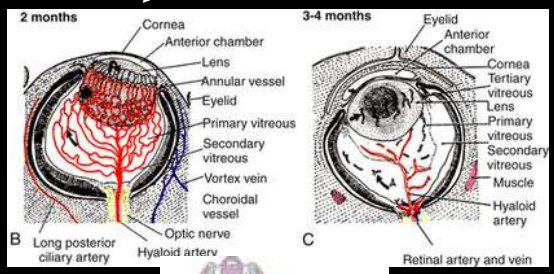
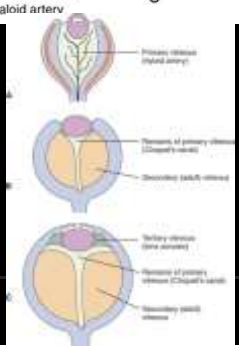
Persistent Fetal Vasculature (PFV)

Mariam AL-Feky, MD, FRCS (Glasgow)
Lecturer Ain Shams University
Watany Eye Hospital

Definition





- congenital anomaly
- failure of embryological primary vitreous and hyaloid vasculature to regress or in the development of the secondary vitreous
- ccc persistence of various portions of the primary vitreous (embryonic hyaloid vascular system) with hyperplasia of the associated embryonic connective tissue, and associated with microphthalmia, cataract and glaucoma

Genetics (OMIM)

- AR persistent hyperplastic primary vitreous (PHPVAR) is caused by homozygous mutation in the ATOH7 gene on chromosome 10q21 or by homozygous deletion in the regulatory region of the ATOH7 gene.

Non-syndromic , (+/- consanguinity), Ghiasvand et al. (1998)

AS dydgenesis, Severe FEVR, Retinal dysplasia

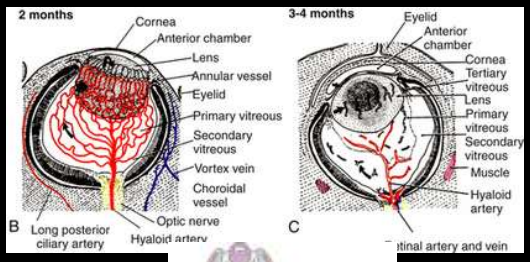
- AD form of PHPV has been described (PHPVAD)

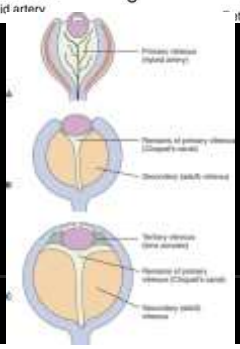
Lin et al. (1990) Galal et al. (2006)

- PHPV shares phenotypic overlap with Norrie disease esp if sporadic with no systemic associations Pendergast et al. (1998)

NDP gene which encodes norrin, on Xp11 (XLR)

Classifcation





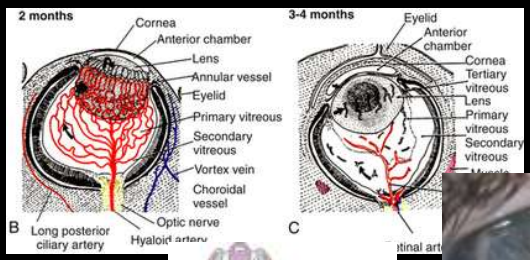
Anatomical Classification

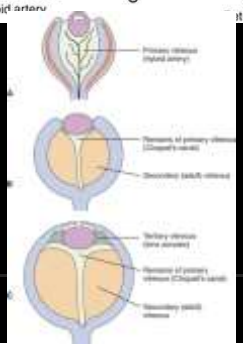
- Anterior
- Posterior
- Combined or complex:
  - I shape
  - Y shape
  - Inverted Y shape
  - X shape

US classification

- Can be isolated (unilateral) or associated with other anomalies or systemic diseases (bilateral)

Classifcation



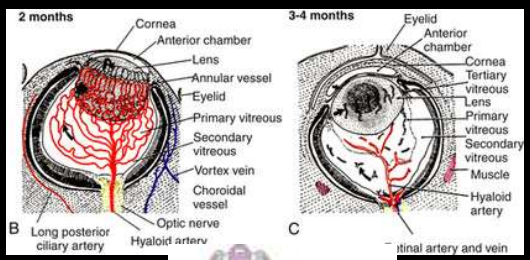


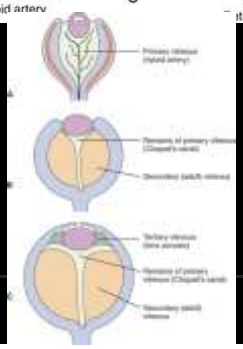
Anatomical Classification

- Anterior
  - Persistent pupillary membrane (persistent tunica vasulosa lentis)

  TTT: No/ Mydriatic / Surgical excision or YAG Lysis

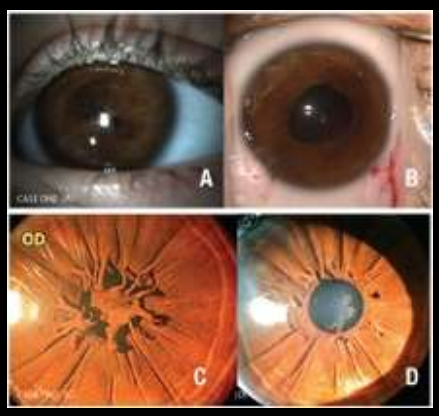
Classifcation



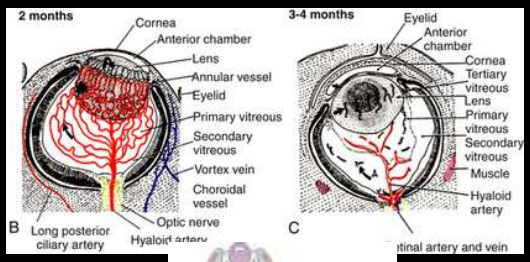


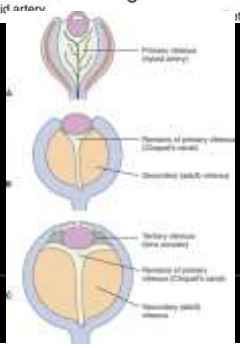
Anatomical Classification

- Anterior
  - Persistent pupillary membrane (persistent tunica vasulosa lentis)
  - TTT: No/ Mydriatic / Surgical excision or YAG Lysis



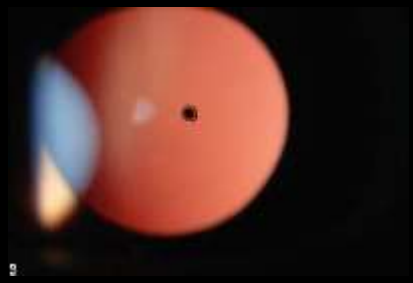
Classifcation



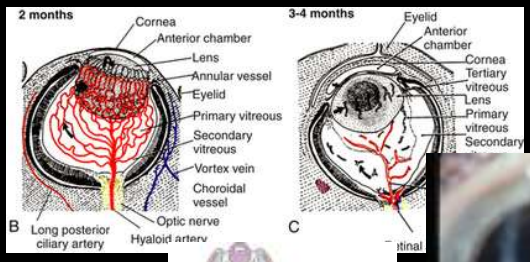


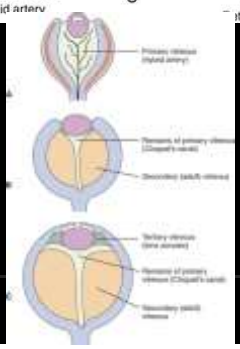
Anatomical Classification

- Anterior
  - Mittendorf dot (small, circular opacity on the posterior lens capsule, classically nasal in location, which represents the anterior attachment of the hyaloid artery)



Classifcation

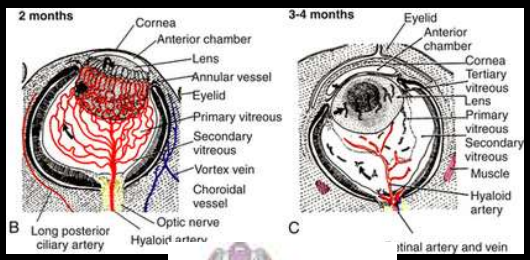


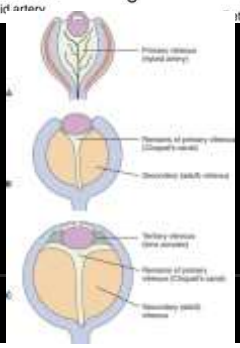


Anatomical Classification

- Anterior
  - The anterior end of the hyaloid remnant divides as it attaches to the back of the lens forming a so-called starfish

Classifcation

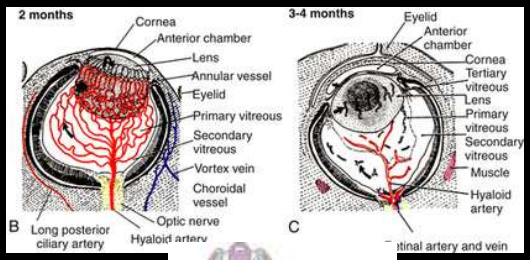


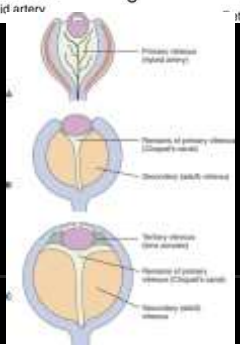


Anatomical Classification

- Anterior
  - Posterior polar cataract (Surgey)

Classifcation

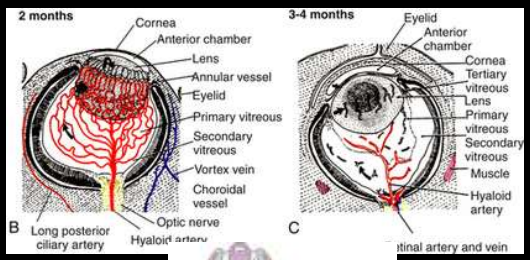


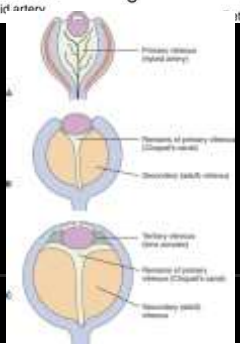


Anatomical Classification

- Anterior
  - Notched edge of the lens or Coloboma (disruption in zonule due to PFV)

Classifcation

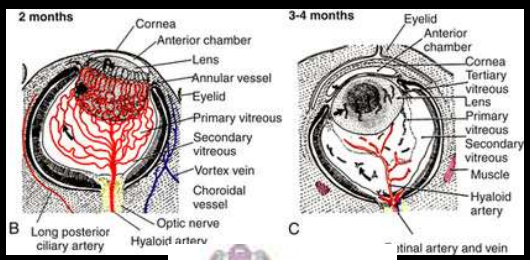


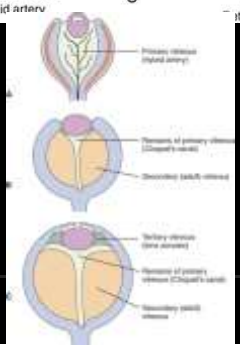


Anatomical Classification

- Anterior
  - Dense retrolental membrane (either white or vascularized) with prominent ciliary processes

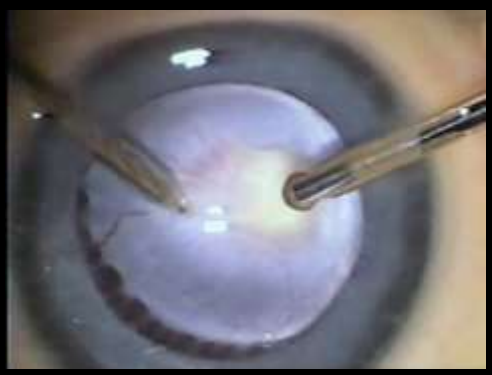
Classifcation



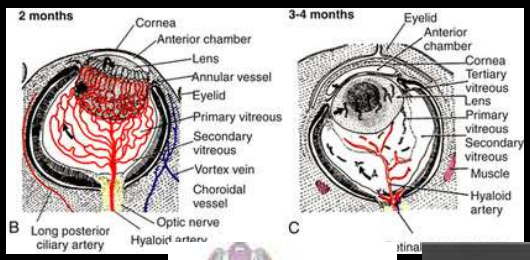


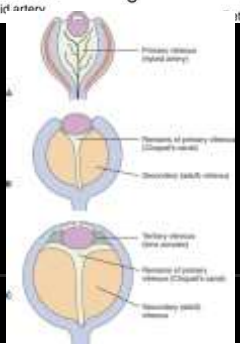
Anatomical Classification

- Anterior
  - Dense retrolental membrane (either white or vascularized)



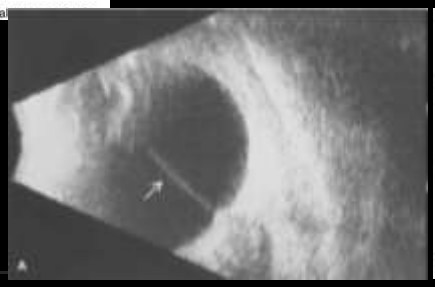
Classifcation





Anatomical Classification

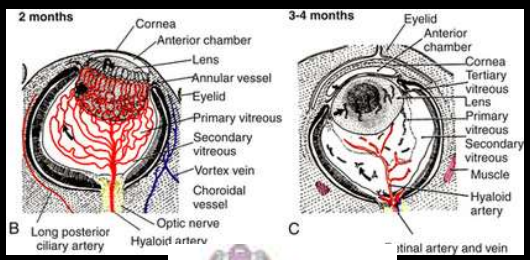
- Anterior
  - Dense retrolental membrane…. B-scan is mandatory to exclude posterior form and to exclude other pathology (RB, ROP, FEVR)

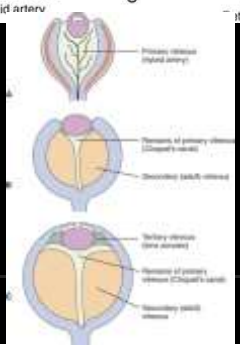


Persistent hyaloid

Persistent hyaloid + RD

Classifcation

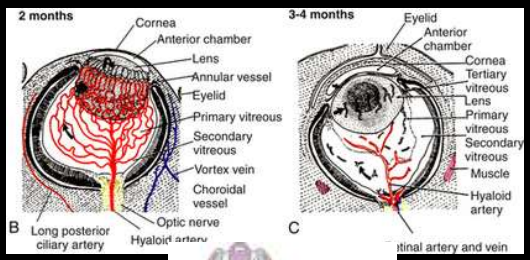


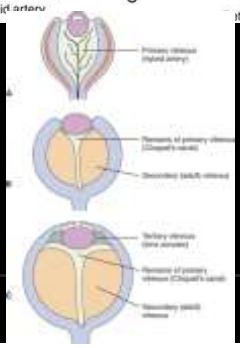


Anatomical Classification

- Anterior
  - In cataract take care the hyaloid may still be attached it has to be cauterized

Classifcation

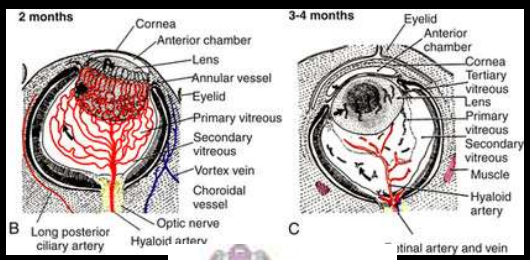


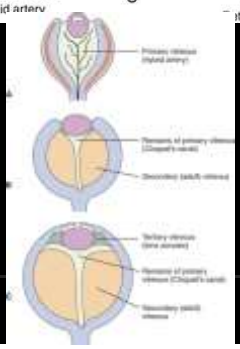


Anatomical Classification

- Anterior
  - Dense retrolental membrane (cut and removed with vitrector after B-scan evaluation)

Classifcation

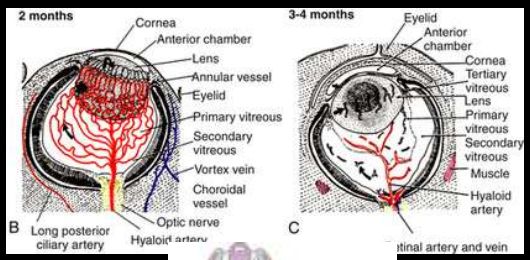


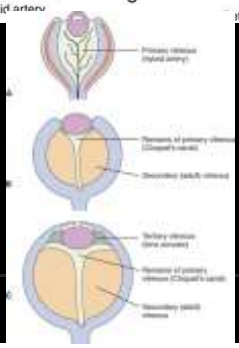


Anatomical Classification

- Anterior
  - Dense retrolental membrane (cut and removed with vitrector after B-scan evaluation)

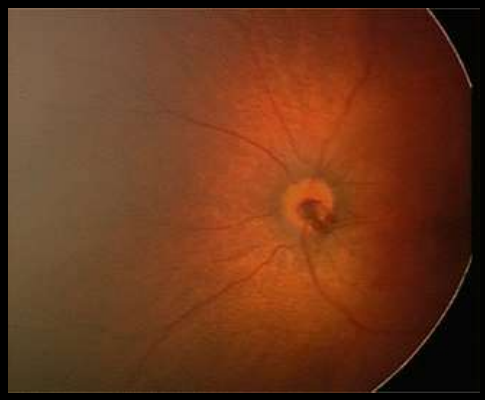
Classifcation



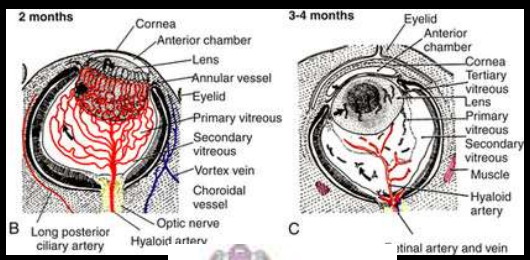


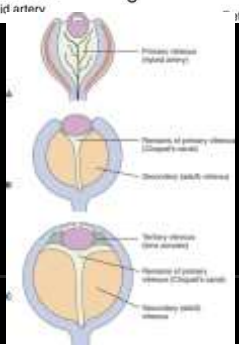
Anatomical Classification

- Posterior
  - Persistent hyaloid (simple)



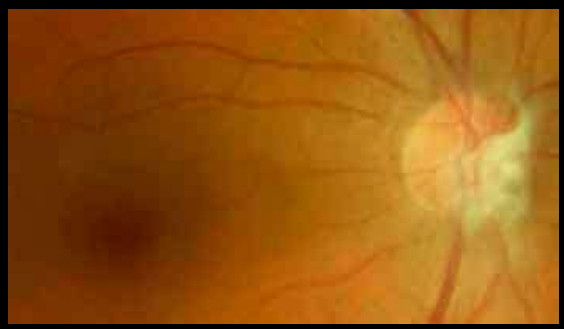
Classifcation



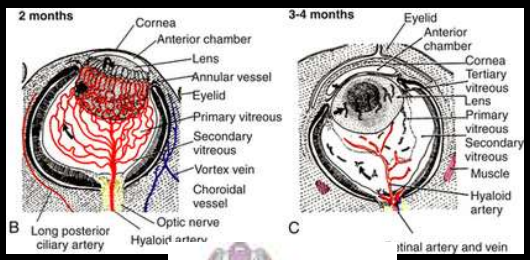


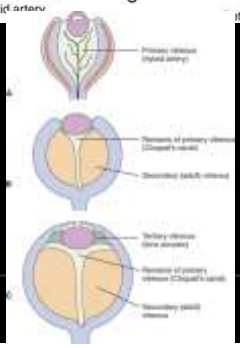
Anatomical Classification

- Posterior
  - Burgmeister papilla (simple)



Classifcation





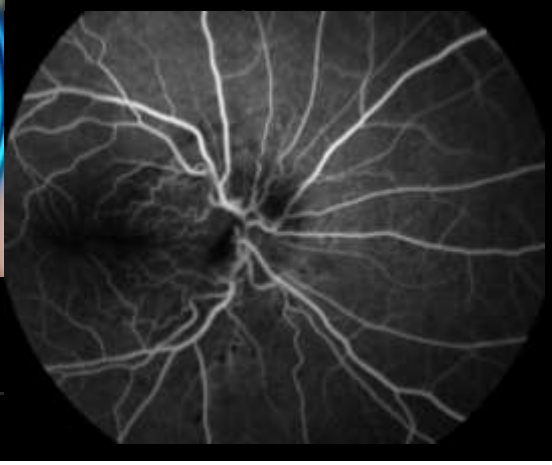
Anatomical Classification

- Posterior
  - Burgmeister papilla (macular dragging)

Classifcation

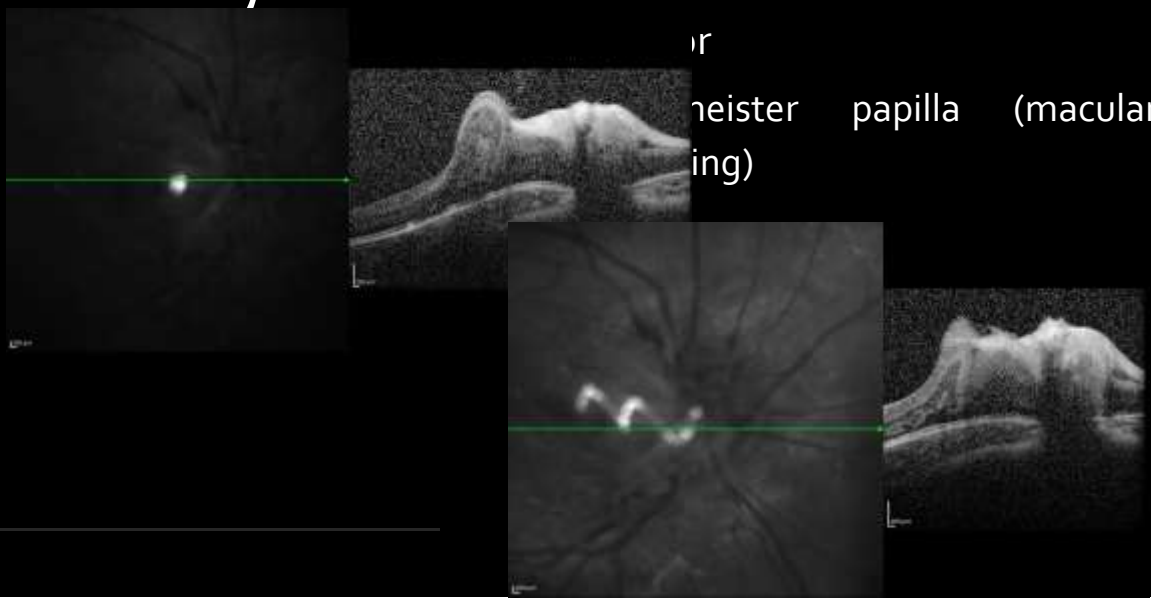
Anatomical Classification

- Posterior
  - Burgmeister papilla (macular



Classifcation

Anatomical Classification



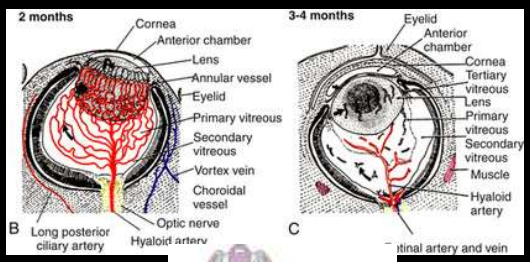
r

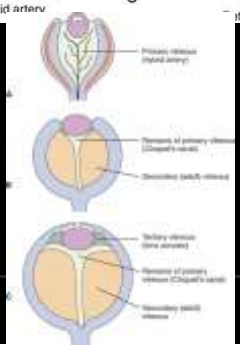
neister papilla (macular

ing)

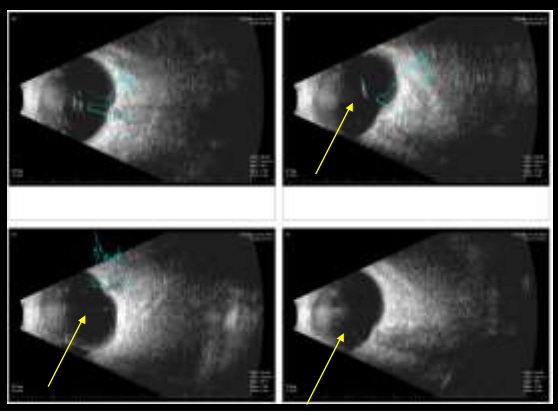
Classifcation

Anatomical Classification





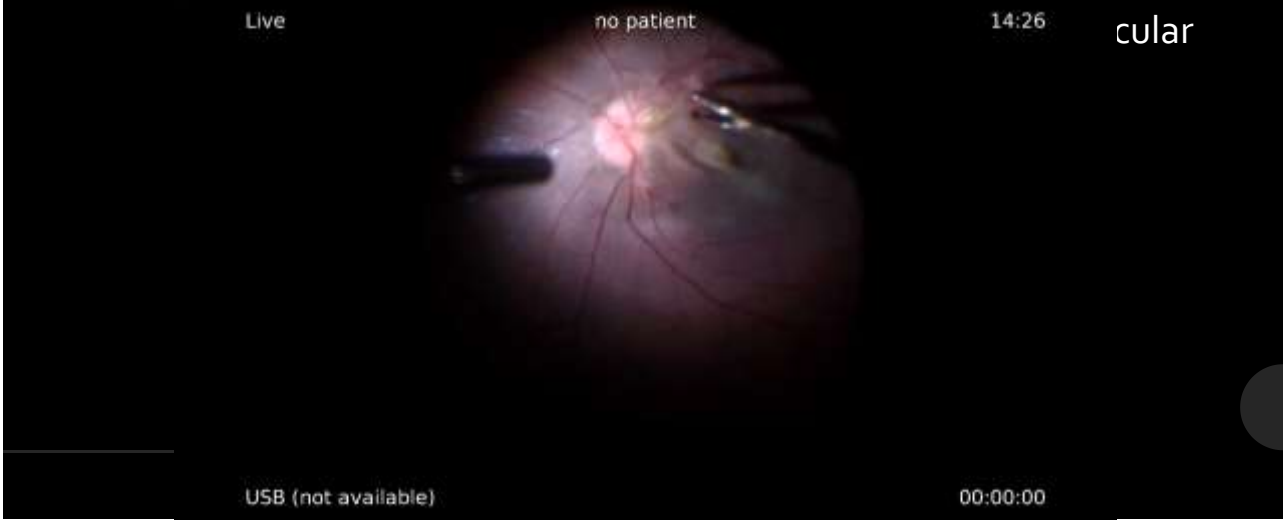
- Posterior
  - Burgmeister papilla (macular dragging)



Classifcation

Anatomical Classification

- Posterior



cular

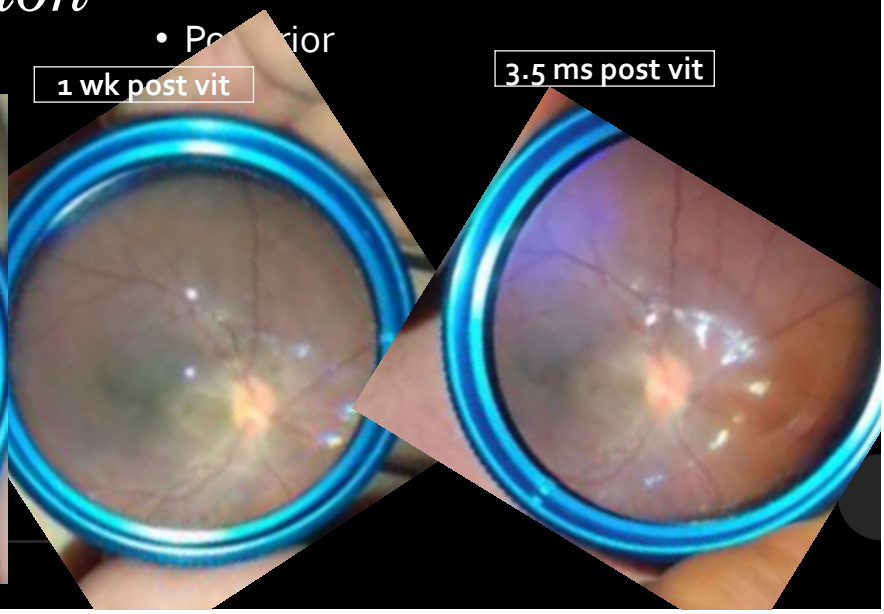
Classifcation

Anatomical Classification

- Po rior

Pre

1 wk post vit



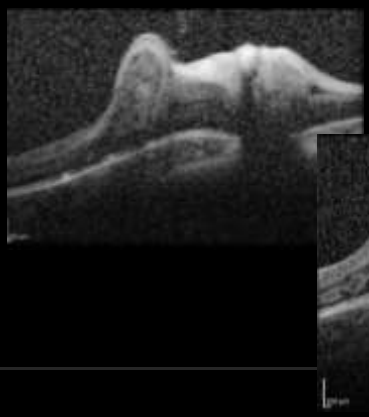
3.5 ms post vit

Classifcation

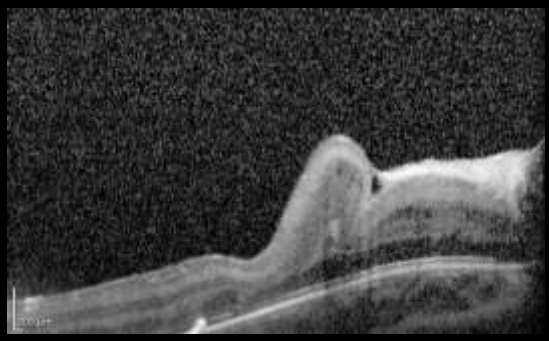
Anatomical Classification

- Posterior

Pre



1 .5 ms post vit



Classifcation

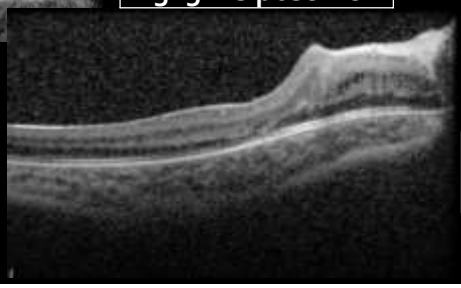
Anatomical Classification

- Posterior

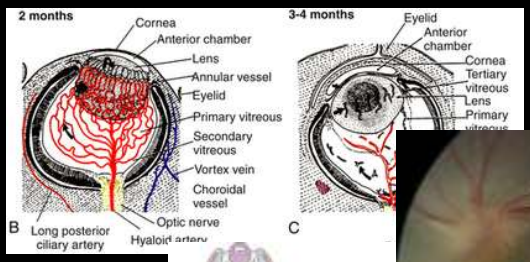
Pre

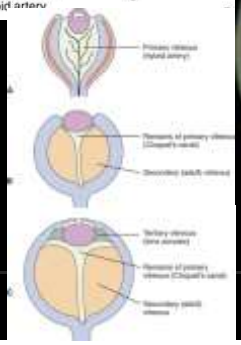
1 .5 ms post vit

3 .5 ms post vit



Classifcation

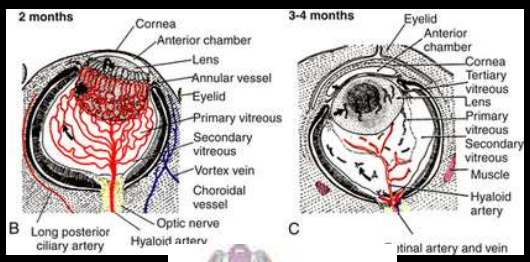


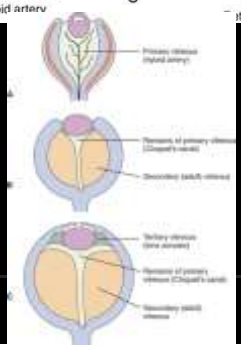


Anatomical Classification

- Posterior
  - With retinal dragging but stalk not reaching the lens

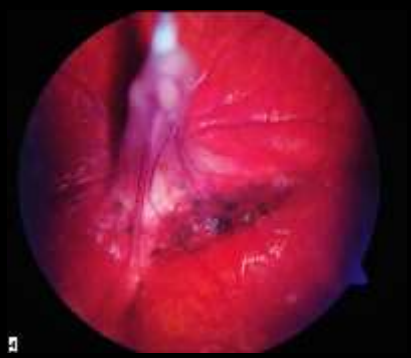
Classifcation





Anatomical Classification

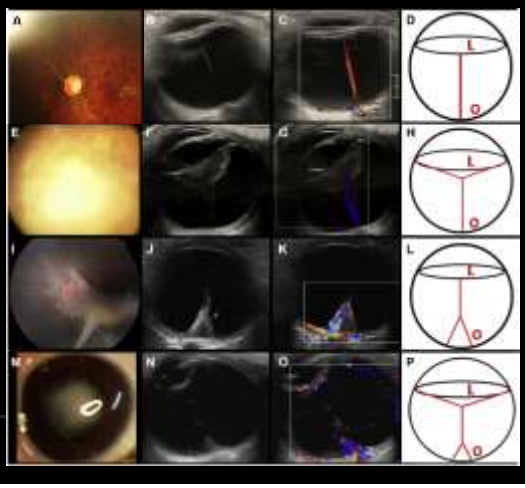
- Posterior
  - With retinal dragging but stalk not reaching the lens



Classifcation

Anatomical Classification

- Combined
  - Stalk extending from lens to retina

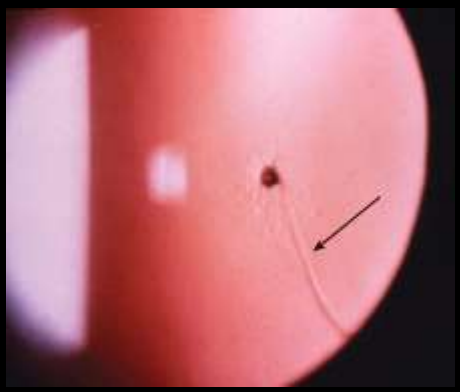


Classifcation

Anatomical Classification

- Combined
  - Stalk extending from lens to retina

    I shape

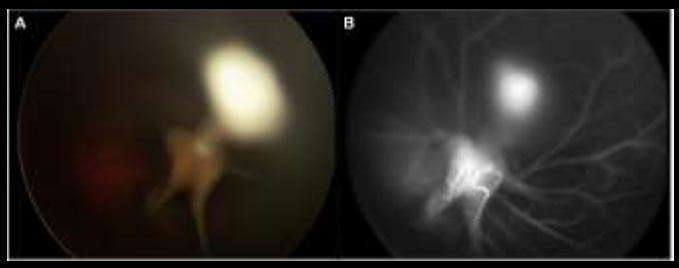


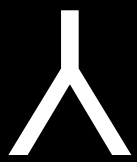
Classifcation

Anatomical Classification

- Combined
  - Stalk extending from lens to retina

Inverted Y

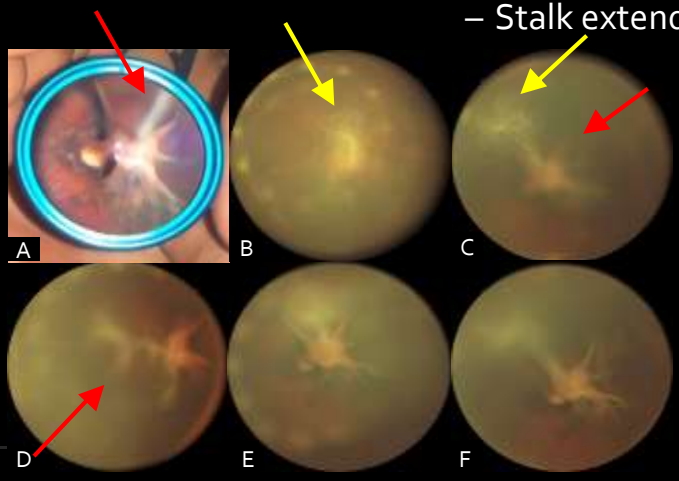




Classifcation

Anatomical Classification

- Combined
  - Stalk extending from lens to retina



Classifcation

Anatomical Classification

- Posterior and combined forms

  PPV +/- Lensectomy

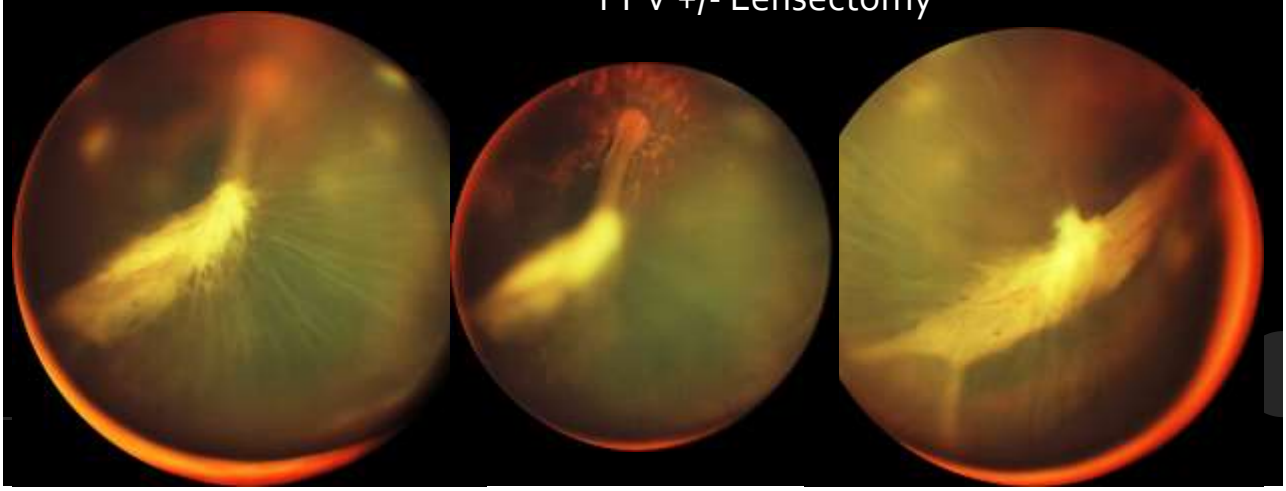
Prof. Singuel Ozdek case

Classifcation

Anatomical Classification

- Posterior and combined forms

  PPV +/- Lensectomy



Classifcation

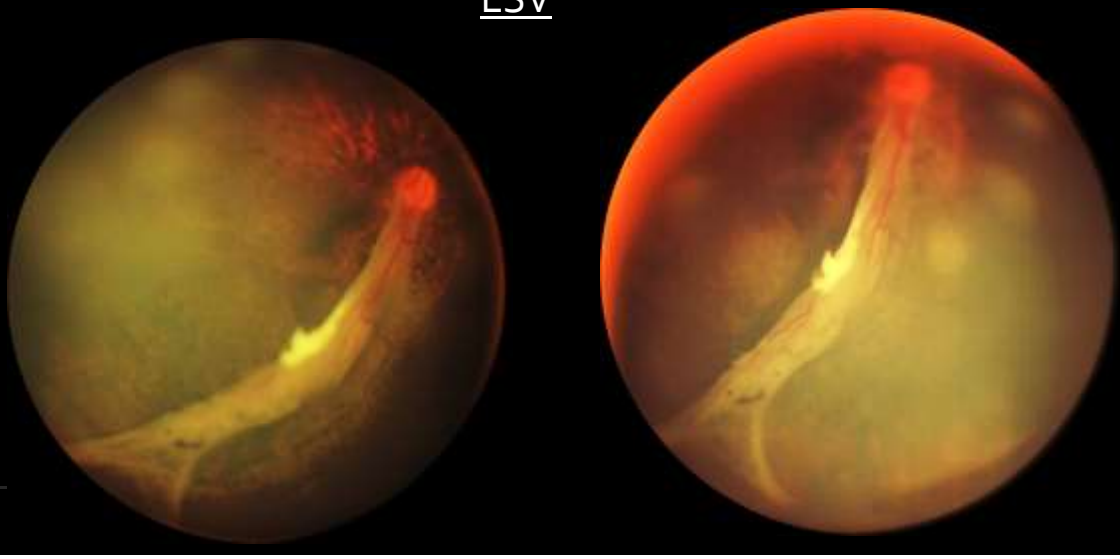
Anatomical Classification

LSV

Classifcation

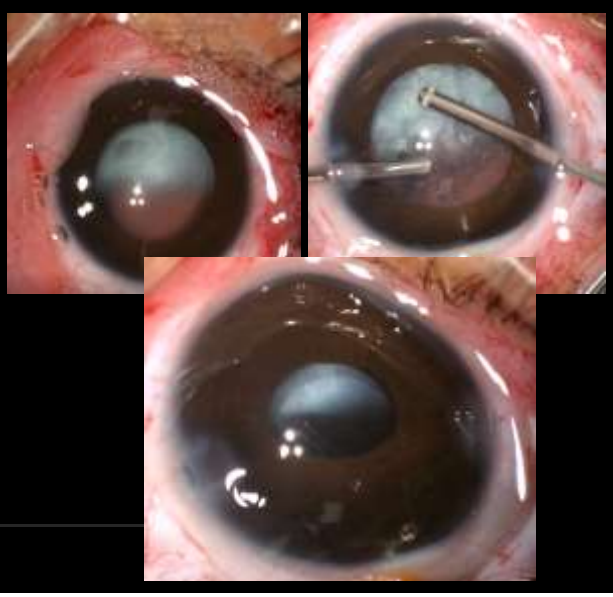
Anatomical Classification

LSV

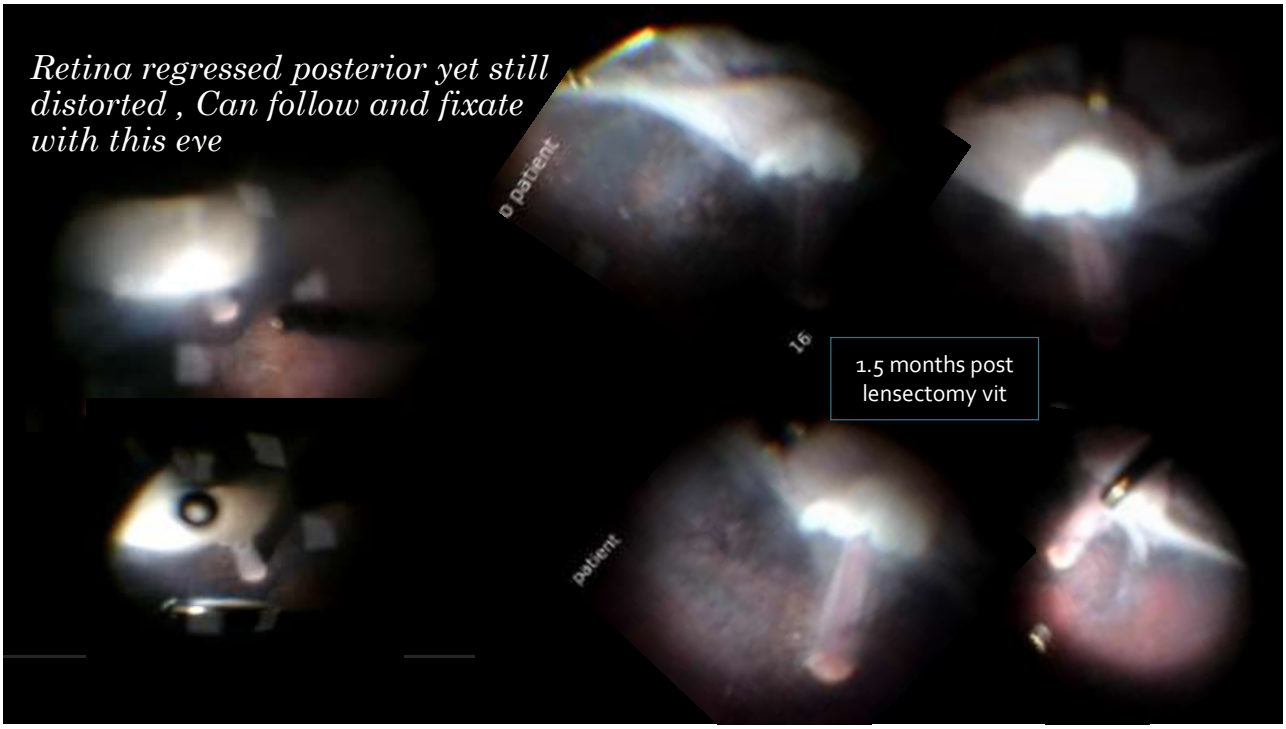


Severe form

Lensectomy + PPV + Membranectomy



Retina regressed posterior yet still distorted , Can follow and fixate with this eve



Classifcation

Anatomical Classification

- Posterior and combined forms

  PPV +/- Lensectomy

  ❖Ant. Insersion of the Ora serrata

Classifcation

Anatomical Classification

- Posterior and combined forms

  PPV +/- Lensectomy

  ❖Ant. Insersion of the Ora serrata

Classifcation

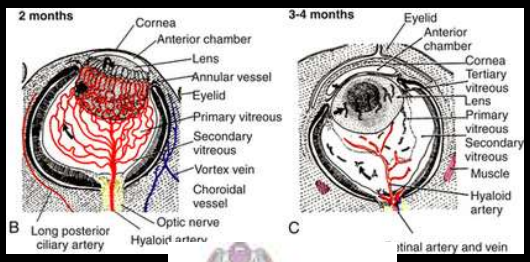
Anatomical Classification

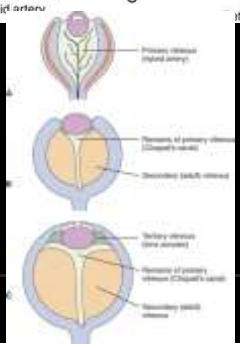
- Posterior and combined forms

  PPV +/- Lensectomy

  ❖Ant. Insersion of the Ora serrata

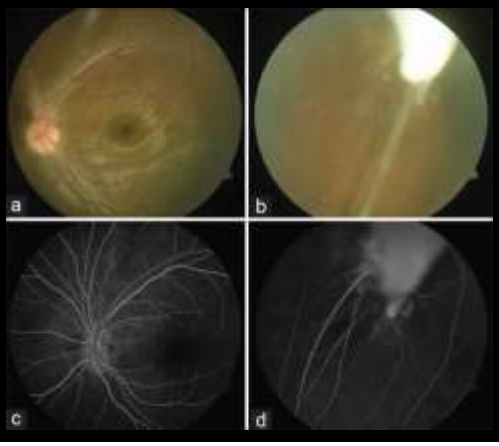
Classifcation



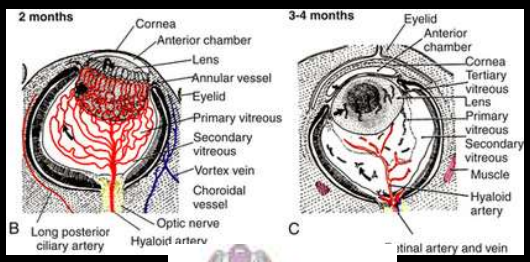


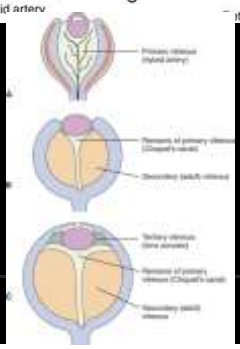
Anatomical Classification

- Atypical forms
  - Stalk extending from disc to peripheral retina



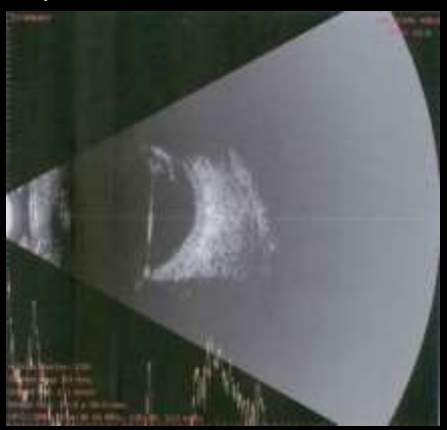
Classifcation





Anatomical Classification

- Atypical forms
  - Stalk extending from disc to peripheral retina

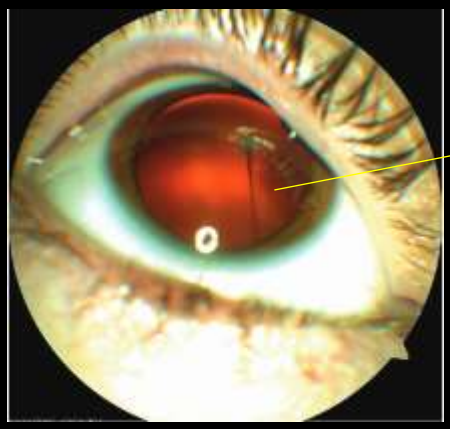


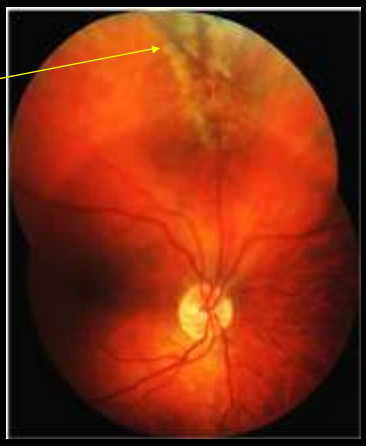
Classifcation

Anatomical Classification

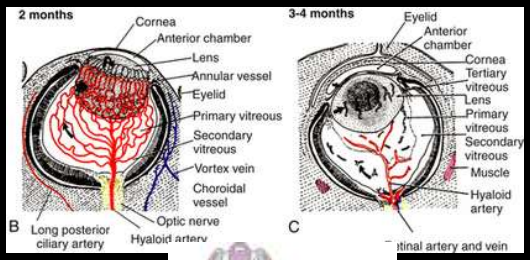
- Atypical forms

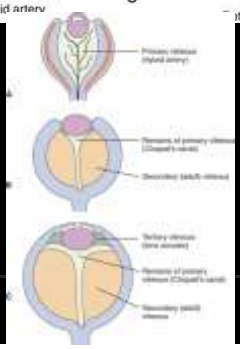
there are residues of persistent hyaloid artery with fibrotic tissue, causing local retinoschisis of an older date





Classifcation





Anatomical Classification

- Atypical forms

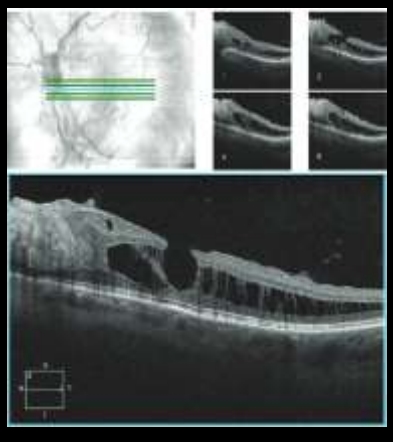
there are residues of persistent hyaloid artery with fibrotic tissue, causing local retinoschisis of an older date

Classifcation

Anatomical Classification

- Atypical forms

there are residues of persistent hyaloid artery with fibrotic tissue, causing local traction



Classifcation

Anatomical Classification

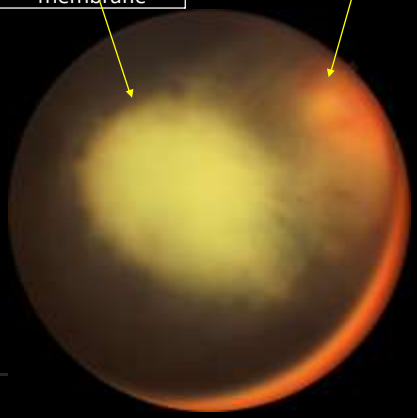
- Atypical forms

Retinal dystrophy (Norrie disease or other syndromes) especialy if bilateral

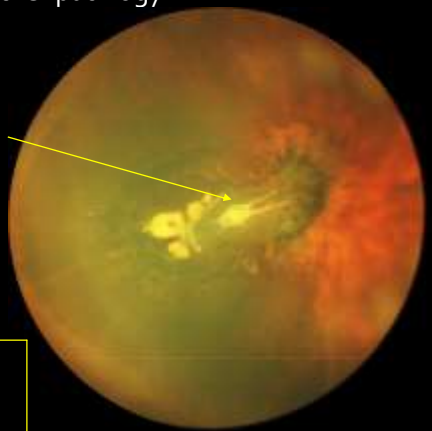
Associated with other pathlogy

Cataract + Retrolental membrane

Retina behind the lens



Central stalk dragging the disc with TRD, subretinal exudation and distorted retina



Bilateral + Dystrophic retina :
? Norrie disease

Classifcation

Anatomical Classification

- Atypical forms

PFV + Multiple congenital anomalies (hydrocephalus, renal impairment)

Retinal dystrophy (Norrie disease or other syndromes) especialy if bilateral

Associated with other pathlogy (ROP, Morning glory, coloboma)

Vascularized retrolental membrame

Vitreous cavity full of opacified strands attached to retina and causing traction

Classifcation

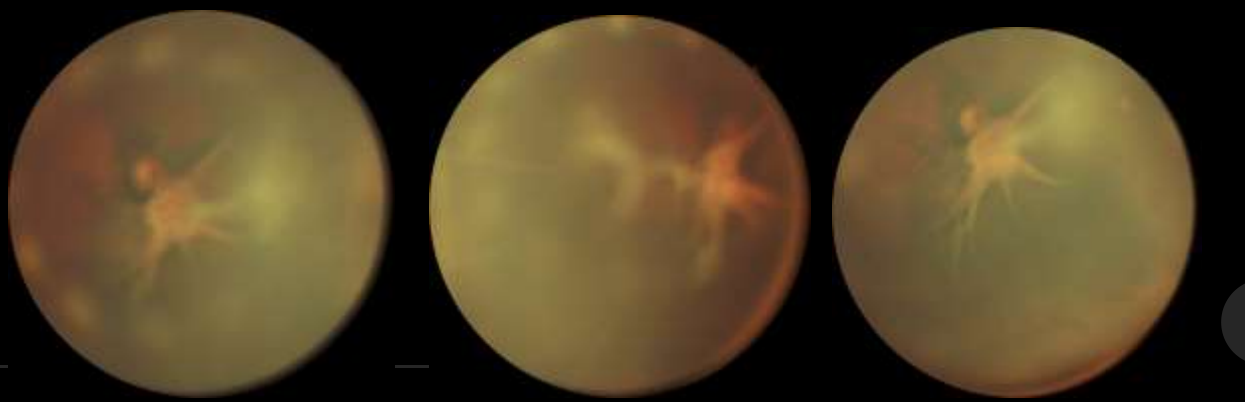
PFV + ROP

Anatomical Classification

- Atypical forms

Retinal dystrophy (Norrie disease or other syndromes) especialy if bilateral

Associated with other pathlogy (ROP, Morning glory, coloboma)



Classifcation

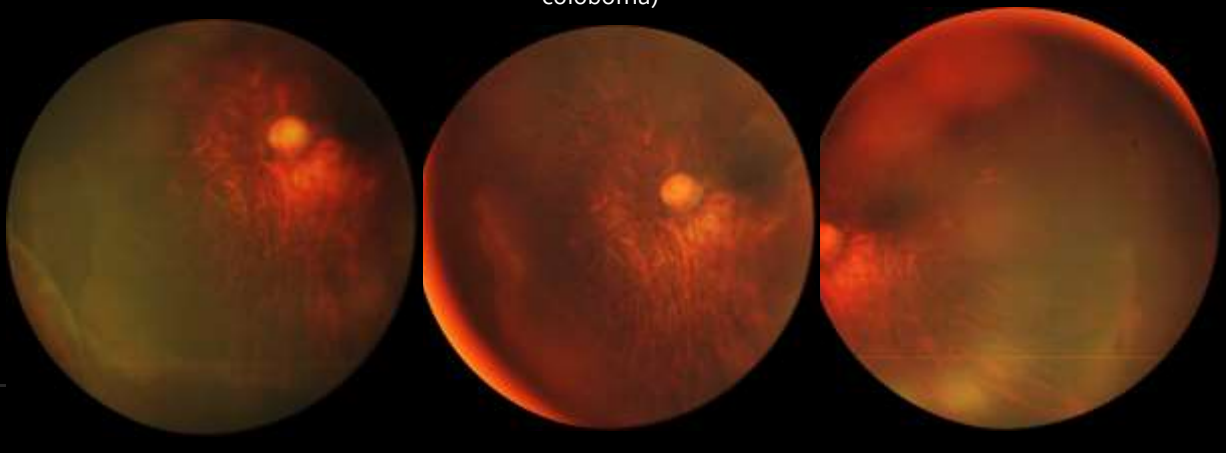
PFV + ROP

Anatomical Classification

- Atypical forms

Retinal dystrophy (Norrie disease or other syndromes) especialy if bilateral

Associated with other pathlogy (ROP, Morning glory, coloboma)



Classifcation

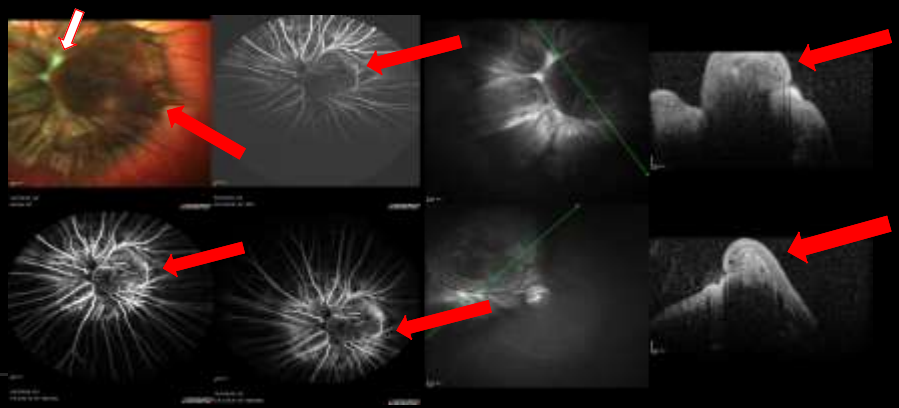
PFV + Morning glory syndrome

Anatomical Classification

- Atypical forms

Retinal dystrophy (Norrie disease or other syndromes) especialy if bilateral

Associated with other pathlogy (ROP, Morning glory, coloboma)



Take home Msg

- PFV has 3 types (AR, AD)
- Can be associated with other congenital anomalies
- If bilateral exclude systemic diseases
- If with retinal dystrophy suspect Norrie disease
- In PPV, examine periphery well