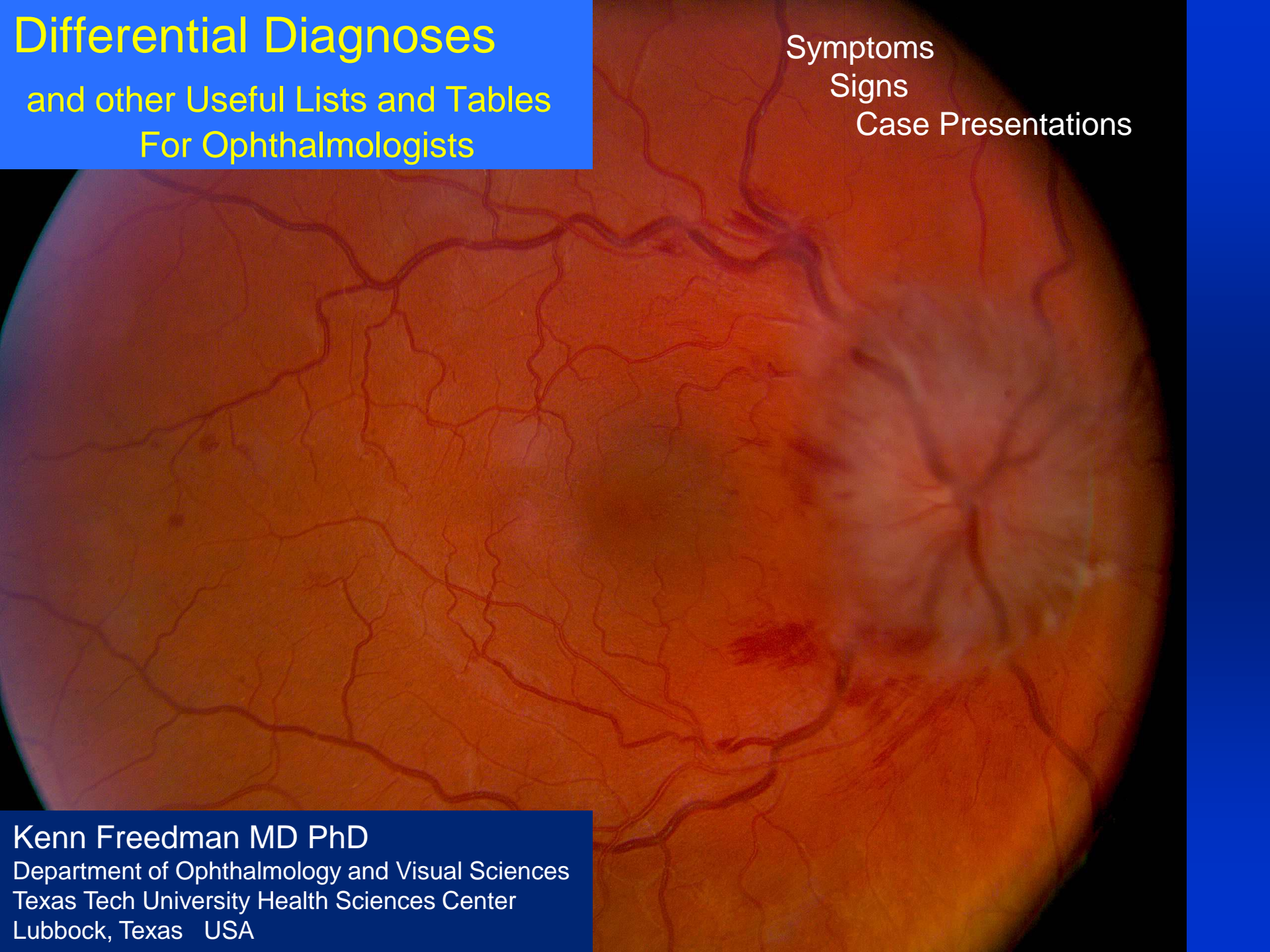


# Differential Diagnoses

and other Useful Lists and Tables  
For Ophthalmologists

Symptoms  
Signs  
Case Presentations



Kenn Freedman MD PhD  
Department of Ophthalmology and Visual Sciences  
Texas Tech University Health Sciences Center  
Lubbock, Texas USA

# Disclaimer and Acknowledgments

*The differential diagnoses and lists contained herein are not meant to be exhaustive, but are to give in most cases the most common causes of many ocular / visual symptoms, signs and situations. Included also in these lists are also some less common, but serious conditions that must be “ruled-out”. These lists have been based on years of experience, and I am grateful for God’s help in developing them. I also owe gratitude to several sources\* including Roy’s classic text on Ocular Differential Diagnosis.*

*This of course will continue to be a work in progress and any concerns or suggestions as to errors or omissions or picture copyrights will be considered. Please feel free to contact me at [kenn.freedman@ttuhsc.edu](mailto:kenn.freedman@ttuhsc.edu)*

*Kenn Freedman*

*Lubbock, Texas*

*October 2018*

\* Please see references at end of document

# Use of this Presentation

*The lists are divided into three main areas*

- 1. Symptoms*
- 2. Signs from the Eight Point Eye Exam*
- 3. Common Situations and Case Presentations*

*The index for all of the lists is given on the following 3 pages. The lists follow in the presentation in the order shown in the index. Each entry in the index (blue) is also a link and when clicked will take to you that specific list in the document.*

*If you want to go to another list within the document you will find a link on each page (shown below) which will take you back to the original index of links*

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*At the end is also a list of abbreviations used in the presentation*

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# 1. Symptoms

Loss of Vision

Transient Visual Phenomena

Floaters

Flashes, Photopsias

Diplopia – Monocular and Binocular

Oscillopsia

Night Blindness

Transient Visual Loss

Photophobia

Headache

Eye and Face Pain

Epiphora

Foreign Body Sensation

Problems Opening Eyes

Chronic Red Eye

## Terms to describe visual loss

*Blurred / Darkening / Fading* – loss of acuity or visual field

*Cannot Focus* – can apply to decreased VA, but also sometimes diplopia

Loss of Function: Reading, Driving, Disorientation

## Loss of Vision (General Considerations)

### Color Vision Loss

**Red-Green** (Protan and Deuteran)

- non-specific, seen with Hereditary, Maculopathies, Retinopathies and some acquired optic neuropathies

**Blue- Yellow** (Tritan)

specific for vascular retinopathies, papilledema, glaucoma and ADOA

**Monochromatic** – hereditary – or end stage of any condition above

### Contrast Sensitivity

Decreased contrast sensitivity often decreases before VA decreases. Cause includes causes of decreased VA.

In light of the multitude of etiologies leading to decreased contrast sensitivity, contrast sensitivity tests are useful in the characterization and monitoring of dysfunction, and less helpful in diagnosis of disease.

## Considerations

**Loss of Visual Acuity from:** Optical Errors – regular and irregular  
Media Opacities – Cornea, Lens and Vitreous  
Retinopathies and Optic Neuropathies  
CNS – Amblyopia, CVA

**VF loss – Scotomas / Anopsias** - visual pathway lesions (retina to occipital lobes)  
external obstruction (e.g. eyelids)

**Loss of Color Vision** - Hereditary and Acquired

**Loss of Contrast Sensitivity**

**Motility and Alignment Problems** - Diplopia and Visual Confusion  
Oscillopsia

**Other Visually Related CNS Disorders**

Agnosia (inability to recognize color, faces, objects)

Alexia and Dyslexia (reading problems)

Spatial and Movement Perception Problems – e.g. Visual Neglect

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1. **Negative - see Transient Visual Loss – “seeing less”**
  - loss of Visual Acuity, Visual Field or Color Vision, Blurring of Vision
  - Scotomas or Anopsias, Dimming or Loss of Brightness
  
2. **Positive - “seeing more” – distortions of / or additional objects**
  - a) Normal Phenomena – e.g. afterimages, physiologic diplopia
  - b) Entopic Phenomena – seeing own eye structures – WBCs, retinal vessels, floaters
  
  - c) Illusions - Misperceptions of external objects – *close eyes and image is gone*
    - e.g. alterations in size (aniseikonia) , shape (metamorphopsia), color (chromatopsia- tinting)
    - number (diplopia, multiplopia – consider optical and alignment problems) and
    - Palinopsia – migraine, psychoactive drugs, medications (e.g. topiramate, acetazolamide, clomiphene),
    - head trauma, lesions in parietal occipital visual pathways, metabolic
  
  - d) Hallucinations - Sensory experiences not based on incoming information - *close eyes and image is still there*
    - e.g. Flashes (see Photopsias) , formed and unformed objects
    - Psychiatric disturbances, Psychoactive medications and Rx drugs (see ocular effects of medications)
    - Cortical Lesions – Palinopsia
    - Charles Bonnet Syndrome (in cases of severe loss of vision- e.g. ARMD, Optic atrophy)
    - Migraine Phenomena (Aura’s etc.)

# Floaters

- Vitreous Syneresis
- R/O Retinal Detachment
  - Especially in the case of new floaters!*
- Vitreous Detachment (e.g. PVD)
- Vitreous Hemorrhage
- Posterior or Intermediate Uveitis
- Other sources of Vitreous Cells
  - e.g. Masquerade Syndrome for Uveitis: Lymphoma or Tumor (RB, Melanoma)
- Other Unusual Causes (in Vitreous)
  - Asteroid Hyalosis, Amyloidosis, Cholesterol Crystals- Synchrony scintillans



# Flashing Lights Photopsias



## Monocular

### Vitreo-Retinal Traction

Normal – rapid eye movements (dark), oculo-digital stimulation

Posterior Vitreous Detachment (PVD)

*R/O Retinal Tear and Detachment*

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### Retinitis / Choroiditis

Outer Retinal Disorders: MEWDS, AIBSES, Multifocal Choroiditis, etc

Retinal Ischemia: Impending CRVO, DR, OIS

Optic Nerve Disease: AION- (*Pre- , during and Post*), Papilledema

### Ocular / Retinal Migraine

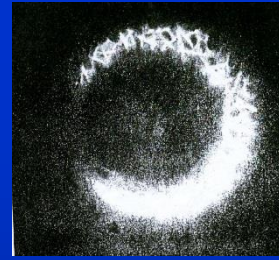
#### Optical “Dysphotopsias”

(Not true Photopsias, optical effects)

- IOL small diameter/ larger pupil; edge glare  
(may see a shadow or light in temporal field)
- IOL shutter early post op period
- RGP edge glare
- Posterior Capsule folds - can produce a Maddox Rod effect \*

Photopsias Continued →

# Flashing Lights Photopsias



## Binocular

Migraine Aura

Epilepsy – Occipital Lobe epilepsy can mimic migraine

CNS lesion – Tumor, MS, AVM, Ischemia and CVA\* (often expect VF loss also)

Vertebro-Basilar Insufficiency

Non-Ketotic Hyperglycemia

Other: e.g. Midbrain Infarct

## Other Monocular or Binocular

Retinitis Pigmentosa

CARS, MARS

Persistent Positive Visual Phenomena in Migraine (PPVPM)

R/O Altered Mental States and Psychoactive Drugs

(Tricyclic Antidepressants, dopaminergic, adrenergics, anticholinergics, beta blockers, street drugs)

Notes: Migraine onset later in life is possible

Snow like pattern (TV) – think of Outer retinal disorders, PPVPM

\*41% with Retrochiasmal Infarct have Photopsias – many not aware of VF Loss

THUS IS IT A GOOD RULE IN GENERAL TO GET **VF TESTING** IN PHOTOPSIAS

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Use **Cover Testing** to Separate into  
Monocular and Binocular Diplopia

**Monocular Diplopia** – Diplopia noted in just one eye at a time\*

Refractive Error – uncorrected, astigmatism – regular or irregular

Optic Irregularities – tear film, cornea, iris/ pupil, lens, vitreous

Only Rarely – Retinal or CNS

**Binocular Diplopia** – *Diplopia noted when binocular, and disappears when you cover either eye*

1. Optical – e.g. Unnecessary or Unwanted Prism in glasses - prescribed or induced  
(e.g. downgaze in patient with anisometropia)

2. Due to Ocular Misalignment – Strabismus

CNS – Supranuclear Palsies (e.g. INO, Skew)

or Cranial Nerve Palsies (e.g. Ischemic, MS, Compressive)

Orbital – e.g. Tumor, Pseudotumor, EOM Restriction: Graves Ophthalmopathy, Trauma, Depositions (Amyloid/ MM, WBC) age related degeneration of Orbital Tissues/ Pulley System → Cyclo-vertical deviations

Systemic – e.g. Myasthenia Gravis

Decompensated Strabismus – e.g. Intermittent XT, Monofixation Syndrome, Fixation Switch Diplopia



# Monocular (Usually Optical) Diplopia

## 1. Uncorrected Refractive Error

e.g. regular astigmatism, wrong glasses or CTLs



Optical: Often have this “Ghost Image”

## 2. Corneal problem/ distortion

e.g. Asymmetric and Irregular Astigmatism

(keratoconus, trauma, CTL warpage, - see complete list)

Corneal deposits or dystrophies

Transient corneal deformation on downgaze by LL

## 3. Cataract, Other Media Opacity

e.g. PSC or Oil Droplet NS, Vitreous

## 4. Other Optical

IOL Related - e.g. PCO or Dislocation of IOL

Iris – Polycoria, defects

## 5. Retinal / Neurologic (not optical, not common)

Macular Disorder: ERM, ARMD, etc.

CNS- e.g. MS, Palinopsia

Functional

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# Binocular Diplopia – Causes of Ocular Misalignment

## 1. Neurologic

Supranuclear – e.g. Inter Nuclear Ophthalmoplegia (INO)

Cranial Nerves – 3,4,6 (Palsy or rarely spasm)\*

Structure Abnormality – e.g. Chiari Malformation, Posterior Fossa Mass

Systemic - e.g. polyneuropathy GBS, MFS



Two distinct images

## 2. Myogenic

Systemic – e.g. Myasthenia Gravis, Mitochondrial Myopathies,  
Medications (e.g. Fluoroquinolones, Statins, ...)

Local – Muscle Damage and/ or Contracture , Previous Surgery (recession or resection)

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## 3. Orbital Disease

e.g. Graves Disease, Orbital Fracture, Orbital Tumor, Orbital Inflammation, Conjunctival Scarring

With aging - degeneration of Orbital Tissues and *Pulley System* → Cyclo-vertical Deviations

## 4. Loss or Distortion of Vision

Foveal Displacement / Traction (e.g. ERM) can induce binocular diplopia (Central Peripheral Rivalry)\*\*

Sensory Deviation (but rarely diplopia) – e.g. Cataract, Optic Atrophy, ROP, Retinal Dystrophy

## 5. Strabismus - Primary, Familial, *Decompensated* – diplopia infrequent (e.g. suppression)

e.g. Congenital / Infantile Esotropia, Intermittent Exotropia, IO Overaction, Accommodative Esotropia,  
Monofixation Syndrome , Fixation Switch

## 1. Transient Posterior CNS Circulation Ischemia

(Careful eye exam does not reveal any ocular motility or neurologic findings, or signs of eyelid / orbital disease).

- **Vertebro-basilar Insufficiency**: Embolic (Cardiac, Plaques), Vertebro-basilar Stenosis, Subclavian steal
- Decreased cardiac output – e.g. **Arrhythmias**, Heart failure

## 2. Incipient Neurologic, Orbital or Myogenic Disease

(Condition early in development does not have obvious manifestations, and a careful search for more subtle signs will be needed)

e.g. Early CNP, INO, MS, Graves Orbitopathy, Orbital Pseudotumor, MG, High ICP, GCA, Ophthalmoplegic Migraine, Myotonic Dystrophy

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## 3. In some of the above and other conditions diplopia may only be noticed intermittently in eccentric positions of gaze or at near. e.g. early 6<sup>th</sup> CNP, Convergence Insufficiency (near), and orbital conditions like TED or Myositis.

## 4. Decompensating Strabismus - With increasing angle of deviation, possible moving out of suppression scotoma at times, e.g. Intermittent Exotropia, Monofixation Syndrome. Also Phorias that “escape” fusion – breakdown periodically, due to aging, fatigue, CNS depressants, Bitemporal VF loss (Need to check carefully for phorias on ACT)

## 5. Other: **Consider Dry Eye, Convergence Insufficiency, Intracranial Hypotension**

**Ocular Neuromyotonia (ONM)**, Spasm of Near Triad, **Superior Oblique Myokymia**,

Toxicities (Drug abuse, toluene, Wernicke's)

Medications (Anesthetic injections – brainstem circulation, Meds that induce / mimic MG)

Metabolic (some drugs at peak levels, High K+, hepatic encephalopathy)

Transient Optical / Monocular Diplopia possible (e.g. transient corneal deformation on downgaze by LL)

**Exercised Induced Transient Diplopia** (case report of transient ET)

# Oscillopsia



- *Acquired Nystagmus* (see list) , but not Congenital  
CNS lesion or inflammation, Metabolic, Drug or Toxin Related
- Other Involuntary Eye Movements- e.g. Opsoclonus
- Superior Oblique Myokymia  
Idiopathic, Trauma, Vascular Compression
- Vestibular Problem
- Lid Orbicularis Myokymia
- Head Tremor or “Titubations” (MS)
- IOL - loose - “Flutter”
- Intermittent Exotropia?

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Before considering true Nyctalopia

## Consider other problems people encounter at night:

Uncorrected refractive errors maybe first present as night problems

e.g. Early Myopia, Latent Hyperopia

Pupils: Miotic Pupils (e.g. Pilocarpine , surgical) - limit light in

Normal Pupillary Dilation in dark can bring out any optical aberrations

e.g. uncorrected refractive error, cataract, corneal

Glare and Dazzle – symptoms of optic aberrations, mistaken for night blindness when driving

### 1. Congenital Stationary Night Blindness\*

### 2. High Myopia

### 3. Progressive Chorio-retinal disorders:

Retinitis Pigmentosa - various forms including Retinitis Punctata Albescans

Previously undiagnosed LCA with incomplete penetrance

Chorioretinal degenerations- Choroideremia, Gyrate atrophy

Medications: - phenothiazines, chloroquines, quinine

Vitamin A Deficiency

Siderosis and other Toxicities

CARS and MARS (Cancer and Melanoma Associated Retinopathy Syndrome)

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### 4. Other: Advanced glaucoma, S/P PRP, other retinopathies (Pigmentary, vascular, etc.)

Optic Neuropathies could present with Nyctalopia



# Transient Visual Loss (TVL)

## 1. Transient Visual Obscurations (TVO)

mono or binocular - few seconds

- usually from disc edema or other optic nerve problem (sheath meningioma, impending AION or GCA, Drusen, papilledema - *high ICP, sometimes low ICP – Intracranial Hypotension*) or ischemia- e.g. Postural hypotension, anemia

## 2. Amaurosis Fugax

- monocular - lasting 5-10 minutes, painless
  - usually thrombo-embolic and need to consider carotid or cardiac source

## 3. Vertebro-Basilar Insufficiency

- binocular – minutes to seconds, maybe have also diplopia or other brainstem Sx

Causes: embolic, Vertebro-basilar stenosis, subclavian steal, decreased cardiac output – e.g. arrhythmias

## 4. Migraine Phenomenon

- binocular, usually 20-30 minutes, associated with or without headache, photopsias / scintillating or fortification Scotoma

## 5. Other Considerations - hours to days:

Monocular (angle closure attack, Hyphema, corneal edema)

Transient Cortical Blindness (binocular)

Transient Myopia – S/P Blunt Ocular Trauma (monocular); Medications such as Topiramate (binocular)

Key History Points:

*Mono or Binocular?*

*Time Frame*

*Pain?*

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# Monocular TVL

## Carotid Disease

- Emboli or plaque coupled with hypoperfusion
- Carotid dissection (pain, Horner's)
- Ocular Ischemic Syndrome (OIS) – TVL with /after exposure to bright light

## Heart Disease

- Hypoperfusion or source of emboli
- Valvular disease, endocarditis, arrhythmia, cardiomyopathy, Atrial myxoma

## Other Vascular

- Aortic Arch (e.g. Takayasu Disease)
- Primary or Secondary Postural Hypotension
- Impending ION, *e.g. GCA – TVL proceed AAION*

## CNS / Neurologic

- Cortical Ischemia ( though usually binocular), Uhthoff's Phenomenon, Epilepsy

## Vasospasm (Dx of exclusion – needs workup)

- Retinal / Ophthalmic migraine (Hx of Migraines)
- Idiopathic Monocular TVL – Retinal artery **Vasospasm** in younger adults

## Think also of Transient Intra-Ocular Problems:

- Hyphema, Elevated IOP, Corneal Edema, Angle Closure Glaucoma, Dry Eye, UGH Syndrome, Reduced Ocular Perfusion, Transient Myopia

## Other

- Hypercoagulable States or High Viscosity States
- TVO – monocular disc problem – *e.g. edema, drusen, papilledema*
- Orbital Tumor – Gaze Evoked TVL
- Exercise Induced Visual Loss
- Transient Monocular Vision Loss on Awakening (Benign- JNO 2017; 37:122)

# Binocular TVL

Could be any of problems listed at left, but think primarily of:

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## Migraine Aura

## Cortical Ischemia

e.g. Vertebro-Basilar Insufficiency

## Papilledema

## Medications: Digitalis, Viagra

## Transient Cortical Blindness

- Pre-eclampsia
- Post-ictal states
- Metabolic: e.g. Hyperglycemia
- Trauma, Contusion Occipital Lobes

## Migraine Mimickers

- Lesion- AVM or tumor
- Occipital Seizures – can last up to days
- SLE, Meningitis, SAH

- workup can include: **Carotid** (U/S, MRA, CTA, etc.), **Cardiac** (Echo (standard or TEE), EKG), **CNS ischemia** (MRI with DWI), **Vascular / Inflammation** (GCA-ESR, CRP, CBC/Platelets), **Hypercoagulable / Hyperviscosity** – Anticardiolipin, Antiphosphotidyl choline, ANA, PTT, SPE, VDRL, Protein S and C

# Photophobia (pain with bright lights)

Think about Ocular and Intracranial Trigeminal Irritation



## Ocular Trigeminal Irritation

- Uveitis and other ocular inflammatory: Keratitis, Graft rejection – relief by topical cycloplegics?  
even some patients just with external disease – Dry Eye, Blepharitis, Pterygium, Graves Ophthalmopathy
- Corneal Epitheliopathy or Neuropathy: Relieved by topical anesthetic?

*Any ocular or visual abnormality could be associated with this symptom*

## Intracranial Trigeminal Irritation

- Meningitis, Subarachnoid Hemorrhage, Trigeminal Neuralgia, Pituitary Apoplexy, Intracranial Hypotension  
Parasellar tumors - Basal meninges of sella richly innervated and tumors in this area can give photophobia

*(In patients with normal eye exam consider: Internal Trigeminal Irritation, Achromatopsia, Optic Neuritis, Migraine)*

## Other

- Migraine, BEB, some Psychiatric: Anxiety / Depression, PSP, TBI, Thalamic lesions
- Dilated Pupils, Iris Atrophy, Aniridia – (too much light), S/P CE
- Retinal?: Albinism, Achromatopsia, Photoreceptor Degeneration, Cone Dystrophy, RP,
- Optic Neuritis, Papilledema
- Some Older and Fair Eyed Patients
- Photo - Oculodynia Syndrome: photophobia without signs of inflammation; h/o surgery or trauma, sympathetic in nature\*
- Medications: Haloperidol, Barbiturates, Benzodiazepines, Chloroquine, Lithium

## Glare or Dazzle – sometimes mistaken for photophobia

Think of: Cataracts, Dilated Pupils, Disorders of Light Adaptation (pupils or cone disorders), Thalamic Infarct

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# Headache



## Things in Particular an Ophthalmologist should think of and look for\*

Asthenopia – with near vision  
High IOP - including Angle Closure  
Uveitis, Scleritis  
Orbital Tumor  
Orbital Cellulitis / Pseudotumor  
Subclinical / Occult Dacryocystitis  
Dissection of Carotid artery – look for **Horner's**  
Chiari Malformation – HA, ET or downbeat nystagmus  
Pituitary Tumor or Apoplexy  
Other Parasellar Tumor  
Cranial Nerve Palsy – even Microvascular  
Aneurysm Compression / **SAH** - (e.g. **3<sup>rd</sup> CNP**)  
High ICP - Papilledema  
GCA (Temporal Arteritis)  
Optic Neuritis  
Carotid Ischemia / Ocular Ischemic Syndrome  
Supra Orbital Neuralgia – palpate Supra -Orbital notch

### New Headache and Anisocoria:

Horner's (Carotid Dissection)  
3<sup>rd</sup> CNP – Aneurysm  
Angle Closure Glaucoma  
Cluster Headache

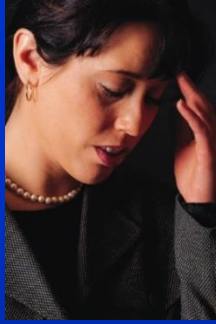
### Also Think of:

Tension Headache  
Migraines  
Medication Overuse Headaches  
Trigeminal Autonomic Neuralgia\*  
Sinusitis  
Ear or Mastoid Infection  
Meningitis  
Post-Traumatic HA  
Post Concussion Syndrome  
Post Herpetic Neuralgia  
Fibromyalgia  
Trigeminal Neuralgia  
Occipital Neuralgias  
Cervical Neuralgias  
Dental Problem  
Other Intracranial Tumors  
Nasopharyngeal Carcinoma  
Low ICP (orthostatic HA, see intracranial hypotension)  
Sub-Dural hematoma  
Sub-Arachnoid Hemorrhage (SAH)  
See Also - List for Eye / Periorbital Pain

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\* Includes: Cluster HA, Paroxysmal Hemicrania, etc.  
Unilateral, Horner Syndrome, Conj injection, epiphora

# Eye and Peri-Orbital Pain



First do a good **8-point eye exam** looking for:

refractive error      VF loss  
high IOP              ocular and orbital ischemia  
inflammation        Neoplasia

Then consider eye and adjacent problems with maybe more subtle findings, e.g.

Asthenopia  
Recurrent Erosion Syndrome  
Occult / Sub-clinical NL infection: Dacryocystitis, Canaliculus  
Supraorbital Neuralgia  
Tenosynovitis - trochlea  
Chronic uveal irritation from IOL  
Ocular Ischemic Syndrome  
Orbital Infarction Syndrome  
Occipital CVA – 15% have some referred pain  
Dental or Sinus Disease, Ear Infection  
Early CN Palsy – e.g. Diabetic 3<sup>rd</sup> or 6<sup>th</sup>  
Infarction – Thalamus (sensory center) , Medulla (Wallenberg)  
Trigeminal Neuralgia  
Post- Herpetic Neuralgia  
Raeder's Syndrome – Horners Syndrome, rhinorhea, upper facial and scalp pain  
Carotid dissection – pain, Horner syndrome  
Ramsay Hunt Syndrome: Zoster of external auditory canal, facial n. palsy  
TMJ Syndrome - pain not limited to jaw joint region  
(Temporomandibular Joint)

Both associated with carotid obstruction and can have dull aching pain

Consider also problems that are **early** in their course and subclinical - no signs yet on presentation

## Early :

Uveitis, Corneal Graft Rejection  
Optic neuritis, myositis, - pain on eye movement  
Orbital: pseudotumor, post. scleritis, cellulitis, Mucor  
Dacryocystitis, LG malignancy  
GCA  
HZO –Prodrome - can be in a lot of pain, can mimic GCA

## **Other Considerations:**

Fibromyalgia  
Unilateral HA- migraine or cluster headache  
Referred orbital pain:  
occipital neuralgia, cervical disease/ neuralgia,  
Chiari Malformation, nasopharyngeal CA, Occipital CVA

## Pain on Eye Movement

Optic Neuritis  
Orbital Inflammatory Process  
Brown's Syndrome – Trochleitis  
Posterior Scleritis

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## Epiphora is a very non-specific Symptom

Before Direct Assessment of the Nasolacrimal Drainage System

Consider External Conditions or other irritants that can evoke tearing

# Epiphora (Tearing)



e.g. Uncorrected Refractive Error

Ocular Allergies

Chronic Blepharconjunctivitis, Dry Eye, Corneal FB or Abrasion, RES, Keratitis, Uveitis, etc.

Glaucoma – High IOP

Trichiasis, Lid Foreign Body (e.g. concretion)

Lid Malpositions (Entropion, Ectropion , Punctal Eversion, Retraction , Centurion Syndrome)

Lower Lid Laxity (sometimes lid tightening procedures can stop Epiphora)

Dermatochalasis – “Upper Eyelid Wick Syndrome” – JAMA Oph 2012;130:1007

7<sup>th</sup> Nerve Palsy (poor pump function and lid laxity)

Jaw winking

Crocodile tearing (e.g. after Bell’s Palsy)

Lacrimal Gland inflammation, mass

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## Nasolacrimal obstructions

- punctal stenosis or conjunctiva blockage or megalocaruncle
- canalicular stenosis (e.g. canaliculitis, HSV, Radioactive Iodine, Docetaxel )
- relative obstruction- with edema of epithelium, chronic allergic, mucous fishing syndrome
- sac (stone, tumor, recurrent dacryocystitis - scarring)
- duct - previous sinus disease or surgery, nose trauma, acquired NLDO
- Congenital malformation – punctal atresia, Canalicular dysgenesis, NLD

NL probing and irrigation – if system is patent, consider punctal stenosis and see if the punctal dilation provides relief for even a few days – if so then punctoplasty maybe helpful

# Foreign Body Sensation (FBS)

Corneal or conjunctival (bulbar, fornix, or palpebral) foreign body

Foreign body on undersurface of eyelid – e.g. concretion

Trichiasis or Distichiasis

Dry Eye or Tear Film problem (lipid – Meibomian, mucous- Goblet cell, etc)

Blepharitis

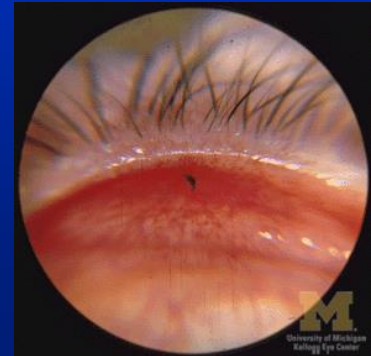
Epithelial defect(s) – Abrasion (fresh or healing) , punctate epitheliopathy

Recurrent Erosion Syndrome

Lid Problem - entropion, ectropion, lid imbrication (upper lid overrides lower lid)  
floppy eyelid syndrome, lid retraction

Corneal Ulcer - think about possible early infection

Conjunctivitis



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## Problem Opening Eyelid(s)

- Blepharospasm – eyelids squeeze (see list)
- Apraxia of Eyelid Opening\*
  - neurological – problem initiating eyelid opening
- Ptosis (though usually not complete closure)
  - Neurologic (3<sup>rd</sup> Nerve)
  - Muscle or Neuromuscular Disorder
  - Congenital, Mechanical, Inflammatory
- Dry Eyes/Blepharitis
- Recurrent Erosion Syndrome (RES)
  - eyes often stuck in the mornings



*Need to get a good history*

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# Chronic Red Eye



## Chronic Conjunctivitis

- Allergic, mucous fishing syndrome  
Irritation from smoke, chemical, topical meds
- Chronic use of any eye drop - medicamentosa  
topical anesthetics, *preservatives* even in ATs  
“red out drops”, atropine, antivirals, alpha agonists
- Chlamydial – AIC, Trachoma
- Molluscum Contagiosum
- Parinaud's Ocular glandular syndrome – cat scratch,  
tularemia, mycobacterial
- Blepharoconjunctivitis, Acne Rosacea
- Superior Limbic Keratoconjunctivitis (SLK)

**Conjunctival Mass, Tumor** – Pingueculae, Pterygium  
Papilloma, OSSN, infiltrative malignancy  
(e.g. *sebaceous cell CA*, *Lymphoma*)

## **Chronic Ocular Inflammation:**

Corneal: Stromal Keratitis, Neurotrophic keratopathy  
Uveitis – Ciliary Flush  
Dry Eye

## **Two important points:**

1. Don't just think infection as most are self-limited and need to think about some other process.
2. Don't let corneal signs focus you too much on the cornea, e.g. chronic epithelial defects can be a sign of chronic dry eyes, chronic allergic disease, chronic eyelid problems, Neurotrophic, etc.

## Orbital Disease

- TED - Congestive Stage
- IOIS – including *posterior scleritis*, dacryoadenitis, myositis
- Orbital **Tumors** including **LG**, **Lymphoma**,...
- Carotid Cavernous Fistula / Dural AV Shunts

Lacrimal - NLO, Dacryocystitis, Canaliculitis (chronic)  
Lacrimal Gland Ductulitis (*look at palpebral lobe*)

Eyelid Problems - Malpositions, Trichiasis,  
Lid imbrication (upper lid overrides lower lid),  
Floppy Eyelid syndrome

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Autoimmune Disease w/ related *episcleritis*, *scleritis*

Reiter's, Wegener's, Relapsing Polychondritis, SLE,  
Sjogrens Syndrome

**OCP** , **Epidermolysis Bulosa (Symblephara)**

Graft versus Host Disease

Loss of 7<sup>th</sup> and 5<sup>th</sup> CN Function- e.g. skull based tumor

Sign of Systemic Conditions: Polycythemia, Sickle Cell,  
Fabry's Disease, Telangiectasia, Alcohol, Cannabis

## 2. Signs (Based on the 8-point eye exam)

Loss of Visual Acuity and Refractive Issues

Loss of Visual Field

Eyelids and Orbit

Motility

Pupils

Anterior Segment

IOP

Fundus



# Loss of Visual Acuity and Refractive Issues

*Think “Front to Back”  
(Glasses to Brain)*

- **Refractive Error:**

Myopia, Hyperopia, Astigmatism (Regular and Irregular)

- **Media Opacity:**

Tear Film, Corneal Opacification (Edema, Infiltrates, Deposits, Dystrophies) ,  
Cataract, Vitreous Hemorrhage or Opacities

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- **Macula:**

Edema from: RVO, Diabetic or Hypertensive Retinopathy, Post CE, Uveitis, etc.  
Detachment (Rhegmatogenous, Exudative, Tractional)  
Hemorrhage, ARMD, ERM, Macular Hole, Degenerative Processes, Dystrophies, Toxins

- **Optic Nerve:**

Edema or Atrophy from  
High ICP, Ischemia (ION), Compression, Demyelination (Optic Neuritis), Toxic (e.g. Methanol)  
Hereditary (e.g. Leber’s Hereditary Optic Neuropathy, Dominant Optic Atrophy)

- **CNS – Chiasm, Optic Tracts, Optic Radiations and Occipital Cortex:**

Demyelination, Ischemia, Hypoxia, Metabolic, Hemorrhage, Toxic,  
Tumor Compression or Infiltration

- **Other: Amblyopia, Nystagmus, Functional (Hysterical or Malingering)**

# Refractive Changes and Shifts

## Acquired *Myopia*

### or a *Myopic* Shift (**more minus**)

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Excessive or Spasm of Accommodation

Lengthening of the Eye:

Growth and Development – e.g. often during pre- and pubescent years

Scleral Buckle

Steepening of Corneal Curvature – e.g. tight radial sutures (more often induce astigmatism)

Lens Hydration Changes – (DM, Pregnancy, Toxemia)

Refractive Index Changes – e.g. Advancing *Nuclear Sclerosis* of Lens

Look for Oil Droplet Changes

CB effusion / edema → pushing lens-iris diaphragm anteriorly

Medications – e.g. Topical: Miotics like Pilocarpine; Systemic – Sulfonamides, Diuretics, **Topiramate**

Lens Subluxation- like seen in Homocystinuria, Marfan's Syndrome

Lens anterior dislocation\* - (Crystalline lens or IOL)

Ciliary Muscle Spasm – Young patients, cholinesterase inhibitors

Seen in patients with ROP, Stickler Syndrome, Congenital Glaucoma, CSNB

Transient Myopia (days) seen after blunt eye trauma

Post CE – early or late capsular dissolution syndrome

## Acquired *Hyperopia*

or a *Hyperopic Shift* (more plus)

- Loss of Accommodation (See Upcoming List)

Can cause a Hyperopic shift in existing refraction  
or can reveal latent Hyperopia

- Posterior Dislocation of Lens (Crystalline or IOL)\*

- Aphakia

- **Drugs and Medications:** Phenothiazides, Antihistamines, Chloroquine, Anticholinergics, Cannabis

- S/P RK or other corneal surgery

with subsequent flattening of Cornea Curvature

- Shortening of distance from cornea to macula

Orbital Mass pressing on Posterior Globe

Central Serous Retinopathy (CSR)

Short or Small Eye

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- Shift in Astigmatism

change in **cylinder power or axis**

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## Corneal – changes in anterior or posterior curvature:

Normal / Common Variations in Regular Astigmatism

Shift to Against the Rule Astigmatism with Aging

Eyelid Weight on Cornea – Ptosis, Chalazion, etc.

Lesions at or near Limbus inducing astigmatism – e.g. pterygium

Shifts due to Corneal Incisions (e.g. after CE, RK, AK, PK, etc.)

Radial Sutures through sclera or corneal wounds

Corneal Ectatic Degenerations (e.g. Keratoconus) – especially for high cylinder

Trauma – laceration

Inflammation (e.g. ulcer, marginal degenerations, etc.)

Contact Lens Wear (extended)

## Lens (Lenticular)

Changes in the Crystalline Lens - Physical, Metabolic or Position

IOL – shifts/ tilts in position

# Asymmetric and Irregular Corneal Astigmatism

Optical /Refractive Error arising from the Cornea that improves with pinhole testing, but cannot be fully corrected with conventional spectacle sphere and cylinder lenses. Symptoms include blurred vision or monocular diplopia

## Causes

### 1. Ectatic Corneal Dystrophies

Keratoconus\*, Keratoglobus

### 2. Corneal Warpage from

Longstanding CTL wear

Excessive Eye Rubbing

Prolonged Downgaze (e.g. reading- with deformation by LL) – usually transient

### 2. Corneal Marginal Disorders

Pellucid Marginal Degeneration

Terrien's or Mooren's Ulcers, RA

### 3. Contact Lens

Warpage

Poor Fitting CTL

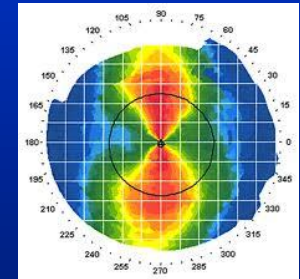
### 4. Corneal Scarring/ Irregularities after

Corneal Ulcer

Corneal Trauma / Laceration

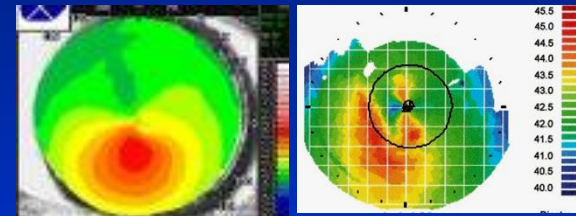
Corneal Refractive Surgery (LASIK, RK; ectasia)

### 5. Other: External Disease, Pterygia, Ocular Dermoid, LG tumor



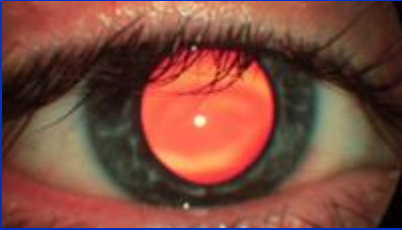
Regular

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Irregular

# Dull or Abnormal Retinoscope Reflex



- High Spherical Error – Myopic or Hyperopic
- High and / or Oblique Astigmatism
- Irregular astigmatism
  - e.g. Keratoconus, Corneal Scarring, etc.
- Media Opacity
  - Including Subtle cataract changes
    - e.g. - star / flower pedal nuclear cataracts
    - oil droplet NS changes
- Posterior Defect – e.g. Coloboma



# Poor Near Visual Acuity Despite Good Distance VA

## 1. Loss of Accommodation

- *Presbyopia* - natural loss of lens accommodation  
(onset usually in mid-forties)

### Other causes of Loss

Aphakia, Pseudophakia

Cataract

Lens Subluxation

Head Trauma

Eye and Orbital Trauma

Encephalitis and Meningitis

Midbrain Disease

Oculomotor Palsy

Tonic Pupils – e.g. Adie's Syndrome

Diabetes

Pharmacologic and Toxic agents: e.g. Atropine, other anticholinergics

Infections: syphilis

Systemic Medications: Amitriptyline

Glaucoma

Myasthenia Gravis

Uveitis

Retinal/ Scleral Treatments (laser or cryo)

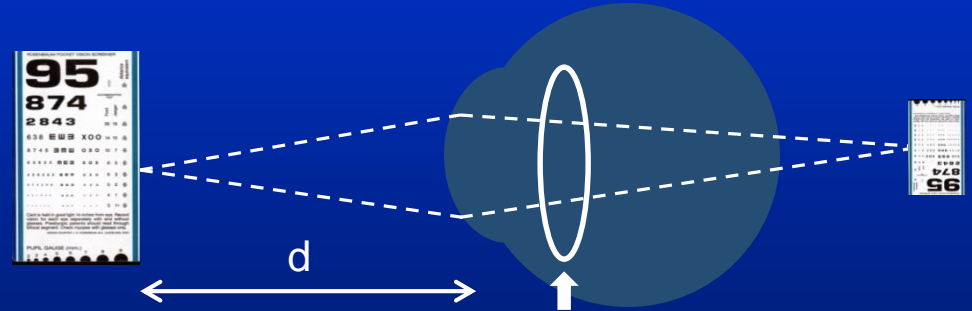
Pregnancy

Functional (Non-Organic)

Benign Syndrome of Transient Loss of Accommodation in Young Patients

( Idiopathic, lasting months – JAMA Oph 2008; 126:1643)

Other: Botulism, Diphtheria, Viral Diseases : Influenza, Chicken Pox and some other Exanthems, GBS



$1/d(m) = \text{accommodative power needed}$   
(if hyperopic need to add distance plus (+) power as well)

## 2. Other Considerations:

Central Cataract – e.g. PSC with Miosis (Recall Near Triad)

Glasses Problems: e.g. Bifocal Segment

See Also “Problems with Reading” List

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# Problems with Glasses

Patients Complaint's



1. Was Refraction / Prescription (Rx) Correct?
2. Were glasses made correctly to Rx?
3. High Refractive Error – Vertex Distance Issues  
(Try over-refraction over old glasses)
4. *Over- Minused* Correction – can happen in younger accommodating patients
5. Astigmatism – was there a significant *change in axis* from last Rx? Often not tolerated
6. Optical Center (OC) – check with respect to the pupil, PD and bifocal segment
7. Pantoscopic Tilt – e.g. minus lenses (tilt can induce cylinder)
8. Optical Aberrations – “waves” in lens sometimes happen when grinding
9. Induced Prism – causing Hypertropia and Diplopia (recall Prentice’s Rule  $P=hD$ )

OC- Optic Center  
PD- Pupillary Distance  
P- Prism Power  
h – displacement from center  
D- Diopters of Lens Power in the  
axis of concern

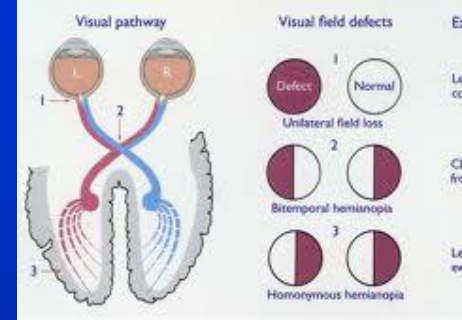
## Problems with Near

1. Bifocal Segment – not enough or too much add power  
- position: top should be a lower lid level. Some are too low
2. Progressive Bifocals - too narrow or patient has to look too far down to get full add
3. Anisometropia – with large differences in vertical induced prism → Diplopia - may need SLAB OFF

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# Loss of Visual Field

# Visual Field Defects and Localizing Lesions



- Unilateral – Retina, Optic Nerve (rarely early Chiasmal)
- Bilateral – 1. Bilateral retinal or optic nerve disease (*but usually there is little symmetry*)  
2. Chiasm or farther posterior (*usually some kind of symmetry*)
- Vertical Midline respect – *suggests lesion at Chiasm or Posteriorly*
- Horizontal Midline respect – *usually optic nerve or retinal disease, **but** can be a lesion of the optic radiations or just the superior or inferior portions of occipital lobe*
- Homonymous Hemianopsia w/ other symptoms :

Occipital Lobe – *often silent with no other non-visual symptoms*

Parietal Lobe (*hemiparesis, visual perception and spatial problems, right left confusion*)

Temporal Lobe (*seizures, formed visual hallucinations, agnosias*)

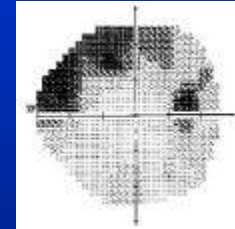
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**Recall** the Optic Nerve / Tract Exam Signs  
that suggest a lesion anterior to Lateral Geniculate Nucleus (LGN)\*:  
e.g. RAPD, Pallor / Atrophy, Edema

# VF Defects respecting the *Horizontal Midline*

Suggestive of Optic Nerve Problem

Glaucoma  
AION  
Optic neuritis  
Papilledema- chronic  
Disc Drusen and other disc abnormalities



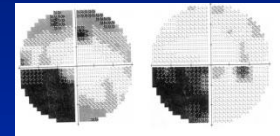
*Examples*  
*Altitudinal*  
*Arcuate*  
*Nasal Step*  
*Temporal Wedge*

Sometimes retinal pathology:

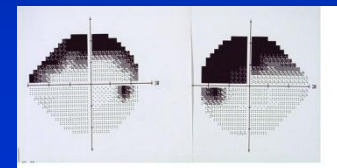
BRAO  
BRVO  
hemi-retinal vein or artery occlusion

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Some retro - chiasmal defects can respect vertical midline ***and horizontal***, but also often are bilateral and have symmetry, e.g. Specific lesions to the Parietal or Temporal lobe radiations or to the superior or inferior portions of the occipital lobes.



Superior loss from: Eyelid Ptosis, UL Dermatochalasis,  
Deep Set Eye in Orbit, Frontal Bossing  
can give appearance of horizontal midline respect

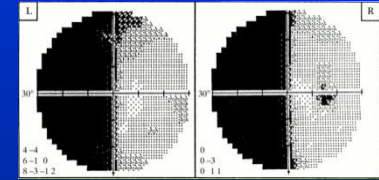


# VF Defects respecting the Vertical Midline

Suggestive of a Lesion at the Level of the Optic Chiasm or More Posteriorly

## Homonymous Hemianopsia

CVA, Tumor, Demyelination posterior to chiasm  
(See separate list)

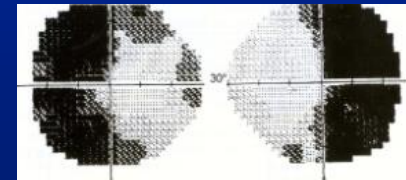


The following Defects more suggestive of lesion at or about the optic chiasm

e.g. Tumors, Hemorrhage (Apoplexy), Aneurysms

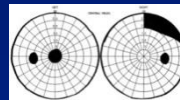
## Bitemporal Hemianopsia

Chiasmatal, Sellar and Parasellar lesions  
(See separate list)

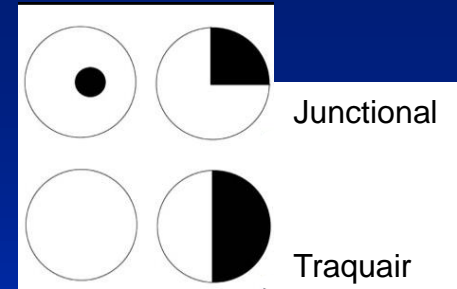


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## Junctional Scotoma



Specific lesion at junction of optic nerve with chiasm (Von- Willebrand's Knee)

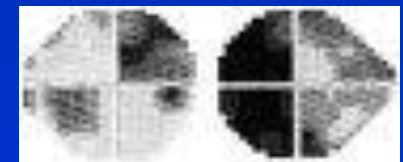


## Isolated Temporal Hemifield Defect

Junctional Scotoma of Traquair

Tilted Disc Syndrome

Unappreciated loss of central vision in contralateral eye of Junctional Scotoma



Other: Nasal Hemianopsia(s) Possible

# Visual Field Defects that Respect the Vertical Midline

# Bitemporal Hemianopsia

## Chiasmal or Supra- or Para- Sellar Lesions:

Chiasmal Tumor – e.g. Glioma

Pituitary Tumor, Pituitary Apoplexy

Other Parasellar Tumors – meningioma, craniopharygioma, Rathke's (pars intermedia) cyst, etc.

Demyelination (MS) - "Chiasmitis" (seen in ONTT)

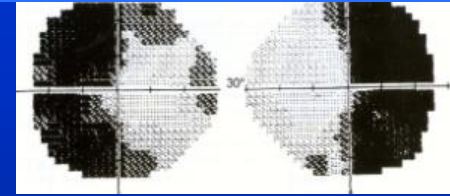
Aneurysms – ICA, branches off circle of Willis

Chiasmal Trauma from Head Injury

Trans-sphenoidal Basal Encephalocele

Other: Meningitis, Sarcoidosis, Ischemia?

Toxicities: Ethambutol



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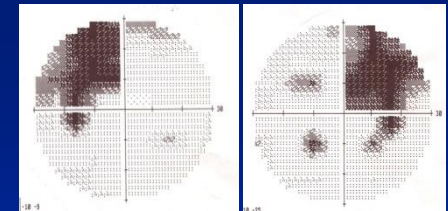
## Incomplete Bitemporal Defects

Any of above can produce this picture

Dermatochalasis with Lateral Hooding

**Tilted or Anomalous Discs** can produce temporal defects

Nasal Staphyloma(s)

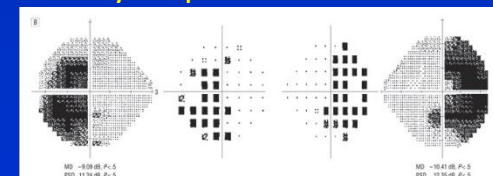


Dermatochalasis with Lateral Hooding

## Centrocecal Scotomas

Things that produce relatively large cecal and centrocecal defects can sometimes artificially respect the vertical midline and produce a Bitemporal Hemianopsia - like picture\*

e.g. Toxicities (Ethambutol, Chloroquine), Deficiencies, Hereditary Optic Neuropathies





# VF Defects respecting the *Vertical Midline*

## Homonymous Hemianopsia

Complete or Incomplete



### Lesions of

- optic tracts (bowtie atrophy)
- LGN
- temporal, parietal or occipital lobes
  - From: CVA (thrombosis, embolism, vasculitis)
  - Tumor (Compression or Infiltration)
  - MS or other Demyelinating Process
  - Trauma
  - Vascular Malformation

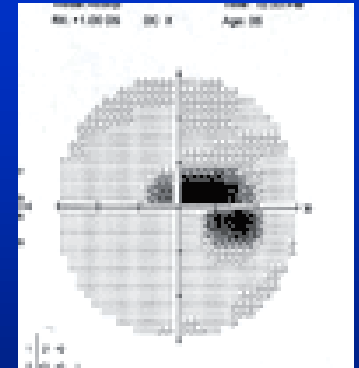
### If there is no apparent Lesions on Neuroimaging, Consider:

- Creutzfeld – Jacob Disease
- Some Degenerative Dementias : Alzheimers, Pick disease
- Subtle Occipital ischemia or hypoxia – not seen on CT or MRI
- Non-ketotic hyperglycemia or other metabolic derangement
- Migraine
- Toxic- e.g. Carbon Monoxide (see cortical visual loss)
- Functional

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## Central and Centrocecal Scotomas

- Macular Disease – ARMD, Diabetic Macula Edema, Cystoid Macular Edema and including subtle ones like CSR, Cone dystrophy
- Optic Nerve Pit with serous detachment of the macula
- Toxic Optic Neuropathies – medications, heavy metals, Methanol, Chemotherapy
- Tobacco-Alcohol-Nutritional Amblyopia
- Hereditary Optic Neuropathies e.g.. Dominant Optic Atrophy (DOA), LHON
- Orbital apex tumor or process (e.g. metastatic disease, pseudotumor)
- Sometimes compression/ infiltration : e.g. Tuberculum Sella meningioma
- Previous Optic Neuritis, AION



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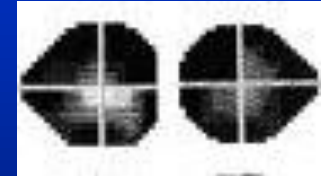
## Enlarged Blind Spot (Cecal Scotoma)

- Papilledema - early
- MEWDS, AIBSES and other outer retinopathies
- Disc Edema in association with Uveitis, Papillophlebitis
- Significant Peri-Papillary Atrophy
- Myelinated Nerve Fiber Layer about Disc
- Disc Coloboma or other disc abnormality



## Severe Constriction / Tunnel VF's

Non-Organic / Functional Visual Loss  
Hysterical (Conversion) or Malingering



Chronic Papilledema

RP or other Tapetoretinal disorders

Bilateral occipital lobe infarctions with macular sparing

CRAO with Cilioretinal artery sparing

End stage glaucoma

s/p PRP

Other: medications, toxins, metabolic (see cortical visual loss / unexplained visual loss)

# Eyelids and Orbit

## Primary- Benign Essential Blepharospasm (BEB)

Associations: Apraxia of eyelid opening, Meige's Syndrome and other cranial/cervical dystonias  
Extrapyramidal disorders (Parkinson, Huntington, and basal ganglia infarction)

## Secondary Blepharospasm

Medications: antihistamines, dopaminergics, nasal decongestants

External Disease, Foreign Body, Keratitis, Dry Eye

Consider any cause of *Photophobia* (see list)

5th CN Irritation\* – Ocular (Uveitis, etc.) or Meningeal (meningitis, parasellar tumor), Trigeminal Neuralgia

Paraneoplastic Syndrome – e.g. Anti-Hu / small Cell CA



Myotonic Dystrophy

Aberrant Facial Nerve Regeneration – after peripheral facial nerve palsy

Hemi- Facial Spasm - Low, but possible risk if CPA tumor or aneurysm

Orbicularis Myokymia - Usually only an upper or lower lid, as opposed to true Blepharospasm

Facial Myokymia - pontine glioma, MS, Neurodegenerative diseases: e.g. ALS, Huntington's Chorea

Tardive Dyskinesia - Multiple Meds can cause– not just neuroleptics (JNO 1998; 18:153)

Eyelid Nystagmus

Torrette's Syndrome

Excessive Blinking

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### May Need to Differentiate from Just a Problem of Opening Eyelid(s)

- Apraxia of Eyelid Opening

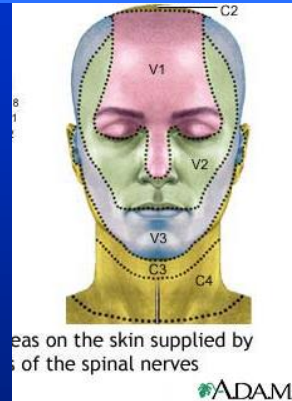
Associated with BEB, PSNP, Parkinson's, Huntington's, CNS Lesion - Frontal (and Parietal?) Lobe, Brainstem, Thalamus

- Dry Eye / Blepharitis / RES - Lids stuck to each other or cornea

- Ptosis

# Causes of Loss of or Impaired Sensation on Face and around Eye

- Damage to Branches of 5<sup>th</sup> Cranial Nerve
  - e.g. Orbital Floor Fracture – damage to V2
  - Shingles (Zoster) - most commonly V1 distribution
- *Facial Carcinomas (even occult ones)*
  - can track along nerves (*perineural invasion*)
  - e.g. BCCA, SCCA
- Tumor in Brainstem, Cavernous Sinus, Orbit or Skull base
  - e.g. CPA angle Acoustic Neuroma
- CNS (cerebral cortex or brainstem) lesion – CVA, MS, etc.
- Recall loss of Corneal Sensation can be related:
  - e.g. Keratitis – active or past Acanthamoeba, Herpetic, ...
  - See list for loss of Corneal Sensation



# Ptosis

## Levator (Dehiscence)

- Aging, Trauma, Post-op (e.g. CE), Post-Inflammation, CTL wear

## Congenital, Hereditary

- **Levator Mal-development**, Marcus Gunn Jaw Winking, Blepharophimosis (BPES)
- Congenital Cranial Dysinnervation Syndromes (e.g. Congenital Fibrosis)



## Neurological

- 3<sup>rd</sup> Nerve Palsy, Horner Syndrome
- Hemispheric Stroke (unilateral or bilateral – associated with hemiparesis)
- Migraine – Isolated Ptosis? “seen with Hemicrania Continua” – can have associated isolated ptosis
- Immune Mediated Polyneuropathies –e.g. Guillain – Barre Syndrome

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## Orbital Disease

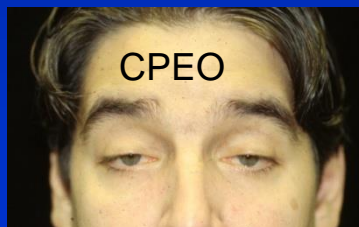
- Inflammatory: Cellulitis, Pseudotumor, Graves
- Tumor: Lymphoma, etc.

## Mechanical

- Eyelid Tumor (e.g. NF), Chalazion
- Excessive Dermatochalasis and/or Brow Ptosis
- Floppy Eyelid Syndrome (Laxity, Lash Ptosis)

## Myogenic

- **Myasthenia Gravis**, Lambert-Eaton Myasthenic Syndrome
- CPEO
- Muscular Dystrophies  
e.g. Oculopharyngeal MD, Myotonic MD



## Inflammatory

- Eyelid, Orbit, Uveitis, Conjunctivitis, Keratitis (e.g. SLK)

## Other

- Prostaglandin (Topical) Associated Orbitopathy
- Observed associations with isolated ptosis: elevated BP

## Pseudo-Ptosis

- Enophthalmos ( see list)
- Phthisis or small globe or Anophthalmos
- Blepharospasm, Dermatochalasis or Brow Ptosis Mistaken for ptosis
- Hypertropia, Hypotropia





## Madarosis (Loss of Lashes)

- R/O Carcinoma – e.g. BCCA, Sebaceous Cell CA
- Chronic infection – e.g. Herpetic, Staph, Fungal, Mites, Blepharitis
- Endocrine – e.g. Hyper and hypo parathyroid and thyroid, hypopituitism
- Dermatoses - e.g. Dermatitis ( atopic, contact), ichthyosis, lichen planus,... (many)
- Trauma – radiation, chemical, Thermal, tattooing, surgery, cryo
- Congenital disorders - multiple
- Drugs and Toxins - e.g. Arsenic, Chemotherapy, Botulinum, ...
- Systemic Conditions – e.g. Parry-Rhombert, VKH, Lupus, Sarcoidosis,...

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## Hypertrichosis (Excess Lashes = Trichomegaly)

- multiple congenital / genetic causes
- frequent manipulation
- Paraneoplastic syndrome
- malnutrition, anorexia, pregnancy, thyroid problems, lupus, uveitis
- Drugs: prostaglandin analogs (e.g. bimatoprost)

# Eyelid Malpositions Entropion and Trichiasis

## Lower Lid Entropion and Trichiasis

Involitional (Senile) – can have spastic (orbicularis) component

Acute Spastic Entropion – after trauma or surgery

Cicatricial (see below)

Congenital / Developmental – e.g. Epiblepharon

Distichiasis – abnormal lashes growing from posterior lid margin (meibomian orifices)  
could be hereditary or from inflammatory process (see below)



## Upper Lid Entropion and Trichiasis

Mechanical – excessive Dermatochalasis

Cicatricial (see below)

Distichiasis

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## **Cicatricial Causes** (Most cases due to secondary scarring and contracture of posterior lamella )

Previous Trauma or Surgery at or near eyelid margin

Chemical Burn

HZO

Chronic Blepharo-conjunctivitis - e.g. Acne Rosacea

Trachoma

Stevens-Johnson Syndrome, Ocular Cicatricial Pemphigoid

***\*Sometimes Orbital Disease can present with eyelid malpositions***

## Lower Lid Ectropion

Senile – with horizontal laxity, check for Medial or Lateral Canthal Tendon laxity

Cicatricial (below)

Combination of both above

Paralytic – 7<sup>th</sup> nerve palsy, MG

Mechanical – Tumor or Big Festoons

Congenital – Ichthyosis; Euryblepharon – excess horizontal skin



## Upper Lid Ectropion

Cicatricial Processes (below)

Congenital – e.g. Ichthyosis

Floppy Eyelid Syndrome – Horizontal Laxity – no true ectropion

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## Cicatricial Changes (of anterior lamella)

Trauma to Eyelids and Face

Burns- Thermal and Chemical

Sun Damage, Carcinoma

Previous Eyelid and Adnexal Surgery

Chronic Inflammation: Rosacea, Atopic Dermatitis, HZO, Infection

**\*Sometimes Orbital Disease can present with eyelid malpositions**

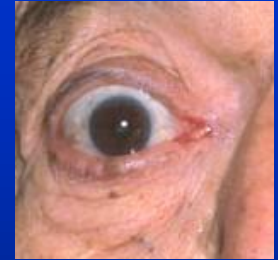
# Eyelid Retraction

Graves Ophthalmopathy- #1 – unilateral or bilateral

Other Causes of Hyperthyroidism

Other Orbital Inflammatory or Neoplastic Conditions

– Orbital Pseudotumor, FB, Granulomatous Inflammation, Neoplasm



Cicatricial Process

Skin or Posterior Lamellar (Trauma, Burns, Systemic or Local Inflammatory Disorders)



Trauma / Post-Operative

*Entrapped Inferior Rectus*

*Vertical Rectus Muscle Recession Surgery*

*S/P Eyelid or Conjunctival Surgery*

Neurologic

- e.g. Dorsal midbrain syndrome (Collier's sign) , aberrant regeneration of the 3<sup>rd</sup> CNP

Metabolic (thyroid, cirrhosis, uremia, Cushing's syndrome, hypokalemia)

Pharmacologic – sympathomimetics, corticosteroids

Congenital – persistent or periodic unilateral retraction reported

Physiologic / Normal Variant – about 2% of population has MRD>5.3mm

Pseudo-retraction

– Contralateral Ptosis (Herring's Law)

- Proptosis

- Lower Lid Laxity

- Large Myopic Eye, prominent glaucoma filtering bleb

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# Lagophthalmos

Inability to Close Eyelids



Neurological

Seventh Nerve Palsy

Cicatricial (Scarring)

Trauma

Burns

Surgery

Blepharoplasty, Ptosis Surgery

Tumor resection

Orbital Condition

Proptosis: Graves Ophthalmopathy, etc. ( see list)

Orbital Inflammatory or Neoplastic Processes

Myogenic – MG, Muscular Dystrophies, CPEO

Botulinum Injections

Don't Confuse with

**Lid Lag on Downgaze**

Congenital Ptosis

Graves Ophthalmopathy

Aberrant Regeneration after 3<sup>rd</sup> CNP

Neurologic and Muscular Disease

- Supranuclear Palsy

- Myotonic Dystrophy

- MG?

Post-op Upper Eyelid Procedures

Possible Sign of Other Orbital Disease

See *Exposure Keratitis*

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# Seventh Nerve Palsy

## Hemifacial Paralysis with Lagophthalmos

- Motor Strip Lesion (Upper Motor Neuron) → Contralateral Lower Face Paralysis
- Peripheral Nerve Palsy – Ipsilateral Upper and Lower Face Paralysis
  - CPA Tumor – e.g. Acoustic Neuroma
  - Other tumors – Parotid, Skull based, temporal bone, external auditory canal
  - Trauma – facial, skull base (temporal bone), birth
  - Lyme Disease – *B. Burgdorferi*
  - HIV infection
  - Central – CVA (e.g. superior cerebellar a. infarct – deafness, Horner's, 7<sup>th</sup> CNP)
    - Parkinson's
  - Ramsay-Hunt Syndrome* (Herpes Zoster Oticus)
  - Mastoiditis / Otitis – 6<sup>th</sup> and 7<sup>th</sup> CNP possible
  - External Auditory Canal and Middle Ear – surgery, tumor
  - Other - Neuro-Sarcoidosis, Leprosy, Pregnancy (3<sup>rd</sup> Trimester), MS
  - Vasculitis, DM, Uremia

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and **Bell's Palsy** (Idiopathic 7<sup>th</sup> CNP)

Most Common 7<sup>th</sup> Nerve Palsy, but better to put Bell's Palsy down at bottom the list – to make you think of other things first

A 7<sup>th</sup> Nerve Palsy is not necessarily a Bell's Palsy!

# Eyelid Mass / Lesions

## Cystic Like / Fluid Filled

Hydrocystoma /Sudoriferous Cysts – clear fluid

Sebaceous Cyst , Epithelial Inclusion Cyst – both usually have white/yellow appearance

Blister, Bulla, Vesicle (e.g. HSV), Pustule



## Pedunculated

Papilloma, Skin Tag, Cutaneous Horn

## Darker / Pigmented

Nevus

Melanoma – often irregular pigmentation and borders

Seborrheic Keratosis (SK) – sessile, stuck on appearance

Xanthelasma – yellowish – often medial canthal skin

Kaposi's Sarcoma

Some Cysts will have dark appearance clinically: e.g. apocrine cyst, some inclusions cysts

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## Nodular - Commonly at Lid Margin

Intradermal Nevus

BCCA

Hair Follicle Tumor



## Vascular

Hemangioma

Cherry Angioma – Bright red

Varix

Other: Kaposi's Sarcoma, Pyogenic Granuloma

Recall signs of Malignancies

- 1) lash loss
- 2) ulceration, bleeding
- 3) telangiectasias
- 4) irregular pigmentation
- 5) distortion or destruction of eyelid anatomy

## Crater / Ulcerated

Carcinomas (BCCA, SCCA, etc)

Keratoacanthoma

Moluscum Contagiosum



- **Don't Forget:** Chalazion, Hordeolum and their Mimics (e.g. Sebaceous Cell CA)



# Signs Suggesting Orbital Disease

- Proptosis, and other  
Globe Displacements
- Chemosis, Conjunctival vessel congestion
- Limited Motility
- Pupil Defect – e.g. RAPD
- Eyelids
  - Edema, Ecchymosis
  - Lid Mass or Fullness, UL sulcus defect (asymmetry)
  - Sometimes: Ptosis, Lid Retraction, Ectropion and Entropion
- Loss of Vision
- Fundus Findings: Disc edema, Vessel Congestion



Left hypoglobus



Lid deformity in  
orbital NF



# Eyelid Edema

## Inflammatory Appearance (red, warm, etc.)

First Consider Underlying Orbital Disease

Orbital Cellulitis, Pseudotumor, Wegener's

Graves Ophthalmopathy, Orbital Varix

Orbital Tumors that can mimic inflammatory process: Lacrimal Gland CA, Lymphoma, Lymphangioma, etc.

Lacrimal Gland – Dacryoadenitis or tumor

Sinus Mucocele

Preseptal Cellulitis

– also think of early -HSV, HZO, or erysipelas (rapid strep),

Periorbital necrotizing fasciitis ( b-hemolytic strep, staph A., pseudomonas)

Dacryocystitis / Dacryocystocele

Blepharitis

Contact Dermatitis – e.g. Neomycin, Gentamicin, **Glaucoma Drops**- Chronic Use

Urticaria / Angioedema

Conjunctivitis with contiguous lid edema

Insect Bite

Lid Tumors: Hordeolum / Chalazion, CA, **Cutaneous Lymphoma**

Melkersson-Rosenthal Syndrome – (Granulomatous inflammation)



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## Without Inflammatory Appearance, consider above but also...

Allergic Eyelid Edema

Hormonal Shifts

Systemic Disorder – Cardiac, Renal, Hepatic, Thyroid with edema

Graves Ophthalmopathy – can just have lid edema w/o inflammatory appearance

Lymphedema after trauma, surgery to lids or orbit (e.g. lymphatics in lateral canthus)

Traumatic Leak of CSF into upper eyelid (JAMA Oph 2014;312:1485)

Blepharochalasis

## Not True Edema but might mimic it:

Dermatochalasis

Hidden Eyelid or  
Sub-Conjunctival Mass

Prolapsed Orbital Fat

# Conjunctival Hemorrhage



# 1 Spontaneous

Trauma – ocular (even minor e.g. rubbing eye), orbital, head

Valsalva Maneuver (sudden venous congestion)

Sometimes Orbital Hemorrhage

Trauma, Retrobulbar Injection

Tumor: Neuroblastoma , Rhabdomyosarcoma,  
Leukemia, Lymphangioma

Acute viral/ bacterial conjunctivitis

Systemic - Clotting disorder, febrile illness, acute HTN

# Peri-Orbital Ecchymosis



Needs follow up– probably benign, but  
could be sign of underlying disease

Eyelid or Orbital Trauma  
Orbital Contusion (“Black Eye”)

**Bad Sub-Conjunctival Hemorrhage can dissect  
into eyelids**

Orbital Hemorrhage associated with:

Trauma

Retrobulbar Injection

**Neuroblastoma , Rhabdomyosarcoma** and other tumors

Amyloidosis, Multiple Myeloma

Leukemia

Sub-Periosteal Hemorrhage

Valsalva : Labor, Vomiting, Coughing

Bleeding Disorders, Liver Disease, Scurvy

Sinus Disease

Venous Congestion

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# Proptosis

Forward Displacement of the Eye

1. **Graves Orbitopathy** (#1 Cause)
2. Orbital Cellulitis
3. Orbital Pseudotumor — including Posterior Scleritis
4. Other Inflammatory - Wegener's, Sarcoidosis, see also Orbital Inflammation
5. Orbital Tumors, Sinus Tumors
6. Orbital Hemorrhage- Trauma, Lymphangioma  
Subperiosteal Hemorrhage
7. Orbital Trauma and Compartment Syndrome
8. Vascular Abnormality —e.g. Varix /Venous Malformations (Congenital)\*, Carotid Cavernous Fistula
9. Bony Orbital Malformation — e.g. congenital
10. Meningo-encephalocele
11. Sinus Mucoceles
12. Orbital Apex and Cavernous Sinus – Tumor, Inflammation, Vascular



## Pseudo-Proptosis

Large Globe- Glaucoma, High Myopia  
Lid Retraction w/o real forward displacement,  
Relative Enophthalmos

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## Age Category

## Orbital Tumors to Consider

### 1. Children



Rhabdomyosarcoma

Neuroblastoma

Lymphangioma

Capillary Hemangioma

Glioma of Optic Nerve

Dermoid Cyst

Neurofibromas (NF1 and NF2)

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### 2. Adults



Cavernous Hemangioma

Lymphoma or other WBC tumor

Orbital Venous Malformations, Varices

Optic Nerve Sheath Meningioma

Lacrimal Gland CA and other tumors

Neural and Fibrous Tumors

Adjacent Invasive Sinus Tumor

Metastasis

Granulomatous Inflammation - think about GPA

# Enophthalmos

Posteriorly Displaced Globe often with superior sulcus deformity



- Orbital Blow-out Fracture
- Congenital – asymmetry in bony orbits
- Bony defects, erosion, expansion in orbit due to:  
tumor, chronic infection, vascular malformations (*e.g. Varix*), radiation, Sinus Infection, Gorham's Disease, NF
- Neoplastic –e.g. Metastatic Scirrhous CA (e.g. Breast)
- Surgery
- Silent Sinus Syndrome
- Starvation
- Aging
- Horner's Syndrome
- Orbital Fat Atrophy
  - **Parry- Romberg Syndrome** (Idiopathic Hemifacial Atrophy, Sympathetic Input?)
  - Past inflammation, Past Trauma, Linear Scleroderma
  - Orbital tumor removal – tumor caused pressure atrophy of the fat
  - **Prostaglandin Associated Periorbitopathy** ( from Glaucoma Prostaglandin Analogues)
- Intracranial Hypotension?
- Pseudo-Enophthalmos
  - Microphthalmos, Phthisis, Contralateral Exophthalmos, Ptosis, Isolated Superior Sulcus Deformity

## Superior Sulcus Defect

- Any of Conditions listed (Enophthalmos)
- Anophthalmos
- S/P enucleation or evisceration with a too small orbital implant
- Too aggressive removal or superior fat pad with Blepharoplasty

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# Signs of Orbital Inflammation

Proptosis, Chemosis, Eyelid Edema, etc.

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Need to R/O  
Orbital Cellulitis / Malignancy  
Imaging of the Orbits

## 1. Orbital Cellulitis

## 2. Orbital Pseudotumor – Dacryoadenitis, Myositis, Diffuse, Posterior Scleritis, IgG4-RD\*

## 3. Graves Ophthalmopathy

## 4. Orbital Lymphoma

## 5. Reactive Inflammation

Foreign Body, Ruptured Dermoid Cyst, Adjacent Sinus Inflammation

## 6. Trauma

## 7. Systemic Disease

Wegener's Granulomatosis, Sjogrens , TB, Sarcoidosis, Syphilis, IgG4-ROD

## 8. Medications: e.g. Biphosphonates

## 9. Vascular etiologies

Vasculitis (e.g. GCA), CC Fistula, Cavernous Sinus or Superior Orbital Vein Thrombosis (Sepsis, Coagulopathies, etc)

## 10. Inflammatory Mimicking Orbital Tumors

Rhabdomyosarcoma, Lacrimal Gland Carcinoma, Lymphangioma

## 11. Orbital Hemorrhage

Work-up could include - CT orbits +

TED: TFTs, TSI

IOIS: CBC (Eosinophilia), ANA, IgG4,

Also: ANCA, ESR, Glucose, UA, CXR, RF

Testing for TB / Sarcoid + possible Orbital Biopsy

## Mass in Superior Temporal Orbit

Dermoid Cyst

Neurofibromas

Prolapsed Orbital Fat

Dermatolipoma

## Lacrimal Gland Problem / Enlargement

Benign and Malignant Tumors

Benign Mixed Tumor (Pleomorphic Adenoma)

Lacrimal Gland CA, Lacrimal Gland Lymphoma

Dacryops

Dacryoadenitis

Idiopathic, Viral, Pseudotumor (IOIS), IgG4-RD, Sarcoidosis, TB, Sjogrens

Graves Ophthalmopathy

Lymphoma

Prolapse of Lacrimal Gland

Dacryops – Lacrimal duct cyst

## Masses in Nasal and Superior Nasal Quadrant

Dermoid Cysts

Meningocele and Encephalocele

Sinus Mucocele

Lacrimal Sac: Tumor, Dacryocystocele, Dacryocystitis

Neurofibromas, Capillary Hemangiomas

Bulging Nasal Fat Pad

## Apparent Mass under Lower Eyelid

Orbital Tumors yes, but also need to consider:

Festoons - Prolapsed Orbital Fat and and/or Redundant folds of skin

Inflammatory Disease – Orbital Cellulitis, Graves Disease, Chalazion / Abscess

Lymphedema, Allergic Reactions

Lymphoproliferative Disorders - Lymphoma, Orbital Pseudotumor, etc.

Allergic "Shiners" edema – often responsive to treatment

# Visible and Palpable Masses in and around the Orbit



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## Hypertelorism

- increased orbital separation and increased interpupillary distance

seen in congenital craniofacial anomalies such as  
Crouzon's Syndrome



## In Contrast

## Telecanthus

- normal orbit separation, but large separation of the medial canthi

seen with:

BPES

Other Congenital Facial abnormalities

Down's Syndrome

Facial Trauma – e.g. avulsion of MCT

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## Hypotelorism

- decreased orbital separation and decreased interpupillary distance

seen with:

FAS – Fetal Alcohol Syndrome

Congenital Anomalies:

e.g. holoprosencephaly





## Small, Misshapen or apparently Absent Globe

– can be interpreted as an orbital problem



MAC Spectrum – Microphthalmos, Anophthalmos, Coloboma – optic fissure closure defects. Can often be associated with systemic defects as well

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Other associated congenital defects:

Goldenhar (Oculoauriculovertebral) syndrome, Trisomy 13-15

Phthisical Eye – after trauma, surgery, or severe inflammatory conditions

Other: Congenital Rubella, toxoplasmosis; high Hyperopia, maternal Vitamin A deficiency

## Large, Buphthalmic Eye

Congenital and Juvenile Glaucoma

Anterior Segment Dysgenesis (e.g. Rieger's), Megalocornea, High Myopia

Intraocular Tumor – RB, Uveitic – Rubella, Toxocara, Herpetic

Systemic: Lowe's Syndrome (oculocerebrorenal synd.), Neurofibromatosis, Sturge-Weber

End Stage Glaucoma



# Motility and Alignment

# Types of Motility Problems

Can be seen alone or in combination

## 1. Loss of Normal Eye Movements

Versions – e.g. Convergence Insufficiency, Parinaud's Syndrome (Upgaze)

Ductions - e.g. 6<sup>th</sup> CNP (Abduction), INO (Adduction), IR Restriction (Elevation)

## 2. Involuntary Eye Movements

Nystagmus, Square Wave Jerks

Ocular Neuromyotonia, Opsoclonus, Dysmetria, SO Myokymia

Oculogyric Crisis

## 3. Misalignments

1. “Strabismus” (“Primary”, Childhood, Decompensated)

2. Related to Neurologic, Myogenic, Orbital or Sensory Problems

### 1. Neurologic

Supranuclear – e.g. Inter Nuclear Ophthalmoplegia (INO), Parinaud's Syndrome, Skew Deviation

Cranial Nerves – 3,4,6 Palsy or Spasm\*

Structure CNS Abnormality – e.g. Chiari Malformation (e.g. Downbeat Nystagmus), Dolichoectatic Vessels

Congenital Dysinnervation Syndromes – e.g. Duane's Syndromes, CFEOM's Type 1 and 2

Systemic – e.g. Immune Polyneuropathies (GBS, MFS)

Neurodegenerative Disorders – Friedreich Ataxia, Spino-cerebellar Ataxia

### 2. Myogenic

Systemic – e.g. Myasthenia Gravis, Mitochondrial Myopathies (e.g. CPEO), Muscular Dystrophies

Local – Muscle Damage and/ or Fibrosis, Previous Surgery

### 3. Orbital Disease

e.g. Graves Disease, Orbital Fracture, Orbital Tumor, Orbital Pseudotumor

### 4. Loss of Vision – Sensory Deviation(Strabismus or Nystagmus)

e.g. Cataract, Optic Hypoplasia or Atrophy, ROP, Retinal Dystrophy

### 5. Strabismus (Primary, Familial, Congenital, Decompensated)

e.g. Congenital / Infantile Esotropia, Intermittent Exotropia,

IO Overaction, Accommodative Esotropia, Monofixation Syndrome

Risk population for Common Comitant Strabismus not associated with Neurologic Disorders\*

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## 1. Sixth Nerve Palsy

Microvascular , DM, MS, High ICP, Tumor (orbit, supra-orbital fissure, Cavernous Sinus, Parasellar, Posterior Fossa), Pseudotumor, Head and Orbital Trauma, Post-immunization, Post Viral, Congenital, Shunt failure, Aneurysm (e.g. Cavernous sinus), Meningitis, sometimes intracranial hypotension

## 2. Graves Ophthalmopathy (MR restriction)

## 3. Duane's Syndrome (Type 1 associated with esotropia, retraction / fissure narrowing on adduction, fissure widening with attempted abduction and upshoots/ downshoots with adduction. More commonly female and left eye. Comparison with 6<sup>th</sup> CNP: Duane's has greater abduction deficit , but less ET in primary). Type 3 Duane's - bilateral abduction loss and some limitations of adduction).

## 4. Myasthenia Gravis

## 5. Tight MR – from long term ET or previous MR Resection

## 6. Also Consider

Myositis

Orbital Tumor

Medial Orbital Wall Fracture with entrapment

Past LR recession

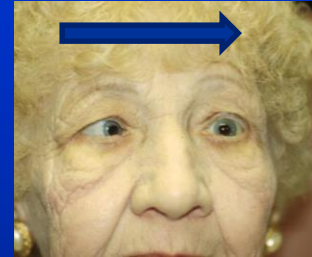
Scleral Buckle

Conjunctival Scarring / Restriction

Spasm of Near Reflex

Congenital Esotropia with Cross Fixator (not real abduction deficit)

Moebius Syndrome\*



Looking Left

# Internuclear Ophthalmoplegia

MS (younger)

CVA (older)

Traumatic INO – definitely possible

**Other:** Infectious (e.g. Syphilis), Tumor, Nutritional, Metabolic, Toxic (toluene),  
Drugs (Li, barbiturates, tricyclic antidepressants, etc), Paraneoplastic syndrome,  
GCA, Meningo-encephalitis, Arnold Chiari malformation, Hydrocephalus

# Adduction deficit



Looking Right

Myasthenia Gravis\* - adduction deficit common motility problem seen with MG

## And Then:

Third Nerve Palsy – but rare to have isolated MR palsy

Congenital Fibrosis of EOM Type 2 (Ptosis and loss of adduction, elevation and depression can mimic 3<sup>rd</sup> CNP)

MR recession

Orbital Disease – Graves, Myositis, Tumor

Post-op: Scleral Buckle

Conjunctival Scarring

Previous EOM surgery- e.g. tight LR

Ocular Neuromyotonia of 6<sup>th</sup> CN

Myotonic Dystrophy

Duane's Syndrome

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# Acquired Esotropia

## 1. Sixth Nerve Palsy – maybe early or partial

Think about: Microvascular, Head Trauma, Post Viral, MS, Tumor, etc.  
(see abduction deficit list )



## 2. Divergence Insufficiency (ET at distance > ET at near)

Usually Benign Condition, unless other neurologic symptoms or signs present, then need further investigation  
Older Patients Consider: Microvascular, CVA, Progressive Supranuclear Palsy, Cerebellar or Brainstem Lesions  
Younger Patients: Pseudotumor Cerebri, Arnold Chiari Malformation, Meningitis

## 3. Graves Ophthalmopathy (tight MR muscle(s))

## 4. Myasthenia Gravis

## 5. Sensory Visual Loss

## 6. Decompensated Esodeviation, e.g. Monofixation Syndrome

(factors: Hydrocephalus, shunt failure, trauma, ...)

## 7. Acquired Comitant ET (neurologically isolated, stable) - give full Cycloplegic Rx and do **prism adaptation**

## 8. After Neurologic Insult – encephalitis, meningitis, trauma (not necessarily 6<sup>th</sup> CNP)

## 9. Previous Muscle Surgery – consecutive ET or recurrent ET

## 10. Other: Over-Minused Correction (glasses or CTLs) (ET near > ET far)

## 11. Any other causes of Abduction Deficit

## 12. Not Previously diagnosed:

Congenital or Childhood Esotropia

Duane's Syndrome (ET less in Primary c/w 6<sup>th</sup> CNP)

Pseudo-Esotropia – Epicanthal folds, Negative angle kappa, Hypotelorism, Telecanthus

Latent Hyperopia with Accommodative ET

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# Acquired Exotropia

## Intermittent Exotropia

Decompensated Exophoria

Types: Basic, Divergence Excess, Convergence Insufficiency

Can Decompensate further to a Constant Exotropia



## INO

Consider: MS, CVA, Drugs, Metabolic, Paraneoplastic, GCA, Trauma

## Third Nerve Palsy

Hydrocephalus, Shunt Failure

Convergence Insufficiency (XT at near >XT at distance)

## Myasthenia Gravis

Orbital Disease: Tumor, Myositis, Pseudotumor, Graves

## Sensory

## Myopia

Previous EOM Surgery – Consecutive or Recurrent Exotropia

Other Ocular Surgeries: Scleral Buckle, large IR recessions

Ocular Neuromyotonia – e.g. of 6<sup>th</sup> CN- after prolonged lateral gaze

Previously Undiagnosed: Duane's Syndrome, Pseudo-Exotropia – positive angle kappa, Hypertelorism

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# Pseudo –Strabismus

## Apparent Horizontal Strabismus

### 1. Pseudoesotropia

- Epicanthal folds, Hypotelorism, Telecanthus
- Apparent Esotropia - Negative angle kappa\*



Normal – mild positive angle kappa

### 2. Pseudoextropia

- Hypertelorism
- Apparent Exotropia – Positive Angle Kappa\*



### How to differentiate?

Angle Kappa from Strabismus: Cover Testing vs Hirschberg test

Detecting Hyper and Hypotelorism: Interpupillary distance

Hirschberg and/or Cover Testing – can then be used to assess whether any strabismus is present

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\**Angle kappa* is the angle between the visual axis and the anatomical pupillary axis of the eye

# Hypertropia / Hypotropia and Vertical Diplopia

Fourth Cranial Nerve (SO) Palsy

Graves Ophthalmopathy (IR or other vertical muscle involvement)

Orbital Trauma / Fracture

Post-op Eye Surgery –e.g. post CE (Local Injection) or Scleral Buckle

Myasthenia Gravis

**Skew Deviation** (comitant or non-comitant vertical deviation often associated with vestibular / brainstem / thalamic problems – e.g. balance problems / ataxia). Incomitant Skew Deviations could include the Including Ocular Tilt reaction (**OTR\***) or mimic of IR palsy (e.g. RHT worse on Right gaze)



Age Related degeneration of Orbital Pulley System for EOMs → Cyclo-Vertical deviations

Brown's Syndrome

Inferior Oblique Over-Action

Third Nerve Palsy or aberrant regeneration

Orbital Tumors

Myositis, Orbital Pseudotumor

Glasses – Anisometropia, Optical Centers off, Induced Prism in eccentric gaze, etc.

Monocular Elevation Paresis (Double Elevator Palsy)

Congenital Absence of IR or Fibrosis

SO Myokymia

Sixth Nerve Palsy – sometimes has an associated vertical misalignment

GCA – can produce isolated EOM palsies

**Other:** Hemifield Slip, Dissociated Vertical Deviation (DVD), Wernicke's, Guillain-Barre, Central Inhibition of Fusional Reserves

**Inferior Oblique Palsy** – isolated IO palsy not likely, so consider: OTR, Orbital Tumor, Brown's Syndrome, MG, and IO damage after a LL Blepharoplasty

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\***OTR** - a Skew Deviation subtype characterized by: Skew Deviation, Ocular Torsion and a Head tilt. Some OTR pts. can have a pattern (3 Step Test) similar to 4<sup>th</sup> CNP, but ocular torsion is different, not the typical excyclotorsion seen with 4<sup>th</sup> CNP. OTR also can mimic other conditions like IO palsy. How to differentiate SD from other vertical strabismus? – Upright – Supine Test (JAMA Oph 2011; 129:1570)\*

## Elevation Deficit



Graves Ophthalmopathy (e.g. IR fibrosis)

Orbital Floor Fracture with entrapment

Third Nerve Palsy (Superior Division)

Double Elevator Palsy (in abduction and adduction)\*

Brown's Syndrome (in adduction)

CFEOM Type 1

Myositis (e.g. IR)

Myasthenia Gravis

Midbrain – Diencephalic Lesion

Isolated Palsy of SR or IO

Previous SR Recession

Other Mechanical: Orbital Mass, Glaucoma  
Implant, Fat Adherence Syndrome

## Upgaze Deficit



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Old Age

Parinaud's Syndrome

Thalamic Infarction

Progressive Supranuclear Palsy (PSP)

Hydrocephalus

Hemispheric Infarctions

Metabolic – Niemann- Pick Disease

Myasthenia Gravis

***Bilateral*** - CFEOM or any of the other  
conditions on the right is possible

# Depression Deficit



SR restriction

Graves

Fibrosis

Myositis

IR Ischemia

3rd CNP – Inferior Division

Myasthenia Gravis

Midbrain and  
Diencephalic Lesions

# Downgaze Deficit



Progressive Supranuclear Palsy

Parkinson– Like Syndromes

Midbrain Lesions

Myasthenia Gravis

Head Trauma

Oculogyric Crisis

(tonic deviation of the eye – usually upward)

Post-encephalitic Parkinsons

Drugs: neuroleptics, benzodiazepines, many cited

Cannabis, MS, Neuro-Syphilis

Lesions of 3<sup>rd</sup> and 4<sup>th</sup> Ventricles, Trauma

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## Convergence Insufficiency

Comitant Exotropia greater at near than distance. Decreased Near Point of Convergence (NPC)

- Primary
- Secondary
  - After Head Injury
  - Parkinson's Disease and other CNS degenerative disorders

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## Divergence Insufficiency

Comitant ET at distance, less or none at near and full ductions

- Primary Divergence Insufficiency  
(Neurologically Isolated\*, usually older patients)
- Secondary Divergence Insufficiency  
(Usually not Neurologically Isolated)
  - Cerebellar Ischemia / Stroke
  - Chiari Malformation
  - Demyelinating Disease
  - Idiopathic Cerebellar Degeneration
  - Progressive Supranuclear Palsy
  - Temporal Arteritis
  - High ICP / Pseudotumor Cerebri

Consider also:

6<sup>th</sup> CNP with spread of comitance over time  
MG

# Ophthalmoplegia – One Eye

(More likely Local Lesions)



## Multiple Cranial Nerve Palsies

### Orbital apex, Superior Orbital Fissure, Cavernous Sinus Lesions:

Tumor (e.g. meningioma, extension of pituitary tumor)

Aneurysm, AVM

Orbital Pseudotumor / IOIS (e.g. **Myositis**, Tolosa - Hunt Syndrome)

Orbital Cellulitis

Consider Fungal including **Mucormycosis**, **Aspergillosis**

Other Inflammatory:

Wegener, Sarcoidosis, TB, Syphilis, GCA, etc.

HZO (can see Ophthalmoplegia up to 3 months after onset)

Cavernous Sinus Thrombosis

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## Other Causes: Mechanical, Orbital Infiltration of Tumor, Ischemia, etc.

Trauma- edema and or orbital hemorrhage

Peri-neural Spread of Cutaneous Carcinoma

Ischemia – Saturday Night Ophthalmoplegia, Orbital Infarction Syndrome\*

Graves Ophthalmopathy – multiple muscles

High ICP, DM – could potentially cause multiple CNP (e.g. 3<sup>rd</sup> and 6<sup>th</sup>) at same time

Third Nerve Palsy – mistake for total Ophthalmoplegia

*\*Systemic Conditions could initially present unilaterally – e.g. MG, Guillain-Barre, ...*

*SEE NEXT LISTING*

# Ophthalmoplegia - Both Eyes

(More likely Systemic Conditions)



**CPEO** – usually bilateral (Kearns Sayre Syndrome = CPEO + pigmentary retinopathy and heart block), usually ptosis, *but not always*

**Myasthenia Gravis** – unilateral or bilateral

**Lambert-Eaton Myasthenic Syndrome**

**Muscular Dystrophies** – e.g. Myotonic Dystrophy ( can see an Iridescent Cataract), Oculo-pharyngeal D.

**Guillain-Barre Syndrome** – autoimmune, various subtypes, triggered by acute infection

AIDP (Acute inflammatory Demyelinating Polyneuropathy) – ascending paralysis

Miller Fisher Syndrome - Ophthalmoplegia, descending paralysis, areflexia, ataxia

Bickerstaff's Brainstem encephalitis

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Head / Brain Trauma

Stroke – Hemorrhage (e.g. putamen), Ischemic (e.g. horizontal gaze center)

Progressive Supranuclear Palsy

Spinocerebellar Ataxias (+ FHx, onset usually childhood and young adulthood)

Paraneoplastic Syndromes

Wernicke's Encephalopathy (Alcohol, Thiamine, Emergency)

Toxins: Organophosphates

Pituitary lesion (Unlikely but possible for bilateral), bilateral cavernous sinus disease

Graves Ophthalmopathy

IOIS, Orbital Fibrosis Syndrome

Medications: e.g. Valproate- Vertical gaze palsy , Statins

Other: Botulism, Meningitis, MS, High ICP, GCA, Whipple Disease, Neuro-Syphilis, Congenital Cranial

Dysinnervation syndromes



# Head Turns or Tilts Torticollis

## 1. Orthopedic / Muscular – most common causes

- e.g. Congenital muscular torticollis: damaged sternocleidomastoid muscle
- Cervical spine – damage / deformity : Fracture, TB, Scoliosis
- Tonsillitis, retropharyngeal abscess, drugs – neuroleptics, dystonias

## 2. Neurologic – not common

## 3. Ocular – not uncommon

- Incomitant Strabismus:
  - CNP: 4<sup>th</sup> (Superior Oblique) or 6<sup>th</sup> (Abducens)
  - Duane Syndrome, Browns Syndrome
  - A and V Patterns
  - DVD (Dissociated Vertical Deviation)
- Nystagmus
  - Congenital Nystagmus with Null Point
  - Spasmus Nutans
- Ptosis - often chin up head position
- Astigmatism



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### Aid in Diagnosis:

Patch one eye and if torticollis resolves then suggests strabismus is cause of torticollis

## 4. Down's Syndrome -25% have head posturing. Significant number have ocular cause (e.g. incomitant strabismus like an ET.) Some Down's patients have no definitive cause.



# Causes of Nystagmus

1. **Idiopathic / Congenital** – Typical Features – Conjugate, No Oscillopsia, Dampens at near and with Convergence, Null Point, Latent Nystagmus
2. **Sensory Visual Loss** – e.g. Deprivational Amblyopia, Optic Nerve, Retinal Disease, Albinism.  
*Need thorough 8 Point eye exam. Sometimes further testing, e.g. ERG.*
3. **Acquired later in Life – Often Associated with Symptoms** – e.g. Oscillopsia, blurring in eccentric gaze
  - CNS lesion – CVA, Tumor, MS (Most commonly in brainstem or cerebellum, less likely in parasellar region and cerebral hemispheres)
  - Spasmus Nutans vs. Tumor of Anterior Visual pathways
  - CNS malformation – e.g. Chiari Malformation
  - CNS inflammation – post viral (e.g. cerebellum) , post immunization, encephalitis, Lupus, MS
  - CNS: Associated with Seizure Activity in Cerebrum (Epileptic Nystagmus)
  - Associated with antibodies - e.g. antiganglioside Abs (anti- GD1b, GMI, GQ1b) – seen in GBS, MS
  - CNS Degenerative Disorders (e.g. Spino-cerebellar Ataxias)
  - Vestibular - e.g. Labyrinthitis
  - Metabolic – Mg and Vitamin B12, B1 (Thiamine) deficiencies
  - Toxicity – Phenytoin, Lithium, alcoholism, street drugs, glue sniffing (toluene)
  - Paraneoplastic Syndrome – associated with e.g. Lung CA and antibodies anti - Hu, Ri, Yo, Tr
  - Multiple Different Congenital Syndromes
  - Latent Nystagmus – manifesting later after some (monocular) loss of vision

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# Acquired Nystagmus

Long DDX but, Think first of:

1. Undiagnosed Congenital Nystagmus – *no Oscillopsia*
2. Drug Toxicity – e.g. Phenytoin, Lithium, Aspirin, Topiramate, etc.
3. Sedatives (e.g. Barbiturates) and Alcohol (e.g. Wernicke)
4. Toxins – e.g. Toluene (glue sniffing)
5. Encephalitis or Post Viral Syndrome – e.g. cerebellar involvement
6. Metabolic - hypothyroidism, magnesium or thiamine deficiency
7. MS – Multiple Sclerosis
8. Paraneoplastic Syndrome – e.g. Lung CA; CXR, Antibody testing (e.g. Hu, Ri, Yo, Tr)
9. Vestibular – Central or Peripheral (e.g. Labyrinthitis) – *Vertigo Present!*
10. Brainstem or Cerebellar Lesions or Malformations  
(e.g. Arnold Chiari, brainstem tumor or infarct) or Parasellar Lesions, rarely cerebral hemispheric lesions.



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*So before jumping to order a MRI consider #1-9, and ...  
Ask about Oscillopsia (not usually present in Congenital)  
Ask about Vertigo (Vestibular Nystagmus)*

# Downbeat Nystagmus

## Cranio-cervical Structural Disorders

Arnold-Chiari spectrum, Platybasia, Basilar Invagination, Syringobulbia  
Dolichoectasia of Vertebrobasilar artery system compressing caudal brainstem  
Tumors compressing caudal brainstem

Downbeat Nystagmus -  
Not always readily seen  
in primary gaze, but often  
noted in eccentric gaze  
positions



## Brainstem /Cerebellar Disease

Spino-cerebellar degenerations (e.g. spinocerebellar ataxias)  
Cerebellar disease/atrophy (e.g. Alcoholism)  
Posterior Fossa Strokes or tumors  
Paraneoplastic syndromes – e.g. cerebellar degeneration associated with Anti – Hu, Yo, Ri, and Tr antibodies  
Hydrocephalus, Trauma  
Encephalitis, Post-Viral Syndrome  
MS – Demyelinating diseases  
Anti-GAD antibodies – (GAD- Glutamic Acid Decarboxylase: Glu → GABA) – associated with Ataxia

## Metabolic, Drug, Toxin

Wernicke's encephalopathy (B1 deficiency), B12 deficiency, Magnesium (Mg) deficiency, Hypothyroidism  
Medications ( Lithium, Phenytoin, Carbamazepine, Felbamate, Morphine-barbituate combo)  
Toluene (glue sniffing) abuse

Exercise induced? JNO 2002;22:127

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Idiopathic – 20% or more cases, assess vasculopathic risk factors - JNO 2001; 21:39-41

Pupils

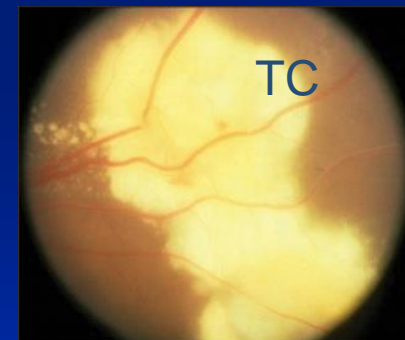
# Leukocoria

## Is it Retinoblastoma (RB)?



1. R/O RB
2. Uveitis – active or quiescent - e.g. Toxocariasis (TC)
3. Cataract
4. Asymmetric or High Refractive Error
5. Coats Disease
6. ROP- Retinopathy of Prematurity
7. Vitreous Hemorrhage –old
8. Retinal Detachment
9. Disc or Choroidal Coloboma
10. Myopic Degeneration
11. Persistent Fetal Vasculature:

e.g. Persistent Hyperplastic Primary Vitreous (PHPV) –  
small / microphthalmic eye, cataract, glaucoma, progressive RD, non-hereditary



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### Aids to help diagnose RB

1. Age of presentation (most cases diagnosed by age 2, 95% by age 5 yo)
2. B-Scan – Mass
3. CT – may have calcifications

# Corectopia

Irregular Pupil Shape and /  
or Abnormal Location

## 1. Congenital and Developmental

Congenital Iris Coloboma, Ectropion Uvea

Ectopia Lentis et pupillae

*Neurofibromatosis – nodules, ectropion uvea*

Anterior Dysgenesis (e.g. Axenfeld- Rieger, Peter's Anomaly)

Iridocorneal Endothelial Syndrome (ICE Syndrome) e.g. essential iris atrophy

Posterior Polymorphous Corneal Dystrophy

## 2. Inflammatory (e.g. uveitis – posterior synechiae; HZO-segmental atrophy)

## 3. Neovascular – NVI (see list)

## 4. Neoplastic (e.g. iris, ciliary body melanoma)

## 5. Damage or Distortion From:

Trauma (e.g. iris dialysis, iris prolapse, scarring, epithelial down-growth)

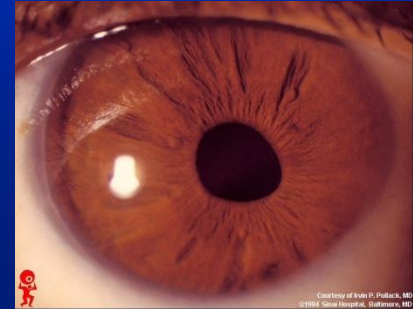
Surgery (e.g. Iridectomy, CE)

Peaked pupil from Vitreous Strand, Blood or synechiae in the angle

Angle Closure attacks

IOL capture, dislocation

Iris Atrophy – age, diabetes, essential, ischemia



Courtesy of Kevin P. Puffack, MD  
© 1998, Second Harvest, Baltimore, MD

## 6. Neurological - Permanent or Transient Irregular Pupils

Midbrain lesion - MS, Ischemia, Tertiary Syphilis (Argyll Robertson)

### Transient Irregular Pupil in what appear to be healthy patients

“Tadpole Pupil”

Migraine patient

Early Horner's

Young Healthy Women

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# Poor Pupil Mobility

Poor Reactions to Light, Dark or other stimuli

- **Age**

- **Diabetes Mellitus**

- **Damaged or Paralyzed Pupil**

Uveitis, Angle Closure, Surgery, Trauma

Topical: Mydriatics (e.g. atropine), Miotics (Pilocarpine), Natural Products

- **Afferent or Efferent Input Defect**

Damage to any part of Pupillary Light Reflex Pathways

RAPD

Parasympathetic or Sympathetic Defect: Third Nerve Palsy, Adie's, Horner's Syndromes

- **Systemic**

Medications or Toxins

Mydriatic - Anticholinergics , Adrenergics (Methamphetamine),

Miotic - Narcotics

Systemic / Neuro-Muscular Conditions

Lesions at level of Tectum (dilated), Midbrain (mid-dilated) , or Pons (miotic)

Myotonic Dystrophy – miotic

Fisher variant of G/B Syndrome or Riley-Day Syndrome - mydriatic

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# Anisocoria (Unequal pupils)

1. Physiologic - 20% of population

2. CNS and Efferent Nerve Input Problem

Lesions at level of Tectum , Midbrain, or Pons – not common

Sympathetic Defect - Horner's Syndrome, Pourfour du Petit syndrome – opposite, stimulated sympathetics\*

Parasympathetic Defect

Third Cranial Nerve Palsy, Aberrant Regeneration

Ciliary Ganglion / **Tonic** Pupil: Post-viral (e.g. HZO), Orbital Tumor, Trauma, Idiopathic (also called Adie's Tonic Pupil)

3. Pharmacologic / Toxic: Stimulation or Blockage of Sympathetic or Parasympathetic Receptors in one eye

Dilation: Anticholinergics : e.g. Atropine, Natural alkaloids (some pollens). Adrenergic- Phenylephrine – Red top Drops

Miosis: Pilocarpine , Anticholinesterases (e.g. Insecticides , Echothiophate)

4. Asymmetric Damage or Atrophy of the Iris and Pupil

Eye Trauma

Surgical / Trauma

Inflammatory / Uveitis

High IOP (e.g. paralytic mydriasis)

Pigmentary Dispersion Syndrome

Other Causes of Iris Atrophy: Asymmetric Atrophy of Irides

Old Age, Essential Iris Atrophy – progressive,  
Ischemia (trauma, past high IOP, Hb SC disease, etc.)  
Syphilis, Diabetes, Idiopathic?

5. Other:

Anisometropia (e.g. Unilateral High Myopia)

Amaurotic (Blind) Eye

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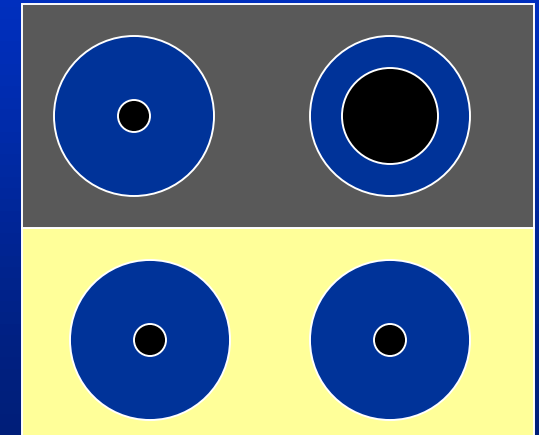
Usually one abnormally small (miotic) pupil or one abnormally large (mydriatic) pupil



### Abnormally Miotic Pupil

(Anisocoria worse in the dark – poor dilation in dark)

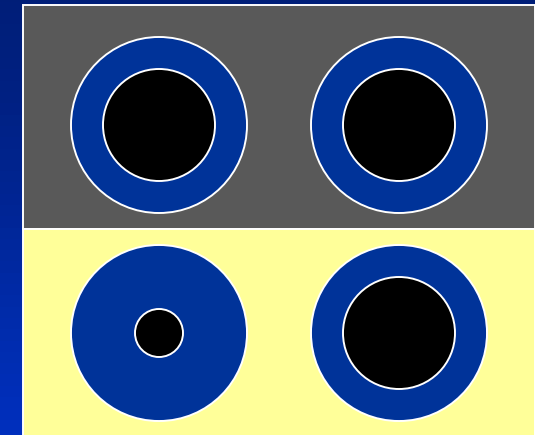
- Iris scarring from Uveitis, Trauma, Surgery
- Horner's Syndrome
- Chronic Pilocarpine Use, Toxins such as Insecticides
- Argyll-Robertson Pupil



### Abnormally Mydriatic Pupil

(Anisocoria worse in the Light – poor constriction to light)

- Tonic Pupil
- Third Nerve Palsy
- Pharmacologic – atropine, natural alkaloids, phenylephrine
- Iris Damage – trauma, intraocular surgery, uveitis  
pigmentary dispersion
- Iris Atrophy - Asymmetric



# Dilated Pupil(s)

Not an emergency most of the time  
Especially if an Isolated Finding

## Third Cranial Nerve Palsy

(A dilated Pupil should be accompanied by a loss of motility and ptosis – unless patient is comatose)

Basal Meningitis

Supratentorial mass, hemorrhage, edema → transtentorial uncal herniation \* (usually accompanied by stupor or coma)

Aneurysm - e.g. posterior communicating artery

Ischemia, Parasellar Mass or Inflammation

## Midbrain Damage

- Ventral – 3rd CN fascicle, Dorsal (usually both pupils)

## Ciliary Ganglion Damage or Damage to SPCNs

- CG: Post-viral (e.g. HZO), Orbital Tumor, Trauma, Idiopathic (Adie's)

- SPCN's – injections, ocular surgery – e.g. SB

## Iris Damage / Atrophy

– old age, trauma, inflammatory (including Herpes Virus infections e.g. HZO, syphilis (tabes dorsalis)), essential iris atrophy, high IOP (paralytic mydriasis), Intraocular Surgery (Castroviejo Syndrome)  
Pigmentary dispersion syndrome

## Pharmacologic

Anticholinergics: e.g. atropine, natural alkaloids (jimson weed, corn)

Adrenergic: Phenylephrine

Pourfour du Petit syndrome – opposite of Horner's – stimulated sympathetics

Sometimes a blind or nearly blind eye (e.g. Severe Optic Neuropathy)

## Transiently Dilated Pupil

- Migraine patients,
- Benign Episodic Mydriasis in Young Women
- Angle Closure Glaucoma

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## Bilateral Dilated Pupils- Think of:

Any of above if Bilateral

Systemic Medications or Drugs (e.g. atropine, amphetamines, cocaine), Exposure to natural products (weeds, corn pickers)

Dorsal Midbrain (Tectal) Lesions, Severe brain anoxia, Bilateral Iris damage or atrophy, Bilateral Adie's Pupil, Bilateral 3<sup>rd</sup> CNP

# *Transient Pupillary Dilation*

- Migraine Patients (EPDYW)
- Benign Episodic Mydriasis in Young Women
- Tadpole pupils
- Pourfour de Petit Syndrome
- Neck-carotid trauma
- Seizure disorder, postictal state
- Episodic angle closure
- First sign of early third?  
Midbrain Corectopia

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## Unilateral Miotic Pupil

### Horner's Syndrome

Idiopathic, Traumatic (head, neck, shoulder), Cluster Headache, Carotid Dissection  
Lesion-sympathetic pathway – including Brainstem, Cervical Spinal Cord, Apical Lungs, Neck, Thyroid,  
Local Anesthetic Blocks – Head, neck, brachial plexus; Central lines / Jugular Venipuncture

### *Pharmacologic – Topical Medications, Toxins*

Pilocarpine, Cholinesterase Inhibitor (e.g. Echothiophate, Organophosphates)

### *Damaged Pupil*

Uveitis , Trauma, Surgery (e.g. Posterior synechiae)

*Rare: Lesion in Midbrain, Unilateral Argyll Robertson Pupil\**

## Constricted Pupil(s)

## Bilateral Miotic Pupils

Opioids (e.g. Morphine)

Drugs with Cholinergic Activity - antipsychotics, antidepressants, antihistamines

Cholinesterase Inhibitors:

Snake Venom, Biochemical Warfare Agents

Systemic Medications - e.g. For MG like Pyridostigmine (Mestinon)

Organophosphate Poisoning ( Insecticides like Malathion, Herbicides)

Pontine Hemorrhage

Lesions in Hypothalamus , Dorsal Midbrain

Argyll Robertson Pupils (syphilis)

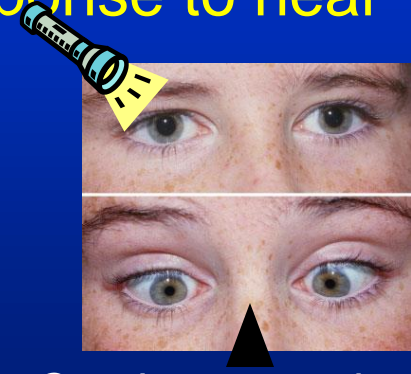
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# Light Near Dissociation (LND)

Poor light reaction, but good constriction response to near

## 1. Dorsal Midbrain Syndrome

Pupils round, usually equal, but bilateral LND  
Pineal Tumor, Hydrocephalus, CVA, etc.



Can be one or both pupils

## 2. Rostral Mid-brain Lesions – can be unilateral LND

## 3. Argyll-Robertson Pupils – often irregular pupil(s), often smaller

## 4. Damaged Ciliary Ganglion – e.g. Tonic Pupil

## 5. Severe Afferent Defect (RAPD)

## 4. Other – **Diabetes Mellitus**, alcoholism, encephalitis

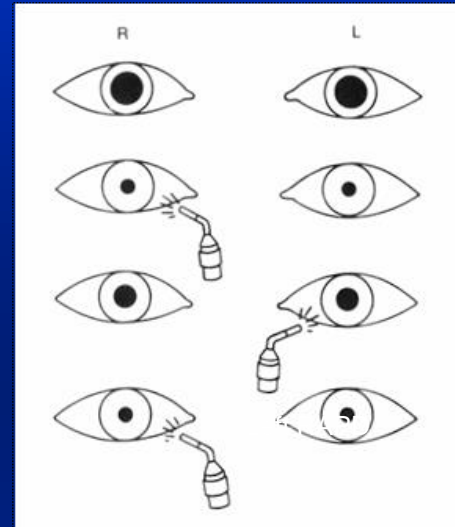
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# RAPD

Relative Afferent Pupillary Defect  
Look for in the Setting of Visual Loss

## More Common

1. Optic Neuritis
2. Ischemic Optic Neuropathy – AION, PION
3. CRAO
4. Traumatic Optic Neuropathy
5. Optic Nerve Tumor –e.g. Glioma, Meningioma
6. Compressive Neuropathy  
    Adjacent Tumor, Graves Ophthalmopathy, Orbital Hemorrhage
7. Ischemic CRVO
8. Optic Atrophy – from previous or ongoing insult to optic nerve  
    Unilateral or asymmetric atrophy- e.g. asymmetric glaucoma damage



## Much Less Common

1. Extensive Retinal Detachment or Damage
2. Contralateral Optic Tract Lesion
3. Asymmetric Chiasmal Lesion
4. Specific lesion in pre-Tectal area\*
5. Dense cataract can cause APD in contralateral eye
6. Mild RAPD - sometimes seen with vitreous hemorrhage, amblyopia, RD, BRAO

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## 1. Bilateral Miosis

Systemic Medications: e.g. Narcotics (e.g. Morphine),

Cholinesterase Inhibitors (Snake venom, biochemical warfare, pesticides, MG treatments)

– early effects excessive salivation and tearing

Topical Medications: e.g. Pilocarpine

Pontine Lesions – pinpoint pupils

Damage to Sympathetic Pathways in Hypothalamus

Diencephalon Lesions – small but reactive

Metabolic or Medical Encephalopathy / Coma – small pupils but still reactive

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## 2. Bilateral Mydriasis

Systemic Medications: anticholinergic (atropine)

sympathomimetics (amphetamines, cocaine, etc.)

\*note some  
confusion and  
possible variations  
of pupils in  
medical coma.

Topical Medications or Natural Products – atropine, cyclopentolate, Jimson Weed pollen

Familial Bilateral Congenital Mydriasis

Tectal and Midbrain Lesions – inflammatory, neoplastic

Severe Anoxia of Brain (e.g. cardiac arrest) – sympathetic effect

Can be seen in Coma from: Alcoholism, Uremia, Epilepsy, Meningitis, Apoplexy

*There are many other causes such as Intraocular Inflammation, Trauma and Surgery, Congenital and Developmental defects, and Lesions that cause disrupted Sympathetic or Parasympathetic input - but usually these do not produce symmetrically bilaterally large or small fixed pupils.*

# Anterior Segment and IOP



# Follicles

*Think of Viral Infections, especially:*

*Adenoviruses – more often bilateral, acute, associated with URI*

*Herpes simplex (primary infection)*

*Molluscum contagiosum*

*Acute Hemorrhagic – e.g. enterovirus*

*Recent URI related*

*Chlamydia – AIC = Adult Inclusion Conjunctivitis(unilateral , chronic), Trachoma*

*Reactions to topical ophthalmic medication,*

*e.g. atropine, alpha agonists, antivirals, miotics*

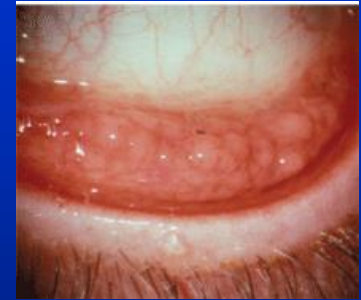
*Other: Parinauds Ocular Glandular Syndrome (e.g. Cat Scratch, Tularemia, Mycobacterial Infections)*

*Lyme, Newcastle, sometimes some bacterial: Strep, Neisseria, Moraxella*

*Children – benign lymphoid folliculosis*

*Conjunctival Lymphoma*

# Conjunctival Bumps



## Conjunctival Papillary Reaction

Non-specific, seen with inflammation from allergic and infectious conjunctivitis, topical medication toxicity, staph marginal disease, mucous fishing, and many other sources.



## Giant Papillary Reaction

Contact Lens Wear (Giant Papillary Conjunctivitis)

Atopic Keratoconjunctivitis

Vernal Keratoconjunctivitis – Palpebral and Limbal

FB- **Ocular Prosthesis** or Suture End



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## R/O Orbital Disease

Inflammatory – Orbital Cellulitis, Contiguous Sinus Inflammation

Orbital Pseudotumor

**Graves Ophthalmopathy**

Orbital Tumor, Surgery (Post op)

Orbital Trauma, Fracture, Open Globe

Blocked Orbital Lymphatics – surgery, radiation

# Chemosis



Conjunctivitis – allergic (local allergen) , local irritant – chemical, insect sting, infectious, HSV, some viral or bacterial are severe enough

Nearby Corneal , Eyelid, Sinus, Meningeal or Lacrimal inflammation

Vascular (Venous) Congestion – Apex or Cavernous Sinus Process, CC fistula

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## Other

Lymphedema (Chronic Hereditary), Myxedema

Angioneurotic Edema

Systemic – R. Heart Failure, Nephrotic syndrome ( Plasma protein low), Chronic Ventilator Patient

Conjunctivochalasis – not to be confused with chemosis – redundant conjunctiva secondary to aging, but also associated with Blepharitis and lid malpositions. More commonly seen inferiorly, but can be 360° - Symptoms – irritation, tearing

# Injected, Congested or Prominent Vessels on the Globe

Usually Conjunctival Vessels, but look for underlying Episcleral/Scleral Vessels

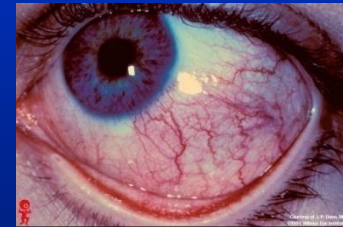
## Conjunctivitis and other External Problems

- allergic, irritation (e.g. smoke), chemical , infectious, dry eye and exposure issues, Blepharoconjunctivitis, Acne Rosacea, Trichiasis, Lid Malpositions, eye drops and preservatives, etc.

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## Ciliary Flush (Circumcorneal Congestion)

- uveitis, acute glaucoma, corneal FB or keratitis



## Episcleritis or Scleritis

Diffuse, but more often focal area of injection

## Systemic

- Polycythemia, Graves disease, Carcinoid, HTN, Renal Failure
- Alcoholism, Cannabis
- Hyperviscosity – e.g. Multiple Melanoma, Sickle Cell
- Fabry's Disease, Ataxia Telangiectasia



## Local Factors

- chronic topical vasodilators ("red out drops"), chronic topical anesthetics
- sympathetic irritation, trauma, FB, insect bite
- local tumor with feeder vessels
- local vascular malformations or vascular tumors

## Vascular Obstruction or Malformation

- Orbit or Cavernous Sinus mass, Carotid-Cavernous Fistula
- Orbital Inflammatory or Infiltrative Process
- Orbital / Peri-Orbital Venous Malformations →



Venous Malformation –  
JAMA Ophthalmology

# Symblepharon and Conjunctival Scarring



- Chemical Burns or Physical Trauma
- **Mucous Membrane Pemphigoid**
  - with ocular involvement called **OCP** (Ocular Cicatricial Pemphigoid)\*
- **Stevens Johnson Syndrome** (bilateral and acute)
- Other Dermatologic Conditions:
  - Erythema Multiforme, Toxic Epidermal Necrolysis, Epidermolysis Bullosa, Pemphigus
  - Also: Think of Paraneoplastic Processes
- Surgery – e.g. post Pterygium Surgery, incomplete resection of Eyelid CA
- CA - undiagnosed BCCA, SCCA, or SEBACEOUS Cell CA of lid
- Chronic Conjunctivitis:
  - Atopic or Vernal Keratoconjunctivitis**, Reiter's, Scleroderma, Graft versus Host, Lichen Planus
  - Infectious: EKC (**Adenovirus**), Beta-hemolytic Strep, Diphtheria, **Trachoma**
  - Topical Drugs - Echothiophate, Epinephrine, Pilocarpine, Timolol, Idoxuridine
- Dry eyes, Sjogrens syndrome
- Acne Rosacea
- Other: Sarcoidosis, Wegener's Granulomatosis, Radiation

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# Conjunctival or Epibulbar Mass / Lesion



**Choristomas** - usually congenital, **Dermoid, Lipodermoid** , Ectopic Lacrimal Gland.

**Epithelial malignancy** – OSSN: Conjunctival Intraepithelial Neoplasia (CIN), Invasive Squamous cell CA, Basal Cell or Sebaceous Cell CA invading Conjunctiva, Amelanotic melanoma

**Benign epithelial** – Squamous Papilloma (can be due to Human Papilloma Virus)  
- Pingueculum, Pterygium (beware of atypical pterygia with CIN)  
- Benign hereditary intraepithelial hyperplasia, Papillomatosis\*

**Pigmented Lesions** - Racial Melanosis (usually bilateral, should not grow), Conjunctival Nevus (often cysts in it, no feeder vessels)  
- PAM (Primary Acquired Melanosis- can undergo malignant transformation), Conjunctival Melanoma,  
- some OSSN can have abnormal pigmentation  
- Deposits: cosmetics, FB's, Hemosiderin after hemorrhage, Addison's, Silver, Systemic Tetracyclines

**Sub-epithelial Tumors** - Lymphoproliferative (e.g. Lymphoma, Lymphoid Hyperplasia, Multiple Myeloma – “Salmon Patch”  
Hemangiomas, Lymphangiomas, Karposi's Sarcoma, Mxyoma, fibroma, mets, neuroma, amyloid

**Cysts** - epithelial, ductal, inclusion



**Inflammatory Lesion** - Pyogenic Granuloma, Orbital Pseudotumor, Pingueculitis,  
- Traumatic lesion, Phlyctenule, Nodular episcleritis,  
- Limbal follicles of Vernal KC  
- Granuloma (Infectious, Allergic, Sarcoidosis, Foreign body, Cat Scratch disease, TB)

**Bitot's spots** – e.g. Vitamin A deficiency, xerosis

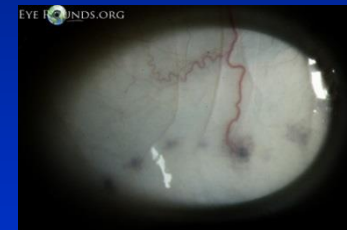
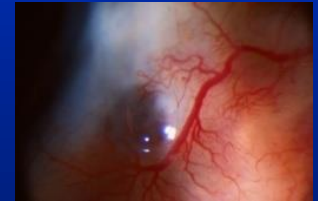


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# Spots on the “White of the Eye”

## Scleral Lesions:

- **Scleral Dellen** – depression or dimple, loss of epithelium over defect in sclera. Seen after surgery (e.g. Pterygium, EOM surgery), Caustery Trauma, medication toxicity, autoimmune disorders
- **Scleral thinning** - e.g. aging- typically in front of rectus muscle insertions, areas of dehydration, previous surgery
- **Staphyloma** – thinning, bulge in sclera
- **Axenfeld Loops**
- **Medication Deposits**
- **Congenital Melanosis Oculi (CMO)** increased melanocytes in episclera, sclera or uvea. Bluish/gray patches suggest increased underlying uveal pigment. Can undergo malignant transformation later in life.
- **Nevus of Ota (CMO +lid nevus)**
- **Scleral discoloration** – also periocular tissues, nails, teeth, auricles - from tetracyclines (e.g. minocycline)



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# Scleral Thinning, Episcleritis and Scleritis

## Scleral Thinning:

- Area of dehydration
- Dell (depression with loss of overlying epithelium)
- Staphyloma
- Age Related Changes – e.g. anterior to EOM insertions
- Episcleritis, Scleritis

## Episcleritis

Mild pain, diffuse or nodular, **rare** uveitis or peripheral keratitis or thinning

Causes include:

Idiopathic

Related to autoimmune disease →

Associated with toxic effects of external diseases such as staph Blepharitis –  
local effect –Episcleritis in region of lid disease

## Scleritis

Fire engine red, not blanching with Neo2.5%. – Possible uveitis and peripheral corneal effects

- Local or diffuse, Nodular, -- usually moderately painful, tender
- Posterior Scleritis – thickening seen on B-scan, moderate to severe pain
- Necrotizing Scleritis – extreme pain, vasculitis, thinning , perforation

Consider:

- Post-Surgical Necrotizing/Infectious Scleritis – e.g. pterygium surgery (e.g. pseudomonas, actinomyces, fungal, mycobacterial, gram + cocci)
- Idiopathic, but...
- 50% of time associated with a systemic disease, see list

## Diseases associated with Episcleritis and Scleritis:

Rheumatoid Arthritis

Herpes Viruses (e.g. Simplex, Zoster)

Other Infectious: Staph, Strep, Syphilis,  
Pseudomonas, Mycobacterium,  
Acanthamoeba

Relapsing Polychondritis (check ears, nose)

Orbital Pseudotumor

Wegener's Granulomatosis

Sarcoidosis

Medications: Fosamax, Aredia

Lupus (SLE)

Inflammatory Bowel Disease

Spondyloarthropathies

### Possible Testing:

ANCA Panel ,

Rheumatoid Factor, FTAbS, SSA/SSB,  
ESR(GCA), PPD, ANA

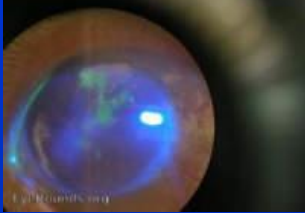
Urinalysis, CXR, ACE, HLA testing

Scleral Biopsy or Trial of Oral Acyclovir

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# Corneal Fluorescein Staining or Pooling



- Epithelial Defect: Abrasion, Ulcer, RES, etc. (see list of Corneal Epithelial Defects)
- Healing epithelial defects – can see pooling and punctate staining around them
- Punctate Staining (PEK) – Dry Eye, etc (see upcoming list)
- Epithelial Disease – e.g. Herpetic Ulcer
- Peripheral Corneal Thinning

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## Acute Peripheral Corneal Thinning:

Physical or Chemical Trauma – associated with swelling of conjunctiva / episclera at limbus

Gonococcal, Streptococcal Infections

Mooren's Ulcer (associated with Hepatitis C)

Connective Tissue / Autoimmune Disorders – Rheumatoid Arthritis

## Dellen

A transient shallow depression in the cornea near the limbus which is caused by a local dehydration of the corneal stroma, leading to a compression of its lamellae (thinning). It can occur as a result of strabismus surgery, cataract surgery, swelling at the limbus (as in chemosis, episcleritis or pterygium), rigid contact lens wear or senility\* Respond to patching or lubrication.





# Corneal Epithelial Defects- Chronic or Non Healing



- **Trauma** (Mechanical, Thermal, Chemical) -most common cause, but when the history is unclear consider:
- **Exposure** - consider tear volume and quality; eyelid malposition, trichiasis and lagophthalmos  
Neuro-paralytic – e.g. 7th N., can be painful, unless CN V also involved  
Dry Eye - these pts can have what appear to be spontaneous "abrasions"
- **Recurrent Erosion Syndrome**
- **Chronic Herpetic Keratitis – e.g. disciform HSV keratitis**
- **Medical Toxicity** - e.g. anti-virals , topical anesthetic abuse, preservatives
- **CHRONIC EYELID PROBLEMS:** ectropion, entropion, **lagophthalmos**, **trichiasis**, **floppy eyelid**, lid imbrication\*, sunken upper eyelid in Enophthalmos.
- Early Corneal Ulcer - early there may not be much of an infiltrate, but there can be some stromal edema as a clue. **THINK - HSV**, fungal, bacterial, Acanthamoeba
- Shield Ulcer - Vernal, Atopic conjunctivitis
- Corneal Dystrophies – can cause RES
- Diabetic Epitheliopathy
- Neurotrophic Ulcer - not painful is big clue – all have corneal hypoesthesia (see list) – **HZO**, HSV, leprosy, neurosurgery, Acoustic Neuroma. Also DM, radiation, Corneal dystrophies, P/O – LASIK,PKP, Burns, CTL Over Wear
- Bullous Keratopathy
- Corneal Melts - e.g. Autoimmune / Connective Tissue diseases
- Chronic inflammatory external diseases: OCP, Atopic/Vernal, Epidermolysis Bulosa
- Secondary or Idiopathic **Limbal Stem Cell Deficiency** (“Conjunctivalization” of Corneal specific sign)

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**Management:** Lubrication, patching, eliminating unnecessary toxic drops, autologous serum, bandage CTL, punctal occlusion, Tarsorrhaphy + punctal occlusion (for most difficult cases – impending perforations, neurotrophic) , lid tightening procedures

# Corneal Haze/ Opacification Loss of Clarity

## Corneal Edema

– stromal or epithelial - see following lists

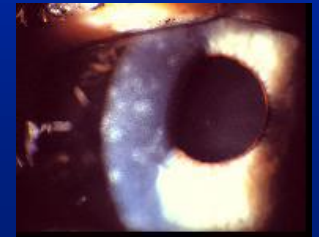
## Corneal Infiltrates

### Infectious:

HSV, HZO, Bacterial, Fungal, TB, Syphilis, Acanthamoeba

### Non-Infectious:

Marginal –Staph, Auto-immune (e.g. RA), Mooren's Ulcer  
CTL Related  
Atopic  
Chemical, Exposure and Dry Eyes  
Neurotrophic



## Interstitial Keratitis (IK) - Stromal vessels and haze/ necrosis

HSV, Congenital Syphilis, Cogan's (IK + auditory and vestibular problems), TB, Sarcoidosis



## Increased IOP - Angle Closure, Post CE, Neovascular Glaucoma, etc.

## Corneal Degenerations: Salzmann's Nodular Degeneration\* , Band Keratopathy

## Corneal Dystrophies

## Corneal Deposits, Crystals

## Corneal Whorls (Verticillata): Fabry Disease, Medications: Amiodarone,

## Corneal Surface Growths – Pterygium, Squamous Neoplasia,

## S/P Laser Refractive Surgery – e.g. PRK



## Congenital / Developmental Problem

e.g. Anterior Segment Dysgenesis, Amyloidosis, Congenital Glaucoma and Secondary Infantile Glaucomas

*"STUMPED": Sclerocornea, Tears in Descemet's (Congenital Glaucoma), Ulcers (e.g. intrauterine – viral), Metabolic*

*(e.g. Mucopolysaccharidoses), Peter's Anomaly, Edema ( Congenital Hereditary Endothelial Dystrophy), Dermoid*

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# Causes of *Acute* Corneal Edema

Trauma – corneal abrasion, chemical or thermal burn,  
intraocular surgery, radiation

Exposure – proptosis, 7<sup>th</sup> Nerve Palsy, lid malpositions

Hypoxia - CTL over wear

Hydrops – Keratoconus

Osmotic – irrigation with hypotonic solutions

High IOP

Infectious – Viral, Bacterial, Fungal, Acanthameoba (sometimes no clear infiltrate)

**Think of HSV!** – disciform, stromal keratitis

Inflammatory – Uveitis

Endothelial Decompensation

in patient with corneal dystrophy – e.g. Fuch's

also in ICE, Descemet's Membrane detachment after CE

Anterior Segment Ischemia

Other

Vitreous Touch, AC Foreign Body, Neurotrophic, Idiopathic cyclic corneal edema

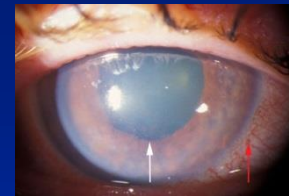
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Keratitis



Angle  
closure



“Steamy appearance”

# Causes of *Chronic* Corneal Edema

## Chronic Compromise of Conjunctival Function

→ Chronic Epithelial Edema, Haze

From: Trauma, External Disease, Exposure, Tumor involving Conjunctiva

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## Loss or Damaged Endothelial Cells

Trauma – Mechanical, Chemical, CE, Phaco

Toxins – Intraocular injection,

TASS – Toxic Anterior Segment Syndrome after CE

## Corneal Dystrophies

Fuch's Dystrophy

Posterior Polymorphous Dystrophy

Iridocorneal Endothelial Syndrome (ICE)\*

Retains lens Fragment(s)

Chronic Inflammation

corneal, uveitis, multiple traumas, surgeries

Signs:

Loss of Endothelial Cells, Guttata

Loss of Transparency

Thickened Stroma

Epithelial cyst formation



PBK – pseudophakic  
bullous keratopathy

# Punctate Corneal Staining (SPK or PEK)

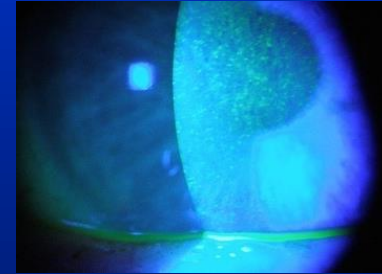
## 1. Tear Film Problem

Dry Eye – aqueous, poor tear production – old age most common cause, but also consider:

Connective Tissue Diseases: e.g Sjogrens (primary or secondary), RA  
Medications (Isoretinoin, antihistamines, antidepressants, BCP)  
HIV

Meibomian Dysfunction- Blepharitis, Staph related, Acne Rosacea

Mucin – goblet cell loss – conjunctival disease



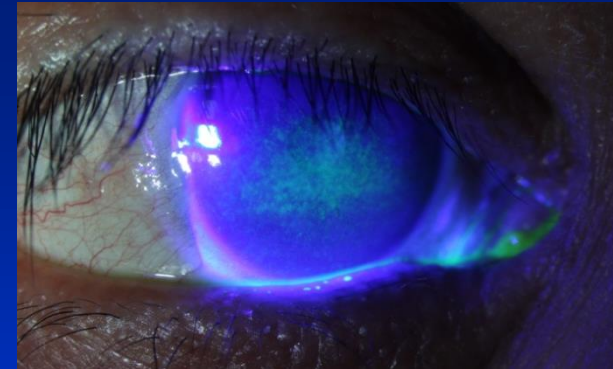
## 2. Exposure Problems

Eyelid malpositions – entropion, trichiasis, ectropion, lid retraction, notching, Floppy eyelid, etc.

Lagophthalmos – 7<sup>th</sup> nerve palsy, Cicatricial, etc.

Proptosis – e.g. Graves, Orbital Tumor, etc.

Damage to V1 – e.g. Herpetic



3. Trauma – including chemical, ultraviolet light

4. Toxicity of Topical Medications or Preservatives

5. Healing Corneal Abrasion

6. Contact Lens Over Wear

7. External Infectious / Inflammatory – blepharitis, conjunctivitis, keratitis

8. Keratitis: Viral (Herpetic – HZO, HSV, Molluscum, Adenovirus), Staph related, Thygeson's SPK

9. Neurotrophic Keratopathy (CN 5)

# Corneal Infiltrates

## Infectious Corneal Ulcers

## Other Infiltrates

Larger Infiltrate

Smaller

Often single lesion

Multiple

Very painful

Less Painful

More Central

More Peripheral

Cells in A/C

No A/C cells

Conjunctival Injection  
more generalized

More segmental / focal  
injection

Infectious:

Viral, Bacterial, Fungal, Acanthamoeba,  
HSV, TB, Syphilis, Hepatitis C

Non-Infectious:

Marginal –Staph sensitivity, Mooren's Ulcer  
Autoimmune PUK's

CTL Related

Atopic

Chemical

Exposure and Dry Eyes

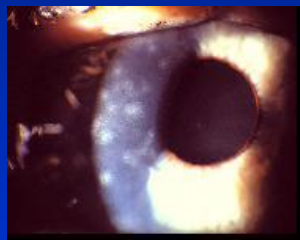
Neurotrophic

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**PEDAL** – to help distinguish non-infectious from infectious  
Infectious more associated with **P**ain, **E**pithelial defects, **D**ischarge, **A**nterior  
chamber reaction and more *central* **L**ocation



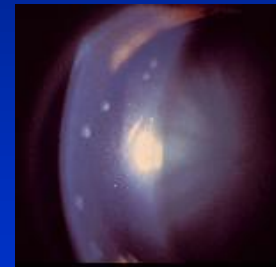
Infectious Corneal Ulcer



Adenovirus



Staph Marginal  
Keratitis



CTL over wear  
Keratitis



Peripheral / Marginal  
Keratitis



# Corneal Ulceration and Thinning

## Central

Infectious – more common

Bacterial, Fungal

Viral (HSV, HZO)

Acanthamoeba

Autoimmune – much less common

- e.g. RA

Other – Neurotrophic, Vernal KC, Trauma, post-op, Exposure, CTL, etc.



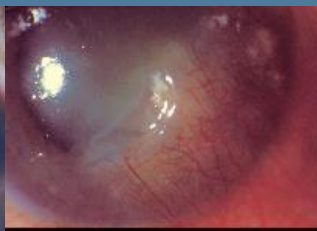
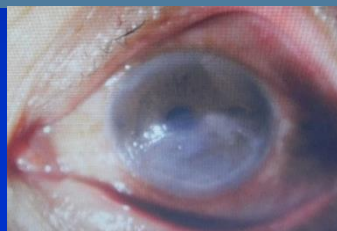
## Peripheral Ulceration and Thinning

If *inflammatory* then need to distinguish between Peripheral Ulcerative Keratitis (PUK) and Mooren's Ulcer

### PUK causes

- Autoimmune Vasculitides (consider as potentially lethal)
- RA, PAN, GPA, SLE etc. see listing
- Other Autoimmune- e.g. GVH, IBD
- Infectious (less common)- Staphylococcus, Gonococcus, TB, Syphilis, HSV, HZO, Hepatitis C, Dengue, Acanthamoeba, Fungal
- Other: Trauma, Post-op, Exposure, CTL, Carcinoma, SJS, OCP, KC Sicca, Blepharitis, Pellucid

Other causes of peripheral thinning without inflammation: Terrien's Marginal degeneration (lipid deposits), and Furrow degeneration near arcus.



# Corneal Vessels / Pannus

- Contact Lens Over Wear - hypoxia
- Staphylococcal Disease – Blepharitis
- Phlyctenular KC - TB, Staph aureus, Chlamydia, Neisseria, ...
- Acne Rosacea
- Previous Corneal Ulcer
- Trachoma or Adult Inclusion Conjunctivitis (AIC)
- Chronic HSV Corneal Disease – stromal vessels
- PUK– e.g. Mooren's, Autoimmune / Vasculitis
- Interstitial Keratitis – HSV, Congenital Syphilis, Cogan's
- Degenerated Blind Eyes often associated with band keratopathy
- Severe Dry Eyes
- SLK – Superior Limbic Keratoconjunctivitis
- Bullous Keratopathy
- Vernal Conjunctivitis
- Vitamin A deficiency
- Undiagnosed Corneal Degenerations and Dystrophies
- ALSO CONSIDER:
  - *Chronic Exposure - lagophthalmos*
  - Lid Malpositions: Ectropion, Entropion, Trichiasis, Floppy Eyelid, Lid Imbrication
  - Cicatricial Conjunctival Disease: OCP, Stevens-Johnson, Epidermolysis Bulosa, Radiation, Trauma, Chemical Burns, S/P Pterygium Surgery



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# Decreased Corneal Sensation Hypesthesia



Common to all cases of  
Neurotrophic Keratopathy is  
corneal hypesthesia

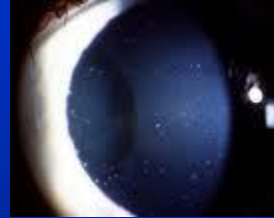
- Space occupying lesion:
  - Cerebellopontine (CPA) angle tumor
  - Cavernous sinus or Superior Orbital Fissure lesion
- Herpes Simplex or Zoster
- Corneal Edema
- S/P CE, PK surgery or LASIK
- Corneal Dystrophies
- Diabetics
- Radiation, Thermal burn, Chemical Burns
- Chronic CTL wear
- Medications: Anesthetics, Timolol, Sulfacetamide, etc.
- Syndromes: Riley- Day, Goldenhar, Mobius
- Misc: Leprosy, Vitamin A deficiency, MS
- Increasing Age

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# Keratic Precipitates (KP)

## Non-Granulomatous Uveitis Marked by:

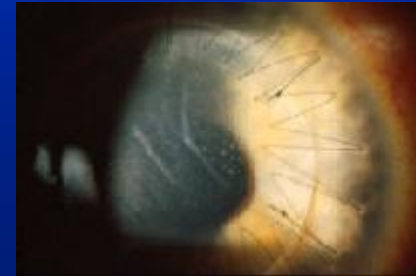
More acute onset      More injection  
Fine KP                  Pain, Photophobia



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## Non-Granulomatous - think of:

HLA- B27 associated Uveitis  
TINU  
Post-Infectious or Drug (antibiotic) induced uveitis  
Idiopathic



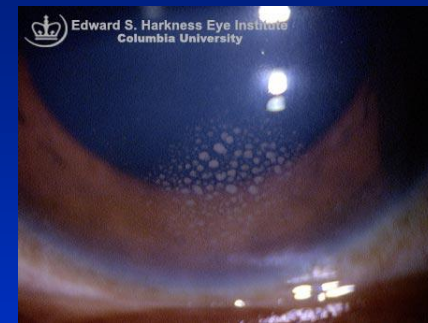
Corneal Graft Rejection  
emedicine Ophthalmology

## Granulomatous Uveitis suggested by :

Chronic Uveitis              Little Vessel Injection  
Mutton Fat KP              Less symptoms

## Granulomatous? Think of:

- Sarcoidosis
- Syphilis
- Sympathetic Ophthalmia
- Herpetic Uveitis
- Intra Ocular FB
- Toxoplasmosis
- Tuberculosis
- VKH
- Uveitis associated with MS
- Lens Induced Uveitis



Large White or Yellow  
“Greasy”  
“Mutton Fat” KP

# Anterior Uveitis

Injection, Flare, KP, Synechiae,  
Cells in AC and/or Anterior Vitreous

1. HLA-B 27 related  
(Ankylosing Spondylitis, IBD, Reiters (Reactive Arthritis), Psoriatic Arthritis)
2. Behcet's (HLA-B5)
3. Juvenile Idiopathic Arthritis (JIA)  
(also known as Juvenile Rheumatoid Arthritis - JRA)
4. Post – Viral
5. Lens Induced (e.g. phacolytic)
6. Medications: biphosphonates -alendronic acid (fosamax), rifabutin, sulfonamides, cidofovir, pamidronic acid, metipranolol, streptokinase
7. Trauma
8. Post CE – early endophthalmitis?
9. Infectious –Herpetic kerato-uveitis (VZV, HSV, EBV), HIV, Rubella Virus, Bartonella, Toxoplasmosis, Whipples disease, Syphilis\*, TB, Mycoplasma pneumoniae
10. Systemic/ Autoimmune – MS, Wegener's, Sarcoidosis, Immune recovery after AIDS treatments, TINU (Tubulo -interstitial nephritis and uveitis syndrome)
11. Uveomeningoencephalitic Syndrome
12. Ocular Conditions: Posner -Schlossman (PS) Syndrome, Fuch's Heterochromic Iridocyclitis, Idiopathic
13. Masquerade Syndrome – Lymphoma, Intraocular Tumors

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## Anterior Uveitis and High IOP

Think of:

HSV or HZO with trabeculitis\*

Glaucomacyclitic Crisis (PS Syndrome)

Lens Induced

IOL- UGH Syndrome

Steroid Responder

(e.g. uveitis being treated with corticosteroid)

Other: Sarcoidosis, Fuchs, JRA

## Also Consider

– not real uveitis, but cells in AC

Pigment Dispersion Syndrome

Retinal Detachment

Ghost Cell Glaucoma

Masquerade Syndrome

– RB, Lymphoma, etc.

Anterior Uveitis:

Get a good Review of Systems

e.g. Mouth Ulcers, Lung Disease, TB, Herpes  
Diarrhea, IBD, Genital Ulcers, Kidney Disease  
Arthritis, **Back Pain (Lower?) – High Yield**  
Shingles? Rashes, Atopic Symptoms

Main Work-up Test Considerations:

HLA- B27 (and any others  
FTABS, RPR/VDRL\* on the left that  
CXR seem indicated)  
ACE/ Lysozyme  
PPD - if Chronic

- ✓ Uveitis
  - Anterior, Panuveitis, Endophthalmitis
- ✓ Keratitis
  - e.g. Secondary to Bacterial Corneal Ulcer
- ✓ Iris Pigment
  - Pigmentary Dispersion, IOL
- ✓ Retinal Detachment
  - usually pigmented cells in Vitreous and AC
- ✓ RBC – see causes of Hyphema, Ghost Cells
- ✓ R/O Masquerade Syndrome
  - e.g. RB, Lymphoma, etc,
- ✓ OIS



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# Hypopion

- ✓ Severe Uveitis  
e.g. Behcet's Disease, HLA-B27 related, etc.
- ✓ TASS - usually within 24 h of intraocular surgery\*
- ✓ Endophthalmitis – usually later than 2h hours after surgery, usually much pain and injection
- ✓ Retained Intraocular FB
- ✓ Corneal Ulcer (can have Hypopion and not necessarily Endophthalmitis)
- ✓ Masquerade Syndrome  
e.g. **RB**, Lymphoma, Leukemia, Metastasis,  
Triamcinolone (or other particulate injections)



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# Hyphema



## 1. Trauma

Blunt and Penetrating, IOFB

## 2. *R/O Intraocular Tumor*

RB, Melanoma, Hemangioma

## 3. Post Intraocular Surgery

Cataract, Laser PI, Trabeculectomy, UGH Syndrome (AC-IOL)

## 4. Iris Abnormalities

*NVI* (*Ischemia, uveitis, etc – see list*), Varices, Telangiectasias

Vascular tumors: Juvenile Xanthogranuloma (JXG), Angioma, RB

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## 5. Uveitis

e.g. HZO, HSV, Fuch's Heterochromia, Reiter's syndrome, Ankylosing Spondylitis, etc

## 6. Systemic Conditions

e.g. Coagulopathies, Leukemia (Child), Sickle Cell

# Neovascularization of the Iris = NVI Rubeosis

## 1. Retinal Ischemia:

CRVO (Central Retinal Vein Occlusion)

PDR (Proliferative Diabetic Retinopathy)

OIS – (Ocular ischemic syndrome, carotid disease)

CRAO – (Central Retinal Artery Occlusion – less common)

Anterior Segment Ischemia

Blood Disorders: Sickle Cell

Vasculitis, ROP, Coats, PHPV

Neurofibromatosis - Gliomas, large and small vessel problems

## 2. Chronic Retinal Detachment

## 3. Chronic Uveitis, Endophthalmitis, Hypotony

## 4. Previous Trauma, Surgery or other Insult (e.g. Radiation)

## 5. Post-op

## 6. Intra-Ocular Tumors of the Iris, Choroid, RB, etc.



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NVI Mimic: Dilated iris vessels – usually radial, seen with active inflammation

# Lesions of the Iris

1. **Nevi** – usually flat, variable pigmentation
2. **Melanoma of Iris or Ciliary Body**
3. **Nodules** - raised: Lisch nodules (Neurofibromatosis)

Granulomatous Uveitis  
Scarred Foreign Body



4. **Iris Cysts** – traumatic, post-op, congenital or acquired stromal cysts, pigmented epithelial
5. **Infectious:** (Nodules, Papules) TB, Leprosy, Syphilis

6. **Primary Tumors:** Juvenile Xanthogranuloma, Hemangiomas, Neurofibromas, Choristoma (Ectopic Lacrimal Gland)



Stromal cyst

7. **Metastatic:** Carcinomas, Leukemia, Lymphoma
8. **Segmental Melanosis Oculi**
9. **Things mistaken for masses:**

Ectropion Uvea, Segmental Iris Atrophy  
(e.g. HZO, CE), Iris Defects, Synechiae (trauma, surgery)



Segmental Iris  
Atrophy

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# Defects of the Iris

## Iris Transillumination Defects:

Albinism

Essential Iris Atrophy

HZO - uveitis

Pseudoexfoliation Glaucoma

Trauma

Intraocular Surgery / Phaco

Pigmentary Dispersion/ Glaucoma



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## Heterochromia

Horner Syndrome

Sturge Weber Syndrome

Uveitis – including Fuch's and Posner Schlossman

Pigment Dispersion

Use of Prostaglandin analogs

Trauma and Surgery

R/O Benign Heterochromia

Parry Romberg Syndrome

Other Syndromes

Pigmented Iris Tumors

Siderosis Bulbi



# Lens Opacities and associations

## Adult

**Nuclear Sclerosis** – myopic shift, subtle oil droplet

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**Cortical** - traumatic

**Posterior Subcapsular** - corticosteroid use, atopic dermatitis

**Less Common** - Posterior Polar, Anterior Subcapsular,  
Diabetic Snowflake, Polychromatic (myotonic dystrophy)

## Pediatric

**Punctate Blue Dot** - common, AD, multiple small blue spots, not progressive or visually significant

**Anterior Polar**

**Nuclear** – often central 3 mm, rubella

**Posterior Polar**

**Zonular (Lamellar)** - 50% of visually significant

### When to do basic

#### Pedi Cataract Workup

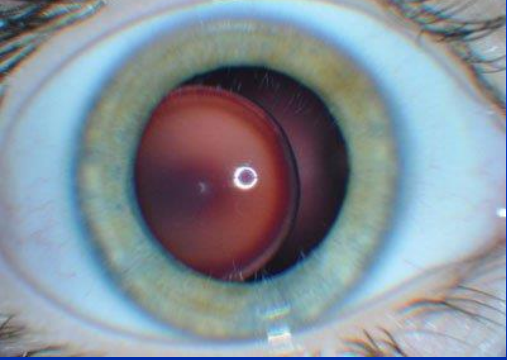
Bilateral Congenital , + FHx – no testing

Bilateral Congenital, - FHx, do W/U

Acquired Bilateral older – do W/U

Unilateral Congenital or Older and  
Healthy – no testing

# Lens Abnormalities and associations



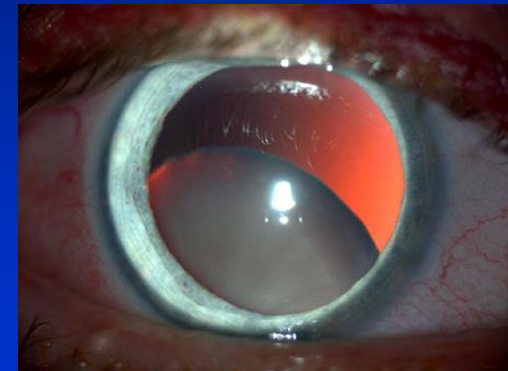
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1. **Aphakia** – acquired or congenital (rare)
2. **Spherophakia** – high myopia, Weill- Marchesani Synd.
3. **Lens Coloboma** - often associated with other ocular colobomas
4. **Ectopic Lens**

Trauma, Surgery

Ectopia Lentis et pupillae – can be associated with other ocular abnormalities.

Marfan's Syndrome, Homocystinuria,  
Hyperlysinemia, Ehlers-Danlos Syndrome.



# Acute High Intraocular Pressure

Any form of Glaucoma can result in High Pressures  
but the Most Common Causes Include:



- Acute Angle Closure Attack or other causes of Pupillary Block *(See upcoming lists)*
- Post Intraocular surgery – CE, PK, DMEK/DSEK\*
  - Acute: Viscoelastic, Air Bubble, Pupillary Block
  - Later: Non-pupillary block Pseudophakic /Aphakic Glaucoma, UGH Syndrome (IOL, especially AC IOLs)
- Uveitic Glaucoma
  - HSV, HZO, Glaucomatocyclitic crisis (PS Syndrome), UGH, Phacolytic, Fuchs Heterochromic Uveitis, JIA, Sarcoidosis
- Steroid Induced High Pressure
- NVG – Neovascular Glaucoma
- Trauma Related – after Hyphema, Angle Recession, etc.
- Orbital Trauma- Hemorrhage, Compartment Syndrome

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\* DMEK / DSEK = Descemet's Membrane/ Stripping Endothelial Keratoplasty

## 1. Primary Angle Closure Glaucoma (ACG)

## 2. Secondary Angle Closure or Narrow Angles

### Anterior Pulling

Peripheral Anterior Synechiae - Inflammation, Trauma

Neovascularization of the Iris and angle

ICE Syndrome

Epithelial Downgrowth

### Posterior Pushing

Posterior Synechiae (secluded pupil, iris Bombay)

Phacomorphic

Tumors

Choroidal Effusions – after surgery, PRP, CRVO

Malignant Glaucoma

# Angle Closure Glaucoma and Pupillary Block

## With Pupillary Block

Primary ACG

Secondary ACG

Phacomorphic

Ectopia lentis

Pseudophakic (IOL)

Aphakic (anterior vitreous face)

Posterior Synechiae (with secluded pupil, iris bombe)

## Without Pupillary Block

Neovascular Glaucoma

ICE – abnormal corneal endothelial growth

Intra-ocular tumors

Uveitis – Peripheral Anterior Synechiae

Plateau Iris

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Primary - (POAG)\* - usually symmetric

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Secondary – more often unilateral

1. Clogging of TM - RBC, lens protein and macrophages, pigment cells, tumor cells, viscoelastics
2. Alteration of TM - Inflammation (Trabeculitis), Trauma (physical and chemical)  
FB – siderosis, Corticosteroids
3. Blockage of drainage after the TM
  - Sickle Cells in Schlemm's Canal (even in Sickle trait)
  - Increased episcleral pressure – Carotid Cavernous (CC) Fistula, Orbital tumor /inflammation, venous backup

# Open angle Glaucoma

By Disease Process

1. Primary Open Angle (POAG)
2. Secondary Open Angle (SPLIT mnemonic)  
Substances: Steroids, Viscoelastics, Siderosis (Iron IOFB)

Pigmentary

Pseudoexfoliation

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Lens – phacolytic, IOL (UGH)

Inflammatory – Uveitic Glaucoma. e.g. PG syndrome, HZO

Increased episcleral pressure – e.g. CC fistula

Tumor – e.g. melanoma

Trauma – angle recession, Hyphema



# Flat or Shallow AC



## With High IOP

Acute Angle Closure Glaucoma (AACG)

Other Pupillary Block – Aphakic, Pseudophakic,  
Phacomorphic, Ectopia Lentis

Synechia closure

anterior

posterior – pupillary block, iris bombay

Malignant Glaucoma

Mature, Intumescent (Swollen) Lens

## With Low IOP:

Post op Wound leaks – CE, PK, Trabeculectomy

Trauma with leakage (open globe)

Corneal Perforation – Corneal Ulcer or other inflammation

Excessive Filtration after Trabeculectomy

Choroidal detachment – serous or hemorrhagic – surgery, trauma, inflammation

Post-traumatic cyclodialysis

## Flat AC: Grades

– contact with corneal endothelium  
and ...

I – peripheral iris

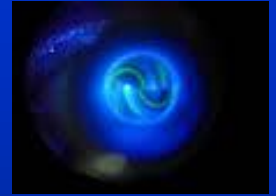
II – iris up to pupil

III – lens (surgical urgency)

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## Causes and Associations

# Hypotony (Low IOP)



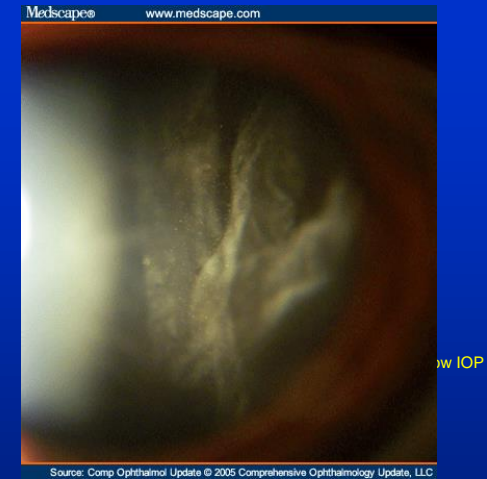
- Post-op (CE, Trabeculectomy, Scleral suture) Wound Leak
- Excessive Filtration after Trabeculectomy, Drainage Devices/Implants
- Choroidal Detachment
- Ciliary Body Detachment (e.g. traction), sometimes large Retinal Detachment
- Uveitis (Ciliary Body Shutdown)
- Trauma with leakage (open globe), Post-traumatic cyclodialysis
- Corneal Perforation – Corneal Ulcer or other inflammation
- Ocular Ischemia
- S/ P Cyclodestructive Procedures, Phthisis Bulbi
- Systemic: Hypertonicity, Acidosis, Uremia, Hyperglycemia, Osmotic Agents – like Mannitol
- Myotonic Dystrophy
- Congenital Anomalies: microphthalmos, aniridia , coloboma

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# Fundus – Vitreous, Retina and Disc

# Vitreous Cells, Pigment or Debris

- Blood (RBC and ghost RBCs) – see vitreous hemorrhage
- White Blood Cells (WBC)
  - Uveitis: intermediate, posterior or panuveitis
  - Masquerade ( actually tumor cells - e.g. lymphoma, RB)
- Pigmented Cells or Granules – can be a sign of RD
- Post Vitrectomy or Other RD Surgeries
- Larger Particles (Crystals) that could be mistaken for cells
  - Cholesterol Crystals- Synchysis scintillans (past injury or inflammation)
  - Asteroid Hyalosis - particles made of calcium and phospholipids
    - usually unknown cause, but associated with DM, HTN , hypercholesterolemia



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If cells are clumped  
think more intraocular  
tumors like CNS  
Lymphoma or Mets

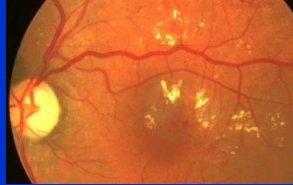


Pigment in Anterior vitreous

# Whitish or Yellowish Retinal or Sub-Retinal Lesions

## Discrete Borders

Hard Exudates



Vessel Plaques

Certain retinal/choroidal tumors

Drusen



Macular Dystrophies

- AD – pattern vary- look like ARMD in younger people – e.g. Adult-onset Vitelliform, Butterfly, Reticular, Stargardt's like Dystrophies.

Tracts – nematode, ophthalmomyiasis (fly larva- maggot – direct invasion)

## Less Defined Borders

Cotton Wool Spots

Myelination of NFL

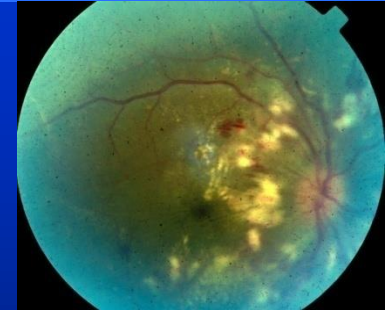
Infiltrates:

- Inflammatory
- Primary Tumors
- Metastasis to retina or choroid
  - Breast and Lung most common
- Lymphoma
  - Uveal (or Vitreo-retinal) lymphoma
- Granuloma – Choroidal Tuberculoma in TB pts

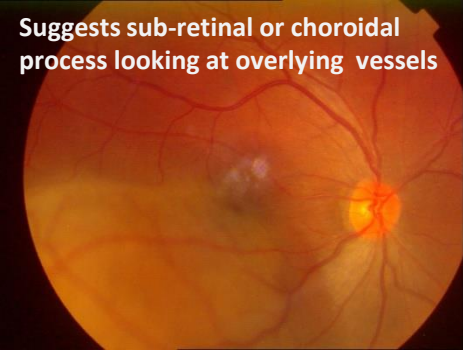
White Dot Syndromes\*

AIBSES, AZOOR, MEWDS, AMPEE, POHS, birdshot retinochoroidopathy, serpiginous choroiditis, etc

Edema of retina – CRAO, BRAO, posterior uveitides, etc.

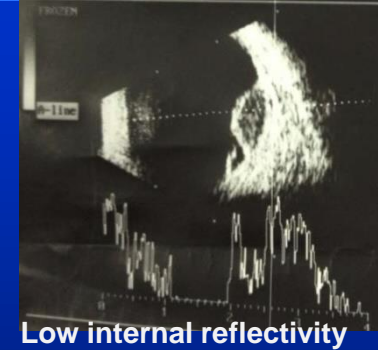


Suggests sub-retinal or choroidal process looking at overlying vessels



# Apparent Mass in Retina / Choroid

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## DDX

### Choroidal Tumor

- R/O Choroidal Melanoma
- Metastatic Tumor, Lymphoid Tumor
- Choroidal Hemangioma
- Choroidal Osteoma

Choroidal Detachment – effusion, hemorrhage

Nodular Posterior Scleritis

Choroidal Nevus

CHRPE

Exudative Retinal Detachment

From: Tumor, Post. Uveitis, Systemic (e.g. Toxemia), etc.

Subretinal Hemorrhage

Retinal Tumor:

- e.g. Retinoblastoma, Astrocytoma, Granuloma (TB, Sarcoidosis), Vascular tumors (e.g. NF1, TS)

### Evaluation and Management

A, B-scan, FA, CT of Orbits

### Assessing for Systemic Involvement

- e.g. Choroidal Melanoma for metastasis
- Choroidal Metastasis for primary site

CXR, Liver Enzymes, Colonoscopy  
CT or MRI of Body or **PET-CT**

### If suspect melanoma or metastasis

– do a systemic evaluation

If negative systemic work-up

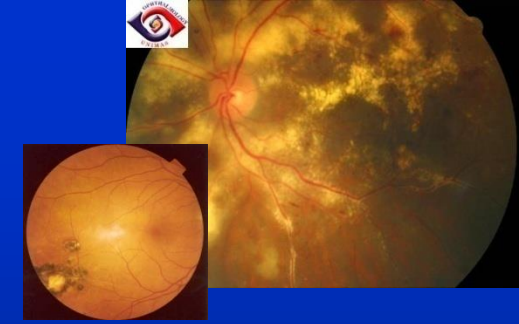
- consider local Rx- radiation, laser, cyro, plaque, or enucleation depending on size

If there is metastasis or primary disease elsewhere

- consider palliative radiation, injections or other local Rx as enucleation is not necessary

# Posterior Uveitis

Signs: Vitreous Cells, Retinal Infiltrates, Vascular Sheathing, CR Scars



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- **Toxoplasmosis** – active infiltrate, often an adjacent CR scar, Treatment\*
- **Toxocariasis** – Granuloma – often up in vitreous, usually no CR seen
- **Syphilis** –front (keratitis) to back (chorioretinal, optic nerve), panuveitis
- **HSV, VZV, CMV: Acute Retinal Necrosis (ARN), Progressive Outer Retinal Necrosis (PORN)**  
**ARN**- relatively immunocompetent- associated with inflammatory signs (AC or Vitreous Cells, with retinal infiltrates+ necrosis)  
**PORN** – immunocompromised - little if any inflammatory response – just retinal necrosis
- **Other Infectious: Lyme Disease, Tuberculosis, Typhus, Whipple’s, West Nile Virus, Rubella, Nematode (DUSN - sub-retinal)**
- **Candida**
- **Sarcoidosis**
- **Intraocular FB, Siderosis**
- **Sympathetic Ophthalmia**
- **Cat Scratch Disease (Bartonella)**
- **Vogt – Koyanagi –Harada Disease (VKH)**
- **Behcets Disease**
- **Uveitis associated with MS**
- **Eales Disease – posterior peripheral phlebitis**
- **Pars Planitis – more intermediate uveitis**
- **Multifocal Idiopathic Inflammatory Conditions of Retina and Choroid (“White dot syndromes”)**  
APMPPE and PPM, Serpiginous Choroiditis, Birdshot Retinochoroidopathy, Multiple Evanescent White Dot Syndrome (MEWDS), Multifocal Choroiditis and Panuveitis (MCP), Presumed Ocular Histoplasmosis Syndrome, Acute Idiopathic Blind Spot Enlargement Syndrome, Acute Retinal Pigment Epithelitis
- **Sometimes Associated with Encephalitis: e.g. VKH, Herpes, West Nile Virus, Toxoplasmosis (HIV)...**
- **Remember - Masquerade Syndrome – Lymphoma, Retinoblastoma, etc.**

## Work up Considerations

Much more based on clinical findings

FTABS, VDRL  
PPD , Quantiferon Gold, CXR

Titers for Toxoplasmosis and Toxocara of no real use (cannot definitely DX or R/O) since there is a wide prevalence of positive titers in the general population

Important to consider infectious causes of uveitis such as syphilis, TB, Herpes (e.g. ARN), Whipplés, before giving corticosteroids which can worsen condition and blindness

# Cotton Wool Spots

- DM
- HTN
- OIS – Carotid or Ophthalmic Artery Disease
- AIDS retinopathy

Radiation retinopathy

Interferon Therapy

Pregnancy (Toxemia)

Purtscher's retinopathy

Collagen vascular disease (e.g. SLE)

Severe Anemias, other Blood Disorders (Leukemia, Multiple Myeloma, etc)

Coagulopathies

Sepsis/ Sub-Acute Bacterial Endocarditis

Fat emboli, CO poisoning, Renal Diseases



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# Hard Exudates

(Lipoprotein deposits leaked from damaged vessels, e.g. microaneurysms)

Discrete Yellow-White deposits

## #1 Diabetic retinopathy

Then think:

- Hypertensive maculopathy
- Neuroretinitis
- Macular degeneration – wet
- Coat's Disease
- Parafoveal Telangiectasis
- Other: angiomas (Von Hippel-Lindau), retinal artery aneurysms, vasculitides, vasculopathies, FEVR



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# Retinal Infiltration or Edema

Can look similar

- Ischemia – CRAO, BRAO
- Tumor or Metastasis
- Trauma – Commotio retinae/ Berlin's Edema
- Uveitis – e.g. AMPPE, ARN
- Vasculitis – e.g. SLE, GCA, Sarcoidosis
- Multiple Cotton Wool Spots
- Purtscher's retinopathy



## CRAO or similar picture

Carotid or Ophthalmic artery disease

Embolic Phenomenon- carotid or heart sources

Vasculitis- e.g. GCA

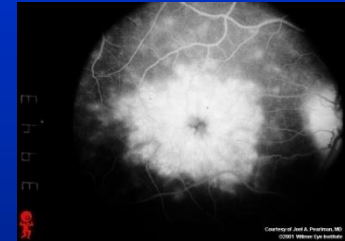
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# Macular Edema or Thickening

Often detected with Contact Lens, FA or OCT

- **Cystoid Macular Edema**

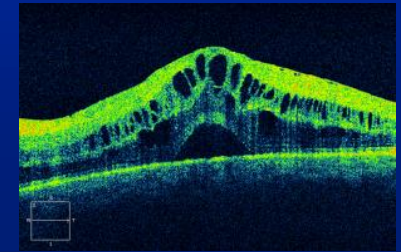
- e.g. Post Cataract Extraction, Broken Capsule, Vitreous Loss
- Vitreal Macular Traction (VMT), ERM, Retinitis Pigmentosa
- associated at times with ARMD, DR, Uveitis, RVO
- Medications: Niacin, Fingolimod, Tamoxifen...



- **Diabetic Macular Edema**

- associated with microaneurysms and hard exudates

- **Uveitis** – anterior, posterior or pars planitis
- **CRVO or Branch RVO**
- **Trauma** – Commotio Retinae, Berlin's Edema
- **Microcystic Macular Edema** – seen in association with optic neuropathies
- **Vascular Tumors of Retina**
- **Retinal Telangiectasia**
- **Choroidal Tumor** - underlying
- **Medications: Hydrochlorothiazide**
- **Other: Juvenile Retinoschisis** –cystic spaces, not really edema, separation of inner retina from other layers



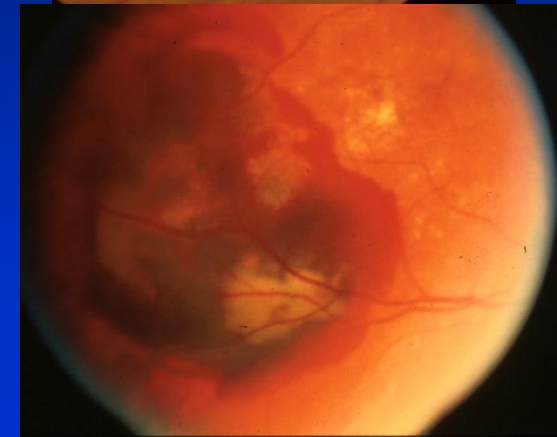
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# Posterior Hemorrhages

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## Types

- Pre-retinal Hemorrhage  
or Vitreous Hemorrhage  
(blocks view of vessels)
- Intra-Retinal Hemorrhages
- Sub-Retinal Hemorrhage  
( can see vessels over it)



# Pre-Retinal or Vitreous Hemorrhage

Trauma – blunt or penetrating, eye or head

Post-op

Neovascularization – disc, retinal or sub-retinal (see causes next slide)

Vitreous Detachment or Traction

Retinal Tear, Break, or Detachment

Chronic Uveitis

Tumor

Macroaneurysm

Terson's Syndrome – Sub - Arachnoid Hemorrhage and High ICP

Valsalva

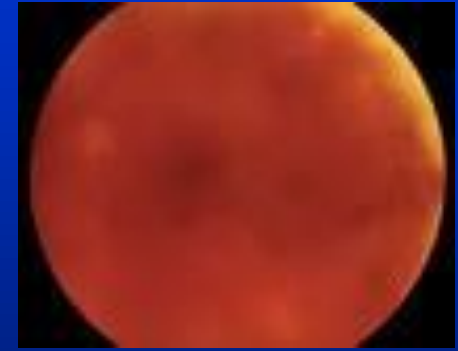
IOL

SRNVM – rare but possible

Retinal or Sub-Retinal Tumor

X-linked Retinoschisis – young males

Other: Persistent Hyaloid Artery, Retinoschisis, Purtscher's Retinopathy, Coats Disease, Anemias, Lupus and other Autoimmune Connective Tissue Diseases



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# Neovascularization

## Disc ,Retina, or Sub-Retinal

- Proliferative Diabetic Retinopathy
- CRVO
- CRAO- rarer than CRVO
- Ocular Ischemic Syndrome
- Chronic Uveitis
- Sub-retinal or Retinal Tumor
- Hemoglobinopathies: Sickle Cell (SA,SS or SC Disease), Thalassemia
- Retinal Vasculitis
- Coat's Disease
- SRNVM
- FEVR – temporal peripheral retina
- Other Vascular Problems?: CC Fistula, Aortic Arch, GCA
- Eales Disease – rule out diagnosis



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# Intra-Retinal Hemorrhages

Trauma, Valsalva, Shaken Baby Syndrome, Normal Baby after Vaginal Delivery, Sudden ocular decompression (IOP)

Vascular Diseases: DM, HTN, OIS, HIV, Vasculitis, Sickle Cell (and other Hemoglobinopathies), Purtscher's Retinopathy, High Altitude, Radiation Retinopathies

Obstructions – Retinal (CRVO, BRVO), Congestion (Head and Neck)- Newborn, Hanging, Choking, Papilledema

Hyperviscosity syndromes – (dysproteinemia, leukemia, polycythemia – think also tortuous vessels)

Systemic – febrile illness (endocarditis), anemia, thrombocytopenia, profound electrolyte disturbances (e.g. hypernatremia in infant), blood transfusion reaction, anticoagulant use

Vascular abnormalities – Macroaneurysm, Familial Retinal Arteriolar Tortuosity

Hemorrhages with white spots (Roth Spots): SBE (Subacute Bacterial Endocarditis), Leukemia, Anemia, Anoxia, CO poisoning, Intracranial hemorrhage, shaken baby, Cerebral malaria, toxoplasmosis, Diabetes

# Sub-retinal Hemorrhages

ARMD with SRNVM (CNVM)

Other causes of CNVM

High Myopia

Angioid Streaks

Histoplasmosis

Trauma

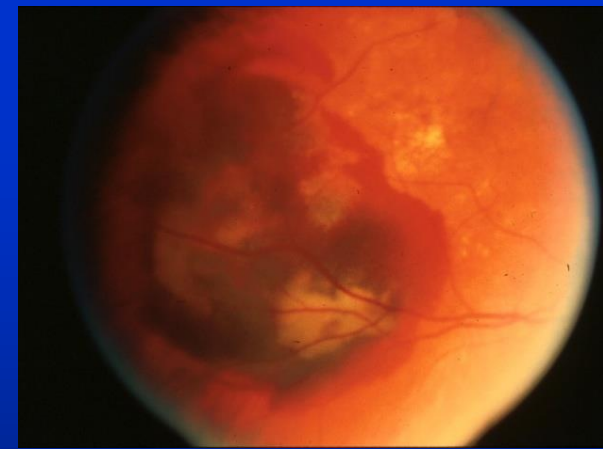
Macroaneurysm

Disc Edema, Papilledema, Disc Drusen (Peri-papillary heme)

Neoplasm – e.g. Melanoma, Choroidal Hemangioma, etc.

( B-scan may be needed in seemingly spontaneous Sub-Retinal hemorrhage)

Peripapillary Sub-retinal Hemorrhages – see list under Disc Hemorrhages



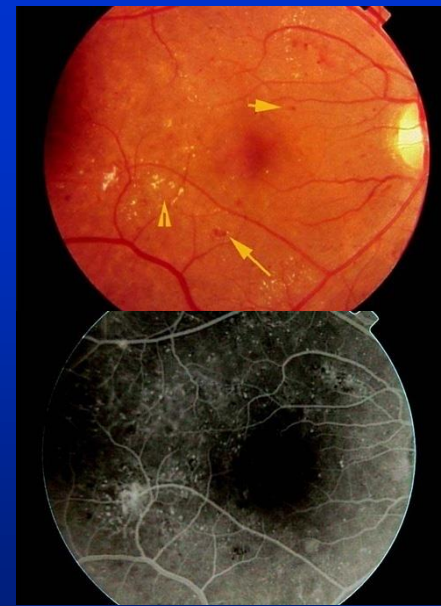
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# Vascular “Retinopathies”

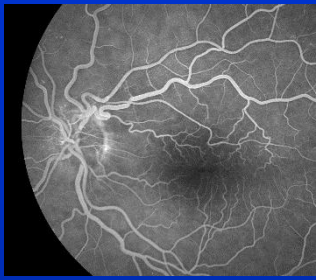
(hemorrhages, exudates, microaneurysms, vessel changes, C/W spots)

- **Hypertension**
- **Diabetes Mellitus**
- **Vasculitis – GCA, SLE, Bechet’s, etc.**
- **Ischemia – OIS (Ocular Ischemic Syndrome)**
  - carotid and ophthalmic artery disease.
- **Blood Disorders**
  - e.g. Sickle Cell, Leukemia
- **Radiation Damage**
- **AIDS**
- **Purtscher’s – bone/ skull trauma, systemic diseases (e.g. pancreatitis)**
- **Other: FEVR (ROP-like in older pts – with temporal avascular retina)**



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# Tortuous or Enlarged Retinal Vessels



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## Tortuous Vessels

- Associated with Congenital Anomalous Disc
- Fabry's Disease
- Retinal Venous Stasis or Obstruction
- Associated with Retinopathy
  - e.g. DR, Sickle Cell disease
- Blood Disorders
- Vascular Abnormality –
  - Retinal Vascular Malformation and or Shunt
  - Carotid Cavernous Fistula
  - Moya Moya Disease
- Membranes – e.g. Epi-Retinal Membrane (ERM)
- Coarctation of Aorta
- Chronic Respiratory Insufficiency
- Newborn fundus

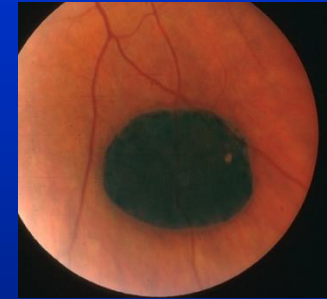
Retinal Venous Malformations  
(Congenital Retinal Macro-vessel)  
Associated frequently with Venous  
Malformations in the Brain  
JAMA Oph 2018;136:372

## Enlarged Retinal Veins

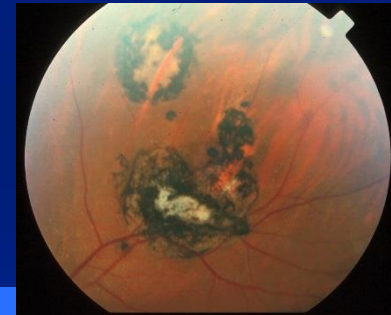
- Impending CRVO
- Papillophlebitis
- Uveitis
- Venous Obstruction – posterior tumor, infiltrative process, Cavernous Sinus Thrombosis
- Cardiovascular – atherosclerosis, decreased arterial supply from carotid blockage, congenital heart disease, temporal arteritis, cardiac insufficiency
- **Increased ICP** – Papilledema
- Blood Diseases – Polycythemia, Sickle Cell and SC Disease, Some Anemias, Leukemias, Macroglobulinemias, (Hyperviscosity)
- Other Systemic – Acute Febrile Illness/ Infections, **Diabetic Retinopathy**, Lupus, Dyslipidemia, Cigarette Smoking, Obesity
- Younger Patients

# Dark or Pigmented Retinal Lesions

- Choroidal Nevus
- CHRPE (multiple lesions suggestive of adenomatous polyposis of the colon)
- Melanoma, and other tumors of Retina or Choroid
- Pigmentary Retinopathy e.g. Bone Spicules (see next list)
- Chorioretinal Scars\*
- Retinal or Choroidal Detachment
- Sub-Retinal Hemorrhage, Choroidal Hemorrhage
- ARMD
- Hemorrhagic Cyst
- Melanocytoma of the Disc
  
- Macula – Bull's Eye



CHRPE



## \*Chorioretinal Scars

Past Chorioretinitis, vasculitis

- Infectious – e.g. Toxoplasmosis

- Autoimmune

Exudative/ Younger - FEVR, Coats, Stargardt's

Exudative / Older – Wet ARMD, Macroaneurysm

Past Retinal Detachment

Past Eye Trauma

Response to Neoplasm, Past Ischemia?

Mimic- Congenital Lesions, Myopic Degeneration

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# Pigmentary Retinopathies

## Congenital

Infectious (e.g. TORCH), Leber Congenital Amaurosis

## Drug Toxicity

e.g. Phenothiazines, Iron overload / Transfusions

## Posterior Uveitis

e.g. Syphilis, *Toxoplasmosis*, VKH, Serpiginous Choroiditis, Nematode (DUSN)

## Retinitis Pigmentosa

Familial - about 50%

Simplex (Sporadic) - about 50%

Associated with Systemic Disease and often Hearing Loss: Usher Syndrome, Refsum Disease (elevated phytanic acid)

**Neoplastic Related:** CARS, MARS, BDUMP (Bilateral Diffuse Uveal Melanocytic Proliferation) – 3 Paraneoplastic syndromes

**Primary Intraocular Lymphoma**, Choroidal Metastatic Disease

**Other:** Vitamin A Deficiency, Siderosis, CPEO, Juvenile Retinoschisis, Metabolic errors

**Hypertensive Retinopathy** - Elschnig's spots (choroidal infarcts)

## Pathologic Myopia

## Bull's Eye Maculopathies →

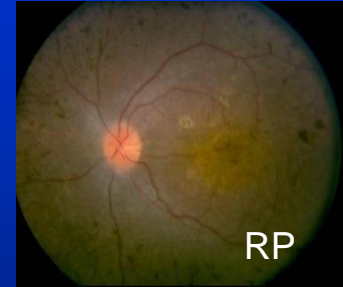
Chloroquine Toxicity

Hydroxy -Chloroquine Toxicity (Plaquenil)

Cone and Cone/Rod Dystrophies

Cone Degenerations

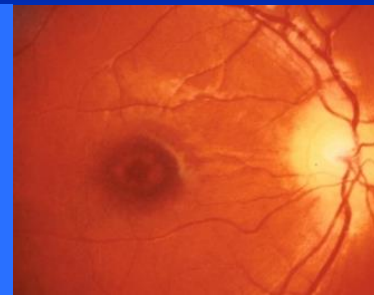
Myotonic Dystrophy



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Bull's Eye Maculopathy



# Rhegmatogenous

- due to retinal break, tear, dialysis
- Associated with trauma, intraocular surgery, high myopia, Choroiditis, Aphakia, Traction

# Tractional

- PDR and Other proliferative retinopathies
- Hemoglobinopathies: Sickle Cell (SSD, SCD), Thalassemia, Eales Disease
- Ischemic CRVO
- PVR (proliferative vitreoretinopathy)
- Fibrosis after prolonged Vitreous Hemorrhage
- Trauma – Penetrating, Surgery
- Other - FEVR

# Exudative

- Systemic Disease : HTN, Toxemia, Renal, SLE, Multiple Myeloma
- Inflammatory Disease: VKH, Scleritis, Sympathetic Ophthalmia, Infectious (TB, Lyme, Syphilis, Dengue, Nematode), Contiguous Orbital Inflammation
- Tumors: melanoma, hemangioma, RB, metastatic disease
- Vascular: macroaneurysm, hemangioma
- Coat's Disease
- Serous : Central Serous Retinopathy (CSR), Leukemia, Juxtafoveal Telangiectasia, Unilateral Acute Idiopathic Maculopathy (acute visual loss and viral prodrome e.g. coxsackie virus)

# Retinal Detachment



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# Retinal / Choroidal - Folds/ Striae

- Hyperopia
- Orbital Tumor
- Thyroid Ophthalmopathy
- ***Posterior Scleritis***\* and sometimes other forms of Orbital Pseudotumor
- Hypotony
- **Chronic Disc Edema, Chronic Papilledema**
- Choroidal - Tumor, Neovascularization, Detachment
- ERM



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# Optic Atrophy / Disc Pallor

Seen with Damage to the Retina (NFL / Ganglion Cells),  
Optic nerve, Optic Chiasm or Optic Tract

## Causes:

**Ischemia** - e.g. past AION or PION

**Compression** - e.g. Pituitary Tumor, Carotid artery, Hydrocephalus, Graves Ophthalmopathy

**Chronic Papilledema** - compression +/- ischemia – see *high ICP list*

**High IOP** - e.g. Glaucoma, Ischemic

**Inflammation** - e.g. Optic Neuritis, MS, Meningitis, Sarcoidosis, Autoimmune, Vasculitis, Infectious

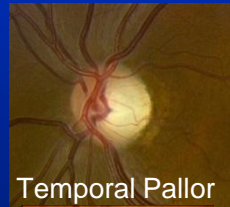
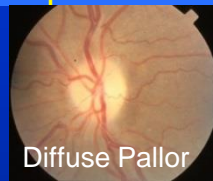
**Trauma** – direct or indirect traumatic optic neuropathy, optic chiasm or tracts

**Toxic / Nutritional Deficit** – e.g. Medications: Ethambutol, Chloramphenicol, Amiodarone, Chemotherapy +/- Radiation, Linezolid, Sildenafil, Anti-TNF $\alpha$ , Interferon, Tacrolimus  
Methanol, Ethylene Glycol, Heavy Metals (Pb, Hg, As, Co, Th), CO, CCl<sub>4</sub>  
Nutritional Deficiencies (B<sub>1</sub> (Thiamine), B<sub>12</sub>, Folate, niacin), Copper (Bariatric Surgery)

**Congenital / Hereditary** – e.g. **Isolated**: Autosomal Dominant Optic Atrophy (ADOA), Leber's (LHON)  
**Non-isolated**: Metabolic, neurodegenerative diseases, Behr's Syndrome  
Friedreich's and Spino- Cerebellar Ataxias  
Associated Hearing Loss: Wolfram's Syndrome (DIDMOAD), Some ADOA

**Degenerative Processes** – e.g. Alzheimer Disease, Vanishing White Matter Disease

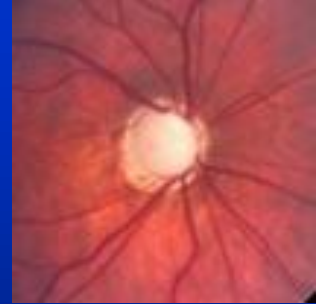
**Retinal Damage** - (False Ocular Localizing Sign) - CRAO, CRVO, Ischemic PDR, S/P PRP, Retinitis; Degeneration (RP)



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## Causes of Optic Disc Cupping or Apparent Cupping

- High Intraocular Pressure (Glaucoma)
- True Normal or Low Tension Glaucoma
- Mimics of Normal / Low Tension Glaucoma
  - e.g. anterior visual pathways lesions, LHON, ADOA, etc.  
(See also slide for Low Tension Glaucoma)
- S/P some Acute Optic Neuropathies – e.g. AION – especially AAION



## Congenital Considerations:

- Morning Glory and other Dysplastic Discs
- Coloboma of Disc
- Tilted Disc
- Megalopapilla – (larger diameter and with same number of axons hence larger cup)
- Optic Nerve Hypoplasia

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## Causes

High ICP - Papilledema

Ischemia – AION

Inflammation / Optic Neuritis

- Classic Demyelinating **Optic Neuritis** associated with or without MS
- Post Viral, Meningitis, Vasculitis, Neuroretinitis, Autoimmune, Infectious (e.g. Toxoplasmosis)

**Infiltrative** – e.g. Leukemia, Sarcoidosis, Lymphoma, Gammopathy

**Compression** – Tumors (e.g. orbital or sellar / parasellar), Large Muscles(e.g. Graves) or Vessel (e.g. carotid a.)

**Hereditary** : e.g. - LHON

**Toxic** - e.g. Methanol, Ethylene Glycol, Chemotherapy, Ethambutol, Anti- TNFa

**Other** – VPT (vitreo-papillary traction- can see disc edema, hemorrhages – OCT of disc)

**Ocular** / False Localizing Sign\*: e.g. not directly optic nerve disease, but coexisting eye problem

Venous stasis (CRVO, BRVO), Hypotony

*Posterior Scleritis, Uveitis*

Acute Multifocal Placoid Pigment Epitheliopathy (AMPPE)

Multiple Evanescent White Dot Syndrome (MEWDS)

**OR**

**MAYBE NOT EDEMA, BUT SOMETHING THAT LOOKS LIKE IT**

e.g. **Anomalous Congenital Disc Elevation** or

Abnormal Disc Vessels or Growths on Disc (see upcoming lists)

# Disc Edema

See also Acute Optic Neuropathy



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# Disc Hemorrhages

## Splinter / NFL / Linear Hemorrhages off Disc

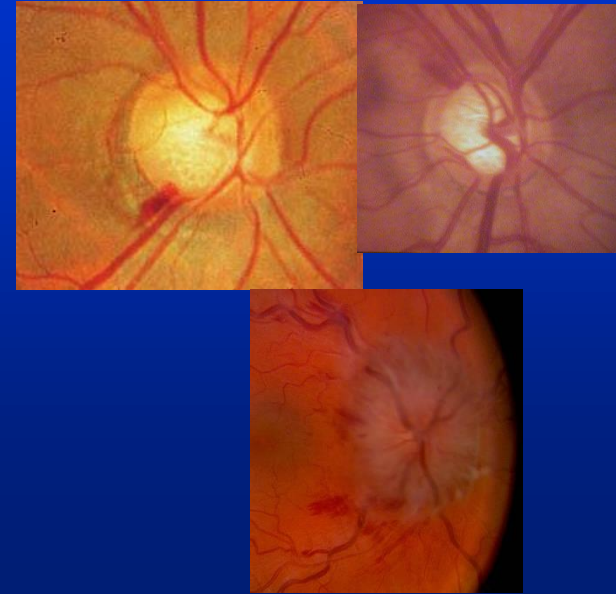
### - Glaucoma – *unstable, progressing*

In patients with Ocular Hypertension a disc hemorrhage suggests increased risk of developing glaucoma

In patients with normal IOP and cupping suggestive of Normal Tension Glaucoma

In healthy pts – suggests possible future glaucoma and should follow them

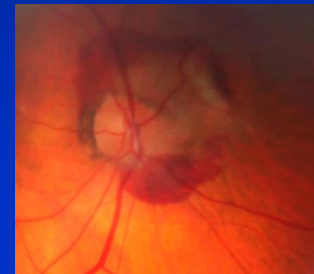
- Disc Edema- e.g. Ischemic Optic Neuropathy, Papilledema
- Optic Disc Drusen
- Local retinal disease – e.g. RVO



## Peripapillary Subretinal Hemorrhages

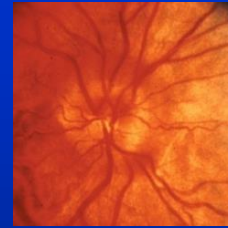
- Disc Edema (papilledema, Uveitis, etc.)
- Disc Drusen and other anomalous discs (e.g. myopic, tilted)
- Juxtapapillary Subretinal Neovascularization (sometimes seen with chronic papilledema)
- Subarachnoid Hemorrhage
- Trauma
- Isolated Peripapillary Subretinal Hemorrhage  
often seen in young myopic women with crowded discs

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# Differentiating between *Congenital* and *Acquired* Disc Elevation

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| Feature            | Congenital        | Acquired                                 |
|--------------------|-------------------|--|
| Nerve Fiber Layer  | Clear             | Opacified                                |
| Large Disc Vessels | Anomalous         | Normal                                   |
| Small Disc Vessels | Normal            | Telangiectatic                           |
| NFL Hemorrhage     | Rare              | Frequent                                 |
| Physiologic Cup    | Small or absent   | Normal<br>(But may be obscured by edema) |
| Drusen             | Sometimes present | Absent                                   |

# Abnormal Disc Vessels and Growths

## Abnormal Vessels

1. ON Sheath Meningioma ( with opto-ciliary shunt vessels)
2. S/P CRVO, BRVO
3. NVD
4. Congenital disc anomaly
5. ONH Drusen (can be differentiated from edema by OCT and FA)\*
6. Disc Edema??
7. ERM or other scarring process



Opto-ciliary  
Shunt  
Vessels

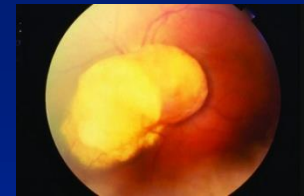


NVD

ERM  
dragging  
disc  
vessels

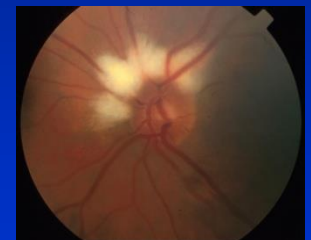
## Abnormal Growths / Remnants

1. Vascular – Hemangiomas – e.g. associated with von Hippel - Lindau Syndrome
2. Meningiomas
3. Gliomas e.g. Astrocytomas – e.g. associated with Tuberous Sclerosis
3. Melanocytomas – usually darkly pigmented
4. Glial Remnants (Bergmeister's Papilla)
5. Adjacent NFL Myelination



Astrocytoma

Transactions of American  
Ophthalmological Society 2004; 102



NFL Myelination

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# Possible Causes of Optic Nerve Inflammation

## Demyelinating:

Multiple Sclerosis, Neuromyelitis Optica = NMO (Devic's Disease)

Post-Viral / Immunization : Acute Disseminated Encephalomyelitis (ADEM)

## Idiopathic

## Viral, Post-Viral, Post-immunization

**Systemic Autoimmune:** Lupus, Behcets, Sjogrens syndrome, "Simple" Autoimmune Optic Neuropathy  
Paraneoplastic Optic Neuritis

## Contiguous Inflammation :

Encephalitis, Meningitis (high ICP and inflammation)

Pachymeningitis – (Idiopathic, intracranial hypotension, Meningeal carcinomatosis, vasculitides, infections such as p. acnes)

Orbit (orbital pseudotumor – e.g. optic perineuritis), IgG4-ROD

Sinuses -Infectious (including Fungal – e.g. mucor, aspergillus), Wegener's Granulomatosis

**Infectious:** Herpes: HSV, VZV, CMV, Syphilis, **Toxoplasmosis**, Cryptococcus, Mycoplasma pneumoniae, RMSF  
Hepatitis A, B, and C, Bartonella / Cat Scratch, Lyme, TB, Measles, Primary HIV, Typhus

**CNS Vasculitis?** – Secondary: Autoimmune, Infectious, Toxic, Neoplastic, **Post-Radiation**  
Primary : Primary Angiitis of the CNS

**Other:** Sarcoidosis, Chronic relapsing Inflammatory Optic Neuropathy (CRION)



Retrobulbar

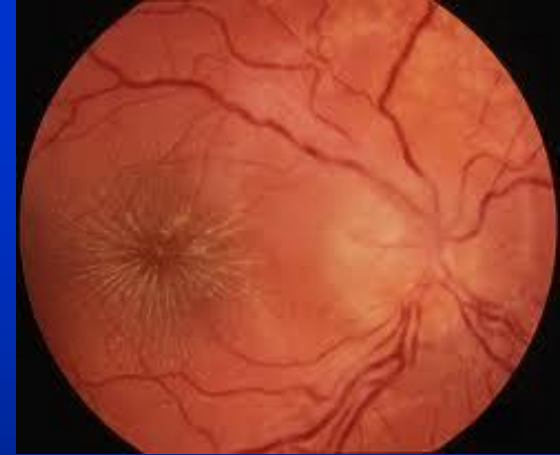
Papillitis

*Remember certain acute optic neuropathies can resemble optic neuritis:  
like: NAION, Compressive Lesions, LHON*

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# Neuroretinitis

## Optic Disc Swelling and Macular Exudates (often a “Macular Star”)



- Leber’s **Idiopathic** Stellate Neuroretinitis
- Cat Scratch Disease (CSD) – from Bartonella Hensalae
- Post-Viral
- Malignant Hypertension, Diabetes, Nephritis
- Sarcoidosis
- Other Infectious besides Bartonella: Syphilis, **TB**, Lyme, **Toxoplasmosis**, HSV, Toxocararisis, West Nile Virus, Nematode (Diffuse Unilateral Subacute Neuroretinitis), Mumps, Salmonella
- Sometimes seen with:  
Papilledema, AION, BRVO, Non-specific Uveitis or other cause of severe disc swelling

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\*Neuroretinitis can be **recurrent** and sometimes require Immunomodulatory or immunosuppressive therapy

- A Negative acute titer is insufficient to exclude diagnosis of CSD. Acute titers can be negative , and so may need to check convalescent titers later – JNO 2012;32:243
- Also Neuroretinitis can occur later after episode of B Henslae infection suggesting autoimmune process. So Idiopathic and Bartonella Neuroretinitis may be related.

# Papilledema and Causes of Increased Intracranial Pressure



1. Hypertension – Malignant
2. Intracranial Tumor (by mere mass effect or by causing obstructive hydrocephalus), Carcinomatous Meningitis
3. Medications -Vitamin A, Accutane, Tetracyclines, Contraceptives, Corticosteroid withdrawal, Thyroid Replacement, Growth Hormone Supplement, ? Fluoroquinolones
4. Toxic: Ethylene Glycol, Lead (Pb), Hyper-ammonemia
5. CNS Inflammation: Meningitis (Bacterial, Viral, Autoimmune / Vasculitis or Medication Induced), Encephalitis (e.g. Lyme, HIV, post-Varicella, Malaria, Abscess) - also HaNDL Syndrome
6. Trauma, Hematomas, Sub - Arachnoid hemorrhage
7. Vascular abnormalities: AVM, AV fistula (e.g. dural AV fistulas after longstanding venous sinus thrombosis)
8. Obstruction to Venous Drainage – Jugular Vein (e.g. paragangliomas-glomus tumors), Vena Cava, Venous Sinus Thrombosis – hypercoagulable states, middle ear or mastoid infections, Erythropoietin abuse
9. Hydrocephalus, Significant Chiari Malformation or Dandy Walker Syndrome, Craniosynostosis
10. Endocrine: Addisons, Hypoparathyroidism, Weight Gain
11. Other: **Sleep Apnea**, Anemia, Thyroid dysfunction, elevated protein levels, POEMS
12. **Idiopathic: Pseudotumor Cerebri or Idiopathic Intracranial Hypertension (IIH)**  
- seen most frequently in young women of child bearing age – including with onset of puberty  
first need to rule out above conditions as well as possible and if you make the diagnosis of IIH,  
then you need to stay open to other causes (like Thrombosis) if pt does respond well to treatment

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## Bilateral Disc Edema

- High ICP / Papilledema
    - maybe, but it could also be...
  - Congenital anomalous discs  
(not edema, mimic)
- 
- Toxic – e.g. Methanol , Ethylene Glycol
  - Medications
    - Amiodarone, Ethambutol, Chemotherapy
  - Compression, Infiltration – but less likely bilateral
  - Simultaneous Bilateral “Optic Neuritis”
    - e.g. Post –Viral, Post-Immunization, Sarcoidosis
  - Simultaneous or Rapidly Sequential AION
    - Shock, Post-op, Trauma, GCA
    - just happens sometimes

## Unilateral Disk Edema

- AION
  - Optic Neuritis  
(but for classic / demyelinating Optic Neuritis  
<1/3 have disc edema)
- 
- Compressive – more likely unilateral
  - Infiltrative
  - Ocular (false localizing sign)
    - e.g. RVO, Hypotony, Uveitis
  - Asymmetric Papilledema - is possible

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# 3. Miscellaneous Case Situations and other Lists

Visual Loss

Reading Problems

Pediatric Presentations: Visual Loss, ET, Glaucoma, Nystagmus

Pregnancy

Systemic Medical Conditions

CN Palsies

Elevated ESR and Concern for Temporal Arteritis

Low Tension Glaucoma

Ocular Effects of Systemic Medications

Imaging Findings- CT, MRI

Dizziness

# Differing Scenarios of Visual Loss

- Longstanding Visual Loss – Undiagnosed
- Acute/ Sudden Visual Loss
- Acute Optic Neuropathy
- Chronic or Progressive Visual Loss
- Unexplained Visual Loss
- Problems with Reading

Of course in any case of visual loss a complete history and exam are necessary and all diagnoses need to be considered, but the following lists can give direction for each scenario.

# Longstanding Visual Loss

Since Childhood or Young Adulthood

## Major Considerations:

### “Front to Back”

- **Refractive**

Unappreciated Refractive Error: especially high Myopia, high Astigmatism, Anisometropia

- **Media Opacities**

Congenital or Developmental Cataracts, Persistent Fetal Vasculature: e.g. PHPV

- **Retinal**

Undiagnosed: Leber's Congenital Amaurosis, Retinal Dystrophies, Achromatopsia, Retinopathy of Prematurity, Congenital and Developmental Inflammations / Scars (e.g. Toxoplasmosis), Myopic Degeneration

- **Optic Nerve**

Optic Nerve Dysplasia, Coloboma, Hypoplasia, Optic Atrophy from Perinatal Insult (e.g. Hypoxia) or hereditary factors  
- e.g. from Dominant Optic Atrophy or Leber's Hereditary Optic Neuropathy

- **CNS**

Amblyopia (Deprivation, Anisometropia or Strabismus),

Perinatal CNS Insult – e.g. hypoxia, intraventricular hemorrhage, etc.

- **Trauma** – to eye or visual pathways:

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# Sudden / Acute Loss of Vision

Developing over Hours to Days

Major Considerations: “Front to Back”

**Refractive:** high glucose level, shift in lens / iris diaphragm, Lens / IOL Dislocation

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**Media Opacities:** Corneal Inflammation or Edema, (e.g. hydrops, high IOP, ulcer), Lens Opacity (e.g. Trauma), Uveitis Cells/ Debris, Endophthalmitis, Vitreous Hemorrhage

**IOP:** High IOP (see previous list) - causing corneal edema, vascular occlusion or low IOP (hypotony)

**Retinal:** Vascular (CRVO, CRAO), Detachment (Rhegmatogenous or Exudative), Inflammatory, SRNVM, Macular Edema, Macular Hemorrhage (sub-retinal, intra-retinal or pre-retinal) Acute Macular Neuroretinopathy

## Timing

Abrupt – ION, LHON

Subacute – optic neuritis

Insidious – compressive or metabolic

**Optic Nerve:** Ischemic Optic Neuropathy, Optic Neuritis, High ICP, Trauma, Compression, Leber’s Hereditary Optic Neuropathy, Toxic

## Character

Dark spot – optic neuropathy

Metamorphopsia - maculopathy

**CNS:** Chiasmal, Tracts or Cortical

CVA (Ischemic or Hemorrhage), Inflammatory, Compressive, Trauma

**Trauma** (Ocular, Orbital, Head Trauma)

**Post-Op** – Endophthalmitis, Retinal Detachment, Cystoid Macular Edema

\*RAPD – very useful in evaluation, especially unilateral visual loss, *see next list*

| Condition   | + RAPD             | - RAPD |
|---|--------------------|--------|
| Classic Optic Neuritis                                  | ★                  |        |
| Retinal Detachment                                      |                    | ★      |
| CRVO  | ★<br>Ischemic CRVO | ★      |
| AION<br>Anterior Ischemic Optic Neuropathy              | ★                  |        |
| CSR<br>Central Serous Retinopathy                       |                    | ★      |
| CRAO  | ★                  |        |
| ARMD<br>Wet ARMD – SRNVM                                |                    | ★      |
| Papilledema<br>High Intracranial Pressure               |                    | ★      |
| Optic Nerve Compression<br>e.g. Tumor or Graves Disease | ★                  |        |
| Vitreous Hemorrhage<br>or Pre-Retinal Hemorrhage        |                    | ★      |
| Traumatic Optic Neuropathy                              | ★                  |        |
| Neuro-retinitis   |                    | ★      |

**RAPD and Acute Unilateral Visual Loss**

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# Acute Optic Neuropathy

(As evidenced by unexplained VA loss, VF loss, RAPD, Disc Changes)

## “Classic” Demyelinating Optic Neuritis:

Related to MS or NMO, Idiopathic, ADEM

## Other Optic Neuritis (Often not classic course / “Atypical”)

Post Viral or Immunization

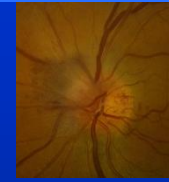
Autoimmune (40-60 yo, responsive to steroids)

Contiguous Inflammation (Meninges, Orbit, Sinuses- e.g. Sphenoid Sinus)

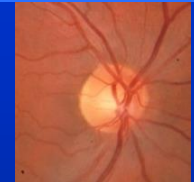
CNS Vasculitis

Infectious: HSV, VZV, Toxoplasmosis, HIV, Bartonella, Cryptococcus, Hepatitis, Syphilis, TB

Other: Sarcoidosis, Optic Perineuritis (IOIS), IgG4-ROD, GBS (rare)



With



or Without

Disc Swelling

## Ischemic

Non-Arteritic Anterior Ischemic Optic Neuropathy - NAION

Arteritic Anterior Ischemic Optic Neuropathy – AAION (GCA)

Posterior Ischemic Optic Neuropathy - PION (peri-operative, arteritic, non-arteritic)

Post-op CE or PPV

## Compressive

e.g. Pituitary Apoplexy, Thyroid Orbitopathy, Carotid Artery, Tumor ...

## Hereditary: LHON

## Acute High ICP

**Traumatic:** Head (Forehead, Temple), Orbit, Globe

**Paraneoplastic:** Associated often with Small Cell Lung CA and CRMP-5 protein

**Medications / Toxins:** e.g. Ethambutol, Chemotherapy, Methanol, Ethylene Glycol

**Radiation Optic Neuritis:** can see months to years after treatment

## Timing

Abrupt – ION, LHON

Subacute – optic neuritis

Insidious – compressive or metabolic

## Character

Dark spot – optic neuropathy

Metamorphopsia - maculopathy

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# Chronic / Progressive Loss of Vision

Developing over Months to Years

## Major Considerations: “Front to Back”

- **Refractive:** regular or irregular astigmatism, progressive myopia, loss of accommodation, nuclear oil droplet changes causing refractive shifts – e.g. myopic
- **Media Opacities:** Most Cataracts, Chronic Corneal Edema, PCO
- **IOP:** Chronically elevated – POAG, Intermittent or Chronic Angle Closure
- **Retinal:** Dry ARMD, Diabetic Retinopathy, Chorio-Retinal degenerations (e.g. RP, choroideremia, gyrate atrophy), Choroidal Tumors – Melanoma, Hemangioma
- **Optic Nerve:** Progressive Atrophy associated with Toxicities, Nutritional Deficiencies, Compression, Drusen, Hereditary: e.g. Autosomal Dominant Optic Atrophy, Glaucoma (high or normal IOP) , Chronic High ICP
- **CNS:** Chiasmal, Tracts or Cortical
  - Compressive or infiltrative neoplasm
  - Neurodegenerative Disorders

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# “Unexplained” Visual Loss”

Loss of Visual Acuity and/or Visual Field without Clear Findings on Exam

## Consider:

- 1. Missed Refractive Errors or Subtle Media Opacities or Distortions**  
*Use Refraction, Retinoscope, Direct Ophthalmoscope, Keratometer, Corneal Topographer*
- 2. Past Retinal Events (RAO, RVO, Commotio, Paraneoplastic) and Past or Subtle Present Macular Disease** (e.g. CME, Dystrophies, Ischemia, Degenerative, ERM, VMT)  
*Use Amsler Grid, OCT and Auto- Fluorescence, Fluorescein Angiography*
- 3. Subtle Optic Neuropathies (often without disc changes early on)**  
- Inherited, Compressive, Toxic, Deficiency, Trauma, Neuro-Degenerative Diseases, Paraneoplastic  
*Use Color Plates, RAPD, Automated VF testing, Orbital or Neuro-imaging , VEP= Visually Evoked Potentials*
- 4. Cortical Visual Loss**  
**CVA**, *Anoxia, Toxic, Metabolic, Vasculitis, Trauma, Migraine, Encephalitis, Meningitis, Dementias, Trauma (SBS)*  
*Use VF Testing, Neuroimaging, LP*
- 5. Other: Amblyopia** (e.g. Anisometropia, Monofixation syndrome)  
**Non-organic / Functional** - *Conversion, Hysteria, Malingering*  
*Use: Prism testing, Tangent VF testing, OKN Drum, Stereoacuity*

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## Color Vision Loss

**Monochromatic** – hereditary – or end stage of any condition below

**Red-Green** (Protan and Deuteran)

- non-specific, seen with Hereditary, Maculopathies and some acquired optic neuropathies

**Blue- Yellow** (Tritan)

specific for vascular retinopathies, papilledema, glaucoma and DOA

Note: Non-organic often associated with some other real ocular problem and the functional loss is an overlay or embellishment of the problem – if focus the patient on treating that associated problem – they may “get better” over time



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## 1. Refractive and Media

High Regular Astigmatism

Irregular Astigmatism – e.g. Keratoconus, see list

Subtle Cataract e.g. Oil Droplet Nuclear Sclerosis, Lens dislocation, High Glucose

## 2. Retinal ( Amsler Grid and ERG Helpful)

Early Pigmentary Retinopathy (RP, toxic), Undiagnosed Leber’s Congenital Amaurosis, Vitamin A deficiency

MARS – melanoma associated retinopathy syndrome or CARS - cancer associated (e.g. lung cancer) – paraneoplastic

Old Trauma with resolved Commotio Retinae, After Vitrectomy (e.g. macular hole surgery), After RAO, RVO (disc shunt vessels?)

**Subtle Maculopathies:** foveal hypoplasia, early macular hole; ERM, CSR, CME – present or **past**, Dry ARMD

Vitreous-macular traction (VMT – OCT ), Ischemia from DR, HIV; Choroidal Infarct, Myopic Maculopathy

Medications – chloroquines, phenothiazines. Cone Degenerations, Cone and Cone/Rod Dystrophies, Cone or Rod Monochromat,

## 3. Optic Nerve, Chiasm, Tracts

Subtle Optic Neuropathies: Low Tension Glaucoma, Dominant Optic Atrophy, LHON, Tobacco/Alcohol

Paraneoplastic, HIV, Subtle Papilledema, Optic Neuropathies associated with Neurodegenerative disorders like Alzheimer’s

Previously unrecognized drusen or hypoplasia, past AION with little pallor

Toxicities: e.g. Methanol, Hg, Pb, As, Bee Sting to face

Medications: Ethambutal, INH, Amiodarone, Tacrolimus, Deferoxamine, Chemotherapy +/- Radiation

Carotid Compression, Meningioma- ONS, sphenoid wing, tuberculum sellae, Optic Nerve Glioma, Compression with Thyroid Orbitopathy

Chiasmal Glioma, MS, Trauma. Optic Tracts – (shunt tips can impinge on structures)

## 4. Cortical Visual Loss: (many problems will have no or minimal findings on Neuroimaging)

CVA not appreciated on MRI/CT, Anoxia, CPR, Hypotension, Carbon Monoxide poisoning, Brain contusion, Toxemia, Meningitis, Encephalitis, Vasculitis – Radiation Vasculitis, Autoimmune. Sarcoidosis, Electrocutation, Occult Neoplasm, Dementias (Alzheimer’s and possibly other causes), White Matter Diseases, CJD= Creutzfeldt-Jakob disease, PML = Progressive Multifocal

Leukoencephalopathy, AIDS, Sleep Apnea; MELAS-Mitochondrial myopathy Encephalopathy Lactic Acidosis and Stoke Syndrome,

Non-ketotic hyperglycemia, Dementias, Procedures: cerebral angiography, ventriculography, blood transfusions. Uremia, porphyria,

syphilis, endocarditis, SSPE – subacute sclerosing panencephalitis – related to measles (rubeola), sudden change in ICP,

Hypoglycemia, correction of hyponatremia, Epilepsy , Chemo and Meds: cis-platinum, tacrolimus, methotrexate, methamphetamine,

vincristine, vindesine, interferon. Posterior Leukoencephalopathy (PLE) seen on MRI seen with hypertensive encephalopathy and

cyclosporine toxicity, Toxic: Nitrous Oxide, ethanol, Pb, Hg, Organophosphates . Migraine patients with permanent VF defects,

occult CVA.. Remote Effect (paraneoplastic) -anywhere along visual pathways. PRES – posterior reversible encephalopathy

syndrome

## 5. Other

Amblyopia – especially think about anisometropic amblyopia, monofixation syndrome

**and consider: Non-Organic (Functional) Visual Loss**

# Unexplained Bilateral or Quickly Sequential Acute Visual Loss

Rapid loss of vision in both eyes simultaneously or sequentially with minimal ocular findings

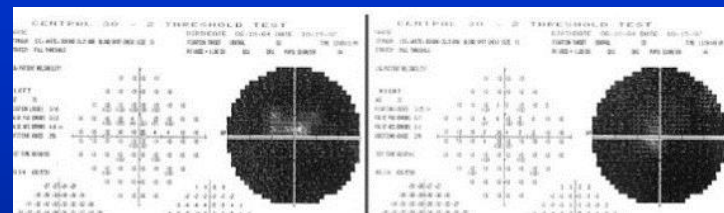
## Vascular

Hypotension – e.g. PION after trauma, surgery, code

Severe Systemic Hypertension

Vertebrobasilar Insufficiency

Temporal Arteritis – e.g. PION



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## Retinal

Paraneoplastic: MARS and CARS\*

## Optic Nerve

LHON

Bilateral / Sequential Retrobulbar Optic Neuritis (e.g. Neuromyelitis Optica (NMO) , MS not as likely)

Other Inflammatory – Post-infectious, Autoimmune, Infectious ON, Meningitis, Vasculitis, Sarcoidosis, GBS

Other Optic Neuropathy – Toxic (e.g. Methanol, Chemo), Nutritional, infiltrative

Paraneoplastic Optic Neuropathy\* (e.g. small cell Lung CA)

PION – e.g. post-op, trauma, shock

## CNS

Migraine

Compressive Lesion – e.g. rapidly expanding like pituitary apoplexy

Cortical Blindness – hypoxia, hypotension, PRES\*, see more complete list under unexplained visual loss

## Other

Sudden Refractive Changes: e.g. loss of accommodation, high Blood Glucose, etc.

### 1. Loss of **Accommodation**

Presbyopia, Diabetes, Latent Hyperopia, see earlier list

### 2. **Centralized Media Opacity** – with miosis e.g. PSC Cataract

### 3. **Strabismus**

e.g. Convergence Insufficiency (primary, head trauma, Parkinson's Disease, ...)

4<sup>th</sup> CN Palsy and other strabismus worse in downgaze, ....

### 4. Problems with **Glasses**

– e.g. bifocal segment position (e.g. too low)

glasses induce diplopia in downgaze (induced prism)

### 5. **Ptosis** – worse on downgaze – typical for levator dehiscence

### 6. **VF loss**

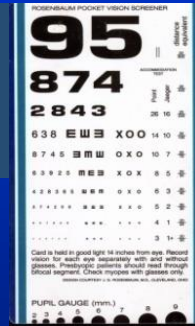
– e.g. Central scotomas, central island of vision or Homonymous Hemianopsia

### 7. **Alexia** – acquired reading problem: from CVA/ lesion in CNS areas of Visual Interpretation

### 8. **Dyslexia** – developmental reading disability

### 9. Other: **Irlen Syndrome** (Scotopic Sensitivity Syndrome)

If NVA is OK, then consider further problems 3-7



### Recall Near Triad

- Accommodation
- Convergence
- Miosis

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# Suspected Loss of Vision in an Infant

## Signs of Poor Vision in Infant:

Absence of blink response to bright light, Overlooking, Oculodigital sign

Poor Fixation, Loss of Opto-Kinetic Nystagmus (OKN) (when spinning with child)

“Congenital Nystagmus” – sign of anterior pathway disease (retinal dystrophy, optic nerve, chiasm, optic tract).  
Onset usually not for 8-12 weeks. Usually in Cortical Visual loss there is no Nystagmus.

Roving Eye Movements – Usually sign of afferent patients with vision less than 20/400



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## First do a complete eye exam looking for more obvious causes

– e.g. refractive error, media opacities, strabismus / amblyopia , retinal changes, optic nerve Atrophy, Hypoplasia, etc. If no obvious findings then consider....

## Considerations:

Delayed Maturation of Visual System

Motor Dysfunction – e.g. Cerebral Palsy with poor eye movements

Leber's Congenital Amaurosis

Retinal Dystrophies e.g. Achromatopsia, Congenital Stationary Night Blindness

Cone Dystrophy

Albinism / Foveal Aplasia

Subtle Optic Neuropathy – Hypoplasia? Atrophy?

Cortical Visual Loss: Peri-natal Hypoxia or Hemorrhage, Hydrocephalus,  
Abuse, Metabolic (hypoglycemia, CO poisoning, uremia)  
Encephalitis, Malformations – Encephaloceles,  
neurodegenerative disorders

These would give appearance of poor vision, yet could still have good vision in each eye

ERG appropriate in suspected Retinal Dystrophy

Neuroimaging needed when see an optic nerve abnormality or cannot explain cortical visual loss

# High IOP Suspect in an Infant

Presenting with Large eye (Pseudo-Proptosis), tearing, corneal clouding

## WHAT DO YOU CONSIDER??

1. Congenital or Juvenile Glaucoma (70% bilateral)
2. Anterior Segment Dysgenesis  
e.g. Peter's Anomaly (Central opacity, 80% bilateral, >50% glaucoma)  
Axenfeld- Reiger's , Aniridia
3. Systemic: Sturge -Weber, Neurofibromatosis
4. Secondary Infantile Glaucomas from  
*Intra-ocular Tumor – e.g. RB (don't forget possibility)*  
*Uveitis / Infectious Process – e.g. Rubella, Toxocara, Herpetic*
5. *Ocular Problems – that may or may not be associated with high IOP:*  
*Megalocornea ( congenital), High myopia*  
*Congenital Corneal Opacity: STUMPED: Sclerocornea, Tears in Descemet's (Congenital Glaucoma),*  
*Ulcers (e.g. intrauterine – viral), Metabolic (e.g. Mucopolysaccharidoses)*  
*Peter's Anomaly, Edema ( Congenital Hereditary Endothelial Dystrophy), Dermoid*



*Don't call all cases like this congenital glaucoma!!*

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*Tearing: Also think: Punctal/ Canalicular Dysgenesis, NLDO, Dacryocystitis, Conjunctivitis*

# Child with Esotropia



1. Congenital / Infantile ET
2. Pseudo-Esotropia – Epicanthal folds, Negative angle kappa, Hypotelorism, Telecanthus

## 3. Hyperopia / Accommodative ET

## 4. Acquired Non-Accommodative ET\*

(most will not have a CNS lesion, but need to consider)

## 5. Duane's Syndrome

6. Cyclic ET – ET on a couple/few days and then off, often pre-school years, sort of like intermittent that eventually becomes constant.

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## 7. Sixth Nerve Palsy – maybe early or partial

Think about high ICP, MS, tumor (orbit, SOF, Cavernous Sinus (inc. aneurysm), Parasellar, Posterior Fossa, Pseudotumor, Microvascular, Head Trauma, Post-immunization, Post Viral

8. Sensory ET ( Visual Loss – think about and look for RB, optic atrophy, etc.)

9. Decompensated Esodeviation, e.g. Monofixation Syndrome (precipitating factors: Hydrocephalus, shunt failure, trauma, ...)

10. Previous Muscle Surgery – consecutive ET or recurrent ET

11. Over Minused Correction (ET near > ET far)

12. Other: Spasm of Near Reflex, Myasthenia Gravis, Any other causes of Abduction Deficit

### Condition

Congenital / Infantile

Accommodative

Late Onset \*

### Common Age of Presentation

< 1 year

18 mo to 3 years

>3-4 years

•need to be concerned for 6<sup>th</sup> CNP - incomitant abduction deficit

•Need to be concerned for AC1 – comitant

•Also suspect high ICP, brainstem / cerebellar lesions, etc.

## 1. Congenital / Infantile Nystagmus

Characteristics – History of Early Onset- sometimes at birth but usually at 2-3 months

Absence of Oscillopsia, Vertigo or Imbalance. Absence of Neurologic abnormalities outside of Visual System

Predominately Horizontal Conjugate Jerk or Pendular Nystagmus, Dampens at Near/Convergence

Increases with attempted Fixation or pursuit. Often a Null point – central or eccentric gaze

## 2. “Eye” Problems – Sensory Visual Loss

e.g. Deprivational Amblyopia,

Congenital Cataracts, Anterior Segment Dysgenesis (e.g. Peter’s Anomaly)

PHPV, ROP, Albinism, Foveal Hypoplasia, Macular Dystrophy or Scar

Retinal Dystrophies: LCA, CSNB, Cone-Rod Dystrophy, Achromatopsia

Optic Nerve Anomaly (e.g. Optic Nerve Hypoplasia, Optic Atrophy)

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## 3. “Neurologic” Problems

Spasmus Nutans – Benign Condition – nystagmus, head nodding and torticollis

Anterior Visual Pathway tumors – hypothalamic, chiasm, optic nerve

Encephalitis, Post Viral Syndrome

Disorders in Posterior Fossa – Tumor, Cerebellar Inflammation

Arnold – Chiari, Hydrocephalus, Spinal Cerebellar degeneration

Neurodegenerative Disorders- e.g. Leigh Disease, Pelizaeus-Merzbacher Disease, Joubert Syndrome

Metabolic: Malnutrition, Maple Syrup Urine, Hypothyroid

Other: Trauma, Down’s Syndrome, Medications and Toxins

Ospoclonus – not true nystagmus, can be herald of Neuroblastoma, acute cerebellar ataxia

# Pregnancy and Vision and Eye Conditions

## Exacerbated

CVA  
Retinal Vascular Occlusion } Thrombo -  
Embolic  
Meningioma- e.g. Sphenoid wing  
Pituitary Apoplexy  
High ICP: Venous Sinus Thrombosis

Pseudotumor Cerebri Exacerbated

Migraine

Diabetic Retinopathy (DME)

CSR

Cranial Neuropathies: 7<sup>th</sup> most common

Orbital

Spontaneous Orbital Hemorrhage

Orbital Varices

Refractive Errors

Myopic Shifts

Loss of Accommodation

Relative Immunocompromised

(Reactivation of Uveitis?)

## Pre-Eclampsia and Eclampsia

Retinopathy and

Serous/Exudative RD

High ICP

AION

CVA

Cortical Blindness

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## Post-Partum

Post Partum Optic Neuritis

Intracranial Hypotension and

Acquired Chiari - 1 Malformation  
(Headache, Diplopia)

## Associations For Baby

Prematurity – ROP, ET, XT

Maternal Smoking – ET and XT

Maternal Alcohol Use → Fetal alcohol syndrome

- Telecanthus, ON Hypoplasia, Strabismus



# Important Medical Conditions and their Associated Eye Pathology

## 1. Smoking (Tobacco) - exacerbates or increases risk of developing ..

ARMD, Graves Ophthalmopathy, Optic Nerve Atrophy, AION, DR, Cataracts, Uveitis

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## 2. Diabetes Mellitus

Early Presbyopia, Non reactive pupil, CNP (especially 3<sup>rd</sup> and 6<sup>th</sup>), Cataract, DR, AION

## 3. Uncontrolled Systemic Hypertension - Retinopathy / Maculopathy, High ICP and Papilledema, CN Palsies

## 4. Obstructive Sleep Apnea

AION, RVO, High ICP, OAG, Floppy Eyelid Syndrome

## 5. Extended Use of Systemic Corticosteroids

Cataracts, High IOP, CSR , High ICP (with withdrawal)

Potential of Some Viral, Bacterial, Fungal Infections

## 6. Liver Disease: Conjunctival and periocular skin icterus; Hep C – association with MALT lymphoma of ocular adnexa; hepatocellular CA metastatic to orbit, angiomas of periocular skin in cirrhosis. Wilson Disease; KF rings, sunflower cataract, motility problems

## 7. Immunosuppression (Alcoholism / poor nutrition ; Inherited and Acquired (e.g. HIV))

– Corneal Ulceration, Severe Viral (e.g. CMV, ARN), Fungal (e.g. Mucor) and Protozoal (e.g. Toxoplasmosis) Infections

# Non-Pathologic changes in the Eye with Aging

1. Neuroretinal age-related loss (measured by confocal scanning and OCT - RNFLT) – occurs and could explain apparent progression in some glaucoma patients ( see Oph 2015;122:2392 for incidence and rate)
2. Pupil Size - decreasing pupil size (linear) in both light and dark with age. Pupil diameter gets down between 2-4 mm under normal lighting situation for elderly. See IOVS 1994;35:1132
3. 10-15% of Caucasian population experience change in eye color as they age – due to loss of melanin pigmentation in the iris

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# Cranial Nerves Palsies

Related to Ocular Motility

## Possible Causes

**Microvascular** (older age, High BP, DM)

**Diabetic** – mainly 3<sup>rd</sup> and 6<sup>th</sup>

Trauma – closed head, basilar skull fracture

Compressive – Aneurysm/Herniation – especially 3<sup>rd</sup>

Tumor – Compression or Infiltration

Ischemic CVA – Brainstem

High Intracranial Pressure – especially 6<sup>th</sup>

Low ICP (Intracranial Hypotension) – 6<sup>th</sup> most common

CNS Inflammatory – e.g. Meningitis,...

MS / Demyelination – especially 6<sup>th</sup>

Congenital – especially 4<sup>th</sup> Nerve

Vasculitis – e.g. GCA

Migraines\*

Systemic - immune mediated polyneuropathy (e.g. GBS, MFS)

Shunt Failure

## Cranial Nerve

3<sup>rd</sup> Oculomotor

4<sup>th</sup> Trochlear

6<sup>th</sup> Abducens

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**Approach:** If it is **isolated** (no other neurologic signs or symptoms) , then observation is often OK, but **Follow-up** of the patient to look for improvement or stability is key. If the diagnosis is a microvascular CNP you must make sure on follow-up that the palsy is resolving or resolved in 2-4 months. IF the patient has more **pain, progression or pupil dilation**, then consider early neuroimaging. A 3<sup>rd</sup> or 6<sup>th</sup> CN palsy in a younger person (<50 yo) is also a reason to do neuroimaging earlier. **In older patient with significant pain ask about Symptoms of GCA and get ESR.**

# Cranial Nerve Palsies

More Specific Causes

## 3<sup>rd</sup> CNP

1. Microvascular
2. R/O PCOM or other aneurysm
3. Traumatic
4. Compressive / Tumor
5. Herniation
6. Ophthalmoplegic Migraine
7. Other: GCA, Pituitary Apoplexy
8. Congenital, Cyclic 3<sup>rd</sup> CNP Paresis

## 6<sup>th</sup> CNP

1. Microvascular
2. Traumatic
3. Demyelinating
4. High ICP, sometimes Low ICP
5. Post Viral, Post Immunization
6. Compressive - Tumor – Cavernous Sinus, Clivus / Skull based tumor
7. Carotid Cavernous Aneurysm
8. Pontine Hemorrhage or CVA
9. Other: Meningitis, GCA
10. Congenital

## 4<sup>th</sup> CNP

1. Traumatic
2. Congenital
3. Microvascular
4. Other Causes - uncommon

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# Elevated ESR

Ophthalmologists are confronted with patients sometimes in whom they **suspect Temporal Arteritis or Giant Cell Arteritis (GCA)**

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1. *R/O GCA (usually older patients >60 yo)*
2. **Anemia** - anemic patient can have a artificially elevated ESR secondary to anemia
3. **Systemic inflammatory conditions** – Polymyalgia Rheumatica, Lupus, Vasculitis  
*Younger Patients with an Autoimmune Disease can present with an Acute Optic Neuropathy*
4. **Cancer** - may need work-up for an occult CA,  
(note some patient with cancer are anemic)
5. **Hyper –proteinemias** - e.g. Multiple Myeloma, Gammaglobulinemias
6. **Active Infection:** Sinusitis, Ear Infection, Mastoiditis, Dental (all could produce pain in area of Temporal Arteritis)  
Wound infection, Prostatitis, Osteomyelitis
7. **Other:** End Stage Renal Disease (ERSD) / Dialysis, Idiopathic Hypertrophic Cranial Pachymeningitis

Suggested work-up for patient with no obvious cause for an elevated ESR:

C-Reactive Protein, CBC (with platelets) , Urinalysis, BUN/Cr, alkaline phosphatase,  
Serum Protein Electrophoresis, Chest X-Ray, CT of Sinuses, *or a temporal artery biopsy?*

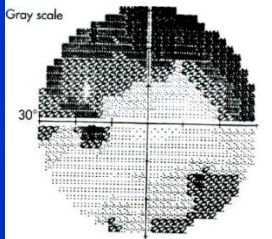
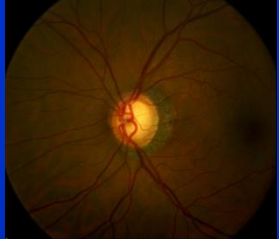
## Temporal and Forehead Pain and Tenderness

Think about GCA yes, but also consider:

- Early / Prodrome VZV / Shingles
- Trigeminal Neuralgia
- Sinus, Ear, Mastoid, Dental Infections
- Other neuralgias – Occipital, Cervical, Supra Orbital
- Unilateral Headaches – Migraine, Cluster, Tension

## Beware of Artificially Low ESR values in Patients with Biopsy Positive GCA

e.g. Pts on Statins and Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) can have lower ESR, but not CRP. (JNO 2011;31:135)



# Glaucomatous Disc Changes and / or Glaucomatous VF loss in the face of normal IOP

Before making the diagnosis of

## - Low / Normal Tension Glaucoma\*

### consider also

- *Anterior Visual Pathway Lesion*  
*e.g. Parasellar Tumors, ON or sheath tumors, etc.*
- *Previous Optic Nerve Damage from:*  
*ACG, Previous Open Angle Glaucomas,*  
*AION, AAION, Papilledema or Optic Neuritis, BRAO, LHON*
- *Carotid Artery Disease*
- *Congenital Disc Defects – pits, drusen, tilted or myopic discs*
- *Temporal Disc Pallor can be mistaken for cupping:*  
*Hereditary (DOA) or Acquired Optic Atrophies (Toxic, Nutritional, Tobacco)*

Evaluation should include:

Good History can eliminate several possibilities

Careful exam of disc

Rim pallor more suggestive of another optic neuropathy

Could also include:

- Corneal Pachymetry – thin cornea could give falsely low IOP
- Diurnal Curve- IOP might fluctuate during day in patient with POAG

More likely Glaucoma if :

- Older patient
- Disc hemorrhages
- VF defects that respect the horizontal midline

Increased suspicion of a CNS lesion if: 1) VF loss respecting the vertical midline, 2) rim pallor, 3) mismatch of amount of cupping with VF loss, 4) RAPD, 5) younger patients 6) pituitary dysfunction, 7)

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- Illusions and Hallucinations:

- Psychogenic Medications and Drugs -

- Dopaminergics: levadopa, bromocryptine,

- Anticholinergics: atropine, scopolamine, cyclopentolate

- Tricyclic Antidepressants, Beta-Blockers

- Adrenergic – albuterol, Phenylephrine, Theophylline, Amphetamine, Cocaine

- Alcohol

- Misc: Benzodiazepines, corticosteroids, NSAIDs, Ca Channel blockers, Narcotics, anticonvulsants,...

- Trazodone, Nefazodone, **Topiramate**, Risperidone, Clomiphene

- Illicit Drugs: LSD, Cannabis, Methamphetamine, Psilocybin (mushrooms)

- Transient Refractive Shifts

- Myopic – Topiramate

- Hyperopic - Phenothiazides, Antihistamines, Chloroquine, Anticholinergics, Cannabis

- Cataract

- Corticosteroids (PSC), Statins, Phenothiazines (Chlorpromazine)

- Angle Closure Glaucoma – increased risk

- Ant-histamines, Anti-psychotics, Tri-cyclic Antidepressants, Ephedrine

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- Retinal Pigmentary Changes

- Phenothiazines - Thioridazine (Mellaril), Chlorpromazine (Thorazine), Chloroquine, Hydroxychloroquine

- Tamoxifen, Indomethacin, Deferoxamine

- **Papilledema / Increased Intracranial Pressure**

Vitamin A , Retinoids e.g. Isotretinoin (Accutane) , Tetracyclines: Doxycycline, Minocycline, Lithium, Human Growth Hormone, Thyroid supplementation, Nalidixic Acid, *Oral Contraceptives*, *Withdrawal of Corticosteroids*,

- **Nystagmus**

Phenytoin, Barbituates, Lithium, Salicylates, Antihistamines

- **Loss of VF and/ or NFL**

Vigabatrin

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- **Optic Neuropathy**

Amiodarone, Ethambutol, INH, Quinine, Sulfonamides, Chloramphenicol, SSRI's

Anti TNF Ab (Remicade, Embrel), Tacrolimus

Interferon; Chemotherapy (Doxorubicin, Cisplatin, Methotrexate, Vincristine) especially with Radiation;

Lindane Shampoo (absorbed)

- **Myasthenia Gravis (Induced, Aggravated or Mimic)**

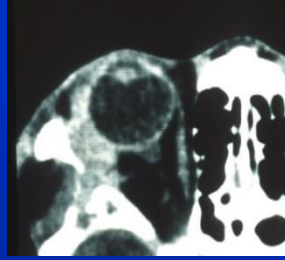
- Statins (3HMG CoA reductase inhibitors)

Also consider: Ca Channel Blockers (e.g. Verapamil), Beta Blockers (e.g. Propranolol or Timolol), Anti-Arrhythmics (e.g. procainamide), Quinine, chloroquine, Penicillamine, Corticosteroids, Antibiotics (e.g. Fluoroquinolones, Aminoglycosides, Macrolides), Anti-Seizure (e.g. phenytoin, carbamazepine and Gabapentine), Chemotherapy, Interferon, Some IV Anesthetics,



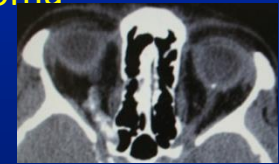
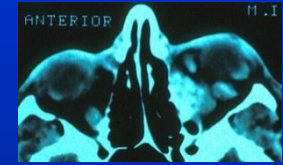
## Boney Erosion / Destruction

- LG Carcinomas
- Recurrent Pleomorphic Adenoma
- Neurofibromatosis
- Metastatic Disease in Orbit
- Dermoid Cyst
- Sinus Mucocele
- Histiocytosis (e.g. Eosinophilic Granuloma)
- Wegener's (GPA) involving sinus or orbit
- Angiosarcoma
- Infectious: TB, Syphilis
- Hyperparathyroidism
- Gorham- Stout Disease (vanishing bone, osteolysis)



## Calcifications

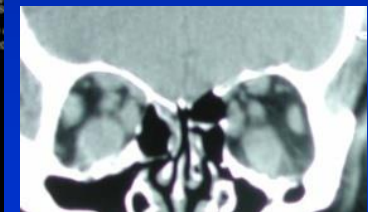
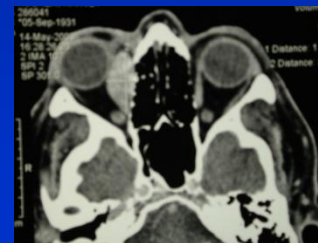
- Orbital Varix (Phleboliths)
- Optic Nerve Sheath Meningioma
- Ocular Tumor – e.g. RB
- Optic Nerve Head Drusen



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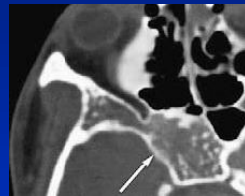
## Enlarged Extra Ocular Muscle(s)

- Graves Ophthalmopathy (IR, MR > SR most common)
- Orbital Pseudotumor (Myositis)
- Post- Viral Myositis (Children), Cysticercosis
- Lymphoma, CLL
- Metastatic Disease – breast, melanoma
- Primary Tumor – e.g. hemangioma, fibrous / Amyloid



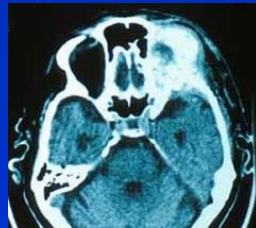
## Boney Growth / Change / Expansion

Fibrous Dysplasia →



Sarcoma of the bone

Metastatic Disease to the Bone →

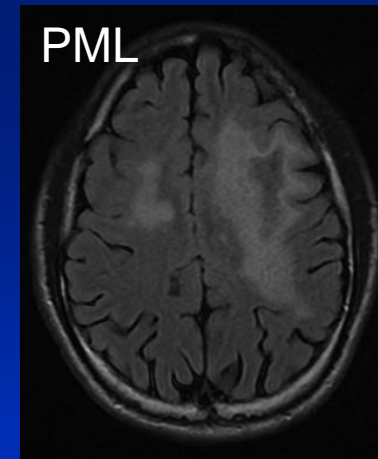
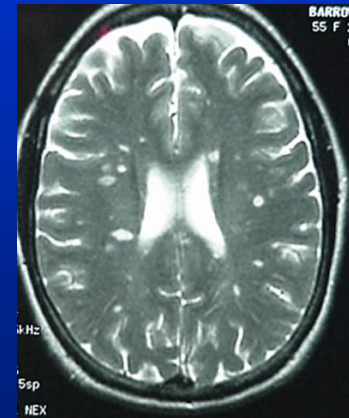


Paget's Disease

# MRI of the Brain (White Matter Lesions)

## White Matter Lesions

- Demyelinating Disease
  - MS
  - ADEM = Acute Disseminated Encephalomyelitis ( post viral or immunization)
- Older patients with any small vessel disease particularly with DM, HTN
- Migraine patients, Chronic Migraine
- Vasculitis -acute and chronic small vessel disease of brain
- Younger patients with uncontrolled or poorly controlled HTN
- Anoxic Encephalopathy – e.g. CO poisoning
- Primary Angiitis of the CNS
- Anti - Phospholipid Antibody Syndrome
- Rapid Correction of Hyponatremia (Osmotic Demyelination Syndrome)
- AIDS
- PML (Progressive Multifocal Leukoencephalopathy – viral disease in immunocompromised)
- Radiation



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## Vertigo - Peripheral or Central causes – associated with nystagmus

**Peripheral:** (BPV) (seconds < minute associated with head posturing, no tinnitus, hearing OK)

Acute Labyrinthitis (hours to 2 weeks, no tinnitus, hearing OK)

Meniere's Disease (hours, +hearing loss, +tinnitus)

Drug Toxicity (Aminoglycosides, anti-seizure meds, alcohol, ASA)

Superior Canal (labyrinth semicircular) Dehiscence Syndrome

- Loud noises cause patient to suffer sudden transient vertigo and blurred vision

**Central:** CPA tumor e.g. Acoustic Neuroma (hearing loss, tinnitus, other CN deficits)

Ischemia, Stroke involving Vertebral basilar circulation

MS

Acute Cerebellar Disease

Vestibular Migraine

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## Disequilibrium – elderly issues like fear of walking, musculoskeletal problems, peripheral neuropathy

Consider also eye Problems: visual loss or causes of oscillopsia and diplopia or problem with glasses

**Pre-syncope** – Orthostatic Hypotension from medications, arrhythmias, vaso-vagal

**Psychiatric** – anxiety, hyperventilation, depression, substance abuse

**Multifactorial or unknown**

**Acute Vestibular Syndrome** – rapid onset of vertigo, N/V, nystagmus, unsteady gait, head motion intolerance, most commonly due to vestibular neuritis or posterior fossa stroke.

**Shopping Cart Syndrome** (B. Farris – COS 2013) – symptoms of dizziness in patients (with prior BPV or labyrinthitis) – when in an environment which can induce OKN – such as pushing shopping cart down visually busy grocery aisle.

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## List of Abbreviations

AC - Anterior Chamber  
AC1 – Arnold Chiari Malformation Type 1  
ACE – Angiotensin Converting Enzyme  
ACG – Angle Closure Glaucoma  
AD – Autosomal Dominant  
ADOA – Autosomal Dominant Optic Atrophy  
AIBSES – Acute Idiopathic Blind Spot Enlargement Syndrome  
AIDS – Acquired Immune Deficiency Syndrome  
AION – Anterior Ischemic Optic Neuropathy  
AAION– Arteritic AION (GCA)  
AK – Astigmatic Keratotomy  
AMPPE – Acute Multifocal Posterior Placoid Epitheliopathy  
ANA – Anti-Nuclear Antibody (Lupus Diagnosis)  
AR – Autosomal Recessive  
ARMD – Age Related Macular Degeneration  
ARN – Acute Retinal Necrosis  
AVM – Arterial Venous Malformation  
BEB – Benign Essential Blepharospasm  
BCCA – Basal Cell Carcinoma  
BCP – Birth Control Pills, Contraceptives  
BDUMP – Bilateral Diffuse Uveal Melanocytic Proliferation  
BP – Blood Pressure  
BPES - Blepharophimosis- Ptosis- Epicanthus Inversus Syndrome  
BPV – Benign Positional Vertigo  
BRAO- Branch Retinal artery Occlusion  
BRVO- Branch RVO  
CA – Carcinoma  
CARS – Cancer Associated Retinopathy Syndrome  
CC – Carotid Cavernous  
CE – Cataract Extraction (Surgery)  
CFEOM – Congenital Fibrosis of Extraocular Muscles  
CHRPE – Congenital Hypertrophy of the RPE  
CIN – Carcinoma in Situ  
CLL – Chronic Lymphocytic Leukemia

CME – Cystoid Macular Edema  
CMV – Cytomegalovirus  
CN- Cranial Nerve  
CNP – CN palsy  
CNS – Central Nervous System  
CPA – Cerebellar Pontine Angle  
CPEO – Chronic Progressive External Ophthalmoplegia  
CR – Chorioretinal  
CRAO- central retinal *artery* occlusion, CRVO- central ret. *vein* occlusion  
CSR – Central Serous Retinopathy  
CSNB – Congenital Stationary Night Blindness  
CT- Computerized Tomography  
CTA – CT Angiogram  
CTL – Contact Lens  
CVA – Cerebral (CNS) Vascular Accident (Stroke)  
CXR – Chest X-Ray  
DDX – Differential Diagnosis  
DOA – Dominant Optic Atrophy  
DR – Diabetic Retinopathy  
DME- Diabetic Macular Edema  
DM – Diabetes Mellitus  
DUSN – Diffuse Unilateral Subacute Neuroretinitis from Nematode  
EOM – Extra Ocular Muscle  
ERG – Electroretinogram  
ERM – Epi - Retinal Membrane  
ESR – Erythrocyte Sedimentation Rate  
ET – Esotropia  
FA- Fluorescein Angiogram  
FB- Foreign Body  
FBS – Foreign Body Sensation  
FEVR - Familial exudative vitreoretinopathy  
FHx – Family Medical History  
GBS- Guillain-Barre Syndrome  
GCA – Giant Cell Arteritis (aka Temporal Arteritis)

GPA- Granulomatosis with Polyangiitis (Wegener's)

HA - Headache

HIV – Human Immunodeficiency Virus (AIDS)

HLA – Human Leukocyte Antigen

HSV – Herpes Simplex Virus

HT – Hypertropia

HoT - Hypotropia

HTN – Hypertension (high blood pressure)

HZO- Herpes Zoster Ophthalmicus

IBD - Inflammatory Bowel Diseases

ICE – Iridocorneal Endothelial (Syndrome)

ICP - Intracranial Pressure

IgG4-ROD – Immunoglobulin G4 Related Ophthalmic Disease

IIH – Idiopathic Intracranial Hypertension (pseudotumor cerebri)

INO – Internuclear Ophthalmoplegia

IO – Inferior Oblique

IOFB – Intra-Ocular Foreign Body

IOIS – Idiopathic Orbital Inflammatory Syndrome

IOL – Intraocular lens

ION- Ischemic Optic Neuropathy

IOP – Intraocular Pressure

IR – Inferior Rectus

JIA – Juvenile Idiopathic Arthritis

JRA - Juvenile Rheumatoid Arthritis

KC – Keratoconjunctivitis

KP – Keratoprecipitates

LASIK – Laser Assisted In Situ Keratomileusis

LCA – Leber's Congenital Amaurosis

LHON – Leber's Hereditary Optic Neuropathy

LG – Lacrimal Gland

LL – Lower Lid (Lower Eyelid)

LLL – Left Lower Eyelid

LP – Lumbar Puncture (Spinal Tap)

LR – Lateral Rectus

LSCD- Limbal Stem Cell Deficiency

LUL – Left Upper Lid

MARS – Melanoma Associated Retinopathy Syndrome

MEWDS – Multiple Evanescent White Dot Syndrome

MFS – Miller Fisher Syndrome (polyneuropathy)

MG – Myasthenia Gravis

MM – Multiple Myeloma

MR – Medial Rectus

MRA – Magnetic Resonance Angiography

MRD – Margin Reflex Distance (Upper Eyelids)

MRI - Magnetic Resonance Imaging

MRV- MR Venography

MS – Multiple Sclerosis

NAION- Non-Arteritic AION

NF - Neurofibromatosis

NFL – Nerve Fiber Layer

NLO – Nasolacrimal Obstruction

NLDO – NL Duct Obstruction

NPDR – Non-Proliferative Diabetic Retinopathy

NVA- Near Visual Acuity

NVD – Neovascularization of the Disk

NVI – Neovascularization of the Iris

OAG – Open Angle Glaucoma

OCP – Ocular Cicatricial Pemphigoid

OCT – Ocular Coherence Tomography

OIS – Ocular Ischemic Syndrome

OKN – Opto-Kinetic Nystagmus

ONH – Optic Nerve Head

OMG – Ocular Myasthenia Gravis

ONM – Ocular Neuromyotonia

OSSN – Ocular Surface Squamous Neoplasia

PAN – Polyarteritis Nodosa

PC – Posterior Chamber

PCO - Posterior Capsular Opacity (Pseudophakic eye)

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PDR – Proliferative Diabetic Retinopathy  
PEK – Punctate Epithelial Keratopathy = SPK  
PET-CT - Positron emission tomography–computed tomography  
PHPV – Persistent Hyperplastic Primary Vitreous  
PPM – Persistent Placoid Maculopathy  
PPV – Pars Plana Vitrectomy (Surgery)  
PI – Peripheral Iridectomy or Iridotomy  
PION – Posterior Ischemic Optic Neuropathy  
PK – Penetrating Keratoplasty (corneal transplant)  
POAG – Primary Open Angle Glaucoma  
PON – Paraneoplastic Optic Neuropathy  
PORN - Progressive Outer Retinal Necrosis  
PRK- Photo- Refractive Keratectomy  
PRP- Pan-Retinal Photocoagulation  
PSC – Posterior Sub-Capsular (Cataract)  
PUK- Peripheral Ulcerative Keratitis  
PVD – Posterior Vitreous Detachment  
RA – Rheumatoid Arthritis  
RAO – Retinal Artery Occlusion  
RAPD – Relative Afferent Pupillary Defect  
RB – Retinoblastoma  
RBC – Red Blood Cells  
RD – Retinal Detachment  
RES – Recurrent Erosion Syndrome  
RLL – Right Lower Lid  
RGP – Rigid Gas Permeable (CTL)  
RK – Radial Keratotomy  
RMSF- Rocky Mountain Spotted Fever (Rickettsiae)  
R/O – Rule Out  
ROP- Retinopathy of Prematurity  
RP- Retinitis Pigmentosa  
RPE – Retinal Pigment Epithelium

RUL – Right Upper Eyelid  
RVO – Retinal Vein Occlusion  
SAH – Subarachnoid Hemorrhage  
SBS – Shaken Baby Syndrome  
SCH – Sub-Conjunctival Hemorrhage  
SCCA – Squamous Cell Carcinoma  
SJS – Stevens Johnson Syndrome  
SLK – Superior Limbic Keratoconjunctivitis  
SLE- Systemic Lupus Erythematosus  
SNP – Supranuclear Palsy  
SO – Superior Oblique  
S/P - Status Post  
SPK - Superficial Punctate Keratitis  
SR – Superior Rectus  
SRNVM – Sub-Retinal Neovascular Membrane  
TASS – Toxic Anterior Segment Syndrome  
TB – Tuberculosis  
TBI- Traumatic Brain Injury  
TM – Trabecular Meshwork  
TORCH – (Toxoplasmosis, Other, Rubella, Cytomegalovirus, Herpes Simplex)  
  
UA – Urine Analysis  
UGH – Uveitis Glaucoma Hyphema Syndrome (Pseudophakic eye AC or PC IOL)  
  
UL – Upper Lid (Eyelid)  
URI – Upper Respiratory Infection  
VA – Visual Acuity  
VF - Visual Field  
VKH - Vogt-Koyanagi-Harada Syndrome  
VMT - Vitreo -Macular Traction  
VPT – Vitreo-Papillary Traction  
VZV - Varicella -Zoster Virus  
WBC – White Blood Cells  
W/U – work up  
XT - Exotropia

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Return To Links](#)

# Major References

|   | <u>Medium</u> |
|---|---------------|
| Hampton Roy, Ocular Differential Diagnosis  | Book          |
| American Academy of Ophthalmology – Basic and Clinical Science Series                   | Monograph     |
| Emedicine Ophthalmology – Multiple articles from website<br>EyeRounds.org, AAO- EyeWiki | Internet      |
| Survey of Ophthalmology – Many of the Major Reviews                                     | Journal       |

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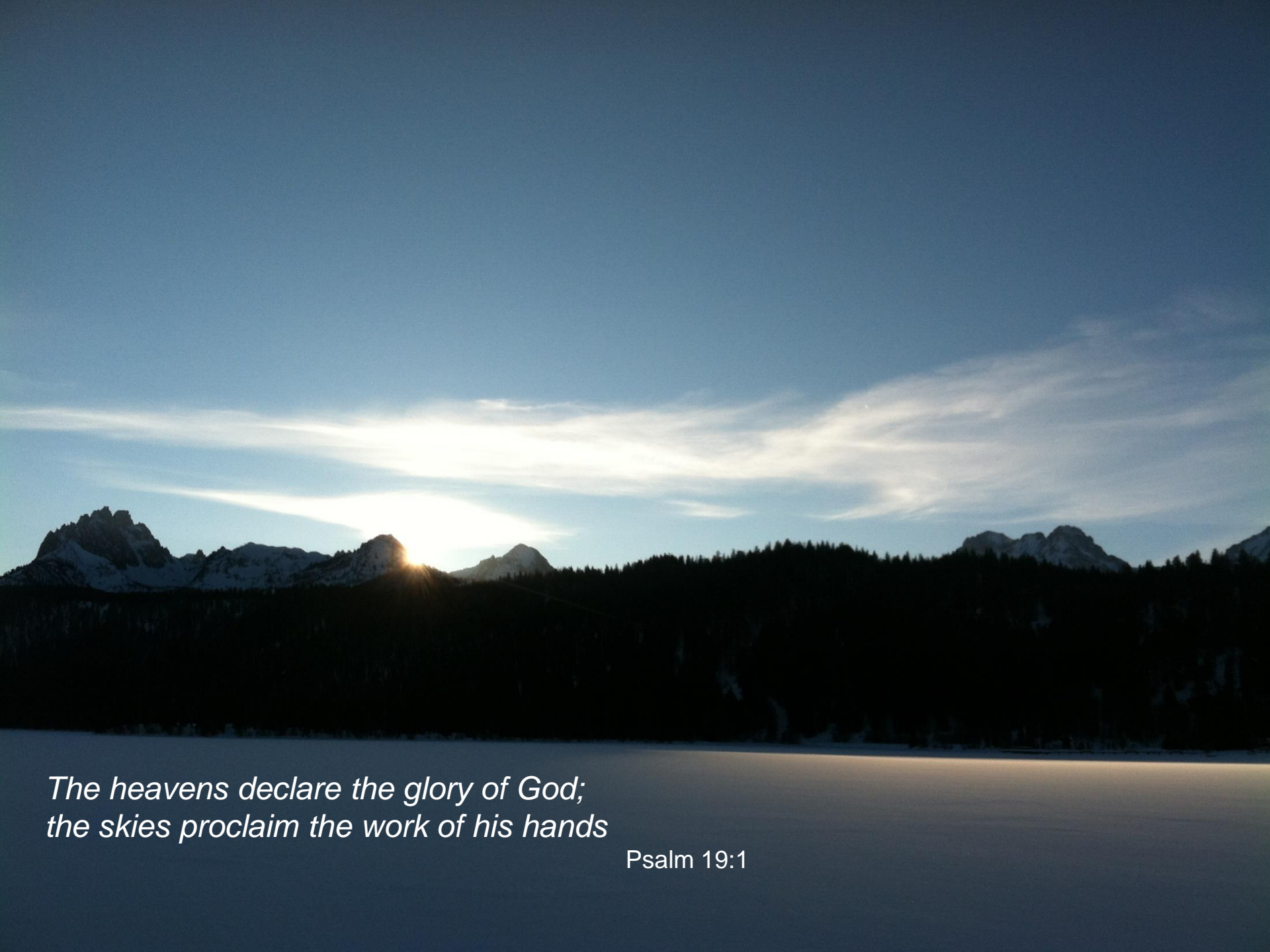
- 1) Personal Collection
- 2) University of Iowa site – uiowa.edu
- 3) Redatlas.org
- 4) University of Michigan , Kellogg Eye Center
- 5) Some images seem to be in wide circulation and have no credit given, but if any image is seen and recognized to be copyrighted and needs to be removed or given proper credit – please contact me.



# Journal References

|  | <u>Abbreviation Used</u> |
|--|--------------------------|
| Ophthalmology (AAO Journal)                    | OPH                      |
| Survey of Ophthalmology                        | Survey                   |
| JAMA Ophthalmology (Archives)                  | JAMA Oph                 |
| American Journal of Ophthalmology              | AJO                      |
| Journal of Neuro-ophthalmology                 | JNO                      |
| Investigative Ophthalmology and Visual Science | IOVS                     |





*The heavens declare the glory of God;  
the skies proclaim the work of his hands*

Psalm 19:1