RETINA.

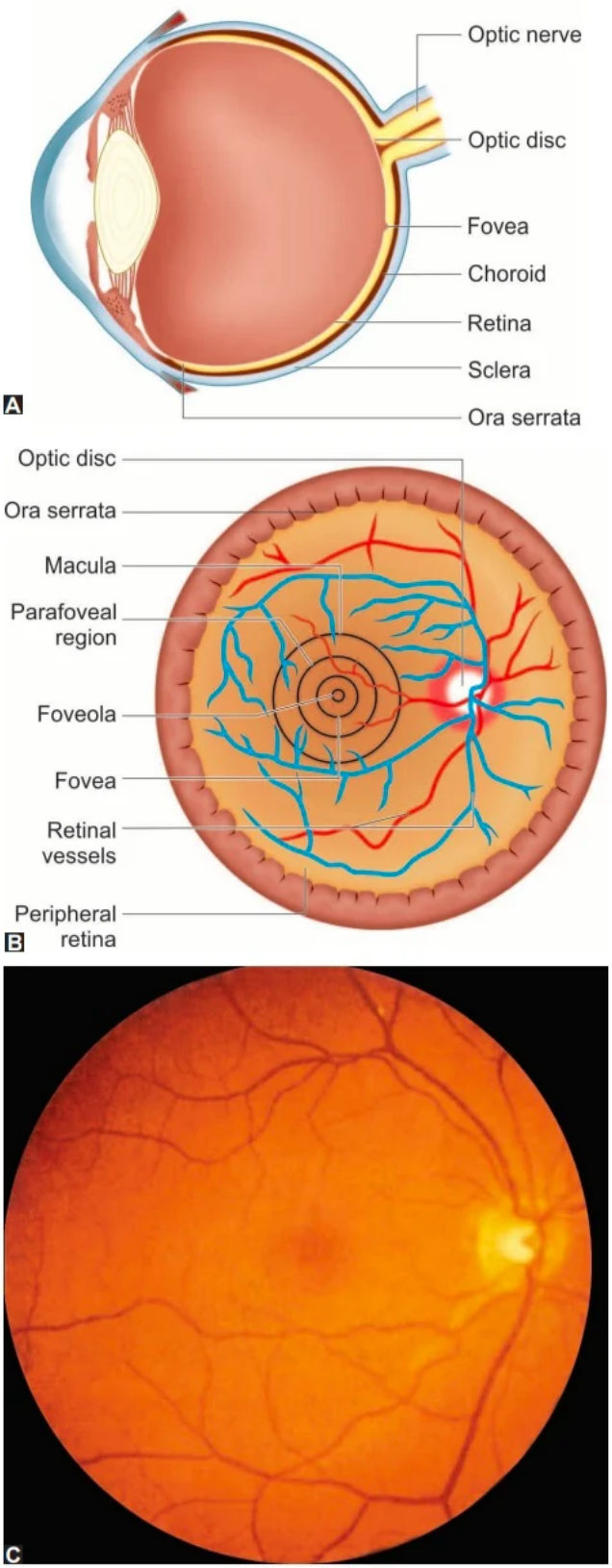
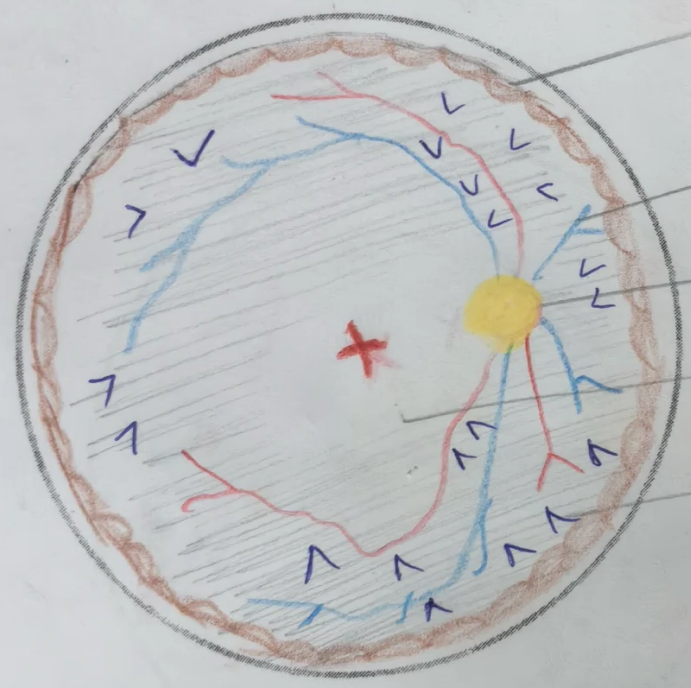


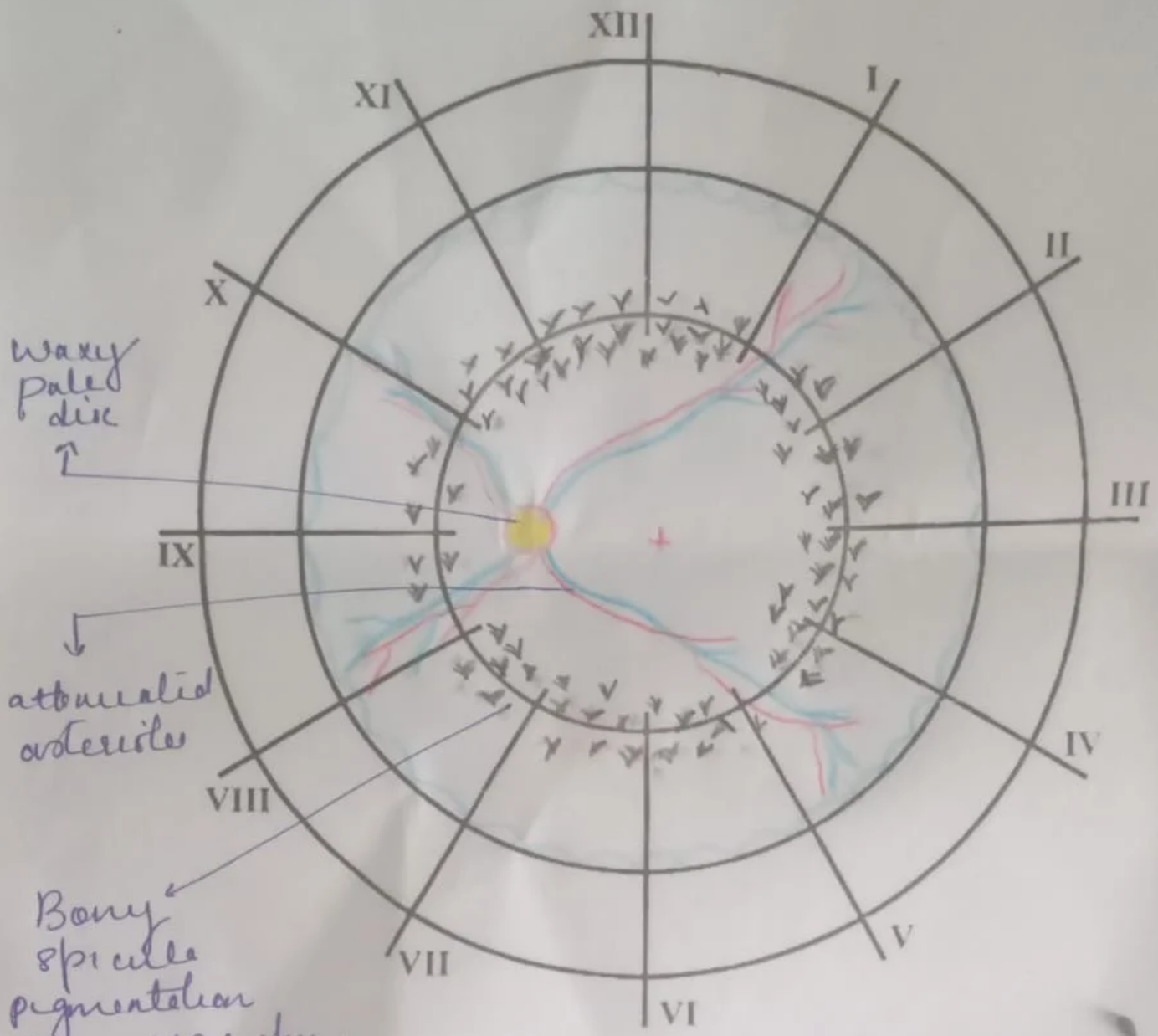
Fig. 12.1 *Gross anatomy of the retina: A, parts of retina in horizontal section at the level of fovea; B, diagrammatic fundus view; C, fundus photograph*

18. RETINITIS PIGMENTOSA



O.D.	O.S. ✓

Date :



Retinitis Pigmentosa

Retinitis Pigmentosa (RP) — Simple Notes

Definition

Retinitis pigmentosa is a **slowly progressive hereditary disease of retina** affecting mainly the **rod cells** first and later cone cells.

Etiology / Causes

- Usually hereditary
- More common with **consanguineous marriage**
- Starts in childhood
- Slowly progresses → blindness in middle age
- Both eyes affected equally
- Males affected more

Types of Inheritance

- Autosomal dominant (best prognosis)
- Autosomal recessive
- X-linked recessive (worst prognosis)

Associated Conditions

Eye Associations

- Myopia
- Open-angle glaucoma
- Keratoconus
- Posterior subcapsular cataract

Systemic Associations

- Usher syndrome
- Refsum syndrome
- Cockayne syndrome

Symptoms

1. Night blindness (Earliest symptom)

Patient cannot see properly in dim light.

2. Tubular vision

Peripheral vision is lost → only central vision remains.

Easy way to remember:

"Looking through a tube"

3. Late loss of central vision

Finally complete blindness may occur.

Signs

Fundus Findings

Classic triad of RP:

1. **Bone spicule pigmentation**
   → black pigment spots in retina
2. **Attenuated retinal vessels**
   → thin thread-like arteries
3. **Waxy pale optic disc**
   → due to optic atrophy

Visual Field

- Ring scotoma initially
- Later → tubular vision

Dark Adaptation

- Poor dark adaptation due to rod dysfunction

ERG Findings

ERG becomes:

- Reduced early
- Later completely extinguished

Atypical Types

1. Retinitis pigmentosa sine pigmento

- Symptoms present
- But no visible pigments in fundus

2. Sectoral RP

- Only one part of retina affected

Differential Diagnosis

- Congenital syphilis
- Vitamin A deficiency / night blindness

Treatment

No permanent cure currently.

Management

1. Correct refractive error with glasses
2. Low vision aids
   - Magnifying glasses
   - Night vision devices
3. Acetazolamide for cystoid macular edema
4. Rehabilitation
5. Genetic counselling
6. Avoid consanguineous marriage

Prognosis

- Vision gradually worsens
- Central vision becomes poor in later life

Very Short Exam Summary

RP = "Night blindness + Tubular vision + Bone spicule pigmentation"

Classical triad:

- Bone spicule pigmentation
- Thin retinal vessels
- Waxy pale optic disc

റെറ്റിനൈറ്റിസ് പിഗ്മെന്റോസ (Retinitis Pigmentosa)

ഇത് കണ്ണിലെ റെറ്റിനയെ ബാധിക്കുന്ന ഒരു പാരമ്പര്യ രോഗമാണ്.
ഈ രോഗത്തിൽ ആദ്യം രാത്രി കാഴ്ചയ്ക്ക് സഹായിക്കുന്ന **റോഡ് സെല്ലുകൾ (Rod cells)** നശിക്കുന്നു. പിന്നീട് നിറക്കാഴ്ചയും പകൽ കാഴ്ചയും നൽകുന്ന **കോൺ സെല്ലുകളും (Cone cells)** ബാധിക്കും.

രോഗം വളരെ പതുക്കെ പുരോഗമിക്കുന്നതാണ്.

കാരണങ്ങൾ / പ്രത്യേകതകൾ

- സാധാരണയായി പാരമ്പര്യ രോഗം
- ബന്ധുവിവാഹമുള്ള കുടുംബങ്ങളിൽ കൂടുതലായി കാണാം
- ചെറിയ പ്രായത്തിൽ തുടങ്ങാം
- രണ്ട് കണ്ണുകളും ഒരുപോലെ ബാധിക്കും
- പുരുഷന്മാരിൽ കൂടുതലാണ്

രോഗിക്ക് ഉണ്ടാകുന്ന പ്രധാന പ്രശ്നങ്ങൾ

1. രാത്രിയിൽ കാണാൻ ബുദ്ധിമുട്ട് (Night blindness)

ഇതാണ് ആദ്യ ലക്ഷണം.

രാത്രിയിലോ ഇരുണ്ട സ്ഥലങ്ങളിലോ വ്യക്തമായി കാണാൻ കഴിയില്ല.

2. വശങ്ങളിലെ കാഴ്ച കുറയുക

കുറച്ച് കുറച്ച് വശങ്ങളിലെ കാഴ്ച നഷ്ടമാകും.

അവസാനം രോഗിക്ക് മുന്നിലെ ചെറിയ ഭാഗം മാത്രം കാണും.

ഇതിനെ:

- Tunnel vision
- Tubular vision

എന്ന് പറയുന്നു.

ഉദാഹരണം:

ഒരു ട്യൂബിലൂടെ നോക്കുന്നതുപോലെ മാത്രം കാണുക.

3. പിന്നീട് മുന്നിലെ കാഴ്ചയും കുറയും

വർഷങ്ങൾക്കു ശേഷം central vision പോലും നഷ്ടപ്പെടാം.

ഡോക്ടർ പരിശോധിക്കുമ്പോൾ കാണുന്ന മാറ്റങ്ങൾ

Fundus examination ൽ

1. Bone spicule pigmentation

റെറ്റിനയിൽ കറുത്ത നിറത്തിലുള്ള പിഗ്മെന്റ് കുത്തുകൾ കാണും.

2. രക്തക്കുഴലുകൾ ചെറുതാകുക

റെറ്റിനയിലെ blood vessels വളരെ നേർന്നതാകും.

3. Optic disc pale ആകുക

Optic disc വെളുത്ത മെഴുകുപോലുള്ള നിറമാകും.

Visual field changes

ആദ്യം:

- വശങ്ങളിലെ കാഴ്ച കുറയും

പിന്നീട്:

- Tunnel vision

അവസാനം:

- Blindness

ERG test

റെറ്റിനയുടെ പ്രവർത്തനം പരിശോധിക്കുന്ന ടെസ്റ്റാണ്.

ഈ രോഗത്തിൽ:

- ERG കുറയുകയും
- പിന്നീട് ഇല്ലാതാകുകയും ചെയ്യും.

Treatment

ഈ രോഗത്തിന് പൂർണ്ണമായ ചികിത്സ ഇല്ല.

പക്ഷേ രോഗിയെ സഹായിക്കാൻ:

- കണ്ണട
- Low vision aids
- Night vision devices
- Rehabilitation
- Genetic counselling

എല്ലാം നൽകാം.

Prognosis

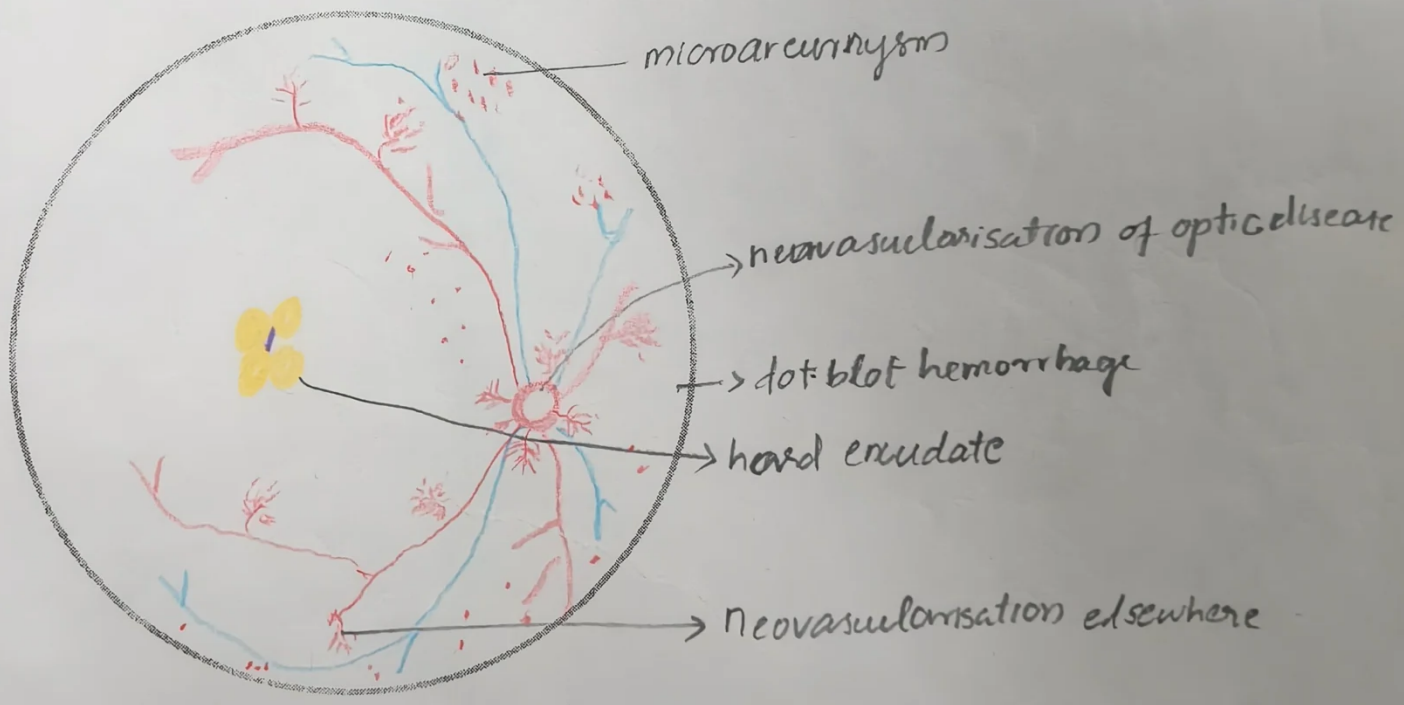
രോഗം പതുക്കെ worsen ചെയ്യും.
അവസാനം കാഴ്ച വളരെ കുറയാം.

വളരെ short ആയി ഓർക്കാൻ

RP =

- Night blindness
- Tunnel vision
- Bone spicule pigmentation

21. DIABETIC RETINOPATHY

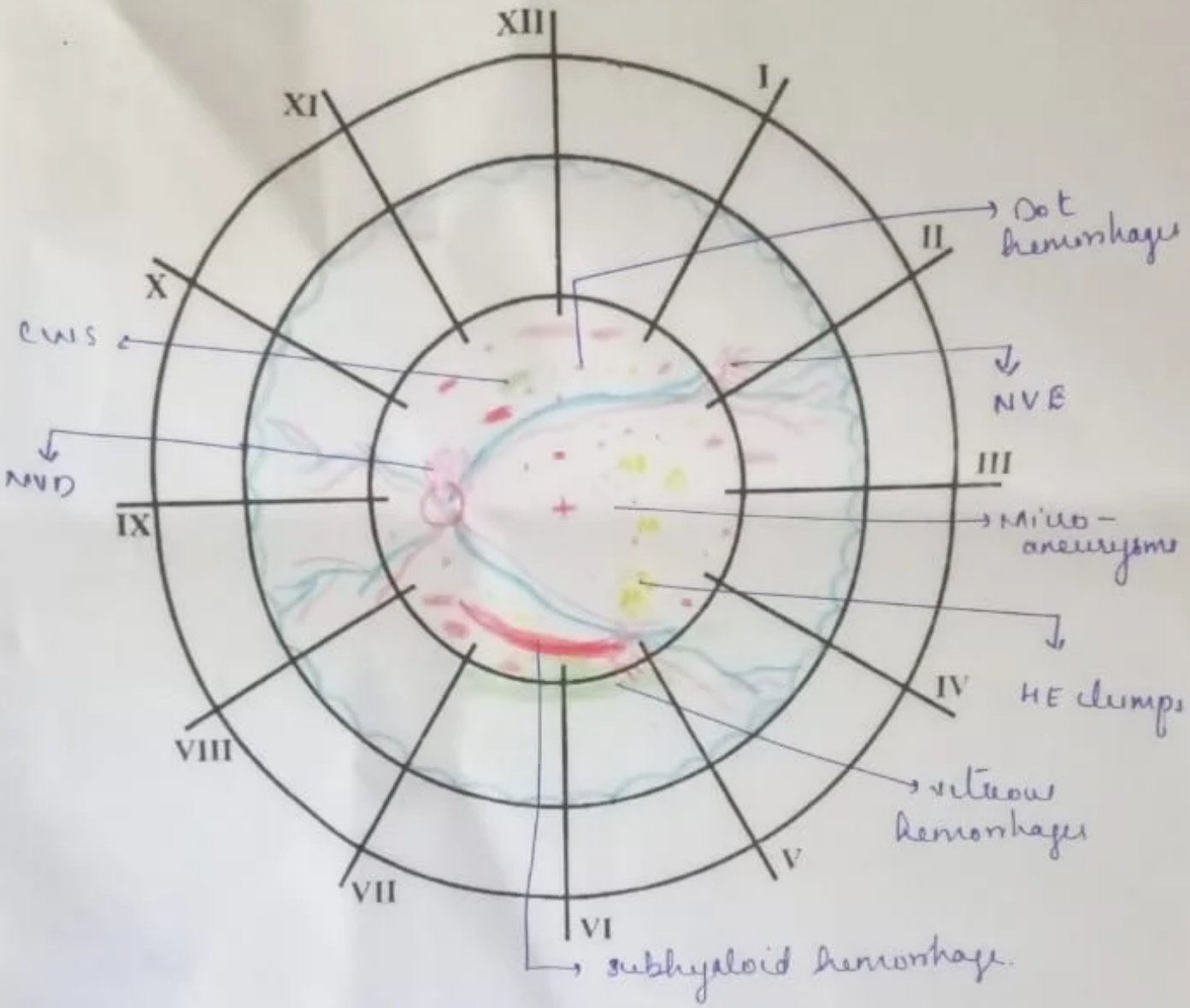


36/591/2014. 50x100. G.P.S.

FUNDUS CHART

O.D.	O.S. ✓

Patient :
Date :



Proliferative Diabetic Retinopathy

DIABETIC RETINOPATHY (DR)

DEFINITION



Diabetic retinopathy (DR) refers to retinal changes seen in patients with diabetes mellitus.

RISK FACTORS







1. Duration of diabetes after the onset of puberty is the most important determining factor.
2. Sex: Females > males (4:3).
3. Poor metabolic control
4. Heredity: It is transmitted as autosomal recessive trait.
5. Other risk factors include:
   - Pregnancy
   - Hypertension (HTN)
   - Smoking
   - Obesity
   - Anaemia
   - Hyperlipidaemia



PATHOGENESIS



Hyperglycemia, in uncontrolled diabetes mellitus, is the starting point for development of DR.

CLASSIFICATION OF DIABETIC RETINOPATHY

1. Non-proliferative Diabetic Retinopathy (NPDR)



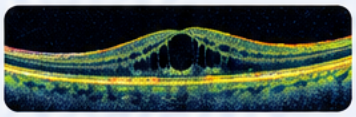
It can be:
- Mild
- Moderate
- Severe
- Very severe

2. Proliferative Diabetic Retinopathy (PDR)



Occurrence of neovascularization over the changes of very severe NPDR is the hallmark of PDR.

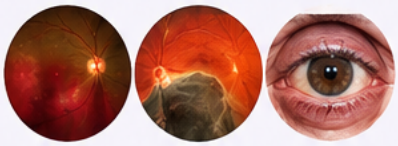
3. Diabetic Maculopathy
(Changes in macular region)



Diabetic Macular Oedema (DME) occurs due to increased permeability of retinal capillaries.
It can be:
- Clinically significant macular oedema (CSME)
- Clinically non-significant macular oedema (N-CSME)

4. Advanced Diabetic Eye Disease (ADED)

It is the end result of uncontrobled PDR. Marked by complications such as persistent vitreous haemorrhage, tractional retinal detachment, and neovascular glaucoma.

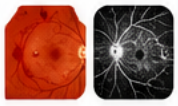


Vitreous Haemorrhage | Tractional Retinal Detachment | Neovascular Glaucoma

1. NON-PROLIFERATIVE DIABETIC RETINOPATHY (NPDR)

OPHTHALMOSCOPIC FEATURES

1. Microaneurysms
- In the macular area, the earliest detectable lesion.
- Appear as red dots, leak fluid and fluorescein dye on FFA.
- Look like clusters of grapes at end of vascular twigs.



2. Retinal Haemorrhages
- Both deep (dot & blot haemorrhages – in outer plexiform layer) and superficial haemorrhages (flame-shaped – in nerve fibre layer) occur from rupture of microaneurysms.



Dot & Blot Haemorrhages | Flame-shaped Haemorrhages

3. Retinal Oedema



4. Hard Exudates
(Lipid deposition)
- Seen in macular area.



5. Cotton-wool Spots
- Represent areas of nerve fibre infarcts.



6. Venous Abnormalities
- Beading
- Looping
- Dilatation of veins adjacent to area of capillary non-perfusion.



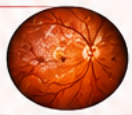
7. Intraretinal Microvascular Abnormalities (IRMA)
- Seen as fine irregular red lines connecting arterioles with venules.
- Represent A-V shunts.



2. PROLIFERATIVE DIABETIC RETINOPATHY (PDR)

Occurrence of neovascularization over the changes of very severe NPDR is the hallmark of PDR. It results in the formation of:

1. Fibrovascular Epiretinal Membrane
Formed due to condensation of connective tissue around the new vessels.



2. Vitreous Detachment and Vitreous Haemorrhage

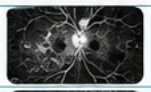


3. DIABETIC MACULOPATHY (CHANGES IN MACULAR REGION)

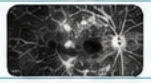
Diabetic Macular Oedema **(DME)** occurs due to increased permeability of retinal capillaries.
It can be:
- Clinically significant macular oedema (CSME)
- Clinically non-significant macular oedema (N-CSME)

CLINICOANGIOGRAPHIC CLASSIFICATION OF DIABETIC MACULOPATHY

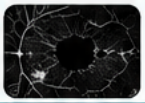
1. Focal Exudative Maculopathy
FFA reveals focal leakage with adequate macular perfusion.



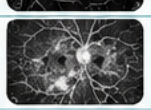
2. Diffuse Exudative Maculopathy
FFA reveals diffuse leakage.



3. Ischaemic Maculopathy
Occurs due to microvascular blockage. FFA shows areas of non-perfusion in the form of foveal avascular zone (FAZ).

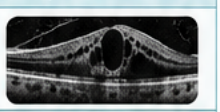


4. Mixed Maculopathy
Combined features of ischaemic maculopathy & exudative maculopathy.

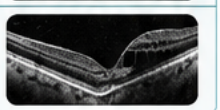


OCT (OPTICAL COHERENCE TOMOGRAPHY) CLASSIFICATION OF DME

1. Non-tractional DME
Cystoid macular oedema (CME)



2. Tractional DME
Vitreo-foveal traction (VFT)



SCREENING OF DIABETIC RETINOPATHY

1. **First examination**
   - 5 years after diagnosis of type 1 DM
   - At the time of diagnosis in type 2 DM
2. **Every year**
   If there is no diabetic retinopathy or there is mild NPDR.
3. **Every 6 months**
   In moderate NPDR.
4. **Every 3 months**
   In severe NPDR.
5. **Every 2 months**
   In PDR with no high-risk characteristics.

INVESTIGATIONS IN A CASE OF DR

Urine Examination
- Urine examination
- 24-hour urinary protein

Blood Investigations
- Blood sugar
- Lipid profile
- HbA1C

Imaging
- FFA (Fluorescein Angiography)
- Optical coherence tomography (OCT)



TREATMENT OF DIABETIC RETINOPATHY

1. Metabolic Control of Diabetes Mellitus & Associated Risk Factors



- **Target blood glucose level:**
  Fasting: <120 mg% **Post-prandial:** <180 mg% HbA1C: <7%
- **Target lipid profile:**
  Cholesterol: <200 mg% Triglycerides: <150 mg%
  HDL: >50 mg% hDL: <150 mg%
- **Control of associated anaemia:** Target haemoglobin >10 g%
- **Control of associated hypertension:** Target BP 130/80 mm Hg
- **Lifestyle changes:** Regular exercises, stop smoking, avoid alcohol consumption

2. Intravitreal Anti-VEGF Drugs (Vascular endothelial growth factor drugs)

Drugs: Bevacizumab (1.25 mg), Ranibizumab (0.5 mg)
- Effects of anti-VEGFs last for 4–6 weeks and frequent injections are warranted.
- There is risk of endophthalmitis.



3. Intravitreal Steroids

Drug: Triamcinolone acetonide (IVTA) (20 mg)
- Restores inner retinal barrier
- Has some anti-VEGF effects as well
- Used along with anti-VEGFs in recalcitrant cases
- Risks: Glaucoma, steroid-induced cataract, endophthalmitis



4. Laser Therapy

- **In DME:** Focal laser for focal DME. Grid laser for diffuse DME.
- **Mechanism:** Laser helps by stimulating the RPE pump mechanism and inhibiting VEGF release.
- **Laser therapy types:** Double frequency YAG laser 532 nm, Argon green laser, Diode laser.



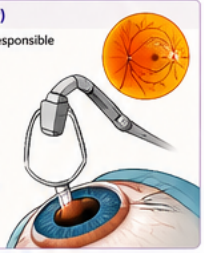
5. Macular Photocoagulation (Focal / Grid)



6. Panretinal Photocoagulation (PRP)

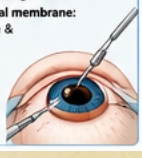
- PRP causes destruction of hypoxic retina responsible for production of vasoformative factors.

Indications for PRP:
1. PDR with HRC:
   - Neovascularization of disc (NVD)
   - Neovascularization elsewhere (NVE)
   - Severe NPDR associated with poor compliance for follow-up
2. Before cataract surgery / YAG capsulotomy
3. Renal failure
4. One-eyed patient
5. Pregnancy



7. Surgery – Vitrectomy (Pars Plana Vitrectomy – PPV)

Indications (Following cases):
- **Tractional DME with NPDR:** PPV + removal of posterior hyaloid.
- **Advanced PDR with dense vitreous haemorrhage:** PPV + removal of opaque vitreous gel & endophotocoagulation
- **Advanced PDR with extensive fibrovascular epiretinal membrane:** PPV + removal of fibrovascular epiretinal membrane & endophotocoagulation
- **Advanced PDR with tractional retinal detachment:** PPV with endophotocoagulation and reatachment of detached retina + scleral buckling
- **Intravitreal** injection of perfluorocarbon liquid or heavy liquid may be used.



Diabetic Retinopathy (DR) – Simplified Notes

Definition

Diabetic retinopathy is the damage that occurs in the retina due to diabetes mellitus.

Risk Factors

1. Long duration of diabetes after puberty → most important factor
2. Females > males
3. Poor diabetic control
4. Heredity
5. Pregnancy
6. Hypertension (HTN)
7. Smoking
8. Obesity
9. Anaemia
10. Hyperlipidaemia

Pathogenesis (How it occurs)

High blood sugar damages retinal capillaries → causes:

- Thickening of capillary basement membrane
- Endothelial cell damage
- Loss of pericytes
- Increased blood viscosity

This leads to:

- Capillary blockage
- Retinal ischemia
- Leakage from vessels
- Microaneurysms
- Hemorrhages
- Retinal edema
- Hard exudates
- IRMA
- Neovascularization

Classification of Diabetic Retinopathy

1. Non-Proliferative Diabetic Retinopathy (NPDR)

Can be:

- Mild
- Moderate
- Severe
- Very severe

Fundus Features

1. Microaneurysms

- Earliest detectable lesion
- Seen as tiny red dots
- Leak fluid and fluorescein dye on FFA
- Look like "cluster of grapes"

2. Retinal Hemorrhages

Two types:

a) Dot & blot hemorrhage

- Present in deeper retinal layers

b) Flame-shaped hemorrhage

- Present superficially in nerve fiber layer
- Occur due to rupture of microaneurysms

3. Retinal edema

- Swelling of retina due to fluid leakage

4. Hard exudates

- Yellow lipid deposits
- Commonly seen in macular area

5. Cotton wool spots

- Due to nerve fiber infarcts

6. Venous abnormalities

- Venous beading
- Looping
- Dilatation

7. IRMA

(Intraretinal microvascular abnormalities)

- Fine irregular red lines
- Connect arterioles and venules
- Represent A-V shunts

2. Proliferative Diabetic Retinopathy (PDR)

Hallmark:

Neovascularization (new vessel formation)

These new vessels can cause:

- Fibrovascular membrane formation
- Vitreous hemorrhage
- Vitreous detachment

3. Diabetic Maculopathy / Diabetic Macular Edema (DME)

Occurs due to increased permeability of retinal capillaries.

Types

1. Focal exudative maculopathy

- Focal leakage on FFA
- Macular perfusion is adequate

2. Diffuse exudative maculopathy

- Diffuse leakage on FFA

3. Ischemic maculopathy

- Due to microvascular blockage
- FFA shows non-perfusion areas

4. Mixed maculopathy

- Features of both ischemic and exudative types

OCT Classification of DME

1. Non-tractional DME

Example:

- Cystoid macular edema (CME)

2. Tractional DME

Example:

- Vitreofoveal traction (VFT)

4. Advanced Diabetic Eye Disease (ADED)

End stage of uncontrolled PDR.

Complications:

- Persistent vitreous hemorrhage
- Tractional retinal detachment
- Neovascular glaucoma

Screening Recommendations

- Type 1 DM → first eye exam after 5 years
- Type 2 DM → eye exam at diagnosis

Follow up:

- Every year → no DR or mild NPDR
- Every 6 months → moderate NPDR
- Every 3 months → severe NPDR
- Every 2 months → PDR without high-risk features

Investigations

- Urine examination
- 24-hour urinary protein
- Blood sugar estimation
- Renal function test (RFT)
- Lipid profile
- HbA1c
- FFA
- OCT

Treatment of Diabetic Retinopathy

1. Control Diabetes & Risk Factors

Target sugar levels

- Fasting <120 mg/dL
- Postprandial <180 mg/dL
- HbA1c <7%

Lipid targets

- Cholesterol <200 mg/dL
- Triglycerides <150 mg/dL
- HDL >50 mg/dL
- LDL <150 mg/dL

Other measures

- Treat anaemia
- Control BP (around 130/80 mmHg)
- Exercise regularly
- Stop smoking and alcohol

2. Intravitreal Anti-VEGF Drugs

Examples:

- Bevacizumab
- Ranibizumab

Uses:

- Reduce neovascularization and edema

Complication:

- Endophthalmitis

3. Intravitreal Steroids

Example:

- Triamcinolone acetonide

Uses:

- Restores retinal barrier
- Helps in resistant cases

Complications:

- Glaucoma
- Steroid cataract
- Endophthalmitis

4. Laser Therapy

Types

a) Focal laser

- For focal DME

b) Grid laser

- For diffuse DME

c) Panretinal photocoagulation (PRP)

PRP destroys ischemic retina → decreases VEGF production.

Indications for PRP

- PDR with high-risk characteristics
- Neovascularization of iris (NVI)
- Severe NPDR with poor follow-up
- Before cataract surgery/YAG capsulotomy
- Renal failure
- One-eyed patient
- Pregnancy

5. Surgical Treatment

Pars plana vitrectomy (PPV)

Indications

1. Tractional DME with NPDR

- PPV + removal of posterior hyaloid

2. Advanced PDR with dense vitreous hemorrhage

- PPV + removal of vitreous gel + endophotocoagulation

3. Extensive fibrovascular membrane

- PPV + membrane removal + endophotocoagulation

4. Tractional retinal detachment

- PPV + retinal reattachment
- Scleral buckling
- Silicone oil/SF6 gas tamponade

Diabetic Retinopathy (DR) — Very Simple Explanation

What is Diabetic Retinopathy?

Diabetes damages the small blood vessels of the retina (back of eye).
This damage is called **Diabetic Retinopathy**.

If sugar is uncontrolled for many years → retina gets damaged → vision decreases.

Why does it happen?

High blood sugar damages retinal blood vessels.

This causes:

- Blood vessel leakage
- Small bleeding
- Swelling of retina
- Less blood supply (ischemia)
- New weak abnormal vessels form

Risk Factors

Who gets DR more?

- Long duration diabetes → MOST IMPORTANT
- Poor sugar control
- High BP
- Smoking
- Obesity
- Pregnancy
- High cholesterol
- Anaemia

Stages of Diabetic Retinopathy

1. NPDR

Non-Proliferative Diabetic Retinopathy

(Early stage)

Old retinal vessels become weak and leaky.

Features

1. Microaneurysm

- Earliest sign
- Tiny balloon-like swelling in capillaries
- Seen as small red dots

Think: "Small vessel bulges"

2. Retinal Hemorrhage

Bleeding inside retina

Two types:

- Dot & blot hemorrhage → deep retina
- Flame-shaped hemorrhage → superficial retina

3. Retinal edema

Fluid leakage → retinal swelling

4. Hard exudates

Yellow fat deposits due to leakage

5. Cotton wool spots

White fluffy patches due to retinal ischemia

6. Venous changes

Veins become:

- Dilated
- Beaded
- Looped

7. IRMA

New abnormal channels inside retina

Body tries to bypass blocked vessels.

2. PDR

Proliferative Diabetic Retinopathy

(Advanced dangerous stage)

Because retina gets less oxygen → eye forms NEW blood vessels.

This is called:

Neovascularization

These new vessels are:

- Weak
- Fragile
- Easily bleed

Problems caused by PDR

Vitreous hemorrhage

Bleeding into vitreous → sudden vision loss

Fibrovascular membrane

Scar tissue forms with vessels

Tractional retinal detachment

Scar tissue pulls retina → retina detaches

3. Diabetic Macular Edema (DME)

Macula = central vision area

Fluid collects in macula due to leakage.

Result:

- Blurred central vision

Types of Maculopathy

Focal

Leakage from one small area

Diffuse

Leakage from many areas

Ischemic

Poor blood supply to macula

Mixed

Combination of both

Advanced Diabetic Eye Disease

Very severe uncontrolled DR.

Complications:

- Persistent vitreous bleed
- Retinal detachment
- Neovascular glaucoma

Screening

- Type 1 DM → eye check after 5 years
- Type 2 DM → eye check at diagnosis

Follow-up:

- Mild disease → yearly
- Moderate → every 6 months
- Severe → every 3 months

Investigations

- Blood sugar
- HbA1c
- Lipid profile
- Kidney tests
- FFA
- OCT

Treatment

1. Control Diabetes

MOST IMPORTANT

Control:

- Blood sugar
- BP
- Cholesterol
- Smoking

2. Anti-VEGF injections

Examples:

- Bevacizumab
- Ranibizumab

Purpose:

- Reduce new vessels
- Reduce swelling

3. Steroid injections

Reduce retinal swelling

4. Laser Treatment

Focal/Grid laser

Used for macular edema

PRP laser

(Panretinal photocoagulation)

Burns ischemic retina → decreases VEGF → reduces new vessel formation

Used in:

- PDR
- Severe NPDR

5. Surgery (PPV)

Done in:

- Vitreous hemorrhage
- Retinal detachment
- Severe PDR

Easy Flow to Remember

High sugar →

Blood vessel damage → Leakage + bleeding → Retinal ischemia → New vessel formation → Bleeding + scar → Retinal detachment + vision loss

One-Line Summary

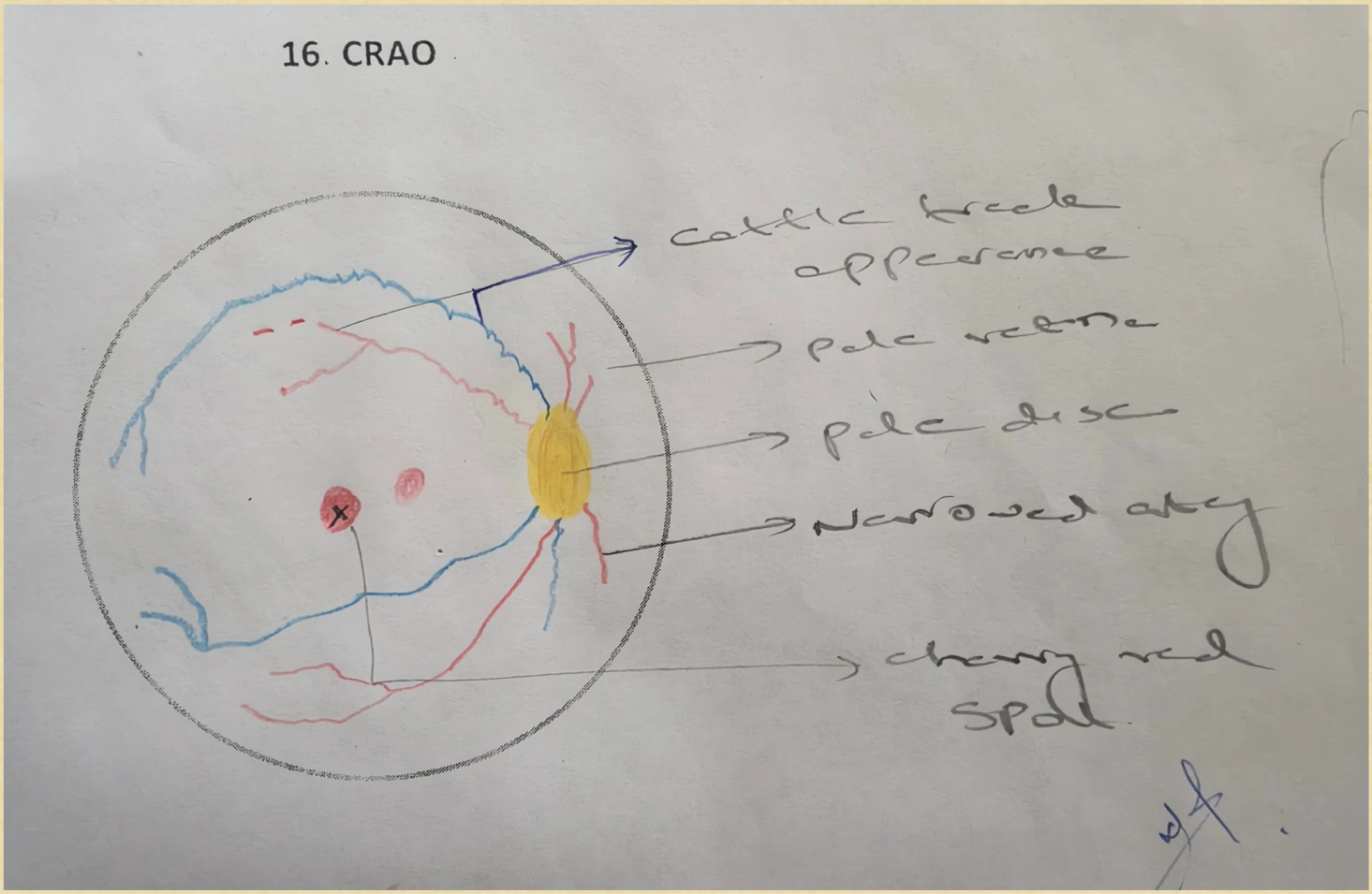
NPDR = leaking vessels

PDR = new vessels forming

DME = macular swelling

PRP laser = destroys ischemic retina to stop new vessels

16. CRAO

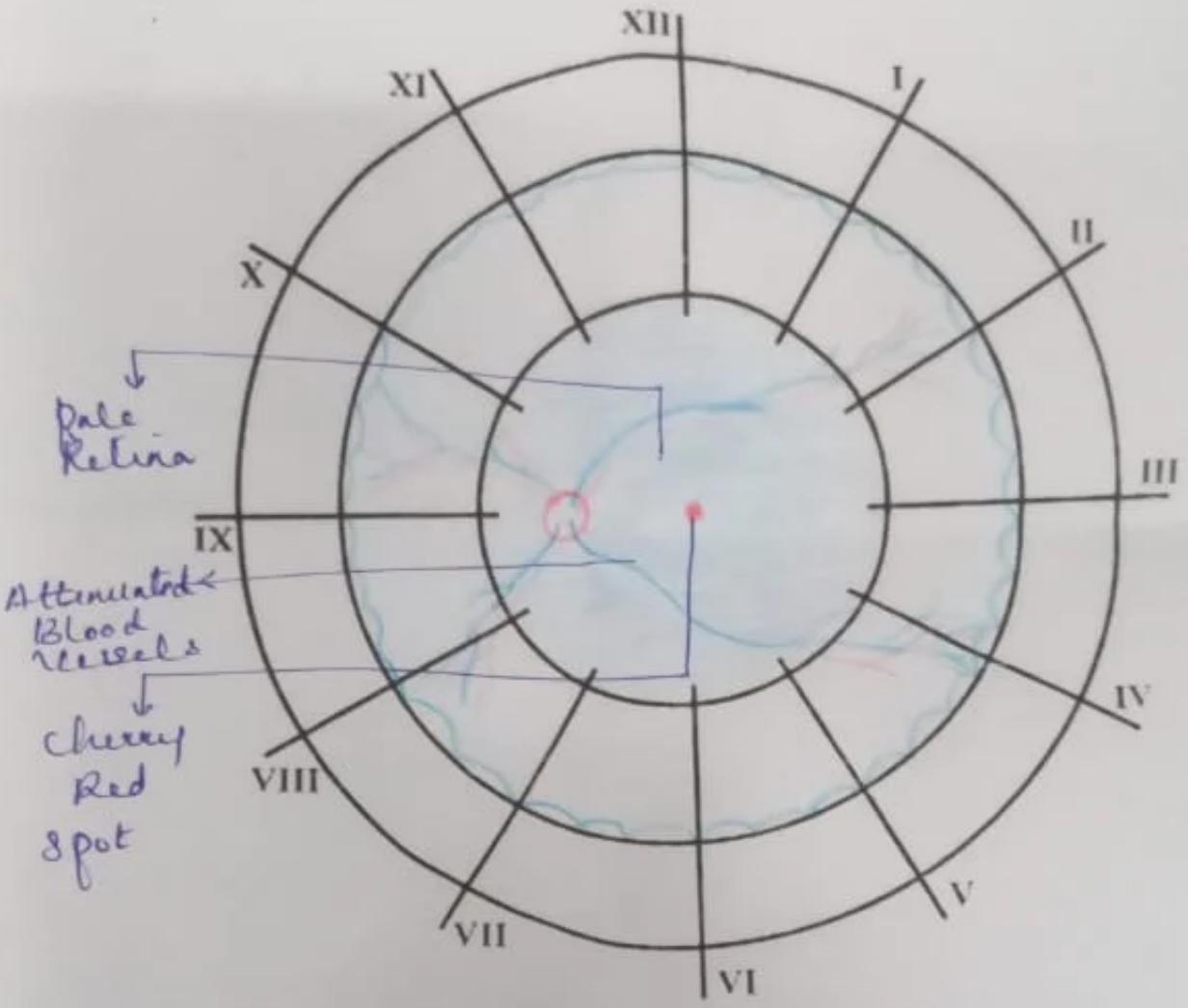

cattle trade appearance
pale retina
pale disc
Narrowed artery
cherry red spot

FUNDUS CHART

O.D.	O.S.

Patient :

Date :

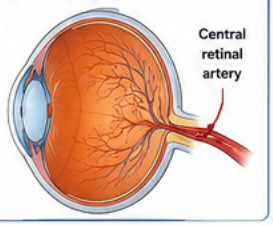


CRAO

CENTRAL RETINAL ARTERY OCCLUSION (CRAO)

DEFINITION

- CRAO is an **ocular emergency** caused by blockage of the **central retinal artery.**
- The central retinal artery supplies the **inner 6 layers of retina.**
- It forms about **60% of all retinal artery occlusions.**
- Blockage usually occurs at the level of **lamina cribrosa.**
- Usually **unilateral.**
- Bilateral cases are rare **(1–2%).**
- Common in:
  - Elderly people
  - Males
  - Patients with:
    - Hypertension
    - Cardiovascular disease



SYMPTOMS

1. Sudden painless loss of vision

- Hallmark symptom
- Occurs suddenly within seconds



2. Amaurosis fugax

- Temporary transient visual blackout before permanent loss
- Acts as a warning symptom



FUNDUS EXAMINATION

1. Retinal oedema

- Retina appears:
  - Pale
  - Milky white
- Due to ischaemic oedema

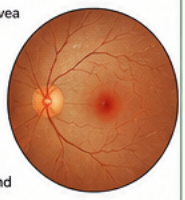


2. Cherry-red spot

- Bright red spot at macula/fovea

Reason

- Fovea is very thin
- No inner nerve fiber layer
- Underlying choroidal blood supply becomes visible
- Surrounding retina is pale and oedematous → makes red spot more obvious



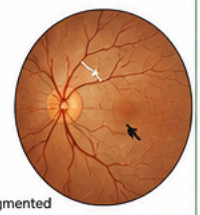
3. Vascular changes

Arteries

- Severely narrowed
- Thread-like

Veins

- Show **cattle tracking**
- Blood column appears segmented



4. Late atrophic changes (after 4–6 weeks)

- Retinal oedema disappears
- Retina becomes atrophic
- Vessels become severely attenuated
- Optic disc becomes pale due to:
  - Consecutive optic atrophy



ETIOLOGY / CAUSES

1. Emboli (Most common cause)

Emboli usually come from:

- **Carotid artery atheroma**
- **Cardiac valves**



Types of emboli

- **Cholesterol emboli**
  - Called Hollenhorst plaques
- **Calcium emboli**
- **Platelet-fibrin emboli**



2. Thrombosis

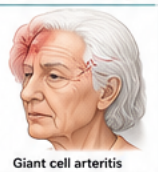
- Due to atherosclerosis
- Usually occurs at lamina cribrosa



3. Retinal arteritis

Inflammatory diseases causing CRAO:

- **Giant cell arteritis**
- **Periarteritis**
  - Seen in:
    - Systemic lupus erythematosus (SLE)
    - Polyarteritis nodosa



4. Other causes

- Angiospasm
- Markedly raised intraocular pressure
- Thrombophilic disorders
- Inherited anticoagulant defects
- Oral contraceptive pill use



CLINICAL SIGNS

1. Visual acuity

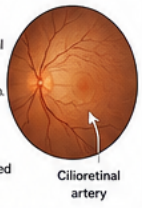
- Vision becomes severely reduced
- Usually:
  - Less than **3/60**
  - Counting fingers (CF)
  - Hand movements (HM)

Seen in about **90% cases.**



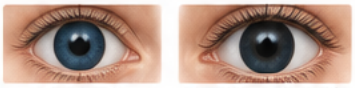
2. Cilioretinal artery effect

- About 5% people have a congenital cilioretinal artery.
- It arises from posterior ciliary circulation.
- It supplies the macula.
- In these patients:
  - Central vision may be preserved despite CRAO.



3. Pupillary reaction

- Direct light reflex absent
- **RAPD present**
  - Relative afferent pupillary defect
  - Also called **Marcus Gunn pupil**



INVESTIGATIONS

1. Fundus Fluorescein Angiography (FFA)

Shows:

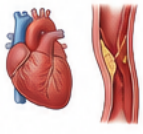
- Delayed arterial filling
- Masking of normal choroidal fluorescence
- Due to opaque oedematous retina



2. Systemic work-up

Necessary to identify:

- Cardiovascular disease
- Carotid artery disease
- Giant cell arteritis
- Hypercoagulable states



Purpose

- Prevent future embolic episodes

MANAGEMENT

Important point

- Retina cannot survive complete ischaemia beyond:
  - **90–100 minutes**
- Visual recovery is usually poor
- Treatment should begin within **24 hours**



3. Fibrinolytic therapy

- Used in selected cases
- Attempts to dissolve clot



4. Intravenous corticosteroids

Indication

- Suspected giant cell arteritis

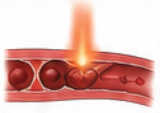
Purpose

- Protect fellow eye



5. Laser photodisruption

- Laser used to:
  - Break embolus
  - Dislodge embolus



TREATMENT

1. Immediate lowering of intraocular pressure (IOP)

Purpose

- Reduce resistance to blood flow
- Help move embolus distally

Methods

a) Digital ocular massage

- Intermittent pressure over closed eye



b) IV hyperosmotic agents

Example: Mannitol

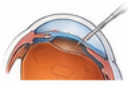


c) IV acetazolamide

- Dose: **500 mg IV**

d) Anterior chamber paracentesis

- Needle used to remove aqueous humour
- Causes sudden drop in IOP
- May flush embolus forward



2. Vasodilators and oxygenation

Methods

- Inhalation of:
  - 95% oxygen + 5% carbon dioxide
- Or breathing into polythene bag

Purpose

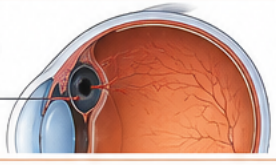
- Relieve angiospasm
- Improve oxygen supply



COMPLICATION

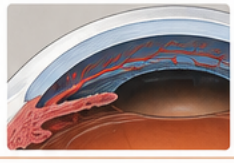
Neovascular glaucoma

- Occurs in about **2–6%** cases



Mechanism

- Severe retinal ischaemia releases angiogenic factors
- New vessels form in anterior chamber angle
- Causes severe rise in intraocular pressure



Time

- Can occur months after CRAO



Central Retinal Artery Occlusion (CRAO)

Definition

- CRAO is an **ocular emergency** caused by blockage of the **central retinal artery**.
- The central retinal artery supplies the **inner 6 layers of retina**.
- It forms about **60% of all retinal artery occlusions**.
- Blockage usually occurs at the level of **lamina cribrosa**.
- Usually **unilateral**.
- Bilateral cases are rare (**1–2%**).
- Common in:
  - Elderly people
  - Males
  - Patients with:
    - Hypertension
    - Cardiovascular disease

Etiology / Causes

1. Emboli (Most common cause)

Emboli usually come from:

- **Carotid artery atheroma**
- **Cardiac valves**

Types of emboli

- **Cholesterol emboli**
  - Called **Hollenhorst plaques**
- **Calcium emboli**
- **Platelet-fibrin emboli**

2. Thrombosis

- Due to **atherosclerosis**
- Usually occurs at **lamina cribrosa**

3. Retinal arteritis

Inflammatory diseases causing CRAO:

- **Giant cell arteritis**
- **Periarteritis**
  - Seen in:
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4. Other causes

- Angiospasm
- Markedly raised intraocular pressure
- Thrombophilic disorders
- Inherited anticoagulant defects
- Oral contraceptive pill use

Symptoms

1. Sudden painless loss of vision

- Hallmark symptom
- Occurs suddenly within seconds

2. Amaurosis fugax

- Temporary transient visual blackout before permanent loss
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- About 5% people have a congenital **cilioretinal artery**.
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- Direct light reflex absent
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- Bright red spot at macula/fovea

Reason

- Fovea is very thin
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- Thread-like

Veins

- Show **cattle tracking**
- Blood column appears segmented

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Indication

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- Protect fellow eye

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- Laser used to:
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  - Dislodge embolus

Complication

Neovascular glaucoma

- Occurs in about **2–6% cases**

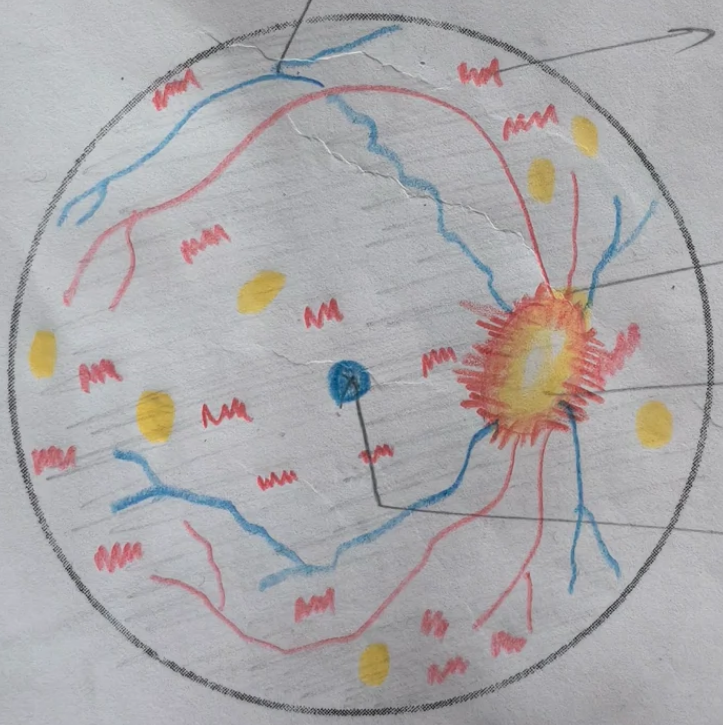
Mechanism

- Severe retinal ischaemia releases angiogenic factors
- New vessels form in anterior chamber angle
- Causes severe rise in intraocular pressure

Time

- Can occur months after CRAO

17. CRVO

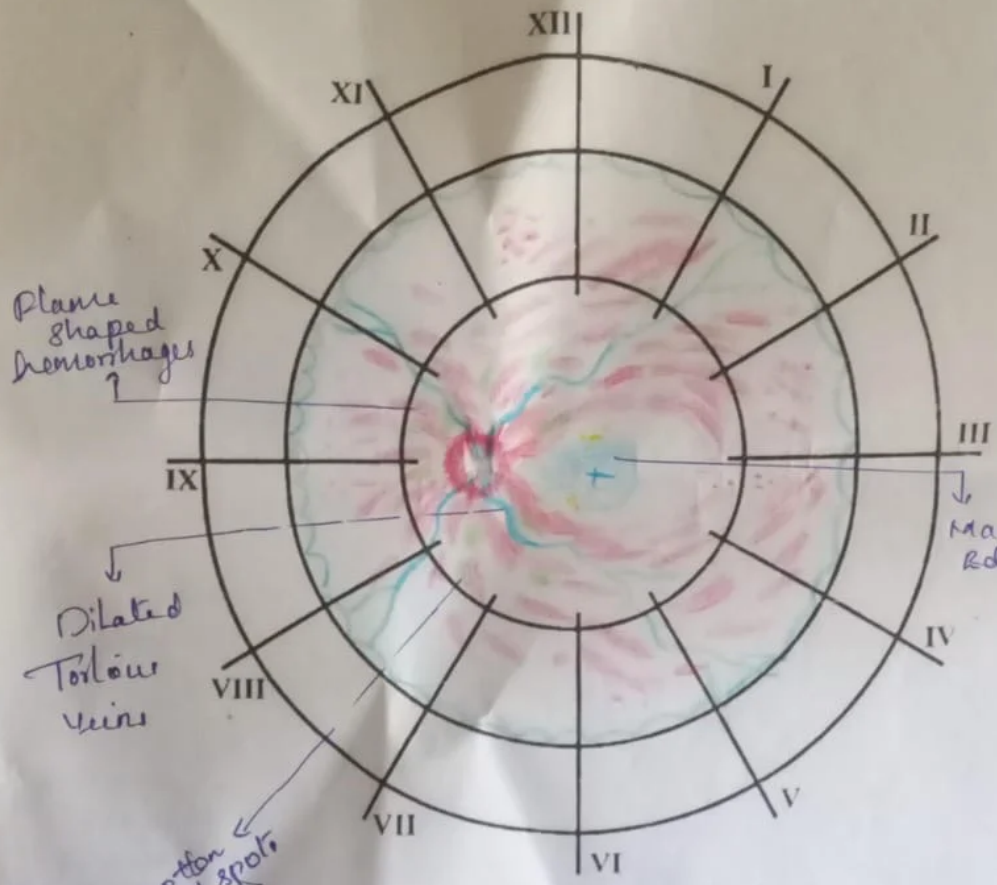

Tortuous vein
splinter hemorrhage
Disc edema
Optic margin blurred
Macular edema

FUNDUS CHART

O.D.	O.S. ✓

Patient :

Date :



ISCHEMIC CRVO

CENTRAL RETINAL VEIN OCCLUSION (CRVO)

DEFINITION

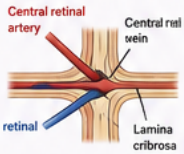
- CRVO is a common retinal vascular disorder caused by blockage of the **central retinal vein.**
- The blockage usually occurs just behind the **lamina cribrosa.**
- Commonly affects:
  - Elderly patients
  - 6th–7th decade of life



ETIOPATHOGENESIS

Main mechanism

- Central retinal artery and vein share a common adventitia.
- Atherosclerotic retinal artery compresses the retinal vein.
- This causes:
  - Venous obstruction
  - Venous stasis
  - Retinal haemorrhages
  - Retinal ischaemia



RISK FACTORS

A) Systemic Causes

1. **Hypertension (HTN)**
2. **Diabetes Mellitus (DM)**
3. **Hyperviscosity states**

   Seen in:
   - Polycythemia
   - Leukaemia
   - Macroglobulinemia
   - Multiple myeloma
4. **Hypercoagulable / thrombophilic disorders**






B) Ocular Causes

1. **Raised intraocular pressure (IOP)**

   Common in:
   - Primary Open-Angle Glaucoma (POAG)



2. **Local orbital causes**
   - Orbital cellulitis
   - Orbital tumours
   - Cavernous sinus thrombosis



CLASSIFICATION

1. Non-Ischaemic CRVO

Also called:
- Venous stasis retinopathy

Features
- Most common type
- About 75% cases
- Mild retinal ischaemia



2. Ischaemic CRVO

Also called:
- Haemorrhagic retinopathy

Features
- Severe retinal ischaemia
- Complete venous occlusion
- More dangerous type



SYMPTOMS



Main symptom
- Sudden painless diminution of vision



In Non-Ischaemic CRVO
- Mild to moderate visual loss



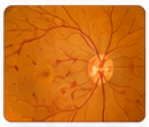
In Ischaemic CRVO
- Sudden severe visual loss
- Marked decrease in vision

CLINICAL SIGNS

A) Non-Ischaemic CRVO

Fundus findings

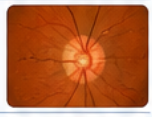
Veins
- Mild venous congestion
- Mild venous tortuosity



Haemorrhages
- Few superficial flame-shaped haemorrhages
- More in peripheral retina



Optic disc
- Mild papilloedema

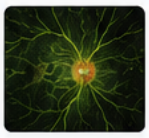


Retina
- Retina relatively normal



RAPD
- Absent

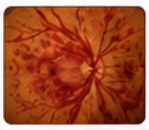
Late findings (6–9 months)
- Venous sheathing
- Cilioretinal collaterals around optic disc



B) Ischaemic CRVO

Fundus findings

Veins
- Massive venous engorgement
- Severe congestion
- Extreme tortuosity

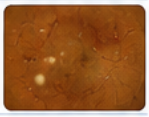


Retina
- Extensive retinal haemorrhages

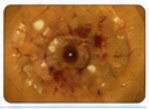
Classic appearance
- "Splashed tomato appearance"



Cotton wool spots
- Numerous soft exudates
- Usually >6–10 spots



Macula
- Severe macular oedema
- Macular haemorrhages present

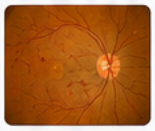
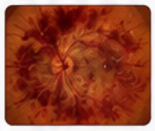


Optic disc
- Marked disc oedema
- Hyperaemia present



RAPD
- Present
- Important sign of ischaemic CRVO

PATTERN COMPARISON

Non-Ischaemic CRVO	Ischaemic CRVO
	
• Mild changes	• Severe changes
• Few haemorrhages	• Extensive haemorrhages
• Mild tortuosity	• Marked tortuosity

INVESTIGATIONS

1. Ocular Examination

Includes:





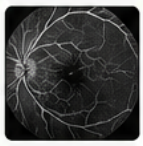
Visual acuity | IOP measurement | Gonioscopy

Purpose:
- Detect POAG
- Detect neovascularization of angle (NVA)

2. Fundus Fluorescein Angiography (FFA)

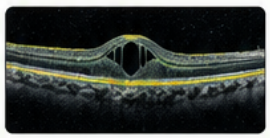
Done after: Retinal haemorrhages settle

Findings in Ischaemic CRVO
- Large capillary non-perfusion (CNP) areas
- Usually >10 disc areas



3. Optical Coherence Tomography (OCT)

Detects:
- Cystoid macular oedema (CME)
- Subretinal fluid
- Epiretinal membrane



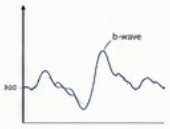
4. Electrophysiology and Perimetry

Goldmann perimetry
- Detects visual field defects



Electroretinography (ERG)

In ischaemic CRVO:
- Reduced b-wave amplitude



5. Systemic Workup

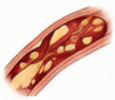
Investigate for:



Hypertension



Diabetes mellitus



Hyperlipidaemia



Homocysteinaemia (Especially in young patients)

MANAGEMENT

1. Observation and Follow-up

Non-Ischaemic CRVO
- Mild/moderate cases may only need observation

Prognosis
- 50% improve spontaneously
- Vision may become almost normal

Important point



- 15% cases may convert to ischaemic CRVO
- Usually within 4 months
- Requires close follow-up

2. Medical Treatment

A) Intravitreal Anti-VEGF Injections

Examples:
- Bevacizumab 1.25 mg
- Ranibizumab 0.3 mg



Uses:
- Reduce macular oedema
- Reduce neovascularization

B) Intravitreal Steroids

Example:
- Triamcinolone acetonide



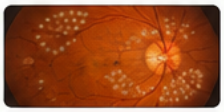
Use:
- Severe cystoid macular oedema (CME)

3. Laser Treatment

Panretinal Photocoagulation (PRP)

Indications

If neovascularization develops:
- NVI – Neovascularization of iris
- NVA – Neovascularization of angle
- NVD – Neovascularization of disc
- NVE – Neovascularization elsewhere



4. Surgical Treatment

Pars plana vitrectomy

Indications
- Persistent vitreous haemorrhage
- Tractional retinal detachment
- Epiretinal membrane

Glaucoma Drainage Device (GDD)

Indication: Intractable neovascular glaucoma

Method: Inserted through pars plana

COMPLICATIONS

1. Neovascular Glaucoma (NVG)

Important complication of ischaemic CRVO

Occurs in
- About 20% cases

Time
- Usually within 3 months

Also called
- "90-day glaucoma"

Other Complications

- Vitreous haemorrhage
- Subhyaloid haemorrhage
- Permanent blindness if untreated

KEY POINTS

CRVO is due to blockage of central retinal vein behind lamina cribrosa.

Non-ischaemic (75%) has better prognosis than ischaemic.

FFA is gold standard to detect non-perfusion in ischaemic CRVO.

Anti-VEGF / Steroids help reduce macular oedema.

Close follow-up is essential to detect complications like NVG early.

CENTRAL RETINAL VEIN OCCLUSION (CRVO)

Definition

- CRVO is a common retinal vascular disorder caused by blockage of the **central retinal vein**.
- The blockage usually occurs just behind the **lamina cribrosa**.
- Commonly affects:
  - Elderly patients
  - 6th–7th decade of life

Etiopathogenesis

Main mechanism

- Central retinal artery and vein share a common adventitia.
- Atherosclerotic retinal artery compresses the retinal vein.
- This causes:
  - Venous obstruction
  - Venous stasis
  - Retinal haemorrhages
  - Retinal ischaemia

Risk Factors

A) Systemic Causes

1. Hypertension (HTN)

2. Diabetes Mellitus (DM)

3. Hyperviscosity states

Seen in:

- Polycythemia
- Leukaemia
- Macroglobulinemia
- Multiple myeloma

4. Hypercoagulable / thrombophilic disorders

B) Ocular Causes

1. Raised intraocular pressure (IOP)

- Common in:
  - Primary Open-Angle Glaucoma (POAG)

2. Local orbital causes

- Orbital cellulitis
- Orbital tumours
- Cavernous sinus thrombosis

Classification

1. Non-Ischaemic CRVO

Also called:

- Venous stasis retinopathy

Features

- Most common type
- About 75% cases
- Mild retinal ischaemia

2. Ischaemic CRVO

Also called:

- Haemorrhagic retinopathy

Features

- Severe retinal ischaemia
- Complete venous occlusion
- More dangerous type

Symptoms

Main symptom

- Sudden painless diminution of vision

In Non-Ischaemic CRVO

- Mild to moderate visual loss

In Ischaemic CRVO

- Sudden severe visual loss
- Marked decrease in vision

Clinical Signs

A) Non-Ischaemic CRVO

Fundus findings

Veins

- Mild venous congestion
- Mild venous tortuosity

Haemorrhages

- Few superficial flame-shaped haemorrhages
- More in peripheral retina

Optic disc

- Mild papilloedema

Retina

- Retina relatively normal

RAPD

- Absent

Late findings (6–9 months)

- Venous sheathing
- Cilioretinal collaterals around optic disc

B) Ischaemic CRVO

Fundus findings

Veins

- Massive venous engorgement
- Severe congestion
- Extreme tortuosity

Retina

- Extensive retinal haemorrhages
- Classic:
  - **"Splashed tomato appearance"**

Cotton wool spots

- Numerous soft exudates
- Usually >6–10 spots

Macula

- Severe macular oedema
- Macular haemorrhages present

Optic disc

- Marked disc oedema
- Hyperaemia present

RAPD

- Present
- Important sign of ischaemic CRVO

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Includes:

- Visual acuity
- IOP measurement
- Gonioscopy

Purpose

- Detect POAG
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Investigate for:

- Hypertension
- Diabetes mellitus
- Hyperlipidaemia
- Homocysteinaemia
    - Especially in young patients

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Method

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Occurs in:

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Time

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Other Complications

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- Subhyaloid haemorrhage
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20. HYPERTENSIVE RETINOPATHY

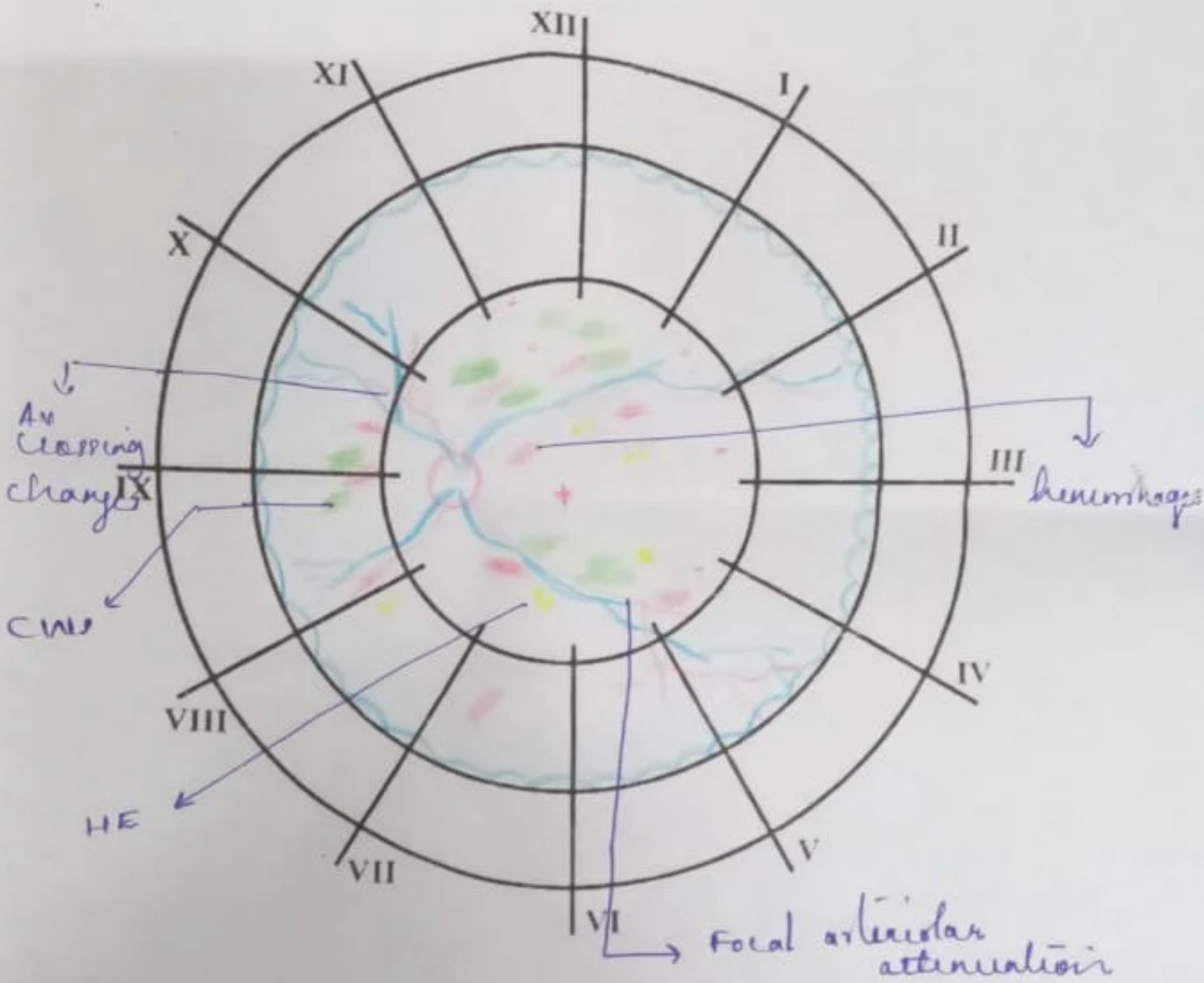


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FUNDUS CHART

O.D.	O.S.

Patient :	
Date :	



Hypertensive Retinopathy

Hypertensive Retinopathy — Very Simple Notes

What is it?

Damage to the retina due to **high blood pressure (HTN)**.

Pathogenesis (How damage happens)

1. Vasoconstriction

Blood vessels become narrow → less blood supply → retinal damage.

2. Arteriosclerosis

Long-standing HTN makes vessels thick and hard.

3. Increased vascular permeability

Blood and fluid leak out → hemorrhages, exudates, retinal edema.

Clinical Types

1) Simple hypertension (without sclerosis)

Usually in **young patients**.

Features

- General narrowing of retinal arteries
- Flame-shaped hemorrhages
- Cotton wool spots may be present

2) Hypertension with senile sclerosis

Usually in **elderly (>50 years)**.

Important signs

Gunn's sign

Vein gets compressed at AV crossing.

Salus sign

Vein bends at AV crossing.

Bonnet sign

Vein becomes banked/distended near crossing.

Other features

- Hard exudates
- Retinal hemorrhages

3) Chronic hypertension with compensatory arteriolar sclerosis

Also called:

"Albuminuric" or "Renal retinopathy"

Features

- Narrow tortuous vessels
- AV nicking
- Multiple retinal hemorrhages
- Cotton wool spots
- Hard exudates
- Vision reduced

4) Malignant (acute) hypertensive retinopathy

Most severe form.

Features

- Severe arteriolar narrowing
- Retinal edema
- Papilloedema
- Flame hemorrhages
- Cotton wool spots
- Hard exudates
- Macular star may occur
- Vision severely affected

Special lesions

- Elschnig spots
- Siegrist streaks

Prognosis

Poor if BP not controlled quickly.

Keith & Wegner Grading

Grade 1

- Mild arteriolar narrowing
- Broadening of light reflex

Grade 2

- More narrowing
- AV crossing changes (Salus sign)

Grade 3

Grade 2 +:

- Copper wiring
- Gunn sign
- Bonnet sign
- Flame hemorrhages
- Cotton wool spots
- Hard exudates

Grade 4

Grade 3 +:

- Silver wiring
- Papilloedema

Treatment

Mild

- Control BP

Moderate

- BP control
- Reduce cholesterol/risk factors

Malignant HTN

- Emergency treatment
- Lower BP slowly

Pregnancy-Induced HTN Retinopathy

Features

- High BP
- Proteinuria
- Edema
- Narrow vessels
- Hemorrhages
- Cotton wool spots
- Retinal edema

Treatment

- Usually improves after delivery
- Severe cases may need termination of pregnancy

Retinal Detachment (RD)

Retinal detachment = separation of **neurosensory retina** from the **retinal pigment epithelium (RPE)**.

Classification

1. **Rhegmatogenous RD** → due to retinal break/tear
2. **Tractional RD** → due to vitreoretinal traction pulling retina
3. **Exudative (Solid) RD** → due to fluid accumulation under retina without tear

1. Rhegmatogenous Retinal Detachment (Most common)

Cause

A **hole or tear** forms in retina → liquefied vitreous enters beneath retina → retina gets detached.

Risk Factors

- Age: 40–60 years
- Myopia
- Trauma
- Aphakia / pseudophakia
- Posterior vitreous detachment (PVD)
- Peripheral retinal degenerations:
  - Lattice degeneration
  - Snail-track degeneration

Symptoms

- Floaters (black spots)
- Photopsia (flashes of light)
- Visual field defect ("curtain coming down")
- Sudden painless loss of vision if macula involved

Signs

On Fundus Examination

- Retina appears:
  - Greyish
  - Elevated
  - Folded
  - Mobile and oscillates with eye movement
- Retinal vessels appear dark
- Retinal tear may be seen
- In total RD → funnel-shaped retina

Visual Field

- Scotoma corresponding to detached retina

ERG

- Reduced or absent

Ultrasonography (USG B-scan)

Useful when fundus cannot be visualized:

- Cataract
- Vitreous hemorrhage
- Corneal opacity

Configuration

- **Convex**

Treatment

Seal retinal break

- Laser photocoagulation
- Cryotherapy
- Diathermy

Reattach retina

- Scleral buckling
- Drainage of subretinal fluid
- Pars plana vitrectomy

2. Tractional Retinal Detachment

Cause

Fibrous tissue in vitreous contracts and mechanically pulls retina away.

Causes / Risk Factors

- Proliferative diabetic retinopathy
- Retinopathy of prematurity
- Trauma
- Sickle cell retinopathy
- Vitreoretinal traction syndromes
- Toxocariasis
- Plastic uveitis

Symptoms

Usually:

- Gradual visual loss
- No flashes or floaters

Signs

- Vitreoretinal traction bands seen
- Retina less mobile
- Shifting fluid absent

Configuration

- **Concave**

Treatment

- **Pars plana vitrectomy**
- Cut tractional bands
- Internal tamponade:
  - Silicone oil
  - Long-acting gas

3. Exudative (Solid) Retinal Detachment

Cause

Fluid accumulates beneath retina due to inflammation, tumor, or vascular leakage.

Causes

Systemic

- Hypertension
- Preeclampsia
- Blood dyscrasias

Ocular

- Choroidal tumors
- Posterior scleritis
- Harada disease
- Coats disease
- Choroidal neovascularization

Important Features

- No retinal tear
- No photopsia
- Shifting fluid present (very important)
- Retina appears smooth and bullous

Signs

- Shifting subretinal fluid
- Transparent detached retina
- Source of fluid may be seen on FFA

Configuration

- **Convex**

Treatment

Treat underlying cause:

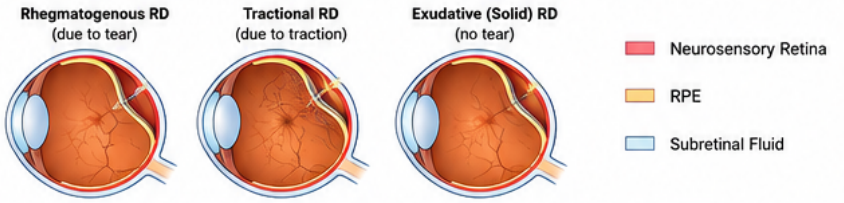
- Control inflammation
- Treat tumor
- Manage vascular disease

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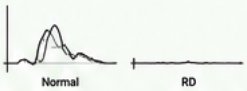
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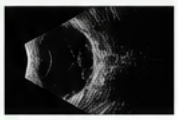
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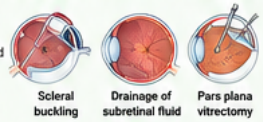
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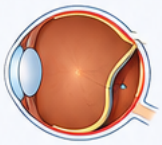
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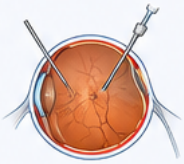


Treatment

- Pars plana vitrectomy
- Cut tractional bands

Internal tamponade:

- Silicone oil
- Long-acting gas





Silicone oil



Long-acting gas

3. EXUDATIVE (SOLID) RETINAL DETACHMENT

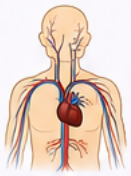
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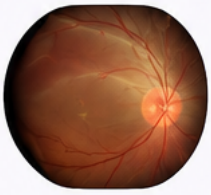
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Important Features

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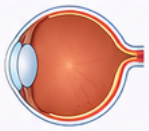
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- Treat tumor
- Manage vascular disease



Control inflammation



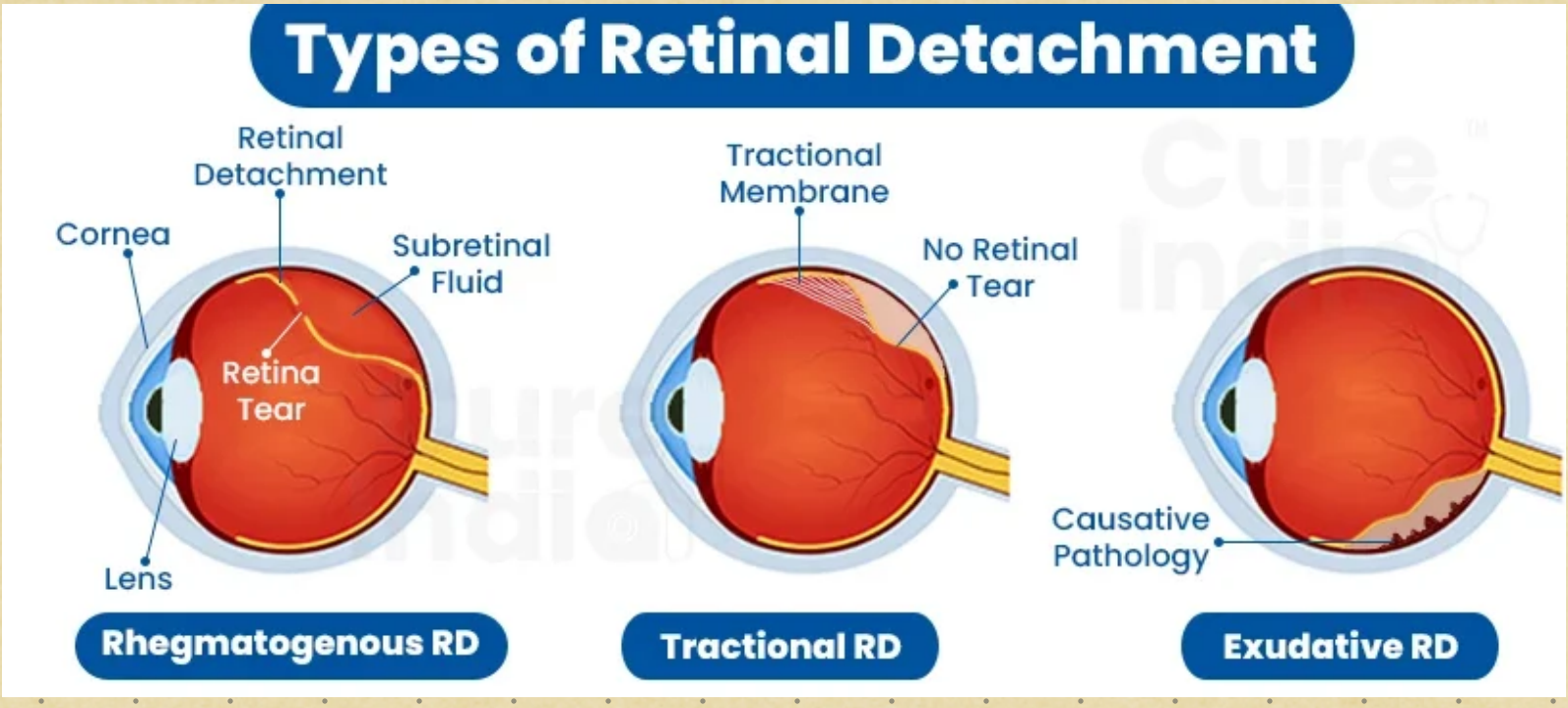
Treat tumor



Manage vascular disease

QUICK COMPARISON

Feature	Rhegmatogenous RD	Tractional RD	Exudative (Solid) RD
Cause	Retinal break → fluid entry	Vitreoretinal traction	Fluid from inflammation, tumor, or vascular leakage
Tear	Present	Absent	Absent
Photopsia / Floaters	Present	Absent	Absent
Shifting Fluid	Absent	Absent	**Present**
Retina Appearance	Greyish, elevated, folded	Traction bands, less mobile	Smooth, bullous, transparent
Configuration	Convex	Concave	Convex


Types of Retinal Detachment
Retinal Detachment
Cornea
Subretinal Fluid
Retina Tear
Lens
Rhegmatogenous RD
Tractional Membrane
No Retinal Tear
Tractional RD
Causative Pathology
Exudative RD



RETINOBLASTOMA

THE COMMONEST MALIGNANT INTRAOCULAR TUMOUR IN CHILDREN



1. DEFINITION

Retinoblastoma is a malignant tumour arising from the immature neuroepithelial cells (retinoblasts) of the developing neurosensory retina in one or both eyes.

It is the **most common intraocular tumour of childhood** and the **second most common malignant intraocular tumour after uveal melanoma.**

Key Point
Retinoblastoma is a potentially curable tumour when detected early.

2. EPIDEMIOLOGY

- **Prevalence:** 1 in 15,000–20,000 live births
- Accounts for about 3% of all childhood cancers
- **Age:** Usually occurs between 1–2 years
- **Sex:** No sex predisposition
- **Laterality:**
  - Unilateral in 70–75%
  - Bilateral in 25–30%
  - One eye is usually affected earlier and more extensively.

3. GENETICS & HEREDITY

Retinoblastoma gene (RB) located on **chromosome 13q14 is a tumour suppressor** (anti-oncogenic) gene.

Knudson two-hit hypothesis:
Deletion/inactivation of both alleles of RB gene leads to tumour.

13q14

1. Heritable (Germline) Retinoblastoma
- 40% of cases
- Autosomal dominant inheritance
- Usually bilateral, multifocal
- Presents earlier

2. Non-heritable (Somatic) Retinoblastoma
- 60% of cases
- Usually unilateral, unifocal
- Sporadic, presents later

4. PATHOLOGY

Origin
Arises as malignant proliferation of immature retinal neural cells.

Histopathology
Tumour composed of small round blue cells with scant cytoplasm and large nuclei.

Rosettes (Characteristic)
- **Flexner-Wintersteiner rosettes:** Highly specific. Central lumen surrounded by tumour cells.
- **Homer-Wright rosettes:** No lumen, tumour cells arranged around neurofibrils.
- **Fleurettes:** Photoreceptor differentiation.
- **Pseudorosettes:** Tumour cells arranged around blood vessels; indicate poor differentiation.



5. CLINICAL FEATURES

I. QUIESCENT STAGE
- Leukocoria (60%) (white pupillary reflex/ "cat's eye reflex")
- Strabismus
- Defective vision
- Nystagmus (usually in bilateral cases)


Leukocoria

II. GLAUCOMATOUS STAGE (PAINFUL RED EYE)
- Secondary / neovascular glaucoma
- Redness, watering
- Corneal haze, raised IOP
- Enlarged eyeball (buphthalmos)
- Orbital cellulitis-like picture


Glaucomatous eye

III. STAGE OF EXTRAOCULAR EXTENSION
- Extension through sclera
- Fungating orbital mass
- Proptosis
- Orbital cellulitis-like features


Orbital extension

IV. STAGE OF DISTANT METASTASIS
- Lymphatic spread
- Direct extension to optic nerve and brain


Brain metastasis

6. TYPES OF RETINOBLASTOMA

1. **Endophytic:** Grows inward into vitreous cavity. White, creamy mass; vitreous seeding common; "cottage cheese" appearance.
2. **Exophytic:** Grows outward; causes exudative retinal detachment.
3. **Mixed:** Features of both endophytic and exophytic growth.
4. **Diffuse infiltrating:** Placoid retinal thickening; diagnosed late.

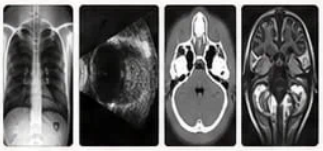
7. DIFFERENTIAL DIAGNOSIS OF LEUKOCORIA (PSEUDOGLIOMA)

- Congenital cataract
- Persistent hyperplastic primary vitreous (PHPV)
- Retinopathy of prematurity
- Toxocariasis
- Coats disease
- Endophthalmitis



8. DIAGNOSIS

- Examination under anaesthesia (EUA):
  - Fundus examination
  - Indirect ophthalmoscopy
  - IOP measurement
- Plain X-ray orbit: Calcification in ~75% cases.
- **B-scan ultrasonography:** Detects tumour mass and calcification.
- **CT scan:** Detects calcification, shows extent of intraocular and extraocular spread.
- **MRI:** Best for optic nerve involvement, intracranial extension and pinealoblastoma.


X-ray (Calcification) | B-scan USG | CT scan | MRI (Optic nerve involvement)

9. INTERNATIONAL CLASSIFICATION OF RETINOBLASTOMA (ICRB)

Group	Description
A	Small tumour confined to retina
B	Larger tumour, may be near fovea or disc
C	Local subretinal / vitreous seeds
D	Diffuse subretinal / vitreous seeds
E	Extensive tumour (≥50% retina) or any with high-risk features (poor prognosis)

10. TREATMENT

A. CONSERVATIVE (GLOBE SALVAGE)
Used in Groups A–D

1. Chemoreduction (CVE regimen)
- Carboplatin
- Vincristine
- Etoposide

2. Focal Therapies
- Cryotherapy – small anterior tumours
- Laser photocoagulation – posterior tumours
- Thermotherapy – small posterior tumours
- Plaque radiotherapy – localized vitreous disease

3. External Beam Radiotherapy (EBRT)
Now used less due to risk of secondary malignancy.

B. ENUCLEATION
Indications
- Group E tumours
- Optic nerve involvement
- Neovascular glaucoma
- Extensive retinal involvement

Optic nerve should be cut with maximum length.



C. PALLIATIVE THERAPY
Indications
- Orbital extension
- Intracranial spread
- Distant metastasis

Includes
- Chemotherapy
- Radiotherapy
- Orbital exenteration

11. METASTASIS

Spread occurs by:
- Lymphatic spread (preauricular & neighbouring nodes)
- Direct extension via optic nerve to brain

12. PROGNOSIS

Good prognosis:
- When detected early and treated before extraocular extension.

Poor prognostic factors:
- Optic nerve invasion
- Massive choroidal invasion
- Undifferentiated tumour cells
- Extraocular extension
- Late stage at presentation

13. IMPORTANT PEARLS

- ★ Leukocoria is the most common presenting sign.
- ★ Bilateral disease is usually heritable and earlier onset.
- ★ Flexner-Wintersteiner rosettes are pathognomonic.
- ★ Calcification on imaging is highly suggestive.
- ★ ICRB guides treatment and prognosis.
- ★ Early diagnosis improves globe salvage and survival.

14. SUMMARY

Retinoblastoma is a highly curable intraocular cancer of childhood when detected early and managed appropriately. A multidisciplinary approach with accurate staging, appropriate therapy and regular follow-up is essential for preserving life, vision and the eye.



Abbreviations: RB – Retinoblastoma gene; IOP – Intraocular pressure; CT – Computed Tomography; MRI – Magnetic Resonance Imaging; USG – Ultrasonography; CVE – Carboplatin, Vincristine, Etoposide; EBRT – External Beam Radiotherapy; ICRB – International Classification of Retinoblastoma

★ Dr Ashik Sureshkumar ★

Retinoblastoma — Very Simple MBBS Note

Definition

Retinoblastoma is the **most common malignant eye tumour in children**.
It arises from the **retina** and usually occurs in **early childhood**.

Etiology / Cause

Origin

- Tumour develops from **immature retinal cells** that fail to mature normally.

Genetics

- Due to mutation of **RB gene** on **chromosome 13q14**.
- Explained by **Knudson's two-hit hypothesis**.

Types

1. **Hereditary type**
   - About **10%**
   - **Autosomal dominant**
   - Usually **bilateral**
2. **Sporadic type**
   - About **90%**
   - Usually **unilateral**

Microscopy (Histology)

Two types of tumour cells are seen:

1. Poorly differentiated cells

- Large dark nuclei
- Little cytoplasm
- Necrosis common

2. Well differentiated cells

They form special arrangements called:

Rosettes

- **Flexner-Wintersteiner rosettes**
  - Specific for retinoblastoma
- **Homer Wright rosettes**
  - Not specific

Fleurettes

- Flower bouquet appearance
- Specific for retinoblastoma

Pseudorosettes

- Tumour cells arranged around blood vessels

Clinical Features

Symptoms

1. Leukocoria (most common)

- White pupillary reflex
- Called **"amaurotic cat's eye reflex"**





2. Squint (strabismus)

- Due to defective vision

3. Poor vision

4. Pain

- Due to raised intraocular pressure

5. Enlarged eyeball

- Advanced disease

Signs

Fundus findings

- Multiple yellow-white retinal masses

Vitreous seeding

- Tumour cells spread into vitreous

Pseudohypopyon

- White tumour cells in anterior chamber

Secondary glaucoma may occur

Types of Growth

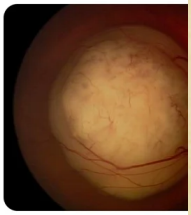
1. Exophytic type

- Grows outward
- Causes retinal detachment

2. Endophytic type

- Grows inward into vitreous





Stages of Retinoblastoma

1. Quiescent stage

- Lasts months to 1 year

2. Glaucomatous stage

- Enlarged painful eye
- Increased intraocular pressure

3. Extraocular extension stage

- Tumour spreads outside eyeball

4. Metastatic stage

Spread occurs through:

- Optic nerve
- Blood
- Lymphatics

Common metastasis sites

- Bone
- Liver

Differential Diagnosis (Causes of Leukocoria)

Pseudoglioma conditions

- Congenital cataract
- Toxocariasis
- Retrolental fibroplasia
- Persistent hyperplastic primary vitreous

Others

- Coats disease
- Retinal dysplasia
- Choroidal coloboma

Diagnosis

Eye examination

- Dilated fundus examination

Raised LDH

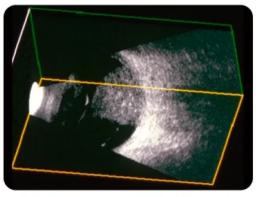
- In aqueous humour

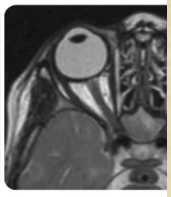
X-ray orbit

- Shows calcification

CT/MRI

- Confirm diagnosis
- Detect optic nerve or brain spread





Treatment

1. Chemotherapy + Radiotherapy

Common drugs:

- Carboplatin
- Vincristine
- Etoposide

2. Cryotherapy

- Small anterior tumours

3. Laser photocoagulation

- Small posterior tumours

4. Enucleation (treatment of choice)

- Removal of whole eyeball
- Done if:
  - Large tumour
  - Optic nerve involvement
  - Glaucoma present

5. Exenteration

- Removal of orbital contents
- Used in advanced stage

Prognosis

Good prognosis

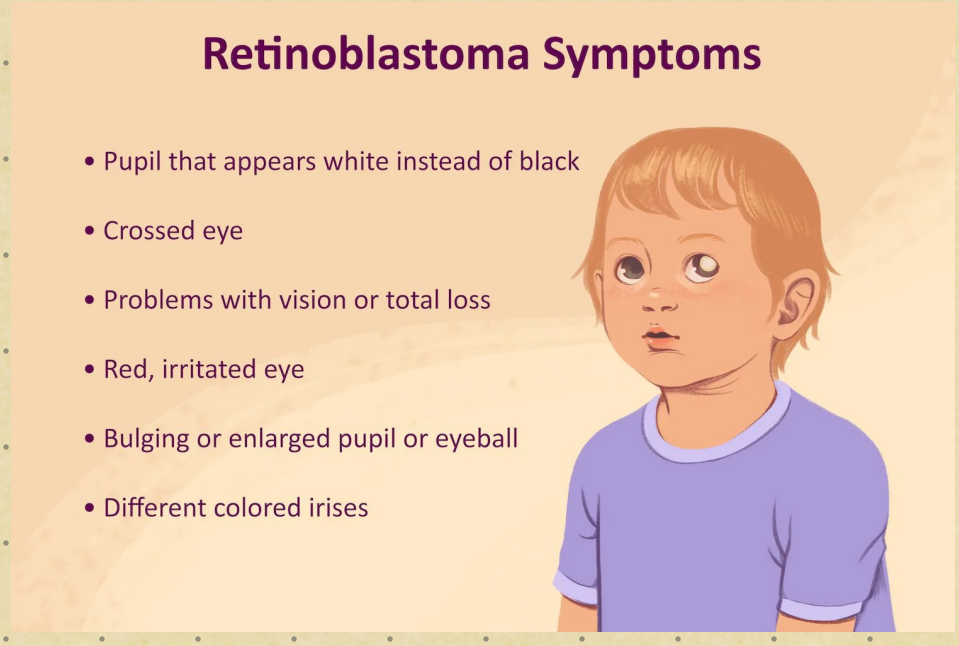
- If detected early
- Before optic nerve spread

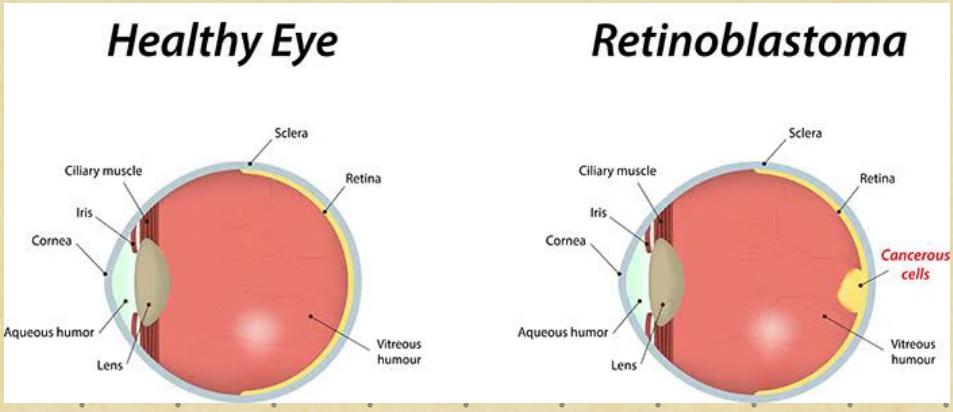
Poor prognosis

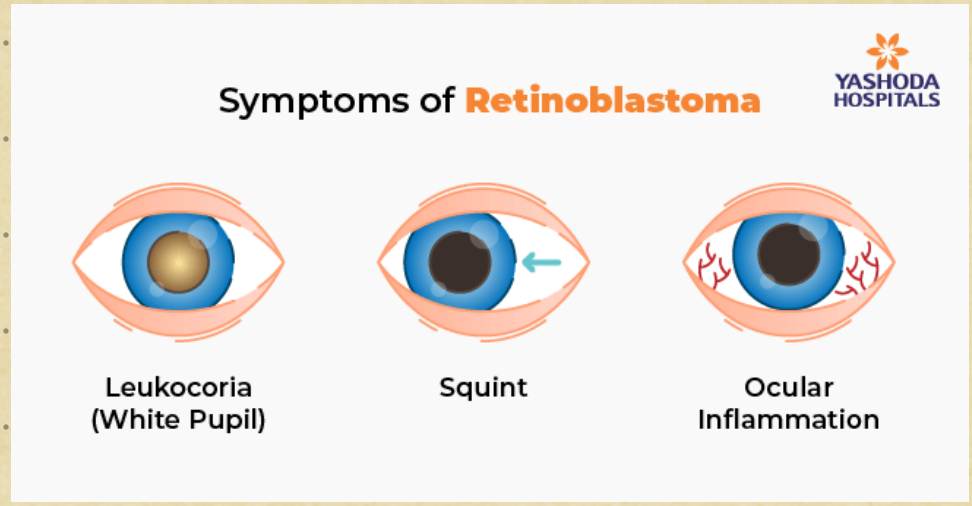
- Optic nerve involvement
- Extraocular spread
- Metastasis

One-line Exam Summary

Retinoblastoma = common malignant retinal tumour in children presenting with leukocoria ("cat's eye reflex"), diagnosed by fundus exam and imaging, treated mainly by enucleation and chemotherapy.

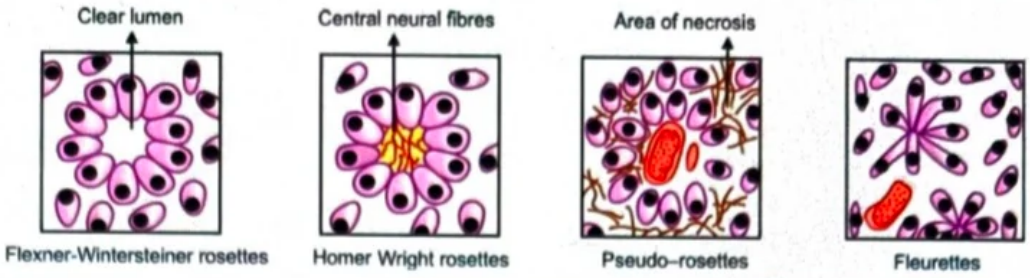






- **Microscopic examination** shows 2 types of cellular characteristics:
  1) Poorly differentiated cells with large hyperchromatic nuclei and scanty cytoplasm along with necrosis. They resemble the nuclear layer of the retina.
  2) Well-differentiated tumour cells may be arranged in 2 special forms: **a.** Rosettes **b.** Fleurettes.
     - **a. Rosettes**
       - ⇒ Flexner-Wintersteiner rosettes—specific for retinoblastoma.
       - ⇒ Homer Wright rosettes— not specific for retinoblastoma (can also be seen in neuroblastoma & medulloepithelioma).

- ⇒ Pseudorosettes—tumour cells are clustered around blood vessels in necrotic retinoblastoma.
- **b. Fleurettes**: specific for retinoblastoma – flower bouquet type aggregation of tumour cells.



49. Causes of vitreous hemorrhage (Give Precise Answers – 2023 Feb, 2010 Scheme):
- Diabetic retinopathy
- Retinal tear with posterior vitreous detachment (PVD)

50. Two causes of vitreous haemorrhage (Give Precise Answers – 2023 Aug):
- Trauma
- Central retinal vein occlusion

51. Two causes of cherry red spot in macula (Give Precise Answers – 2021 Dec):
- Central retinal artery occlusion (CRAO)
- Commotio retinae (Berlin's edema)

52. Two causes of cherry red spot in retina (Give Precise Answers – 2024 June):
- Tay–Sachs disease
- Niemann–Pick disease

53. Two causes of sudden loss of vision (Give Precise Answers – 2024 Feb):
- Central retinal artery occlusion (CRAO)
- Vitreous hemorrhage

54. Mention two causes for sudden loss of vision (Give Precise Answers – 2014 Mar):
- Acute angle-closure glaucoma
- Central retinal vein occlusion

55. Name four retinal findings following blunt trauma to the eye (Give Precise Answers – 2014 Sep):
- Commotio retinae (Berlin's edema)
- Retinal hemorrhages
- Retinal tear
- Retinal detachment

8. Enucleation

Definition

Enucleation is **removal of whole eyeball after cutting optic nerve, while extraocular muscles and orbital contents are left behind**.

Indications

- Retinoblastoma
- Painful blind eye
- Severe ocular trauma
- Malignant intraocular tumors

Complications

- Orbital infection
- Implant extrusion
- Socket contracture

9. Two Indications of Enucleation

1. Retinoblastoma
2. Painful blind eye

Other indications:

- Severe trauma
- Sympathetic ophthalmia prevention

10. Leukocoria

Definition

White pupillary reflex seen through pupil.

Causes

- Congenital cataract
- Retinoblastoma
- Retinopathy of prematurity
- Persistent hyperplastic primary vitreous (PHPV)

11. Two Differential Diagnoses of Leukocoria

1. Retinoblastoma
2. Congenital cataract

20. Two Macular Function Tests

1. Amsler grid test
2. Maddox rod test

Other tests:

- Photostress test
- Laser interferometry

21. Mention Two Causes for Sudden Loss of Vision

1. Central retinal artery occlusion (CRAO)
2. Retinal detachment

Other causes:

- Central retinal vein occlusion
- Vitreous hemorrhage
- Optic neuritis

22. Mention Two Syndromes Associated with Retinitis Pigmentosa

1. Usher syndrome
2. Laurence-Moon-Biedl syndrome

Important Viva Point

Classical triad of retinitis pigmentosa

1. Night blindness
2. Bone-spicule pigmentation
3. Attenuated retinal vessels