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

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October 2018

* Please see references at end of document
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.

At the end is also a list of abbreviations used in the presentation.
Symptoms
Loss of Vision
Transient Visual Phenomena
Floaters
Flashes, Photopsias
Diplopia
Monocular Diplopia
Binocular Diplopia – Ocular Misalignment
Transient Diplopia
Oscillopsia
Night Blindness
Transient Visual Loss
Photophobia
Headache
Eye and Face Pain
Epiphora
Foreign Body Sensation
Problems Opening Eyes
Chronic Red Eye

Loss of Visual Acuity and Refractive Issues
Decreased Distance Visual Acuity
Refractive Shift – Myopic
Refractive Shift – Hyperopic
Refractive Shift – Astigmatic
Asymmetric and Irregular Astigmatism
Dull or Abnormal Retinoscope Reflex
Poor Near Visual Acuity
Problems with Glasses

Loss of Visual Field (VF)
Visual Field Defects and Localizing Lesions
VF Defects Respecting the Horizontal Midline
VF Defects Respecting the Vertical Midline
Bitemporal Hemianopsia
Homonymous Hemianopsia
Central, Centrococcal and Cecal VF Defects
Severe Constriction of VF, Tunnel VF

Eyelids and Orbit
Blepharospasm
Loss of Sensation, Numbness of Face around Eye
Ptosis
Eyelashes – Loss or Excess
Eyelid Malpositions – Entropion and Trichiasis
Eyelid Malpositions – Ectropion
Eyelid Retraction
Lagophthalmos and Lid Lag
Seventh Nerve Palsy
Eyelid Mass / Lesion
Signs Suggesting Orbital Disease
Eyelid Edema
Conjunctival Hemorrhage and Peri-Orbital Ecchymosis
Proptosis
Orbital Tumors
Enophthalmos
Orbital Inflammation
Visible and Palpable Orbital Masses
Orbital and Facial Distortions
Distortions of the Globe
Motility and Alignment Problems
Types and Causes of Motility Problems
Abduction Deficit
Adduction Deficit
Acquired Esotropia
Acquired Exotropia
Apparent Horizontal Strabismus
Hypertropia / Hypotropia
Elevation and Upgaze Deficits
Depression and Downgaze Deficits
Convergence and Divergence
Ophthalmoplegia – One Eye
Ophthalmoplegia – Both Eyes
Head Turns and Tilts / Torticollis
Causes of Nystagmus
Acquired Nystagmus
Downbeat Nystagmus

Pupils
Leukocoria
Corectopia
Poor Pupil Mobility
Anisocoria
Anisocoria – Which Pupil is Abnormal?
Dilated Pupil(s)
Transient Pupil Dilation
Constricted Pupil(s)
Light Near Dissociation
Relative Afferent Pupillary Defect
Bilateral Miotic or Mydriatic Pupils

Anterior Segment
Conjunctival Bumps - Papillae and Follicles
Chemosis
Injected, Congested or Prominent Conjunctival Vessels
Symblepharon
Conjunctival or Epibulbar Mass
Spot on the White of Eye, Scleral Lesion
Scleral Thinning, Episcleritis, and Scleritis
Corneal Fluorescein Staining or Pooling
Corneal Epithelial Defects – Chronic or Non-Healing
Corneal Haze or Opacification
Acute Corneal Edema
Chronic Corneal Edema
Punctate Corneal Staining
Corneal Infiltrates
Corneal Ulcer
Corneal Vessels and Pannus
Decreased Corneal Sensation
Keratic Precipitates (KP)
Anterior Uveitis
Cells in the Anterior Chamber
Hypopyon
Hyphema
Neovascularization of the Iris
Lesions of the Iris
Defects of the Iris
Lens Opacification
Lens Abnormalities
**Fundus - Vitreous, Retina and Optic Nerve**
- Vitreous Cells, Pigment or Debris
- Yellow or Whitish Spots on Retina
- Apparent Mass in Retina / Choroid
- Posterior Uveitis
- Cotton Wool Spots
- Hard Exudates
- Retinal Infiltrates or Edema
- Macular Edema or Thickening
- Posterior Hemorrhages – Types
- Pre-Retinal or Vitreous Hemorrhages
- Neovascularization of the Disc, Retinal or Sub-Retinal
- Intra-Retinal Hemorrhages
- Sub-Retinal Hemorrhages
- Vascular Retinopathies
- Tortuous or Enlarged Retinal Vessels
- Dark or Pigmented Retinal Lesions
- Pigmentary Retinopathies
- Retinal Detachment
- Retinal Folds / Striae
- Optic Atrophy / Disc Pallor
- Optic Disc Cupping
- Optic Disc Edema
- Disc Hemorrhages
- Differentiating Between Acquired and Congenital Disc Elevation
- Abnormal Disc Vessels and Growths
- Causes of Optic Nerve Inflammation
- Neuroretinitis
- Papilledema and Increased Intracranial Pressure
- Bilateral vs. Unilateral Disc Edema

**IOP and Glaucoma**
- Acute Elevated Intraocular Pressure
- Narrow Angles or Angles Closed
- Angle Closure Glaucoma and Pupillary Block
- Open Angle Glaucoma Mechanisms
- Open Angle Glaucoma by Disease Process
- Flat or Shallow AC with High and Low IOP
- Hypotony – Low IOP

**Miscellaneous Case Situations and Lists**
- Longstanding Visual Loss
- Sudden / Acute Loss of Vision
- RAPD and Acute Visual Loss
- Acute Optic Neuropathy
- Chronic Progressive Loss of Vision
- Unexplained Visual Loss
- Bilateral Unexplained Acute Visual Loss
- Problems with Reading
- Suspected Visual Loss in a Infant / Child
- High Pressure Suspect (Large Cloudy Tearing Eye) in Infant Child with Esotropia
- Nystagmus in a Child
- Pregnancy and its effect on Eye Conditions

**LIST OF ABBREVIATIONS**
- IOP and Glaucoma
- Acute Elevated Intraocular Pressure
- Narrow Angles or Angles Closed
- Angle Closure Glaucoma and Pupillary Block
- Open Angle Glaucoma Mechanisms
- Open Angle Glaucoma by Disease Process
- Flat or Shallow AC with High and Low IOP
- Hypotony – Low IOP
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

**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

**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.
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- Synchysis scintillans
Monocular

Vitreo-Retinal Traction
Normal – rapid eye movements (dark), oculo-digital stimulation
Posterior Vitreous Detachment (PVD)
R/O Retinal Tear and Detachment

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 *
**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 PHOTOPSISAS**
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

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
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

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)

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).
   - **Vertebral-basilar Insufficiency**: Embolic (Cardiac, Plaques), Vertebral-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

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 6th 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?
Before considering true Nyctalopia

Consider other problems people encounter at night:

Uncorrected refractive errors may be 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)

4. Other: Advanced glaucoma, S/P PRP, other retinopathies (Pigmentary, vascular, etc.)
   Optic Neuropathies could present with Nyctalopia
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)
**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

**Binocular TVL**

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

**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 / Hyperviscocity – 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?

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

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
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)
- 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. 3rd CNP)
- High ICP - Papilledema
- GCA (Temporal Arteritis)
- Optic Neuritis
- Carotid Ischemia / Ocular Ischemic Syndrome
- Supra Orbital Neuralgia – palpate Supra-Orbital notch

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

- Asthenopia – with near vision
- Unilateral, Horner Syndrome, Conj injection, epiphora
- Things in Particular an Ophthalmologist should think of and look for*
- 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
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- Other Parasellar Tumor
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- Aneurysm Compression / SAH - (e.g. 3rd CNP)
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- 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)
- 3rd 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

* Includes: Cluster HA, Paroxysmal Hemicrania, etc. Unilateral, Horner Syndrome, Conj injection, epiphora
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 3rd or 6th
- Infarction – Thalamus (sensory center) , Medulla (Wallenberg)
- Trigeminal Neuralgia
- Post- Herpetic Neuralgia
- Raeder's Syndrome - Horner Syndrome, rhinorhea, upper facial and scalp pain
- Carotid dissection – pain, Horner syndrome
- Ramsay Hunt Syndrome: Zoster of external auditory canal, facial n. palsy

**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

Both associated with carotid obstruction and can have dull aching pain

Click to Return To Links
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

e.g. Uncorrected Refractive Error

Ocular Allergies

Chronic Blepharoconjunctivitis, 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

7th Nerve Palsy (poor pump function and lid laxity)

Jaw winking

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

Lacrimal Gland inflammation, mass

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
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 (3rd 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*
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
- Parainaud’s Ocular glandular syndrome – cat scratch, tularemia, mycobacterial
- Blepharoconjunctivitis, Acne Rosacea
- Superior Limbic Keratoconjunctivitis (SLK)

Conjunctival Mass, Tumor – Pingeuculae, 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

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

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

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 7th and 5th 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
• **Refractive Error:**
  Myopia, Hyperopia, Astigmatism (Regular and Irregular)

• **Media Opacity:**
  Tear Film, Corneal Opacification (Edema, Infiltrates, Deposits, Dystrophies), Cataract, Vitreous Hemorrhage or Opacities

• **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)
Acquired *Myopia*

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

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 dissention 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
• **Shift in Astigmatism**

  change in **cylinder power or axis**

**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
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
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

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

\[ \frac{1}{d(m)} = \text{accommodative power needed} \]

*(if hyperopic need to add distance plus (+) power as well)*
Problems with Glasses

Consider

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  \( P=hD \))

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 \( \rightarrow \) Diplopia - may need SLAB OFF
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*)

**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

Sometimes retinal pathology:
- BRAO
- BRVO
  - hemi-retinal vein or artery occlusion

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 Eyen 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**
Chiasmal, Sellar and Parasellar lesions
(See separate list)

**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
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

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)

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

Click to Return To Links
Central and Centroccecal 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

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

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

Click to Return To Links
Causes of Loss of or Impaired Sensation on Face and around Eye

- **Damage to Branches of 5th 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
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
- 3rd 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

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

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

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

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)
- Ptosis 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-Rhomberg, VKH, Lupus, Sarcoidosis,…

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)

* Comprehensive Listing : Survey 2006; 51:550
Eyelid Malpositions
Entropion and Trichiasis

**Lower Lid Entropion and Trichiasis**
- Involutional (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

**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*
Eyelid Malpositions - Ectropion

**Lower Lid Ectropion**
- Senile – with horizontal laxity, check for Medial or Lateral Canthal Tendon laxity
- Cicatricial (below)
- Combination of both above
- Paralytic – 7th 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

**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*
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 3rd 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

Click to Return To Links
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

See Exposure Keratitis

Don’t Confuse with
Lid Lag on Downgaze

Congenital Ptosis
Graves Ophthalmopathy
Aberrant Regeneration after 3rd CNP
Neurologic and Muscular Disease
  - Supranuclear Palsy
  - Myotonic Dystrophy
  - MG?
Post-op Upper Eyelid Procedures
Possible Sign of Other Orbital Disease
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, 7th CNP)
    - Parkinson’s
  - *Ramsay-Hunt Syndrome* (Herpes Zoster Oticus)
  - Mastoiditis / Otitis – 6th and 7th CNP possible
  - External Auditory Canal and Middle Ear – surgery, tumor
  - Other - Neuro-Sarcoidosis, Leprosy, Pregnancy (3rd Trimester), MS
    - Vasculitis, DM, Uremia

and *Bell’s Palsy* (Idiopathic 7th CNP)

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

A 7th 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

Nodular - Commonly at Lid Margin
- Intradermal Nevus
- BCCA
- Hair Follicle Tumor

Vascular
- Hemangioma
- Cherry Angioma – Bright red
- Varix
- Other: Kaposi’s Sarcoma, Pyogenic Granuloma

Crater / Ulcerated
- Carcinomas (BCCA, SCCA, etc)
- Keratoacanthoma
- Moluscum Contagiosum

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

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

**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

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

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

Click to Return To Links
Proptosis
Forward Displacement of the Eye

1. **Graves Orbitopathy** (#1 Cause)
   - **Orbital Cellulitis**
   - **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
Orbital Tumors to Consider

**Age Category**

1. **Children**
   - Rhabdomyosarcoma
   - Neuroblastoma
   - Lymphangioma
   - Capillary Hemangioma
   - Glioma of Optic Nerve
   - Dermoid Cyst
   - Neurofibromas (NF1 and NF2)

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

[Click to Return To Links]
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
  - Microophthalmos, 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

Click to Return To Links
Hypotony
Causes and Associations

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

**Signs of Orbital Inflammation**
Proptosis, Chemosis, Eyelid Edema, etc.

**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
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

Hypotelorism
– decreased orbital separation and decreased interpupillary distance
seen with: FAS – Fetal Alcohol Syndrome
Congenital Anomalies:
e.g. holoprosencenphaly
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

Other associated congenital defects:
Goldenhar (Oculoauriculo-vertebral) 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. 6th 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*
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 6th 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*
**Adduction deficit**

- 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

**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 3rd CNP)
- MR recession
- Orbital Disease – Graves, Myositis, Tumor
- Post-op: Scleral Buckle
- Conjunctival Scarring
- Previous EOM surgery- e.g. tight LR
- Ocular Neuromyotonia of 6th CN
- Myotonic Dystrophy
- Duane's Syndrome

*Click to Return To Links*
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 6th 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 6th CNP)
    - Pseudo-Esotropia — Epicanthal folds, Negative angle kappa, Hypotelorism, Telecanthus
    - Latent Hyperopia with Accommodative ET
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 6th CN- after prolonged lateral gaze

Previously Undiagnosed: Duane’s Syndrome, Pseudo-Exotropia – positive angle kappa, Hypertelorism
1. Pseudoesotropia
   - Epicanthal folds, Hypotelorism, Telecanthus
   - Apparent Esotropia - Negative angle kappa*

2. Pseudoeextropia
   - 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

*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

*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 4th CNP, but ocular torsion is different, not the typical excyclotorsion seen with 4th 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)*
<table>
<thead>
<tr>
<th>Elevation Deficit</th>
<th>Upgaze Deficit</th>
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<tbody>
<tr>
<td>Graves Ophthalmopathy (e.g. IR fibrosis)</td>
<td>Old Age</td>
</tr>
<tr>
<td>Orbital Floor Fracture with entrapment</td>
<td>Parinaud’s Syndrome</td>
</tr>
<tr>
<td>Third Nerve Palsy (Superior Division)</td>
<td>Thalamic Infarction</td>
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<tr>
<td>Double Elevator Palsy (in abduction and adduction)*</td>
<td>Progressive Supranuclear Palsy (PSP)</td>
</tr>
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<td>Brown’s Syndrome (in adduction)</td>
<td>Hydrocephalus</td>
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<td>CFEOM Type 1</td>
<td>Hemispheric Infarctions</td>
</tr>
<tr>
<td>Myositis (e.g. IR)</td>
<td>Metabolic – Niemann-Pick Disease</td>
</tr>
<tr>
<td>Myasthenia Gravis</td>
<td>Myasthenia Gravis</td>
</tr>
<tr>
<td>Midbrain – Diencephalic Lesion</td>
<td><strong>Bilateral</strong> - CFEOM or any of the other conditions on the right is possible</td>
</tr>
<tr>
<td>Isolated Palsy of SR or IO</td>
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<tr>
<td>Previous SR Recession</td>
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<tr>
<td>Other Mechanical: Orbital Mass, Glaucoma Implant, Fat Adherence Syndrome</td>
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</tbody>
</table>
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 3rd and 4th Ventricles, Trauma
Convergence and Divergence

Convergence Insufficiency
Conmitant Exotropia greater at near than that distance. Decreased Near Point of Convergence (NPC)

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

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:
- 6th 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

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. 3rd and 6th) 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

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: 4th (Superior Oblique) or 6th (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

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.

Aid in Diagnosis:
Patch one eye and if torticollis resolves then suggests strabismus is cause of torticollis
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
Hypotony

Causes and Associations

Low IOP

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.

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

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

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

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

Click to
Return To Links
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

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
• **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
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
**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)

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 3rd CNP

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

Click to Return To Links
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
**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**

**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)
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.

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

Can be one or both pupils
**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
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

2. **Bilateral Mydriasis**

**Systemic Medications:** anticholinergic (atropine)

sympathomimetics (amphetamines, cocaine, etc.)

**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

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

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
Conjunctival Bumps

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, Neisirria, Moraxella

Children – benign lymphoid folliculosis
Conjunctival Lymphoma

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
Chemosis

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

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

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
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.

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, Cannibus
Hyperviscocity – 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
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
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)
- Pingeuelum, 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, Phylectenule, 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

[Click to Return To Links]
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), Cautery 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)
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 effect:
- 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
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
  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)

**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 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
Causes of *Acute* Corneal Edema

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

**Exposure** – proptosis, 7th 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
Causes of *Chronic* Corneal Edema

**Chronic Compromise of Conjunctival Function**

→ Chronic Epithelial Edema, Haze

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

**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
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 – 7th 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)**
<table>
<thead>
<tr>
<th><strong>Infectious Corneal Ulcers</strong></th>
<th><strong>Other Infiltrates</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Larger Infiltrate</td>
<td>Smaller</td>
</tr>
<tr>
<td>Often single lesion</td>
<td>Multiple</td>
</tr>
<tr>
<td>Very painful</td>
<td>Less Painful</td>
</tr>
<tr>
<td>More Central</td>
<td>More Peripheral</td>
</tr>
<tr>
<td>Cells in A/C</td>
<td>No A/C cells</td>
</tr>
<tr>
<td>Conjunctival Injection</td>
<td>More segmental / focal injection</td>
</tr>
</tbody>
</table>

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

**PEDAL** – to help distinguish non-infectious from infectious

Infectious more associated with **Pain**, **Epithelial defects**, **Discharge**, **Anterior chamber reaction** and more **central Location**
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.
• Contact Lens Over Wear - hypoxia
• Staphylococcal Disease – Blepharitis
• Phylectenular 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
Decreased Corneal Sensation

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
Keratic Precipitates (KP)

Non-Granulomatous Uveitis Marked by:
- More acute onset
- More injection
- Fine KP
- Pain, Photophobia

Non-Granulomatous - think of:
- HLA- B27 associated Uveitis
- TINU
- Post-Infectious or Drug (antibiotic) induced uveitis
- Idiopathic

Granulomatous Uveitis suggested by:
- Chronic Uveitis
- Little Vessel Injection
- Mutton Fat KP
- Less symptoms

Granulomatous? Think of:
- Sarcoidosis
- Toxoplasmosis
- Syphilis
- Tuberculosis
- Sympathetic Ophthalmia
- VKH
- Herpetic Uveitis
- Uveitis associated with MS
- Intra Ocular FB
- Lens Induced Uveitis

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

Corneal Graft Rejection

edmedicine Ophthalmology
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**

**Anterior Uveitis**

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

**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 Yields  
  - Shingles? Rashes, Atopic Symptoms

**Main Work-up Test Considerations:**
- HLA- B27  
  (and any others)
- FTABS, RPR/VDRL* on the left that  
  (on the left that)
- CXR  
  (seem indicated)
- ACE/ Lysozyme
- PPD - if Chronic
Cells in the A/C

- **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**
✓ **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)
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

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.

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)
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)
Iris Transillumination Defects:

- Albinism
- Essential Iris Atrophy
- HZO - uveitis
- Pseudoexfoliation Glaucoma
- Trauma
- Intraocular Surgery / Phaco
- Pigmentary Dispersion/ Glaucoma

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
### Adult

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

* 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
**Primary** - (POAG)* - usually symmetric

**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
   - 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

Synechiae 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)
Hypotony (Low IOP)

Causes and Associations

- 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
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

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

Click to Return To Links
**Apparent Mass in Retina / Choroid**

**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

*Suggests sub-retinal or choroidal process looking at overlying vessels*
Hypotony
Causes and Associations

- **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

**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), Whipples, before giving corticosteroids which can worsen condition and blindness

- **Sometimes Associated with Encephalitis:** e.g. VKH, Herpes, West Nile Virus, Toxoplasmosis (HIV)...
- **Remember - Masquerade Syndrome – Lymphoma, Retinoblastoma, etc.**
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
**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
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
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
Posterior Hemorrhages

**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
Hypotony
Causes and Associations

- 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

Neovascularization
Disc, Retina, or Sub-Retinal
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), Purtcher’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
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
- Purtcher’s – bone/skull trauma, systemic diseases (e.g. pancreatitis)
- Other: FEVR (ROP-like in older pts – with temporal avascular retina)
Tortuous or Enlarged Retinal Vessels

**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

**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

---

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

*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

Click to Return To Links
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

Click to Return To Links
Retinal Detachment

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

**Tractional**
- PDR and Other proliferative retinopathies
- Hemoglobinopathies: Sickle Cell (SSD, SCD), Thalassemia, Eales Disease
- Ischemic CRVO
- PVR (proliferative vitreous retinopathy)
- 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)
• 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
Optic Atrophy / Disc Pallor

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-TNFα, Interferon, Tacrolimus, Methanol, Ethylene Glycol, Heavy Metals (Pb, Hg, As, Co, Th), CO, CCI₄, Nutritional Deficiencies (B₁ (Thiamine), B₁₂, 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)
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
Disc Edema

See also Acute Optic Neuropathy

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)
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
<table>
<thead>
<tr>
<th>Feature</th>
<th>Congenital</th>
<th>Acquired</th>
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<tr>
<td>Nerve Fiber Layer</td>
<td>Clear</td>
<td>Opacified</td>
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<tr>
<td>Large Disc Vessels</td>
<td>Anomalous</td>
<td>Normal</td>
</tr>
<tr>
<td>Small Disc Vessels</td>
<td>Normal</td>
<td>Telangietatic</td>
</tr>
<tr>
<td>NFL Hemorrhage</td>
<td>Rare</td>
<td>Frequent</td>
</tr>
<tr>
<td>Physiologic Cup</td>
<td>Small or absent</td>
<td>Normal</td>
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<tr>
<td></td>
<td></td>
<td>(But may be obscured by edema)</td>
</tr>
<tr>
<td>Drusen</td>
<td>Sometimes present</td>
<td>Absent</td>
</tr>
</tbody>
</table>
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

**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
4. Melanocytomas – usually darkly pigmented
5. Glial Remnants (Bergmeister’s Papilla)
6. Adjacent NFL Myelination

* Click to Return To Links

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Opto-ciliary Shunt Vessels

NVD

ERM dragging disc vessels

Astrocytoma

Transactions of American Ophthalmological Society 2004; 102

NFL Myelination
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, Bechets, Sjogrens syndrome, “Simple” Autoimmune Optic Neuropathy
- Paraneoplastic Optic Neuritis

Contiguous Inflammation:
- Encephalitis, Meningitis (high ICP and inflammation)
- Pachymeninigitis – (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)

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

*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


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
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
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.
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:
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

**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

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

**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
<table>
<thead>
<tr>
<th>Condition</th>
<th>+ RAPD</th>
<th>- RAPD</th>
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<tbody>
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<td>e.g. Tumor or Graves Disease</td>
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<tr>
<td>or Pre-Retinal Hemorrhage</td>
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<tr>
<td>Neuro-retinitis</td>
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</tbody>
</table>
“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)

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
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
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

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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
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
     - Vitreal-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 sella, 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. Sarcoïdosis, Electrocution, 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, vindesin, 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
Vascular
- Hypotension – e.g. PION after trauma, surgery, code
- Severe Systemic Hypertension
- Vertebrobasilar Insufficiency
- Temporal Arteritis – e.g. PION

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.
Problems with Reading

Do 8-point Eye Exam (to be sure no obvious ocular problems) and Check NVA

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, ...)
   - 4th 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)
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 affixational patients with vision less than 20/400

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 could 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
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
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

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

Don’t call all cases like this congenital glaucoma!!
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.

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

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### Condition | Common Age of Presentation
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- **Congenital / Infantile** | < 1 year
- **Accommodative** | 18 mo to 3 years
- **Late Onset*** | >3-4 years

*need to be concerned for 6th 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. Deprivation 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)

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
- Meningioma - e.g. Sphenoid wing
- Pituitary Apoplexy
- High ICP: Venous Sinus Thrombosis
  - Pseudotumor Cerebri Exacerbated
- Migraine
- Diabetic Retinopathy (DME)
- CSR
- Cranial Neuropathies: 7th 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

**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
1. **Smoking (Tobacco)** - exacerbates or increases risk of developing ..
   ARMD, Graves Ophthalmopathy, Optic Nerve Atrophy, AION, DR, Cataracts, Uveitis

2. **Diabetes Mellitus**
   Early Presbyopia, Non reactive pupil, CNP (especially 3rd and 6th), 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)
   Potentiation 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
Possible Causes

**Microvascular** (older age, High BP, DM)
**Diabetic** – mainly 3rd and 6th
**Trauma** – closed head, basilar skull fracture
**Compressive** – Aneurysm/Herniation – especially 3rd
**Tumor** – Compression or Infiltration
**Ischemic CVA** – Brainstem

**High Intracranial Pressure** – especially 6th
**Low ICP (Intracranial Hypotension)** – 6th most common
**CNS Inflammatory** – e.g. Meningitis,…
**MS / Demyelination** – especially 6th
**Congenital** – especially 4th Nerve
**Vasculitis** – e.g. GCA
**Migraines** *
**Systemic - immune mediated polyneuropathy** (e.g. GBS, MFS)
**Shunt Failure**

**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 3rd or 6th 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.**

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**Cranial Nerves Palsies**
**Related to Ocular Motility**

**Cranial Nerve**
3rd Oculomotor
4th Trochlear
6th Abducens
### Cranial Nerve Palsies

#### More Specific Causes

**3rd 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 3rd CNP Paresis

**6th 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

**4th CNP**
1. Traumatic
2. Congenital
3. Microvascular
4. Other Causes - uncommon
Elevated ESR

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

1. R/O GCA (usually older patients >60 yo)

2. Anemia - anemic patient can have an 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 patients 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 a 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)
Illusions and Hallucinations:

Psychogenic Medications and Drugs -

- Dopaminergics: levodopa, bromocryptine,
- Anticholinergics: atropine, scopolamine, cyclopentolate
- Tricyclic Antidepressants, Beta-Blockers
- Adrenergic – albuterol, Phenylephrine, Theophyline, Amphetamine, Cocaine
- Alcohol
- Misc: Benzodiazepines, corticosteroids, NSAIDS, Ca Channel blockers, Narcotics, anticonvulsants,...
  - Trazodone, Nefazodone, Topiramate, Risperidone, Clomiphene
  - Illicit Drugs: LSD, Cannibus, Methamphetamine, Psylocibin (mushrooms)

Transient Refractive Shifts

- Myopic – Topiramate
- Hyperopic - Phenothiazides, Antihistamines, Cholorquine, Anticholinergics, Cannabis

Cataract

- Corticosteroids (PSC), Statins, Phenothiazines (Chlorpromazine)

Angle Closure Glaucoma – increased risk

- Ant-histamines, Anti-psychotics, Tri-cyclic Antidepressants, Ephedrine

Retinal Pigmentary Changes

- Phenothiazines - Thioridazine (Mellaril), Chlorpromazine (Thorazine), Chloroquine, Hydroxychloroquine
- Tamoxifen, Indomethacine, Deferoxamine
Ocular Effects of Medications

- **Papilledema / Increased Intracranial Pressure**
  - Vitamin A, Retinoids e.g. Isotretinoin (Accutane), Tetracylines: 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

- **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

**Boney Growth / Change / Expansion**

- Fibrous Dysplasia
- Sarcoma of the bone
- Metastatic Disease to the Bone
- Paget’s Disease

**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
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
Dizziness

**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

**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.
<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Full Form</th>
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<tr>
<td>AC - AC1</td>
<td>Anterior Chamber - Arnold Chiari Malformation Type 1</td>
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<tr>
<td>ACE</td>
<td>Angiotensin Converting Enzyme</td>
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<td>ACG</td>
<td>Angle Closure Glaucoma</td>
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<td>AD</td>
<td>Autosomal Dominant</td>
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<tr>
<td>ADOA</td>
<td>Autosomal Dominant Optic Atrophy</td>
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<td>AK</td>
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<td>AMPPE</td>
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<td>ANA</td>
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<td>ARMD</td>
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<tr>
<td>BCP</td>
<td>Birth Control Pills, Contraceptives</td>
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<tr>
<td>BDUMP</td>
<td>Bilateral Diffuse Uveal Melanocytic Proliferation</td>
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<tr>
<td>BP</td>
<td>Blood Pressure</td>
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<td>BPES</td>
<td>Blepharophimosis- Ptosis- Epicanthus Inversus Syndrome</td>
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<td>BPV</td>
<td>Benign Positional Vertigo</td>
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<td>BRAO</td>
<td>Branch Retinal artery Occlusion</td>
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<td>BRVO</td>
<td>Branch RVO</td>
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<tr>
<td>CA</td>
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<td>Cancer Associated Retinopathy Syndrome</td>
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<td>CC</td>
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<td>CE</td>
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<td>CFEOM</td>
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<td>CMV</td>
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<td>CN</td>
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<td>CN palsy</td>
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<td>CNS</td>
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<tr>
<td>CPA</td>
<td>Cerebellar Pontine Angle</td>
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<td>CPEO</td>
<td>Chronic Progressive External Ophthalmoplegia</td>
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<tr>
<td>CR</td>
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<tr>
<td>CRAO</td>
<td>Central retinal artery occlusion, CRVO- central ret. vein occlusion</td>
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<td>CSN</td>
<td>Central Serous Retinopathy</td>
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<td>CSNB</td>
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<td>CT</td>
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<td>CTA</td>
<td>CT Angiogram</td>
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<tr>
<td>CTL</td>
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<td>CVA</td>
<td>Cerebral (CNS) Vascular Accident (Stroke)</td>
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<td>Chest X-Ray</td>
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<td>DR</td>
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<td>DME</td>
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<tr>
<td>DM</td>
<td>Diabetes Mellitus</td>
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<td>DUSN</td>
<td>Diffuse Unilateral Subacute Neuroretinitis from Nematode</td>
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<td>EOM</td>
<td>Extra Ocular Muscle</td>
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<tr>
<td>ERG</td>
<td>Electroretinogram</td>
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<tr>
<td>ERM</td>
<td>Epi - Retinal Membrane</td>
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<tr>
<td>ESR</td>
<td>Erythrocyte Sedimentation Rate</td>
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<td>ET</td>
<td>Esotropia</td>
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<tr>
<td>FA</td>
<td>Fluorescein Angiogram</td>
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<tr>
<td>FB</td>
<td>Foreign Body</td>
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<tr>
<td>FBS</td>
<td>Foreign Body Sensation</td>
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<tr>
<td>FEVR</td>
<td>Familial exudative vitreoretinopathy</td>
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<tr>
<td>FHx</td>
<td>Family Medical History</td>
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<tr>
<td>GBS</td>
<td>Guillain-Barre Syndrome</td>
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<tr>
<td>GCA</td>
<td>Giant Cell Arteritis (aka Temporal Arteritis)</td>
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</tbody>
</table>
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)
<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
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<tr>
<td>PDR</td>
<td>Proliferative Diabetic Retinopathy</td>
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<tr>
<td>PEK</td>
<td>Punctate Epithelial Keratopathy</td>
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<tr>
<td>PET-CT</td>
<td>Positron emission tomography–computed tomography</td>
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<tr>
<td>PPHV</td>
<td>Persistent Hyperplastic Primary Vitreous</td>
</tr>
<tr>
<td>PPM</td>
<td>Persistent Placoid Maculopathy</td>
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<tr>
<td>PPV</td>
<td>Pars Plana Vitrectomy (Surgery)</td>
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<tr>
<td>PI</td>
<td>Peripheral Iridectomy or Iridotomy</td>
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<tr>
<td>PION</td>
<td>Posterior Ischemic Optic Neuropathy</td>
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<tr>
<td>PK</td>
<td>Penetrating Keratoplasty (corneal transplant)</td>
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<tr>
<td>POAG</td>
<td>Primary Open Angle Glaucoma</td>
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<td>PON</td>
<td>Paraneoplastic Optic Neuropathy</td>
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<td>PORN</td>
<td>Progressive Outer Retinal Necrosis</td>
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<td>PRK</td>
<td>Photo-Refractive Keratectomy</td>
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<td>PRP</td>
<td>Pan-Retinal Photocoagulation</td>
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<td>PSC</td>
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<td>PUK</td>
<td>Peripheral Ulcerative Keratitis</td>
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<td>PVD</td>
<td>Posterior Vitreous Detachment</td>
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<tr>
<td>RA</td>
<td>Rheumatoid Arthritis</td>
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<td>RAO</td>
<td>Retinal Artery Occlusion</td>
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<td>RAPD</td>
<td>Relative Afferent Pupillary Defect</td>
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<td>RB</td>
<td>Retinoblastoma</td>
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<td>RBC</td>
<td>Red Blood Cells</td>
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<td>RD</td>
<td>Retinal Detachment</td>
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<td>RES</td>
<td>Recurrent Erosion Syndrome</td>
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<td>RLL</td>
<td>Right Lower Lid</td>
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<td>RGP</td>
<td>Rigid Gas Permeable (CTL)</td>
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<td>RK</td>
<td>Radial Keratotomy</td>
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<td>RMSF</td>
<td>Rocky Mountain Spotted Fever (Rickettsiae)</td>
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<td>R/O</td>
<td>Rule Out</td>
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<td>ROP</td>
<td>Retinopathy of Prematurity</td>
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<td>RP</td>
<td>Retinitis Pigmentosa</td>
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<td>RPE</td>
<td>Retinal Pigment Epithelium</td>
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<td>RUL</td>
<td>Right Upper Eyelid</td>
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<tr>
<td>RVO</td>
<td>Retinal Vein Occlusion</td>
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<tr>
<td>SAH</td>
<td>Subarachnoid Hemorrhage</td>
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<td>SBS</td>
<td>Shaken Baby Syndrome</td>
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<td>SCH</td>
<td>Sub-Conjunctival Hemorrhage</td>
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<td>SCCA</td>
<td>Squamous Cell Carcinoma</td>
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<td>SJS</td>
<td>Stevens Johnson Syndrome</td>
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<td>SLK</td>
<td>Superior Limbic Keratoconjunctivitis</td>
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<td>SLE</td>
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<td>SNP</td>
<td>Supranuclear Palsy</td>
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<td>Superior Oblique</td>
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<td>Status Post</td>
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<td>SR</td>
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<td>SRNVM</td>
<td>Sub-Retinal Neovascular Membrane</td>
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<td>TASS</td>
<td>Toxic Anterior Segment Syndrome</td>
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<td>TB</td>
<td>Tuberculosis</td>
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<td>TBI</td>
<td>Traumatic Brain Injury</td>
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<td>TM</td>
<td>Trabecular Meshwork</td>
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<tr>
<td>TORCH</td>
<td>(Toxoplasmosis, Other, Rubella, Cytomegalovirus, Herpes Simplex)</td>
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<td>UA</td>
<td>Urine Analysis</td>
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<td>UGH</td>
<td>Uveitis Glaucoma Hyphema Syndrome</td>
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<td>UL</td>
<td>Upper Lid (Eyelid)</td>
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<td>URI</td>
<td>Upper Respiratory Infection</td>
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<td>VA</td>
<td>Visual Acuity</td>
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<td>VF</td>
<td>Visual Field</td>
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<td>VKH</td>
<td>Vogt-Koyanagi-Harada Syndrome</td>
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<td>VMT</td>
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<td>VPT</td>
<td>Vitreo-Papillary Traction</td>
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<td>Varicella-Zoster Virus</td>
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<td>Work up</td>
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<td>Exotropia</td>
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Major References

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<td>American Journal of Ophthalmology</td>
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<tr>
<td>Journal of Neuro-ophthalmology</td>
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<tr>
<td>Investigative Ophthalmology and Visual Science</td>
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The heavens declare the glory of God; the skies proclaim the work of his hands

Psalm 19:1