

# PEDIATRIC VITREORETINAL SURGERY ROP, FEVR, COATS, PFV.....



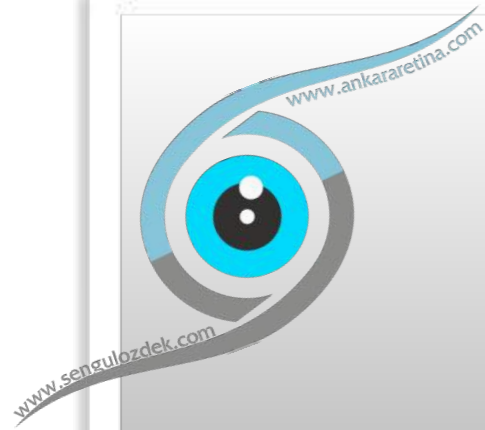
[sengulozdek@gmail.com](mailto:sengulozdek@gmail.com)  
[sozdek@gazi.edu.tr](mailto:sozdek@gazi.edu.tr)  
Ankara, TURKEY





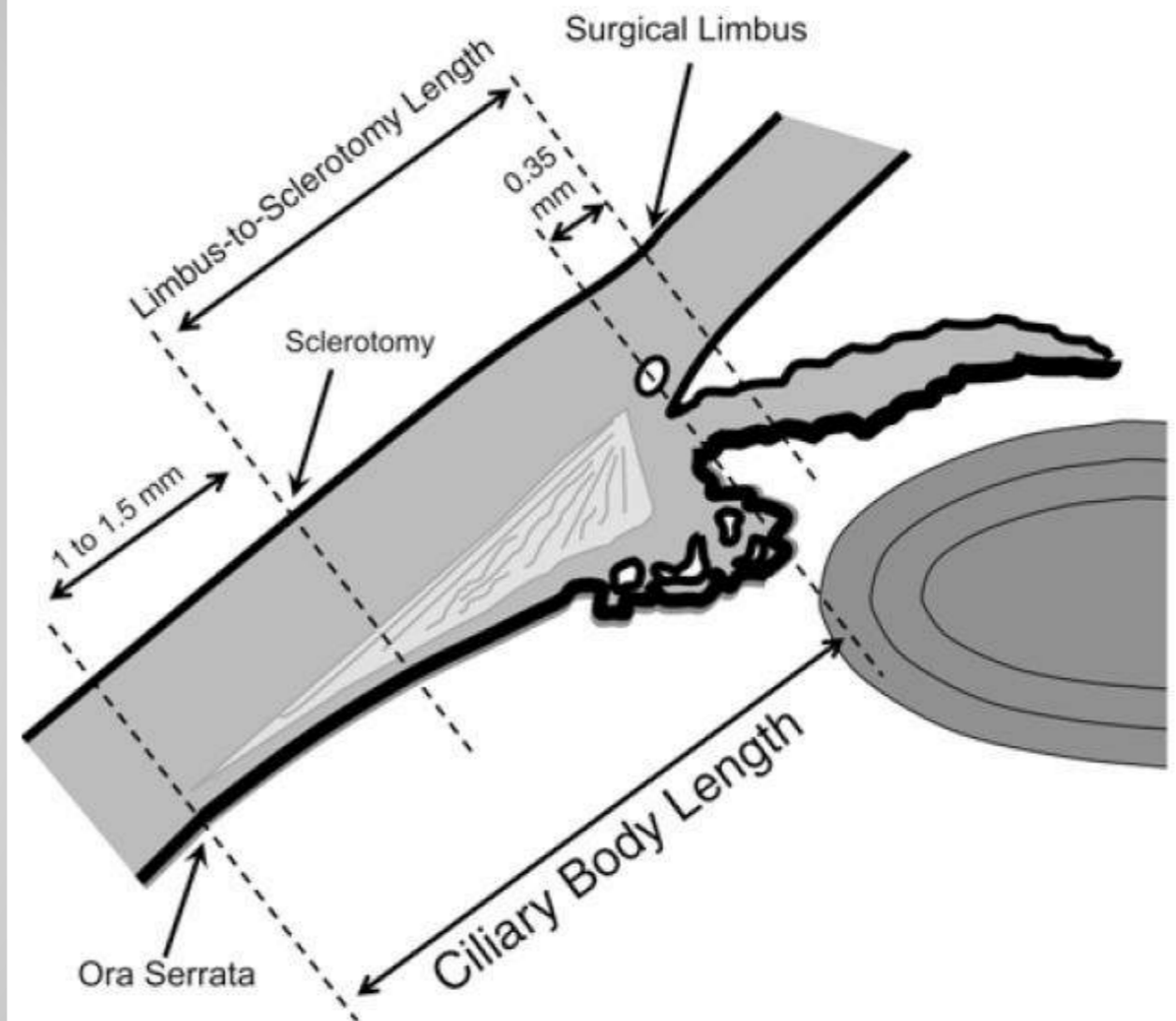
# PEDIATRIC VITREORETINAL SURGERY

- Its own rules
- Its own complications
- Does not forgive faults
- Does not accept apologies



# PEDIATRIK VRS ANATOMY

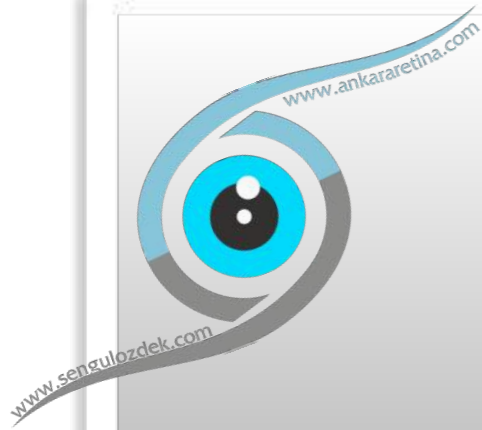
- Very small eye: Narrow space for maneuvers (15-19mm)
- Lens is relatively large
- Vitreous-Retina relation!
- Pars plana has not developed.
- Sclera is thin and elastic





# PEDIATRIC VRS Features

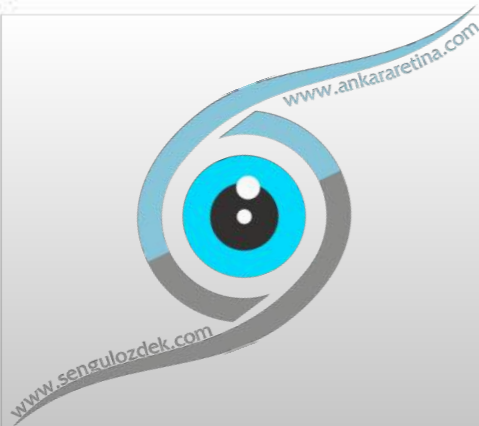
- Retinal breaks usually ends up with surgical failure
- PFCL and Silicon oil are rarely used.
- Children do not obey the head positioning!
- PVR is frequent and severe!
- Amblyopia problem!



# VITRECTOMY

## Sclerotomy Construction

- Pars plicata, Trans iris-root, Limbal
- 2-3 port entry
- 20G-23G-25G-27G surgeries
- Lens sparing vitrectomy (LSV) if possible!
- Lensectomy-Vitrectomy



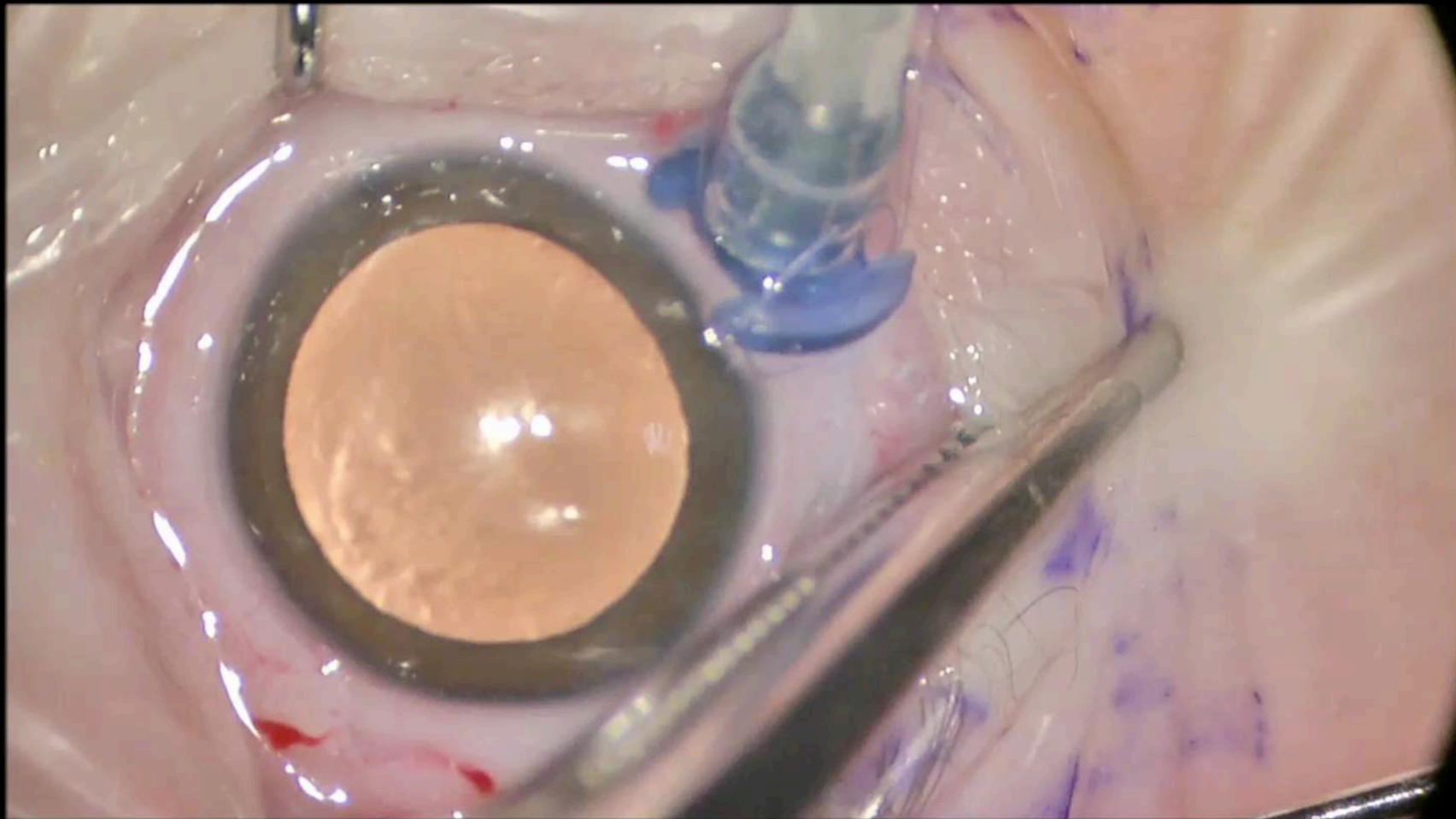
# Sclerotomy

DATA USED TO PLAN SCLEROTOMY LOCATION

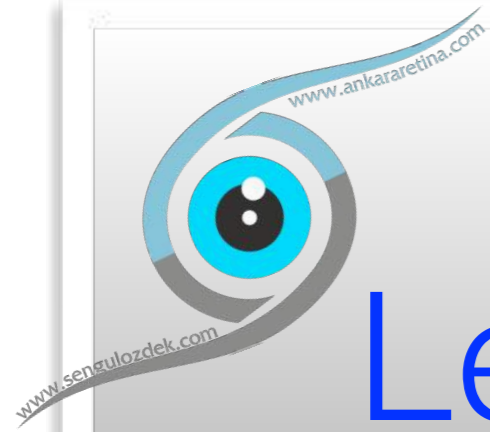
AGE	MINIMUM CILIARY BODY LENGTH (AIELLO ET AL <sup>1</sup> )	MINIMUM LIMBUS-TO-ORA SERRATA DISTANCE	LIMBUS-TO-SCLEROTOMY DISTANCE	
			CALCULATED	APPLIED <sup>*</sup>
0–6 mo	2.60 mm	2.95 mm	1.45 mm	1.5 mm <sup>†</sup>
6–12 mo	2.86 mm	3.21 mm	1.71 mm	2.0 mm
1–2 yr	3.28 mm	3.63 mm	2.13 mm	2.5 mm
2–3 yr	3.75 mm	4.10 mm	2.60 mm	3.0 mm <sup>‡</sup>
Adult	4.60 mm	4.95 mm	3.45 mm	3.5 mm <sup>§</sup>

Aiello AL, Tran VT, Rao NA. Postnatal development of the ciliary body and pars plana: a morphometric study in childhood. Arch Ophthalmol 1992;110:802-805.

# At the very beginning...



Narrow space can be widened with Canthotomy



# When Lensectomy is Planned

1. Iris root entry: Iris problems

2. Limbal entry:

Corneal distortion, endothelial damage

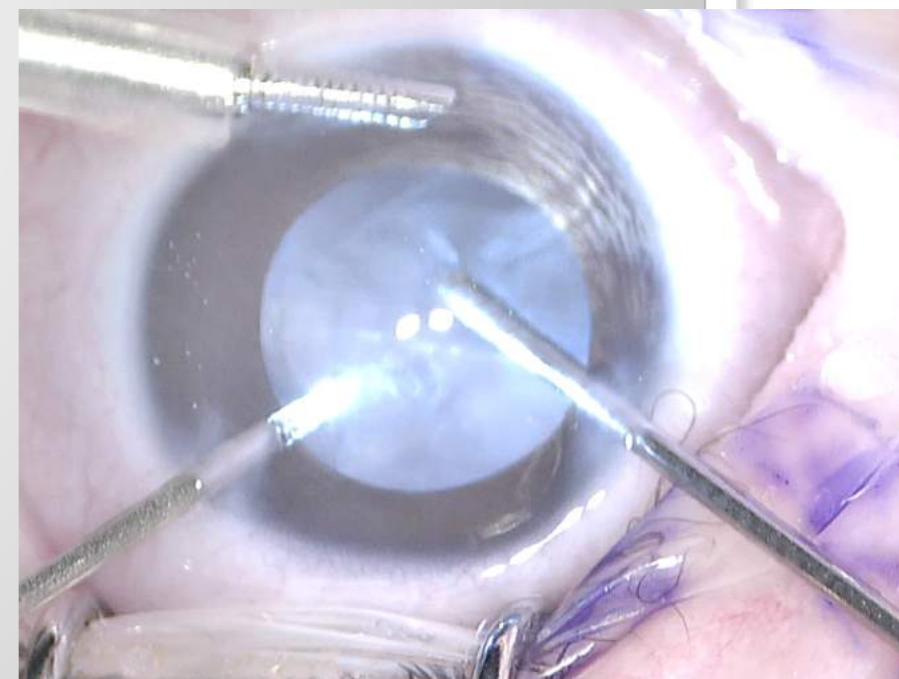
10/0 suture needed (EUGA for removal)

Advantages:

Chantotomy not needed

Conjunctiva is protected

Peripheral retinal damage is avoided

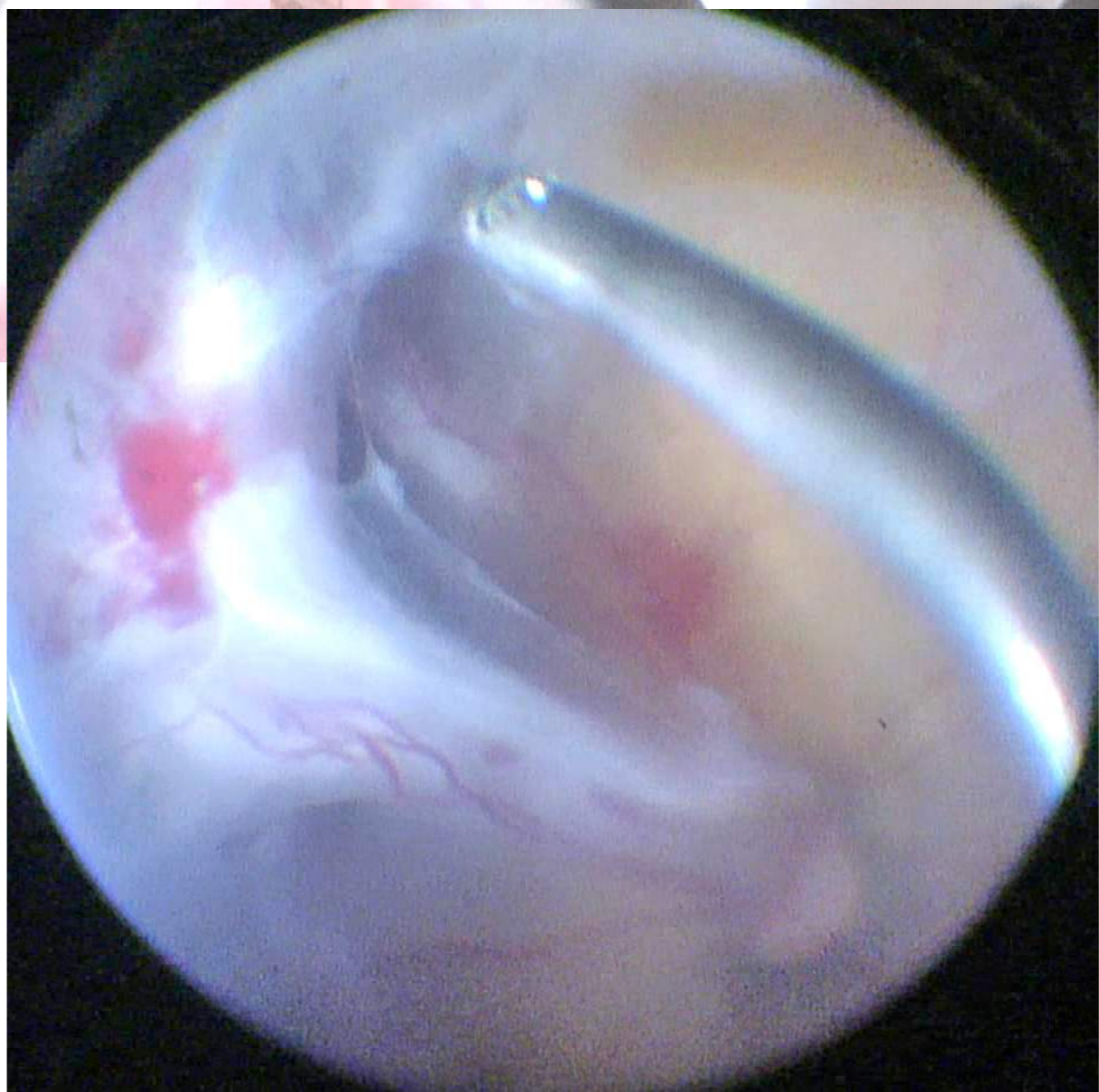
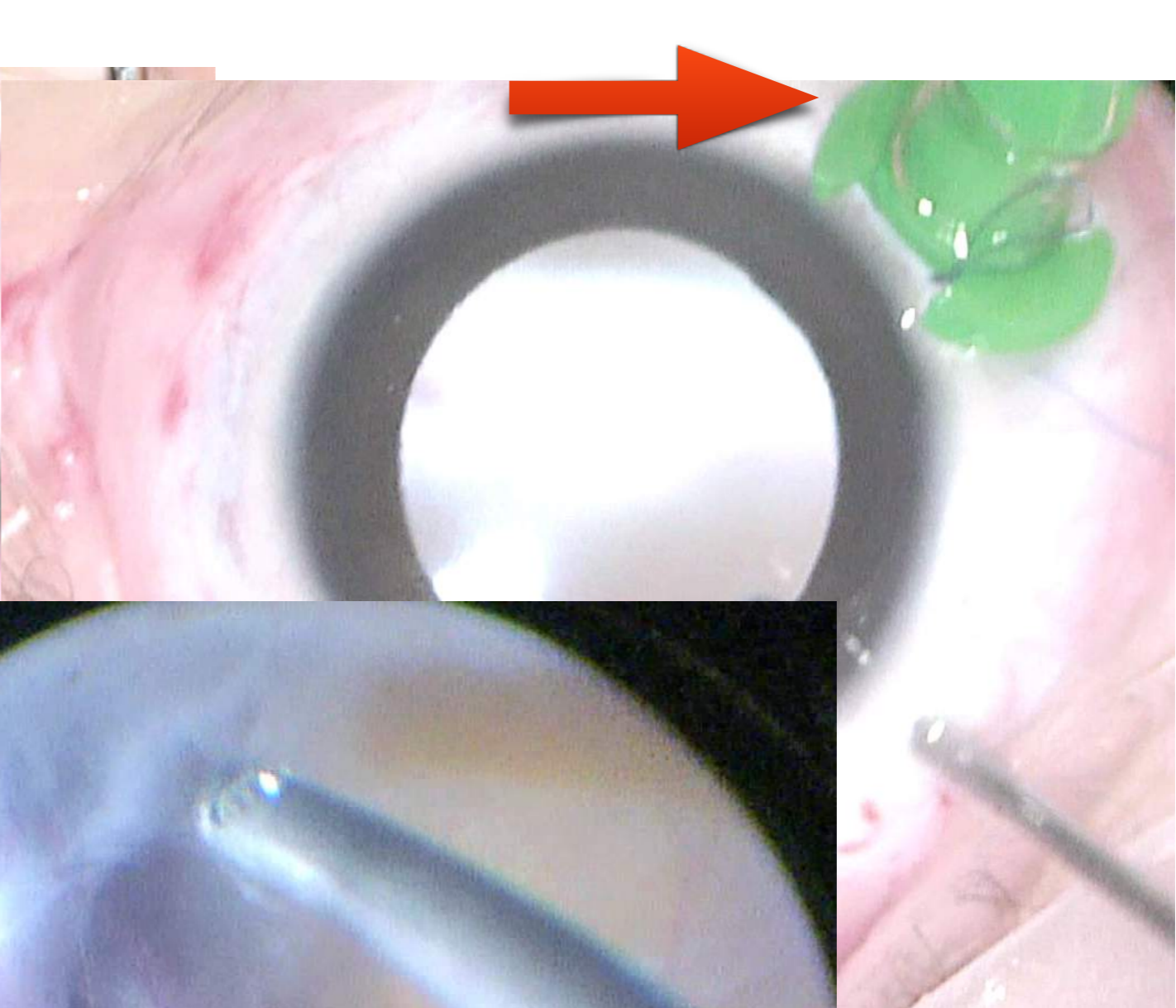






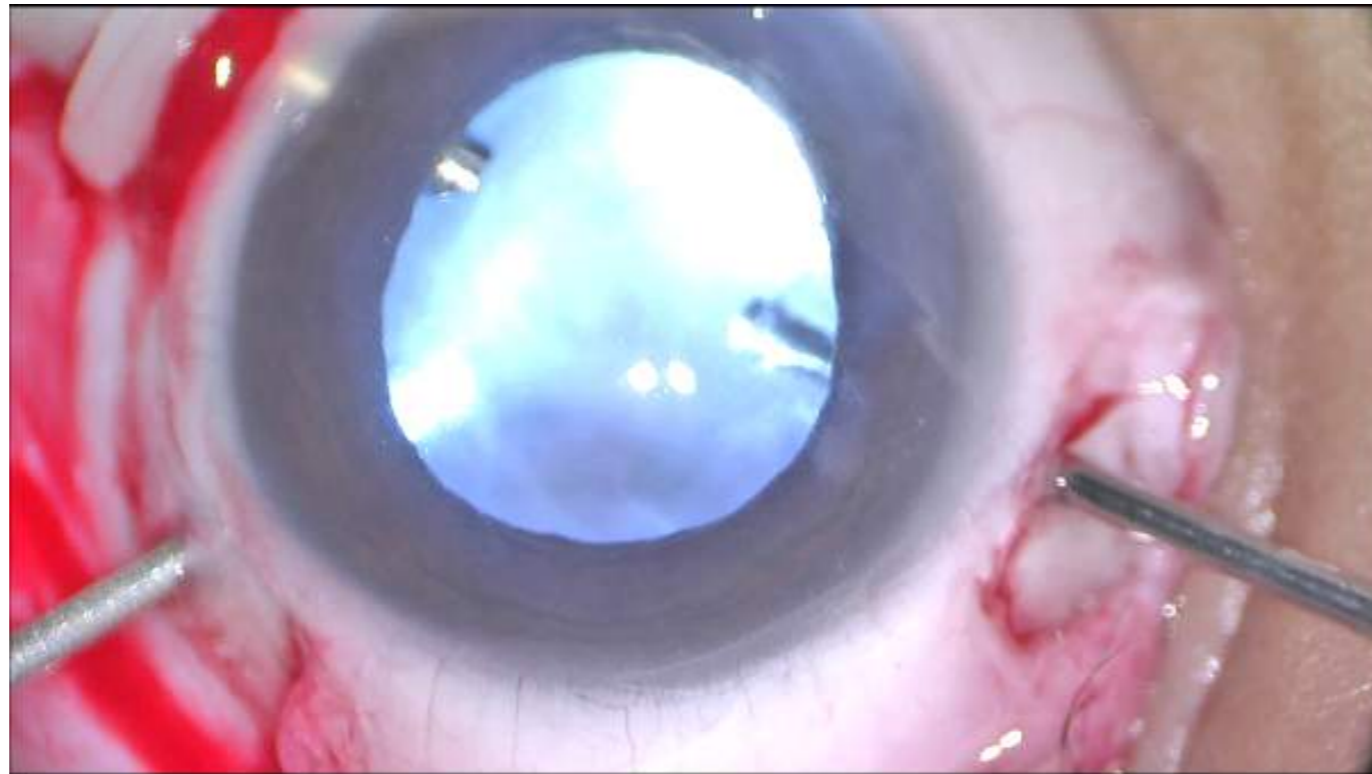
# Choosing Sclerotomy Sites

- Conjunctival peritomy
- Avoid damage to anterior fibrovascular tissue and retina
- Suture all sclerotomies

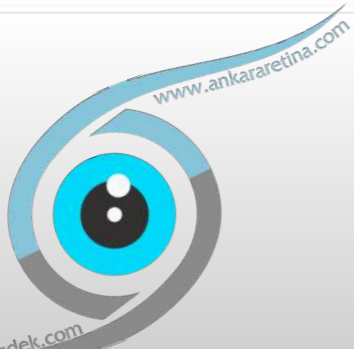




# 3 ports sclerotomy



- Adult type instrumentation, with/without trockars
- Infusion cannula: 23G sutured (4mm)



# PEDIATRIC VRS

ROP

PFV

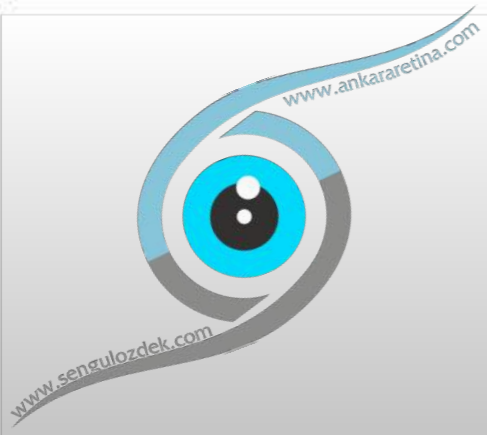
FEVR

COATS

SHAKEN BABY

CXLR

others



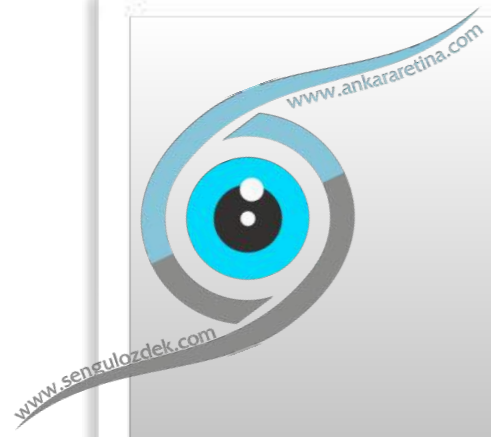
# ROP

STAGE 4A

STAGE 4B

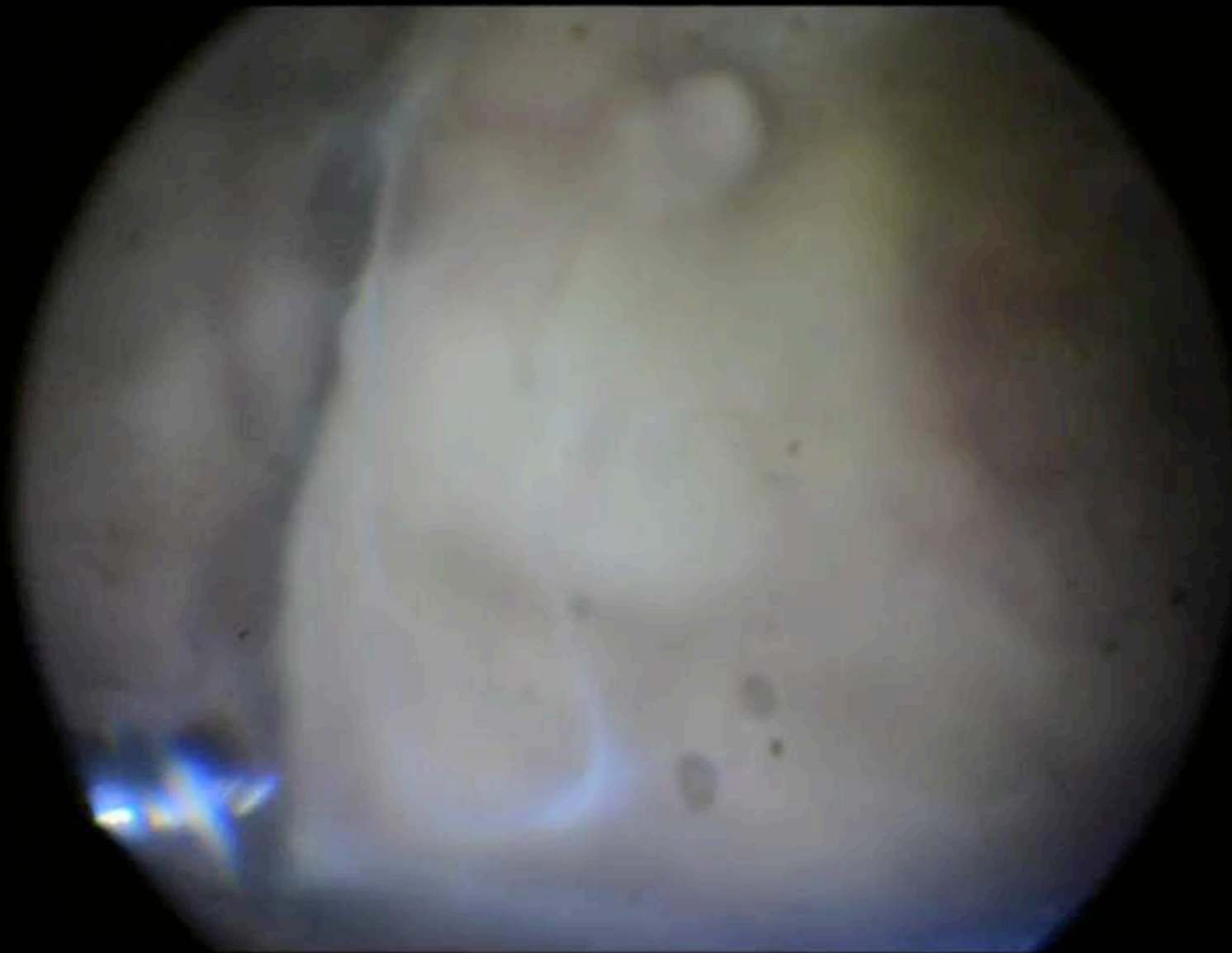
STAGE 5

SCATRICIAL

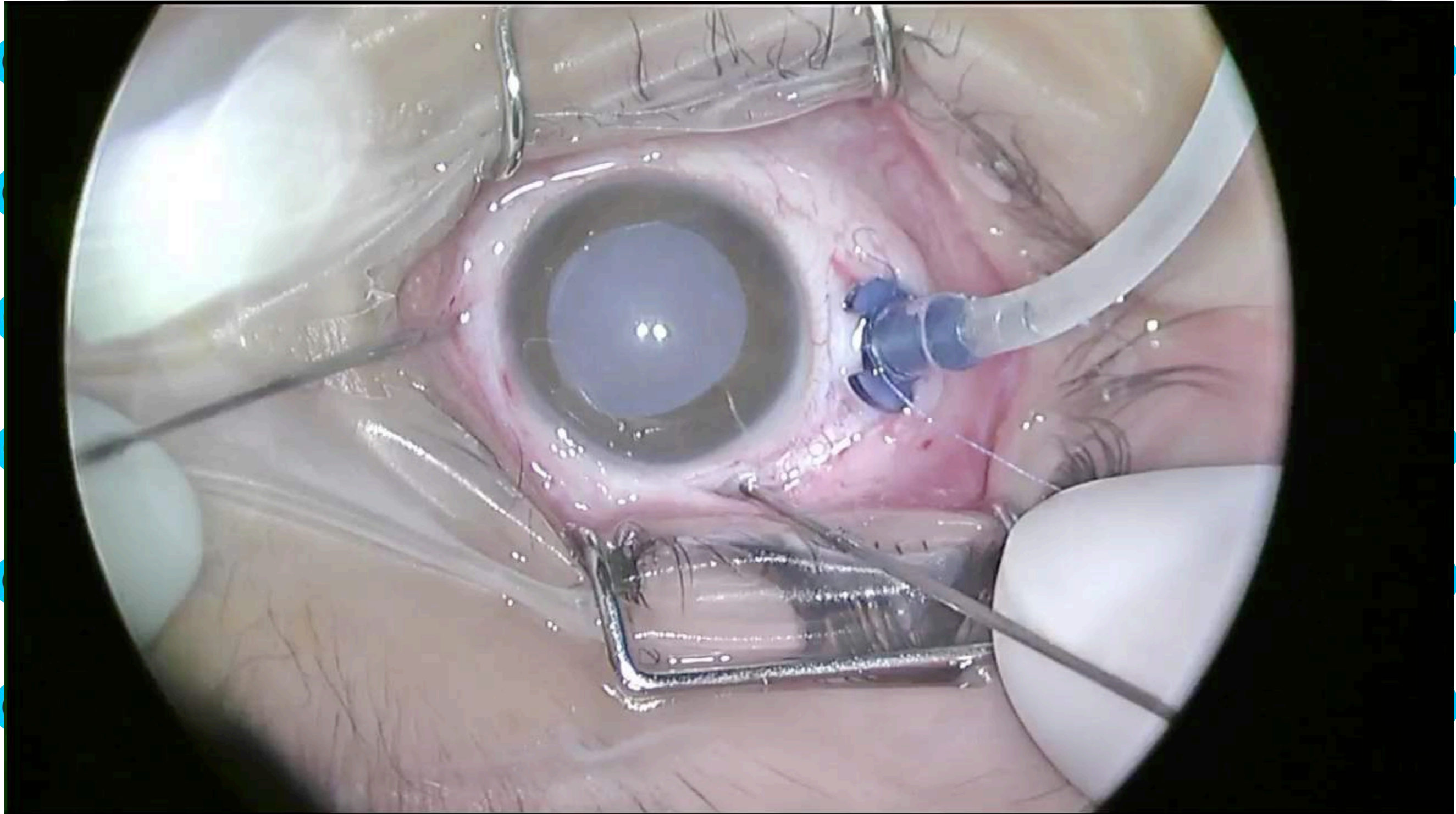


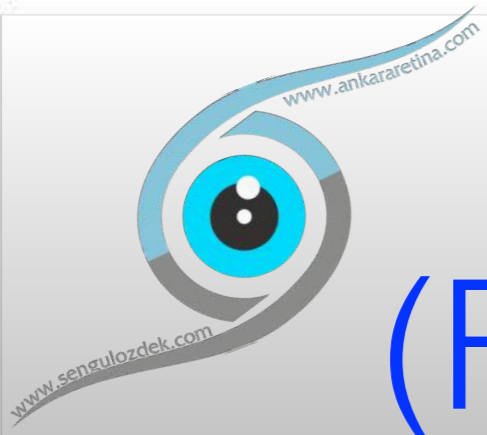
# 23G-with cannula Stage 4a ROP

Prof. Dr. Şengül Özdek

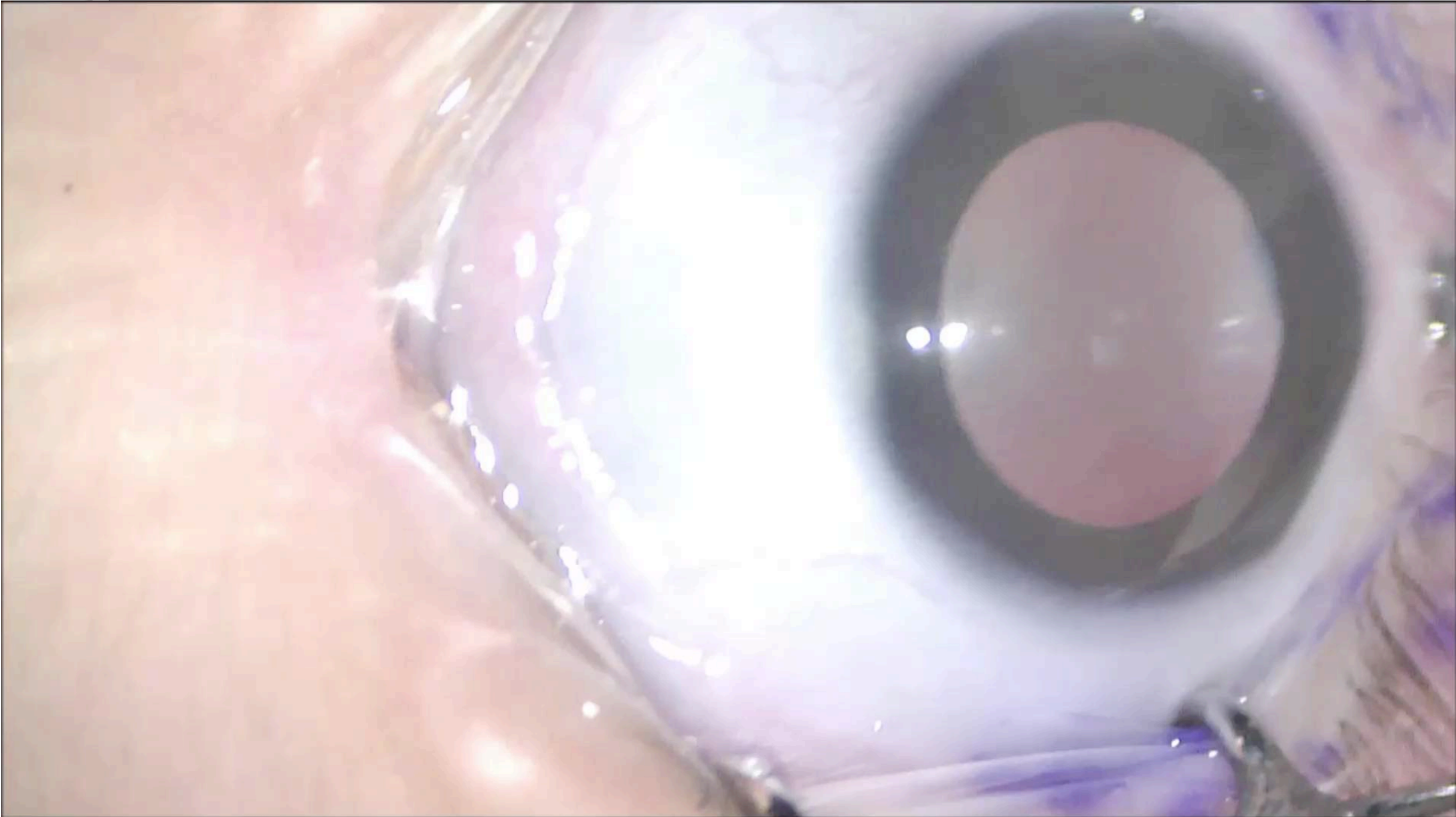


# Stage 4A-without trockars





# Stage 4B (Fellow eye stage 5)

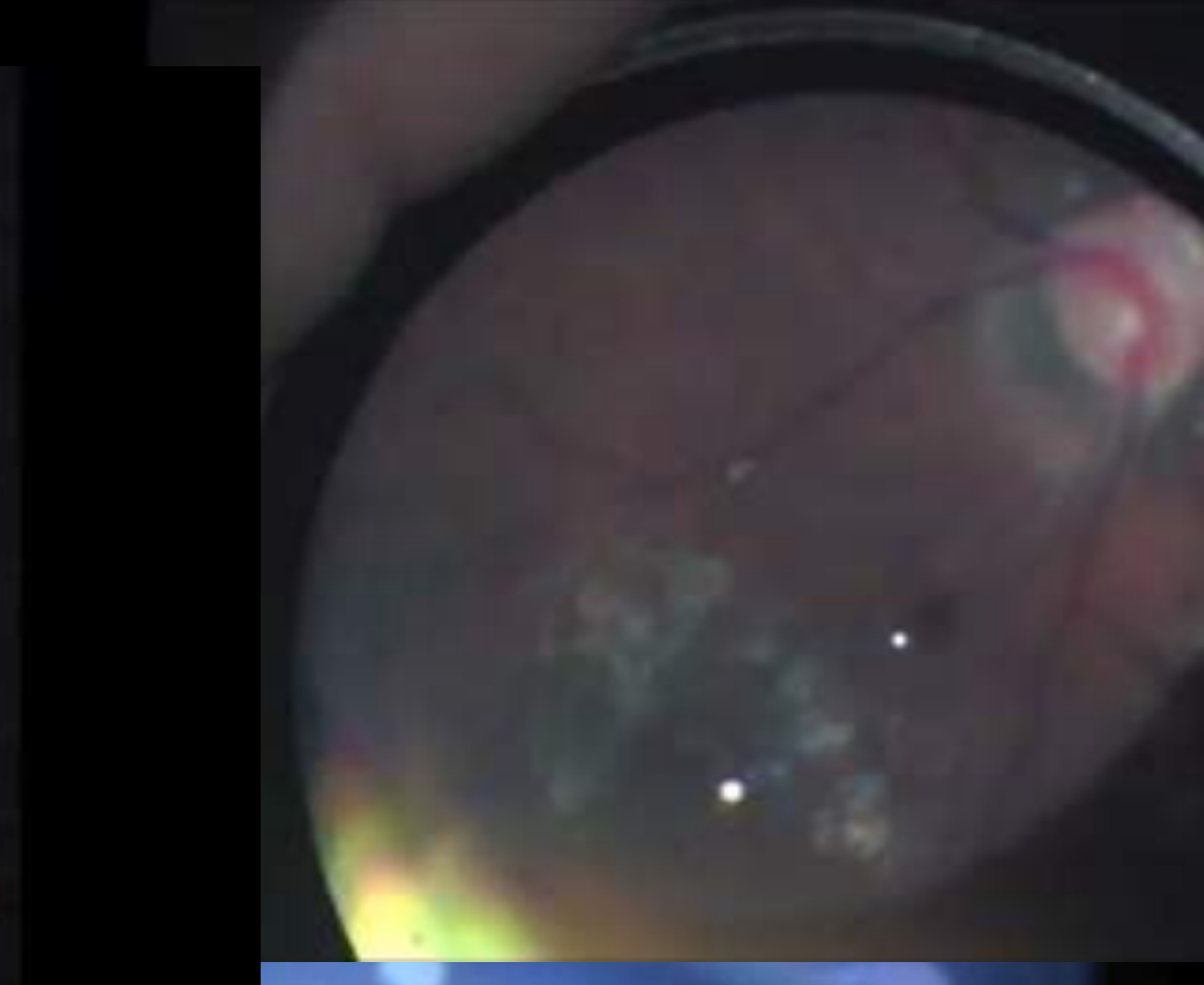
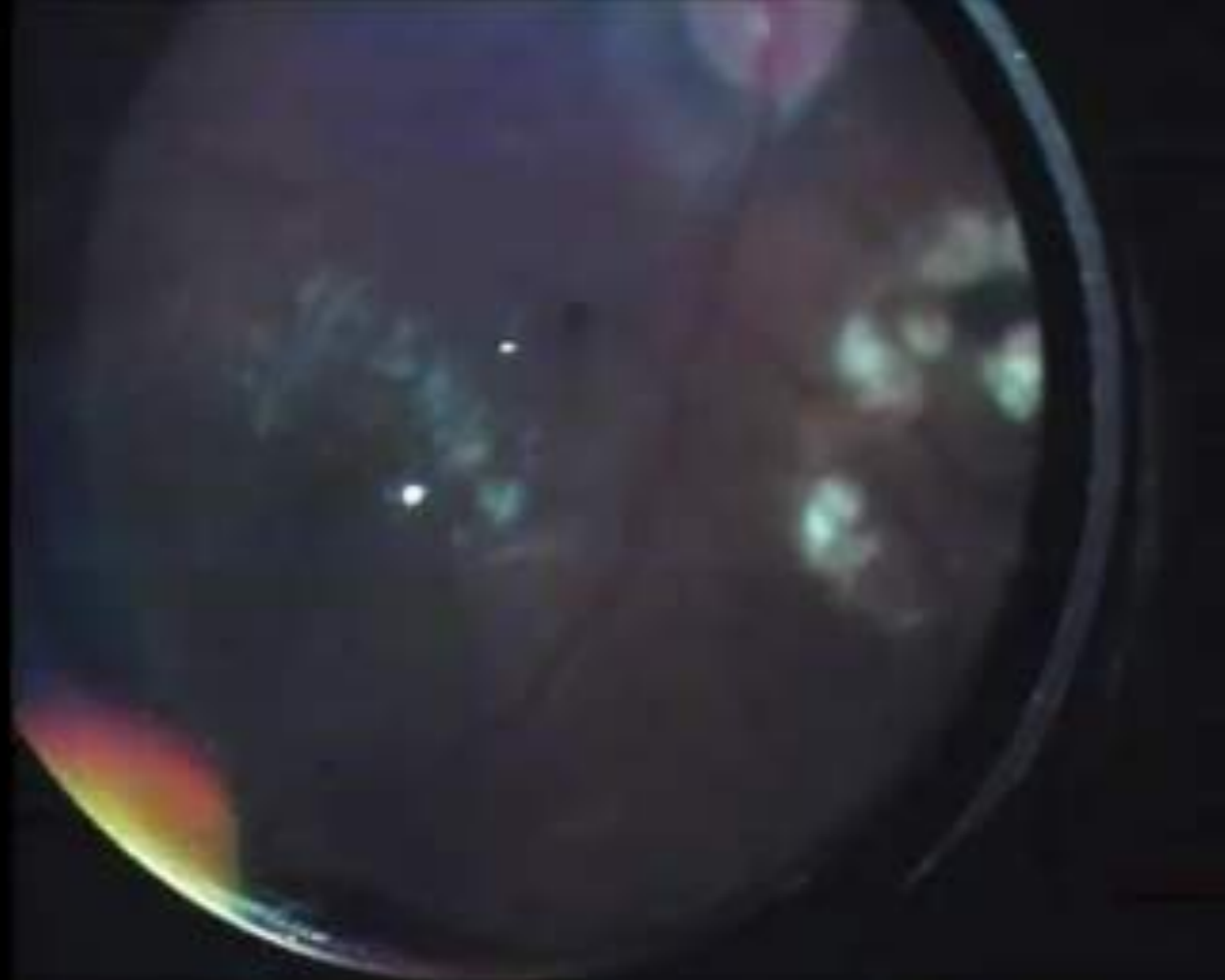


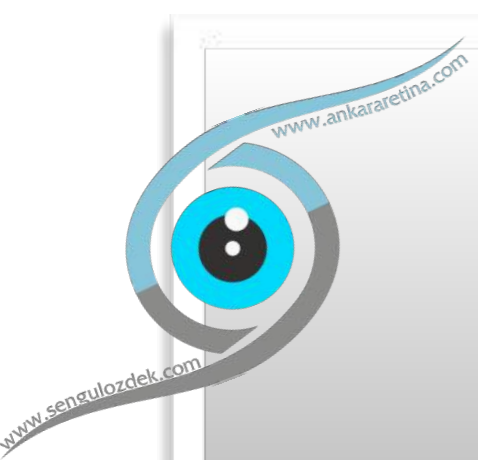




# ROP Surgery

- This is the most typical surgery where the surgeon should know where to stop!
- “Perfect” is the enemy of “good”
- Do not let any iatrogenic retinal break!





# If a retinal break occurs...

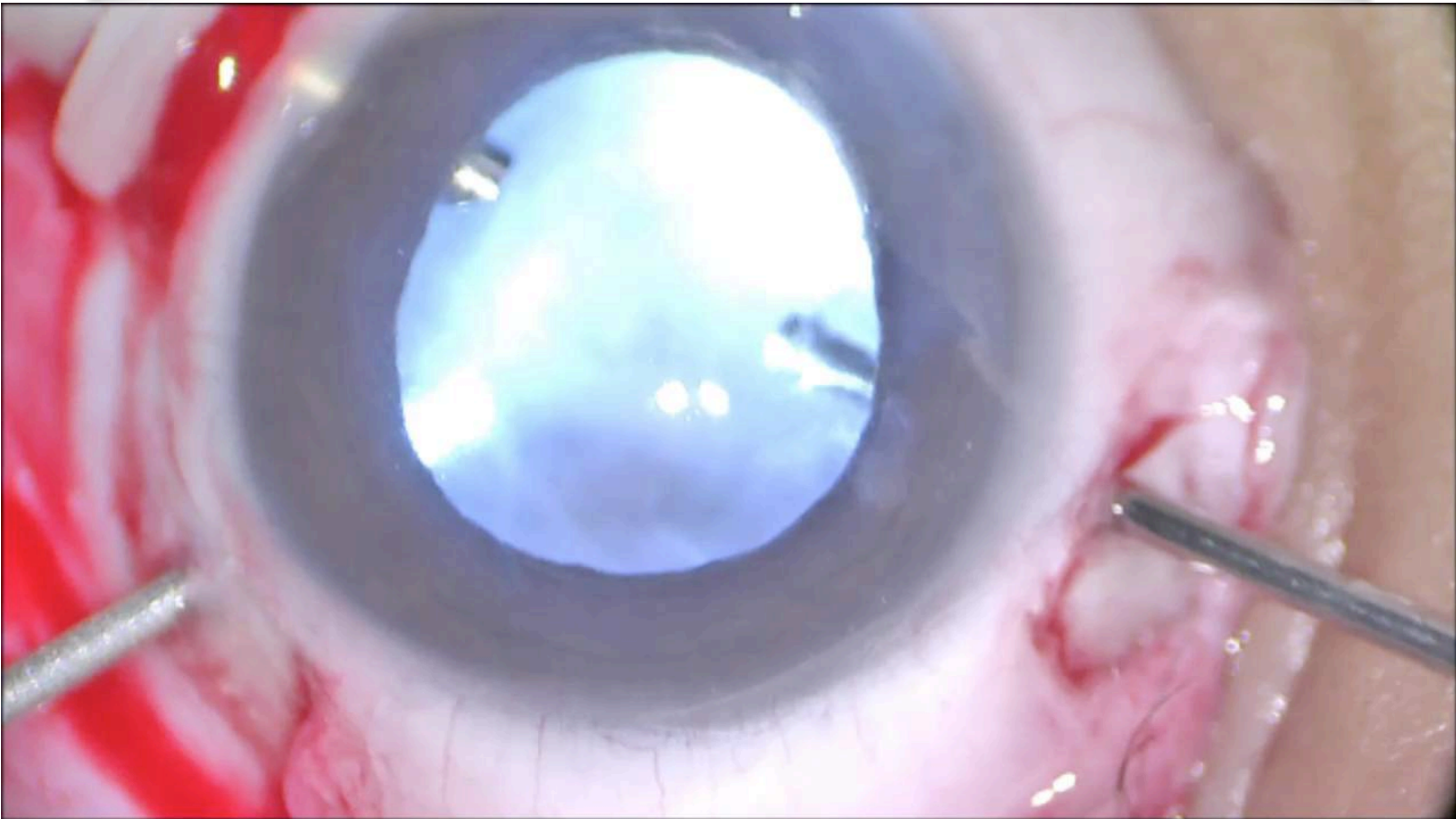
- In a silent area... continue with standard techniques
- In the most proliferative area... abandon the surgery



# SURGERY FOR STAGE 5

- SURGERY?
- VERY LOW VISUAL EXPECTATIONS
- PRESERVATION OF LIGHT PERCEPTION/  
AMBULATORY VISION???
- ANATOMICAL PURPOSES
- IF FELLOW EYE IS GOOD.....MOSTLY NOT  
OPERATED

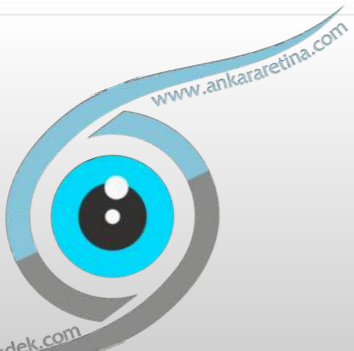
# Stage 5



# SURGERY FOR ROP

To decrease risk of post op hemorrhage

- Preop avastin to eyes with plus disease?
- Do not let hypotony at the end of surgery.
- F-A-X
- Fill with Viscoelastics?

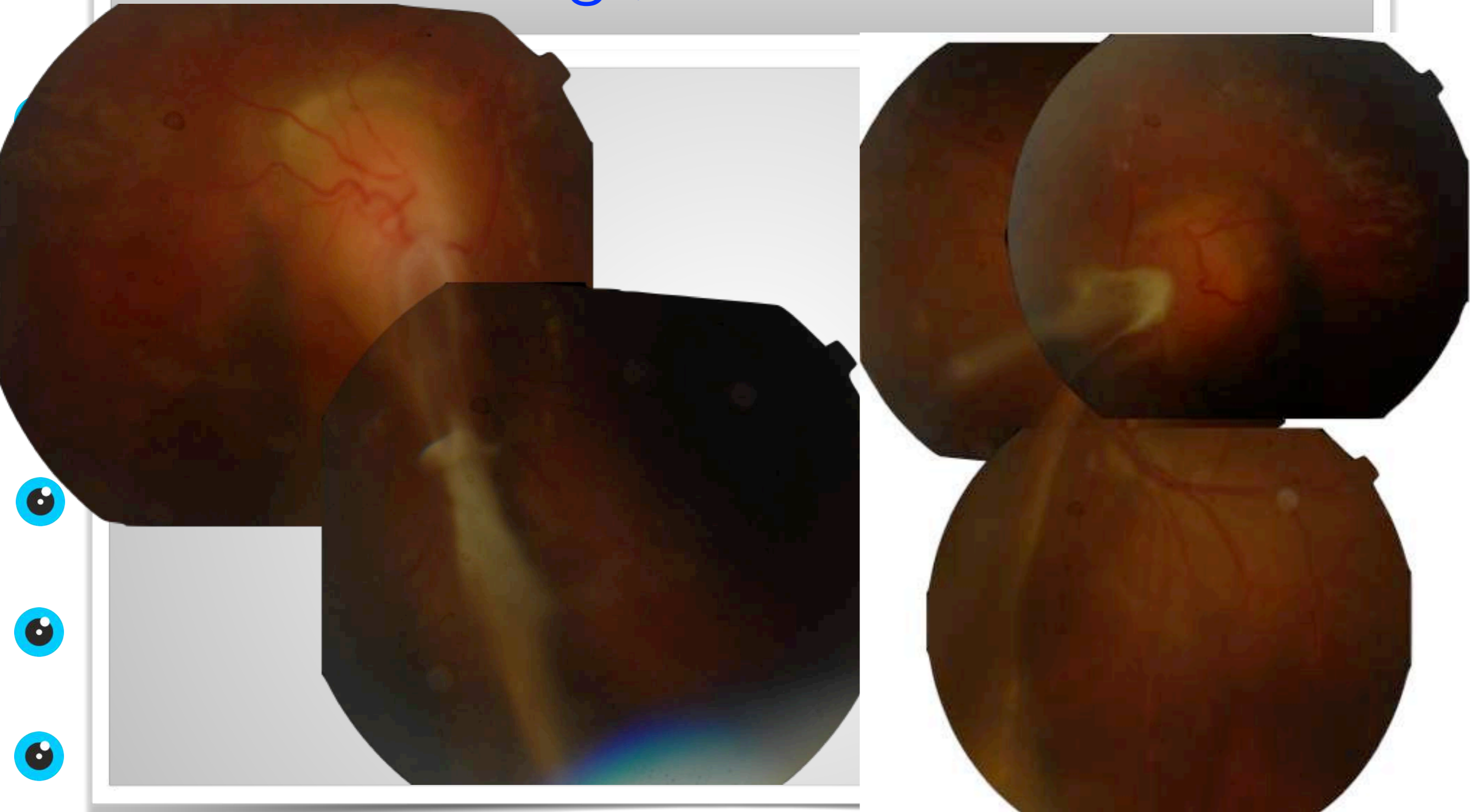


# Surgery for Cicatricial ROP

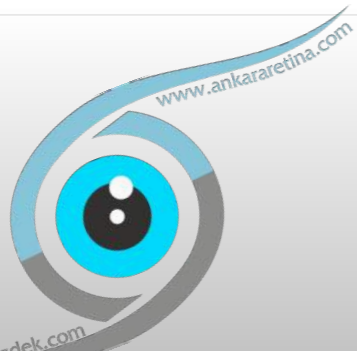
Olgu/Cinsi yet/Göz	Doğum Haftası/Kilosu (hft/gr)	Cerrahi Yaşı (ay)	Preop GK	Preop Tedavi	Cerrahi	Komplikasyon	Takip Süresi(ay)	Anatomik Başarı	Maküler katlantı Preop/Postop	Postop GK
1/E/Sol	28/1060	6	-	-	PPV+PPL	-	24	Kısmi Başarı	+/-	-
2/K/Sol	25/650	38	IOT(-)	LFK	PPV	-	5	+	-/-	1mps
3/E/Sol	29/1100	59	PPEH	LFK	PPV+PPL	latrojenik delik	1	-	-/-	PPEH
4/K/Sol	30/1370	10	IOT(-)	-	PPV	-	3	Kısmi Başarı	+/+	IOT(+)
5/K/Sol	28/1500	11	IOT(-)	-	PPV+PPL	-	20	+	+/-	0.1
6/E/Sağ	28/850	79	0.2	-	PPV	-	40	+	+/-	0.7
6/E/Sol	28/850	81	0.05	-	PPV	-	40	+	+/-	0.3
7/K/Sağ	28/1800	4	IOT(-)	-	PPV	-	24	+	+/-	0.05
8/K/Sağ	30/1290	104	1mps	-	PPV	-	6	Kısmi Başarı	+/+	0.05
9/K/Sağ	28/990	2	IOT(-)	LFK+İVB	PPV+PPL	-	19	Kısmi Başarı	-/-	IOT(-)
9/K/Sol	28/990	2	IOT(-)	LFK+İVB	PPV+PPL	Geç Vit Hem/RRD	19	-	-/-	IOT(-)
10/K/Sağ	28/900	213	1mps	-	PPV+PPL	-	12	Kısmi Başarı	+/+	1mps

6,5 y, M, VA:0.1/0.15

28 wk 1300 gr, No treatment before







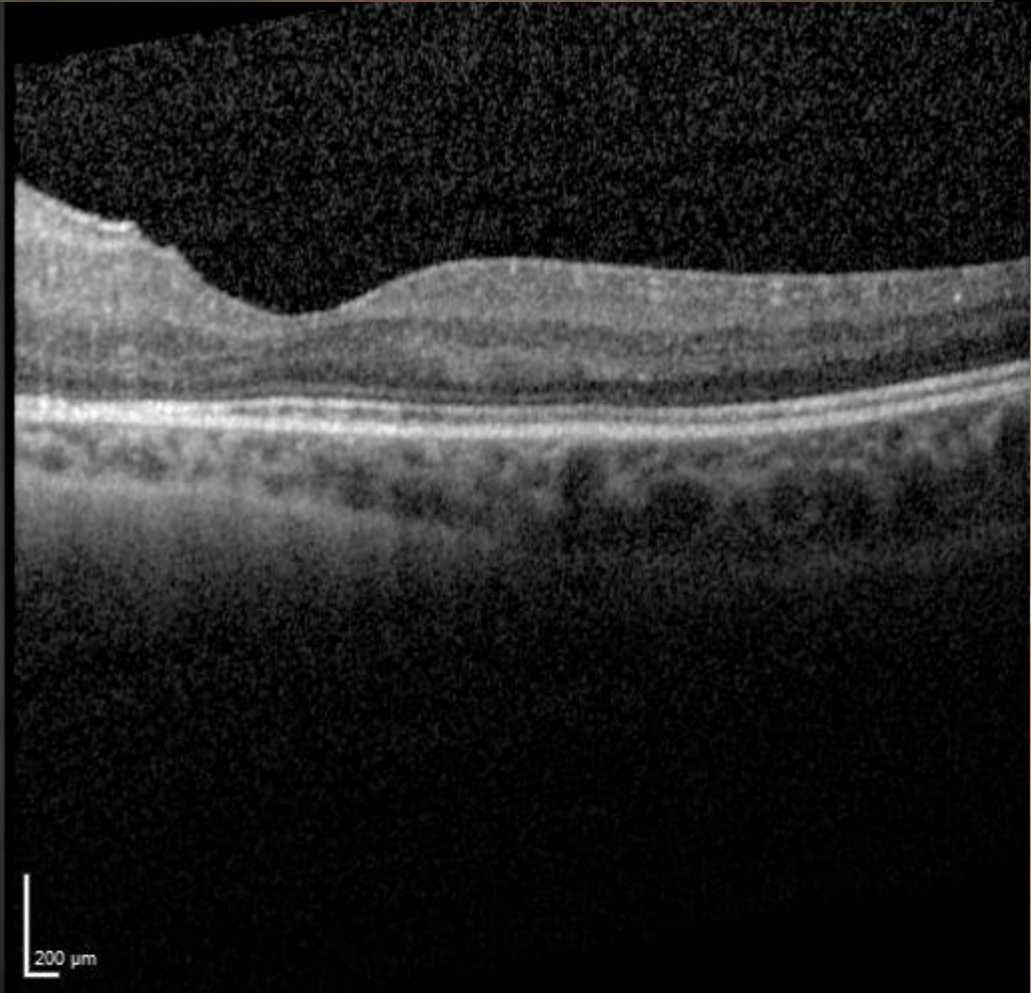
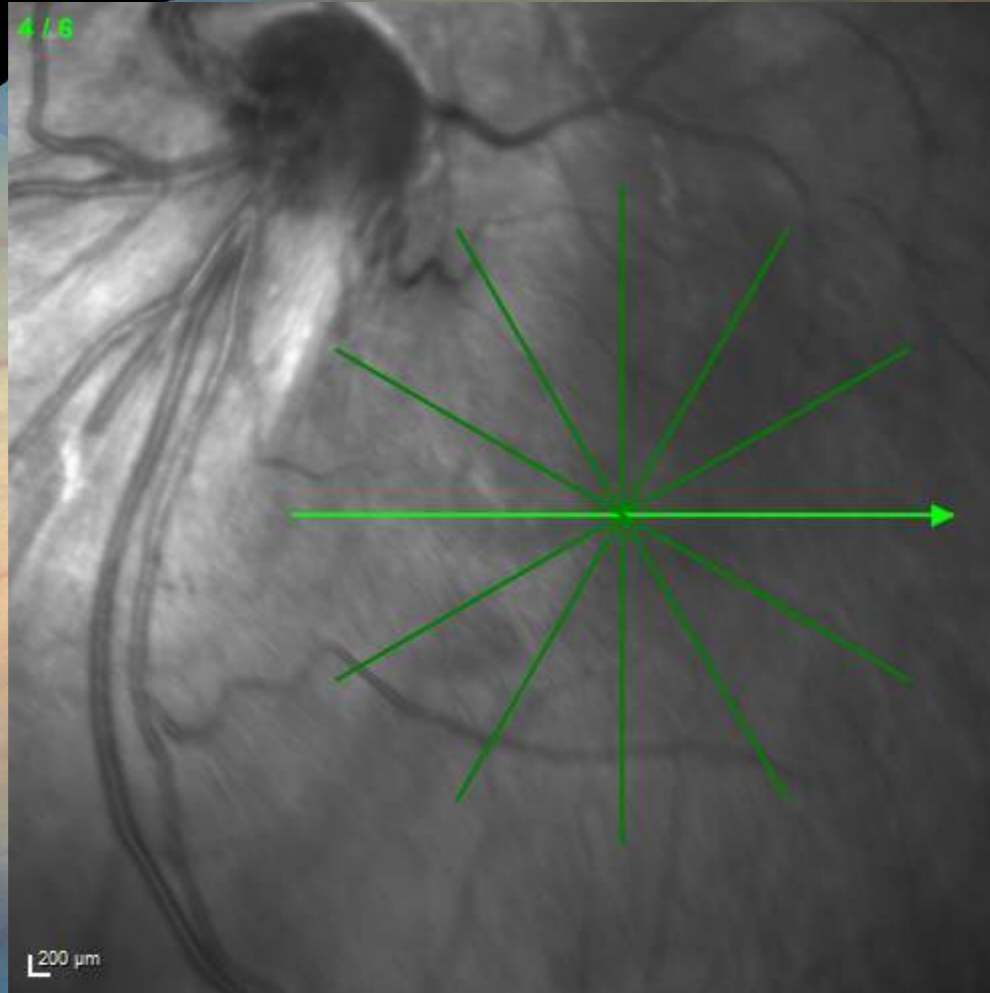
[www.ankararetina.com](http://www.ankararetina.com)

[www.sengulozdek.com](http://www.sengulozdek.com)

# Cicatricial ROP



Postop 3rd year  
VA: 0.7

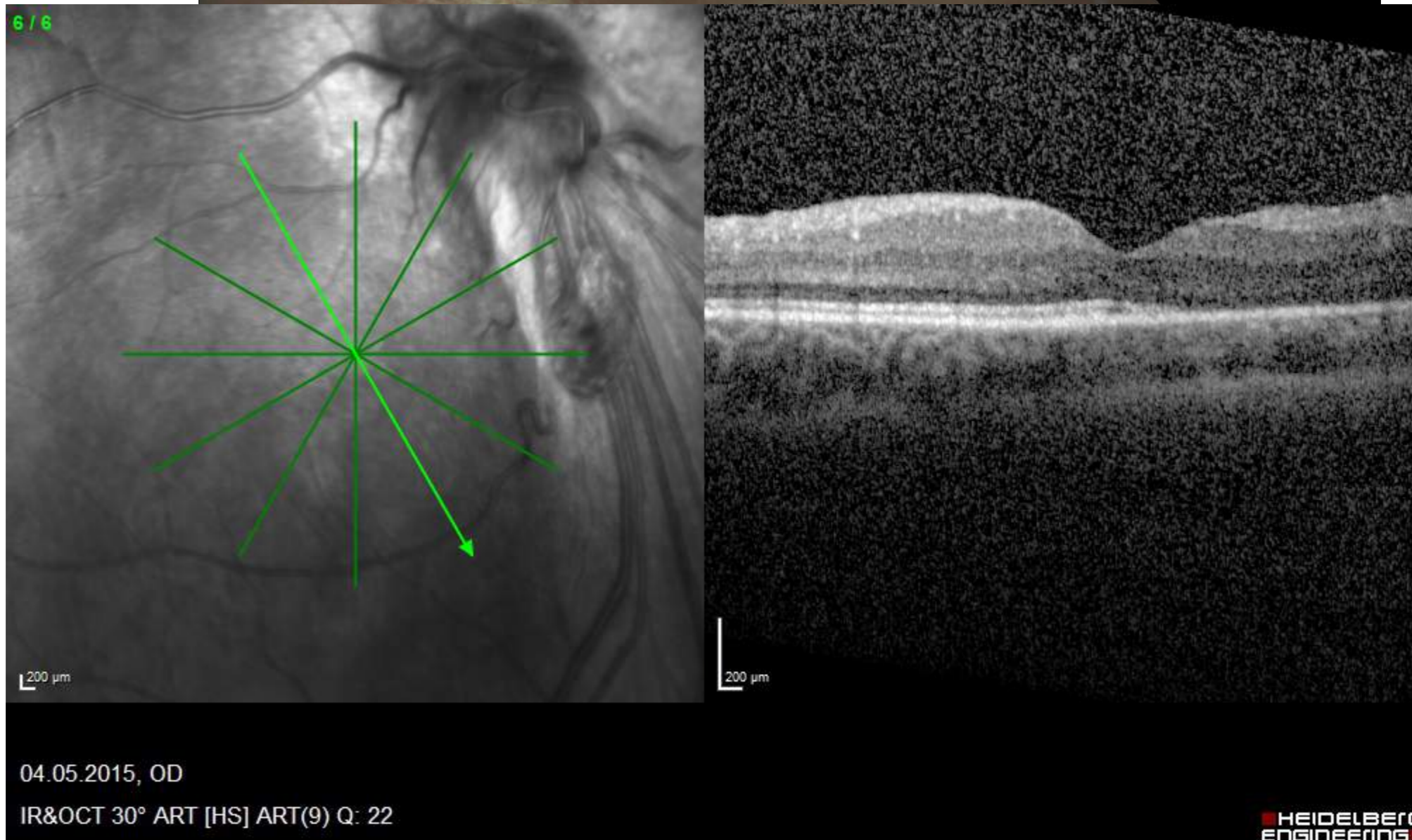


04.05.2015, OS  
IR&OCT 30° ART [HS] ART(10) Q: 25

HEIDELBERG  
ENGINEERING

GK: 0.7

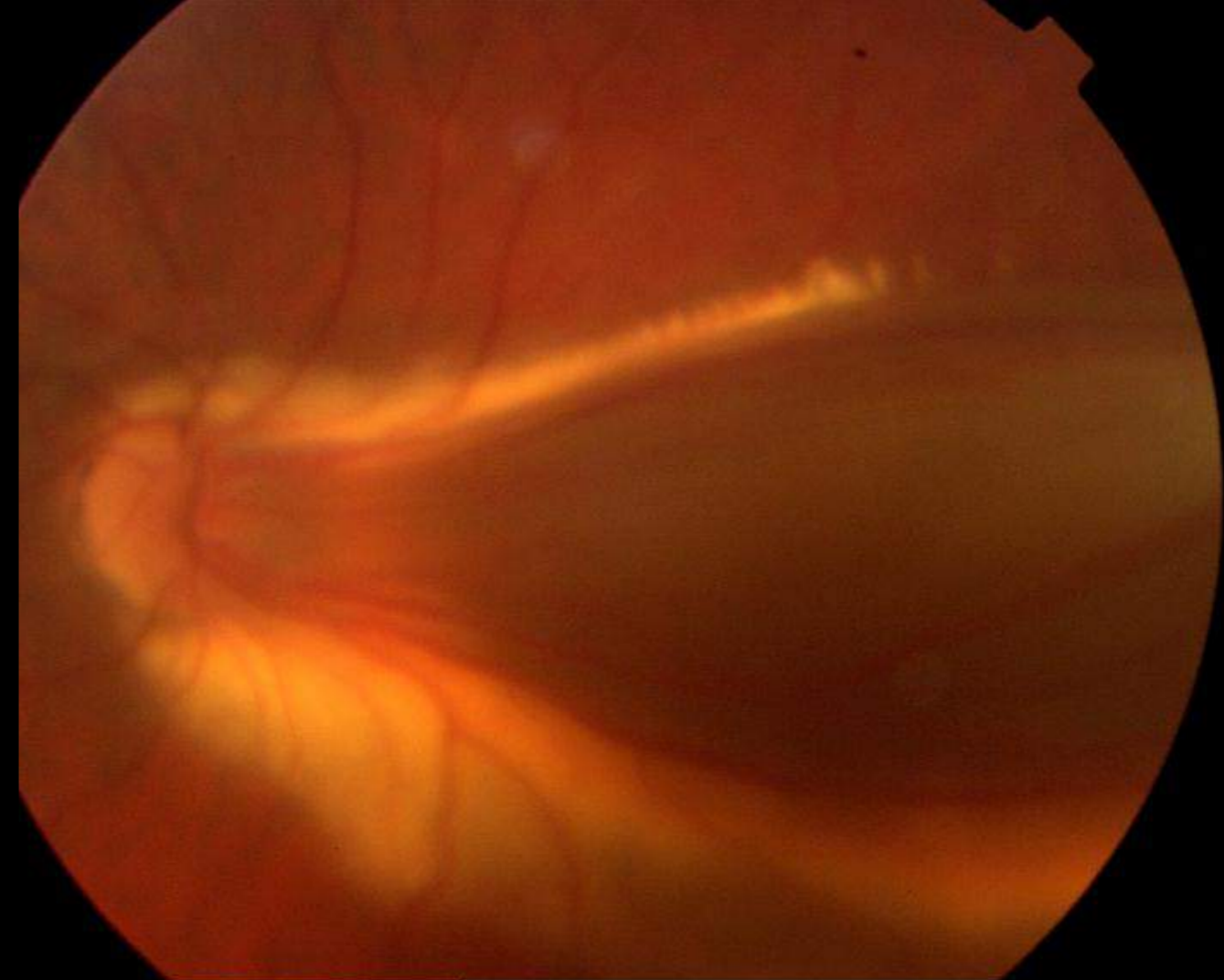
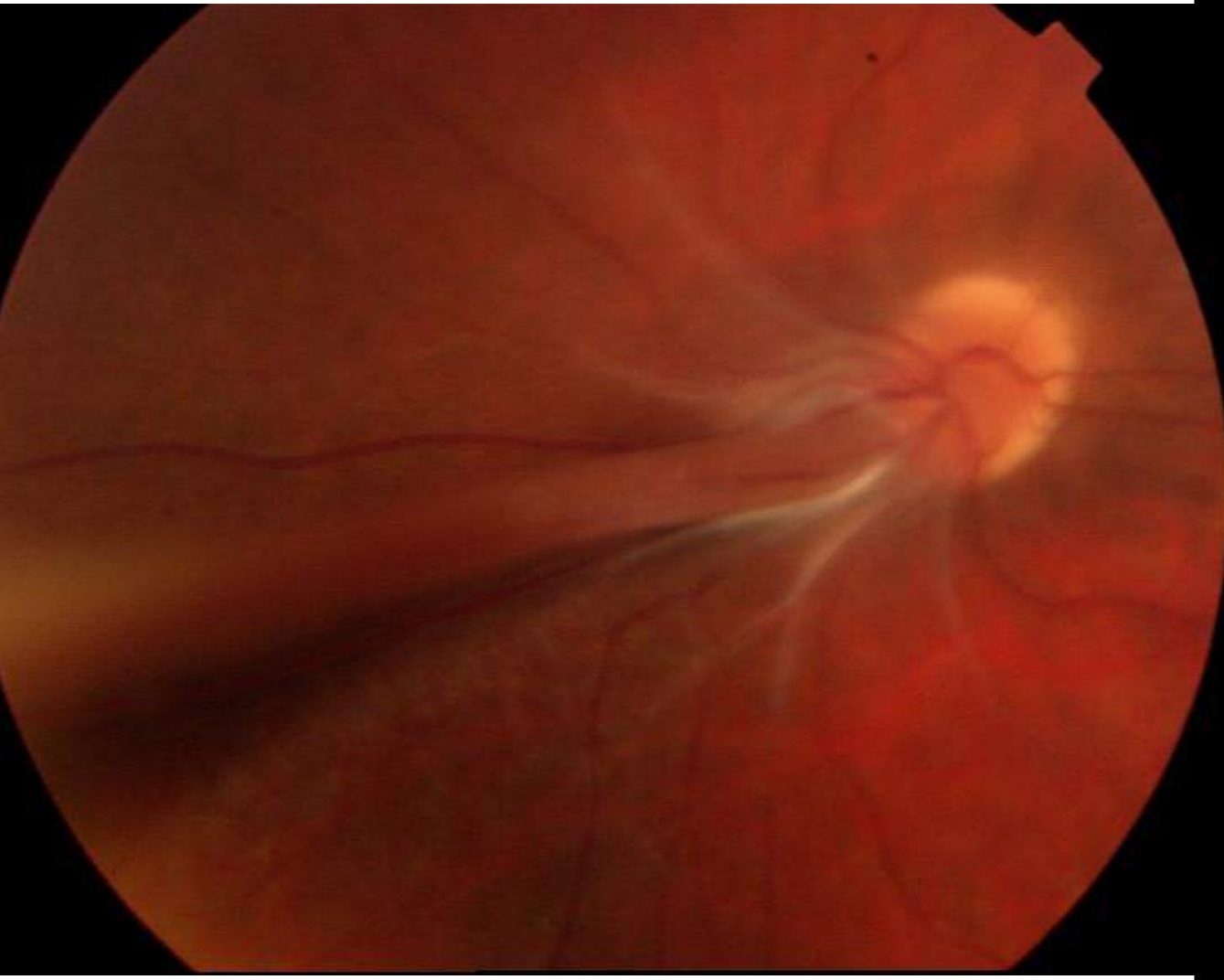
Postop 3rd  
year: VA: 0.3



GK: 0.3

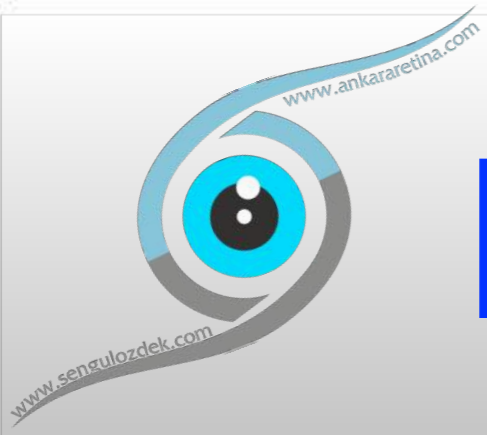
# Cicatricial ROP Surgery

- VRS may lead to very good anatomical and functional results in some of scatricial ROP cases

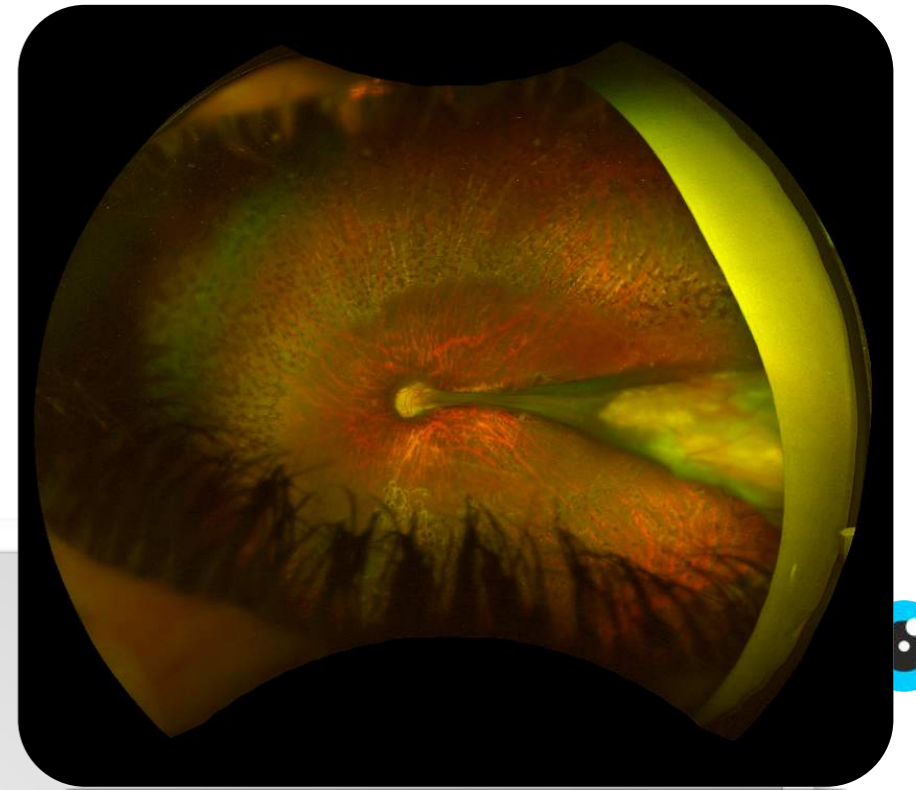


# FEVR





# FEVR



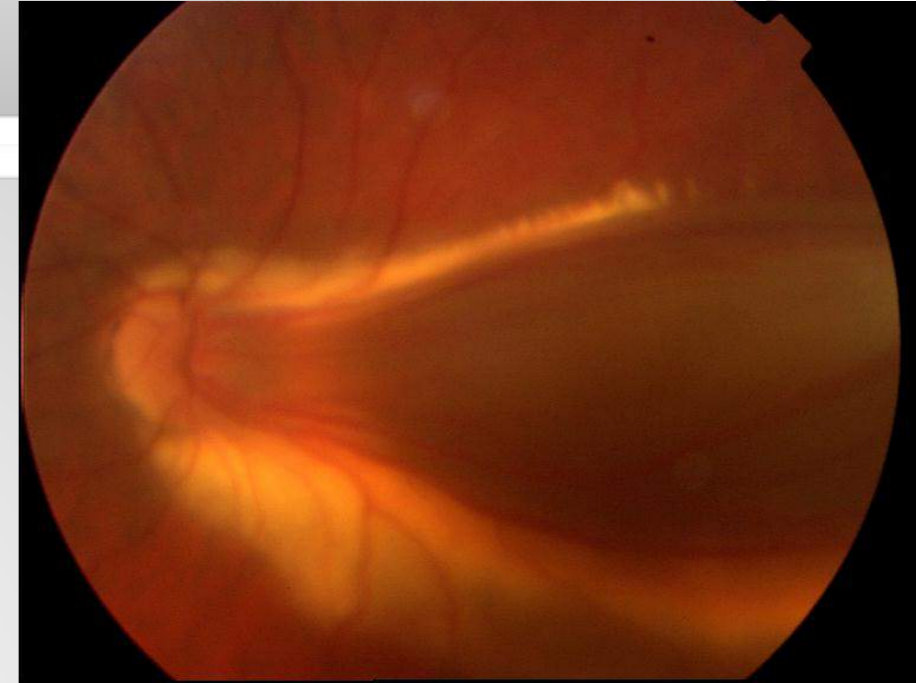
- Mostly **AD inheritance**
- NDP, FZD4, LRP5, TSPAN12, ZNF408: (Wnt-**NORRIN signal pathway**) **defective genes**
- Retinal angiogenesis is defective: Vascular differentiation is insufficient
- Peripheral retinal vascularization is incomplete
- 21- 64% RD: Tractional or exudative



# FEVR-Treatment

- LASER

- STAGE 1: Peripheral avascular retina
- STAGE 2: Retinal NVE



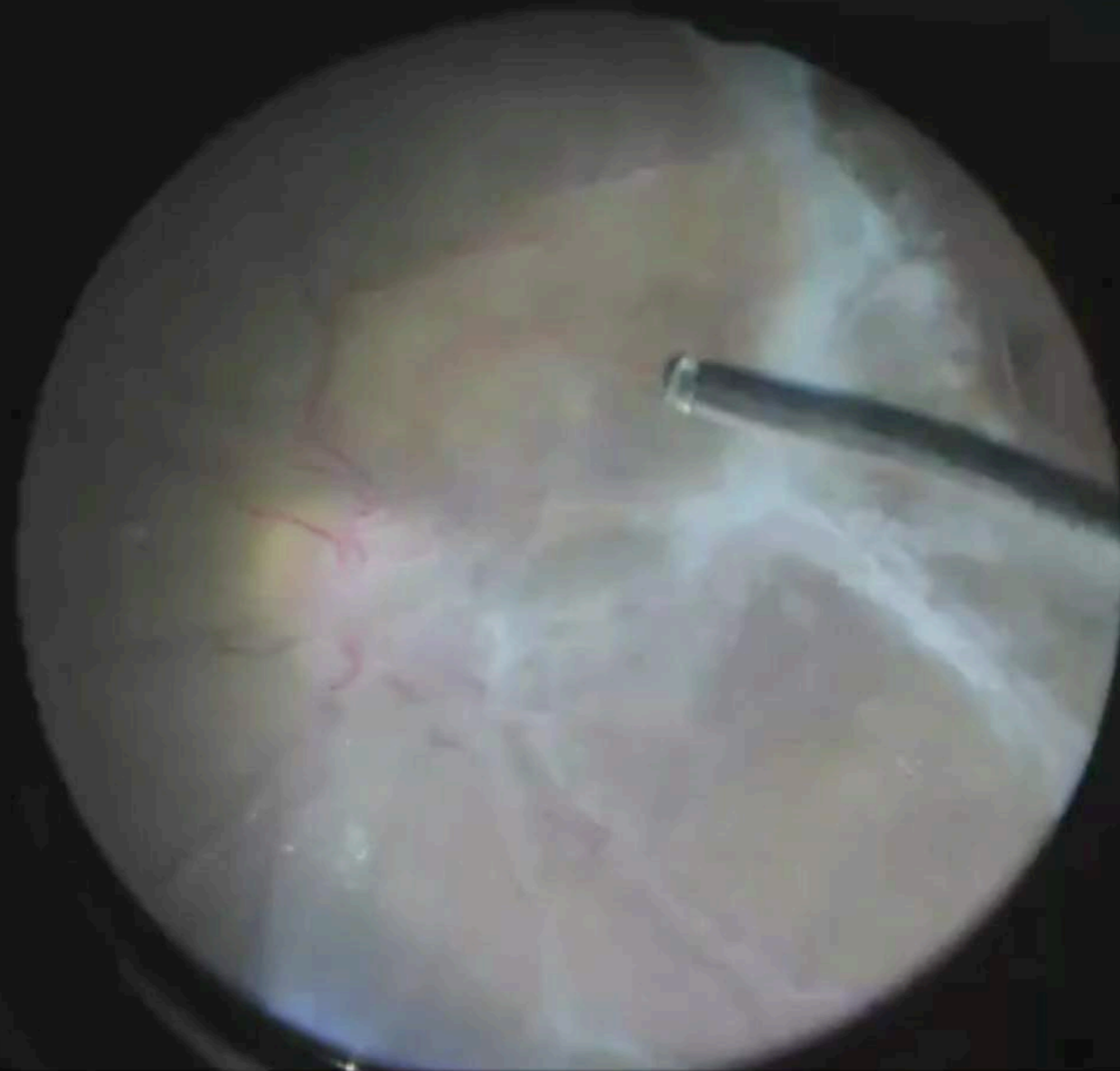
- SURGERY

- STAGE 2: Retinal NVE-Vit Hem
- STAGE 3: Macula on RD (macula ectopic)
- STAGE 4: Macula off RD (macular fold)
- STAGE 5: Total RD



# FEVR-Treatment

6y old, F, bilat TRD, VA: 0.05

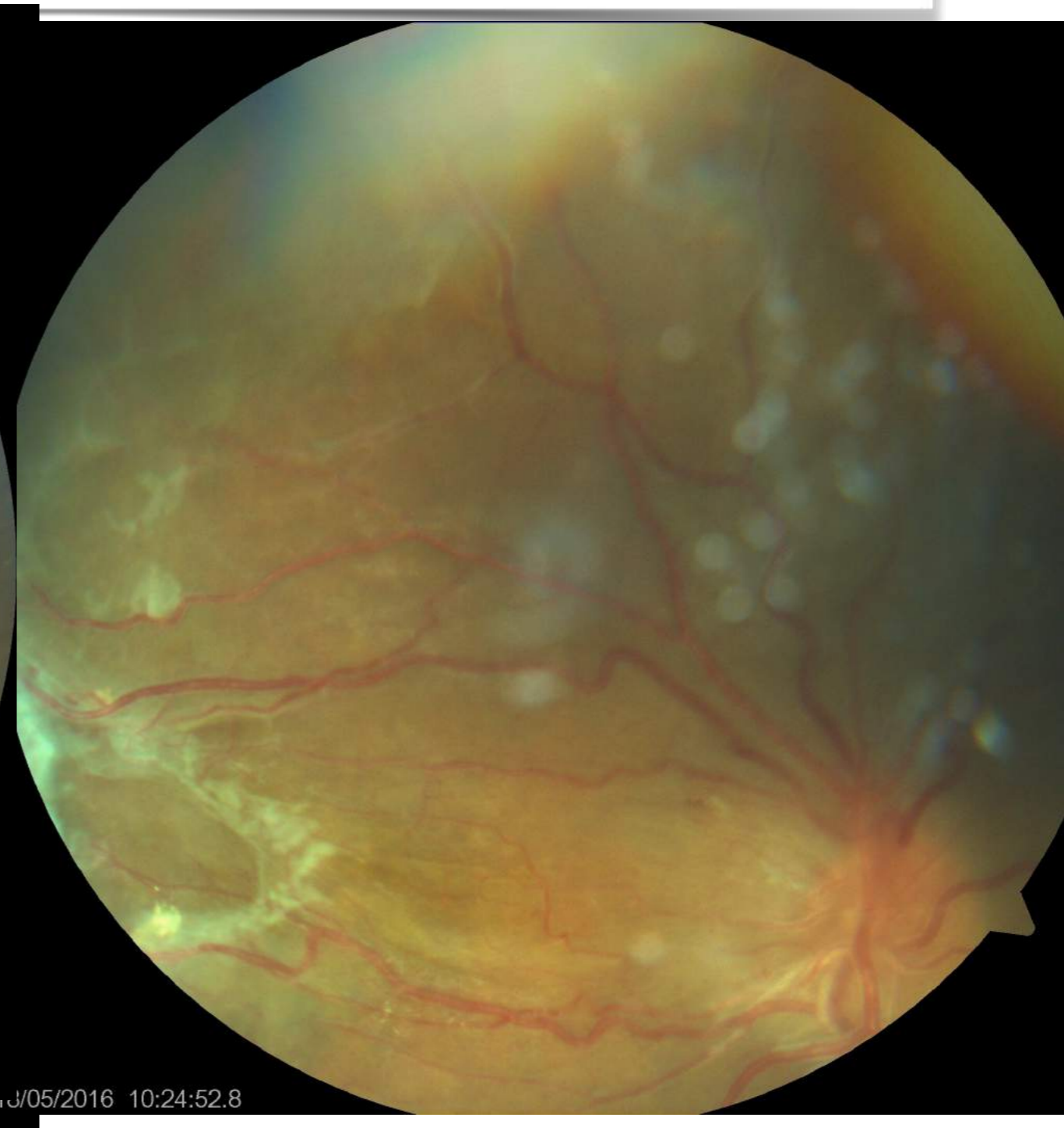
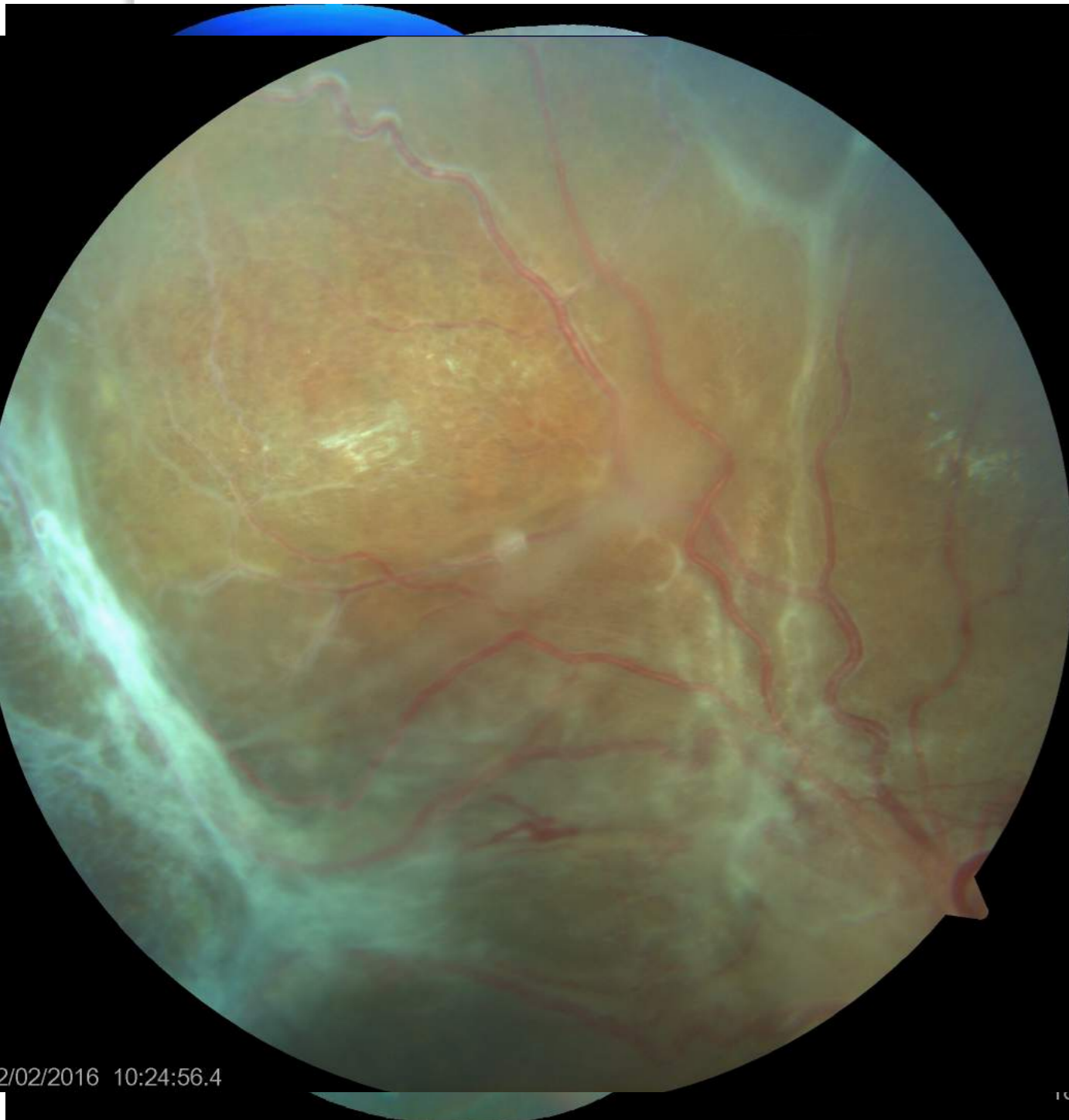






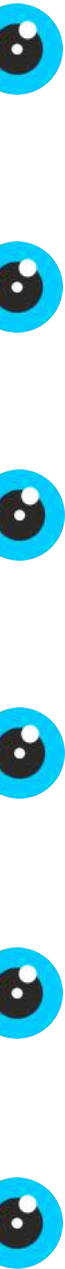
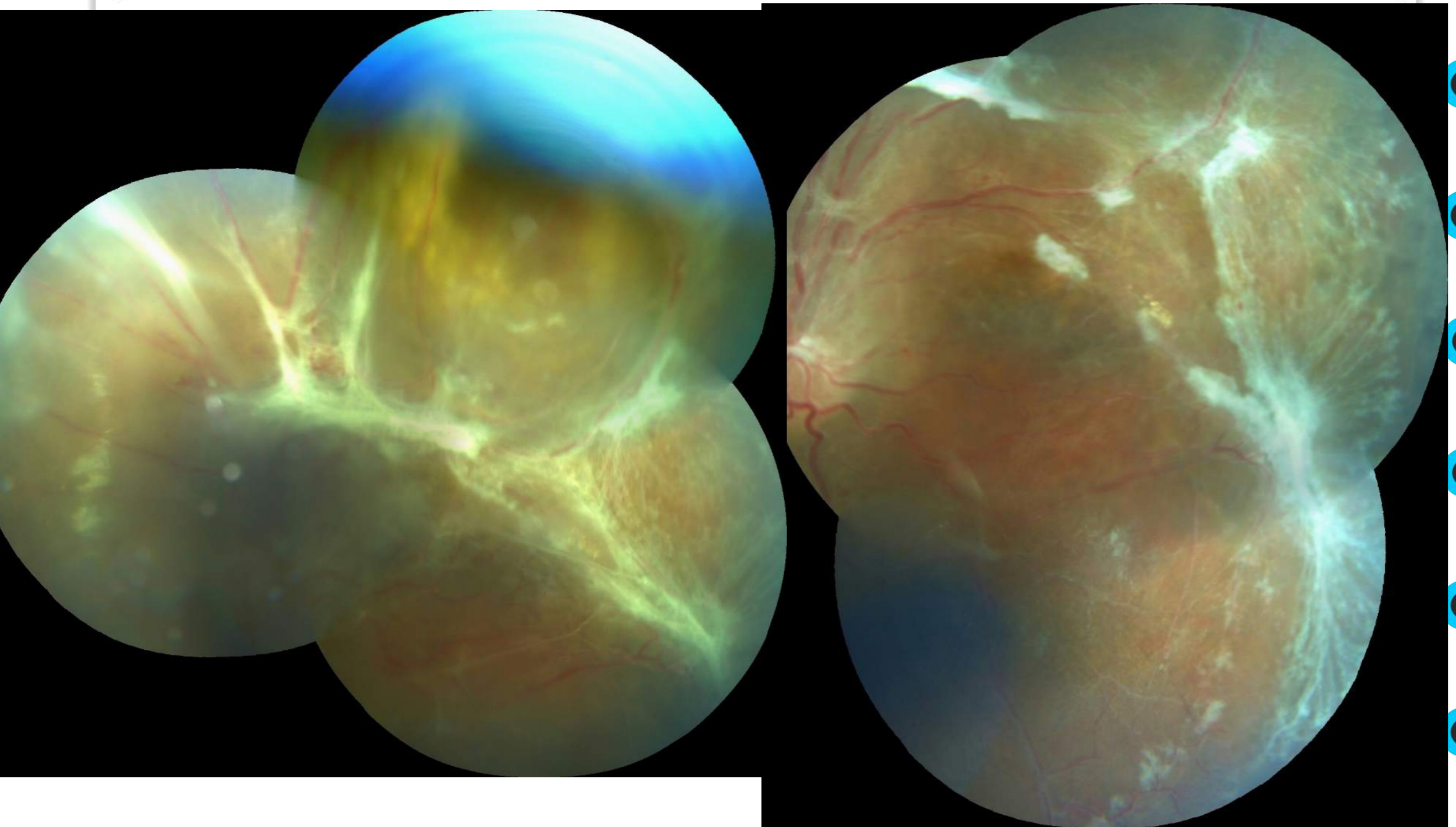
# FEVR-VRS

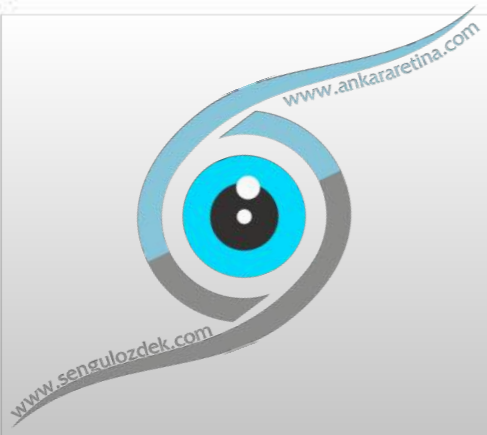
VA: Preop: 0.05 Postop: 0.4



# FEVR-Treatment

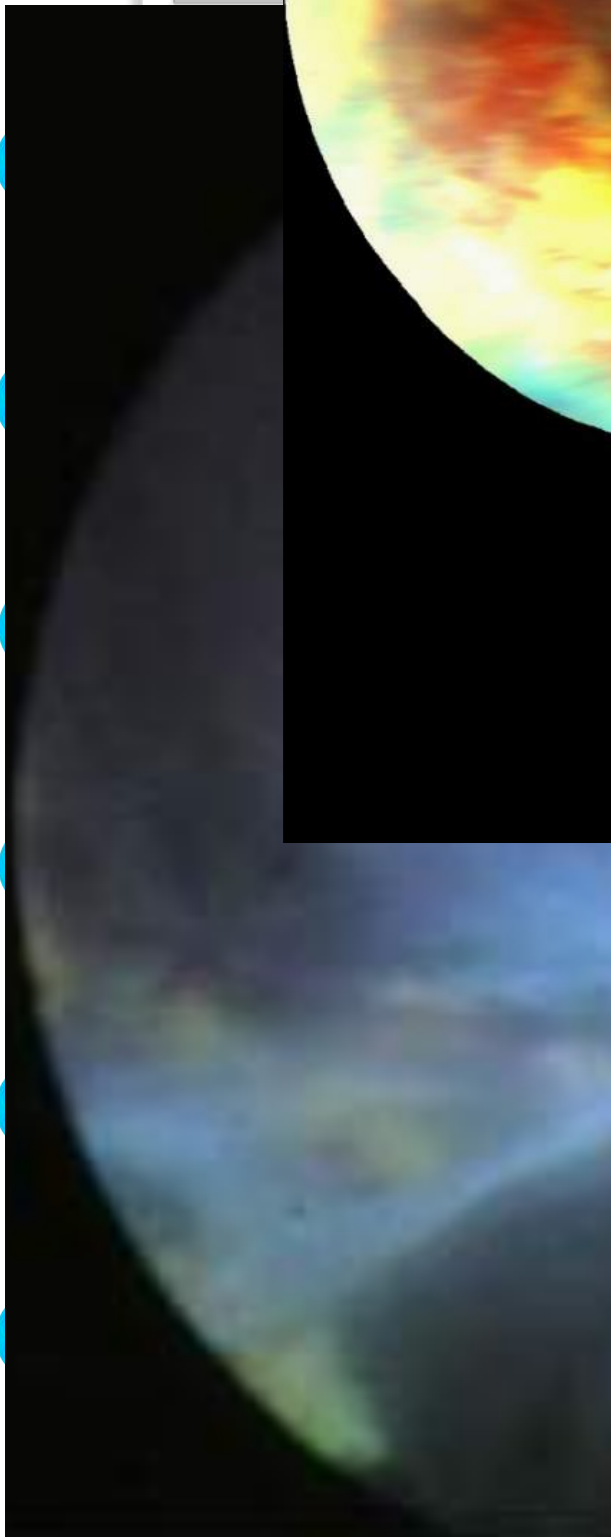
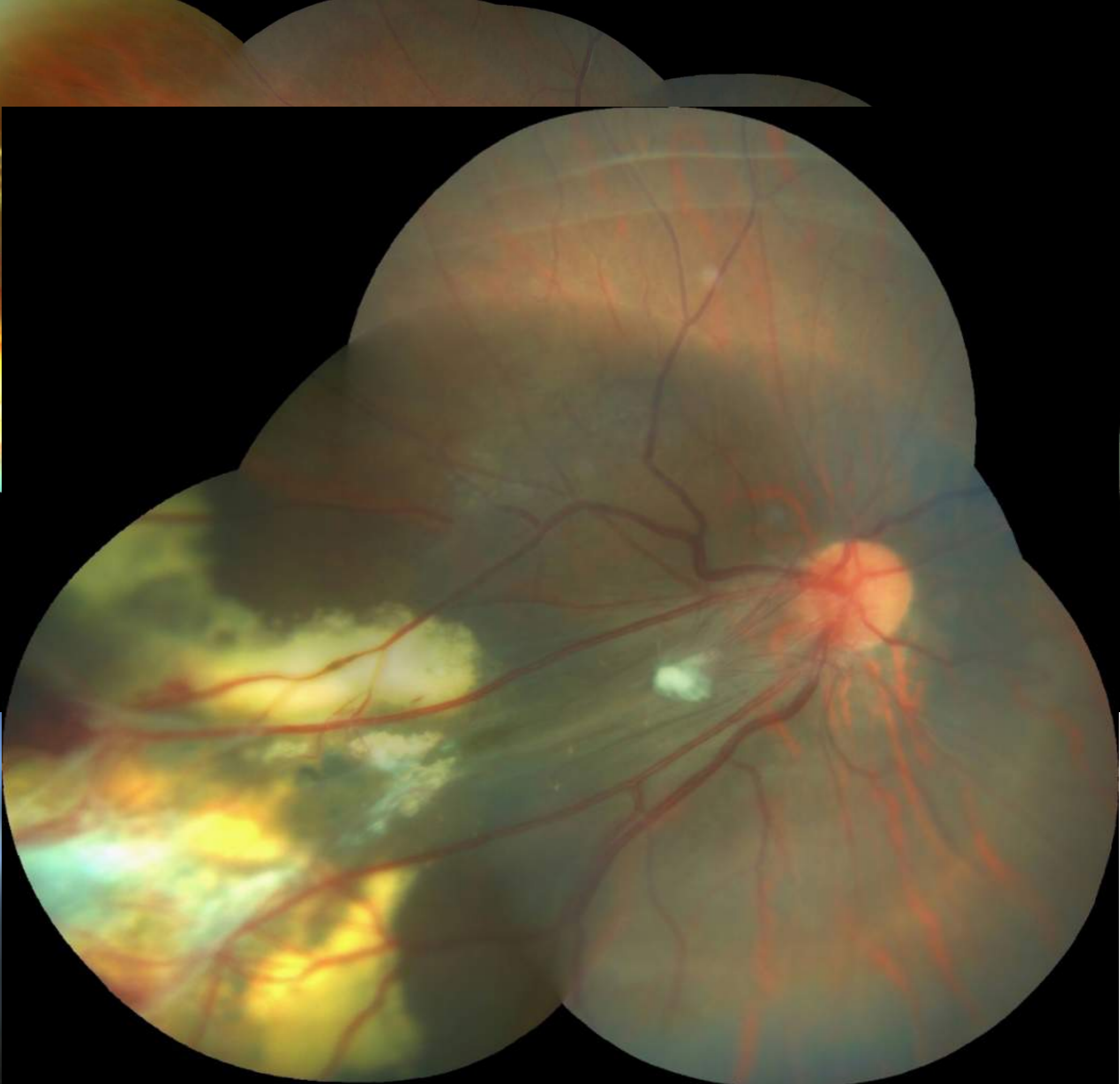
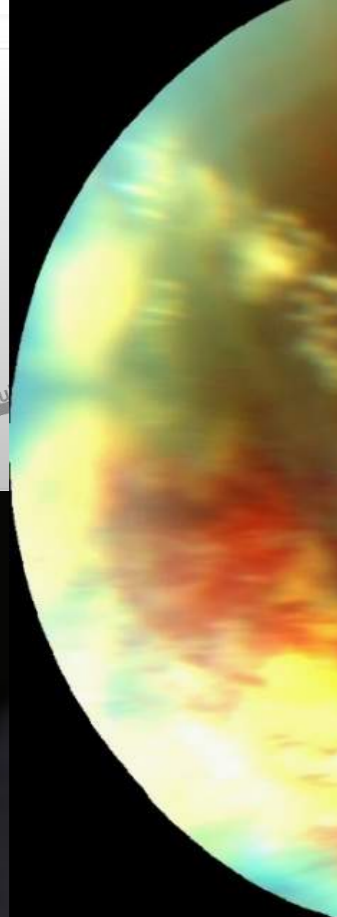
VA: Preop:0.05 Postop: 0.3

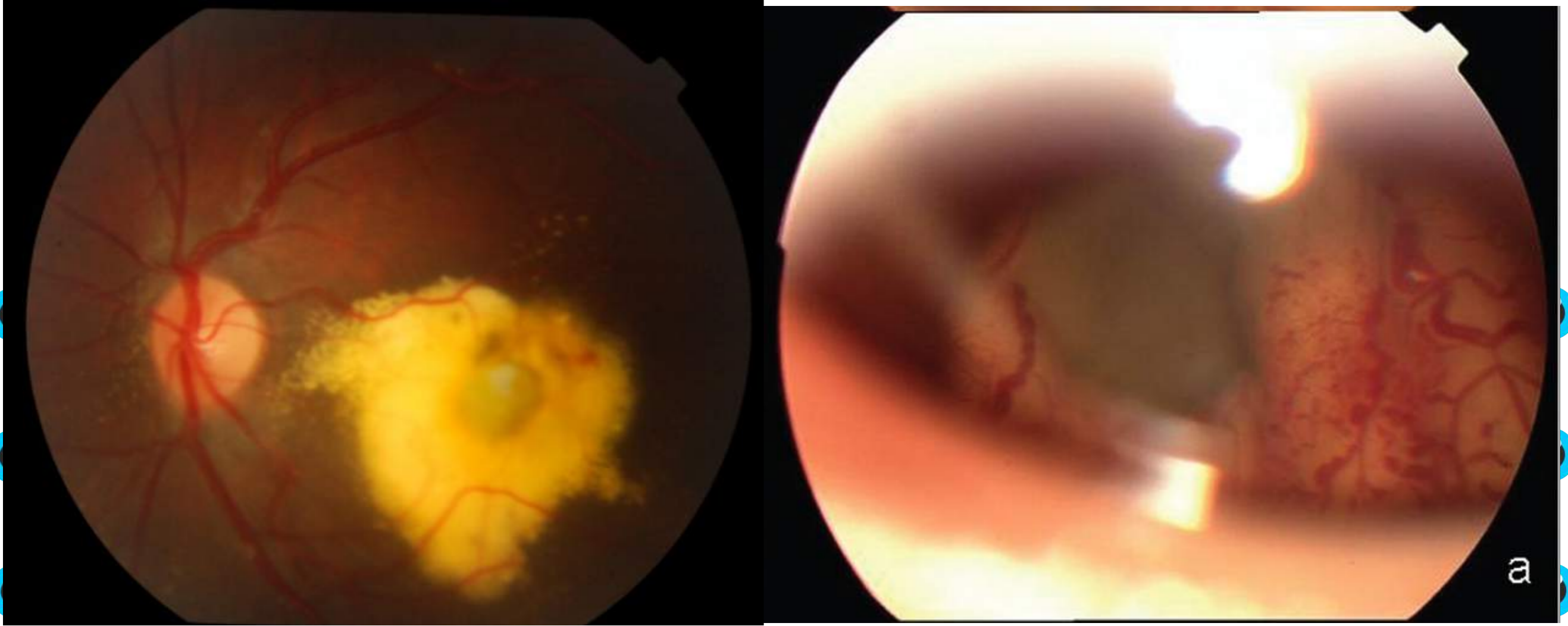




# FEVR VRS





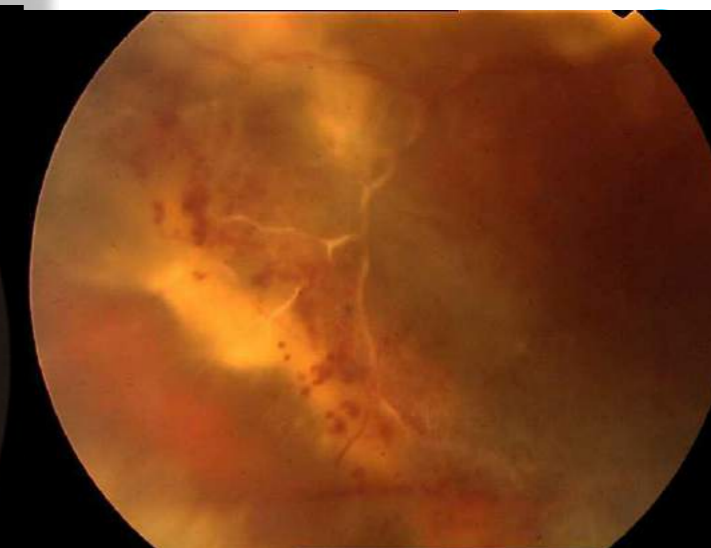
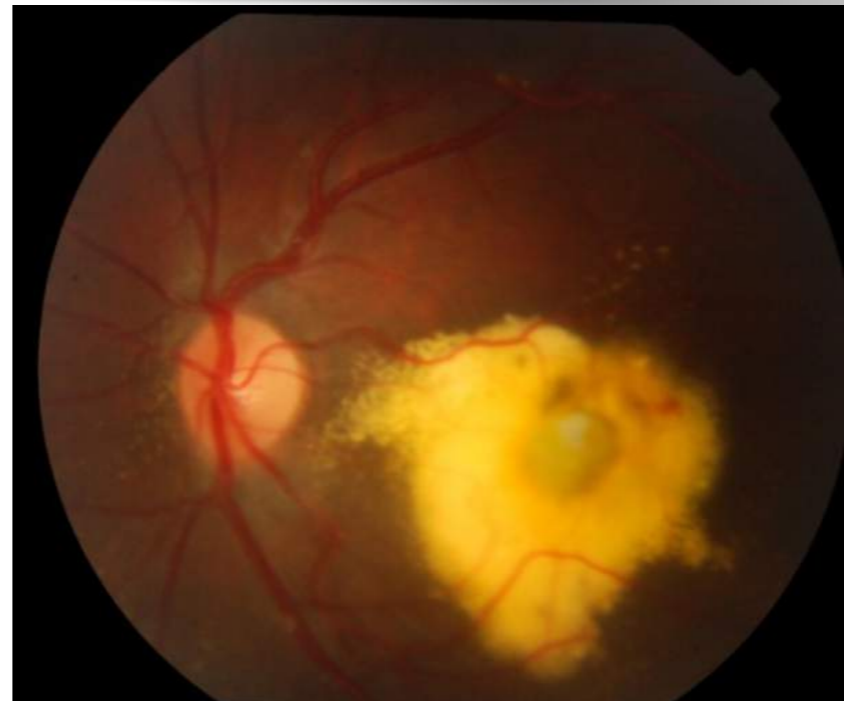
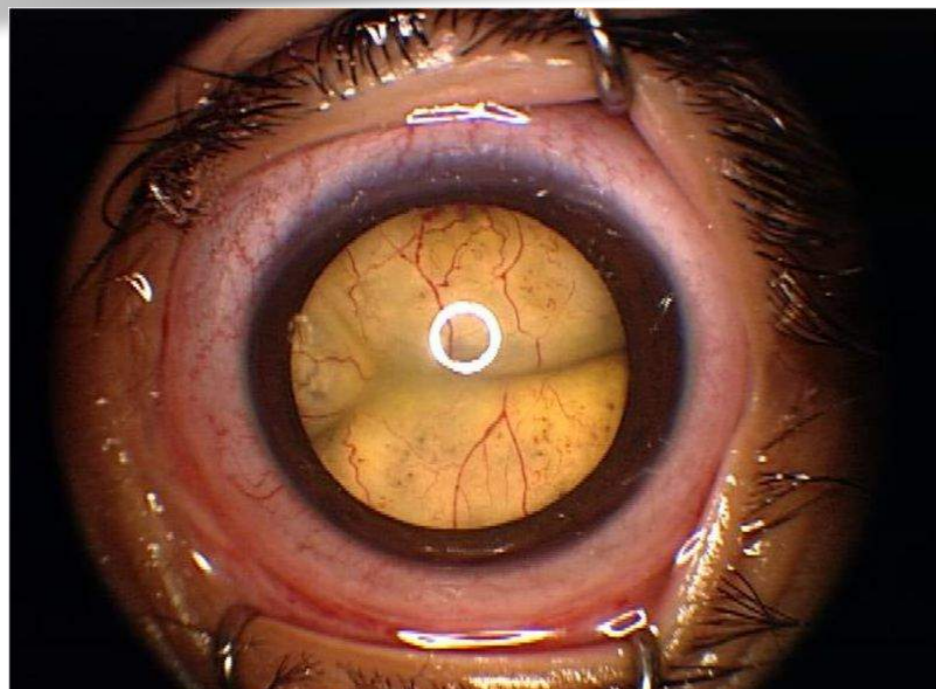
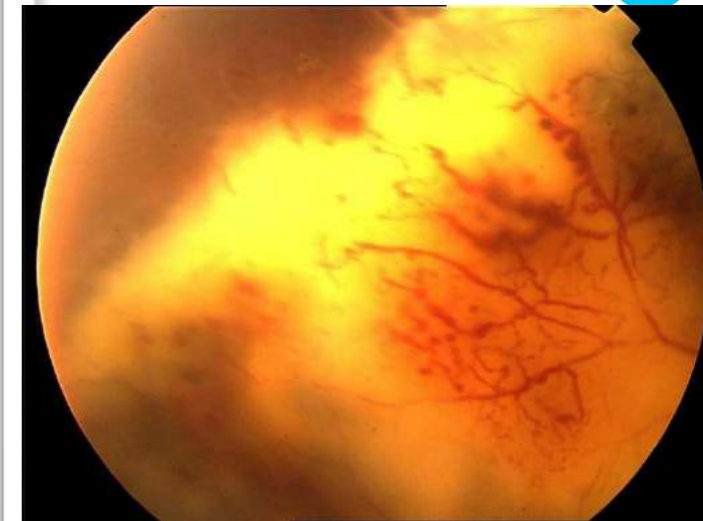


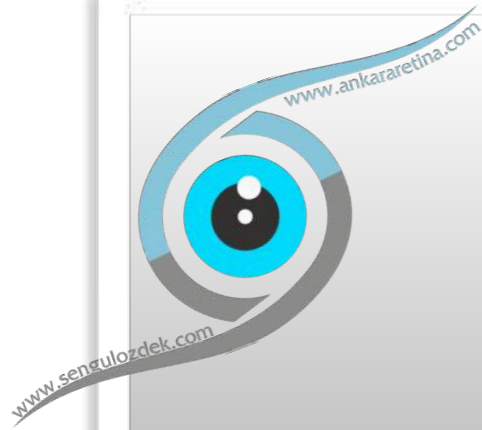
# COATS' DISEASE



# COATS DISEASE

- Endothelium and pericyte disease:  
Inner blood retina barrier defect
- Exudative (Tractional) RD

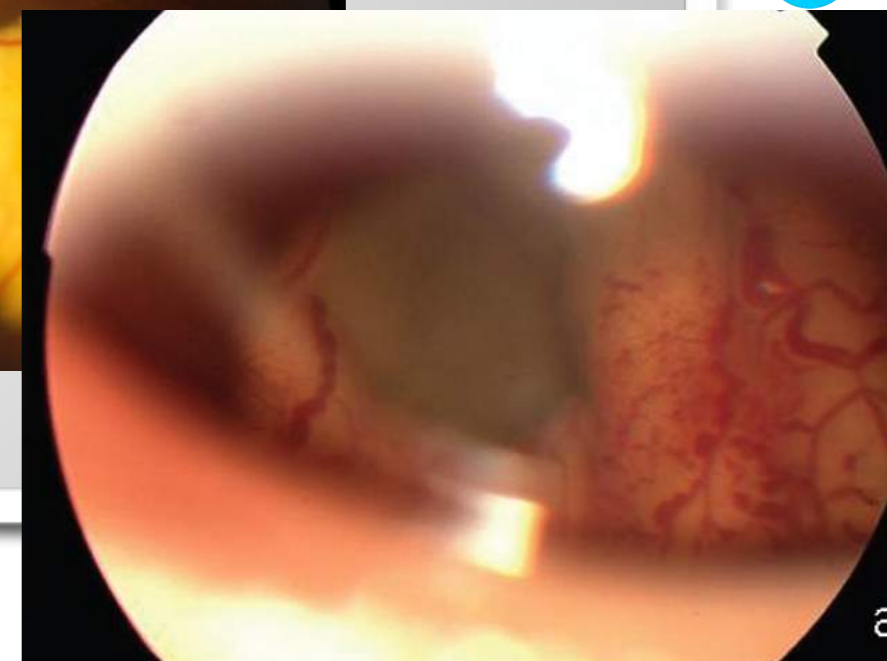
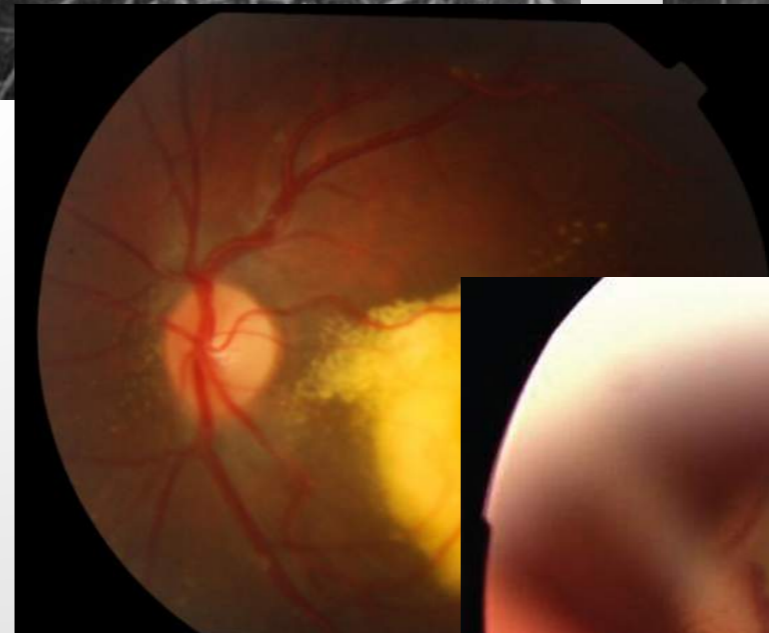
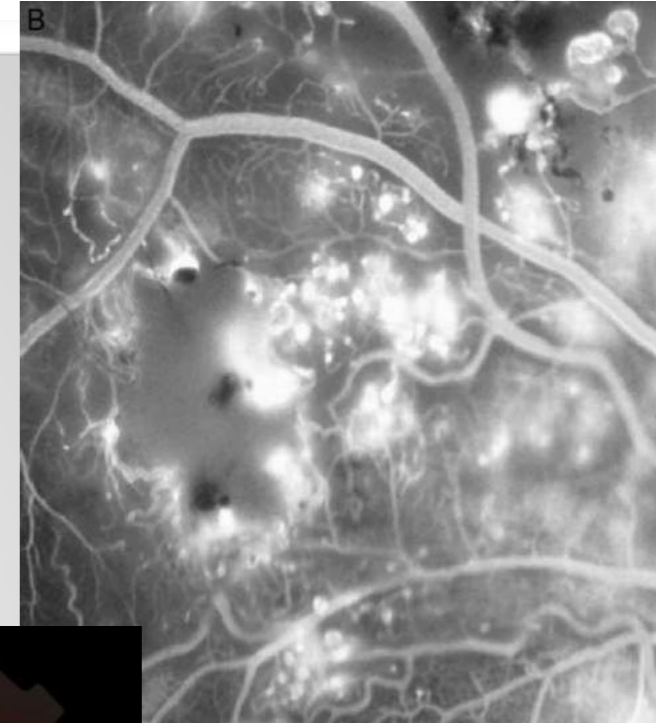
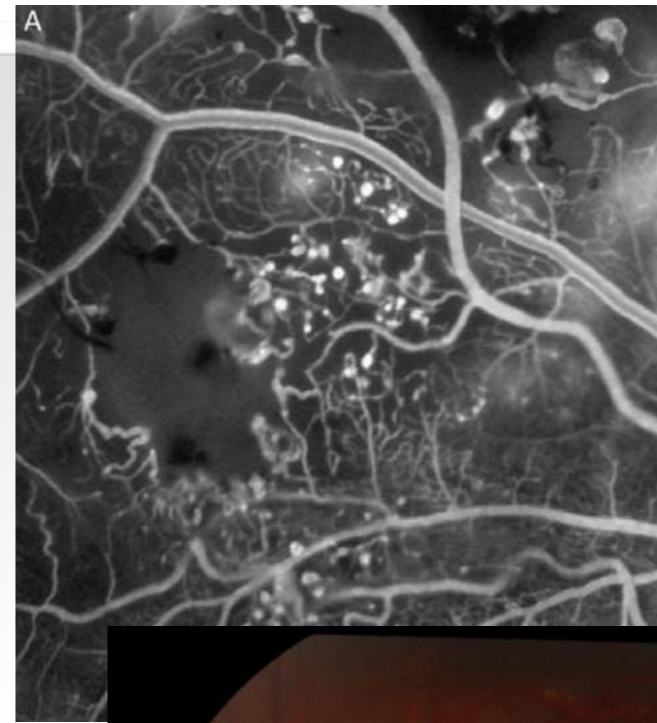


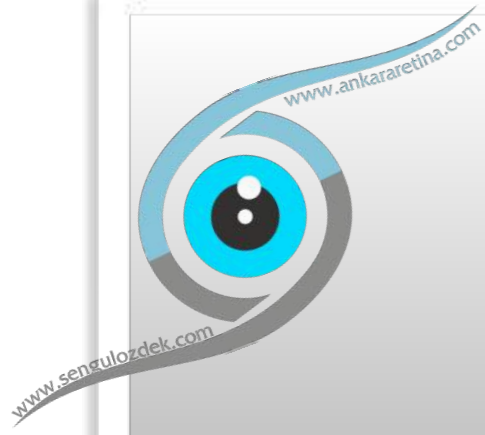


# COATS

## Classification-Shields

1. ONLY TELANGIECTASIS
2. TELANGIECTASIS & EXUDATION
  - A. Extrafoveal Exudation
  - B. Foveal Exudation
3. + EXUDATIVE RD
  - A. subtotal
  - B. total
4. + NVG
5. END STAGE DISEASE (Painful red eye)





# COATS-Treatment

- Primary Treatment: Laser (Stage 1,2,3)
- Cryotherapy: Stage 3
- Surgery: Stage 3-4
  - External Drainage Cryo or Laser
  - Anti-VEGF or Ozurdex
  - PPV-External Drainage
  - Retinotomy for internal drainage should absolutely be avoided!



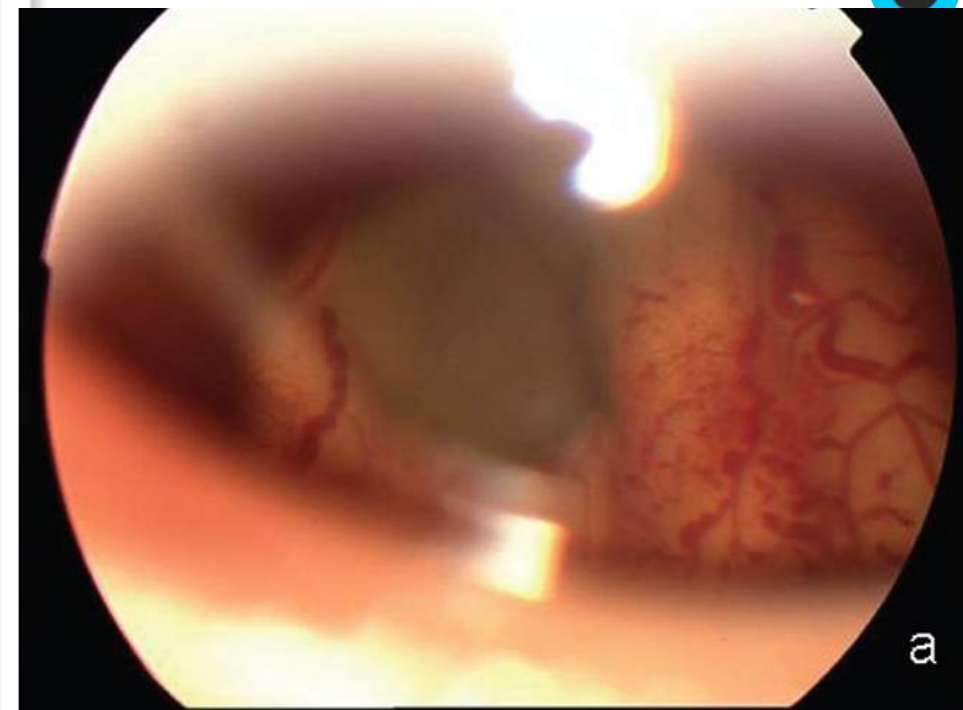


# COATS Treatment

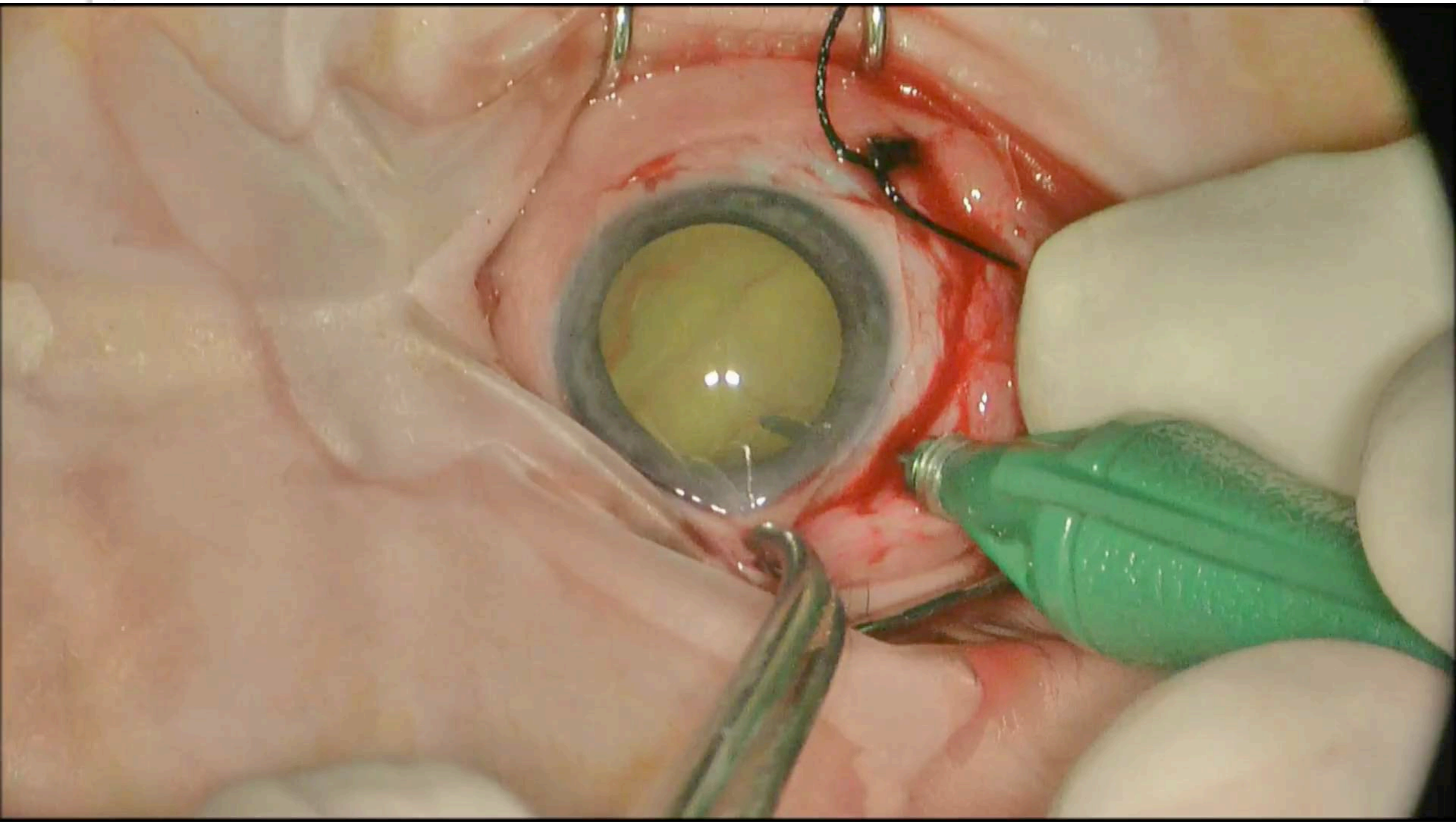
## End stage



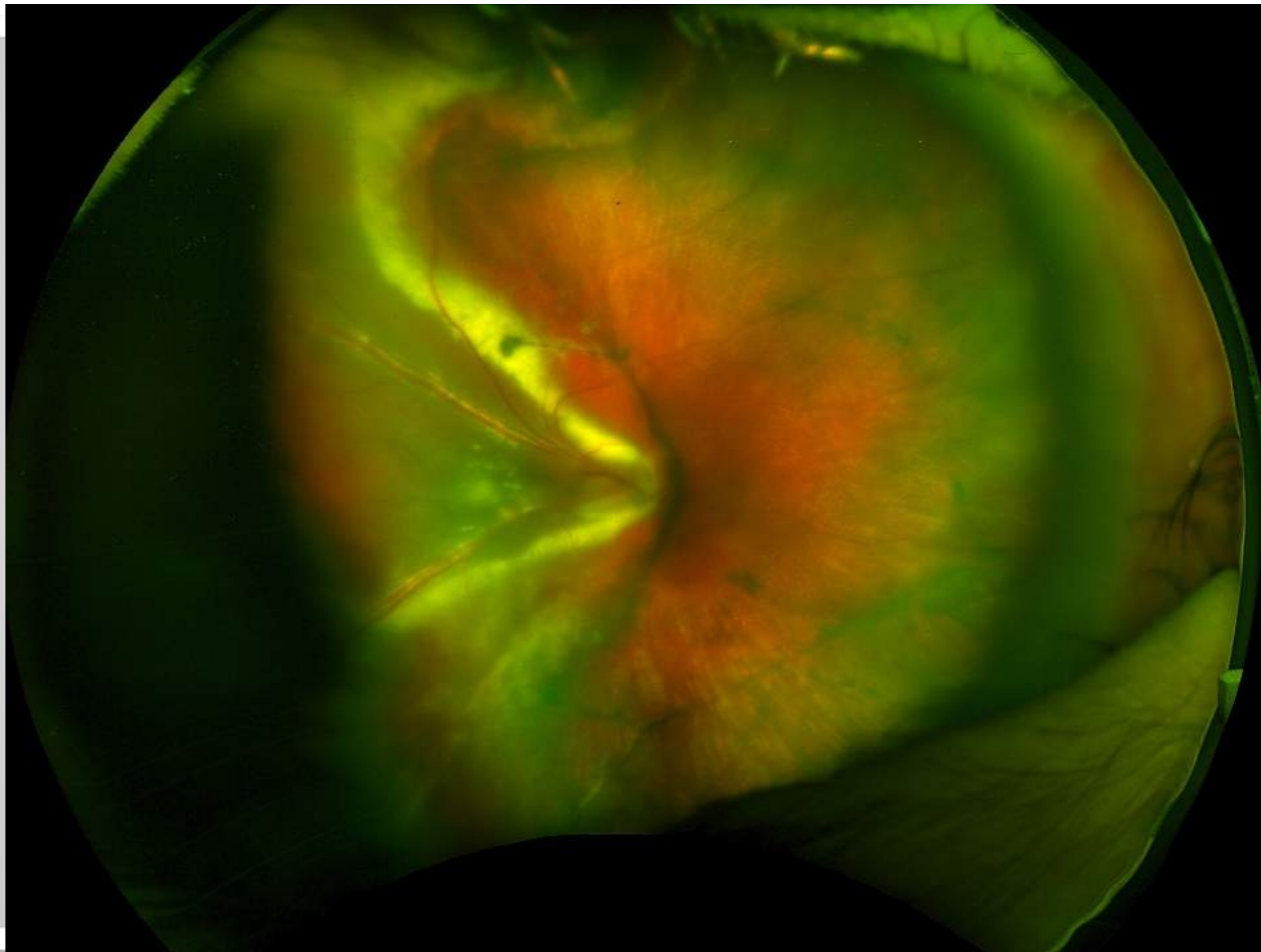
- Surgery to preserve the globe in advanced cases (for cosmesis and comfort)
- 16% of eyes in Coats disease are enucleated because of painful eye.



# 9 mo old boy, Stage 4 Coats Drainage Cryo



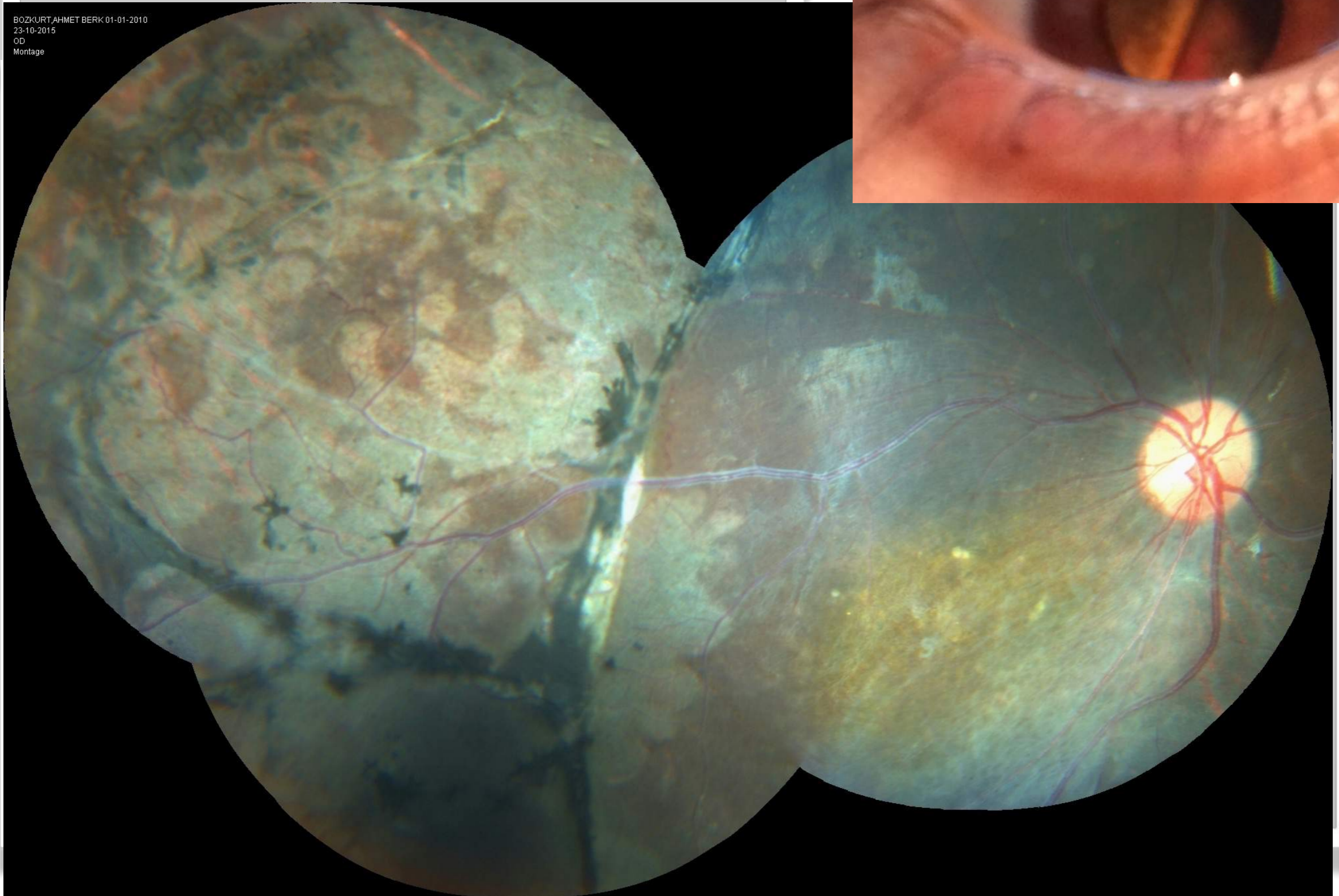
# 9 mo old boy, Stage 4 Coats Drainage Cryo



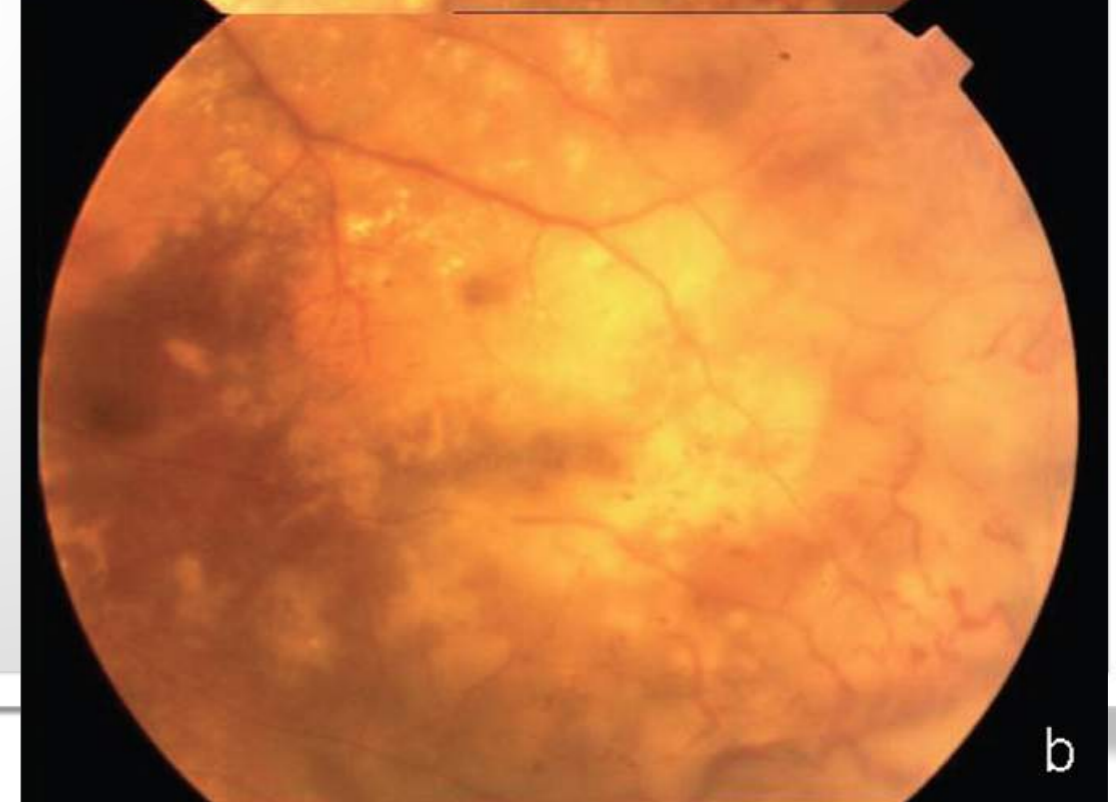
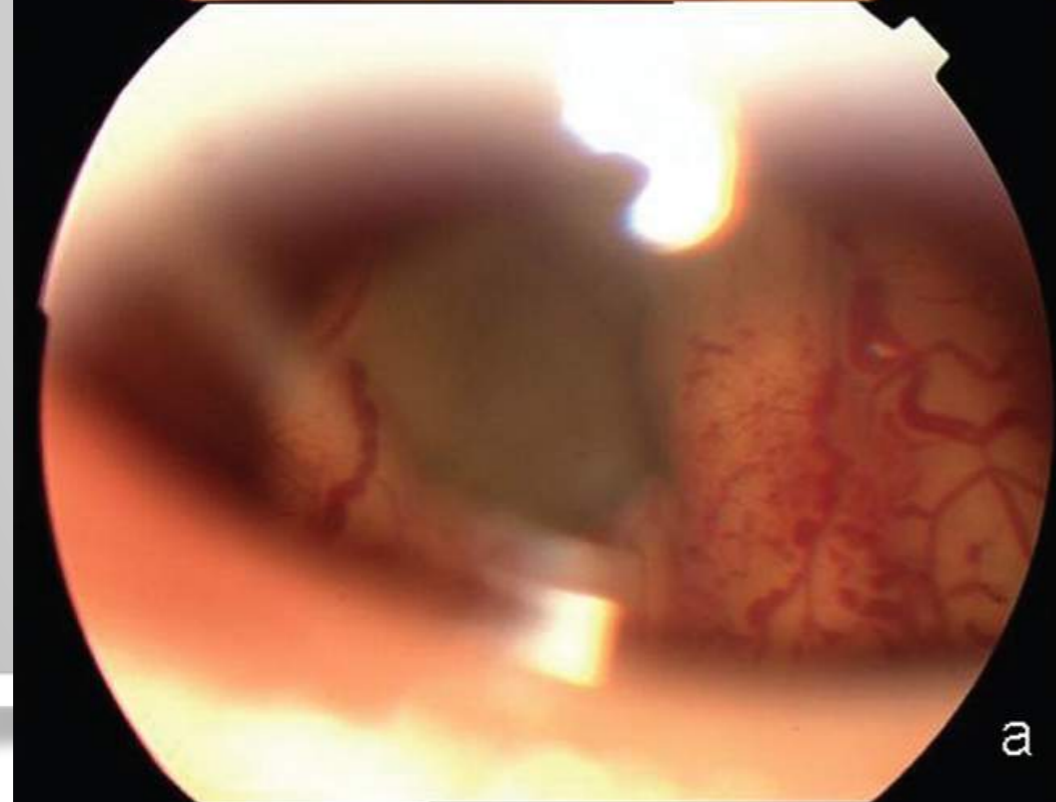
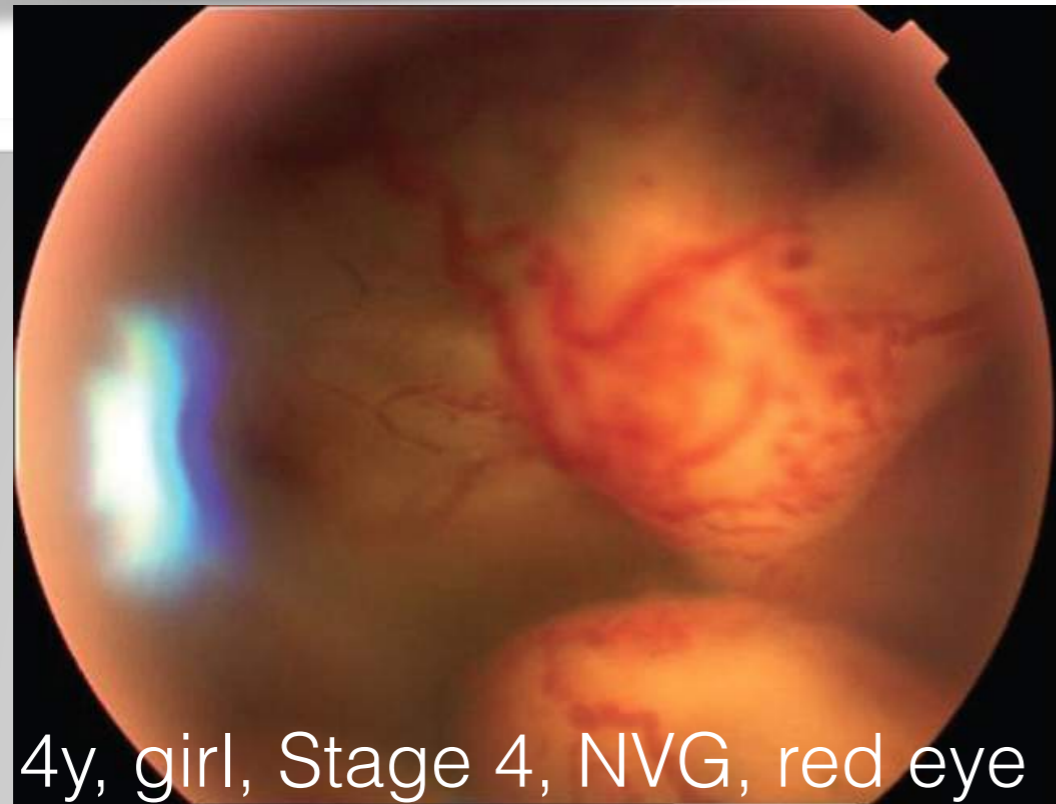
# Stage 4 Coats

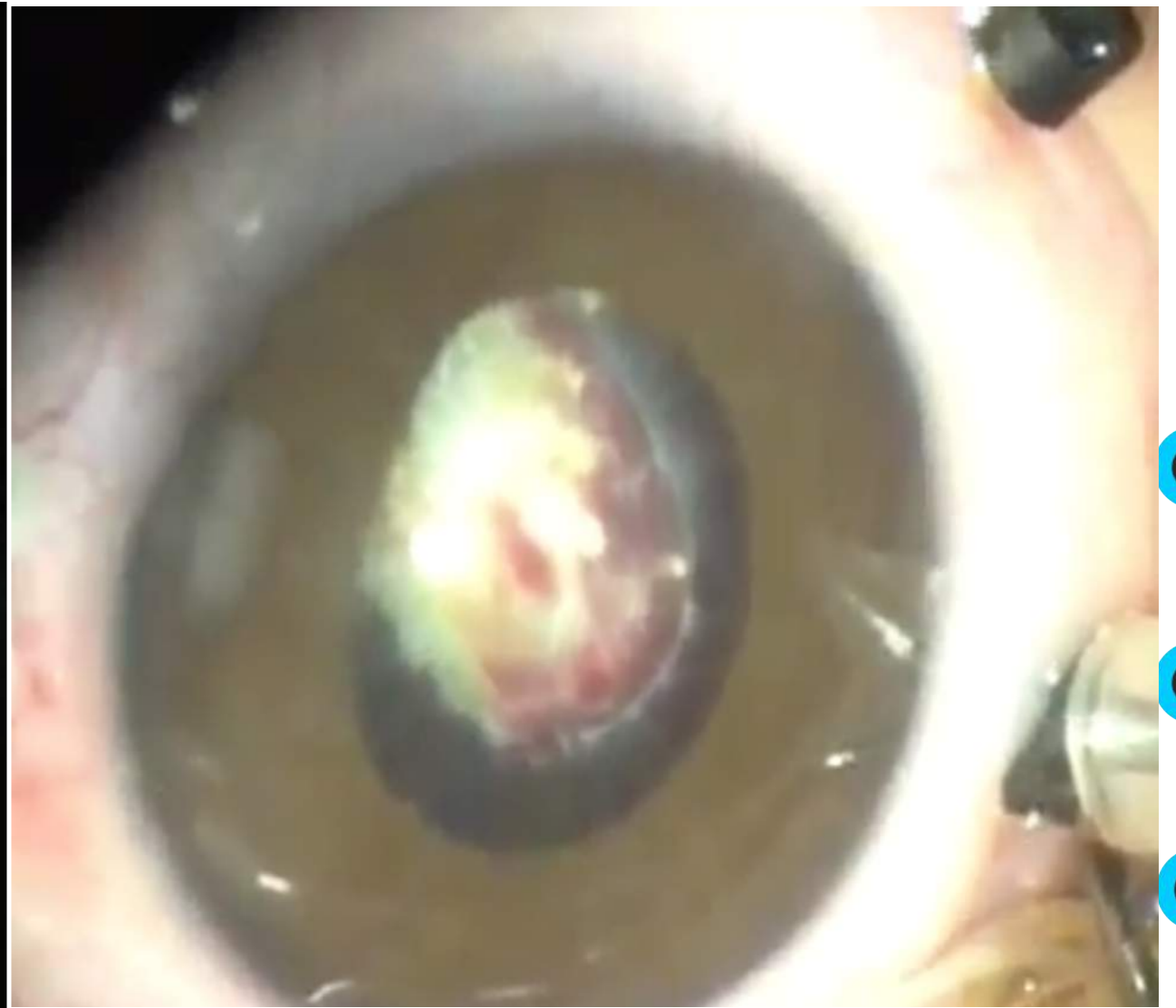
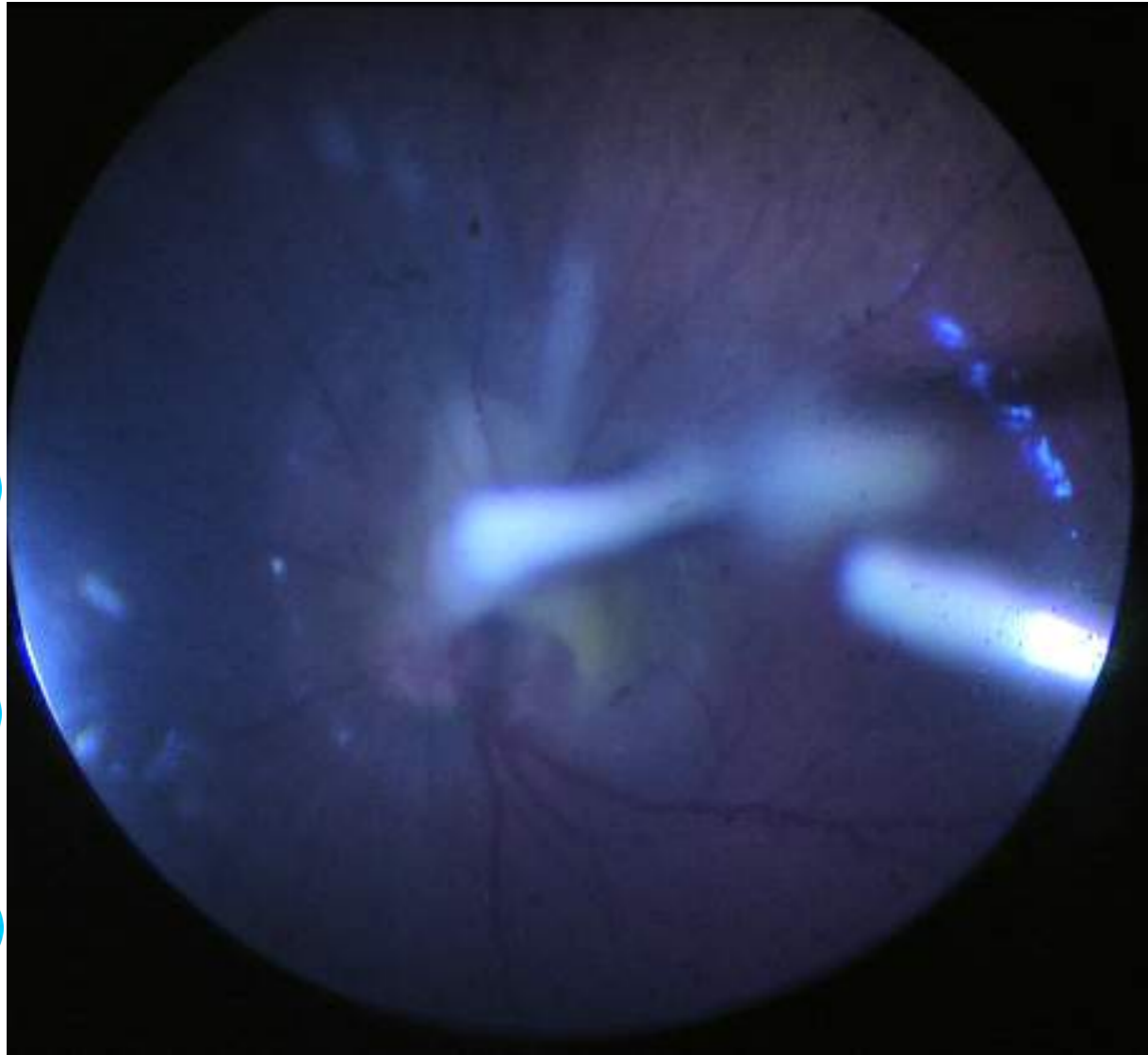


BOZKURT AHMET BERK 01-01-2010  
23-10-2015  
OD  
Montage



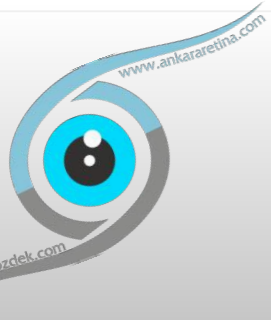
Ozdek S et al. Spontaneous Regression in Two Unusual Cases of Advanced Coats' Disease. J Pediatr Ophthalmol Strabismus. 2010 Mar 26:1-4.





# Persistent Fetal Vasculature





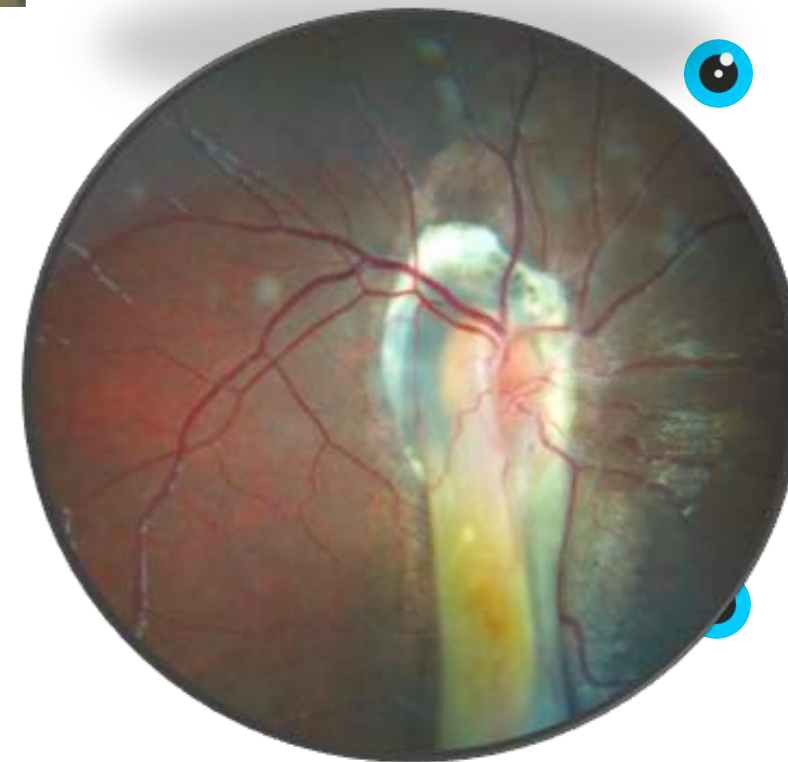
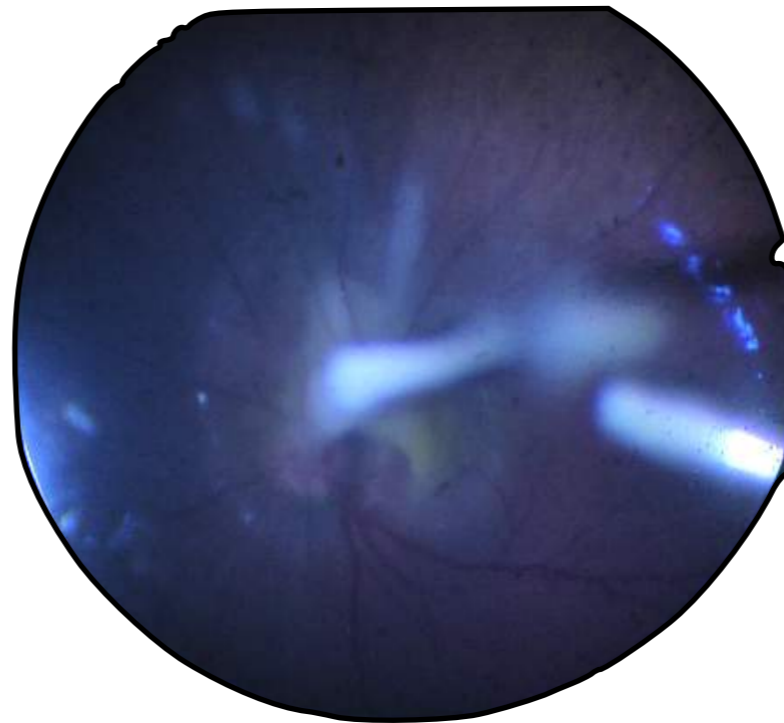
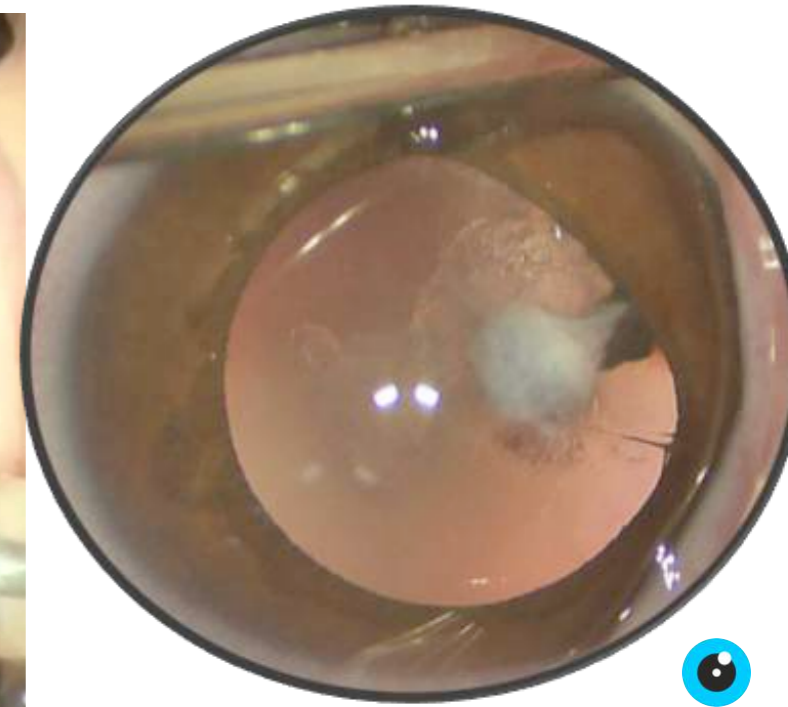
# PFV

Anterior

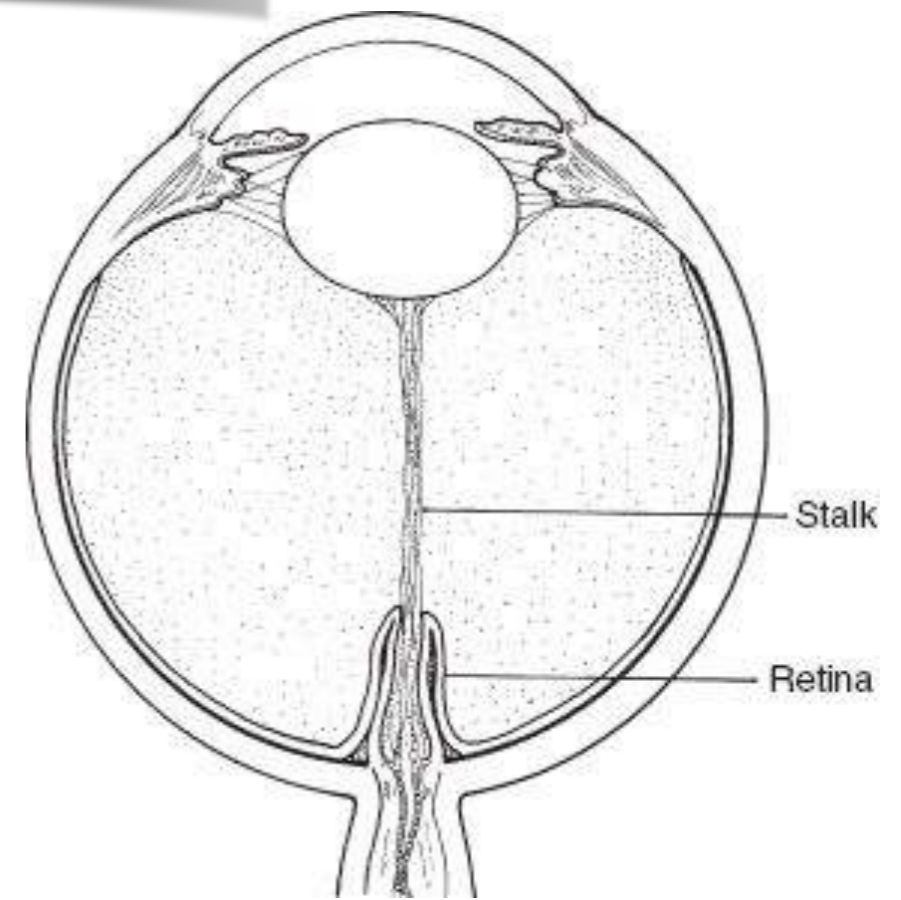
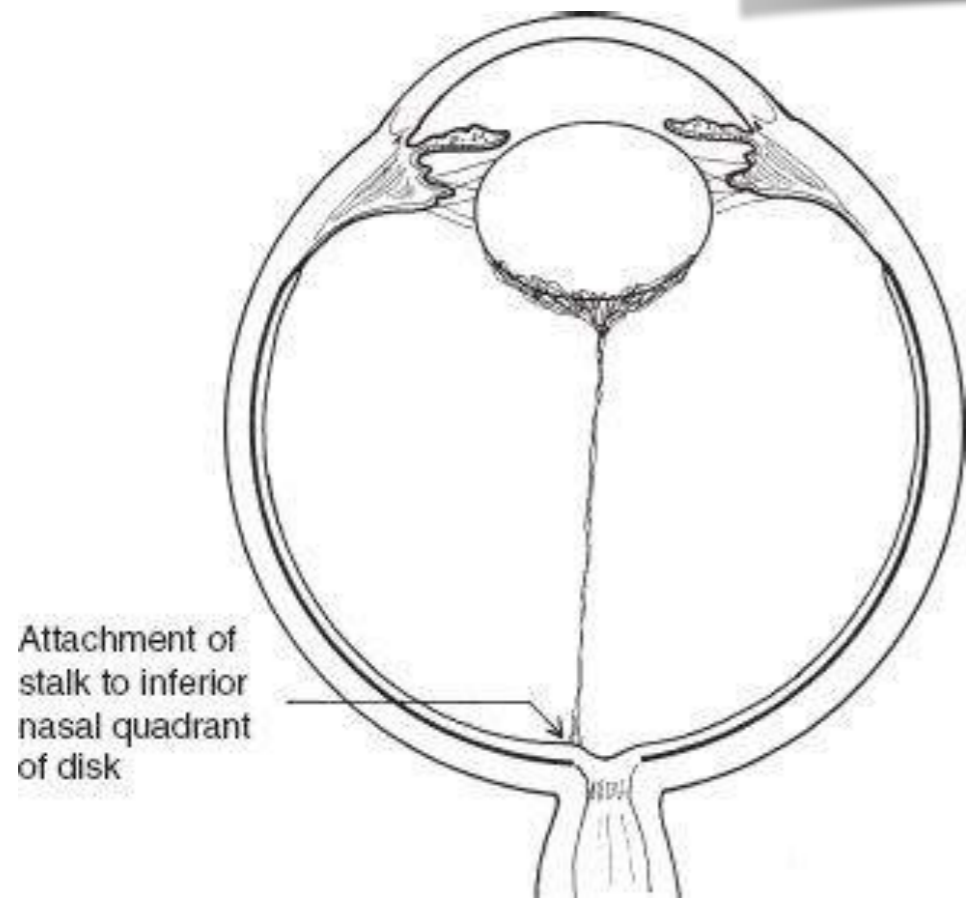
Posterior

Mixed

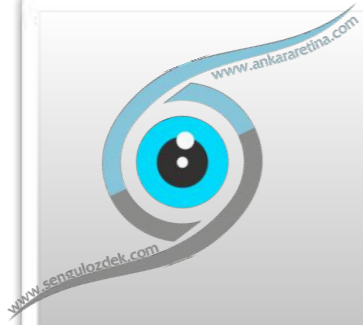
Combined with other  
retinal pathologies



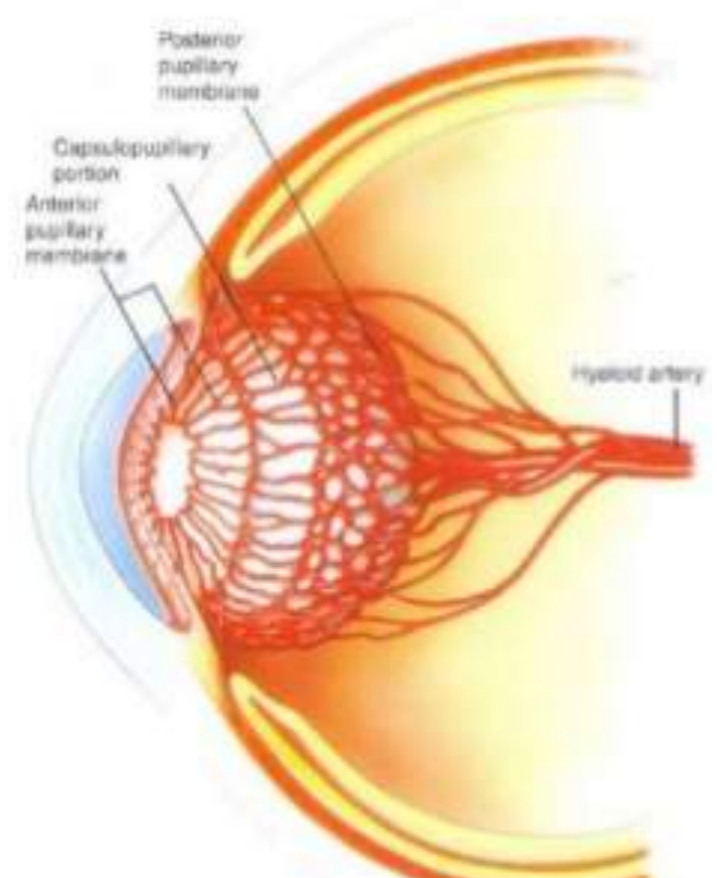
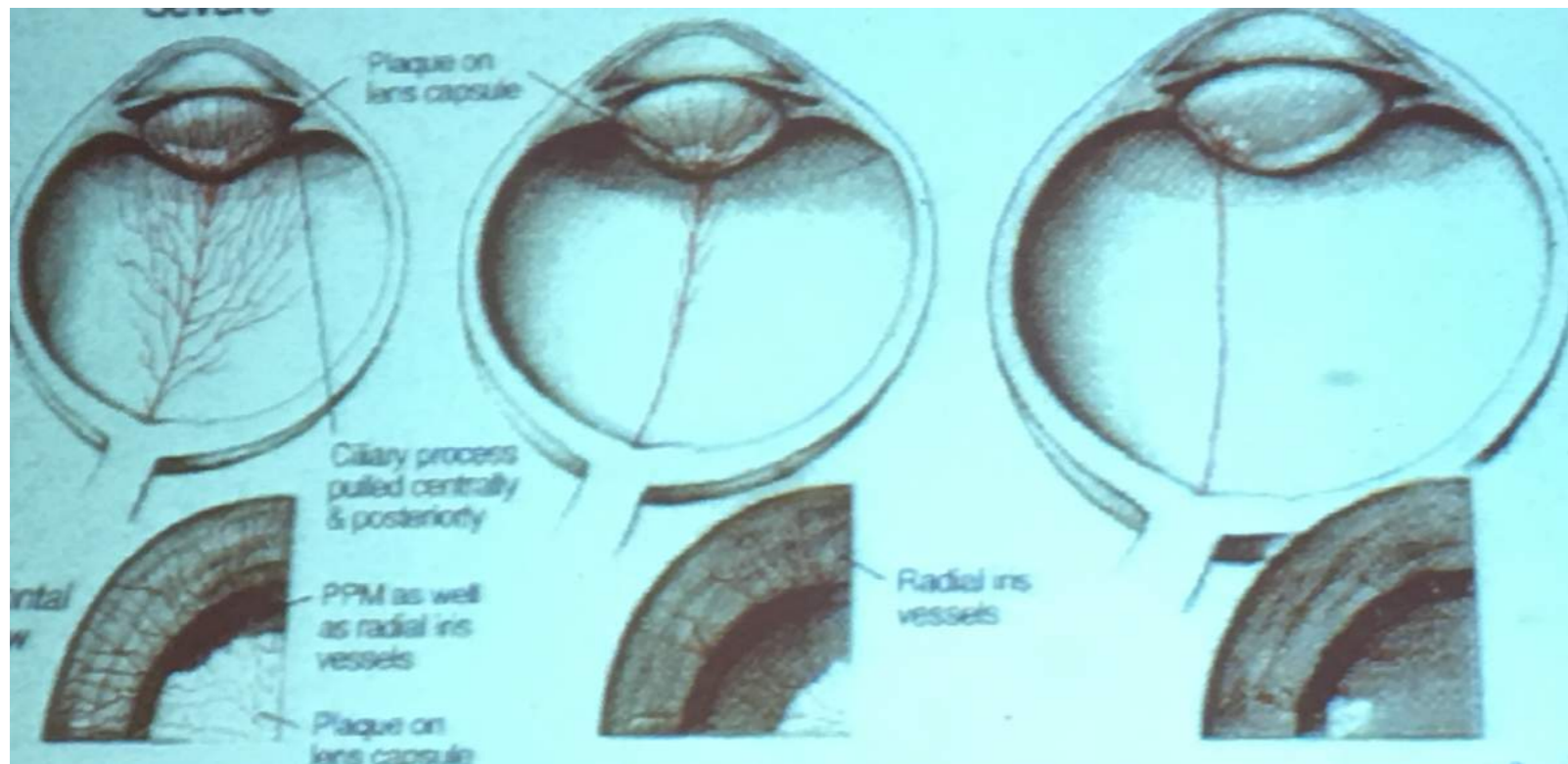
# PFV

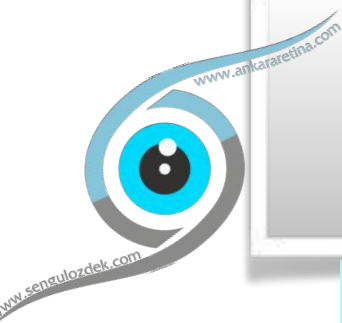




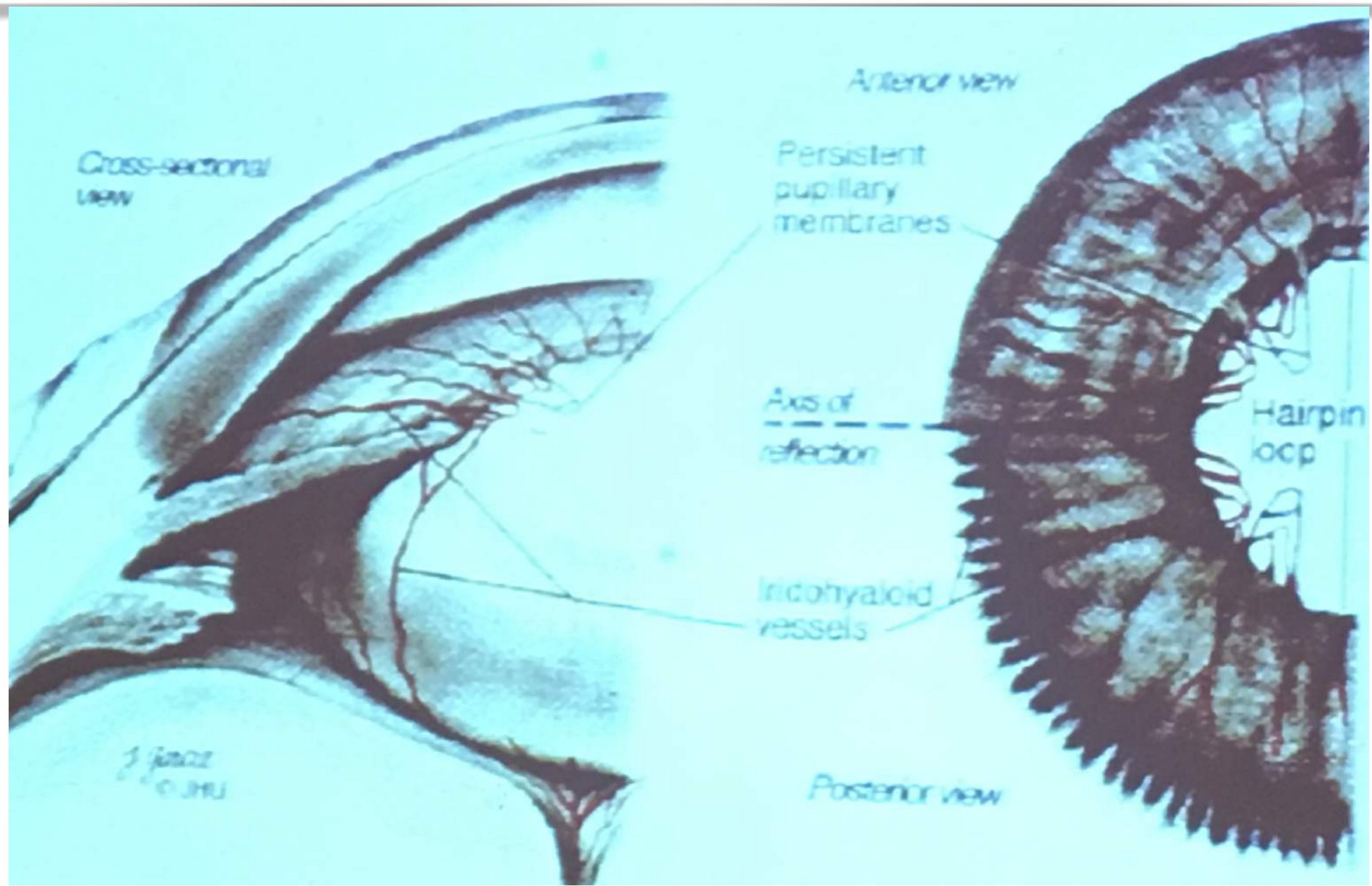


# PFV-anterior





# Radial iris vessels (hairpin-loops)



# Goals of Surgery for PFV

To get acceptable anatomical and functional results

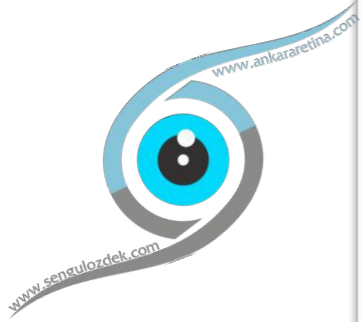
- Clear the media in order to prevent amblyopia
- Relieve tractional forces
  - To prevent TRD, glaucoma, Phthisis bulbi
  - To let the eye to have the opportunity to grow

# Anatomical differences in Anterior PFV

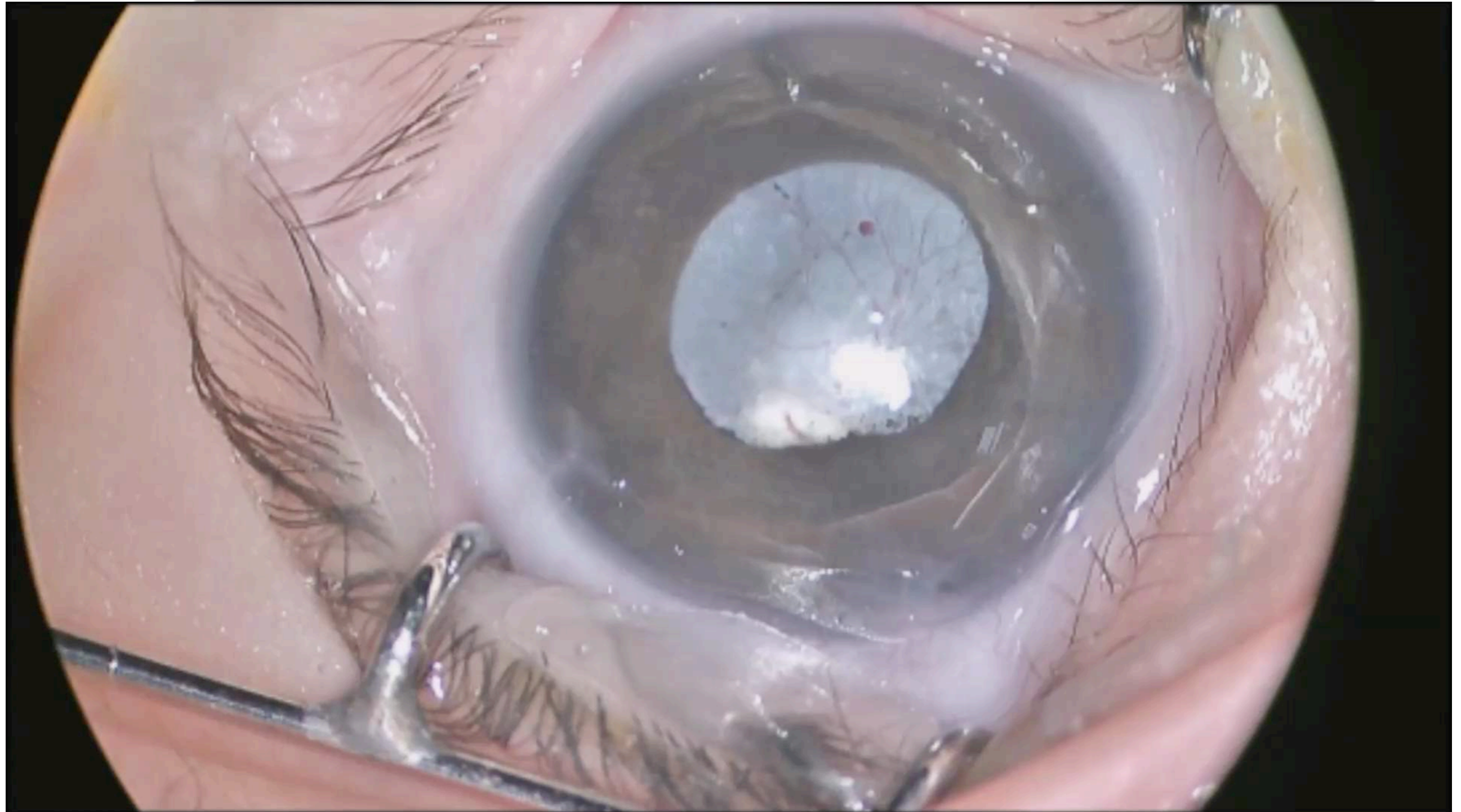
- PP not developed,
- No zonules,
- Elongated ciliary processes



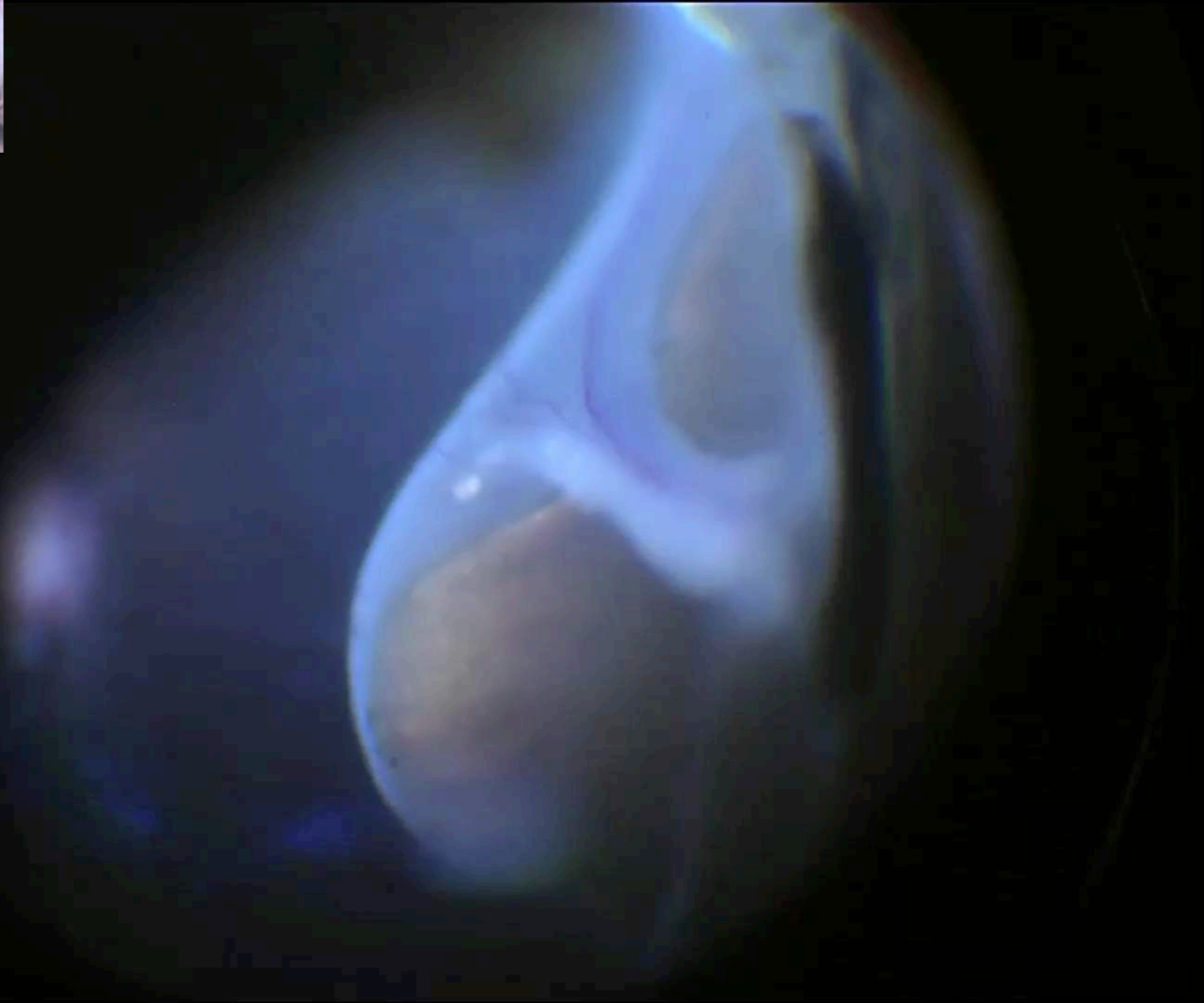
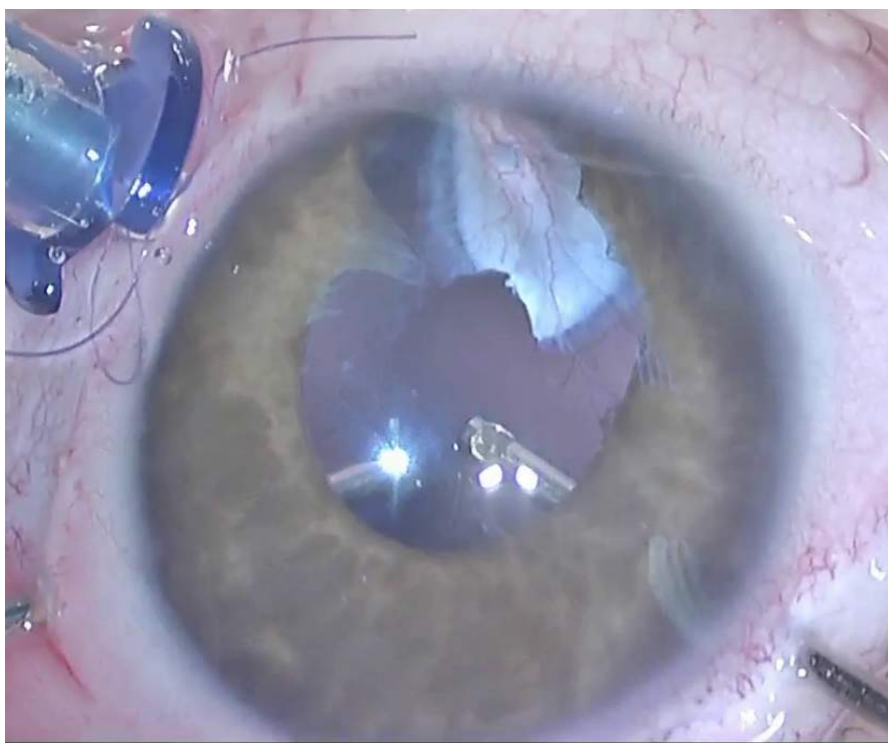
Anterior fibrovascular structure may be continuous with retina!

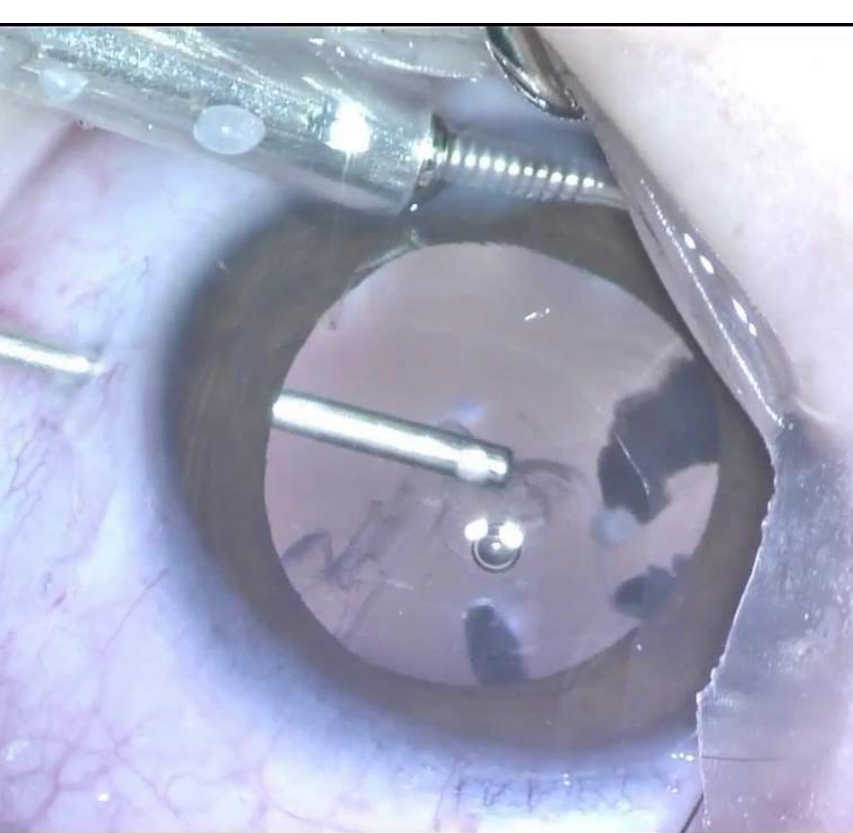


# PFV-Bad surprise!

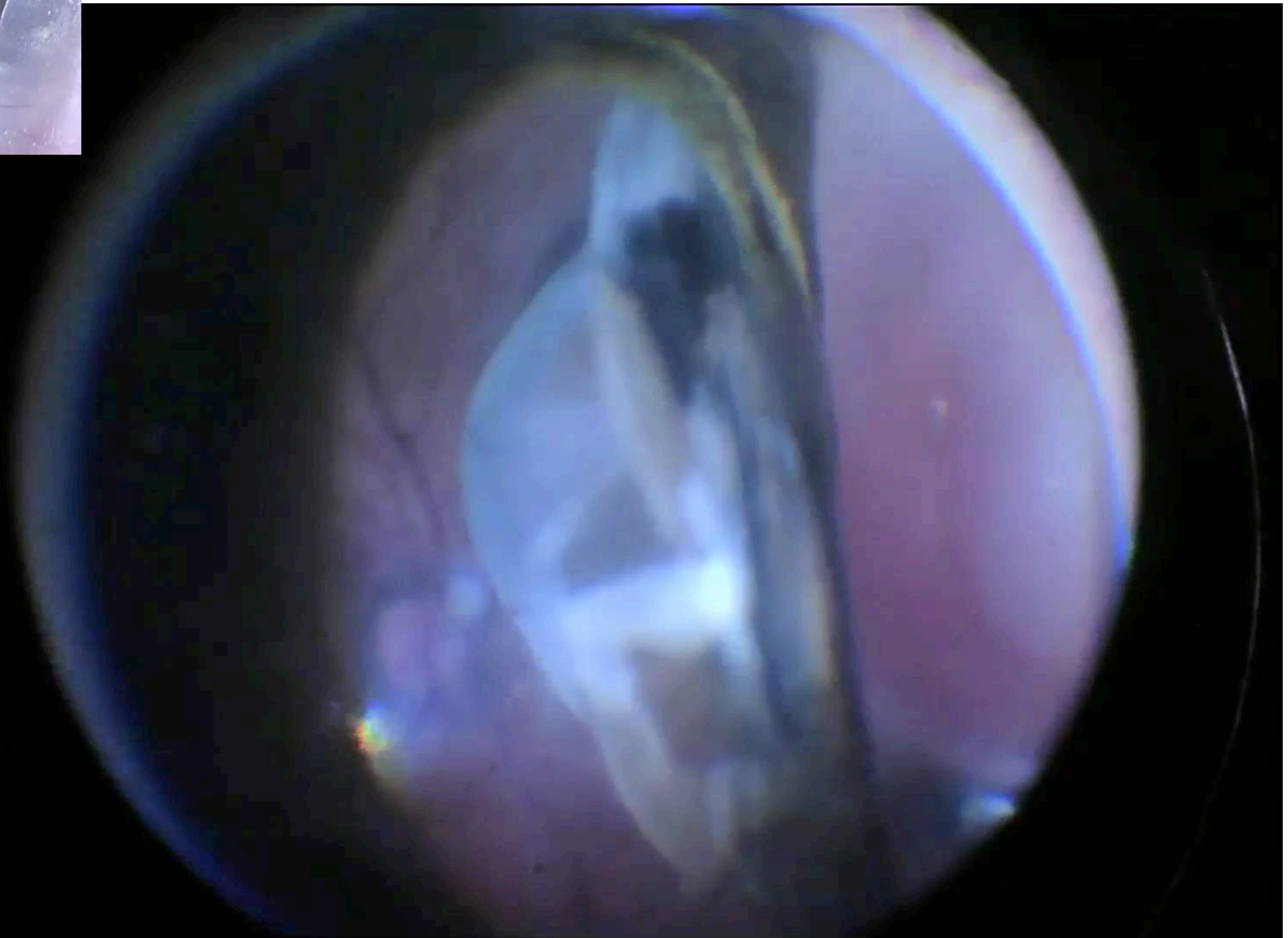


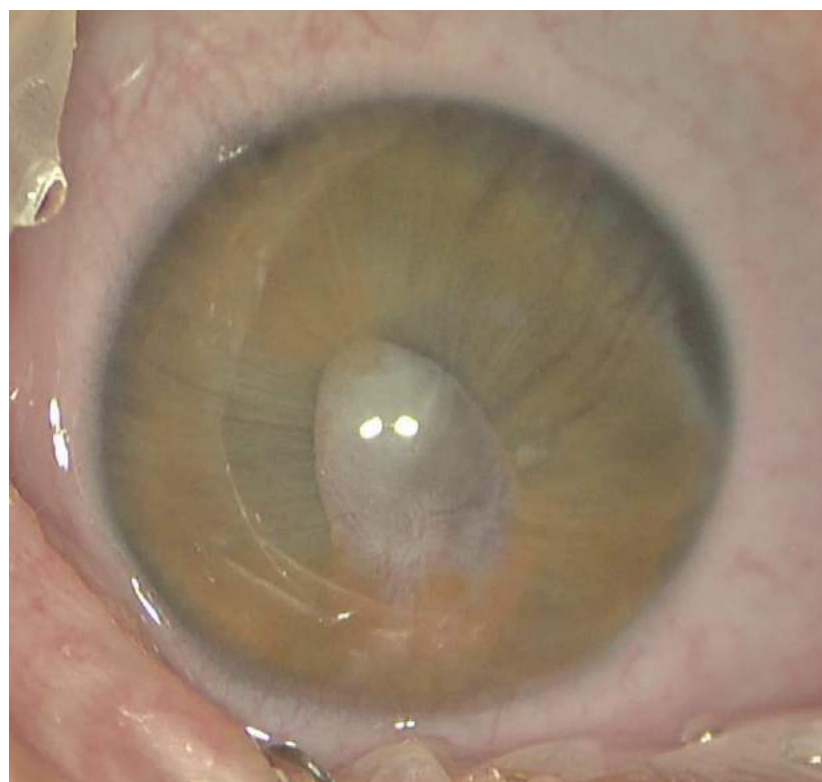
# PFV-anterior retinal elongation



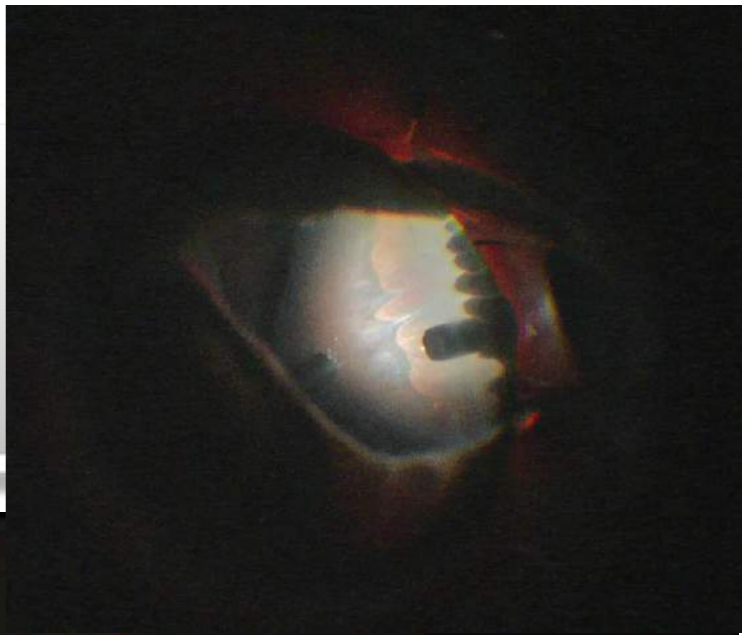


Tips: Pigmented plaques  
Nasal and inferior location



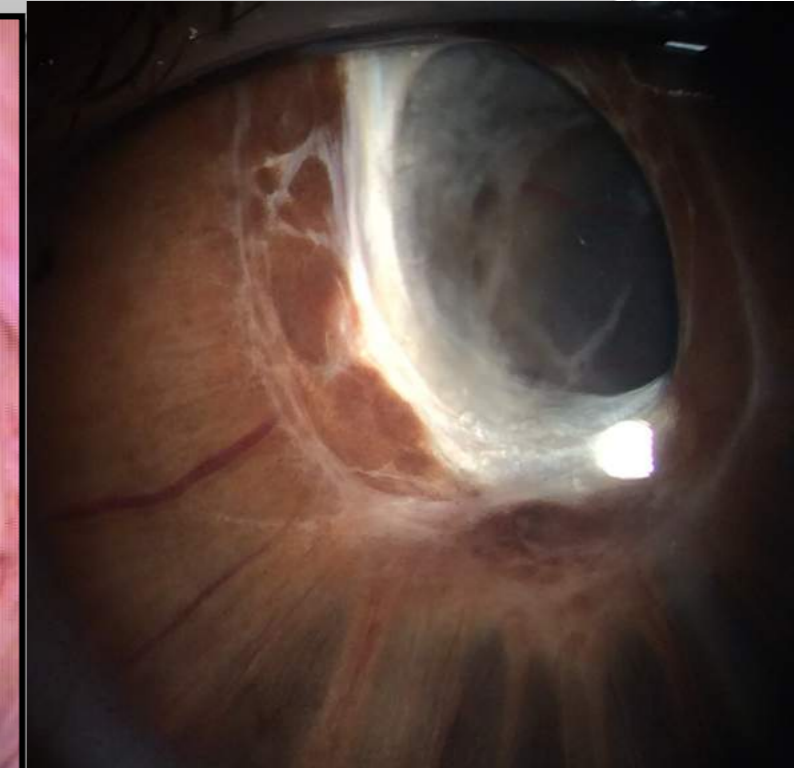


PFV-anterior retinal elongation

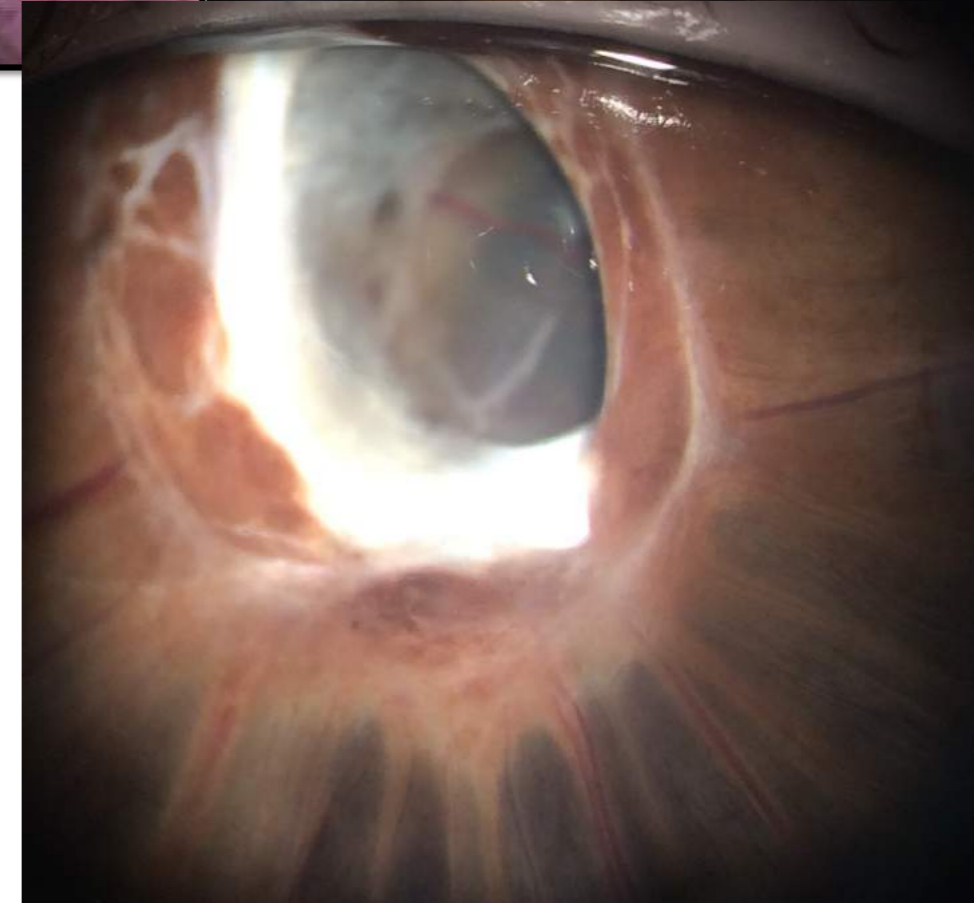




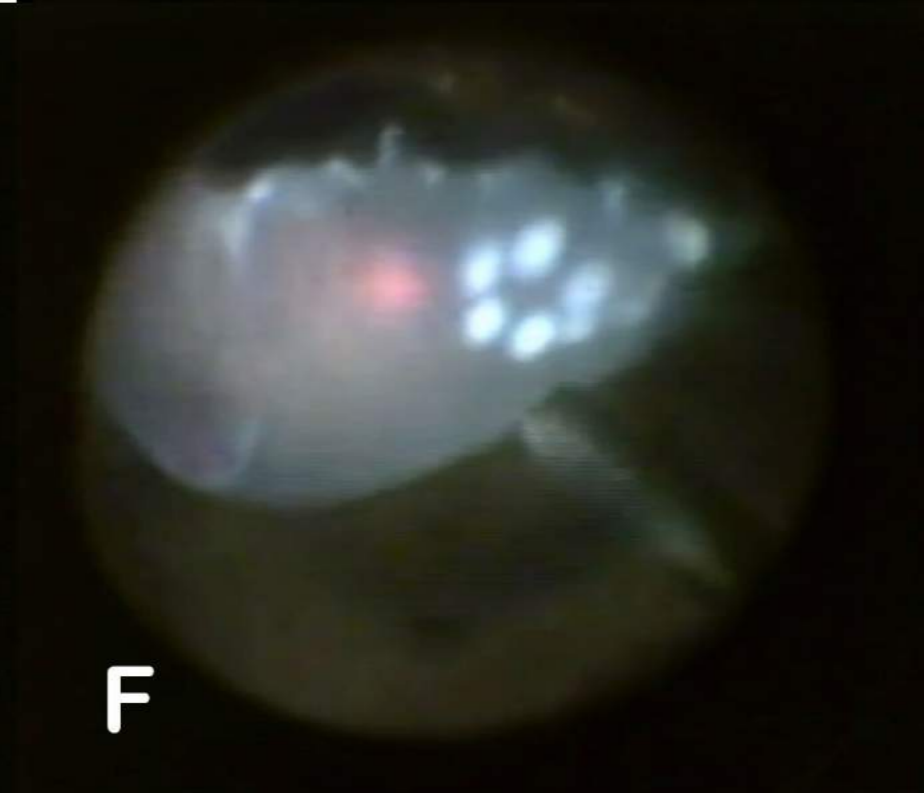
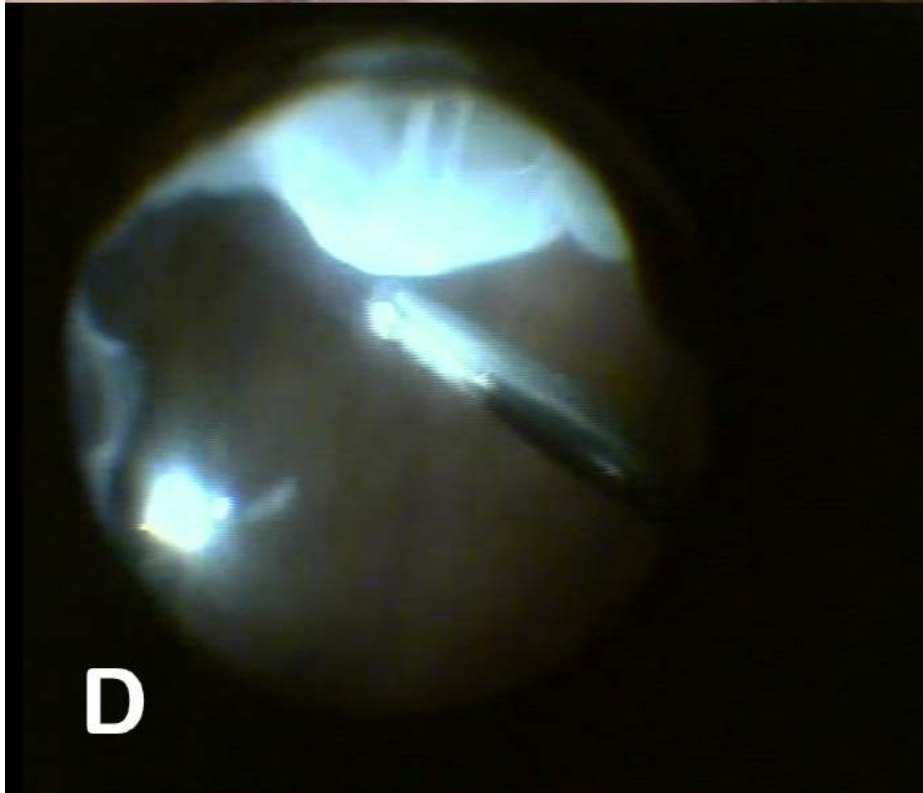
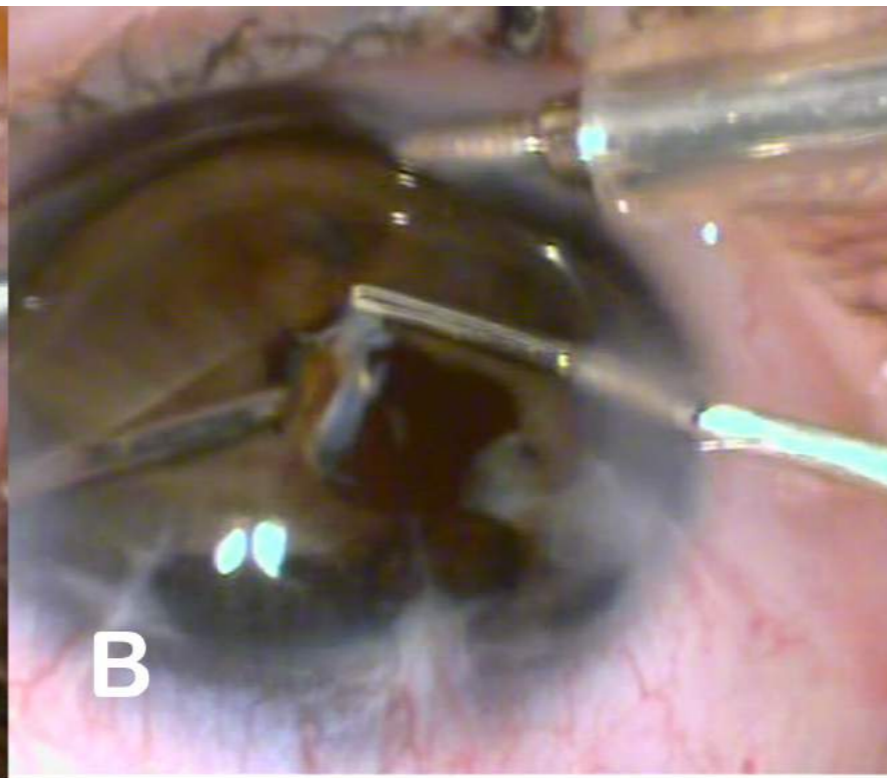
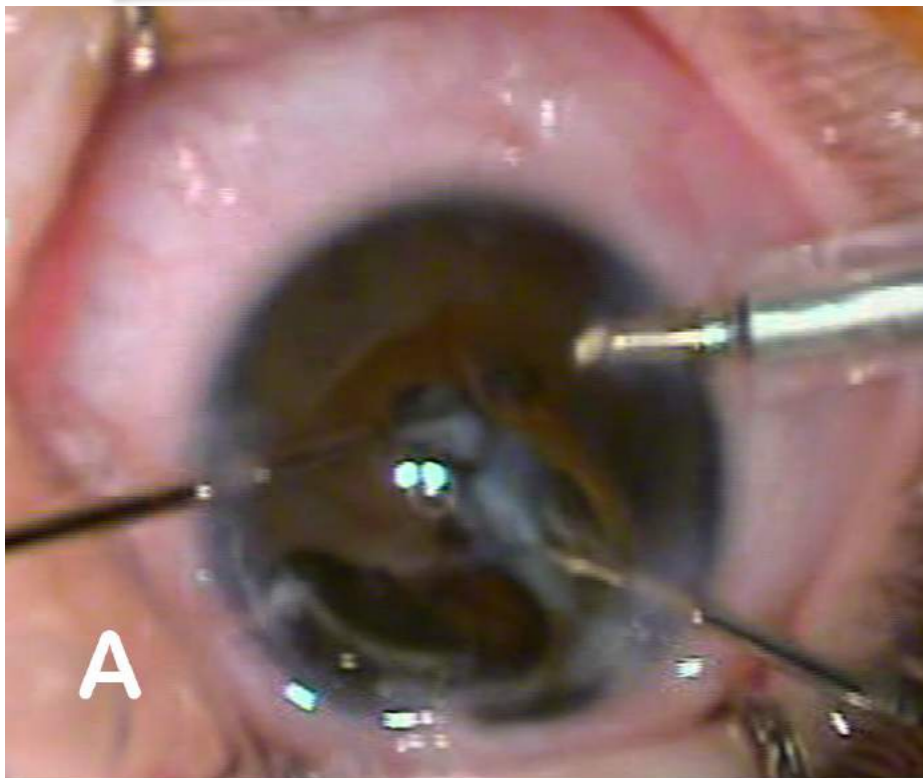
# Total excision vs leaving some part of FV tissue?

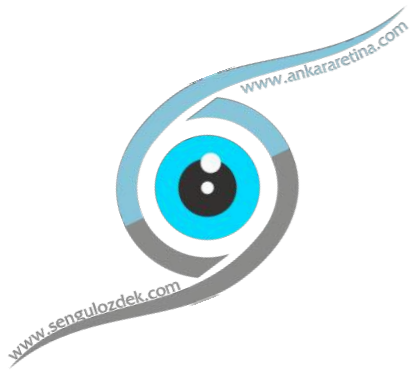


FV tissue contracts and causes pupillary obliteration or Peripheral RD within years leading total TRD



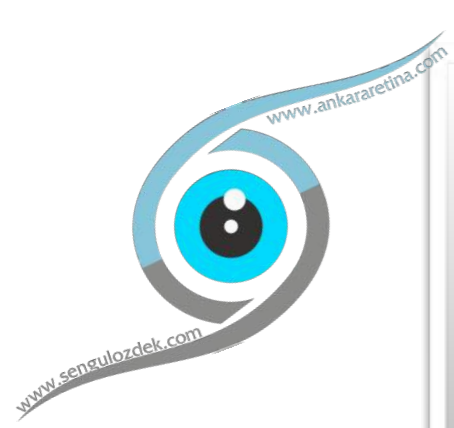
# Leaving peripheral part of FV tissue....pupillary obliteration





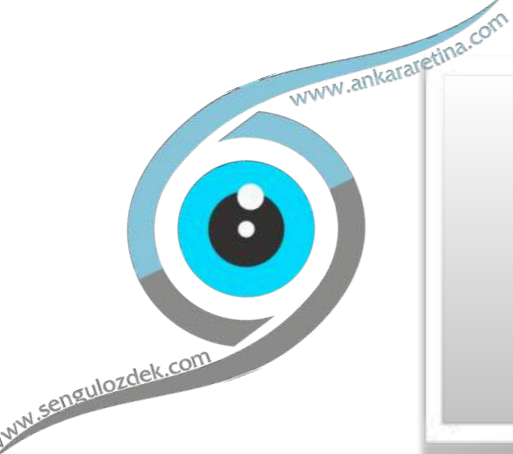
# Patients and Methods

- 29 eyes of 28 patients with PFV (last 6 years)
- 19 (67.1%) were male
- 14 eyes (47.7%): Anterior PFV
- 10 eyes (34.5%): Posterior PFV
- 5 eyes (17.2%): Mixed type
- Median follow-up: 14 months (6 months to 5.5 years)



# Patients and Methods

- Anterior retinal elongation: 64.3% of anterior PFV.
- 84.6% of anterior PFV cases resulted in anatomic success.
- Final VA: 20/200 or better in 38.5% of anterior PFV cases.



# Complications

**Table 2. Complications**

	Retinal complications		p-value <sup>†</sup>		Anterior segment complications		p-value <sup>†</sup>	
	Yes n (%)	No n (%)	Univariate	Multivariate	Yes n (%)	No n (%)	Univariate	Multivariate
Extend of lens opacity (n=19)*								
Less than half (n=8)	0 (0)	8 (100)	0.018 <sup>†</sup>	0.009 <sup>‡</sup>	0 (0)	8 (100)	0.045 <sup>†</sup>	0.026 <sup>‡</sup>
More than half (n=11)	6 (54.5)	5 (45.5)			5 (45.5)	6 (54.5)		

\*includes anterior and combined cases.

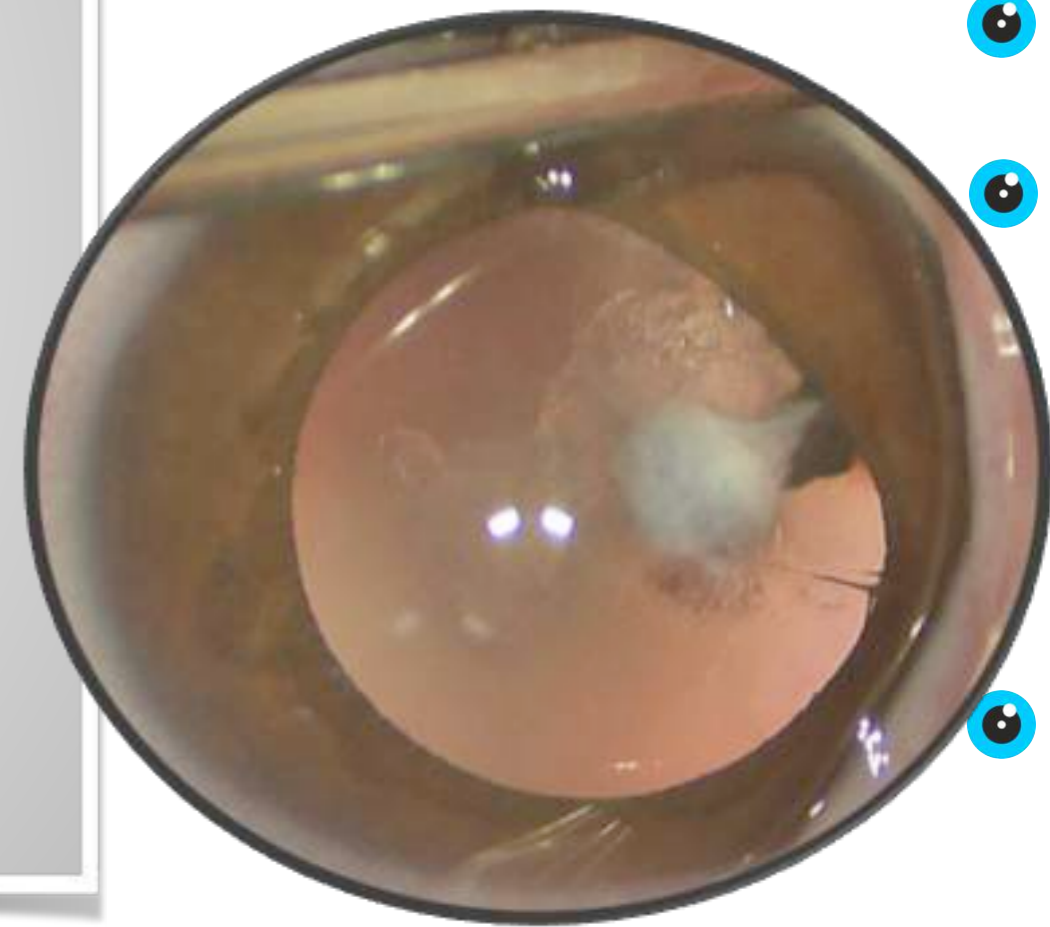
<sup>†</sup>Fischer exact test.

<sup>‡</sup>OLS

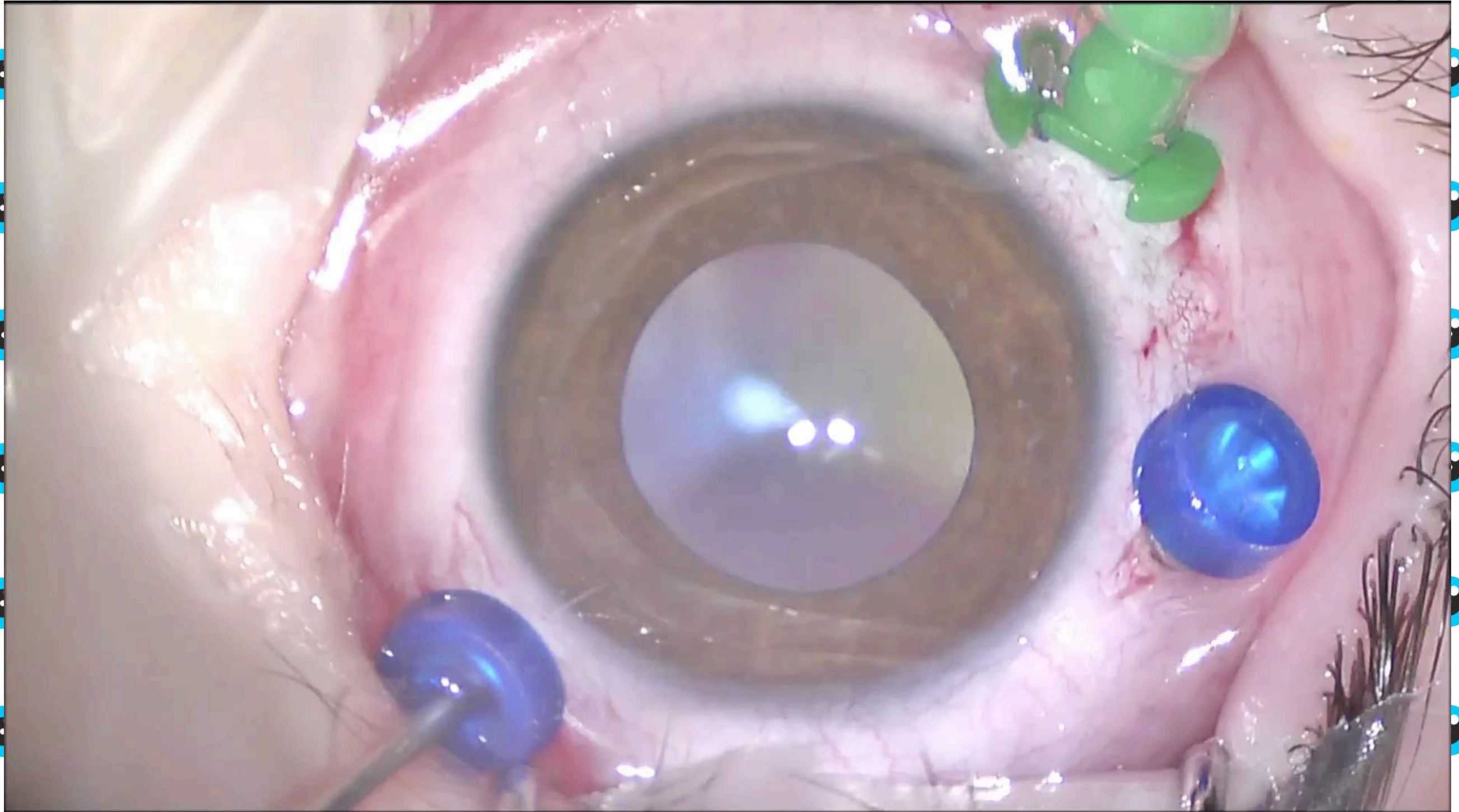


# Results

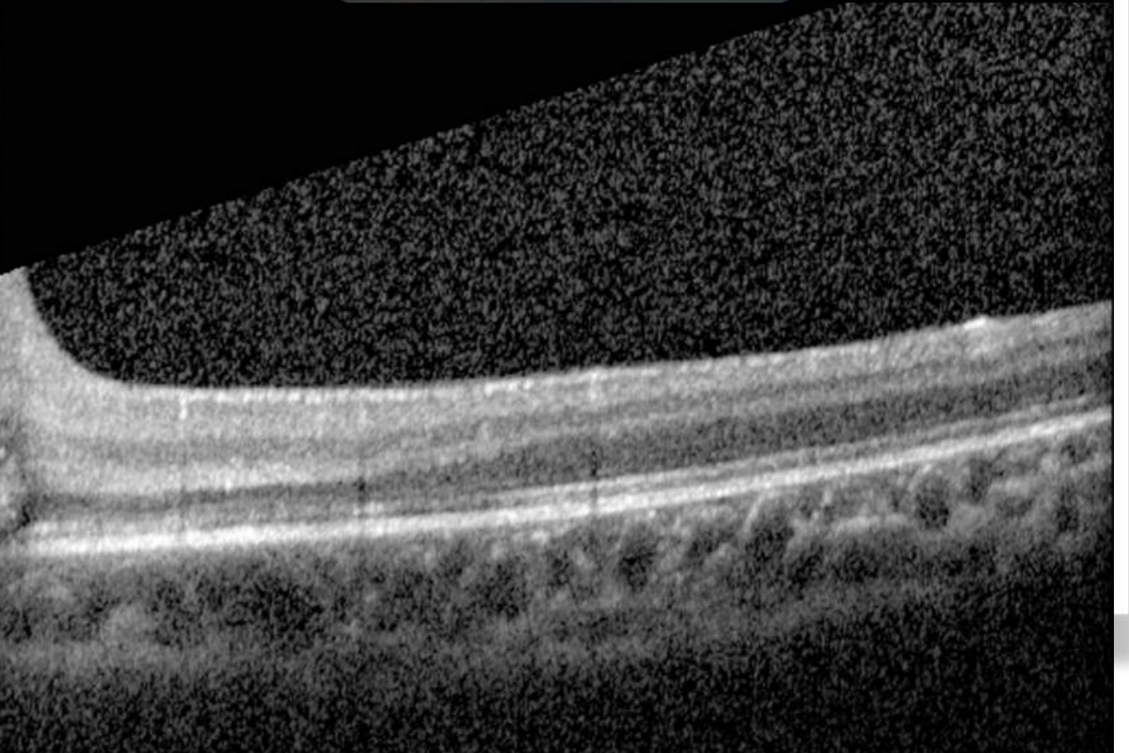
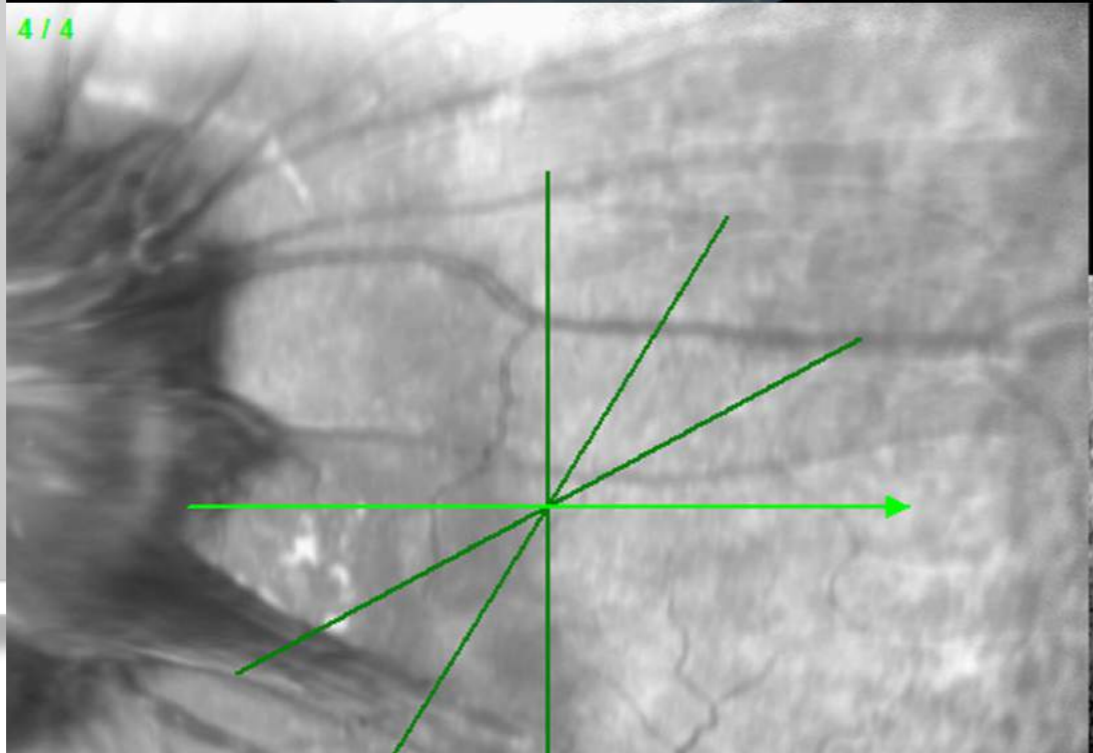
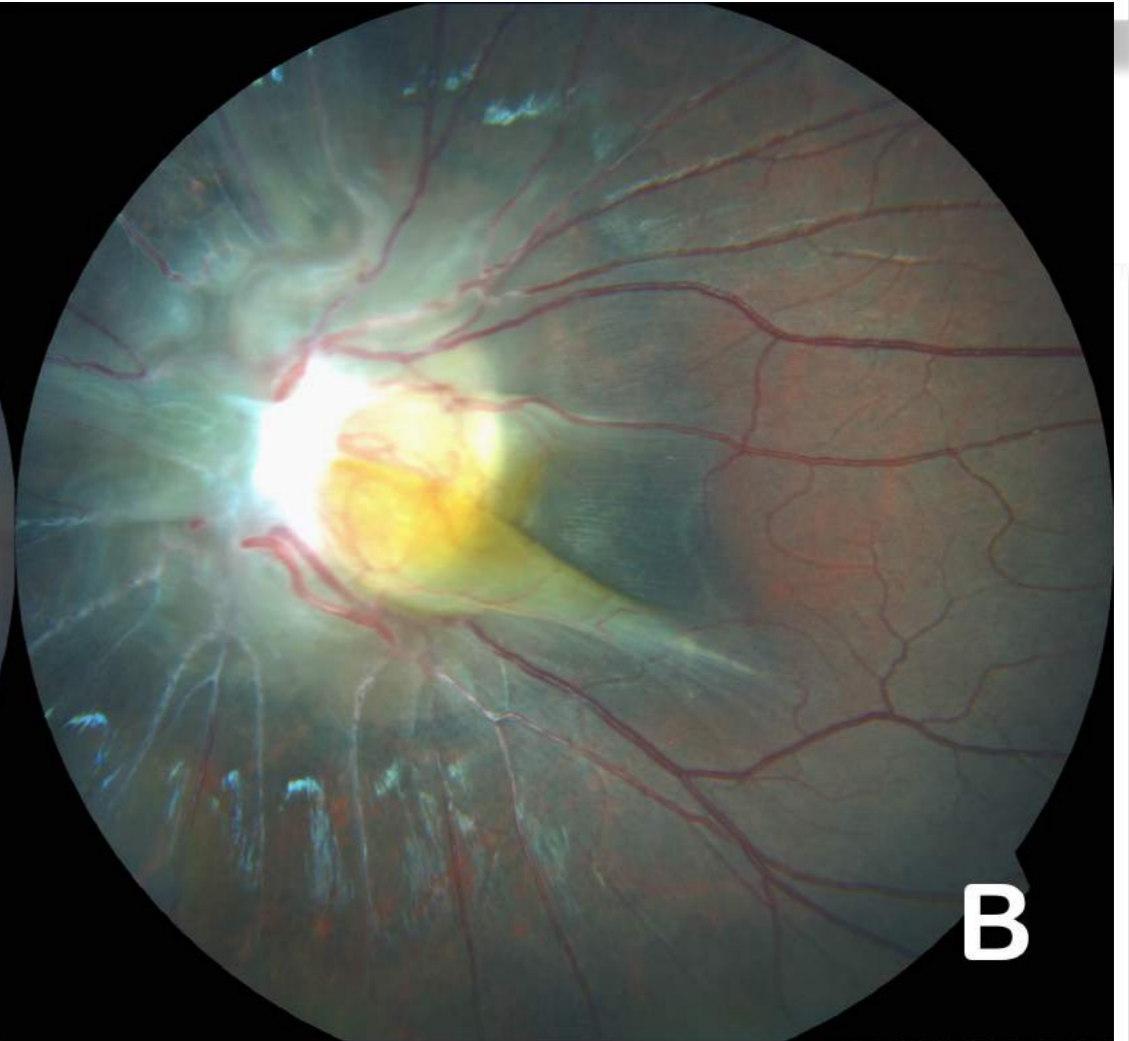
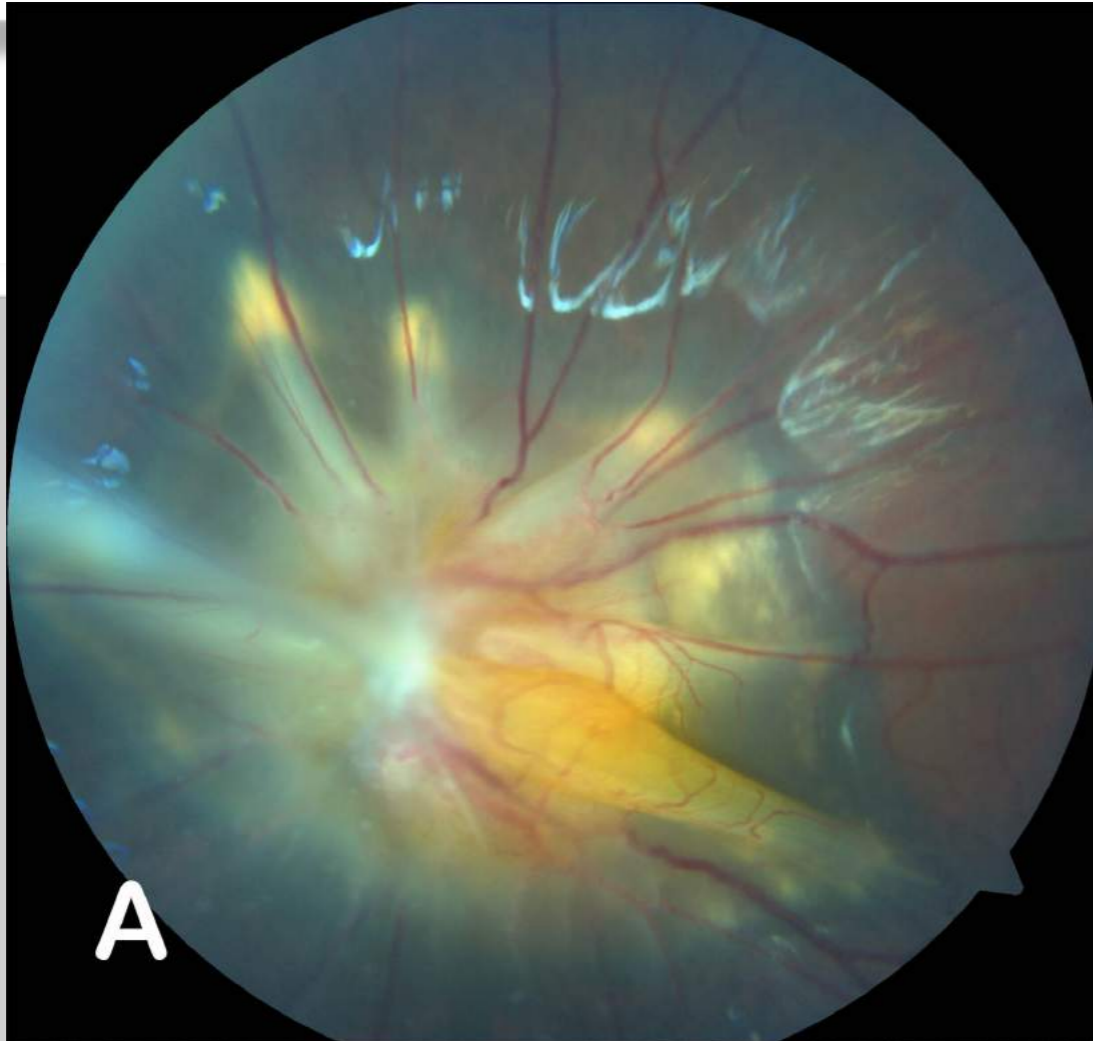
- PFV patients who has more extensive anterior disease tend to have a higher risk of overall complications than patients with localized fibrovascular tissue.
- 
- 
- 
- 



# PFV-posterior

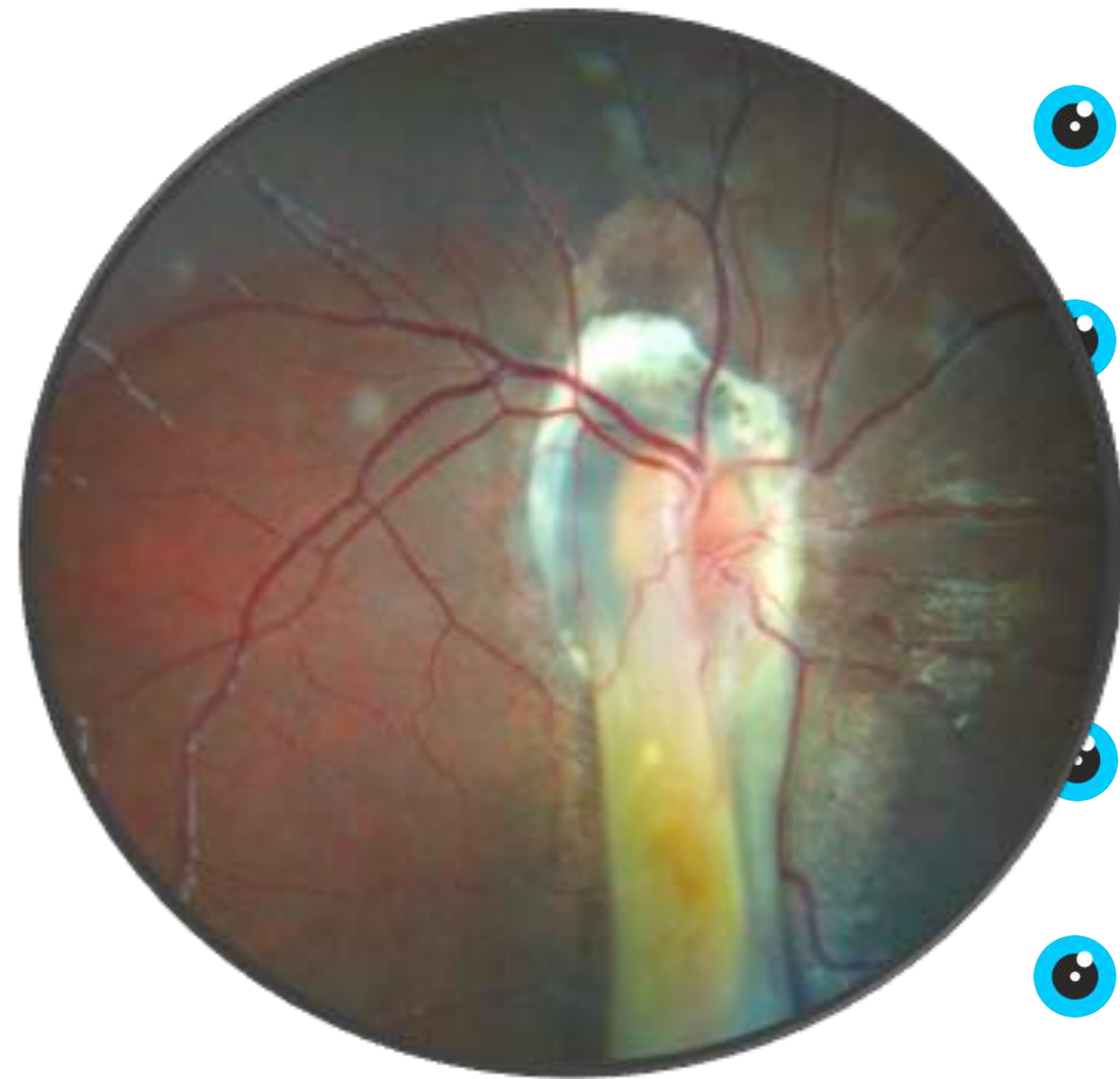
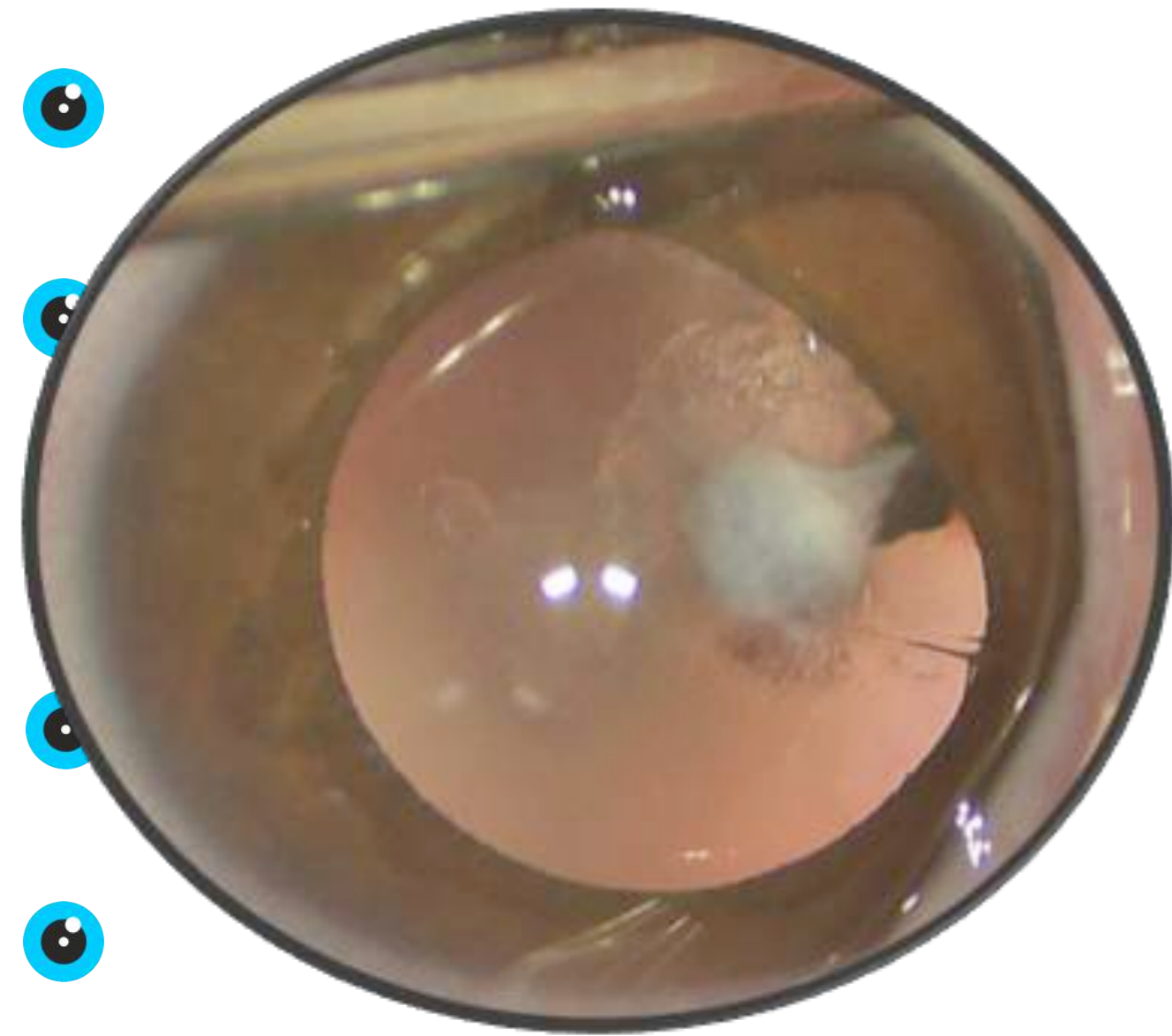


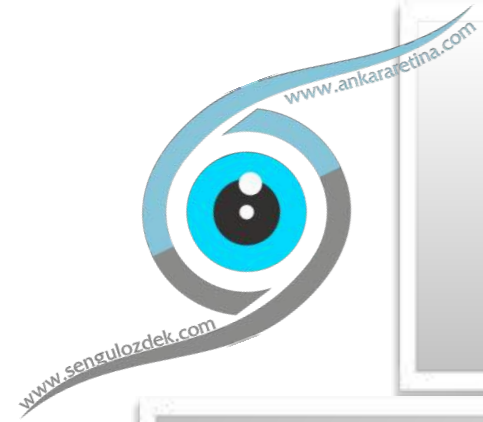
# PFV-posterior-postop





# PFV-mixed





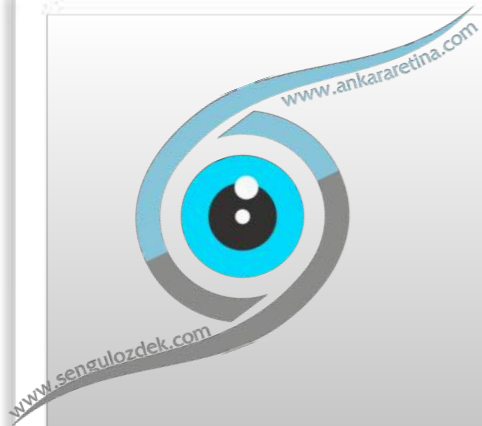
# Anterior PFV-Take home messages

- Aware of peripheral retinal extensions through pars plana esp in Ant PFV.
- Do not leave peripheral vascular fibrotic tissue during surgery: May cause severe complications
- Be prepared for RD surgery.



# CONGENITAL X-LINKED RETINOSCHISIS CXL

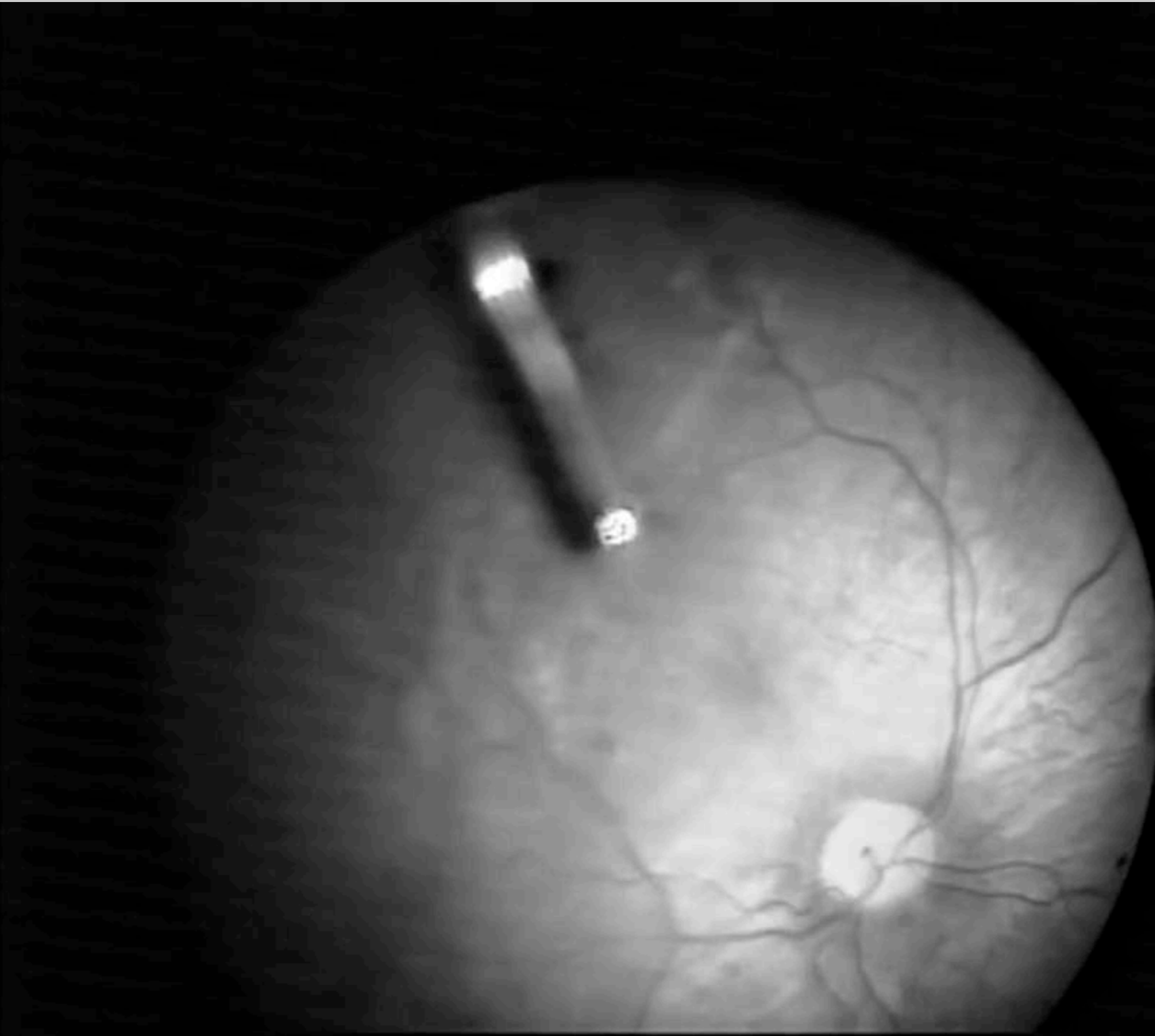




# CXLR

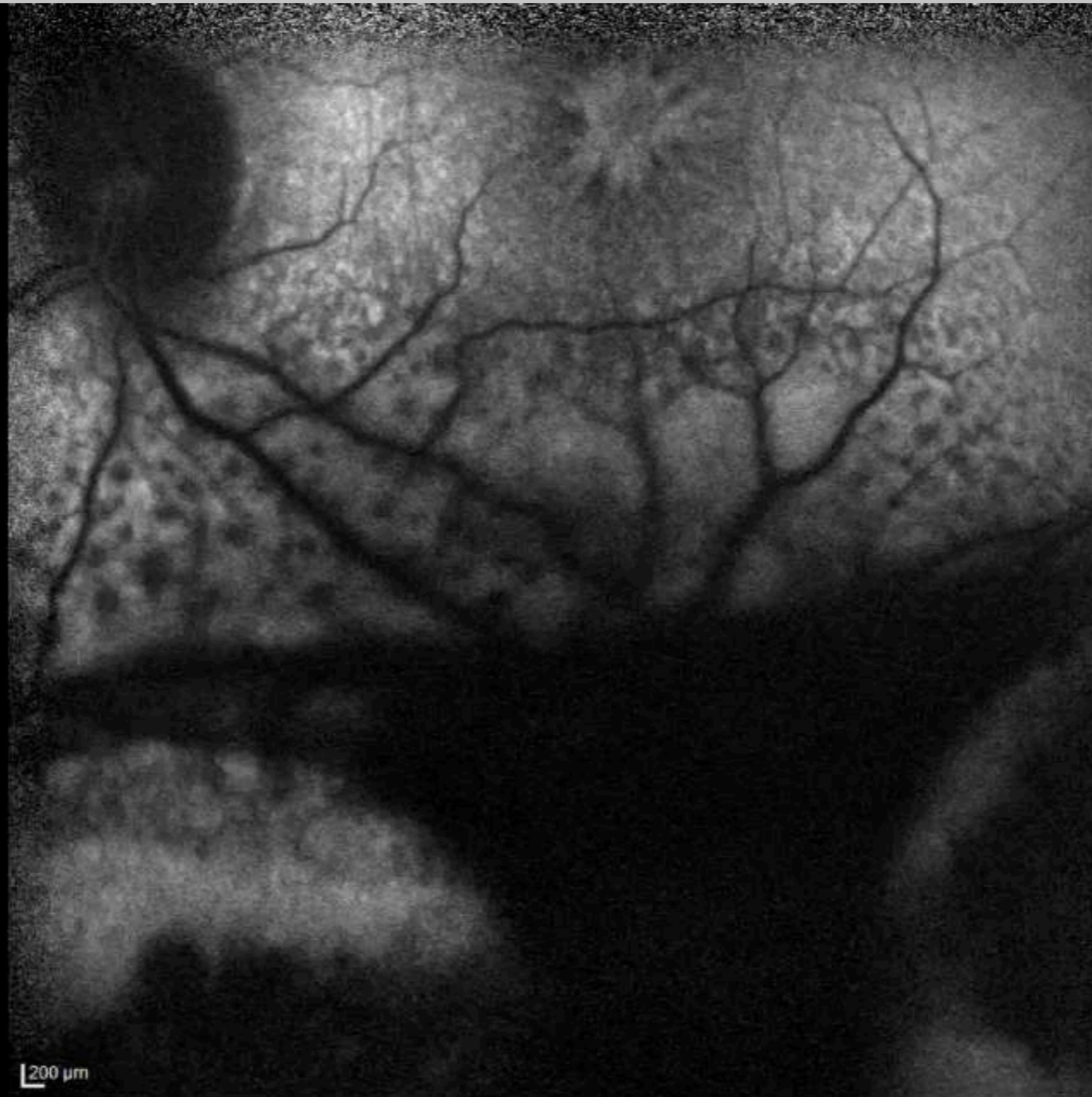
- Males
- Bilateral, peripheral+ foveal retinoscisis
- Recurrent vitreous hemorrhages
- RRD

8 mo old, M, Bilateral Vit Hem,



21y, M, VA: 0.3 (LE)

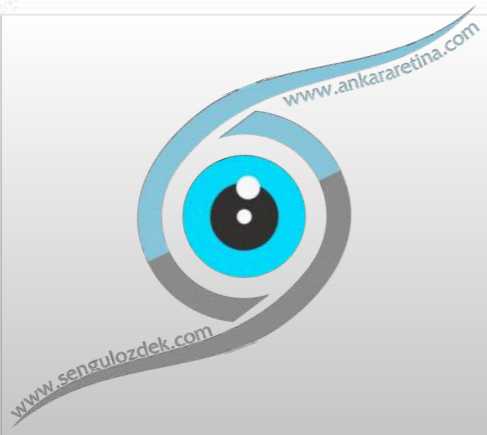
Bilat PPV 15 yr earlier, RE: FB



200  $\mu$ m

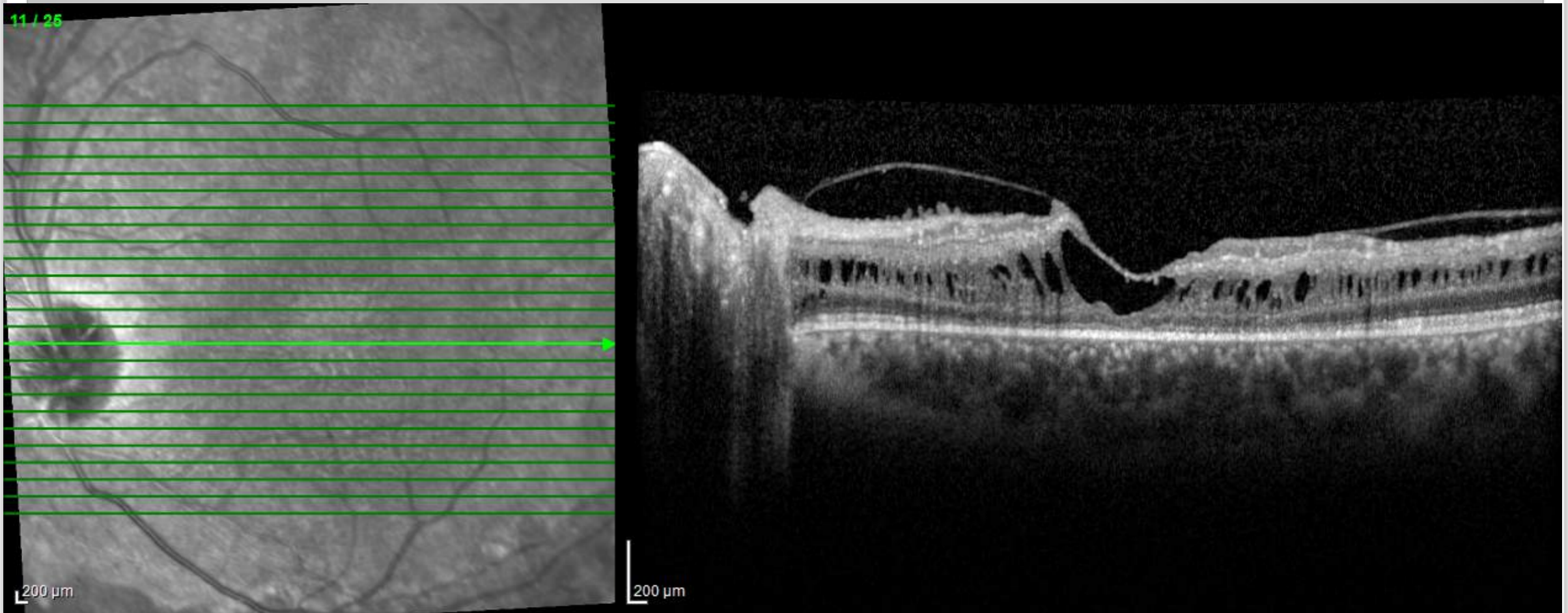
04.07.2014, OS  
BAF 30° ART(44)

HEIDELBERG  
ENGINEERING



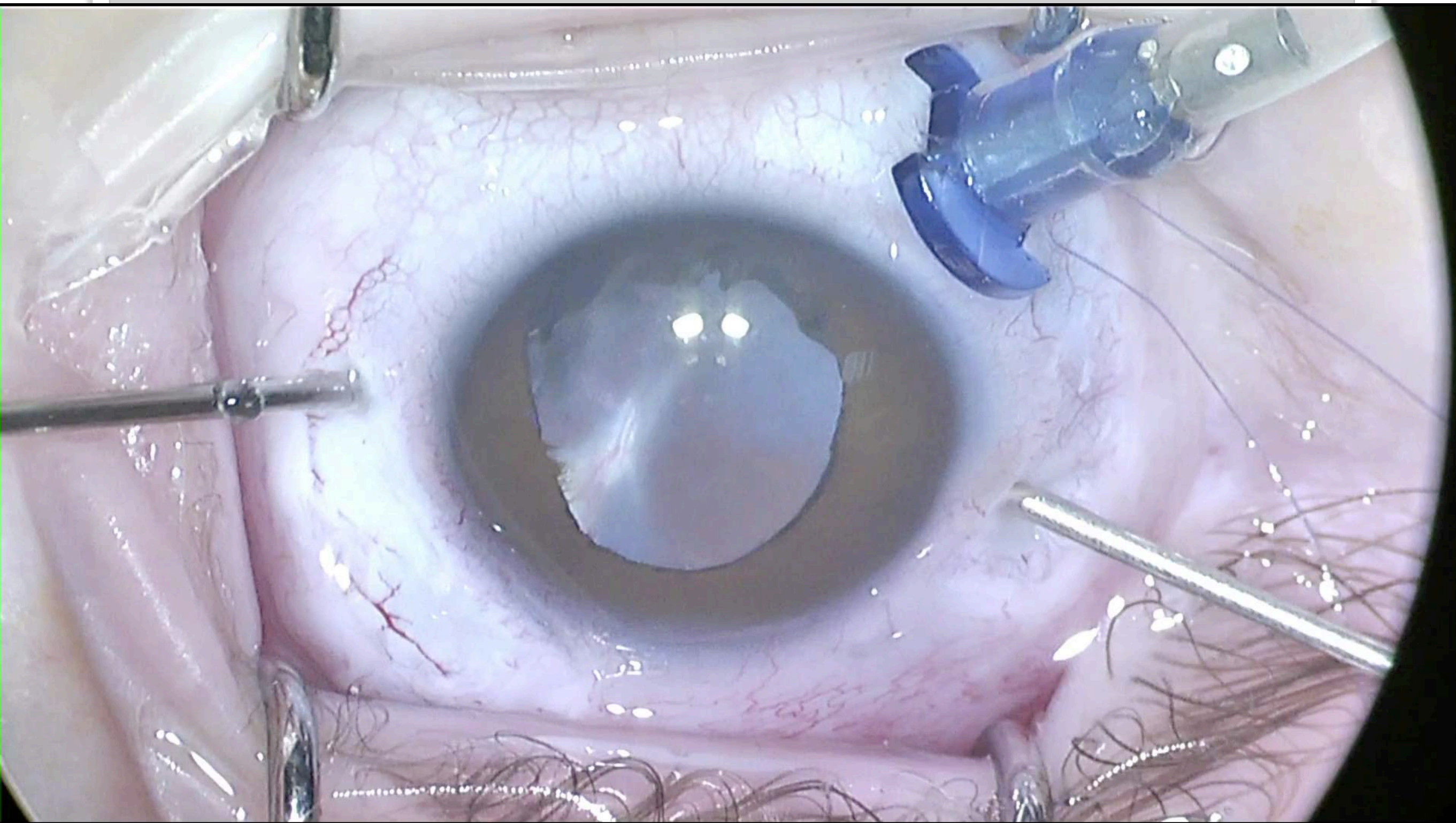
# Postop 5 mo

## VA: 0.4



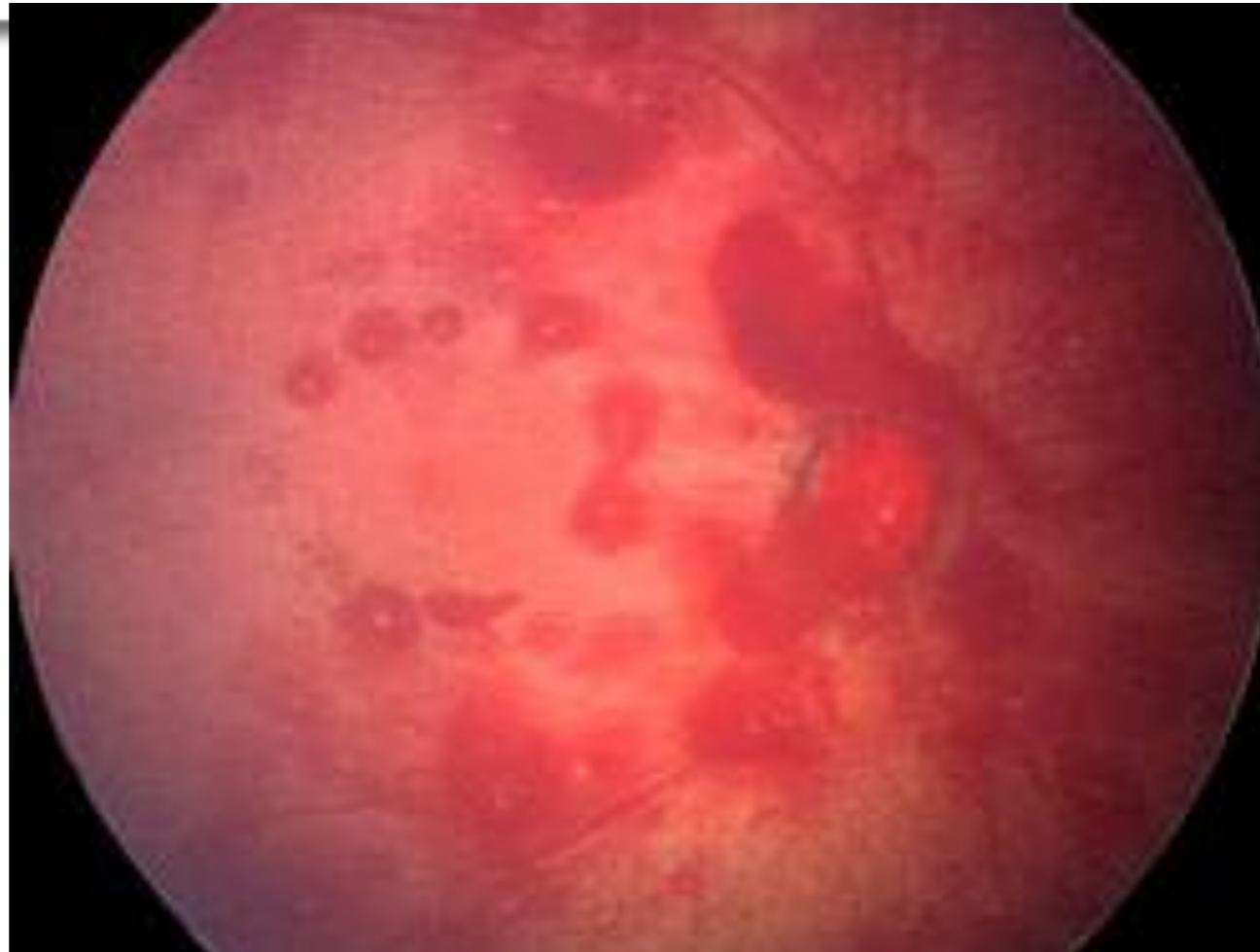
11 / 25  
200 μm  
200 μm  
15.12.2015, OS  
IR&OCT 30° [HS] ART(9) Q: 31

HEIDELBERG  
ENGINEERING





# PEDIATRIC TRAUMA

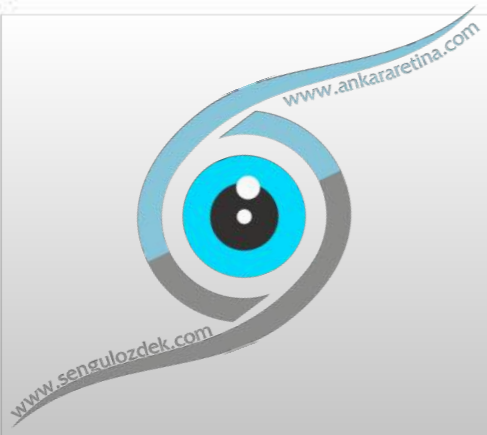




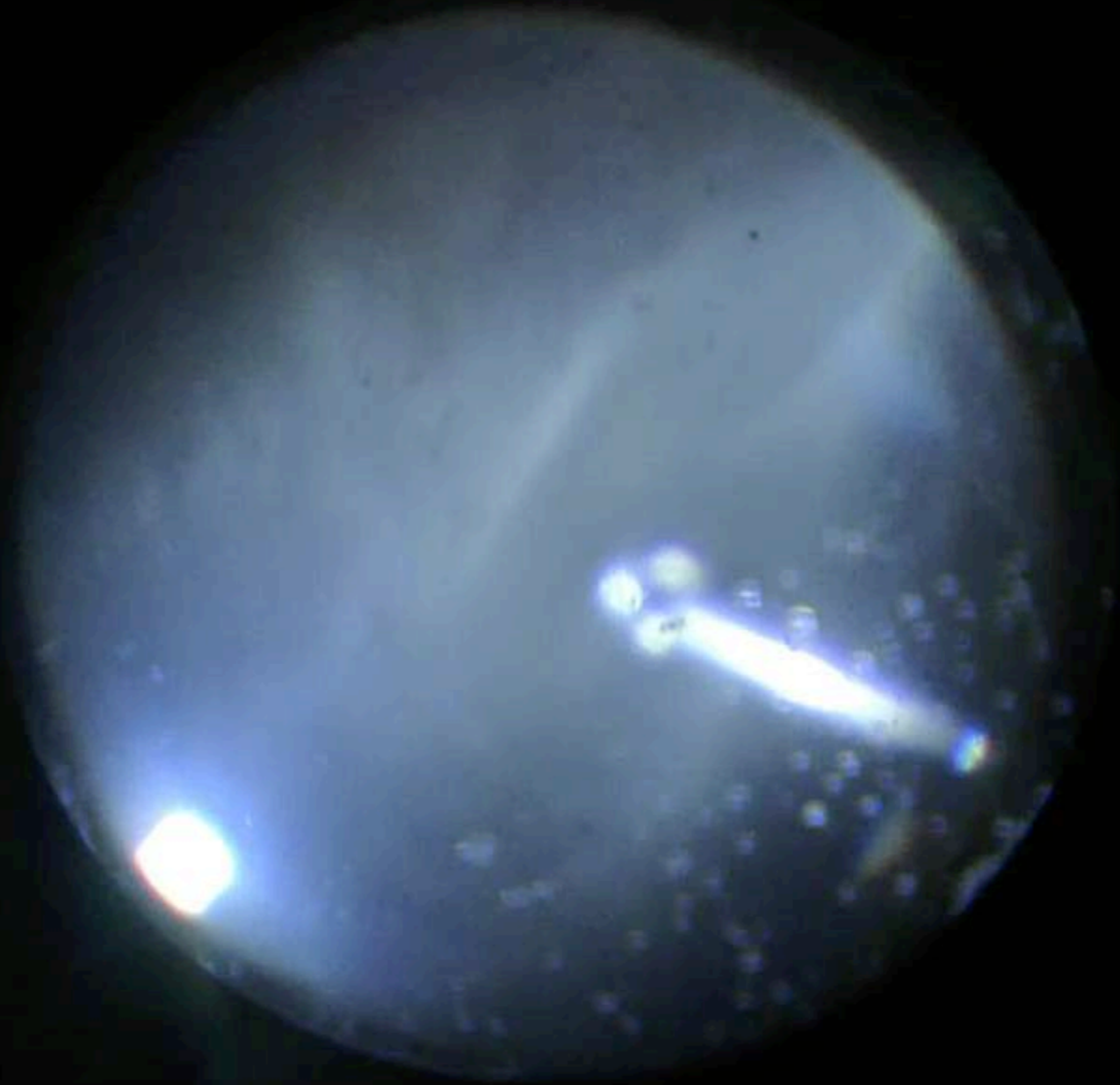
# TRAUMA

## Penetrating Injury

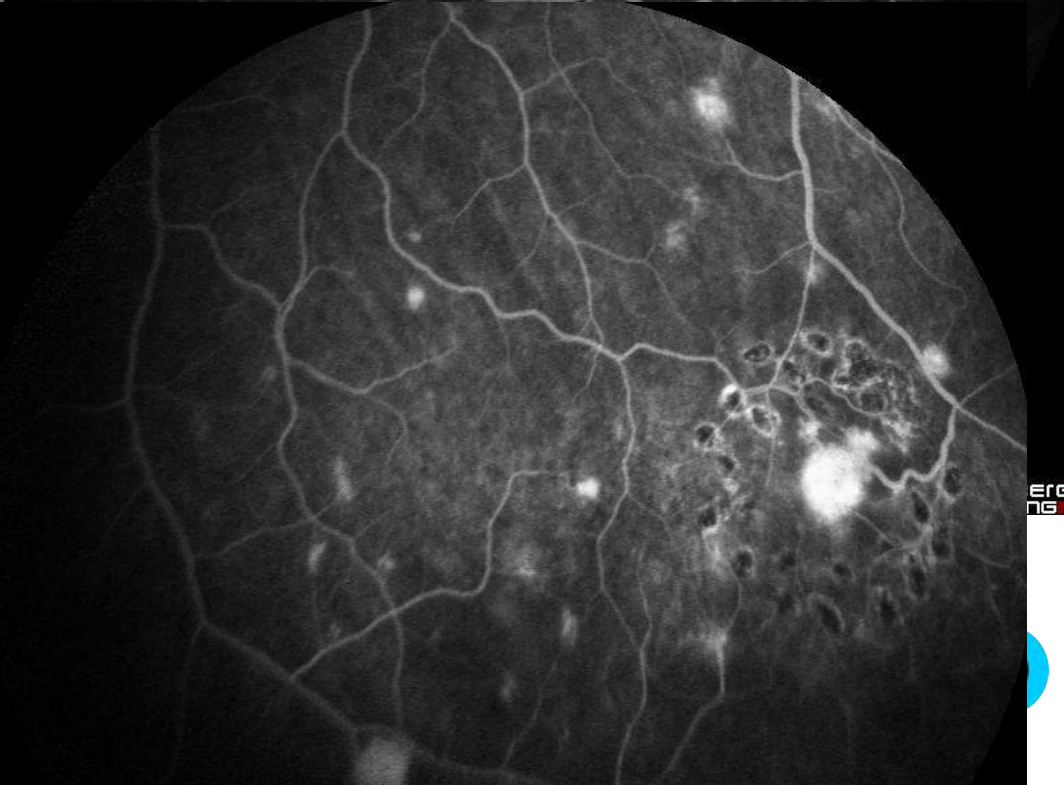
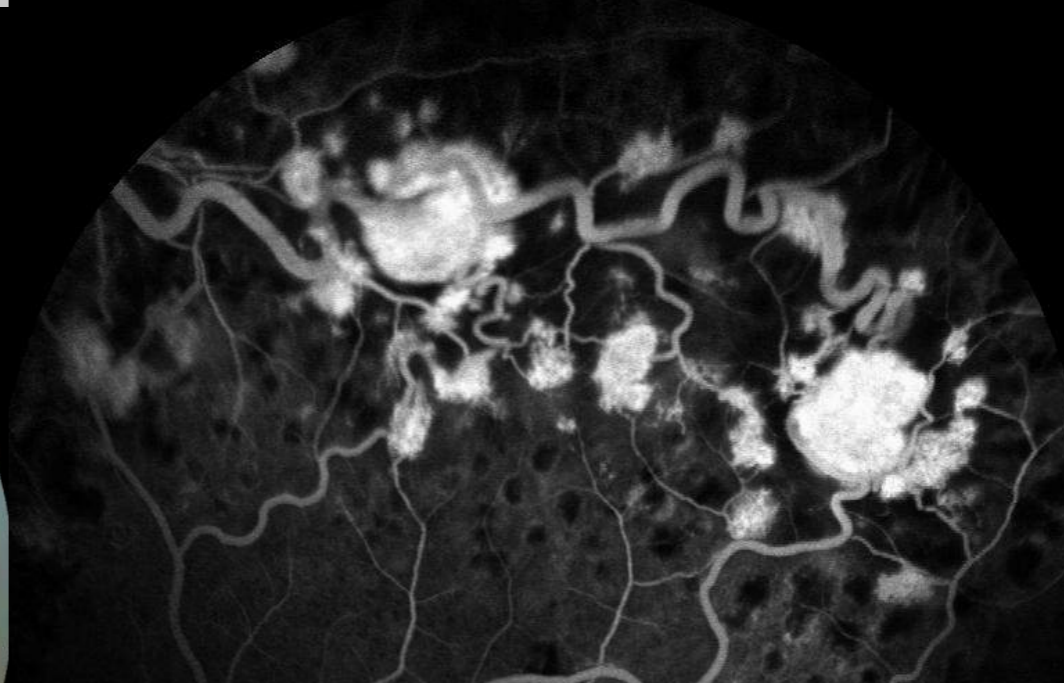
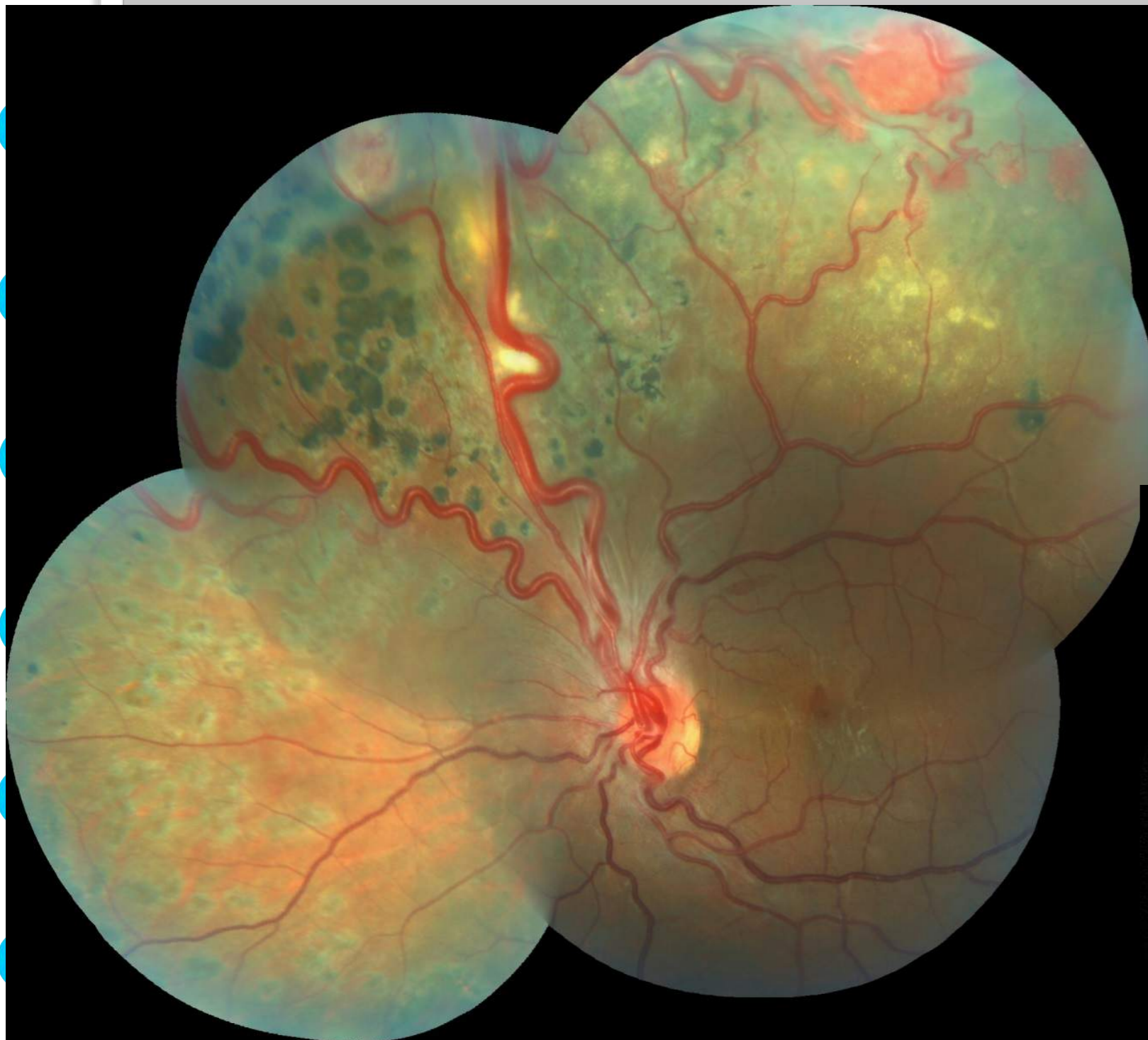
- Delayed diagnosis,
- Risk of Endophthalmitis high
- Wound healing: PVR risk high
- Follow up examination are really difficult, assessment is hard.
- Ambliopia!



# SHAKEN BABY



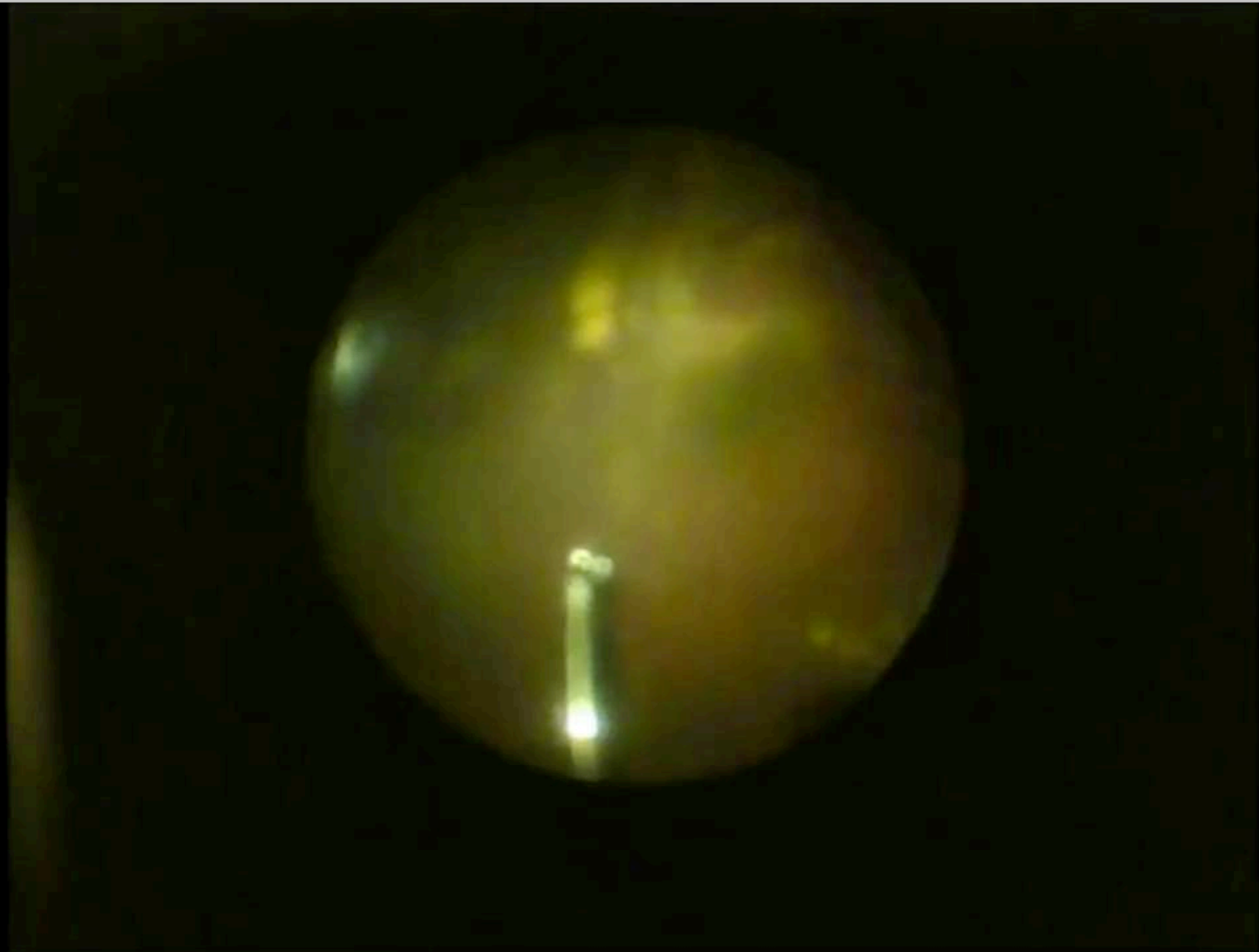
# RETINAL HEMANGIOBLASTOMA



14 y old, Cerebellar hemangioma (operated)

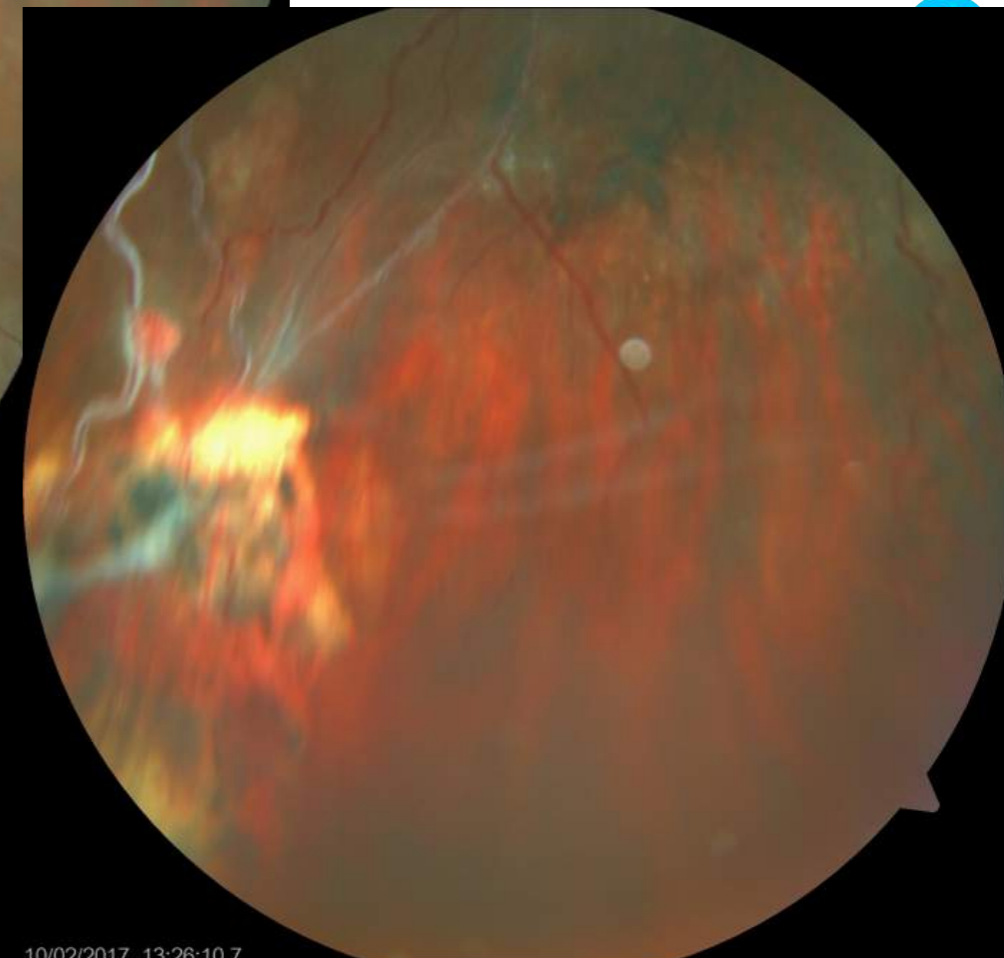
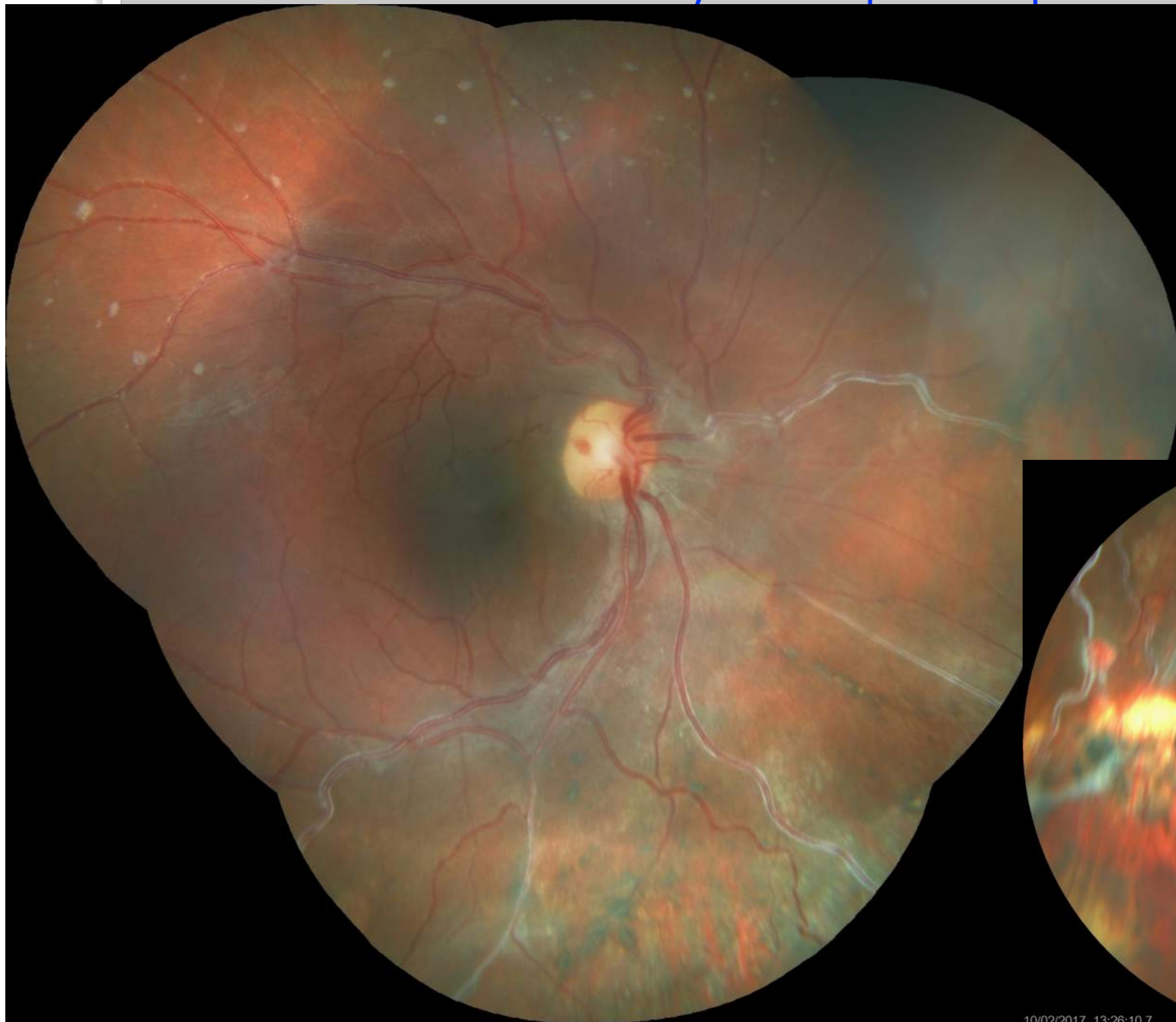
VA: HM

Fundus: Vit Hem-TRD



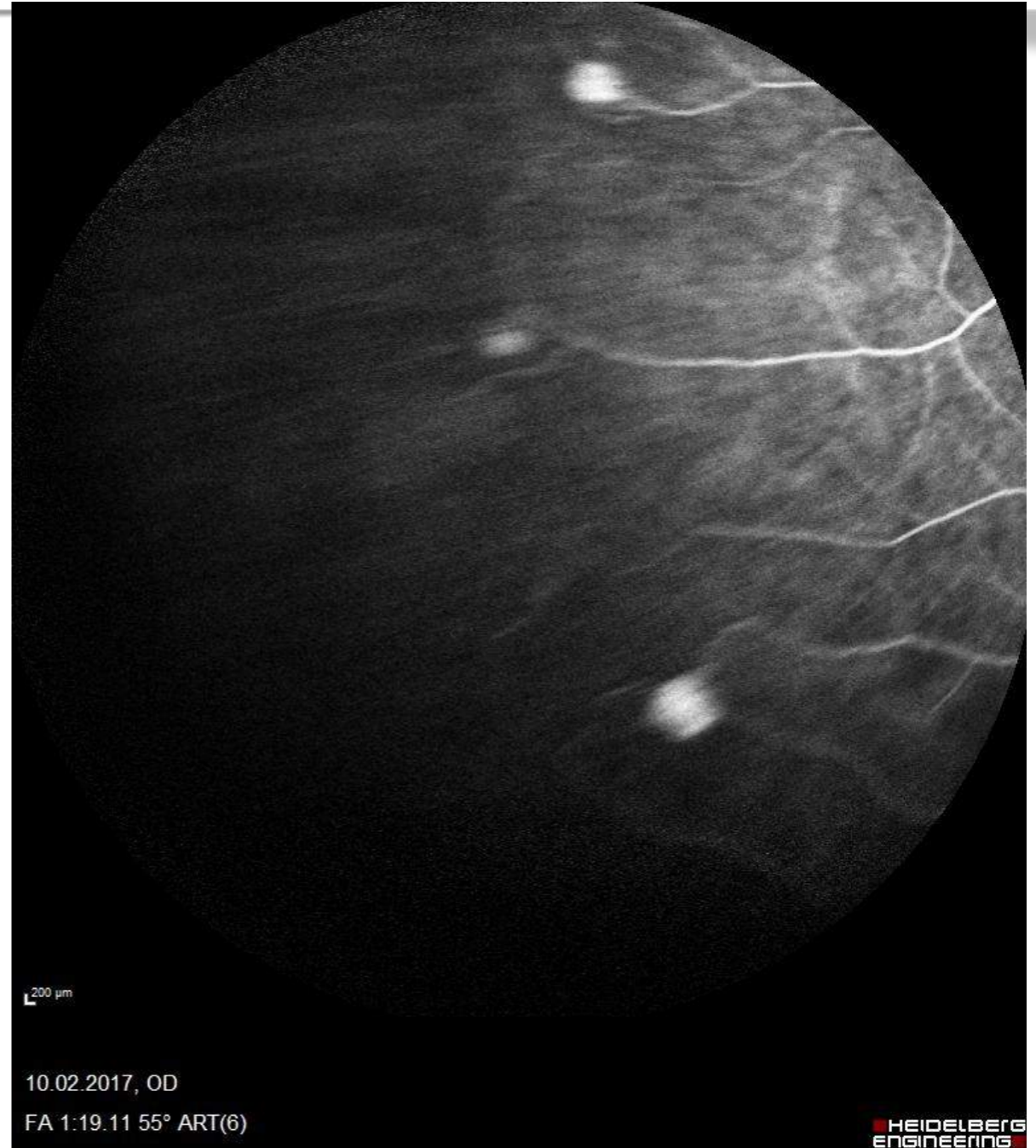
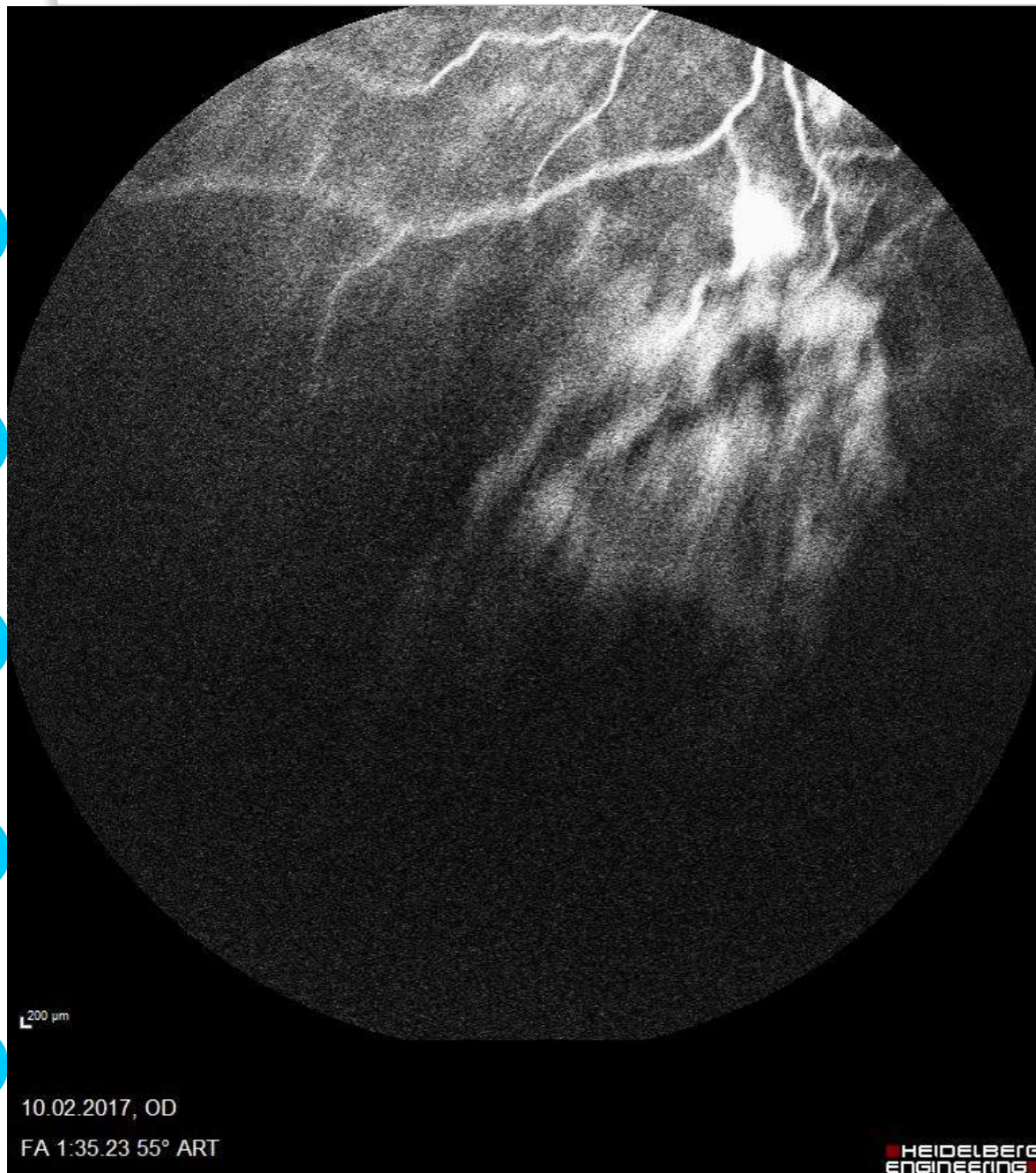
# RETINAL HEMANGIOBLASTOMA

3 years postop



# RETINAL HEMANGIOBLASTOMA

Follow ups with FA every 4-6 months



# SURGERY IS AN ART



[www.sengulozdek.com](http://www.sengulozdek.com)  
[sengulozdek@gmail.com](mailto:sengulozdek@gmail.com)

 **Prof. Dr.**  
**Şengül ÖZDEK**  
[www.ankararetina.com](http://www.ankararetina.com)