A Road Trip through the Visual Pathways of the Brain

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Principles of visual field testing

- Test one eye at a time
- Control fixation
- Test central +/- peripheral fields
- Chart carefully as the patient sees it
- Consider stimuli options
  - Size, intensity, color
Field testing methods

- Confrontation
- Amsler grid
- Tangent screen
- Goldmann kinetic perimetry
- Supra-threshold screening perimetry
- Static threshold perimetry
Confrontation fields

- Use your hands
- Avoid moving targets
- Use quadrants
- Count fingers or use red tops or hands
- Check for hemianopic desaturation
Amsler grid

Tangent screen
Perimeters
Stimulus size

- **Goldmann perimeter**
  - $0 = 0.625 \text{ mm}^2$
  - $I = 0.25 \text{ mm}^2$
  - $II = 1 \text{ mm}^2$
  - $III = 4 \text{ mm}^2$
  - $IV = 16 \text{ mm}^2$
  - $V = 64 \text{ mm}^2$

- **Humphrey/Octopus**
  - **Standard stimulus** = Goldmann III
Units of light intensity

- Maximum stimulus intensity
  - Humphrey = 10,000 asb
  - Octopus and Goldmann = 1,000 asb

- Background intensity = 31.5 asb

- Projected stimuli attenuated by neutral density filters
  - Attenuation in dB
  - 0 dB = no attenuation = 10,000 asb
  - 1 log unit of attenuation = 10 dB (1/10 of original intensity)
Stimulus intensity scales
Interpreting the HVF

- Reliability indices
  - Fixation losses
  - False neg/pos rates
- Total deviation
  - Compared to age-corrected normals
- Pattern deviation
- **Mean deviation**
- Pattern standard deviation
- Short term fluctuation
FIG. 6-5. GLOBAL INDICES. The MD and PSD indices are derived from the numbers in the total deviation plot. The SF index is derived from the preselected 10 locations that have duplicate threshold determinations. The CPSD index is calculated from the PSD and SF indices.
Perimetric Mean Deviation (PMD) = Average loss per test location weighted for the central points

Average loss = -5.13 dB

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<th>-4.41 dB</th>
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Choice of perimeters

- **Goldmann**
  - Kinetic and static testing
  - Excellent for very young or old patients, malingering, retrochiasmal lesions

- **Static threshold (Humphrey/Octopus)**
  - Requires an alert, cooperative patients
  - Excellent for long-term monitoring
    - Optic neuritis, PTC, glaucoma, pituitary tumors
  - Prone to testing artifact
Pearls of visual field testing

• Choose the field to answer a clinical question
• Check the central 30 degrees for neurological disease
• Always check the vertical meridian for a step
• Suspect results? Repeat the fields on another day or do a different type of field to confirm defect
Monocular visual field defects

- Optic nerve or retinal etiology
- Most retinal lesions have fundus findings
- Optic nerve defects have associated impaired acuity, color vision, and often relative afferent pupillary defects
Fig. 1-5. Nerve fiber pattern of retina.
Enlarged blind spot

• Optic nerve disease
  – Posterior staphyloma
  – Myelinated NFL at disc margin
  – Papillophlebitis
  – Papilledema

• Retinal disease
  – Big blind spot syndromes
Staphyloma
Papillophlebitis
Early Papilledema
“My left eye is flashing”

- 42-year-old woman with 2 month history of shimmering in OS
- Film over OS with washed out colors
- No eye pain at onset
Neuro-ophthalmic exam

- Visual acuity: 20/15, 20/15
- Color plates: 10/10, 5/10
- Pupils: Normal, Trace RAPD
- Fundus: Normal, Normal
Follow Up

- Patient followed now for 8 years
- Serial MRI scans unchanged
- BCVA OS now 20/60
- Fundus exam unremarkable
Repeat HVF OS
mf ERG Results
mf ERG color map
Optic nerve type VF defects

• Papillomacular bundle type
  – Central scotoma
  – Centrocecal scotoma
  – Paracentral scotoma
Central scotoma case
“I’ve lost vision in my left eye”

• 15-year-old boy elbowed in the left eye in a wrestling match
• Loss of central vision OS 2 days later
• Normal brain MRI
• 2 months later loss of vision OD
• No pain with onset of visual loss
• Normal brain and orbital MRI scan
Neuro-ophthalmic exam

- Visual acuity: 20/200 CF
- Color plates: Control Solids
- Pupils: Normal Mild RAPD
- Motility: Normal Normal
- Normal general neurological exam
Goldmann Visual Fields
Further Testing

- Brain MRI with gad negative
- No response to 3 days of IVMP 1 gm per day
- Subsequently blood testing positive for LHON 11,778 mutation
'My left eye is blurry’’

- 10-year-old girl with viral syndrome with fever
- Painless loss of central vision OS
- Left axillary node swelling mentioned to family doctor
Neuro-ophthalmic exam

- Visual acuity: 20/20 CF
- Color plates: 14/14 Control
- Pupils: Normal RAPD
- Motility: Normal Normal
Cat Scratch antibody testing

- *B. henslæ IgG*  1:128   1:256
- *B. henslæ IgM*  1:20   <1:20
- *B. quitana IgG*  1:64   1:256
- *B. quitana IgM*  <1:20   <1:20
Optic nerve type VF defects

• Arcuate NFB defects
  – Bjerrum or arcuate scotoma
  – Seidel scotoma
  – Nasal step
• Altitudinal defects
• Nasal nerve fiber bundles
  – Wedge shaped defects from blind spot temporally
Etiology of arcuate and altitudinal VF defects

• Optic nerve diseases
  – Glaucoma
  – Papilledema
  – ONHD
  – Optic neuropathies (AION, optic neuritis, etc.)

• Retinal disease
  – BRAO
“My right eye’s vision is worsening”

- 42-year-old woman with blurry vision OD for 2 months
- No eye pain
- Uses OS now to focus camera
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<tr>
<td>Pupils</td>
<td>RAPD</td>
<td>Normal</td>
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<tr>
<td>Fundus</td>
<td>OD cupped with temporal pallor</td>
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ophthalmic a. aneurysm
“I’ve lost vision in the top half of my right eye!”

- 23-year-old man in dirt bike accident with LOC
- Orbital CT scan: no fractures
- Marked orbital swelling OD prevented initial eye exam for 6 days
Neuro-ophthalmic exam

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Initial fundus appearance

Later fundus appearance
Binasal VF defects

• Think GLAUCOMA
• Most nasal defects are due to arcuate scotomas – PTC, ONHD
• Not a result of chiasmal compression
• Rarely secondary to compression of temporal aspect of optic nerve
“I failed my visual field test”

• 65-year-old woman who failed screening VF test at optometrist's office
• Patient rarely drives
• No major complaints regarding peripheral vision loss
Neuro-ophthalmic exam

- Visual acuity: 20/20 20/20
- Color plates: 8/10 6/10
- Pupils: Normal Normal
- Motility: Normal Normal
Optic Chiasm Visual Field Defects
Etiology of chiasmal VF loss

- Tumors
- Aneurysms
- Other processes
  - Sarcoidosis, sphenoid sinus mucocele, empty sella syndrome
- Demyelination, ischemia, radionecrosis, trauma, arachnoiditis
Epiphenomena of chiasmal defects

- Impaired vision
- Dyschromatopsia
- Post-fixation blindness
- Visual release hallucinations
- Hemifield slip (diplopia)
- See-saw nystagmus (craniopharyngioma)
Principles of the optic chiasm

- Nasal fibers crossed, temporal fibers uncrossed (53/47)
- Lower retinal fibers > lateral in optic tracts
- Upper retinal fibers > medial position
Wilbrand’s knee

- Inferonasal retinal fibers cross in the chiasm and course anteriorly for 4 mm in the contralateral optic nerve before turning back to join uncrossed inferotemporal fibers in the optic tract.
- Results in junctional scotoma:
  - Ipsilateral optic nerve defect and a superior-temporal VF loss in the contralateral eye.
“I can’t see very well”

- 28 yo woman with eye ache OD
- Subsequent visual blurring, OD then OS
- Unremarkable past medical history
# Neuro-ophthalmic exam

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<tr>
<td><strong>Fundus</strong></td>
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Optic chiasmal neuritis

Nancy J. Newman, MD; Simmons Lessell, MD; and Jacqueline M.S. Winterkorn, PhD, MD

Article abstract—In four of six patients with clinical optic chiasmal neuritis, MRI demonstrated abnormalities of the chiasm. Optic chiasmal neuritis may be the initial manifestation of multiple sclerosis, a reflection of established CNS demyelination, or an isolated clinical finding.

Recent advances in MRI allow detection of demyelinating lesions in the intraorbital, intracanaliculair, and intracranial portions of the optic nerves. Abnormal signal intensities and gadolinium enhancement may appear in these structures and correlate with symptoms and signs of optic neuritis. Demyelinating lesions of the chiasm may be present pathologically and suggested clinically. While in some cases MRI has shown chiasmal involvement, this has only rarely been correlated with clinical findings. We present six patients with clinical evidence suggesting demyelination of the optic chiasm. In four, MRI showed abnormal signal intensities and gadolinium enhancement. Extensive medical evaluation was unrevealing, including complete blood count, routine serum chemistries, erythrocyte sedimentation rate, and negative serologic tests for syphilis and Lyme disease. Thyroid function studies, serum cortisol, follicle-stimulating hormone, prolactin levels, angiotensin converting enzyme level, complement levels, antineutrophil cytoplasm antibodies, and anti-double-stranded DNA antibodies were normal. Antinuclear antibody titer was positive at a low level (1:40) and there was a slightly abnormal band in the gamma region on serum immunoelectrophoresis. The CSF was under normal pressure, acellular, and contained normal levels of protein and glucose. No myelin basic protein or oligoclonal bands were found. Brainstem auditory and
Patient follow up

• Patient initially treated as optic neuritis and diagnosed with MS
• Subsequently tested positive for NMO antibody
Principles of the optic chiasm

- Chiasm is mostly macular fibers
- Look for central defects, eg. central bitemporal hemianopia
- Check visual field carefully in the “normal” eye in a patient with monocular poor vision.
- Look for the upper temporal defect of a junctional scotoma!
“I got in a fight”

- 25-year-old man in a fist fight
- Left eye was grabbed by assailant
- Now no light perception OS
- Orbit explored OS-avulsed left optic nerve
Neuro-ophthalmic exam

- Visual acuity: 20/15 NLP
- Color vision: 12/12 Sutured
- Pupils: Dilated with drops OD
- Motility: Normal
Chiasmal VF defects

• Any VF defect that arises in the temporal field, comes to the midline, and stops is chiasmal until proven otherwise, even if only in one eye!
“My left eye’s blind spot is getting bigger.”

- 34-year-old man who covered right eye and noted a blind spot in OS
- Gradual worsening over 1-2 years
- No eye pain or headache
- No flickering lights
Neuro-ophthalmic exam

- Visual acuity: 20/20, 20/30
- Color plates: 7/10, Control
- Pupils: Normal, RAPD
- Motility: Normal, Normal
Chiasmal VF defects

- Complete or quadrant bitemporal defects
- Temporal Bjerrum scotoma to midline
- Bitemporal hemicentral scotomas
- Monocular temporal defects
- Junctional pattern
“My left eye has been blurry for a week”

- 37 yo man with gradual blur OS
- Slight ache with movement OS
- No hx of prior neurological symptoms
# Neuro-ophthalmic Exam

- **Visual Acuity**: 20/20, 20/50
- **Color Vision**: 12/12, Control
- **Pupils**: Normal, RAPD
- **Motility**: Normal, Normal
- **Focus**: Normal, Normal
Follow up

• Prolactin initially = 26,000
• One month on parlodel, prolactin = 475
• Visual acuity  20/20  20/20
• Color vision    Full    Full
• Pupils        Normal   mild RAPD
Post-treatment HVF
Classic bow-tie atrophy
Lesions that mimic chiasmal VF defects

- Retinopathies (RP/RD)
- Choroidal folds
- Tilted discs (defects cross midline)
- Hysteria
- Centrocecal scotomas
- Big blind spots
- Focal hypoplasia of the disc
Tilted optic discs
Dominant Optic Atrophy
Big blind spot syndrome

• Without disc elevation (Fletcher et al., *Arch Ophthalmol* 1988; 106:44-49)
  – Presumed to be a retinopathy
  – Peripapillary RPE alterations with focal ERG abnormalities
  – Similar self-limited enlargement of the blind spot
Retrochiasmal VF defects

- All retrochiasmal lesions result in contralateral HH
- Unilateral defects-no drop in acuity
- Incongruous vs congruous defects
- Upper vs lower field defects
  - Pie in sky
  - Parietal lobe
  - Occipital lobe (quadrantic defects)
Posterior visual pathways
Optic tract syndrome

- Field defects vary
  - Incongruous contralateral partial homonymous hemianopia
  - Complete fixation-splitting contralateral HH
  - No effect on visual acuity
- Contralateral RAPD may occur
- Ipsilateral temporal pallor, contralateral bow-tie atrophy
Optic tract incongruous homonymous hemianopia
Right tract demyelinating lesion
Retrogeniculate VF defects

• Meyer’s loop (inferior retinal fibers)
  – Pie in the sky pattern
• Superior retinal fibers course through the parietal lobe
  – Contralateral inferior homonymous quadrantanopsia or partial HH
“My eyes are not clear or comfortable”

- 28 yo myopic student with difficulty seeing the board
- Optometrist prescribed glasses, and found abnormal screening VF
- She denies any difficulty with peripheral vision
Neuro-ophthalmic exam

- Visual acuity: 20/20, 20/20
- Color vision: 16/17, 16/17
- Pupils: Normal, Normal
- Motility: Normal, Normal
- Fundus: Normal, Normal
Occipital Lobe Field Defects and Higher Cortical Function

Figure 7.01. The areas occupied by the primary visual cortex and the prestriate areas according to Brodmann’s classification. A, mesial surf.; B, lateral surface. (From R. Lindenberg, F.B. Walsh, and J.G. Sacks. Neuropathology of Vision. Philadelphia. Lea & Febiger, 1973.)
Vascular supply to visual cortex
Occipital lobe visual field defects

• Rarely: incongruous defects
• Maybe: complete hemianopia
• Probably: complete homonymous quadrantanopia
• Definitely:
  – Temporal crescent
  – Congruous homonymous scotomas
  – Complete hemianopia with sparing of fixation
Isolated homonymous hemianopia
Trobe et al., Arch Ophthalmol 1973;89:377

- Retrospective review of 104 cases in men 50-70 years old
  - 92 (89%) secondary to cerebral infarction
- 88% static or improved clinical course
- 18% demonstrated visual field improvement
- Sudden onset without prodrome
Occipital lobe VF defects

• Pearl: Reduced central acuity CANNOT be attributed to a macula-splitting hemianopia
“I’m having a migraine and lost my right visual field”

• 44 yo physician with history of migraine with visual aura (scintillating scotoma in hemifield)
• Patient awoke without headache with loss of both right hemifields persisting for 4 hours
• 2 nifedipine at onset did not abort the attack
Neuro-ophthalmic exam

- Visual acuity: 20/20 20/20
- Color plates: Misses digits in R hemifield
- Pupils: Normal Normal
- Motility: Normal Normal
- Fundus: Normal Normal
Occipital lobe infarction
Pessin et al., Ann Neurol 1987;21:290

- 35 consecutive patients with occipital stroke and homonymous VF defect
  - Embolism was the #1 stroke mechanism
    - Unknown source (11 patients)
    - Cardiac source (10)
    - Vertebrobasilar atheroma (6)
    - Migraine (6)
    - Systemic illness with coagulopathy (3)
  - TIA’s rare (3), headache common (21)
Temporal crescent
Benton et al., Brain 1980;103:83

• An area in the field of each eye for which there are no corresponding visual points in the other eye
• Perceived by a nasal crescent ot retina -> contralateral visual cortex in the anterior portion of the mesial surface of the occipital lobe
• Homonymous heminopia with sparing of the temporal crescent -> occipital lobe lesion
“I see colored lights to my left”

- 48 yo woman with history of migraine with aura
- Aura consists of fortification spectra
- Right-sided headache with distortion of left hemifield
- No response to dilantin trial for seizure
Macular sparing

- At least 5 degrees of the central field must be spared in both eyes on the side of the hemianopia
- Dual supply of macular cortex by MCA/PCA terminal branches
- Central homonymous hemianopia-hypoperfusion states
- Bilateral homonymous hemianopias with macular sparing result in constricted fields
“I can’t see to my left”

- 74 yo man with loss of left hemifields at 12:30 am
- Upon awakening next am, mild bifrontal headache
- Periodic multicolored flashing light in left hemifields
- Hx of asthma and gastric ulcer
**Neuro-ophthalmic exam**

- **Visual acuity**
  - 20/25  
  - 20/25

- **Color vision**
  - Normal  
  - Normal

- **Pupils**
  - Normal  
  - Normal

- **Motility**
  - Normal  
  - Normal

- **Fundus**
  - Normal  
  - Normal
Cortical blindness

- Occipital lesions may be asymmetric or sequential
- 22% of patients with unilateral occipital infarction may develop contralateral infarction within 3 years
- Symmetric visual acuity, normal pupils, normal fundi
Cortical blindness etiology

- Stroke
- Hypotension
- Ictal
- MS
- Adrenal leukodystrophy
- Congenital malformation
- Mercury
- Ethanol
- Intermittent porphyria
- Hypoglycemia

- Post-CABG
- Post-angiography
- Meningitis
- CJD
- Brain tumor
- Head trauma
- Lead
- Cis-platinum
- MELAS
- SSPE
Cortical blindness
Aldrich et al., Ann Neurol 1987;21:149

- Common causes of CB in 35 patients
  - Ischemic stroke (32%) - uniformly poor visual recovery
  - Cardiac surgery (20%)
  - Cerebral angiography (12%)

- Poor outcome associated with cognitive, language, memory impairment or bi-occipital lucencies on CT scan
“I became blind this morning.”

- 57-year-old truck driver with transient bilateral visual loss 2 weeks earlier
- Trouble seeing to right since Fall 1998
- Hx hypertension and diabetes
- Smoker
Neuro-ophthalmic exam

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Cortical blindness

• Stages of recovery
  – Perception of motion
  – Recovery of central vision
  – Recovery of binocular perception
  – Disappearance of visual agnosia, spatial disorientation

• Anton’s syndrome (rare with trauma) with amnesia and agitated delirium
Bilateral constricted VF

- Glaucoma
- Optic disc drusen
- Post-papilledema optic atrophy
- Retinitis pigmentosa
- Functional VF loss
“Everything is dark, like nighttime during the day.”

- 32-year-old woman s/p quinine overdose as suicide attempt
- Initial hearing loss and blindness (NLP)
- Exam findings
  - 20/30 BCVA OU
  - Control color plates only OU
  - Pupils 8 mm OU, minimally reactive
The visual variant of Alzheimer’s Disease
Levine DN, et al., Neurol 1993;43:305

- Difficulty reading, locating, and identifying items by sight
- Normal visual acuity, color vision
- Impaired contrast sensitivity for low spatial frequencies
- Early intact memory and intellect
- NF tangles and SP in occipitoparietal area
- MRI-only atrophy seen