Paediatric Retinal Diseases

Symptoms of Retinal Diseases

- Diminution of vision without pain
- Night blindness
- Photopsia- *sparks or lightening flashes*
- Metamorphopsia- *distorted images*
  micropsia & macropsia
- Peripheral constriction of visual field
- Patient unaware of symptoms

Symptoms of Retinal Diseases in Paediatric cases

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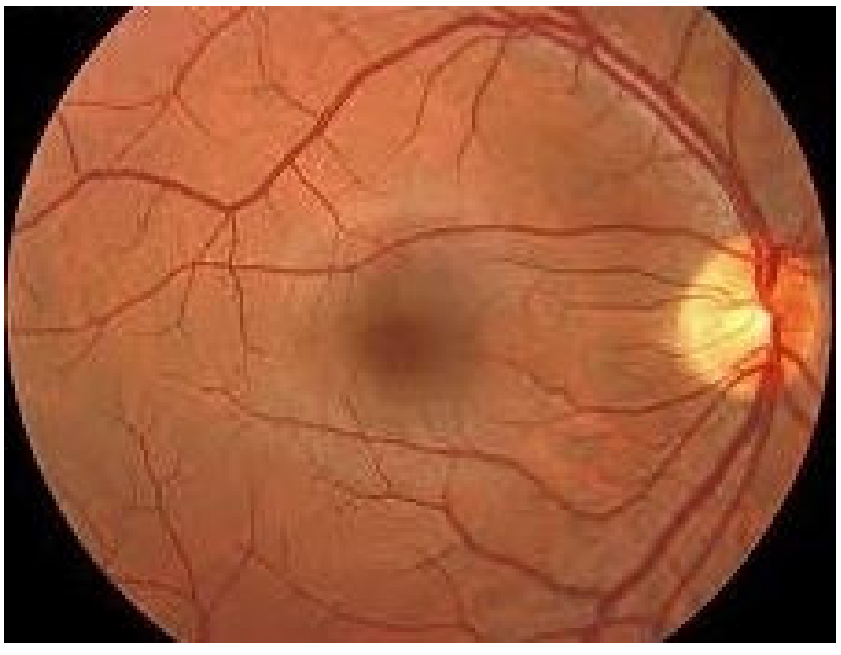
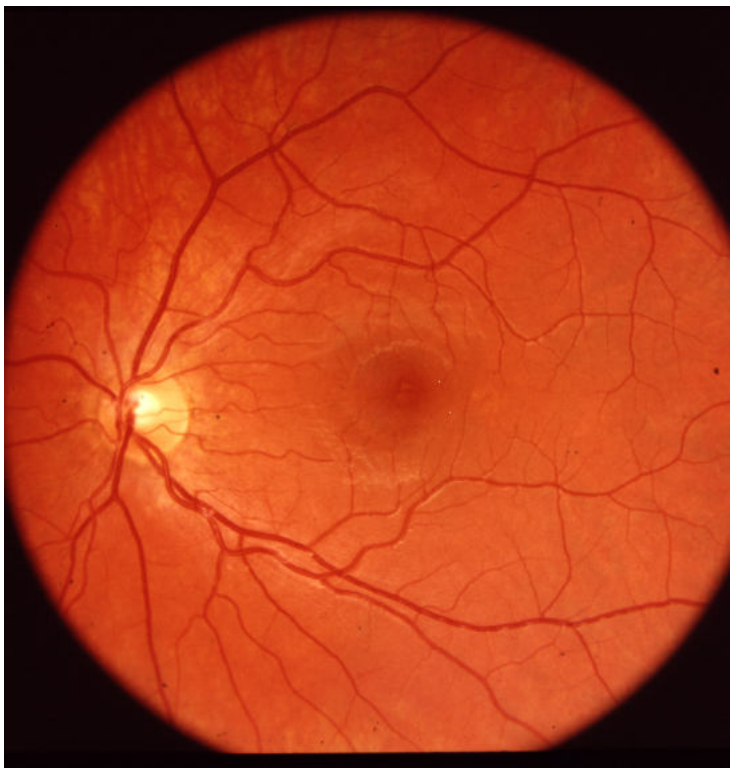
- Night blindness
- Photopsia- sparks or lightening flashes
- Metamorphopsia- distorted images
- micropsia & macropsia
- Peripheral constriction of visual field
- Patient unaware of symptoms
- Jerky movements of eyes[Nystagmus]

Normal Fundus





What do we examine in fundus?

- Fundal glow/ ocular media
- Optic disc
- Retinal blood vessels
- General (Background) fundus
- Macula & foveal reflex
- Peripheral fundus

Examination of Fundus (Ophthalmocopy)

- Distant direct ophthalmoscopy
- Direct ophthalmoscopy
- Indirect ophthalmoscopy
- Slit lamp assisted Fundus examination-

i] Hruby lens

ii] +90 D / +78 d lens

iii] 3-mirror contact lens

Types of Retinal diseases in Paediatric age group

- Retinopathy of Prematurity
- Retinitis Pigmentosa
- Phakomatosis
- Coat's disease
- Hereditary Dystrophies of central retina and choroid
- Myelinated nerve fibres

Retinopathy of Prematurity(ROP)

- B/L abnormality
- Retinal neovascularization
- Premature infants

➢ <1500 gm

➢ <32 week gestational age

➢ Given high conc of Oxygen during first 10 days of life

ROP -Signs

Dilatation of retinal vessels

+

hazy white patches in peripheral retina (temporal)

↓

Fibrous tissue

↓

Proliferation & rolling over (Pseudo glioma)

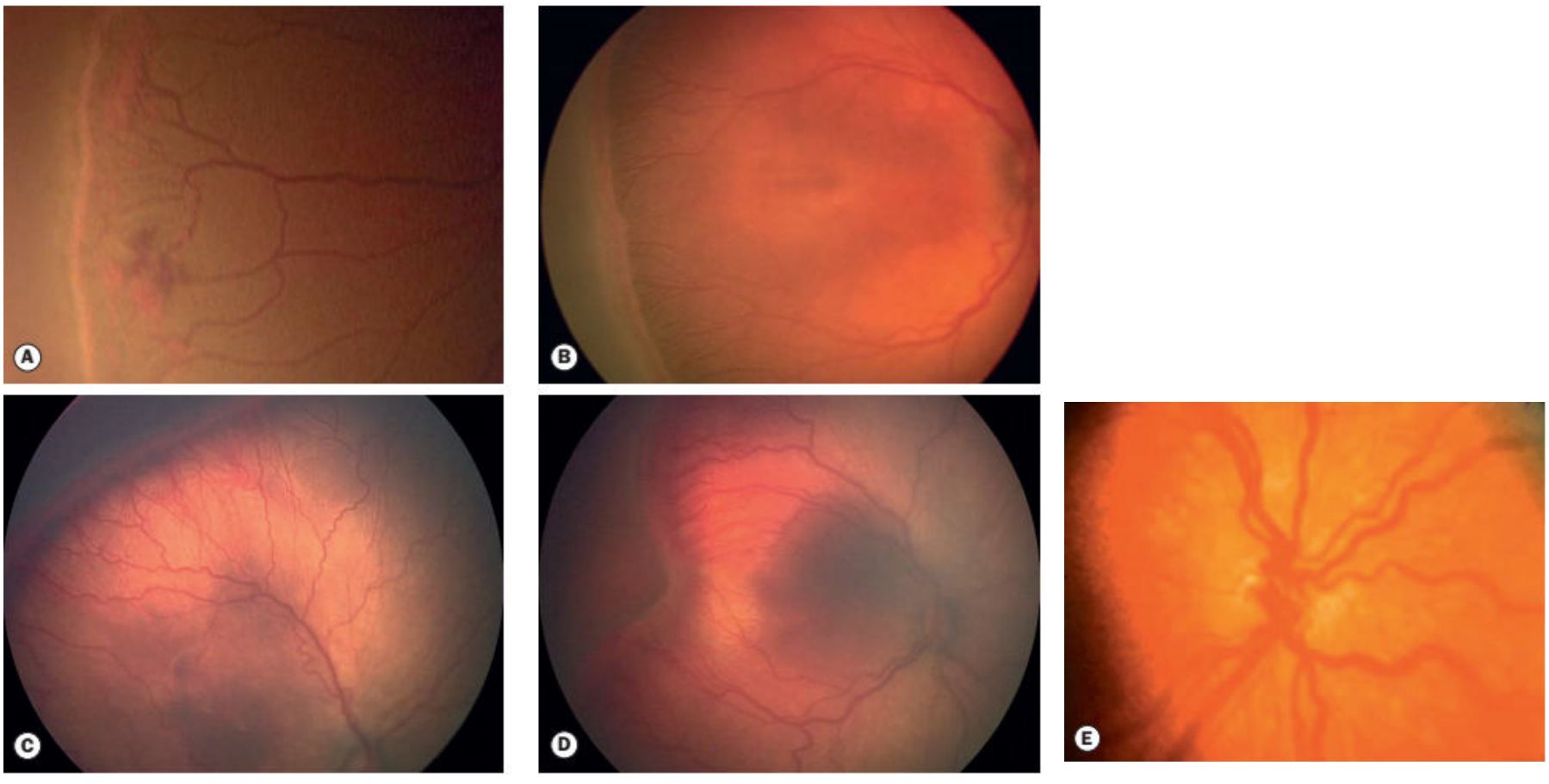
↓

Retinal detachment (Lost vision)

ROP Staging

- Stage 1= a faint demarcation line
- Stage 2= an elevated ridge
- Stage 3= extraretinal fibrovascular tissue
- Stage 4= sub-total retinal detachment
- Stage 5= total retinal detachment

ROP



ROP- Treatment

- 80% infants spontaneous regression
- Treatment required if disease progresses

  i] Photocoagulation/ Cryotherapy of avascular, immature retina

  ii] Scleral buckling for RD
- Prophylaxis is most important

ROP Prophylaxis

- Monitor umblical arterial Pa Oxygen level (50-100 is normal)
- Examine temporal periphery of retina before the newborn/infant is discharged from hospital (look for threshold disease)
- All premature infants <32 week or <1500 gm should be screened
- If ROP develops, follow-up exam

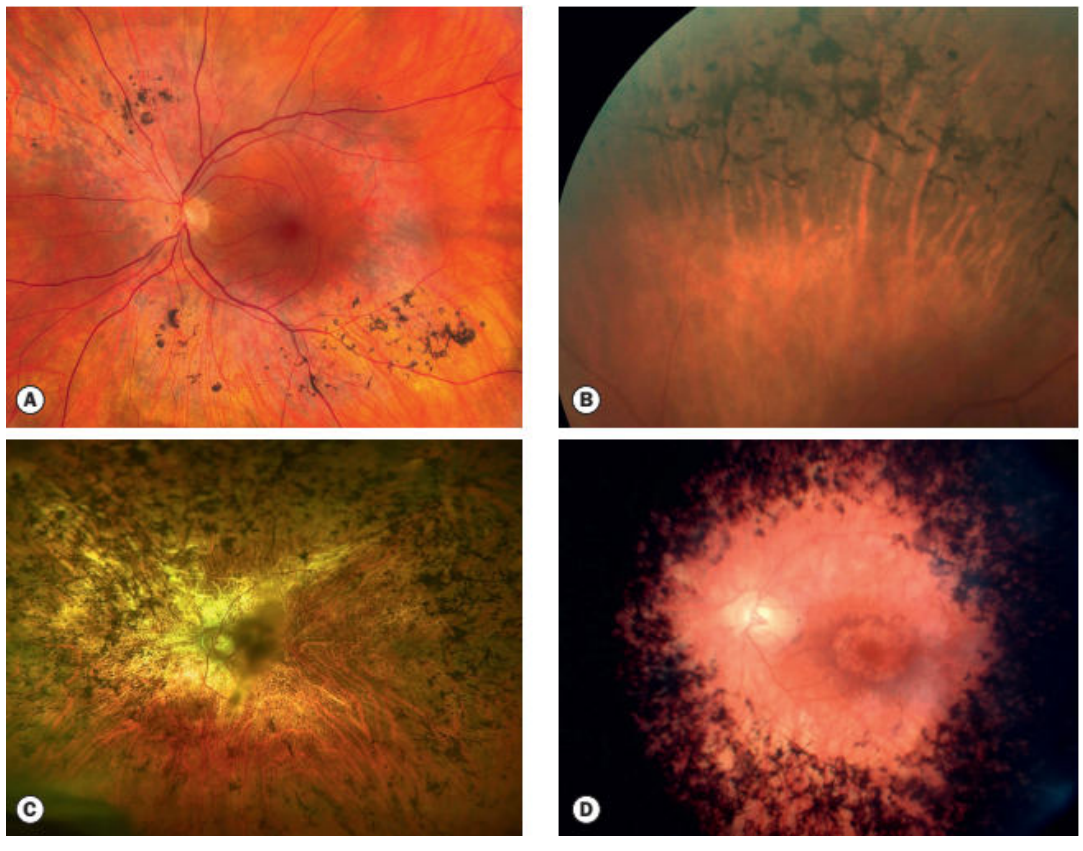
Retinitis Pigmentosa

- Hereditary disease
- Inheritance-Autosomal recessive
- Consanguinity of parents
- Progressive night blindness
- Constriction of visual field
- B/L
- Symmetrical , diffuse pigmentary retinal dystrophy which affects Rods
- Mostly symptoms appear in young age

Retinitis Pigmentosa

Signs-

- Bony specule pigmentation
- Arteriolar attenuation
- Waxy pallor of disc



Retinitis Pigmentosa

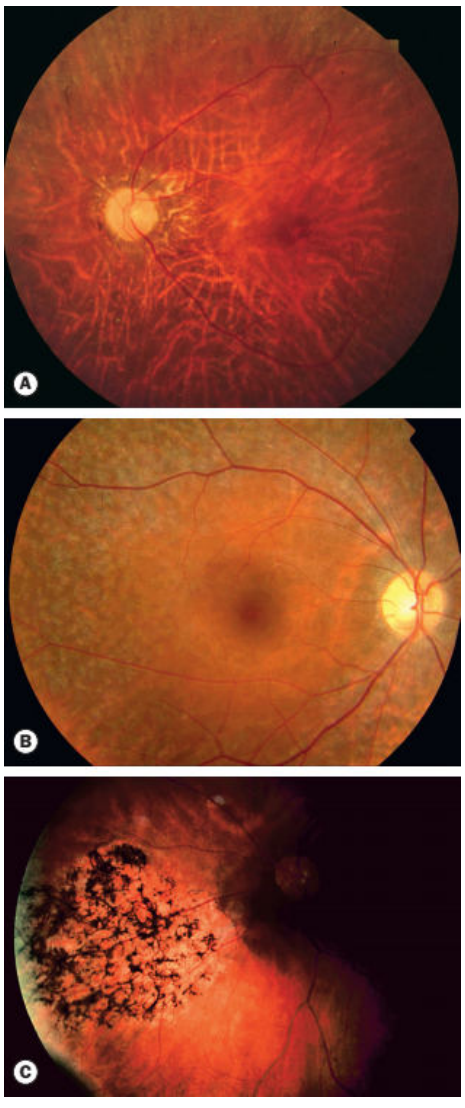
Investigations-

- Electro-retinography- ⇩ amplitude
- Electro-oculography- absence of light peak
- Visual fields- ring scotoma

Retinitis Pigmentosa-Variants

Atypical RP

i. Retinitis pigmentosa sine pigmento
ii. Retinitis puntata albescens
iii. Sector retinitis pigmentosa



RP associated with systemic diseases-

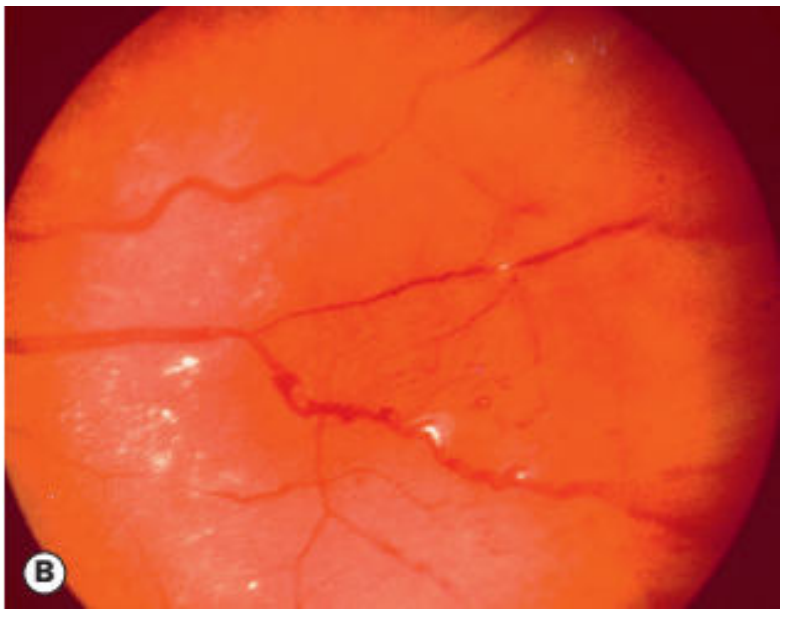
I. Laurence-moon-biedl syndrome(Bardet Biedle)
II. Bassen-kornzweig syndrome (Abeta lipoproteinemia)
III. Refsum's syndrome
IV. Usher's syndrome
V. Cockayne's syndrome
VI. Kearns-Sayre syndrome
VII. Friedreich's ataxia
VIII. NARP syndrome

Persistent Hyperplastic Primary Vitreous

- Failure of structures within primary vitreous to regress
- Unilateral white pupilary reflex in full term infant
- May be Anterior/ Posterior
- May be associated with cataract, Glaucoma, Micro ophthalmos, persistent hyaloid artery
- Bilateral cases may be associated with Patau's syndrome or Norries disease.

Coat's Disease

- Chronic, progressive, vascular abnormality
- Telangiectic retinal vessels leak fluid
- Exudative bullous RD
- Boys 18months -18 years
- Unilateral
- White pupillary reflex
- Tt: early photocoagulation /cryotherapy



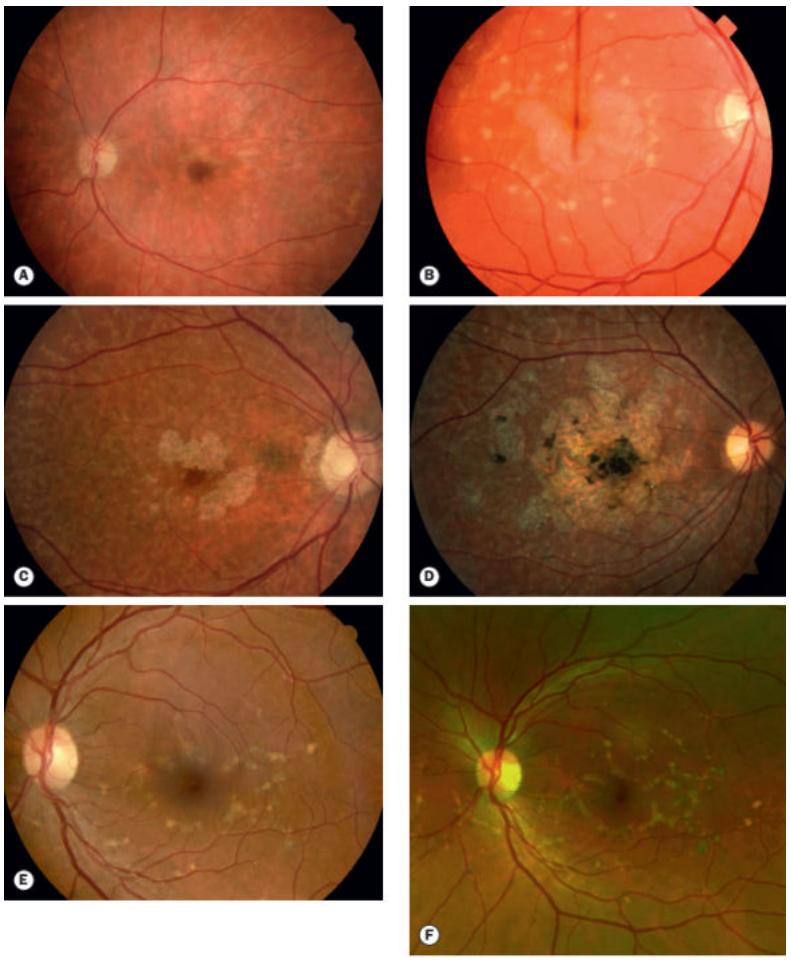
Stargardt disease

- Recessive, progresstive tapetoretinal dystrophy of central retina
- Age 8-14 years
- Beaten bronze atrophy of fovea
- Extensive chorioretinal atrophy
- Poor vision

Hereditary Dystrophies of central retina and choroid

Stargardt disease/ fundus flavimaculatus

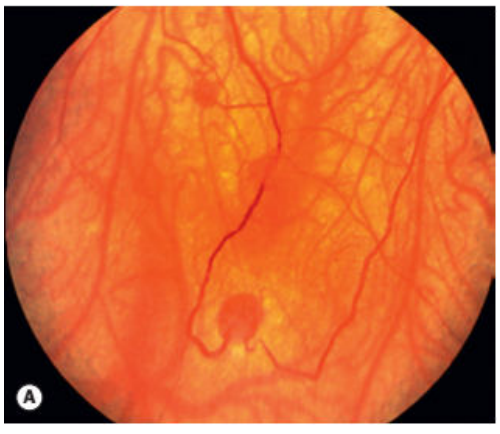
- Most common macular dystrophy
- Gradual impairment of central vision

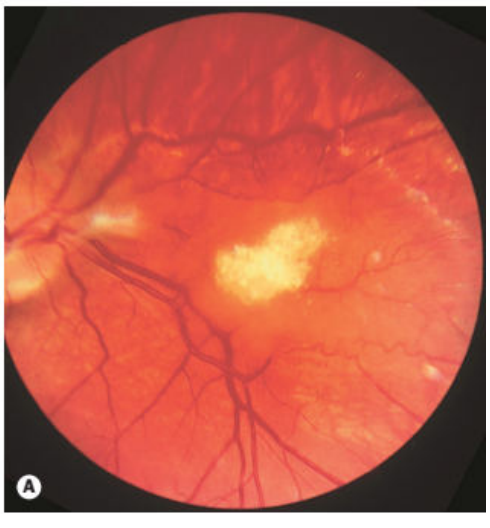


Phakomatosis

(Systemic associations present)

- Angiomatosis of retina with cerebellar haemangioblastoma (Von Hippel Lindau disease)
- Tuberous sclerosis (Bourneville disease)
- Neurofibromatosis (Von Recling hausen disease)

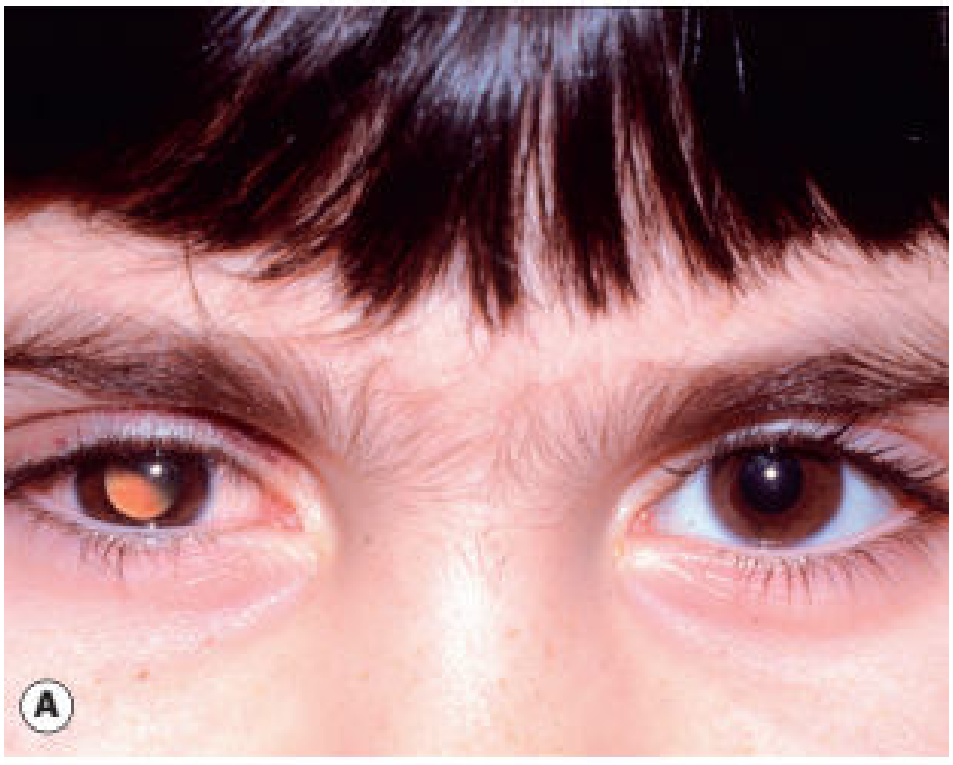




Phakomatosis

- Sturge-weber's syndrome- port wine stain along distribution of trigeminal nerve of affected side

White pupillary reflex (Leukocoria)



- Retinoblastoma- dealt in separate lecture

White pupillary reflex (Leukocoria)

i. Retinoblastoma
ii. ROP
iii. Coats disease
iv. Congenital cataract
v. Toxocara infestation
vi. Persistant hyperplastic vitreous
vii. Retrolental hyperplasia
viii. Retrolental inflammatory membrane due to Uvitis

Acknowledgement

Photgraphs from Kanski, Parson and Archives Dept of Ophthalmology AIIMs Rishikesh

MCQs

1] While working in a neonatal ICU your team delivers a premature infant at 27 weeks of gestation and weighing 1500 gm. How soon will you request fundus examination by an ophthalmologist?

(a) Immediately

(b) Three to four weeks after delivery

(c) At 34 weeks' gestational age

(d) At 40 weeks' gestational age

MCQs

- With which of the following is PHPV associated?

(a) Patau's Syndrome

(b)Down syndrome

(c) Tuberous sclerosis

(d) Sturge Weber syndrome

MCQs

Amaurotic cat's eye reflex is seen in:

a.Papilloedema

b. Retinoblastoma

c. Papillitis

d. Retinitis