

# Ocular Manifestations of Systemic Diseases

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# Systemic Diseases

- Endocrine
- Neurology
- Cardiovascular
- Rheumatology
- Dermatology
- Genitourinary
- Gastrointestinal
- Respiratory
- Nephrology
- Treatment of systemic disease

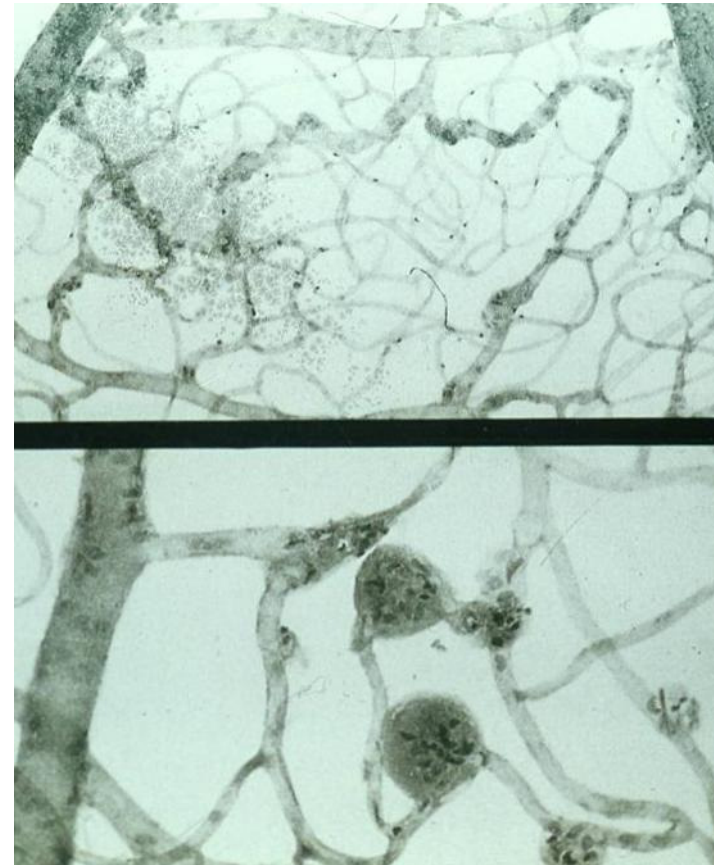
MANY SYSTEMIC DISEASES CAN HAVE AN OCULAR COMPLICATION OR ASSOCIATION

# Endocrine - Diabetes Mellitus

- Major cause of blind in people < 65 years in the western world

# Non-proliferative Diabetic Retinopathy

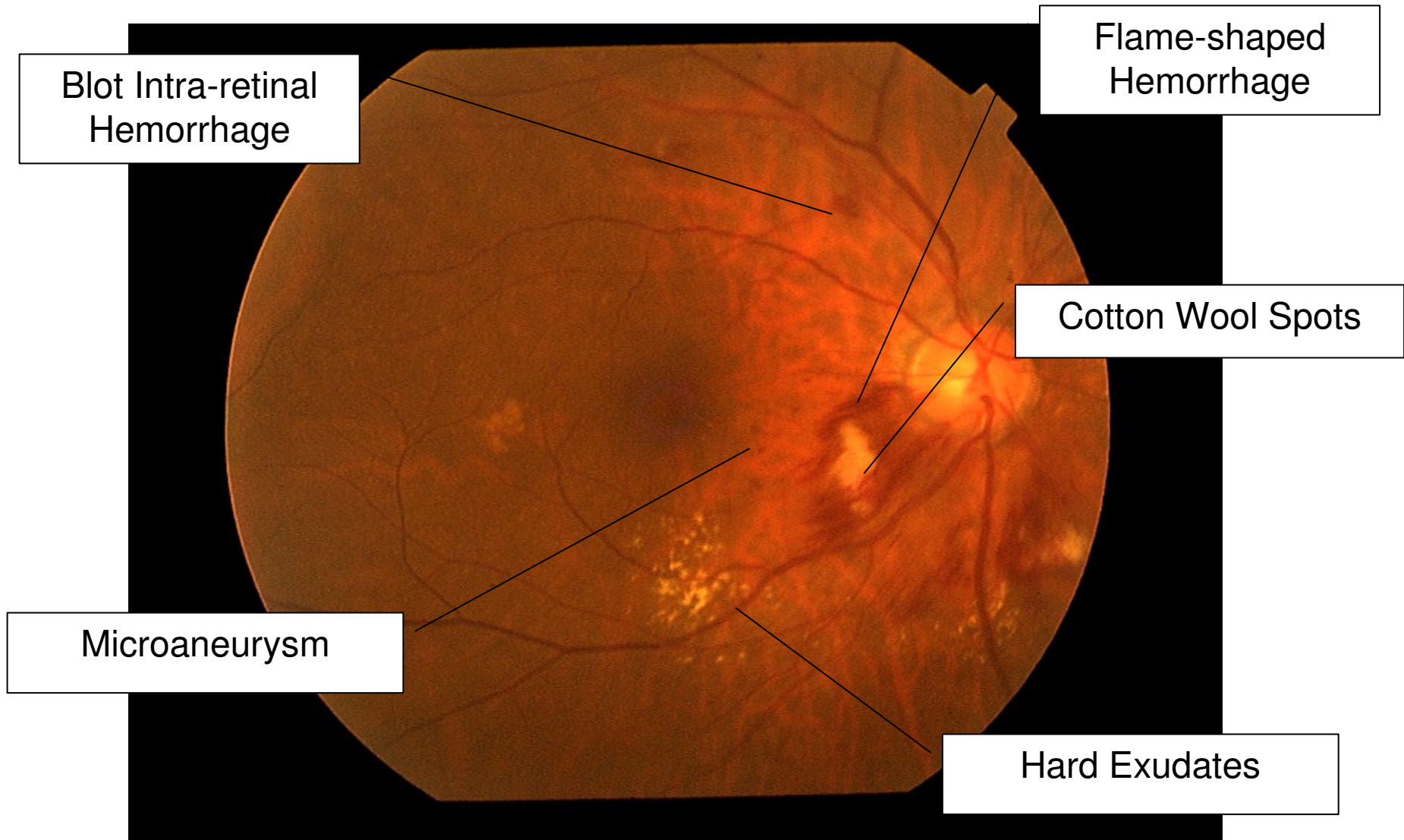
- Capillary basement membrane thickening
- Loss of pericytes
- Distention of retinal capillary walls
  - microaneurysm formation
- Breakdown of blood-retinal (endothelial) barrier
  - Leakage of plasma constituents into retina
    - Clinically significant macular edema (CSME)
  - Lipid exudation



# Diabetic Retinopathy

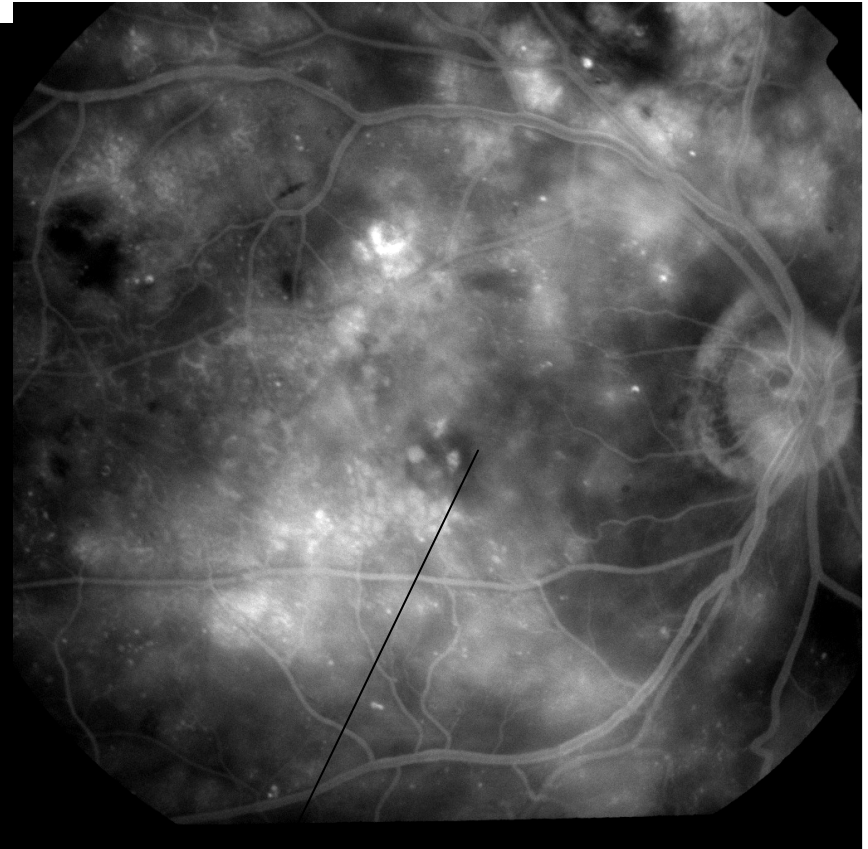
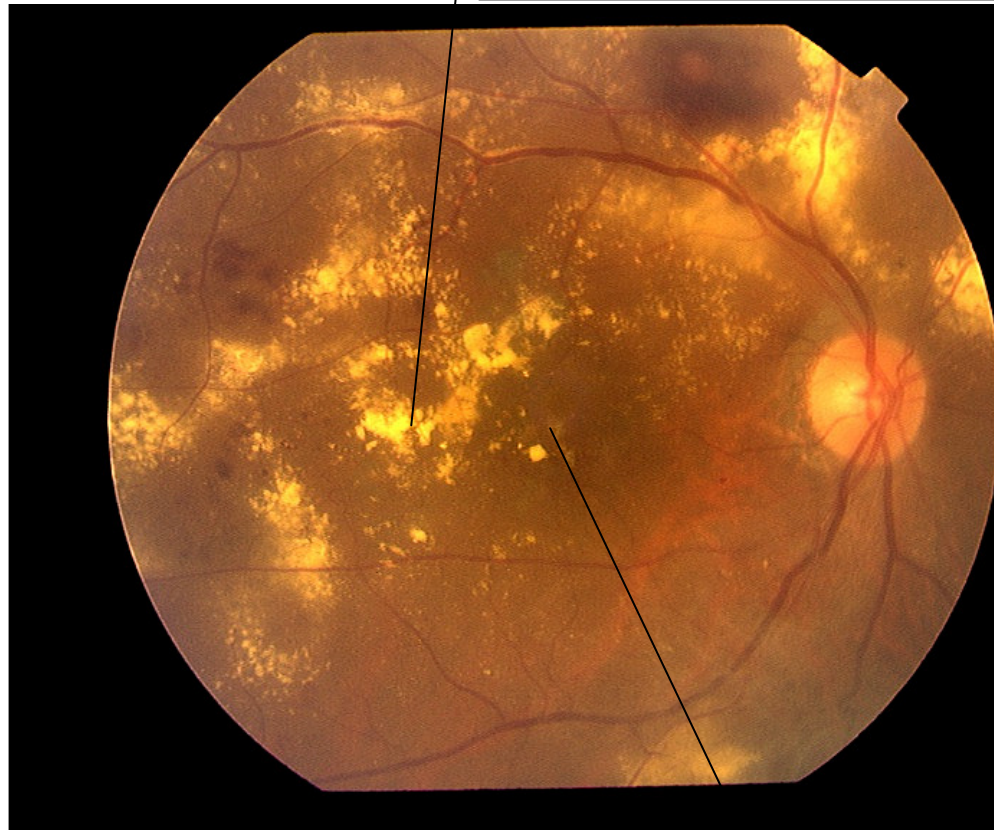
- Intraretinal hemorrhages
  - Usually visually insignificant
- Hard exudates
  - Lipoproteins following partial / complete resorption of retinal edema
- Cotton wool spots
  - Focal areas of ischemic infarcts
- Venous beading
  - Increasing ischemia
- Intraretinal microvascular abnormalities (IRMA)
  - Pre-neovascularization retinal vascular abnormalities

# Non-proliferative Diabetic Retinopathy



# Non-proliferative Diabetic Retinopathy

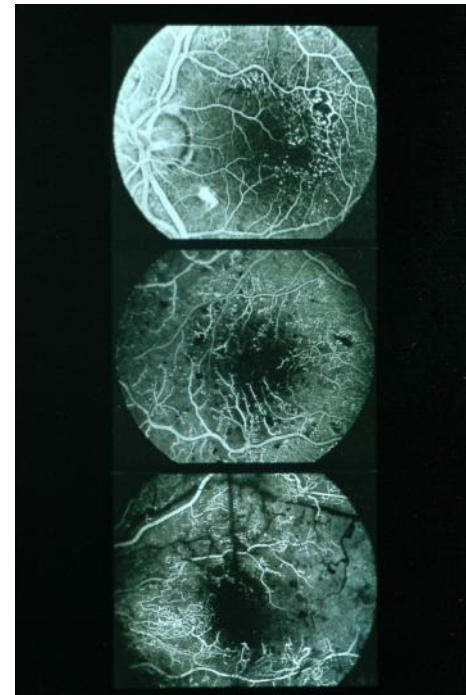
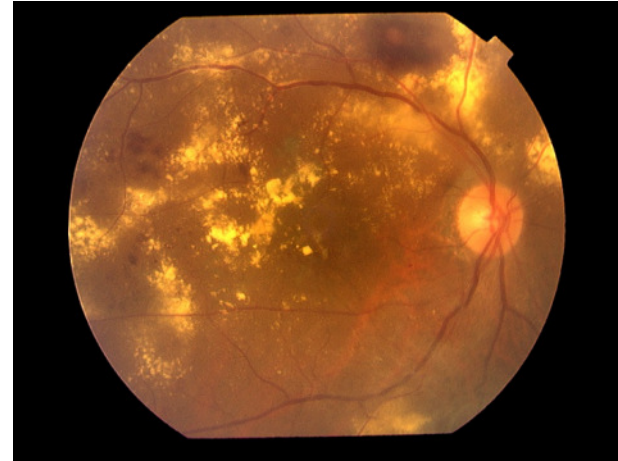
Hard Exudates



Macular Edema

# Non-proliferative Diabetic Retinopathy

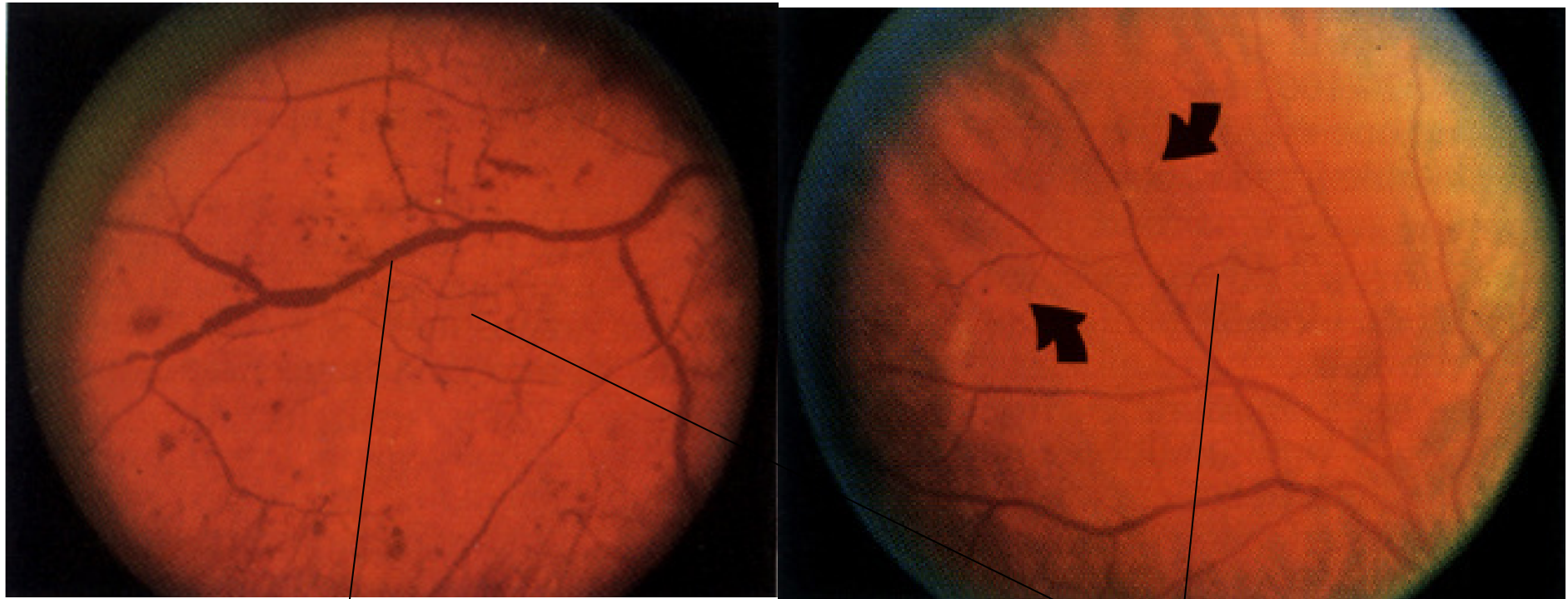
- Visual Loss – Diabetic Maculopathy
  - Macular edema
  - Macular ischemia



# Diabetic Retinopathy

- Microvascular occlusion
- Retinal capillary non-perfusion
- Retinal ischemia
- Arteriovenous shunts
  - Significant capillary occlusion (“dropout”)
  - Intraretinal microvascular abnormalities (IRMA)
- Pre-retinal neovascularization

# Diabetic Retinopathy

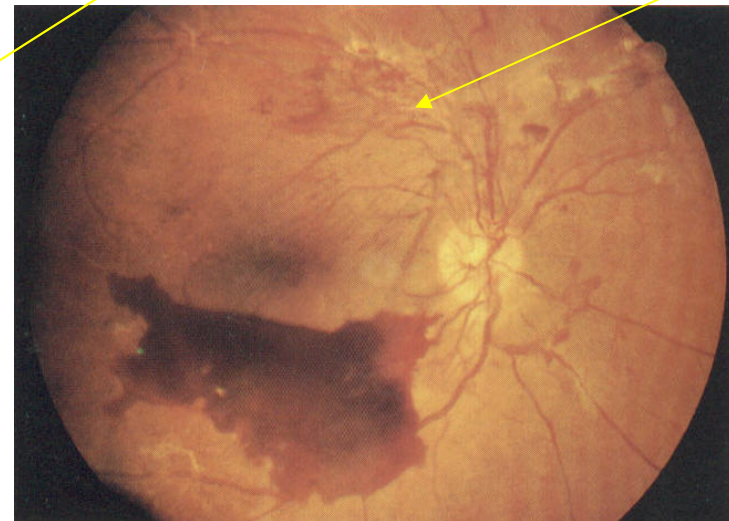
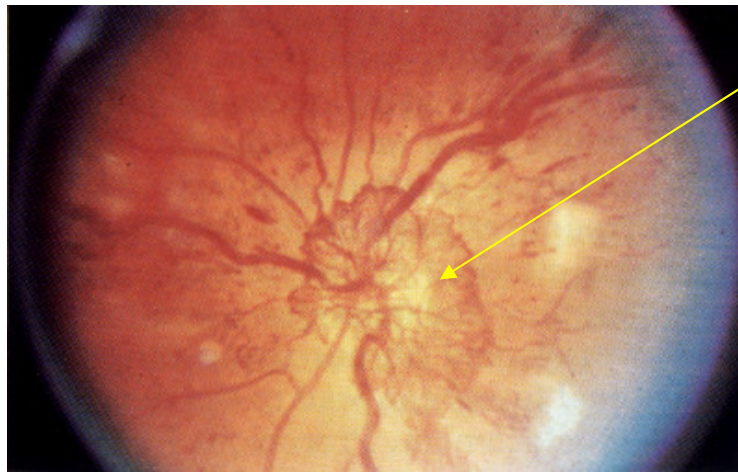


Venous Beading

“IRMA”

# Proliferative Diabetic Retinopathy

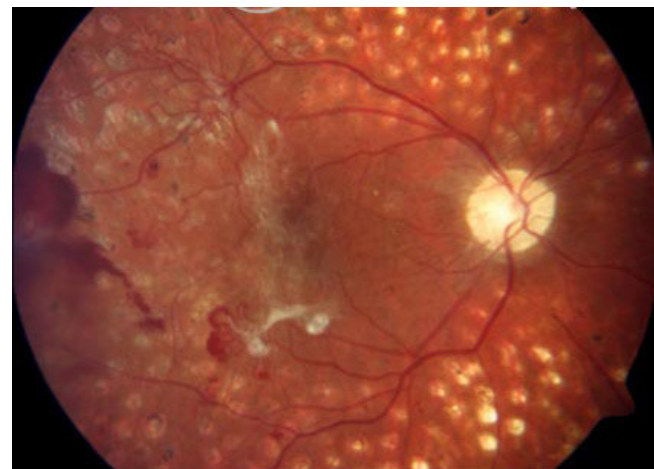
- Preretinal Neovascularization (NV)
  - Retinal ischemia
  - Elaboration of vasoproliferative substances (VEGF)
  - NV develops from venous end of retinal vasculature
  - NV grows on vitreous
  - Neovascularization of the disc (NVD) and elsewhere (NVE)



# Proliferative Diabetic Retinopathy

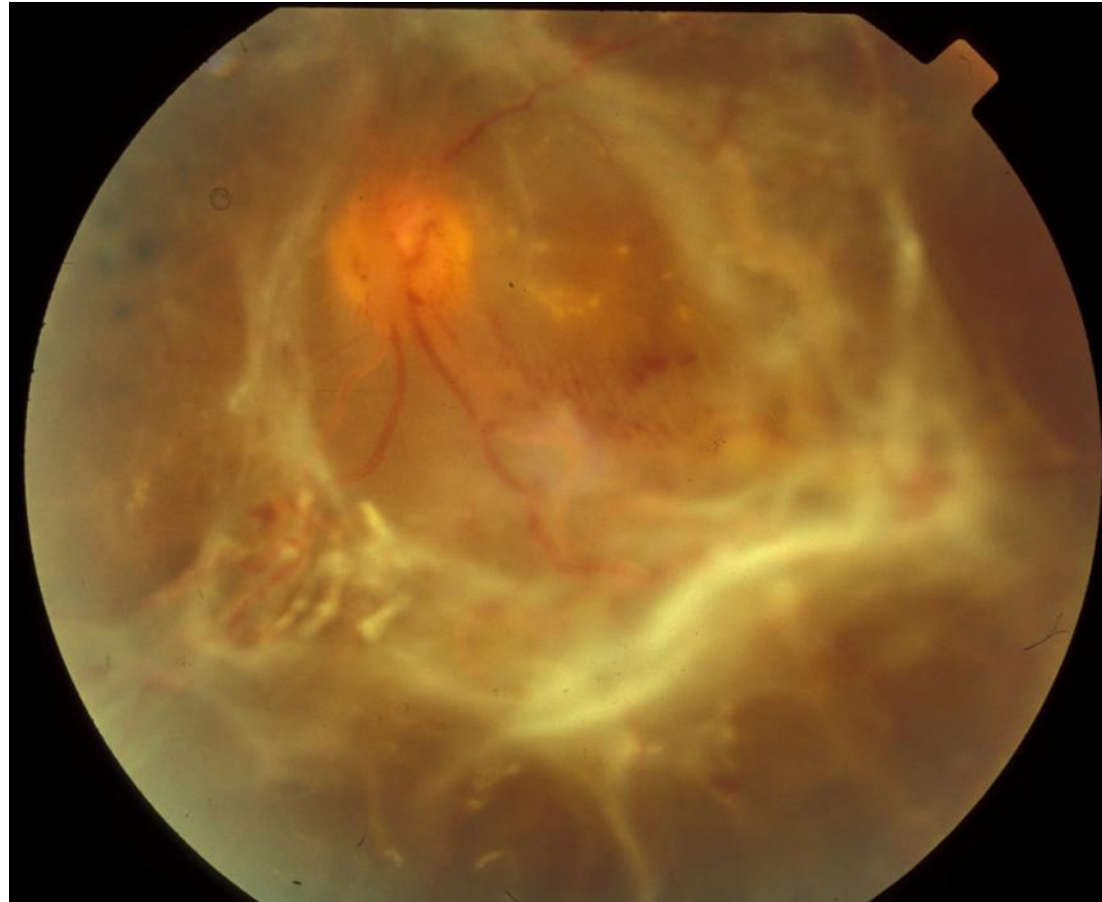
- Complications of NVD / NVE
  - Pre-retinal or vitreous hemorrhage
  - Tractional retinal detachment  $\pm$  rhegmatogenous retinal detachment
  - Neovascular glaucoma
  - Blindness and phthisis bulbi

# Vitreous Hemorrhage



Proliferative Diabetic Retinopathy

# Tractional Retinal Detachment



Proliferative Diabetic Retinopathy

# Diabetic Retinopathy

Retinal Microangiopathy

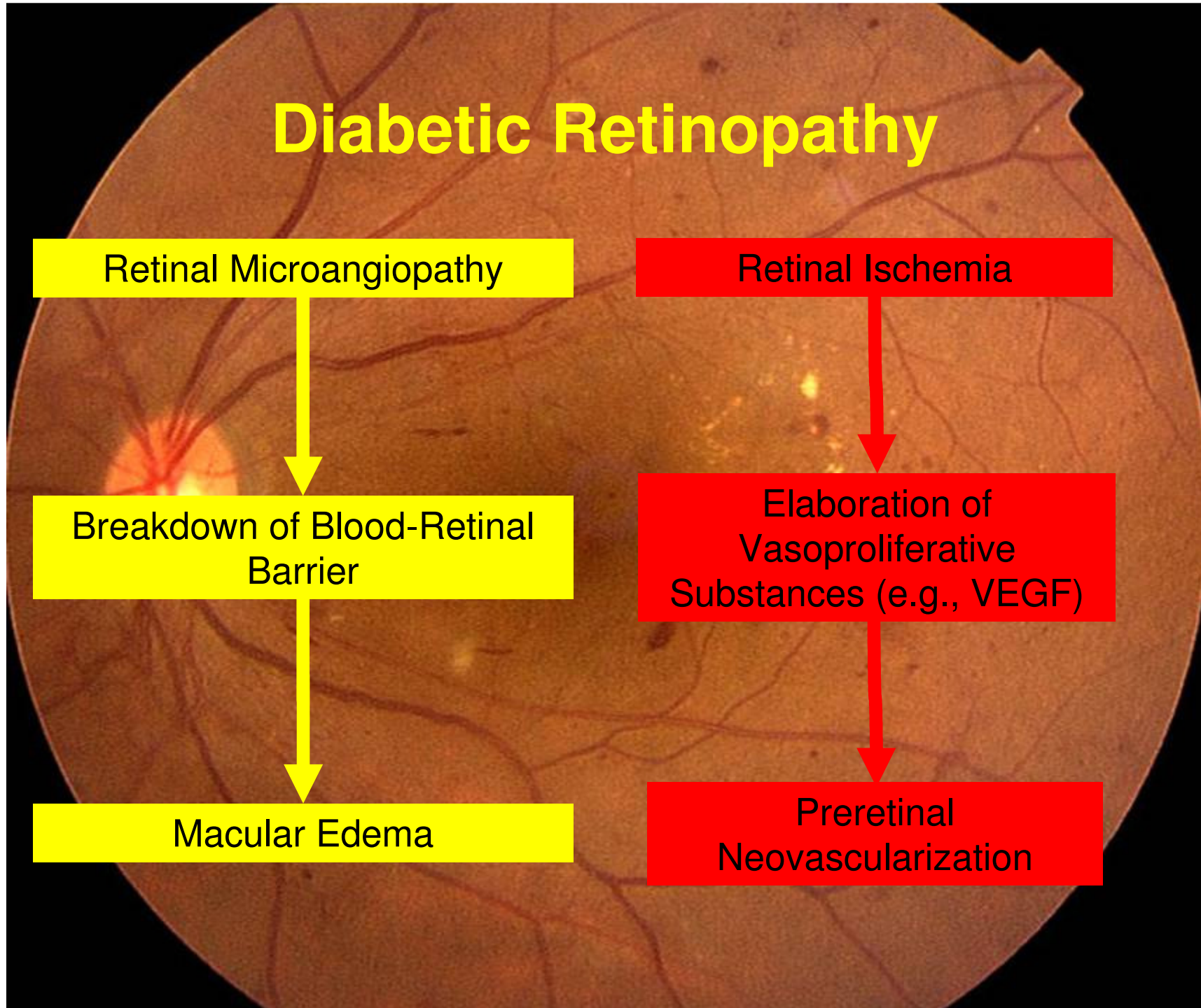
Retinal Ischemia

Breakdown of Blood-Retinal Barrier

Elaboration of Vasoproliferative Substances (e.g., VEGF)

Macular Edema

Preretinal Neovascularization



# Endocrine - Thyroid

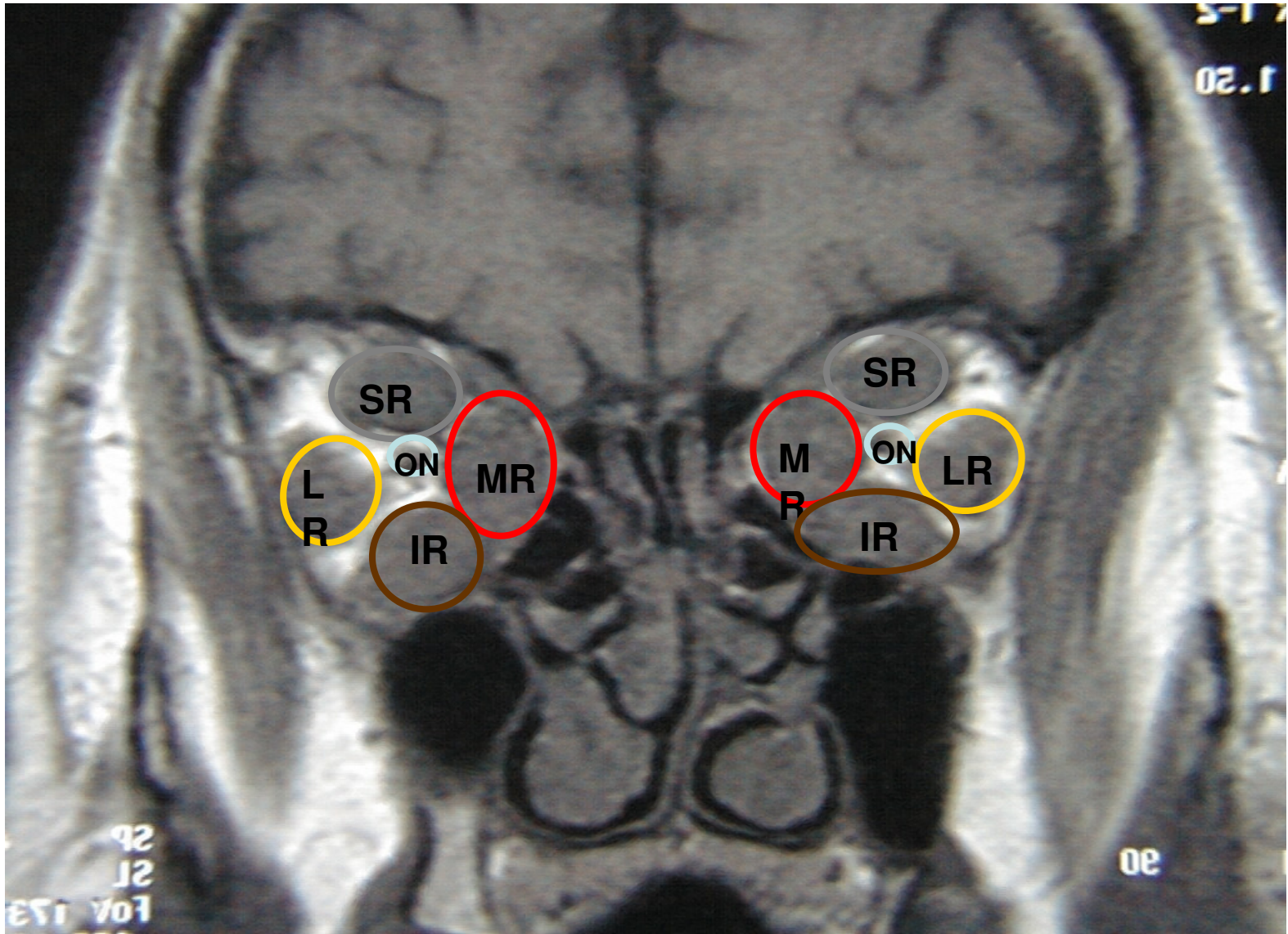
- Eye and orbital changes usually in association with hyperthyroidism (Graves' disease)
- Patient may be clinically and biochemically euthyroid - ophthalmic Graves' disease
- Immunologically mediated disease

# Endocrine - Thyroid

- Exophthalmos (proptosis)
- Grossly swollen extraocular muscles - esp. medial and inferior recti
- Lid retraction and lid lag
- Major complications include:
  - Optic nerve compression
  - Corneal perforation due to exposure keratopathy

# Thyroid-Associated Ophthalmopathy





# Neurology

- Ophthalmology comprises the eye AND visual system
- Any lesion that interferes with the visual pathway
- III, IV, VI cranial nerve palsies
- Horner's syndrome
- Demyelinating disease
- Myasthenia gravis
- Myotonic dystrophy

# CN III Palsy



# CN VI Palsy

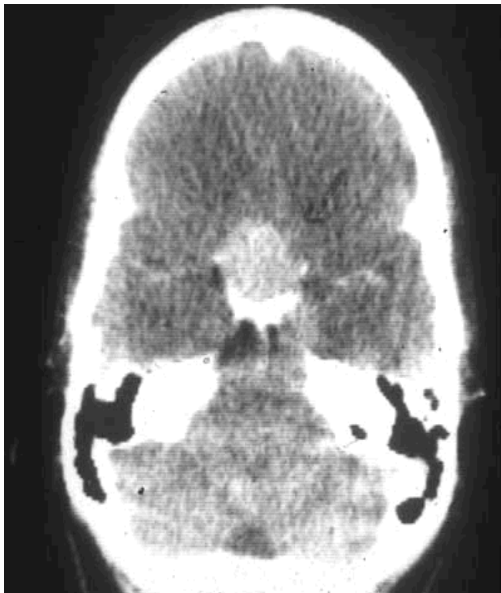


# Horner's Syndrome

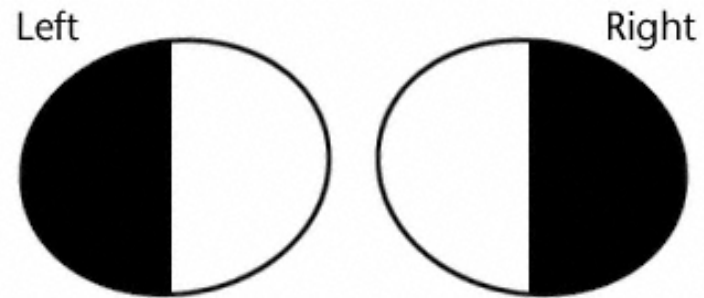
- Ptosis
- Miosis
- Anhydrosis
- Iris heterochromia (congenital cases)



# Pituitary Adenoma



- Bitemporal hemianopia



# Myotonic Dystrophy

- Ptosis
- Ophthalmoplegia



# Cardiovascular

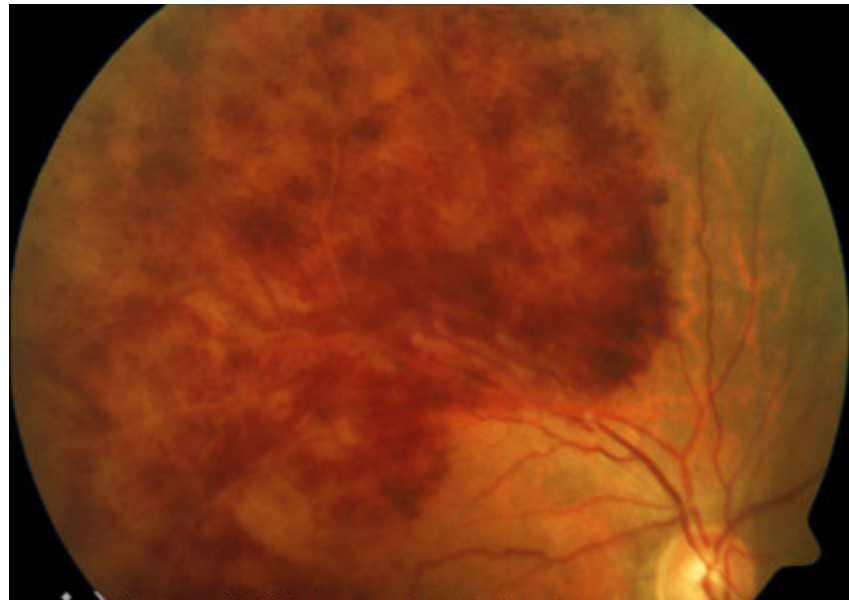
- Hypertension
- Atherosclerosis
- Arteriosclerosis
- Hyperlipidemia
- Embolic phenomena

# Cardiovascular

- Retinal vascular occlusion
  - Central retinal artery occlusion
  - Branch retinal artery occlusion
  - Central retinal vein occlusion
  - Branch retinal vein occlusion
- Anterior ischemic optic neuropathy
- Hypertensive retinopathy

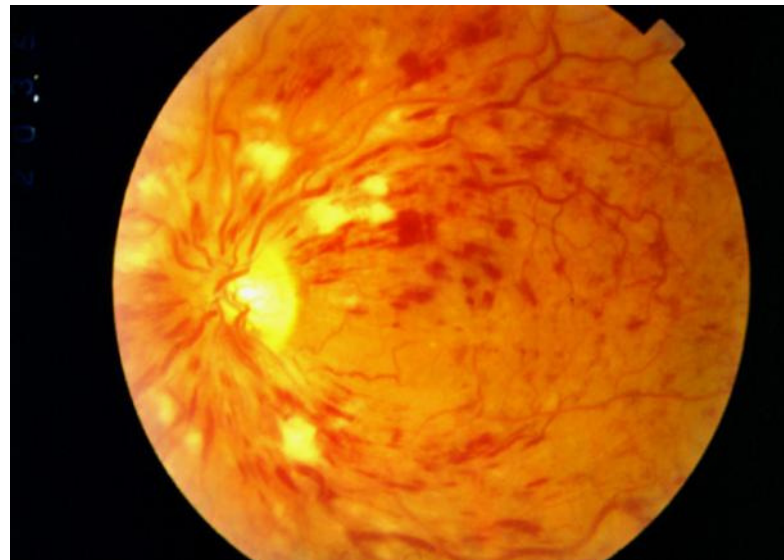
# Venous Occlusive Diseases

- **Branch Retinal Vein Occlusion (BVO)**
  - Arteriolosclerosis → retinal vein compression → thrombotic occlusion
  - Hypertension, diabetes mellitus, arteriosclerosis, hyperopia
  - Retinal hemorrhages, macular edema, ischemia, neovascularization

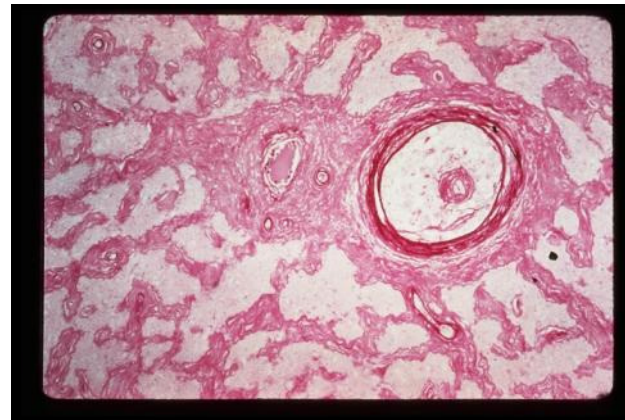
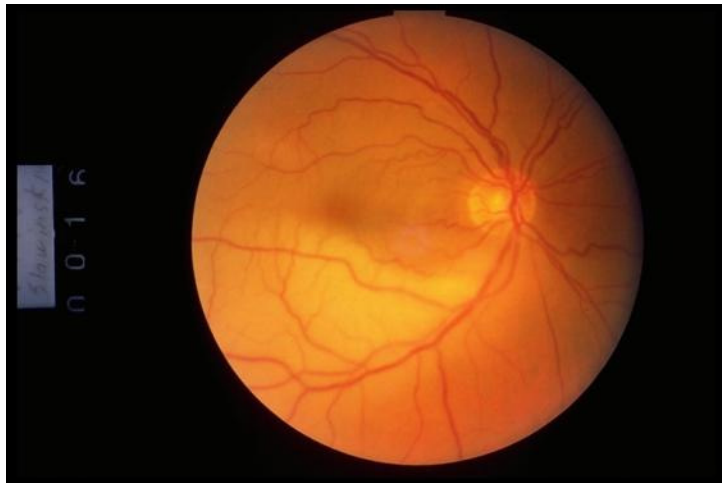


# Venous Occlusive Diseases

- **Central Retinal Vein Occlusion (CVO)**
  - Thrombus in central retinal vein at & posterior to lamina cribrosa
  - Hypertension, diabetes mellitus, hyperviscosity syndrome, blood dyscrasias, open angle glaucoma, oral contraceptives
  - Retinal hemorrhages, macular edema, ischemia, neovascularization



# Retinal Artery Occlusion



Retinal Arterial Occlusion

# Rheumatology

- Rheumatoid arthritis
- Seronegative spondyloarthropathies
  - HLA-B27 +ve
  - Ankylosing spondylitis, Reiter's disease, psoriatic arthritis
- Sjogren's syndrome
- Juvenile idiopathic arthritis
- Behçet's disease
- Sarcoidosis
- Systemic lupus erythematosus
- Wegener's granulomatosis
- Giant cell arteritis

# Rheumatology

- Dry eyes
  - RA
  - Sjogren's
- (Epi)Scleritis
  - Mainly RA
- Uveitis
  - Behçet
  - Sarcoidosis
  - Juvenile idiopathic arthritis
- Vascular occlusion
  - Systemic lupus erythematosus
- Orbital disease
  - Wegener's
- Anterior ischemic optic neuropathy
  - Central retinal artery occlusion
  - Giant cell arteritis

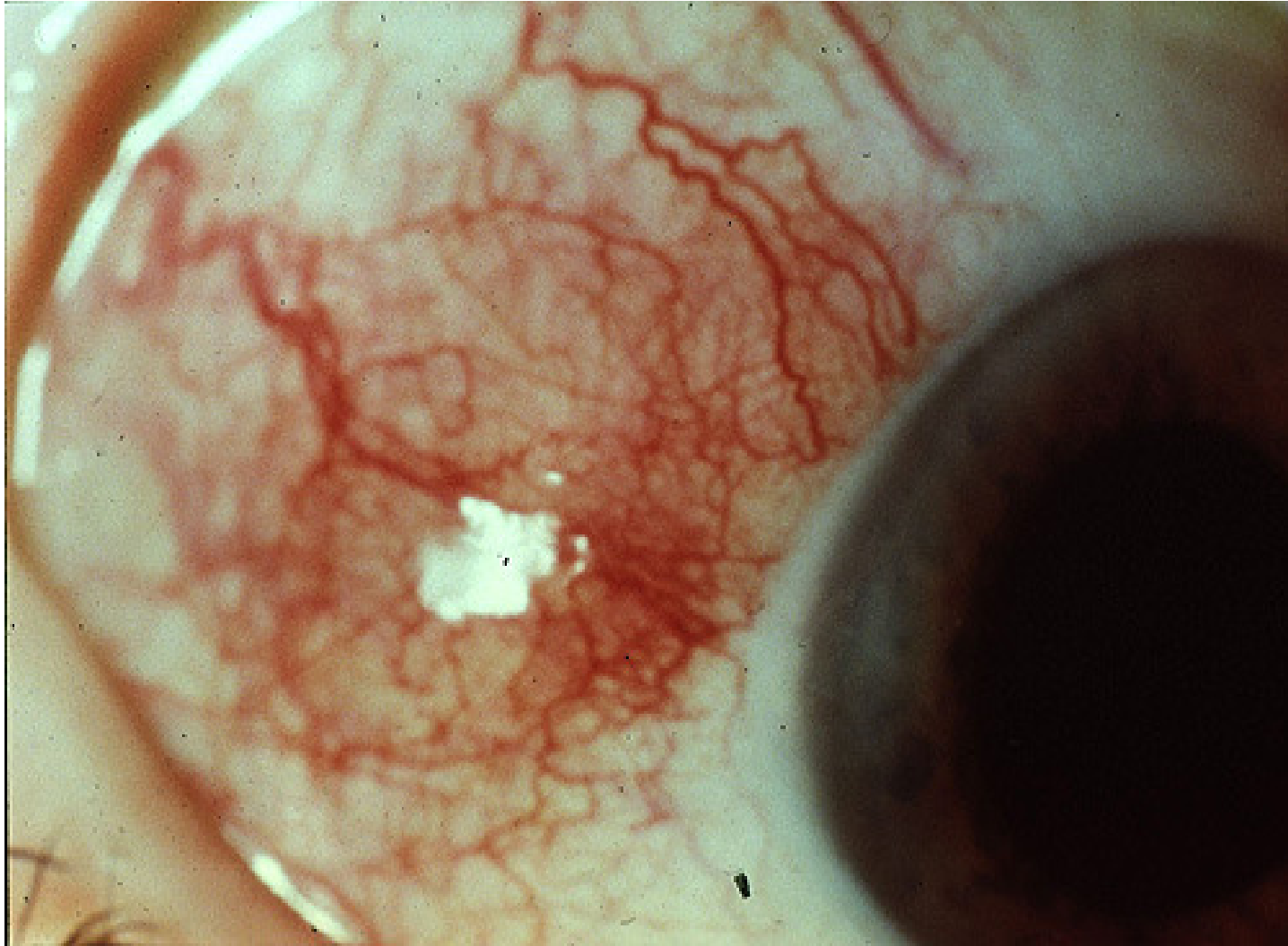
# Rheumatoid Arthritis



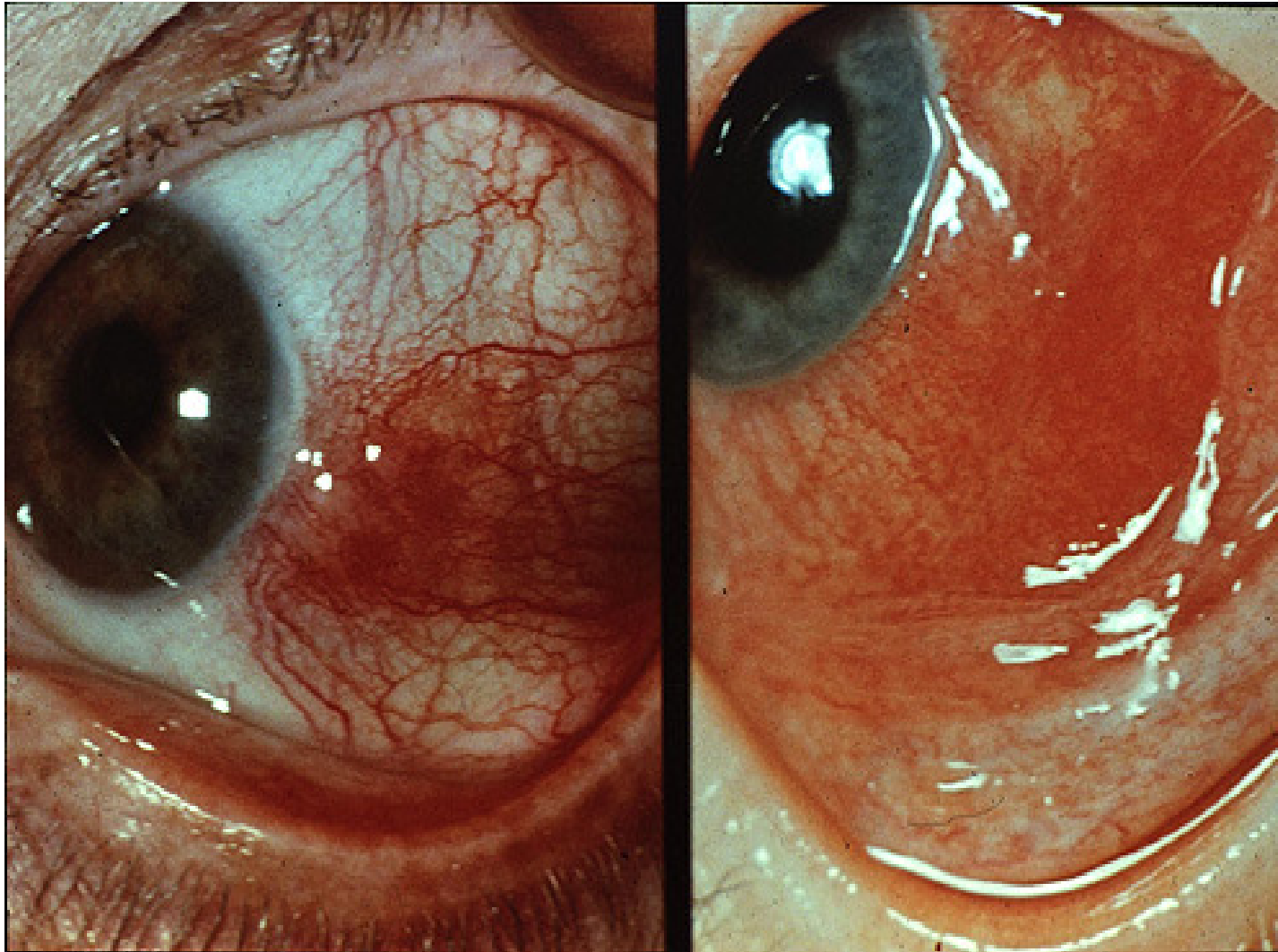
# Sjogren's Syndrome



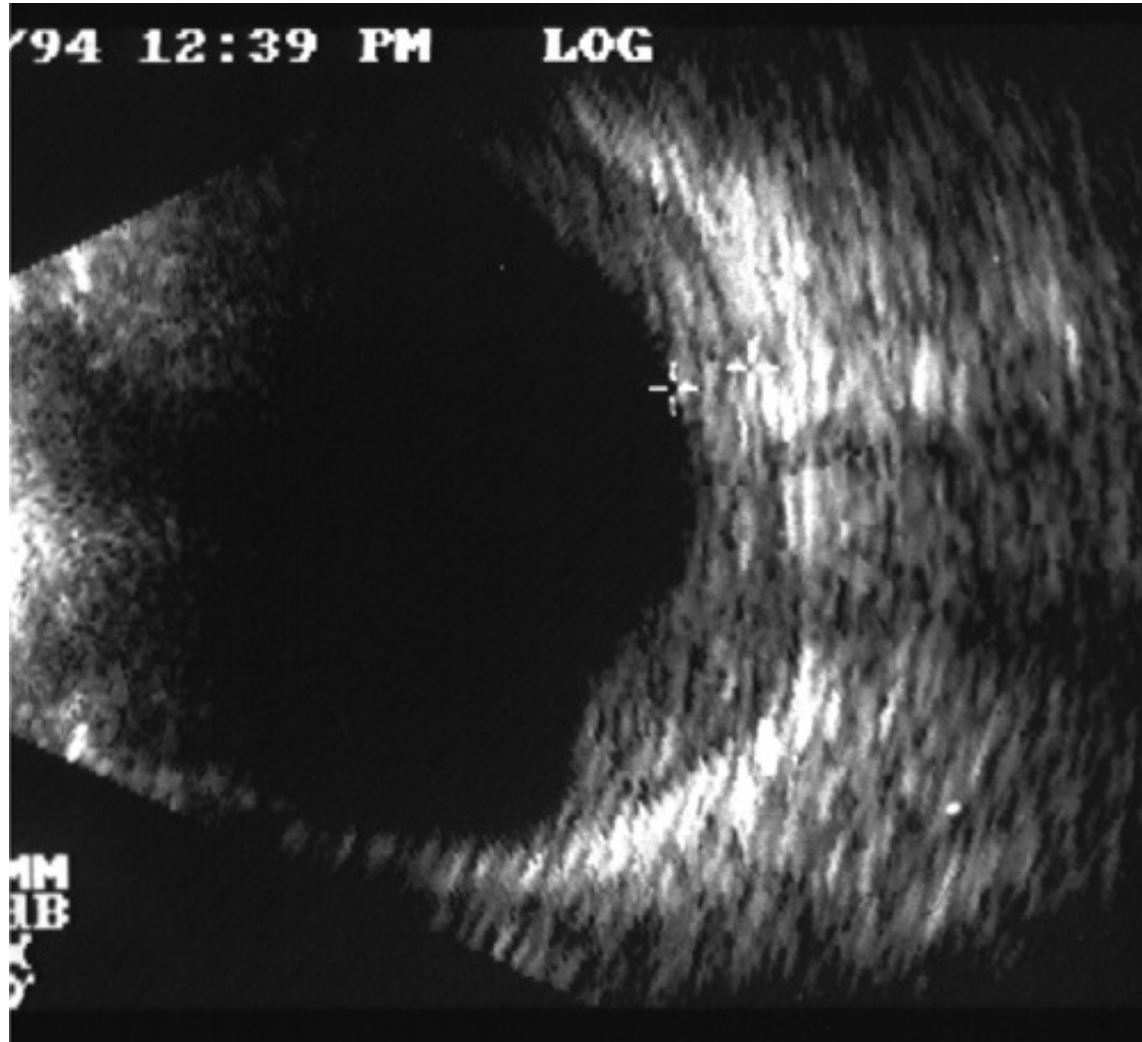
# Episcleritis



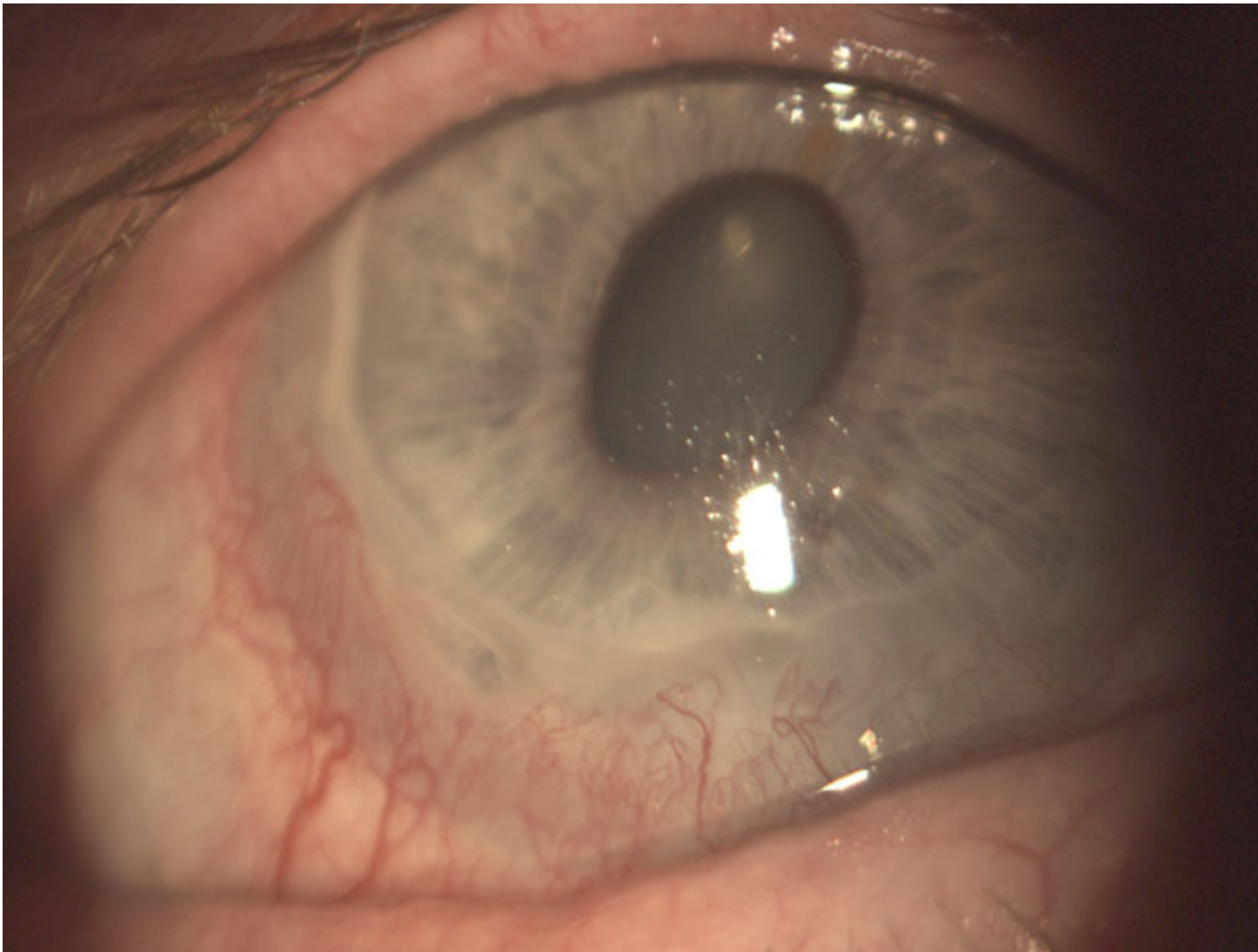
# Anterior Scleritis



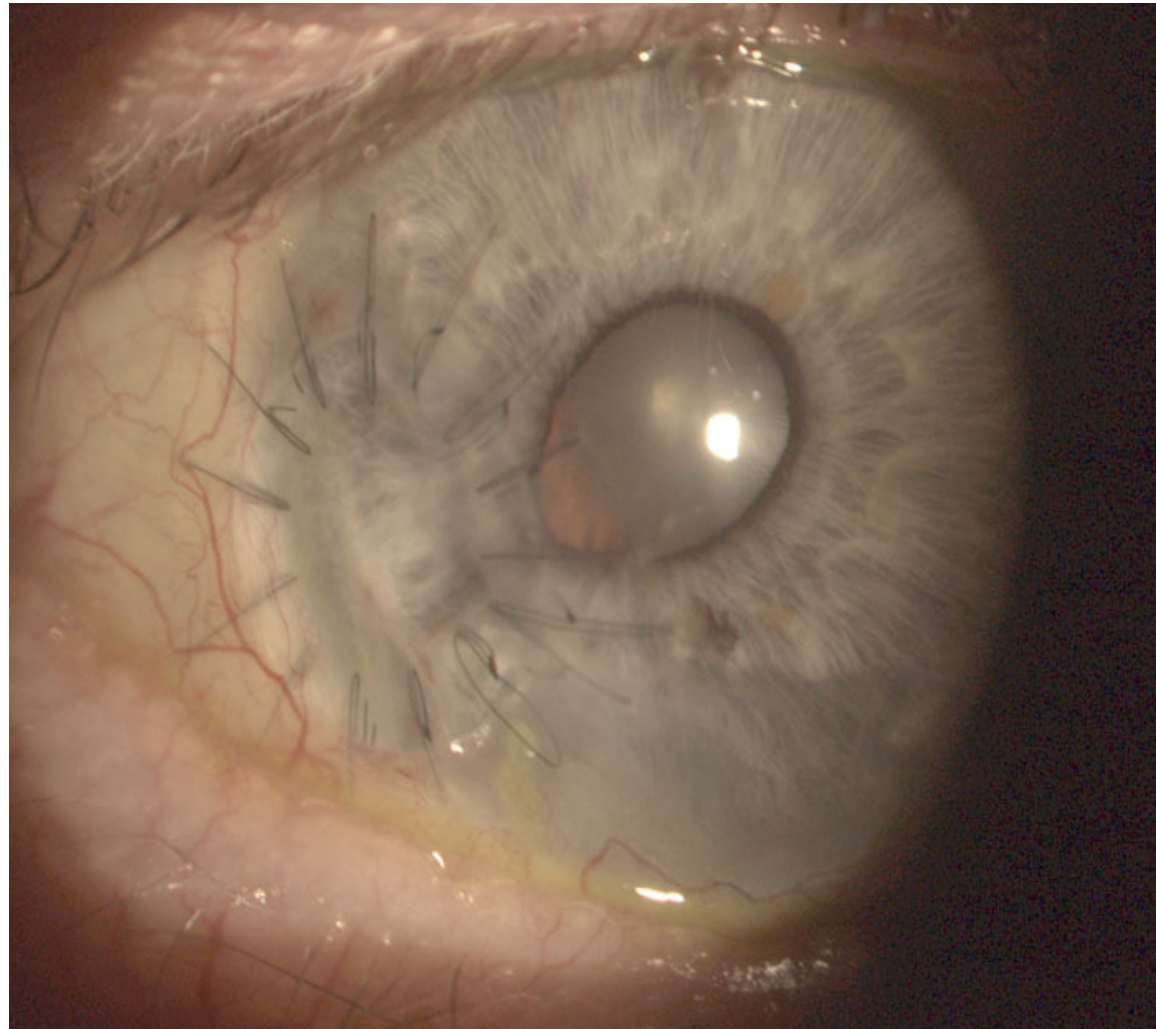
# Posterior Scleritis



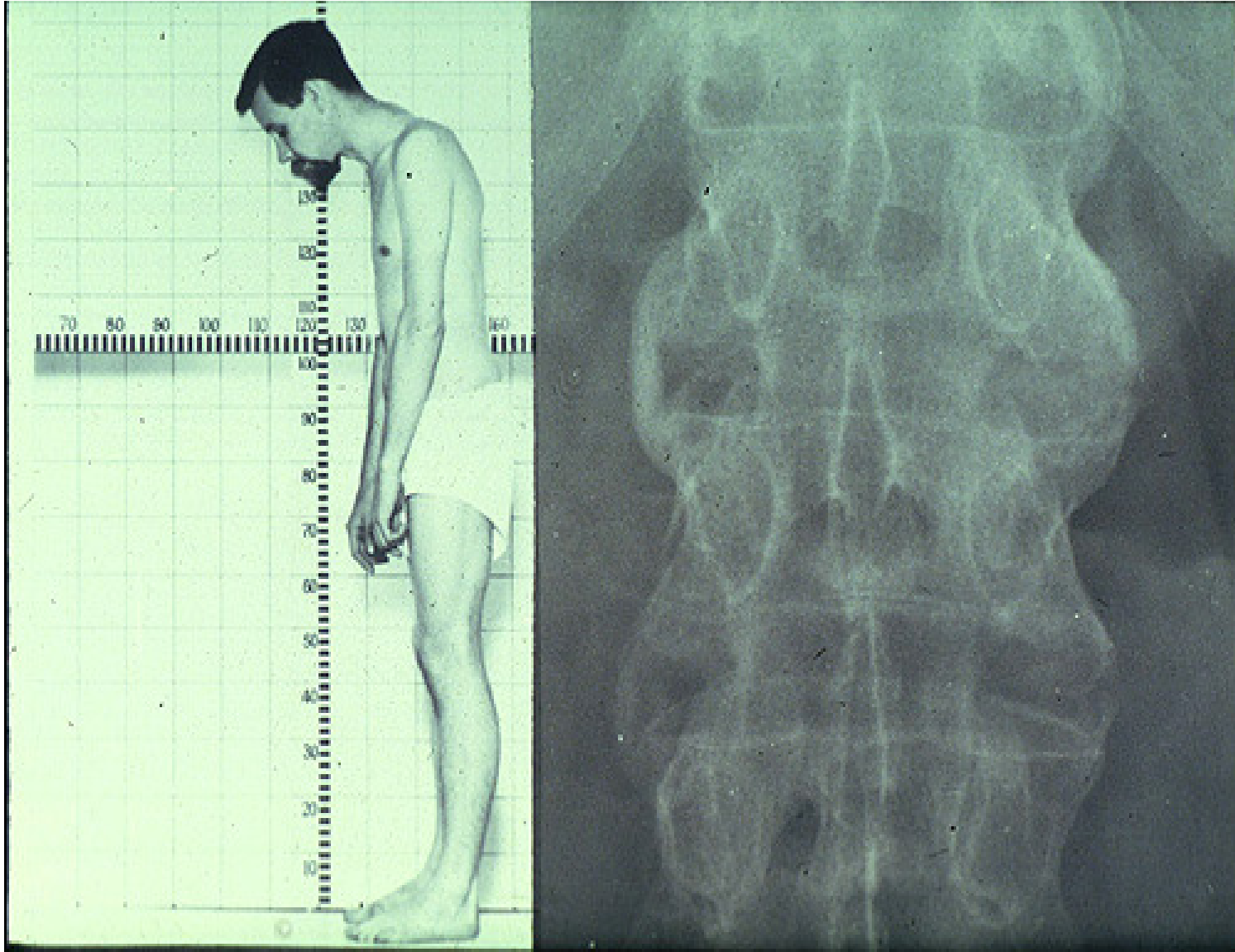
# Corneal Melt



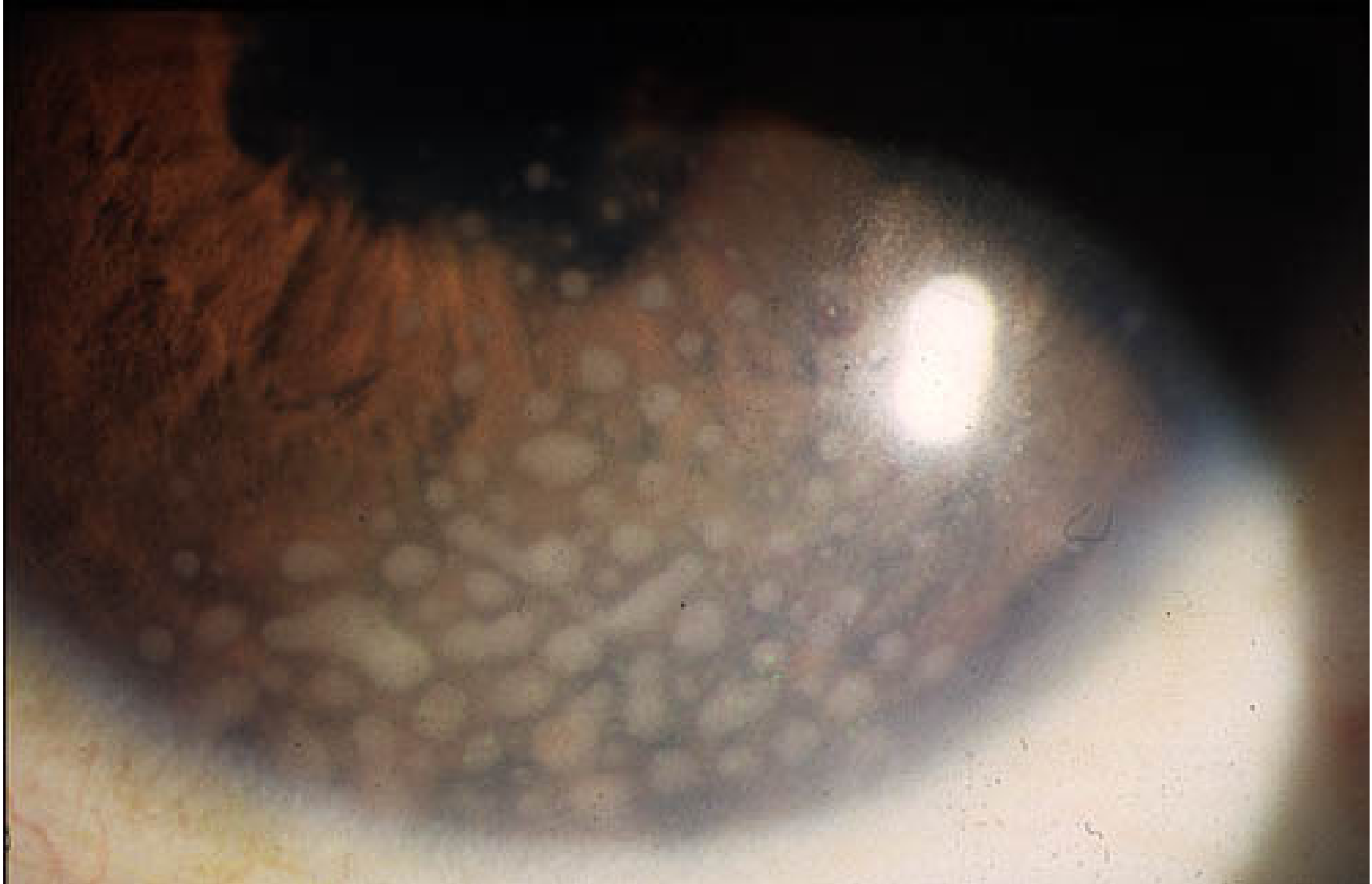
# Corneal Graft for Melt



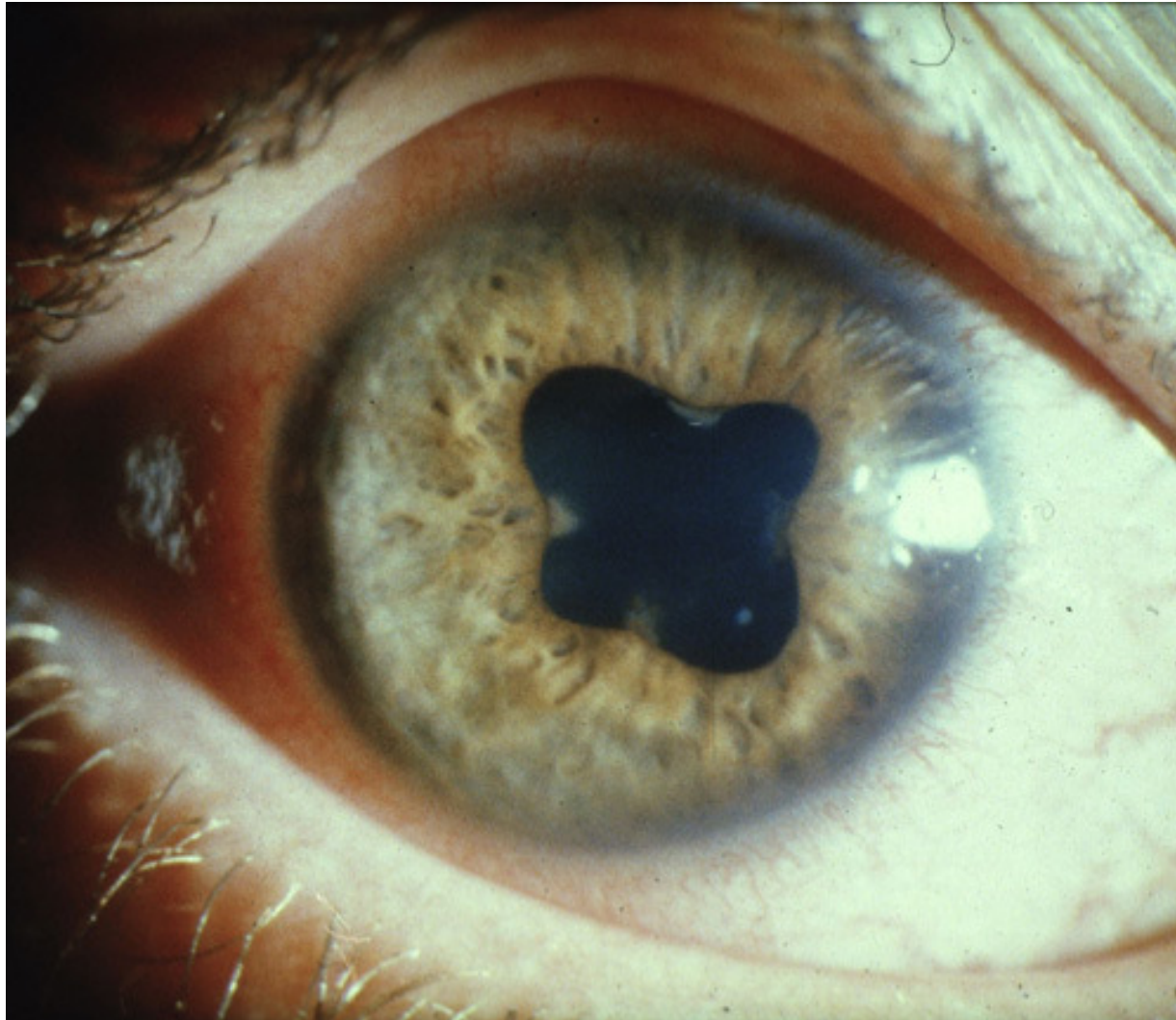
# Ankylosing Spondylitis



# Uveitis – Keratic Precipitates



# Uveitis – Posterior Synechiae

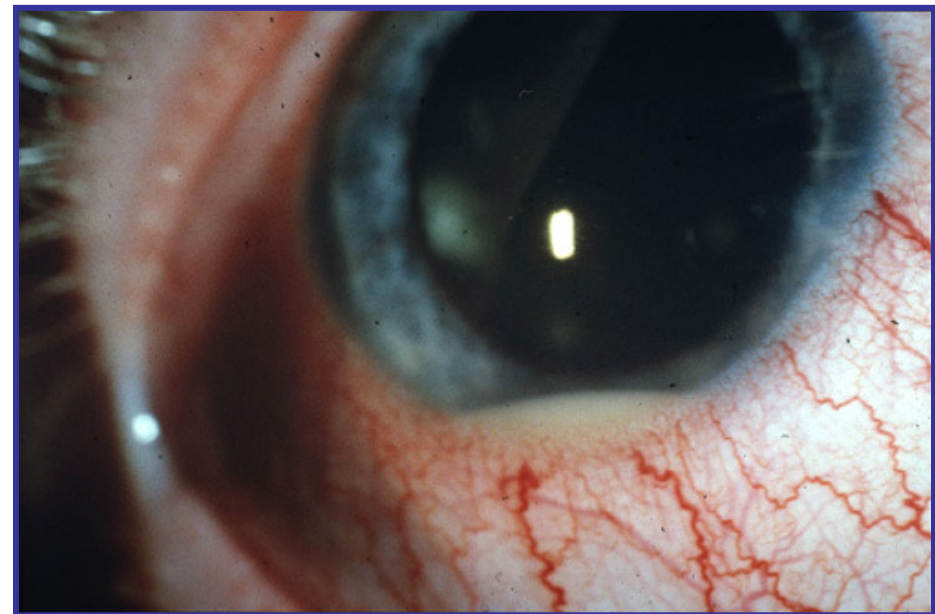
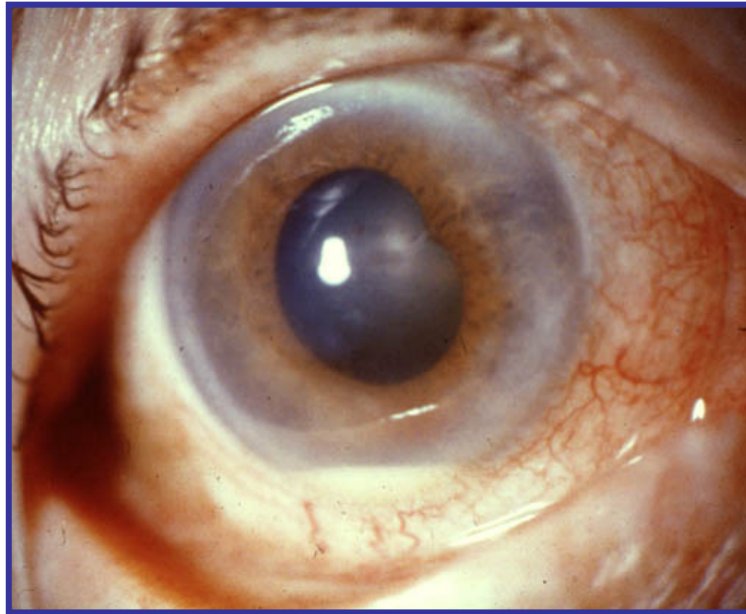


# Behçet's Disease

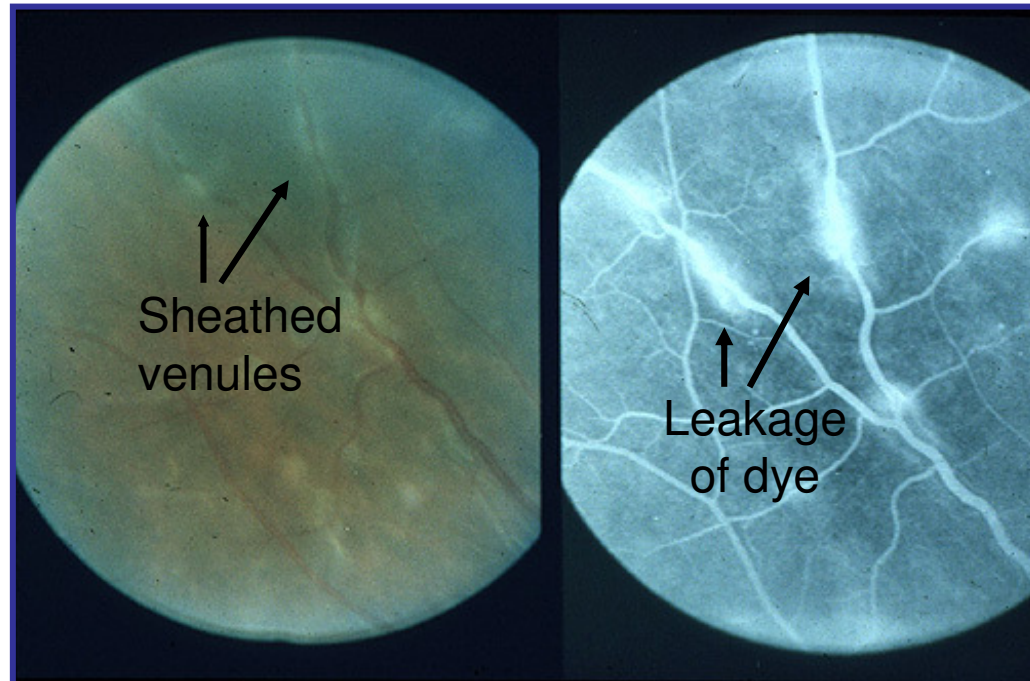
- Idiopathic multisystem disorder
- Occlusive vasculitis - eyes involved in 75%
- Presents in the 3rd and 4th decades
- Diagnosis: Oral ulceration with two of
  - Recurrent genital ulceration
  - Skin lesions - folliculitis, erythema nodosum
  - Positive pathergy test
  - Eye involvement
    - Acute anterior uveitis
    - Vitritis, vasculitis, retinitis
- Treatment: systemic immunosuppression

Devastating, potentially blinding  
condition of young adults

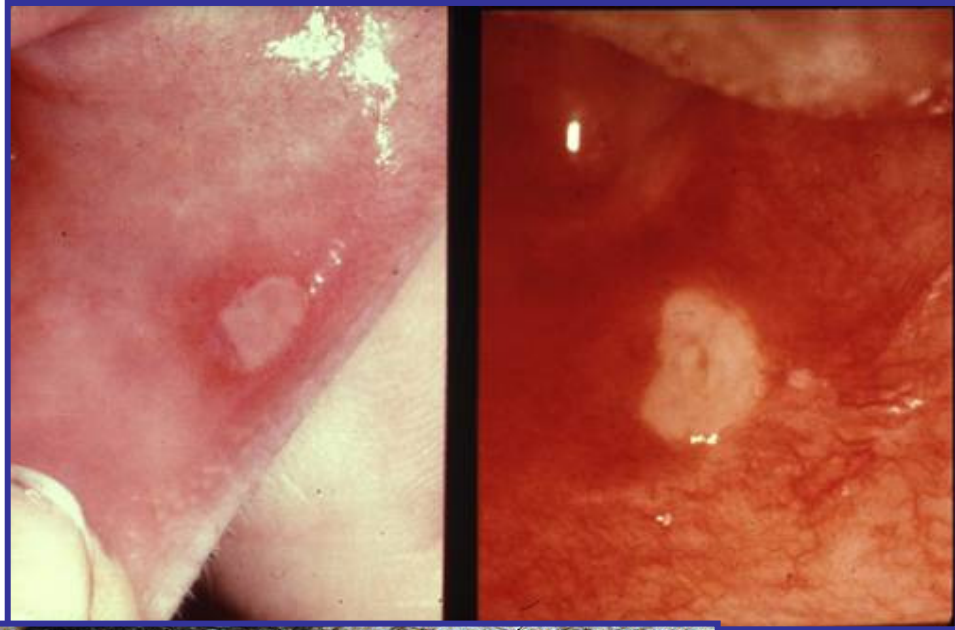
# Hypopyon



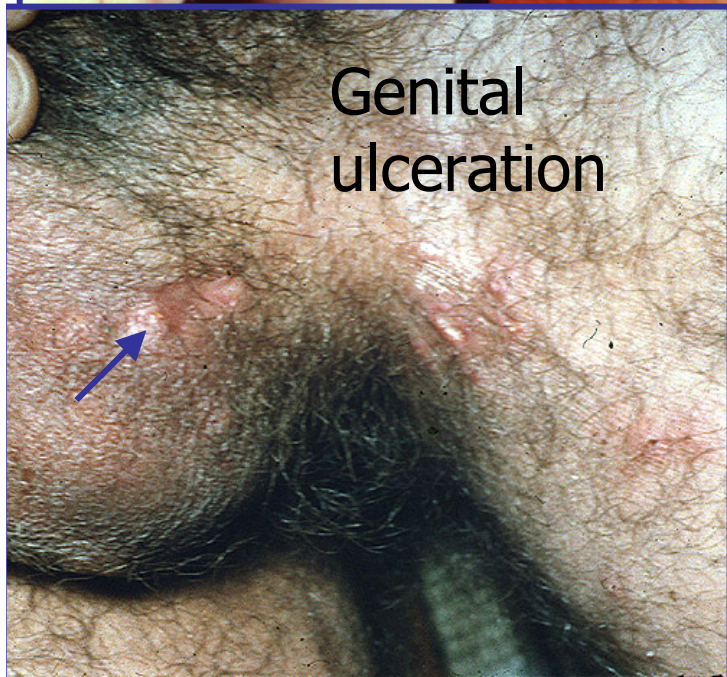
Retinal  
vasculitis



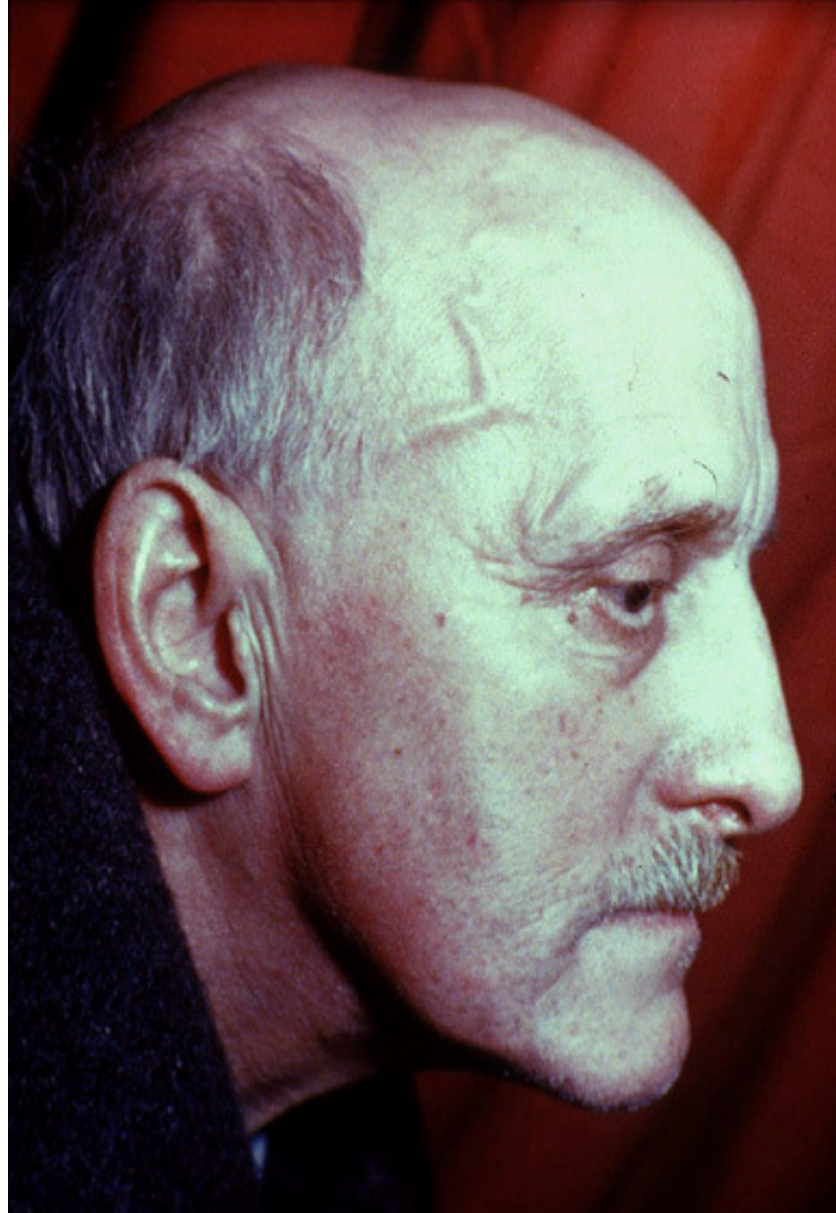
# Aphthous oral ulceration



+ve Pathergy test

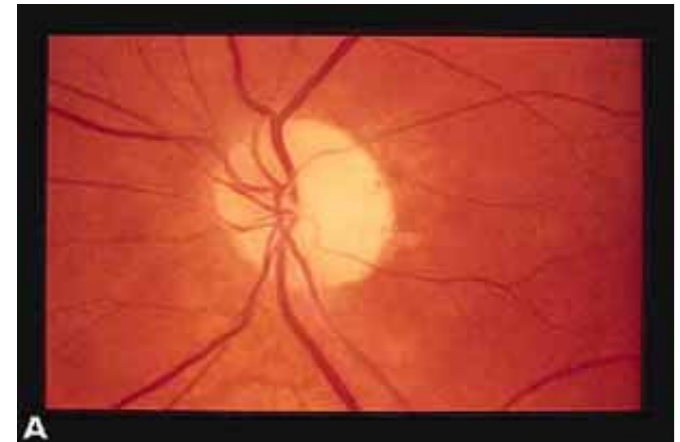


# Giant Cell Arteritis



# Acute Visual Loss

- **Ischemic Optic Neuropathy**
  - **Arteritic**
    - Afferent pupillary defect
    - Devastating visual loss
    - Pale swollen optic nerve, often with flame shape hemorrhage
    - Optic atrophy as ON edema resolves
    - Visual field defects
    - Immediate ESR, C-reactive protein
    - Temporal artery biopsy
    - High dose steroids



# Dermatology

- Acne rosacea
- Topical allergy
- Stevens-Johnson syndrome (erythema multiforme)
- Psoriasis
- Herpes zoster ophthalmicus
- Mucous membrane pemphigoid
- Sturge-Weber
- Lid tumors
  - BCC, SCC

# Acne Rosacea



- **Blepharitis**

- Hyperemic, crusty & thickened lid margins
- Prominent blood vessels
- Inspissated oil glands at lid margins



- **Complications**

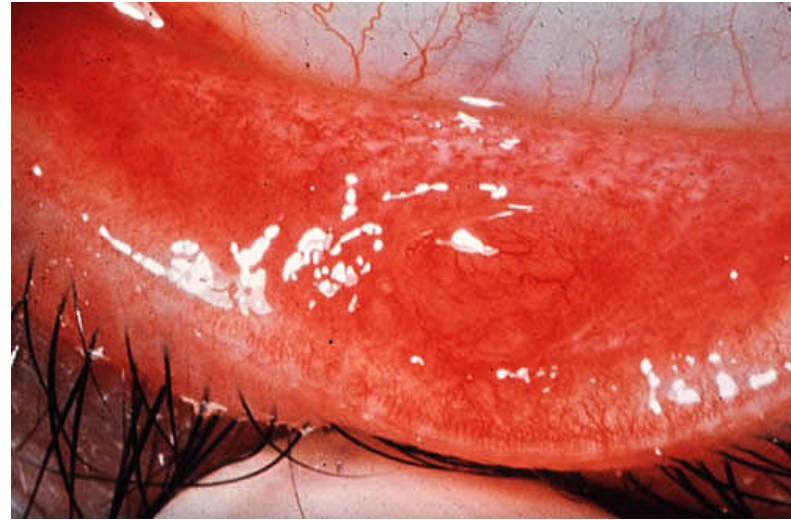
- Conjunctivitis
- Marginal keratitis
- Meibomian cysts



# Stye (Hordeolum)



# Chalazion



# Stevens Johnson Syndrome

- Acute, self limiting, males > females
- Hypersensitivity reaction to drugs and infection resulting in an acute vasculitis
- Presentation: fever, malaise, sore throat, arthralgia
- Eye: pseudomembranous conjunctivitis, cicatrization of the conjunctiva and lids
- Skin: symmetrical erythematous maculopapules, vesicubullous lesions
- Mouth: bullae, erosions, haemorrhagic crusts



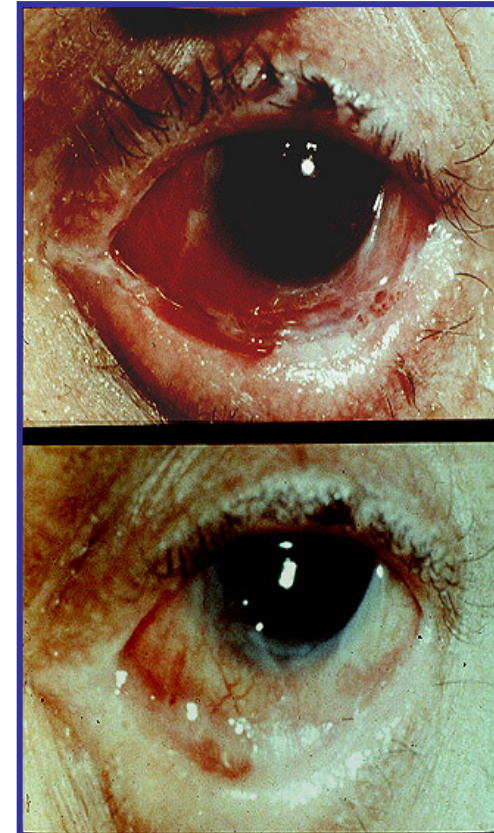
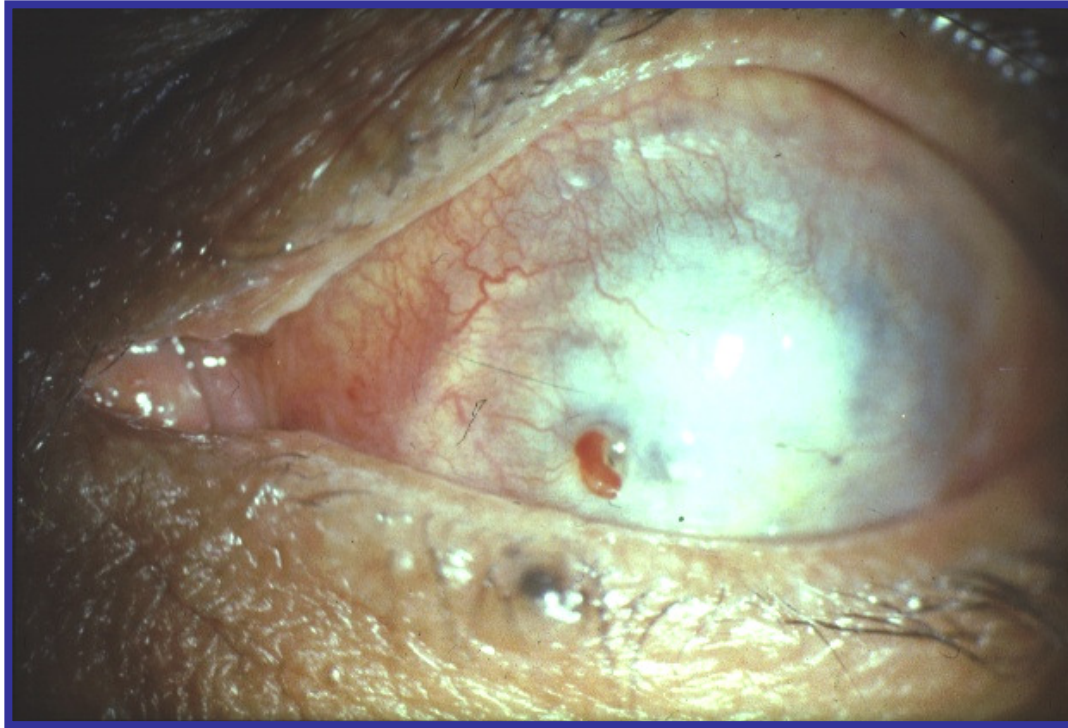
# Herpes Zoster Ophthalmicus



# Cicatricial Mucous Membrane Pemphigoid

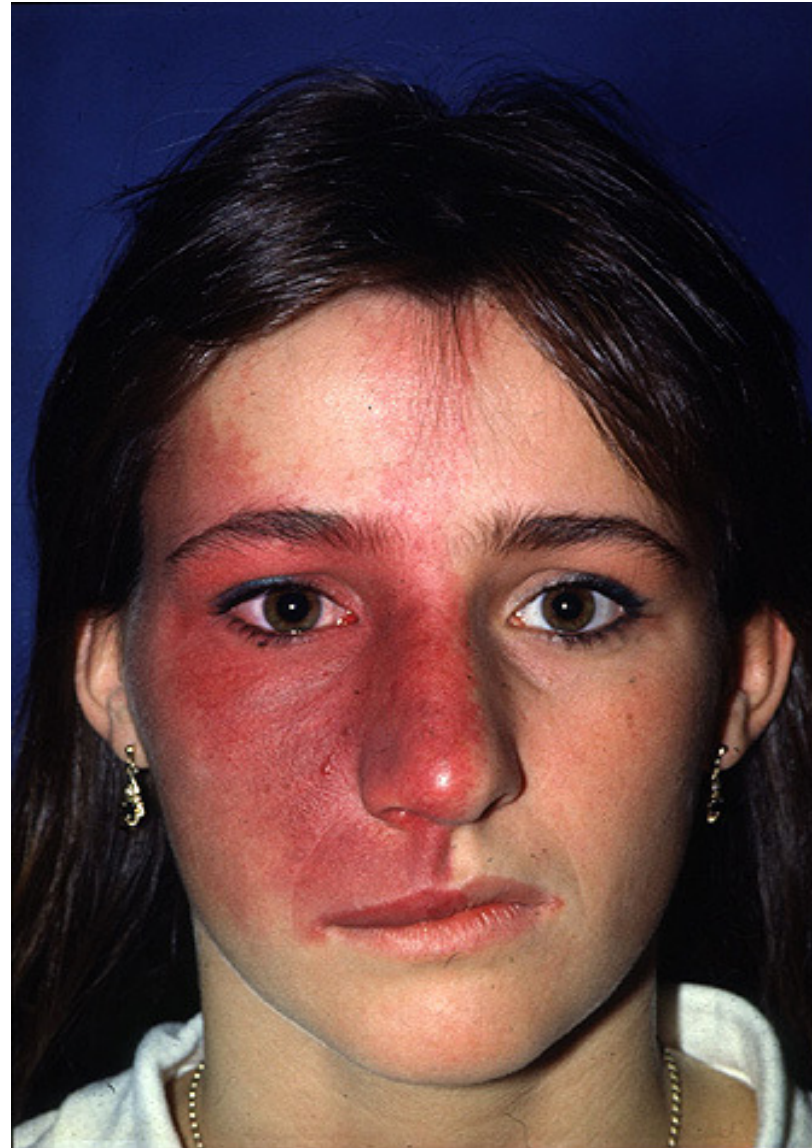
- Rare, idiopathic, chronic progressive, females>males
- Inflammation followed by scarring
- Skin: 25%
  - Type I: recurrent vesicobullous, non-scarring
  - Type II: Localised erythematous plaques, with recurrent vesicles and bullae, healing to small atrophic scars

- Mouth: 80%
  - Submucosal blisters leading to erosions, scars and strictures
- Eye: 50-70%
  - Dry eye, symblepharon, ankyloblepharon, keratopathy
  - Treatment with topical and systemic steroids, immunosuppressives, silicone contact lenses, lid surgery



# Sturge-Weber Syndrome

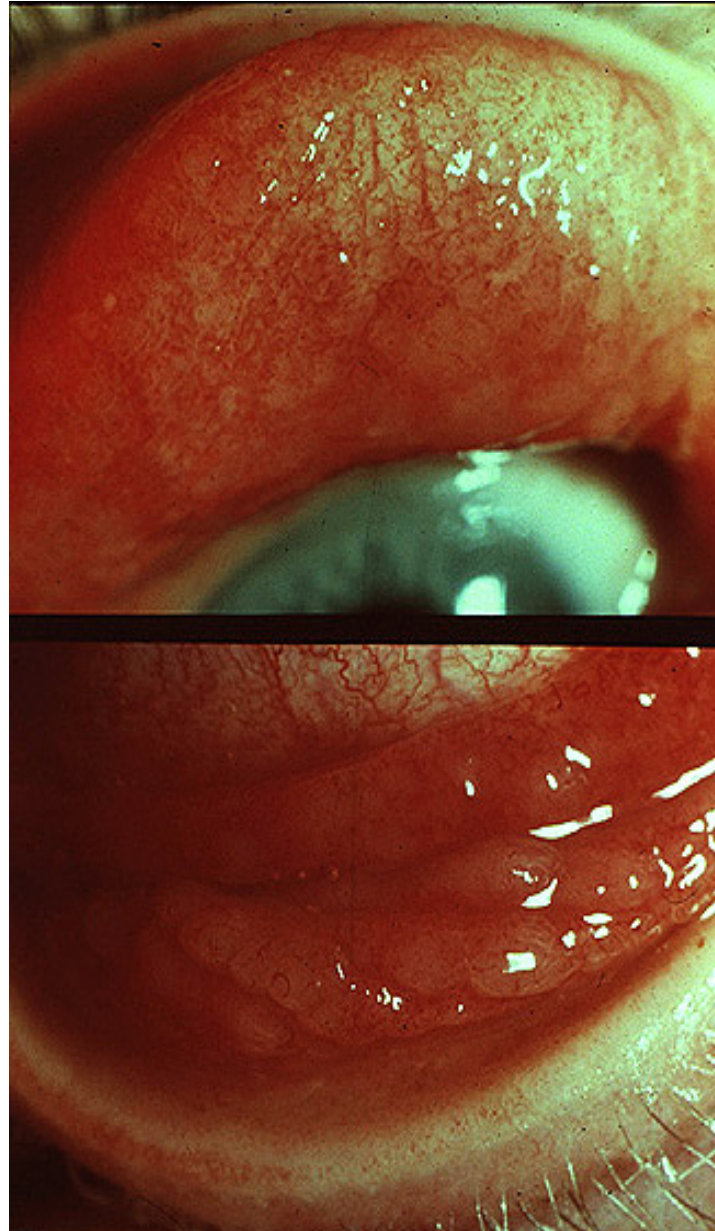
- Nevus flammeus (port wine stain)
- Congenital glaucoma
- Diffuse choroidal hemangioma
- Serous retinal detachment



# Genitourinary

- Syphilis
  - Masquerade syndrome
- Chlamydia
  - Conjunctivitis
- Gonorrhoea
  - Conjunctivitis
- HIV
  - Opportunistic infections - e.g., cytomegalovirus (CMV) retinitis

# Chlamydial Conjunctivitis



Oral Hairy Leukoplakia



CMV Retinitis



HIV

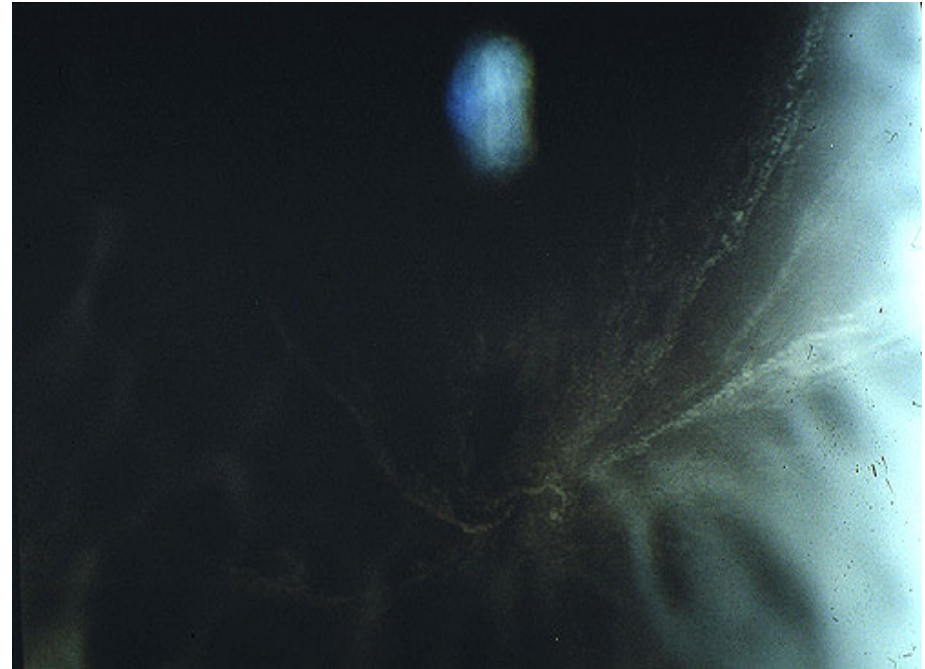
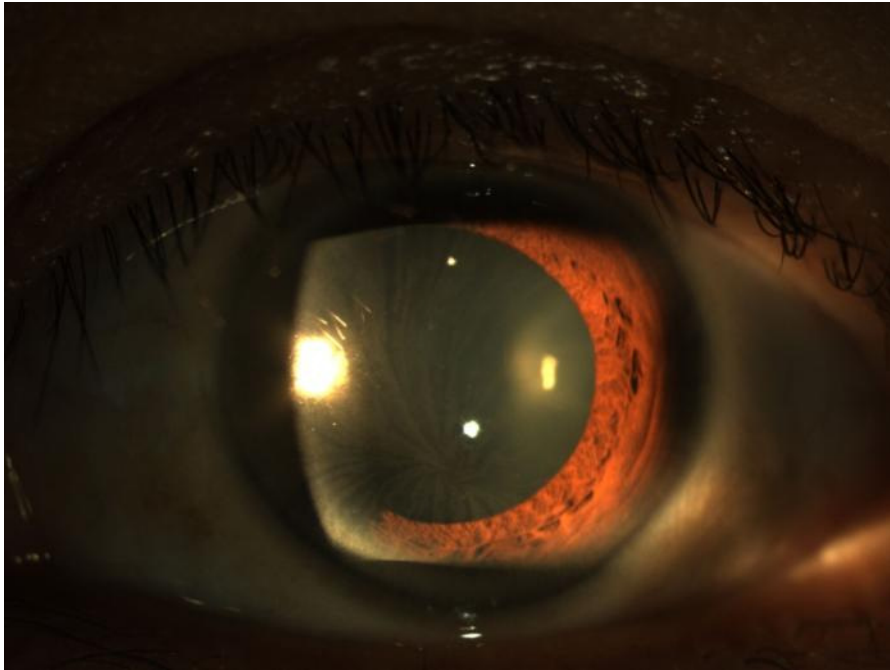
# Others

- Gastrointestinal
  - Inflammatory bowel disease (Crohn's disease, ulcerative colitis)
    - Uveitis, episcleritis and scleritis
- Respiratory
  - Sarcoidosis
    - Masquerade syndrome
- Nephrology
  - Systemic lupus erythematosus
  - Wegener's

# Medications for Systemic Diseases

- Corticosteroids
  - Posterior subcapsular cataract
  - Secondary glaucoma
- (Hydroxy)Chloroquine
  - Maculopathy
- Desferrioxamine
  - Maculopathy
- Tamoxifen
  - Maculopathy
- Quinine
  - Optic atrophy, arteriolar narrowing
- Amiodarone
  - Corneal change
- Vigabatrin
  - Visual field loss

# Amiodarone Keratopathy



Corneal verticillata  
(Vortex keratopathy)