2.9 Retina: Acquired maculopathies, Dystrophies, Retinal detachment, tumors

Plan

Anatomy

- What is fundus and what are retinal layers

Acquired macular disorders

- Age related macular degeneration (ARMD)
- Central serous chorioretinopathy
- Macular hole

Retinal dystrophies

- Retinitis pigmentosa

Retinal detachment

- Types and treatment

Retinal tumours

- Types, Retinoblastoma & Malignant melanoma

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Anatomy

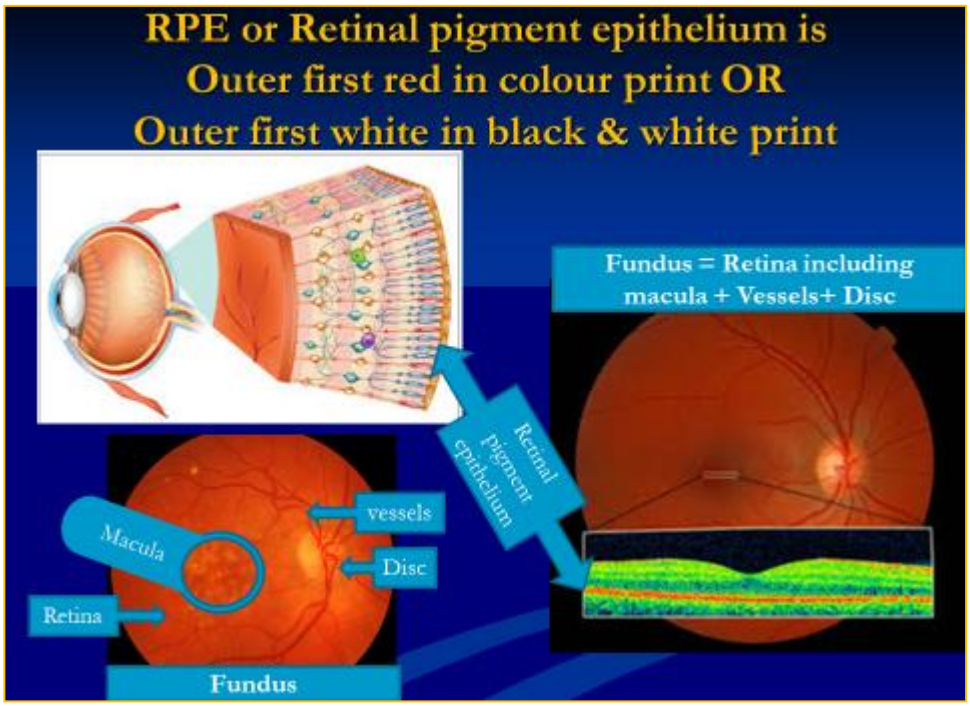
Fundus = Retina including macula + vessels + optic disc

Retina consists of RPE & neurosensory retina

RPE = Retinal pigment epithelium is single layer attached to neurosensory retina

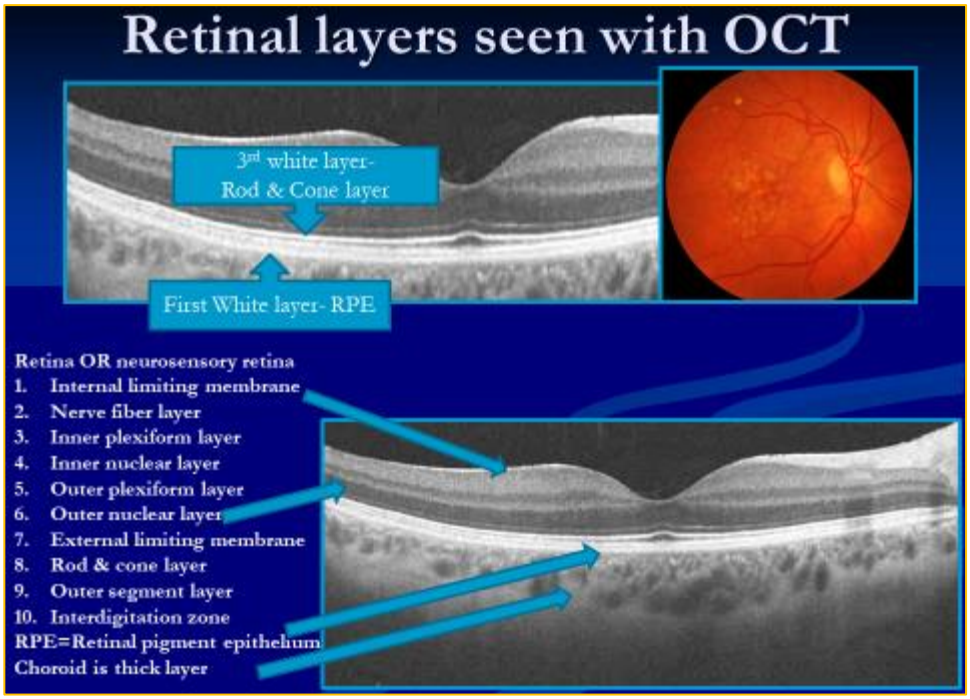
Neurosensory Retina = Layers 1-10 in OCT is retina or neurosensory retina.

Separation of RPE and neurosensory retina is retinal detachment



First outer most layer on OCT in retinal pigment epithelium or RPE.

Third outer most is inner segment outer segment layer (IS/OS) made of rods & cones



Acquired maculopathies: ARMD

What is ARMD

- **ARMD** is **A**ge **R**elated **M**acular **D**egeneration
- Most common cause of visual loss/blindness in world
- Affects people over age 50
- Females more than males
- Caucasians (whites) more than Asian
- Africans rarely affected

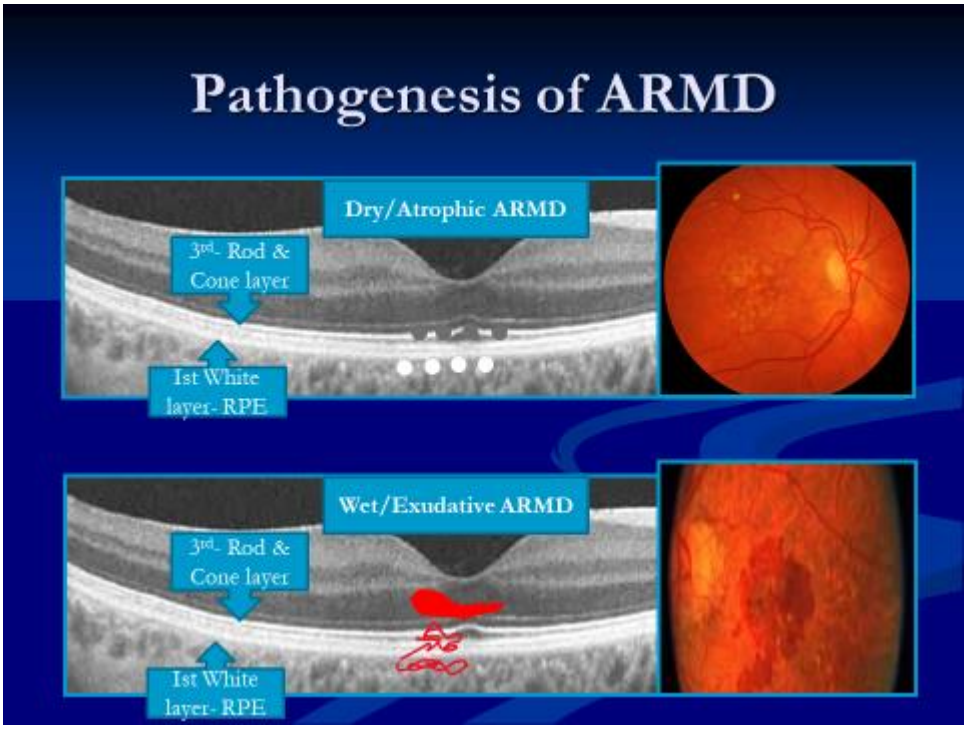
First clear concept of retinal layers, RPE and choroid in retinal anatomy section

Risk factors for ARMD

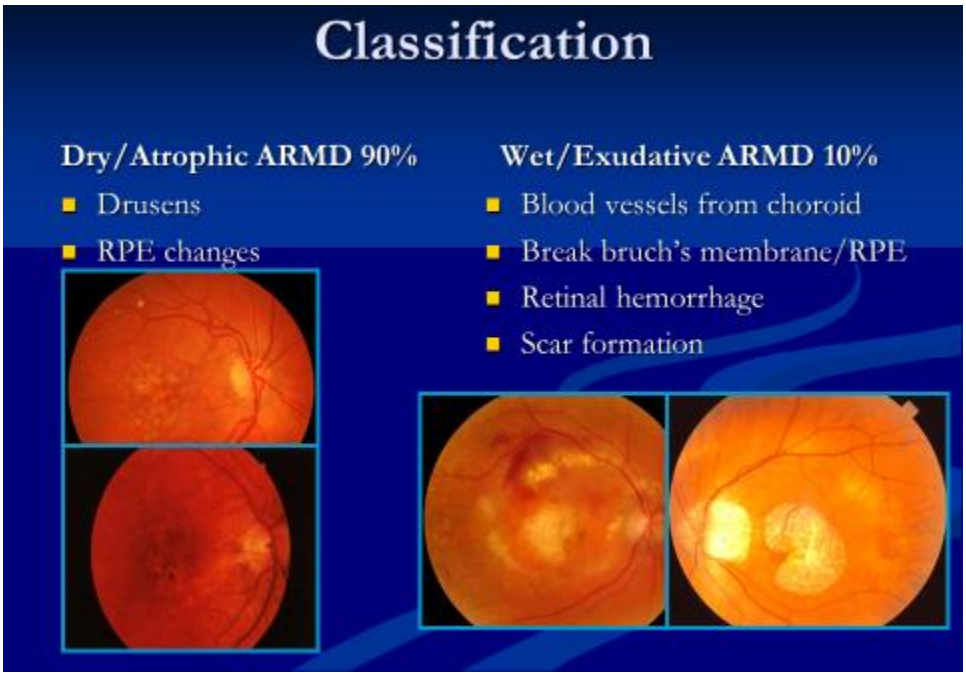
- Caucasians race
- Positive family history of ARMD
- Female sex
- Cigarette smoking
- Hypertension and raised cholesterol
- Hypermetropia
- History of previous high exposure of UV light
- Cataract surgery

Pathogenesis

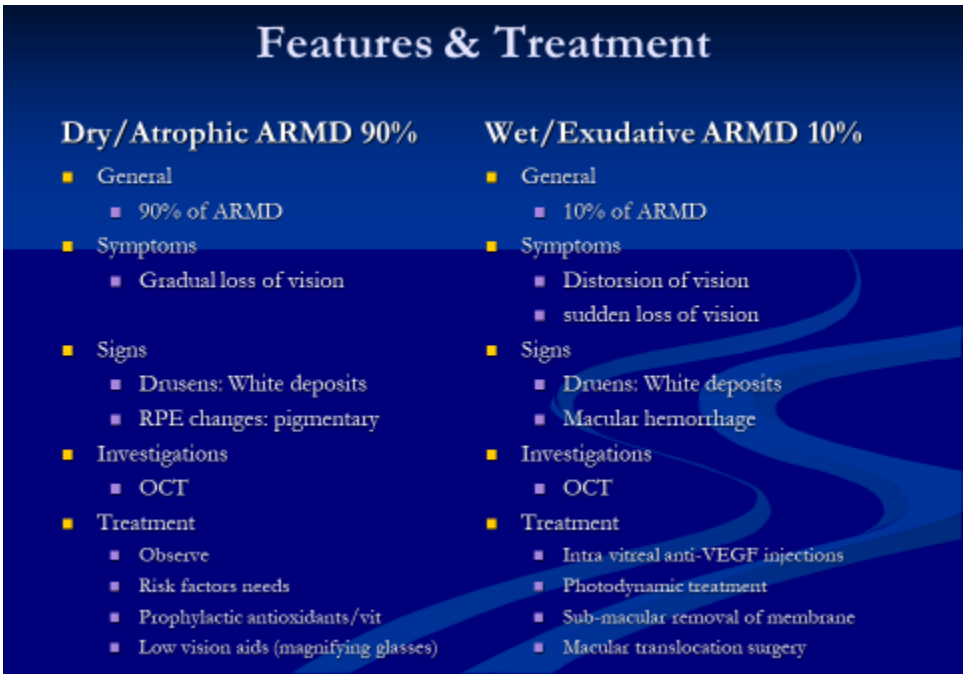
Dry ARMD: White drusens appear behind RPE and gradually cause damage to IS/OS layer and so gradually cause damage to vision.
Wet ARMD: Blood vessels develop behind RPE, break through RPE and cause retinal bleeding which later convert in to scar. Loss of vision is sudden.



Classification of ARMD



Clinical features and treatment



Acquired maculopathies: CSCR

Definition

There is fluid between RPE and neurosensory retina

Pathogenesis

It is because of exudation of fluid from choroid or para foveal capillaries

Clinical features

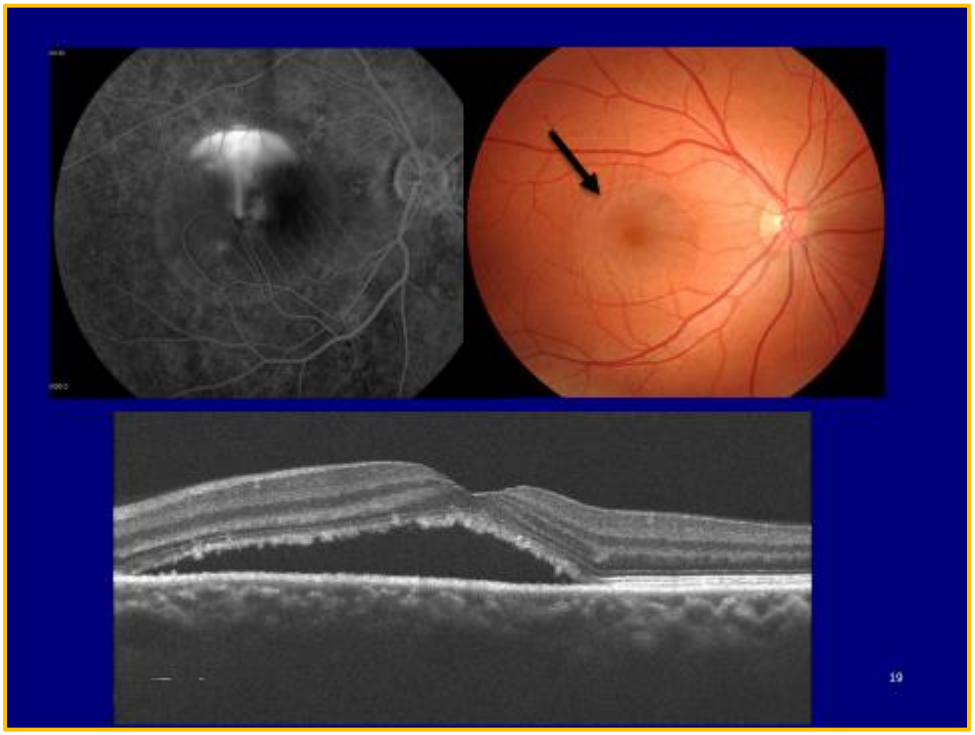
Mostly young adults affected
Blurry vision
Black patch in central vision
Micropsia (things appear smaller in affected eye)
Circular swelling seen in macular area (as in photo)

Investigations

FFA: Leakage of fluorescein dye on fundus fluorescein angiography (see photo)
OCT: Fluid between RPE (outer most white layer) and retina (see photo)

Treatment

Mostly self healing but takes months
Many other options like Laser and anti VEGF injections are available



Acquired maculopathies: Macular hole

Definition

It is hole in retinal layers at macula.

Symptoms & signs

Symptoms:

Blurry vision

Signs

Partial or full thickness hole in retinal layers (Not in RPE)

Surrounding fluid may be present

Causes

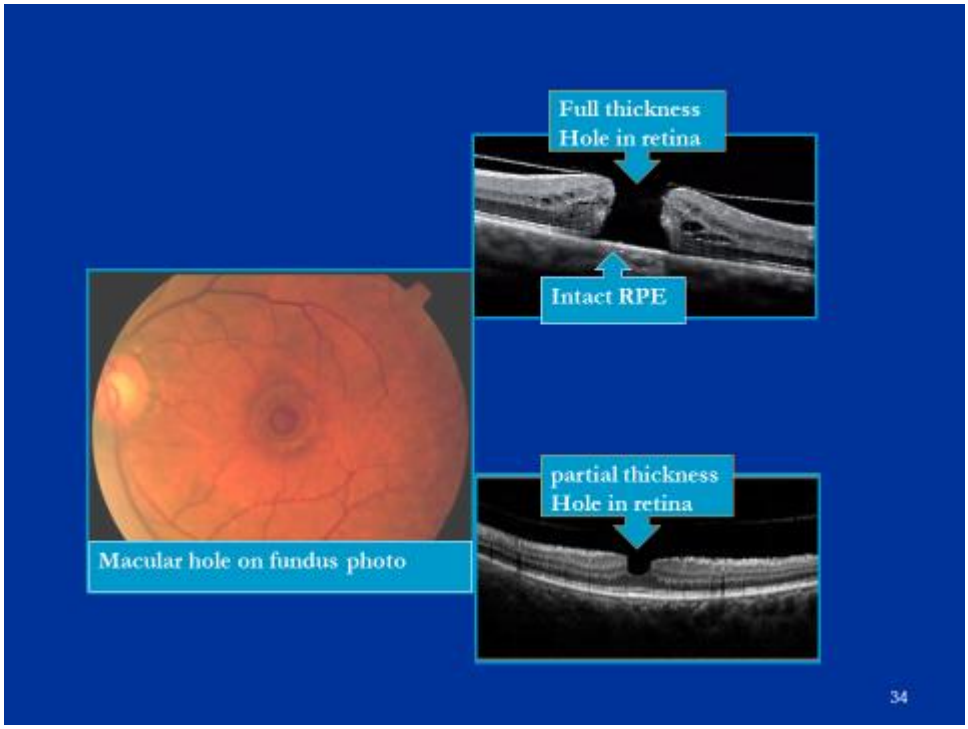
- Mostly idiopathic
- Trauma
- Myopia

Investigations

OCT will show partial (inner layers) or full thickness hole (all layers but not RPE).

Treatment

Par plana vitrectomy (PPV) with gas or silicone oil tamponade.



Retinal dystrophies: Retinitis pigmentosa

Introduction

Retinitis pigmentosa is group of diseases which affects rods and cones.
Age of onset depends on mode of inheritance and usually starts in childhood.
Disease may be sporadic or Inheritance may be AD, AR or XLR. XLR is least common but most severe while AD is most common and least severe.

Symptoms & signs

Classical symptom is defective night vision because of early damage to rods. Later vision is affected because of damage to cones.
Classical signs are triad of

- Bone spicule retinal pigmentation
- Attenuated arterioles
- Consecutive optic atrophy or waxy pale disc

Complications

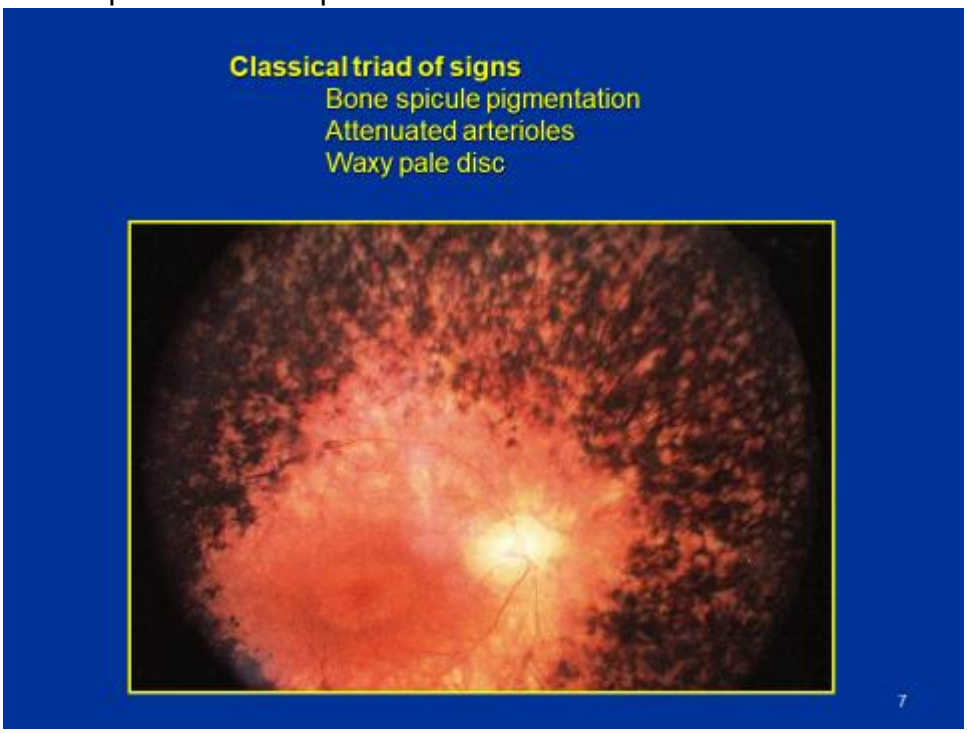
- Posterior capsular cataract
- Pen angle glaucoma
- Keratoconus

Investigations

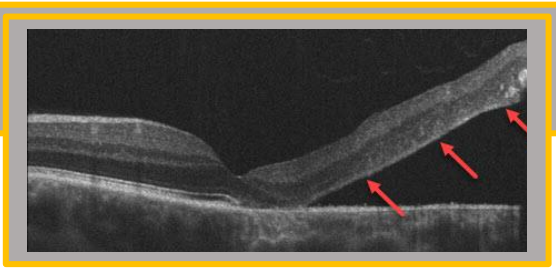
ERG or electro retino gram

Treatment

N specific treatment but gene therapy may hold future
Regular follow ups to treat complications and low vision aids.



Retinal detachment

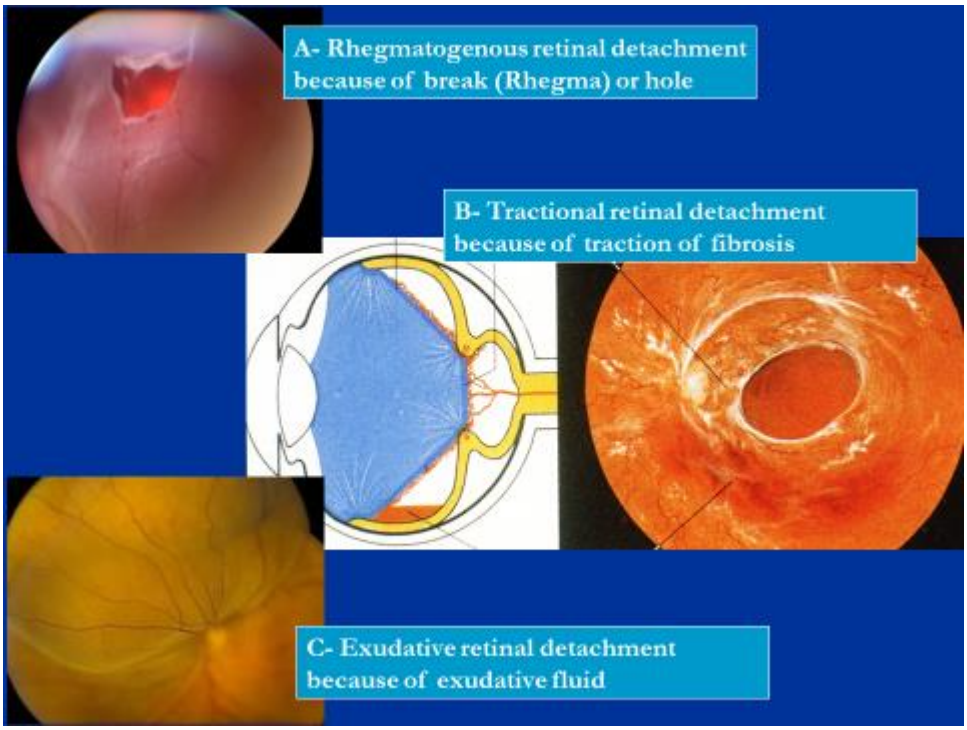


Definition

It is separation of neurosensory retina from retinal pigment epithelium (RPE).

Types, pathogenesis & treatment

Types of detachment	Pathogenesis	Treatment
A- Rhegmatogenous retinal detachment (Most common) Causes: Breaks in retina because of degenerations or trauma	Because of hole or break (Rhegma) in retina. Fluid enters through break (Rhegma) and **pushes** neurosensory retina to separate from RPE	External method: Sclera is pushed forward to attach retina Internal method (PPV): Retina in pushed towards RPE/sclera
B- Tractional retinal detachment Causes: Proliferative diabetic retinopathy Trauma	Fibrosis **pulls** retina and hence separate it from RPE	External method Internal method
C- Exudative Retinal detachment (Least common) Causes: Inflammation like uveitis & scleritis Retinal tumours	Exudative fluid **pushes** neurosensory retina and separates it from RPE	Treat the cause Occasional drainage of fluid



Retinal tumours

Benign conjunctival tumours like conjunctival nevus, papilloma and limbal dermoid.
Malignant cnjunctival tumours like conjunctival melanoma.
Iris tumours like iris nevus and iris melanoma.
Ciliary body tumours like ciliary body melanoma
Retinal vascular tumours like capillary haemangioma.
Neural retinal tumours like **retinoblastoma.**
Choroidal tumours like choroidal nevus and **choroidal melanoma.**

Retinoblastoma is most common tumour in children.
It is usually unilateral but may be bilateral in 25% cases.
Presentation is usually as white pupil or leukocoria.
May present as enlargement of globe or hypopyon.
Differential diagnosis is congenital cataract & retinopathy of prematurity.
Diagnosis is examination, B scan, CT scan & MRI.
Treatment depends on size of tumour and may be with chemotherapy, radiotherapy, laser and enucleation.


White pupil as leukocoria

Malignant melanoma is most common adult tumour.
Most cases are sporadic.
Presentation: usually no symptoms and tumour is detected by chance as elevated lesion. Large tumours can cause blurry vision.
Diagnosis is with FFA, B scan, C scan & MRI.
Treatment depends on size and may be laser, excision, radiotherapy and enucleation.


Large elevated tumour


Tumour on B scan