EyeToday

COATS DISEASE

Dr. R.Ram Sudarshan
Vitreo-retinal fellow
Aravind eye hospital

HISTORY

- George Coats first described the disease in 1908
- Initial Coats classification consisted of three stages
- Third stage later confirmed as a separate entity – Retinal Hemangioblastoma by Eugen Von Hippel
- Leber's Disease (after theodre Von Leber) – Earlier stage of coats (Milder form of coats)

GENETICS

- Part of spectrum of genetic disorders known as “Retinal Hypovasculopathies”

- The genes that have been associated are:

1. Norrie disease protein
2. CRB1
3. PANK 2

PATHOGENESIS

Breakdown of BRB

↓

Leak of plasma into vessel wall

↓

Thickening ,necrosis of vessel wall leading to sausage like shape of vessel

Abnormal pericytes and endothelial cells causes abnormal retinal vasculature and formation of microaneurysms and closure of vessels - ISCHEMIA

↓

Leakage of Cholesterol rich exudates ,hemorrhage, cysts ,edema, lymphocytic infiltration and fibrin deposition leading to thickening and edema

CLINICAL PRESENTATION

- Age : 8-16 years
- Sex : Males (three times more affected)
- Laterality : Unilateral (80-95%)
- Race : No racial or ethnic predilliction

SYMPTOMS

- Decreased VA
- Strabismus
- Leukocoria
- Pain
- Hetrochromia

SIGNS

Anterior Segment (mostly normal)

- Cataract
- Iris neovascularisation (later stages)
- Choloesterol in anterior chamber
- Megalocornea

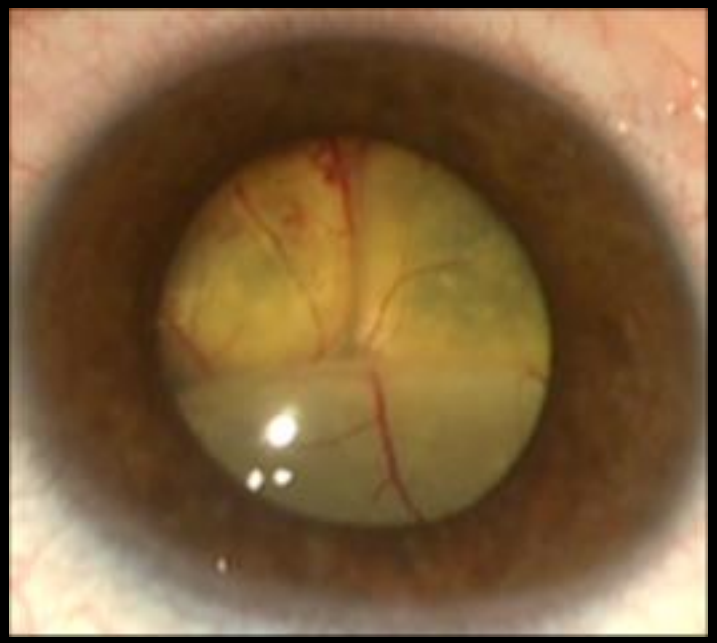
Posterior Segment

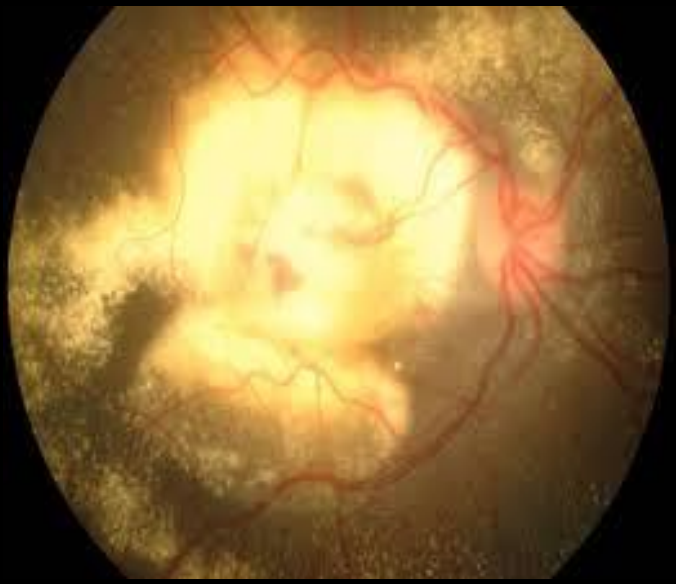
- Retinal Telengectasia
- Aneurysms
- Intraretinal exudation
- Exudative retinal detachment
- Retinal macrocyst

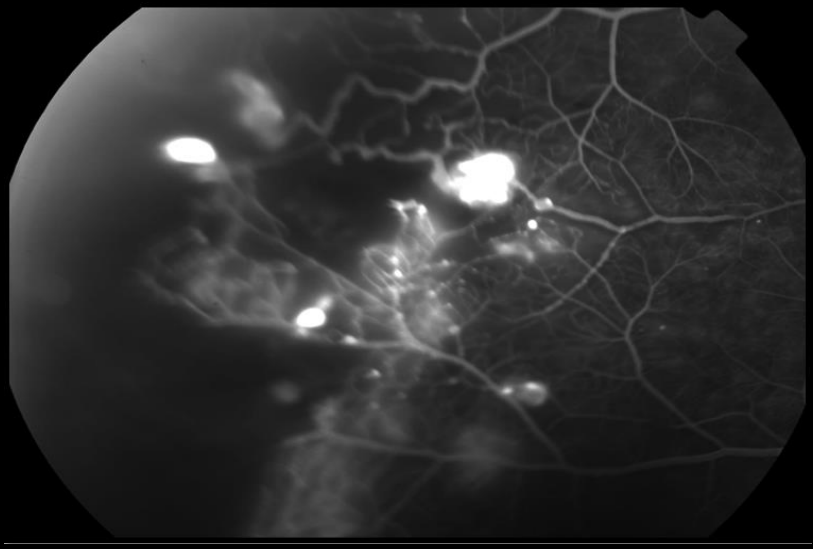
Typical Features:

Localized subretinal exudation with adjacent vascular anomalies

Exudation more common during active form of disease

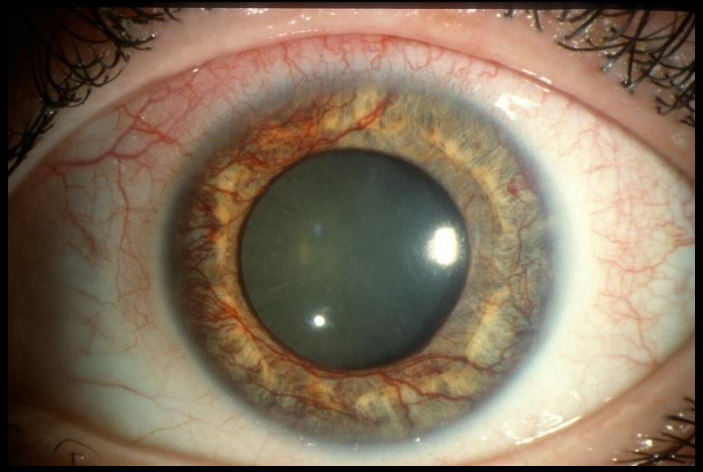






END STAGE DISEASE

- Neovascular glaucoma



- Exudative retinal detachment

- Pthisis Bulbi

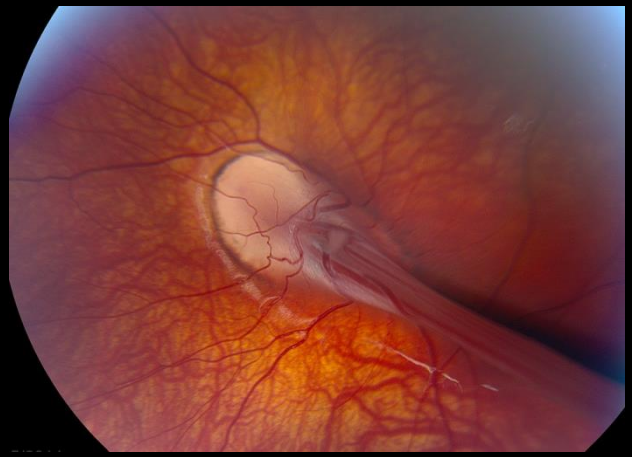
STAGING (SHIELDS et al ;2000)

STAGES	SIMPLIFIED FORMAT	CRITERIA
1	T only	Retinal telengectesia
2 2A 2B	T+E Extrafoveal Foveal	Telengectesia+Exudation
3A A1 A2 3B	T+E+D Extrafoveal detachment Foveal detachment Total Exud detachment	Telengectesia+Exudation +Eexudative RD
4	T+E+D+G	Total RD+ sec glaucoma
5	T+E+D+G+P	End stage with Pthisis bulbi

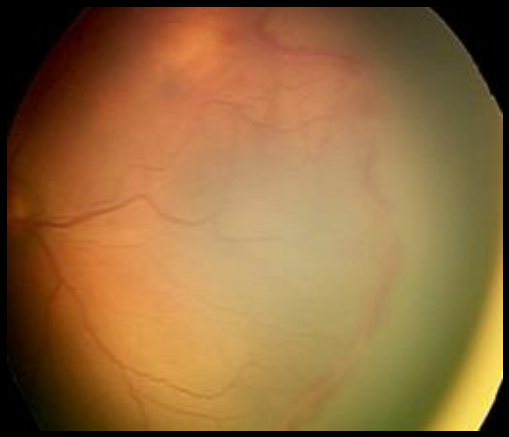
DIFFERENTIAL DIAGNOSIS

DIAGNOSIS	DIFFERENTIAL POINTS
RETINOBLASTOMA	Mass lesion + calcification Misdiagnosis can lead to wrong treatment regimen
RETINOPATHY OF PREMATURITY	Very early presentation Bilateral presentation, presence of avuscular zone , demarcation line/ridge Risk factors
FEVR	Bilateral presentation Presence of retinal drag
PHPV	Microphthalmos Retrolental fibrovascular membranes Family h/o+
OCULAR TOXOCARIASIS	Due to Toxocara canis Presence of anterior uveitis, vitris , fundus granuloma

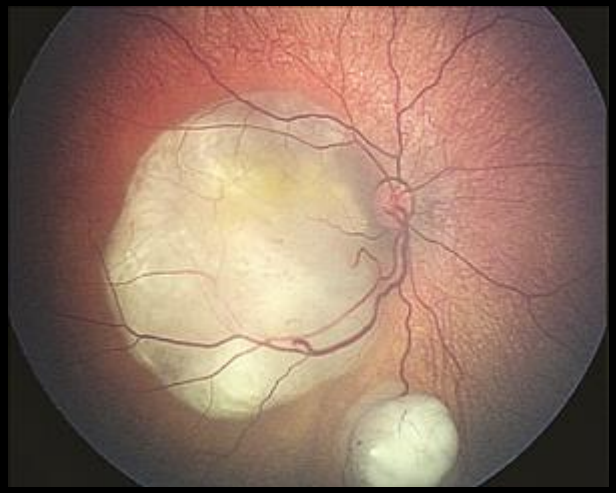
FEVR



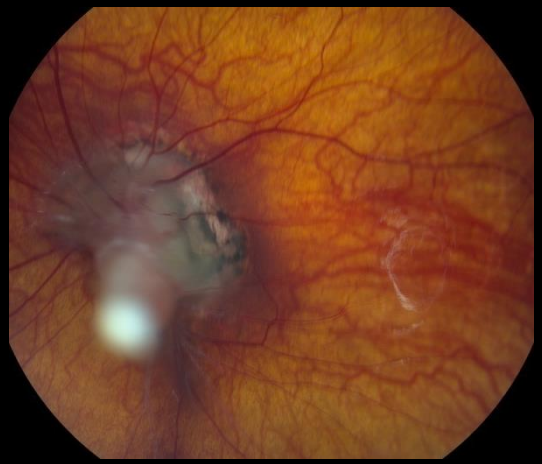
ROP



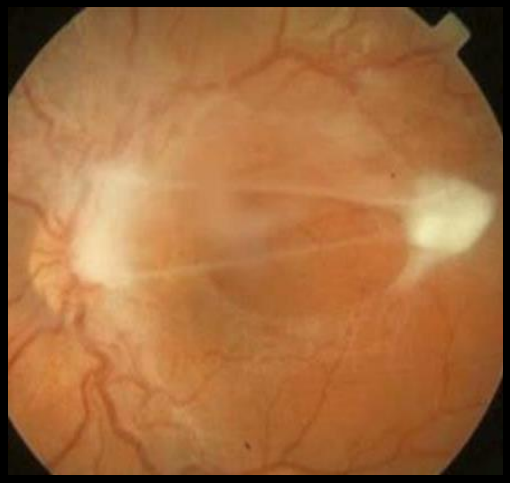
RB



PHPV



TOXOCARIASIS



ATYPICAL PRESENTATION OF COATS

VARIANTS	FEATURES
ADULT ONSET COATS:	Smaller area of involvement Slower progression Large haemmorhage near vessel dilatations Frequently associated with Hypercholestermia ,which is not found in the juvenile form
COATS PLUS SYNDROME :	Associated with a) Skeletal disorders b) Extrapyramidal and Cerebellar movement disorder c) Seizures c) Postnatal Growth Failure

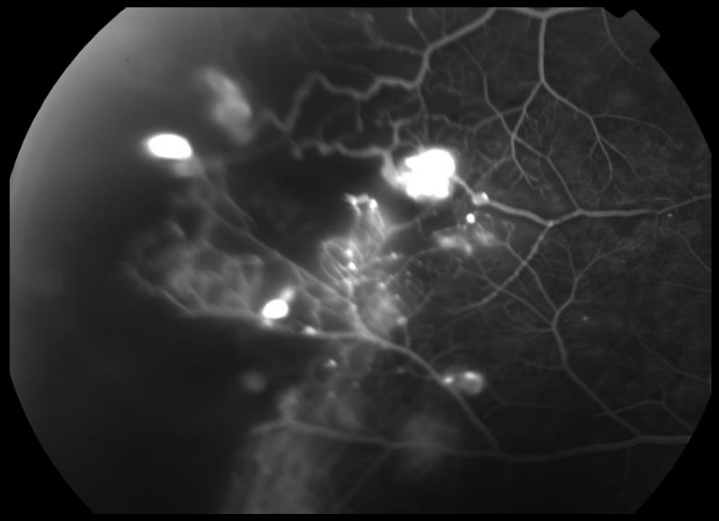
CONDITIONS THAT CAN PRESENT WITH COATS LIKE RETINAL FINDINGS

SYSTEMIC CONDITIONS	**Turner syndrome** **Epidermal nevus syndrome** **Alport syndrome**
OCULAR CONDITIONS (OTHER THAN DD MENTIONED BEFORE)	Congenital cataract Norrie disease Vasoproliferative tumor Macular dystrophy
OCULAR CONDITIONS THAT MIMIC COATS AT ANY AGE	BRVO Eale's disease Vasculitis

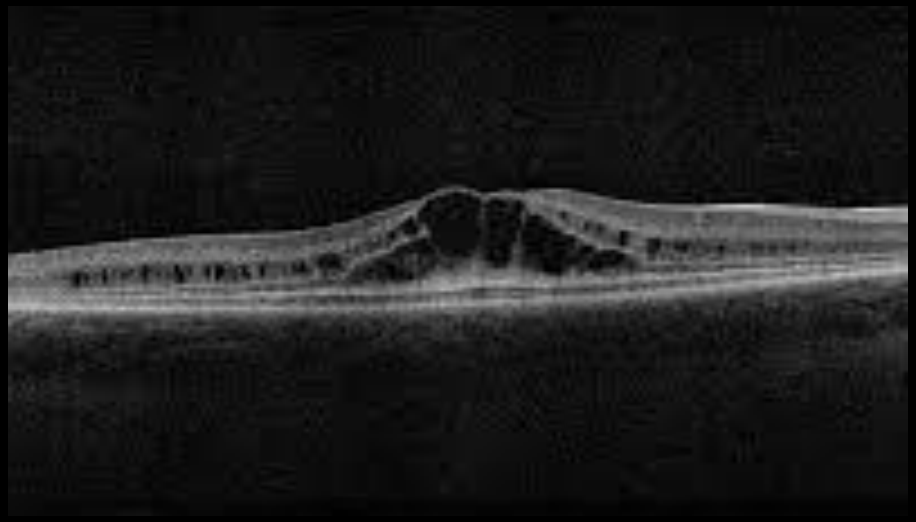
INVESTIGATIONS

FFA	**Early hyperfluorescence (telengictactic vessels) ,blocked Fluorescence due to exudation.** **Areas of capillary non perfusion Arteriolar and Venular anomalies can be seen near areas of capillary non perfusion. Extensive hyperfluorescence due to leakage and staining in late phase.**
OCT	**Prominent Intraretinal hyperreflective material s/o hard exudates** **Cystoid edema** **Useful in monitoring macular involvement**
CT	**Helps in r/o RB (look for presence of calcification)**
MRI	**Superior to CT in ruling out retinoblastoma** **Studies have shown MRI to be superior in differntiating a subretinal exudation from a mass**

FFA - showing hyperflouresence of telengectasia , leakage and staining

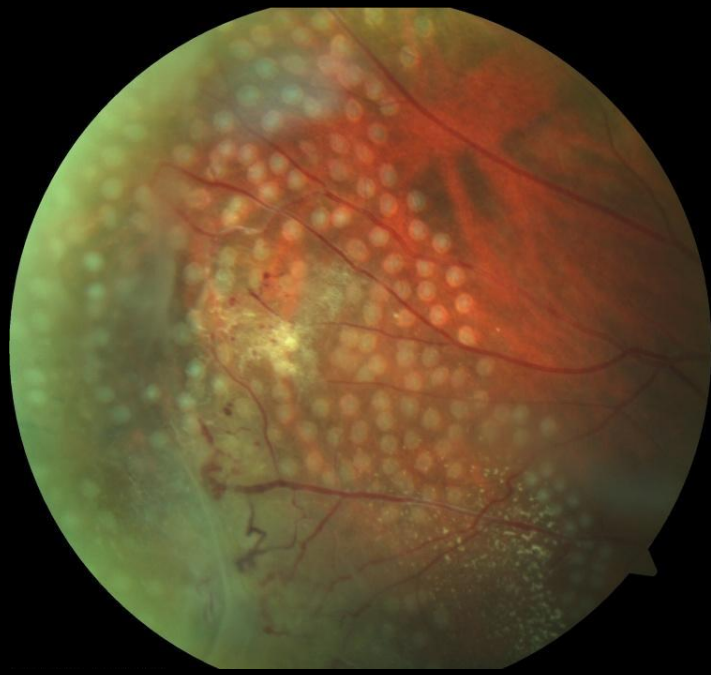


OCT – showing intraretinal exudates and cystoid macular edema



TREATMENT

STAGES	TREATMENT
Stage 1	Documentation(Colour fundus photo /Wide field FFA)observation
Stage 1,2 (mild disease with liitle exudation)	Laser photocoagulation Cryotherapy(double freeze thaw cryotherapy)
Stage 3,4	Vitreoretinal Surgery
Stage 5 With pain No pain	 Ennucleation Observation



INTRAVITREAL INJECTIONS

MODALITY	OUTCOMES
IV STEROIDS (TRIAMCINOLONE /OZURDEX)	IVTA/Ozurdex have been combined with other treatment modalities with good outcomes.
ANTI - VEGF	VEGF levels are higher in coats disease Studies have shown anti–VEGF reduces subretinal exudation and macular exudation when given alone or with laser But ,risk of TRD due to vitreous fibrosis