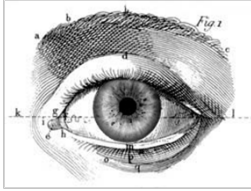

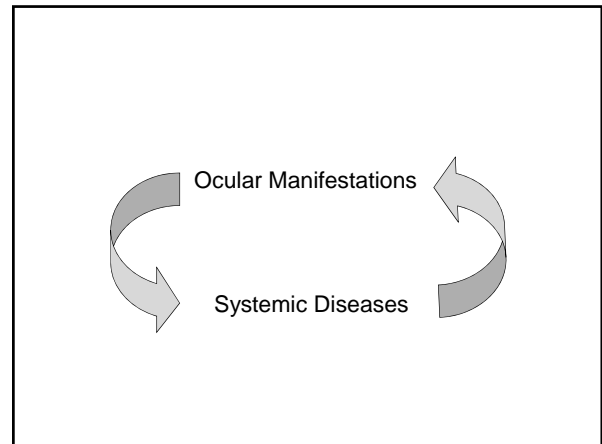


Ophthalmic Manifestations of Selected Systemic Diseases

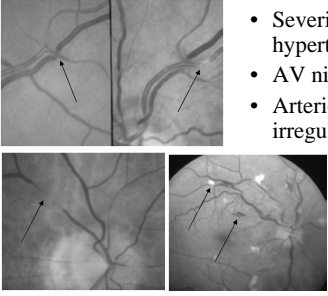


Patrick Sibony, MD
March, 2012

- ### Categories of Systemic Diseases
-
- Congenital
 - Genetic
 - Trauma
 - Vascular
 - Neoplastic
 - Autoimmune
 - Idiopathic
 - Infectious
 - Metabolic /
 - Endocrine
 - Drugs / Toxins

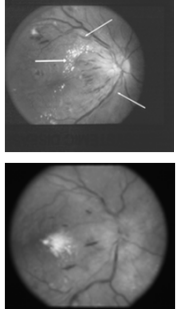
Chronic Hypertensive Retinopathy



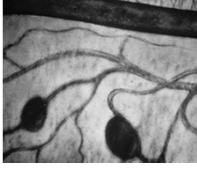
- Severity and duration of hypertension.
- AV nicking
- Arteriolar narrowing and irregularity
- Sclerotic vessels
- Blot/flame heme
- Cotton wool spots

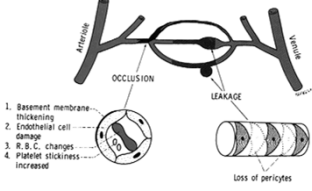
Acute hypertensive retinopathy

- Malignant, accelerated, hypertensive crisis
- BP >200/120
- Renal disease, toxemia of pregnancy, vasculitis
- Usually associated with vision loss
- Severe exudative changes, disc edema, hemorrhages

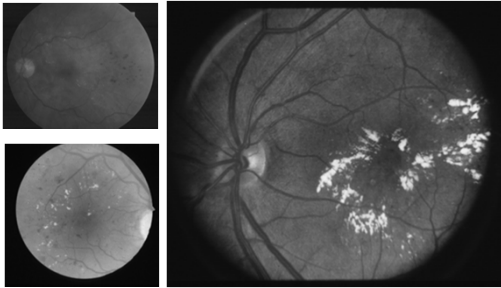


Diabetic Retinopathy

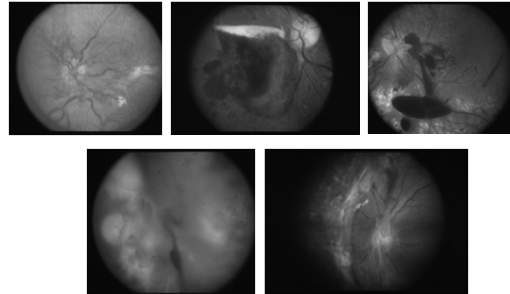




Non proliferative Diabetic Retinopathy



Proliferative Diabetic Retinopathy



Factors that lead to progression

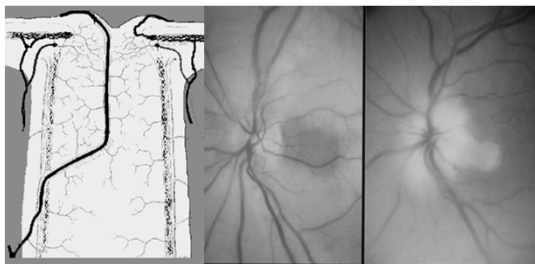
- **Puberty and pregnancy**
- Systolic and diastolic **blood pressure**
- **Hyperlipidemia** : hard exudates in the macula and high risk of visual loss.
- Poor control of **serum glucose**
- **ASA and smoking has no effect.**

Evaluation of Diabetics

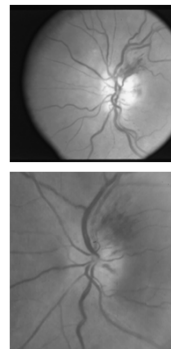
- * May progress without visual symptoms.
- * Florid neovascularization and still maintain perfect 20/20 vision.
- Insulin dependent, juvenile onset:
 - Needs exam during first 4 years, then yearly
- Non insulin dependent, adult onset:
 - Needs exam at the time of diagnosis, then yearly
- Diabetes prior to pregnancy:
 - Needs exam prior to or early in first trimester, then every trimester

Ocular Circulation

Central retinal a. Posterior ciliary a

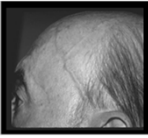
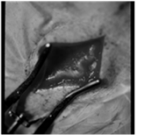



Ischemic Optic Neuropathy



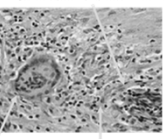
- Sudden , painless vision loss with signs of optic neuropathy.
- Non arteritic
 - HBP or DM
 - Blood loss / Anemia
- Arteritic
 - Cranial arteritis

Cranial Arteritis







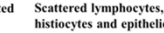
**Disruption of the internal elastic lamina
Intimal hyperplasia**




Granulomatous inflammation



Luminal stenosis

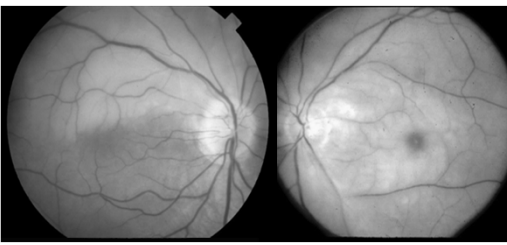


**Multinucleated
Giant cell**





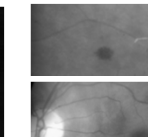
**Scattered lymphocytes,
histiocytes and epithelioid cells**

Retinal Artery Occlusion



- Carotid stenosis, cardiogenic emboli, vasculitis, and hypercoagulability
- Ophthalmic emergency: paracentesis
- Urgent evaluation to prevent further events

Retinal Emboli

**Cholesterol
(Hollenhorst plaque)**

Carotid
(aorta, heart valves)

Asymptomatic

Calcific

Cardiac

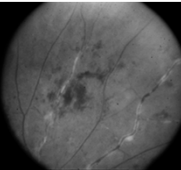
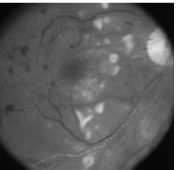
BRAO

Platelet – fibrin

Carotid or cardiac

BRAO

Retinal Vasculitis

Retinal periphlebitis
Sarcoidosis

Primary Ocular conditions

Idiopathic obliterative peripheral retinovasculopathy (Eale's disease)

Frosted branch angiitis

Birdshot chorioretinopathy

Idiopathic retinal vasculitis

Optic disc vasculitis

Systemic Associations.

Lupus

Rheumatoid arthritis

Microangiopathy of retina/brain (Susacs)

Inflammatory bowel disease

Wegeners, Polyarteritis

Acute retinal necrosis (herpes zoster)

Retinal arteriolitis
Lupus

Giant cell arteritis

Sarcoidosis

Behcets

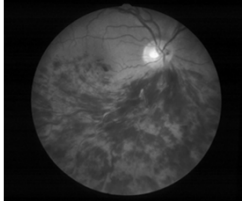
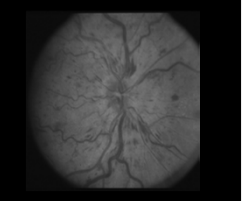
Multiple sclerosis

syphtilis, TB

Toxoplasmosis

CMV (HIV)

Retinal Vein Occlusion

Branch vein occlusion

Hypertension, glaucoma, hypercoagulability, anticardiolipin antibody Syndrome, hyperviscosity, myeloproliferative disorders, anemia

Central retinal vein occlusion

Hypertension, glaucoma, hypercoagulability, anticardiolipin antibody Syndrome, hyperviscosity, myeloproliferative disorders, anemia

Transient Vision Loss

Monocular

Transient Visual Obscurations

Optic disc edema (papilledema, uveitis, tumor)

Seconds

Transient Monocular Blindness [Amaurosis Fugax]

Carotid stenosis
Cardiogenic
Vasculitis
Hypercoagulable

2-10 m

Binocular

Migraine

15- 45 m

Vertebrobasilar TIA


2-10 m

45 yo WM with a visual disturbance.

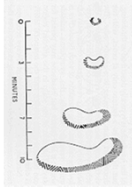
DATE	EX	DESCRIPTION
1/1/12	11min	A bilateral scintilla in the 2nd° when looking in a horizontal up gaze. The patient reported a "shimmering" effect and that the vision was "flickering" and "blurred" and sometimes it was "very bright" and "white" and "like a star" and "like a sun" and "like a fire" and "like a light" and "like a glow" and "like a shimmer" and "like a sparkle" and "like a glimmer" and "like a gleam" and "like a glint" and "like a glaze" and "like a gloss" and "like a sheen" and "like a luster" and "like a radiance" and "like a brilliance" and "like a splendor" and "like a glory" and "like a radiance" and "like a brilliance" and "like a splendor" and "like a glory".

Doesn't matter whether either or both eyes are open, closed, look left or right, up or down
 Day or Night - **Symmetrical**


During a conversation in the afternoon - Kaleidoscope effect
 Evening or morning in bed - 30-10 min duration
 eyes open or closed - gradually moves out
 used to occur 1/2 year - of view shimmering
 now 5/6 times a year - 70 seconds of pain



Migraine

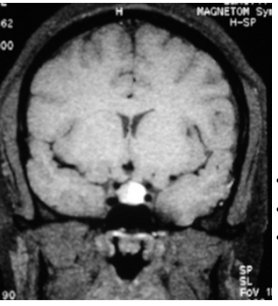
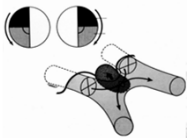


Binocular, scintillations ± hemianopic scotoma
 15-45 minutes
 Fortification spectra
 "Spectral march" across the visual field
 with or without headache
 prior h/o migraine headaches
 strong family history of headaches





Bourlange : Dutch fortress. 1577 - 1821

Pituitary tumors

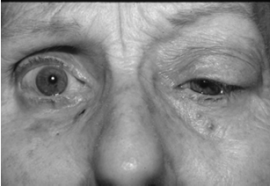
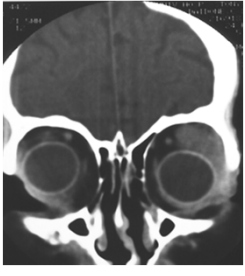
- ACTH: Cushing's
- GH: Acromegaly
- Prolactin:
 - F: amenorrhea, galactorrhea
 - M: impotence, gynecomastia

Orbital Lymphoma

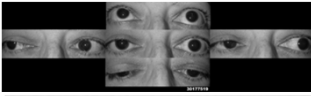

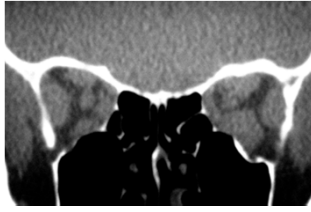

- Extranodal B cell lymphomas
- 50-70 yo
- Unilateral (bilateral)
- Proptosis, anterior congestion, ophthalmoplegia, ptosis
- 40% associated with systemic involvement.
- May infiltrate any of the orbital structures or present as a molding, non displacing mass.
- Immunohistochemistry - to distinguish from benign lymphoid hyperplasia

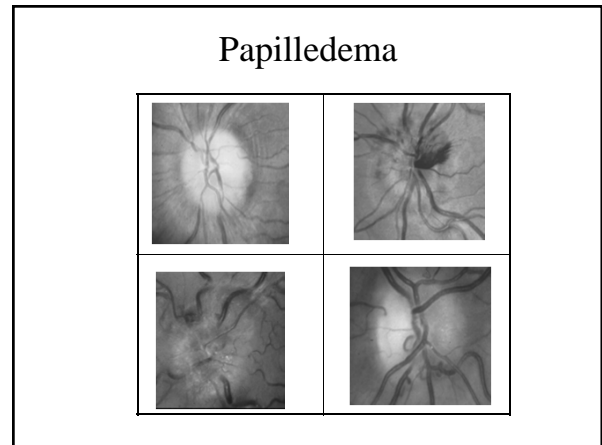
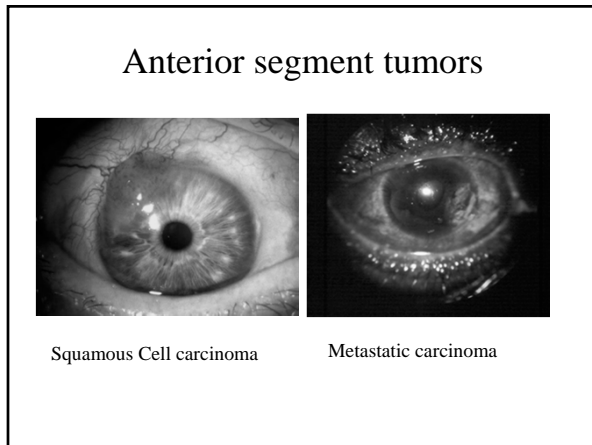
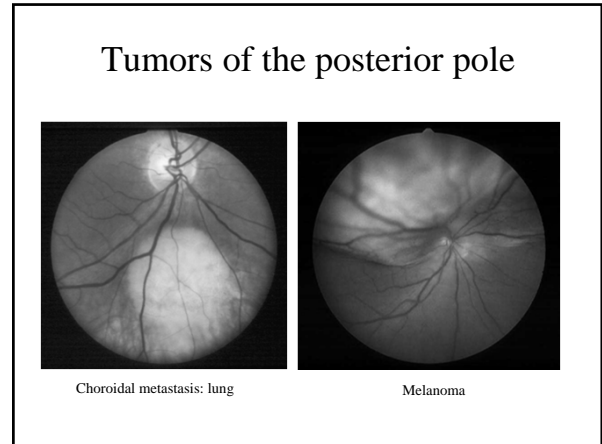
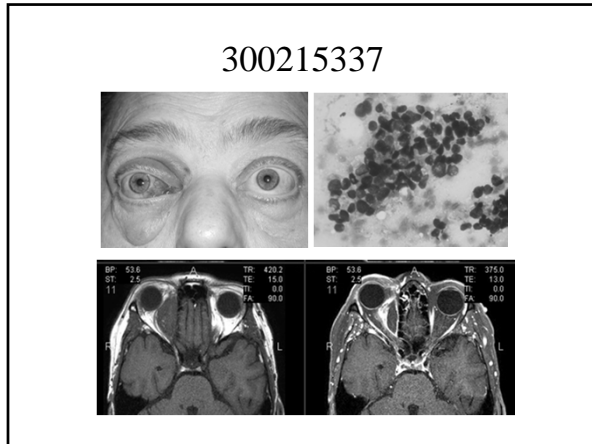
Orbital Lymphoma

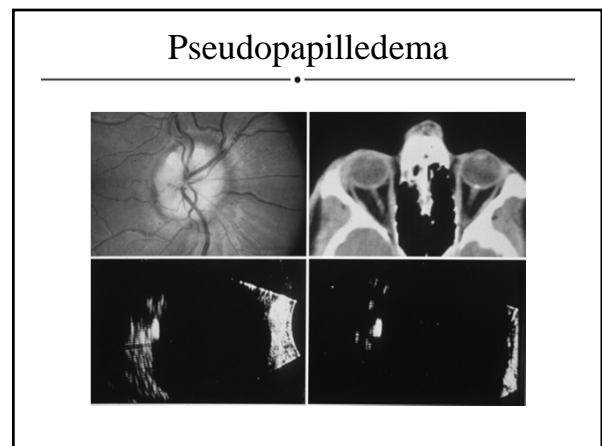
Orbital Myeloma

30177519

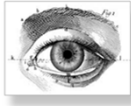


- Papilledema
- Idiopathic intracranial hypertension
 - Brain tumors
 - Venous sinus thrombosis
 - Obstructive hydrocephalus
 - Meningitis
 - Cerebral edema
 - Subarachnoid hem



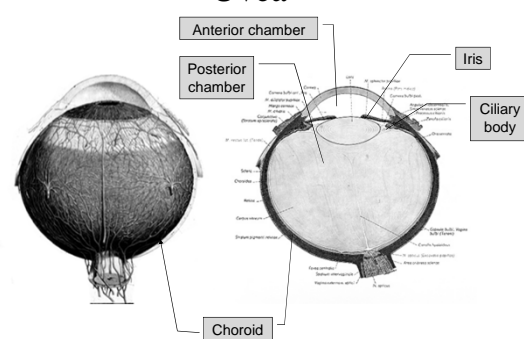
Part 2

Ophthalmic Manifestations of Selected Systemic Diseases



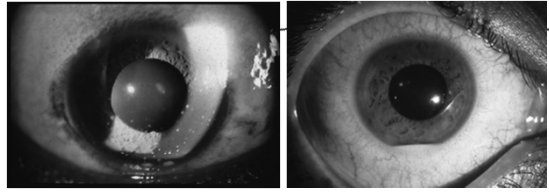
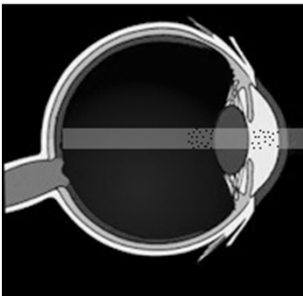
Patrick Sibony MD
March 2012

Uvea



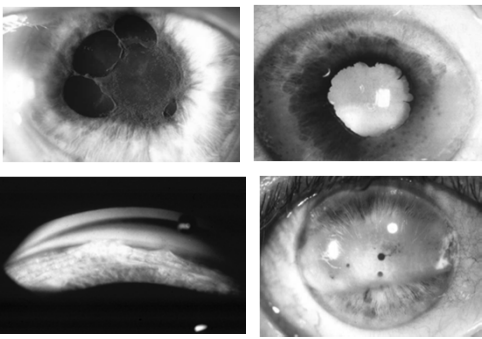
Anterior chamber
Posterior chamber
Iris
Ciliary body
Choroid

Cells and flare

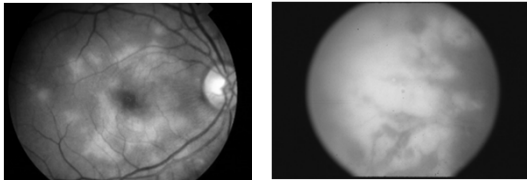


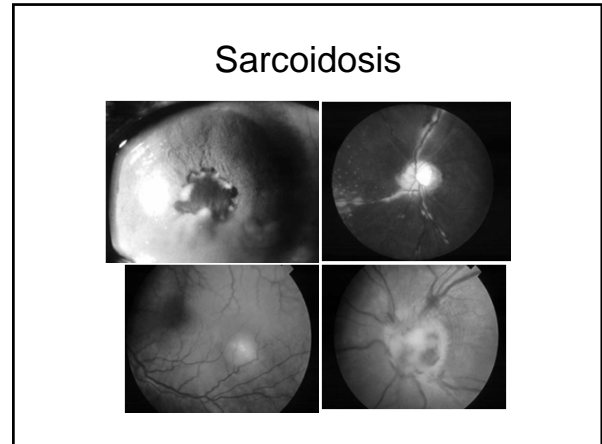
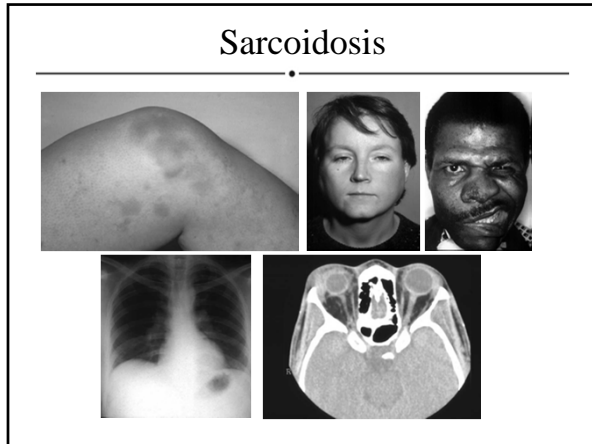
Keratic precipitates Hypopyon

Complications of Uveitis



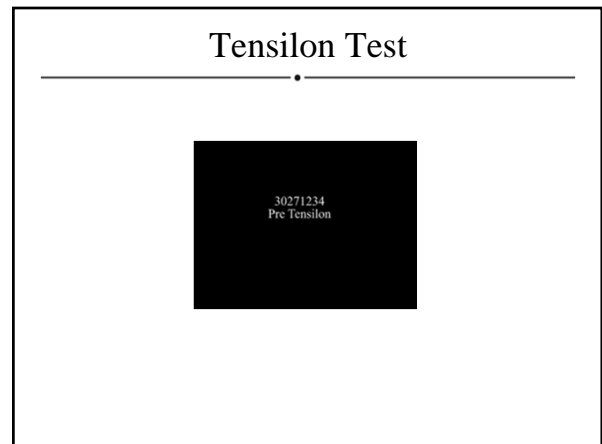
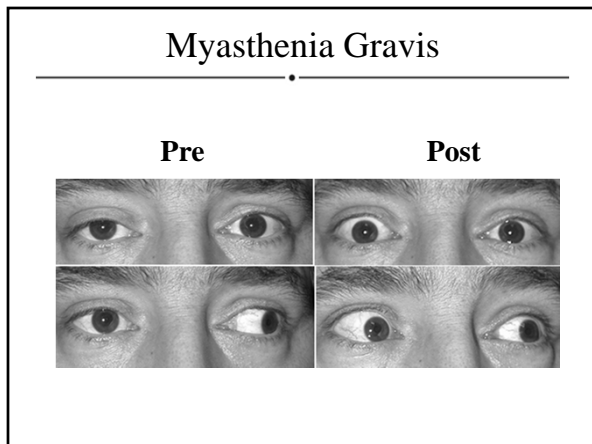
Posterior Uveitis





- ### Uveitis
- Infectious
 - Toxoplasmosis
 - Syphilis
 - Lyme
 - Viral
 - TB
 - Herpes zoster
 - Nematodes
 - CMV
 - Toxocara canis (dog roundworm)
 - Autoimmune
 - Ankylosing spondylitis
 - Reiters syndrome
 - MS
 - Inflammatory bowel disease
 - Sarcoidosis
 - Vogt Koyanagi Harada
 - Vasculitis
 - Behcets
 - Idiopathic
 - Masquerade syndrome
 - Lymphoma
 - Ocular ischemia
 - Retinoblastoma

- ### Scleritis
-
- 50% with systemic disease
 - Rheumatoid arthritis
 - Wegeners
 - Polyarteritis
 - Lupus
 - Relapsing polychondritis.
- A photograph showing a patient's eye with scleritis, characterized by a localized area of redness and swelling of the sclera.

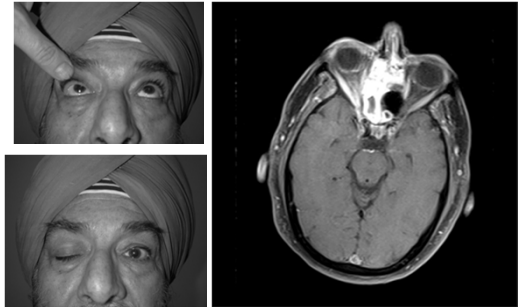


Orbital Cellulitis

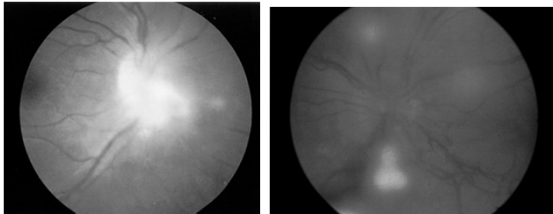


- Bacterial infection of the orbit secondary to sinusitis
- Erythema, swelling, proptosis, Ophthalmoplegia , vision loss
- fever, leucocytosis

Infection: sinusitis

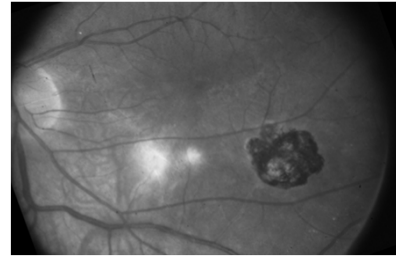


Candida endophthalmitis



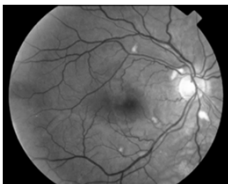
- Opportunistic infection
- Diabetics, burn patients, chronic iv antibiotics, iv drug abuse, patients receiving parenteral nutrition

Toxoplasmosis

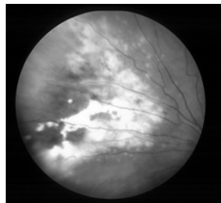


- Protozoan infection due to *t. gondii*; Host: cat
- Ingestion of contaminated uncooked meat or reactivation from prior transplacental in utero exposure

HIV



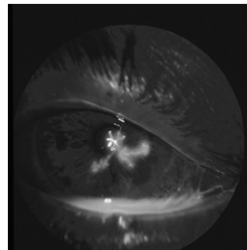
HIV retinopathy



CMV retinitis

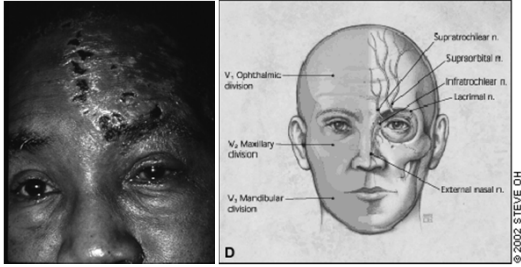
- usu seen with CD4 <50
- Rx. Gancyclovir, Foscarnet, Cidofovir

Herpes Simplex Keratitis



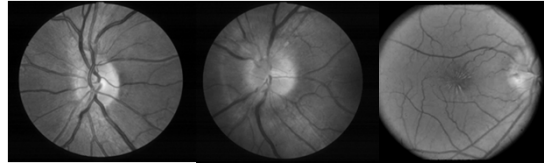
- Intracellular DNA virus
- Usually Type I
- Primary infection usually in children
- Neonatal (type II)
- Recurrent forms (type I), trigeminal ganglion reservoir

Zoster ophthalmicus



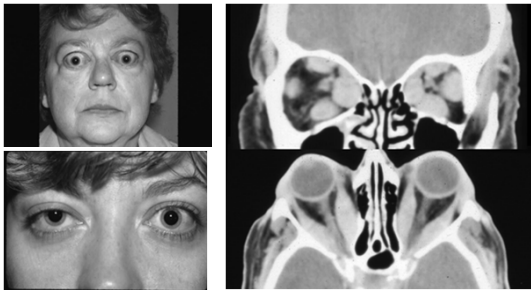
Optic Neuritis

Acute, painful vision loss with decreased acuity, abnormal color vision, APD and Central scotoma on visual fields. Fundus findings consist of three types:

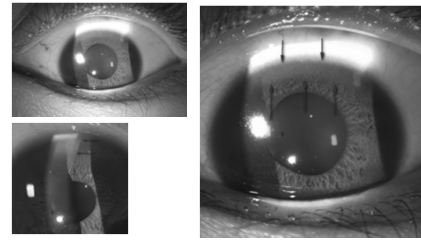


Retrobulbar Optic neuritis MS, Idiopathic
 Papillitis MS, Virus
 Neuroretinitis Virus

Dysthyroid orbitopathy

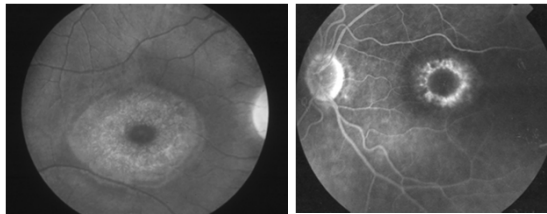


Keyser-Fleischer Ring



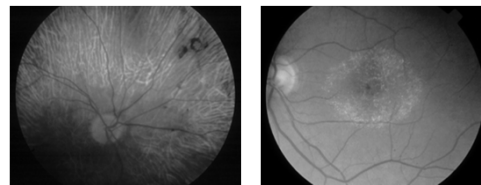
Wilson's disease
 Inherited disturbance in copper metabolism
 Neurological problems especially basal ganglia

Bulls eye maculopathy



Placquinil (chloroquine) : risk of toxicity with cumulative doses of 300 gm

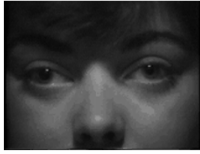
Toxic retinopathies



Phenothiazines

Tamoxifen

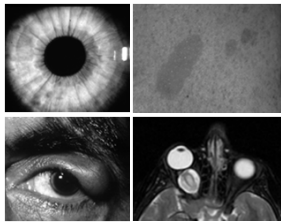
Toxic reactions



Genetic Disorders

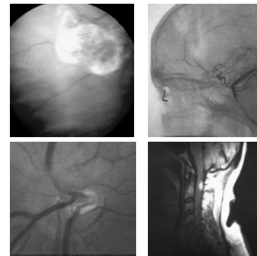
- Phakomatoses
 - Neurofibromatosis
 - Tuberous sclerosis
 - Von Hippel Lindau
- Mitochondrial myopathies
- Coloboma
- Down's syndrome
- Marfan syndrome

Neurofibromatosis



- Dominantly inherited
- 3:10,000
- Lisch nodules of the iris
- Café au lait spots (>5)
- Cutaneous neurofibroma
- Optic n gliomas
- Intertriginous freckles
- osseous lesions (sphenoid dysplasia)

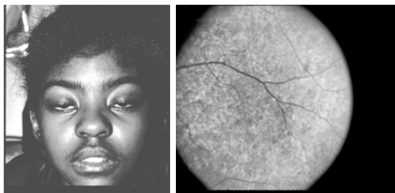
von Hippel Lindau



- Dominantly inherited
- Capillary angiomas of the disc and retina
- CNS tumors : hemangioblastomas
- Abdominal or visceral tumors e.g. renal carcinoma, pheochromocytomas

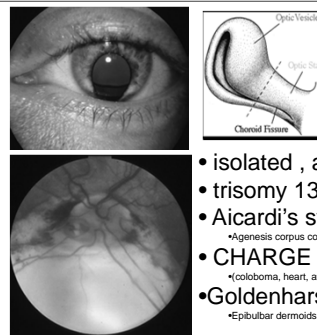
Kearns-Sayre

Chronic progressive external ophthalmoplegia



- Mitochondrial DNA disease
- Chronic progressive external ophthalmoplegia
- Cardiac conduction disturbance
- Retinitis pigmentosa

Coloboma



- isolated , autosomal dominant
- trisomy 13
- Aicardi's syndrome
 - *Agenesis corpus callosum, EEG abnrl, choroidal lacunae, retardation
- CHARGE
 - * (coloboma, heart, atresia, retardation, genital, Ear)
- Goldenhars syndrome
 - * Epibulbar dermoids, maldevelopment ears, mouth and jaw