# INFANTILE RETINAL VASCULAR DISORDERS

Adel Alei Eldin
Professor of pediatric Ophthalmology
Research institute Of Ophthalmology
Cancer Children Hospital
Shereen Hassan
Lecturer Pediatric ophthalmology
Fayoum University
Cancer Children Hospital

## INFANTILE RETINAL VASCULAR DISORDERS

- There is a group of disorders which mainly affect the infantile vascular system of the retina. These disorders are ranging from developmental one like persistent fetal vasculature and Norrie's disease.
- While other entities are acquired like ROP in premature infants which could be saved on early screening to avoid a severe damage if left undiagnosed.

# INFANTILE RETINAL VASCULAR DISORDERS

- Coats' disease is a retinal vascular disorder that characterized by retinal telangiectasia, intraretinal exudation, and exudative retinal detachment and when it presents in infants it is usually unilateral and always has a poor prognosis if not discovered early and can be treated with cryo, laser and suprachoroidal drainage in cases with marked exudative detachment.

- FEVR is a retinal vascular disease in which the peripheral retinal vessels fail to grow into the far peripheral retina, leaving area of avascular retina It is a lifelong active retinal vascular disease with variable periods of quiescence.

PERSISTANT FEATAL VASCULATURE
PFV

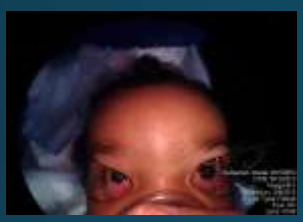
# PHPV MRI

-Retrolental soft tissue

-Evidence of retinal detachment retinal hges

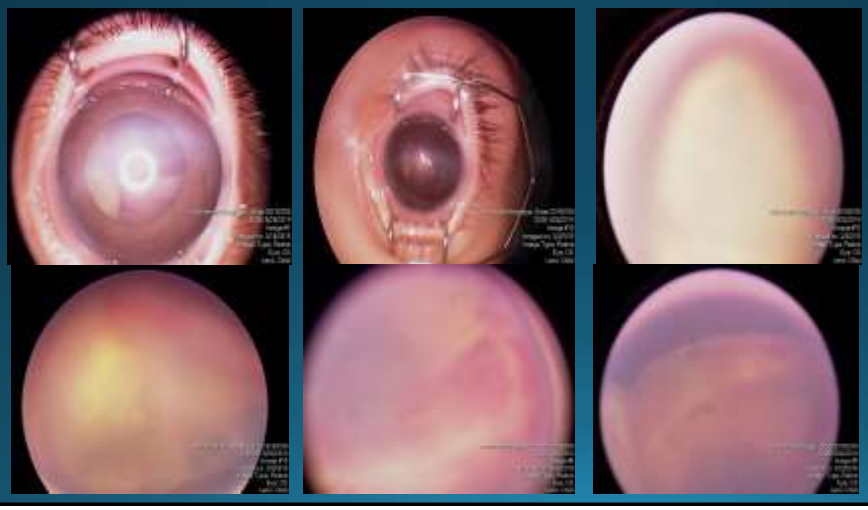
-No definite retinal masses

# PERSISTANT FEATAL VASCULATURE (PFV)



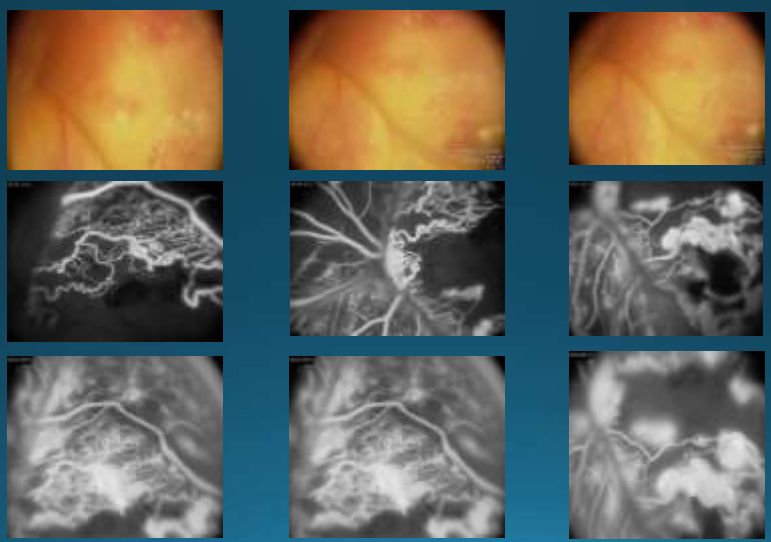
## Coat's disease

It is a retinal vascular disorder characterized by retinal telangiectasia, intraretinal exudation, and exudative retinal detachment



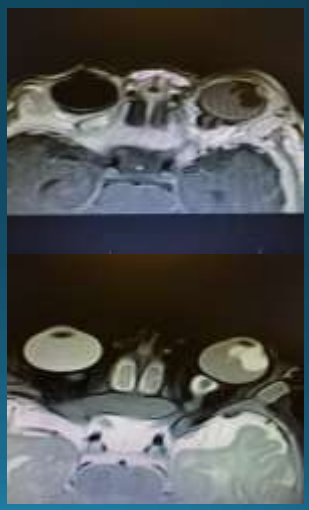
# Coat's

FA Criteria

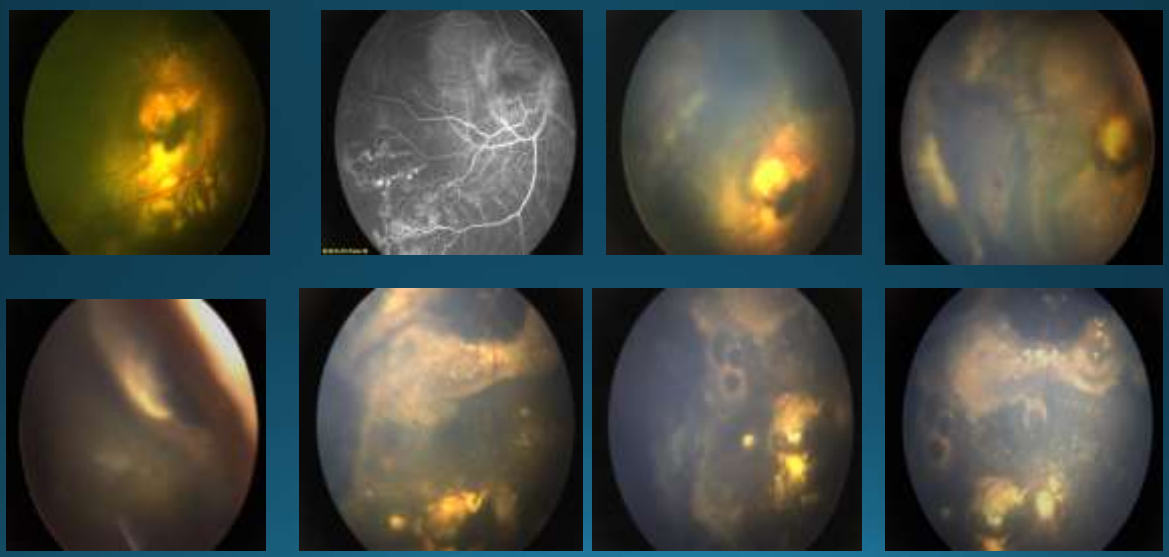


## Coat's MRI Criteria

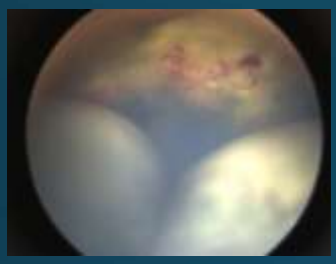
- Abnormal signals of vitreous with low signal intensity in T2 and high signals in T1
- Irregular areas of high signal intensity in T2



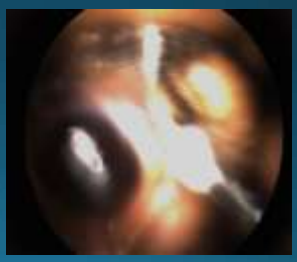
## Coat's prognosis



## Coat's prognosis



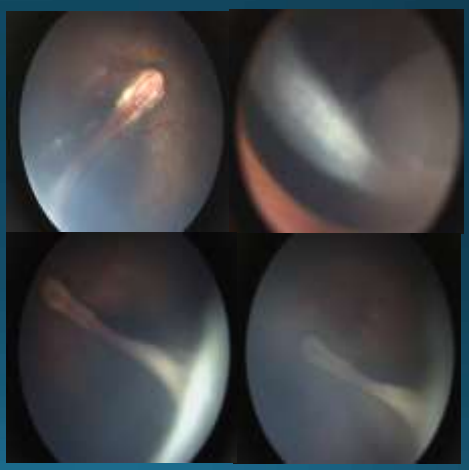




After 1 year of surgery

## FEVR

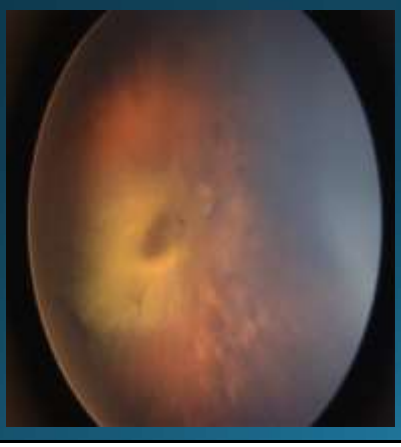
- It is a retinal vascular disease in which the peripheral retinal vessels fail to grow into the far peripheral retina,leaving area of avascular retina
- It is a life long active retinal vascular disease with variable periods of quiescence



# Cases of the year

Infant 4 ms of age presenting with leukocoria.There is no history of prematurity

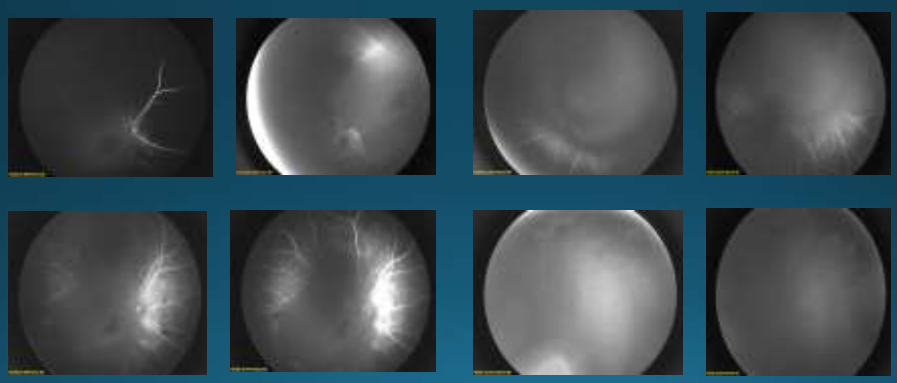
OD



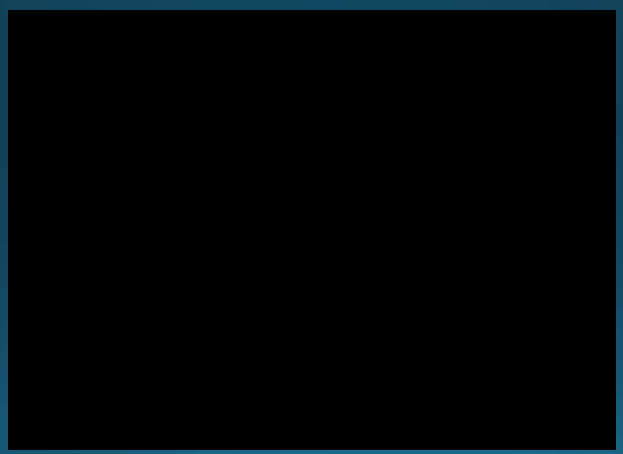
OS

# FA

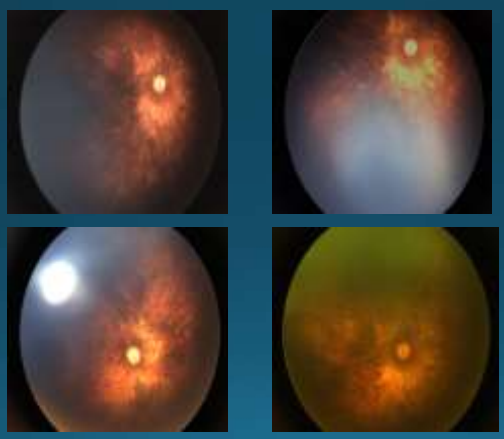
OD OS



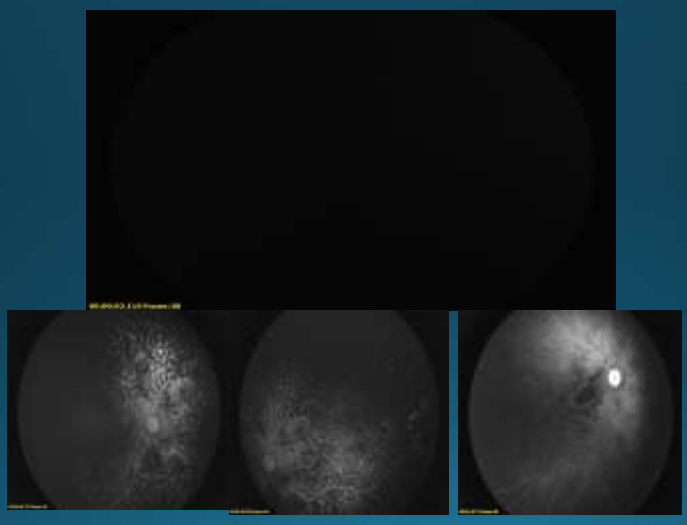
# Vitrectomy



# Post operative photos

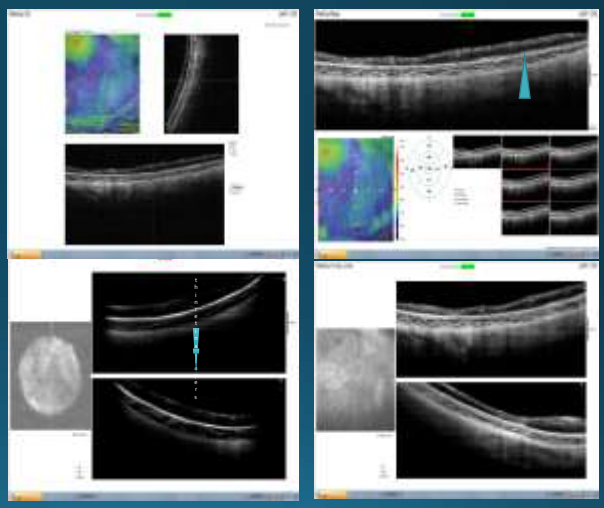


# FA



## OCT findings

- No definite boundaries of external limiting membrane ,IS and OS
- Thin retinal layers and macula
- It could be a retinal dystrophy



## ERG Report

**Name:** Infant Mohamed Ahmed Mohamed Elsayed
**Birth date:** 08/06/2017 **Date of examination:** 07/12/2017
**ID no.:** 17-11574
**Referred by:** Professor Dr Adel Aley Eldin

**Summary:**
Flash ERG examination was done for both eyes under photopic conditions and after 2 minutes of dark adaptation. It showed the following:

**Right eye:**
There were normal a, and b waves under photopic conditions.
There was a following response to 30 Hz flicker.
There were normal a, and b waves under scotopic conditions.

**Left eye:**
There were normal a, and b waves under photopic conditions.
There was a following response to 30 Hz flicker.
There were normal a, and b waves under scotopic conditions.

**Conclusion:**
Flash ERG examination shows no definite abnormality of rod and cone mediated functi on both sides. Follow up flash ERG examination is recommended

**Signature:** Amira El Gohary, MD
ISCEV Member

## VEP Report

**Name:** Infant Mohamed Ahmed Mohamed Elsayed
**Birth date:** 08/06/2017 **Date of examination:** 07/12/2017
**ID no.:** 17-11574
**Referred by:** Professor Dr Adel Aley Eldin

**Summary:**
Flash VEP examination was done for each eye separately according to the ISCEV Standards (latest update).
The examination was done by the Ganzfeld stimulator using a flash frequency of 8 Hz.
The examination was done twice for each eye to test for reproducibility of the response and it showed the following:

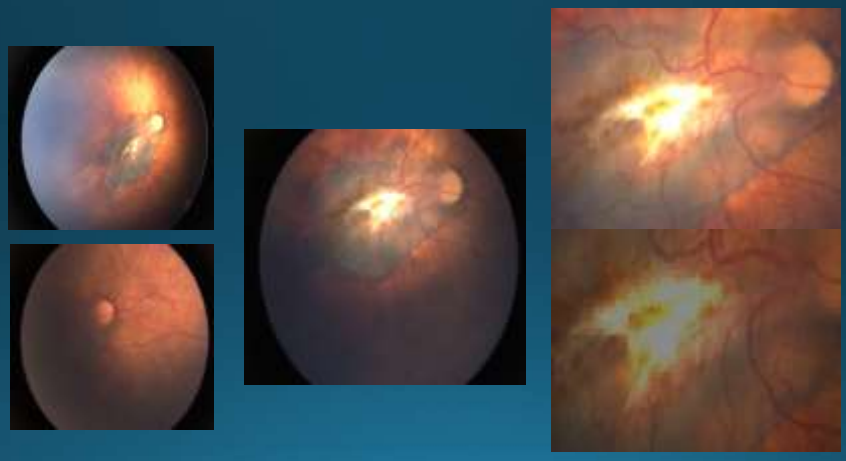
**Right eye:**
There were no consistent or reproducible responses.

**Left eye:**
There were consistent and reproducible responses.

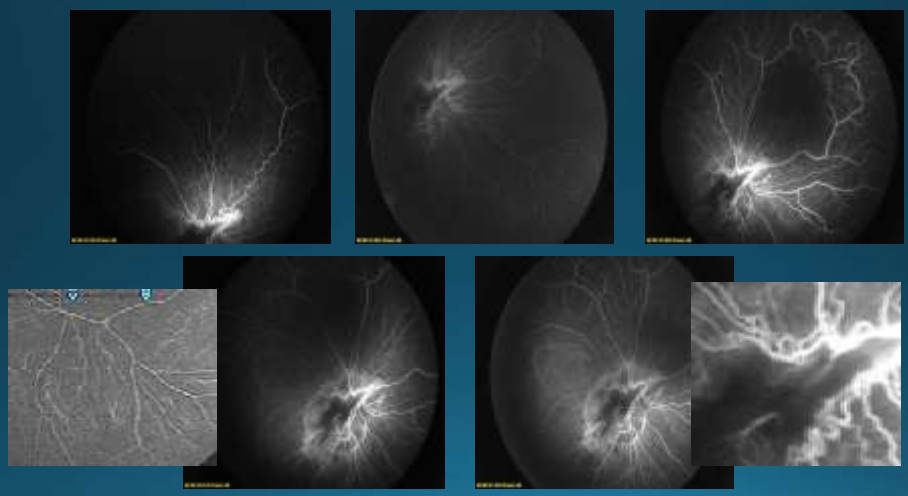
**Conclusion:**
Flash VEP examination shows evidence of an affection of the visual pathway on stimulating the right eye. Stimulating the left eye by flash evokes within normal responses To be correlated with the clinical condition. Follow up VEP examination is recommended

**Signature:** Amira El Gohary, MD
ISCEV Member

Infant aged 6 ms
Presenting with Rt leukocoria



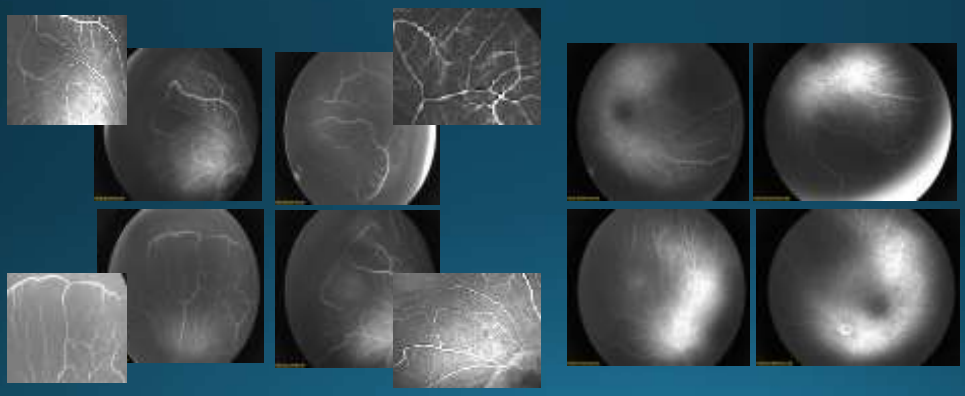
# FA



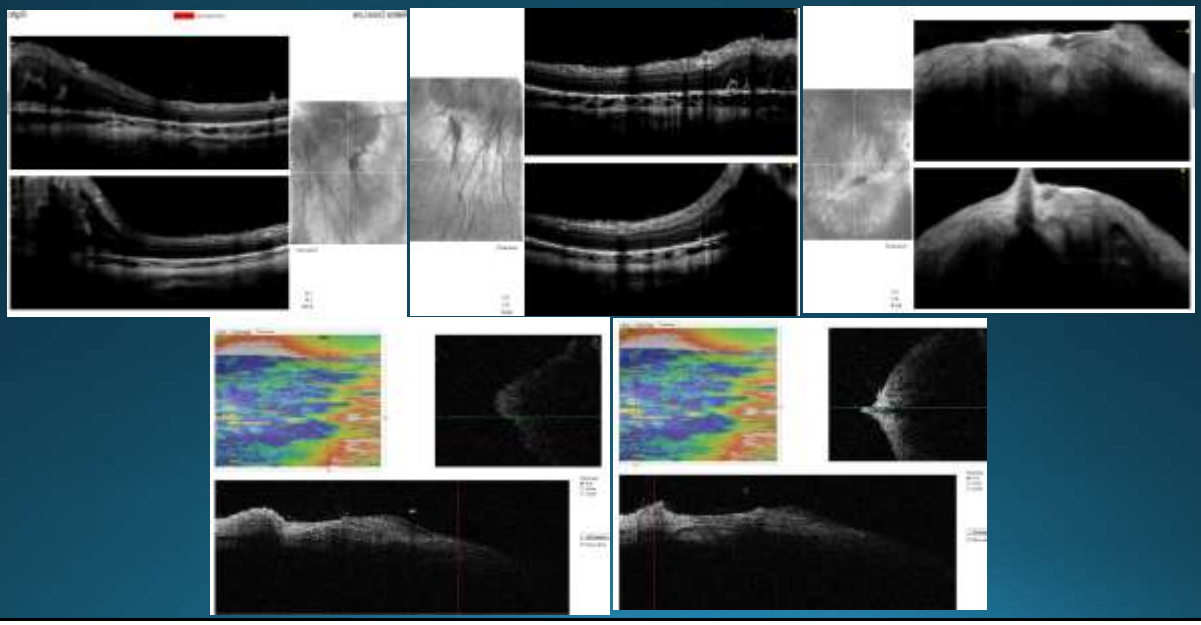
# Abnormal Peripheral vascular pattern

OD

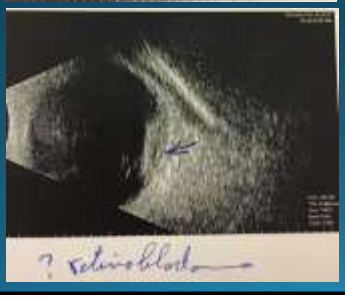
OS NORMAL



# OCT



# US




# Vitreous traction band

- ERG

**Summary:**
Flash ERG examination was done for both eyes under photopic conditions and after 2
minutes of dark adaptation. It showed the following:

**Right eye:**
There were normal a, and b waves as regards latency and amplitude under photopic conditions.
There was a following response to 30 Hz flicker.
There were normal a, and b waves as regards latency and amplitude under scotopic conditions.

**Left eye:**
There were normal a, and b waves as regards latency and amplitude under photopic conditions.
There was a following response to 30 Hz flicker.
There were normal a, and b waves as regards latency and amplitude under scotopic conditions.

**Conclusion:**
Flash ERG examination shows no detectable abnormality of rod and cone mediated function on both sides.

**Summary:**
Flash VEP examination was done for each eye separately according to the ISCEV Standards (latest update).
The examination was done by the Ganzfeld stimulator using a flash frequency of 8 Hz.
It showed the following:

**Right eye:**
There were consistent and reproducible responses.

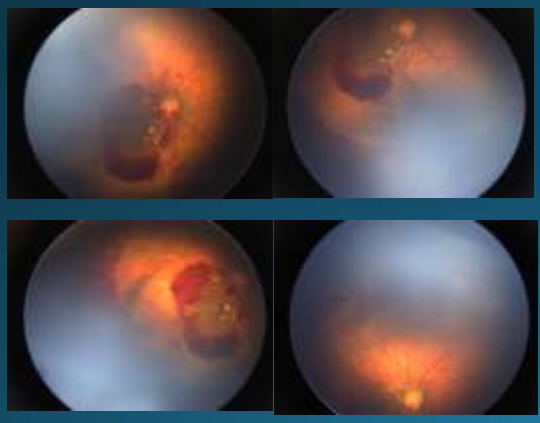
**Left eye:**
There were consistent and reproducible responses.

**There was no significant interside latency or amplitude difference.**

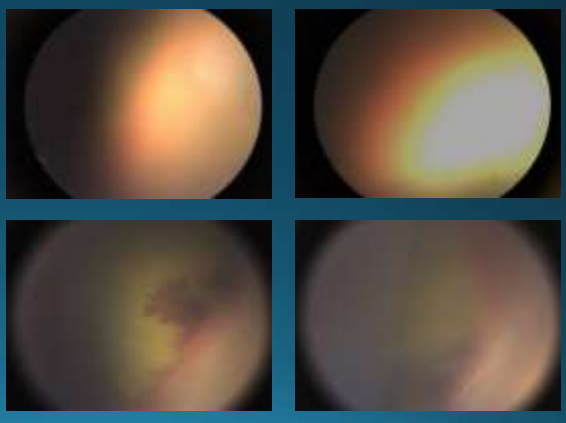
**Conclusion:**
Flash VEP examination shows evidence of a functioning visual pathway on stimulating each eye.

## Infant 4 ms of age presenting with leukocoria

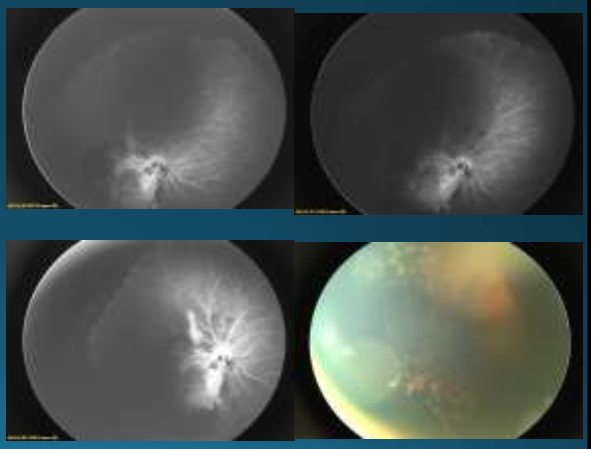
OD



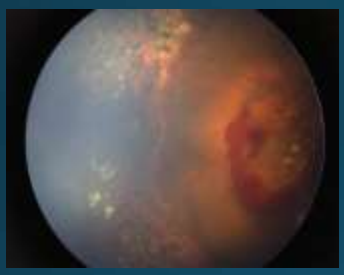
OS

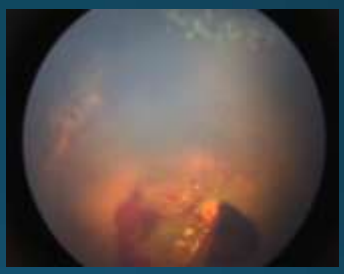


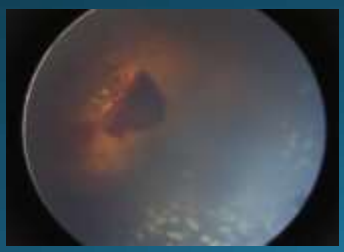
# FA prove diagnosis (FEVR)



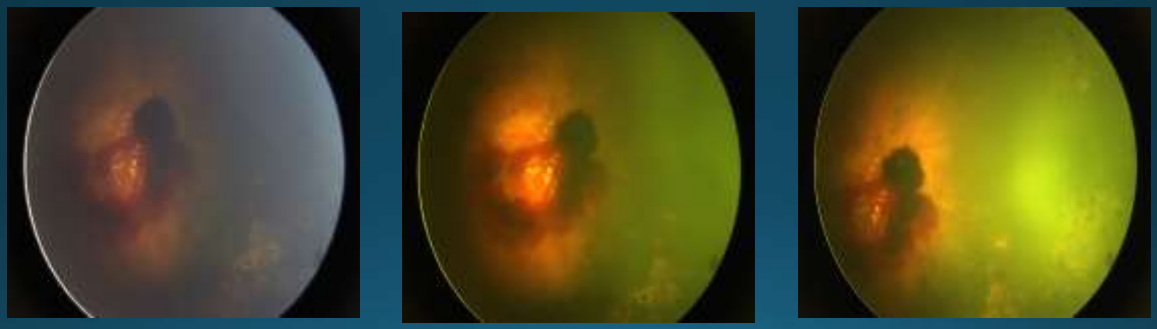
# Continuous laser treatment within 3 weeks



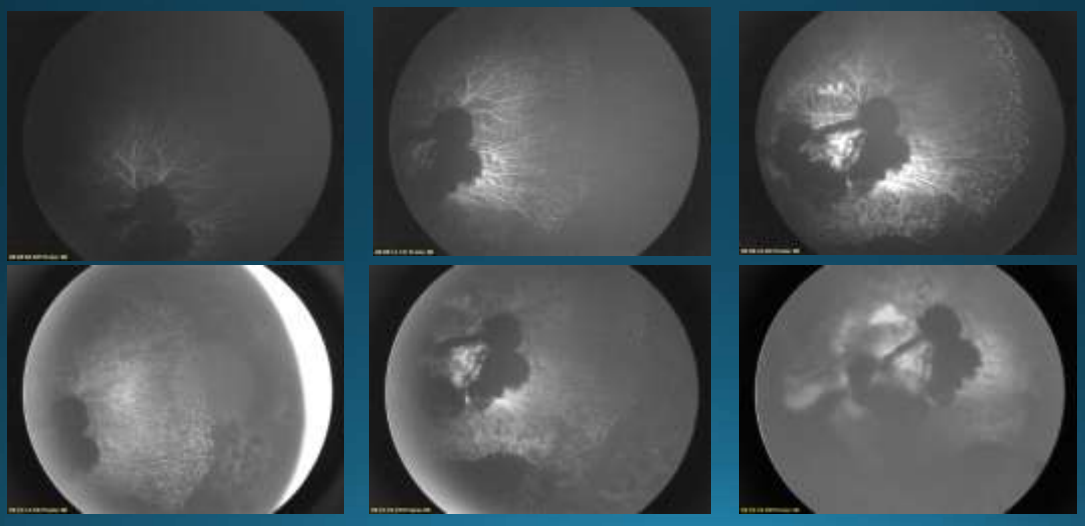




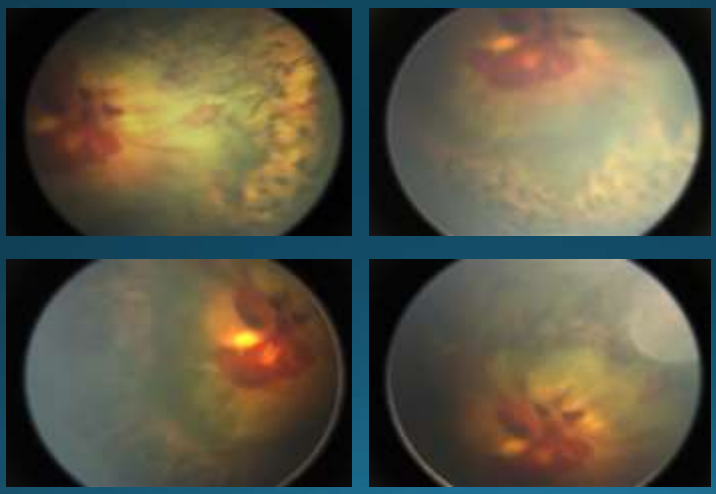
# End of treatment in 3 weeks



# FA



# Follow up another 2 weeks more



# VITRECTOMY

# Thank You