Neuro-Ophthalmology: Case After Case…

Nancy J. Newman, MD
Valérie Biousse, MD
Emory University School of Medicine, Atlanta, GA

- **Disclosures:**
  - Consultant for Gensight, Santhera and Stealth
  - Research support: Gensight, Santhera and NIH
  - Data Safety Committee: Quark NAION clinical trial
  - Most illustrations are from **Neuro-Ophthalmology Illustrated**, Thieme 2019. 3rd edition

1

Neuro-Ophthalmology: Case After Case…

Nancy J. Newman, MD
Valérie Biousse, MD
Emory University School of Medicine, Atlanta, GA

2

19 yo woman with visual loss

- Past medical history: Obese
- Meds: None
- No tobacco, no alcohol

- Family history: Unremarkable

3

19 yo woman with visual loss

- Past 2 weeks:
  - severe headaches
  - Rapidly progressive visual loss OU
  - Diplopia

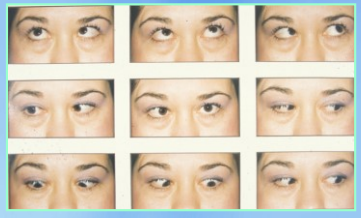
4

Examination

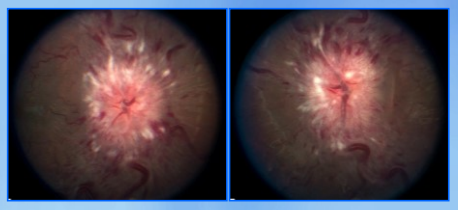
	Right eye	Left eye
Visual acuity	20/30	20/20
Color vision	13/14	14/14
SLE/IOP	Normal	Normal
Pupils	No RAPD	No RAPD
Lids	Normal	Normal

5

Extraocular Movements: Abduction deficit / Esotropia

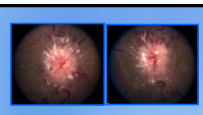


6

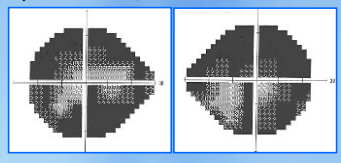


What next?

7



- Visual field testing
- Blood pressure 118/66



8

She needs

- Neurologic examination
- General examination
- Blood pressure
- Brain MRI with Gadolinium
- Brain MRV with Gadolinium
- Lumbar puncture if imaging normal

- Immediately

9

Send her to Emergency Department

- In hospital with:
  - Neurology
  - Neurosurgery
  - Neuroradiology
  - Available 24/7
  - And… Neuro-ophthalmologist

- Warn Neuro-Ophthalmologist and ED

10

Evaluation

- Normal neurologic examination
- Blood pressure: 122/72
- Brain MRI with Gadolinium: Normal
- Brain MRV: Normal (signs of raised ICP)
- Lumbar puncture:
  - Opening pressure: 63 cm H2O
  - Normal CSF contents
- No anemia, no sleep apnea, no medications

11

Fulminant Idiopathic Intracranial Hypertension

- Acetazolamide 500 mg bid
- Dramatic improvement of headaches and diplopia after lumbar puncture
- Consult Neurosurgery

12

Fulminant Idiopathic Intracranial Hypertension

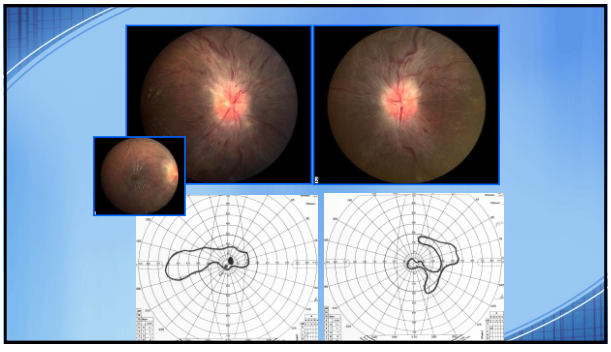
- 12 hours later: recurrence of headaches and diplopia
  - VA: 20/400 right eye, 20/50 left eye
  - Lumbar drain
- Lumbo-peritoneal shunt next day

13

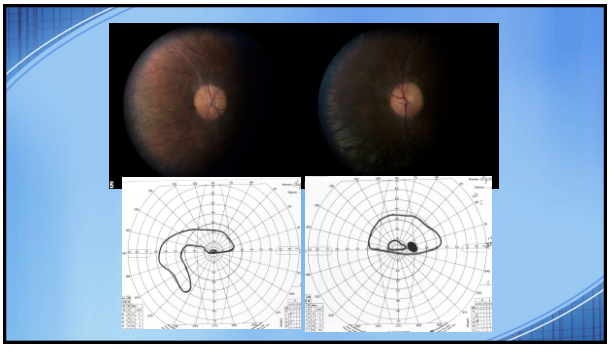
Examination 1 week later

	Right eye	Left eye
VA	20/50	20/40
Color vision	14/14	14/14
SLE/IOP	Normal	Normal
Pupils	No RAPD	No RAPD
EOMs	Full	Full

14

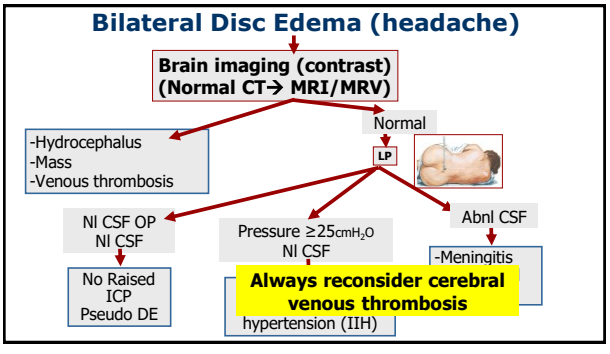


15



16

Bilateral Disc Edema (headache)



17

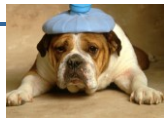
Idiopathic Intracranial Hypertension

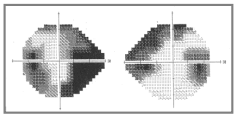
- **Isolated intracranial hypertension**
  - Headaches
  - Papilledema
  - Diplopia (VIth)
  - Tinnitus
- **MRI rules out intracranial process and venous thrombosis**
- **Lumbar puncture confirms high CSF OP and normal CSF contents**

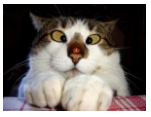
18

Idiopathic Intracranial Hypertension
Evaluation

- Severity of headaches
- Diplopia
- Visual function







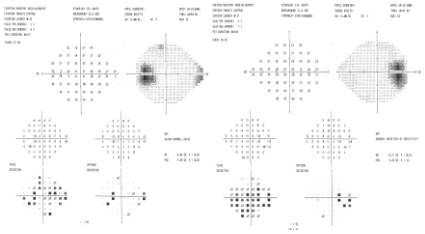
19

Idiopathic Intracranial Hypertension
Management

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

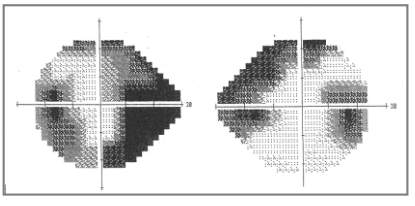
20

Raised ICP and Papilledema
Visual Field Testing



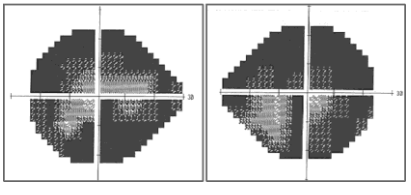
21

Raised ICP and Papilledema
Visual Field Testing

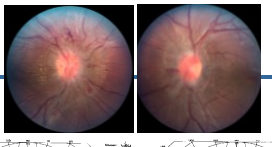


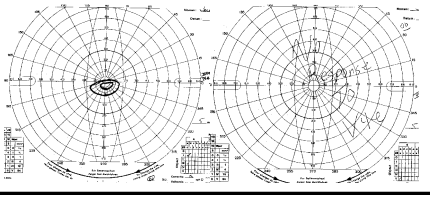
22

Raised ICP and Papilledema
Visual Field Testing



23





24

Idiopathic Intracranial Hypertension Management

- **Follow-up/Treatment**
  - Lumbar puncture(1st treatment)
  - Correct precipitation factors
    - Drug, anemia, sleep apnea, …
  - Weight loss (long term)
  - Acetazolamide (1-3 grams/day)

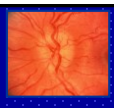
25

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

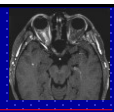
Treatment efficacy
Visual function (**visual field**)

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

26



Optic Neuritis: Is It or Isn't It ?



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

EMORY EYE CENTER

27

A 39 y/o man with visual loss right eye

Past Medical History: hypertension, cigs

Medications: atenolol

Family History: Unremarkable

28

5 weeks ago:
- progressive, painless visual loss OD over 4-5 days

No previous visual loss or neurologic symptoms

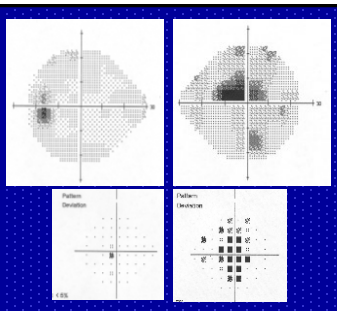
No improvement

29

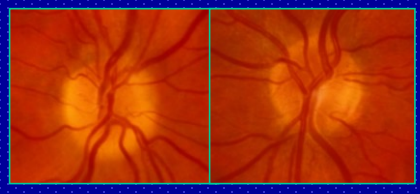
Examination:

Vision:	20/200	20/20
Color:	Control	14/14
Orbits:	Normal	Normal
SLE:	Normal	Normal
IOPs:	14	14
Pupils:	Pharmacologically Dilated	

30



31



32

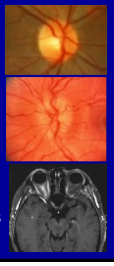
Is It Optic Neuritis?

- Macular disease
- A different cause of optic neuropathy

33

Optic Neuritis
Typical Idiopathic

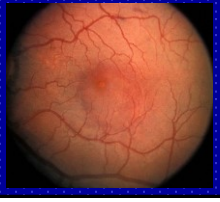
- Inflammation of the optic nerve
- F:M 3:1
- Age: 15-45
- Pain on eye movement
- Normal or swollen disc
- Spontaneous improvement
- Associated with multiple sclerosis



34

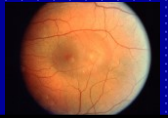
Optic Neuritis Mimickers

- Macular disease



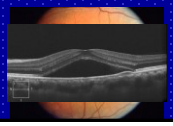
35

	Optic Neuritis	CSR
Sex	Female	Male
Pain	Yes	No
Field	Central	Central
Pupil	RAPD	No RAPD
Fundus	Nerve nl/edema	Blister



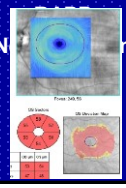
36

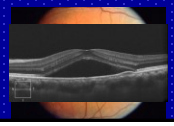
	Optic Neuritis	CSR
Sex	Female	Male
Pain	Yes	No
Field	Central	Central
Pupil	RAPD	No RAPD
Fundus	Nerve nl/edema	Blister



37

	Optic Neuritis	CSR
Sex	Female	Male
Pain	Yes	No
Field	Central	Central
Pupil		No RAPD
Fundus	N ma	Blister





38

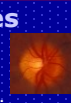
Is It Optic Neuritis?

- A different cause of optic neuropathy

39

Optic Neuropathy Causes

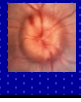
- Inflammatory
- Vascular
- Compressive/Infiltrative
- Hereditary
- Toxic/Nutritional
- Traumatic
- Elevated intracranial pressure
- Elevated intraocular pressure



40

Optic Neuritis DDx

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



41

Optic Neuritis Mimickers

- Anterior ischemic optic neuropathy
- Compressive optic neuropathy
- Leber hereditary optic neuropathy
- Other inflammatory/infectious optic neuritis

42

	Optic Neuritis	NAION
Sex	Female	Either
Age	Younger	Older
Pain	Yes	No
Field	Central	Altitudinal
Fundus	Nerve nl/edema	Nerve edema
MRI	Nerve enhances	Nerve nl
Course	Improves	No change

43

	Optic Neuritis	Compressive
Sex	Female	Either
Age	Younger	Any
Pain	Yes	No
Field	Central	Central/Temporal
Fundus	Nerve nl/edema	Nerve nl/edema
MRI	Nerve enhances	Nerve tumor
Course	Improves	Same/worse

44

	Optic Neuritis	Leber HON
Sex	Female	Male
Pain	Yes	No
Field	Central	Central
Bilateral	No	Yes
Course	Improves	Severe loss
MRI	Nerve enhances	Nerve nl
Fundus	Nerve nl/edema	Pseudoedema

45

	Op Neuritis	NAION	Compressive	Leber HON
Age	Younger	Older	Any	Younger
Gender	F > M	M = F	M = F	M > F
Fam Hx	No or MS	No	No	Yes
Bilateral?	<10% at 1yr	<10% at 1yr	No	100% at 1yr
Onset	Rapid	Rapid	Slow	Subacute
Pain?	++ (w EOM)	Rare	No	No
Color	Poor	Often spared	Poor	Poor
Visual Field	Central	Altitudinal	Central/temp	Cecocentral
Optic Nerve	N /edema	Edema	Nl/edema	Nl/pseudo
MRI	Abnl ON	Normal	Tumor	Normal
Prognosis	Good	Poor	Poor (if no Rx)	Poor
Systemic	MS	HTN/DM	None	None

46

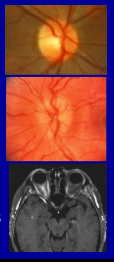
	Op Neuritis	NAION	Compressive	Leber HON
Age	Younger	Older	Any	Younger
Gender	F > M	M = F	M = F	M > F
Fam Hx	No or MS	No	No	Yes
Bilateral?	<10% at 1yr	<10% at 1yr	No	100% at 1yr
Onset	Rapid	Rapid	Slow	Subacute
Pain?	++ (w EOM)	Rare	No	No
Color	Poor	Often spared	Poor	Poor
Visual Field	Central	Altitudinal	Central/temp	Cecocentral
Optic Nerve	N /edema	Edema	Nl/edema	Nl/pseudo
MRI	Abnl ON	Normal	Tumor	Normal
Prognosis	Good	Poor	Poor (if no Rx)	Poor
Systemic	MS	HTN/DM	None	None

47

Optic Neuritis

Typical Idiopathic

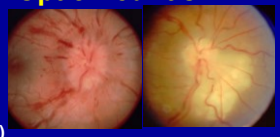
- Inflammation of the optic nerve
- F:M 3:1
- Age: 15-45
- Pain on eye movement
- Normal or swollen disc
- Spontaneous improvement
- Associated with multiple sclerosis



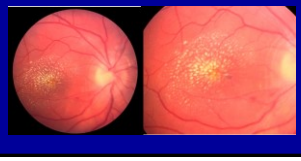
48

Atypical Optic Neuritis

- Bilateral
- Intra-ocular cells
- Disc swelling
- Hemorrhages
- Exudates ("star")



- Sarcoidosis
- Bartonella
- Syphilis
- Tuberculosis
- Viral



49

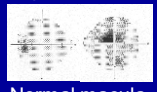
Atypical Optic Neuritis

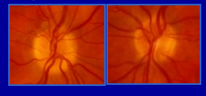
- Bilateral
- Severe
- No recovery
- Recurrent

- AQP4 (NMO) Abs
- MOG Abs

50

A 39 y/o man with painless visual loss OD to 20/200 over 5 days, past history of HTN, no family history of visual loss, and no improvement over the past 5 weeks.

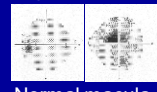




- Normal macula
- Right RAPD
- Normal MRI orbits/brain with contrast
- No vision recovery OD and no vision loss OS for > 1 year

51

A 39 y/o man with painless visual loss OD to 20/200 over 5 days, past history of HTN, no family history of visual loss, and no improvement over the past 5 weeks.



- Normal macula
- Right RAPD
- Normal MRI orbits/brain with contrast
- No vision recovery OD and no vision loss OS for > 1 year

Nonarteritic Anterior Ischemic Optic Neuropathy

52

- 37-yo woman with visual loss OS
- PMHx:
  - 2 normal pregnancies
  - 2 abortions
- Fam Hx:
  - Unremarkable

53

- Visual loss OS:
  - Central shadow
  - Progression over 5 days
  - Pain with eye movements
- No associated neurologic or systemic symptoms

54

Examination

- BP: 116/74, RR 67
- Neurologic examination normal

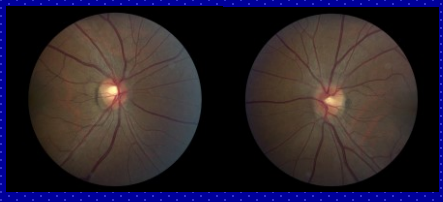
55

Examination

	OD	OS
VA	20/20	Count Fingers
Color	14/14	No control
Orbits	Normal	Normal
Lids	Normal	Normal
IOP	14	15
SLE	Normal	Normal
Pupils	Normal	++ RAPD
EOM	Full	Full

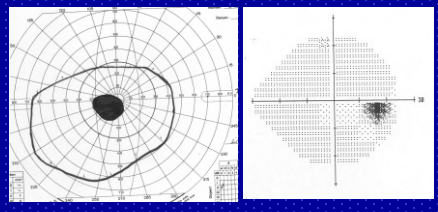
56

Fundus



57

Visual Fields



58

Diagnosis ?

- Optic neuropathy OS
- Retrobulbar
- Inflammatory

- « Idiopathic optic neuritis »

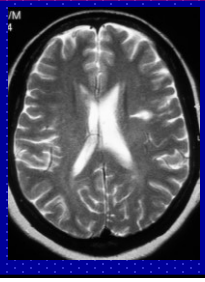
59

What next ?

- Refer

60

Brain MRI



61

Work-up

- Negative syphilis testing
- Normal ACE
- Negative AQP4/MOG antibodies

62

Management-

- 3 days IV methylprednisolone (250 mg q6 hours)
- 10 days oral prednisone (taper)

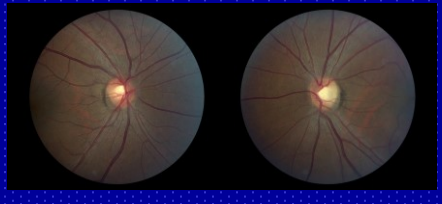
63

Follow-up: 6 weeks later

	OD	OS
• VA	20/20	20/40
• Color	14/14	2/14
• Orbits	Normal	Normal
• Lids	Normal	Normal
• IOP	14	15
• SLE	Normal	Normal
• Pupils	Normal	+ RAPD
• EOM	Full	Full

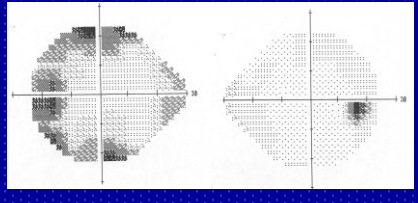
64

Follow-up: 6 weeks later



65

Follow-up: 6 weeks later



66

Patient's Questions…

- Are you sure it is an optic neuritis?
- Is it idiopathic demyelinating optic neuritis?
- Do I have MS ?
- What is my risk of MS ?
- How should I be treated ?

67

What next ?

- Refer

68

- 51 yo woman with visual loss in left eye
- PMHx:
  - Breast cancer 11 years prior (lumpectomy, radiation, chemotherapy, neg lymph nodes)
  - Hypertension, borderline diabetes
  - Migraine headaches (no aura)
- Medications:
  - Hydrochlorothiazide, aspirin 81, vitamins, ibuprofen prn

69

HPI

- Followed by neurologist for episodes of tingling of both legs and occasionally left arm shaking
  - Normal brain MRI
  - Scheduled for electromyography
- Saw optometrist for annual visit:
  - Decreased vision left eye
  - Left optic nerve pallor
  - => "Left optic neuritis"

70

- Neurologist:
  - Cancelled electromyography
  - Repeated brain MRI (normal)
  - Planned LP for possible multiple sclerosis
- Patient:
  - Panicked
  - Refused LP
  - Second opinion

71

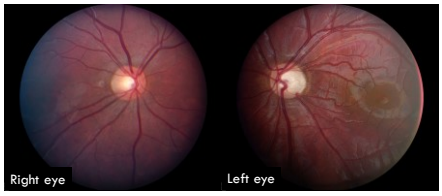
Neuro-Ophthalmology

- BP: 148/92. BMI 32

	Right eye	Left eye
Visual acuity	20/20	20/40-
Color vision	14/14	3/14
Slit lamp	Mild cataracts	
IOP	12	13
Pupils	Normal	RAPD++
Eye movements	Full	Full

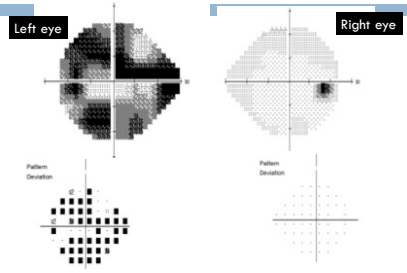
72

Fundus Examination



73

Humphrey Visual Fields



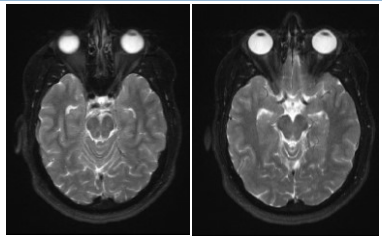
74

Diagnosis

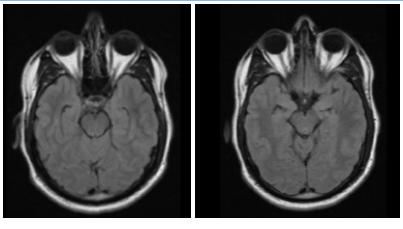
- Left optic neuropathy (chronic)
- Incidentally found
- No pain

75

Review MRI

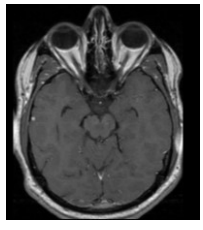


76



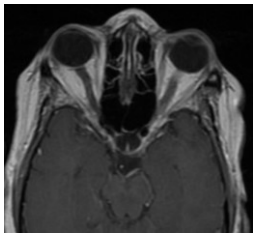
77

With contrast



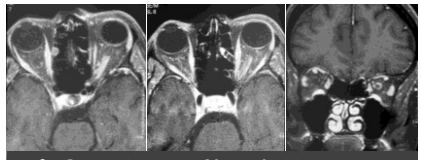
78

MRI **Orbits** with Contrast



79

MRI **Orbits** with Contrast **and Fat Suppression**



Left Optic Nerve Sheath Meningioma

80

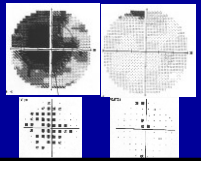
- 20-yo WM with visual loss in both eyes
- PMHx
  - Unremarkable
- Fam Hx:
  - Unremarkable
- College student – no ETOH or drugs

81

- Age 8: told he had « swelling OU » during routine examination
  - Asymptomatic
  - Observed yearly, without change

82

- Age 20 (8 months prior seeing us)
  - Sudden, painless visual loss OS
    - VA: 20/20 OD; 20/200 OS
    - "Swelling" OS

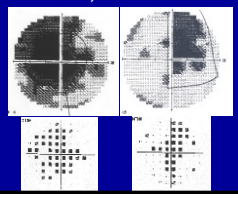


83

- MRI brain/orbits: normal
- CBC, bartonella, toxo, RPR, FTA: normal or negative

84

- 3 months later:
  - Visual loss OD
  - VA: 20/100 OD; CF OS



85

- Repeat MRI: normal
- « More blood tests » : all normal
- Lumbar puncture:
  - OP: 16 cm
  - CSF contents: normal

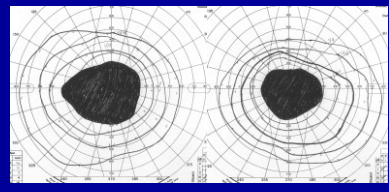
86

Examination 2 months later

	OD	OS
VA	CF	CF
Col Vis	No control	No control
Orbit	Normal	Normal
Lid	Normal	Normal
IOP	14	15
SLE	Normal	Normal
Pupils	Normal	1.2 RAPD
EOM	Full	Full

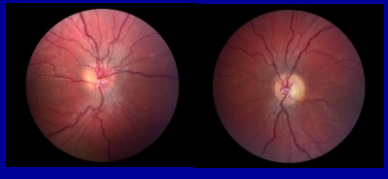
87

Goldmann Visual Fields



88

Fundus



89

Work-up

- Leber's hereditary optic neuropathy:
  - 3 primary mutations:
    - mt DNA 11778
    - mt DNA 14484
    - mt DNA 3460
- Negative

90

Work-up

- **Blood sent to lab with expertise in LHON**
  - **Complete sequencing of mtDNA**
    - **10 mutations found**
    - **Including, novel mtDNA mutation at np 12848** (heteroplasmic)
      - Alters complex 1

-

91

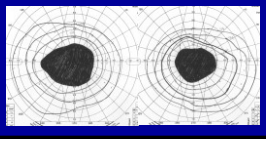
Leber's Hereditary Optic Neuropathy

- Subacute sequential bilateral central visual loss
- Age of onset typically 18-30 (range 1-87)
- Male predominance (80-90%)
- Progression in each eye over weeks to months
- Recognized interval between eyes in 50% (days to months)
- > 97% bilateral within 1 year

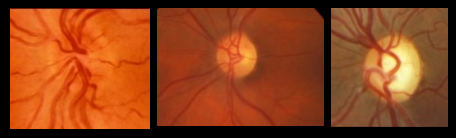
92

Leber's Hereditary Optic Neuropathy

- Acuity usually worse than 20/200
- Color vision affected early
- Central or cecocentral defects



93

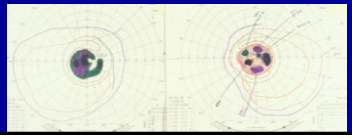


94

Leber's Hereditary Optic Neuropathy

Spontaneous Recovery

- May occur years later
- 4% - 71%
- Depends on the mtDNA mutation
- More likely if visual loss before age 20 (esp <10)



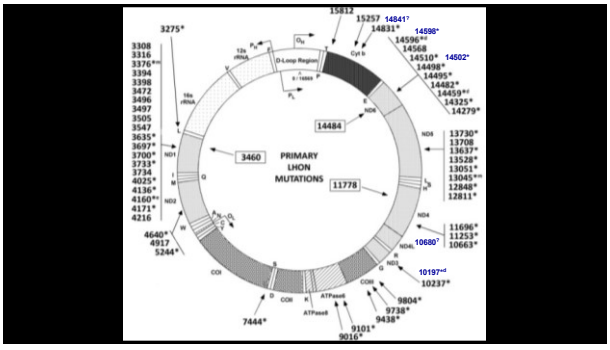
95

Leber's Hereditary Optic Neuropathy

Associated Findings

- Cardiac conduction defects
- Minor neurologic abnormalities
- Multiple sclerosis-like illness
- More severe neurologic syndromes

96

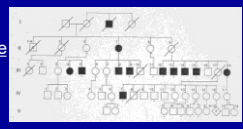


97

Leber Hereditary Optic Neuropathy

Determinants of Phenotype

- Genotype
  - the mutation
  - heteroplasmy
- MtDNA factors
- Nuclear factors
- Environmental factors



98

Hereditary Optic Neuropathies

Treatment

- Genetic counseling
- Symptomatic
- Disease-modifying
  - Mitochondrial diseases
  - Hereditary optic neuropathies
- Idebenone (900mg/d) (ongoing clinical trial)
- Gene therapy (ongoing clinical trials)

99

Mitochondrial Diseases

Symptomatic Treatment

- Improve quality of life: reading, navigating, communication, employment, driving
- Low-vision aids
- Avoid mitochondrial toxicity:
  - Tobacco use
  - ? Heavy alcohol use
  - Meds with mitochondrial toxicity
  - Environmental toxins

100

Hereditary Optic Neuropathies

Treatment

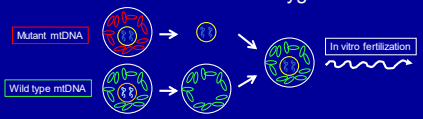
- Genetic counseling
- Symptomatic
- Disease-modifying
  - Mitochondrial diseases
  - Hereditary optic neuropathies
- Idebenone (900mg/d) (ongoing clinical trial)
- Gene therapy (ongoing clinical trials)

101

MtDNA Mitochondrial Disorders

Treatment – Nuclear Transfer

- Oocyte nuclear spindle replacement
- Pronuclear transfer between zygotes



(Tachibana M, et al. Nature 2009;461:367-372)
(Craven L, et al. Nature 2010;465:82-85)

102

The New York Times

Birth of Baby With Three Parents' DNA Marks Success for Banned Technique

The Washington Post

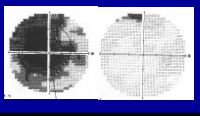
A U.S. doctor went to Mexico to help a Jordanian couple conceive a baby with 3 genetic parents. What's next?

103

Leber Hereditary Optic Neuropathy

Treatment

- Ideal "laboratory" for testing treatment efficacy
  - Sequential visual loss: therapeutic window
  - Accessibility via topical or intravitreal route
  - Implications for other optic neuropathies

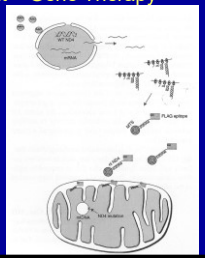


104

Leber Hereditary Optic Neuropathy

Treatment – Gene Therapy

- Allotopic Rescue



105

RESCUE & REVERSE Phase 3 Trials:

Time-based strategy to assess GS010 efficacy

Different patient inclusion criteria	Same design	Same endpoints at Week 48
REVERSE Onset of vision loss 6 months to ≤ 1 yr; 37 patients; Fully enrolled Feb 2017	• One eye randomized to GS010, other eye received sham injection, "best" eye treated half the time	**Primary** • Mean difference change from baseline, ETDRS letters, drug treated eyes vs. sham treated eyes (LogMAR)
RESCUE Onset of vision loss ≤ 6 months; 39 patients; Fully enrolled July 2017	Group 1: GS010 in right eye / SHAM in left eye; Group 2: SHAM in right eye / GS010 in left eye	**Secondary** • SD-OCT, visual field, color and contrast vision • Responder analysis: ◦ Gain from baseline of ≥ 15 ETDRS letters ◦ Snellen acuity ≥ 20/200 • Treated vs. sham eyes BCVA for best-seeing and worst-seeing eyes

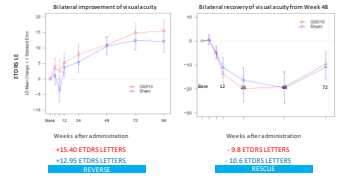
106

REVERSE 6 – 12 months

Results REVERSE and RESCUE

RESCUE ≤ 6 months

GS010 and Sham eye groups keep tracking together



+15.40 ETDRS LETTERS
+12.95 ETDRS LETTERS
REVERSE

- 9.8 ETDRS LETTERS
- 10.6 ETDRS LETTERS
RESCUE

107

REFLECT Phase 3 trial:

Assess efficacy and safety of bilateral injection

Patient inclusion criteria	Design	Endpoints at week 48
• **98 subjects** with vision loss ≤ 1 year • 49 subjects received bilateral GS010 • 49 subjects received GS010 IVT one eye, placebo in the other • Fully enrolled July 2019	Group 1: GS010 in 1st affected eye / GS010 in 2nd affected eye; 1st affected eye always treated with GS010; 2nd affected eye randomized between GS010 and placebo; GS010 in 1st affected eye / Placebo in 2nd affected eye; Group 2	**Primary** • Difference in change of vision compared to baseline between GS010 treated eye vs. placebo eye in 2nd affected/not yet affected eyes (LogMAR) **Secondary** • Pelli Robson contrast sensitivity • Humphrey visual fields • Responder analyses ◦ Gain from baseline of ≥ 15 ETDRS letters ◦ Snellen acuity ≥ 20/200 • Spectral domain OCT • Quality of life assessments

108

From a Patient's Perspective

- Social media growth makes it easier for LHON patients and families to connect
  - Global LHON Facebook (4,000+)
  - Clinical database (3,400+ entries)
- Website and social media facilitate study trial recruitment with just a "click"

109

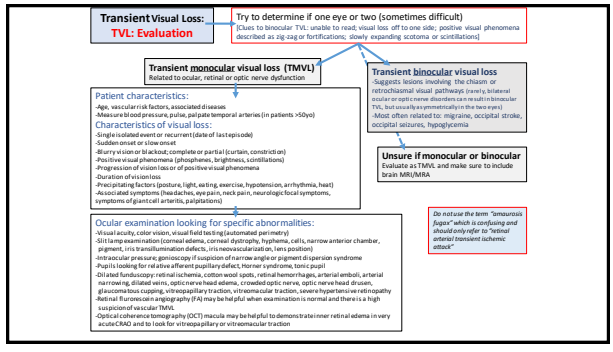
61 yo man

- With acute visual loss in one eye…

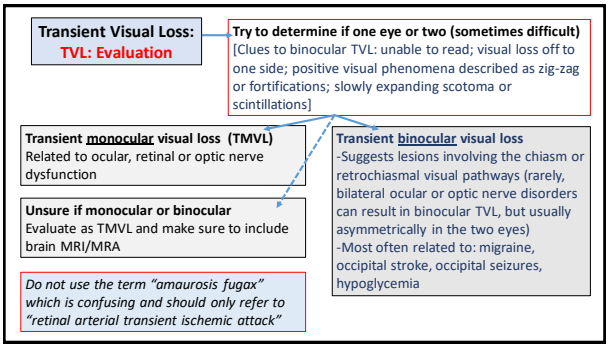
110

"My Vision Blacked Out!"

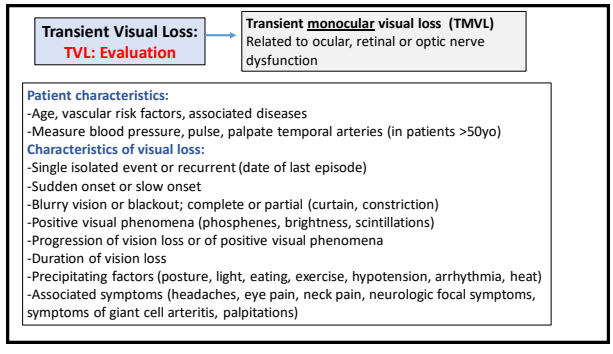
111



112



113



114

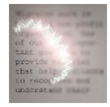
Transient Visual Loss: TVL: Evaluation

Transient monocular visual loss (TMVL)
Related to ocular, retinal or optic nerve dysfunction

Ocular examination looking for specific abnormalities:
-**Visual acuity, color vision, visual field testing** (automated perimetry)
-**Slit lamp examination** (corneal edema, corneal dystrophy, hyphema, cells, narrow anterior chamber, pigment, iris transillumination defects, iris neovascularization, lens position)
-**Intraocular pressure**; gonioscopy if suspicion of narrow angle or pigment dispersion
-**Pupils** looking for relative afferent pupillary defect, Horner syndrome, tonic pupil
-**Dilated funduscopy**: retinal ischemia, cotton wool spots, retinal hemorrhages, arterial emboli, arterial narrowing, dilated veins, optic nerve head edema, crowded optic nerve, optic nerve head drusen, glaucomatous cupping, vitreopapillary traction, vitreomacular traction, severe hypertensive retinopathy
-**Retinal flurorescein angiography** (FA) may be helpful when examination is normal and there is a high suspicion of vascular TMVL
-**OCT** macula helpful to demonstrate inner retinal edema in very acute CRAO and to look for vitreopapillary or vitreomacular traction

115

Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss



TMVL is NOT migraine!
.Migrainous visual aura is always binocular (originates in the occipital cortex: cortical depression). Often described as "on the side"
.Positive visual phenomena: expanding zig-zag lines often followed by an enlarging scotoma (scintillating scotoma) highly suggestive of occipital origin
.Lasts 20-30 minutes; changes sides with attacks; followed by headaches but may remain isolated. Typically recurrent with no sequelae

116

Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss

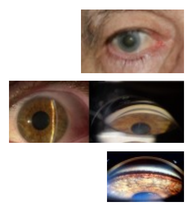
Ocular causes of TMVL (non-vascular)
Related to ocular, retinal or optic nerve dysfunction
-**Refractive error** (accommodative spasm, hyperglycemia)
-**Ocular surface disease** (dry eyes, blepharitis, epiphora, contact lens)
.Blurry vision, fluctuates, worse when focusing, better when blinking
-**Corneal edema, corneal dystrophy**
.Blurry vision, common upon awakening
-**Anterior chamber inflammation/hyphema***
-**Phakodonesis***
-**Intermittent angle closure**
.Ocular pain common, but may be absent
-**Pigment dispersion syndrome***
.Young, myopic, blurry vision with exercise
.Krukenberg spindles on cornea, pigmented angle
-**Vitreous hemorrhage*, vitreous floaters***
-**Incomplete retinal detachment**
.Moving curtain
-**Vitreomacular traction**
-**Vitreopapillary traction***
-**Nocturnal pressure on the eye** (obstructive sleep apnea)
-**Smartphone blindness**
-**Orbital mass (or thyroid eye disease)***
-**Orbital vascular malformation**
-**Abnormal optic nerve head** (papilledema, optic nerve head drusen, crowded disc)
.Transient visual obscurations*: very brief (seconds) blackouts of vision (often bilateral, but usually asymmetrically in the two eyes)
-**Previous optic neuropathy (usually optic neuritis)**
.Uhthoff's phenomenon: decreased vision (minutes to hours) precipitated by increased body heat (hot bath, exercise)
Blurry vision precipitated by changes in position or eye movements

This is NOT migraine!
.Migrainous visual aura is always binocular (originates in the occipital cortex: cortical depression). Often described as "on the side"
.Positive visual phenomena: expanding zig-zag lines often followed by an enlarging scotoma (scintillating scotoma) highly suggestive of occipital origin
.Lasts 20-30 minutes; changes sides with attacks; followed by headaches but may remain isolated. Typically recurrent with no sequelae

Vascular causes of TMVL
Related to arterial or venous ischemia of the retina, choroid, optic nerve or the entire eye.
-**Retinal arterial transient ischemic attack**
.Brief (minutes, not seconds), sudden, often complete blackout of vision, or curtain or constriction
.Spasm of central retinal artery may produce isolated brief (minutes, not seconds), recurrent episodes of complete blackout of vision in young healthy people (diagnosis of exclusion)
-**Choroidal ischemia**
.TMVL is often described as "rain", "fog", "web", sometimes with purple colors (lasts often longer than retinal arterial ischemia)
.Retinal fluorescein or indocyanine green (ICG) angiography helpful
.Highly suggestive of vasculitis such as giant cell arteritis (GCA)
-**Impending central retinal vein occlusion**
.TMVL is longer (20-30 min) and incomplete, often described as "fog" or diffusely blurry
.Dilation of retinal veins with often normal retina and optic nerve
-**Ocular ischemic syndrome**
.TMVL lasts longer (30 min) and often precipitated by orthostasis, hypotension, post-prandial, exercise, exposure to bright light.
.Phosphenes, impaired dark adaptation common
.May have ocular pain improving when lying down
-**Ischemic optic neuropathy**
.Optic nerve head edema if anterior ischemic optic neuropathy
.Highly suggestive of vasculitis such as GCA (non-arteritic ischemic optic neuropathy never presents with TMVL)
.May be isolated or with cotton wool spots, choroidal ischemia (GCA)

117

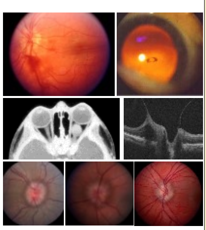
Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss



Ocular causes of TMVL (non-vascular)
Related to ocular, retinal or optic nerve dysfunction
-**Refractive error** (accommodative spasm, hyperglycemia)
-**Ocular surface disease** (dry eyes, blepharitis, epiphora, contact lens)
.Blurry vision, fluctuates, worse when focusing, better when blinking
-**Corneal edema, corneal dystrophy**
.Blurry vision, common upon awakening
-**Anterior chamber inflammation/hyphema***
-**Phakodonesis***
-**Intermittent angle closure**
.Ocular pain common, but may be absent
-**Pigment dispersion syndrome***
.Young, myopic, blurry vision with exercise
.Krukenberg spindles on cornea, pigmented angle

118

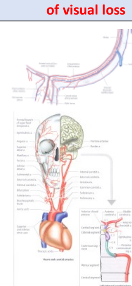
Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss



Ocular causes of TMVL (non-vascular)
-**Vitreous hemorrhage*, vitreous floaters***
-**Incomplete retinal detachment**
.Moving curtain
-**Vitreomacular traction**
-**Vitreopapillary traction***
-**Nocturnal pressure on the eye** (obstructive sleep apnea)
-**Smartphone blindness**
-**Orbital mass (or thyroid eye disease)***
-**Orbital vascular malformation**
-**Abnormal optic nerve head** (papilledema, optic nerve head drusen, crowded disc)
.Transient visual obscurations*: very brief (seconds) blackouts of vision (often bilateral, but usually asymmetrically in the two eyes)
-**Previous optic neuropathy (usually optic neuritis)**
.Uhthoff's phenomenon: decreased vision (minutes to hours) precipitated by increased body heat (hot bath, exercise)
Blurry vision precipitated by changes in position or eye movements

119

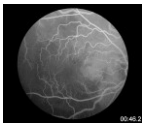
Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss



Vascular causes of TMVL
Related to arterial or venous ischemia of the retina, choroid, optic nerve or the entire eye.
-**Retinal arterial transient ischemic attack**
.Brief (minutes, not seconds), sudden, often complete blackout of vision, or curtain or constriction
.Spasm of central retinal artery may produce isolated brief (minutes, not seconds), recurrent episodes of complete blackout of vision in young healthy people (diagnosis of exclusion)

120

Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss

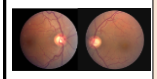


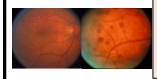
Vascular causes of TMVL
Related to arterial or venous ischemia of the retina, choroid, optic nerve or the entire eye.

-Choroidal ischemia
.TMVL is often described as "rain", "fog", "web", sometimes with purple colors (lasts often longer than retinal arterial ischemia)
.Retinal fluorescein or indocyanine green (ICG) angiography helpful
.Highly suggestive of vasculitis such as giant cell arteritis

121

Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss





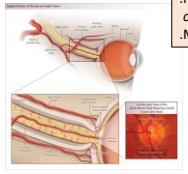
Vascular causes of TMVL
Related to arterial or venous ischemia of the retina, choroid, optic nerve or the entire eye.

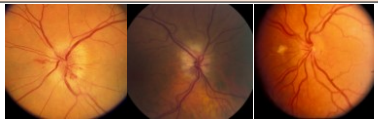
-Impending central retinal vein occlusion
.TMVL is longer (20-30 min) and incomplete, often described as "fog" or diffusely blurry
.Dilation of retinal veins with often normal retina and optic nerve

-Ocular ischemic syndrome
.TMVL lasts longer (30 min) and often precipitated by orthostasis, hypotension, post-prandial, exercise, exposure to bright light.
.Phosphenes, impaired dark adaptation common
.May have ocular pain improving when lying down

122

Transient Monocular Visual Loss
Mechanisms and diagnosis of visual loss





Vascular causes of TMVL
Related to arterial or venous ischemia of the retina, choroid, optic nerve or the entire eye.

-Ischemic optic neuropathy
.Optic nerve head edema if anterior ischemic optic neuropathy
.Highly suggestive of vasculitis such as **GCA** (*non-arteritic ischemic optic neuropathy never presents with TMVL*)
.May be isolated or with cotton wool spots, choroidal ischemia

123

Transient Monocular Visual Loss TMVL: Management

X **Do NOT make a falsely reassuring diagnosis of migraine!**
Do NOT assume it is central retinal artery vasospasm until other causes are ruled out

Slow down and take the time to take a good history (see Evaluation)
Ask patients to draw what they saw: if they draw fortifications/zig-zags, it may be binocular (migraine)

Slow down and perform a detailed ocular examination
Check the pupils yourself! Obtain a visual field if examination is normal. Consider retinal FA and OCT

Look for associated symptoms: eye pain, headache, neck pain, giant cell arteritis, neurologic symptoms
Measure blood pressure, check pulse for arrhythmia

Ocular or orbital cause

Patient > 50 yo

Think giant cell arteritis
Obtain immediate CBC, platelets, ESR, CRP

Abnormal

Normal

Treat as GCA
.Intravenous high dose steroids
.Temporal artery biopsy

Vascular causes of TMVL – Is it?:
-Retinal arterial transient ischemic attack
-Choroidal ischemia
-Impending central retinal vein occlusion
-Ocular ischemic syndrome / Venous stasis retinopathy
-Ischemic optic neuropathy (GCA)

Retinal examination normal or shows retinal emboli, retinal ischemia, ocular ischemic syndrome

Retinal arterial transient ischemic attack

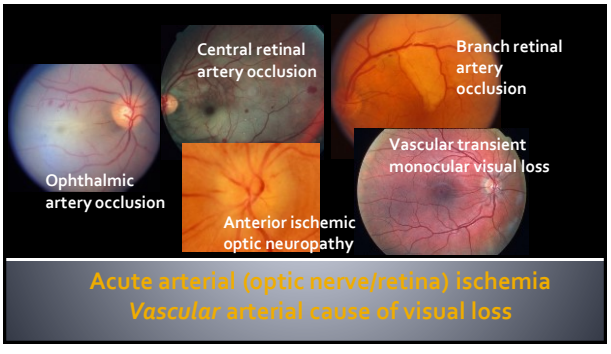
Follow the guidelines!
Immediate referral to closest emergency Stroke Center
Find it at: https://hospitalmaps.heart.org/#/AHAMAP/map/qmaps up

124

71 yo man

- With acute visual loss in one eye...

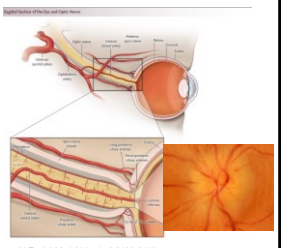
125



126

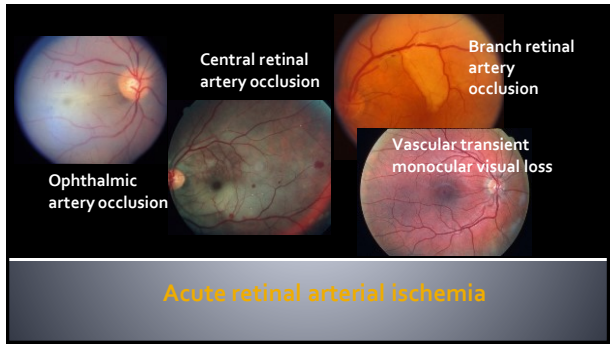
Anterior Ischemic Optic Neuropathy

- **Not the same kind of "stroke"**
  - Small vessel disease
- **Think giant cell arteritis:**
  - Older than 50 yo
  - Transient or permanent visual loss

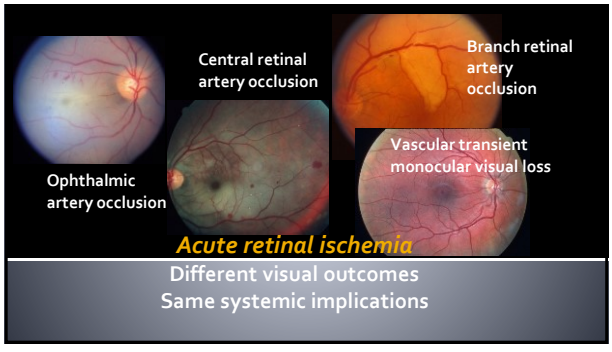


N Engl J Med 2015; 372:2428-2436

127



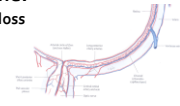
128

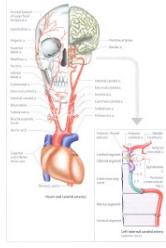


129

Acute Retinal Arterial Ischemia

- **It's all the same:**
  - **Transient visual loss**
  - **BRAO**
  - **CRAO**
  - **OAO**

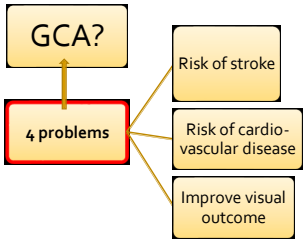




- Same vascular territory as brain (anterior circulation)
- Same mechanisms and causes as cerebral ischemia

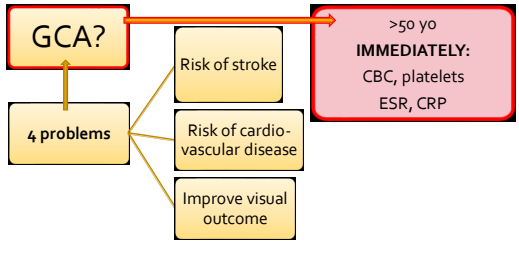
130

Acute Retinal Arterial Ischemia



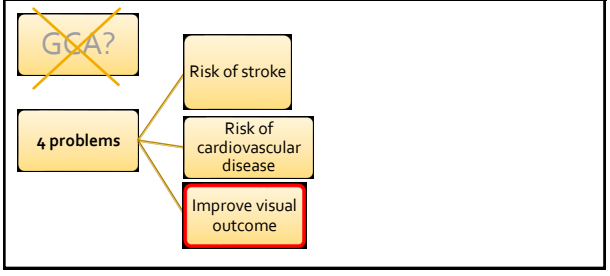
131

Acute Retinal Arterial Ischemia



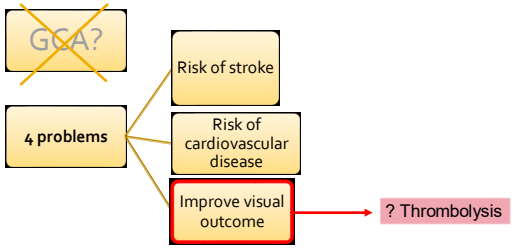
132

Acute Retinal Arterial Ischemia



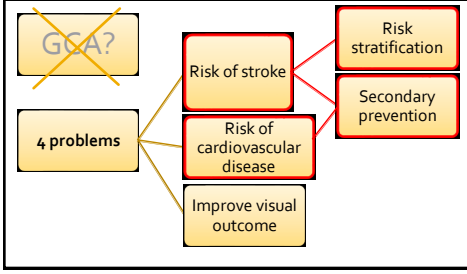
133

Acute Retinal Arterial Ischemia



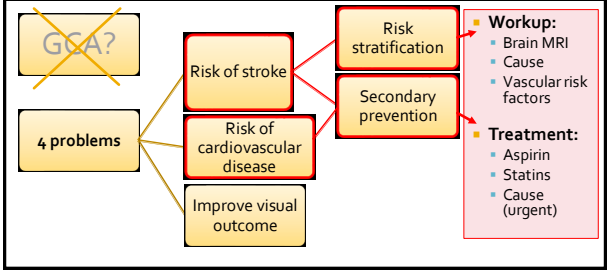
134

Acute Retinal Arterial Ischemia



135

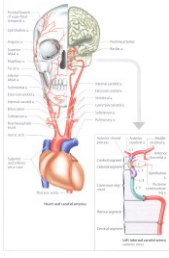
Acute Retinal Arterial Ischemia



136

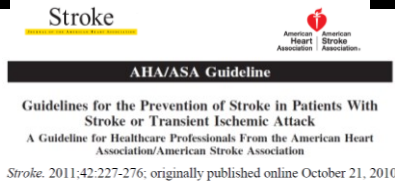
Acute Retinal Ischemia (OAO/CRAO/BRAO/TVL)

- **Risk of stroke highest within a few days after visual loss**
- Urgent workup allows immediate identification of major cause associated with highest risk of stroke
  - Carotid atheromatous stenosis
  - Carotid dissection
  - Cardiac source of emboli (atrial fibrillation)



137

International Guidelines



Stroke

AHA/ASA Guideline

Guidelines for the Prevention of Stroke in Patients With Stroke or Transient Ischemic Attack

A Guideline for Healthcare Professionals From the American Heart Association/American Stroke Association

Stroke. 2011;42:227-276; originally published online October 21, 2010;

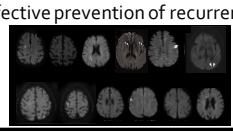
"Any patient with suspected TIA or those with acute retinal ischemia should be evaluated urgently in order to identify those at high risk of immediate cerebral infarction and cardiac ischemia""

138

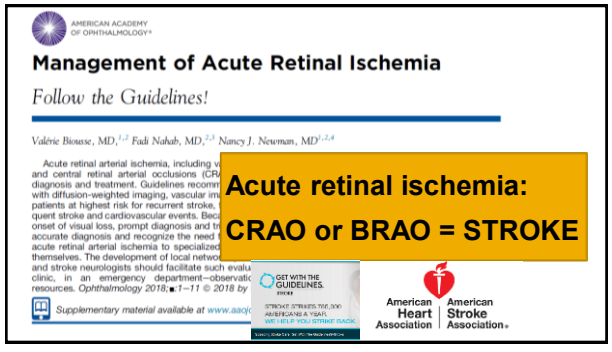
139

DWI-MRI In Acute Retinal TIA/Ischemia

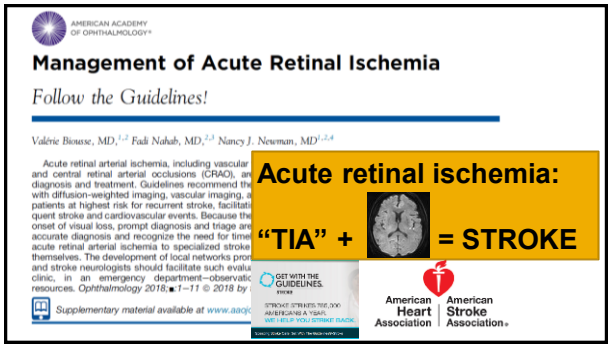
- Up to 53% CRAO, 31% BRAO, 18% TMVL patients have a positive DWI-MRI
- DWI-MRI identifies a subgroup of patients at very high risk of major stroke
- DWI-MRI needs to be performed within 24/48 hours of visual loss to allow for effective prevention of recurrent stroke



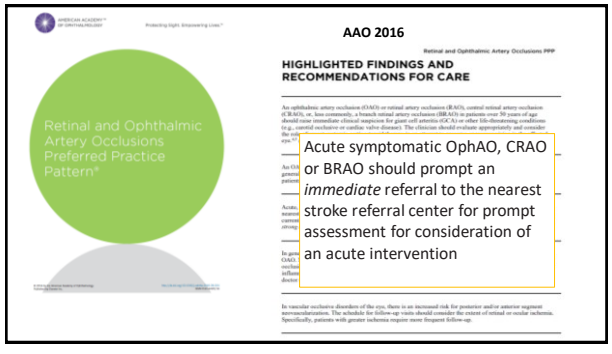
140



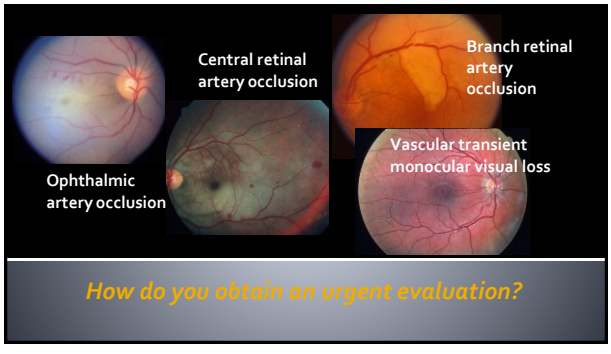
141



142



143



144

Disposition of the patient

Acute Stroke Care 24/7

American Heart Association American Stroke Association CERTIFIED Primary Stroke Center

Models vary among hospitals

Emergency Department

- Hospitalization
- Emergency Department Observation Unit
- Rapid Access TIA Clinic

145

Tell the patient:

- "Go to the Emergency Department"

American Heart Association American Stroke Association CERTIFIED Primary Stroke Center

- "Tell them you had a stroke in the eye"
- **Do NOT send these patients to their primary care physician, cardiologist, neurologist, retina specialist, neuro-ophthalmologist**
- **Do not try to obtain the workup yourself**

146

Call the Emergency Department Triage Provider:

- "I am sending you a patient who had
  - A **stroke** in the eye
  - For immediate stroke workup and treatment by stroke neurology"

147

Acute Retinal Ischemia: What to Do?

- Educate and help your colleagues
- Establish a network with closest Stroke Center and local stroke neurologist

Management of Acute Retinal Ischemia

Follow the Guidelines!

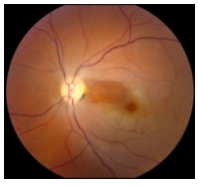
- **Simple message:**

1) Make the correct diagnosis
2) Send the patient immediately to a Stroke Center

148

71 yo man

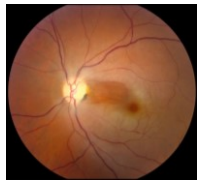
- With acute visual loss in one eye…



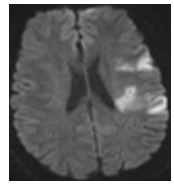
149

The next 24 hrs in the Emergency Department…

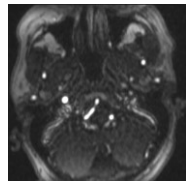
Seen in the Emergency Department 13 hours after acute visual loss
-Normal GCA labs
-No thrombolysis



Cardiac monitoring, observation, brain MRI/MRA, Neurologist, echo
-3 hours later, right hemiparesis and aphasia



Left carotid occlusion and left cerebral (MCA) infarction
-Thrombectomy / thrombolysis
-Good neurologic outcome



150

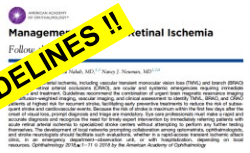
Acute Retinal Ischemia: What to Do?

- Educate and help your colleagues
- Establish a network with closest Stroke Center and local stroke neurologist

- **Simple message:**

1) Make the correct [illegible]osis

2) Send the pa[illegible]mediately to a Stroke Cen[illegible]



FOLLOW THE GUIDELINES !!

151

- 78 yo W woman with transient visual loss OD
- PMHx:
  - Atrial fibrillation on Coumadin
  - Recurrent falls
  - s/p cataract extraction/PCIOL OU

152

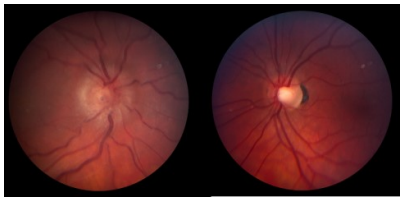
- Oct 30: sudden blurry vision OD
  - Resolved within 30 minutes
- November 2nd: blurry vision OD
  - Improved, but stayed blurry
- Seen on November 5

153

- Nov 5 (Optometrist):

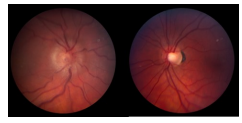
VA:	20/200 OD	20/30 OS
Color	2/14 OD	12/14 OS
Pupils	++ RAPD	Normal
Extraocular movements full		
HVF	Black OD	Diffuse depression
Fundus		

154



155

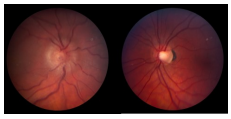
What next?



- 78 yo white woman
- AION
- No disc at risk
- Preceded by transient visual loss

156

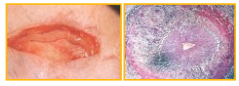
What next?



- 78 yo white woman
- AION
- No disc at risk
- Preceded by transient visual loss
  - **=> Giant cell arteritis very likely**
  - => Emergency department for immediate CBC/ESR/CRP, intravenous steroids, admission, evaluation and treatment

157

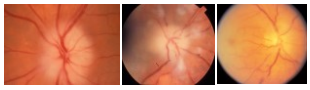
- Elevated ESR and CRP
- Temporal artery biopsy: florid inflammation



158

Visual Loss in Giant Cell Arteritis

- Severe, permanent (AION>PION>CRAO)
- Premonitory transient visual loss or diplopia in 65% within prior 8.5 days
- Second eye involved within 8 days in >70%
- Headaches helpful, but visual loss isolated in 25% of GCA cases



159

Treatment of GCA

- **Steroids treat GCA**
  - **Prototypical steroid-responsive disease**
  - Immediate and dramatic improvement of systemic symptoms and headaches
  - Vision protective

160

GCA: Steroids for Visual Loss

- **Immediately**
  - Visual outcome correlates with how fast steroid treatment is initiated
- High dose
- Route ? (IV vs PO)

161

British Society of Ophthalmology guidelines – *Initial Treatment*

Clinical presentation	Acute treatment protocol
Uncomplicated GCA (no jaw claudication or visual disturbance)	40-60mg oral prednisone daily (not less than 0.75 mg/kg daily)
Evolving visual loss (recent onset visual symptoms over 6-12 hours) **or transient visual loss**	IV methylprednisolone 500 – 1000 mg daily for 3 days before oral steroids
Established visual loss	At least 60 mg prednisolone daily, to protect fellow eye

Dasgupta B, Borg FA, Hassan N, et al. Rheumatology 2010;49:1594-1597

162

Why Intravenous is better:

- **Immediate** treatment
- You know when it happens
- Around the clock (250 mg qid)
- Patient in hospital
  - Monitored / prevention complications
  - Easier organization of consultations / follow-ups
  - Easier for patient and family / better compliance

  => Better (overall) outcome

163

Think Giant Cell Arteritis

- >50 yo
- Visual symptoms/signs
  - Visual loss (optic nerve, retina, choroid)
  - Unexplained visual loss
  - Transient visual loss
  - Diplopia
  - Transient diplopia
  - Cranial nerve palsies
- Headaches

164

47 yo AA Man

- Wakes up with no light perception OS
- Headache
- "Tired"

165

Sees his optometrist

- 20/20 OD; NLP OS
- Left RAPD
- Normal fundus OU

- Does not feel well
- Headache, a little "drowsy"
- BP: 141/95. HR 88

166

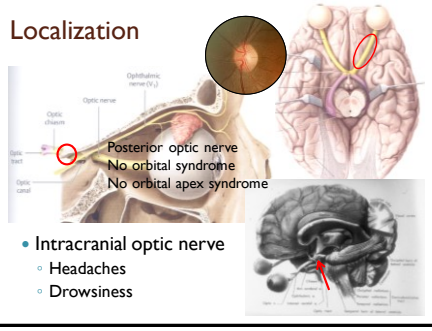
Optometrist calls us

- Do you want to see the patient today or do I send the patient to the ED?

- Emory ED!
  - (I know what he has)
    - Will need admission/treatment in hospital

167

Localization

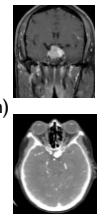


- Intracranial optic nerve
  - Headaches
  - Drowsiness

168

Diagnosis

- Acute posterior optic neuropathy
- Headache
- Drowsiness
  - Pituitary apoplexy
  - (Ophthalmic artery/sellar aneurysm)

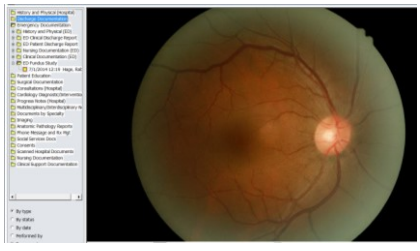


169

2 hours later:

- Patient evaluated in the ED

170

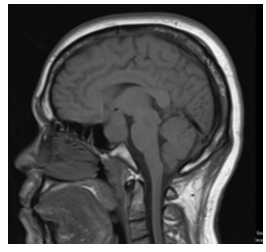


171

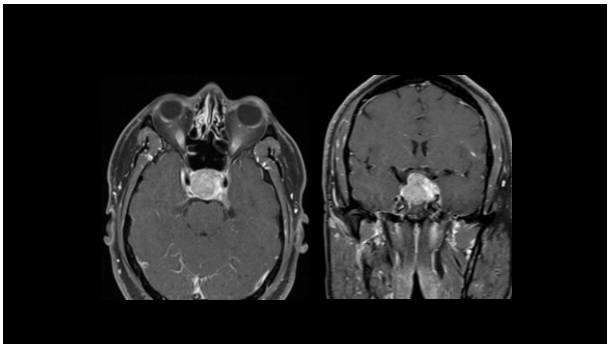
172

ED H&P

173



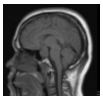
174

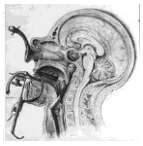


175

Pituitary Apoplexy

- Transphenoidal resection same night



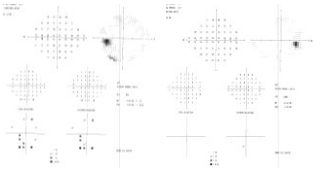


- Path: non-functioning pituitary adenoma
- Post-op: normal endocrine function

176

2 days later

- 20/15 OU
- Trace left RAPD
- Full color vision



177

3 days later

- Discharged on acetaminophen
- Back to work

178

What Went Right?

Better to call for advice than to obtain the wrong test

Better to refer to a specialized center

We knew what we were looking for

179

180

84 yo white man with

- Bilateral optic nerve edema
- PMHx:
  - Pace-maker (3rd degree block)
  - Afib/anticoagulated
  - Diabetes/mild diabetic retinopathy
  - Congestive heart failure
  - s/p cataract surgery – good outcome

181

Routine examination optometrist:

- Bilateral optic nerve head edema
  - No headache
  - No visual loss

182

Ophthalmologist:

- Bilateral disc edema
- => Head CT normal
- Normal/neg ESR, CRP, CBC, ACE, B12, folate, ANA, Bartonella, RPR

183

One month later:

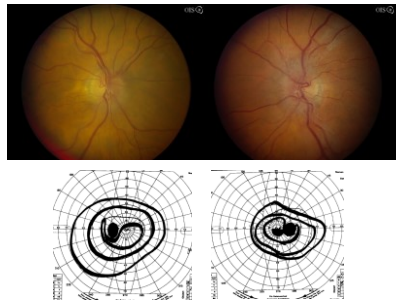
- Mild decreased vision
- Same optic nerve edema
- => Neurologist:
- LP: normal CSF OP, normal contents

184

One month later neuro-oph:

	OD	OS
VA	20/30-	20/30+
Color	12/14	13/14
Pupils	RAPD	
SLE	PCIOL	PCIOL
EOM	Full	Full

185



186

What are we missing?

- Lipitor
- Coreg
- Synthroid
- Coumadin
- Lisinopril
- Pacerone
- Aspirin
- Nexium
- Metformin

187

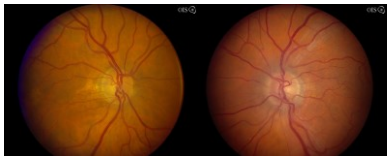
What are we missing?

- Lipitor
- Coreg
- Synthr
- Coum
- Lisinopril
- **Pacerone**
- Aspirin
- Nexium
- Metformin

Amiodarone

188

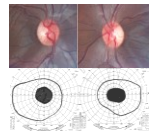
One month after d/c amiodarone



189

Toxic Optic Neuropathies

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



190

What Went Wrong?

Mistakes easily repeated when using electronic med record

- ophthalmologist's note: medication list copied from [illegible] student
- Neurologist's note: medication [illegible]

Always double check with patient possible "toxic" drugs

You only find what you are looking for

191

55 yo woman with diplopia

- PMHx: cholecystectomy, hysterectomy (endometriosis)
- Medications: None
- No tobacco, no alcohol
- Fam Hx: unremarkable

192

55 yo woman with diplopia

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

193

55 yo woman with diplopia

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

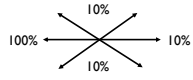
194

Neuro-Op consultation (March 25)…

- Neurologist not sure what to do at this point

195

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

196

Management:

- Right third nerve palsy
- Pupil involved
- Headaches
- Recent worsening
- Normal MRI/MRA

- 11:30AM, Friday
- MRI still at outside hospital

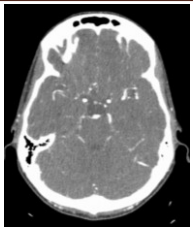
197

Management:

- 11:30AM: Warn neurosurgery
- 11:30AM: Send husband to pick up MRI
- 12:15PM: CT-Angiogram with contrast

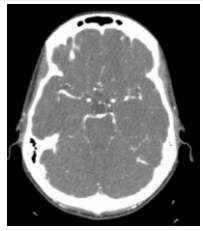
198

CTA:



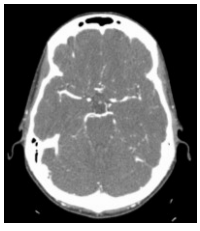
199

CTA:



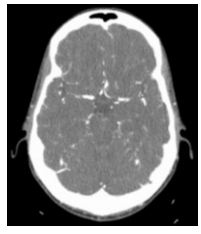
200

CTA:



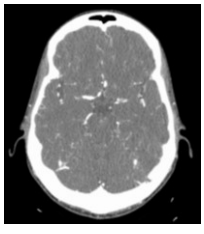
201

CTA:



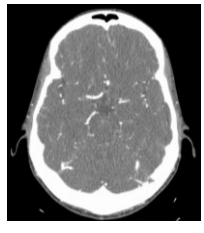
202

CTA:



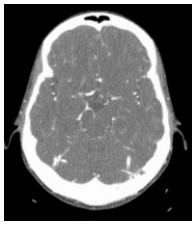
203

CTA:



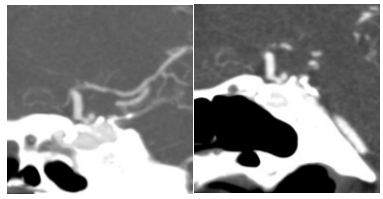
204

CTA:



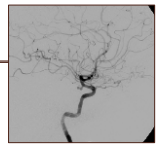
205

CTA:



206

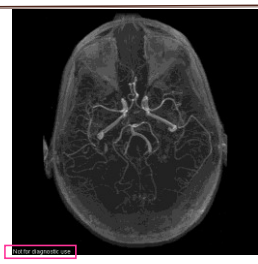
Management:



- 3:00PM: Neurosurgery OR
  - Angiogram in the OR
  - Right frontal craniotomy
    - Aneurysmal rupture at the time of clipping
- Post-op course uncomplicated
- 6 months later: EOM normal OD

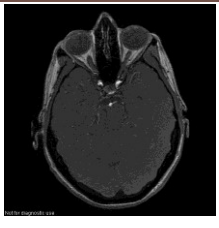
207

MRI/MRA (Husband back at 4PM)



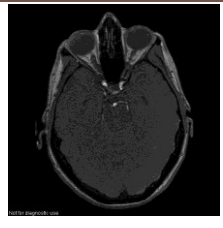
208

MRI/MRA



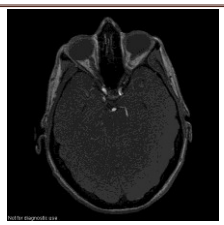
209

MRI/MRA



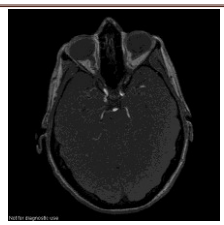
210

MRI/MRA



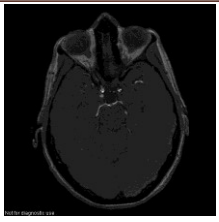
211

MRI/MRA



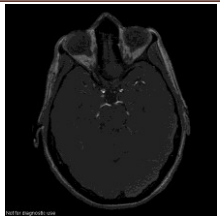
212

MRI/MRA



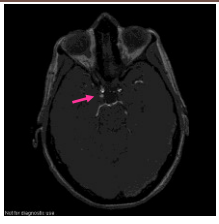
213

MRI/MRA



214

MRI/MRA

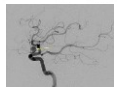


215

Underdiagnosis of Posterior Communicating Artery Aneurysm in Noninvasive Brain Vascular Studies

Valerie I. Elmalem, MD, Patricia A. Hudgins, MD, Beau B. Bruce, MD, Nancy J. Newman, MD, Valérie Biousse, MD

-Know what you are looking for
-Know the neuroradiologist
-Talk to the radiologist



216

- 48 year old man with difficulty reading after coronary artery bypass surgery
- PMHx:
  - Hypertension
  - Hypercholesterolemia
- Had good vision (reading glasses)

217

- Developed chest pain from angina
- Severe systemic hypertension
- => Coronary artery bypass graft (off pump)
- No immediate complication

218

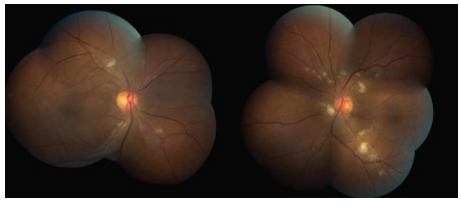
- A few days later:
  - Difficulty reading

219

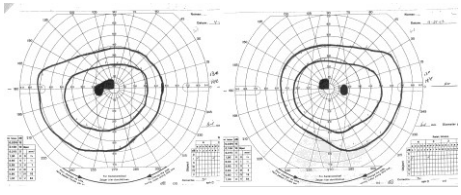
Seen Emergently

- BP 148/86; HR 58, regular
- Mild obesity
- Neurologic examination normal
- Visual acuity, color vision, confrontation visual fields, anterior segment, intraocular pressures, pupils, extraocular movements:
  - Normal

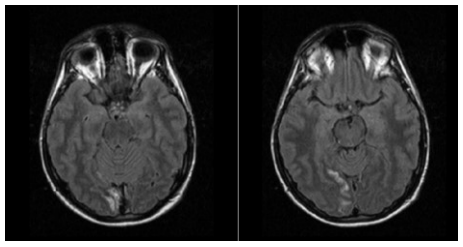
220



221



222



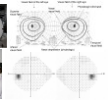
223

Confrontation Visual Fields

224

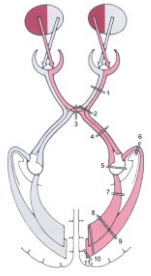
Confrontation Visual Fields

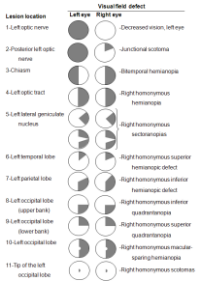
- Overall sensitivity is poor (50-74%)
  - 40% of anterior visual pathway lesions
  - 68-90% of homonymous hemianopias
- Detects only moderate or dense defects
- Using a red target may help sensitivity
- Always consider obtaining formal perimetry when the clinical findings don't fit
- Still do them!



225

Anatomy of Visual Pathways





Lesion location	Visual field defect: Left eye	Visual field defect: Right eye
1-Left optic nerve		Decreased vision, left eye
2-Posterior left optic nerve		Junctional scotoma
3-Chiasm		Bitemporal hemianopia
4-Left optic tract		Right homonymous hemianopia
5-Left lateral geniculate nucleus		Right homonymous sectoranopias
6-Left temporal lobe		Right homonymous superior hemianopic defect
7-Left parietal lobe		Right homonymous inferior hemianopic defect
8-Left occipital lobe (upper bank)		Right homonymous inferior quadrantanopia
9-Left occipital lobe (lower bank)		Right homonymous superior quadrantanopia
10-Left occipital lobe		Right homonymous macular-sparing hemianopia
11-Tip of the left occipital lobe		Right homonymous scotomas

226

Approach to the Interpretation of Visual Fields

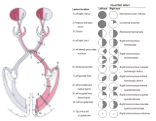
"The Four Questions"

227

Approach to the Interpretation of VFs "The Four Questions"

1) Does the field defect involve one eye or two?

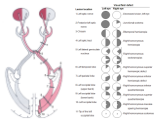
- If one eye: it's in the eye or optic nerve
- If two eyes: it's either bilateral eye/optic nerve or it's chiasmal/retrochiasmal



228

Approach to the Interpretation of VFs "The Four Questions"

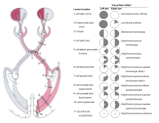
2) If two eyes, does the defect respect the vertical meridian?
- If no, then it's in the bilateral eye/optic nerve
- If yes, then it's in the chiasm/retrochiasm



229

Approach to the Interpretation of VFs "The Four Questions"

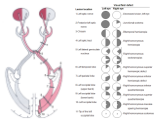
3) If it respects the vertical meridian, are the defects on the same sides of the vertical in each eye (homonymous) or bitemporal?
- If bitemporal, then it's in the chiasm
- If homonymous, then it's retrochiasmal on the other side



230

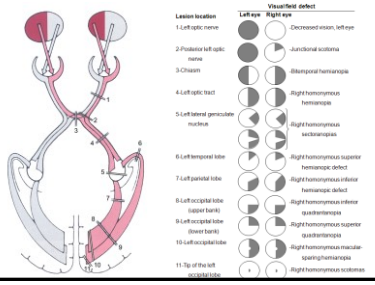
Approach to the Interpretation of VFs "The Four Questions"

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

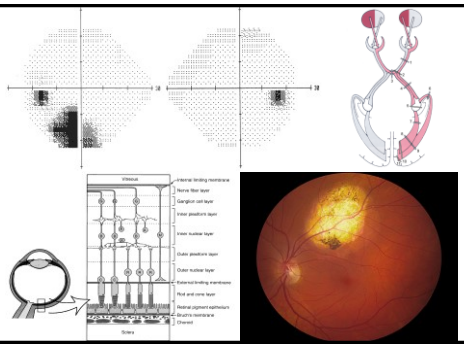


231

Anatomy of Visual Pathways

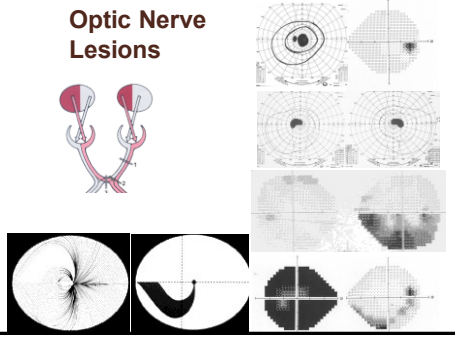


232



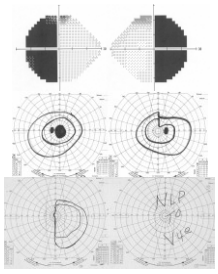
233

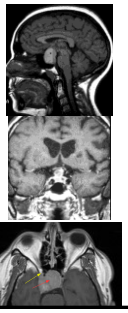
Optic Nerve Lesions



234

Chiasmal Lesions



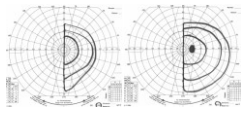


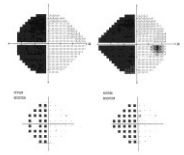
235

Complete Homonymous Hemianopia

Goldmann VF

Humphrey VF

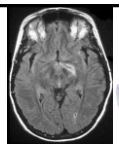


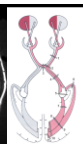


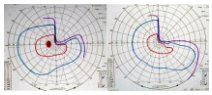
236

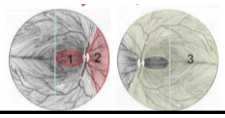
Optic Tract Syndrome

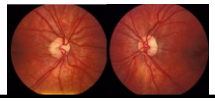
- **Left optic tract lesion:**
  - Right HH
  - Right RAPD
  - Bowtie atrophy of right optic nerve
  - Temporal pallor of left optic nerve





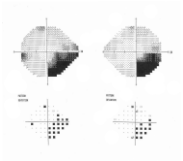


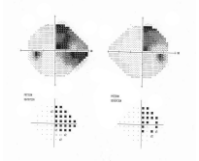


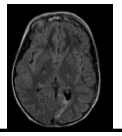


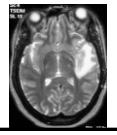
237

Incomplete Homonymous Hemianopia





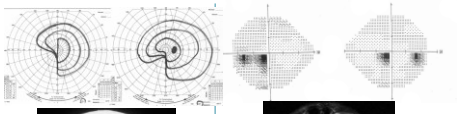


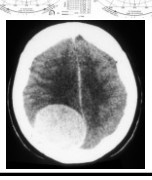


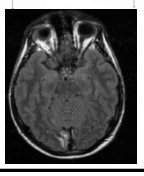
238

Incongruous HH / Congruous HH

Congruous = Posterior (occipital)



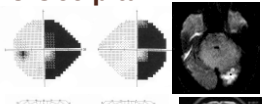




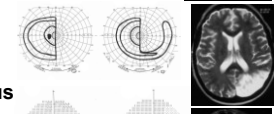
239

Always is Occipital

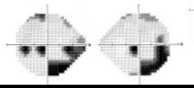
- **Macular sparing**

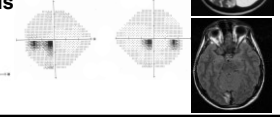


- **Temporal crescent**



- **Homonymous scotomas**

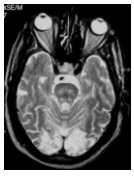




240

Cerebral Blindness

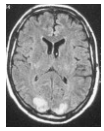
- Bilateral visual acuity loss
  - Equal in both eyes
- Normal pupils
- Normal fundus



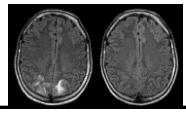
- Anton's: Patients with cerebral blindness say they can see (denial)

241

Bilateral Occipital Lesions
Cerebral Blindness

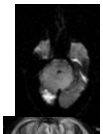


- Vascular:
  - Vertebrobasilar ischemia (PCAs)
  - Cerebral anoxia
  - Cerebral venous thrombosis
- Hypertensive encephalopathy
- Eclampsia
- Posterior Reversible Leukoencephalopathy (PRESS)
- Alzheimer
- PML
- CJD

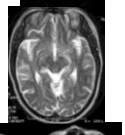


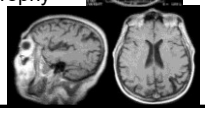
242

MRI "Negative" HH



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





243

Approach to the Interpretation of VFs
"The Four Questions"

1) Does the field defect involve one eye or two?
2) If two eyes, does the defect respect the vertical meridian?
3) If it respects the vertical meridian, are the defects on the same sides of the vertical in each eye (homonymous) or bitemporal?
4) If homonymous, is it complete or incomplete?

244