Vision loss in the White Eye

Daryl Guest
UMeyecare

Introduction

- In daily clinical practice how do we diagnose?
- 18 second rule: that’s the average time it takes a doctor to interrupt you as you’re describing your symptoms. By that point, he/she has in mind what the answer is, and that answer is probably right 80% of the time. Dr Jerome Groopman

Systemized investigation

- Diagnostic sieve LINDOCARF
  - L – locations
  - I – intensity
  - N – nature
  - D – duration
  - C – occurrence
  - O – concurrence
  - A – aggravating factors
  - R – relieving factors
  - F – features

Diagnostic approaches

- Exhaustive method:
  - (Students while observational skills still developing)
  - Collect as much data as possible – correlate with evidence based practice – they hope mass of data offsets observational deficiencies
- Algorithmic method:
  - (NH&MRC glaucoma / diabetes guidelines)
  - Systemized evidence based approach but assumes DDx in many cases
- Differential diagnosis method:
  - (Hope students are here when qualify)
  - Compare targeted data collected with knowledge of disease presentation and epidemiology
- Pattern-recognition:
  - (Experienced clinicians)
  - Recognising the disease because they have seen it before – comes with experience but brings some danger
  - Skip steps in Algorithmic method – is this safe?

5% of US autopsies find lethal diagnostic errors. Diagnostic errors – the next frontier for patient safety. JAMA Volume 301 (10) 11 March 2009, p1060-1062
Ask a silly question.

Need to see beyond the presentation

Think how your question can be interpreted.

Think how your question can be interpreted.

Think how your question can be interpreted – are you getting the answer you think you are?

How thorough is a good history
Decision Path - analysis of Pupils

Consensual > Direct Chiasm to LGN
Hemifield Full Field Optic Atrophy

RAPD

Yes

Irregular Pupil

Other Pupil Defect?

Yes

No

Look for signs of disease in the angle

Is patient on drug that affects pupil reactions?

Miotic Mydriatic

Yes No

Ptosis

Yes No

IIIrd nerve/Horner’s Accommodation?Eye movements?

No

Lesion of the ciliary ganglion eg Dilantin Tofranil Melleril Valium eg Morphine Lavodopa

flow logic for DDx of unilateral swollen disc

Prof AJ Vingrys

unilateral disc edema

Normal VA (≥ 6/7)

Spot OR blur, ↓ CS, etc.

Color/Bright comp. 4 – 7 of 10

RAPD ~ 1-3+

No / minimal VF loss

↓ CS, CV

RAPD ~ 1-3+

major VF loss-

altitudinal

What must I NOT miss ON tumor

Retrobulbar Mass

Retropulsion, Proptosis, Orbital CT/MRI?

RNFL VISIBLE VISION LOSS: RAPD, CV

MDNOR

MINOR

VISION LOSS: RAPD, CV

MDNOR

BLOOD PRESSURE

NORMAL

ELEVATED

ALTERNATIONAL FIELD DEFECT

NE

Ischaemic Temporal arteritis

Papilitis (pain/APD) CN compression

CRVO NO normal papillitis ON Drusen

CT SCANS

Normal

Abnormal

Intracranial mass

CSF flow anomaly

LUMBAR PUNCTURE

Normal

Abnormal

Pseudotumour

Neurroglia

Sarcoïd

Spinal cord tumour

aetiology

causes of vision loss in the white eye

Anne Weymouth
pupils, ocular motility

Behind the white eye... BLOOD SUPPLY

NEURAL SUPPLY

contraceptive

van Noss and Bouman, 1967

contrast sensitivity

van Noss and Bouman, 1967

VA contrast sensitivity (MET)

VA = CS

VA ≠ CS

VA ≠ CS

who cares?

SYMMETRY

CORRELATION WITH VA/PATIENT VISION

VA contrast sensitivity (MET)

VA = CS

R 6/30 L 6/60

R 14 L 13 dB cone dystrophy

VA ≠ CS

R 6/4.8 L 6/4.8

18 dB R & L undiagnosed DM

VA ≠ CS

R 6/4.8 L 6/4.8

R 19 L 21 dB early retinal toxicity Rxs

VA ≠ CS

R 6/7.5 L 6/9.5

R 17 dB L 18 dB retinal dystrophy, paramac cysts

high/low contrast VA

Melbourne Edge Test

van Noss and Bouman, 1967

nyctalopia
**colour vision**

**CHOICE and INTERPRETATION**

If you can see one so nimber but not the other, you have shortsighted and cataracts.

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**perimetry**

Selecting the most useful test field:

- macular,
- central,
- peripheral

Protocol: threshold/suprathreshold

Stimulus colour: SWAP, red-on-white

Spatial-temporal characteristics: static, kinetic, FDT

but
- ceiling & floor effects
- increased variability in disease states
- perimetric test sensitivity vs temporal onset of disease

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**invisible retina: neurons**

- vasculature
- pigment
- nerve bundle
- glia

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**abnormal subjective results in the white eye**

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**perimetry**

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*Spry et al. 2002 IOVS, 42(6), 1404-10 Fig 4*
invisible retina...or not?

is this macula healthy?

testing neurons - ERG

flash ERG

bright flash response

structure vs function

where is the vision loss?

summary
Case 3

Using a diagnosis of exclusion, go through the tests that you would need to do, and the results that would rule out each potential cause of the vision loss outlined below.

A 76-year-old woman is referred to you by her general practitioner for management of an unexplained difference in colour perception between eyes. She is a painter and has noticed this change over the past year. She has seen 6 ophthalmologists since her symptoms began, all of whom advised that her ocular health was normal. She has had bilateral cataract extraction (RE: 4 years ago, LE: 10 years ago), a capsulotomy in the left eye one year ago, and has been using tear supplements four times a day for the past year.

Case 3 GH

Polymyalgia rheumatica + Rheumatoid arthritis > 10yr
Prednisolone 5 mg/day
Antidepressants
Plaquenil 200mg morning
Methotrexate 10-12mg/week 5 months
Drug hypersensitivities

Case 3 VA/Rx

VA R +1.75/-1.50 x 135 (6/7.5) L +1.00/-1.00 x 120 (6/9.5)

Case 3 Contrast

Contrast sensitivity (MET) R 14 dB L 10 dB

Case 3 Colour Vision

T2 Tritan Plate
L’ Anthony’s Desaturated Panel Test
TES R = 52 (AGE 74)
TES L = 138

R normal L Tritan defect
Case 3 perimetry
Medmont Central 30°
L: general depression  
R: WNL

Case 3 perimetry RoW
Visual field  
Humphrey red-on-white (discus 4')
L: WNL, depressed 4 dB cf RE  
R: WNL

Case 3 anterior eye

Case 3 anterior eye

Case 3 fundoscopy

Case 3 ERG
Full field, flash
Scotopic: normal rod function  
Photopic: reduced flicker & flash responses  
Inner retinal cone pathway dysfunction
Diagnosis: mild bilateral Plaquenil retinal toxicity, sustained, stable 12 months
Case 1

Using a diagnosis of exclusion, go through the tests that you would need to do, and the results that would rule out each potential cause of the vision loss outlined below.

A 68-year-old man has had cataract surgery (RE) two weeks previously, and has come to you for this first time for new spectacles. The cataract had been developing for some time, but the surgeon reports that the cataract operation has been a total success. He has an unremarkable history before the cataract surgery. The other eye (LE) underwent cataract surgery the previous year without incident.

Best corrected visual acuities are 6/9+ and left 6/6−, and during the refraction the patient comments on the difference in vision between the two eyes.

Case 1 – Visual Fields

Although automated visual fields in amblyopic eyes typically appear normal, all four types of amblyopia are associated with a generalized depression of light sensitivity, which is proportionately greatest at the fovea and highly correlated with visual acuity loss. In general, amblyopia is not associated with any area of focal loss of threshold light sensitivity. If a focal defect is present in the visual field of the amblyopic eye, organic causes of visual loss should be suspected.

Automated perimetry in amblyopia: a generalized depression.

Case 1 – Colour Vision

Deficient color vision in the amblyopic eyes was not related to the visual acuity and type of amblyopia.

Visual acuity and color vision deficiency in amblyopia.

Therefore, although all six amblyopes performed normally on both the HRR and D-15 color vision tests.

A Comparison of Color and Luminance Discrimination in Amblyopia

Case 1 – Pupils

Relative afferent pupillary defects (RAPD) were detected in 32.3% of patients with amblyopia by a modification of the swinging flashlight test.

Pupillary responses in amblyopia.

• Amblyopia is diagnosed by testing visual acuity in each eye separately, with the person wearing an adequate refractive correction, and after exclusion of ocular pathology.
**Case 2**

Using a diagnosis of exclusion, go through the tests that you would need to do, and the results that would rule out each potential cause of the vision loss outlined below.

An 83 year-old woman presents with sudden onset poor vision in one eye. She is fit and well for her age, although takes medication for hypertension and cholesterol. Visual acuities with pinhole are right 6/7.5 and left 6/36. The eye is slightly painful, but there are no other symptoms.

The only previous ocular history of significance is successful bilateral cataract surgery over a decade earlier.

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**Demyelinating neuropathies**

**Retrobulbar-Neuritis**

No obvious ophthalmoscopic signs 3-6 months Dr. sees nothing Pt sees nothing? OCT defects in 6-8 wks MS mast common cause but MS may also give papillitis mild edema may become apparent (1-2 weeks) due to axoplasmic stasis loss of RNFL and optic atrophy with time 6-8 weeks after onset

Other Neural Eye involvement:
- Retrobulbar-Neuritis
- RAPD when acute
- VF loss normal after event but often persists as restriction to Red on horizontal meridian.
- RG CVD (<5/10 comp or Ishihara) recovers after event
- VER increased peak time (P100 >5 ms delay), even after recovery
- INO or diplopia

Vision loss: mild blur (reduced CS) to HM during attack will return to near normal with time but vague CS loss persists after event

**Devic’s disease**

Fisher’s syndrome

Toxicity
- CO poisoning, metabolic problems

**Multiple sclerosis**

and is caused by:
- Multiple sclerosis most common cause of demyelination and RBN – viral trigger activates T-cells to cross BBB and act against own myelin sheaths. Gives a demyelinated plaque in heavily myelinated regions of CNS: ONs, periventricular white matter, brain stem, and spinal cord.
- Toxicity
- CO poisoning, metabolic problems
- Fisher’s syndrome
- GBS: ataxia, areflexia, ophthalmoplegia
- Guillain-Barré syndrome: (GBS) peripheral polyneuritis
- Devic’s disease

**Gullian-Barré syndrome:** (GBS) peripheral polyneuritis occurs most commonly in the setting of recent upper respiratory or gastrointestinal illness. The symptoms usually appear 5-7 days after the inciting event. The acute symptoms vs chronic = mild to no symptoms?

**Case 2 - Demyelination**

Demyelination usually produces a RBN (75%) and is caused by:
- Multiple sclerosis
- most common cause of demyelination and RBN – viral trigger activates T-cells to cross BBB and act against own myelin sheaths. Gives a demyelinated plaque in heavily myelinated regions of CNS: ONs, periventricular white matter, brain stem, and spinal cord.
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**Retrobulbar-Neuritis signs**

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- **Eye involvement:**
  - Vision loss: mild blur (reduced CS) to HM during attack
  - 50% on presentation, 80% sometime worsens first 7-10 days then improves over 30 days.
  - will return to near normal with time but vague CS loss persists after event
  - VER increased peak time (P100 >5 ms delay), even after recovery
  - RAPD when acute (unless bilateral Dr’s?): normal with recovery? Depends on VA.
  - VF loss normal after event but often persists as restriction to Red on horizontal meridian.
- **Other Neural**
  - **Demyelinating neuropathies**
    - OCT slight thinning of RNFL (~20%)
  - INO or diplopia
  - Also:
    - Uhthoff’s sign excessive fatigue – symptoms worsen with heat?
    - “hermitte” sign tenderness/parasthesia on neck flexion
    - Constipation/urinary retention: ataxia, weakness, numbness or paralysis in limbs

**Demyelinating neuropathies**

**Multiple sclerosis**

requires multiple lesions over multiple episodes (MRI) to identify "clinically definite MS" – CDMS vs CIS = clinically isolated syndrome

**MacDonald criterion:**

- **dissemination in space:** 3+ lesions peri-aqueductal grey + other AND
- **dissemination in time:** (new lesions 3/12 later)

**Chronic Progressive**

- **Probable:**
  - Age 20-50. Multiple neurologic defects (felmished). No other plausible cause and MRI finds isolated CNS lesions.

**Diagnostic criteria**

- Painful, sudden, acute-onset of visual loss = papillitis
- OCT defects found in MS treated pts without ON (n=70) and
- with ON (n=189) vs GON (n=24)

**OCT defects in MS**

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  - 
  - 

**MS pts tend to have generalized reductions with temporal loss of glaucoma - sup loss**

**Visual Field**

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**Clin Neurol Neurosurg 2010**

**OCT**

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**MacDonald**

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**Clinical classifications**

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**CIS = abnormal MRI plus optic neuritis NOT confirmed in time not due to a single isolated event**

**BV + Vision loss = multiple lesions recurrence = multiple events**

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