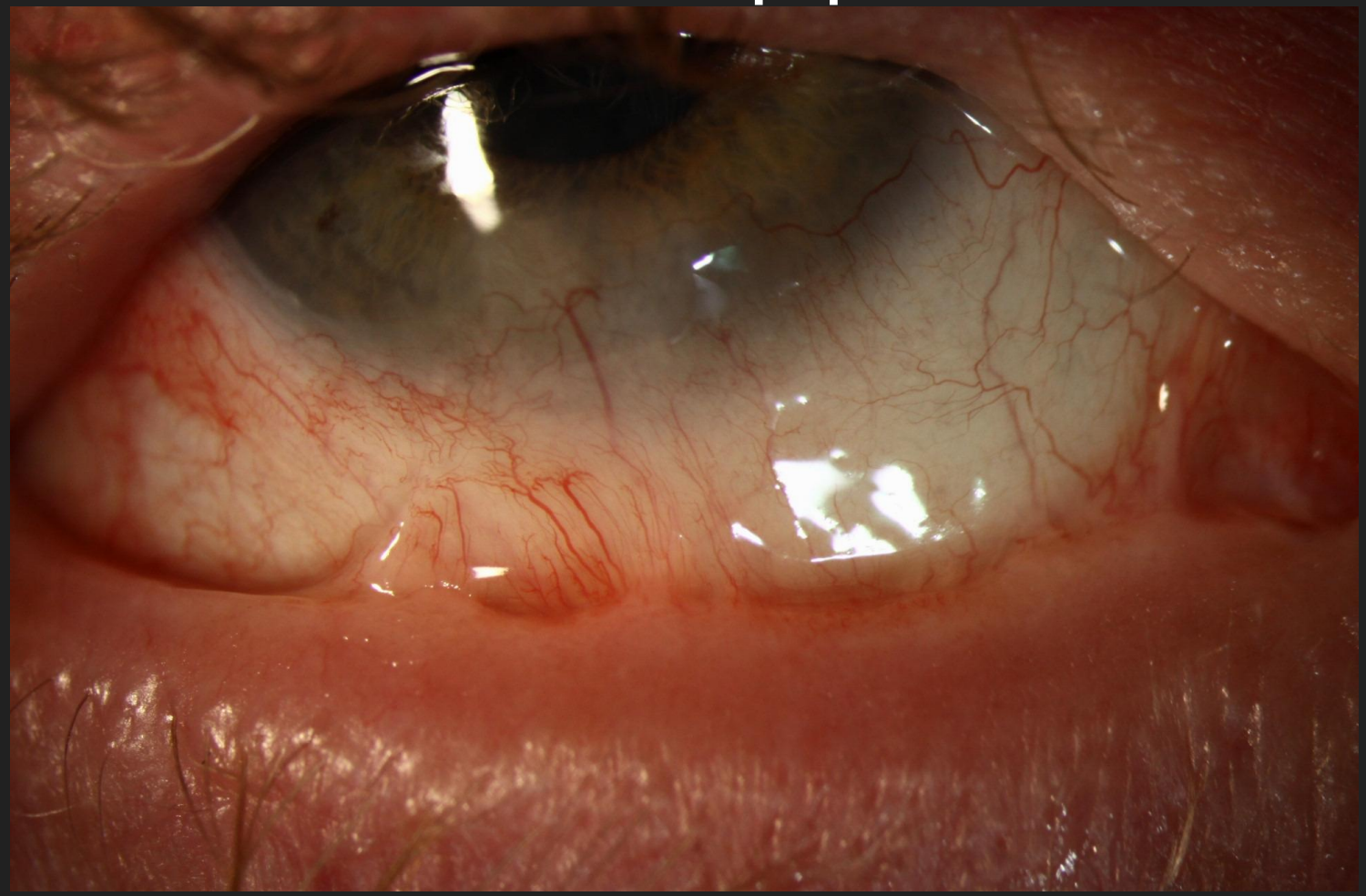
| | |
|---|---|
| **Eyelids/Adnexa** | Thickened lids with trichiasis |
| **Conjunctiva** | Diffuse inferior and superior **symblepharon formation with fusion of lid margin to cornea** |
| **Cornea** | **Complete conjunctivalization** |
| **Anterior Chamber** | No view |
| **Iris** | No view |
| **Lens** | No view |
| **Vitreous** | No View |

# Adnexa Photography

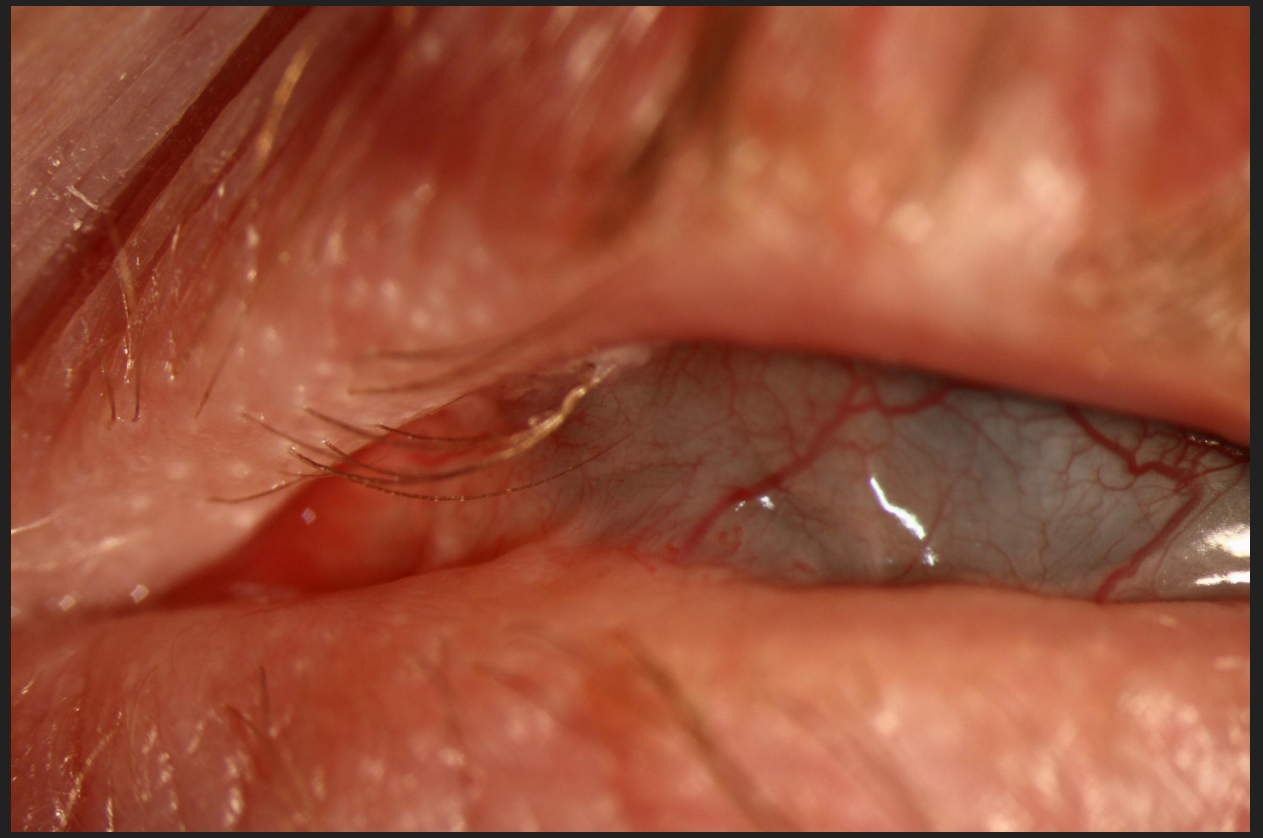
Primary gaze

Opening his eyes as wide as possible

# Initial visit: Slit lamp photo OD



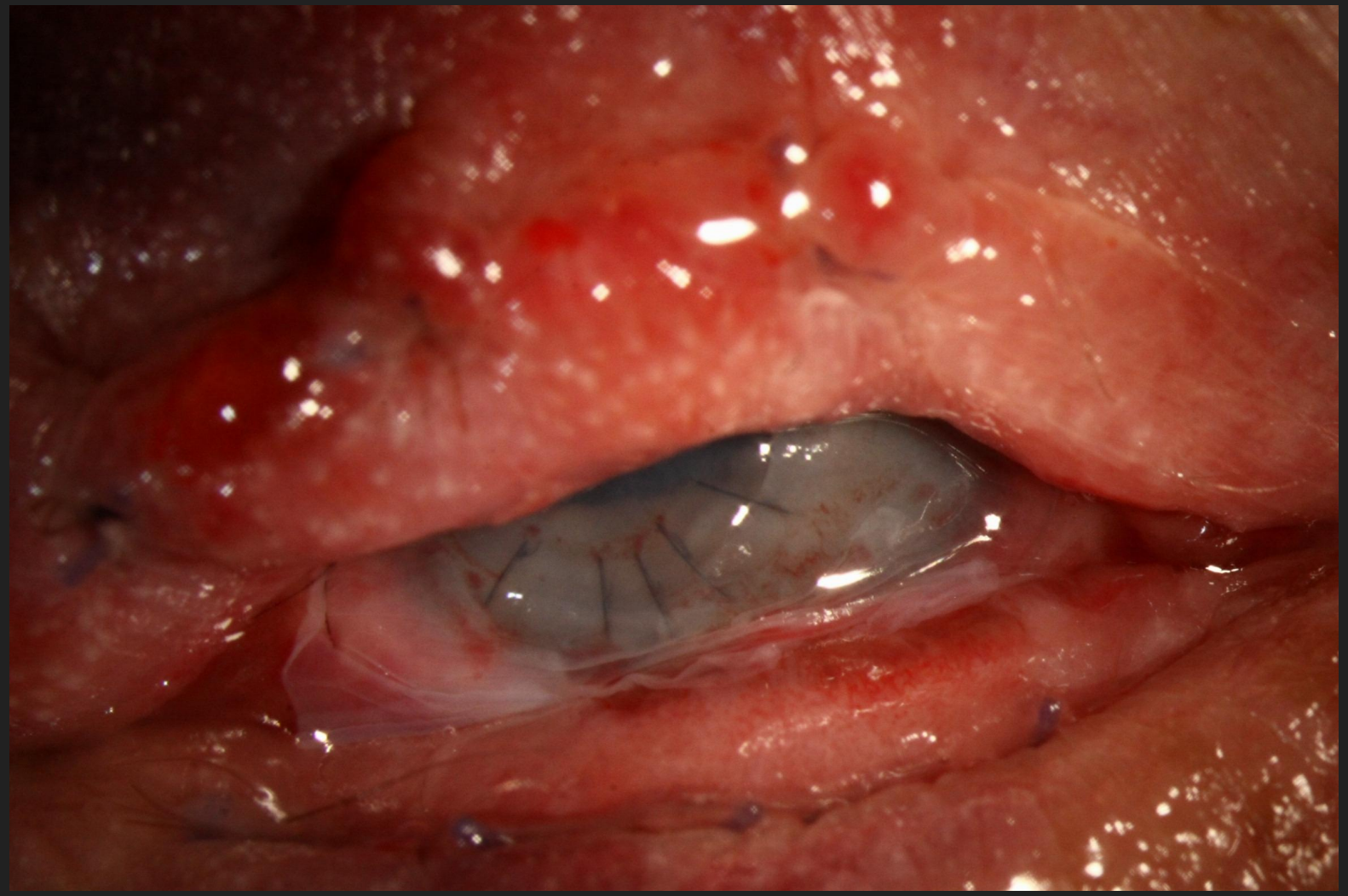
# Initial visit: Slit lamp photos OS



# Well what do you want *me* to do?

- Quickly google to see if I can find out what is happening
- Not a good candidate for CLs (no fornix OD, completely conjunctivalized OS)
- Run out of the room and talk with the oculoplastics team
    - **Maybe we can *create* a fornix (??)**

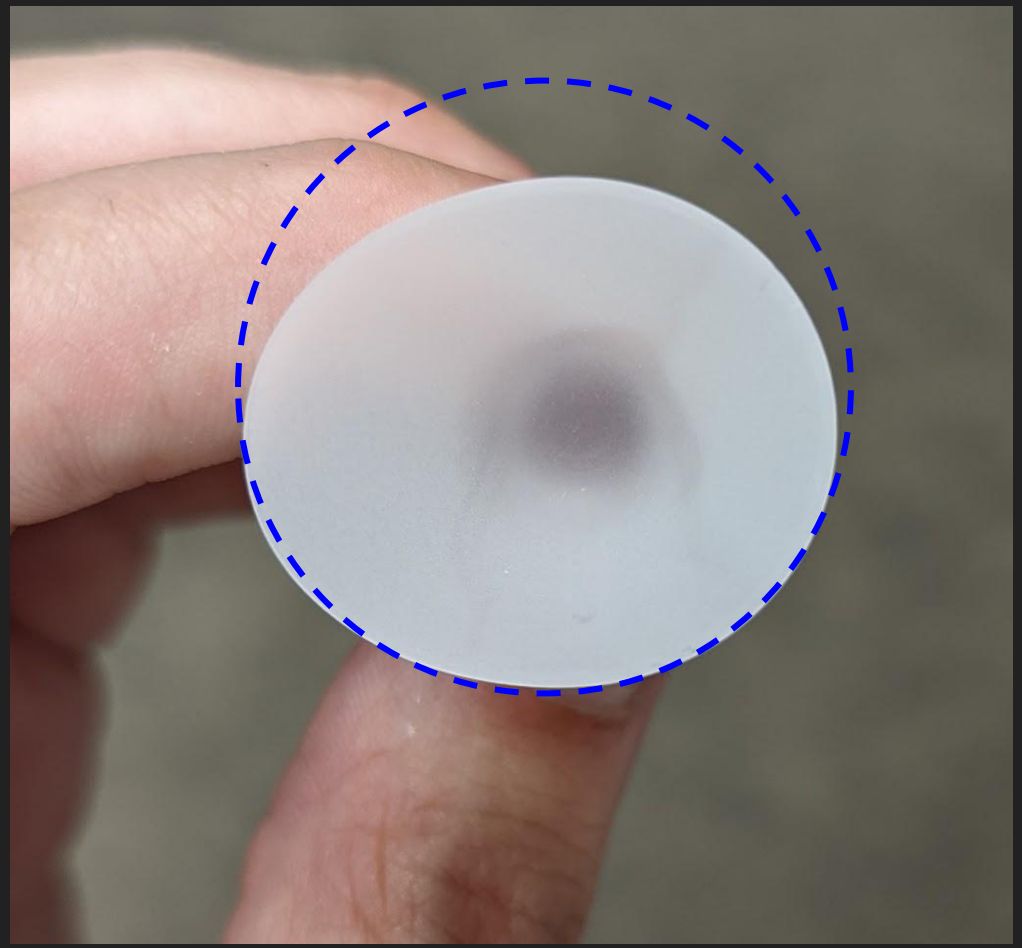
# Post-Fornix Creation Surgery



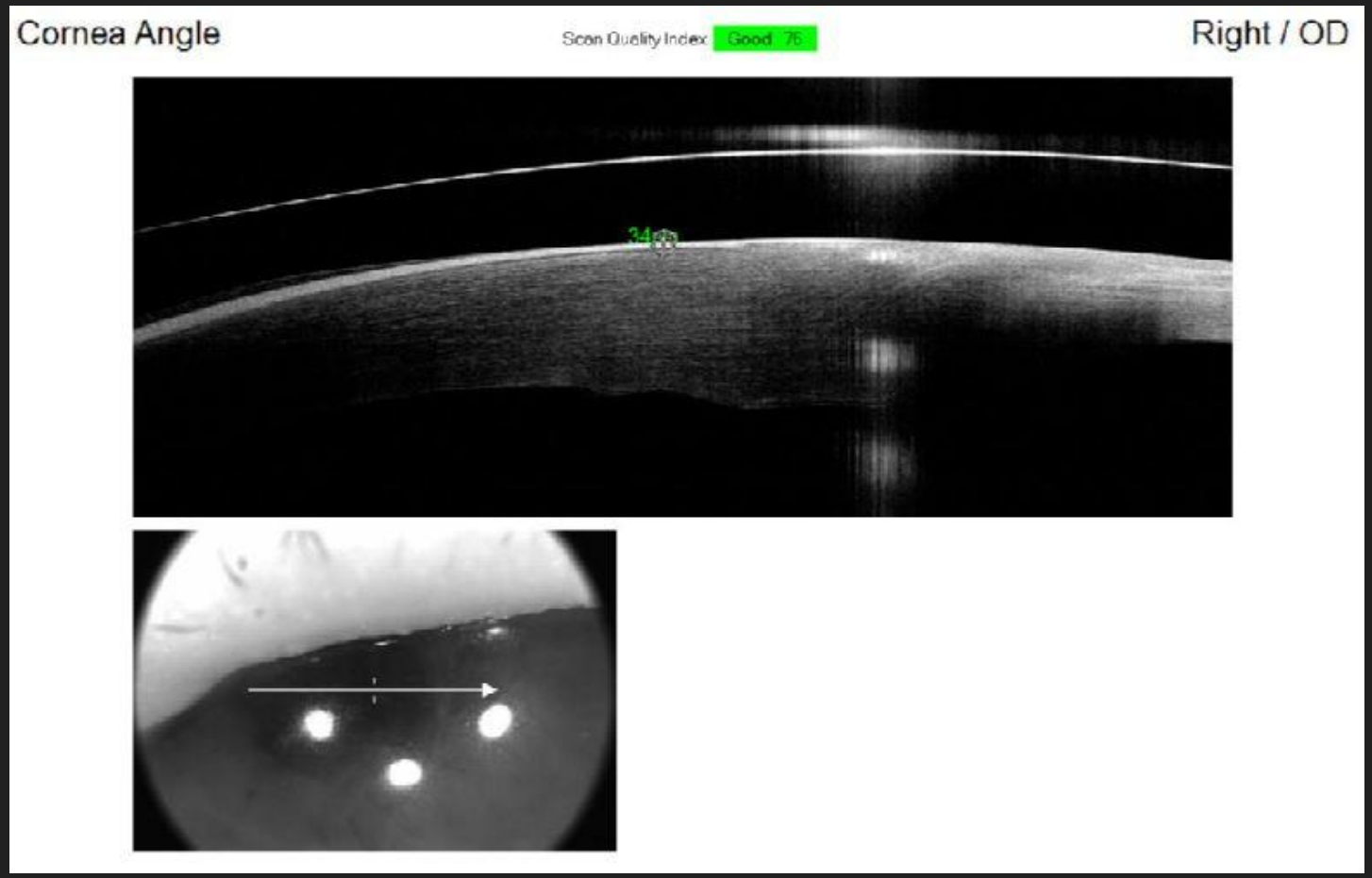
**BCVA: CF 3’**

# I’m supposed to fit that??

# Fitting a modified EyePrint PRO

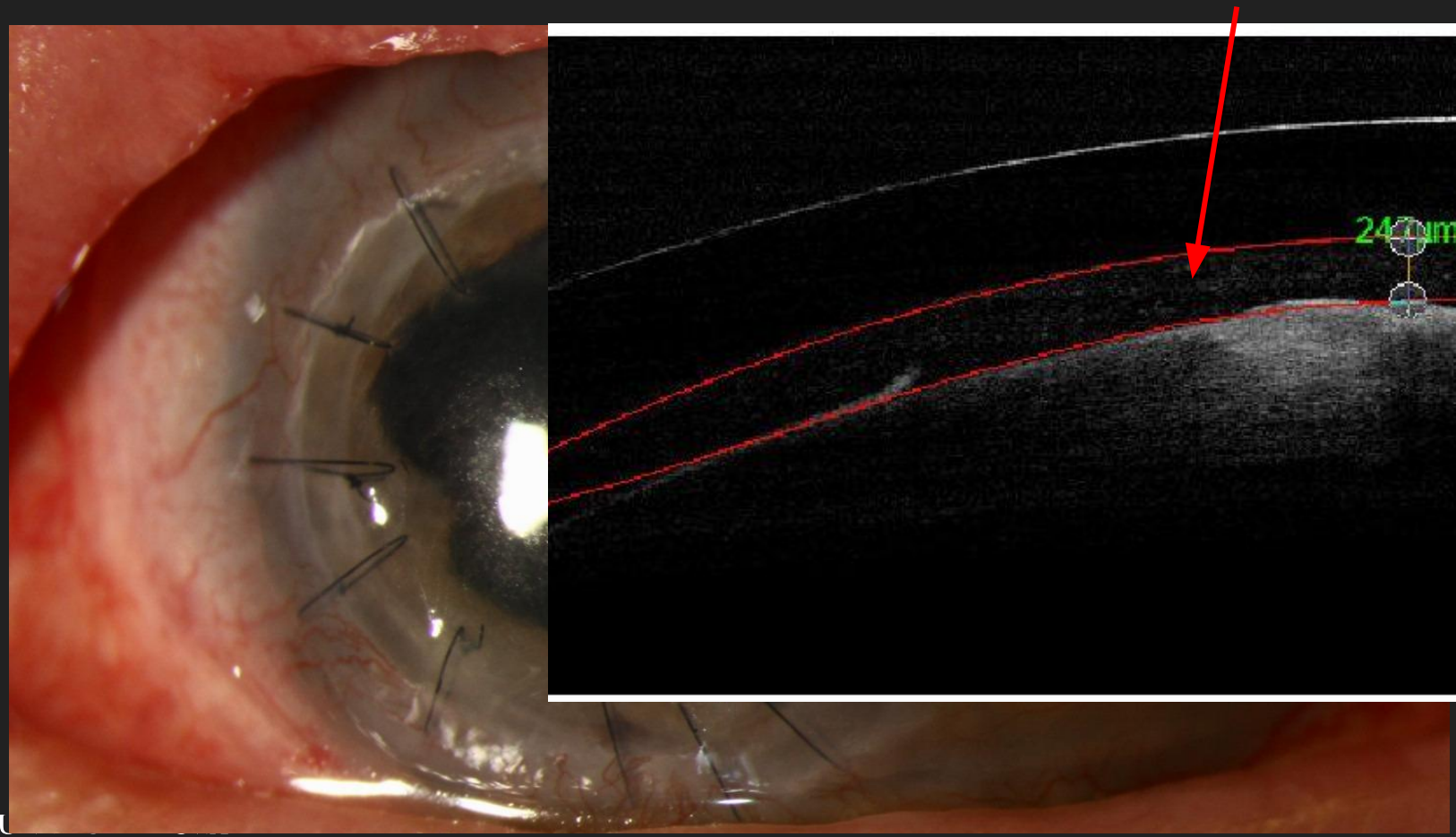


# Lens Dispense

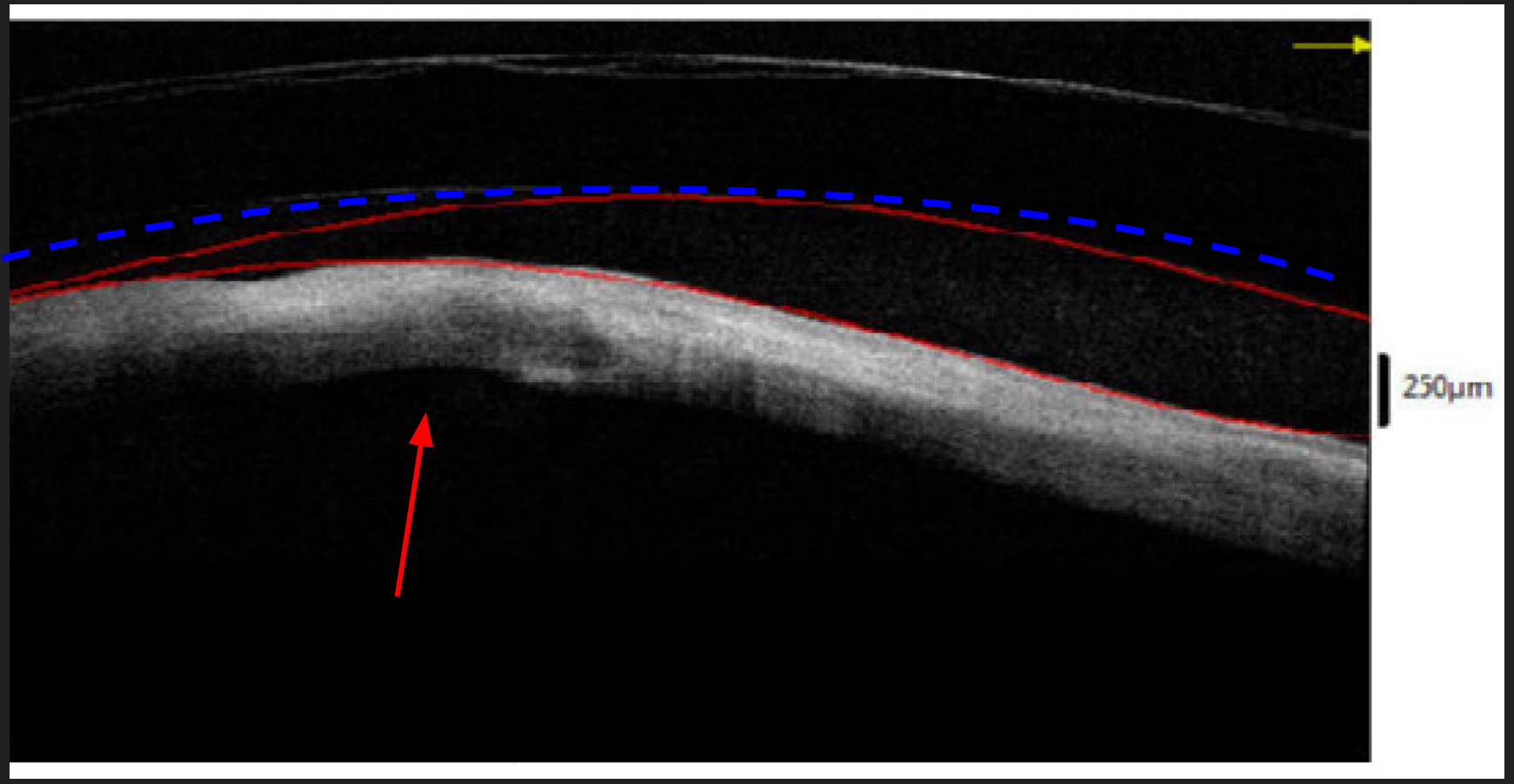



**BCVA 20/300**

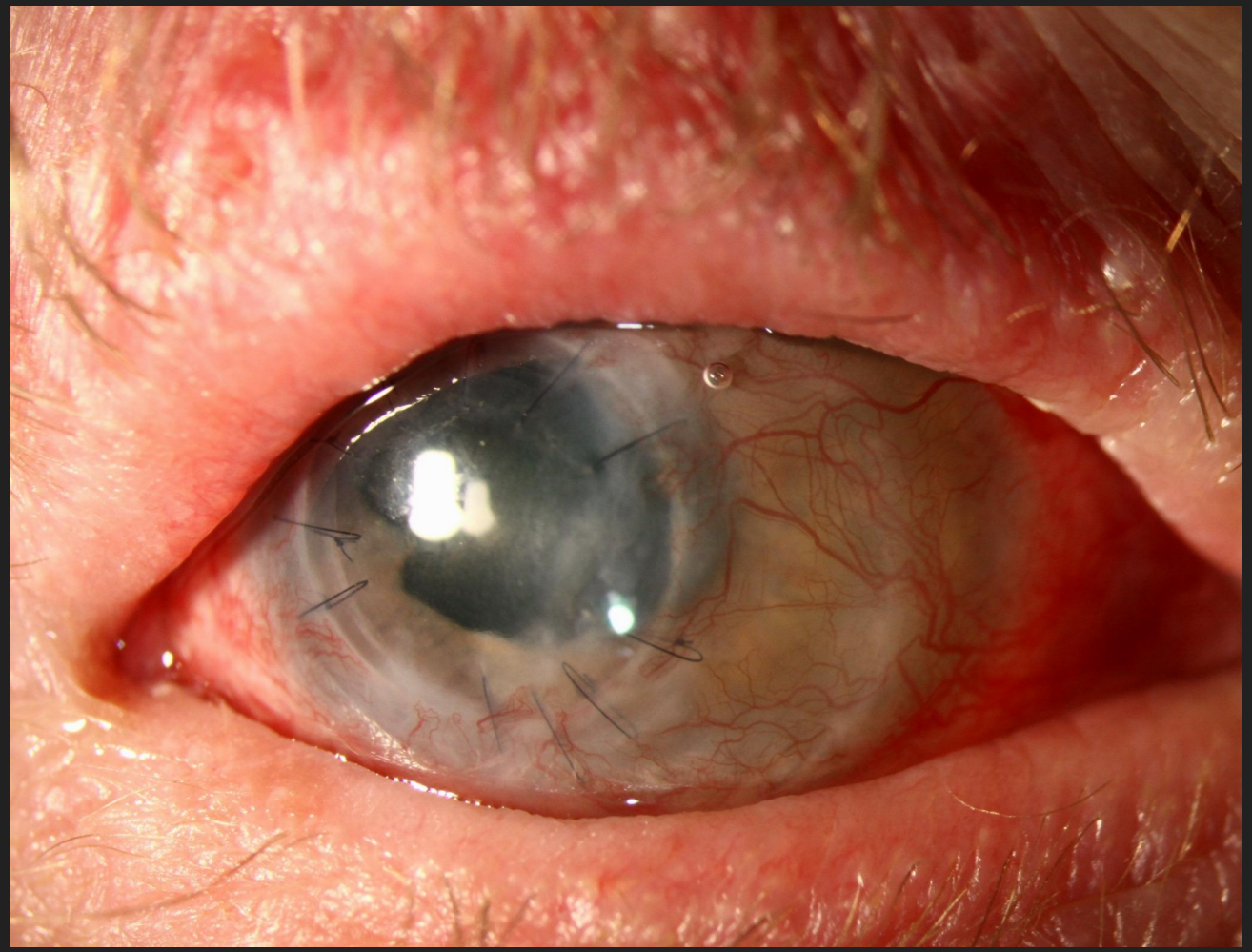
# 1 month (and a few follow-ups) later



# Epi-defect resolved!



# Final Lens

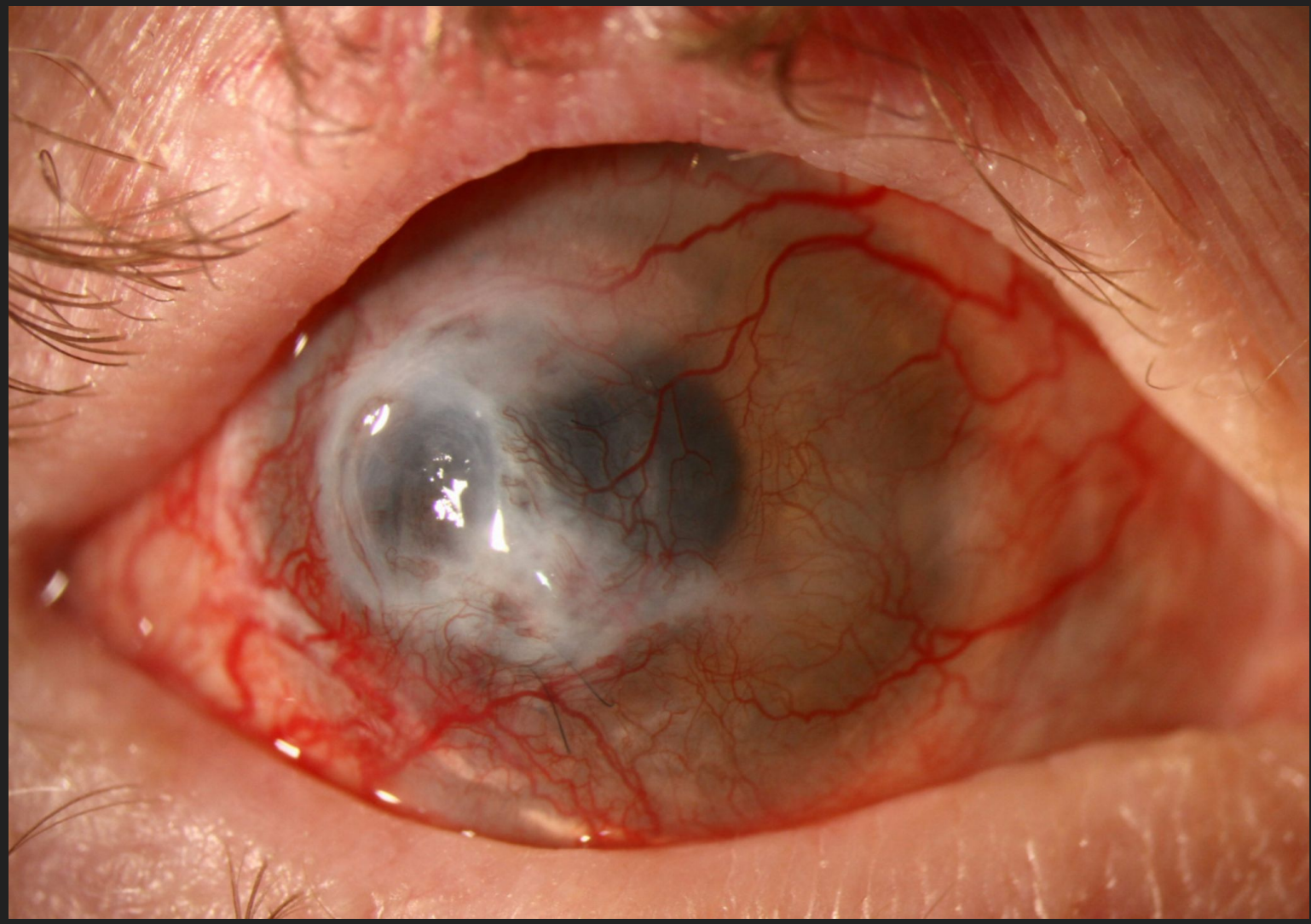


# Final Lens Parameters

| Brand | Overall Diameter | Base Curve | Power | CT | Material | Add-ons |
|---|---|---|---|---|---|---|
| EyePrint PRO (EyePrint Prosthetics) | 15.0mm | 7.273mm | -19.25 DS | 0.3mm | Optimum Infinite | Hydrapeg |

**BCVA: 20/100 → 20/70 → 20/50 → 20/30 !!**

# Approximately 1 year later

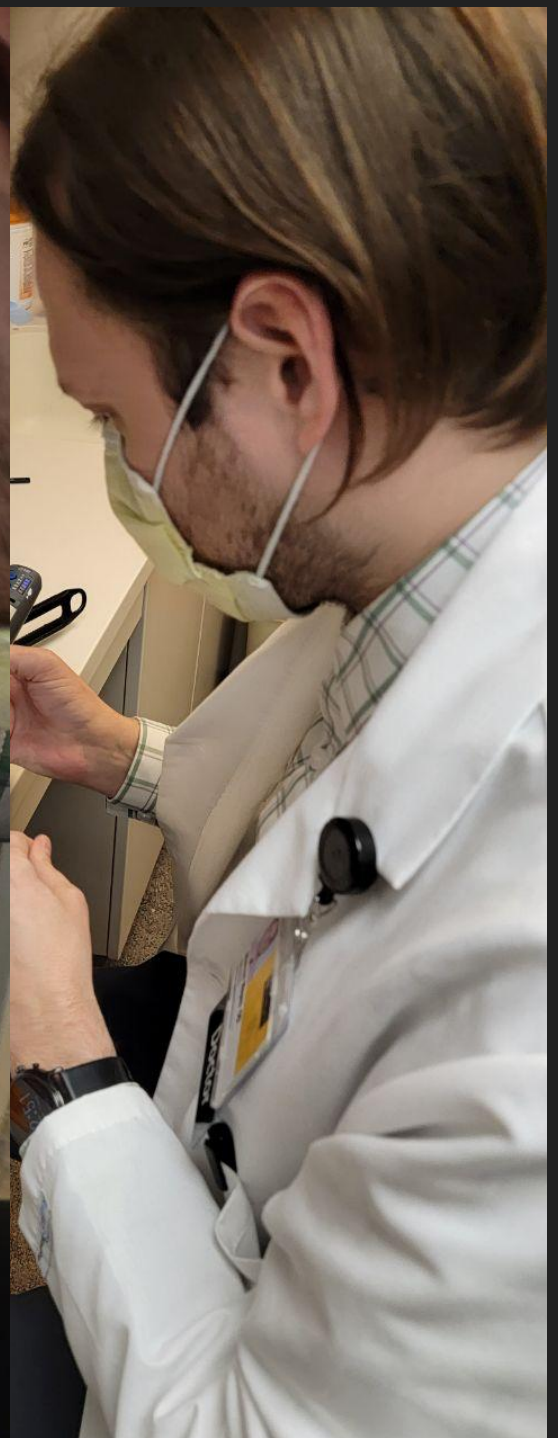


# In-office Lens

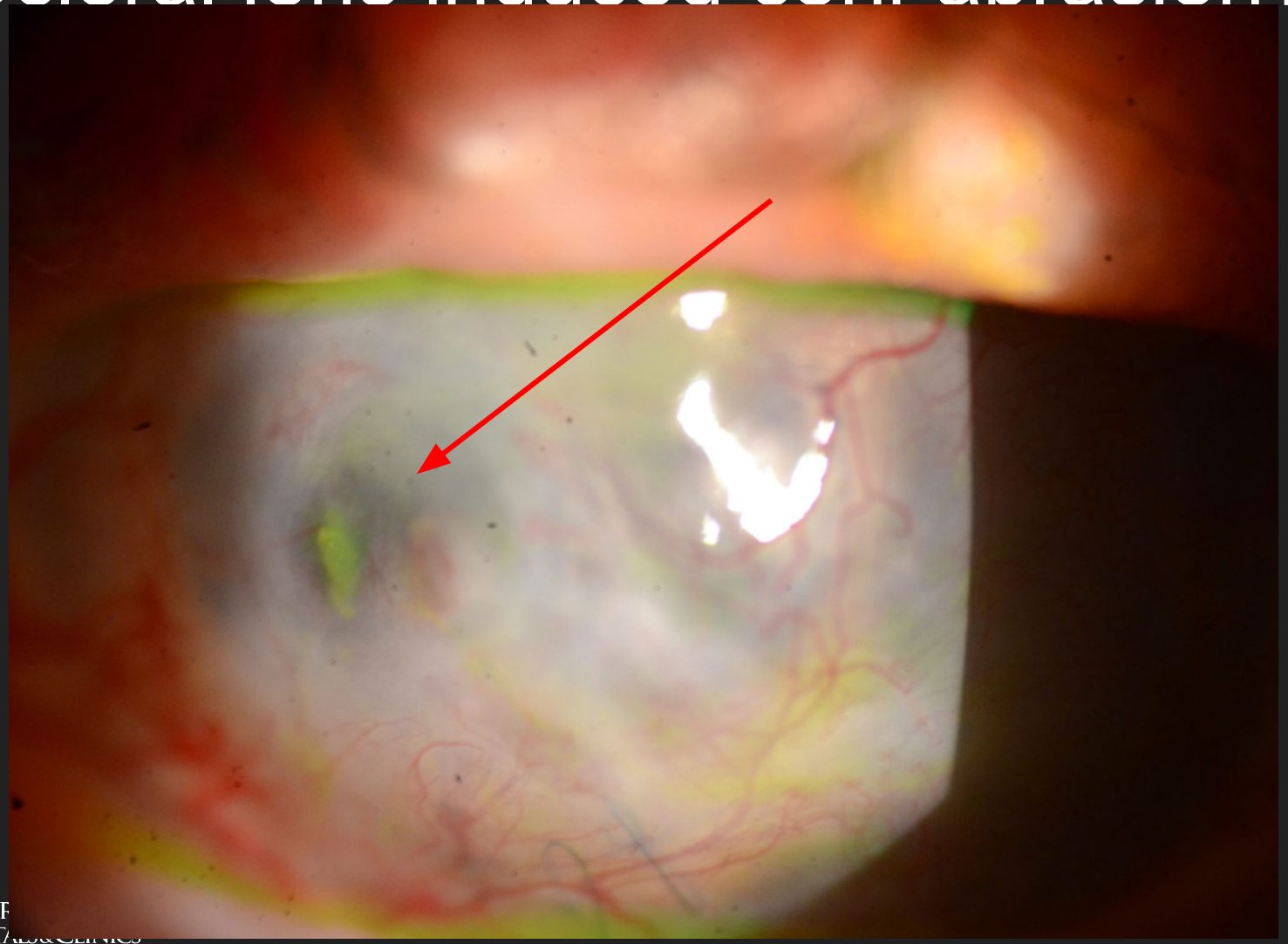
Fenestrated his lens in

improve tear exchange

permissibility

# Scleral lens induced coni-abrasion?

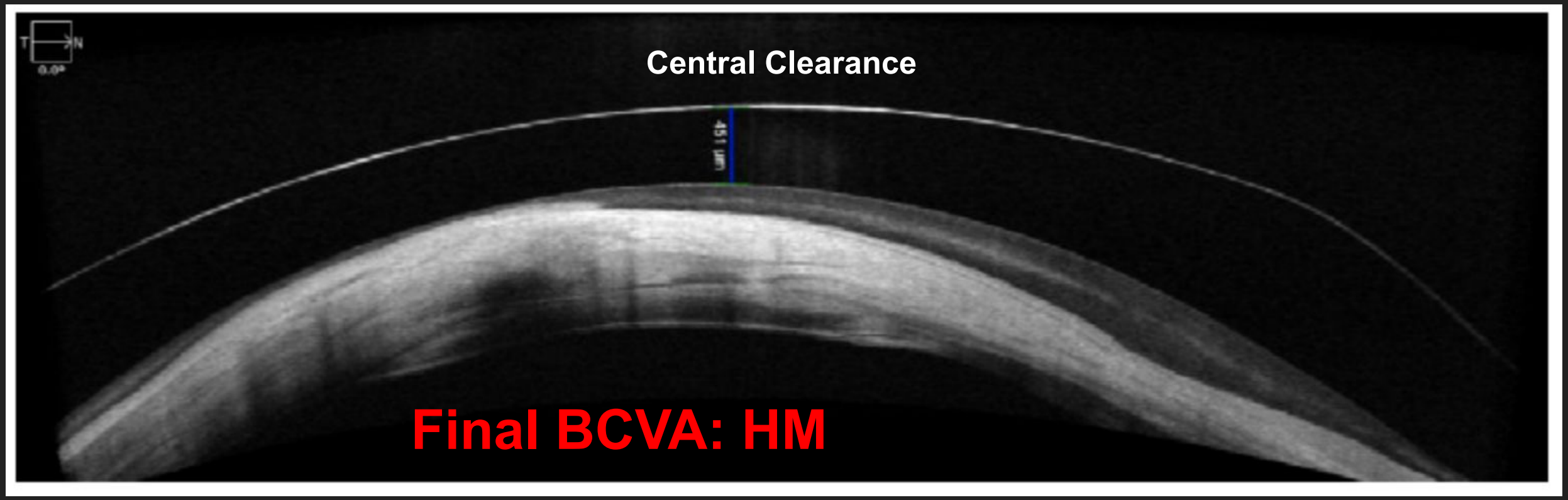


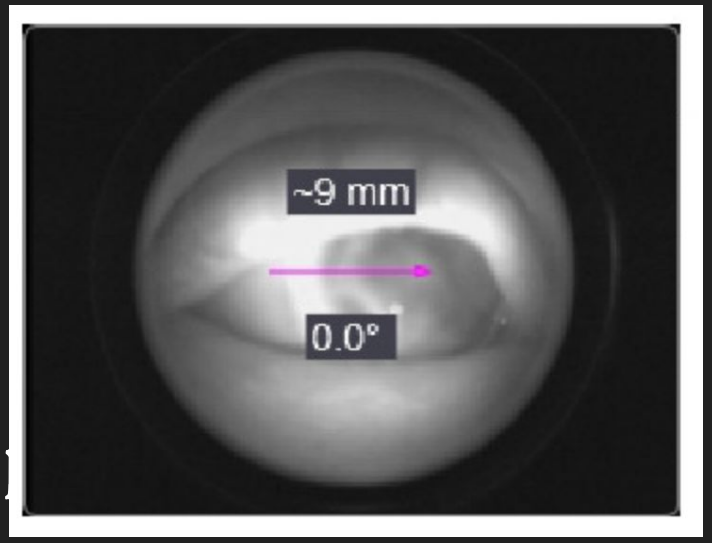
# Back to Scleral or abandon?

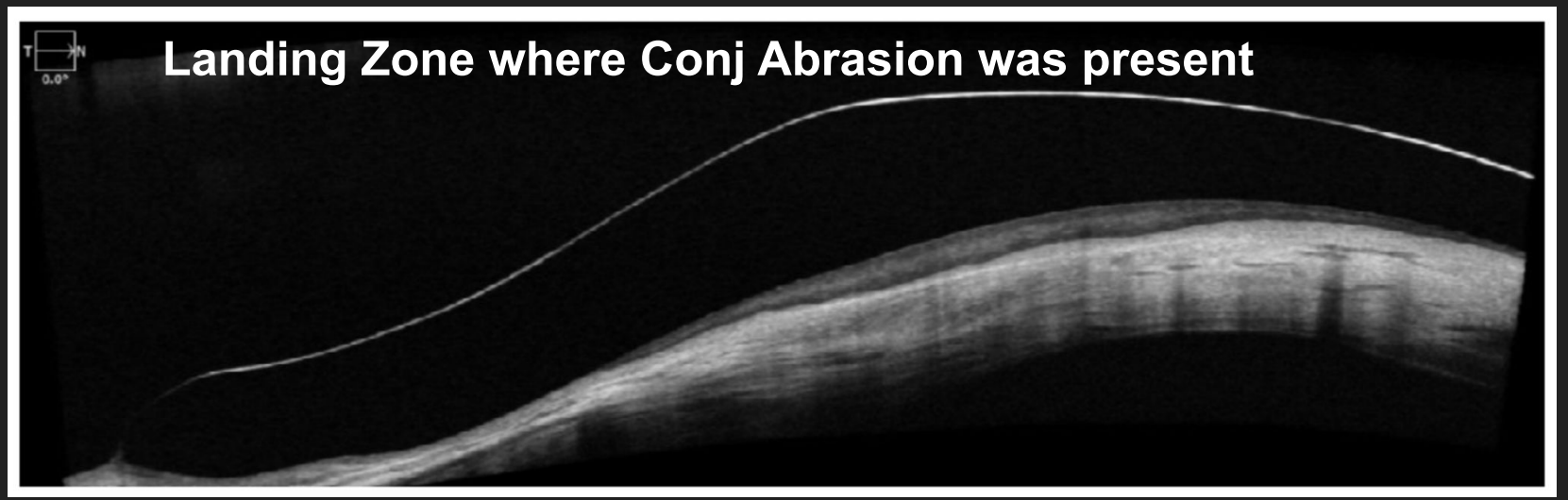
- Chances of salvaging the eye are low
- He has met with Vision Rehab clinic to learn how to function legally blind
- Highly motivated to learn Braille

# One last fit

- Try and keep his eye viable as long as possible
- Impression re-fit with fenestrations


Central Clearance
451 µm
Final BCVA: HM


~9 mm
0.0°


Landing Zone where Conj Abrasion was present

# Lesson Learned

- A lot of the time “the buck stops here”
    - Need to utilize all tools/aspects of scleral lens design and application
- Tailor your expectations accordingly– warn patients if there’s a chance things may head south
- Sometimes “everything” is still not enough– try not to take it personally

# Special Thanks


- Special thanks to David Slater for providing Images

- Also Shante Morgan, Arixa Gibson, Esthera Ansi, Angela Simon, Daren Jamieson, Mariann Sanchez, the Specialty Contact Lens Department, and the Ophthalmic Photography department at the University of Iowa Department of Ophthalmology and Visual Sciences, and EyePrint Prosthetics.

# Thanks!

Questions for me and not for Bryan?