


OKAP Review
Neuro-Ophthalmology

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 NYEEI

Non-organic Visual Loss

- Severe monocular loss
 - RAPD: <20/200
 - **Duochrome Test**
 - Red lens - see both sides of chart
 - Green lens - only see green side
 - Put red lens in front of "bad eye"
 - Reads entire line better than alleged Va



Non-organic Visual Loss

- **Color plates**
 - ICP cannot be seen through green lens
 - Red lens over "bad eye"
 - At least 20/400 if can read the plates

Non-organic Visual Loss ★

- **Prism shift test**
 - 20/50 Snellen Line
 - 4 base-out in front of the "bad eye"
 - Good vision:
 - movement of both eyes towards apex followed by movement of the fellow eye back to center
 - Bad vision: no eye movement
 - with prism over "good eye", binocular shift to apex without compensatory movement of the blind eye to center

Non-organic Visual Loss

- Moderate monocular loss (20/40-100)
 - **Visual acuity testing**
 - 20/100=10/50
 - Distance=Near
 - **Fogging**
 - +6.00D sphere over the good eye
 - +4.00D and -4.00D cylinder aligned on the same axis in both eyes (neutralize each other)
 - Rotate cylinder in "good eye" to 90° of the other

Diplopia

- Binocular vs. Monocular
- Distance vs. near
- Vertical vs. horizontal
- Directionality
- Constant vs. Variable
- Associated symptoms and signs

Diplopia

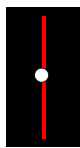
Horizontal	Vertical
CN VI	CN IV
CN III	CN III
Convergence insufficiency	TED
INO	Skew Deviation
MG	MG

Testing Diplopia

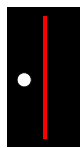
- Maddox Rod: phoria + tropia
- Placed in front of OD, distant light source



Maddox Rod ★



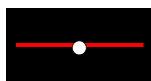
Ortho vertical



ET - uncrossed



XT - crossed



Ortho horizontal



RHT



LHT

Eye Movements

- Horizontal Saccades
 - FEF -> contralateral PPRF
- Pursuit
 - Ipsilateral occipitoparietal region
- Vertical Saccades
 - FEF -> riMLF & INC

CN III - Nuclear ★

- **Bilateral** ptosis
 - Single sub-nucleus
- Ipsilateral mydriasis
- Ipsilateral MR, IR, IO palsy
- **Bilateral** SR palsy
 - Crossed fibers pass through the contralateral SR sub-nucleus w/o synapse and innervate the **contralateral SR**

CN III - Fascicular

- *Claude syndrome*
 - CN III & contralateral ataxia
- *Weber syndrome*
 - CN III & contralateral hemiparesis
- *Benedikt syndrome*
 - CN III & contralateral tremor or involuntary limb movements (**red nucleus**)

CN III - Subarachnoid

- Uncal herniation
 - Ipsilateral pupillary dilation may be the 1st sign
- **PComA** Aneurysm
 - Early pupillary dilation
 - Usually painful
- Basilar Artery Aneurysm
 - CN III or IV

CN III - Internal Ophthalmoplegia

- PComA - ICA junction Aneurysm (early)
- Basilar Aneurysm
- Extrinsic lesions in the interpeduncular cystem (*rare*)
- Basilar meningitis

CN III - Vasculopathic

- DM, HTN, CAD, Smoking, advanced age, Hypercholesterolemia
- **CANNOT** use pain to ddx aneurysmal from vasculopathic
- Other causes include
 - GCA
 - SLE
 - HZO

CN III - Trauma

- Vulnerable to stretch and contusion
 - Posterior clinoid process posterior to CS
 - Frontal head trauma (MVA)
 - Surgery in the parasellar region

CN III - CS & SOF

- Usually cranial polyneuropathy
 - CN III
 - CN IV
 - CN VI
 - Sympathetics (small midposition poorly-reactive pupil)
 - V1, V2
- Proptosis, edema, chemosis
- Pain

CS & SOF

- Steroid responders
 - Tolosa-Hunt syndrome
 - Tumors
 - Aneurysms
- Similar signal intensity on MRI
 - THS
 - Meningioma
 - Lymphoma
 - Metastasis
 - Sarcoidosis

CN III - Orbit

- Enters as 2 divisions
 - Superior (LPS, SR)
 - Inferior (MR, IR, IO, motor root CG)
- Apex
 - optic neuropathy
 - variable proptosis
 - +/- pain

CN III - Evaluation

- *Complete pupil-sparing*
 - observe (MRI if worsen or fail to improve in 8 wks)
- *Complete or partial pupil-involving*
 - MRI/MRA
- *Superior division partial*
 - MRI

CN III - Aberrant regeneration ★

- Eyelid retraction on downgaze (pseudo-Graefe sign)
- Elevation of the eyelid on adduction
- Pupil constriction on adduction (pseudo-Argyll Robertson pupil)
- *Does NOT occur after ischemia*
- Primary aberrant regeneration
 - Rule-out slow growing lesion in the CS (imaging)

CN III - Congenital

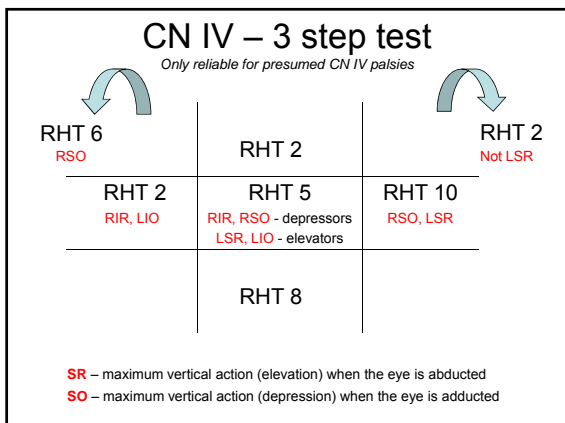
- 1/2 of CN III palsies in children
- No neurologic or systemic abnormalities
- Pupil usually miotic (regeneration ?)

Cyclic Oculomotor Paresis

- Usually congenital & unilateral
- Spasms lasting 10-30 seconds **q2 min**
- Usually persists unchanged
- Birth trauma, infection or unknown cause

CN IV Palsy

- Most common cause of acquired vertical strabismus
- Diplopia greatest in
 - Downgaze
 - To the opposite side
 - **Excyclotorsion** (especially in abduction)
 - DDx from skew deviation – higher eye **intorted**



CN IV - Bilateral

- Common with trauma
- Crossed hypertropia
- Excyclotorsion > 10°
- Large (>25D) pattern

CN IV - Congenital

- Large vertical fusion amplitudes (10-25)
 - Normal 3-6Δ
- Head tilt
- Can decompensate after mild head trauma or with age
- Review old photographs

CN IV - Acquired

- Trauma
 - Fragile nerve with a long subarachnoid course
- Compression
 - Aneurysm: Basilar – Superior Cerebellar j.
- Infiltration, ischemia, inflammation
- Parinaud’s Syndrome
- Aqueductal stenosis & hydrocephalus

CN IV - Localization ★

- Central **Horner’s syndrome**
 - Dorsal brainstem
 - Contralateral
- **RAPD** w/o optic neuropathy or tract syndrome
 - Brachium of the superior colliculus
 - Can be unilateral or contralateral

CN VI Palsy - Nucleus

- Produces a **conjugate gaze palsy** to the ipsilateral side
- Usually accompanied by ipsilateral peripheral CN VII palsy (pons)
- Ischemia, infiltration, trauma, inflammation, compression, metabolic (Wernicke-Korsakoff, hepatic encephalopathy)

CN VI Palsy

- Increased or decreased ICP
- Gradenigo's syndrome – mastoiditis with petrous bone meningeal inflammation +/- facial pain (Gasserian ganglion)
- Pterygopalatine ganglion injury
 - Loss of tearing
 - Ipsilateral eye irritation
 - V3 neuropathy
 - Usually nasopharyngeal carcinoma

CN VI – CS/SOF ★

- Within body of the sinus (not wall)
 - Damage from intracavernous lesions
- Ipsilateral 3rd order **Horner's syndrome**

CN VI - Evaluation

- Neuroimaging
 - No vasculopathic risk factors
 - Younger age (<60)
 - No recovery (3-6 months) or interval worsening
 - Any associated neurological abnormality
 - V1-V3 neuropathy
 - Facial paresis
 - Hearing loss
 - Horner's syndrome
 - Redness, swelling, proptosis

CN VII

- Nuclear Facial Palsy
 - Facial monoplegia
 - Ipsilateral gaze palsy (CN VI nucleus)
 - Frequent ataxia
 - Occasional Horner syndrome
- CPA
 - Co-involvement of CN V, VI, or VIII
- Melkersen-Rosenthal syndrome
 - Recurrent CN VII palsy
 - Recurrent facial swelling
 - Lingua plicata (furlowing of the tongue)

INO ★

- MLF
 - CN VI (pons) -> MR subdivision of the contralateral CN III (midbrain)
- Weakened ipsilateral adduction
- Contralateral abduction nystagmus
- Skew deviation
 - Ipsilateral hypertropia, intorsion
- Posterior INO
 - Convergence intact
- Anterior INO
 - Convergence affected
- Dissociated vertical nystagmus
 - Ipsilateral downbeat, contralateral torsional

1 and 1/2 syndrome ★ (paralytic pontine exotropia)

- CN VI nucleus or PPRF & MLF
- Ipsilateral gaze palsy & INO
- Preserved **contralateral abduction**

Parinaud's syndrome ★

(dorsal midbrain syndrome)

- Limitation of upgaze
- Lid retraction (Collier's sign); rarely ptosis
- Disturbance of downgaze
 - Downward gaze preference ("setting sun" sign)
 - Downbeating nystagmus
 - Impaired saccades or smooth pursuit
- Disturbance of vergence
 - Paralysis or spasm
 - Pseudo-abducens palsy
 - **Convergence-retraction nystagmus** (attempted upgaze)
 - Compression of the superior colliculus

Parinaud's syndrome ★

- Fixation instability
 - Square-wave jerks
- Skew deviation
- Pupillary abnormalities
 - Large pupils with **light-near dissociation**

Parinaud's syndrome - causes

- Tumor – pineal germinoma
- Hydrocephalus – aqueductal stenosis
- Vascular
- Metabolic
- Drugs
- Degenerative – PSP
- Other – MS, Whipple's disease

Skew Deviation

- Vertical misalignment from disturbance of prenuclear inputs/otolithic inputs
 - Vestibular periphery
 - Brainstem
 - Cerebellum
- Comitant or incomitant
- Accompanied by signs of CNS dysfx
- Maybe associated with ocular tilt reaction (**OTR**)
 - Ocular torsion (**intorsion**) and head tilt

Wallenberg's Syndrome

- Lateral medullary infarction
- Ipsilateral Vertebral A or PICA
- Ipsilateral facial pain & temperature loss
- Contralateral trunk/limbs P&T loss
- Central Horner's syndrome
- Dysarthria
- Dysphagia

Wallenberg's Syndrome

- Vertigo
- Body and environmental tilt
- Lateropulsion
 - Towards the side of the lesion
 - Ipsipulsion of saccades
 - Ipsilateral overshoot
 - Contralateral undershoot
- OTR
- Skew deviation
 - Ipsilateral hypotropia

CPEO

- Mitochondrial myopathy
- Generalized skeletal muscle weakness
- Ptosis
 - May precede ophthalmoplegia by months-years
 - Slowly progressive
 - Fixed
- Ophthalmoplegia
 - Most w/o diplopia because symmetric
 - Downgaze relatively intact relative to rest

CPEO - Kearns-Sayre syndrome ★

- Pigmentary retinopathy
 - Before age 20
 - Posterior fundus initially
 - “Salt & Pepper”
- Cardiac conduction disturbances
 - Heart block (prolonged PR on EKG)
- Cerebellar ataxia
- Short stature
- Deafness
- Dementia
- Endocrine abnormalities

Oculopharyngeal Dystrophy

- AD, chromosome 14
- French-Canadian heritage common
- Ptosis
 - Usually precedes dysphagia by years
- Dysphagia
 - Solids -> liquids
- Look at old family photos
- Ophthalmoplegia in most
- Limb-muscle weakness
 - Mild & proximal

Myotonic Muscular Dystrophy

- Frontal balding
- Ptosis
- Hollowing of masseter and temporalis
- Facial weakness
- Testicular atrophy
- Insulin resistance
- Cardiac conduction defects

Myotonic Muscular Dystrophy ★

- Ptosis
- Orbicularis weakness
- Abnormal eye movements
- Sluggish, miotic pupils
- Cataracts
 - Most common ocular abnormality
- Retinal abnormalities
 - Electronegative ERG

Myasthenia Gravis

- Reduced ACh receptors 2/2 Ab's
- Thymic hyperplasia/thymoma
- Bimodal
 - Young women, older men
- LPS, EOM, orbicularis
- Variability
- Fatigue

Myasthenia Gravis

- Ptosis
 - Fluctuating
 - Frequently absent on awakening
 - Cogan's lid twitch
 - Transient eyelid retraction
 - Usually on return from upward to primary gaze

Myasthenia Gravis

- EOM
 - No set pattern
 - MR more frequently affected
 - Mimics INO
- Pupils
 - No clinically significant dysfunction

Myasthenia Gravis - Diagnosis ★

- Sleep test/Ice test
- Tensilon Test
 - Acetylcholinesterase
 - Excess ACh at nicotinic synapses
 - Rapid onset (30 seconds), short duration (5 minutes)
 - Atropine on stand-by
- Single-fiber EMG
 - The most sensitive test
- AChR Ab
- CT chest
- Other AI disorders - thyroid

Myasthenia Gravis - Treatment

- Pyridostigmine (Mestinon)
 - Anticholinesterase
- Thymectomy
- Immunosuppressive Rx
 - Prednisone
 - Azathioprine (Imuran)
 - Cyclosporine

Miller-Fisher syndrome

- Variant of *Guillain-Barre syndrome*
- Ophthalmoplegia & ataxia
- Decreased or absent DTR's
- Elevated CSF protein
- GQ1b IgG serum antibodies
- Usually complete recovery

Ocular Neuromyotonia

- Brief episodic contractions of muscles supplied by CN 3,4,6
- Usually unilateral
- Usually h/o skull base XRT
- W/U with MRI brain
- Carbamazepine may help

Superior Oblique Myokymia

- Monocular blurring
- Tremulous sensation
- Vertical/torsional diplopia/oscillopsia
- Short-lived, recurrent
- Worse with looking in direction of SO fx
- Most have no underlying disease

Ocular Motor Apraxia ★

- Impaired ability to generate saccades
- COMA
 - Thrusting horizontal head movements on attempted change of fixation
 - +/- prominent blinking, eye rubbing
 - **Vertical** eye movements are **normal** in the **congenital** form
 - May have associated CNS anomalies
- Most **acquired** cases cause defects in horizontal & vertical planes

Progressive Supranuclear Palsy (PSP) ★

- Degenerative disease affecting tone and posture
 - Blepharospasm
 - Apraxia of eyelid closing or opening
 - Lid retraction/lag
- Ocular motor defects
 - Impaired vertical saccades (down > up)
 - Square wave jerks
 - May progress to complete ophthalmoplegia

Wernicke's Encephalopathy

- Ophthalmoplegia
- Mental confusion
- Ataxia
- Thiamine deficiency

Nystagmus

- Described according to the quick phase direction
- It is the slow phase that reflects the underlying abnormality
- Retinal, optic nerve, CNS, vestibular etiologies

Gaze-Evoked Nystagmus

- Cannot maintain eccentric gaze
- Etiologies
 - Advanced age
 - Metabolic/Toxic (EtOH, anticonvulsants)
 - BS/Cerebellar lesion (esp. if assymmetric)
 - EOM myopathies
 - MG
 - Extremes of gaze in normals
- Dysfunctional neural integrator
 - Nucleus prepositus hypoglossi near the medial vestibular nucleus at the pontomedullary junction

Acquired Pendular Nystagmus

- Horizontal, vertical and torsional components - one may predominate
- MS (Jell-O)
- Visual loss
 - Unilateral ON disease
- Oculopalatal myoclonus
 - Pendular nystagmus with palatal tremor
 - Often vertical
 - May resemble see-saw nystagmus
 - Brainstem or cerebellar CVA, degenerative
 - Hypertrophy of inferior olivary nucleus

Vestibular Nystagmus

	Peripheral	Central
Vertigo	Severe	Mild
Tinnitus/Hearing loss	Common	Absent
Horizontal with torsion	Typical	Not typical
Pure vertical or torsional	Almost never	Diagnostic
Visual fixation	Damps	No effect
Common causes	Labyrinthitis, Meniere dz	Demyelination, vascular, drugs

Downbeat nystagmus

- Usually in primary position
- Greatest in downgaze (least in upgaze)
- Accentuated in downgaze to the side
- Abnormal smooth pursuit
- Vertical diplopia if associated with skew
- Rx:
 - Clonazepam
 - Baclofen
 - Base-out prisms (induce convergence)

Downbeat nystagmus ★

- C/O oscillopsia, postural instability, difficulty reading
- Etiologies:
 - Cervical-medullary junction (Arnold-Chiari)
 - Cerebellum
 - MS
 - Toxic-metabolic
 - Anticonvulsants
 - Lithium
 - B12 deficiency
 - ETOH

Upbeat Nystagmus

- Greatest in upgaze
- Poor localizing value (posterior fossa)
 - Medulla, Midbrain or Cerebellum (vermis)
- MS
- LCA
- Organophosphate poisoning
- Middle Ear disease
- Congenital

Periodic Alternating Nystagmus (PAN)

- Horizontal
- Primary gaze
- **4 min** cycle, reverses direction every **2 min**
- Etiologies:
 - Chiari malformations
 - MS
 - Cerebellar degeneration or mass
 - BS infarcts
 - Anticonvulsant meds
 - Hepatic encephalopathy
 - Congenital

See-Saw Nystagmus

- Elevation/Intorsion (**UP/IN**) with simultaneous Depression/Extorsion (**DOWN/OUT**)
- Etiologies:
 - **Parasellar masses** (Craniopharyngioma)
 - Septo-optic dysplasia
 - BS stroke
 - Chiari malformations
 - RP
 - Congenital

Congenital Nystagmus

- Recognized in first few months
- Almost always conjugate
- Horizontal + small torsional component
- Can appear as jerk or pendular
- Null point is common - head turn
- Fixation **amplifies** the nystagmus
- **Reversal of OKN** (slow phase in the opposite direction of the drum rotation)
- Absent in sleep

Latent Nystagmus

- Horizontal jerk
- Only in **monocular** viewing conditions
- Fast phase towards the viewing eye (away from occluded eye)
- Almost always associated with strabismus (ET>XT)
- Manifest LN - binocular viewing

Spasmus Nutans

- Bilateral intermittent, horizontal, low-amplitude, rapid ("shimmering")
- Often disconjugate
- Head nodding
- Abnormal head posture/torticollis
- Usually resolves completely by age 6
- **DDX: chiasmal glioma -> get MRI**

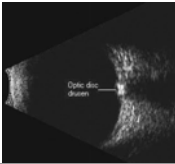
Opsoclonus ★

- Multidirectional (saccadomania)
- Flutter if horizontal, small-amplitude, high-frequency
- Etiologies:
 - Paraneoplastic process
 - Neuroblastoma in children
 - Lung, breast, ovarian cancer in adults
 - MS
- Work-up:
 - Anti-Ri, anti-Hu antibodies

Whipple's Disease

- Mimics PSP
- Abnormal vertical saccades first
- Later, may lose all movements
- **Oculomasticatory myorhythmia**
 - Pendular oscillations
 - Vertical saccade palsy
 - Contractions of masticatory muscles

Optic Nerve Drusen

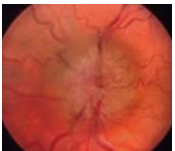


- Calcium deposits
- 0.3-2.0% of the population
- 2/3 bilateral
- AD inheritance
- **Angioid streaks and RP**
- Diagnosis
 - Autofluorescence
 - Red-free photograph
 - OCT
 - A- and B-scan U/S
 - CT scan

ON Drusen

- VF Defects in 75%
 - Arcuate, Quadrantic, Sector
 - BS enlargement
 - Concentric constriction
- +/- RAPD
- Acuity loss - extremely rare
- Pre- or peripapillary hemorrhages
- Serous retinopathy
- ION
- Retinal vascular occlusions
- Transient visual obscurations
- Peripapillary subretinal NV

Papilledema



- By definition, from raised ICP
- VF defects
 - Enlarged BS
 - NLF defects (nasal, arcuate)
- Acuity & color vision usually normal
- Chronic papilledema
 - NFL gliosis
 - Optociliary shunt vessels
 - Refractile bodies of the disc

Papilledema

- Patients c/o:
 - TVOs, often with orthostatic changes
 - Headache
 - N/V
- Etiologies
 - Mass
 - Hydrocephalus
 - CNS infection
 - CNS infiltration (granulomatous or malignant)
 - IIH
 - Dural sinus thrombosis

Idiopathic Intracranial Hypertension (PTC)

- 3rd decade, overweight, obese females
- Si/Sx of elevated ICP
 - Headache
 - Pulsatile tinnitus
 - TVOs
 - CN 6 palsy (non-localizing)
- with:
 - Normal MRI (empty sella)
 - High OP on LP (>25 mmHg) with normal CSF studies
 - No secondary cause for elevated ICP

IIH ★

- Secondary causes
 - Endocrine dysfunction
 - Dural sinus thrombosis
 - Pregnancy
 - Vitamin A toxicity
 - Medications
 - Tetracycline
 - Nalidixic Acid
 - Cyclosporine
 - Use of or withdrawal of steroids
 - OCP's

IIH ★

- Management
 - Weight Loss (6-10% body weight)
 - Diamox
 - High-dose corticosteroids
 - LPS
 - With intractable H/A
 - ONSF
 - With progressive/severe vision loss, does not treat the H/A

ON Hypoplasia



- “Double-ring” sign
- 20/20 to NLP
- Astigmatism
- Maybe sectoral
- Septo-optic dysplasia
 - **Get MRI**
- Maternal IDDM
 - Superior segmental hypoplasia
- Fetal Alcohol Syndrome
- Maternal anticonvulsants, EtOH, drugs, quinine
- Fetal CMV or Hepatitis B

De Morsier Syndrome ★



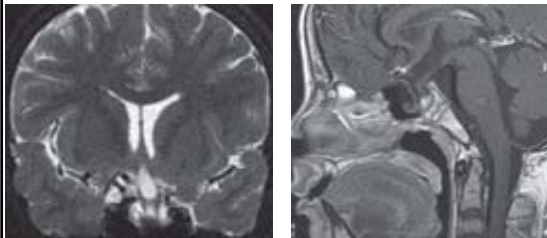
- Small anterior visual pathways
- Absence of septum pellucidum
- Thinning/agenesis of the corpus callosum
- Anterior pituitary hormone deficiencies
 - May evolve over time
 - **Endocrine evaluation**
 - GH most common
 - Risk of sudden death during febrile illness (corticotropin deficiency)
- **Get MRI** with unilateral or bilateral ON hypoplasia

Morning Glory Disc Anomaly

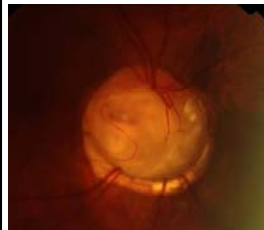


- Usually unilateral
- < 20/200
- Females
- Transsphenoidal basal encephalocele
 - Associated with other midline anomalies
- Hypopituitarism
- Serous RD in 25-40%

Transsphenoidal basal encephalocele



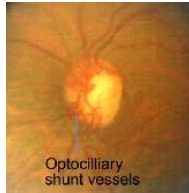
ON Coloboma & Pit



- Coloboma
 - VF defects
 - +/- RAPD
- Pit
 - Mild VF defect
 - Paracentral or arcuate
 - SRD 25-75%

Optociliary Shunt Vessels

- Congenital
 - Connections between the retinal & choroidal circulation
- Chronic ON compression
 - Shunt: retinal -> choroidal venous circulation (bypass obstructed CRV)
- Etiologies:
 - Spheno-orbital meningioma
 - Chronic papilledema
 - Optic glioma
 - ON arachnoid cysts
 - Craniopharyngioma
 - CRVO
 - Orbital vascular malformations



Foster Kennedy Syndrome

- Intracranial lesion producing
 - (Ipsilateral) optic atrophy
 - (Contralateral) papilledema
- Frontal lobe tumors and olfactory groove meningiomas
- Much more common to see pseudo-FKS
 - Non-simultaneous ischemic optic neuropathy
 - Non-simultaneous optic neuritis

NAION

- Painless monocular vision loss, often in AM
 - Compromise of ON microcirculation, “crowding”
- VF loss
 - Inferior arcuate/altitudinal defects
- RAPD
- ON edema -> atrophy in 6-10 weeks
 - Hyperemic and diffuse most common (can be pale and segmental)
- “Disc at risk”
- Contralateral 5-year risk 12-19%
 - “pseudo Foster Kennedy syndrome”
- Delayed IVFA disc filling

NAION

- Risk factors
 - “disc at risk”
 - DM
 - HTN
 - Smoking
 - Vasculitis
 - Hyperlipidemia
 - CRI/dialysis
 - OSA
- 43% improve at least 3 lines
- No proven Rx

GCA

- Older patients
- F>M
- Inflammatory & thrombotic occlusion of the short posterior ciliary vessels
- H/A, temporal/scalp tenderness, jaw claudication, malaise, anorexia, weight loss, fever, joint pain, ear pain
- “Occult” in up to 20%
- May be preceded by amaurosis fugax

GCA

- Exam
 - CWS
 - Pallid ON edema
 - Choroidal ischemia
 - CRAO
- Work-up
 - Elevated ESR (also rises with age, anemia, infection, DM, malignancy), CRP, platelets
 - ESR *Males*=age/2; *Females*=(age+10)/2
 - ESR + CRP >97% specificity
 - TAB
 - >2.5cm specimen bc of “skip lesions”
- Treatment
 - High-dose steroids w/o delay

Transient Monocular Visual Loss (Amaurosis Fugax)

- Ocular
 - ACG or hyphema
 - Palledema
 - Optic disc drusen
- Orbital
 - Mass (gaze-evoked amaurosis)
- Systemic (ischemia or vascular insufficiency)
 - Emboli (carotid, cardiac, great vessels)
 - Vasculitis (GCA)
 - Hypoperfusion (carotid stenosis or dissection)
 - Hyperviscosity or Hypercoagulability

Transient Monocular Visual Loss (Amaurosis Fugax)

- Evaluation
 - Complete exam including gonioscopy
 - Auscultation for carotid bruit
 - Carotid U/S
 - Echocardiography
 - Halter monitor
 - Laboratory studies

Optic Neuritis

- Pain (maybe absent if intracranial or intracranial)
 - Preceding or concurrent with vision loss
 - Worse with EOM
- Photopsias
- Uhthoff phenomenon (not specific)
- Loss of central vision
- Dyschromatopsia (dysproportionate to acuity)
- VF loss
- RAPD
- Disc swelling (Papillitis - 30%)
- Vitritis
- Retinal venous sheathing
 - Pts with uveitis or retinal phlebitis have increased risk of developing MS

Optic Neuritis - MRI

- Establish the diagnosis if clinically unclear
- Prognosis
 - Risk of MS within 5 years
 - 16% with normal MRI
 - 37% with 1 or 2 lesions
 - 51% with more than 2 lesions

Optic Neuritis - Prognosis

- Visual worsening over <2 weeks followed by improvement
- Improvement (w/o treatment) within
 - 3 weeks in 79%
 - 5 weeks in 93%
 - Nearly complete by 5 weeks after onset
- <10% with <20/40

Optic Neuritis - ONTT

- Randomized to
 - PO prednisone x 14 days
 - IV steroids x 3 days then PO x 11 days
 - PO placebo x 14 days
- 1 year after onset - no difference in vision, contrast sensitivity, color or VF
- IV/PO group recovered vision faster (by 2-3 weeks, greatest benefit if begun in first 15 days)
- PO group had increased risk of recurrent ON OU
- IV/PO group had a reduced rate of clinically definite MS during the first 2 years (benefit only seen with abnormal MRI)

Optic Neuritis - other causes

- Post-viral & post-vaccination
 - Onset in 1-3 weeks
 - Typically bilateral and simultaneous
 - More common in children
- Vasculitis
 - SLE, PAN
- Granulomatous
 - Syphilis
 - Sarcoid
- Infectious
 - Lyme disease
 - HIV and cryptococcal meningitis, CMV, HSV, TB

Optic Neuritis - children

- More often anterior - swelling in >70%
- More often bilateral simultaneous (60%)
- 1-2 weeks after viral infection or vaccination
- Less often associated with MS
- Often steroid-sensitive & steroid-dependent

Papillophlebitis

- Subset of CRVO in the young with prominent disc edema
- Unilateral blurring with occasional TVO
- Vision & color usually normal (unless macular edema)
- No RAPD
- Enlarged BS on VF
- Exam
 - marked venous engorgement & hyperemic disc edema
 - +/- retinal hemorrhages
- Resolves spontaneously in 6-12 months
- Rarely associated with systemic vasculitis

Optic Neuropathy - Toxic ★

- Gradually progressive
- Bilateral
 - Maybe asymmetric early
- Painless
- Reduced vision (20/50-200)
- Dyschromatopsia
- Mild-moderate disc swelling
- Optic atrophy (temporal pallor)
- VF defects
 - Cecocentral

Optic Neuropathy - Toxic

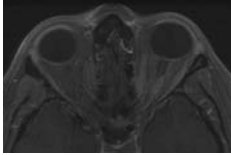
- Methanol & Ethylene glycol
 - Rapid, severe, prominent disc edema
- Deficiencies
 - Vitamin B12 (pernicious anemia)
 - Folate
 - Thiamine
- Ethambutol
 - May improve after withdrawal
 - Check Zinc level

Ethambutol	INH	Chloramphenicol
Chloroquine	Digitalis	Vincristine, Cisplatin
Lead, Mercury	Penicillamine	Amiodarone ?

Infiltrative Optic Neuropathy

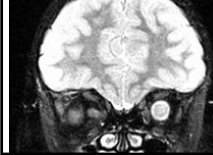
- Inflammatory or neoplastic
- Progressive vision loss
- ON maybe normal or swollen
- Etiologies
 - Meningioma
 - Glioma
 - Leukemia or Lymphoma
 - Metastatic (breast, lung)
 - Granulomatous
 - Sarcoid
 - Syphilis
 - TB
 - Fungal

Optic Nerve Meningioma



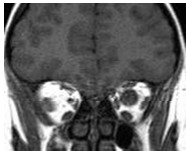
- F>M, 40-50, increased in NF1
- Painless vision loss
- Optic atrophy or edema
- Optociliary shunt vessels
- RAPD
- VF defects
- Imaging
 - Tubular ON enlargement
 - Thick enhanced sheath "tram track"
 - Calcification of the nerve sheath
 - Adjacent bony hyperostosis
- Management
 - XRT

Optic Nerve Glioma



- Most common ON tumor
- 70% 1st decade, 90% 2nd
- NF-1 (maybe more benign)
- Proptosis
- Vision loss
- ON pallor
- ON edema
- Strabismus

Optic Nerve Glioma ★



- Imaging
 - Fusiform ON enlargement
 - Thickening of ON & sheath
 - Kinking or buckling
 - Cystic spaces within the ON
 - No extradural extension
 - No calcification
 - Iso- or hypointense on T1
 - Hyperintense on T2
- Management
 - Observation for intraorbital tumors
 - Surgery for
 - Disfiguring proptosis, severe vision loss
 - Poor vision & intracranial extension

Traumatic Optic Neuropathy ★

- Direct
 - ON avulsion
 - Compression
- Indirect
 - Often frontal head injury
 - Intracanalicular tethered point
- Management is controversial

Dominant Optic Atrophy

- Most common hereditary neuropathy
- Chromosome 3
- First decade
- Insidious onset
- Bilateral, maybe asymmetric
 - 20/60-200
 - Color loss (tritanopia: blue-yellow color blindness)
- VF defects
 - Central/Cecocentral
- Optic atrophy
 - Focal temporal pallor (wedge-shaped temporal excavation)

Lebers Hereditary Optic Neuropathy

- Circumpapillary telangiectatic microangiopathy
- NFL swelling around disc (pseudopapilledema)
- No disc leakage on IVFA
- Classic triad in up two 60% of cases
- Evolves to temporal pallor and NFL dropout

LHON

- Mutations:
 - 11778 makes up 85-90%
 - 14484 & 346010-15%
- All mutations present the same
- 50% bilateral presentation
- Age range 2 - 80, peak 15 -35 years
- 90% cases in USA are male
- Women present later than men
- Vision hits nadir typically by 2 months
- Average interval between eyes 3 months, 97% bilateral by 1 year

LHON

- Relatively spared pupillary light reflexes
- Vision typically worse than 20/200
- Cecocentral scotomas
- Recovery
 - up to 50% in 14484
 - least (4%) in 11778
- 23% of eyes 20/50 or better finally

Visual Fields

- Static - varying intensity (brightness & size)
 - Goal: to determine threshold sensitivity
- Kinetic - fixed intensity
- All points of equal threshold connect to form an isopter

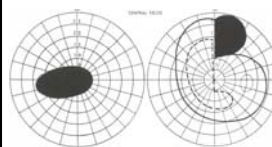
Visual Fields

- Central macular fibers are sensitive to:
 - Toxic
 - Metabolic
 - Hereditary
 - Compressive
 - Inflammatory insults

Hemi-field Slide Phenomenon ★

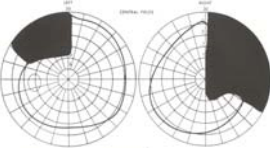
- Bitemporal hemianopic field defects
- Heteronymous altitudinal or broad arcuate defects
- Binocular diplopia
- Loss of the normal partial VF overlap
- Decompensation of a phoria

VF Chiasm - Anterior



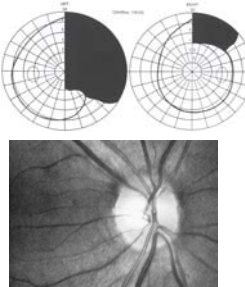
- Junctional Scotoma
- Damage to the ON & anterior chiasm compression
 - Unilateral vision loss
 - Contralateral superior temporal defect

VF Chiasm ★



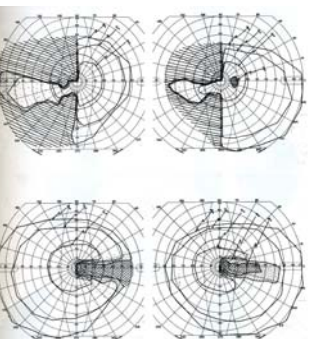
- Pituitary adenoma
 - Enlarge in pregnancy
 - Prolactinoma most common
- Parasellar meningioma
 - Enlarge in pregnancy
- Craniopharyngioma
 - Bimodal age distribution
 - Inferior VF loss
- ICA/ACoM A aneurysm
- Chiasmatic glioma
- 3rd ventricle dilation

VF - Optic Tract



- Stroke
- Mass lesions
- Optic tract syndrome
 - Incongruous contralateral homonymous hemianopia
 - Homonymous retinal NFL & ON atrophy
 - Contralateral "bow-tie atrophy"
 - Mild contralateral RAPD

VF - LGN



Quadruple Sectoranopia
Anterior Choroidal Artery Infarct

Horizontal Homonymous Sectoranopia
Lateral Choroidal Artery Infarct

LGN

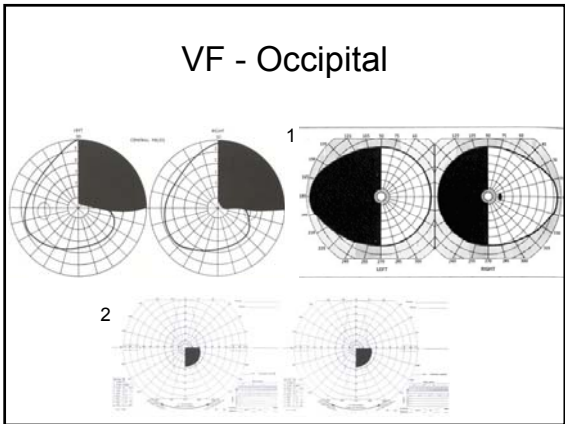
- Most compact portion of the visual system
- Divided into 6 levels
 - 4 superior levels: parvocellular axons
 - Ganglion cells for spatial resolution & color
 - 2 inferior levels: magnocellular axons
 - Ganglion cells for motion

VF ★

- Temporal Lobe
 - Usually tumors
 - Pie in the sky
 - Seizure activity
 - Olfactory & formed visual hallucinations
- Parietal Lobe
 - Usually vascular
 - Contralateral inferior homonymous hemianopia
 - Gersmann syndrome
 - Acalculia, agraphia, finger agnosia & R-L confusion

VF - Occipital

- Posterior cerebral A occlusion
 - Macula-sparing homonymous hemianopia (1)
 - Occipital tip receives dual blood supply
 - Macula supplied by the middle cerebral A
- Systemic hypotension
 - Homonymous paracentral hemianopic scotoma (2)
- Cerebral Blindness
 - Bilateral cerebral lobe dysfunction



- ### Anisocoria
- Dark vs. Light
 - Dark>Light
 - Horner Syndrome
 - Posterior Synechiae
 - Physiologic
 - Pharmacologic
 - Light>Dark
 - CN III
 - Adie's Tonic pupil
 - Trauma to the sphincter
 - Pharmacologic
 - Light & Near reactivity
 - Old photographs

- ### Physiologic Anisocoria
- Most common cause of anisocoria
 - More common in elderly
 - r/o another cause in kids
 - Usually <1.0 mm
 - Dark = or > Light

- ### Horner Syndrome
- Lesion in the oculosympathetic pathway
 - 1st order/central: Hypothalamus to C8-T2
 - 2nd order/pre-ganlionic: C8-T2 to superior cervical ganglion
 - 3rd order/post-ganlionic: ICA (carotid plexus), CS (CN VI), NC branch of V1
 - Ptosis
 - Upper & lower eyelids
 - Miosis
 - Dilation lag
 - Denervation hypersensitivity
 - Iris heterochromia when congenital (lighter)
 - Anhidrosis
 - Central - face
 - Pre-ganlionic - head, face, neck to clavicle
 - Post-ganlionic - absent or forehead only

	Localizing test	Function	Effect	Cause
HS	10% Cocaine	Blocks NE re-uptake	Fails to dilate	
1st order	None			Vascular occlusions (lateral medulla), tumors, cervical disc disease
2nd order	1% Paradrine	Releases NE from the pre-synaptic terminal	Dilates	Apical lung tumors, metastasis, chest surgery, thoracic aortic aneurysm, trauma
3rd order	1% Paradrine		Fails to dilate	Carotid artery disease/dissection), Aneurysms, tumors extending to the skull base, CS or orbital apex, cluster H/A, GCA

Horner Syndrome

1st order	numbness, weakness, ataxia, nystagmus, Wallenberg's syndrome, contralateral CN IV palsy
2nd order	cough, hemoptysis, neck swelling
3rd order	facial numbness, CN VI palsy, neck pain, dysgeusia, visual obscurations or loss
Childhood	neuroblastoma

Tonic Pupils

- Post-ganglionic parasympathetic pupillomotor damage
- Light-Near dissociation
 - Poor response to light
 - Better response to near but may be impaired initially
- Slow (tonic) re-dilation
- Regional palsy on SLE
- Unilateral in 80%, second pupil involved 4% per year
- Cholinergic supersensitivity (pilo 0.1%)
- “little old” Adie

Holmes-Adie Syndrome

- Tonic pupil
- Decreased DTRs
- Orthostatic hypotension
- 70% female

Tonic pupils - systemic

- Herpes Zoster & Varicella
- GCA
- Syphilis
- Orbital trauma
- Dysautonomia
 - Cancer
 - Amyloidosis

Internal Ophthalmoplegia - CN III

- Superficially located pupillary fibers in subarachnoid portion
 - Meningitis: may cause isolated pupillary involvement
 - Aneurysms: usually with external ophthalmoplegia
 - PCom A
 - Basilar A
- Isolated damage to EWN causes bilateral pupillary abnormalities
- Other locations most commonly associated with motility disturbances

LND Cause	Location	Mechanism
Loss of afferent input	Anterior visual pathway	Damage to the retina, ON, chiasm
Loss of input into EWN	Tectum of the midbrain	Compression (Perinaud's - dilated pupils), infection (Argyll Robertson - miotic)
Adie syndrome	Ciliary ganglion	Aberrant reinnervation
CN III aberrant reinnervation	Course of CN III	Damage to long ciliary nerves
PRP	Long ciliary nerve	Aberrant reinnervation

Argyll Robertson Pupil

- Tertiary syphilis (dx with CSF studies)
- Small (<2 mm) & irregular
- Dilate poorly with mydriatics
- Iris atrophy
- Argyll Robertson-like pupils
 - DM
 - Alcoholism
 - Encephalitis
 - After PRP

Hemifacial Spasm

- Unilateral episodic spasm of the facial musculature
- Abnormalities of the CN VII at the root entry zone
 - Compression by abnormal vessels at CPA
 - Tumor at CPA
 - Previous CN VII injury
- Treatment
 - Botox
 - Clonazepam, Baclofen, Carbamazepine
 - Janetta procedure

Essential Blepharospasm

- Progressive bilateral episodic contraction of the orbicularis muscle
- 40-60 years old
- *Meige syndrome* - involvement of other facial muscles
- Exact cause unknown (? Basal ganglia)
- Maybe seen with extrapyramidal d/o
- Rule out reflex blepharospasm
 - Dry eye, intraocular inflammation
- Treatment
 - Botox