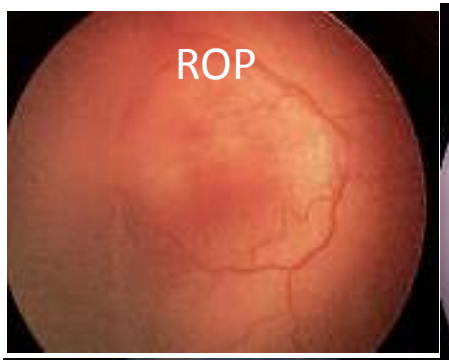
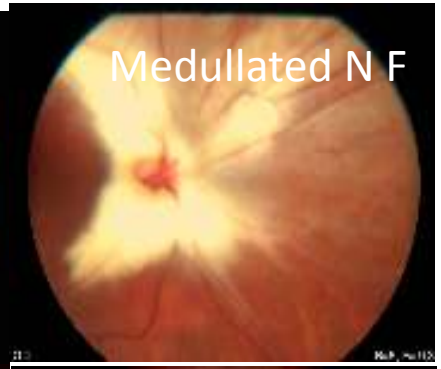
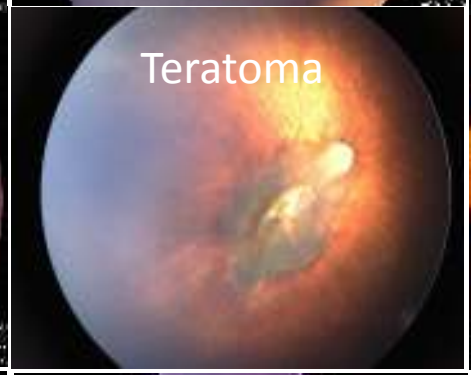
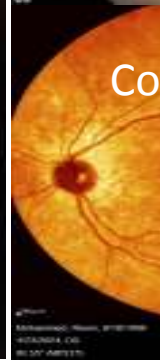
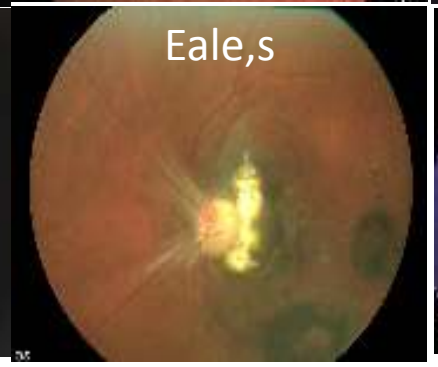
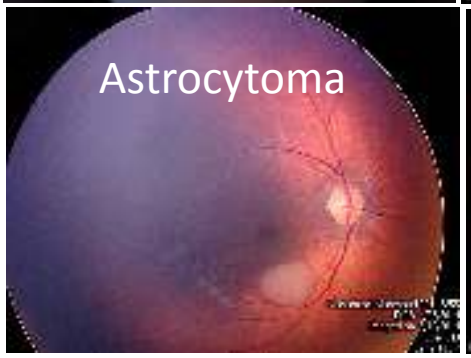
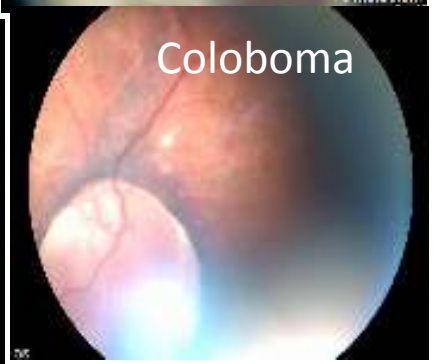
Challenging Pediatric Retinal disorders How to approach

ADEL ALEIELDIN

PROFESSOR OF OPHTHALMOLOGY

RESEARCH INSTITUTE OFOPHTHALMOLOGY

SCOPE OF PEDIATRIC RETINAL DISORDERS

Vascular	Infectious	Tumours	IRD	Developmental
 ROP	 Toxocara	 Retinoblastoma	 XLRS	 Medullated N F
 Coat,s	 Toxoplasmosis	 Teratoma	 Cone dystrophy	 PHPV
 FEVR	 Eale,s	 Astrocytoma	 Best	 Coloboma

SCOPE OF PEDIATRIC RETINAL DISORDERS

Vascular Disorders
ROP
Caot,s
FEVR

Tumours
Retinoblastom
Astrocytic hamartoma

Herdeitary retinal disorders
Leber,s
RP
Cone –Rod dystrophy

Developmental
PFV
Coloboma
Morning glory syndrome

Inflammatory and infectious
Toxoplasmosis
CMV

Tractional and structural
XLRS
RD

Concept of approach

Team of work

- Experienced pediatric ophthamologist
- Clinical genetics
- Investigative ophthalmology
- Low vision facility
- Data system IT

Multidusciplinary Team

- Peditaric subspecialties (e.g Oncology ,Endocrinology….etc)
- Radiodiagnosis
- Intervension radiology
- pathology

Scheme of approach

HISTORY

-Pregnancy

-Prematurity

-Developmantal milestones

-Family history

-Genetic counceling

IMAGING

- Ultrasonography
- Colored fundus imaging
- FA
- OCT
- OCT Angio
- Autofluoresence
- MRI

Pediatric Retinal Vascular Disorders

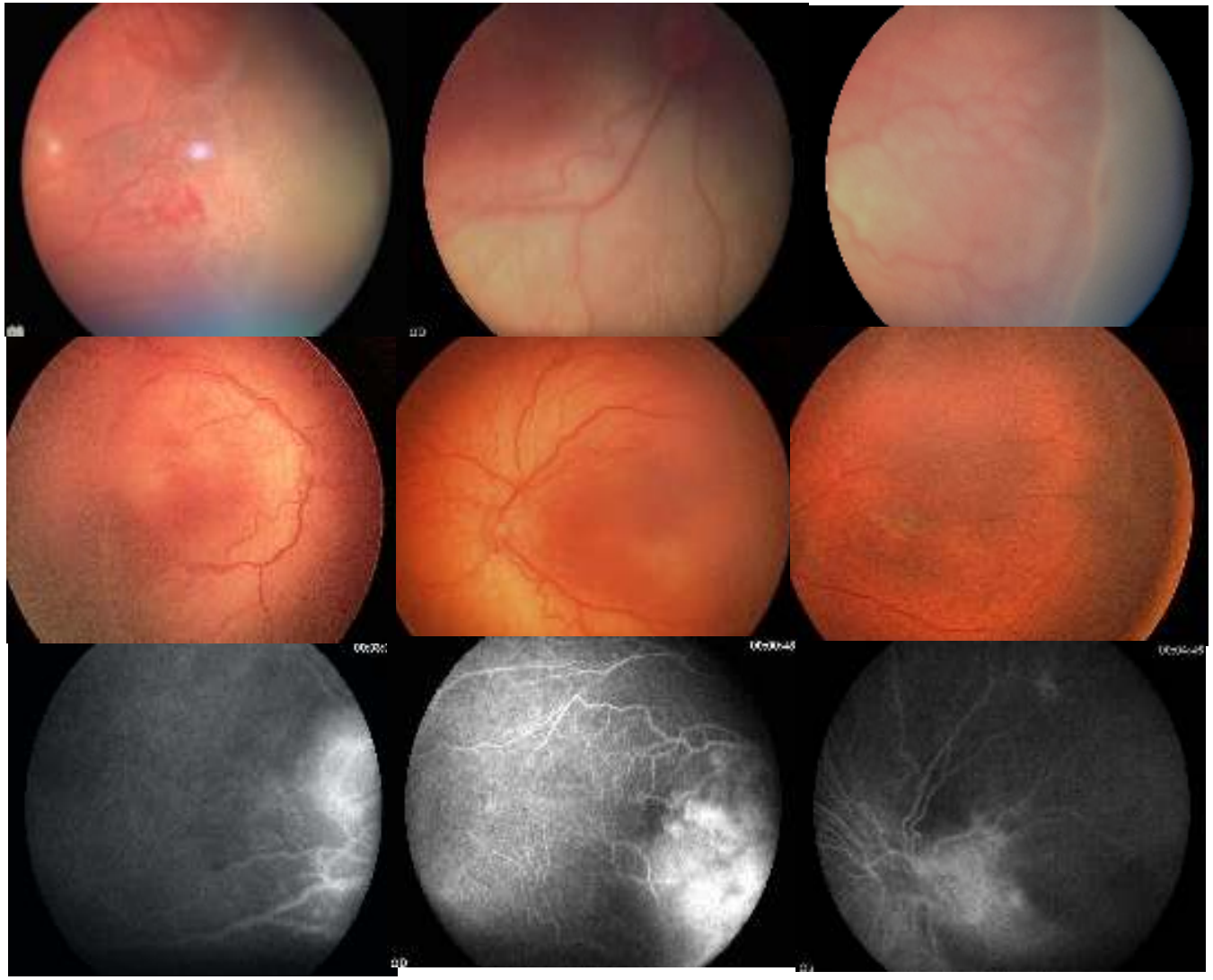
1. Retinopathy of Prematurity (ROP)

- Abnormal retinal vascular development in premature infants.
- Risk factors: Low gestational age, oxygen fluctuations.
- Features: Avascular periphery, ridge, neovascularization.
- Management: Laser photocoagulation, anti-VEGF, surgery.

ROP

Screening guidelines

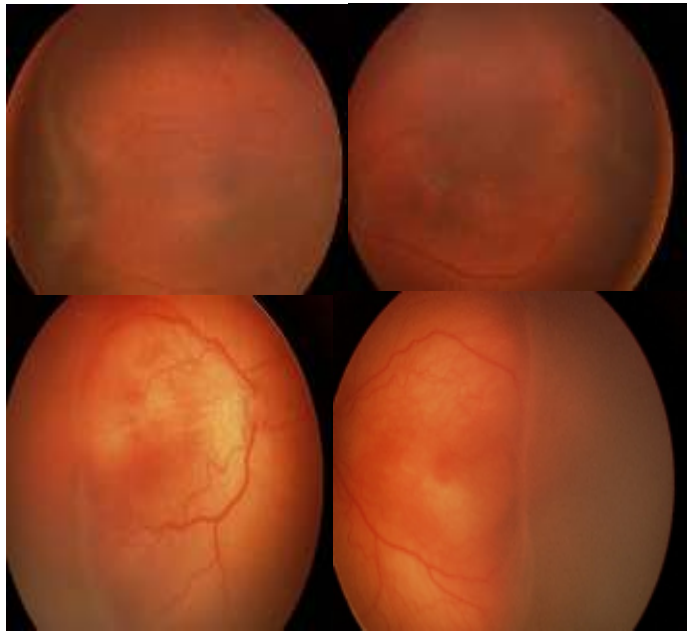
- Documenting early changes
- Fundus Photography to detect missed findings by indirect ophthalmoloscope
- FA detect areas of avascularisation and neovessels



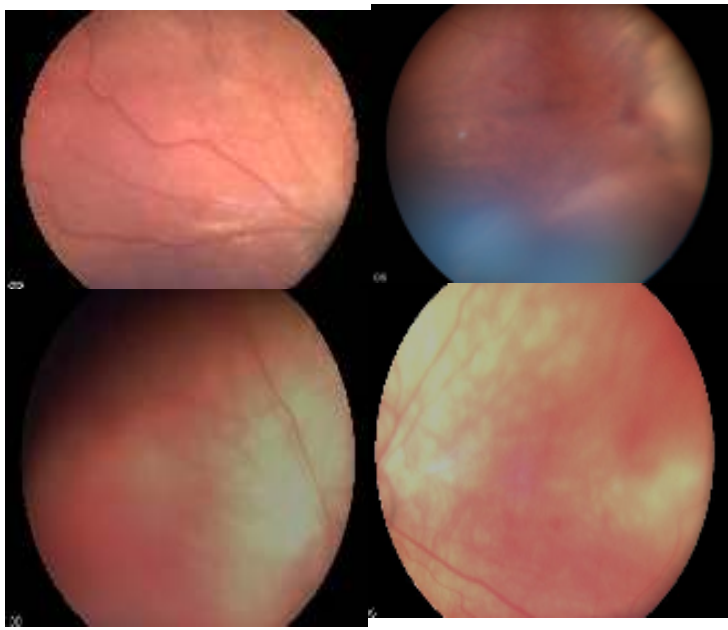
Treatment Guidlines

- ZoneI, any stage ROP with Plus disease.
- ZoneI, stage 3ROP without Plus disease.
- ZoneII, stage 2or3 ROP with Plus disease.

Pretreatment

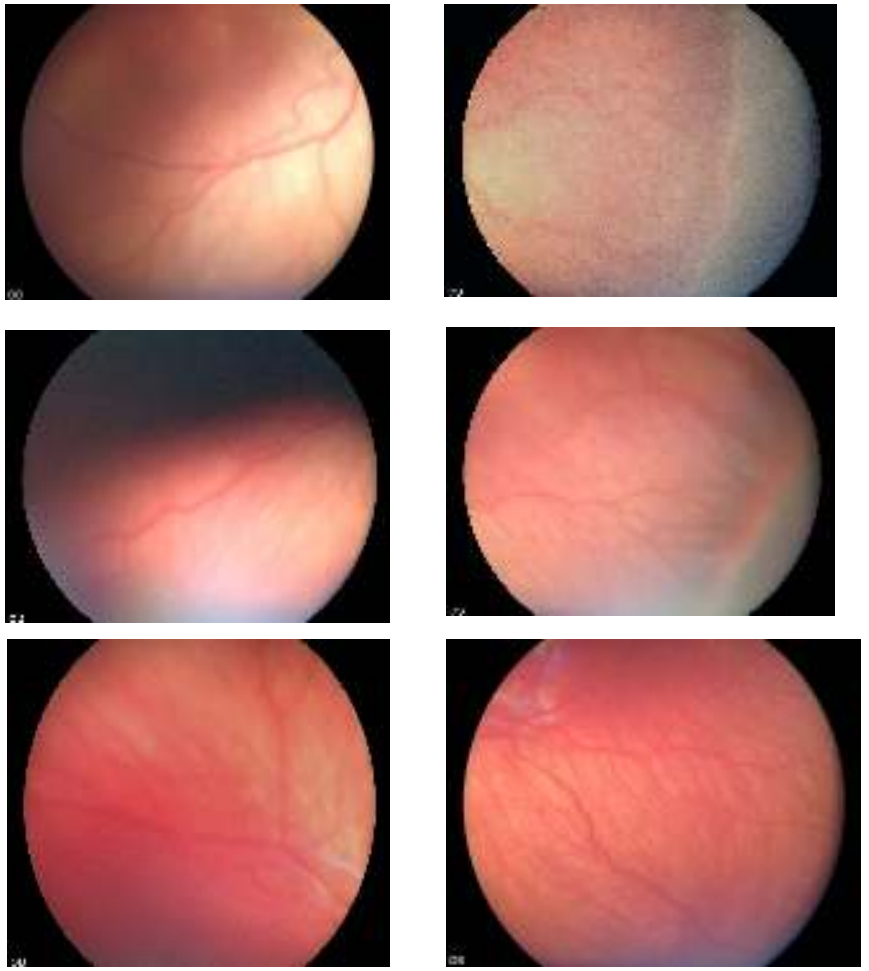


Post treatment



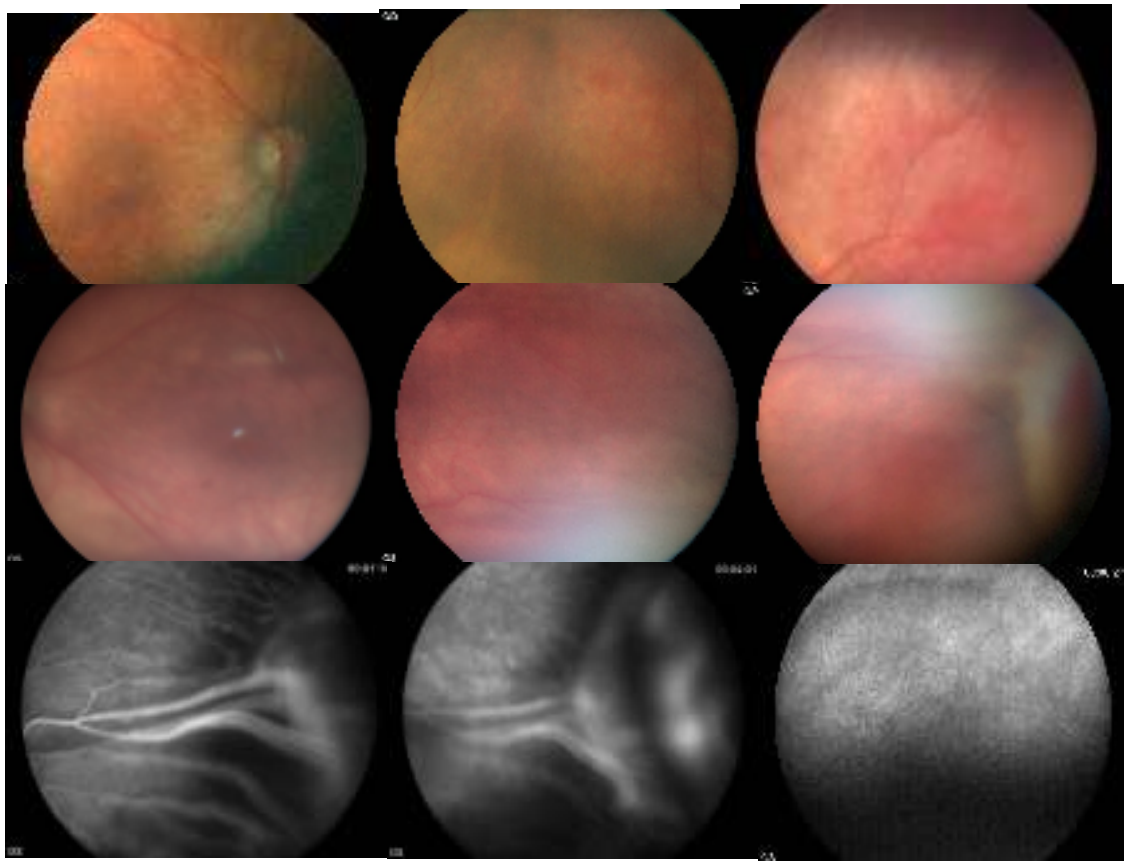
Treatment Guidlines

- Stage 1 FU
- Stage 2 meticulous FU
- Stage 3 treat
- Preplus ?? close FU
- Plus treat
- APROP treat
- Stage 4 a,b surgery
- Stage 5 still questionable



Reactivation Post injection in plus ROP

- Reactivation means still avascular retina that neede laser treatment during Follow up period post injection



Surgical considerations

Axial length

- average axial length of 16.8–17.7 mm and is approximately 70% the size of the adult eye, which averages 23.5 mm
- the lens to globe ratio is greater for children than for adults
- Parsplana issue

	Distance posterior to limbus (mm)						References
	1.0	1.5	2.0	2.5	3.0	3.5	
Age of child	n/a	0–6 months	6–12 months	1–2 years	2–3 years	3 + years	Lemley and Han [5]
	1–3 months	4–7 months	8–12 months	18 months–2 years	3–5 years	6–12 years	Wright et al. [6]

Surgical considerations

Vitreous

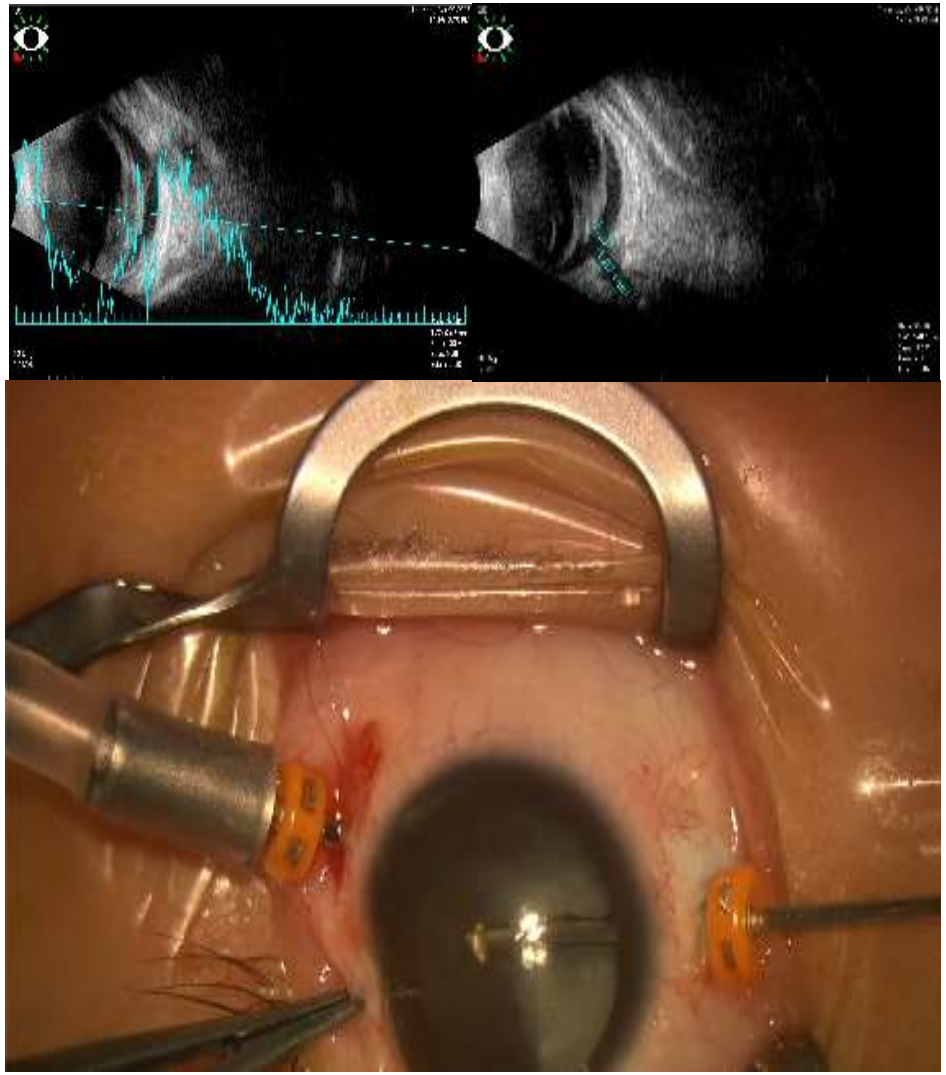
- More formed ,denser and gel like
- More adherent to the retina (base,ON,and macula)
- Posterior hyaloid is tightly adherent
- Risk of poliferative vitreoreinopthy

Surgical Tips In ROP Surgery

Do what is necessary,no more no less avoiding unnecessary surgery or injections

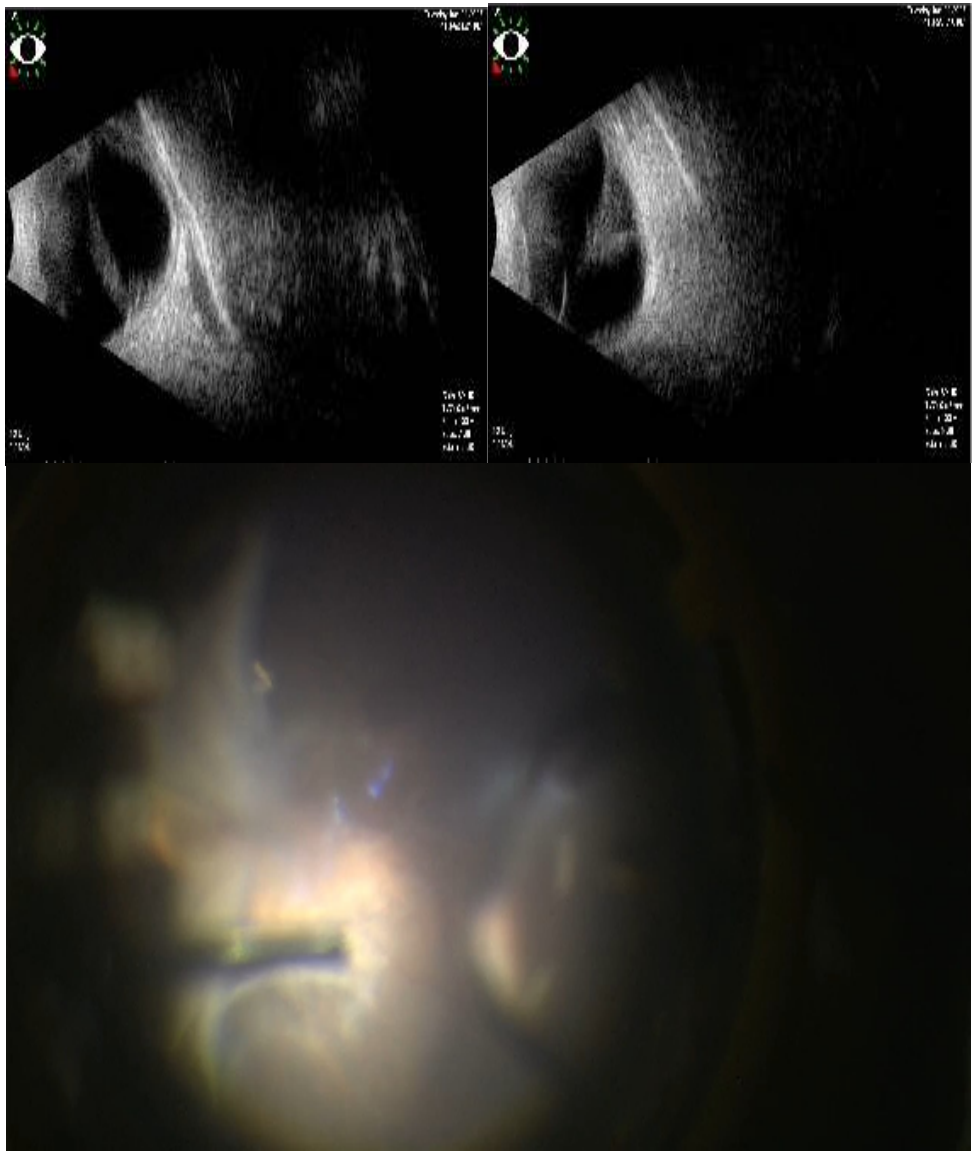
- Relief traction as possible
- Dissection of traction folds

  if it is acsesable

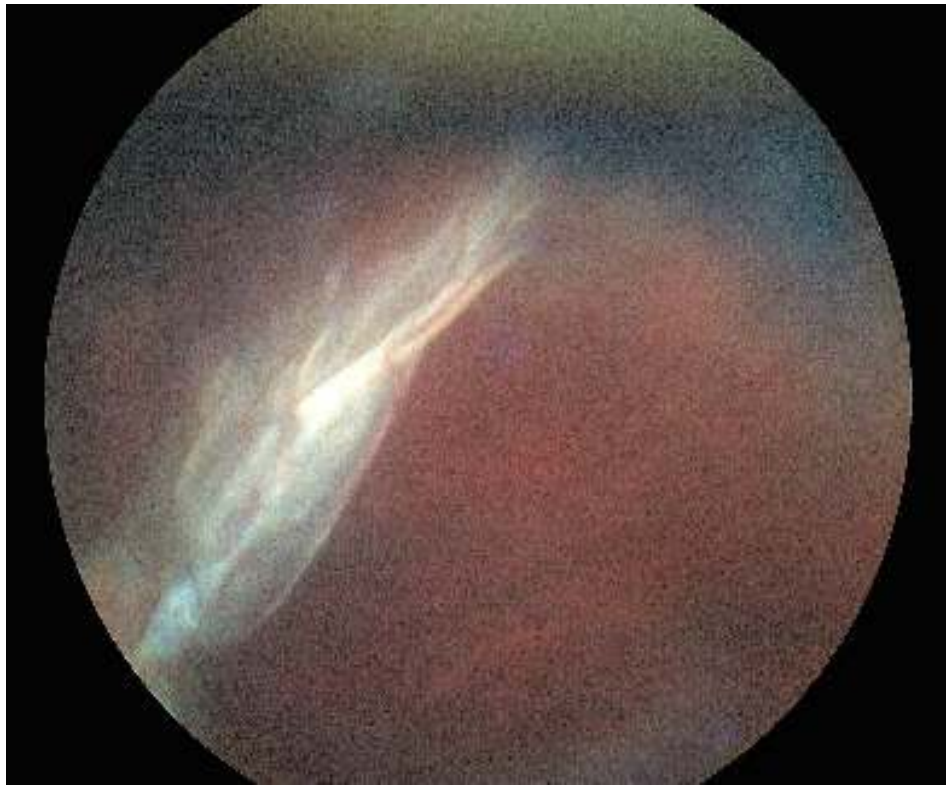


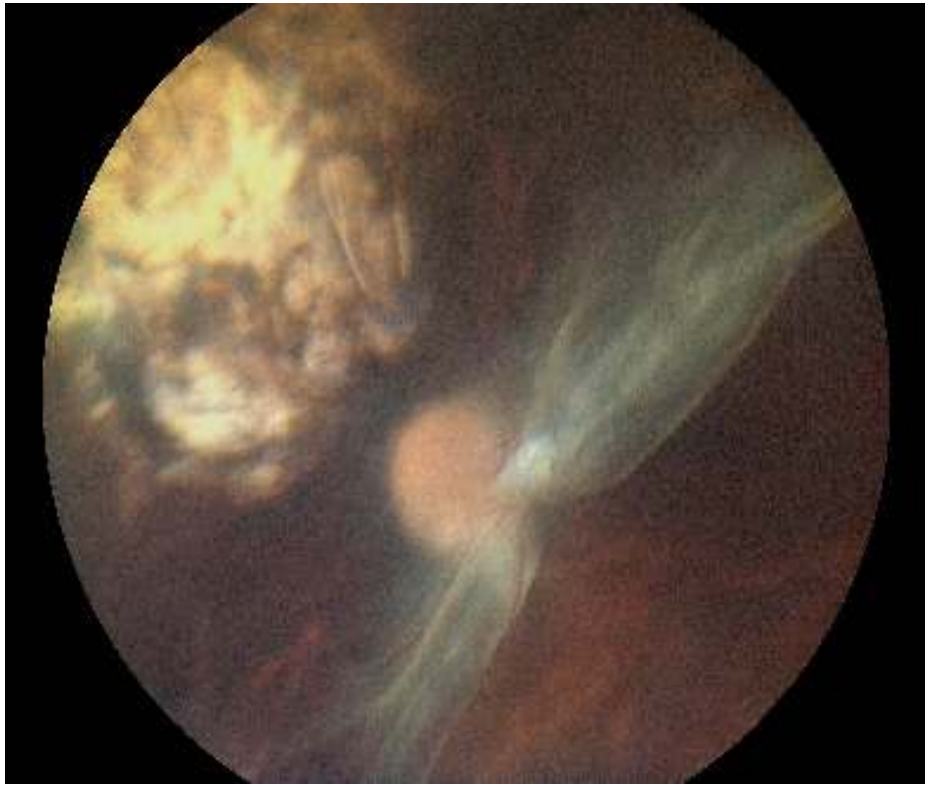
Surgical Tips In ROP Surgery

- Missconcept of PHPV by US
- Complete total vitrectomy is not the main target
- Avoiding iatrogenic breaks
- It can be managed so far no traction and limited break)

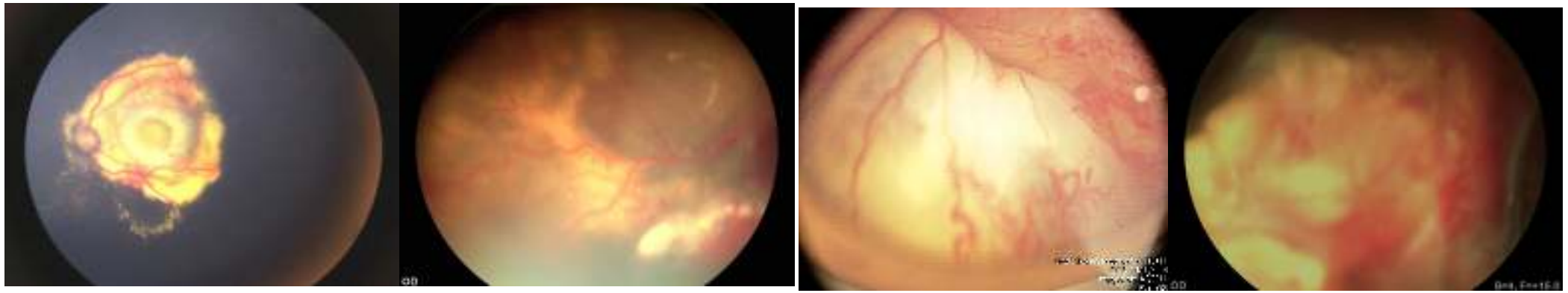


Post Op



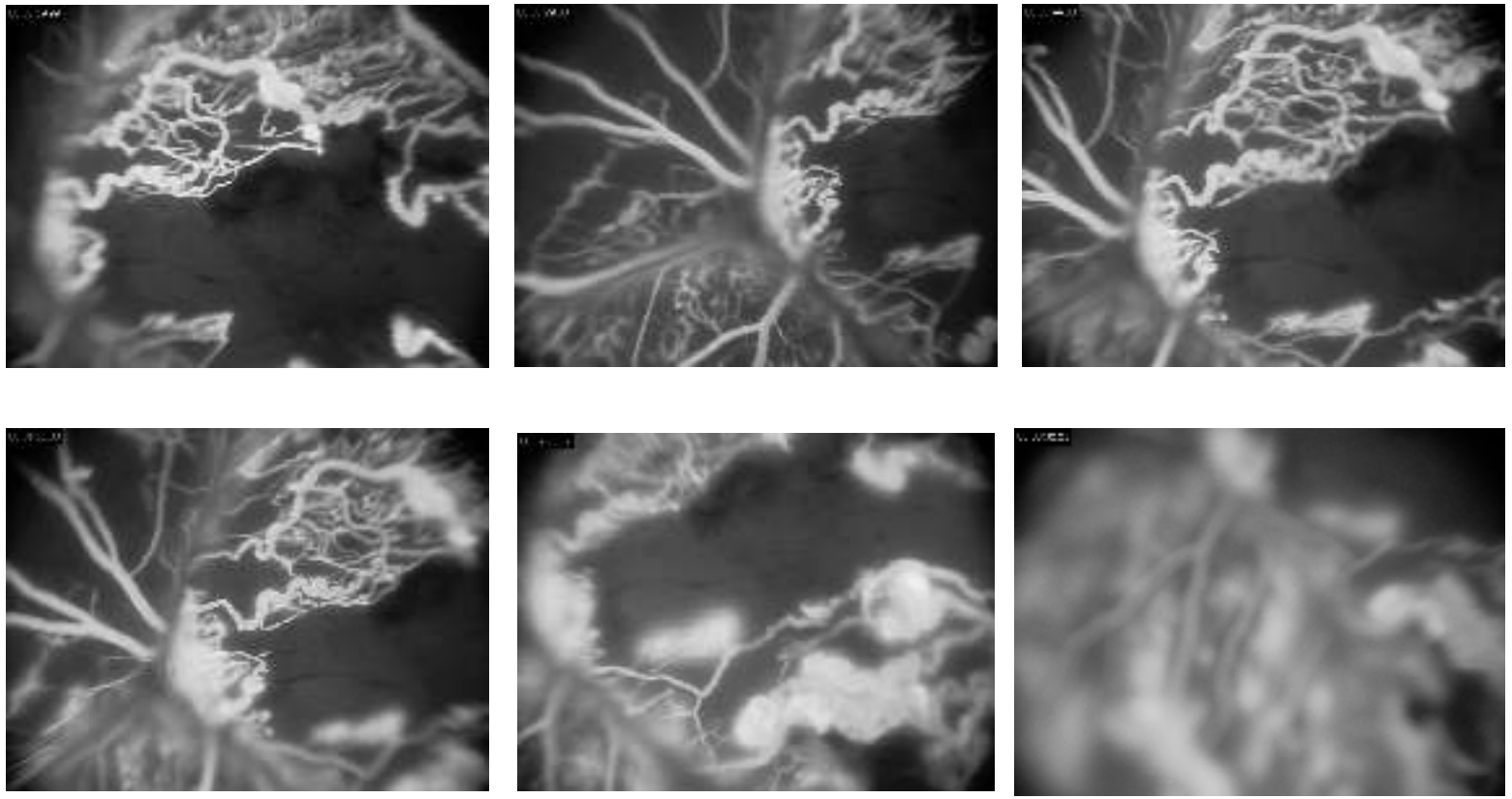


2. Coats' Disease

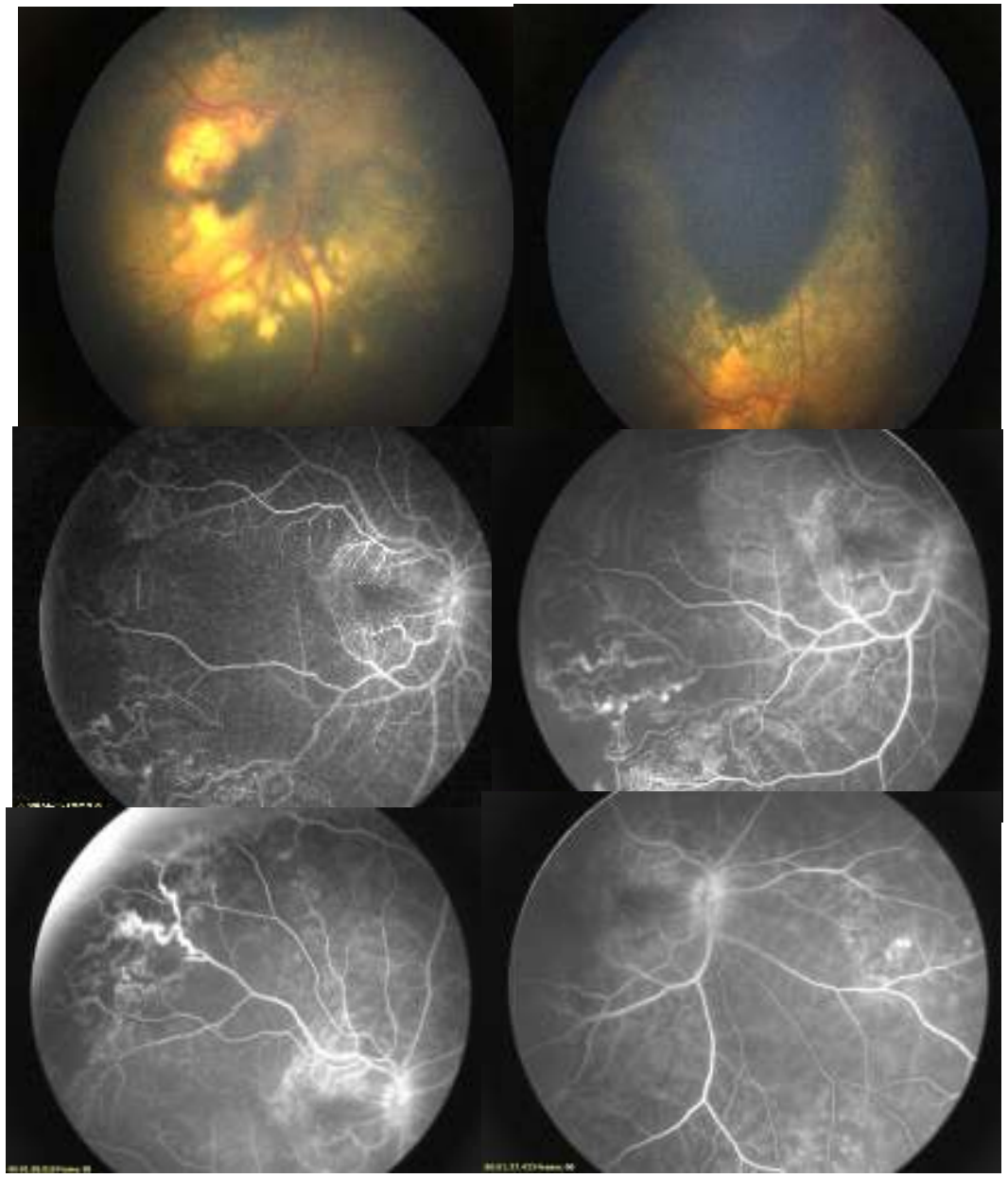


- Idiopathic retinal telangiectasia with exudation (usually unilateral).
- Age: 3–10 years, male predominance.
- Findings: 'Light bulb' telangiectasia, exudation, exudative detachment.
- Treatment: Laser/cryotherapy for telangiectasia, surgery for late stages.

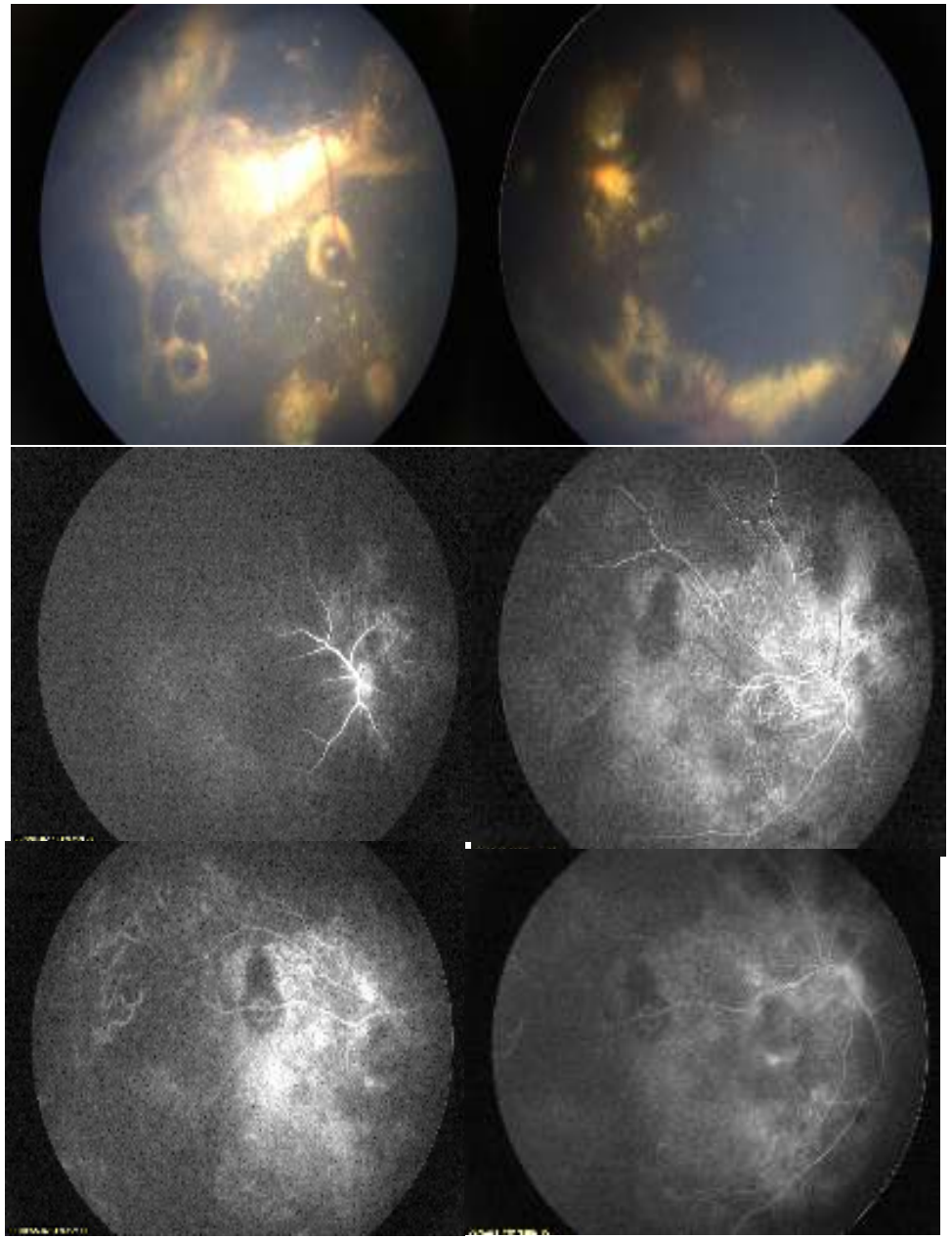
Coat,s



Before treatment

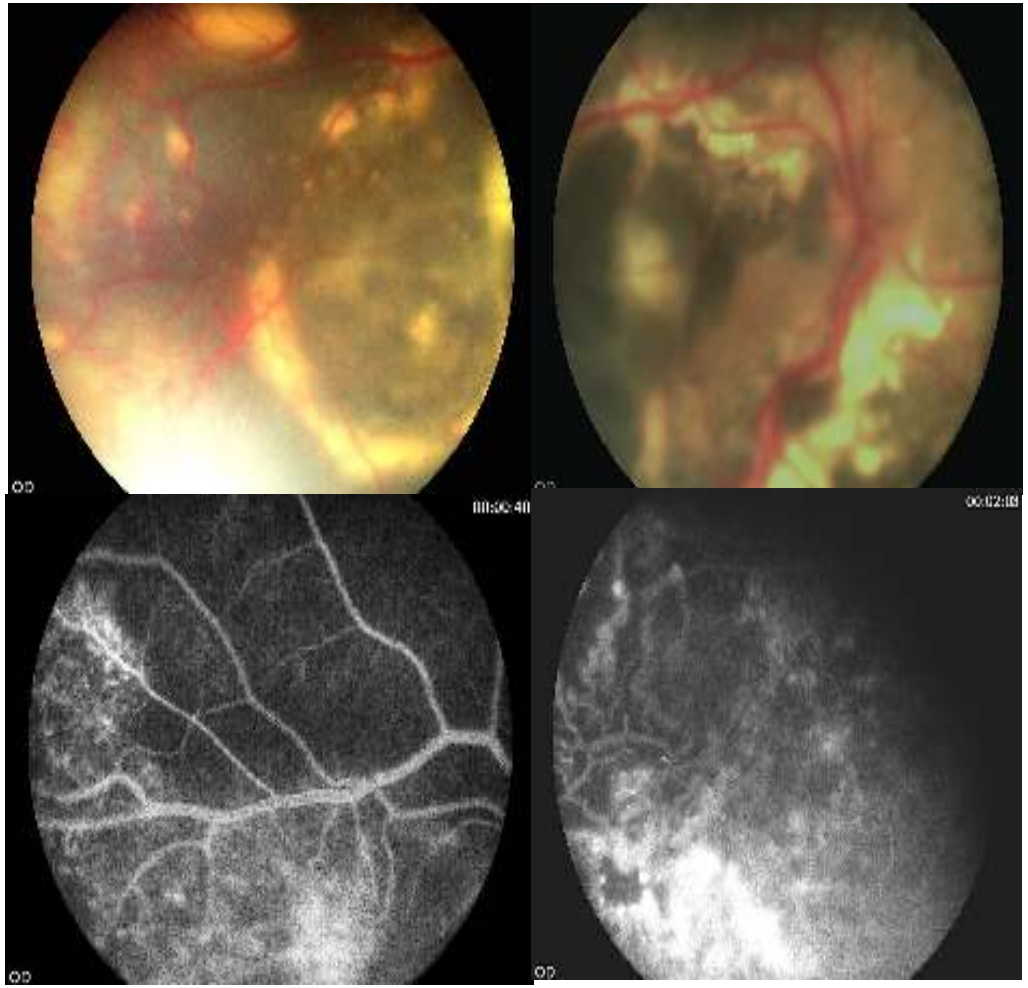


After treatment

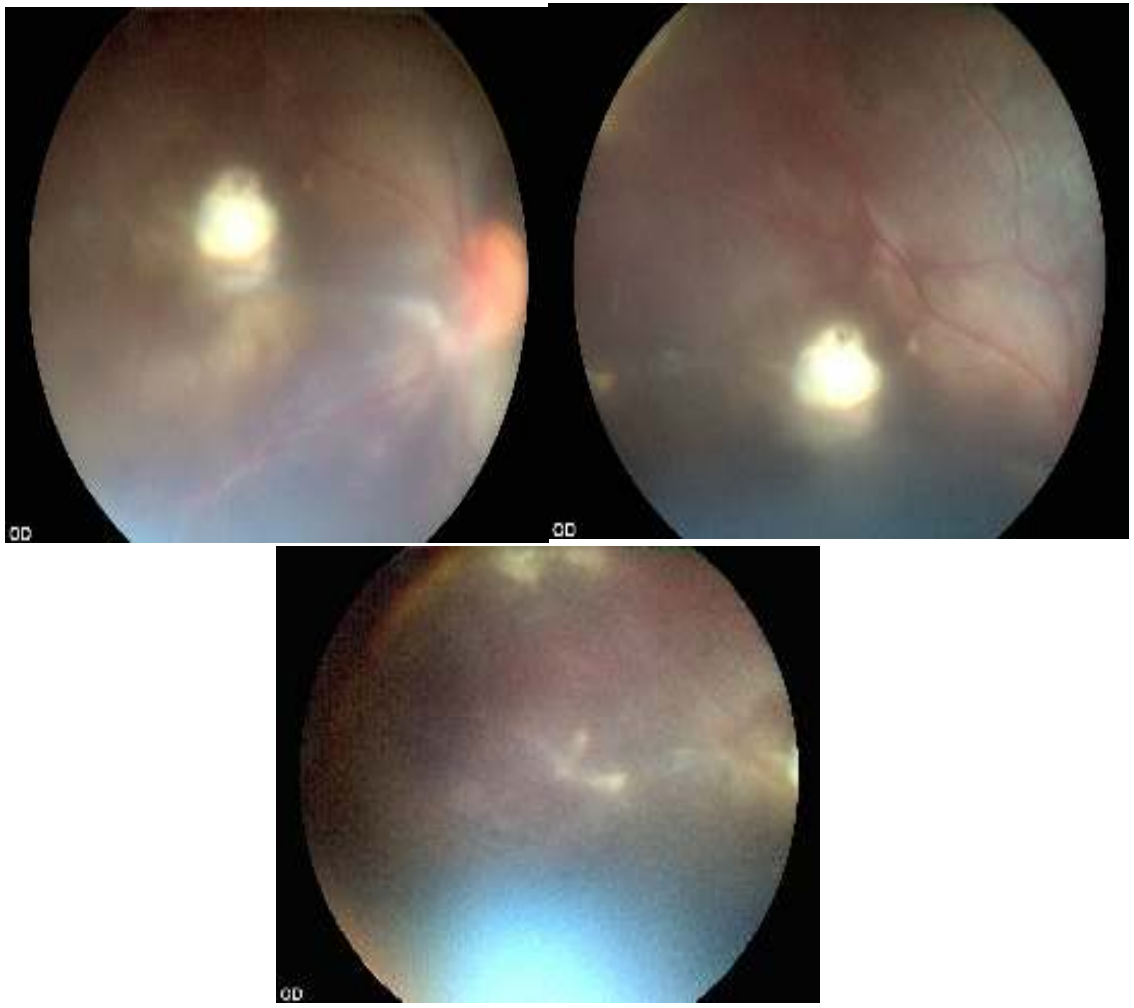


Male child DOB 21/5/2015. Age at ex 3.5 ys

16-9-2018



6/2/2022



Role of Surgery in Coat,s

Prefered non invasive procedure (Ozdek 2020)

1-Evacuation of subretinal exudate through posterior sclerotomy

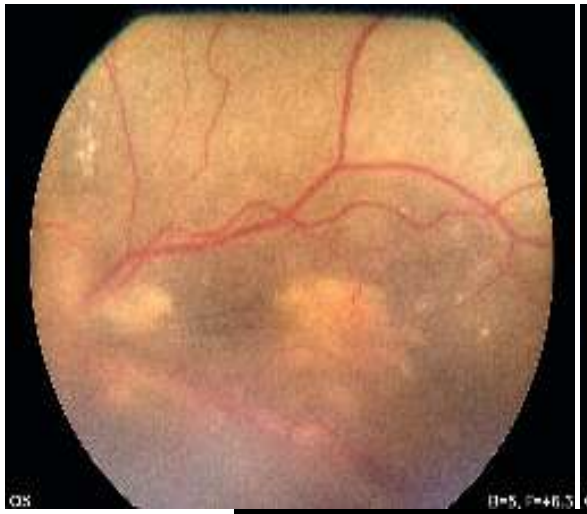
2-continuous formation of the globe through limbal corneal entry with viscoelastic and saline

3-Endoillumination using 25 G Chandelier

4-Dtailed Cryo application throught BIOM viewing system

5-Injection of AVGF

Stage 3 B



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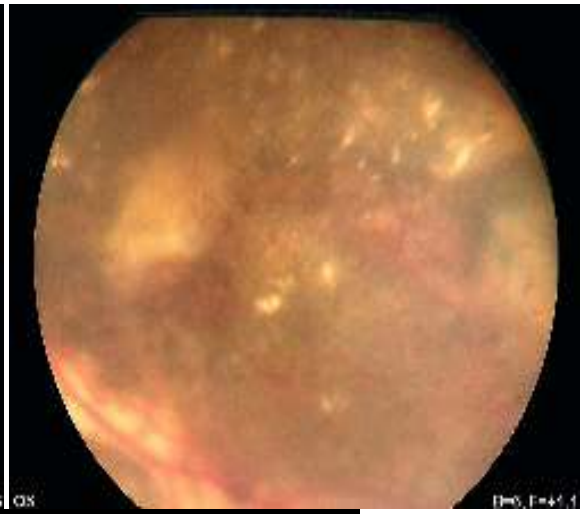
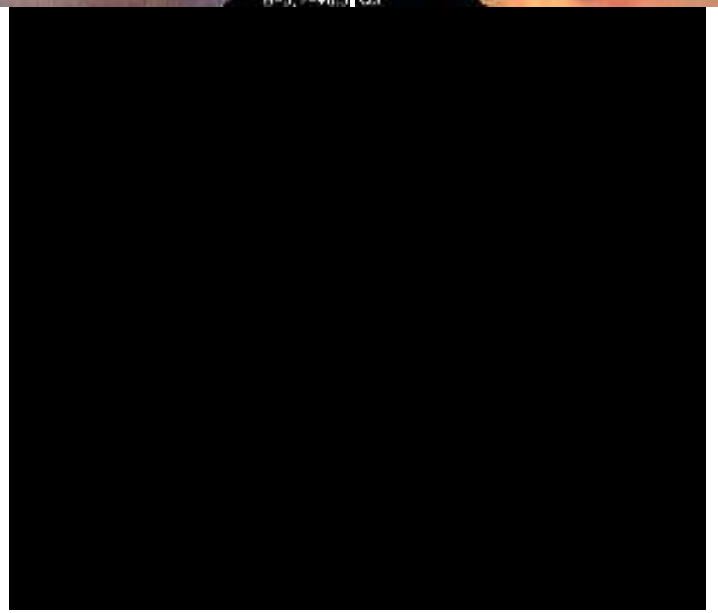
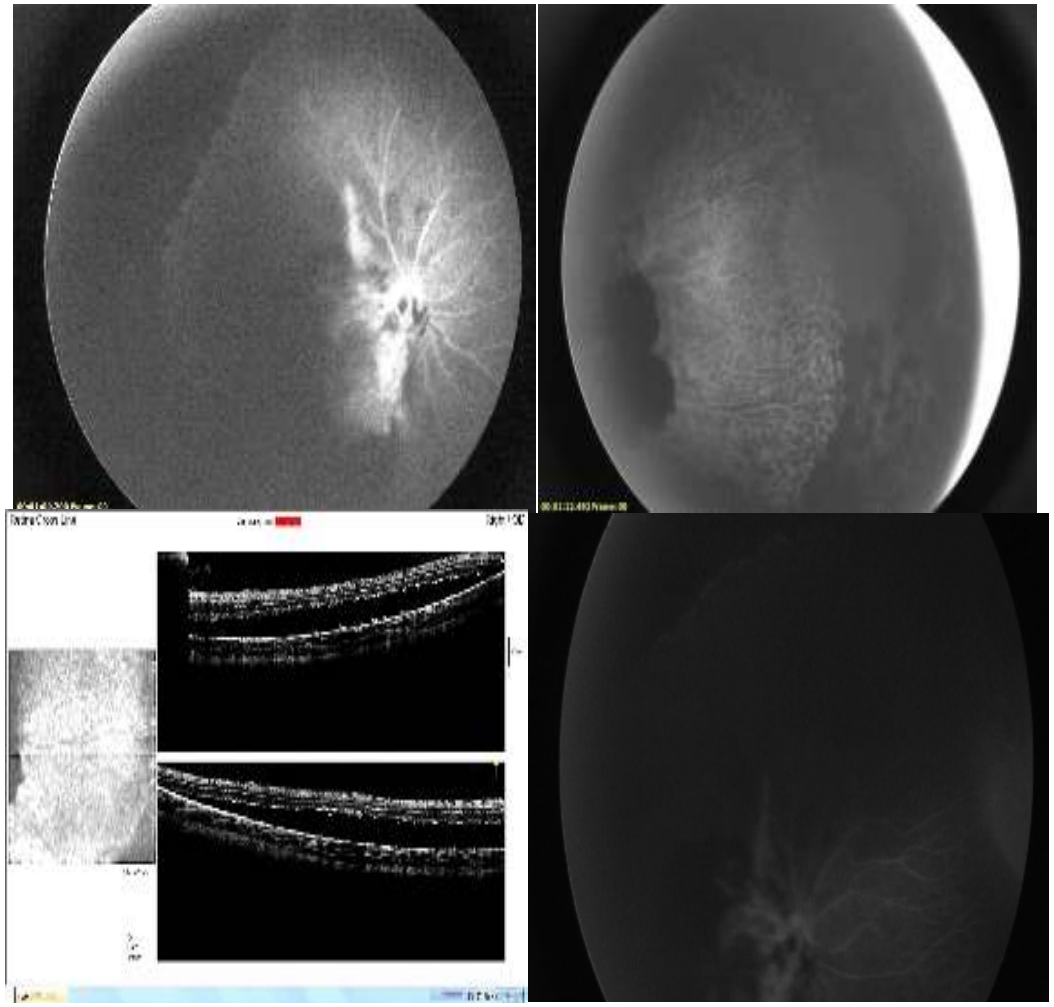




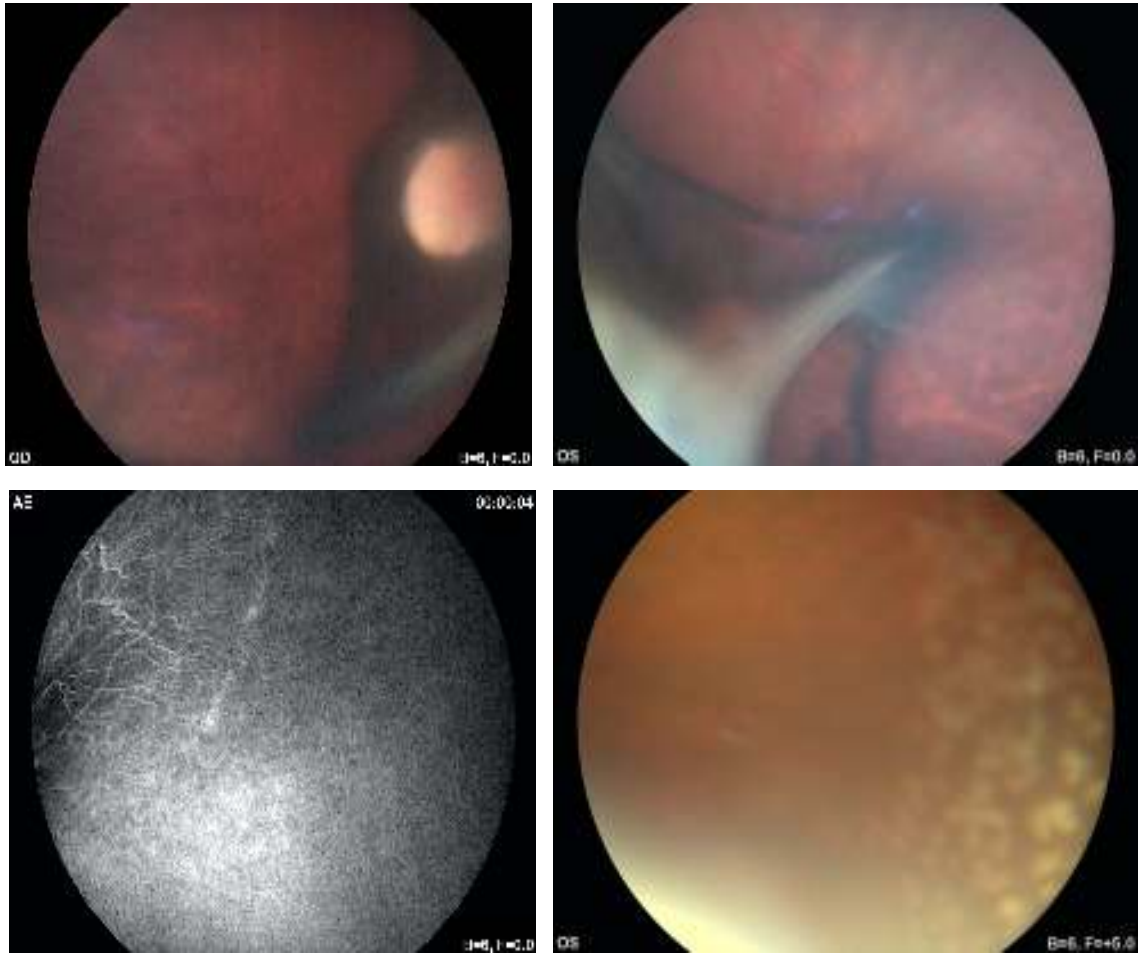
3. Familial Exudative Vitreoretinopathy (FEVR)

- It may be autosomal dominant,autosomal recessive or X linked
- It has several Gene pathway mutations (FZD4, LRP5, NDP).
- Full-term infants with peripheral avascular retina (bilateral).
- Complications: Macular dragging, tractional RD
- OCT (splitting , dragging and epiretinal traction)
- Management: Laser ablation, anti-VEGF, surgery; family screening.



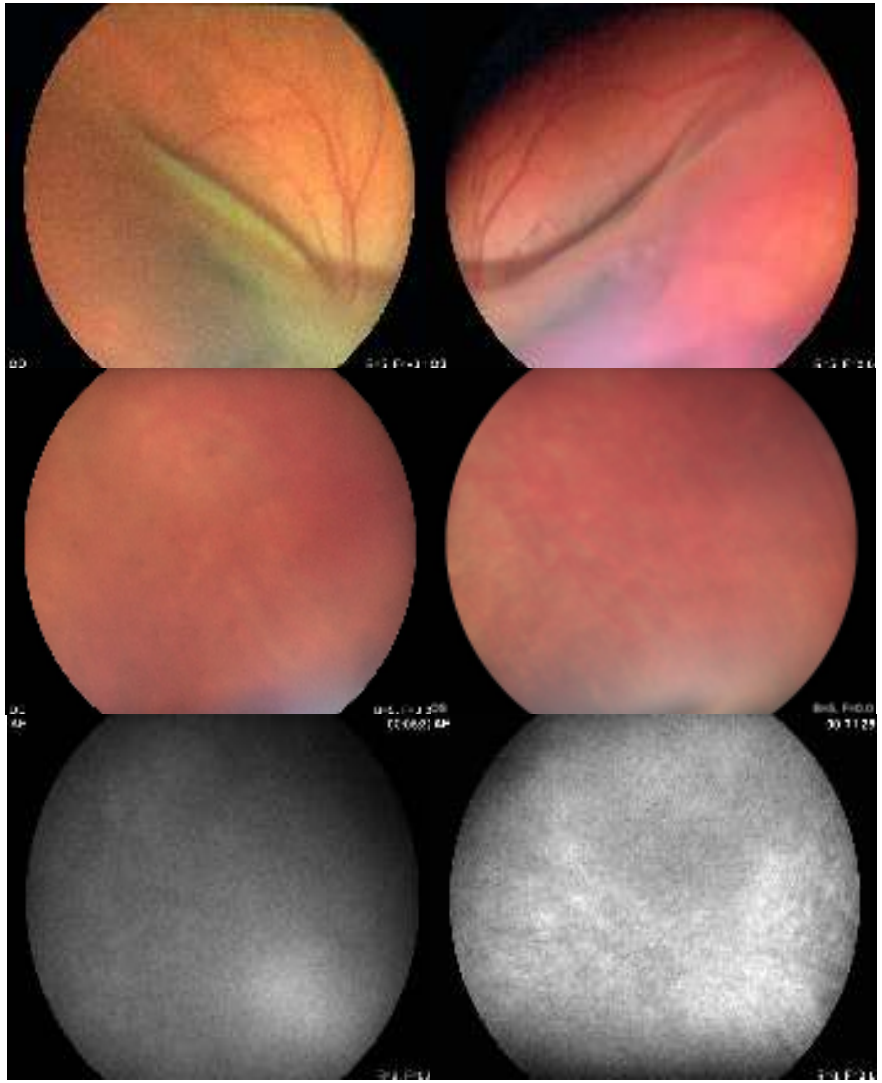
Cases

- 4 ms old infant presenting with Bilateral Leukocoria
- Draging fold s nasally
- Non vascularised areas treated with laser



Cases

- Infant 1 year old
- Presenting with Rt esotropia
- Bilateral tractional macular fold
- Non perfused area treated with Laser



Cases

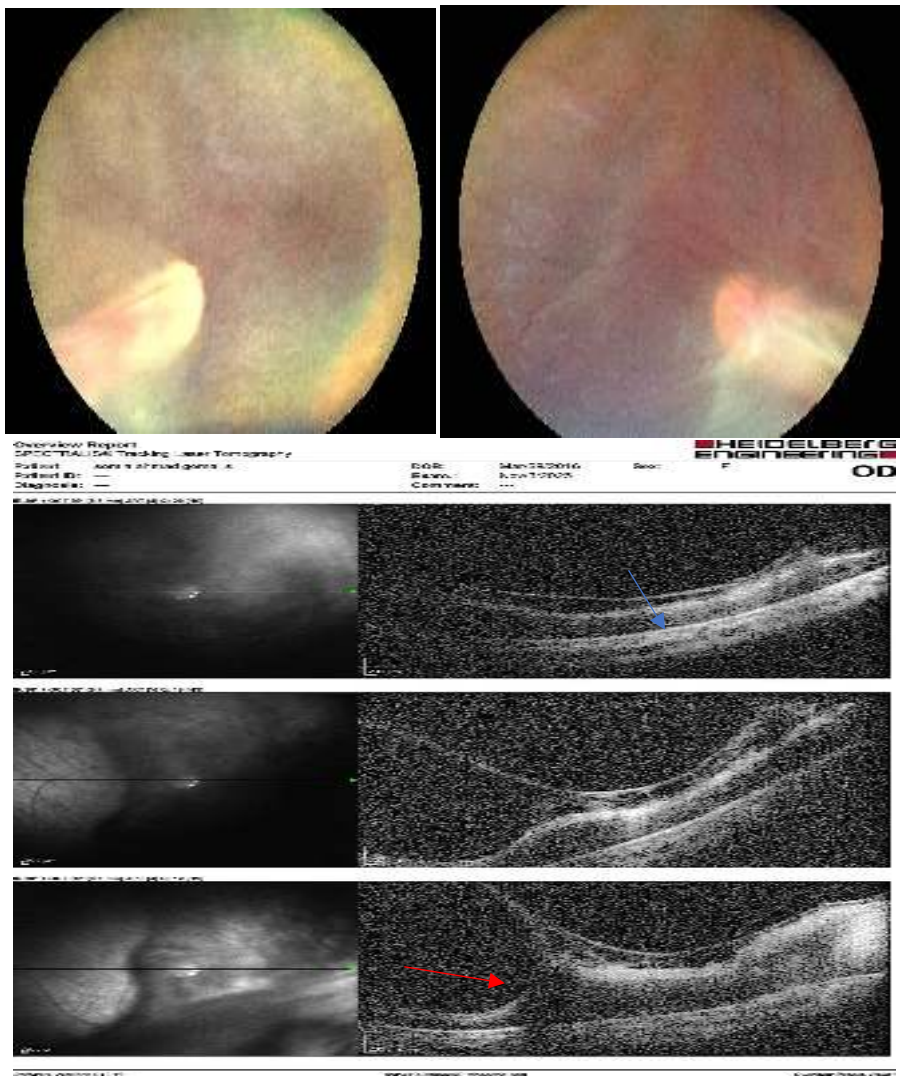
- 2 sisters (X linked)
- Age 6 ,7 ys
- Bilateral FEVR
- Nystagmus
- Advanced in one
- OCT findings

Foveal distorsion

Membrane dragging

Thickened retina

Area of retina splitting



Summary Table

- ROP – Bilateral – Premature – Avascular retina – Laser/Anti-VEGF
- Coats' – Unilateral – Child – Telangiectasia/exudation – Laser/Cryo
- FEVR – Bilateral – Term – Familial avascularity – Laser/Anti-VEGF