SYLLABUS

TC50: Neurological Visual Disturbances

Wednesday, November 16, 2011 14:30 - 18:00 Hall F

Marrakesh, Morocco, November 12-17, 2011

XXth WORLD CONGRESS OF NEUROLOGY

XXth WCN 2011 MARRAKESH World Congress of Neurology

WFN

SMN SOCIETE MAROCAINE DE NEUROLOGIE

WCN Education Program

Wednesday, 16 November, 2011

14:30-18:00

NEUROLOGICAL VISUAL DISTURBANCES

Chairperson: **Tulay Kansu**, *Turkey*

14:30 **VISUAL LOSS**
Tulay Kansu, *Turkey*

15:00 **CASES BY FACULTY**

15:30 **OPTIC NEUROPATHIES**
Nancy J. Newman, *USA*

16:00 *Coffee Break*

16:30 **PAPILLEDEMA**
Valerie Biousse, *USA*

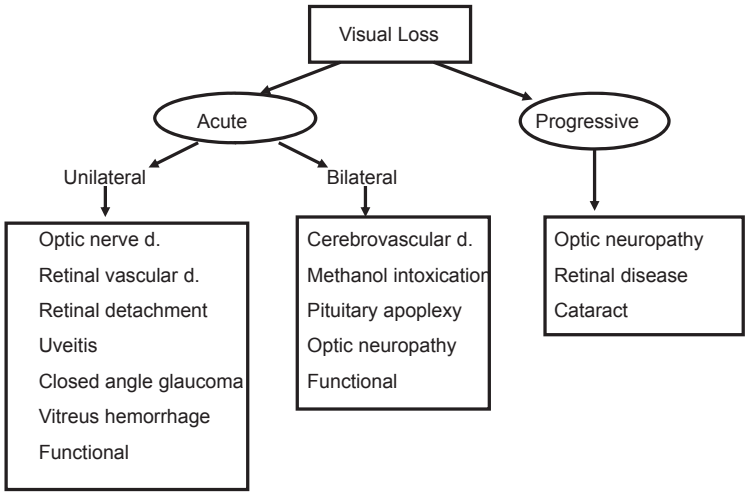
16:55 **VISUAL FIELDS**
Nancy J. Newman, *USA*

17:15 **ANISOCORIA**
Valerie Biousse, *USA*

17:30 **CASES, QUESTIONS**

VISUAL LOSS

Tulay Kansu
Hacettepe University Medical School
Department of Neurology
Neuro-Ophthalmology Unit
Ankara, Turkey



Approach to the patient with visual loss

– History

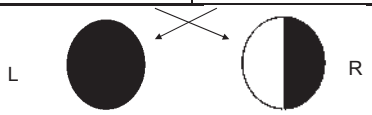
- Unilateral, bilateral
- Acute, progressive
- Central, peripheric
- Transient, permanent
- Painful or not

– examination

- Visual acuity
- Color vision
- Visual fields
- Swinging light test-RAPD?
- Eyeground

1. 58 yo, M, bilateral acute visual loss, severe headache

	R	L
Visual acuity	20/50	Hand motion
Color vision	2/12	0
Eye movements	full	Limited in and up
Light reflex	+	- RAPD+
Fundus	Pale disc	Pale disc
Visual field	Temporal h	complete



Pituitary apoplexia

- Hipophyseal tumor + infarct or hemorrhage
- Normal hipophysis + blood loss, hypotension, thrombosis, ischemia
- Tx: hormon replacement, electrolyte balance, surgical decompression

Murad-Kejbou S, Eggenberger E. Curr Opin Ophthalmol. 2009

2. 62 yo, M, sudden onset difficulty seeing objects on the left

- VA 20/30 20/30
- Color vision 12/12 12/12
- Eye movements Full
- Light reaction + / +
- Fundus Normal
- Visual fields Left homonim hemianopsia w central sparing

3. 75 yo,W, w acute visual loss. She hits the objects but can read the near card

	R	L
VA	20/40 (near)	20/40 (near)
Color vision	Identifies colors	Identifies colors
Eye movement	full	full
Light reflex	++	++
Fundus	Normal	Normal
Visual field	Tubular	Tubular

Tubuler vision (peripheral constriction)

- Bilateral occipital infarct
- Functional

- Advanced glaucoma
- Retinitis pigmentosa

4.68 yo M, w seizure, headache, bilateral visual loss

- Hx of headache for 2 days, visual blurring, nausea, vomiting and a generalized seizure.
- Hypertensive for 10 yrs.
- Somnolent, not cooperative for visual examination. Fundus normal.
- BP: 230 / 75 mm Hg

Posterior Reversible Encephalopathy Syndrome (PRES)

- Reversible Posterior leukoencephalopathy Syndrome (RPLS)
- 39 % present with visual symptoms

Feske SK. Semin Neurol. 2011

5. 73 yo, W, Visual loss noted by relatives
She hits the objects but denies blindness

	R	L
VA	Doesn't follow	light or objects
Color vision	-	-
Eye movements	Full	Full
Light reflex	++	++
Fundus	Normal	Normal
Visual field	Not	cooperated

Cortical blindness

- Ischemia (Bilateral infarct)
- Hypoxia, anoxia (CO)
- Toxic (angiography, contrast?)

- Denial of blindness (Anton syndrome)

Fraser JA, Newman NJ, Biousse V.Disorders of the optic tract, radiation, and occipital lobe. Handb Clin Neurol. 2011

6.38 yo,M, Bilateral visual loss noted on awakening

- VA: Hand motion
- Fundus: Minimal disc edema
- NE normal

Methanol Intoxication

- Loss of vision (bilateral optic neuropathy)
- Putaminal necrosis, death
- Formaldehide and formic acid accumulation
- Treatment
  - Competetive inhibitors of alcohol dehydrogenase
    - Ethyl alcohol: (0.8-2 ml/kg/hr, IV)
    - Fomepizol (Antizol): 15 mg/kg IV slow infusion X 2
  - Hemodialysis

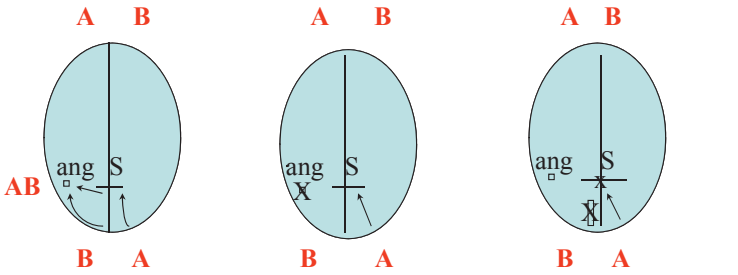
Acute blindness and putaminal necrosis in methanol intoxication. Int Ophthalmol. 1998

7.48 yo M, suddenly unable to read

• VA	20/20	20/20
• Color V	9/12	9/12
• VF	Right homonim hemianopsia	
• EM	Full	
• Pupil	LR + / +	
• Fundus	Normal	

Alexia

ang: anguler gyrus
S: Splenium of Corpus Callosum



Cortical visual disturbances

Cortical blindness
Blindsight
Riddoch phenomena
Visual ataxia
Achromatopsia
Akinetopsia
Hallucinations

Dorsal pathway
Balint syndrome
Visual neglect
Visual allesthesia
Upside-down vision

Ventral pathway
Agnosia
Prosopagnosia
Alexia
Color anomia
Visual amnesia

Barton JJ. Disorders of higher visual processing. Handb Clin Neurol.2011

Transient visual loss How to approach?

>50 yo
- **Carotid artery stenosis**
  - **Hypertension**
  - **Atherosclerosis**
  - **Diabetes, hyperlipemia, smoking**

<50 yo
- **Cardiac (arrhythmia, mitral stenosis)**
- **Oral contraceptives**
- **Migraine**
- **Coagulopathy**

>70 yo
- **Temporal arteritis**

Thurtell MJ, Rucker JC. Transient visual loss. Int Ophthalmol Clin. 2009

Functional bilateral visual loss

- OKN
- Mirror test
- Finger to finger
- Hand writing
- VEP can be misleading

Miller NR. Functional neuro-ophthalmology. Handb Clin Neurol.2011
Bruce BB, **Newman** NJ. Functional visual loss. Neurol Clin. 2010

Unexplained visual loss

- Ocular
  - Anterior segment
  - Retinal diseases
- Neuro-ophthalmic
  - Amblyopia
  - Retrobulbar and intracranial
- Functional

Griffiths PG, Ali N. Medically unexplained visual loss in adult patients. Curr Opin Neurol. 2009

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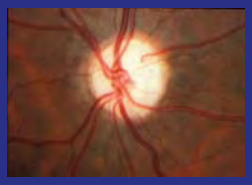
Optic Neuropathy Update

Nancy J. Newman, MD
Emory University
Atlanta, GA, USA

Most illustrations are from:
Biousse V, Newman NJ.
Neuro-Ophthalmology Illustrated. Thieme 2009.
No conflict of interest

Optic Neuropathy

Classic Features



- Decreased visual acuity
- Abnormal visual field
- Relative afferent pupillary defect
- Can see through to the nerve
- Swollen or pale optic nerve

Optic Neuropathy

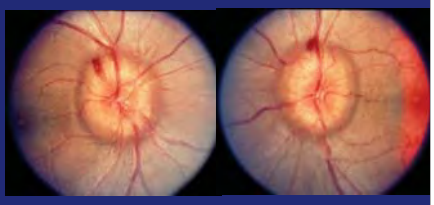
Causes

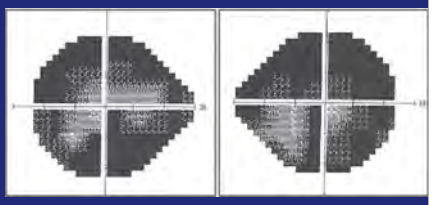
- Inflammatory
- Vascular
- Compressive/Infiltrative
- Toxic/Nutritional
- Hereditary
- Traumatic
- Elevated intracranial pressure
- Elevated intraocular pressure

Optic Neuropathy

Papilledema

- Disc swelling from ↑ intracranial pressure
- Any age
- Painless
- Bilateral
- Spares visual acuity
- Constriction of visual field

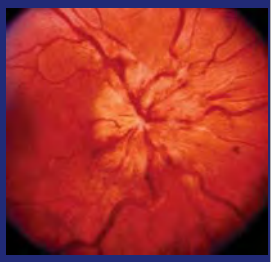




Papilledema

Causes

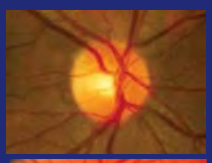
- Intracranial mass lesions
- Hydrocephalus
- Meningeal processes
- Cerebral venous thrombosis
- Idiopathic intracranial hypertension (pseudotumor cerebri)

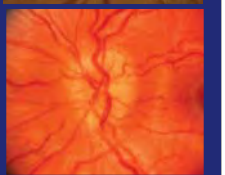


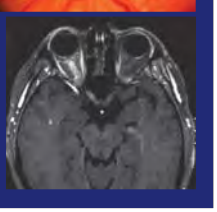
Optic Neuropathy

Typical Optic Neuritis

- Inflammation of the optic nerve
- F:M 3:1
- Age: 15-45
- Pain on eye movement
- Normal or swollen disc
- Spontaneous improvement
- Associated with multiple sclerosis





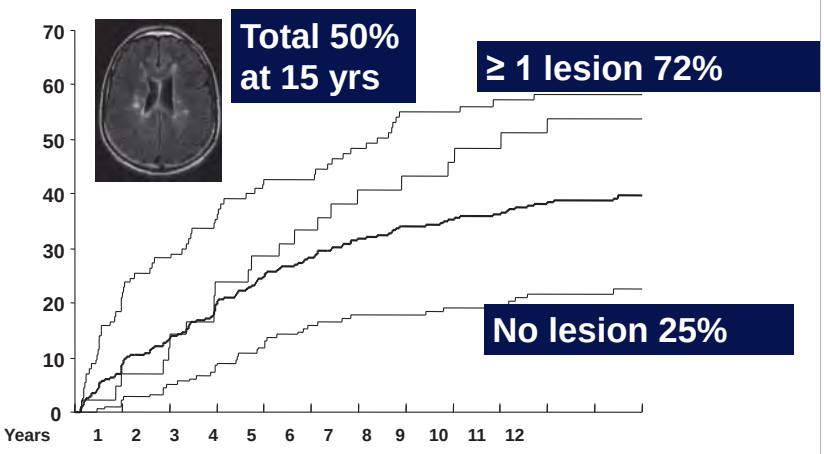


ONTT

- No difference in visual acuity between steroid and placebo groups at 6 months.
- I.V. steroids may accelerate recovery by 2 to 3 weeks.
- Oral prednisone (1mg/kg/d) doubles risk of recurrence

(NEJM 326:581, 1992)

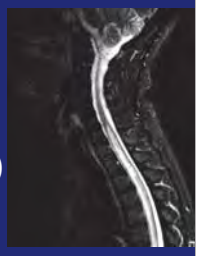
ONTT: MRI predicts the risk of MS



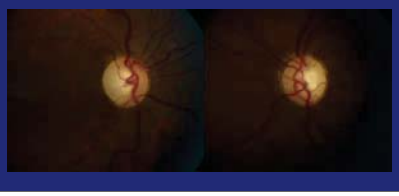
Monosymptomatic Pts and Delay to CDMS

- CHAMPS: IFNβ-1a (Avonex®)
  - –30 µg IM qwk X 2 yrs (35% vs 50%)
- ETOMS: IFNβ-1a (Rebif®)
  - –22 µg SC qwk X 2 yrs (34% vs 45%)
- BENEFIT: IFNβ-1b (Betaseron®)
  - –250 µg SC qod X 2 yrs (28% vs 45%)
- PreCISe:Glatiramer acetate (Copaxone®)
  - 20 mg SC qd X 2 yrs (25% vs 43%)

Neuromyelitis Optica (Devic)



- Severe uni- or bilateral optic neuritis
- Transverse myelopathy
- Abnormal cervical spine MRI (long T2-hypersignal over more than 3 segments)
- Brain MRI normal or with atypical T2-hypersignals
- Positive NMO IgG antibodies (blood test)
- Prognosis poor



Ischemic Optic Neuropathy

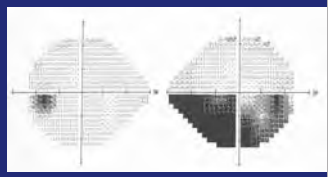
- **Anterior ischemic optic neuropathy (AION):** disc edema
- **Posterior ischemic optic neuropathy (PION):** optic nerve normal acutely
- AION >> PION

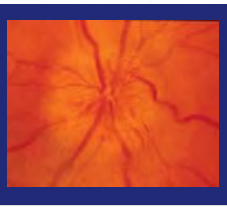
- Local small vessel disease (not embolic!)
- Rule-out giant cell arteritis if >50 years old!!

Optic Neuropathy

Nonarteritic Anterior Ischemic Optic Neuropathy

- Ischemia to the optic nerve head
- M:F 1:1
- Age: older than 50
- Diabetes, hypertension
- Painless
- Altitudinal defect
- Swollen disc
- Permanent visual loss





Perioperative Ischemic Optic Neuropathy

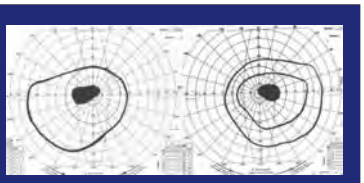
- Anterior optic nerve
  - Acute: swelling of disc
  - > 6 wks: pallor of disc

- Posterior optic nerve
  - Acute: normal fundus
  - > 6 wks: pallor of disc

Compressive Optic Neuropathy

- Progressive, painless, visual loss
- RAPD if unilateral or asymmetric
- Pale nerve (or swollen if orbital mass)
  - Any orbital mass (thyroid) or infiltrative process
  - Any orbital apex lesion
  - Any intracranial mass or infiltrative process compressing the anterior visual pathways

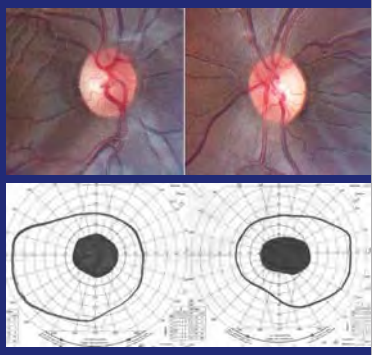
Toxic/Nutritional



- **Vitamin B12 deficiency**
- **Tobacco (cigars > cigarettes)**

- Progressive, bilateral, symmetric central visual loss
- Dyschromatopsia
- Cecocentral scotomas
- Acutely optic nerve normal or appears swollen
- Temporal optic disc pallor may be delayed

Toxic Optic Neuropathies



- **Ethambutol**
  - Dose-related
  - Early dyschromatopsia
- **Linezolide**
  - Dose-related
  - Mild disc edema
  - Peripheral neuropathy
- **Amiodarone**
  - Disc edema (mimics AION)
- **Methanol and ethylene glycol**

Hereditary Optic Neuropathies

- **Isolated optic neuropathies:**
  - Leber hereditary optic neuropathy
  - Dominant optic atrophy
- **Optic neuropathies associated with other neurologic and systemic abnormalities:**
  - DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy, and Deafness)
  - Friedreich ataxia
  - Spinocerebellar ataxia
  - Charcot-Marie Tooth (HMSN type VI)

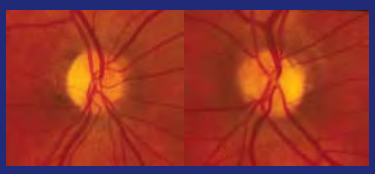
Leber Hereditary Optic Neuropathy

- Bilateral sequential painless central visual loss
- Male >> female, age 15-35
- Hyperemic optic nerve acutely
- Cecocentral scotomas
- Pale optic nerve late
- Poor visual prognosis
- EKG (cardiac conduction abnormalities)

- 3 primary mutations in mitochondrial DNA (11778, 14484, 3460)
- Inherited maternally

Dominant Optic Atrophy (Kjer)

- Bilateral slowly progressive painless visual loss
- Pale optic nerves temporally
- Cecocentral scotomas
- Vision loss relatively moderate
- May have hearing loss



- Autosomal dominant
- Genetic testing (OPA1 gene, chromosome 3)
- Gene codes for mitochondrial protein

Leber Hereditary Optic Neuropathy

Treatment – Idebenone?

- Idebenone Trial (RHODOS)
  - UK/Germany/Montreal
  - 900 mg/day
  - Acute LHON – poor recruitment
  - LHON $\leq$ 5 yrs – 85 pts randomized
    - No adverse side effects
    - Trend for better VA at 6 mos (1 Snellen line)
    - Trend for better VA if Rx'd early

Papilledema

Valerie Biousse, MD

Emory University School of Medicine. Atlanta, GA. USA

Most illustrations are from:
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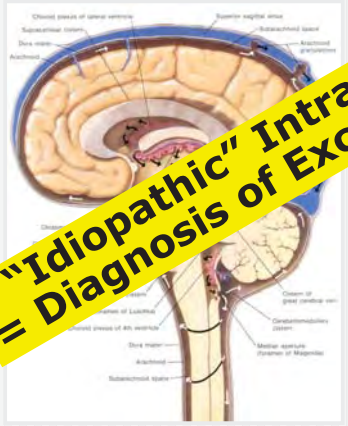
Neuro-Ophthalmology Illustrated. Thieme 2009.
No conflict of interest

Raised Intracranial Pressure

- **Headaches**
- Neurological signs
- Tinnitus
- Diplopia (CN VI)
- Papilledema

Raised ICP

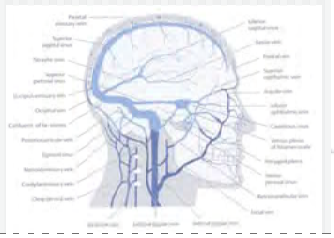


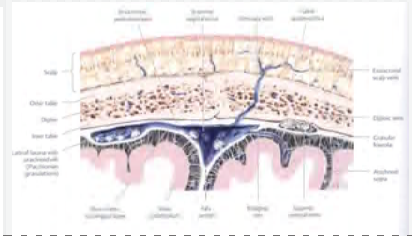
"Idiopathic" Intracranial Hypertension = Diagnosis of Exclusion

- Hy[illegible]us
- [illegible]anial mass
  [illegible]or/abscess/infarction/
  inflammation/blood/AVM…
- Edema
  Toxic/trauma/anoxia
- Meningitis
- **Venous obstruction / dural fistula**
- Increased CSF pressure
- Increased CSF protein

Cerebral Venous Thrombosis

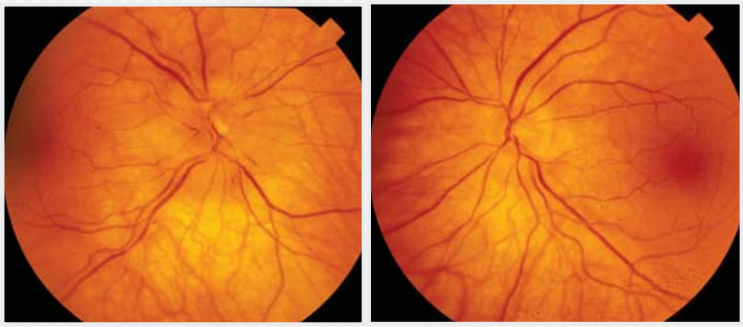
- Transverse and sagittal sinus
  - Coagulation workup
  - Repeat LP (to reduce CSF pressure)
  - Anticoagulation



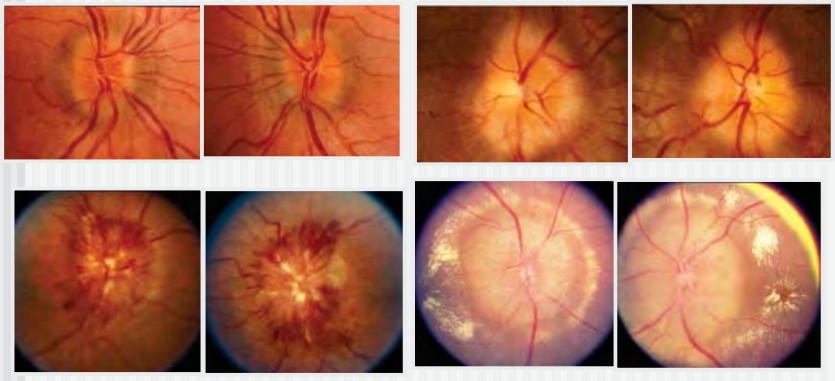


Raised ICP
Papilledema

- No correlation between optic nerve appearance, severity of disc edema and mechanism of raised ICP

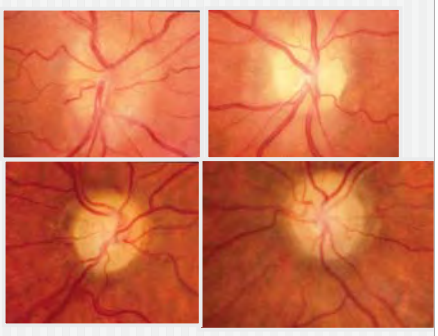


Raised ICP
Papilledema

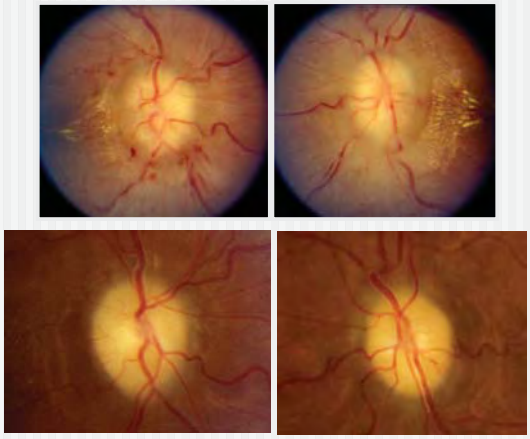


Raised ICP
Optic atrophy

- If raised ICP untreated:
  => Visual loss (optic atrophy)

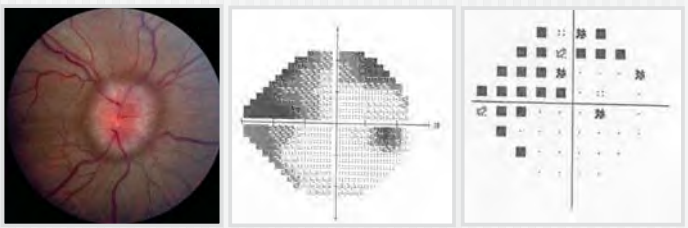


Raised ICP
Optic atrophy

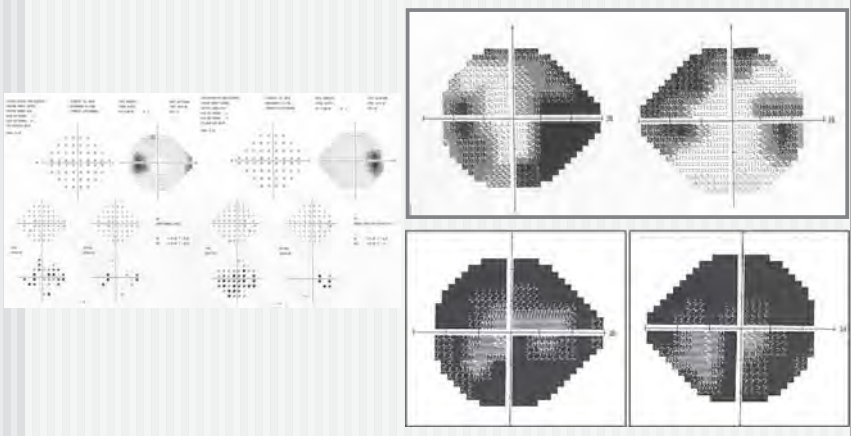


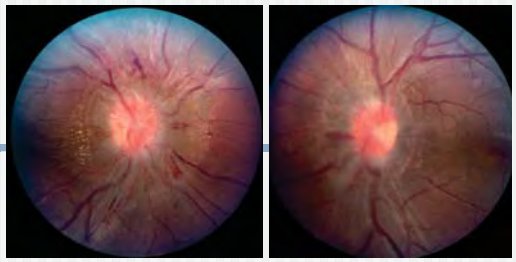
Raised ICP and Papilledema

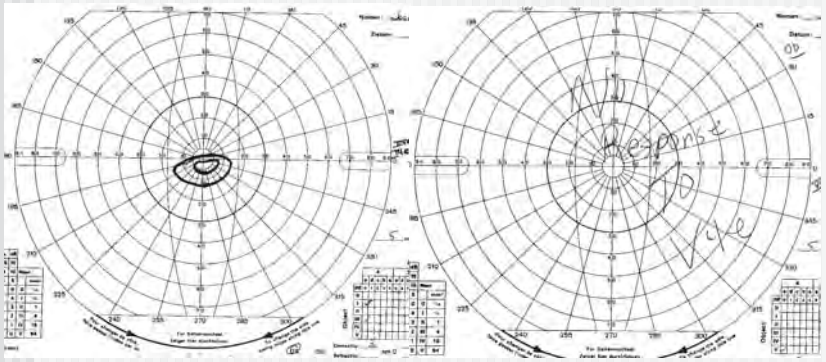
- Visual field defects
- Loss of visual acuity (secondary optic atrophy)



Raised ICP and Papilledema
Visual Field Testing







Bilateral Disc Edema (headache)

Brain imaging (contrast)
(Normal CT→ MRI / MRV)

-Hydrocephalus
-Mass
-Venous thrombosis

Normal

LP

Nl CSF OP
Nl CSF

No Raised ICP
Pseudo DE

Pression ≥25cmH$_2$O
Nl CSF

Abnl CSF

Always reconsider cerebral venous thrombosis

Idiopathic Intracranial Hypertension

- **Isolated intracranial hypertension**
  - Headaches
  - Papilledema
  - Diplopia (Vith)
  - Tinnitus
- **MRI rules out intracranial process and venous thrombosis**
- **LP confirms high CSF OP and normal CSF contents**

Idiopathic Intracranial Hypertension Management

- **Prognosis**
  - Rapid onset
  - Patient's characteristics
    - Drug-induced
    - Severe obesity, race, gender
    - Anemia / sleep apnea syndrome / HTN
  - Visual function
    - Visual acuity, color vision
    - Visual field (automated perimetry, Goldmann perimetry)

Idiopathic Intracranial Hypertension Management

- **Follow-up / Treatment**
  - Lumbar puncture(1st treatment)
  - Correct precipitation factors
    - Drug, anemia, sleep apnea, …
  - Weight loss (long term)
  - No published clinical trial (IIHTT in US
  - Acetazolamide/Topiramate

Idiopathic Intracranial Hypertension
Followup/Treatment (1-2 weeks)

Treatment efficacy
Visual function (**visual field**)

- Improvement
  - Followup (1-2 mo)
  - Followup (3 mos)
  - Followup (6 mos)
- No improvement
  Worsening
  - -Repeat LP
  - -Surgery ?

Slide 49

Idiopathic Intracranial Hypertension
Surgical Treatment

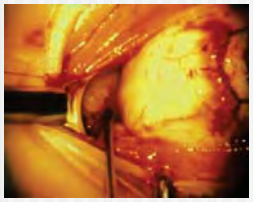
Worsening of vision
Despite "medical" treatment

Sometimes useful emergently:
Lumbar drain, IV steroids

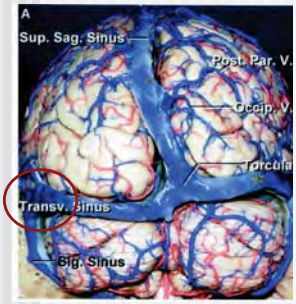
- CSF Shunt (LP or VP)
- Optic nerve sheath fenestration

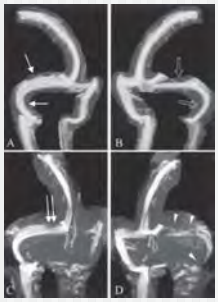
Slide 49

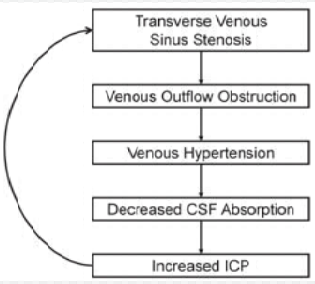
Optic Nerve Sheath Fenestration



Venous Stenting in IIH







Venous Stenting for IIH in 2011

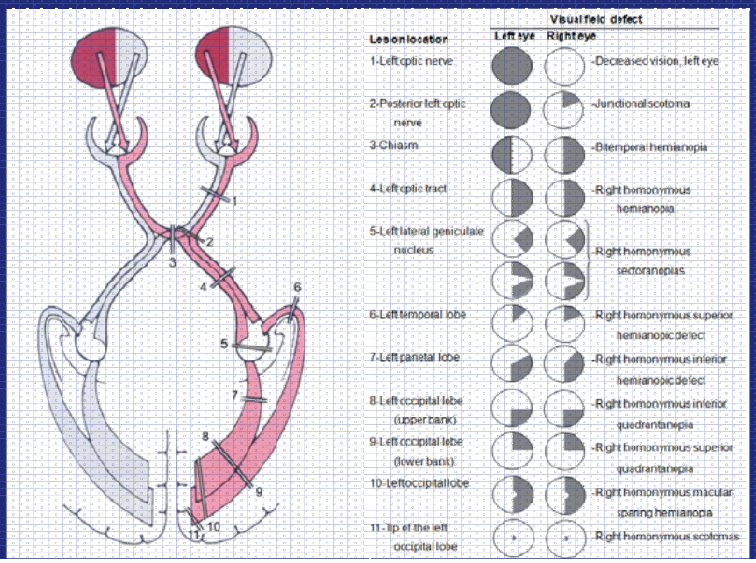
- **Still very controversial**
- **<u>"No"</u> in most cases:**
  - Most cases of IIH are "benign"
  - Venous stenosis often improves or resolves after LP / other treatments of raised ICP
- **<u>"Maybe"</u> in selected severe IIH cases:**
  - With bilateral stenosis (or stenosis of dominant transverse sinus)
  - Resistant to other treatments
  - Surgery not possible

Visual Fields

Nancy J. Newman, M.D.
Emory University School of Medicine
Atlanta, Georgia, USA

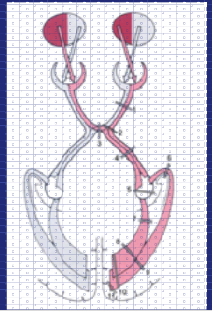
Most illustrations are from:
Biousse V, Newman NJ.
Neuro-Ophthalmology Illustrated. Thieme 2009.
No conflict of interest

Anatomy of Visual Pathways



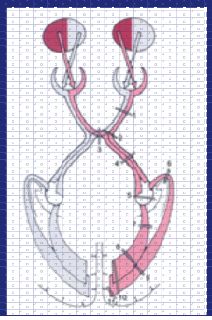
Approach to the Interpretation of VFs "The Four Questions"

1) Does the field defect involve one eye or two?
- **If one eye: it's in the eye or optic nerve**
- **If two eyes: it's either bilateral eye/optic nerve or it's chiasmal/retrochiasmal**



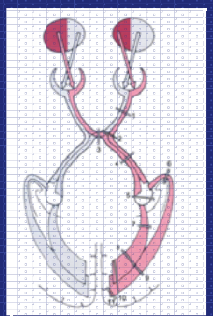
Approach to the Interpretation of VFs "The Four Questions"

2) If two eyes, does the defect respect the vertical meridian?
- If no, then it's in the bilateral eye/optic nerve
- If yes, then it's in the chiasm/retrochiasm



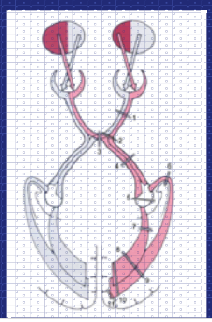
Approach to the Interpretation of VFs "The Four Questions"

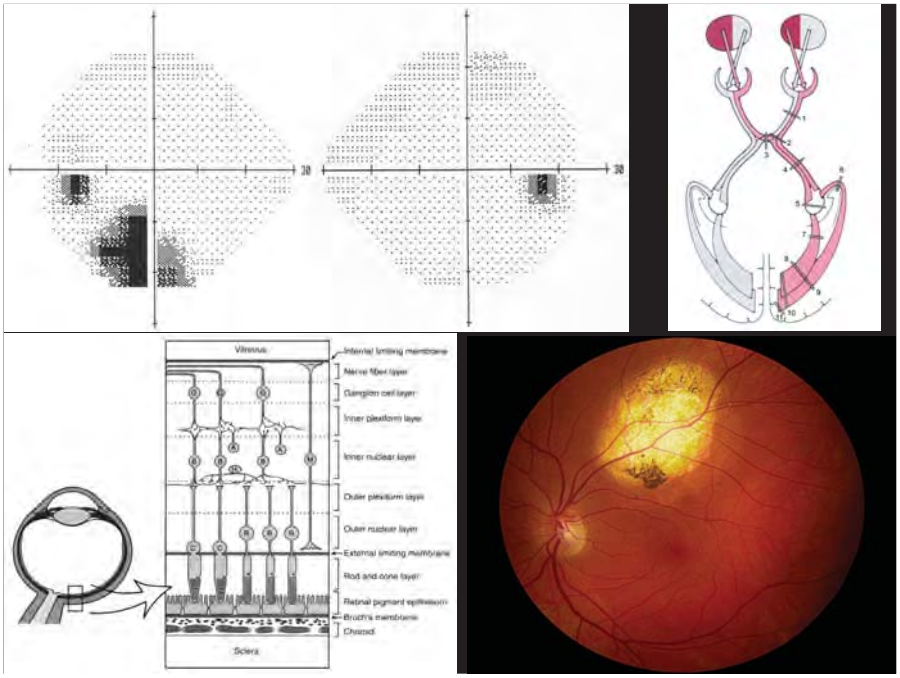
3) If it respects the vertical meridian, are the defects on the same sides of the vertical in each eye (homonymous) or bitemporal?
- If bitemporal, then it's in the chiasm
- If homonymous, then it's retrochiasmal on the other side



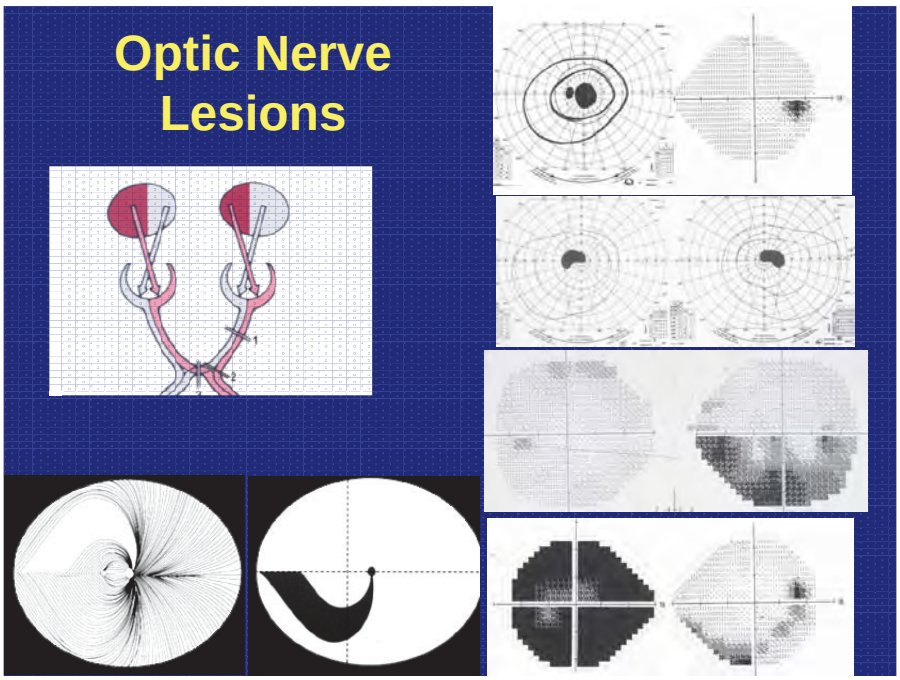
Approach to the Interpretation of VFs "The Four Questions"

4) If homonymous, is it complete or incomplete?
- If complete, it has no further localizing value
- If incomplete, the more congruous, the more posterior

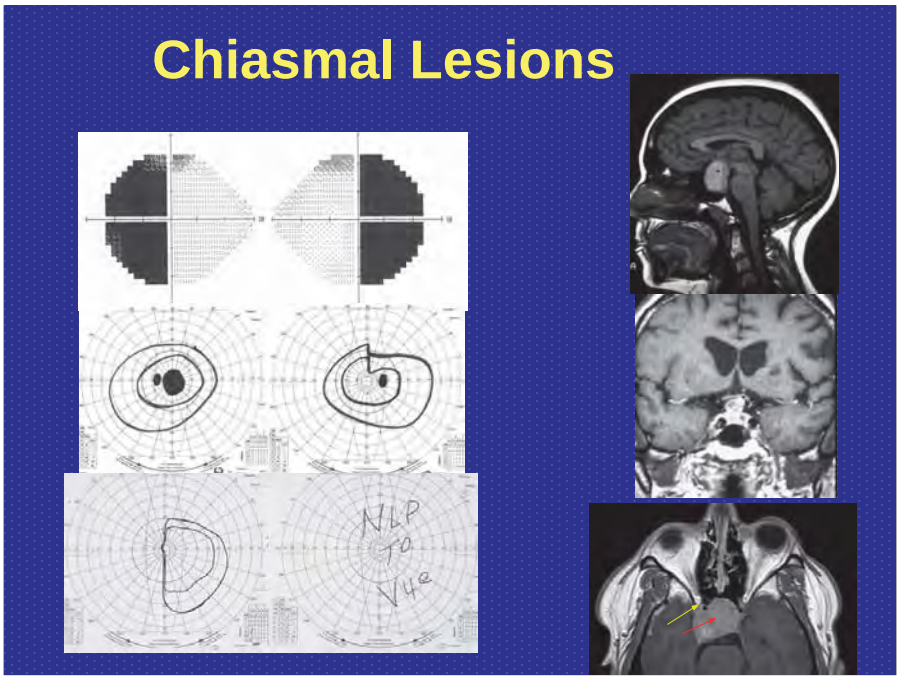




Optic Nerve Lesions



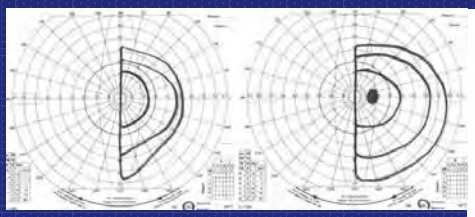
Chiasmal Lesions

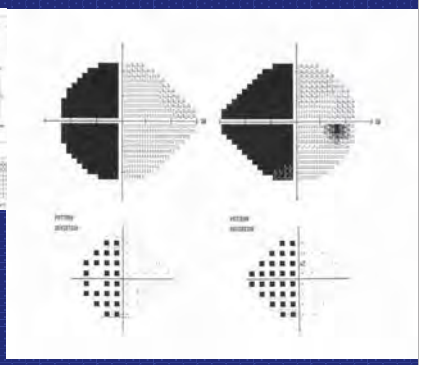


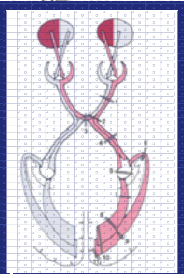
Complete Homonymous Hemianopia

Goldmann VF

Humphrey VF

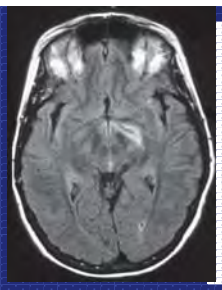


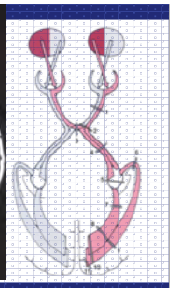


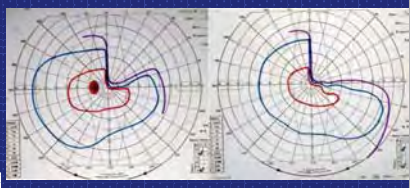


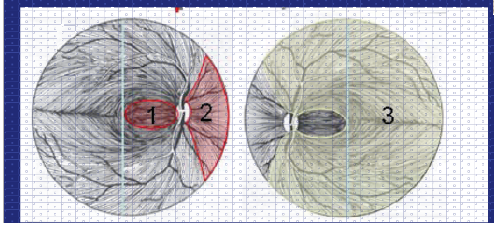
Optic Tract Syndrome

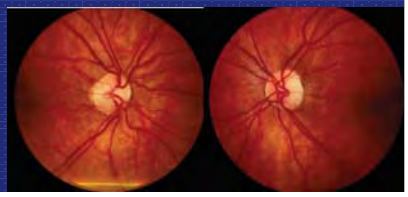
- Left optic tract lesion:
  - Right HH
  - Right RAPD
  - Bowtie atrophy of right optic nerve
  - Temporal pallor of left optic nerve





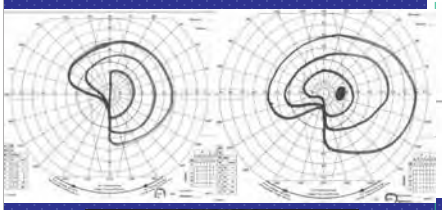


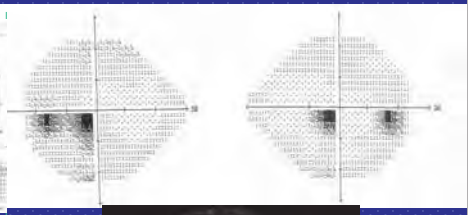


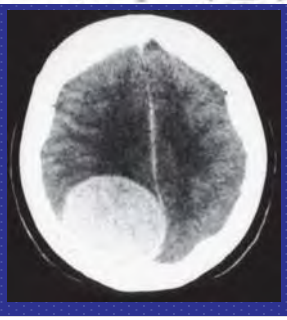


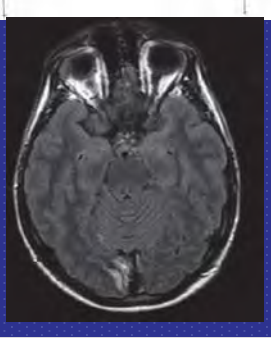
Incongruous HH / Congruous HH

Congruous = Posterior (occipital)

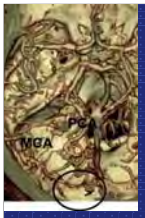




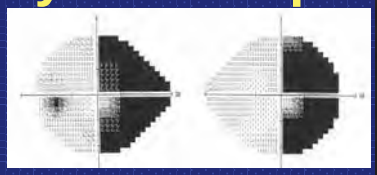


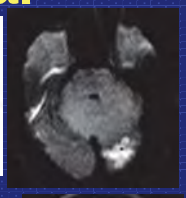


Always is Occipital

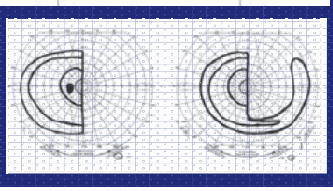


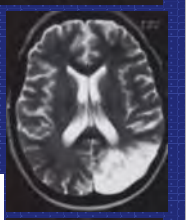
- Macular sparing



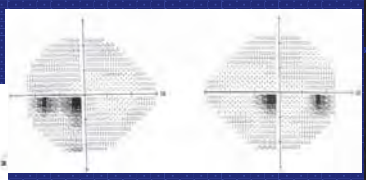


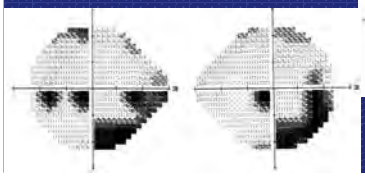
- Temporal crescent

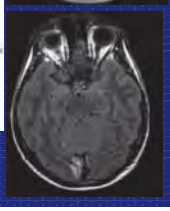




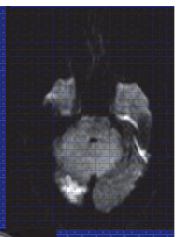
- Homonymous scotomas



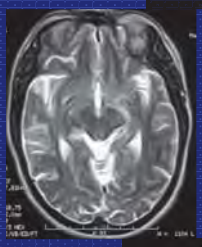


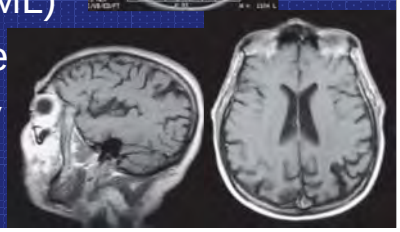


MRI “Negative” HH

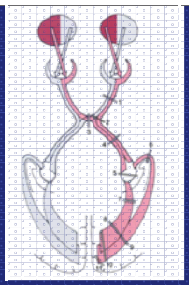


- Wrong technique
- Small or old occipital stroke
- Optic tract lesion
- Progressive multifocal leukoencephalopathy (PML)
- Creutzfeld Jacob disease
- Posterior cortical atrophy (Alzheimer)

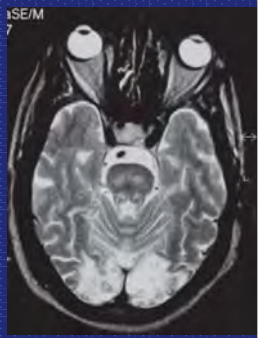




Cerebral Blindness

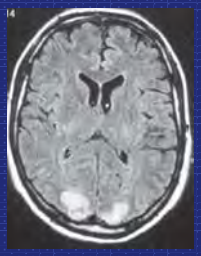


- **Bilateral visual acuity loss**
  - **Equal in both eyes**
- **Normal pupils**
- **Normal fundus**

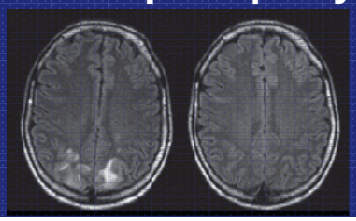


- **Anton’s: Patients with cerebral blindness say they can see (denial)**

Bilateral Occipital Lesions
Cerebral Blindness

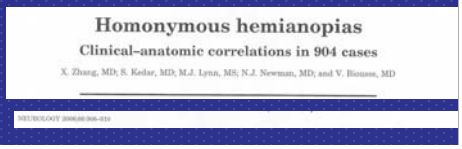


- **Vascular:**
  - Vertebrobasilar ischemia (PCAs)
  - Cerebral anoxia
  - Cerebral venous thrombosis
- **Hypertensive encephalopathy**
- **Eclampsia**
- **Posterior Reversible Leukoencephalopathy (PRESS)**
- **Alzheimer**
- **PML**
- **CJD**



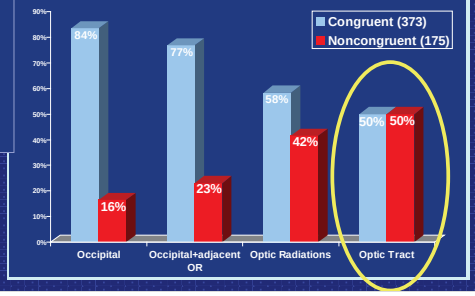
Causes of lesions for each brain lesion location

	Infarct	Hemorrhage	Trauma	Tumor	Others	Total
Occipital (%)	289 (73)	43 (11)	17 (4)	30 (8)	17 (4)	396 (100)
Optic radiations (%)	156 (55)	45 (16)	27 (9.5)	40 (14)	17 (6)	285 (100)
Optic tract (%)	36 (40)	3 (3)	15 (17)	29 (32)	7 (8)	90 (100)
Multiple (%)	28 (28)	4 (4)	64 (64)	2 (2)	2 (2)	100 (100)
Others (%)	23 (70)	3 (9)	0	1 (3)	6 (18)	33 (100)
Total (%)	532 (58.8)	98 (10.8)	123 (13.6)	102 (11.3)	49 (5.4)	904 (100)


Homonymous hemianopias
Clinical-anatomic correlations in 904 cases
X. Zhang, MD; S. Kedar, MD; M.J. Lynn, MS; N.J. Newman, MD; and V. Biousse, MD

Congruency of Homonymous Hemianopia With Respect to Lesion Location

More posterior lesions (occipital) produce more congruent VF defects ($p<.0001$)

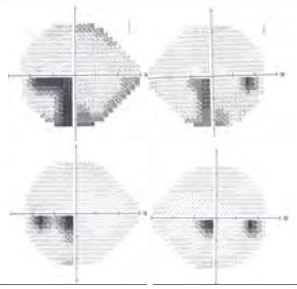


Natural history of homonymous hemianopia

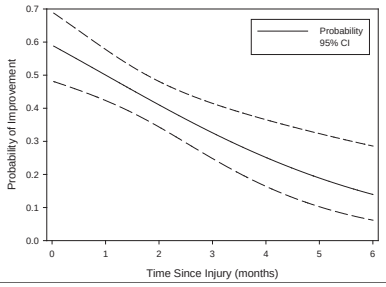
X. Zhang, MD; S. Kedar, MD; M.J. Lynn, MS; N.J. Newman, MD; and V. Biousse, MD

Abstract—*Objective:* To describe the characteristics of spontaneous recovery of homonymous hemianopia (HH). *Methods:* The authors reviewed medical records of all patients with HH confirmed by formal visual field testing and seen in follow-up in their service between 1989 and 2004. Clinical characteristics, causes, neuroradiologic definition of lesion location, final outcome, and evolution of the visual field defects were recorded. The associations among final visual field defect outcome, time from injury, and clinical features were analyzed. *Results:* A total of 254 patients with 263 HH were included in this study. Spontaneous visual field defect recovery was observed in 101 HH (38.4%). The likelihood of spontaneous recovery decreased with increasing time from injury to initial visual field testing ($p = 0.0003$). The probability of improvement was related to the time since injury ($p = 0.0003$) with a 50 to 60% chance of improvement for cases tested within 1 month after injury that decreased to about 20% for cases tested at 6 months after surgery. No other factor was found to correlate with the final outcome of the visual field defects. Improvement after 6 months from injury was mild and usually related to improvement of the underlying disease. *Conclusion:* Spontaneous improvement of homonymous hemianopia is seen in at least 50% of patients first seen within 1 month of injury. In most cases, the improvement occurs within the first 3 months from injury. Spontaneous improvement after 6 months postinjury should be interpreted with caution as it is most likely related to improvement of the underlying disease or to improvement in the patient's ability to perform visual field testing reliably.

NEUROLOGY 2006;66:901–905

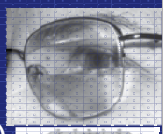


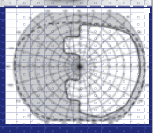
Probability of Improvement vs Time Since Injury



"Rehabilitation" of Homonymous Hemianopia

- Part of post-stroke rehabilitation
- Specific if neglect
- Multiple suggested "therapies"
  - Prisms (displacement of the seeing field)
  - Saccade training
  - Vision stimulation (?"restoration")
    - Enhance cortical plasticity?
    - Use "blindsight"?
    - Increase attention?





Anisocoria
Big Pupil Problem

vs.

Small Pupil Problem

Valerie Biousse, MD

Emory University, Atlanta GA, USA

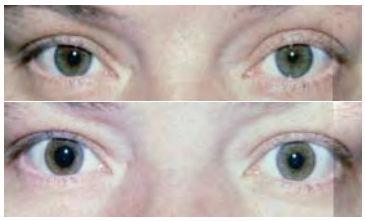
Most illustrations are from: Biousse V, Newman NJ. Neuro-Ophthalmology Illustrated. Thieme 2009.
No conflict of interest

Physiologic Anisocoria

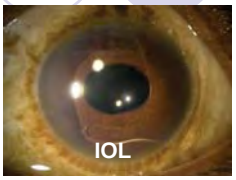
- 10-20% of population has 0.4mm of anisocoria
- Normal light, near and dark reactions

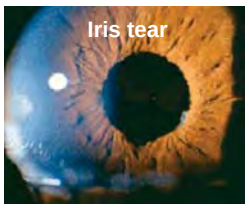
Light

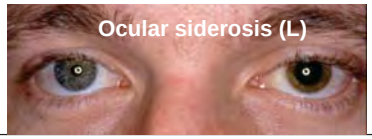
Dark



Anisocoria. Ocular causes

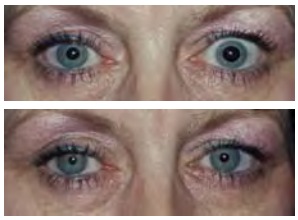


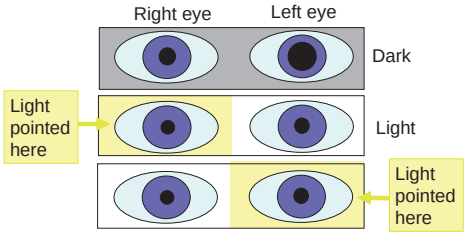




The small pupil is abnormal

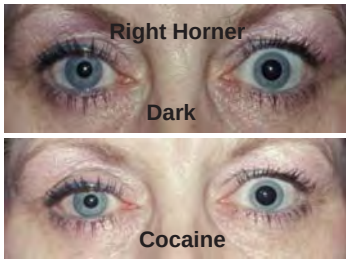
- **The anisocoria is greater in the dark than in the light**
  - Poor pupillary dilation on the abnormal side
  - Abnormality of the sympathetic system.





Pharmacologic Diagnosis of Horner

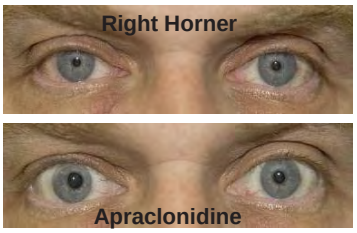
Cocaine (4 or 10%)
-Normal pupil dilates
-Horner pupil dilates poorly
Anisocoria increases



-Cocaine blocks the reuptake of norepinephrine at the sympathetic nerve synapse with the iris dilator:
=> pupillary dilation in eyes with intact sympathetic innervation
=> no effect in eyes with impaired sympathetic innervation, regardless of the lesion location

Pharmacologic Diagnosis of Horner

Apraclonidine (0.5 or 1%)
-Normal pupil does not dilate
-Horner pupil dilates
Anisocoria reverses and palpebral fissure enlarges (apraclonidine reverses the Horner syndrome)



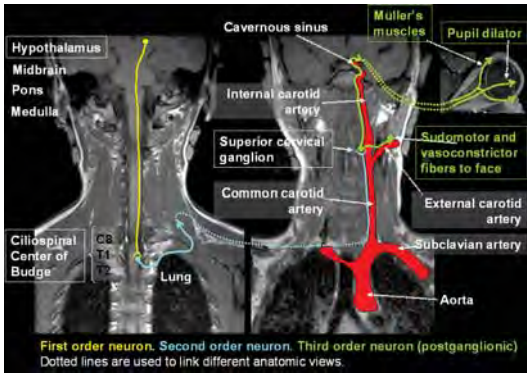
Apraclonidine is a direct α-receptor agonist (strong α2 and weak α1)
=>No effect in eyes with intact sympathetic innervation
=>Mild pupillary dilation in eyes with sympathetic denervation, regardless of the lesion location (with denervation hypersensitivity, α1 effect dilates the Horner pupil)

Horner's Syndrome: Localization

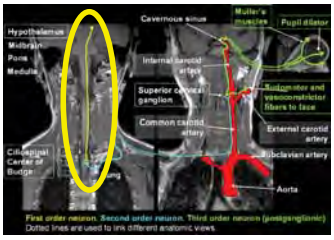
1: 1st order neuron: brainstem/ spine

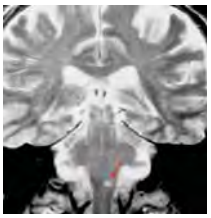
2: 2nd order neuron: brachial plexus/ lung apex

3: 3rd order neuron: carotid dissection

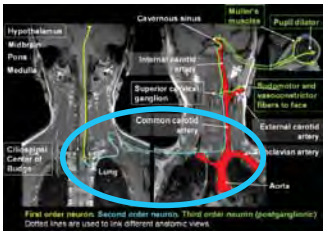


1st Order Horner: Brainstem

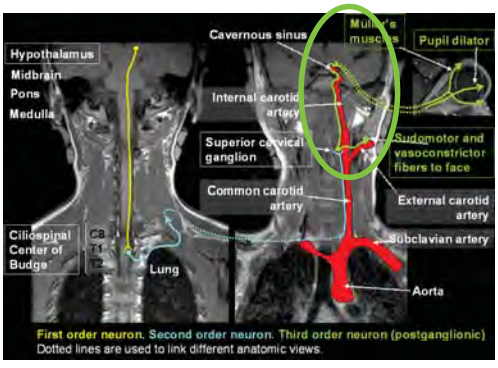


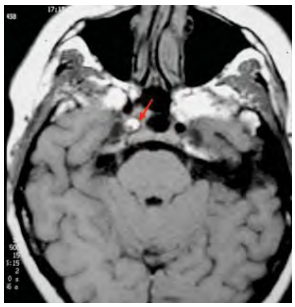


2nd Order Horner: Pancoast Tumor

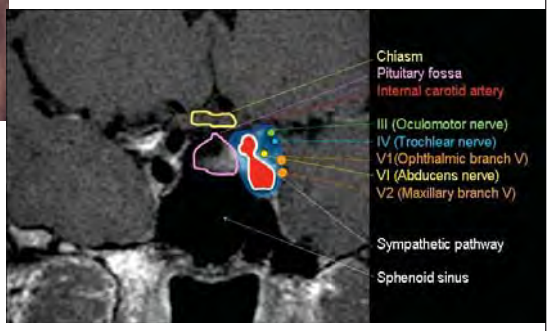


3rd Order Horner: ICA Dissection





Horner + VIth = Cavernous Sinus



Pharmacologic Localization of Horner Hydroxyamphetamine Test

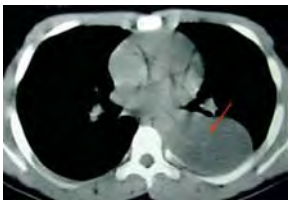
Normal pupil dilates

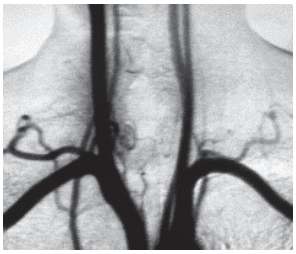
1st or 2nd order dilate

3rd order dilates poorly

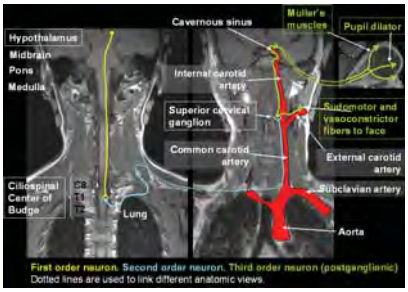
Hydroxyamphetamine releases stored norepinephrine form the postganglionic adrenergic nerve endings. Causes pupillary dilation in eyes with intact sympathetic innervation or intact post ganglionic fibers; has no or partial effect in eyes with impaired sympathetic innervation from lesions involving the post ganglionic fibers

Congenital Horner: Neuroblastoma





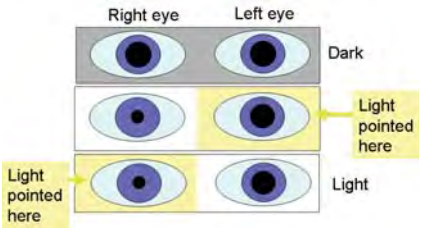
Imaging Horner syndrome



- 1st order-brain and cervical MRI
- 2nd order-MRI spine and neck/ Chest
- 3rd order-MRI brain/MRA head and neck
- Don't know - MR brain, neck and chest X-Ray or CTA head, neck, aortic arch
- Congenital- MRI neck, chest, abdomen

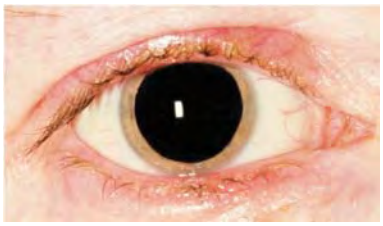
The big pupil is abnormal

- **The anisocoria is greater in the light than in the dark**
  - Poor pupillary constriction on the abnormal side
  - Abnormality of the parasympathetic system.



Pharmacologic Mydriasis

- Very large pupil
- Does not react to light or near
- Poor constriction with Pilocarpine 1%



Pharmacologic Mydriasis

Sphincter blockers

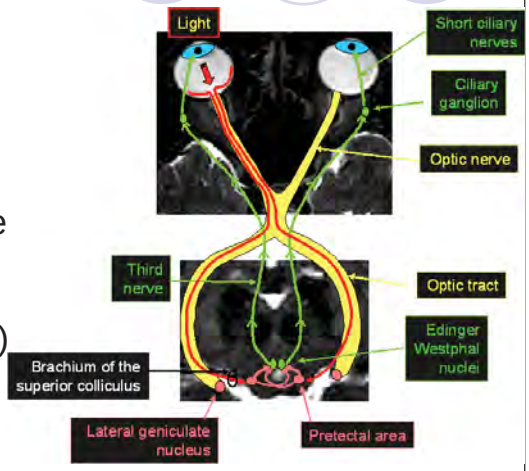
- Belladonna alkaloids
- Atropine
- Scopolamine
- Tropicamide
- Cyclopentolate
- Anticholinergic inhalants
- Gentamycin
- Lidocaine

Dilator Stimulators

- Epinephrine
- Phenylephrine
- Ephedrine
- Hydroxyamphetamine
- Cocaine
- Ocular decongestants
- Adrenergic inhalants

Tonic Pupil

- Initially large, irregular, tonic redilation
- Good near response
- Sensitivity to dilute pilocarpine (0.125%)



Tonic Pupil

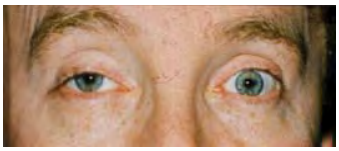
- Light response

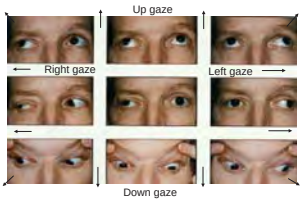
- Near response

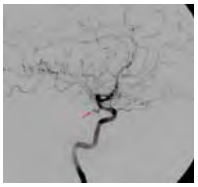
- Constriction with dilute pilocarpine
- Sectoral paralysis, segmental contraction
- Loss of pupillary ruff
- Vermiform movements of iris

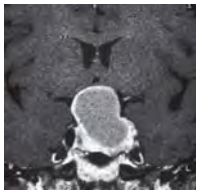
Third Nerve Palsy

- Dilated pupil
- Poorly reactive to light
- Always with ptosis/diplopia
- **PCOM Aneurysm / Pituitary Apoplexy**









Anisocoria :Remember

- **No anisocoria w/ afferent defect**
- **Carotid dissection (Horner)**
- **Posterior communicating artery aneurysm and pituitary apoplexy (IIIrd n. palsy)**

- Case 1

Emergency consult for 66 RHWM with "blown pupils" after CABG

- PMHx:
  - CAD, unstable angina
  - S/p CABG, angioplasties
- Medications:
  - ASA, diltiazem, allopurinol, nitroglycerin
- FamHx:
  - Coronary artery disease (father)
  - Subarachnoid hemorrhage (brother)

HPI

- Coronary artery bypass graft surgery (complicated by angina, intra-aortic balloon pump) - Post-op routine
- Post-op day 2:
- Severe headache, photophobia, blurred vision
  - Rx'd Percocet some help
  - 4 hrs later: Headache and "blown pupils"

Examination

- BP = 120/51, NSR, afebrile
- Somnolent, easily rousable, fully oriented
- Ptosis OU, complete right, partial left
- Near vision: 20/40 OD, 20/30 OS
- Confrontation VFs: full both eyes
- Pupils: 8 mm nonreactive OD, 6 mm trace reactive OS. No RAPD
- Poor adduction, elevation, depression both eyes, right eye worse than left
- Fundi: normal both eyes
- Neurologic exam otherwise normal

- Case 2

48 yo woman with ophthalmoplegia and visual loss

- PMHx: Unremarkable
- Meds: None
- AA, no tobacco, no ETOH
- Fam Hx: stroke, DM

48 yo woman with ophthalmoplegia and visual loss

- Sept 2004: bilateral ptosis, OD deviated out and painful visual loss OD
- Neurologist: "Bilateral IIIrd nerve palsies"
  - Normal brain MRI
- Ophthalmologist:
  - VA 20/400 OD and 20/40 OS

Feb 15, 2005 -- Emory

- Comprehensive Ophthalmology: "My vision is worse OD and I have bilateral third nerve palsies"
- Cornea: Perforated corneal ulcer OD


Neuro-Op consultation

	OD	OS
• VA	CF	20/30
• Pupils	Not seen No RAPD	Normal
• Lids	3 mm ptosis Corneal exposure Lower lid retraction	2 mm ptosis
• Fundus	Not seen	Normal
• EOM	Restricted	Restricted

- Case 3

55 yo woman with diplopia

- PMHx: leg Fx, cholecystectomy, hysterectomy (endometriosis)
- Herpes zoster keratouveitis with chronic uveitis and glaucoma: corneal transplant with cataract extraction Jan 2005
- Meds: None
- No tobacco, no ETOH
- Fam Hx: unremarkable

55 yo woman with diplopia

- Dec 2004: right retro-orbital headaches
  - Episodic, isolated
- Feb 2005: still has episodic pain
  - Brain CT with contrast: normal
- March 2005: Acupuncture for headaches
- March 20, 2005: headaches worse, nausea, diplopia, right ptosis

55 yo woman with diplopia

- March 21: neurologist
  - Ptosis OD, partial adduction OD
  - Right pupil sluggish
  - MRI brain with gad: normal
  - MRA and MRV: normal
  - CBC, ESR, CRP: normal

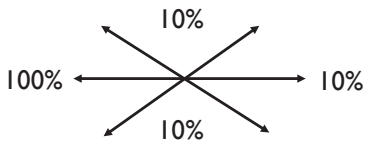
Admitted to hospital

- "Treatment of migraine"
- Prednisone 60 mg PO
- Discharged with diagnosis of "migraine"

Neuro-Op consultation (March 25)…

- The "outside neurologist": "Can you clear the patient for IV steroids?. She has a history of corneal transplant and herpes infection, and I do not know whether these patients can take high doses of steroids"

Neuro-Op consultation

	OD	OS
VA	20/25	20/25
Pupils	Poorly reactive No RAPD	Reactive
Lids	4 mm ptosis	Normal
Fundus	Normal	Normal
EOM		Full

Case 4

37 year old white woman with visual loss right eye

Past Medical History: Unremarkable

Medications: None

Family History: Migraines (mother)

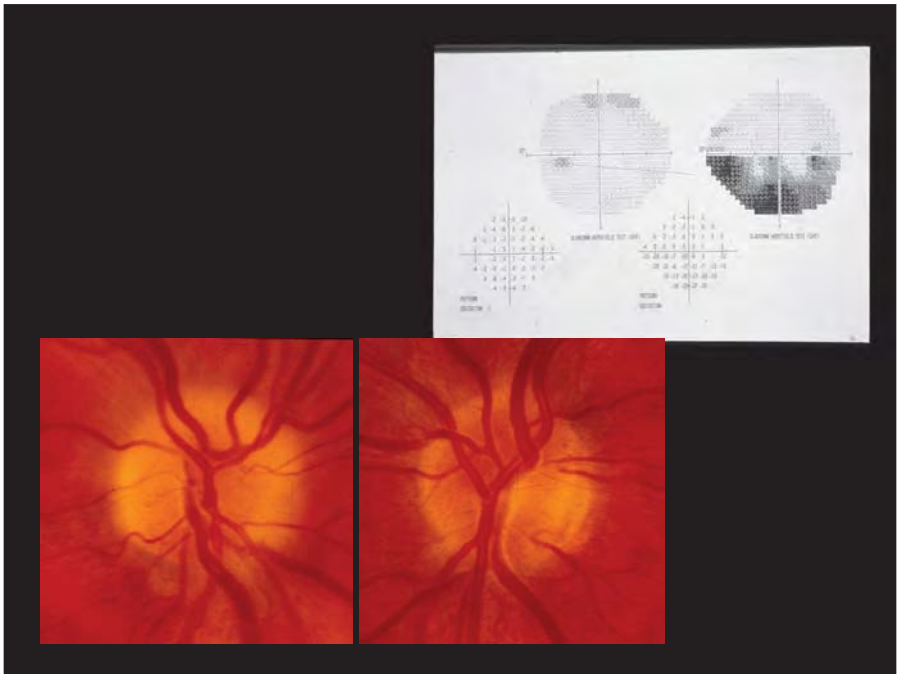
5 weeks prior:
- Irritation/itching right eye
- Better with artificial tears

5 days later:
- Decreasing vision in right eye lower visual field
- Worsened over 4-5 days
- No headache, pain, pain on eye movement

No previous visual loss or neurologic dysfunction
Vision failed to recover over next 3-4 weeks

Examination:

Vision:	20/20	20/20
Color:	14/14 20% red desat	14/14
Orbits:	Normal	Normal
SLE:	Normal	Normal
IOPs:	14	14
Pupils:	+ RAPD	
EOMs:	Full	Full



Case 5

- 79 yo white woman with bilateral visual loss
- Past Medical History:
  - Breast cancer treated with left mastectomy 15 years prior
  - Cataract surgery both eyes 8 years ago

- 3 months prior to referral:
  - Neck pain, neck stiffness and shoulder pain
  - Felt as if she had the “flu”
  - Evaluated by her primary care doctor
    - Steroids for a few days
    - No improvement

- 2 months later:
  - 3 episodes of horizontal binocular diplopia lasting 15 minutes each
  - No headaches or any other neurologic or ocular symptoms
- One week later, awoke with a “shade across the right eye”

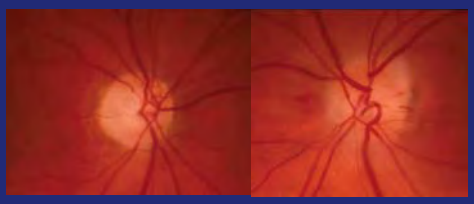
- Evaluated by an ophthalmologist:
  - Head CT normal
  - CBC: normal
  - Platelets: 373,000
  - ESR: 32 mm/1h
  - Glucose: 179

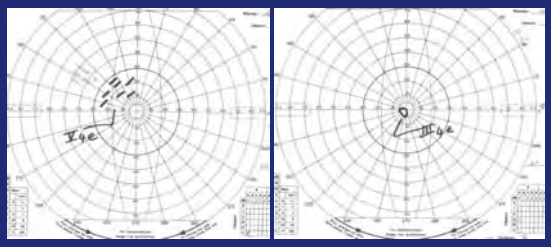
- 2 weeks later:
  - Visual loss right eye
  - Slight pain
- A few days later:
  - Visual loss left eye
- Seen by ophthalmologist, referred to neuro-ophthalmologist

Examination

	OD	OS
VA	20/100-1	Hand Motion
Orbit	Normal	Normal
Lid	Normal	Normal
IOP	14	15
SLE	Normal	Normal
Pupils	+ RAPD	
EOM	Full	Full

Fundus and Goldmann VF





Case 5

66 year old white woman referred with diplopia

PMHx: Non-insulin dependant diabetes mellitus
Hypertension
Hypothyroidism

Meds: Glynase, nifedipine, pentoxifylline, atenolol/chlorthalidone, levothyroxine

FamHx: Noncontributory

9 days prior:

"Sinus pressure"
Scant yellow nasal discharge
Treated with antibiotic, beclomethasone spray

6 days prior:

Horizontal binocular diplopia
Right abduction deficit
CT normal (mild sinus disease)

4 days prior:

Drooping right upper lid
Right ptosis, ophthalmoplegia
Normal pupils and vision
No ocular or facial pain or decreased sensation
No "sinus pressure" or headache
Glucose 250, white count 8,800

Examination: Afebrile, normal general exam

Vision:	CF 1 foot	20/40
Color:	No control	Normal
Extern:	Ptosis	Normal
Exoph:	20	18
Pupils:	4 mm + RAPD	4 mm
SLE:	Normal	Normal
Corneal:	Decreased	Normal
Fundus:	Normal	Normal

Neurologic examination otherwise normal except for: