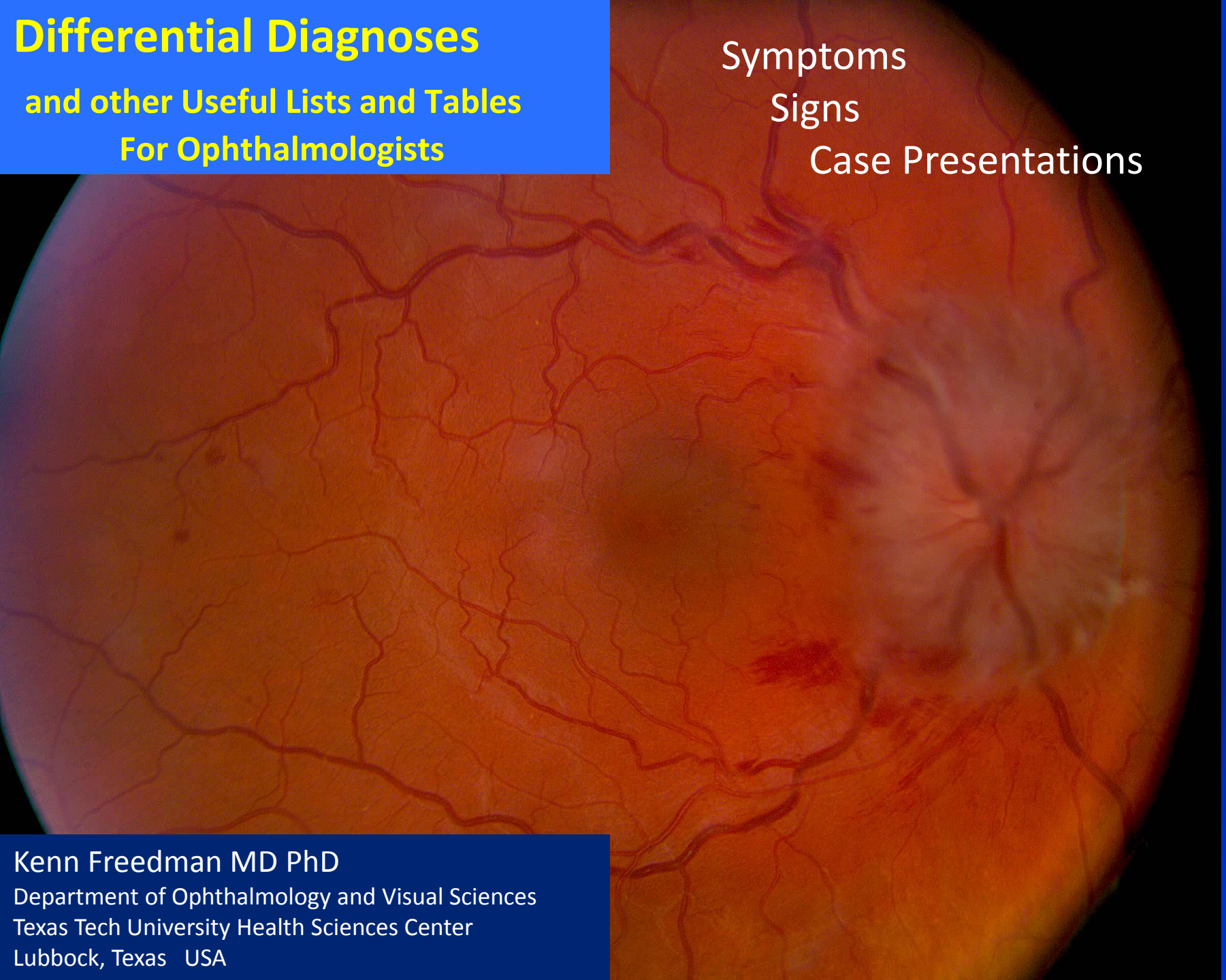
Differential Diagnoses

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.*

* Please see references at end of document

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

Kenn Freedman

Lubbock, Texas - October 2018

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.

Use of this Presentation

The lists are divided into three main areas

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

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

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

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

Symptoms

Loss of Visual Acuity and Refractive Issues

Loss of Visual Field (VF)

Eyelids and Orbit

Index of all Lists with Links

Motility and Alignment Problems

Pupils

Anterior Segment

Index of all Lists with Links

Fundus - Vitreous, Retina and Optic Nerve

LIST OF ABBREVIATIONS

IOP and Glaucoma

Miscellaneous Case Situations and Lists

Index of all Lists with Links

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

Loss of Vision (General Considerations)

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.

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Transient Visual Phenomena

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1. Negative - see Transient Visual Loss – *"seeing less"*

– loss of Visual Acuity, Visual Field or Color Vision, Blurring of Vision

Scotomas or Anopsias, Dimming or Loss of Brightness

2. Positive - *"seeing more"* – distortions of / or additional objects

a) Normal Phenomena – e.g. afterimages, physiologic diplopia

b) Entopic Phenomena – seeing own eye structures – WBCs, retinal vessels, floaters

c) Illusions - Misperceptions of external objects – *close eyes and image is gone*

e.g. alterations in size (aniseikonia) , shape (metamorphopsia), color (chromatopsia- tinting) number (diplopia, multiplopia – consider optical and alignment problems) and Palinopsia – migraine, psychoactive drugs, medications (e.g. topiramate, acetazolamide, clomiphene), head trauma, lesions in parietal occipital visual pathways, metabolic

d) Hallucinations - Sensory experiences not based on incoming information - *close eyes and image is still there*

e.g. Flashes (see Photopsias) , formed and unformed objects

- Psychiatric disturbances, Psychoactive medications and Rx drugs (see ocular effects of medications)
- Cortical Lesions – Palinopsia
- Charles Bonnet Syndrome (in cases of severe loss of vision- e.g. ARMD, Optic atrophy)
- Migraine Phenomena (Aura's etc.)

Floaters

- Vitreous Syneresis
- R/O Retinal Detachment

  Especially in the case of new floaters!
- Vitreous Detachment (e.g. PVD)

- Vitreous Hemorrhage
- Posterior or Intermediate Uveitis
- Other sources of Vitreous Cells

  e.g. Masquerade Syndrome for Uveitis: Lymphoma or Tumor (RB, Melanoma)

- Other Unusual Causes (in Vitreous)

  Asteroid Hyalosis, Amyloidosis, Cholesterol Crystals- Synchysis scintillans

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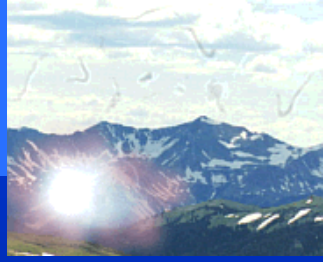
Flashing Lights Photopsias



Monocular

Vitreo-Retinal Traction

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

Posterior Vitreous Detachment (PVD)

R/O Retinal Tear and Detachment

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Retinitis / Choroiditis

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

Retinal Ischemia: Impending CRVO, DR, OIS

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

Ocular / Retinal Migraine

Optical "Dysphotopsias"
(Not true Photopsias, optical effects)

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

Photopsias Continued ➔

Flashing Lights Photopsias

Binocular

Migraine Aura
Epilepsy – Occipital Lobe epilepsy can mimic migraine
CNS lesion – Tumor, MS, AVM, Ischemia and CVA* (often expect VF loss also)
Vertebro-Basilar Insufficiency
Non-Ketotic Hyperglycemia
Other: e.g. Midbrain Infarct

Other Monocular or Binocular

Retinitis Pigmentosa
CARS, MARS
Persistent Positive Visual Phenomena in Migraine (PPVPM)
R/O Altered Mental States and Psychoactive Drugs

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

Notes: Migraine onset later in life is possible
Snow like pattern (TV) – think of Outer retinal disorders, PPVPM
*41% with Retrochiasmal Infarct have Photopsias – many not aware of VF Loss

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

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Diplopia

Use Cover Testing to Separate into Monocular and Binocular Diplopia

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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

Monocular (Usually Optical) Diplopia

1. Uncorrected Refractive Error

   e.g. regular astigmatism, wrong glasses or CTLs

2. Corneal problem/ distortion

   e.g. Asymmetric and Irregular Astigmatism

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

   Corneal deposits or dystrophies

   Transient corneal deformation on downgaze by LL

3. Cataract, Other Media Opacity

   e.g. PSC or Oil Droplet NS, Vitreous

4. Other Optical

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

   Iris – Polycoria, defects

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

   Macular Disorder: ERM, ARMD, etc.

   CNS- e.g. MS, Palinopsia

   Functional

Optical: Often have this “Ghost Image”

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Binocular Diplopia – Causes of Ocular Misalignment

1. **Neurologic**

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

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

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

   Systemic - e.g. polyneuropathy GBS, MFS, Medication induced

E E

Two distinct images

2. **Myogenic**

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

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

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3. **Orbital Disease**

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

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

4. **Loss or Distortion of Vision**

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

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

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

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

Transient Diplopia

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

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

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

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

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

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

Ocular Neuromyotonia (ONM) , Spasm of Near Triad, **Superior Oblique Myokymia**,
Toxicities (Drug abuse, toluene, Wernicke's)
Medications (Anesthetic injections – brainstem circulation, Meds that induce / mimic MG)
Metabolic (some drugs at peak levels, High K+, hepatic encephalopathy)
Transient Optical / Monocular Diplopia possible (e.g. transient corneal deformation on downgaze by LL)
Exercised Induced Transient Diplopia (case report of transient ET)

Oscillopsia

- *Acquired Nystagmus* *(see list)* , but not Congenital

  CNS lesion or inflammation, Metabolic, Drug or Toxin Related

- Other Involuntary Eye Movements- e.g. Opsoclonus
- Superior Oblique Myokymia

  Idiopathic, Trauma, Vascular Compression
- Vestibular Problem
- Lid Orbicularis Myokymia
- Head Tremor or “Titubations” (MS)
- IOL - loose - “Flutter”
- Intermittent Exotropia?

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Night Blindness (Nyctalopia)

Before considering true Nyctalopia

Consider other problems people encounter at night:

Uncorrected refractive errors maybe first present as night problems

e.g. Early Myopia, Latent Hyperopia

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

Normal Pupillary Dilation in dark can bring out any optical aberrations

e.g. uncorrected refractive error, cataract, corneal

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

1. Congenital Stationary Night Blindness*
2. High Myopia
3. Progressive Chorio-retinal disorders:

Retinitis Pigmentosa - various forms including Retinits Punctata Albescans

Previously undiagnosed LCA with incomplete penetrance

Chorioretinal degenerations- Choroideremia, Gyrate atrophy

Medications: - phenothiazines, chloroquines, quinine

Vitamin A Deficiency

Siderosis and other Toxicities

CARS and MARS (Cancer and Melanoma Associated Retinopathy Syndrome)

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

Optic Neuropathies could present with Nyctalopia

Transient Visual Loss (TVL)

1. Transient Visual Obscurations (TVO)

mono or binocular - few seconds

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

Key History Points:
Mono or Binocular?
Time Frame
Pain?

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)

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Monocular TVL

Carotid Disease

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

Heart Disease

- Hypoperfusion or source of emboli
- Valvular disease, endocarditis, arrhythmia, cardiomyopathy, Arial 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:

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Migraine Aura

Cortical Ischemia

e.g. Vertebro-Basilar Insufficiency

Papilledema

Medications: Digitalis, Viagra

Transient Cortical Blindness

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

Migraine Mimickers

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

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

Photophobia (pain with bright lights)

Think about Ocular and Intracranial Trigeminal Irritation

Ocular Trigeminal Irritation

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

Any ocular or visual abnormality could be associated with this symptom

Intracranial Trigeminal Irritation

– Meningitis, Subarachnoid Hemorrhage, Trigeminal Neuralgia, Pituitary Apoplexy, Intracranial Hypotension

Parasellar tumors - Basal meninges of sella richly innervated and tumors in this area can give photophobia

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

Other

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

Glare or Dazzle – sometimes mistaken for photophobia

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

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Headache

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

Asthenopia – with near vision
High IOP - including Angle Closure
Uveitis, Scleritis
Orbital Tumor
Orbital Cellulitis / Pseudotumor
Subclinical / Occult Dacryocystitis
Dissection of Carotid artery – look for **Horner's**
Chiari Malformation – HA, ET or downbeat nystagmus
Pituitary Tumor or Apoplexy
Other Parasellar Tumor
Cranial Nerve Palsy – even Microvascular
Aneurysm Compression **/ SAH** - (e.g. **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)

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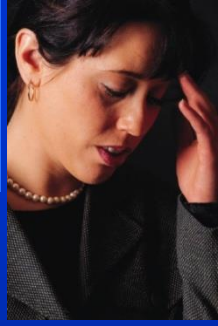
See Also - List for Eye / Periorbital Pain

* 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
high IOP
inflammation
VF loss
ocular and orbital ischemia
Neoplasia

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

Asthenopia
Recurrent Erosion Syndrome
Occult / Sub-clinical NL infection: Dacyrocystitis, Canaliculus
Supraorbital Neuralgia
Tenosynovitis - trochlea
Chronic uveal irritation from IOL
Ocular Ischemic Syndrome
Orbital Infarction Syndrome

Both associated with carotid obstruction and can have dull aching pain

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 - Horners Syndrome, rhinorhea, upper facial and scalp pain
Carotid dissection – pain, Horner syndrome
Ramsay Hunt Syndrome: Zoster of external auditory canal, facial n. palsy
TMJ Syndrome - pain not limited to jaw joint region (Temporomandibular Joint)

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

Early :

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

Other Considerations:

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

Pain on Eye Movement

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

Epiphora is a very non-specific Symptom

Before Direct Assessment of the Nasolacrimal Drainage System

Consider External Conditions or other irritants that can evoke tearing

Epiphora (Tearing)

e.g. Uncorrected Refractive Error

Ocular Allergies

Chronic 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

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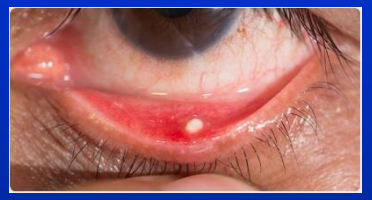
Nasolacrimal obstructions

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

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

Foreign Body Sensation 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

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Problem Opening Eyelid(s)

- Blepharospasm – eyelids squeeze (see list)
- Apraxia of Eyelid Opening*
  – neurological – problem initiating eyelid opening

- Ptosis (though usually not complete closure)
  - Neurologic (3rd Nerve)
  - Muscle or Neuromuscular Disorder
  - Congenital, Mechanical, Inflammatory

- Dry Eyes/Blepharitis
- Recurrent Erosion Syndrome (RES)
  – eyes often stuck in the mornings

Need to get a good history

Click to
Return To Links

Chronic Red Eye

Chronic Conjunctivitis

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

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

Chronic Ocular Inflammation:

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

Two important points:

1. Don't just think infection as most are self-limited and need to think about some other process.

2. Don't let corneal signs focus you too much on the cornea, e.g. chronic epithelial defects can be a sign of chronic dry eyes, chronic allergic disease, chronic eyelid problems, Neurotrophic (CN 5 and or 7 dysfunction), etc.

Orbital Disease

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

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

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

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Autoimmune Disease w/ related episcleritis, scleritis
Reiter's, Wegener's, Relapsing Polychondritis, SLE, Sjogrens Syndrome
OCP , Epidermolysis Bulosa (Symblephara)
Graft versus Host Disease

Loss of 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

Think "Front to Back" (Glasses to Brain)

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- 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)

Refractive Changes and Shifts

Acquired *Myopia*

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

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Excessive or Spasm of Accommodation

Lengthening of the Eye:

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

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

Lens Hydration Changes – (DM, Pregnancy, Toxemia)

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

Look for Oil Droplet Changes

CB effusion / edema → pushing lens-iris diaphragm anteriorly

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

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

Lens anterior dislocation* - (Crystalline lens or IOL)

Ciliary Muscle Spasm – Young patients, cholinesterase inhibitors

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

Transient Myopia (days) seen after blunt eye trauma

Post CE – early or late capsular dissention syndrome

Refractive Changes and Shifts

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, Cholorquine, Anticholinergics, Cannabis

- S/P RK or other corneal surgery
  with subsequent flattening of Cornea Curvature

- Shortening of distance from cornea to macula
  Orbital Mass pressing on Posterior Globe
  Central Serous Retinopathy (CSR)
  Short or Small Eye

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Refractive Changes and Shifts

- Shift in Astigmatism

change in **cylinder power or axis**

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Corneal – changes in anterior or posterior curvature:

Normal / Common Variations in Regular Astigmatism
Shift to Against the Rule Astigmatism with Aging
Eyelid Weight on Cornea – Ptosis, Chalazion, etc.
Lesions at or near Limbus inducing astigmatism – e.g. pterygium
Shifts due to Corneal Incisions (e.g. after CE, RK, AK, PK, etc.)
Radial Sutures through sclera or corneal wounds
Corneal Ectatic Degenerations (e.g. Keratoconus) – especially for high cylinder
Trauma – laceration
Inflammation (e.g. ulcer, marginal degenerations, etc.)
Contact Lens Wear (extended)

Lens (Lenticular)

Changes in the Crystalline Lens - Physical, Metabolic or Position
IOL – shifts/ tilts in position

Asymmetric and Irregular Corneal Astigmatism

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

Causes

1. Ectatic Corneal Dystrophies
   - Keratoconus*, Keratoglobus

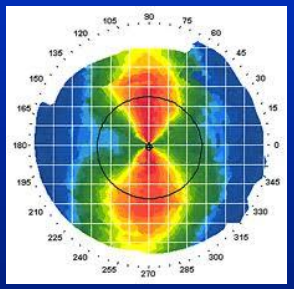
2. Corneal Warpage from
   - Longstanding CTL wear
   - Excessive Eye Rubbing
   - Prolonged Downgaze (e.g. reading- with deformation by LL) – usually transient

2. Corneal Marginal Disorders
   - Pellucid Marginal Degeneration
   - Terrien's or Mooren's Ulcers, RA

3. Contact Lens
   - Warpage
   - Poor Fitting CTL

4. Corneal Scarring/ Irregularities after
   - Corneal Ulcer
   - Corneal Trauma / Laceration
   - Corneal Refractive Surgery (LASIK, RK; ectasia)

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



Regular

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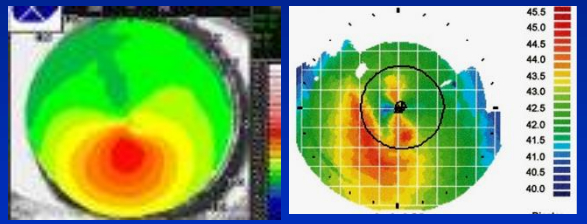

Irregular

Dull or Abnormal Retinoscope Reflex

- High Spherical Error – Myopic or Hyperopic
- High and / or Oblique Astigmatism
- Irregular astigmatism

  e.g. Keratoconus, Corneal Scarring, etc.
- Media Opacity

  Including Subtle cataract changes

  e.g. - star / flower pedal nuclear cataracts

  - oil droplet NS changes
- Posterior Defect – e.g. Coloboma

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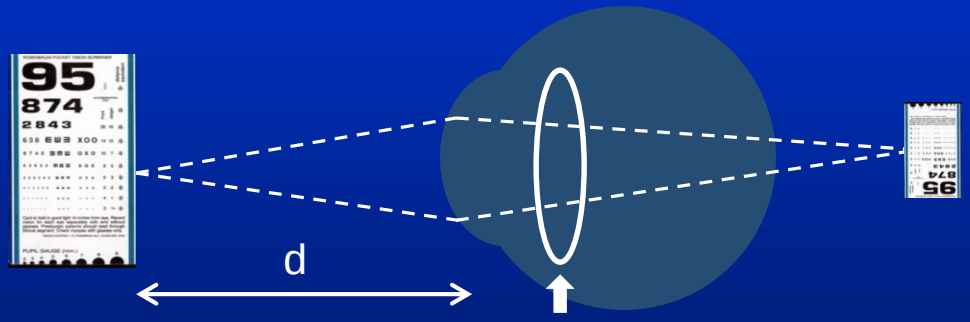
Poor Near Visual Acuity
Despite Good Distance VA

1. Loss of Accommodation

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

Other causes of Loss
Aphakia, Pseudophakia
Cataract
Lens Subluxation
Head Trauma
Eye and Orbital Trauma
Encephalitis and Meningitis
Midbrain Disease
Oculomotor Palsy
Tonic Pupils – e.g. Adie's Syndrome
Diabetes
Pharmacologic and Toxic agents: e.g. Atropine, other anticholinergics
Infections: syphilis
Systemic Medications: Amitriptyline
Glaucoma
Myasthenia Gravis
Uveitis
Retinal/ Scleral Treatments (laser or cryo)
Pregnancy
Functional (Non-Organic)
Benign Syndrome of Transient Loss of Accommodation in Young Patients
(Idiopathic, lasting months – JAMA Oph 2008; 126:1643)
Other: Botulism, Diphtheria, Viral Diseases : Influenza, Chicken Pox and some other Exanthems, GBS



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

2. Other Considerations:

Central Cataract – e.g. PSC with Miosis (Recall Near Triad)
Glasses Problems: e.g. Bifocal Segment
See Also "Problems with Reading" List

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Problems with Glasses

Patients Complaint's

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)

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

Problems with Near

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

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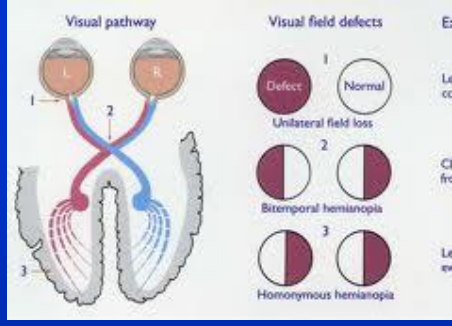
Loss of Visual Field

Visual Field Defects and Localizing Lesions



- Unilateral – Retina, Optic Nerve (rarely early Chiasmal)

- Bilateral – 1. Bilateral retinal or optic nerve disease *(but usually there is little symmetry)*
  2. Chiasm or farther posterior *(usually some kind of symmetry)*

- Vertical Midline respect – *suggests lesion at Chiasm or Posteriorly*

- Horizontal Midline respect – *usually optic nerve or retinal disease, **but** can be a lesion of the optic radiations or just the superior or inferior portions of occipital lobe*

- Homonymous Hemianopsia w/ other symptoms :

  Occipital Lobe – *often silent with no other non-visual symptoms*
  Parietal Lobe *(hemiparesis, visual perception and spatial problems, right left confusion)*
  Temporal Lobe *(seizures, formed visual hallucinations, agnosias)*

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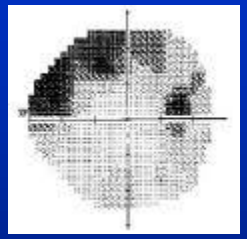
Recall the Optic Nerve / Tract Exam Signs
that *suggest a lesion anterior to Lateral Geniculate Nucleus (LGN)*:*
e.g. RAPD, Pallor / Atrophy, Edema

VF Defects respecting the *Horizontal Midline*

Suggestive of Optic Nerve Problem

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



Examples

Altitudinal
Arcuate
Nasal Step
Temporal Wedge

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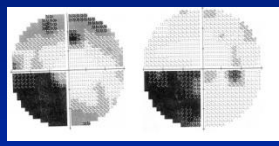
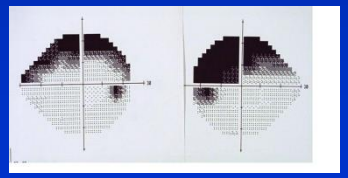
Sometimes retinal pathology:

- 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.



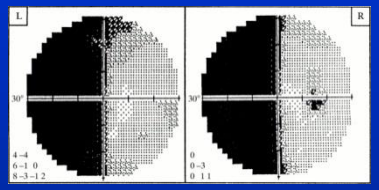
Superior loss from: Eyelid Ptosis, UL Dermatochalasis,
Deep Set Eyen in Orbit, Frontal Bossing
can give appearance of horizontal midline respect



VF Defects respecting the *Vertical Midline*

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

Homonymous Hemianopsia

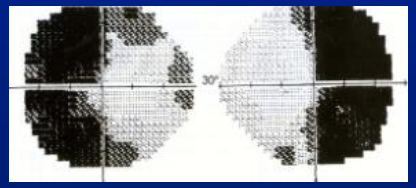


CVA, Tumor, Demyelination posterior to chiasm
(See separate list)

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

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

Bitemporal Hemianopsia



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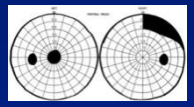
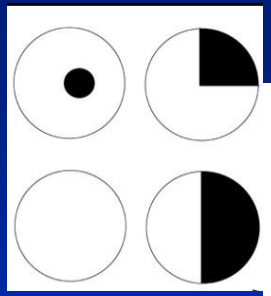
Chiasmal, Sellar and Parasellar lesions
(See separate list)

Junctional Scotoma

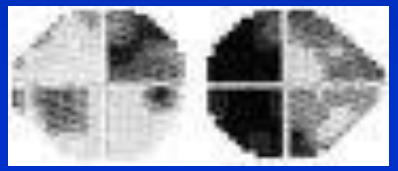


Specific lesion at junction of optic nerve with chiasm (Von- Willebrand's Knee)



Isolated Temporal Hemifield Defect

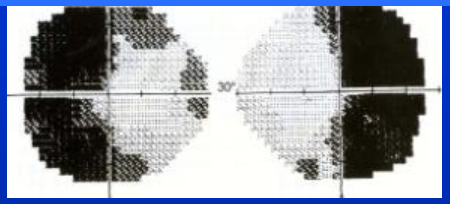
Junctional Scotoma of Traquair
Tilted Disc Syndrome
Unappreciated loss of central vision in contralateral eye of Junctional Scotoma



Other: Nasal Hemianopsia(s) Possible

Visual Field Defects that Respect the Vertical Midline

Bitemporal Hemianopsia



Chiasmal or Supra- or Para- Sellar Lesions:

- Chiasmal Tumor – e.g. Glioma
- Pituitary Tumor, Pituitary Apoplexy
- Other Parasellar Tumors – meningioma, craniopharygioma, Rathke's (pars intermedia) cyst, etc.
- Demyelination (MS) - "Chiasmitis" (seen in ONTT)
- Aneurysms – ICA, branches off circle of Willis
- Chiasmal Trauma from Head Injury
- Trans-sphenoidal Basal Encephalocele
- Other: Meningitis, Sarcoidosis, Ischemia?
- Toxicities: Ethambutol

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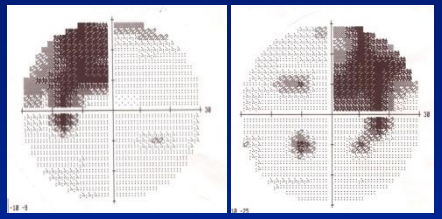
Incomplete Bitemporal Defects

- Any of above can produce this picture
- Dermatochalasis with Lateral Hooding
- ***Tilted or Anomalous Discs*** can produce temporal defects
- Nasal Staphyloma(s)

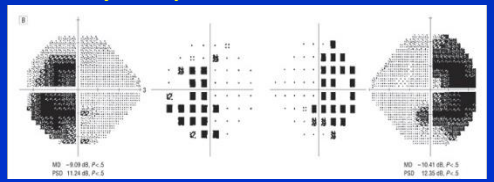


Dermatochalasis with Lateral Hooding

Centrocecal Scotomas

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

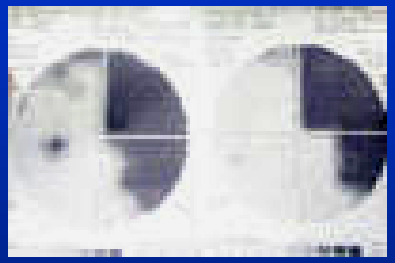
e.g. Toxicities (Ethambutol, Chloroquine), Deficiencies, Hereditary Optic Neuropathies



VF Defects respecting the *Vertical Midline*

Homonymous Hemianopsia

Complete or Incomplete



Lesions of

- optic tracts (bowtie atrophy)
- LGN
- temporal, parietal or occipital lobes

From: CVA (thrombosis, embolism, vasculitis)
Tumor (Compression or Infiltration)
MS or other Demyelinating Process
Trauma
Vascular Malformation

If there is no apparent Lesions on Neuroimaging, Consider:

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

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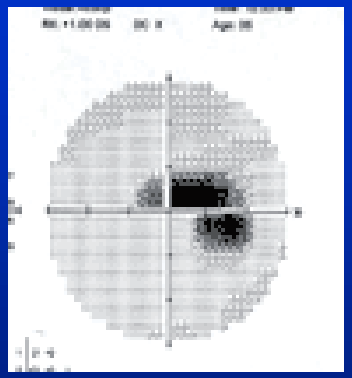
Other Visual Field Defects

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



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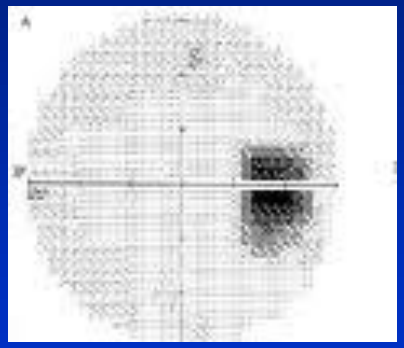
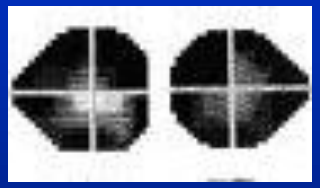
Enlarged Blind Spot (Cecal Scotoma)

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



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)

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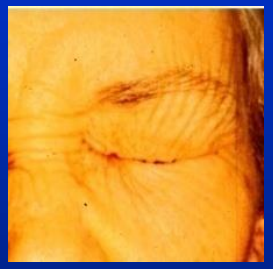
Eyelids and Orbit

Blepharospasm

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 Syndorme

Excessive Blinking

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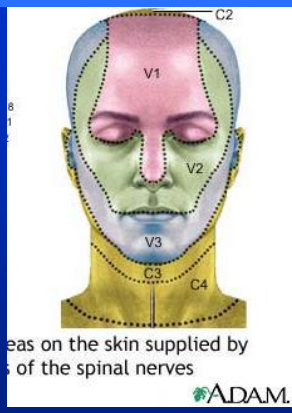
May Need to Differentiate from Just a Problem of Opening Eyelid(s)

- Apraxia of Eyelid Opening
  Associated with BEB, PSNP, Parkinson's, Huntington's, CNS Lesion - Frontal (and Parietal?) Lobe, Brainstem, Thalamus
- Dry Eye / Blepharitis / RES - Lids stuck to each other or cornea
- Ptosis

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



- Damage to Branches of 5th Cranial Nerve
  e.g. Orbital Floor Fracture – damage to V2
  Shingles (Zoster) - most commonly V1 distribution

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

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

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

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

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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

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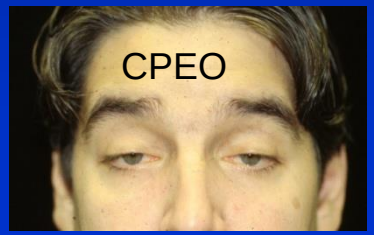
Orbital Disease

- 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

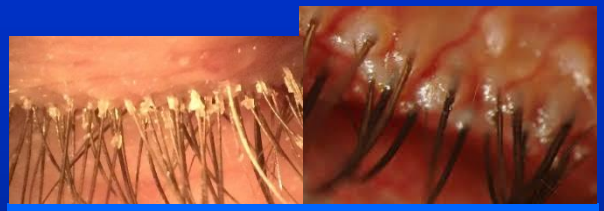
Eyelashes and Eyelid Margin

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

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* Comprehensive Listing : Survey 2006; 51:550

Eyelid Malpositions
Entropion and Trichiasis

Lower Lid Entropion and Trichiasis

- Involutional (Senile) – can have spastic (orbicularis) component
- Acute Spastic Entropion – after trauma or surgery
- Cicatricial (see below)
- Congenital / Developmental – e.g. Epiblepharon
- Distichiasis – abnormal lashes growing from posterior lid margin (meibomian orifices)
  could be hereditary or from inflammatory process (see below)

Upper Lid Entropion and Trichiasis

- Mechanical – excessive Dermatochalasis
- Cicatricial (see below)
- Distichiasis

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

- Previous Trauma or Surgery at or near eyelid margin
- Chemical Burn
- HZO
- Chronic Blepharo-conjunctivitis - e.g. Acne Rosacea
- Trachoma
- Stevens-Johnson Syndrome, Ocular Cicatricial Pemphigoid

Sometimes Orbital Disease can present with eyelid malpositions

Eyelid Malpositions - Ectropion

Lower Lid Ectropion

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

Cicatricial (below)

Combination of both above

Paralytic – 7th nerve palsy, MG

Mechanical – Tumor or Big Festoons

Congenital – Ichthyosis; Euryblepharon – excess horizontal skin

Medications: TNF-α inhibitors

Upper Lid Ectropion

Cicatricial Processes (below)

Congenital – e.g. Ichthyosis

Floppy Eyelid Syndrome – Horizontal Laxity – not true ectropion

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Cicatricial Changes (of anterior lamella)

Trauma to Eyelids and Face

Burns- Thermal and Chemical

Sun Damage, Carcinoma

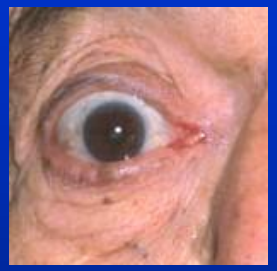
Previous Eyelid and Adnexal Surgery

Chronic Inflammation: Rosacea, Atopic Dermatitis, HZO, Infection

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

Eyelid Retraction

Graves Ophthalmopathy- #1 – unilateral or bilateral



Other Causes of Hyperthyroidism
Other Orbital Inflammatory or Neoplastic Conditions
– Orbital Pseudotumor, FB, Granulomatous Inflammation, Neoplasm

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

Trauma / Post-Operative
Entrapped Inferior Rectus
Vertical Rectus Muscle Recession Surgery
S/P Eyelid or Conjunctival Surgery

Neurologic
- e.g. Dorsal midbrain syndrome (Collier's sign) , aberrant regeneration of the 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

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Lagophthalmos

Inability to Close Eyelids

Neurological

Seventh Nerve Palsy

Cicatricial (Scarring)

Trauma

Burns

Surgery

Blepharoplasty, Ptosis Surgery

Tumor resection

Orbital Condition

Proptosis: Graves Ophthalmopathy, etc. (see list)

Orbital Inflammatory or Neoplastic Processes

Myogenic – MG, Muscular Dystrophies, CPEO

Botulinum Injections

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

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Seventh Nerve Palsy

Hemifacial Paralysis with Lagophthalmos

- Motor Strip Lesion (Upper Motor Neuron) → Contralateral Lower Face Paralysis

- Peripheral Nerve Palsy – Ipsalateral 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

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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

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Nodular - Commonly at Lid Margin

Intradermal Nevus
BCCA
Hair Follicle Tumor

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

Vascular

Hemangioma
Cherry Angioma – Bright red
Varix
Other: Kaposi's Sarcoma, Pyogenic Granuloma

Crater / Ulcerated

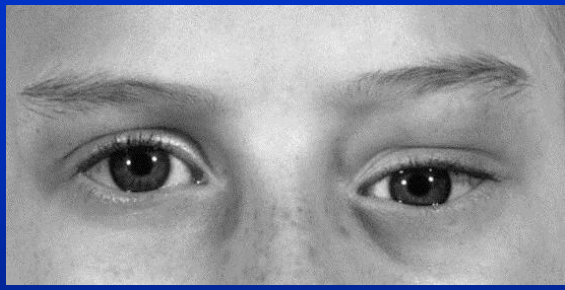
Carcinomas (BCCA, SCCA, etc)
Keratoacanthoma
Moluscum Contagiosum

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

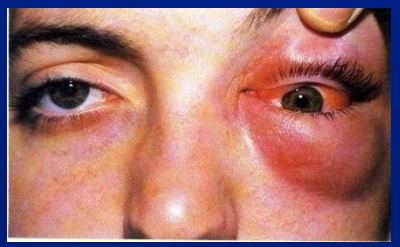
Signs Suggesting Orbital Disease

- Proptosis, and other
  Globe Displacements

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



Left hypoglobus



Lid deformity in orbital NF

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Eyelid Edema

Inflammatory Appearance (red, warm, etc.)

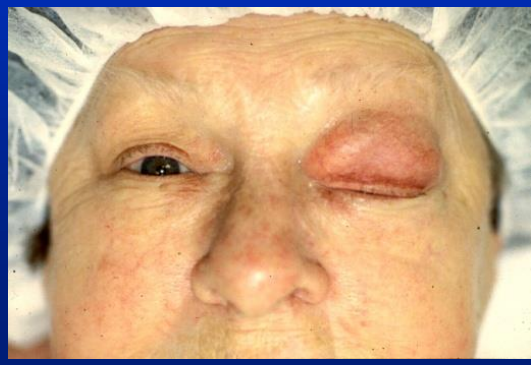
First Consider Underlying Orbital Disease

- Orbital Cellulitis, Pseudotumor, Wegener's
- Graves Ophthalmopathy, Orbital Varix
- Orbital Tumors that can mimic inflammatory process: Lacrimal Gland CA, Lymphoma, Lymphangioma, etc.
- Lacrimal Gland – Dacryoadenitis or tumor
- Sinus Mucocele

Preseptal Cellulitis

– also think of early -HSV, HZO, or erysipelas (rapid strep),
Periorbital necrotizing fasciitis (b-hemolytic strep, staph A., pseudomonas)

Dacryocystitis / Dacryocystocele
Blepharitis
Contact Dermatitis – e.g. Neomycin, Gentamicin, ***Glaucoma Drops-*** Chronic Use
Urticaria / Angioedema
Conjunctivitis with contiguous lid edema
Insect Bite
Lid Tumors: Hordeolum / Chalazion, CA, ***Cutaneous Lymphoma***
Melkersson-Rosenthal Syndrome – (Granulomatous inflammation)



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Without Inflammatory Appearance, consider above but also…

Allergic Eyelid Edema
Hormonal Shifts
Systemic Disorder – Cardiac, Renal, Hepatic, Thyroid with edema
Graves Ophthalmopathy – can just have lid edema w/o inflammatory appearance
Lymphedema after trauma, surgery to lids or orbit (e.g. lymphatics in lateral canthus)
Traumatic Leak of CSF into upper eyelid (JAMA Oph 2014;312:1485)
Blepharochalasis

Not True Edema but might mimic it:

Dermatochalasis

Hidden Eyelid or Sub-Conjunctival Mass

Prolapsed Orbital Fat

Conjunctival Hemorrhage

1 Spontaneous

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

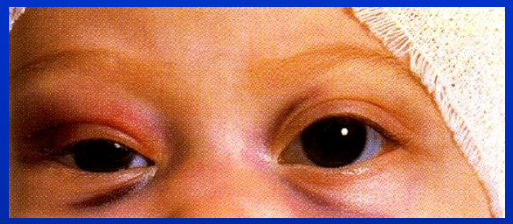
Valsalva Maneuver (sudden venous congestion)

Sometimes Orbital Hemorrhage
- Trauma, Retrobulbar Injection
- Tumor: Neuroblastoma , Rhabdomyosarcoma, Leukemia, Lymphangioma

Acute viral/ bacterial conjunctivitis

Systemic - Clotting disorder, febrile illness, acute HTN

Peri-Orbital Ecchymosis



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

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

Bad Sub-Conjunctival Hemorrhage can dissect into eyelids

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

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

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Proptosis

Forward Displacement of the Eye

1. **Graves Orbitopathy** (#1 Cause)
2. Orbital Cellulitis
3. Orbital Pseudotumor – including Posterior Scleritis

4. Other Inflammatory - Wegener's, Sarcoidosis, see also Orbital Inflammation

5. Orbital Tumors, Sinus Tumors
6. Orbital Hemorrhage- Trauma, Lymphangioma
   Subperiosteal Hemorrhage
7. Orbital Trauma and Compartment Syndrome
8. Vascular Abnormality –e.g. Varix /Venous Malformations (Congenital)*, Carotid Cavernous Fistula
9. Bony Orbital Malformation – e.g. congenital
10. Meningo-encephalocele
11. Sinus Mucoceles
12. Orbital Apex and Cavernous Sinus – Tumor, Inflammation, Vascular

Pseudo-Proptosis

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

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Age Category	Orbital Tumors to Consider
1. Children	Rhabdomyosarcoma Neuroblastoma Lymphangioma Capillary Hemangioma Glioma of Optic Nerve Dermoid Cyst Neurofibromas (NF1 and NF2)
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

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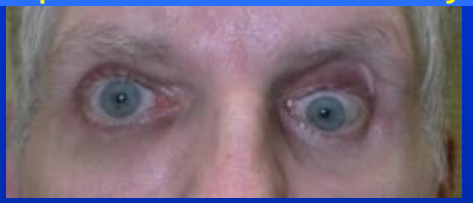
Enophthalmos

Posteriorly Displaced Globe often with superior sulcus deformity



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

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

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Signs of Orbital Inflammation

Proptosis, Chemosis, Eyelid Edema, etc.

1. Orbital Cellulitis
2. Orbital Pseudotumor – Dacryoadenitis, Myositis, Diffuse, Posterior Scleritis, IgG4-RD*
3. Graves Ophthalmopathy
4. Orbital Lymphoma
5. Reactive Inflammation
   Foreign Body, Ruptured Dermoid Cyst, Adjacent Sinus Inflammation
6. Trauma
7. Systemic Disease
   Wegener's Granulomatosis, Sjogrens , TB, Sarcoidosis, Syphilis, IgG4-ROD
8. Medications: e.g. Biphosphonates
9. Vascular etiologies
   Vasculitis (e.g. GCA), **CC Fistula**, Cavernous Sinus or Superior Orbital Vein Thrombosis (Sepsis, Coagulopathies, etc)
10. Inflammatory Mimicking Orbital Tumors
    Rhabdomyosarcoma, Lacrimal Gland Carcinoma, Lymphangioma
11. Orbital Hemorrhage

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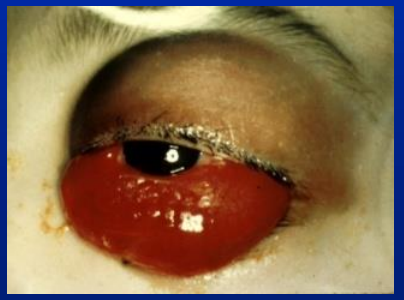


Need to R/O
Orbital Cellulitis / Malignancy
Imaging of the Orbits

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

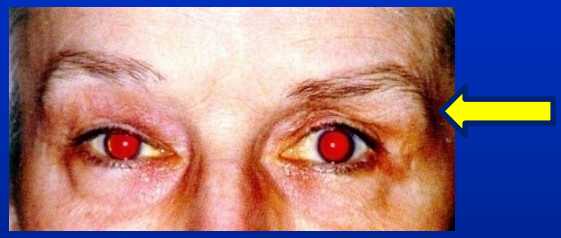
Visible and Palpable Masses in and around the Orbit

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



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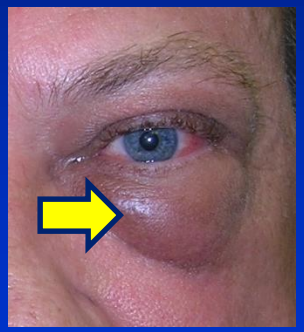
Masses in Nasal and Superior Nasal Quadrant

- Dermoid Cysts
- Meningocele and Encephalocele
- Sinus Mucocele
- Lacrimal Sac: Tumor, Dacryocystocele, Dacryocystitis
- Neurofibromas, Capillary Hemangiomas
- Bulging Nasal Fat Pad

Apparent Mass under Lower Eyelid

Orbital Tumors yes, but also need to consider:
Festoons - Prolapsed Orbital Fat and and/or Redundant folds of skin
Inflammatory Disease – Orbital Cellulitis, Graves Disease, Chalazion / Abscess
Lymphedema, Allergic Reactions
Lymphoproliferative Disorders - Lymphoma, Orbital Pseudotumor, etc.
Allergic “Shiners” edema – often responsive to treatment



Orbital and Facial Distortions.

Hypertelorism

– increased orbital separation and increased interpupillary distance

seen in congenital craniofacial anomalies such as
Crouzon's Syndrome

In Contrast

Telecanthus

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

seen with: BPES
Other Congenital Facial abnormalities
Down's Syndrome
Facial Trauma – e.g. avulsion of MCT

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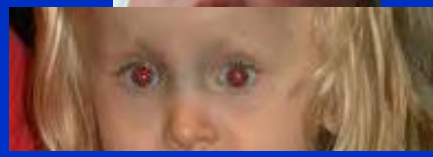
Hypotelorism

– decreased orbital separation and decreased interpupillary distance

seen with: FAS – Fetal Alcohol Syndrome
Congenital Anomalies:
e.g. holoprosencenphaly



Distortions of the Globe

Small, Misshapen or apparently Absent Globe

– can be interpreted as an orbital problem

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

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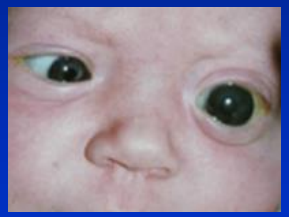
Other associated congenital defects:

Goldenhar (Oculoauriculovertebral) syndrome, Trisomy 13-15

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

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

Large, Buphthalmic Eye



Congenital and Juvenile Glaucoma

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

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

Systemic: Lowe's Syndrome (oculocerebrorenal 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

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Motility and Alignment Problems

A Spectrum of Causes

1. Neurologic

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

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

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

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

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

Neurodegenerative Disorders – Friedreich Ataxia, Spino-cerebellar Ataxia

2. Myogenic

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

Local – Muscle Damage and/ or Fibrosis, Previous Surgery

3. Orbital Disease

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

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

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

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

e.g. Congenital / Infantile Esotropia, Intermittent Exotropia,
IO Overaction, Accommodative Esotropia, Monofixation Syndrome
Risk population for Common Comitant Strabismus not associated with Neurologic Disorders*

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Abduction Deficit

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

Looking Left

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*

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Internuclear Ophthalmoplegia

Adduction deficit

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

Looking Right

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

And Then:

Third Nerve Palsy – but rare to have isolated MR palsy

Congenital Fibrosis of EOM Type 2 (Ptosis and loss of adduction, elevation and depression can mimic 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

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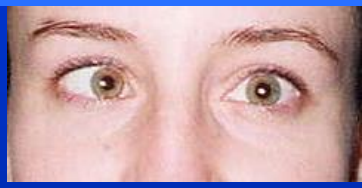
Acquired Esotropia



1. Sixth Nerve Palsy – maybe early or partial
   Think about: Microvascular, Head Trauma, Post Viral, MS, Tumor, etc.
   (see abduction deficit list)

2. Divergence Insufficiency (ET at distance > ET at near)
   Usually Benign Condition, unless other neurologic symptoms or signs present, then need further investigation
   Older Patients Consider: Microvascular, CVA, Progressive Supranuclear Palsy, Cerebellar or Brainstem Lesions
   Younger Patients: Pseudotumor Cerebri, Arnold Chiari Malformation, Meningitis

3. Graves Ophthalmopathy (tight MR muscle(s))
4. Myasthenia Gravis
5. Sensory Visual Loss
6. Decompensated Esodeviation, e.g. Monofixation Syndrome
   (factors: Hydrocephalus, shunt failure, trauma, …)
7. Acquired Comitant ET (neurologically isolated, stable) - give full Cycloplegic Rx and do ***prism adaptation***

8. After Neurologic Insult – encephalitis, meningitis, trauma (not necessarily 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

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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

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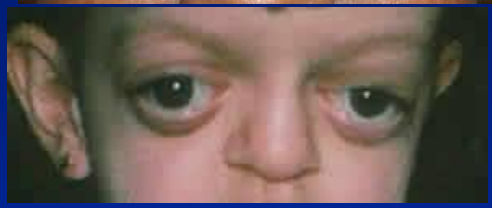
Pseudo –Strabismus

Apparent Horizontal Strabismus

1. Pseudoesotropia
   - Epicanthal folds, Hypotelorism, Telecanthus
   - Apparent Esotropia - Negative angle kappa*

Normal – mild positive angle kappa

2. Pseudoextropia
   - Hypertelorism
   - Apparent Exotropia – Positive Angle Kappa*



How to differentiate?
Angle Kappa from Strabismus: Cover Testing vs Hirschberg test

Detecting Hyper and Hypotelorism: Interpupillary distance
Hirschberg and/or Cover Testing – can then be used to assess whether any strabismus is present

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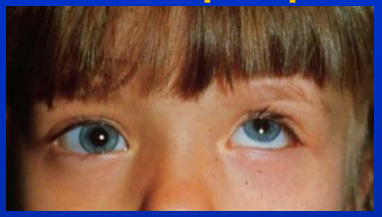
**Angle kappa* is the angle between the visual axis and the anatomical pupillary axis of the eye

Hypertropia / Hypotropia and Vertical Diplopia



Fourth Cranial Nerve (SO) Palsy
Graves Ophthalmopathy (IR or other vertical muscle involvement)
Orbital Trauma / Fracture
Post-op Eye Surgery –e.g. post CE (Local Injection) or Scleral Buckle
Myasthenia Gravis
Skew Deviation (comitant or non-comitant vertical deviation often associated with vestibular / brainstem / thalamic problems – e.g. balance problems / ataxia). Incomitant Skew Deviations could include the Including Ocular Tilt reaction (**OTR***) or mimic of IR palsy (e.g. RHT worse on Right gaze)

Age Related degeneration of Orbital Pulley System for EOMs → Cyclo-Vertical deviations
Brown's Syndrome
Inferior Oblique Over-Action
Third Nerve Palsy or aberrant regeneration
Orbital Tumors
Myositis, Orbital Pseudotumor
Glasses – Anisometropia, Optical Centers off, Induced Prism in eccentric gaze, etc.
Monocular Elevation Paresis (Double Elevator Palsy)
Congenital Absence of IR or Fibrosis
SO Myokymia
Sixth Nerve Palsy – sometimes has an associated vertical misalignment
GCA – can produce isolated EOM palsies
Other: Hemifield Slip, Dissociated Vertical Deviation (DVD), Wernicke's, Guillain-Barre, Central Inhibition of Fusional Reserves
Inferior Oblique Palsy – isolated IO palsy not likely, so consider: OTR, Orbital Tumor, Brown's Syndrome, MG, and IO damage after a LL Blepharoplasty

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

Elevation Deficit

Graves Ophthalmopathy (e.g. IR fibrosis)

Orbital Floor Fracture with entrapment

Third Nerve Palsy (Superior Division)

Double Elevator Palsy (in abduction and adduction)*

Brown's Syndrome (in adduction)

CFEOM Type 1

Myositis (e.g. IR)

Myasthenia Gravis

Midbrain – Diencephalic Lesion

Isolated Palsy of SR or IO

Previous SR Recession

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

Upgaze Deficit

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Old Age

Parinaud's Syndrome

Thalamic Infarction

Progressive Supranuclear Palsy (PSP)

Hydrocephalus

Hemispheric Infarctions

Metabolic – Niemann- Pick Disease

Myasthenia Gravis

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

Depression Deficit

SR restriction
- Graves
- Fibrosis
- Myositis

IR Ischemia

3rd CNP – Inferior Division

Myasthenia Gravis

Midbrain and Diencephalic Lesions

Downgaze Deficit

Progressive Supranuclear Palsy

Parkinson– Like Syndromes

Midbrain Lesions

Myasthenia Gravis

Head Trauma

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

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Convergence and Divergence

Convergence Insufficiency

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

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

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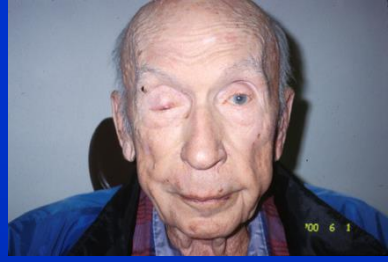
Divergence Insufficiency

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

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

Consider also:
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

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

- Trauma- edema and or orbital hemorrhage
- Peri-neural Spread of Cutaneous Carcinoma
- Ischemia – Saturday Night Ophthalmoplegia, Orbital Infarction Syndrome*
- Graves Ophthalmopathy – multiple muscles
- High ICP, DM – could potentially cause multiple CNP (e.g. 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

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Head / Brain Trauma

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

Progressive Supranuclear Palsy

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

Paraneoplastic Syndromes

Wernicke's Encephalopathy (Alcohol, Thiamine, Emergency)

Toxins: Organophosphates

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

Graves Ophthalmopathy

IOIS, Orbital Fibrosis Syndrome

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

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

Head Turns or Tilts
Torticollis

1. Orthopedic / Muscular – most common causes

   e.g. Congenital muscular torticollis: damaged sternocleidomastoid muscle

   Cervical spine – damage / deformity : Fracture, TB, Scoliosis

   Tonsillitis, retropharyngeal abscess, drugs – neuroleptics, dystonias

2. Neurologic – not common

3. Ocular – not uncommon

   - Incomitant Strabismus:

     CNP: 4th (Superior Oblique) or 6th (Abducens)

     Duane Syndrome, Browns Syndrome

     A and V Patterns

     DVD (Dissociated Vertical Deviation)

   - Nystagmus

     Congenital Nystagmus with Null Point

     Spasmus Nutans

   - Ptosis - often chin up head position
   - Astigmatism

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Aid in Diagnosis:

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

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

Causes of Nystagmus

1. Idiopathic / Congenital – Typical Features – Conjugate, No Oscillopsia, Dampens at near and with Convergence, Null Point, Latent Nystagmus

2. Sensory Visual Loss – e.g. Deprivational Amblyopia, Optic Nerve, Retinal Disease, Albinism.
   Need thorough 8 Point eye exam. Sometimes further testing, e.g. ERG.

3. Acquired later in Life – **Often Associated with Symptoms** – e.g. Oscillopsia, blurring in eccentric gaze

   CNS lesion – CVA, Tumor, MS (Most commonly in brainstem or cerebellum, less likely in parasellar region and cerebral hemispheres)

   Spasmus Nutans vs. Tumor of Anterior Visual pathways

   CNS malformation – e.g. Chiari Malformation

   CNS inflammation – post viral (e.g. cerebellum) , post immunization, encephalitis, Lupus, MS

   CNS: Associated with Seizure Activity in Cerebrum (Epileptic Nystagmus)

   Associated with antibodies - e.g. antiganglioside Abs (anti- GD1b, GMI, GQ1b) – seen in GBS, MS

   CNS Degenerative Disorders (e.g. Spino-cerebellar Ataxias)

   Vestibular - e.g. Labyrinthitis

   Metabolic – Mg and Vitamin B12, B1 (Thiamine) deficiencies

   Toxicity – Phenytoin, Lithium, alcoholism, street drugs, glue sniffing (toluene)

   Paraneoplastic Syndrome – associated with e.g. Lung CA and antibodies anti - Hu, Ri, Yo, Tr

   Multiple Different Congenital Syndromes

   Latent Nystagmus – manifesting later after some (monocular) loss of vision

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Acquired Nystagmus

Long DDX but, *Think first of:*

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

10. Brainstem or Cerebellar Lesions or Malformations

(e.g. Arnold Chiari, brainstem tumor or infarct) or Parasellar Lesions, rarely cerebral hemispheric lesions.

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

Downbeat Nystagmus

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

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

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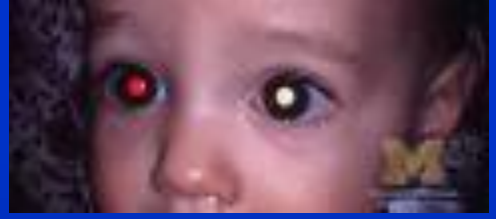
Exercise induced? JNO 2002;22:127

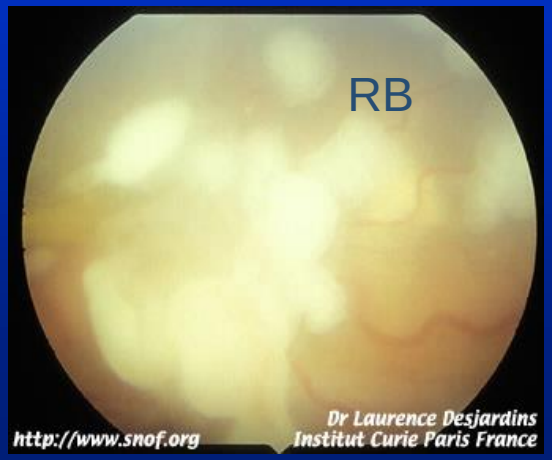
Idiopathic – 20% or more cases, assess vasculopathic risk factors - JNO 2001; 21:39-41

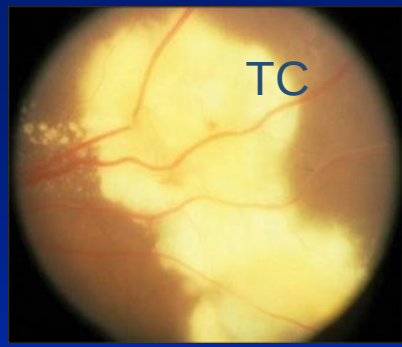
Pupils

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

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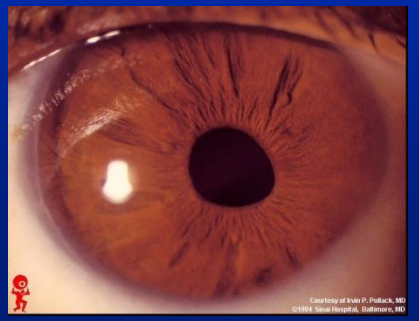
Aids to help diagnose RB

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

Corectopia

Irregular Pupil Shape and / or Abnormal Location

1. Congenital and Developmental
   - Congenital Iris Coloboma, Ectropion Uvea
   - Ectopia Lentis et pupillae
   - *Neurofibromatosis – nodules, ectropion uvea*
   - Anterior Dysgenesis (e.g. Axenfeld- Rieger, Peter's Anomaly)
   - Iridocorneal Endothelial Syndrome (ICE Syndrome) e.g. essential iris atrophy
   - Posterior Polymorphous Corneal Dystrophy
2. Inflammatory (e.g. uveitis – posterior synechiae; HZO-segmental atrophy)
3. Neovascular – NVI (see list)
4. Neoplastic (e.g. iris, ciliary body melanoma)



5. Damage or Distortion From:
   - Trauma (e.g. iris dialysis, iris prolapse, scarring, epithelial down-growth)
   - Surgery (e.g. Iridectomy, CE)
     - Peaked pupil from Vitreous Strand, Blood or synechiae in the angle
   - Angle Closure attacks
   - IOL capture, dislocation
   - Iris Atrophy – age, diabetes, essential, ischemia

6. Neurological - <u>Permanent or Transient Irregular Pupils</u>
   - Midbrain lesion - MS, Ischemia, Tertiary Syphilis (Argyll Robertson)

<u>Transient Irregular Pupil in what appear to be healthy patients</u>
- "Tadpole Pupil"
- Migraine patient
- Early Horner's
- Young Healthy Women

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Poor Pupil Mobility

Poor Reactions to Light, Dark or other stimuli

- **Age**
- **Diabetes Mellitus**
- **Damaged or Paralyzed Pupil**

  Uveitis, Angle Closure, Surgery, Trauma

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

- **Afferent or Efferent Input Defect**

  Damage to any part of Pupillary Light Reflex Pathways

  RAPD

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

- **Systemic**

  Medications or Toxins

  Mydriatic - Anticholinergics , Adrenergics (Methamphetamine),

  Miotic - Narcotics

  Systemic / Neuro-Muscular Conditions

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

  Myotonic Dystrophy – miotic

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

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Anisocoria (Unequal pupils)

1. Physiologic - 20% of population

2. CNS and Efferent Nerve Input Problem

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

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

   Parasympathetic Defect

   Third Cranial Nerve Palsy, Aberrant Regeneration

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

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

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

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

4. Asymmetric Damage or Atrophy of the Iris and Pupil

   Eye Trauma

   Surgical / Trauma

   Inflammatory / Uveitis

   High IOP (e.g. paralytic mydriasis)

   Pigmentary Dispersion Syndrome

   Other Causes of Iris Atrophy: Asymmetric Atrophy of Irides

   Old Age, Essential Iris Atrophy – progressive,

   Ischemia (trauma, past high IOP, Hb SC disease, etc.)

   Syphilis, Diabetes, Idiopathic?

5. Other:

   Anisometropia (e.g. Unilateral High Myopia)

   Amaurotic (Blind) Eye

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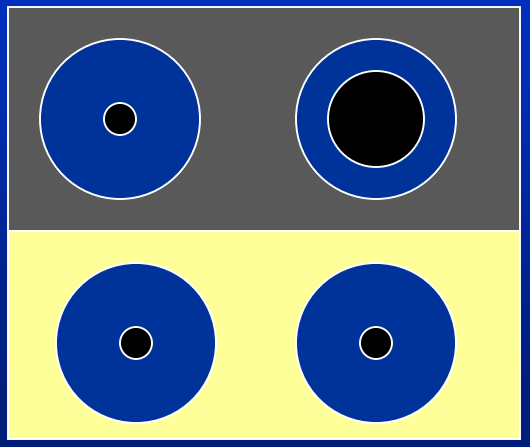
Usually one abnormally small (miotic) pupil or one abnormally large (mydriatic) pupil

Anisocoria
Which Pupil is Abnormal?

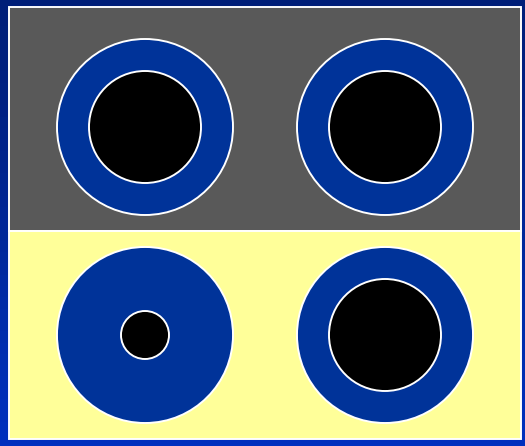
Abnormally Miotic Pupil
(Anisocoria worse in the dark – poor dilation in dark)

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



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Dilated Pupil(s)

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

- Third Cranial Nerve Palsy

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

  Basal Meningitis

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

  Aneurysm - e.g. posterior communicating artery

  Ischemia, Parasellar Mass or Inflammation

- Midbrain Damage

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

- Ciliary Ganglion Damage or Damage to SPCNs

  - CG: Post-viral (e.g. HZO), Orbital Tumor, Trauma, Idiopathic (Adie's)
  - SPCN's – injections, ocular surgery – e.g. SB

- Iris Damage / Atrophy

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

- Pharmacologic

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

  Adrenergic: Phenylephrine

- Pourfour du Petit syndrome – opposite of Horner's – stimulated sympathetics
- Sometimes a blind or nearly blind eye (e.g. Severe Optic Neuropathy)

Transiently Dilated Pupil

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

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Bilateral Dilated Pupils- Think of:

Any of above if Bilateral

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

Dorsal Midbrain (Tectal) Lesions, Severe brain anoxia , Bilateral Iris damage or atrophy, Bilateral Adie's Pupil, Bilateral 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

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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)

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Light Near Dissociation (LND)

Poor light reaction, but good constriction response to near

Can be one or both pupils

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

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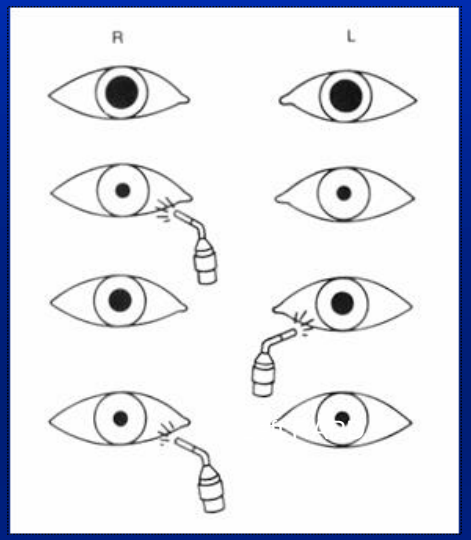
RAPD

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

More Common

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



Much Less Common

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

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Bilateral Miosis or Mydriasis

1. *Bilateral* Miosis

Systemic Medications: e.g. Narcotics (e.g. Morphine),
Cholinesterase Inhibitors (Snake venom, biochemical warfare, pesticides, MG treatments)
– early effects excessive salivation and tearing

Topical Medications: e.g. Pilocarpine

Pontine Lesions – pinpoint pupils

Damage to Sympathetic Pathways in Hypothalamus

Diencephalon Lesions – small but reactive

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

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2. *Bilateral* Mydriasis

Systemic Medications: anticholinergic (atropine)
sympathomimetics (amphetamines, cocaine, etc.)

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

Familial Bilateral Congenital Mydriasis

Tectal and Midbrain Lesions – inflammatory , neoplastic

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

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

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

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

Anterior Segment and IOP

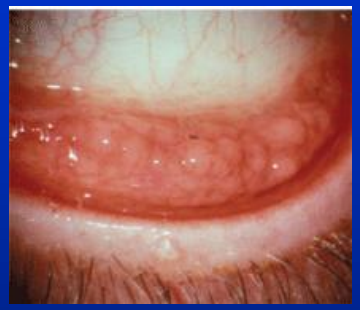
Conjunctival Bumps

Follicles

Think of Viral Infections, especially:

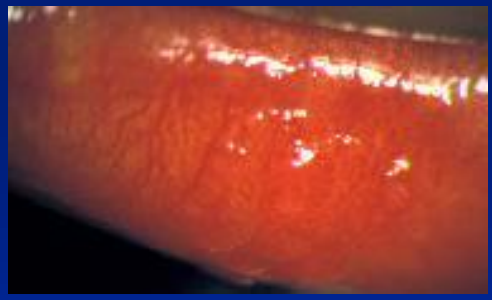
Adenoviruses – more often bilateral, acute, associated with URI
Herpes simplex (primary infection)
Molluscum contagiosum
Acute Hemorrhagic – e.g. enterovirus
Recent URI related

Chlamydia – AIC = Adult Inclusion Conjunctivitis(unilateral , chronic), *Trachoma*
Reactions to topical ophthalmic medication,
e.g. atropine, alpha agonists, antivirals, miotics
Other: Parinauds Ocular Glandular Syndrome (e.g. Cat Scratch, Tularemia, Mycobacterial Infections)
Lyme, Newcastle, sometimes some bacterial: Strep, Neisirria, Moraxella
Children – benign lymphoid folliculosis
Conjunctival Lymphoma



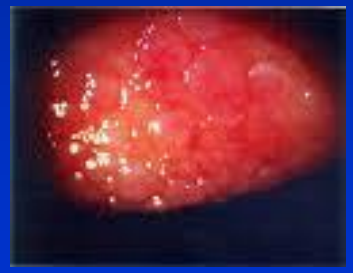
Conjunctival Papillary Reaction

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



Giant Papillary Reaction

Contact Lens Wear (Giant Papillary Conjunctivitis)
Atopic Keratoconjunctivitis
Vernal Keratoconjunctivitis – Palpebral and Limbal
FB- ***Ocular Prosthesis*** or Suture End



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R/O Orbital Disease

Inflammatory – Orbital Cellulitis, Contiguous Sinus Inflammation

Orbital Pseudotumor

Graves Ophthalmopathy

Orbital Tumor, Surgery (Post op)

Orbital Trauma, Fracture, Open Globe

Blocked Orbital Lymphatics – surgery, radiation

Chemosis

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

Nearby Corneal , Eyelid, Sinus, Meningeal or Lacrimal inflammation

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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

Injected, Congested or Prominent Vessels on the Globe

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

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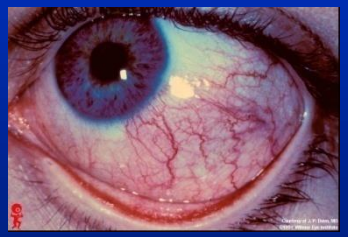
Conjunctivitis and other External Problems

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

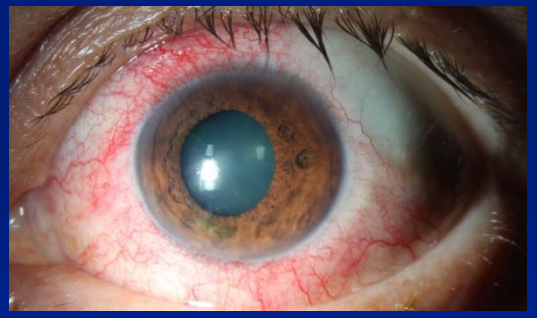


Ciliary Flush (Circumcorneal Congestion)

- 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

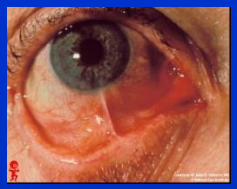
- chronic topical vasodilators ("red out drops"), chronic topical anesthetics
- sympathetic irritation, trauma, FB, insect bite
- local tumor with feeder vessels
- local vascular malformations or vascular tumors

Vascular Obstruction or Malformation

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

Venous Malformation – JAMA Ophthalmology

Symblepharon and Conjunctival Scarring



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- 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.

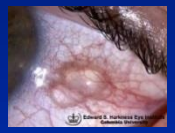
Epithelial malignancy – OSSN: Conjunctival Intraepithelial Neoplasia (CIN), Invasive Squamous cell CA, Basal Cell or Sebaceous Cell CA invading Conjunctiva, Amelanotic melanoma

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 Melanosis- can undergo malignant transformation), Conjunctival Melanoma,
- some OSSN can have abnormal pigmentation
- Deposits: cosmetics, FB's, Hemosiderin after hemorrhage, Addison's, Silver, Systemic Tetracyclines

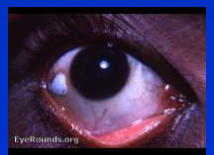
Sub-epithelial Tumors - Lymphoproliferative (e.g. Lymphoma, Lymphoid Hyperplasia, Multiple Myeloma – "Salmon Patch" Hemangiomas, Lymphangiomas, Karposi's Sarcoma, Mxyoma, fibroma, mets, neuroma, amyloid

Cysts - epithelial, ductal, inclusion



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

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



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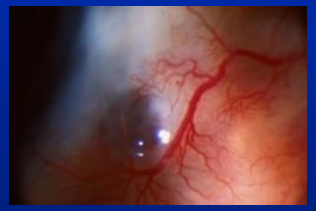
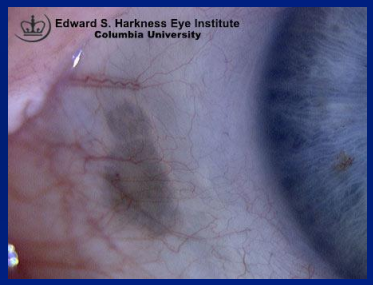
Spots on the "White of the Eye" Scleral Lesions:

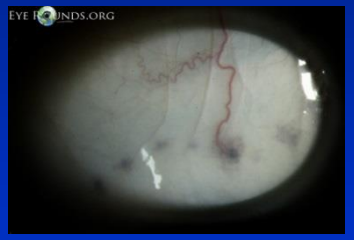
- Scleral Dellen – depression or dimple, loss of epithelium over defect in sclera. Seen after surgery (e.g. Pterygium, EOM surgery), Cautery Trauma, medication toxicity, autoimmune disorders



- 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)

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Scleral Thinning, Episcleritis and Scleritis

Scleral Thinning:

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

Episcleritis

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

Causes include:

Idiopathic

Related to autoimmune disease →

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

Scleritis

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

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

Consider:

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

Diseases associated with Episcleritis and Scleritis:

Rheumatoid Arthritis

Herpes Viruses (e.g. Simplex, Zoster)

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

Relapsing Polychondritis (check ears, nose)

Orbital Pseudotumor

Wegener's Granulomatosis

Sarcoidosis

Medications: Fosamax, Aredia

Lupus (SLE)

Inflammatory Bowel Disease

Spondyloarthropathies

Possible Testing:

ANCA Panel ,

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

Urinalysis, CXR, ACE, HLA testing

Scleral Biopsy or Trial of Oral Acyclovir

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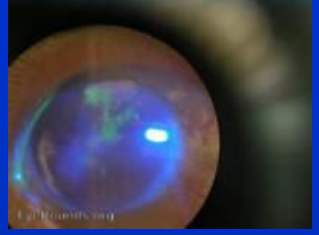

Corneal Fluorescein Staining or Pooling

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

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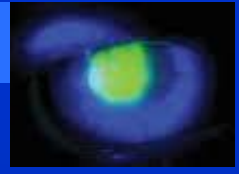
Acute Peripheral Corneal Thinning:

Physical or Chemical Trauma – associated with swelling of conjunctiva / episclera at limbus
Gonococcal, Streptococcal Infections
Mooren's Ulcer (associated with Hepatitis C)
Connective Tissue / Autoimmune Disorders – Rheumatoid Arthritis

Dellen

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

Corneal Epithelial Defects- Chronic or Non Healing



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

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

Corneal Haze/ Opacification
Loss of Clarity

Corneal Edema
– stromal or epithelial - see following lists

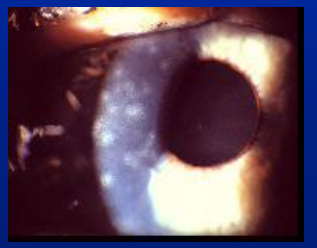
Corneal Infiltrates

Infectious:

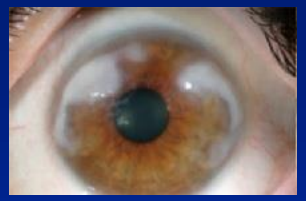
HSV, HZO, Bacterial, Fungal, TB, Syphilis, Acanthamoeba

Non-Infectious:

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



Interstitial Keratitis (IK) - Stromal vessels and haze/ necrosis
HSV, Congenital Syphilis, Cogan's (IK + auditory and vestibular problems), TB, Sarcoidosis



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

Corneal Degenerations: Salzmann's Nodular Degeneration* , Band Keratopathy
Corneal Dystrophies
Corneal Deposits, Crystals
Corneal Whorls (Verticillata): Fabry Disease, Medications: Amiodarone,
Corneal Surface Growths – Pterygium, Squamous Neoplasia,
S/P Laser Refractive Surgery – e.g. PRK

Congenital / Developmental Problem
e.g. Anterior Segment Dysgenesis, Amyloidosis, Congenital Glaucoma and Secondary Infantile Glaucomas
"STUMPED": Sclerocornea, Tears in Descemet's (Congenital Glaucoma), Ulcers (e.g. intrauterine – viral), Metabolic (e.g. Mucopolysaccharidoses) , Peter's Anomaly, Edema (Congenital Hereditary Endothelial Dystrophy), Dermoid

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Causes of *Acute* Corneal Edema

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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

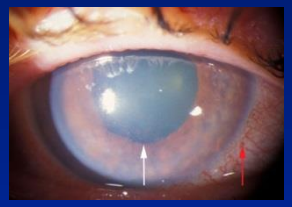
Anterior Segment Ischemia

Other

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

Keratitis ⇧

Angle closure ⇩



"Steamy appearance"

Causes of *Chronic* Corneal Edema

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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

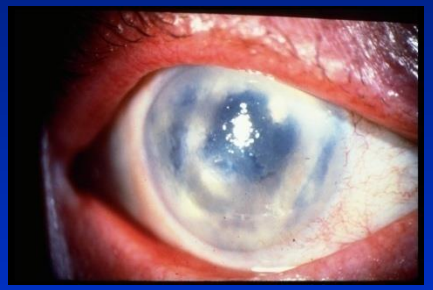
Iridocorneal Endothelial Syndrome (ICE)*

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



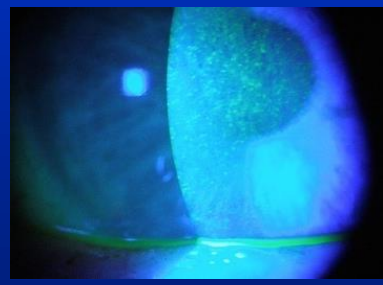
PBK – pseudophakic bullous keratopathy

Punctate Corneal Staining (SPK or PEK)

1. Tear Film Problem

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

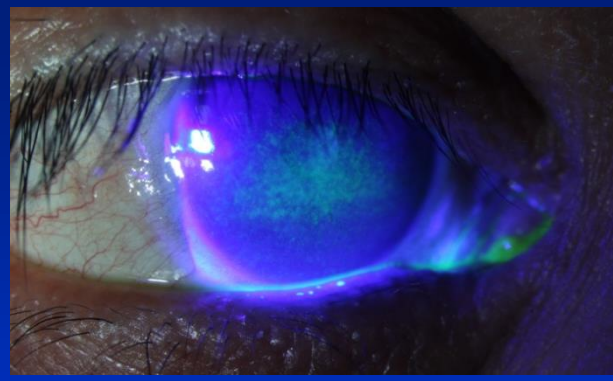
Connective Tissue Diseases: e.g Sjogrens (primary or secondary), RA
Medications (Isoretinoin, antihistamines, antidepressants, BCP)
HIV



Meibomian Dysfunction- Blepharitis, Staph related, Acne Rosacea
Mucin – goblet cell loss – conjunctival disease

2. Exposure Problems

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

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Corneal Infiltrates

Infectious Corneal Ulcers	Versus	Other Infiltrates
Larger Infiltrate		Smaller
Often single lesion		Multiple
Very painful		Less Painful
More Central		More Peripheral
Cells in A/C		No A/C cells
Conjunctival Injection more generalized		More segmental / focal injection

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

Non-Infectious:
Marginal –Staph sensitivity, Mooren's Ulcer
Autoimmune PUK's
CTL Related
Atopic
Chemical
Exposure and Dry Eyes
Neurotrophic

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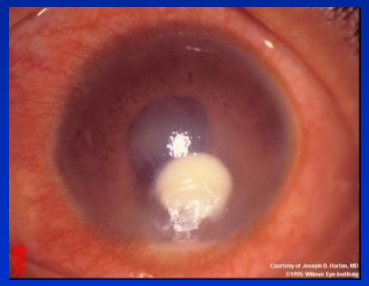
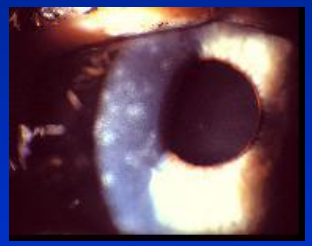
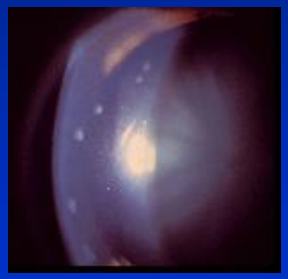
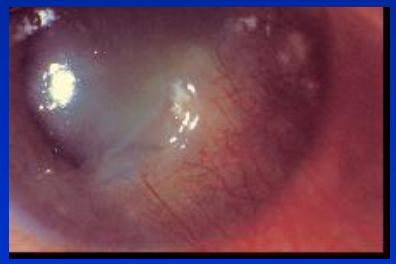
PEDAL – to help distinguish non-infectious from infectious
Infectious more associated with Pain, Epithelial defects, Discharge, Anterior chamber reaction and more *central* Location


Infectious Corneal Ulcer


Adenovirus


Staph Marginal Keratitis


CTL over wear Keratitis


Peripheral / Marginal Keratitis

Corneal Ulceration and Thinning

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Central

Infectious – more common

Bacterial, Fungal

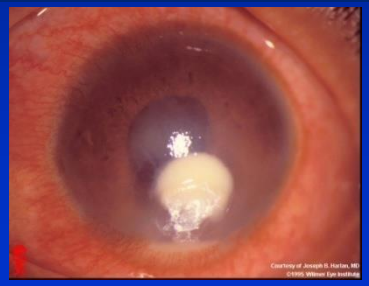
Viral (HSV, HZO)

Acanthamoeba

Autoimmune – much less common

- e.g. RA

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



Peripheral Ulceration and Thinning

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

PUK causes

-Autoimmune Vasculitides (consider as potentially lethal)

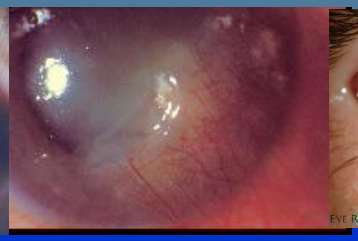
-RA, PAN, GPA, SLE etc. see listing

-Other Autoimmune- e.g. GVH, IBD

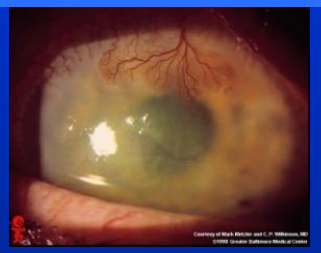
- Infectious (less common)- Staphylococcus, Gonococcus,TB, Syphilis, HSV, HZO, Hepatitis C, Dengue, Acanthamoeba, Fungal

- Other: Trauma, Post-op, Exposure, CTL, Carcinoma, SJS, OCP, KC Sicca, Blepharitis, Pellucid

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



Corneal Vessels / Pannus



- Contact Lens Over Wear - hypoxia
- Staphylococcal Disease – Blepharitis
- 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

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Decreased Corneal Sensation
Hypoesthesia

Common to all cases of Neurotrophic Keratopathy is corneal 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

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Keratic Precipitates (KP)

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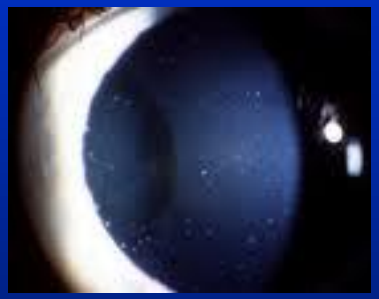
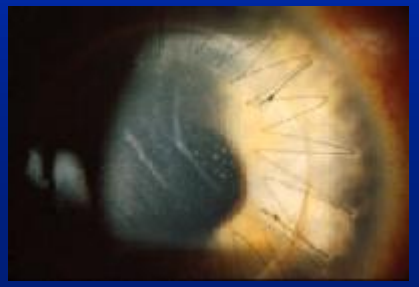
Non-Granulomatous Uveitis Marked by:

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



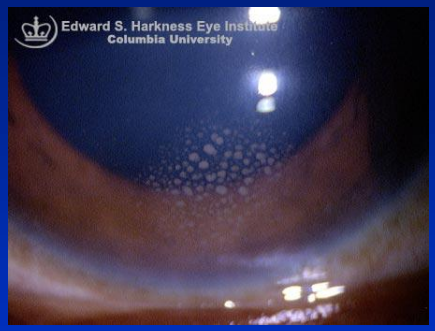
Corneal Graft Rejection
emedicine Ophthalmology

Granulomatous Uveitis suggested by :

Chronic Uveitis — Little Vessel Injection
Mutton Fat KP — Less symptoms

Granulomatous? Think of:

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



Large White or Yellow
"Greasy"
"Mutton Fat" KP

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

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

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Anterior Uveitis and High IOP

Think of:

HSV or HZO with trabeculitis*

Glaucomacyclitic Crisis (PS Syndrome)

Lens Induced

IOL- UGH Syndrome

Steroid Responder

(e.g. uveitis being treated with corticosteroid)

Other: Sarcoidosis, Fuchs, JRA

Also Consider

– not real uveitis, but cells in AC

Pigment Dispersion Syndrome

Retinal Detachment

Ghost Cell Glaucoma

Masquerade Syndrome

– RB, Lymphoma, etc.

Anterior Uveitis:

Get a good Review of Systems

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

Main Work-up Test Considerations:

HLA- B27
FTABS, RPR/VDRL*
CXR
ACE/ Lysozyme
PPD - if Chronic

(and any others on the left that seem indicated)

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

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Hypopion

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

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Hyphema

1. Trauma

   Blunt and Penetrating, IOFB

2. *R/O Intraocular Tumor*

   RB, Melanoma, Hemangioma

3. Post Intraocular Surgery

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

4. Iris Abnormalities

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

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

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

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Neovascularization of the Iris = NVI
Rubeosis

1. Retinal Ischemia:
   - CRVO (Central Retinal Vein Occlusion)
   - PDR (Proliferative Diabetic Retinopathy)
   - OIS – (Ocular ischemic syndrome, carotid disease)
   - CRAO – (Central Retinal Artery Occlusion – less common)
   - Anterior Segment Ischemia
   - Blood Disorders: Sickle Cell
   - Vasculitis, ROP, Coats, PHPV
   - Neurofibromatosis - Gliomas, large and small vessel problems
2. Chronic Retinal Detachment
3. Chronic Uveitis, Endophthalmitis, Hypotony
4. Previous Trauma, Surgery or other Insult (e.g. Radiation)
5. Post-op
6. Intra-Ocular Tumors of the Iris, Choroid, RB, etc.

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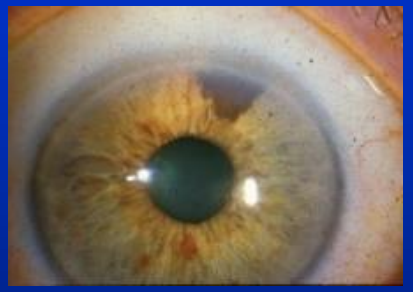
NVI Mimic: Dilated iris vessels – usually radial, seen with active inflammation

Lesions of the Iris

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

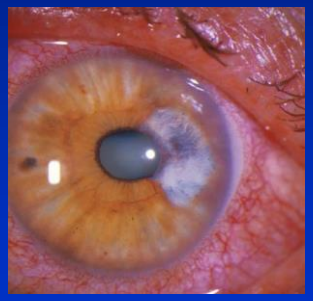


4. Iris Cysts – traumatic, post-op, congenital or acquired stromal cysts, pigmented epithelial cysts
5. Infectious: (Nodules, Papules) TB, Leprosy, Syphilis
6. Primary Tumors: Juvenile Xanthogranuloma, Hemangiomas,
   Neurofibromas, Choristoma (Ectopic Lacrimal Gland)
7. Metastatic: Carcinomas, Leukemia, Lymphoma
8. Segmental Melanosis Oculi
9. Things mistaken for masses:

   Ectropion Uvea, Segmental Iris Atrophy

   (e.g. HZO, CE), Iris Defects, Synechiae (trauma, surgery)

Stromal cyst



Segmental Iris Atrophy

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Defects of the Iris

Iris Transillumination Defects:

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

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Heterochromia

- Horner Syndrome
- Sturge Weber Syndrome
- Uveitis – including Fuch's and Posner Schlossman
- Pigment Dispersion
- Use of Prostaglandin analogs
- Trauma and Surgery
- R/O Benign Heterochromia
- Parry Romberg Syndrome
- Other Syndromes
- Pigmented IrisTumors
- Siderosis Bulbi

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)

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Pediatric

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

Anterior Polar

Nuclear – often central 3 mm, rubella

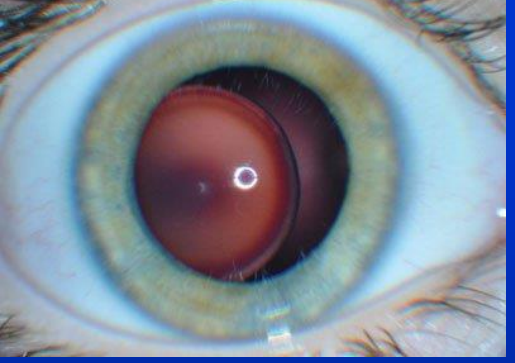
Posterior Polar

Zonular (Lamellar) - 50% of visually significant

When to do basic Pedi Cataract Workup

Bilateral Congenital , + FHx – no testing
Bilateral Congenital, - FHx, do W/U
Acquired Bilateral older – do W/U

Unilateral Congenital or Older and Healthy – no testing



Lens Abnormalities
and associations

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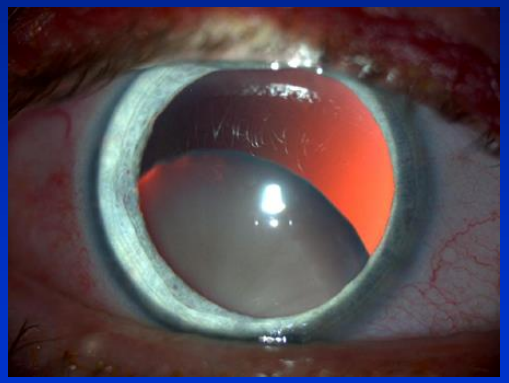
1. Aphakia – acquired or congenital (rare)
2. Spherophakia – high myopia, Weill- Marchesani Synd.
3. Lens Coloboma - often associated with other ocular colobomas
4. Ectopic Lens

   Trauma, Surgery

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

   Marfan's Sydnrome, Homocystinuria,

   Hyperlysinemia, Ehlers-Danlos Syndrome.



High Intraocular Pressure

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

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

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

Narrow Angles or Angle Closure

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

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Angle Closure Glaucoma
and Pupillary Block

With Pupillary Block

Primary ACG

Secondary ACG
- Phacomorphic
- Ectopia lentis
- Pseudophakic (IOL)
- Aphakic (anterior vitreous face)
- Posterior Synechiae (with secluded pupil, iris bombe)

Without Pupillary Block

- Neovascular Glaucoma
- ICE – abnormal corneal endothelial growth
- Intra-ocular tumors
- Uveitis – Peripheral Anterior Synechiae
- Plateau Iris

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Open Angle Glaucoma
Mechanisms

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Primary - (POAG)* - usually symmetric

Secondary – more often unilateral

1. Clogging of TM - RBC, lens protein and macrophages, pigment cells, tumor cells, viscoelastics

2. Alteration of TM - Inflammation (Trabeculitis), Trauma (physical and chemical) FB – siderosis, Corticosteroids

3. Blockage of drainage after the TM
   - Sickle Cells in Schlemm's Canal (even in Sickle trait)
   - Increased episcleral pressure – Carotid Cavernous (CC) Fistula, Orbital tumor /inflammation, venous backup

Open angle Glaucoma

By Disease Process

1. Primary Open Angle (POAG)

2. Secondary Open Angle (SPLIT mnemonic)

Substances: Steroids, Viscoelastics, Siderosis (Iron IOFB)

Pigmentary

Pseudoexfoliation

Lens – 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

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Flat or Shallow AC

With High IOP

Acute Angle Closure Glaucoma (AACG)

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

Synechiae closure
- anterior
- posterior – pupillary block, iris bombay

Malignant Glaucoma

Mature, Intumescent (Swollen) Lens

Flat AC: Grades

– contact with corneal endothelium and …

I –peripheral iris
II – iris up to pupil
III – lens (surgical urgency)

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

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Hypotony (Low IOP)

Causes and Associations

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

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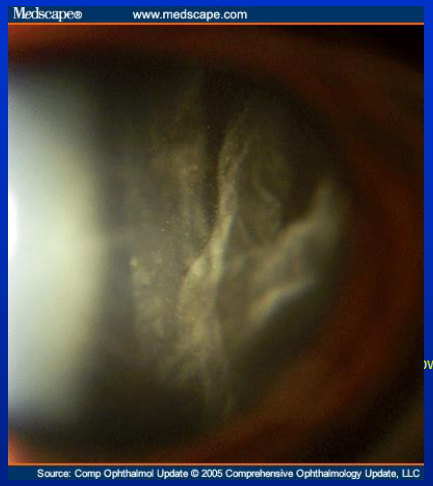
Fundus – Vitreous, Retina and Disc

Vitreous Cells, Pigment or Debris



- Blood (RBC and ghost RBCs) – see vitreous hemorrhage
- White Blood Cells (WBC)

  Uveitis: intermediate, posterior or panuveitis

  Masquerade (actually tumor cells - e.g. lymphoma, RB)
- Pigmented Cells or Granules – can be a sign of RD
- Post Vitrectomy or Other RD Surgeries
- Larger Particles (Crystals) that could be mistaken for cells

  Cholesterol Crystals- Synchysis scintillans (past injury or inflammation)

  Asteroid Hyalosis - particles made of calcium and phospholipids

  - usually unknown cause, but associated with DM, HTN , hypercholesterolemia

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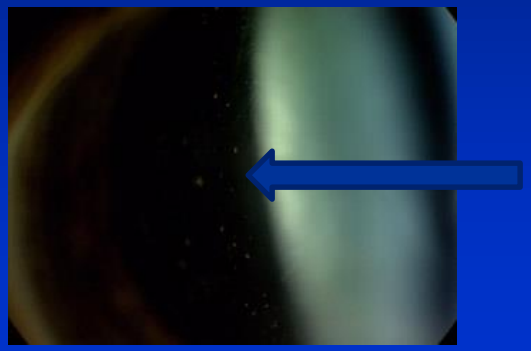
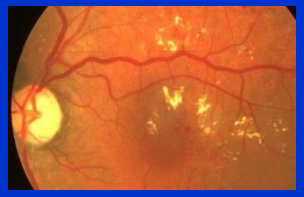
If cells are clumped think more intraocular tumors like CNS Lymphoma or Mets



Pigment in Anterior vitreous

Whitish or Yellowish Retinal or Sub-Retinal Lesions

Discrete Borders



Hard Exudates

Vessel Plaques

Certain retinal/choroidal tumors

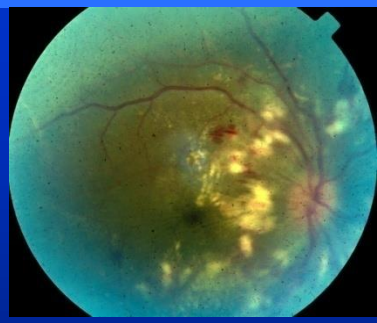
Drusen

Macular Dystrophies

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

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

Less Defined Borders

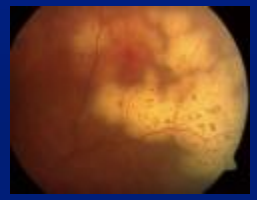


Cotton Wool Spots

Myelination of NFL

Infiltrates:

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



White Dot Syndromes*

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

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

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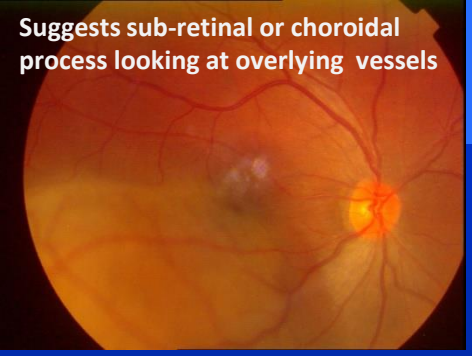
Apparent Mass in Retina / Choroid


Suggests sub-retinal or choroidal process looking at overlying vessels

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Low internal reflectivity

DDX

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

Choroidal Detachment – effusion, hemorrhage

Nodular Posterior Scleritis

Choroidal Nevus

CHRPE

Exudative Retinal Detachment
- From: Tumor, Post. Uveitis, Systemic (e.g. Toxemia), etc.

Subretinal Hemorrhage

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

Evaluation and Management

A, B-scan, FA, CT of Orbits

Assessing for Systemic Involvement

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

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

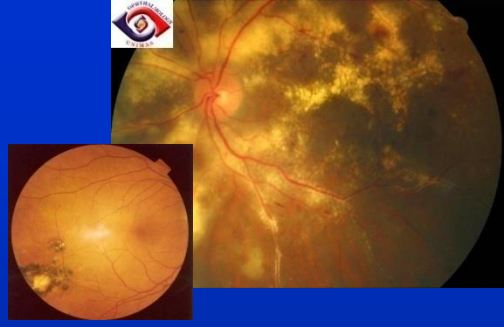
If suspect melanoma or metastasis – do a systemic evaluation

If negative systemic work-up
- consider local Rx- radiation, laser, cyro, plaque, or enucleation depending on size

If there is metastasis or primary disease elsewhere
- consider palliative radiation, injections or other local Rx as enucleation is not necessary

Posterior Uveitis

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



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- *Toxoplasmosis* – active infiltrate, often an adjacent CR scar, Treatment*
- Toxocariasis – Granuloma – often up in vitreous, usually no CR seen
- Syphilis –front (keratitis) to back (chorioretinal, optic nerve), panuveitis
- HSV, VZV, CMV: Acute Retinal Necrosis (ARN), Progressive Outer Retinal Necrosis (PORN)

  ARN- relatively immunocompotent- associated with inflammatory signs (AC or Vitreous Cells, with retinal infiltrates+ necrosis)

  PORN – immunocompromised - little if any inflammatory response – just retinal necrosis
- Other Infectious: Lyme Disease, Tuberculosis, Typhus, Whipple's, West Nile Virus, Rubella, Nematode (DUSN - sub-retinal)
- Candida
- Sarcoidosis
- Intraocular FB, Siderosis
- Sympathetic Ophthalmia
- Cat Scratch Disease (Bartonella)
- Vogt – Koyanagi –Harada Disease (VKH)
- Behcets Disease
- Uveitis associated with MS
- Eales Disease – posterior peripheral phlebitis
- Pars Planitis – more intermediate uveitis
- Multifocal Idiopathic Inflammatory Conditions of Retina and Choroid ("White dot syndromes")

  APMPPE and PPM, Serpiginous Choroiditis, Birdshot Retinochoroidopathy, Multiple Evanescent White Dot Syndrome (MEWDS), Multifocal Choroiditis and Panuveitis (MCP), Presumed Ocular Histoplasmosis Syndrome, Acute Idiopathic Blind Spot Enlargement Syndrome, Acute Retinal Pigment Epithelitis

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

Work up Considerations

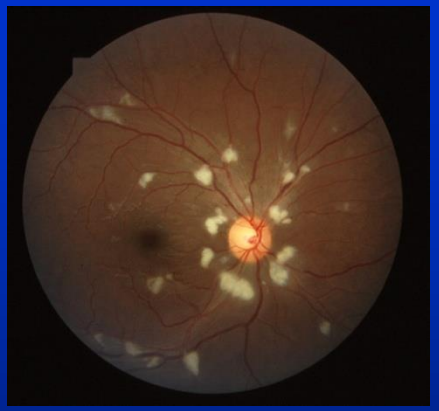
Much more based on clinical findings

FTABS, VDRL
PPD , Quantiferon Gold, CXR

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

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

Cotton Wool Spots



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

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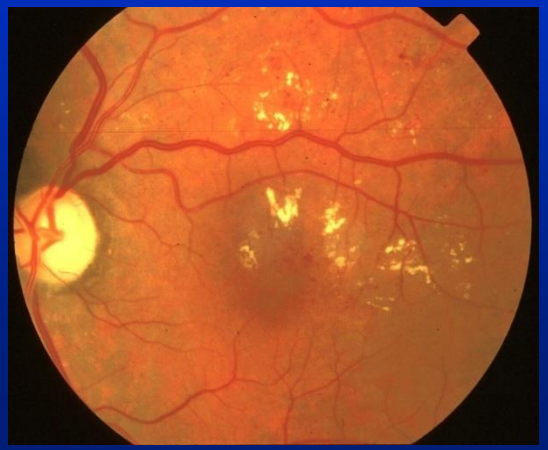
Radiation retinopathy
Interferon Therapy
Pregnancy (Toxemia)
Purtscher's retinopathy
Collagen vascular disease (e.g. SLE)
Severe Anemias, other Blood Disorders (Leukemia, Multiple Myeloma, etc)
Coagulopathies
Sepsis/ Sub-Acute Bacterial Endocarditis
Fat emboli, CO poisoning, Renal Diseases

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 Hipple-Lindau), retinal artery aneurysms, vasculitides, vasculopathies, FEVR

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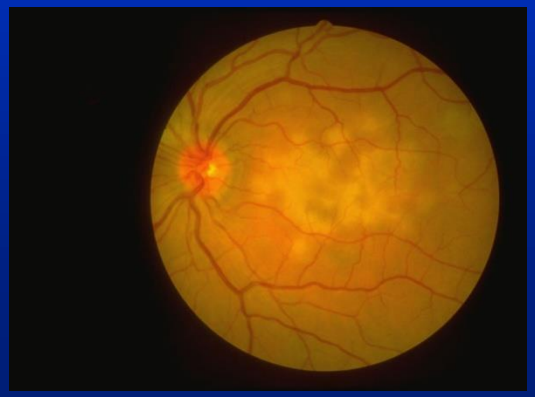
Retinal Infiltration or Edema

Can look similar

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



CRAO or similar picture

Carotid or Ophthalmic artery disease

Embolic Phenomenon- carotid or heart sources

Vasculitis- e.g. GCA

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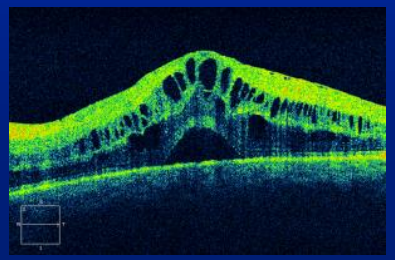
Macular Edema or Thickening

Often detected with Contact Lens, FA or OCT



- Cystoid Macular Edema
  - – e.g. Post Cataract Extraction, Broken Capsule, Vitreous Loss
  - - Vitreal Macular Traction (VMT), ERM, Retinitis Pigmentosa
  - - associated at times with ARMD, DR, Uveitis, RVO
  - - Medications: Niacin, Fingolimod, Tamoxifen…
- Diabetic Macular Edema
  - – associated with microaneurysms and hard exudates
- Uveitis – anterior, posterior or pars planitis
- CRVO or Branch RVO
- Trauma – Commotio Retinae, Berlin's Edema
- Microcystic Macular Edema – seen in association with optic neuropathies
- Vascular Tumors of Retina
- Retinal Telangiectasia
- Choroidal Tumor - underlying
- Medications: Hydrochlorothiazide
- Other: Juvenile Retinoschisis –cystic spaces, not really edema, separation of inner retina from other layers

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Posterior Hemorrhages

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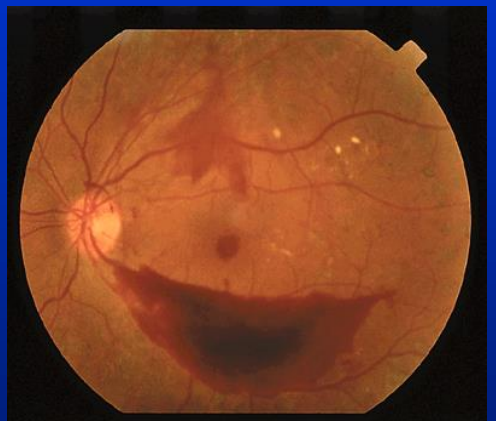
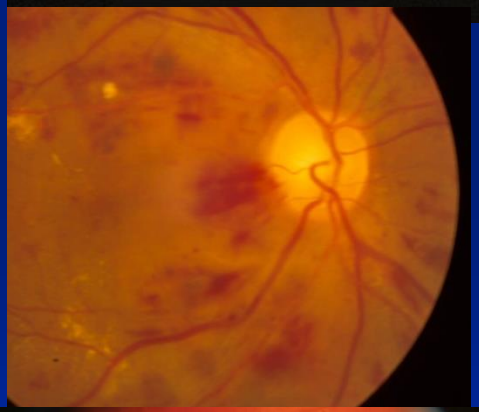
Types

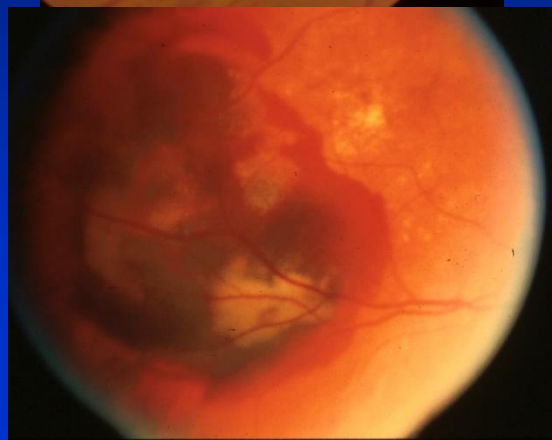
- Pre-retinal Hemorrhage
  or Vitreous Hemorrhage
  (blocks view of vessels)



- Intra-Retinal Hemorrhages



- Sub-Retinal Hemorrhage
  (can see vessels over it)



Pre-Retinal or Vitreous Hemorrhage

Trauma – blunt or penetrating, eye or head

Post-op

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

Vitreous Detachment or Traction

Retinal Tear, Break, or Detachment

Chronic Uveitis

Tumor

Macroaneurysm

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

Valsalva

IOL

SRNVM – rare but possible

Retinal or Sub-Retinal Tumor

X-linked Retinoschisis – young males

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

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Neovascularization
Disc ,Retina, or Sub-Retinal

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

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Intra-Retinal Hemorrhages

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

Vascular Diseases: DM, HTN, OIS, HIV , Vasculitis, Sickle Cell (and other Hemoglobinopathies) , 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

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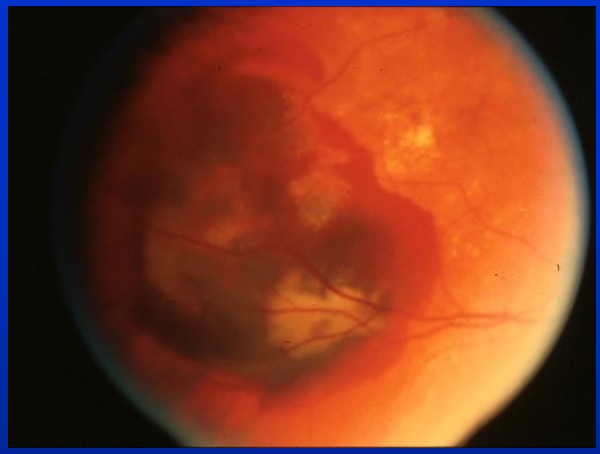
Sub-retinal Hemorrhages



ARMD with SRNVM (CNVM)

Other causes of CNVM

- High Myopia
- Angioid Streaks
- Histoplasmosis

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Trauma

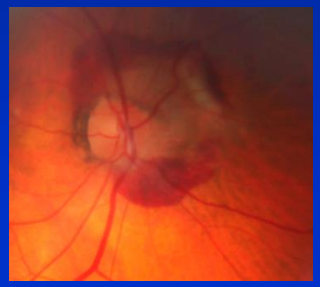
Macroaneurysm

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

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

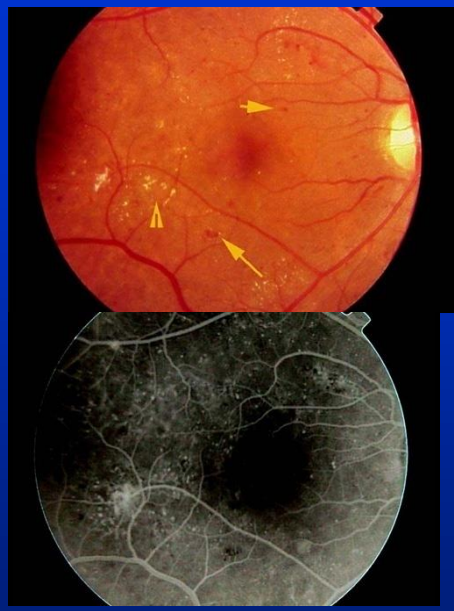
(B-scan may be needed in seemingly spontaneous Sub-Retinal hemorrhage)

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)

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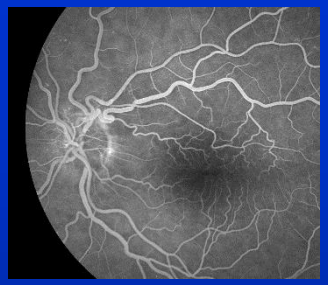
Tortuous or Enlarged Retinal Vessels



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

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

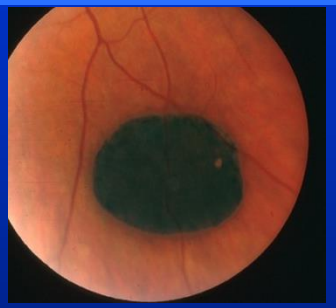
Enlarged Retinal Veins

- 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

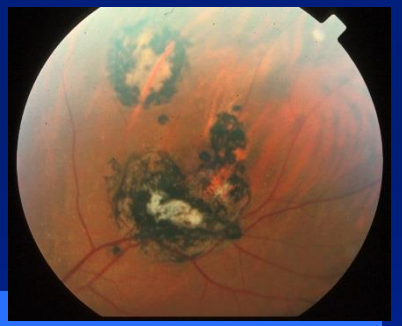
Dark or Pigmented Retinal Lesions

- 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



CHRPE



*Chorioretinal Scars

Past Chorioretinitis, vasculitis
- Infectious – e.g. Toxoplasmosis
- Autoimmune

Exudative/ Younger - FEVR, Coats, Stargardt's
Exudative / Older – Wet ARMD, Macroaneurysm
Past Retinal Detachment
Past Eye Trauma
Response to Neoplasm, Past Ischemia?
Mimics- Congenital Lesions, Myopic Degeneration

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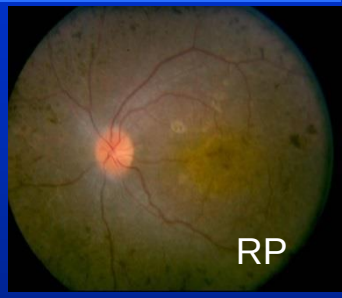
Pigmentary Retinopathies

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

Drug Toxicity
e.g. Phenothiazines, Iron overload / Transfusions

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



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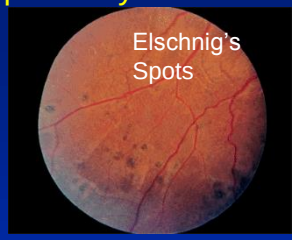
Retinitis Pigmentosa
Familial - about 50%
Simplex (Sporadic) - about 50%
Associated with Systemic Disease and often Hearing Loss: Usher Syndrome, Refsum Disease (elevated phytanic acid)

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

Retinal Detachment

Rhegmatogenous

– due to retinal break, tear, dialysis

- Associated with trauma, intraocular surgery, high myopia, Chorioretinitis, Aphakia, Traction

Tractional

PDR and Other proliferative retinopathies

Hemoglobinopathies: Sickle Cell (SSD, SCD), Thalassemia, Eales Disease

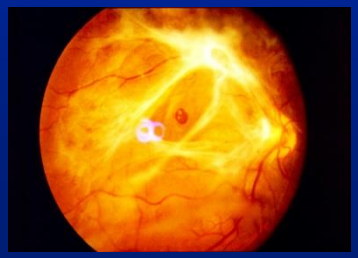
Ischemic CRVO, ROP

PVR (proliferatvie vitreo retinopathy)

Fibrosis after prolonged Vitreous Hemorrhage

Trauma – Penetrating, Surgery

Other - FEVR



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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

Retinal / Choroidal - Folds/ Striae

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

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Optic Atrophy / Disc Pallor

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

Causes:

Ischemia - e.g. past AION or PION

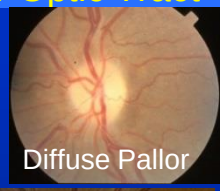
Compression - e.g. Pituitary Tumor, Carotid artery, Hydrocephalus, Graves Ophthalmopathy

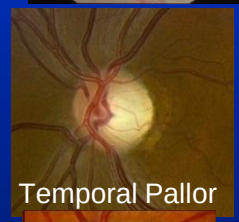
Chronic Papilledema - compression +/- ischemia – *see high ICP list*

High IOP - e.g. 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


Diffuse Pallor


Temporal Pallor


Bowtie Atrophy

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, CCl_4
Nutritional Deficiencies (B_1 (Thiamine), B_{12}, 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

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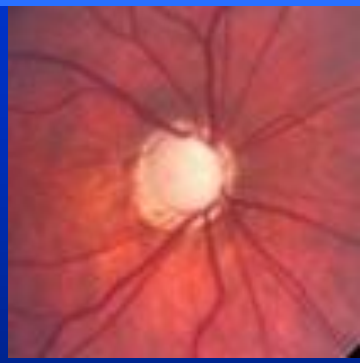
Retinal Damage - (False Ocular Localizing Sign) - CRAO, CRVO, Ischemic PDR, S/P PRP, Retinitis; Degeneration (RP)

Optic Disc Cupping



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

Morning Glory Disc – The American Society of Retina Specialists

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Disc Edema

See also Acute Optic Neuropathy

Causes

High ICP - Papilledema

Ischemia – AION

Inflammation / Optic Neuritis

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

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

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

Hereditary : e.g. - LHON

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

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

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

Venous stasis (CRVO, BRVO), Hypotony

Posterior Scleritis, Uveitis

Acute Multifocal Placoid Pigment Epitheliopathy (AMPPE)

Multiple Evanescent White Dot Syndrome (MEWDS)

OR

MAYBE NOT EDEMA, BUT SOMETHING THAT LOOKS LIKE IT

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

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

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Disc Hemorrhages

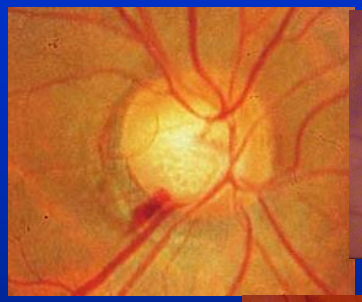
Splinter / NFL / Linear Hemorrhages off Disc

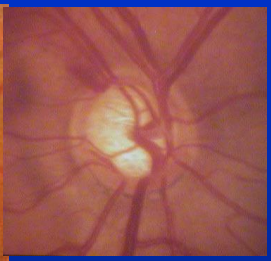
- Glaucoma – ***unstable, progressing***

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

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

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





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

Peripapillary Subretinal Hemorrhages

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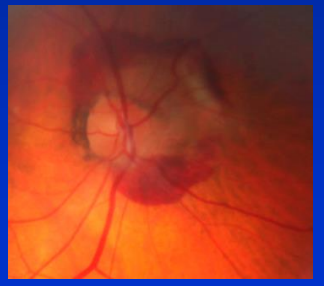
Return To Links

- 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

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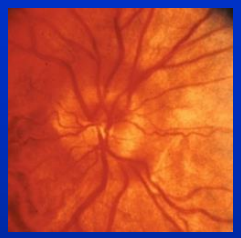
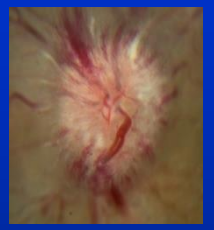


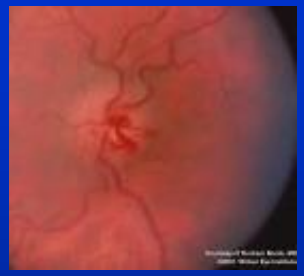


Feature	Congenital	Acquired
Nerve Fiber Layer	Clear	Opacified
Large Disc Vessels	Anomalous	Normal
Small Disc Vessels	Normal	Telangietatic
NFL Hemorrhage	Rare	Frequent
Physiologic Cup	Small or absent	Normal (But may be obscured by edema)
Drusen	Sometimes present	Absent

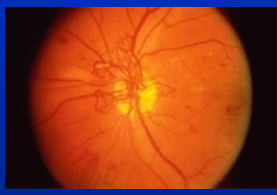
Abnormal Disc Vessels and Growths

Abnormal Vessels

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



Opto-ciliary Shunt Vessels

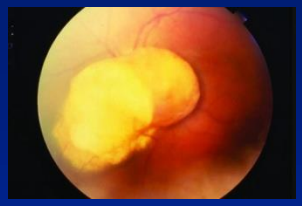


NVD

ERM dragging disc vessels

Abnormal Growths / Remnants

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



Astrocytoma

Transactions of American Ophthalmological Society 2004; 102

NFL Myelination

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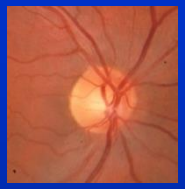
Possible Causes of Optic Nerve Inflammation




Retrobulbar Papillitis

Demyelinating:
Multiple Sclerosis, Neuromyelitis Optica = NMO (Devic's Disease)
Post-Viral / Immunization : Acute Disseminated Encephalomyelitis (ADEM)

Idiopathic

Viral, Post-Viral, Post-immunization

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

Contiguous Inflammation :
Encephalitis, Meningitis (high ICP and inflammation)
Pachymeninigitis – (Idiopathic, intracranial hypotension, Meningeal carcinomatosis, vasculitides, infections such as p. acnes)
Orbit (orbital pseudotumor – e.g. optic perineuritis), IgG4-ROD
Sinuses -Infectious (including Fungal – e.g. mucor, aspergillis), 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)

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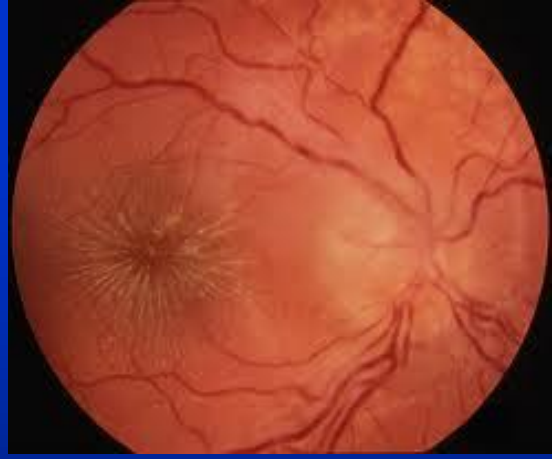
Remember certain acute optic neuropathies can resemble optic neuritis:
like: NAION, PION, Compressive Lesions, LHON, …

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

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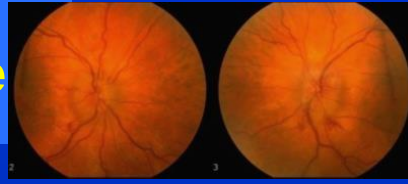
*Neuroretinitis can be ***recurrent*** and sometimes require Immunomodulatory or immunosuppressive therapy

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

Papilledema and Causes of Increased Intracranial Pressure



1. Hypertension – Malignant

2. Intracranial Tumor (by mere mass effect or by causing obstructive hydrocephalus), Carcinomatous Meningitis

3. Medications -Vitamin A, Accutane, Tetracyclines, Contraceptives, Corticosteroid withdrawal, Thyroid Replacement, Growth Hormone Supplement, ? Fluoroquinolones

4. Toxic: Ethylene Glycol, Lead (Pb), Hyper-ammonemia

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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

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3. Miscellaneous Case Situations and other Lists

Visual Loss

Reading Problems

Pediatric Presentations: Visual Loss, ET, Glaucoma, Nystagmus

Pregnancy

Systemic Medical Conditions

CN Palsies

Elevated ESR and Concern for Temporal Arteritis

Low Tension Glaucoma

Ocular Effects of Systemic Medications

Imaging Findings- CT, MRI

Dizziness

Differing Scenarios of Visual Loss

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

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

Longstanding Visual Loss
Since Childhood or Young Adulthood

Major Considerations:

"Front to Back"

- Refractive

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

- Media Opacities

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

- Retinal

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

- Optic Nerve

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

- CNS

  Amblyopia (Deprivation, Anisometropia or Strabismus),

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

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- Trauma – to eye or visual pathways:

Sudden / Acute Loss of Vision

Developing over Hours to Days

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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*

Timing

Abrupt – ION, LHON

Subacute – optic neuritis

Insidious – compressive or metabolic

Character

Dark spot – optic neuropathy

Metamorphopsia - maculopathy

RAPD and Acute Unilateral Visual Loss

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

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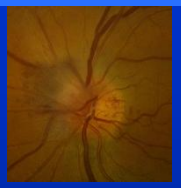
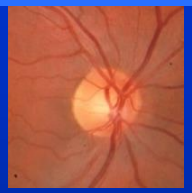
Acute Optic Neuropathy

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



With



or Without Disc Swelling

"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

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Chronic / Progressive Loss of Vision

Developing over Months to Years

Major Considerations: "Front to Back"

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

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"Unexplained" Visual Loss"

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

Consider:

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

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

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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

Color Vision Loss

Monochromatic – hereditary – or end stage of any condition below

Red-Green (Protan and Deuteran)
- non-specific, seen with Hereditary, Maculopathies and some acquired optic neuropathies

Blue- Yellow (Tritan)
specific for vascular retinopathies, papilledema, glaucoma and DOA

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

"Unexplained" Visual Loss

More Comprehensive Listing

Click to Return To Links

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), White Matter Diseases, CJD= Creutzfeldt-Jakob disease, PML = Progressive Multifocal Leukoencephalopathy, AIDS, Sleep Apnea; MELAS- Mitochondrial myopathy Encephalopathy Lactic Acidosis and Stoke Syndrome, Non-ketotic hyperglycemia, Dementias, Procedures: cerebral angiography, ventriculography, blood transfusions. Uremia, porphyria, syphilis, endocarditis, SSPE – subacute sclerosing panencephalitis – related to measles (rubeola), sudden change in ICP, Hypoglycemia, correction of hyponatremia, Epilepsy , Chemo and Meds: cis-platinum, tacrolimus, methotrexate, methampetamine, vincristine, vindesin, interferon. Posterior Leukoencephalopathy (PLE) seen on MRI seen with hypertensive encephalopathy and cyclosporine toxicity, Toxic: Nitrous Oxide, ethanol, Pb, Hg, Organophosphates . Migraine patients with permanent VF defects, occult CVA.. Remote Effect (paraneoplastic) -anywhere along visual pathways. PRES – posterior reversible encephalopathy syndrome

5. Other

Amblyopia – especially think about anisometropic amblyopia, monofixation syndrome

and consider: Non-Organic (Functional) Visual Loss

Unexplained Bilateral or Quickly Sequential Acute Visual Loss

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

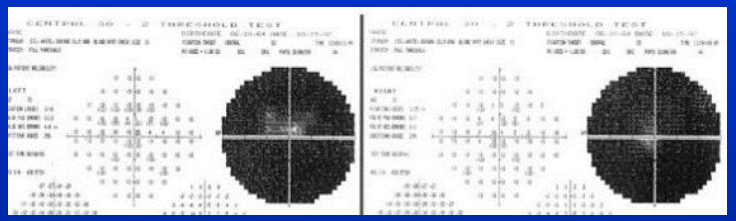
Vascular

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

Severe Systemic Hypertension

Vertebrobasilar Insufficiency

Temporal Arteritis – e.g. PION



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Retinal

Paraneoplastic: MARS and CARS*

Optic Nerve

LHON

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

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

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

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

PION – e.g. post-op, trauma, shock

CNS

Migraine

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

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

Other

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

Problems with Reading

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

1. Loss of **Accommodation**

   Presbyopia, Diabetes, Latent Hyperopia, see earlier list

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

3. **Strabismus**

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

   4th CN Palsy 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)

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

Recall Near Triad

- Accommodation
- Convergence
- Miosis

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Suspected Loss of Vision in an Infant

Signs of Poor Vision in Infant:

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

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

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

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

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First do a complete eye exam looking for more obvious causes

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

Considerations:

Delayed Maturation of Visual System

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

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

Leber's Congenital Amaurosis

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

Cone Dystrophy

Albinism / Foveal Aplasia

ERG appropriate in suspected Retinal Dystrophy

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

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

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

3. Systemic: Sturge -Weber, Neurofibromatosis

4. Secondary Infantile Glaucomas from
   Intra-ocular Tumor – e.g. RB (don't forget possibility)
   Uveitis / Infectious Process – e.g. Rubella, Toxocara, Herpetic

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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

Child with Esotropia

1. Congenital / Infantile ET
2. Pseudo-Esotropia – Epicanthal folds, Negative angle kappa, Hypotelorism, Telecanthus
3. Hyperopia / Accommodative ET
4. Acquired Non-Accommodative ET*
   (most will not have a CNS lesion, but need to consider)
5. Duane's Syndrome
6. Cyclic ET – ET on a couple/few days and then off, often pre-school years, sort of like intermittent that eventually becomes constant.
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.

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Nystagmus in a Child

1. Congenital / Infantile Nystagmus

Characteristics – History of Early Onset- sometimes at birth but usually at 2-3 months
Absence of Oscillopsia, Vertigo or Imbalance. Absence of Neurologic abnormalities outside of Visual System
Predominately Horizontal Conjugate Jerk or Pendular Nystagmus, Dampens at Near/Convergence
Increases with attempted Fixation or pursuit. Often a Null point – central or eccentric gaze

2. "Eye" Problems – Sensory Visual Loss

e.g. Deprivational Amblyopia,
Congenital Cataracts, Anterior Segment Dysgenesis (e.g. Peter's Anomaly)
PHPV, ROP, Albinism, Foveal Hypoplasia, Macular Dystrophy or Scar
Retinal Dystrophies: LCA, CSNB, Cone-Rod Dystrophy, Achromatopsia
Optic Nerve Anomaly (e.g. Optic Nerve Hypoplasia, Optic Atrophy)

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3. "Neurologic" Problems

Spasmus Nutans – Benign Condition – nystagmus, head nodding and torticollis
Anterior Visual Pathway tumors – hypothalamic, chiasm, optic nerve
Encephalitis, Post Viral Syndrome
Disorders in Posterior Fossa – Tumor, Cerebellar Inflammation
Arnold – Chiari, Hydrocephalus, Spinal Cerebellar degeneration
Neurodegenerative Disorders- e.g. Leigh Disease, Pelizaeus-Merzbacher Disease, Joubert Syndrome
Metabolic: Malnutrition, Maple Syrup Urine, Hypothyroid
Other: Trauma, Down's Syndrome, Medications and Toxins
Ospoclonus – not true nystagmus, can be herald of Neuroblastoma, acute cerebellar ataxia

Pregnancy and Vision and Eye Conditions

Exacerbated

- CVA
- Retinal Vascular Occlusion } Thrombo - Embolic
- Meningioma- e.g. Sphenoid wing
- Pituitary Apoplexy
- High ICP: Venous Sinus Thrombosis
  - Pseudotumor Cerebri Exacerbated
- Migraine
- Diabetic Retinopathy (DME)
- CSR
- Cranial Neuropathies: 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

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Post-Partum

- Post Partum Optic Neuritis
- Intracranial Hypotension and
  - Acquired Chiari - 1 Malformation
  - (Headache, Diplopia)

Associations For Baby

- Prematurity – ROP, ET, XT
- Maternal Smoking – ET and XT
- Maternal Alcohol Use → Fetal alcohol syndrome
  - Telecanthus, ON Hypoplasia, Strabismus

Important Medical Conditions and their Associated Eye Pathology

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

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2. Diabetes Mellitus
   Early Presbyopia, Non reactive pupil, CNP (especially 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

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Cranial Nerves Palsies

Related to Ocular Motility

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

3rd	Oculomotor
4th	Trochlear
6th	Abducens

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Approach: If it is ***isolated*** *(no other neurologic signs or symptoms) , then observation is often OK, but* ***Follow -up*** *of the patient to look for improvement or stability is key. If the diagnosis is a microvascular CNP you must make sure on follow-up that the palsy is resolving or resolved in 2-4 months. IF the patient has more* ***pain, progression or pupil dilation****, then consider early neuroimaging. A 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

3rd CNP

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

6th CNP

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

4th CNP

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

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Elevated ESR

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

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

2. Anemia - anemic patient can have a artificially elevated ESR secondary to anemia

3. Systemic inflammatory conditions – Polymyalgia Rheumatica, Lupus, Vasculitis
Younger Patients with an Autoimmune Disease can present with an Acute Optic Neuropathy

4. Cancer - may need work-up for an occult CA,
(note some patient with cancer are anemic)

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5. Hyper –proteinemias - e.g. Multiple Myeloma, Gammaglobulinemias

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

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

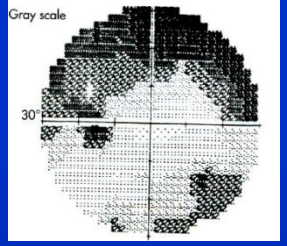
Suggested work-up for patient with no obvious cause for an elevated ESR:
C-Reactive Protein, CBC (with platelets) , Urinalysis, BUN/Cr, alkaline phosphatase,
Serum Protein Electrophoresis, Chest X-Ray, CT of Sinuses, *or a temporal artery biopsy?*

Temporal and Forehead Pain and Tenderness

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

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

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



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

Before making the diagnosis of

- Low / Normal Tension Glaucoma*

consider also

- *Anterior Visual Pathway Lesion*

  e.g. Parasellar Tumors, ON or sheath tumors, etc.

- *Previous Optic Nerve Damage from:*

  ACG, Previous Open Angle Glaucomas,

  AION, AAION, Papilledema or Optic Neuritis, BRAO, LHON

- *Carotid Artery Disease*

- *Congenital Disc Defects – pits, drusen, tilted or myopic discs*

- *Temporal Disc Pallor can be mistaken for cupping:*

  Hereditary (DOA) or Acquired Optic Atrophies (Toxic, Nutritional, Tobacco)

Evaluation should include:

Good History can eliminate several possibilities

Careful exam of disc
Rim pallor more suggestive of another optic neuropathy

Could also include:
- Corneal Pachymetry – thin cornea could give falsely low IOP

- Diurnal Curve- IOP might fluctuate during day in patient with POAG

More likely Glaucoma if :
- Older patient
- Disc hemorrhages
- VF defects that respect the horizontal midline

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

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Ocular Effects of Medications

- 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

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- Retinal Pigmentary Changes

  Phenothiazines - Thioridazine (Mellaril), Chlorpromazine (Thorazine), Chloroquine, Hydroxychloroquine
  Tamoxifen, Indomethacine, Deferoxamine

Ocular Effects of Medications

- Papilledema / Increased Intracranial Pressure

  Vitamin A , Retinoids e.g. Isotretinoin (Accutane) , Tetracylines: Doxycycline, Minocycline, Lithium, Human Growth Hormone, Thyroid supplementation, Nalidixic Acid, *Oral Contraceptives, Withdrawal of Corticosteroids,*

- Nystagmus

  Phenytoin, Barbituates, Lithium, Salicylates, Antihistamines

- Loss of VF and/ or NFL

  Vigabatrin

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- Optic Neuropathy

  Amiodarone, Ethambutol, INH, Quinine, Sulfonamides, Chloramphenicol, SSRI's
  Anti TNF Ab (Remicade, Embrel), Tacrolimus
  Interferon; Chemotherapy (Doxorubicin, Cisplatin, Methotrexate, Vincristine) especially with Radiation;
  Lindane Shampoo (absorbed)

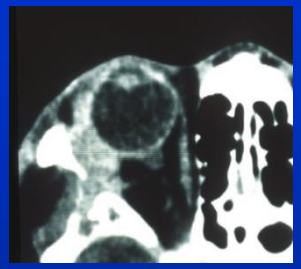
- Myasthenia Gravis (Induced, Aggravated or Mimic)

  - Statins (3HMG CoA reductase inhibitors)

Also consider: Ca Channel Blockers (e.g. Verapamil), Beta Blockers (e.g Propranolol or Timolol), Anti-Arrhythmics (e.g. procainamide), Quinine, chloroquine, Penicillamine, Corticosteroids, Antibiotics (e.g. Fluoroquinolones, Aminoglycosides, Macrolides), Anti-Seizure (e.g. phenytoin, carbamazepine and Gabapentine), Chemotherapy, Interferon, Some IV Anesthetics,

CT of Orbits

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)

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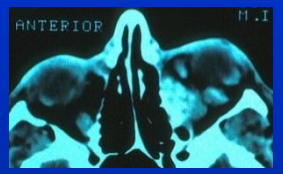
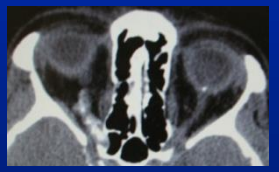
Calcifications

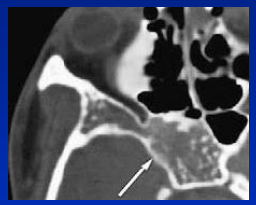


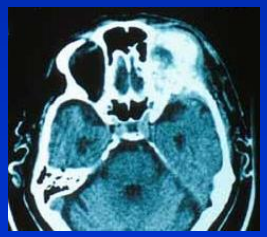
Orbital Varix (Phleboliths)
Optic Nerve Sheath Meningioma
Ocular Tumor – e.g. RB
Optic Nerve Head Drusen



Boney Growth / Change / Expansion

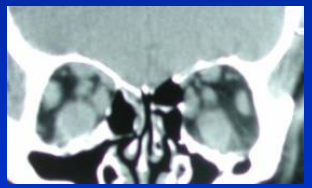
Langerhans Histiocytosis (1-4 yo)
Fibrous Dysplasia (children- young adults) →
Sarcoma of the bone
Sphenoid Wing Meningioma
Paget's Disease (older patients)
Metastatic Disease to the Bone →

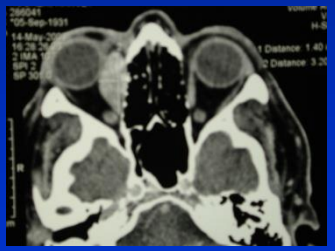




Enlarged Extra Ocular Muscle(s)

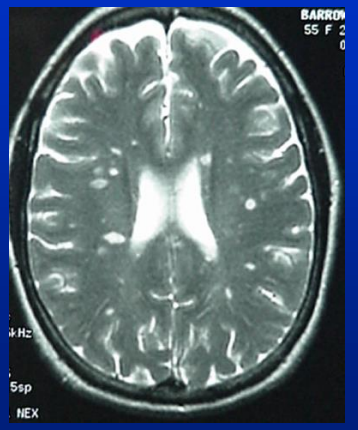
Graves Ophthalmopathy (IR ,MR >SR most common)
Orbital Pseudotumor (Myositis)
Post- Viral Myositis (Children), Cysticercosis
Lymphoma, CLL
Metastatic Disease – breast, melanoma
Primary Tumor – e.g. hemangioma, fibrous / Amyloid





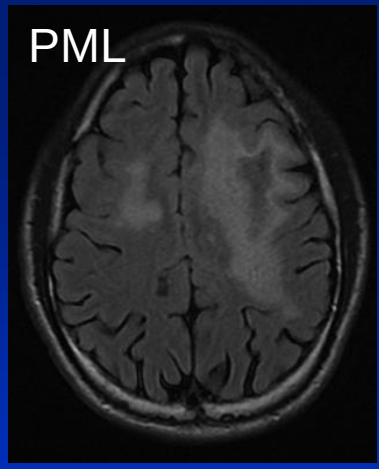
MRI of the Brain
(White Matter Lesions)

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 Leukoencepholopathy – viral disease in immunocompromised)
- Radiation



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Dizziness

Dizzy patients are often referred for eye exams

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

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Disequilibrium – elderly issues like fear of walking, musculoskeletal problems, peripheral neuropathy

Consider also eye Problems: visual loss or causes of oscillopsia and diplopia or problem with glasses

Pre-syncope – Orthostatic Hypotension from medications, arrhythmias, vaso-vagal

Psychiatric – anxiety, hyperventilation, depression, substance abuse

Multifactorial or unknown

- Acute Vestibular Syndrome – rapid onset of vertigo, N/V, nystagmus, unsteady gait, head motion intolerance, most commonly due to vestibular neuritis or posterior fossa stroke.

- ***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.

List of Abbreviations

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AC - Anterior Chamber
AC1 – Arnold Chiari Malformation Type 1
ACE – Angiotensin Converting Enzyme
ACG – Angle Closure Glaucoma
AD – Autosomal Dominant
ADOA – Autosomal Dominant Optic Atrophy
AIBSES – Acute Idiopathic Blind Spot Enlargement Syndrome
AIDS – Acquired Immune Deficiency Syndrome
AION – Anterior Ischemic Optic Neuropathy
AAION– Arteritic AION (GCA)
AK – Astigmatic Keratotomy
AMPPE – Acute Multifocal Posterior Placoid Epitheliopathy
ANA – Anti-Nuclear Antibody (Lupus Diagnosis)
AR – Autosomal Recessive
ARMD – Age Related Macular Degeneration
ARN – Acute Retinal Necrosis
AVM – Arterial Venous Malformation
BEB – Benign Essential Blepharospasm
BCCA – Basal Cell Carcinoma
BCP – Birth Control Pills, Contraceptives
BDUMP – Bilateral Diffuse Uveal Melanocytic Proliferation
BP – Blood Pressure
BPES - Blepharophimosis- Ptosis- Epicanthus Inversus Syndrome
BPV – Benign Positional Vertigo
BRAO- Branch Retinal artery Occlusion
BRVO- Branch RVO
CA – Carcinoma
CARS – Cancer Associated Retinopathy Syndrome
CC – Carotid Cavernous
CE – Cataract Extraction (Surgery)
CFEOM – Congenital Fibrosis of Extraocular Muscles
CHRPE – Congenital Hypertrophy of the RPE
CIN – Carcinoma in Situ
CLL – Chronic Lymphocytic Leukemia
CME – Cystoid Macular Edema
CMV – Cytomegalovirus
CN- Cranial Nerve
CNP – CN palsy
CNS – Central Nervous System
CPA – Cerebellar Pontine Angle
CPEO – Chronic Progressive External Ophthalmoplegia
CR – Chorioretinal
CRAO- central retinal *artery* occlusion, CRVO- central ret. *vein* occlusion
CSR – Central Serous Retinopathy
CSNB – Congenital Stationary Night Blindness
CT- Computerized Tomography
CTA – CT Angiogram
CTL – Contact Lens
CVA – Cerebral (CNS) Vascular Accident (Stroke)
CXR – Chest X-Ray
DDX – Differential Diagnosis
DOA – Dominant Optic Atrophy
DR – Diabetic Retinopathy
DME- Diabetic Macular Edema
DM – Diabetes Mellitus
DUSN – Diffuse Unilateral Subacute Neuroretinitis from Nematode
EOM – Extra Ocular Muscle
ERG – Electroretinogram
ERM – Epi - Retinal Membrane
ESR – Erythrocyte Sedimentation Rate
ET – Esotropia
FA- Fluorescein Angiogram
FB- Foreign Body
FBS – Foreign Body Sensation
FEVR - Familial exudative vitreoretinopathy
FHx – Family Medical History
GBS- Guillain-Barre Syndrome
GCA – Giant Cell Arteritis (aka Temporal Arteritis)

List of Abbreviations

GPA- Granulomatosis with Polyangiitis (Wegener's)
HA - Headache
HIV – Human Immunodeficiency Virus (AIDS)
HLA – Human Leukocyte Antigen
HSV – Herpes Simplex Virus
HT – Hypertropia
HoT - Hypotropia
HTN – Hypertension (high blood pressure)
HZO- Herpes Zoster Ophthalmicus
IBD - Inflammatory Bowel Diseases
ICE – Iridocorneal Endothelial (Syndrome)
ICP - Intracranial Pressure
IgG4-ROD – Immunoglobulin G4 Related Ophthalmic Disease
IIH – Idiopathic Intracranial Hypertension (pseudotumor cerebri)
INO – Internuclear Ophthalmoplegia
IO – Inferior Oblique
IOFB – Intra-Ocular Foreign Body
IOIS – Idiopathic Orbital Inflammatory Syndrome
IOL – Intraocular lens
ION- Ischemic Optic Neuropathy
IOP – Intraocular Pressure
IR – Inferior Rectus
JIA – Juvenile Idiopathic Arthritis
JRA - Juvenile Rheumatoid Arthritis
KC – Keratoconjunctivitis
KP – Keratoprecipitates
LASIK – Laser Assisted In Situ Keratomileusis
LCA – Leber's Congenital Amaurosis
LHON – Leber's Hereditary Optic Neuropathy
LG – Lacrimal Gland
LL – Lower Lid (Lower Eyelid)
LLL – Left Lower Eyelid
LP – Lumbar Puncture (Spinal Tap)
LR – Lateral Rectus
LSCD- Limbal Stem Cell Deficiency
LUL – Left Upper Lid
MARS – Melanoma Associated Retinopathy Syndrome
MEWDS – Multiple Evanescent White Dot Syndrome
MFS – Miller Fisher Syndrome (polyneuropathy)
MG – Myasthenia Gravis
MM – Multiple Myeloma
MR – Medial Rectus
MRA – Magnetic Resonance Angiography
MRD – Margin Reflex Distance (Upper Eyelids)
MRI - Magnetic Resonance Imaging
MRV- MR Venography
MS – Multiple Sclerosis
NAION- Non-Arteritic AION
NF - Neurofibromatosis
NFL – Nerve Fiber Layer
NLO – Nasolacrimal Obstruction
NLDO – NL Duct Obstruction
NPDR – Non-Proliferative Diabetic Retinopathy
NVA- Near Visual Acuity
NVD – Neovascularization of the Disk
NVI – Neovascularization of the Iris
OAG – Open Angle Glaucoma
OCP – Ocular Cicatricial Pemphigoid
OCT – Ocular Coherence Tomography
OIS – Ocular Ischemic Syndrome
OKN – Opto-Kinetic Nystagmus
ONH – Optic Nerve Head
OMG – Ocular Myasthenia Gravis
ONM – Ocular Neuromyotonia
OSSN – Ocular Surface Squamous Neoplasia
PAN – Polyarteritis Nodosa
PC – Posterior Chamber
PCO - Posterior Capsular Opacity (Pseudophakic eye)

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List of Abbreviations

PDR – Proliferative Diabetic Retinopathy
PEK – Punctate Epithelial Keratopathy = SPK
PET-CT - Positron emission tomography–computed tomography
PHPV – Persistent Hyperplastic Primary Vitreous
PPM – Persistent Placoid Maculopathy
PPV – Pars Plana Vitrectomy (Surgery)
PI – Peripheral Iridectomy or Iridotomy
PION – Posterior Ischemic Optic Neuropathy
PK – Penetrating Keratoplasty (corneal transplant)
POAG – Primary Open Angle Glaucoma
PON – Paraneoplastic Optic Neuropathy
PORN - Progressive Outer Retinal Necrosis
PRK- Photo- Refractive Keratectomy
PRP- Pan-Retinal Photocoagulation
PSC – Posterior Sub-Capsular (Cataract)
PUK- Peripheral Ulcerative Keratitis
PVD – Posterior Vitreous Detachment
RA – Rheumatoid Arthritis
RAO – Retinal Artery Occlusion
RAPD – Relative Afferent Pupillary Defect
RB – Retinoblastoma
RBC – Red Blood Cells
RD – Retinal Detachment
RES – Recurrent Erosion Syndrome
RLL – Right Lower Lid
RGP – Rigid Gas Permeable (CTL)
RK – Radial Keratotomy
RMSF- Rocky Mountain Spotted Fever (Rickettsiae)
R/O – Rule Out
ROP- Retinopathy of Prematurity
RP- Retinitis Pigmentosa
RPE – Retinal Pigment Epithelium
RUL – Right Upper Eyelid
RVO – Retinal Vein Occlusion
SAH – Subarachnoid Hemorrhage
SBS – Shaken Baby Syndrome
SCH – Sub-Conjunctival Hemorrhage
SCCA – Squamous Cell Carcinoma
SJS – Stevens Johnson Syndrome
SLK – Superior Limbic Keratoconjunctivitis
SLE- Systemic Lupus Erythematosus
SNP – Supranuclear Palsy
SO – Superior Oblique
S/P - Status Post
SPK - Superficial Punctate Keratitis
SR – Superior Rectus
SRNVM – Sub-Retinal Neovascular Membrane
TASS – Toxic Anterior Segment Syndrome
TB – Tuberculosis
TBI- Traumatic Brain Injury
TM – Trabecular Meshwork
TORCH – (Toxoplasmosis, Other, Rubella, Cytomegalovirus, Herpes Simplex)

UA – Urine Analysis
UGH – Uveitis Glaucoma Hyphema Syndrome (Pseudophakic eye AC or PC IOL)

UL – Upper Lid (Eyelid)
URI – Upper Respiratory Infection
VA – Visual Acuity
VF - Visual Field
VKH - Vogt-Koyanagi-Harada Syndrome
VMT - Vitreo -Macular Traction
VPT – Vitreo-Papillary Traction
VZV - Varicella -Zoster Virus
WBC – White Blood Cells
W/U – work up
XT - Exotropia

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Journal References

	Abbreviation Used
Ophthalmology (AAO Journal)	OPH
Survey of Ophthalmology	Survey
JAMA Ophthalmology (Archives)	JAMA Oph
American Journal of Ophthalmology	AJO
Journal of Neuro-ophthalmology	JNO
Investigative Ophthalmology and Visual Science	IOVS

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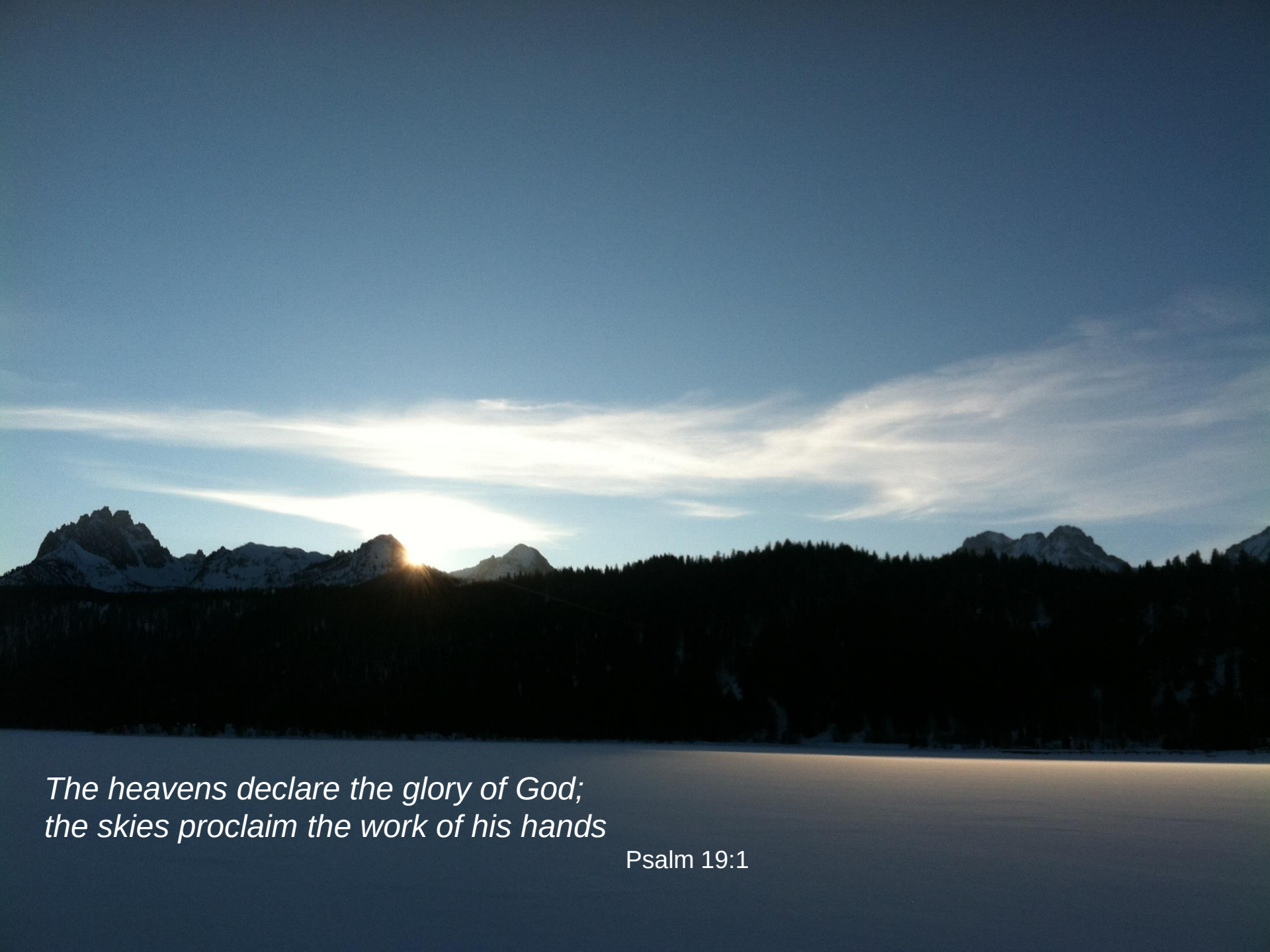
The heavens declare the glory of God;
the skies proclaim the work of his hands
Psalm 19:1