MULTIPLE RETINAL HEMANGIOBLASTOMAS IN VON HIPPEL LINDAU DISEASE: CASE REPORT USING WIDE FIELD ANGIOGRAPHY

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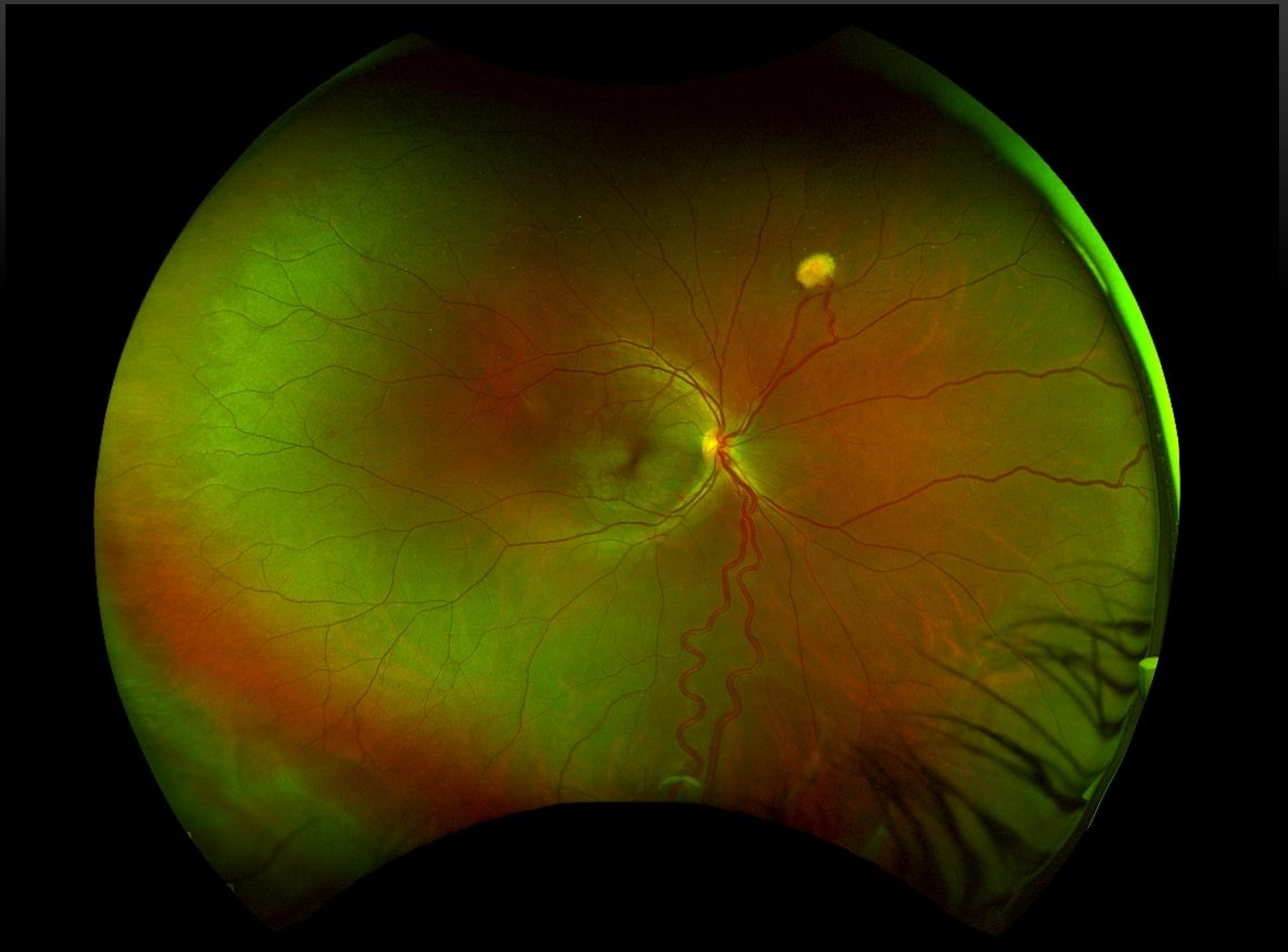
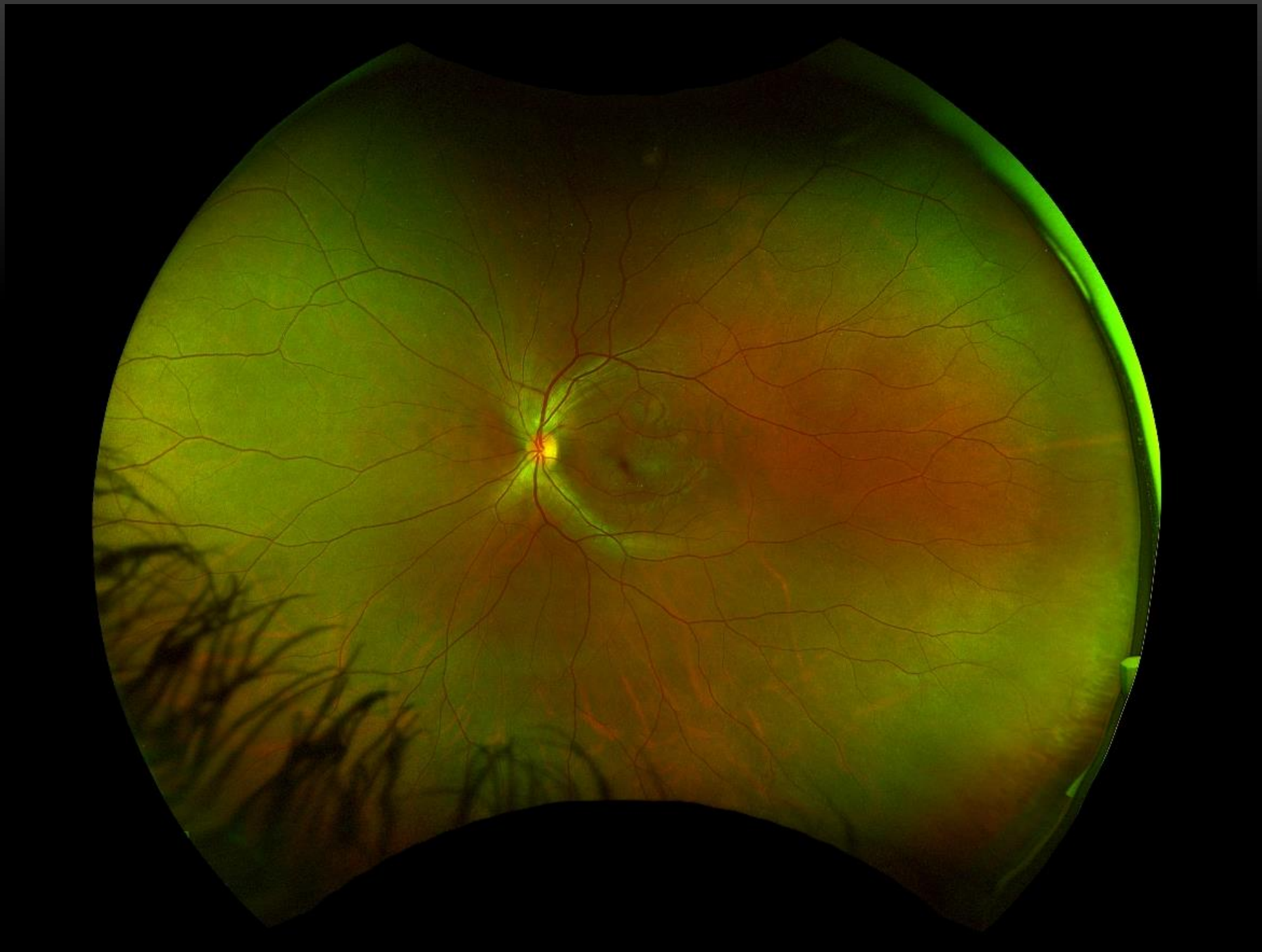
1. FELLOW OF THE RETINA AND VITREOUS SERVICE
2. HEAD OF THE RETINA AND VITREOUS SERVICE

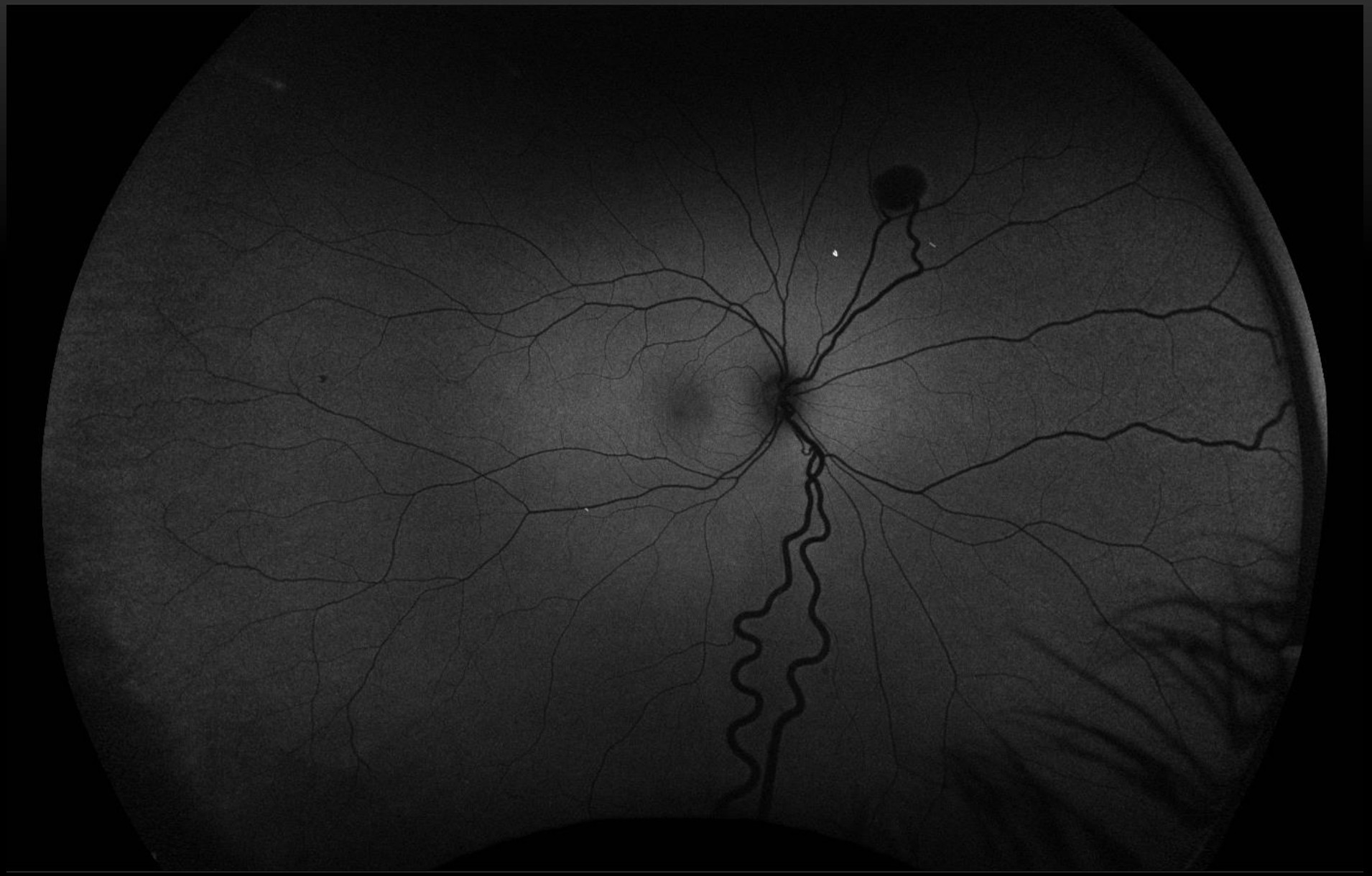
06/25/19

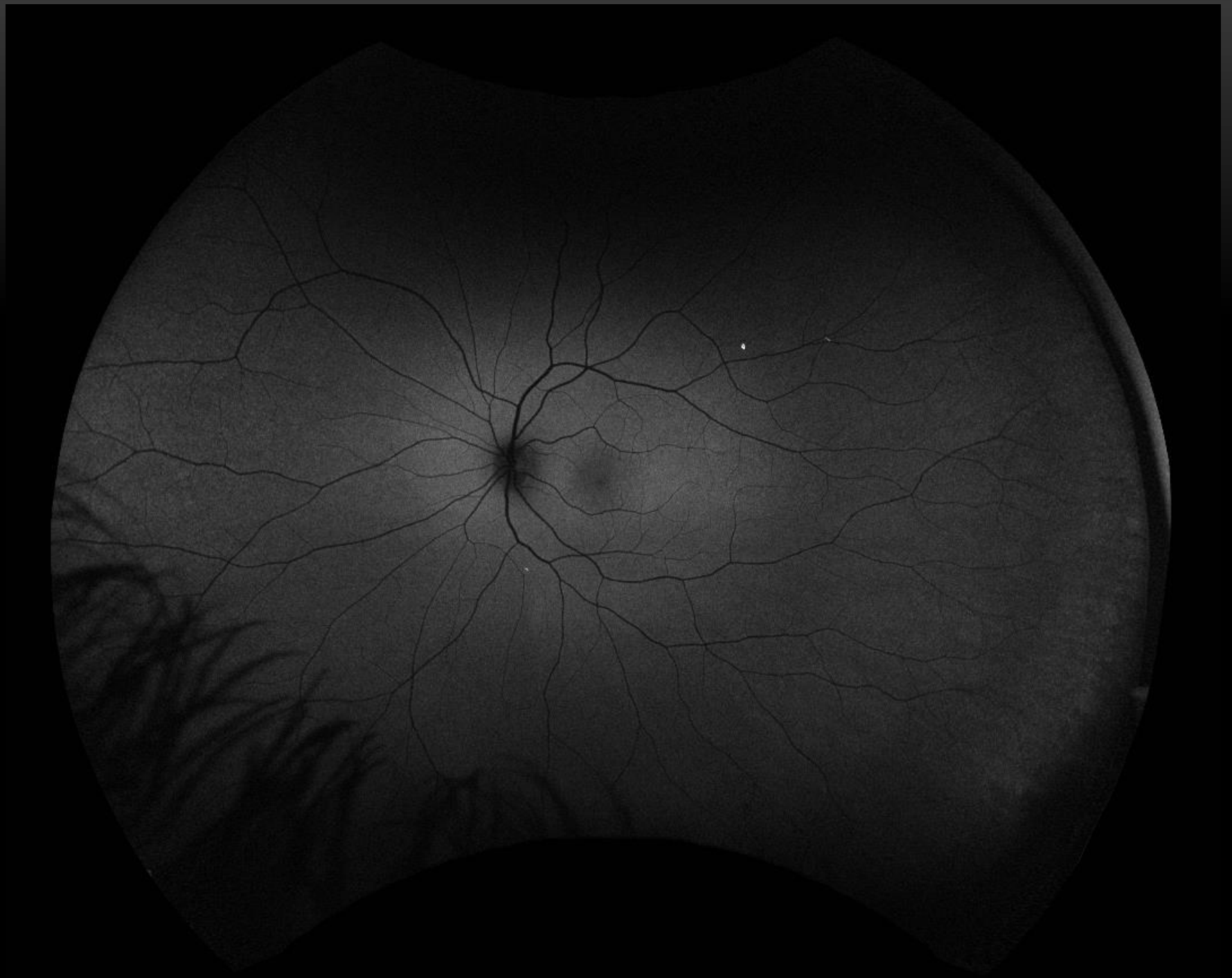
- 22-years-old, white, female
- CC: Patient referred by neurologist for ophthalmology evaluation. Asymptomatic.
- PMx: systemic disease diagnosed 1 year ago
- POx: none
- FMx: sister with the same disease

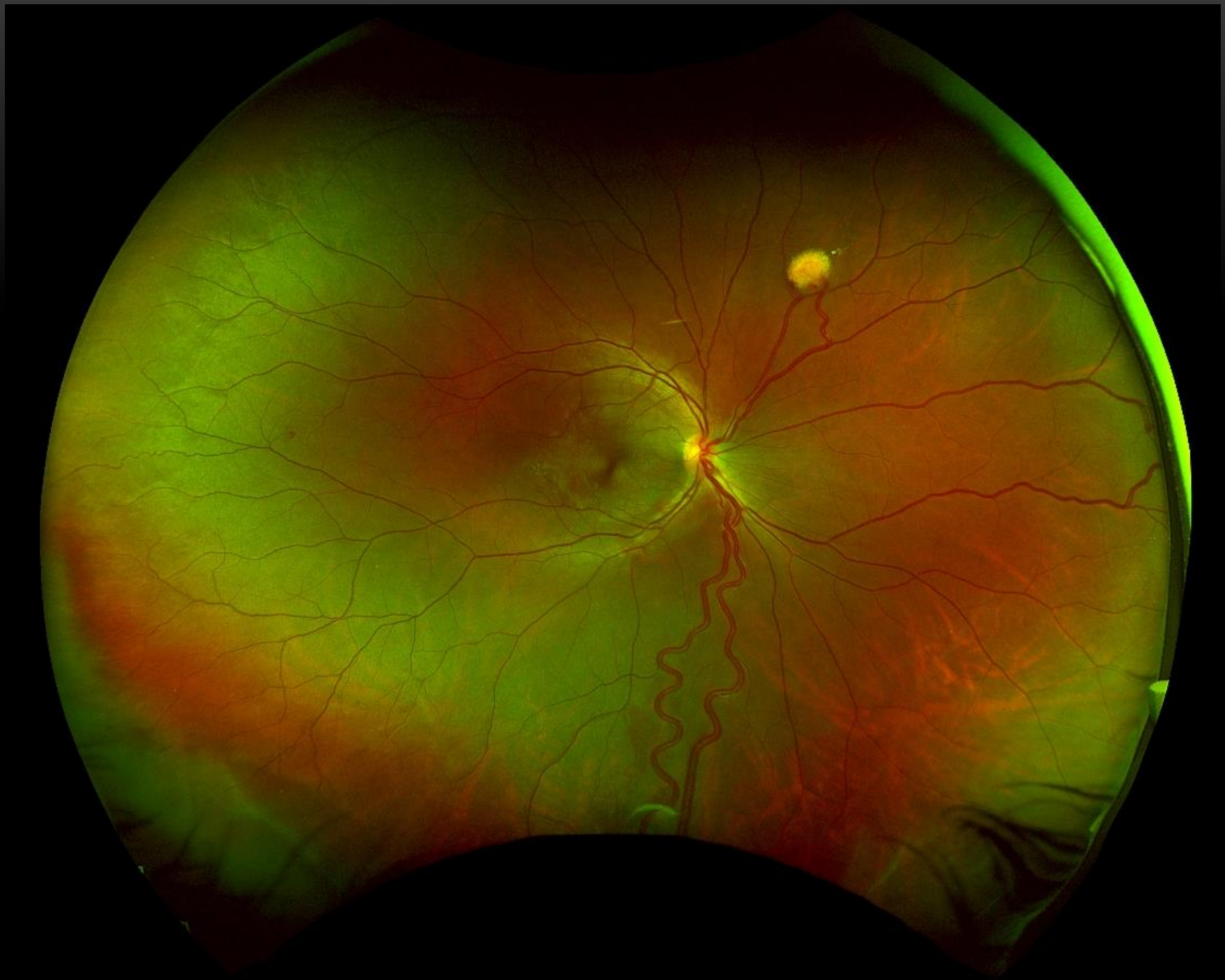
	OD	OS
BCVA	20/20	20/20
BIO	Clear cornea; quiet AC ; clear lens	Clear cornea; quiet AC ; clear lens
IOP	14	14
FUNDUS	Photos	Photos

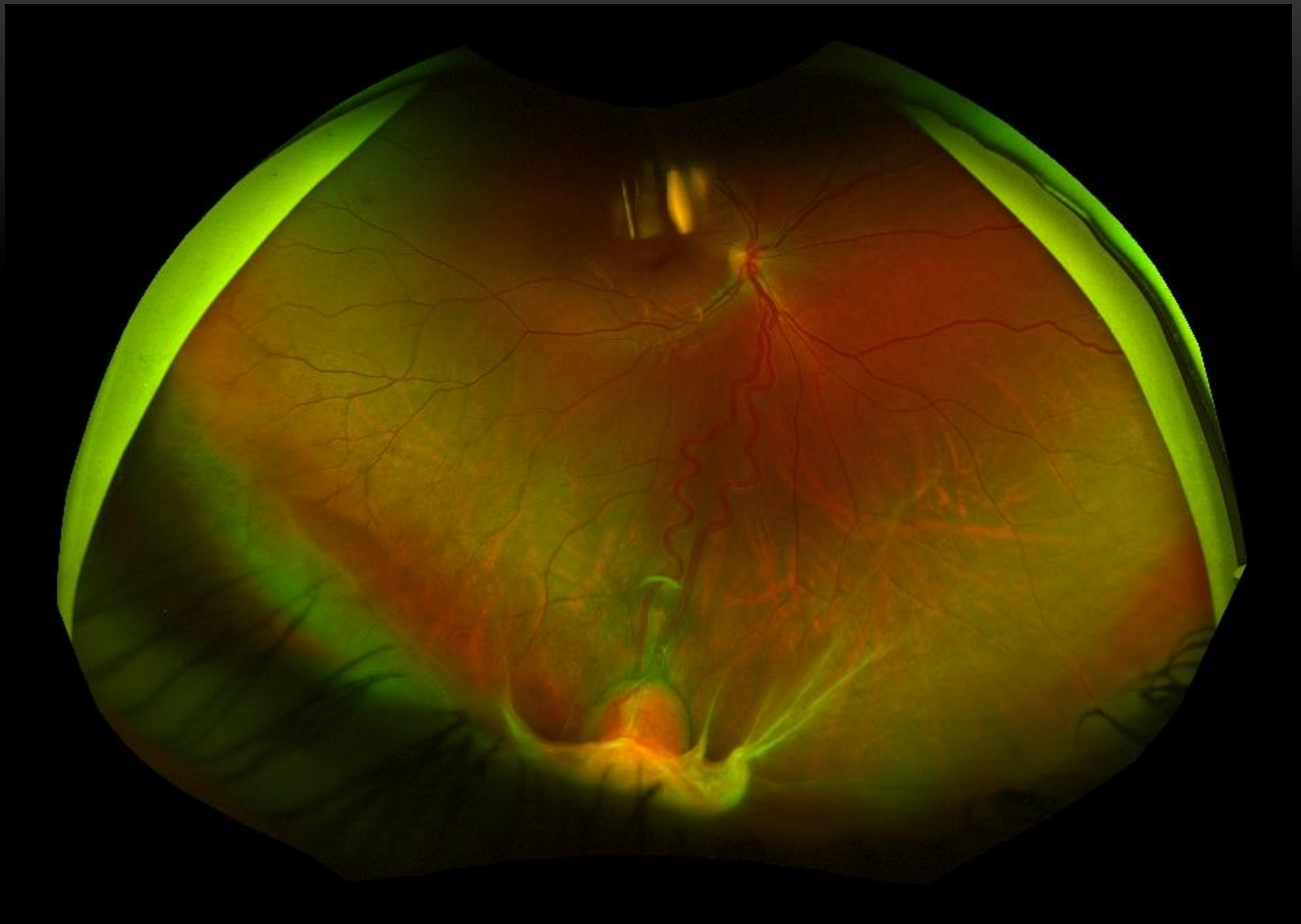


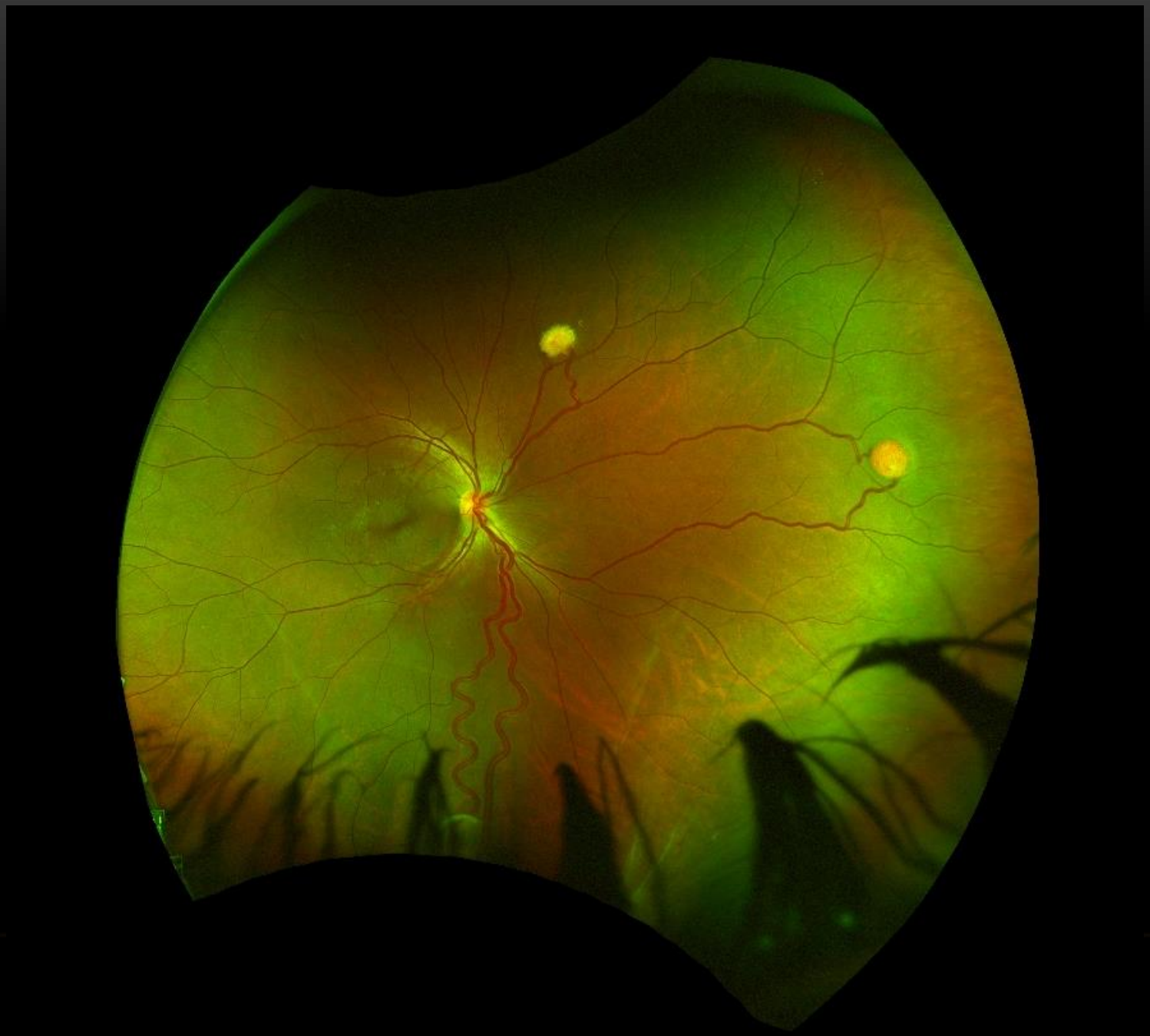


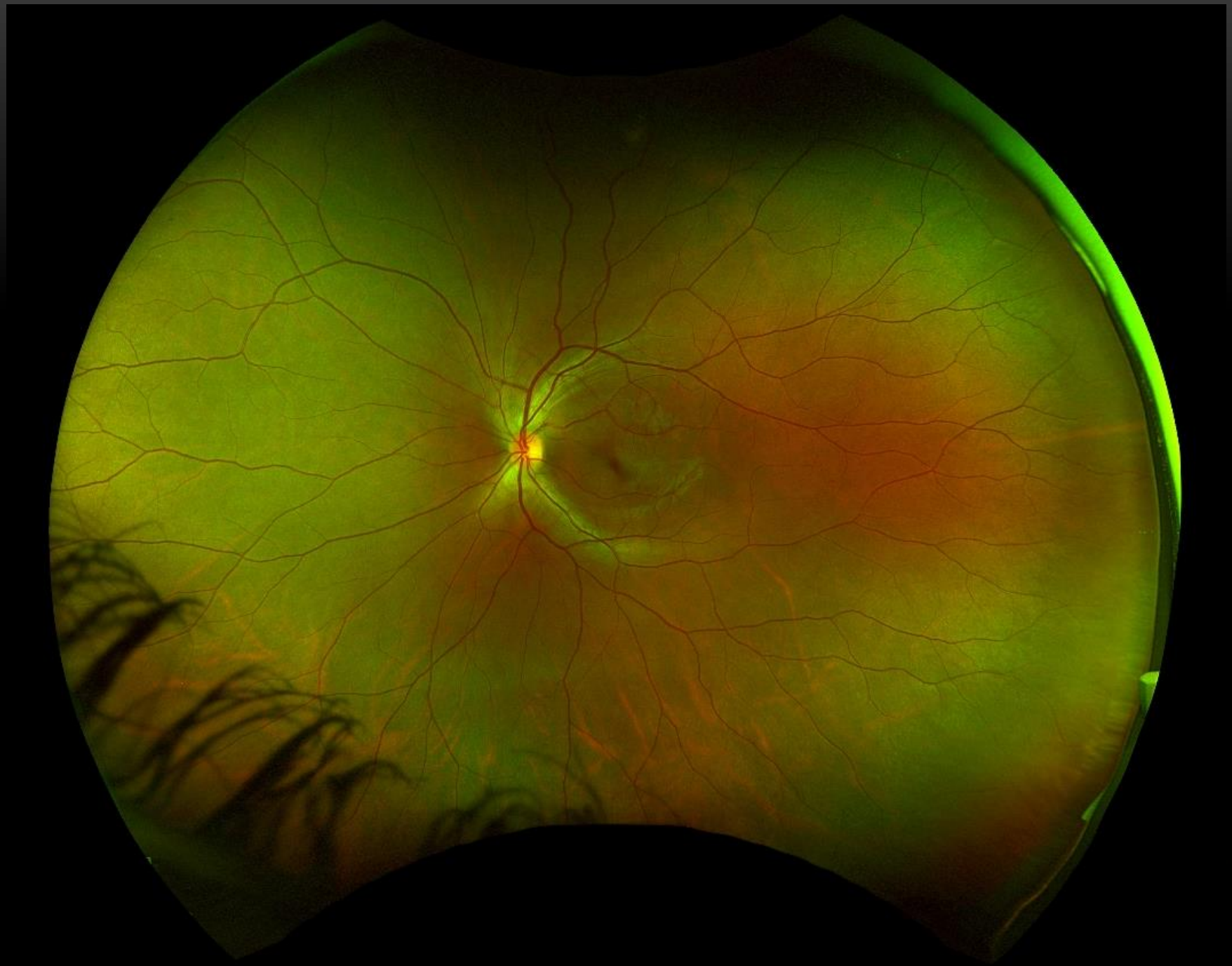


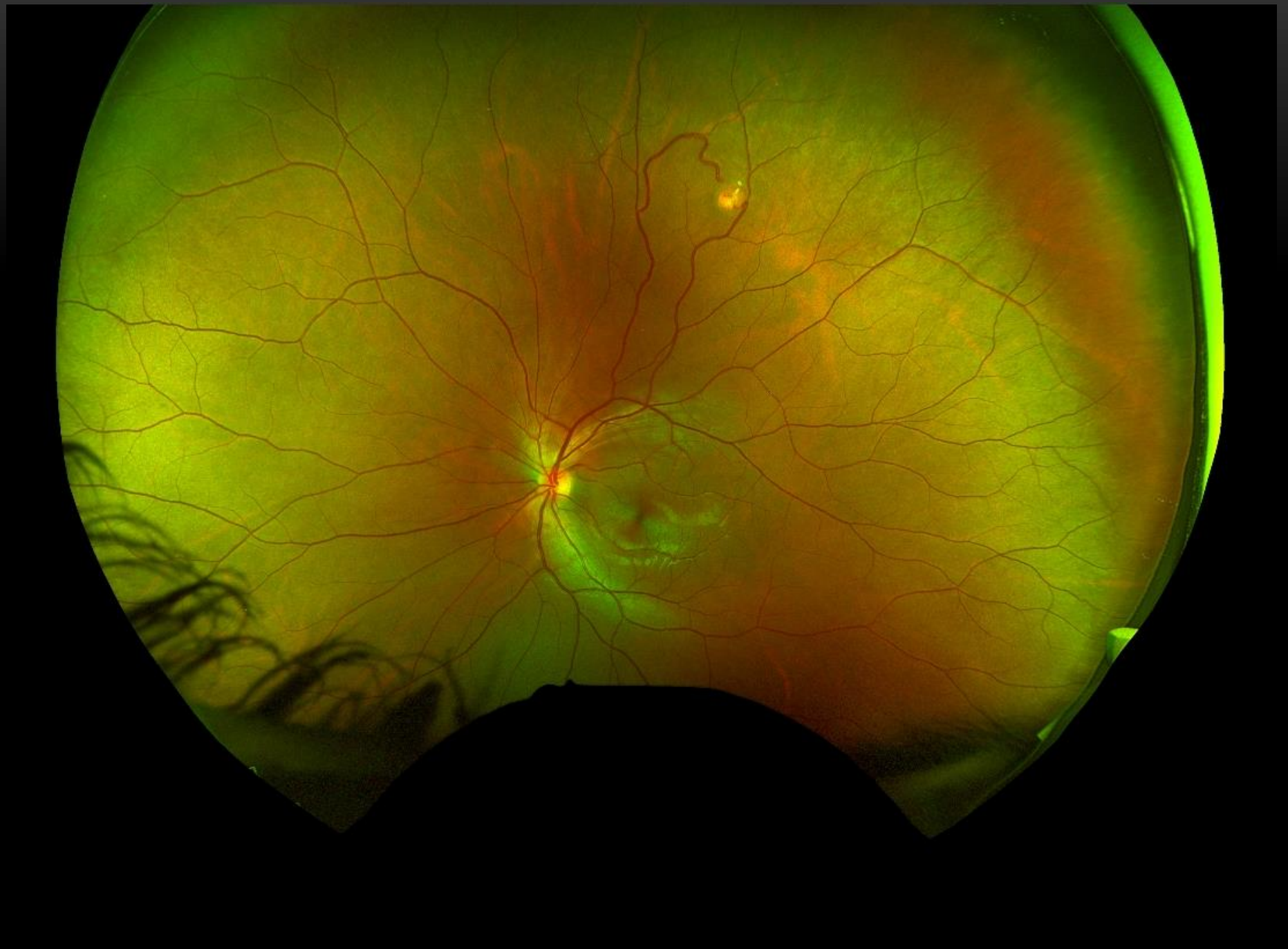


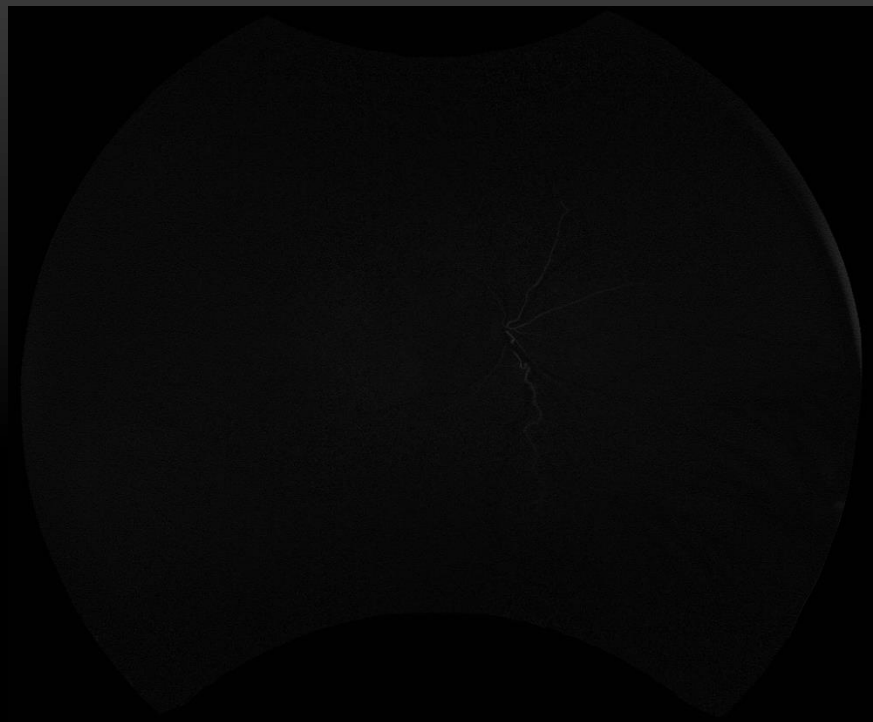
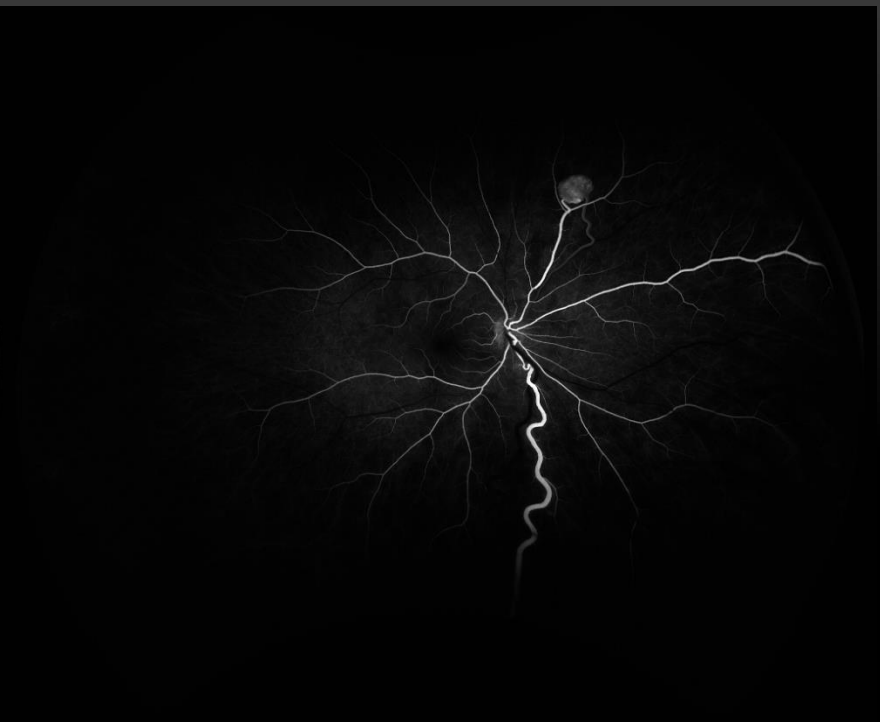
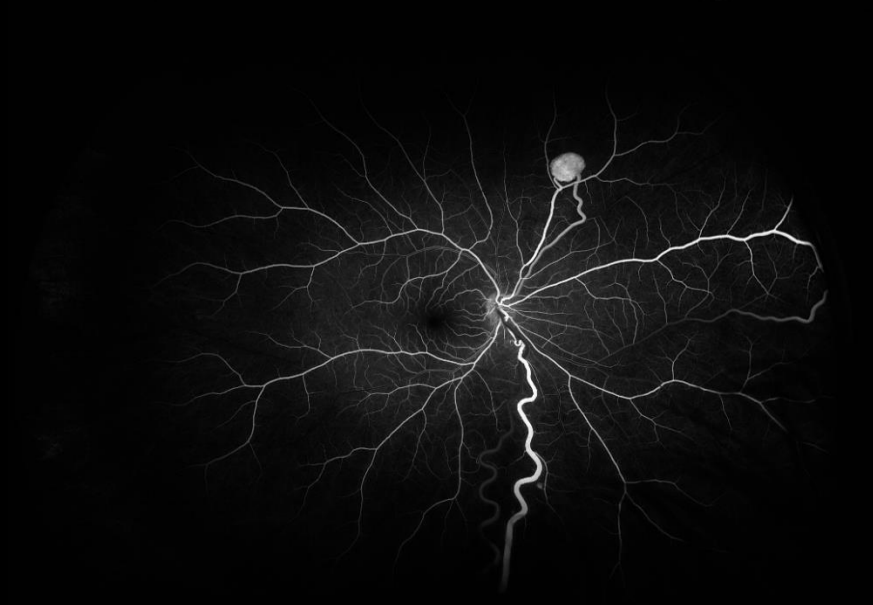
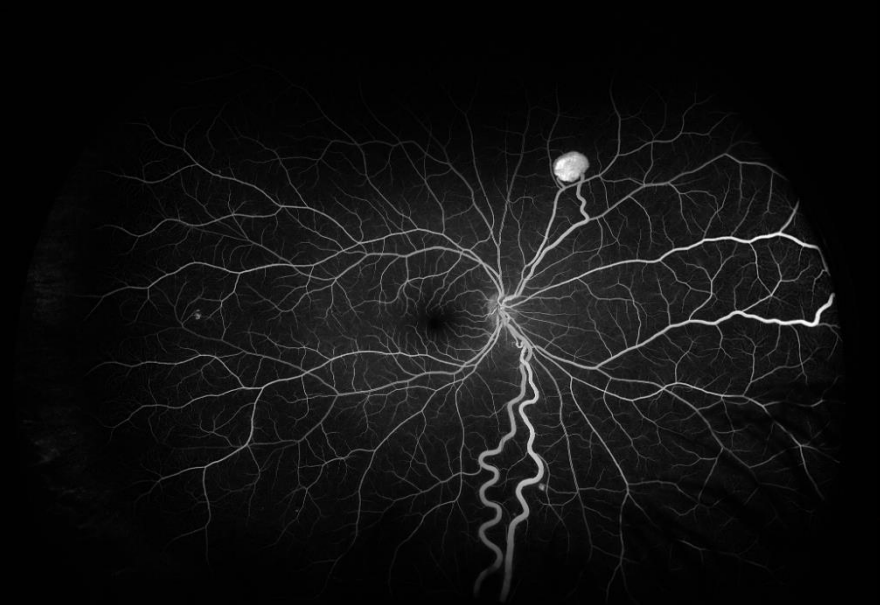


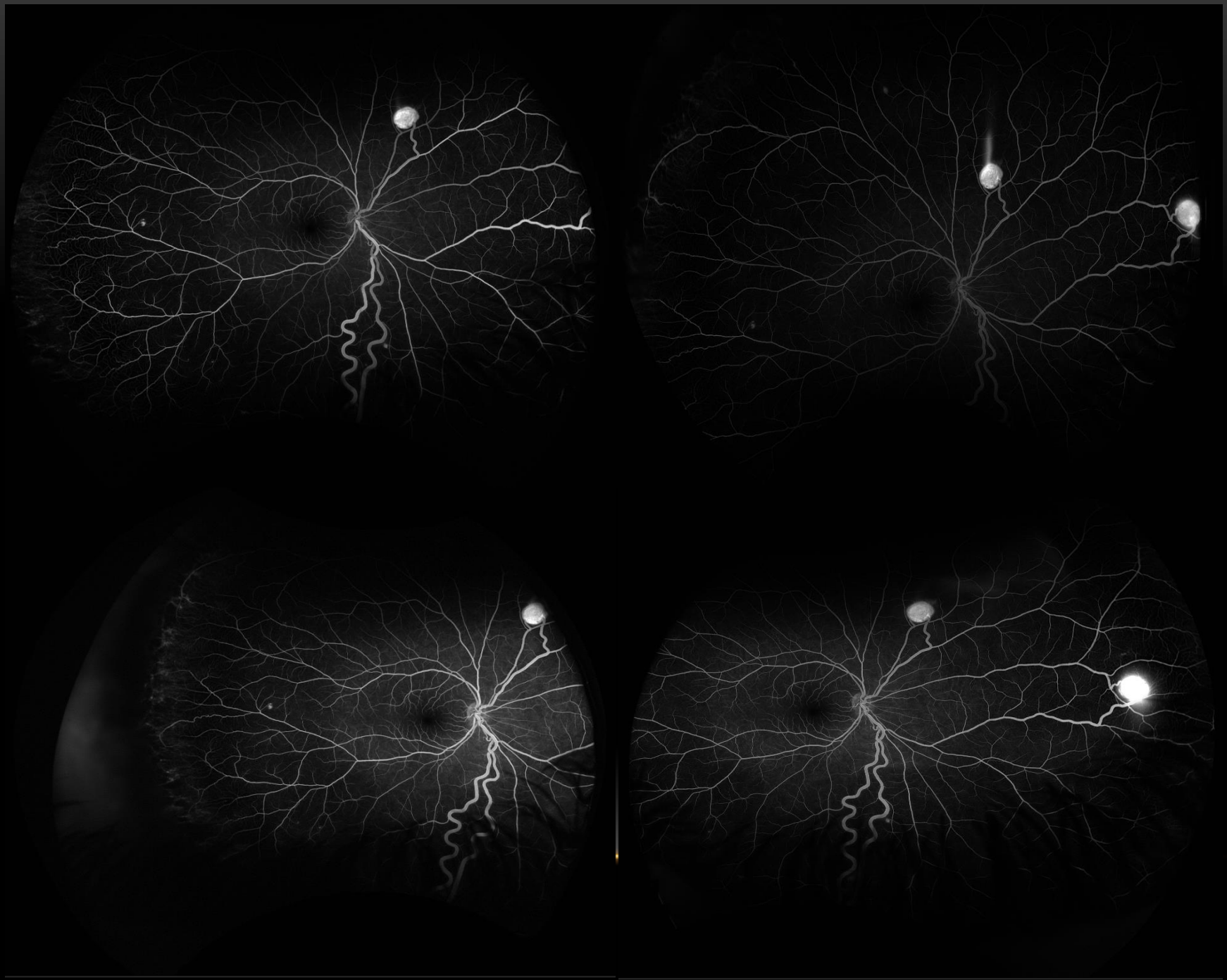


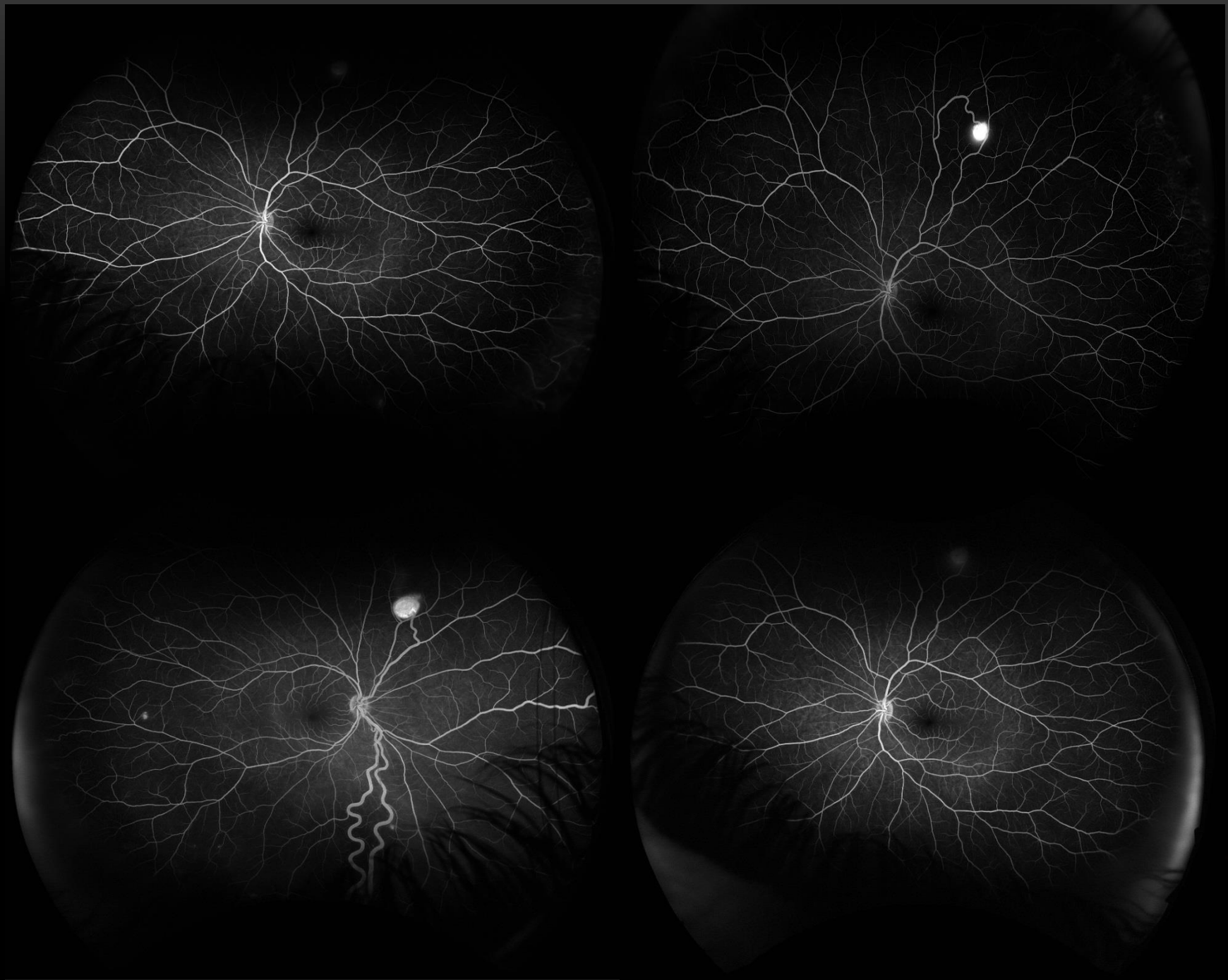


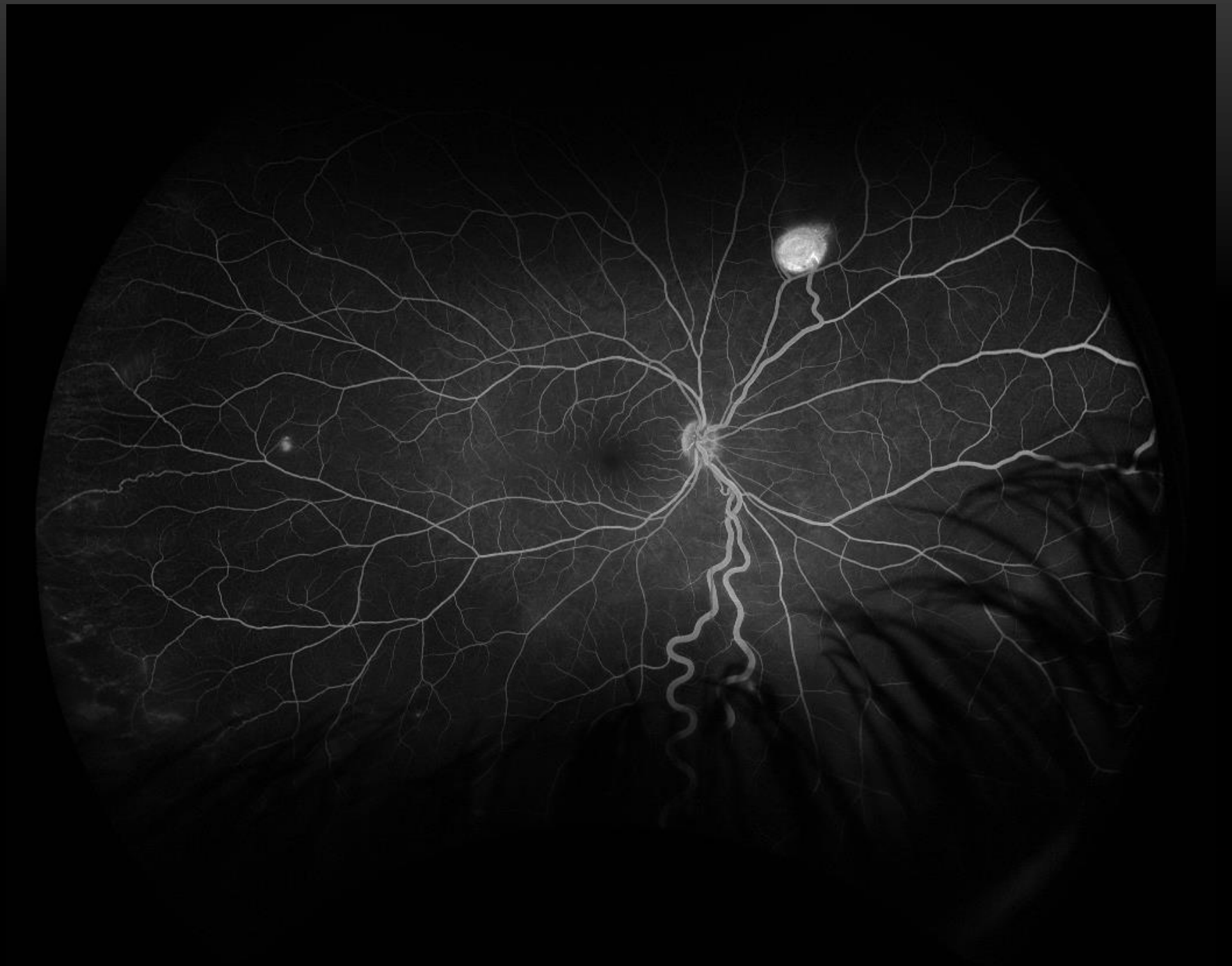


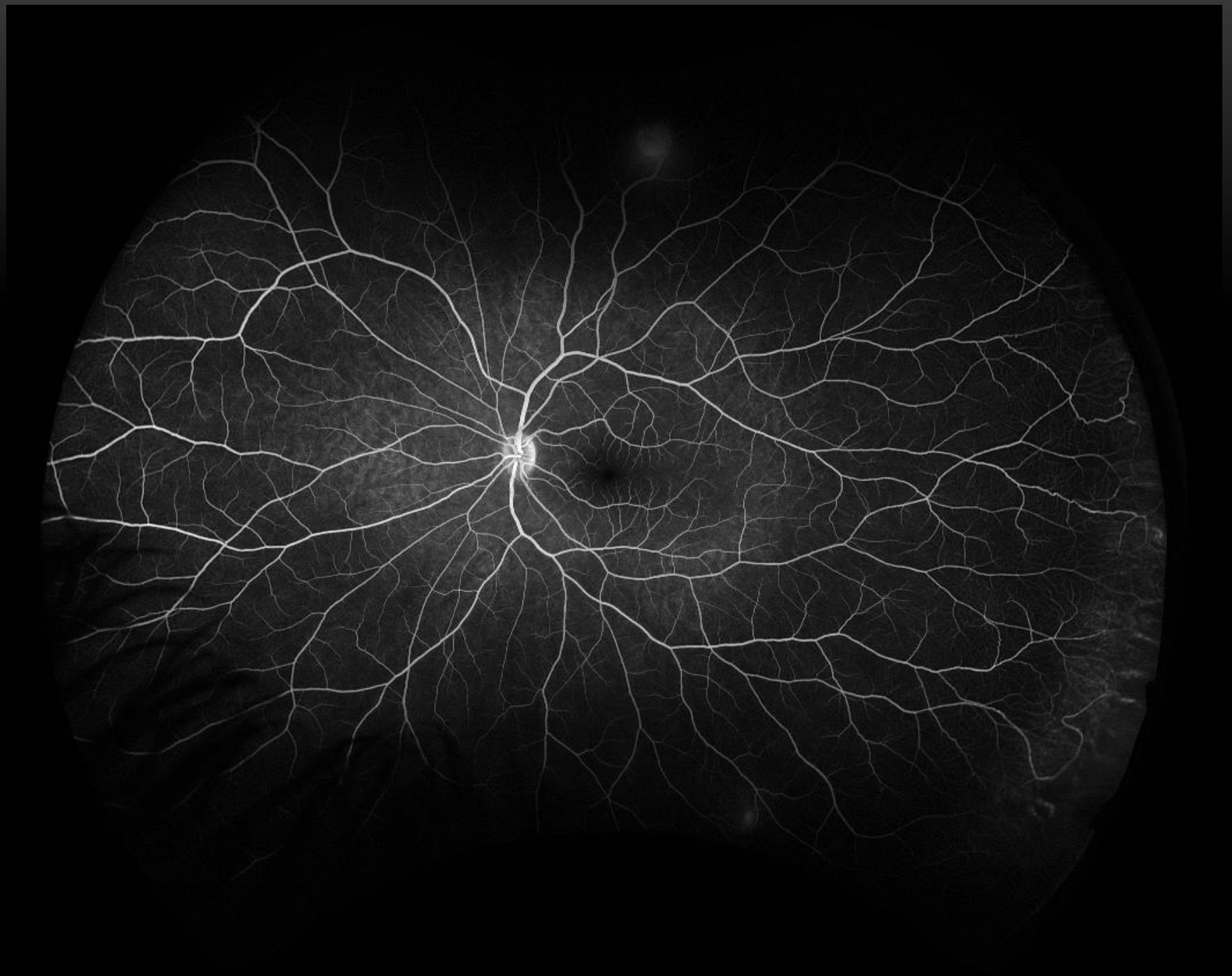




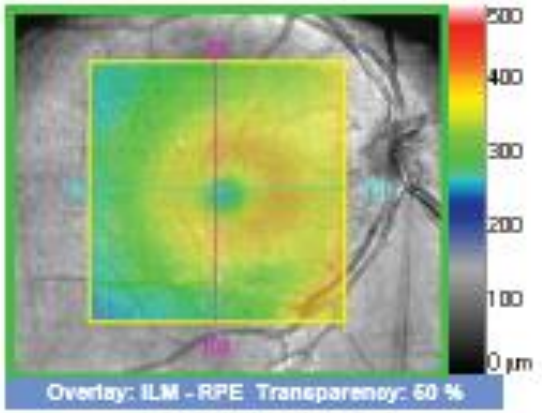




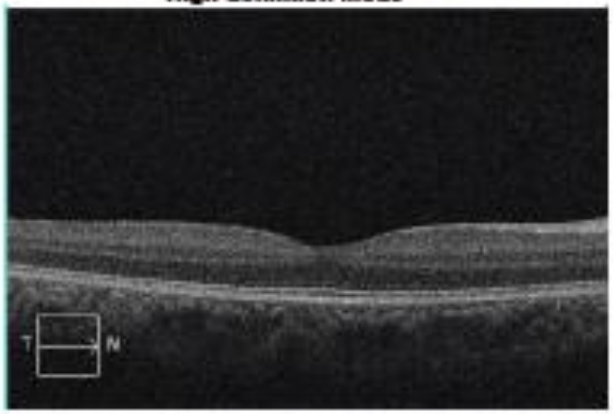


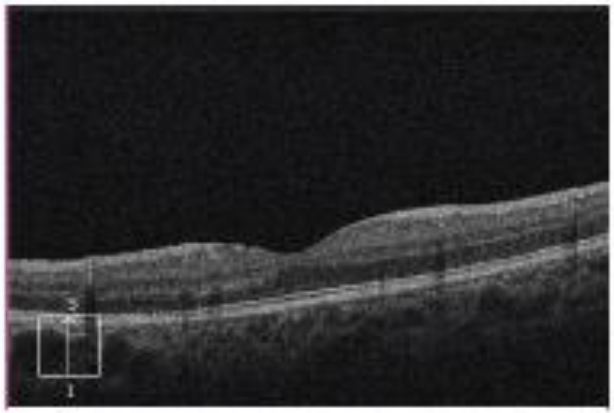
Macula Thickness : Macular Cube 200x200

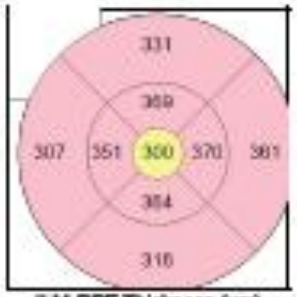
OD ● ○ OS



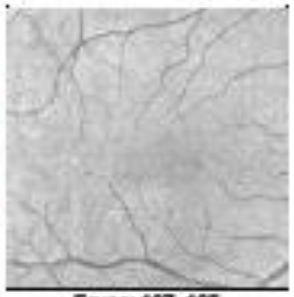
High-definition mode



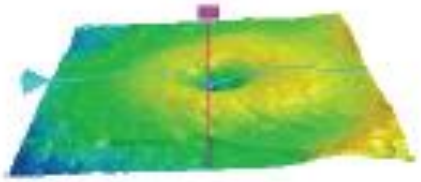




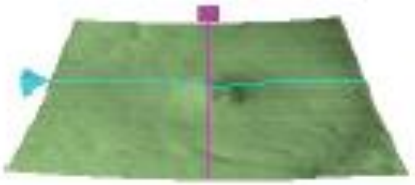
ILM-RPE Thickness (µm)



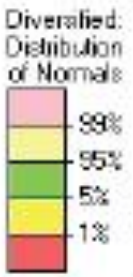
Fovea: 107, 105

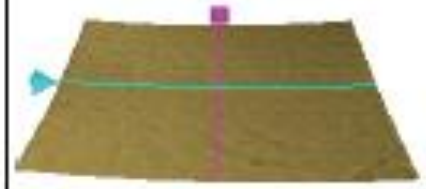


ILM - RPE



ILM



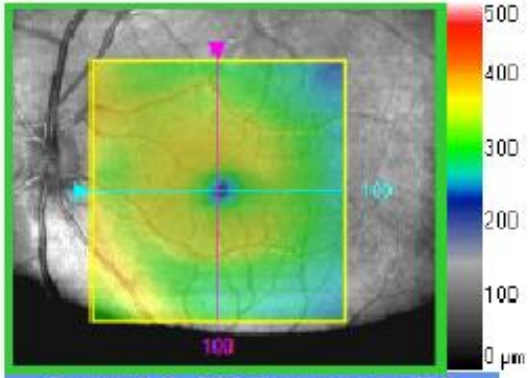


RPE

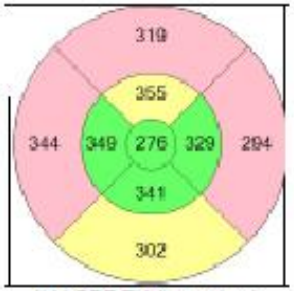
⚠	Central Subfield Thickness (µm)	Cube Volume (mm³)	Cube Average Thickness (µm)
ILM - RPE	300	11.8	329

Macula Thickness : Macular Cube 200x200

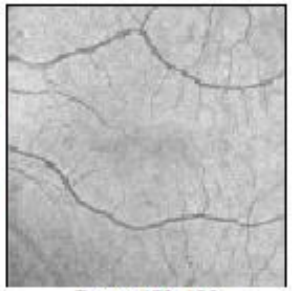
OD ○ | ● OS



Overlay: ILM - RPE Transparency: 50 %

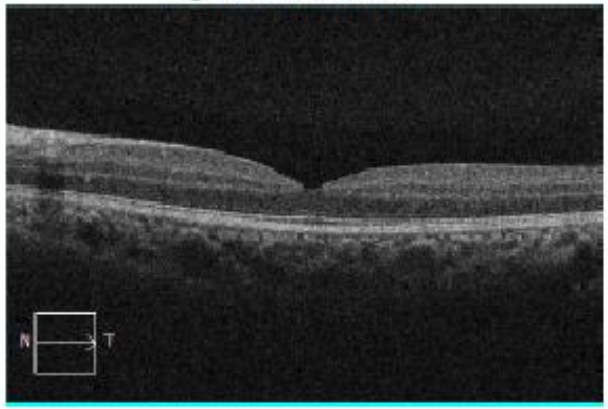


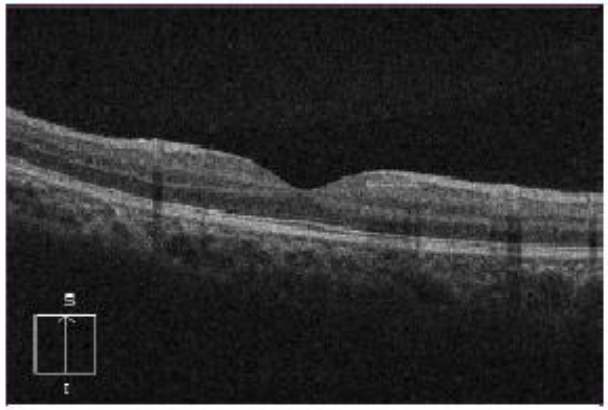
ILM-RPE Thickness (µm)

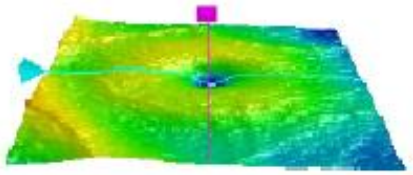


Fovea: 103, 100

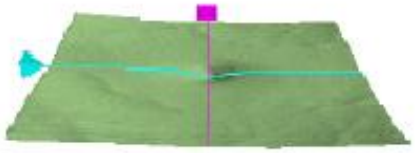
High-definition mode



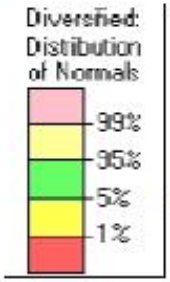


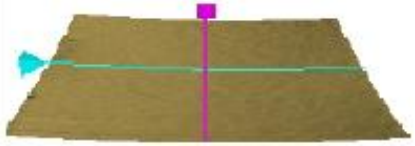


ILM - RPE



ILM



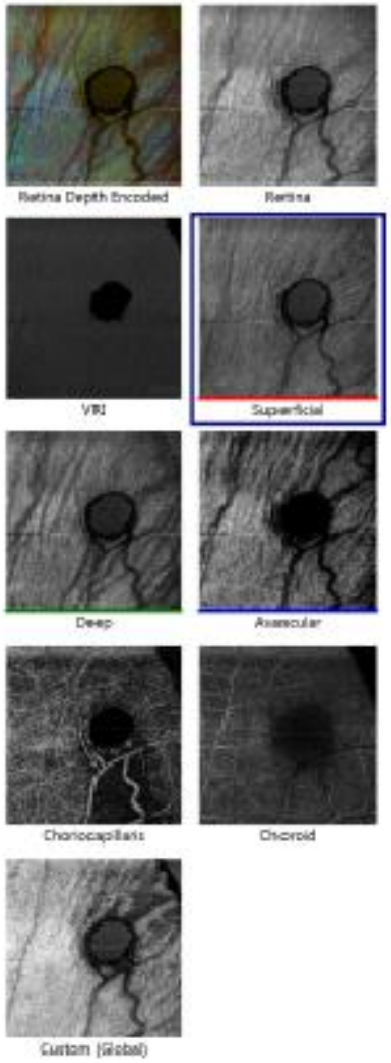


RPE

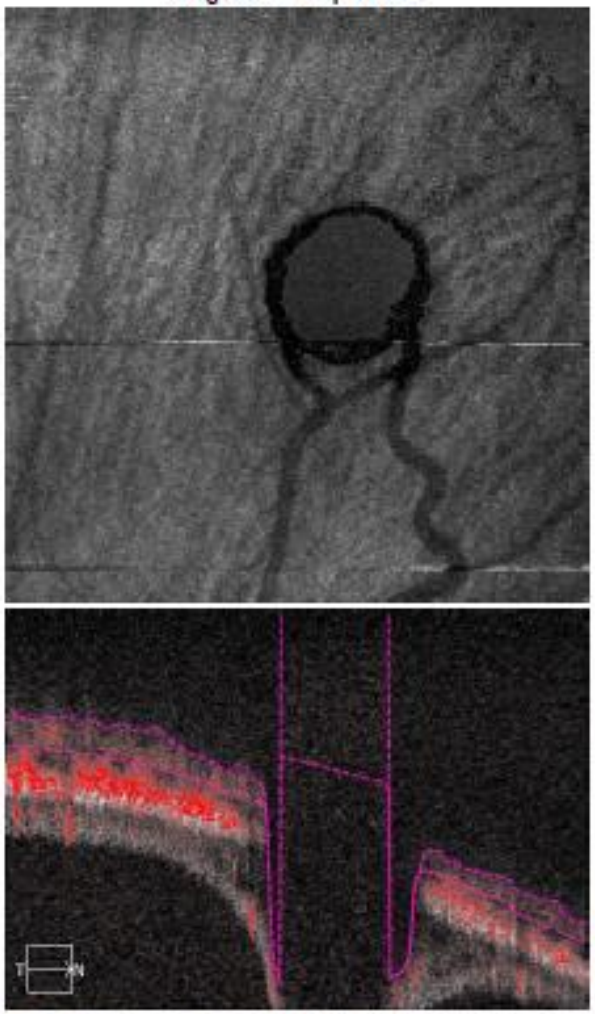
	Central Subfield Thickness (µm)	Cube Volume (mm³)	Cube Average Thickness (µm)
ILM - RPE	276	11.4	317

Angiography Analysis : Angiography 6x6 mm

OD ● | ○ OS

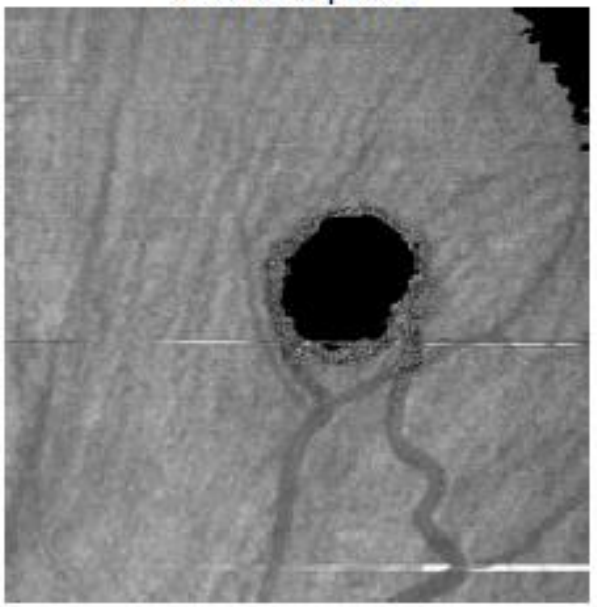


AngioPlex - Superficial



Slice: 175 Top: ILM Bottom: IPL

Structure - Superficial



Overlays
Structure - None
AngioPlex - None

AngioPlex Metrix

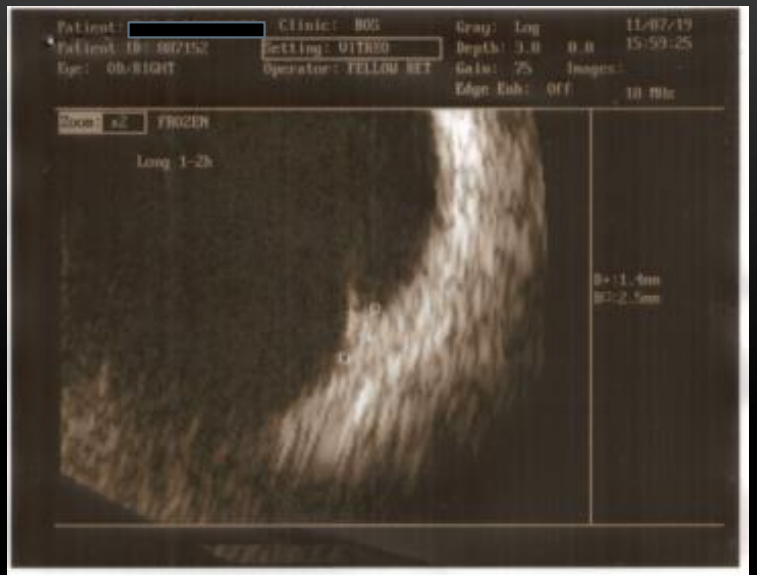
ETDRS - Vessel

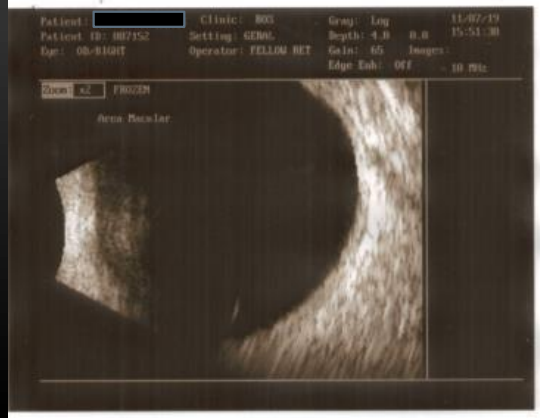
Region	Density
Central	0.0 mm⁻¹
Inner	12.5 mm⁻¹
Outer	17.8 mm⁻¹
Full	16.0 mm⁻¹

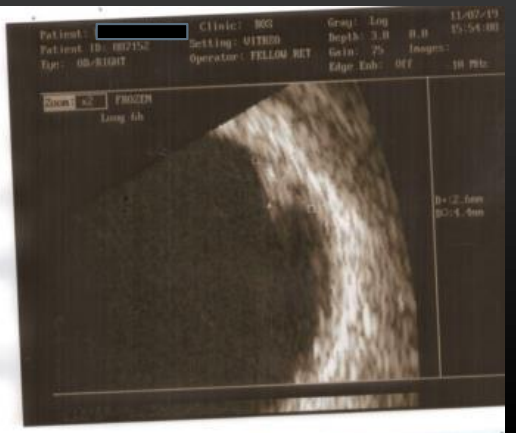
FAZ

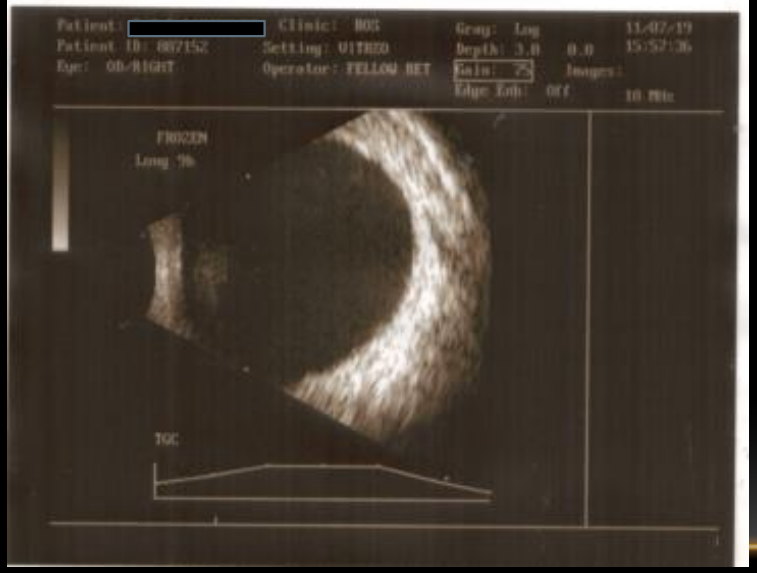
Area	0.06 mm²
Perimeter	0.88 mm
Circularity	0.76

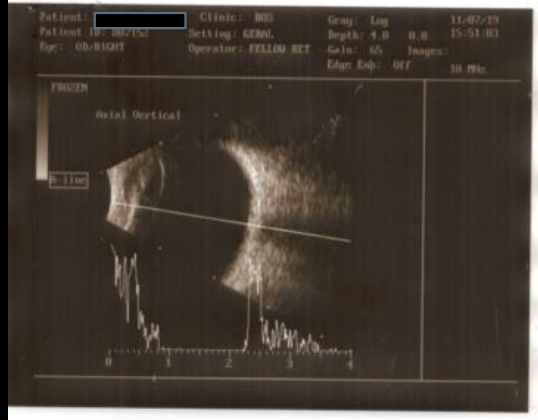
Tracked during scan

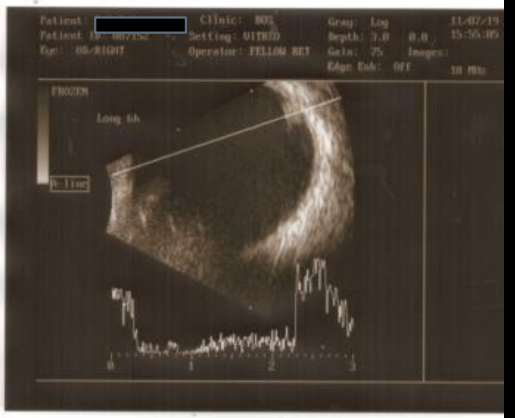

Clinic: BOS
Setting: VITREO
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Long 1-2h

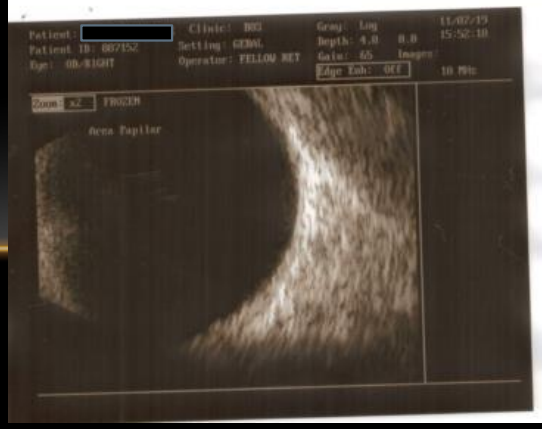

Clinic: BOS
Setting: GENRL
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Area Macular

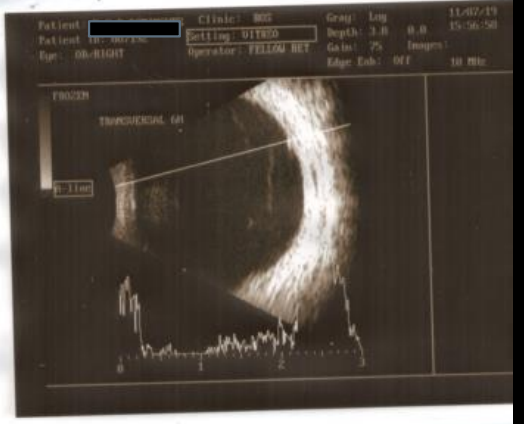

Clinic: BOS
Setting: VITREO
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Long 6h

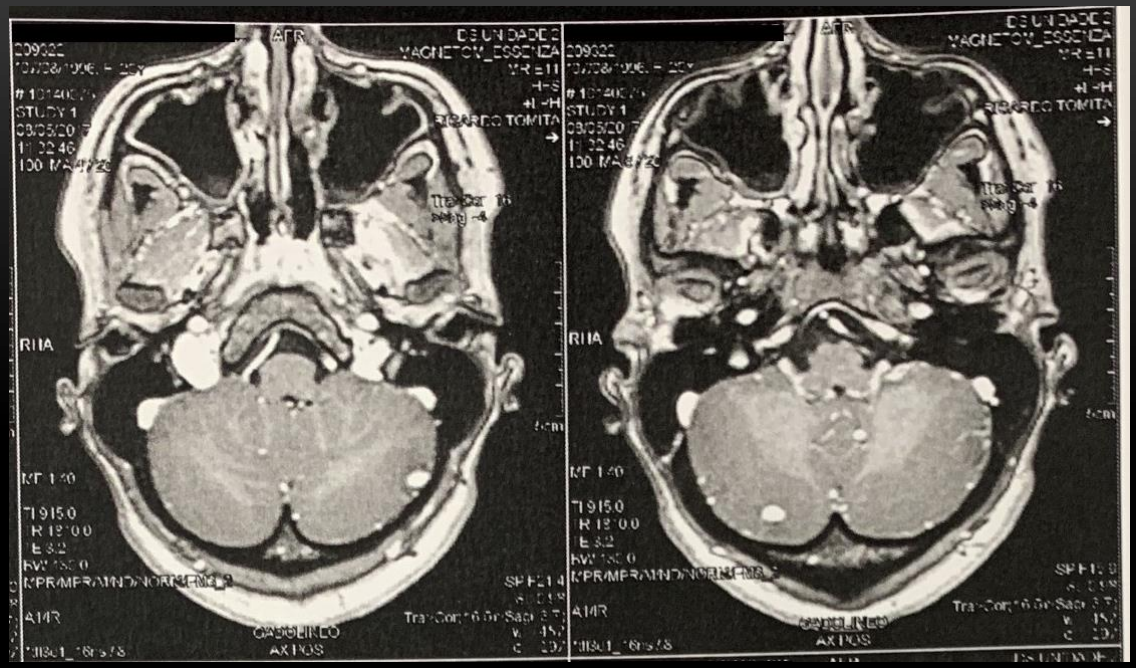

Clinic: BOS
Setting: VITREO
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Long 9h
TGC

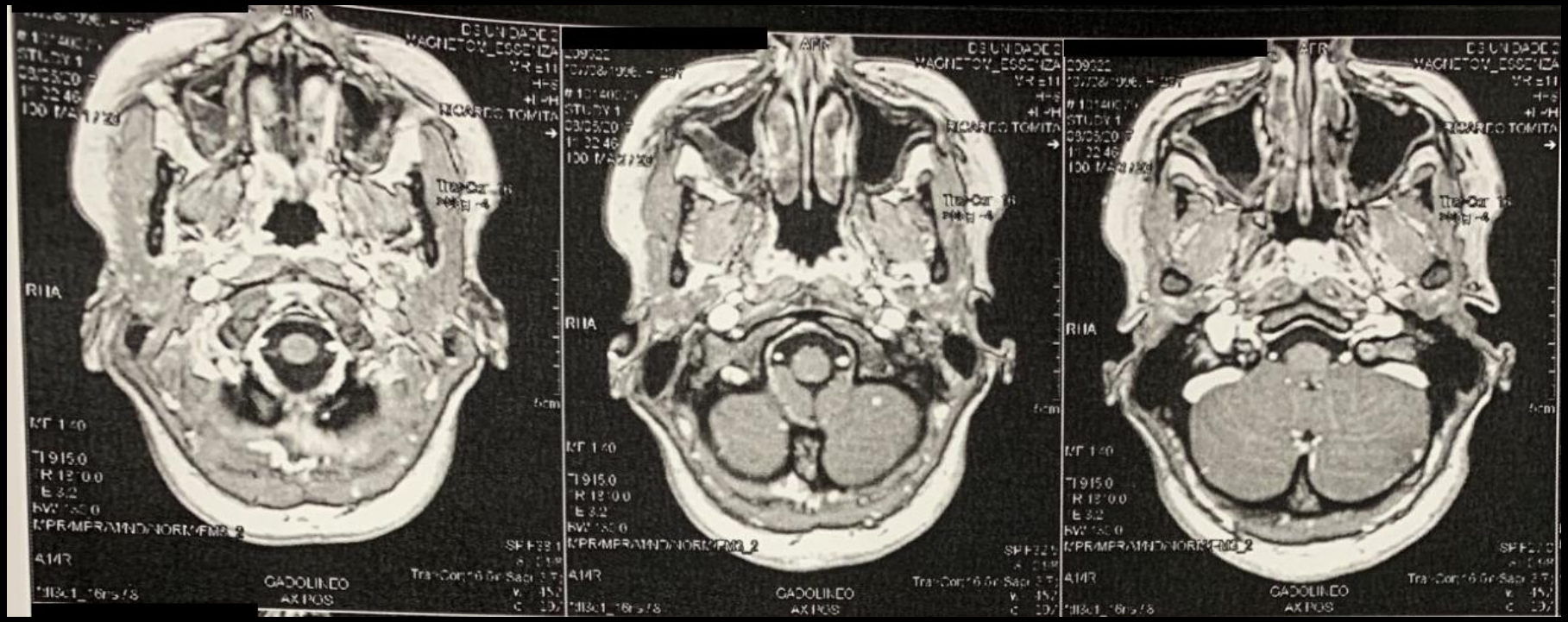

Clinic: BOS
Setting: GENRL
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Axial Vertical


Clinic: BOS
Setting: VITREO
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Long 6h


Clinic: BOS
Setting: GENRL
Operator: FELLOW RET
Eye: OD-RIGHT
FROZEN
Area Papilar

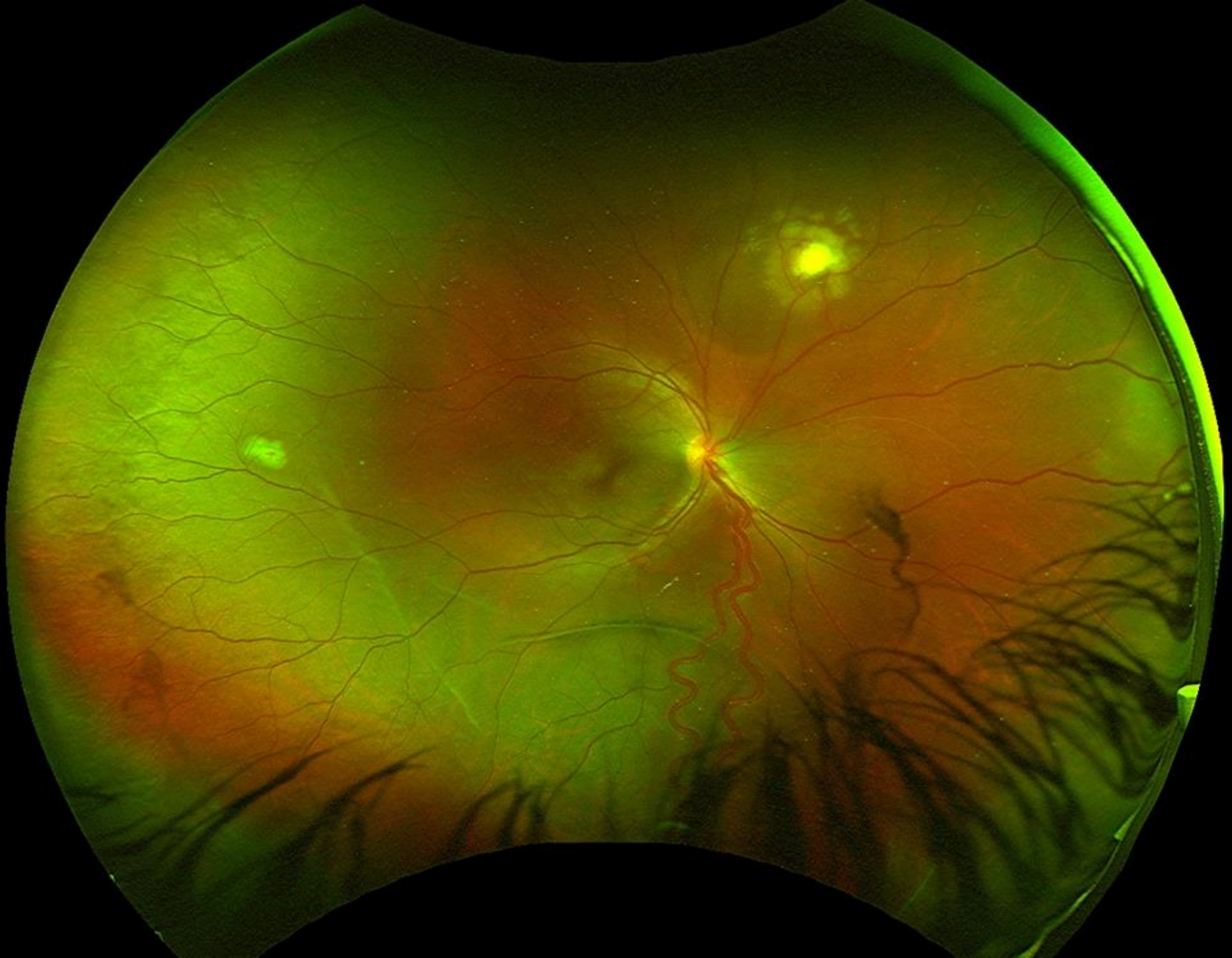

Clinic: BOS
Setting: VITREO
Operator: FELLOW RET
FROZEN

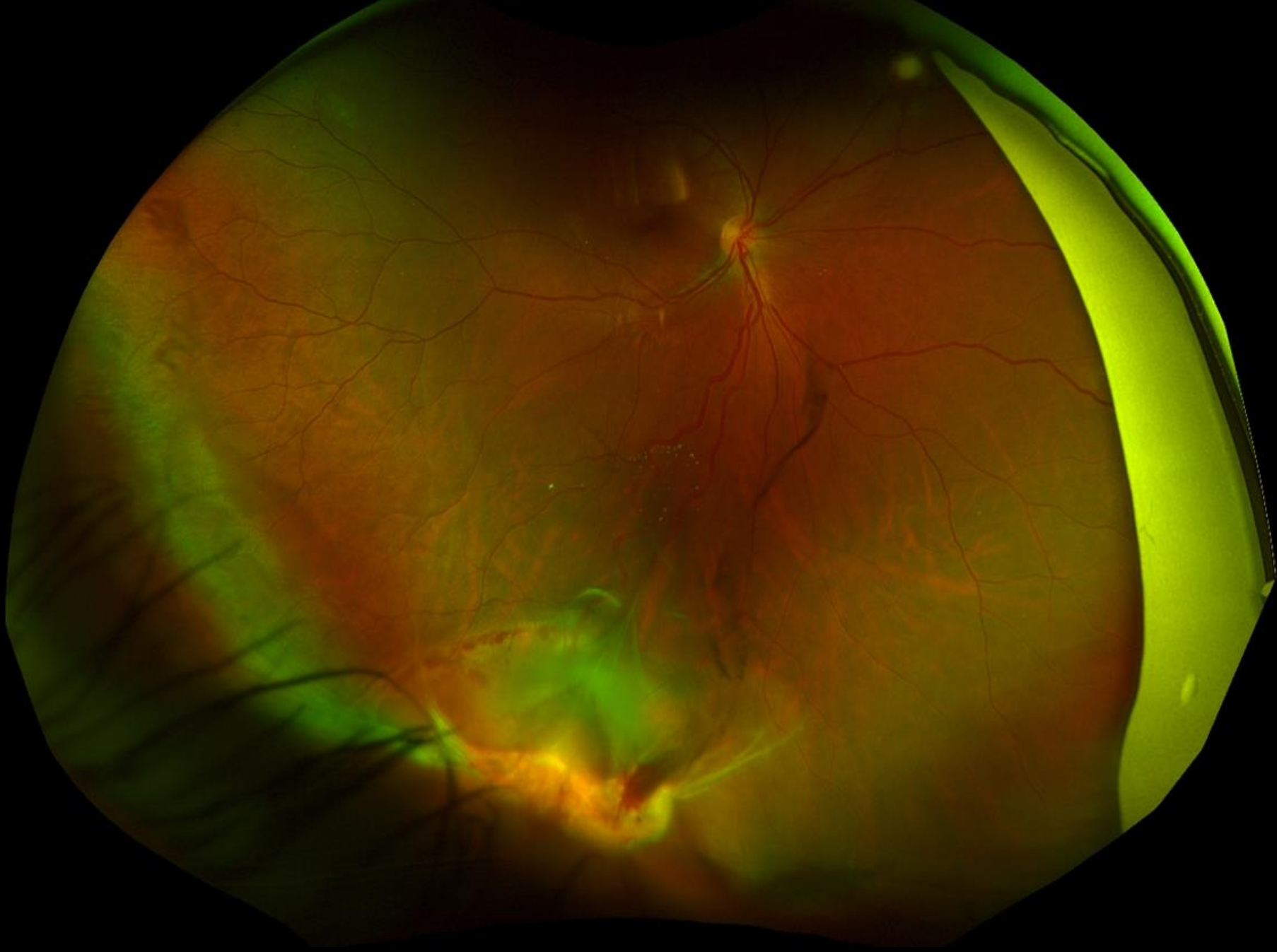

RIIA
GADOLINEO
AXIPOS

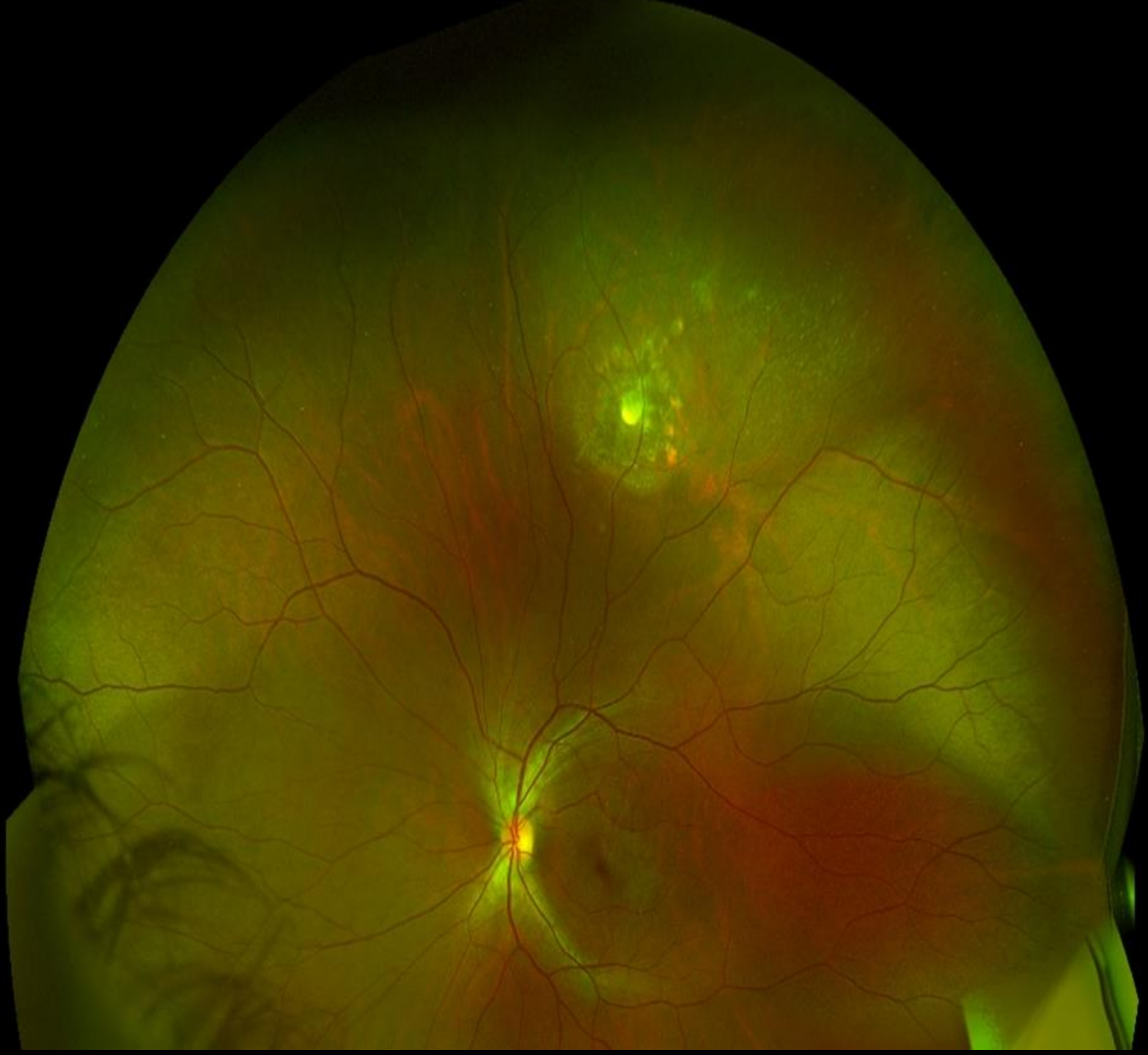

RIIA
GADOLINEO
AXIPOS

TREATMENT

- Laser photocoagulation (small lesions OD and OS)
- Cryotherapy: double freeze-thaw method (6h lesion OD)







MANAGEMENT

- Recognition and diagnosis of VHL disease when present, with appropriate referral for comprehensive care, ideally to a multispecialty team familiar with this complex and potentially lethal systemic condition
- Ocular surveillance for individuals with VHL disease, through regular ophthalmic evaluation, to identify and offer timely treatment for new or active RH as early as possible
- Ablative treatment of RH that can be safely destroyed, to lower risk of vision loss from progressive growth, exudation, and scarring
- Observation or consideration of nonablative treatments for RH that cannot be safely destroyed (such as those growing in proximity to the optic disk and fovea)
- Observation of asymptomatic retinal vascular proliferation, with consideration of vitrectomy for lesions exerting macular traction and consequent effects on vision

REFERENCES

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THANK YOU