Differential Diagnoses
and other Useful Lists and Tables
For Ophthalmologists

Symptoms
Signs
Case Presentations

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Acknowledgments and Disclaimer

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 presentation, 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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Disclaimer: The diagnostic algorithm for the diagnosis and management of Ocular or Neurological Conditions contained in this presentation is not intended to replace the independent medical or professional judgment of the physician or other health care providers in the context of individual clinical circumstances to determine a patient’s care.

* 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 you to 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 and Itching
- 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, Centrocecal and Cecal VF Defects
- Severe Constriction of VF, Tunnel VF

Eyelids and Orbit
- Blepharospasm
- Loss of Sensation, Numbness of Face around Eye
- Ptosis
- Eyelashes and Eyelid Margin
- 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
Hypopion
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
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
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
Basic Differential Diagnosis
CINTAVO* (mnemonic)

C - Congenital / Familial / Genetic
I - Inflammatory: Infectious / Allergic / Autoimmune
N - Neoplastic
T - Traumatic / Toxic
A - Aging: Degenerative
V - Vascular: Ischemia / Malformation / Hemorrhage
O - Other (OMNI-P): Obstruction / Compression
  - Medication
  - Nutritional / Metabolic
  - Iatrogenic
  - Pressure related: Blood, ICP, IOP

*cintavo is a real word: Italian first-person singular, imperfect indicative of cintare - “to enclose or wrap up”
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.
Transient Visual Phenomena

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
Flashing Lights
Photopsias

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

Retinitis / Choroiditis
Outer Retinal Disorders: MEWDS, AIBSES, Multifocal Choroiditis, etc
Retinal Ischemia: Impending CRVO, DR, OIS

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

Ocular / Retinal Migraine

Optical “Dysphotopsias”
(Not true Photopsias, optical effects)
- IOL small diameter/ larger pupil; edge glare
  (may see a shadow or light in temporal field)
- IOL shutter early post op period
- RGP edge glare
- Posterior Capsule folds - can produce a Maddox Rod effect *

Photopsias Continued ➤
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, dopaminergics, 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, Medications (e.g. statins, anti-seizure- e.g. lamotrigine, diet pills, Celecoxib)

   **Decompensated Strabismus** — e.g. Intermittent XT, Monofixation Syndrome, Fixation Switch 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
Binocular Diplopia – Causes of Ocular Misalignment

1. **Neurologic**
   - Supranuclear – e.g. Inter Nuclear Ophthalmoplegia (INO)
   - Cranial Nerves – 3,4,6 (Palsy or rarely spasm)*
   - Structure Abnormality – e.g. Chiari Malformation, Posterior Fossa Mass
   - Systemic – e.g. polyneuropathy GBS, MFS, Medication induced

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

2. **Incipient Neurologic, Orbital or Myogenic Disease**
   (Condition early in development does not have obvious manifestations, and a careful search for more subtle signs will be needed)
   e.g. Early CNP, INO, MS, Graves Orbitopathy, Orbital Pseudotumor, MG, High ICP, GCA, Ophthalmoplegic Migraine, Myotonic Dystrophy

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)
Ocillopsia

• **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 maybe first present as night problems
  e.g. Early Myopia, Latent Hyperopia
  Pupils: Miotic Pupils (e.g. Pilocarpine, surgical) - limit light in
  Normal Pupillary Dilation in dark can bring out any optical aberrations
    e.g. uncorrected refractive error, cataract, corneal

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

1. Congenital Stationary Night Blindness*
2. High Myopia
3. Progressive Chori-o-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 can proceed AAION

**CNS / Neurologic**
- Cortical Ischemia (though usually binocular), Uhthoff’s Phenomenon, Epilepsy

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

**Think also of Transient Intra-Ocular Problems:**
- Hyphema, Elevated IOP, Corneal Edema, Angle Closure Glaucoma, Dry Eye, UGH Syndrome, Reduced Ocular Perfusion, Transient Myopia

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

Binocular TVL

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

**Migraine Aura**

**Cortical Ischemia**
- *e.g.* Vertebro-Basilar Insufficiency

**Papilledema**

**Medications:**
- Digitalis, Viagra

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

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

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

Think about Ocular and Intracranial Trigeminal Irritation

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

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

Any ocular or visual abnormality could be associated with this symptom

(In patients with normal eye exam consider: Internal Trigeminal Irritation, Achromatopsia, Optic Neuritis, Migraine)
Things in Particular an Ophthalmologist should think of and look for*

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

New Headache and Anisocoria:
  Horner’s (Carotid Dissection)
  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
**Eye and Peri-Orbital Pain**

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

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

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

- Asthenopia
- Recurrent Erosion Syndrome
- Occult / Sub-clinical NL infection: Dacrocystitis, 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's 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
- Dacrocystitis, 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

**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 and Itching

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

Itching (Pruritus)

Blepharitis (see list)

Ocular Allergies
  - Vernal, Atopic,
  - Allergic Conjunctivitis
    CTL related – Giant Papillary

Dry Eyes

Eyelid Mass – benign or malignant

Healing after Eye Surgery
Problem Opening Eyelid(s)

- **Blepharospasm** – eyelids squeeze (see list)
- **Apraxia of Eyelid Opening** *
  - neurological – problem initiating eyelid opening
- **Ptosis** (though usually not complete closure)
  - Neurologic (3\textsuperscript{rd} 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
- Paranaud’s Ocular glandular syndrome – cat scratch, tularemia, mycobacterial
- Blepharoconjunctivitis, Acne Rosacea
- Superior Limbic Keratoconjunctivitis (SLK)

Conjunctival Mass, Tumor – Pingeuclae, 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 (CN 5 and or 7 dysfunction), etc.

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

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

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

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
Decreased Distance Visual Acuity

- **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

Click to Return To Links
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
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)

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**Problems with Near**

1. Bifocal Segment – not enough or too much add power
   - position: top should be a lower lid level. Some are too low

2. Progressive Bifocals – too narrow or patient has to look too far down to get full add

3. Anisometropia – with large differences in vertical induced prism → Diplopia - may need SLAB OFF
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
Other Visual Field Defects

Central and Centrocecal Scotomas

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

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
Other Visual Field Defects

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 5\textsuperscript{th} Cranial Nerve**
  - e.g. Orbital Floor Fracture – damage to V2
  - Shingles (Zoster) - most commonly V1 distribution

- **Facial Carcinomas (even occult ones)**
  - can track along nerves (*perineural invasion*)
  - *e.g. BCCA, SCCA*

- **Tumor in Brainstem, Cavernous Sinus, Orbit or Skull base**
  - *e.g. CPA angle Acoustic Neuroma*

- **CNS (cerebral cortex or brainstem) lesion** – CVA, MS, etc.

- **Recall loss of Corneal Sensation can be related:**
  - *e.g. Keratitis – active or past Acanthamoeba, Herpetic, ...*
  - See list for loss of Corneal Sensation
Ptosis

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

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

**Neurological**
- 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)
- Phthisis or small globe or Anophthalmos
- Blepharospasm, Dermatochalasis or Brow Ptosis Mistaken for ptosis
- Hypertropia, Hypotropia
Eyelashes and Eyelid Margin

Madarosis (Loss of Lashes)
- R/O Carcinoma – e.g. BCCA, Sebaceous Cell CA
- Chronic Blepharitis – e.g. Herpetic, Staph, Fungal, Mites …
- Endocrine – e.g. Hyper and hypo parathyroid and thyroid, hypopituitism
- Dermatoses - Dermatitis (atopic, contact), ichthyosis, lichen planus,…
- 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)

Blepharitis
Erythema, injection, telangiectasia, madarosis and lash misdirection, scurf, crusting, collarets, blocked Meibomian glands, erosion of lid margin

Consider Causes:
Staphylococcal
Seborrheic (dermatitis)
Acne Rosacea
Demodex infestation (increases with age)
Contact Dermatoblepharitis

DDX of Signs:
Eyelid Malpositions
Chalazia and Hordeolum

R/O Eyelid margin tumor: especially BCCA and Squamous Cell CA

* Comprehensive Listing : Survey 2006; 51:550
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*
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
- Medications: TNF-α inhibitors

Upper Lid Ectropion

- **Cicatricial Processes (below)**
- Congenital – e.g. Ichthyosis
- Floppy Eyelid Syndrome – Horizontal Laxity – not 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
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 Lagophthalmos 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
• 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
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

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

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

Bad Sub-Conjunctival Hemorrhage can dissect into eyelids

Orbital Hemorrhage associated with:
  - Trauma
  - Retrobulbar Injection
  - Neuroblastoma , Rhabdomyosarcoma and other tumors
  - Amyloidosis, Multiple Myeloma
  - Leukemia

Sub-Periosteal Hemorrhage
  - Valsalva : Labor, Vomiting, Coughing
  - Bleeding Disorders, Liver Disease, Scurvy
  - Sinus Disease
  - Venous Congestion

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

**Pseudo-Proptosis**
- Large Globe- Glaucoma, High Myopia
- Lid Retraction w/o real forward displacement,
  Relative Enophthalmos
Age Category

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

These two lists are not mutually exclusive

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

These two lists are not mutually exclusive
**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 Scirrhoua CA (*e.g.* Breast)
- Surgery
- Silent Sinus Syndrome
- Starvation
- Aging
- Horner’s Syndrome
- Orbital Fat Atrophy
  - Parry-Romberg Syndrome (Idiopathic Hemifacial Atrophy, Sympathetic Input?)
  - Past inflammation, Past Trauma, Linear Scleroderma
  - Orbital tumor removal – tumor caused pressure atrophy of the fat
  - Prostaglandin Associated Periorbitopathy (*from* Glaucoma Prostaglandin Analogues)

- Intracranial Hypotension?
- Pseudo-Enophthalmos
  Microphthalmos, Phthisis, Contralateral Exophthalmos, Ptosis, Isolated Superior Sulcus Deformity
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, Sjogren’s, 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

Imaging of the Orbits

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

Visible and Palpable Masses in and around the Orbit

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 (Oculoauriculovertebral) syndrome, Trisomy 13-15

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

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

Large, Buphthalmic Eye

Congenital and Juvenile Glaucoma
Anterior Segment Dysgenesis (e.g. Rieger’s), Megalocornea, High Myopia
Intraocular Tumor – RB, Uveitic – Rubella, Toxocara, Herpetic
Systemic: Lowe’s Syndrome (oculocerebrorenal syndrome), 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. Misalignment of Visual Axes
   1. “Primary Strabismus” - Childhood, Decompensated in Adult
   2. Related to Neurologic, Myogenic, Orbital or Sensory Problems
Motility and Alignment 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*

Click to Return To Links
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**
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**

Intermittent XT that becomes more frequent – with age, sickness, ...
Can Decompensate to a Constant Exotropia
Types: Basic, Divergence Excess
Diplopia – usually not constant, just occasional – when outside of suppression scotoma

**INO**

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

**Sensory**

Loss of vision in one eye – trauma, RD, Optic Atrophy

**Myasthenia Gravis**

**Convergence Insufficiency (XT at near >XT at distance)**

**Orbital Disease: Tumor, Myositis, Pseudotumor**

**Previous EOM Surgery – Consecutive or Recurrent Exotropia**

Third Nerve Palsy
Hydrocephalus, Shunt Failure
Myopia
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*
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 exyclotorsion 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)*
Elevation Deficit

Graves Ophthalmopathy (e.g. IR fibrosis)
Orbital Floor Fracture with entrapment
Third Nerve Palsy (Superior Division)
Double Elevator Palsy (in abduction and adduction)*
Brown’s Syndrome (in adduction)
CFEOM Type 1
Myositis (e.g. IR)
Myasthenia Gravis
Midbrain – Diencephalic Lesion
Isolated Palsy of SR or IO
Previous SR Recession
Other Mechanical: Orbital Mass, Glaucoma Implant, Fat Adherence Syndrome

Upgaze Deficit

Old Age
Parinaud’s Syndrome
Thalamic Infarction
Progressive Supranuclear Palsy (PSP)
Hydrocephalus
Hemispheric Infarctions
Metabolic – Niemann-Pick Disease
Myasthenia Gravis

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

- SR restriction
  - Graves
  - Fibrosis
  - Myositis

- IR Ischemia

- 3rd CNP – Inferior Division

- Myasthenia Gravis

- Midbrain and Diencephalic Lesions

Downgaze Deficit

- Progressive Supranuclear Palsy

- Parkinson–Like Syndromes

- Midbrain Lesions

- Myasthenia Gravis

- Head Trauma

- Oculogyric Crisis
  (tonic deviation of the eye – usually upward)
  - Post-encephalitic Parkinsons
  - Drugs: neuroleptics, benzodiazepines, many cited
  - Cannabis, MS, Neuro-Syphilis
  - Lesions of 3rd and 4th Ventricles, Trauma
Convergence and Divergence

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

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

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 Fistula or 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
1. Orthopedic / Muscular – most common causes
   e.g. Congenital muscular torticollis: damaged sternocleidomastoid muscle
   Cervical spine – damage / deformity: Fracture, TB, Scoliosis
   Tonsillitis, retropharyngeal abscess, drugs – neuroleptics, dystonias

2. Neurologic – not common

3. Ocular – not uncommon
   - Incomitant Strabismus:
     CNP: 4<sup>th</sup> (Superior Oblique) or 6<sup>th</sup> (Abducens)
     Duane Syndrome, Browns Syndrome
     A and V Patterns
     DVD (Dissociated Vertical Deviation)
   - Nystagmus
     Congenital Nystagmus with Null Point
     Spasmus Nutans
   - Ptosis - often chin up head position
   - Astigmatism

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.
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**

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

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**So before jumping to order a MRI consider #1-9, and ...**

*Ask about Oscillopsia (not usually present in Congenital)*

*Ask about Vertigo (Vestibular Nystagmus)*
Downbeat Nystagmus

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

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
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
Anisocoria (Unequal pupils)

- 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
**Anisocoria**

**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
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
Constricted Pupil(s)

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)**

6. **Other** – *Diabetes Mellitus*, Riley Day Syndrome, Aberrant Regeneration of 3rd Cranial Nerve, alcoholism, encephalitis
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
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
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)
- Pingueculum, Pterygium (beware of atypical pterygia with CIN)
- Benign hereditary intraepithelial hyperplasia, Papillomatosis*

**Pigmented Lesions** - Racial Melanosis (usually bilateral, should not grow), Conjunctival Nevus (often cysts in it, no feeder vessels)
- PAM (Primary Acquired Melanosisis- 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, Kaposi’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
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>Versus</strong></th>
<th><strong>Other Infiltrates</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Larger Infiltrate</td>
<td></td>
<td>Smaller</td>
</tr>
<tr>
<td>Often single lesion</td>
<td></td>
<td>Multiple</td>
</tr>
<tr>
<td>Very painful</td>
<td></td>
<td>Less Painful</td>
</tr>
<tr>
<td>More Central</td>
<td></td>
<td>More Peripheral</td>
</tr>
<tr>
<td>Cells in A/C</td>
<td></td>
<td>No A/C cells</td>
</tr>
<tr>
<td>Conjunctival Injection</td>
<td>more generalized</td>
<td>More segmental / focal injection</td>
</tr>
</tbody>
</table>

**Infectious Corneal Ulcer**
- Pain
- Epithelial defects
- Discharge
- Anterior chamber reaction
- More central location

**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

**Images**
- Infectious Corneal Ulcer
- Adenovirus
- Staph Marginal Keratitis
- CTL over wear Keratitis
- Peripheral / Marginal Keratitis
Corneal Ulceration and Thinning

Central

Infectious – more common
- Bacterial, Fungal
- Viral (HSV, HZO)
- Acanthamoeba

Autoimmune – much less common
- e.g. RA

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

Peripheral Ulceration and Thinning

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

**PUK causes**

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

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

- 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

Common to all cases of Neurotrophic Keratopathy is corneal hypoesthesia
**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
Anterior Uveitis

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 and High IOP

Think of:
- HSV or HZO with trabeculitis*
- Glaucomacyclitic Crisis (PS Syndrome)
- Lens Induced
- IOL- UGH Syndrome
- Steroid Responder
  (e.g. uveitis being treated with corticosteroid)
- Other: Sarcoidosis, Fuchs, JRA

**Also Consider**
- not real uveitis, but cells in AC
  - Pigment Dispersion Syndrome
  - Retinal Detachment
  - Ghost Cell Glaucoma
  - Masquerade Syndrome
    - RB, Lymphoma, etc.

Anterior Uveitis:
Get a good Review of Systems
e.g. Mouth Ulcers, Lung Disease, TB, Herpes Diarrhea, IBD, Genital Ulcers, Kidney Disease Arthritis, **Back Pain (Lower?)** — High Yield Shingles? Rashes, Atopic Symptoms

Main Work-up Test Considerations:
- HLA- B27 (and any others FTABS, RPR/VDRL* on the left that 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

Click to Return To Links
✓ 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
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
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 cysts
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
- Parry Romberg Syndrome
- Sturge Weber Syndrome
- Other Syndromes
- Uveitis – including Fuch’s and Posner Schlossman
- Pigment Dispersion
- Pigmented Iris Tumors
- Use of Prostaglandin analogs
- Siderosis Bulbi
- Trauma and Surgery
- R/O Benign Heterochromia
Lens Opacities and associations

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 **More Common Causes Include:**

- **Acute Angle Closure Attack or other causes of Pupillary Block** *(See upcoming lists)*
- **Open Angle Glaucoma (POAG, secondary forms – see upcoming lists)*
- **Post Intraocular surgery – CE, PK, DMEK/DSEK*, Vitrectomy, Glaucoma**
  
  Acute: Viscoelastic, Air Bubble, Pupillary Block, Aqueous Misdirection
  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**
- **Trabecular Outflow Obstruction – Lens Material, RBC, Tumor Cells**
- **Venous Outflow Problem – Orbital Apex Mass, CC Fistula**

* DMEK / DSEK = Descemet’s Membrane/ Stripping Endothelial Keratoplasty

**Acute IOP Elevation**

With sudden onset of symptoms (HA, N/V, Eye Pain, Decreased VA)
Think of:

**Acute ACG**
Post op –
  - Viscoelastic, Air
  - Aqueous Misdirection
**Uveitic – Lens, Herpetic**
**Hyphema**
**Orbital Causes – Hemorrhage, CCF**

**Click to Return To Links**
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
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
1. Primary Open Angle (POAG)

2. Secondary Open Angle (SPLIT mnemonic)
   - Substances: Steroids, Viscoelastics, Siderosis (Iron IOFB)
   - Pigmentary
   - Pseudoexfoliation
   - Lens – natural – phacolytic
   - Lens – 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

**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)
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
Hypotony

Causes and Associations

Low IOP

- 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

Pigment in Anterior vitreous
Whitish or Yellowish Retinal or Sub-Retinal Lesions

**Discrete Borders**
- Hard Exudates
- Vessel Plaques
- Certain retinal/choroidal tumors
- Drusen
- Macular Dystrophies
  - AD – pattern vary - look like ARMD in younger people – e.g. Adult-onset Vitelliform, Butterfly, Reticular, Stargardt’s like Dystrophies.

**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

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

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

Low internal reflectivity
**Hypotony**

**Causes and Associations**

- **Low IOP**
  - 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 Retinochoroididopathy, 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

- Low IOP
- 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
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

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

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

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

- Macula – Bull’s Eye

*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?
  - Mimics- Congenital Lesions, Myopic Degeneration
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
**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, ROP
- PVR (proliferative vitreal 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** Macular Detachments: Central Serous Retinopathy (CSR), Leukemia, Juxtafoveal Telangiectasia, Optic Disc Pit, Unilateral Acute Idiopathic Maculopathy (acute visual loss and viral prodrome e.g. coxsackie virus)
Sometimes with: posterior uveitis and Scleritis, choroidal neovascularization, Best’s, BDUMP
• 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

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

Causes:

Ischemia  - e.g. past AION or PION

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

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

High IOP   - e.g. OAG, AGG, CC Fistula, Inflammation, etc.

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, CCl₄
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, Neuretinitis, 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
### Differentiating between *Congenital* and *Acquired* Disc Elevation

<table>
<thead>
<tr>
<th>Feature</th>
<th>Congenital</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nerve Fiber Layer</td>
<td>Clear</td>
<td>Opacified</td>
</tr>
<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 (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
Possible Causes of Optic Nerve Inflammation

**Demyelinating:**
- Multiple Sclerosis, Neuromyelitis Optica = NMO (D evic’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)
- Pachymeningitis – (Idiopathic, intracranial hypotension, Meningeal carcinomatosis, vasculitides, infections such as p. acnes)
- Orbit (orbital pseudotumor – e.g. optic perineuritis), IgG4-ROD
- Sinuses -Infectious (including Fungal – e.g. mucor, aspergillus), Wegener’s Granulomatosis

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

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

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

*Remember certain acute optic neuropathies can resemble optic neuritis:*
like: NAION, PION, 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 Idiopathc 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...

- 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

- Also consider
  Congenital anomalous discs
  (not edema, mimic)

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
3. Miscellaneous Case Situations and other Lists

Visual Loss
Reading Problems
Pediatric Presentations: Visual Loss, ET, Glaucoma, Nystagmus
Pregnancy
Systemic Medical Conditions
CN Palsies
Elevated ESR and Concern for Temporal Arteritis
Low Tension Glaucoma
Ocular Effects of Systemic Medications
Imaging Findings- CT, MRI
Dizziness
Differing Scenarios of Visual Loss

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

Of course in any case of visual loss a complete history and exam are necessary and all diagnoses need to be considered, but the following lists can give direction for each scenario.
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:
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>
<tr>
<td>Classic Optic Neuritis</td>
<td>⭐</td>
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<tr>
<td>Retinal Detachment</td>
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<tr>
<td>CRVO</td>
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<tr>
<td>AION</td>
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<td>Anterior Ischemic Optic Neuropathy</td>
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<td>CSR</td>
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<tr>
<td>Central Serous Retinopathy</td>
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<tr>
<td>CRAO</td>
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<td></td>
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<tr>
<td>ARMD</td>
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<tr>
<td>Wet ARMD – SRNVM</td>
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<tr>
<td>Papilledema</td>
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<tr>
<td>High Intracranial Pressure</td>
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<tr>
<td>Optic Nerve Compression</td>
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<td>e.g. Tumor or Graves Disease</td>
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<tr>
<td>Vitreous Hemorrhage</td>
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<tr>
<td>or Pre-Retinal Hemorrhage</td>
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<td>Traumatic Optic Neuropathy</td>
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<tr>
<td>Neuro-retinitis</td>
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</tr>
</tbody>
</table>

*Ischemic CRVO*
“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 – see list
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
Other: Carotid Cavernous Fistula

Timing
Abrupt – ION, LHON
Subacute – optic neuritis
Insidious – compressive or metabolic

Character
Dark spot – optic neuropathy
Metamorphopsia - maculopathy
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
"Unexplained" Visual Loss
Loss of Visual Acuity and/or Visual Field without Clear Findings on Exam

Consider:

1. Missed Refractive Errors or Subtle Media Opacities or Distortions
   Use Refraction, Retinoscope, Direct Ophthalmoscope, Keratometer, Corneal Topographer

2. Past Retinal Events (RAO, RVO, Commotio, Paraneoplastic)
   and Past or Subtle Present Macular Disease (e.g. CME, Dystrophies, Ischemia, Degenerative, ERM, VMT)
   Use Amsler Grid, OCT and Auto-Fluorescence, Fluorescein Angiography

3. Subtle Optic Neuropathies (often without disc changes early on)
   - Inherited, Compressive, Toxic, Deficiency, Trauma, Neuro-Degenerative Diseases, Paraneoplastic
     Use Color Plates, RAPD, Automated VF testing, Orbital or Neuro-imaging, VEP= Visually Evoked Potentials

4. Cortical Visual Loss
   CVA, Anoxia, Toxic, Metabolic, Vasculitis, Trauma, Migraine, Encephalitis,
   Meningitis, Dementias, Trauma (SBS)
   Use VF Testing, Neuroimaging, LP

5. Other: Amblyopia (e.g. Anisometropia, Monofixation syndrome)
   Non-organic / Functional - Conversion, Hysteria, Malingering
   Use: Prism testing, Tangent VF testing, OKN Drum, Stereoacuity

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. Sarcoidosis, Electrocution, Occult Neoplasm, Dementias (Alzheimer’s and possibly other causes),

5. **Other**
   - Amblyopia – especially think about anisometropic amblyopia, monofixation syndrome
   - and consider: Non-Organic (Functional) Visual Loss
Unexplained Bilateral or Quickly Sequential Acute Visual Loss

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

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

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

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 or 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)
- 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

ERG appropriate in suspected Retinal Dystrophy

Neuroimaging needed when see an optic nerve abnormality or cannot explain cortical visual loss
High IOP Suspect in an Infant
Presenting with Large eye (Pseudo-Proptosis), tearing, corneal clouding

WHAT DO YOU CONSIDER??

1. Congenital or Juvenile Glaucoma (70% bilateral)

2. Anterior Segment Dysgenesis
e.g. Peter’s Anomaly (Central opacity, 80% bilateral, >50% glaucoma)
   Axenfeld-Reiger’s, Aniridia


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
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

### Condition | Common Age of Presentation
--- | ---
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 Oscillospia, Vertigo or Imbalance. Absence of Neurologic abnormalities outside of Visual System
   Predominately Horizontal Conjugate Jerk or Pendular Nystagmus, Dampens at Near/Convergence
   Increases with attempted Fixation or pursuit. Often a Null point – central or eccentric gaze

2. **“Eye” Problems – Sensory Visual Loss**
   e.g. Deprivational Amblyopia,
   Congenital Cataracts, Anterior Segment Dysgenesis (e.g. Peter’s Anomaly)
   PHPV, ROP, Albinism, Foveal Hypoplasia, Macular Dystrophy or Scar
   Retinal Dystrophies: LCA, CSNB, Cone-Rod Dystrophy, Achromatopsia
   Optic Nerve Anomaly (e.g. Optic Nerve Hypoplasia, Optic Atrophy)

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
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
Important Medical Conditions and their Associated Eye Pathology

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

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**

**Orbital or Cavernous Sinus Lesion** – tumor, aneurysm, fistula

**Cranial Nerve Palsies**

3rd Oculomotor
4th Trochlear
6th Abducens

**Cranial Nerve**

**3rd** Oculomotor
**4th** Trochlear
**6th** Abducens

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**Migraines***

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

**Shunt Failure**

**Orbital or Cavernous Sinus Lesion** – tumor, aneurysm, fistula

**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.**
Cranial Nerve Palsies
More Specific Causes related to type of Palsy

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

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

4\textsuperscript{th} CNP
1. Traumatic
2. Congenital
3. Microvascular
4. Other Causes - uncommon
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 patient with cancer are anemic)

5. Hyperproteinemias - e.g. Multiple Myeloma, Gammaglobulinemias

6. Active Infection: Sinusitis, Ear Infection, Mastoiditis, Dental (all could produce pain in area of Temporal Arteritis)
   Wound infection, Prostatitis, Osteomyelitis

7. Other: End Stage Renal Disease (ERSD) / Dialysis, Idiopathic Hypertrophic Cranial Pachymeningitis

Suggested work-up for patient with no obvious cause for an elevated ESR:
   C-Reactive Protein, CBC (with platelets), Urinalysis, BUN/Cr, alkaline phosphatase,
   Serum Protein Electrophoresis, Chest X-Ray, CT of Sinuses, or a temporal artery biopsy?

Temporal and Forehead Pain and Tenderness
   Think about GCA yes, but also consider:
   - Early / Prodrome VZV / Shingles
   - Trigeminal Neuralgia
   - Sinus, Ear, Mastoid, Dental Infections
   - Other neuralgias – Occipital, Cervical, Supra Orbital
   - Unilateral Headaches – Migraine, Cluster, Tension

Beware of Artificially Low ESR values in Patients with Biopsy Positive GCA
   e.g. Pts on Statins and Non-Steroidal Anti-Inflammatory Drugs (NSAIDS) can have lower ESR, but not CRP. (JNO 2011;31:135)
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: levadopa, 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, Indomethacin, 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

Enlarged Extra Ocular Muscle(s)

- Graves Ophthalmopathy (IR, MR > SR most common)
- Orbital Pseudotumor (Myositis)
- Post- Viral Myositis (Children), Cysticercosis
- Lymphoma, CLL
- Metastatic Disease – breast, melanoma
- Primary Tumor – e.g. hemangioma, fibrous / Amyloid

Boney Growth / Change / Expansion

- Langerhans Histiocytosis (1-4 yo)
- Fibrous Dysplasia (children- young adults)
- Sarcoma of the bone
- Sphenoid Wing Meningioma
- Paget’s Disease (older patients)
- Metastatic Disease to the Bone
MRI of the Brain
(White Matter Lesions)

White Matter Lesions

- Demyelinating Disease
  - MS
  - ADEM = Acute Disseminated Encephalomyelitis (post viral or immunization)
- Older patients with any small vessel disease particularly with DM, HTN
- Migraine patients, Chronic Migraine
- Vasculitis - acute and chronic small vessel disease of brain
- Younger patients with uncontrolled or poorly controlled HTN
- Anoxic Encephalopathy – e.g. CO poisoning
- Primary Angiitis of the CNS
- Anti - Phospholipid Antibody Syndrome
- Rapid Correction of Hyponatremia (Osmotic Demyelination Syndrome)
- AIDS
- PML (Progressive Multifocal Leukoencephalopathy – viral disease in immunocompromised)
- Radiation
Dizziness

Vertigo - Peripheral or Central causes – associated with nystagmus

Peripheral:
- BPPV* (seconds < minute associated with head posturing, no tinnitus, hearing OK)
- Acute Labyrinthitis/Vestibular Neuritis (hours to 2 weeks, no tinnitus, hearing OK)
- Meniere’s Disease (hours, +hearing loss, +tinnitus)
- Drug Toxicity (Aminoglycosides, anti-seizure meds, alcohol, ASA, chemo)
- 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.

- *BPPV- Benign Paroxysmal Positional Vertigo

- Shopping Cart Syndrome (B. Farris – COS 2013) – symptoms of dizziness in patients (with prior BPPV or labyrinthitis) – when in an environment which can induce OKN – such as pushing shopping cart down visually busy grocery aisle.
AC - Anterior Chamber
AC1 – Arnold Chiari Malformation Type 1
ACE – Angiotensin Converting Enzyme
ACG – Angle Closure Glaucoma
AD – Autosomal Dominant
ADOA – Autosomal Dominant Optic Atrophy
AIBSES – Acute Idiopathic Blind Spot Enlargement Syndrome
AIDS – Acquired Immune Deficiency Syndrome
AION – Anterior Ischemic Optic Neuropathy
AAION – Arteritic AION (GCA)
AK – Astigmatic Keratotomy
AMPPE – Acute Multifocal Posterior Placoid Epitheliopathy
ANA – Anti-Nuclear Antibody (Lupus Diagnosis)
AR – Autosomal Recessive
ARMD – Age Related Macular Degeneration
ARN – Acute Retinal Necrosis
AVM – Arterial Venous Malformation
BEB – Benign Essential Blepharospasm
BCCA – Basal Cell Carcinoma
BCP – Birth Control Pills, Contraceptives
BDUMP – Bilateral Diffuse Uveal Melanocytic Proliferation
BP – Blood Pressure
BPES - Blepharophimosis- Ptosis- Epicanthus Inversus Syndrome
BPV – Benign Positional Vertigo
BRAO- Branch Retinal artery Occlusion
BRVO- Branch RVO
CA – Carcinoma
CARS – Cancer Associated Retinopathy Syndrome
CC – Carotid Cavernous
CE – Cataract Extraction (Surgery)
CFEOM – Congenital Fibrosis of Extraocular Muscles
CHRPE – Congenital Hypertrophy of the RPE
CIN – Carcinoma in Situ
CLL – Chronic Lymphocytic Leukemia
CME – Cystoid Macular Edema
CMV – Cytomegalovirus
CN- Cranial Nerve
CN palsy
CNS – Central Nervous System
CPA – Cerebellar Pontine Angle
CPEO – Chronic Progressive External Ophthalmoplegia
CR – Chorioretinal
CSR – central retinal artery occlusion, CRVO- central ret. vein occlusion
CSNB – Central Serous Retinopathy
CT- Computerized Tomography
CTA – CT Angiogram
CTL – Contact Lens
CVA – Cerebral (CNS) Vascular Accident (Stroke)
CXR – Chest X-Ray
DDX – Differential Diagnosis
DOA – Dominant optic atrophy
DR – Diabetic Retinopathy
DME- Diabetic Macular Edema
DM – Diabetes Mellitus
DUSN – Diffuse Unilateral Subacute Neuroretinitis from Nematode
EOM – Extra Ocular Muscle
ERG – Electroretinogram
ERM – Epi - Retinal Membrane
ESR – Erythrocyte Sedimentation Rate
ET – Esotropia
FA- Fluorescein Angiogram
FB- Foreign Body
FBS – Foreign Body Sensation
FEVR - Familial exudative vitreoretinopathy
FhX – Family Medical History
GBS- Guillain-Barre Syndrome
GCA – Giant Cell Arteritis (aka Temporal Arteritis)
<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
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<tbody>
<tr>
<td>GPA</td>
<td>Granulomatosis with Polyangiitis (Wegener's)</td>
</tr>
<tr>
<td>HA</td>
<td>Headache</td>
</tr>
<tr>
<td>HIV</td>
<td>Human Immunodeficiency Virus (AIDS)</td>
</tr>
<tr>
<td>HLA</td>
<td>Human Leukocyte Antigen</td>
</tr>
<tr>
<td>HSV</td>
<td>Herpes Simplex Virus</td>
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<tr>
<td>HT</td>
<td>Hypertrophia</td>
</tr>
<tr>
<td>HoT</td>
<td>Hypotrophia</td>
</tr>
<tr>
<td>HTN</td>
<td>Hypertension (high blood pressure)</td>
</tr>
<tr>
<td>HZO</td>
<td>Herpes Zoster Ophthalmicus</td>
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<tr>
<td>IBD</td>
<td>Inflammatory Bowel Diseases</td>
</tr>
<tr>
<td>ICE</td>
<td>Iridocorneal Endothelial (Syndrome)</td>
</tr>
<tr>
<td>ICP</td>
<td>Intracranial Pressure</td>
</tr>
<tr>
<td>IgG4-ROD</td>
<td>Immunoglobulin G4 Related Ophthalmic Disease</td>
</tr>
<tr>
<td>IIH</td>
<td>Idiopathic Intracranial Hypertension (pseudotumor cerebri)</td>
</tr>
<tr>
<td>INO</td>
<td>Internuclear Ophthalmoplegia</td>
</tr>
<tr>
<td>IO</td>
<td>Inferior Oblique</td>
</tr>
<tr>
<td>IOFB</td>
<td>Intra-Ocular Foreign Body</td>
</tr>
<tr>
<td>IOIS</td>
<td>Idiopathic Orbital Inflammatory Syndrome</td>
</tr>
<tr>
<td>IOL</td>
<td>Intraocular lens</td>
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<tr>
<td>ION</td>
<td>Ischemic Optic Neuropathy</td>
</tr>
<tr>
<td>IOP</td>
<td>Intraocular Pressure</td>
</tr>
<tr>
<td>IR</td>
<td>Inferior Rectus</td>
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<tr>
<td>JIA</td>
<td>Juvenile Idiopathic Arthritis</td>
</tr>
<tr>
<td>JRA</td>
<td>Juvenile Rheumatoid Arthritis</td>
</tr>
<tr>
<td>KC</td>
<td>Keratoconjunctivitis</td>
</tr>
<tr>
<td>KP</td>
<td>Keratoprecipitates</td>
</tr>
<tr>
<td>LASIK</td>
<td>Laser Assisted In Situ Keratomileusis</td>
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<tr>
<td>LCA</td>
<td>Leber's Congenital Amaurosis</td>
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<tr>
<td>LHON</td>
<td>Leber's Hereditary Optic Neuropathy</td>
</tr>
<tr>
<td>LG</td>
<td>Lacrimal Gland</td>
</tr>
<tr>
<td>LL</td>
<td>Lower Lid (Lower Eyelid)</td>
</tr>
<tr>
<td>LLL</td>
<td>Left Lower Eyelid</td>
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<tr>
<td>LP</td>
<td>Lumbar Puncture (Spinal Tap)</td>
</tr>
<tr>
<td>LR</td>
<td>Lateral Rectus</td>
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<tr>
<td>LSCD</td>
<td>Limbal Stem Cell Deficiency</td>
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<tr>
<td>LUL</td>
<td>Left Upper Lid</td>
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<tr>
<td>MARS</td>
<td>Melanoma Associated Retinopathy Syndrome</td>
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<tr>
<td>MEWDS</td>
<td>Multiple Evanescent White Dot Syndrome</td>
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<tr>
<td>MFS</td>
<td>Miller Fisher Syndrome (polyneuropathy)</td>
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<tr>
<td>MG</td>
<td>Myasthenia Gravis</td>
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<tr>
<td>MM</td>
<td>Multiple Myeloma</td>
</tr>
<tr>
<td>MR</td>
<td>Medial Rectus</td>
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<tr>
<td>MRA</td>
<td>Magnetic Resonance Angiography</td>
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<tr>
<td>MRD</td>
<td>Margin Reflex Distance (Upper Eyelids)</td>
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<tr>
<td>MRI</td>
<td>Magnetic Resonance Imaging</td>
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<tr>
<td>MRV</td>
<td>MR Venography</td>
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<tr>
<td>MS</td>
<td>Multiple Sclerosis</td>
</tr>
<tr>
<td>NAION</td>
<td>Non-Arteritic AION</td>
</tr>
<tr>
<td>NF</td>
<td>Neurofibromatosis</td>
</tr>
<tr>
<td>NFL</td>
<td>Nerve Fiber Layer</td>
</tr>
<tr>
<td>NLO</td>
<td>Nasolacrimal Obstruction</td>
</tr>
<tr>
<td>NLDO</td>
<td>NL Duct Obstruction</td>
</tr>
<tr>
<td>NPDR</td>
<td>Non-Proliferative Diabetic Retinopathy</td>
</tr>
<tr>
<td>NVA</td>
<td>Near Visual Acuity</td>
</tr>
<tr>
<td>NVD</td>
<td>Neovascularization of the Disk</td>
</tr>
<tr>
<td>NVI</td>
<td>Neovascularization of the Iris</td>
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<tr>
<td>OAG</td>
<td>Open Angle Glaucoma</td>
</tr>
<tr>
<td>OCP</td>
<td>Ocular Cicatricial Pemphigoid</td>
</tr>
<tr>
<td>OCT</td>
<td>Ocular Coherence Tomography</td>
</tr>
<tr>
<td>OIS</td>
<td>Ocular Ischemic Syndrome</td>
</tr>
<tr>
<td>OKN</td>
<td>Opto-Kinetic Nystagmus</td>
</tr>
<tr>
<td>ONH</td>
<td>Optic Nerve Head</td>
</tr>
<tr>
<td>OMG</td>
<td>Ocular Myasthenia Gravis</td>
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<tr>
<td>ONM</td>
<td>Ocular Neuromyotonia</td>
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<tr>
<td>OSSN</td>
<td>Ocular Surface Squamous Neoplasia</td>
</tr>
<tr>
<td>PAN</td>
<td>Polyarteritis Nodosa</td>
</tr>
<tr>
<td>PC</td>
<td>Posterior Chamber</td>
</tr>
<tr>
<td>PCO</td>
<td>Posterior Capsular Opacity (Pseudophakic eye)</td>
</tr>
</tbody>
</table>
PDR – Proliferative Diabetic Retinopathy
PEK – Punctate Epithelial Keratopathy = SPK
PET-CT - Positron emission tomography–computed tomography
PHPV – Persistent Hyperplastic Primary Vitreous
PPM – Persistent Placoid Maculopathy
PPV – Pars Plana Vitrectomy (Surgery)
PI – Peripheral Iridectomy or Iridotomy
PION – Posterior Ischemic Optic Neuropathy
PK – Penetrating Keratoplasty (corneal transplant)
POAG – Primary Open Angle Glaucoma
PON – Paraneoplastic Optic Neuropathy
PORN – Progressive Outer Retinal Necrosis
PRK – Photorefractive Keratectomy
PRP – Pan-Retinal Photocoagulation
PSC – Posterior Sub-Capsular (Cataract)
PUK – Peripheral Ulcerative Keratitis
PVD – Posterior Vitreous Detachment
RA – Rheumatoid Arthritis
RAO – Retinal Artery Occlusion
RAPD – Relative Afferent Pupillary Defect
RB – Retinoblastoma
RBC – Red Blood Cells
RD – Retinal Detachment
RES – Recurrent Erosion Syndrome
RLL – Right Lower Lid
RGP – Rigid Gas Permeable (CTL)
RK – Radial Keratotomy
RMSF – Rocky Mountain Spotted Fever (Rickettsia)
R/O – Rule Out
ROP – Retinopathy of Prematurity
RP – Retinitis Pigmentosa
RPE – Retinal Pigment Epithelium
RUL – Right Upper Eyelid
RVO – Retinal Vein Occlusion
SAH – Subarachnoid Hemorrhage
SBS – Shaken Baby Syndrome
SCH – Sub-Conjunctival Hemorrhage
SCA – Squamous Cell Carcinoma
SJ – Stevens Johnson Syndrome
SLK – Superior Limbic Keratoconjunctivitis
SLE – Systemic Lupus Erythematosus
SNP – Supranuclear Palsy
SO – Superior Oblique
S/P – Status Post
SPK – Superficial Punctate Keratitis
SR – Superior Rectus
SRNVM – Sub-Retinal Neovascular Membrane
TASS – Toxic Anterior Segment Syndrome
TB – Tuberculosis
TBI – Traumatic Brain Injury
TM – Trabecular Meshwork
TORCH – (Toxoplasmosis, Other, Rubella, Cytomegalovirus, Herpes Simplex)
UA – Urine Analysis
UGH – Uveitis Glaucoma Hyphema Syndrome
(Pseudophakic eye AC or PC IOL)
UL – Upper Lid (Eyelid)
URI – Upper Respiratory Infection
VA – Visual Acuity
VF – Visual Field
VKH – Vogt-Koyanagi-Harada Syndrome
VMT – Vitreo-Macular Traction
VPT – Vitreo-Papillary Traction
VZV – Varicella-Zoster Virus
WBC – White Blood Cells
W/U – work up
XT – Exotropia
<table>
<thead>
<tr>
<th>Journal Name</th>
<th>Abbreviation</th>
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<tr>
<td>Ophthalmology (AAO Journal)</td>
<td>OPH</td>
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<td>Survey of Ophthalmology</td>
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<td>JAMA Ophthalmology (Archives)</td>
<td>JAMA Oph</td>
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<tr>
<td>American Journal of Ophthalmology</td>
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<td>Investigative Ophthalmology and Visual Science</td>
<td>IOVS</td>
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<td>Medium</td>
<td>Major References</td>
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<tr>
<td>Book</td>
<td>Hampton Roy, Ocular Differential Diagnosis</td>
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<td>Monograph</td>
<td>American Academy of Ophthalmology – Basic and Clinical Science Series</td>
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<td>Internet</td>
<td>Emedicine Ophthalmology – Multiple articles from website</td>
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<td>AAO- EyeWiki</td>
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The heavens declare the glory of God; the skies proclaim the work of his hands

Psalm 19:1