

Ocular Manifestations of Systemic Diseases

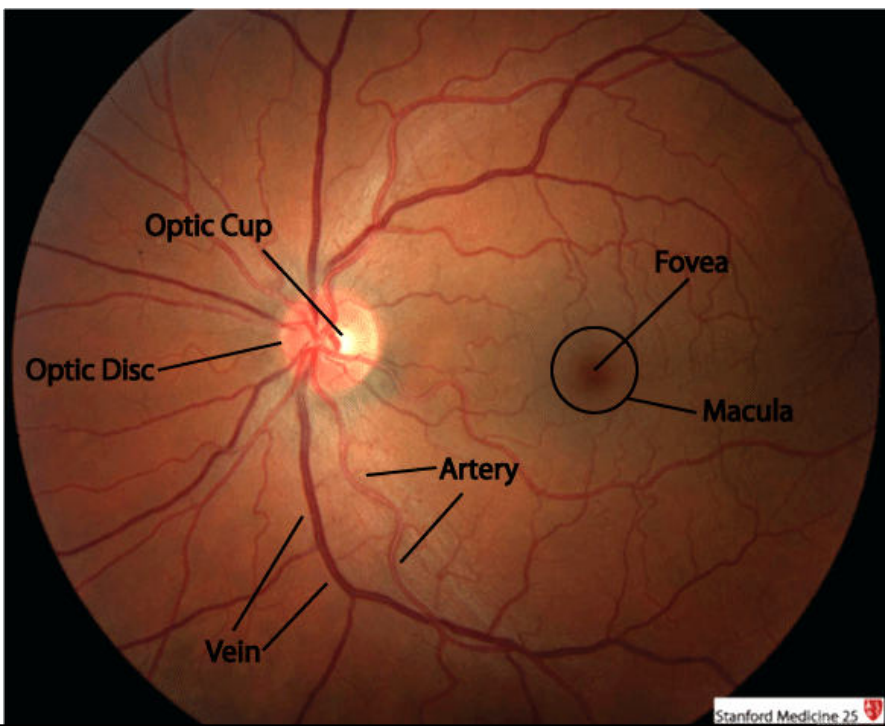
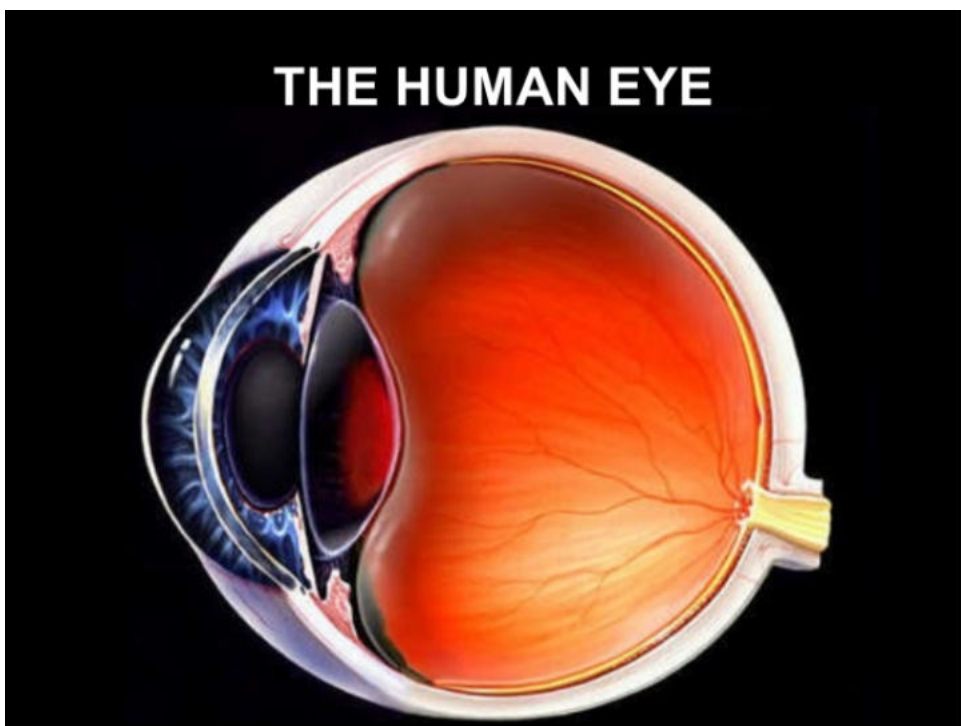
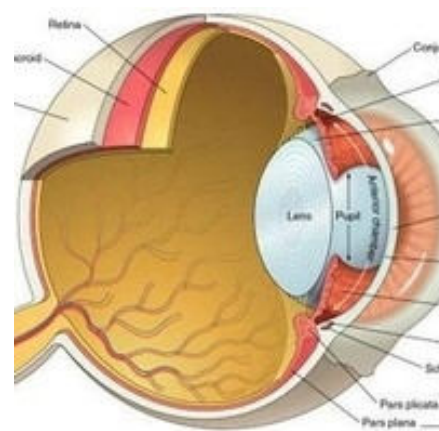
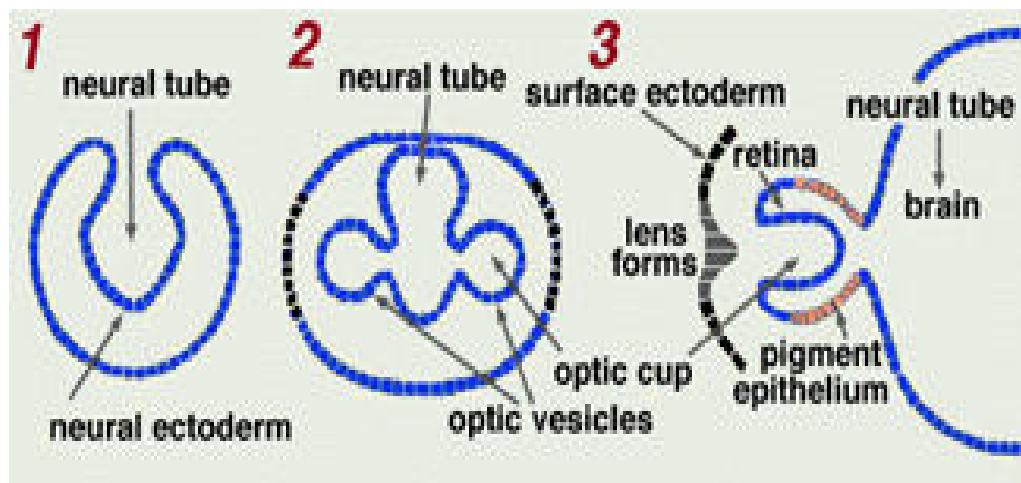
1

Objectives of learning

- Why some systemic disorders can involve eyes?
- What are commoner disorders you can encounter during your clinical postings in other specialities?
- Learn a multi-speciality , judicious cross referral.

The eye is linked with rest of the body by-

- Development
- Blood supply
- Meninges & nerve fibres of brain



Common Systemic Diseases affecting the Eye

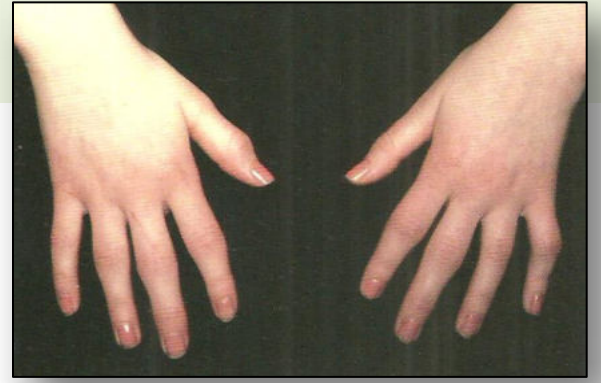
- Autoimmune disorders
- Hematological abnormalities
- Infections
- Endocrine disorders
- Muscular disorders
- Inherited disorders

5

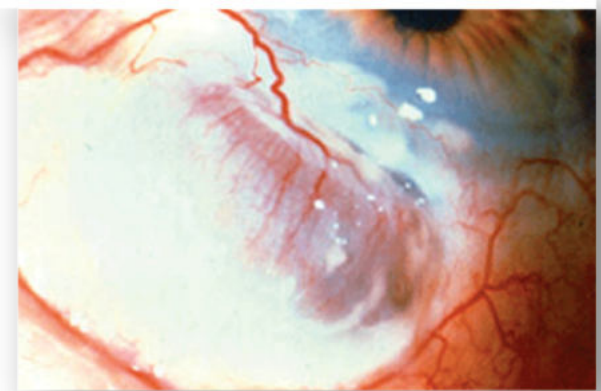
Auto-immune disorders

Rheumatoid Arthritis

- a) Kerato Conjunctivitis Sicca
(Dry eye)- 25% patients
- b) Ulcerative Keratitis
- c) Scleritis



K C S



Scleritis

7

Systemic Lupus Erythematosus

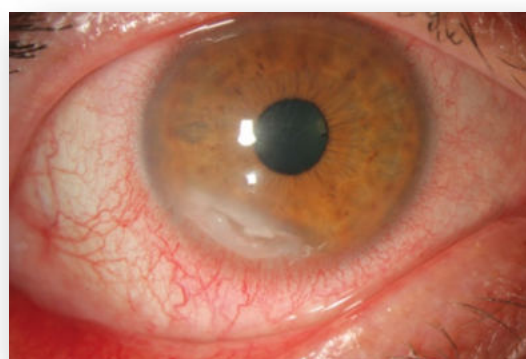
- a) Kerato Conjunctivitis Sicca
(Dry Eye)- 25% patients
- b) Scleritis
- c) Optic Neuropathy



Raynaud's phenomenon



Madarosis



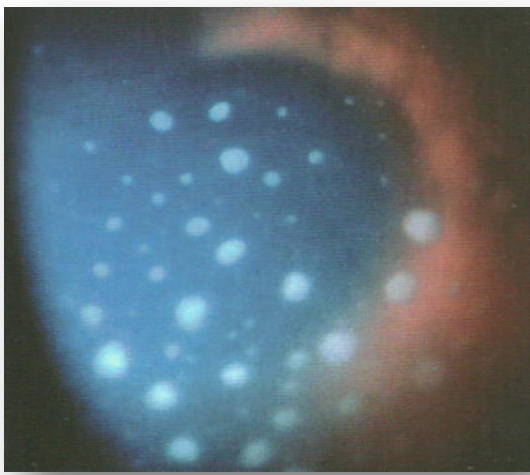
Peripheral ulcerative Keratitis



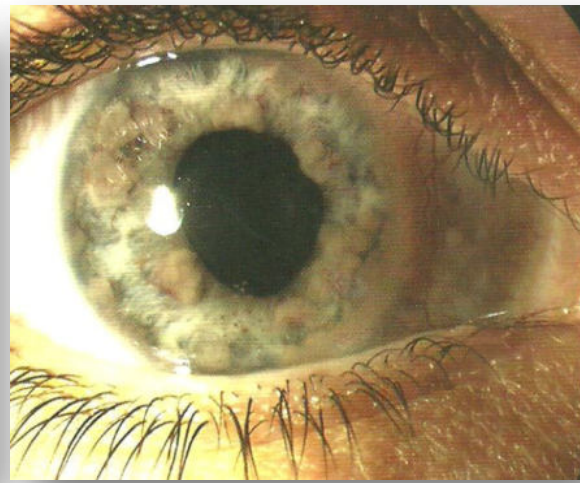
Retinal Vasculitis

SARCOIDOSIS

•25-30% patients have ocular signs



Mutton fat kps



Iris nodule



Phlebitis

Sjogren's Syndrome

Dry Eye
in
30% Patients



Dry fissured tongue

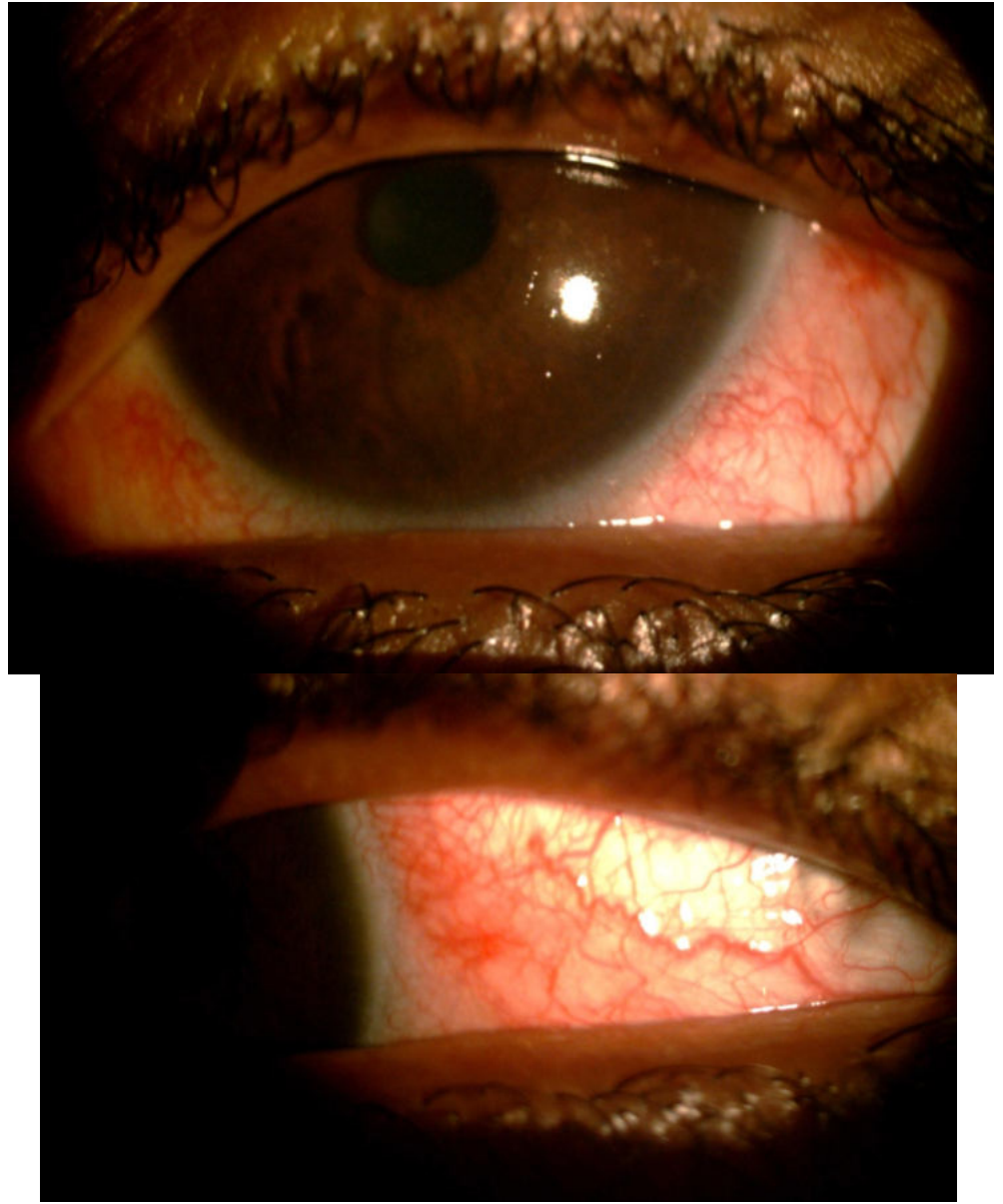


Parotid enlargement



Dental caries

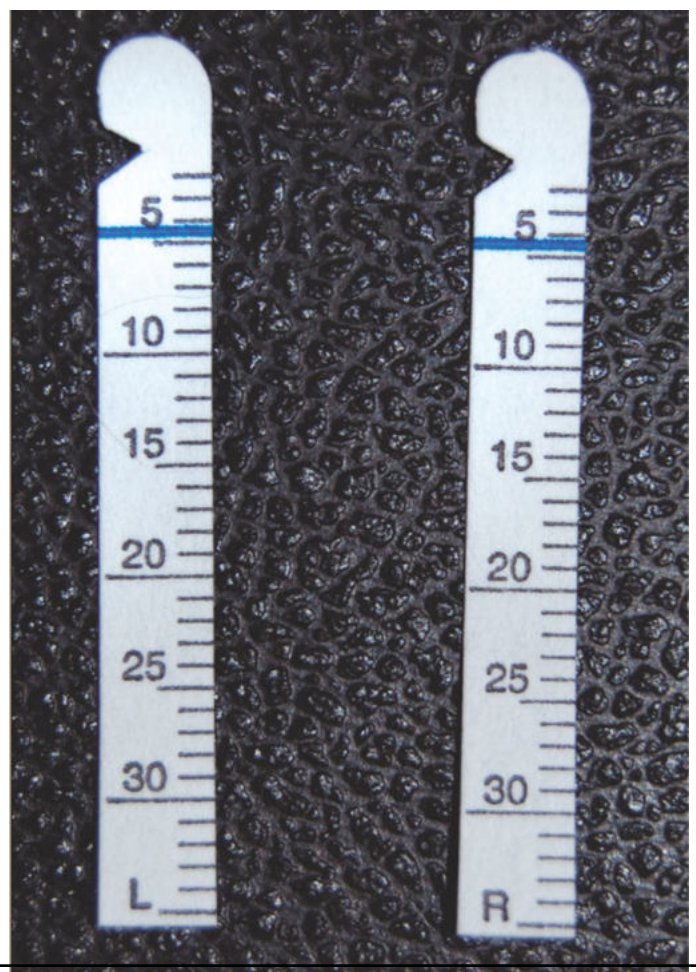
Bulbar conjunctiva
congestion
in Interpalpebral
area



Schirmer test

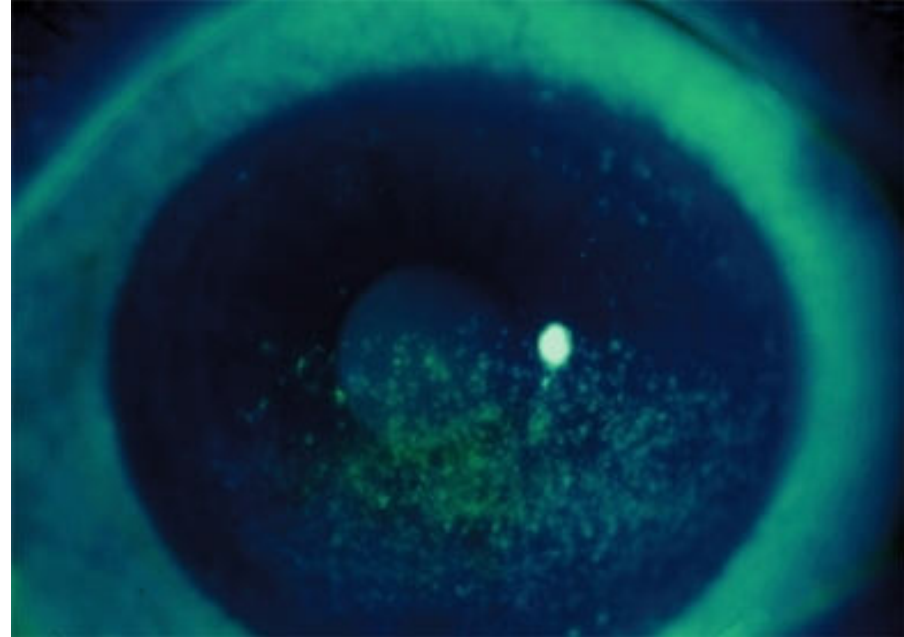
- 5mm end of 5 minutes

SIGNIFY DRY EYE
(less than 10 mm at the
end of 5 min is
suggestive of
abnormality)*



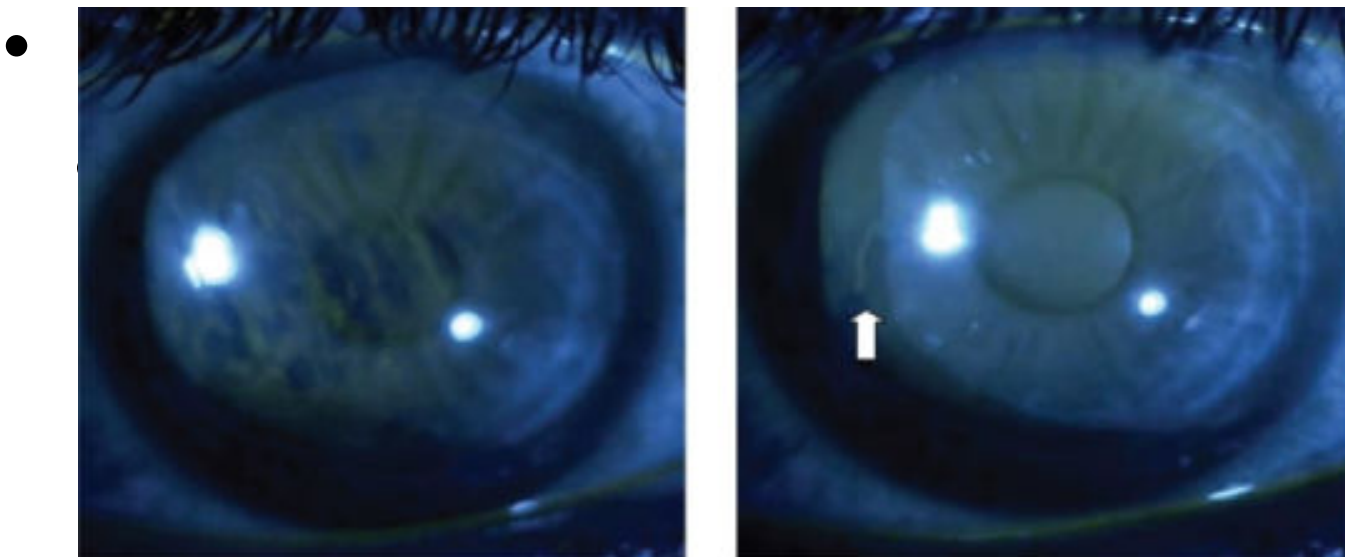
Corneal Staining

- Fluorescein Staining
- Punctate epithelial staining present in lower half of cornea (*interpallebral region*)



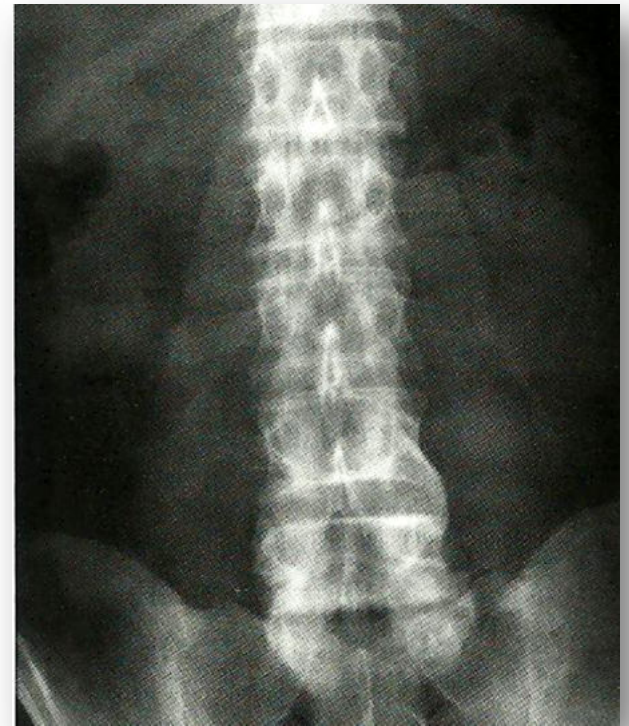
TEAR FILM BREAK UP TIME

- Less than 5 seconds (**SIGNIFY DRY EYES**)



Ankylosing Spondylitis

- Acute anterior uveitis (prevalence of 0.2–1.2%) ,
- *HLA27 is +ve in 75% cases*
- Scleritis

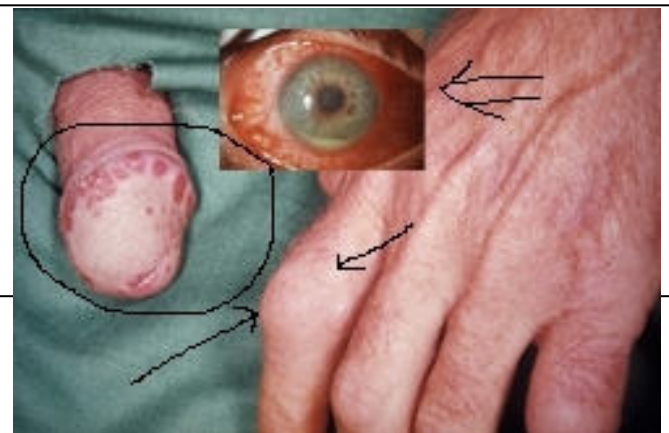


- Bilateral sclerosis
- Erosion of s-i joint
- Bony fusion of spine

15

Reiter Syndrome

Triad → Urethritis
→ Arthritis
↘ Conjunctivitis



Eye Signs:

- Conjunctivitis
- Ac. anterior uveitis
- Numular Keratitis
- Episcleritis
- Scleritis
- Papillitis
- Retinal vasculitis



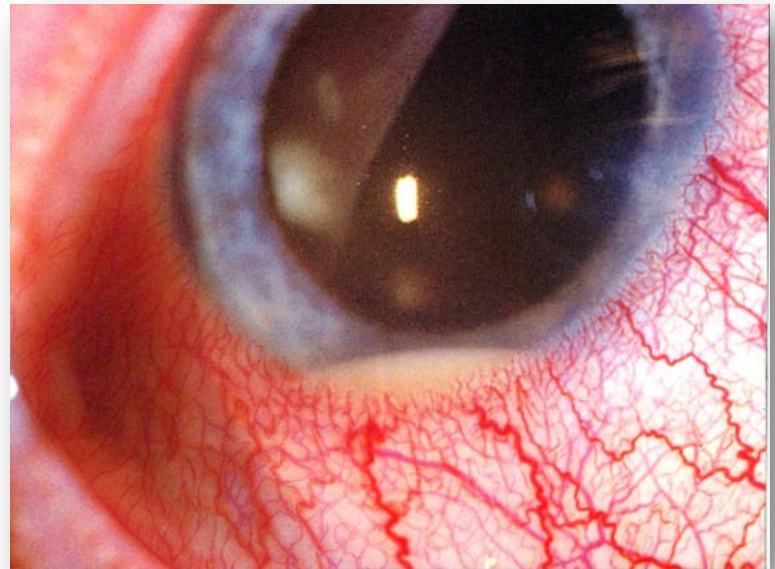
Behcet's Disease

Triad of oral aphthous ulcers, genital ulcers, and uveitis



Eye Signs:

- a) Hypopyon
- b) Uveitis / Iritis



17

Inflammatory Bowel Disease-

Ulcerative Colitis
Crohn's Disease

Ophthalmic manifestations:

- a) Acute anterior uveitis
- b) Episcleritis
- c) Conjunctivitis
- d) Peripheral corneal infiltrates
- e) Scleritis
- f) Papillitis
- g) Retinal Vasculitis



Acute anterior uveitis

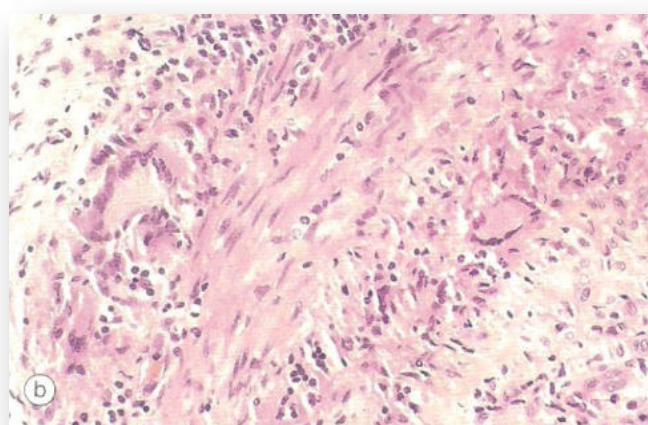


b) Peripheral corneal infiltrates

Giant Cell Arteritis

Predilection for superficial temporal A, ophthalmic A, posterior Ciliary & proximal vertebral A

- a) AION (Arteritic)
- b) TIA , Cilio-retinal Occlusion
- c) Central retinal artery occlusion



Giant cells & small round cells



Superficial temporal arteritis

Vasculitis

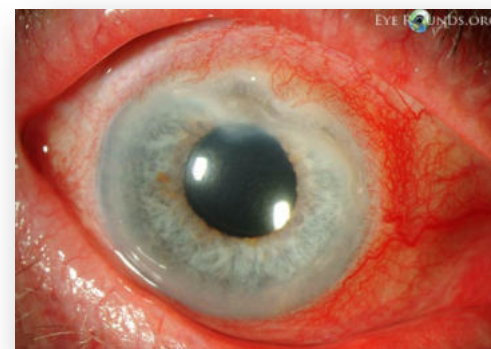
- Wegener granulomatosis
- Relapsing Polyarteritis nodosa
- Polychondritis

Ocular features-

- a) Necrotising scleritis
- b) Peripheral ulcerative Keratitis
- c) Occlusive retinal periarteritis
- d) Peripheral ulcerative Keratitis
- e) Dacryocystitis
- f) Acute anterior uveitis



Dermal infarcts



Peripheral ulcerative Keratitis



Dacryocystitis

Haematological diseases

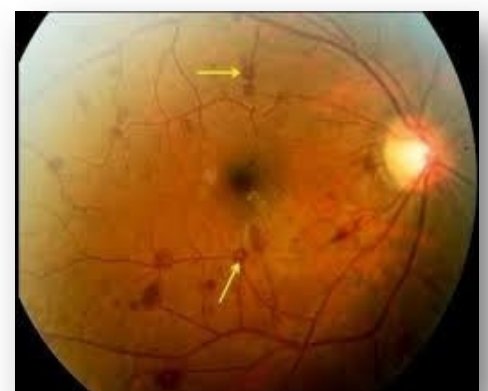
Haematological diseases

- 1) Anaemia
- 2) Leukaemia
- 3) Lymphomas
- 4) Sickle cell Anaemia

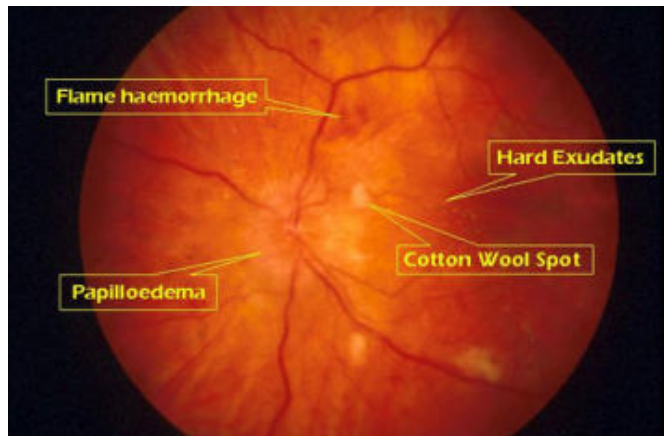
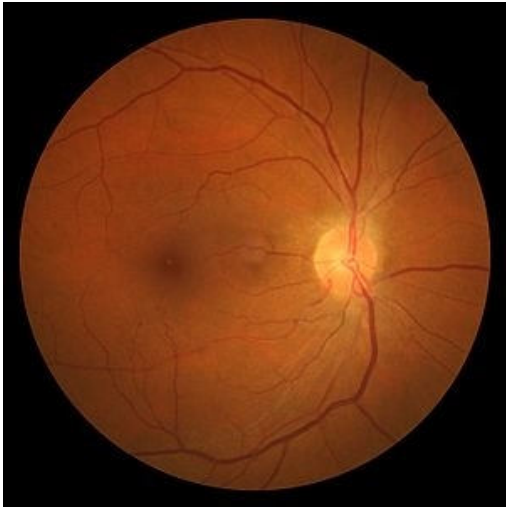
Ocular Presentation:

Haemorrhagic Retinopathy

“ROTHS SPOT”



Hypertensive retinopathy

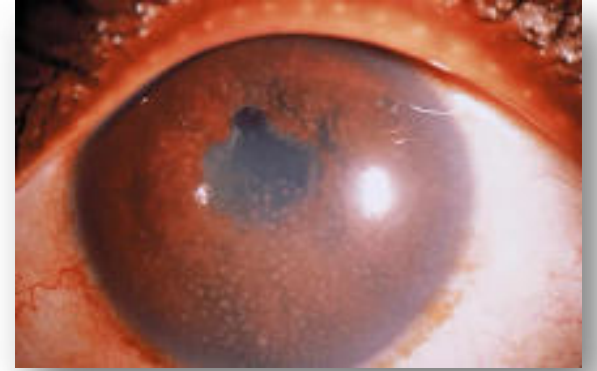


Infectious diseases

1) Tuberculosis

May involve all Structures of Eye except Lens

- a) Uveitis:** chronic granulomatous anterior uveitis, multifocal choroiditis
- b) Eyelids:** lupus vulgaris
- c) Orbit:** dacryoadenitis, dacryocystitis, osteomyelitis
- d) Conjunctiva:** phlyctenular conjunctivitis
- e) Cornea:** phlyctenular keratoconjunctivitis, interstitial keratitis
- f) Sclera:** episcleritis, nodular scleritis
- g) Retina:** exudative retinitis, Vasculitis
- h) Optic nerve:-** papilloedema, ON



2) Leprosy

- a) Facial palsy
- b) Madarosis
- c) Granulomatous iritis
- d) Episleritis/scleritis
- e) Secondary Glaucoma
- f) Cataract

3) Syphilis

Congenital: Acute interstitial keratitis, Diffusely opaque cornea, salt & pepper fundus

Secondary : Iritis, choroiditis, and/or exudates around disc + vessels

Tertiary: Chorioretinitis and/or diffuse neuro-retinitis and vascular sheathing

Viral Infections associated with Ocular Diseases

- Herpes zoster
- Herpes simplex
- CMV
- Rubella- **cataract**
- Measles
- AIDS



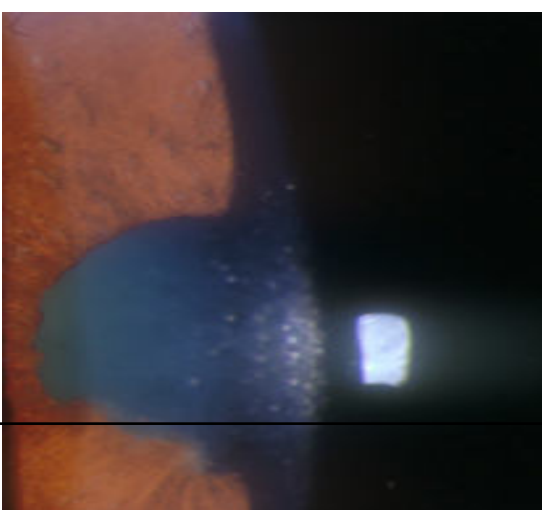
27

AIDS

Kaposi's sarcoma-
most common tumour



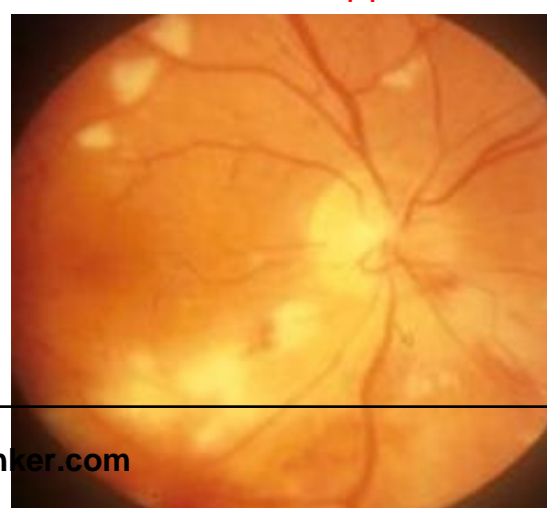
Anterior Uveitis



AIDS retinopathy- 50 to 70%



CMV retinitis-most common
opportunistic infection



Endocrine & Metabolic disorders

Endocrine & Metabolic disorders

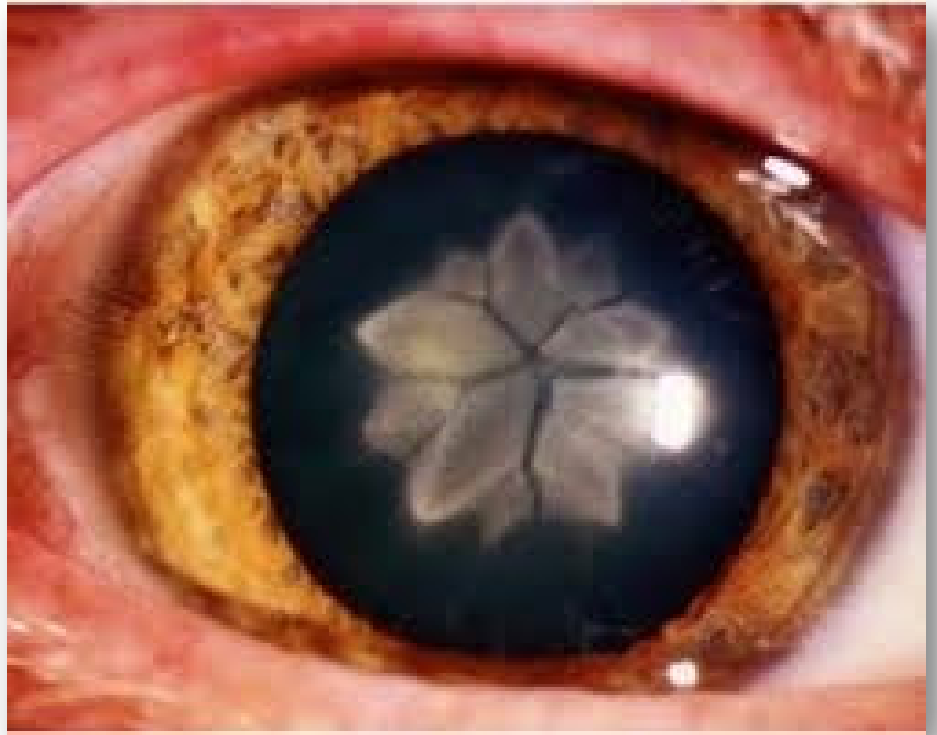
- Diabetes Mellitus
- Muco-polysaccharidoses
- Wilson disease
- Homocystinuria
- Hyperthyroidism
- Hypothyroidism

Metabolic Cataract

Diabetes Mellitus-

- Pre-senile cataract
- Snowflake cataract

[Sorbitol accumulation in lens due to aldose reductase pathway



33

Metabolic Cataract

- **Galactosemic Cataract**

Deficiency of GPUT- Oil droplet cataract

Deficiency of Galactokinase-Lamellar Cataract

Metabolic Cataract

- **Wilson's disease**

Hepatolenticular degeneration

Sun flower Cataract

Kayser-Fleischer Ring

Metabolic Cataract

- **Lowe's syndrome [oculo-cerebral-renal syndrome]**

inborn error of amino acid metabolism

Congenital cataract, microphakia

Posterior lenticonus, glaucoma

Metabolic Cataract

- **Mannosidosis**

Deficiency of alpha-mannosidase → mannose rich oligosaccharides in tissues → spokes like posterior capsular cataract

Metabolic Cataract

- **Fabry's disease**

Deficiency of alpha galactosidase → spoke like cataract & vortex keratopathy

- **Hypocalcemia-**

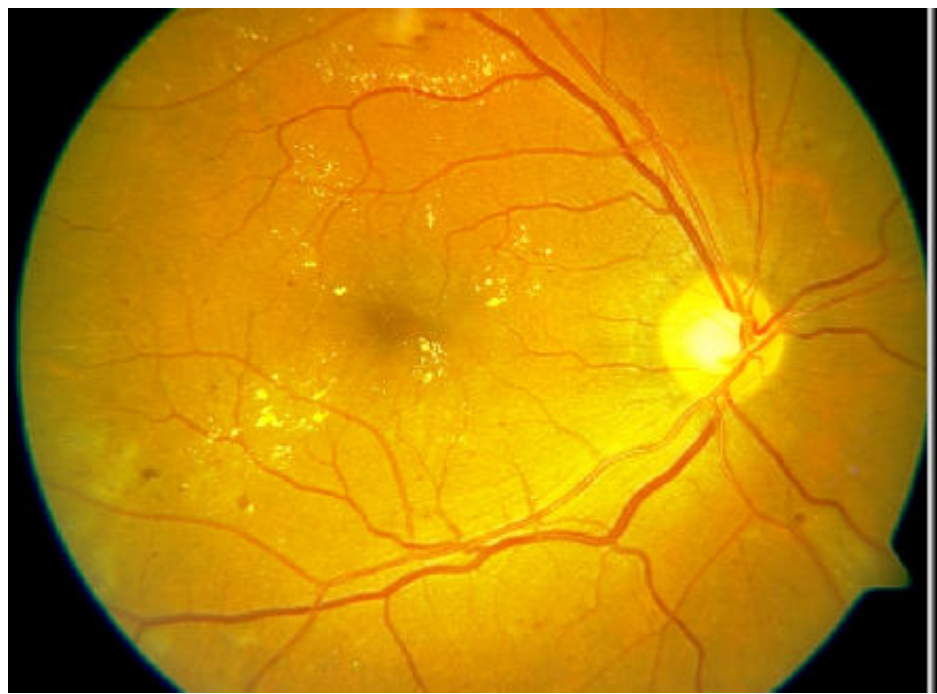
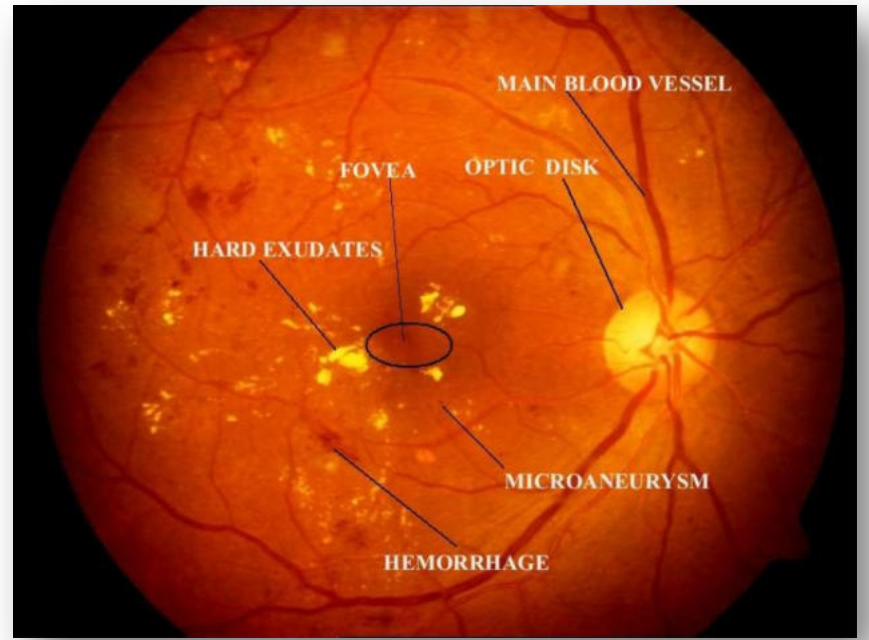
Decreased parathyroid activity leads to cataract

Diabetes Mellitus

- Diabetic Retinopathy
- Diabetic Maculopathy
- Corneal erosions
- Optic nerve Papillopathy
- Transient Myopia

FUNDUS

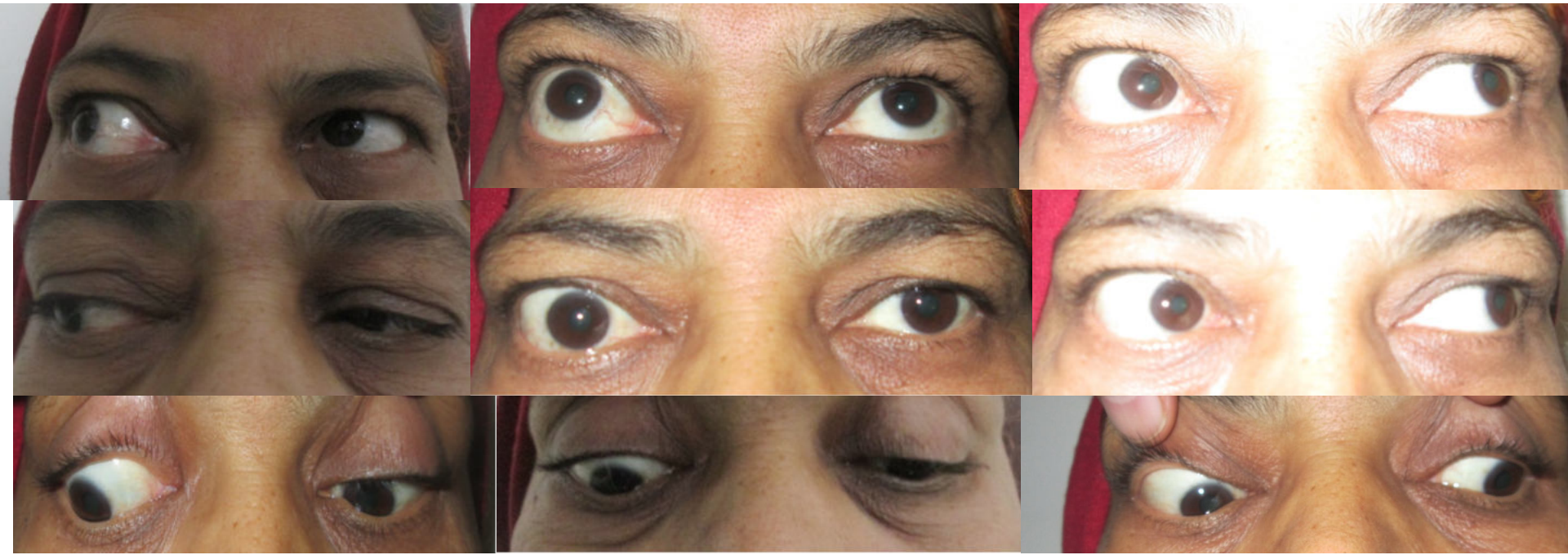
Retinopathy
Maculopathy



THYROID EYE DISEASE

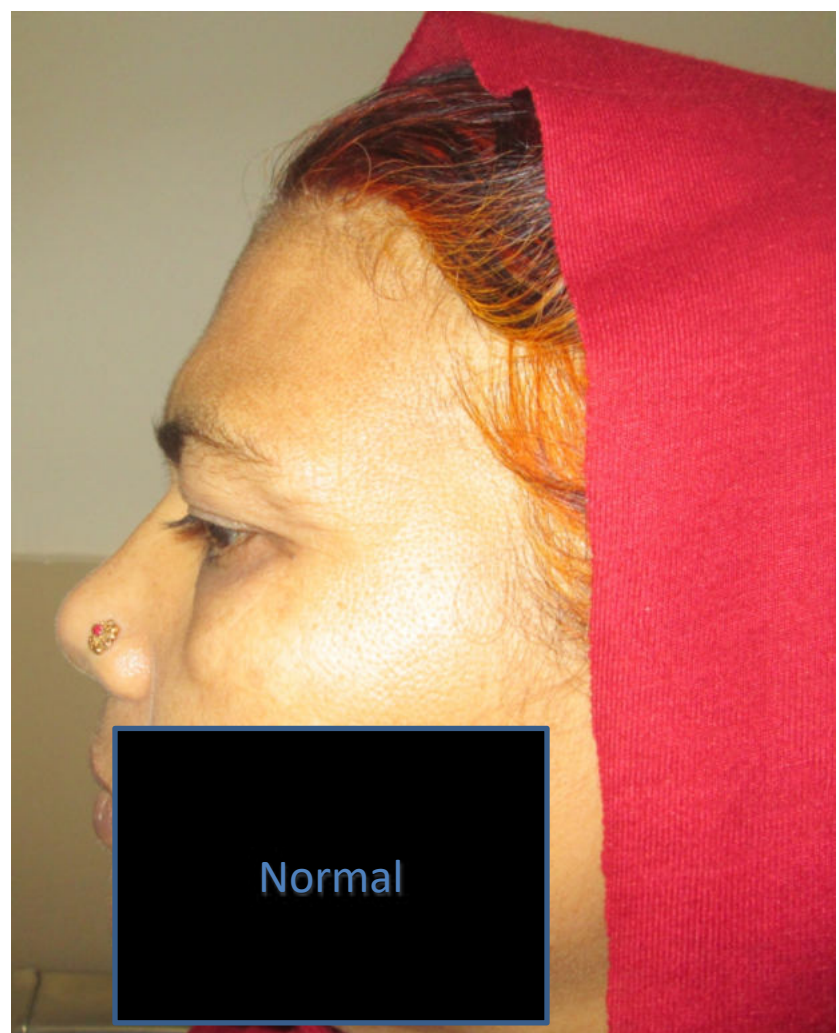


OCULAR MOVEMENTS



Restrictive myopathy

Dalrymple Sign



Naffziger test

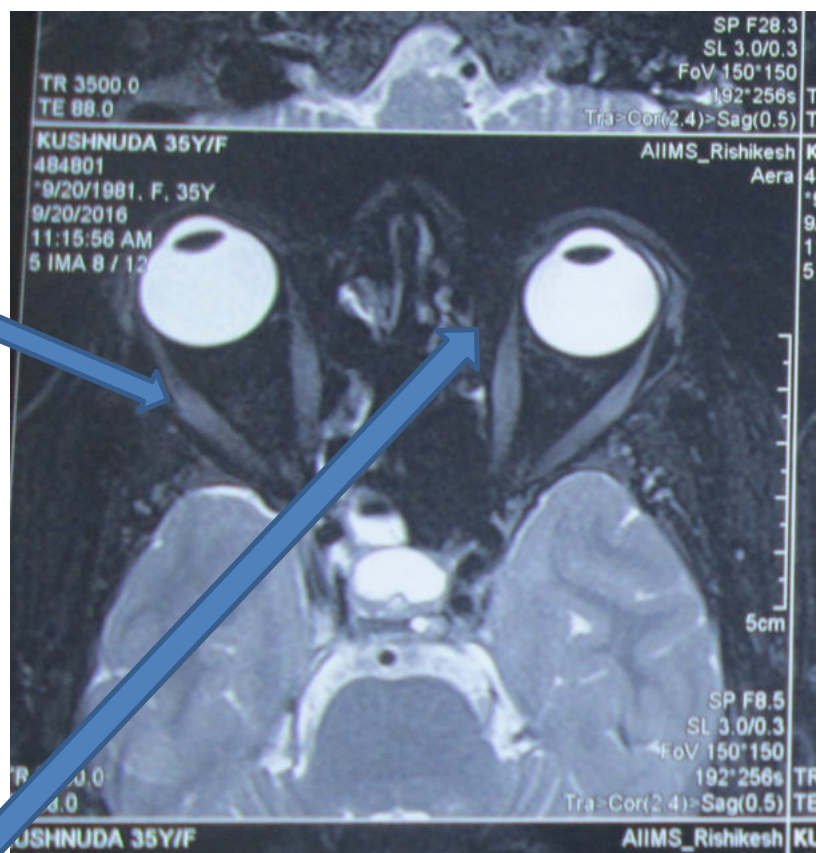


Exophthalmos

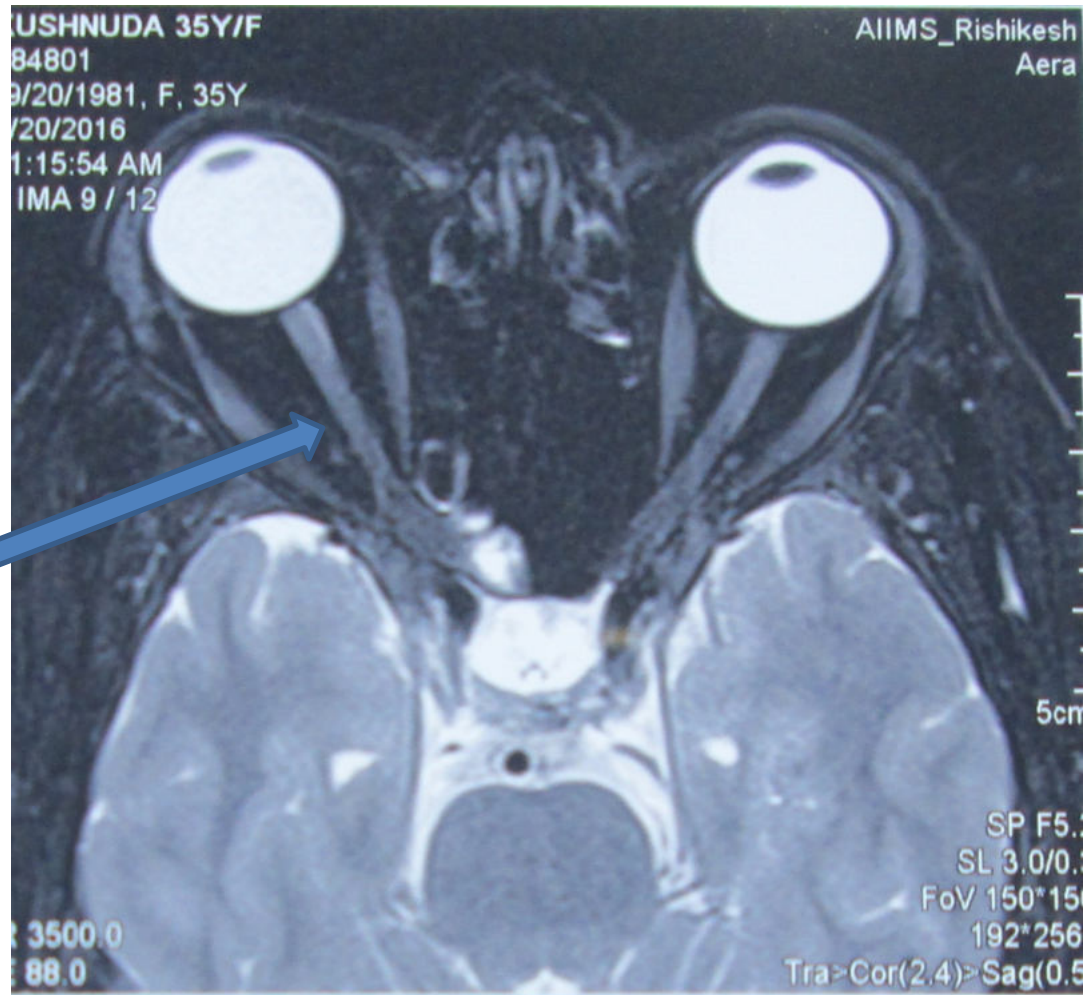


MRI

- Thickening of Muscle belly
- Tendon sparing
- Radiologically both eyes are involved



Slight straightening
of optic nerve in Right
eye



Muscular disorders

Muscular disorders

- Myasthenia gravis
- Muscular dystrophy

Ocular Manifestations:

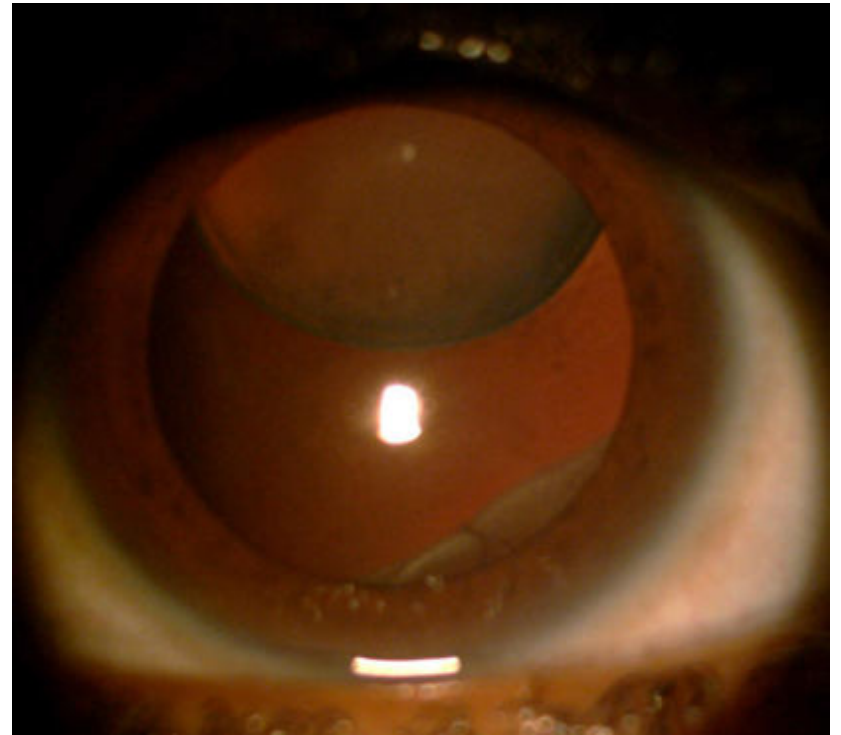
- a) Ptosis,
- b) diplopia,
- c) exophthalmoplegia,
- d) cataract(Christmas-tree cataract)



51

Inherited disorders

- a) Ectopia lentis BE
[superior Lenticular
subluxation]
- b) Microspherophakia
- c) Retinal Detachment
- d) Myopia
- e) Keratoconus

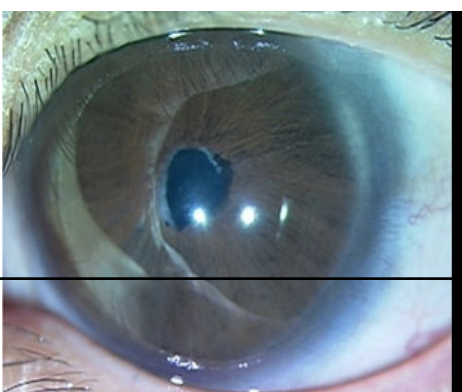


Ectopia lentis

Long spider like fingers



- High arch palate

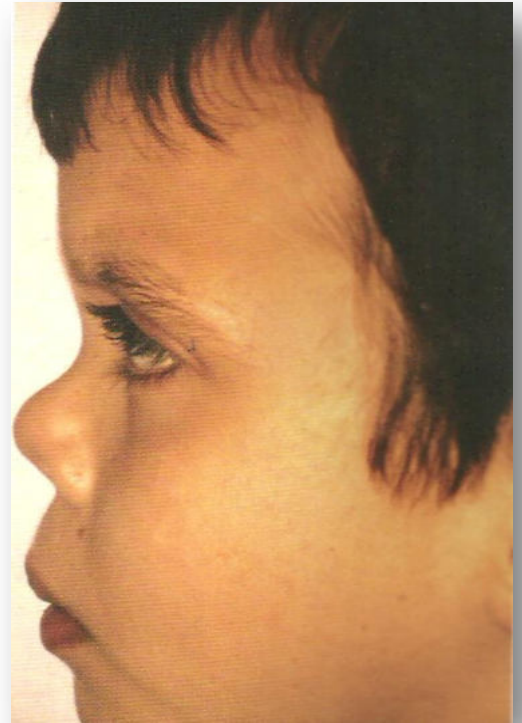


Tall thin stature with
disproportionately long limbs
compared to trunk



Sticklers Syndrome (Hereditary artho-ophthalmopathy)

- Vitreous syneresis
- Retinal Detachment

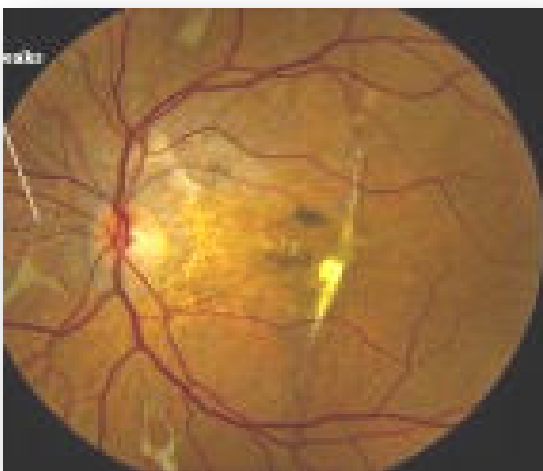


Flat nasal bridge

55

3) Pseudoxanthoma elasticum

Prevalence 1/25000



Angiod streaks



Loose skin folds

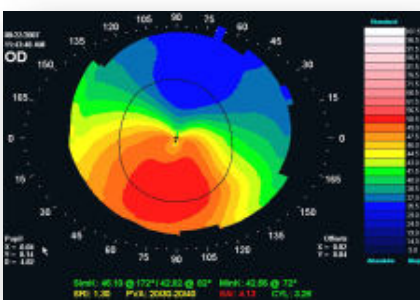
Osteogenesis imperfecta

1/20000 live births

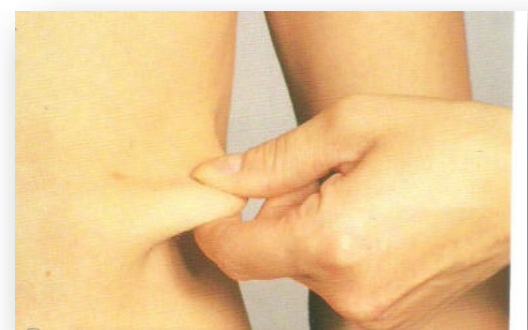
Blue sclera



Ehlers - Danlos Syndrome type 6 (Ocular sclerotic)



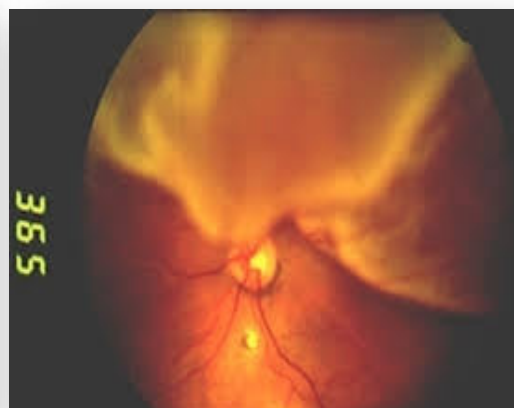
Keratoconus



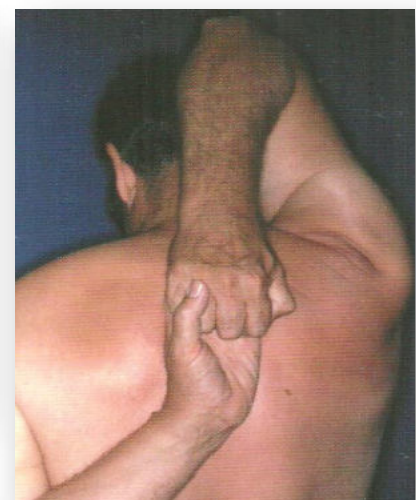
Hyperelasticity of skin



High myopia



RD



Hypermobility of joints

PHACOMATOSES - Neurofibromatosis-Type I (Von Recklinghausen disease)

- Most common phacomatosis
- Affects 1:4000 individuals
- Presents in childhood
- Gene localized to chromosome 17q11

Nodular



**Facial hemiatrophy-
Skeletal defects**



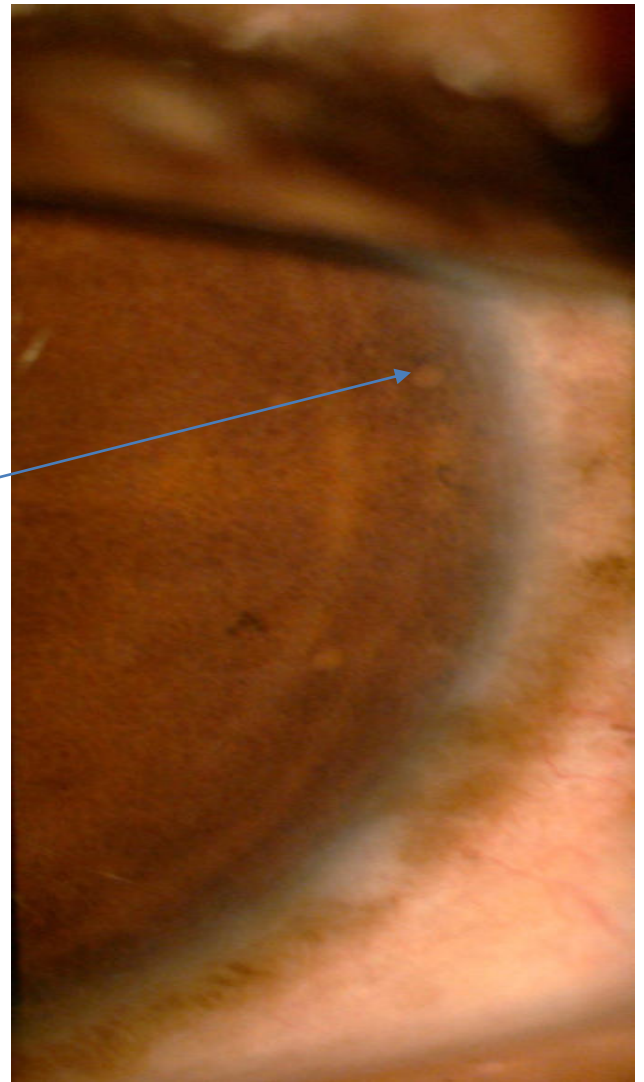
Café-au-lait spots



- C/O mass over right eyelid since birth
- Growing with age
- Painless

Iris-Lisch Nodules

- **LISCH NODULES/hamartoma**
- **Café au lait spot and axillary freckling were also present**

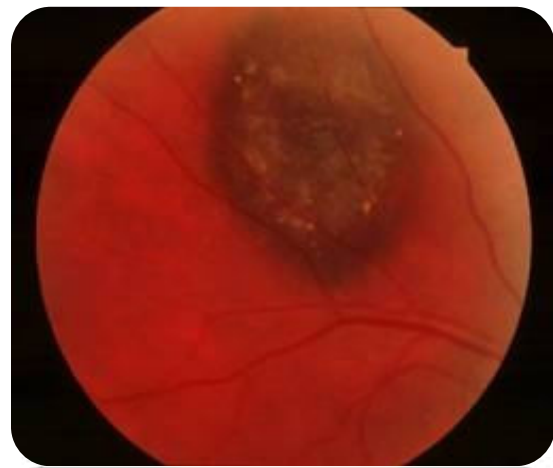


Intraocular lesions in NF-1

Lisch nodules



Choroidal naevi



Family history



Father has multiples neurofibromas all over chest and arm (1st degree relative)

Orbital lesions in NF-1

•**Optic nerve glioma(in 15%) invading hypothalamus**

•**Spheno-orbital encephalocele showing congenital absence of left greater wing of sphenoid bone**

• **Pulsating proptosis without bruit!**



Tuberous sclerosis (Bourneville disease)

- Autosomal dominant
- Triad - *mental handicap, epilepsy, adenoma sebaceum*

Adenoma sebaceum



Shagreen patches



Ash leaf spots



Retinal astrocytomas

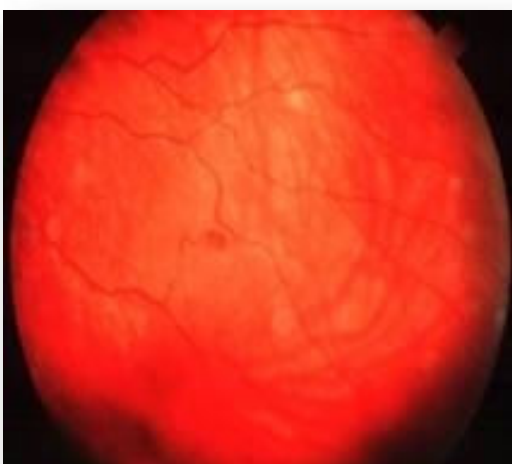


Innocuous tumour present in 50% of patients

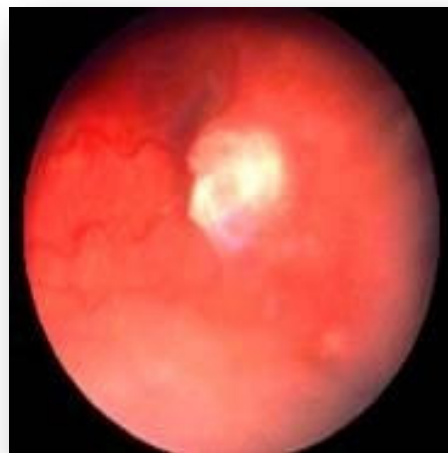
Von-Hippel-Lindau syndrome

- Tumours— renal carcinoma and pheochromocytoma
- Cysts- kidneys, liver, pancreas, epididymis, ovary and lungs
- Polycythaemia
- Retinal capillary haemangioma

Tiny lesion between arteriole and venule



Round orange-red mass



Associated dilatation and tortuosity of feeder vessels



Sturge-Weber syndrome

Involves face, leptomeninges and the eyes

- Arteriovenous malformations of episclera
- Choroidal haemangioma
- Glaucoma (ipsilateral)
- Iris heterochromia

Albinism

Hypopigmented skin and hair with eyes involvement

- Nystagmus
- Translucent iris giving rise to a “pink-eyed” appearance.
- Large choroidal vessels are seen due to lack of pigment in fundus.
- Foveal hypoplasia

Malignancy

Malignancy

- Metastasis
 - ✓ From Breast, lung most common.
 - ✓ Usually localize to choroid but Extra Ocular Muscles,
 - ✓ optic nerve and orbit can also be affected
- Lymphoma, leukemia

Summary

- Ocular examination plays a significant role in diagnosing several systemic illnesses.
- At times, patient present for eye related problem, which then comes out to be a part of systemic disease, requiring inter-departmental reference of the case to Physician/ Paediatrician/Neurologist/ Neurosurgeon/ Orthopaedician/Obstetrician.

Acknowledgement

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

1] Snow-flake cataract is seen in

- a) Diabetes
- b) Galactosemia
- c) Trauma
- d) Rubella

2] Cataract associated with atopic dermatitis

- a) Sunflower cataract
- b) Coronary cataract
- c) Shields cataract
- d) Coralliform cataract

3] Sunflower type cataract is seen in

- (a) Diabetes
- (b) Chalcosis
- (c) Argyrosis
- (d) Tay sach's disease

4. Most common ocular manifestation of AIDS is:

- (a) Cottonwood spots
- (b) Cytomegalovirus retinitis
- (c) Kaposi's sarcoma of conjunctiva
- (d) Pseudotumors

5. Which prominent ocular manifestation is associated with

Marfan's syndrome? :

- (a) Microcornea
- (b) Microspherophakia
- (c) Megalocornea
- (d) Ectopia lentis