



# Optic Neuropathy Outside of MS



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No Disclosures

# Outline

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- Part I – Introduction – What is Optic neuropathy?
  - Definition of optic neuropathy
  - Clinical signs of optic neuropathies
  - Visual field correlates
- Part 2 Cases of Optic Optic Neuritis outside of Multiple Sclerosis
- - Typical optic neuritis vs Atypical optic neuritis
  - CASE 1 - NMO
  - CASE 2 - MOGAD
  - CASE 3 - GCA
  - CASE 4 – SARCOID-MOGAD
  - CASE 5 - Syphilis
- Summary

# What is optic neuropathy?

- Optic neuropathy is a global term describing dysfunction of optic nerve due to various mechanisms

Table 4-3 Mechanisms of Optic Neuropathies

**Papilledema (raised intracranial pressure)**

**Glaucoma**

**Inflammatory (optic neuritis)**

Idiopathic optic neuritis (associated with MS)

Neuromyelitis optica (Devic disease)

Infections

Systemic inflammatory disorders

**Vascular (ischemic optic neuropathy)**

Anterior/posterior

Arteritic/nonarteritic

**Compressive/infiltrative**

Neoplastic

Nonneoplastic

**Hereditary**

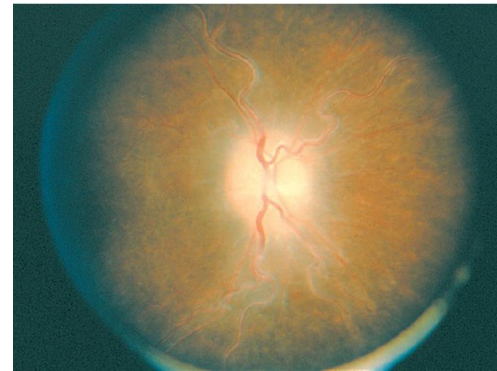
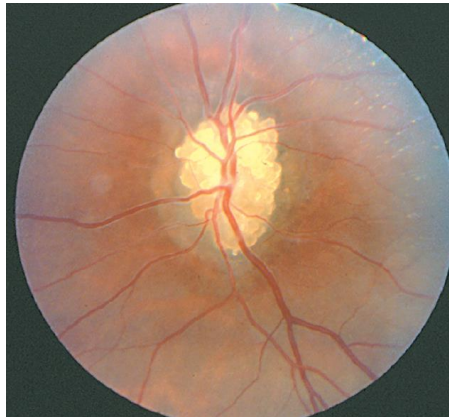
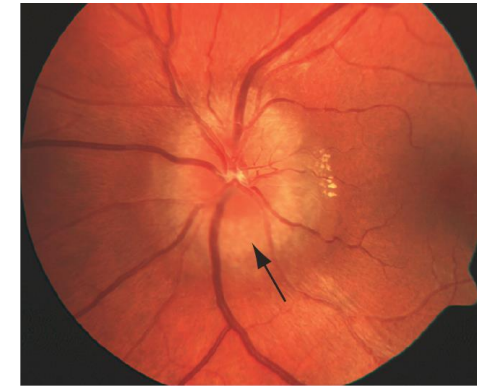
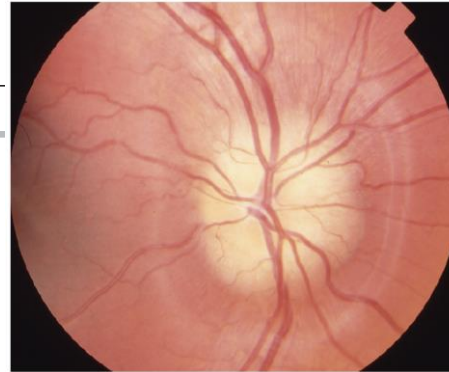
**Toxic/nutritional**

**Traumatic**

**Anomalous optic nerve**

Congenitally anomalous

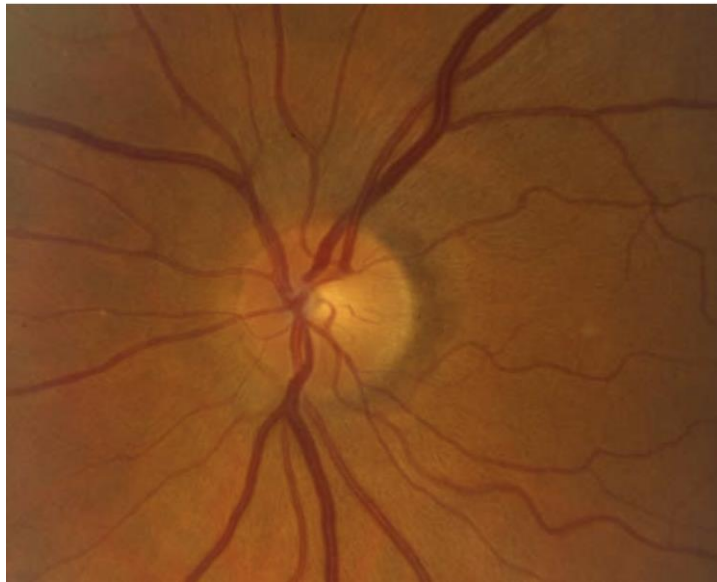
Drusen



MS = multiple sclerosis.

# Pathology of the optic nerve

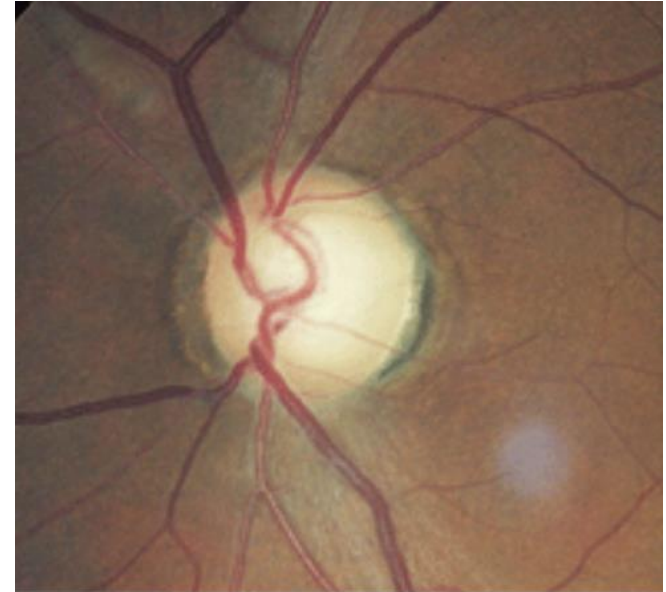
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No Swelling - Normal



Swelling



No swelling - pallor  
(irreversible injury)

These are the two ways in which acute pathology present in the optic nerves

**Swelling or No Swelling**

Evolution of injury: anterograde (7 days) vs retrograde degeneration (3-4 weeks)

Pallor develops 4-6 weeks after vision stabilizes

# Clinical Characteristics of Optic Neuropathies

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- Abnormal visual function (acuity may be normal)
- Acquired dyschromatopsia
- RAPD (if deficit is unilateral or asymmetric)
- Optic nerve can appear normal, edematous, or pale
- Visual field defects vary (can aid in diagnosis)
  - Scotoma may respect horizontal meridian, but not the vertical meridian

# Visual Acuities

Vision 20/200 after pinhole 20/20.

**Record:** 20/200 without correction, 20/20 with pin hole

Pin hole effect:

- Decreases blur

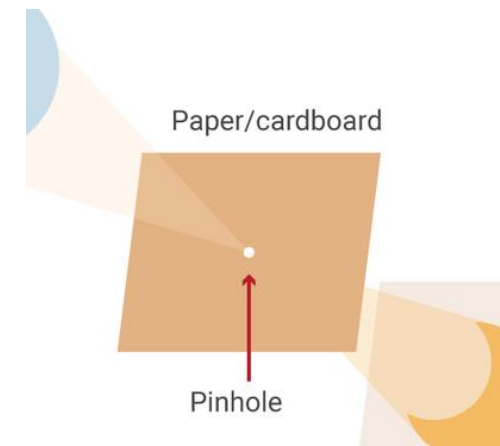
- Removes and reduces distortion (aberration)

- Increases depth of focus

- Decreases field of vision – don't walk around with pin holes!

Pin hole improves vision in cases of vision decrease due to:

- Refractive error
- Corneal disruption ( dry eyes, astigmatism)
- Lens disruption (cataracts)



# Visual Acuities

- Count Fingers and Hand Motion Vision
- When the patient can't see the card at distance or near (40 cm or 16 inches).
- Count fingers: 6 ft → 4 ft → 2 ft → 1 ft → inches
  - **Record:** *CF at @2 ft right eye.*
- Hand Motion: wave hands in each eye at 6 ft → 4 ft → 2 ft → 1 ft → inches
  - **Record:** *HM at 6 inches in upper half of vision, right eye*



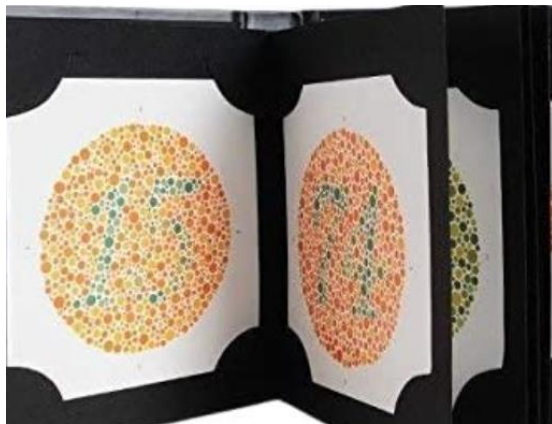
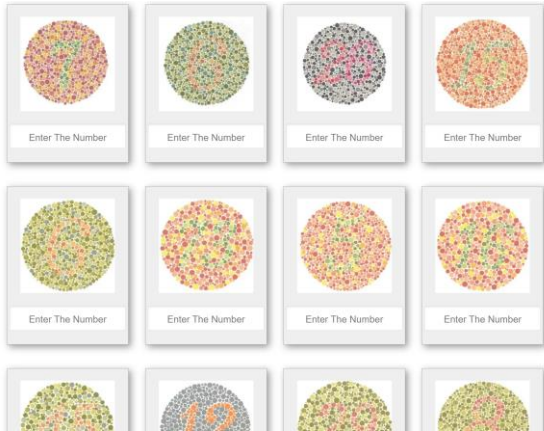


# Visual Acuities

- Light Perception vs Light Projection:
  - When the patient can't see finger counting or hand motion
  - Use pen light or small focused light source; test one eye at a time.
    - 6 ft → 4 ft → 2 ft → 1 ft → inches.
  - Light perception with projection:
    - If the patient can tell that the light is coming from a specific direction.
      - **Record:** *Light projection from upper left corner of right eye*
      - **Record:** *Light perception in the right eye.*
- Unable to see light:
  - **Record:** *NLP = No light perception*

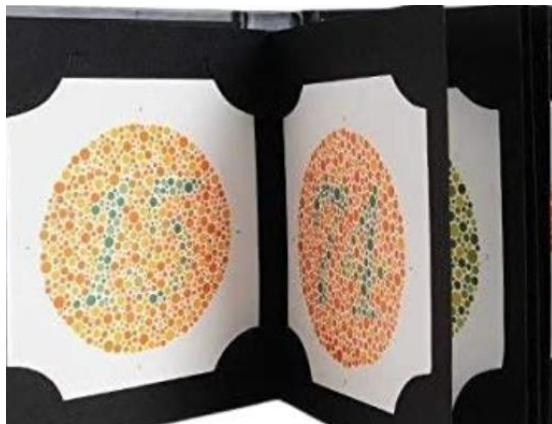
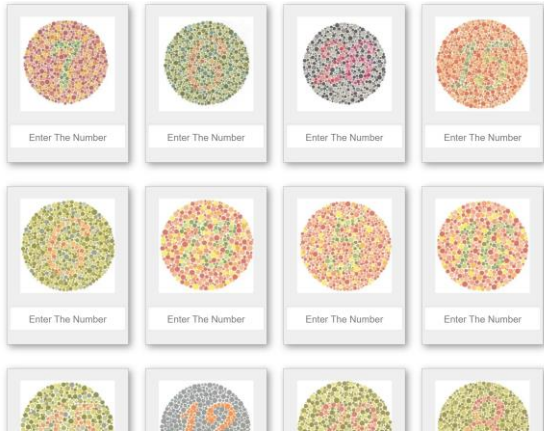


# Dyschromatopsia



- Ishihara color plates, preferred
- Test one eye at a time (right eye then left eye)
- If you have 20/200 or better visual acuity, you should be able to see the test plate
- Purpose of color vision testing:
  - Indicate whether there is dysfunction of the optic nerve or photoreceptors (cones/rods)
  - Localize the dysfunction (asymmetric)
    - Abnormal optic nerve function or injury to the retina causes decrease color vision ipsilaterally
  - Both eyes: congenital vs something acquired

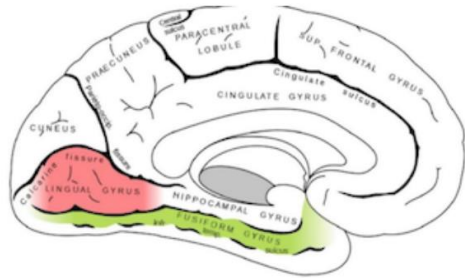
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  - Both eyes: congenital vs something acquired

**Are there brain lesions that can cause color vision loss?**

# Dyschromatopsia



**Yes, cerebral dyschromatopsia can occur due to a medial occipital lobe lesion.**

What about red desaturation?

Helpful to:

- Localize the side of optic neuropathy
- Nonspecific, just like many testing we do, it is about the clinical picture.
- In anything that affects the optic nerve can cause red desaturation



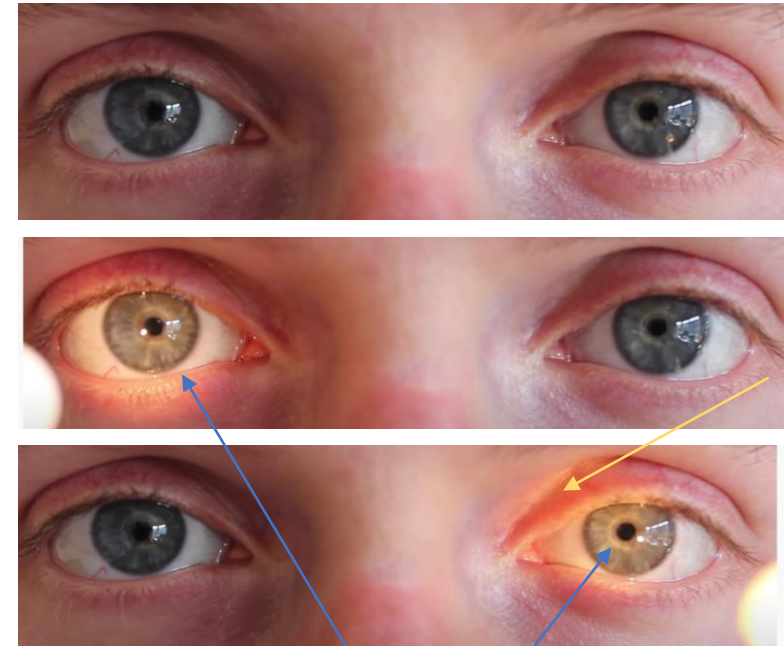
# Pupillary Responses

- **Direct Light & Consensual**

- In a dim room
- Patient fixates at a distance target
- Directly shine a light source on to the right pupil
  - Note the speed of reaction:
    - *brisk, fast, sluggish, minimal, fixed, amaurotic*
- Do this three times in one eye to get a good estimated speed, each 1-3 seconds duration.
- Repeat it in the other eye

- **Light source:**

- Preferably a muscle light or transilluminator
- Pen light
- Flashlight? (not the best) you want some thing focused into the pupil



Consensual  
Response

Direct Response

# Pupils Testing Technique

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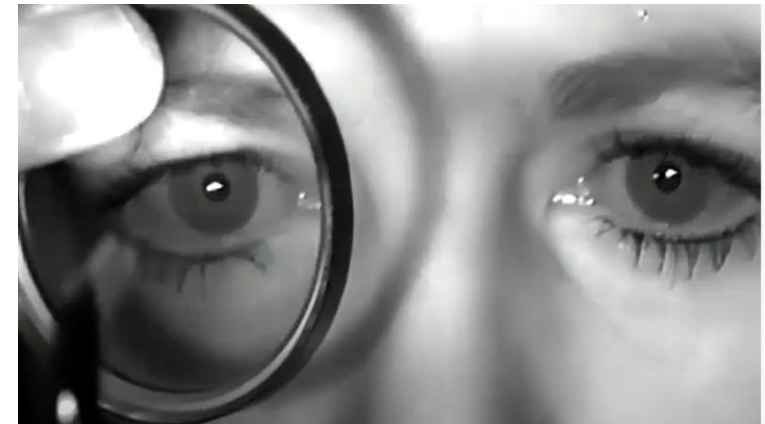
- **Swinging Flashlight Test**
  - Dim room
  - Patient fixates at a distance target
  - Direct a light in the one pupil 1-2 seconds
  - Swing to the other pupil leaving the light same time (1-2 seconds)
- **Evaluate and compare:**
  - Reaction of light between the left and the right.



# Grading RAPD

Clinical Grading of an Afferent Pupillary Defect

| Grade   | Neutral density filter | Pupillary response  |
|---------|------------------------|---|
| Grade 1 | 0.4 log units          | Weak initial constriction followed by greater re-dilation                         |
| Grade 2 | 0.7 log units          | Initial stalling followed by greater re-dilation                                  |
| Grade 3 | 1.1 log units          | Immediate dilation  |
| Grade 4 | 2.0 log units          | Immediate dilation following prolonged illumination of the good eye for 6 seconds |
| Grade 5 | Infinity               | Immediate dilation with no secondary constriction                                 |



## Amaurotic

This is seen when one eye has no perception of light. The pupil of this eye only constricts when light is shone into the other eye. When the light is shone back into the eye with no perception of light the pupil rapidly enlarges against the light.

## 3-4+

The pupil enlarges as soon as the light is swung from the normal eye into the abnormal eye.

## 1-2+

The pupil enlarges, but only after a short delay, after the light is swung from the normal eye into the abnormal eye.

## Subtle/ trace

Sometimes the pupils of both eyes can enlarge in the short time interval between shining the light in the normal eye and the abnormal eye.

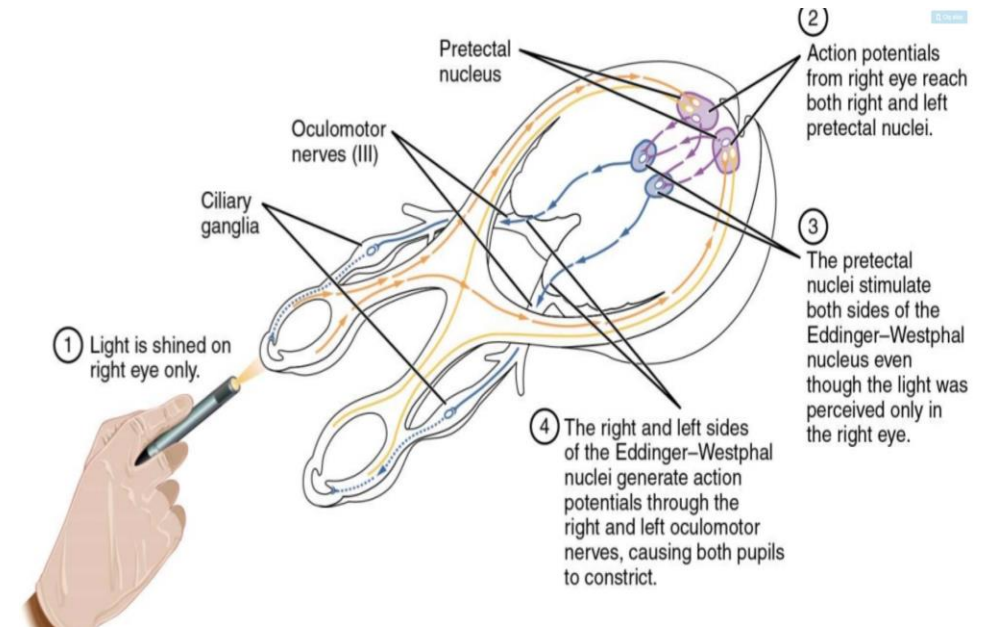
If this happens, the pupil of the abnormal eye may constrict a little bit before enlarging.





# Relative Afferent Pupillary Defect

- Damage at or anterior to the lateral geniculate
  - Optic neuropathy or optic nerve disease.
  - Ipsilateral visual loss
  - Asymmetric bilateral visual loss
- 
- Ocular diseases, such as corneal abnormalities, cataracts, and most retinal disorders do NOT cause RAPDs.



**RAPD cannot be caused by disorders of ocular media or refraction, even in extreme cases**

# What is the pupillary abnormality here?

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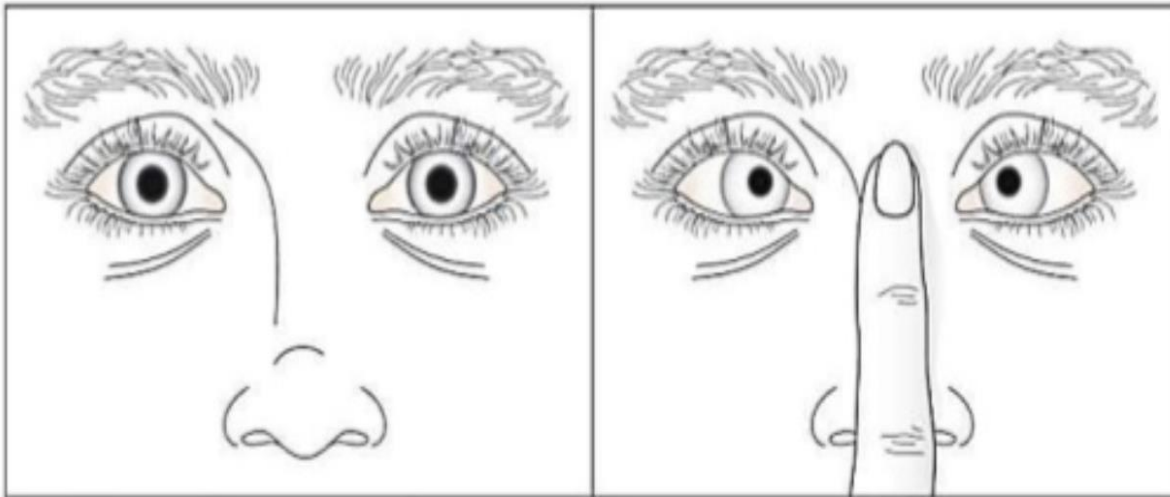


# Pupil Testing

## Technique Accommodation

L+ Clip

- The patient is asked to look at a distant object and then at an object close to his face.
- Both pupils should constrict and dilate again when distant gaze is resumed



- ▶ Document whether there is constriction to accommodation or not.
- ▶ **Light near dissociation:**
  - ▶ Pupils do not constrict to light, but will constrict to near/accommodation
  - ▶ There are a number of associated conditions.

# Light Near Dissociation – associated conditions

**Table 10-3 Causes of Light–Near Pupil Dissociation**

| <b>Cause</b>  | <b>Location</b>   | <b>Mechanism</b>   |
|---|---|--|
| Severe loss of afferent light input to both eyes                  | Anterior visual pathway (ie, retina, optic, nerves, chiasm) | Damage to the retina or optic nerve pathways   |
| Panretinal photocoagulation, retinal cryotherapy, orbital surgery | Short posterior ciliary nerves                              | Aberrant regeneration of accommodative nerve fibers  |
| Peripheral neuropathy   | Short posterior ciliary nerves                              | Disproportionate loss of axons to iris sphincter   |
| Adie tonic pupil  | Ciliary ganglion  | Aberrant regeneration of iris sphincter by accommodative neurons   |
| Aberrant regeneration of CN III                                   | CN III  | Aberrant regeneration of iris sphincter by accommodative neurons or extraocular muscle neurons                   |
| Loss of pretectal light input to Edinger-Westphal nucleus         | Midbrain tectum   | Infection (eg, Argyll Robertson pupils secondary to syphilis) compression (eg, pinealoma), ischemia (eg, stroke) |

CN = cranial nerve.

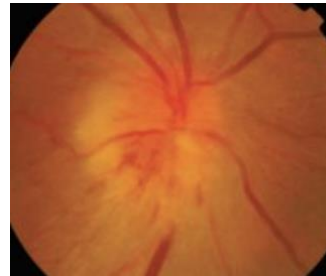
# Appearance of Optic Disc

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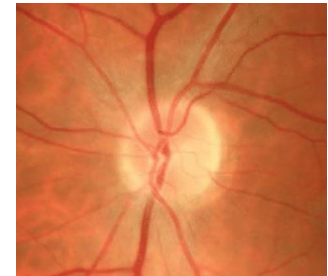
- Acutely the optic nerve may appear normal or edematous
- Evolution of injury: anterograde (7 days) vs retrograde degeneration (3-4 weeks)
  - Pallor develops 4-6 weeks after vision stabilizes
- Chronically the nerve appear pallid



Normal



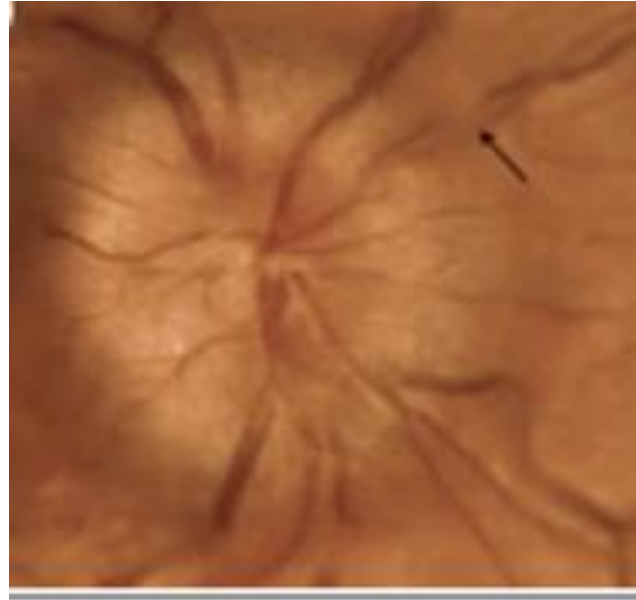
Optic nerve edema due to ischemia



Optic nerve

pallor

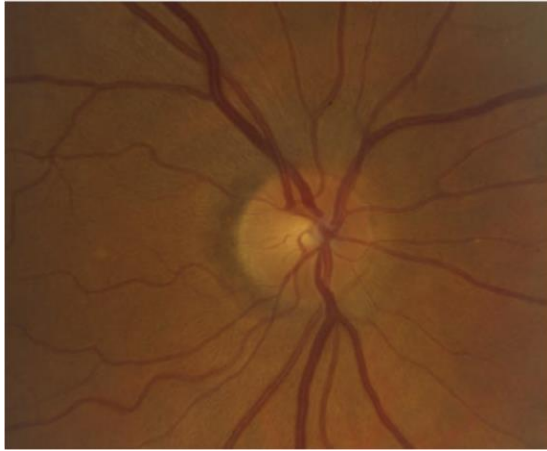
# Optic Nerve Swelling



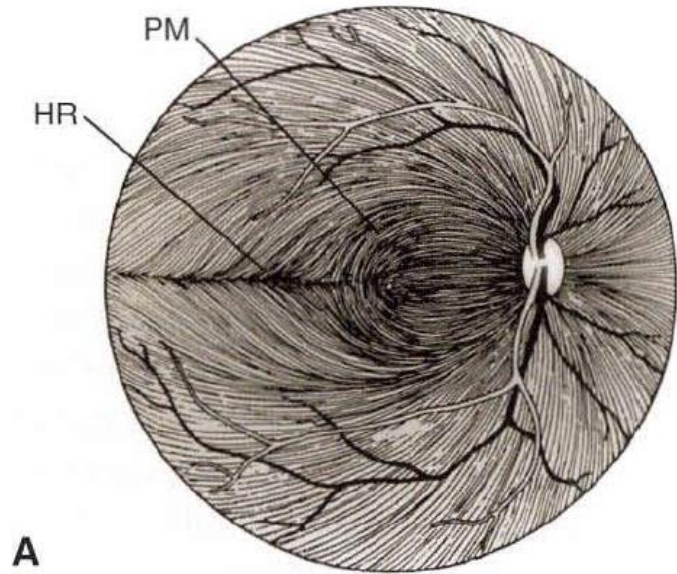
WHAT'S PAPILLEDEMA, AND WHY IS THIS NOT PAPILLEDEMA?

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# Optic Disc



Optic disc diameter 1.5 mm



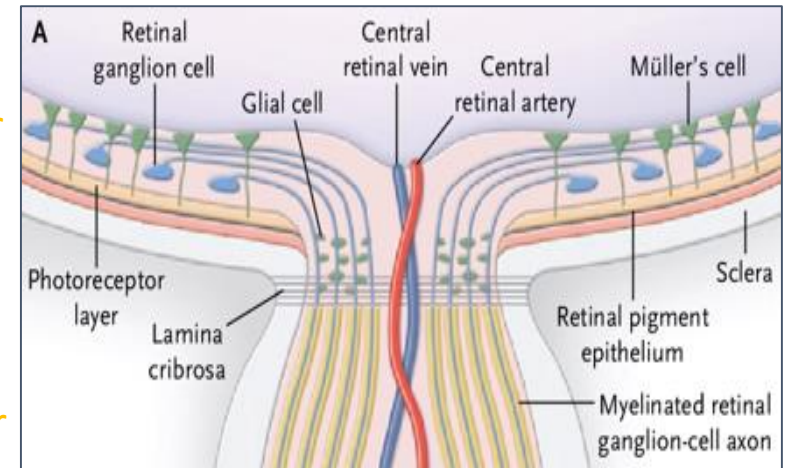
A

- Retinal Ganglion Cell Layer: 1.1 to 1.2 million cells distributed prelaminar zone.
- Nerve Fiber Layer: 1.1 to 1.2 million axons traverse the retina and makes a 90 degree turn towards the bundle of nerves
- Optic disc is the prelaminar zone (size of dime) containing 1.1 to 1.2 axons bundled tightly together.

Prelaminar

Laminar

Retrolaminar



A

# Structure & Function: Evolution of injury



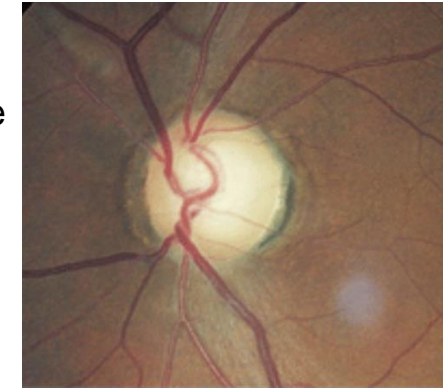
No Swelling - Normal

Acutely:  
Axoplasmic stasis  
Lack of cellular integrity  
Compartment syndrome  
Lack of vascular blood supply

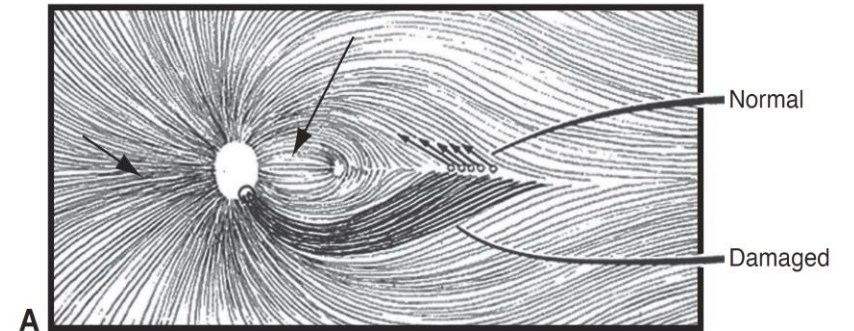
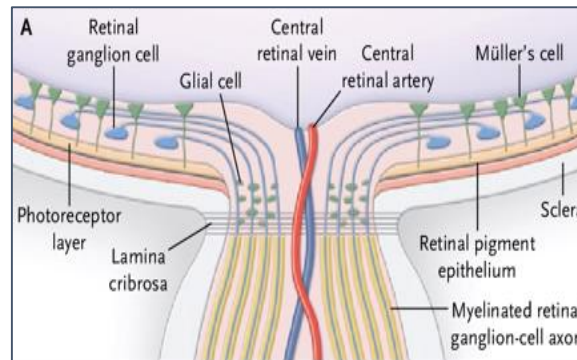
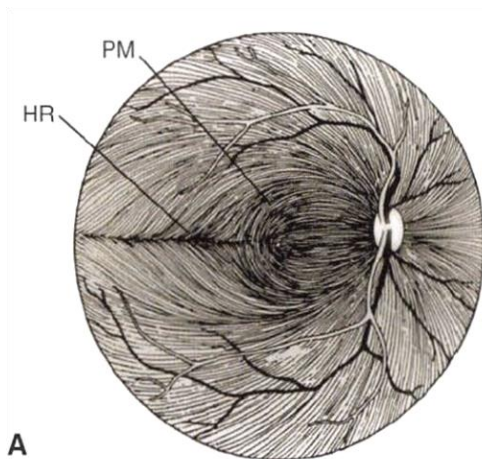


Swelling

Anterograde  
(7 days) or retrograde  
degeneration (3-4  
weeks)  
Causes drop out of  
nerve fiber & loss of  
axons



No swelling - pallor  
(irreversible injury)

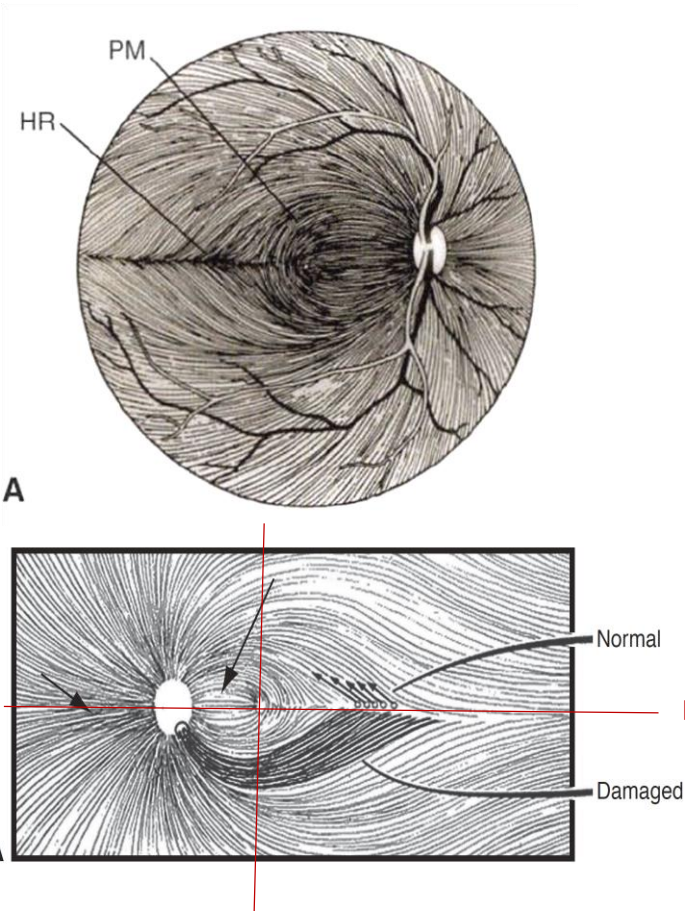


**Loss of axons & ganglion cells result in patterns of visual loss**

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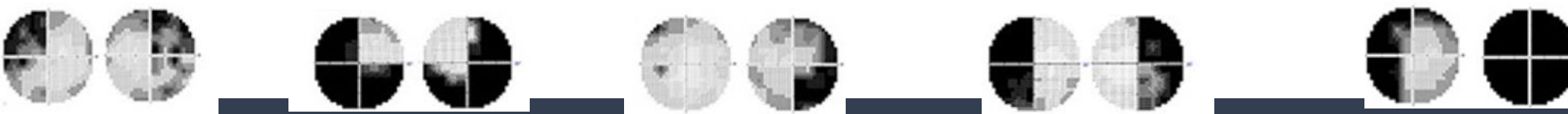


# Visual Field Patterns in Optic Neuropathy



Three types of nerve fiber loss:

1. *papillomacular fibers*:  
 cecocentral scotoma  
 paracentral scotoma  
 central scotoma
2. *arcuate fibers*:  
 arcuate scotoma (nerve fiber bundle defect)  
 altitudinal defect (broader region of arcuate fibers)  
 nasal (step) defect (temporal portion of arcuate fibers)
3. *nasal radiating fibers*: temporal wedge defect

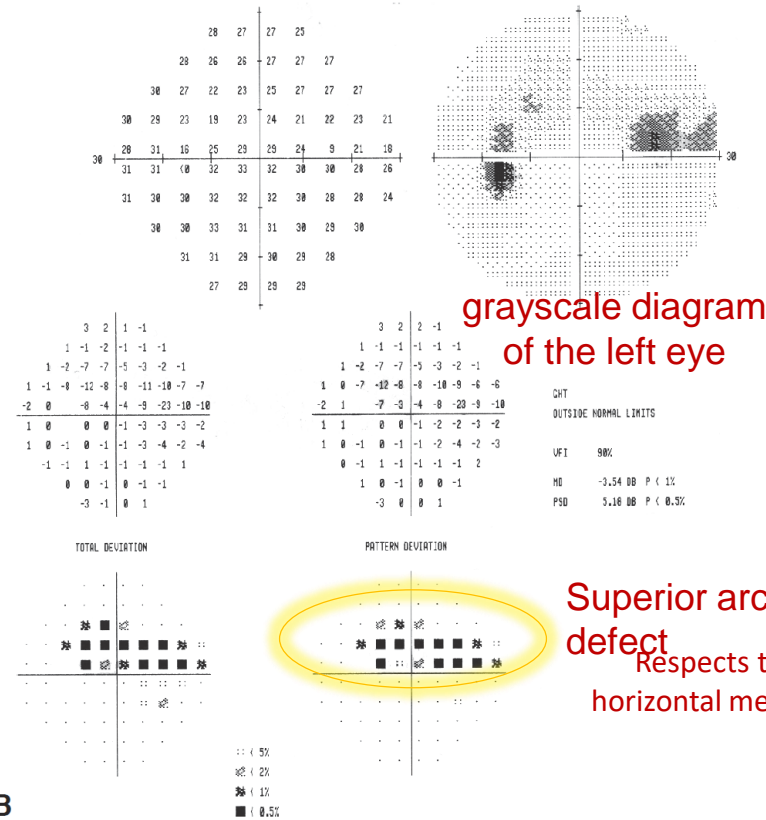
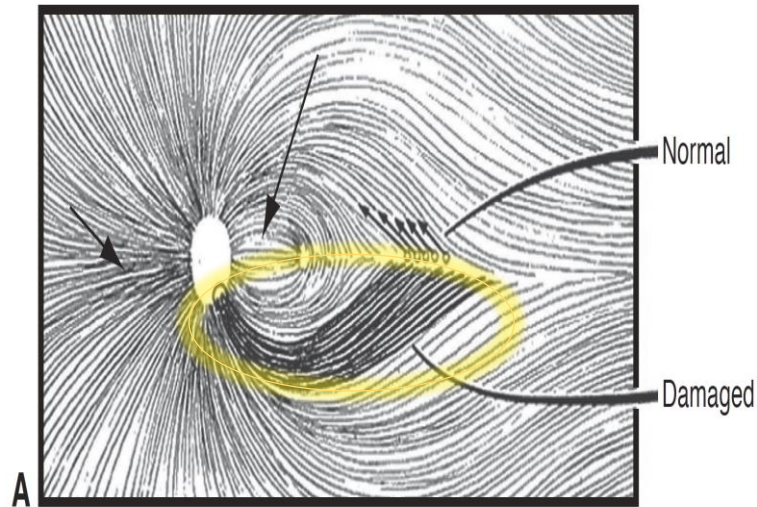


junctional scotoma

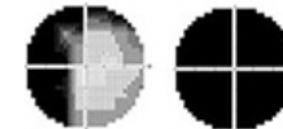
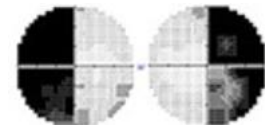
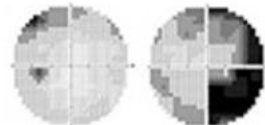
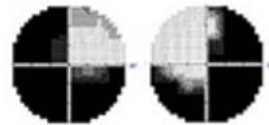
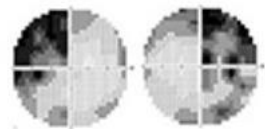
# Visual Field Patterns in Optic Neuropathy

Humphrey 30-2 perimetry:

Optic Neuropathies  
 inferior arcuate nerve fiber bundle damage leading to superior arcuate visual field defect.



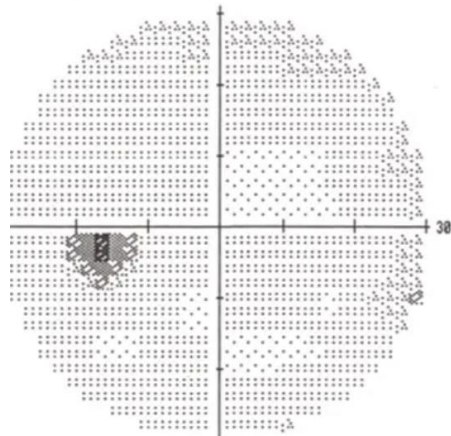
Inferior arcuate nerve fiber bundle damage



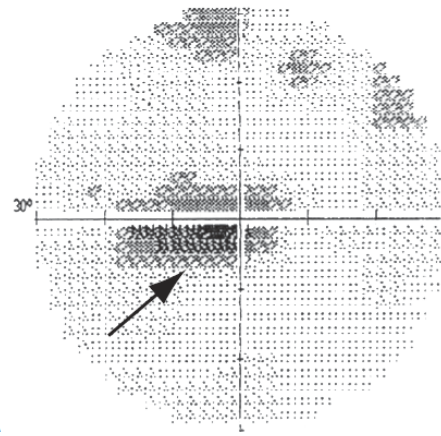
junctional scotoma

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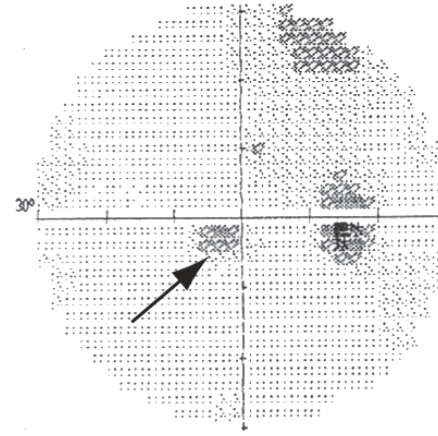
# Visual Field Patterns in Optic Neuropathies



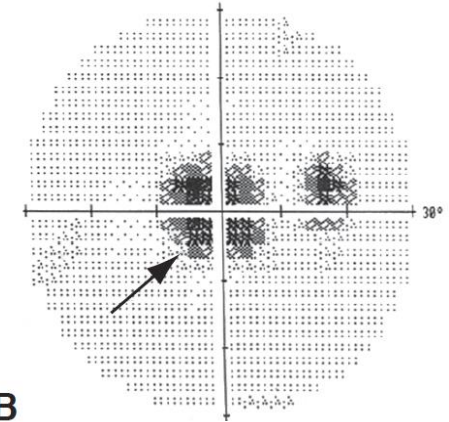
Normal



Cecocentral scotoma

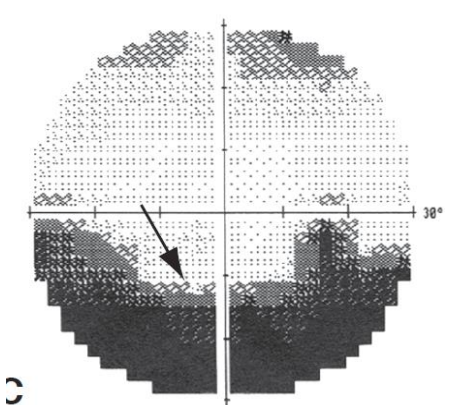


Paracentral scotoma



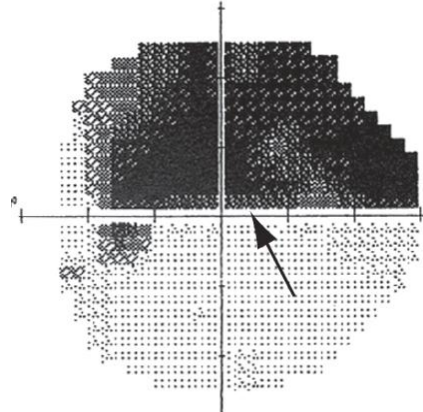
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Central scotoma

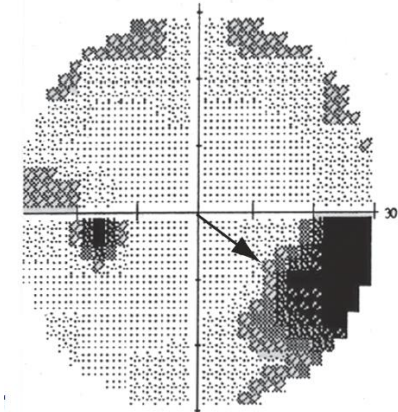


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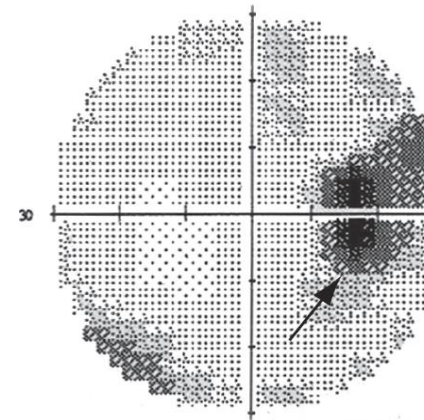
Arcuate  
scotoma



Altitudinal defect



Nasal step



Enlarged blind spot

# Visual Field Patterns in Optic Neuropathies



Normal



Cecocentral scotoma



Paracentral scotoma



Central scotoma



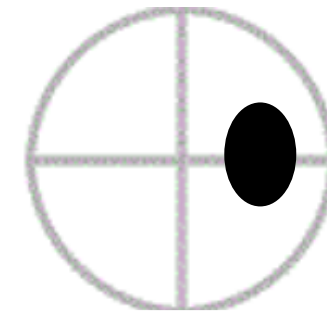
Arcuate  
scotoma



Altitudinal defect



Nasal step



Enlarged blind spot

# History



- Which eye? **>65 years Temporal Arteritis**
  - Onset, timing, duration, frequency
  - Pattern of vision loss
  - Symptoms associated with vision loss
  - Any associated systemic symptoms
    - Painless or painful
    - Pain with eye movement → optic neuritis
    - Facial numbness, diplopia → cavernous sinus lesion
    - Headache + jaw claudication + systemic symptoms → GCA
- Clues of underlying systemic disease process
  - Medications and recent changes
- Family, Surgical and Social history: decreased vision, vision loss at a young age, degenerative neurologic conditions.

Table 4-3 Mechanisms of Optic Neuropathies

|  |
|--|
| Papilledema (raised intracranial pressure)     |
| Glaucoma                                       |
| Inflammatory (optic neuritis)                  |
| Idiopathic optic neuritis (associated with MS) |
| Neuromyelitis optica (Devic disease)           |
| Infections                                     |
| Systemic inflammatory disorders                |
| Vascular (ischemic optic neuropathy)           |
| Anterior/posterior                             |
| Arteritic/nonarteritic                         |
| Compressive/infiltrative                       |
| Neoplastic                                     |
| Nonneoplastic                                  |
| Hereditary                                     |
| Toxic/nutritional                              |
| Traumatic                                      |
| Anomalous optic nerve                          |
| Congenitally anomalous                         |
| Drusen   |

MS = multiple sclerosis.

# Clinical Signs of Optic Neuropathy



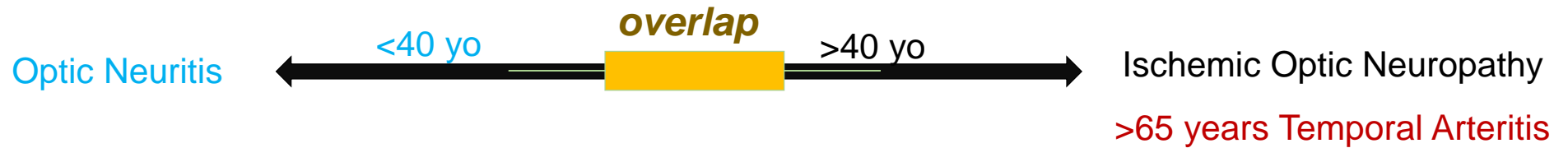
- ✓ Abnormal visual function (acuity may be normal)
- ✓ Acquired dyschromatopsia
- ✓ RAPD (if deficit is unilateral or asymmetric)
- ✓ Optic nerve can appear normal, edematous, or pale
- ✓ Visual field defects vary (can aid in diagnosis) - Scotoma may respect horizontal meridian, but not the vertical meridian



| Optic Neuropathy Pattern Recognition Table |   |
|--|---|
| Change in Vision                           | ✓ |
| Dyschromatopsia                            | ✓ |
| RAPD +                                     | ✓ |
| Optic Disc: normal edema, pale             | ✓ |
| VF defect respects the horizontal meridian | ✓ |

# Apply what you learned

- The stakes are high.



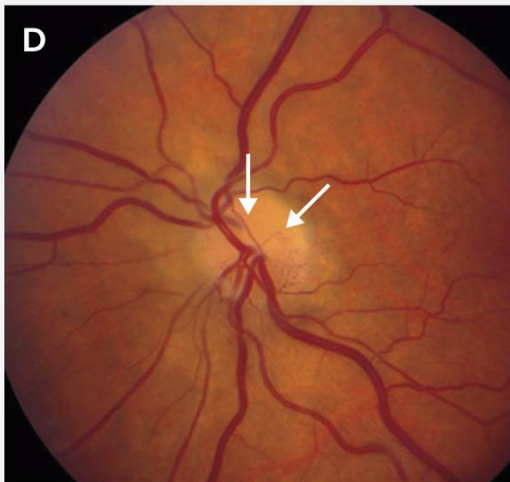
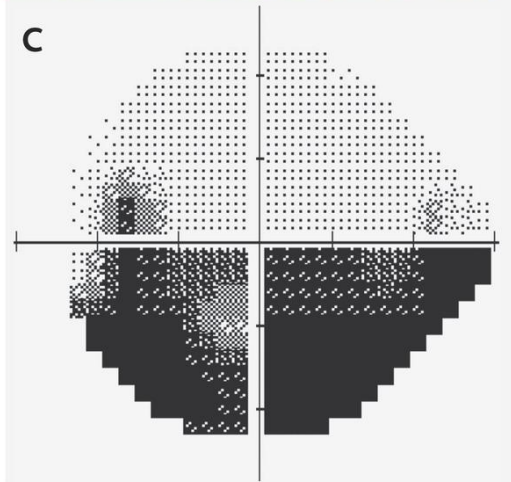
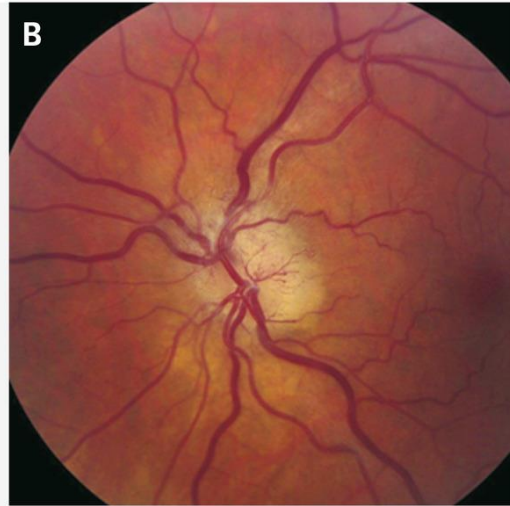
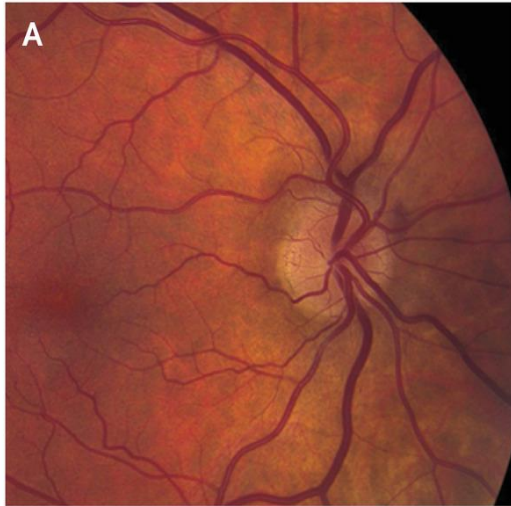
Optic Neuritis

<40 yo

overlap

>40 yo

Ischemic Optic Neuropathy  
>65 years Temporal Arteritis



Acute Onset:

- 70 YO M Blurry vision and decreased vision with loss of visual acuity upon awakening or preceded by amaurosis fugax in left eye.
- Relative afferent pupillary defect, left eye.
- Loss of color vision, left eye
- Visual field defects: altitudinal defect
- Optic nerves are swollen in the acute phase as in view.



Optic Neuritis

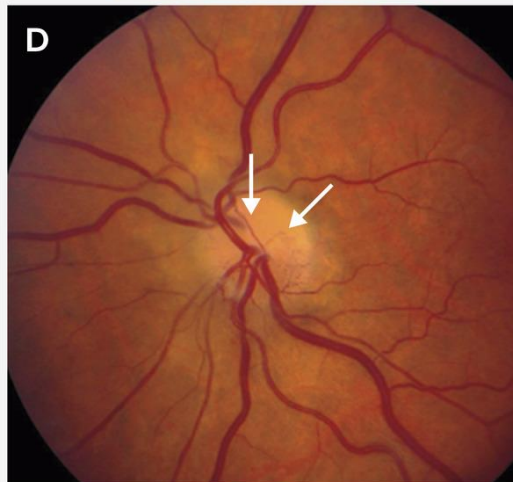
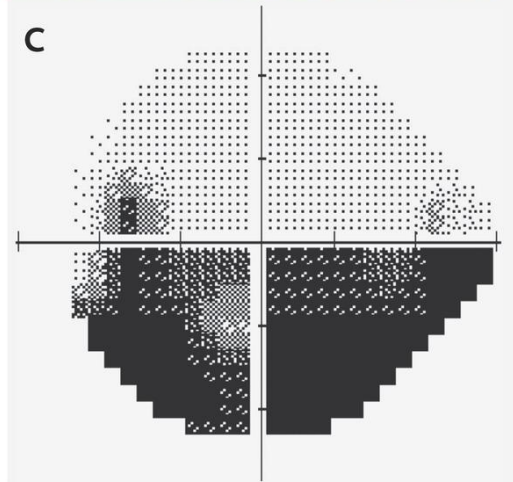
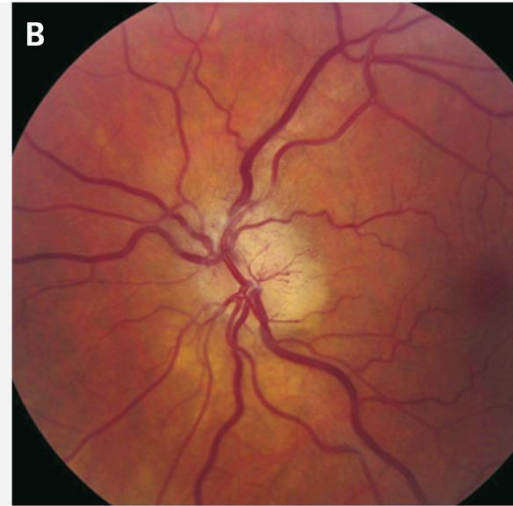
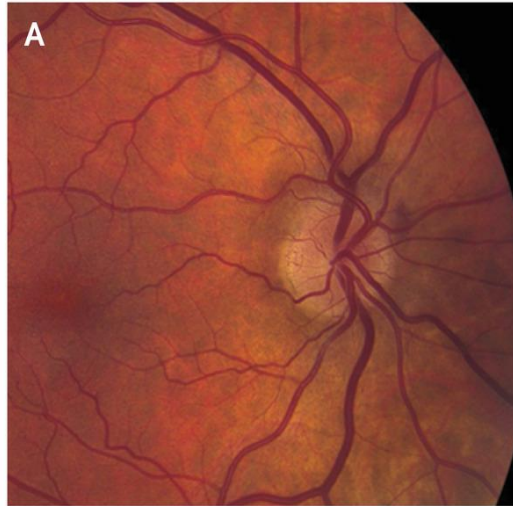
<40 yo

overlap

>40 yo

Ischemic Optic Neuropathy

>65 years Temporal Arteritis



Acute Onset:

- 70 YO M Blurry vision and decreased vision with loss of visual acuity upon awakening or preceded by amaurosis fugax in left eye.
- Relative afferent pupillary defect, left eye.
- Loss of color vision, left eye
- Visual field defects: altitudinal defect
- Optic nerves are swollen in the acute phase as in view.

Optic Neuropathy Pattern Recognition Table

Change in Vision

Dyschromatopsia

RAPD +

Optic Disc: normal edema, pale

VF defect respects the horizontal meridian

Optic Neuritis

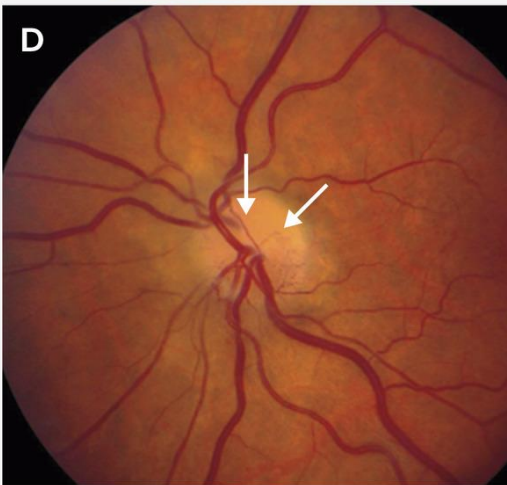
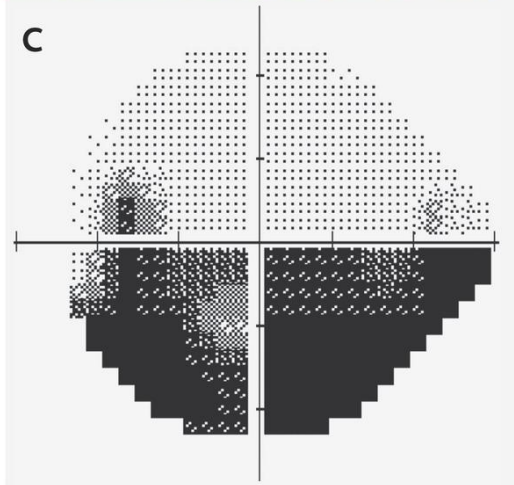
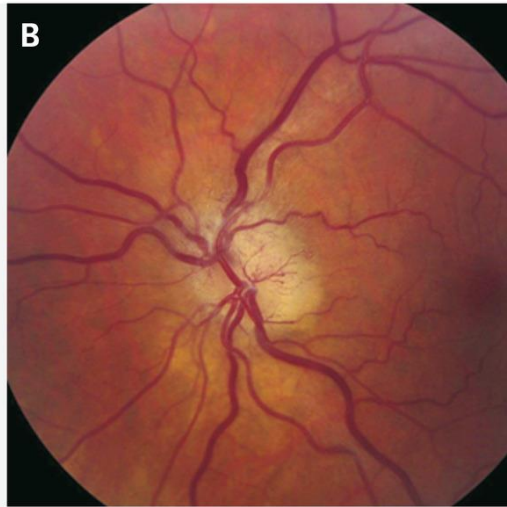
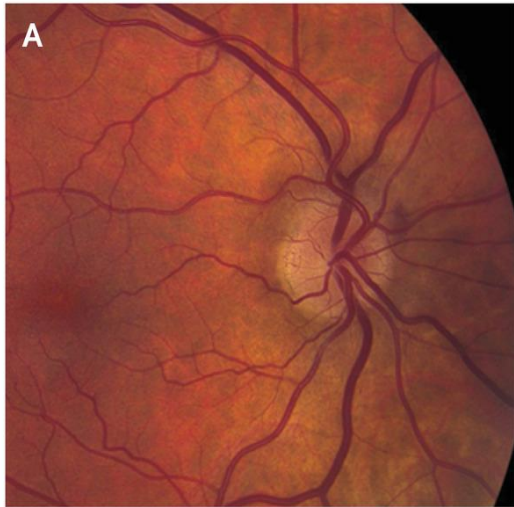
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Optic Neuropathy Pattern Recognition Table

Change in Vision ✓

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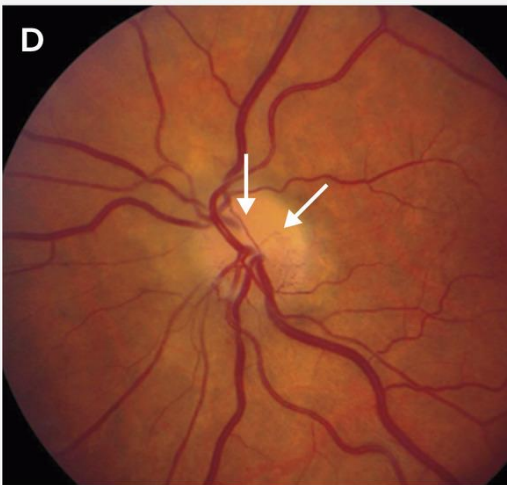
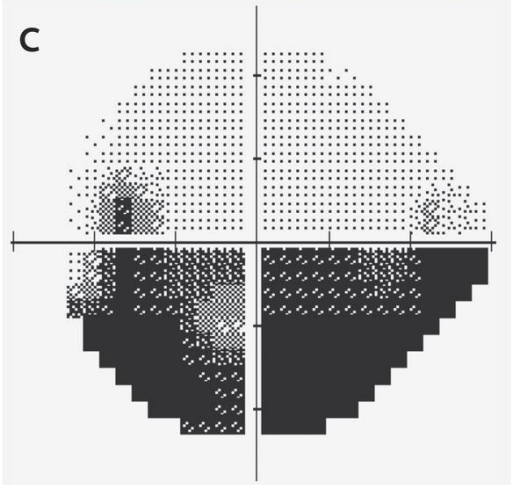
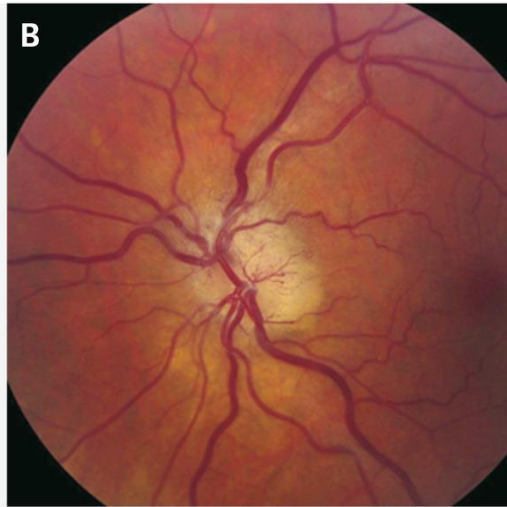
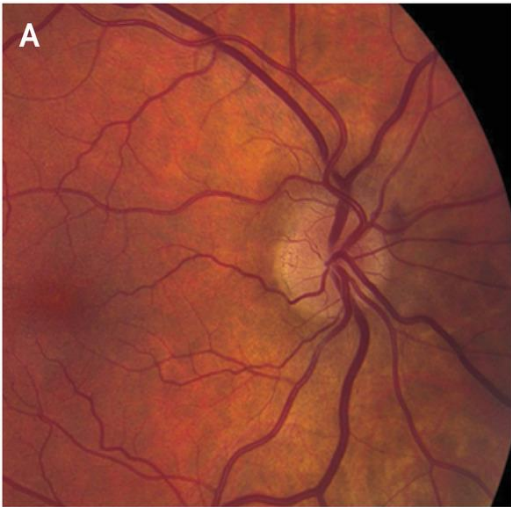
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Ischemic Optic Neuropathy

>65 years Temporal Arteritis



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- Optic nerves are swollen

### Optic Neuropathy Pattern Recognition Table

Change in Vision ✓

Dyschromatopsia ✓

RAPD +

Optic Disc: normal edema, pale

VF defect respects the horizontal meridian

Optic Neuritis

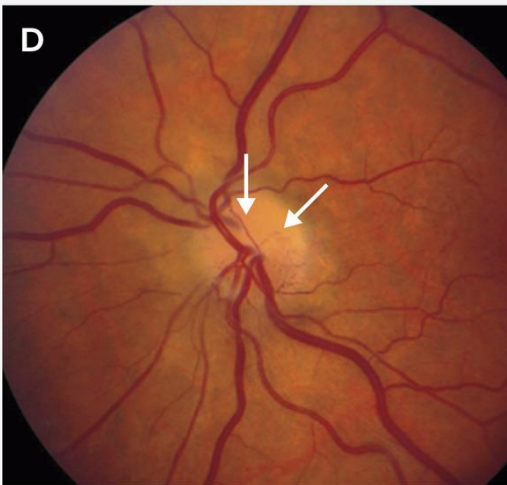
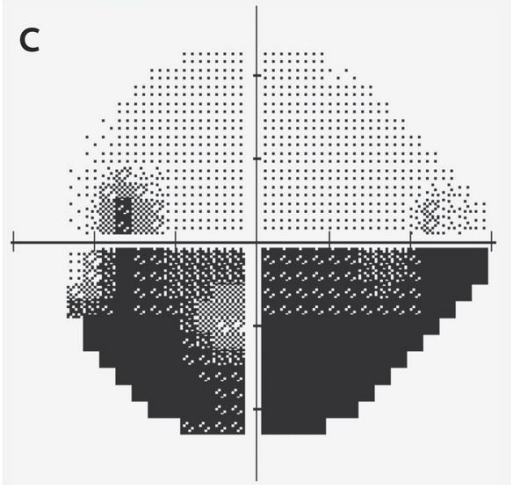
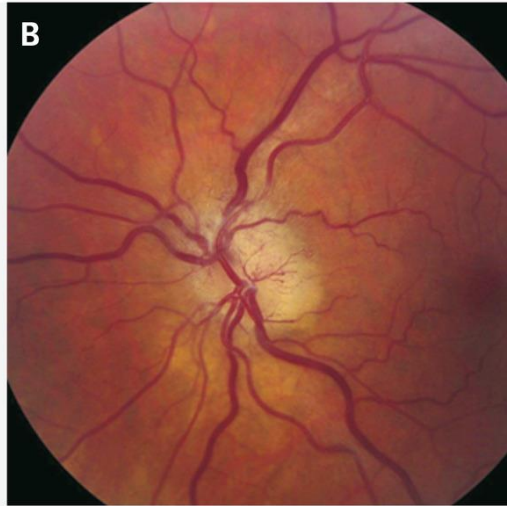
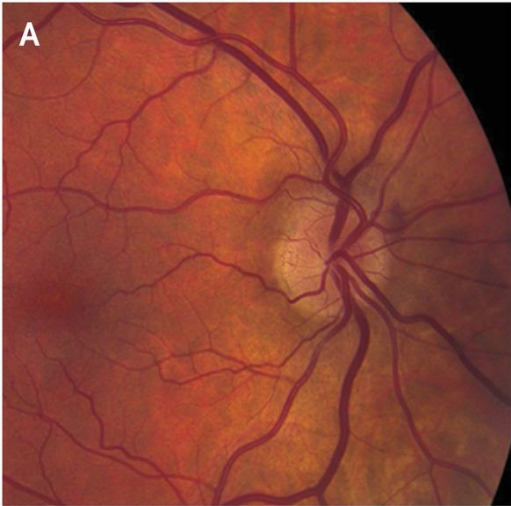
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Ischemic Optic Neuropathy

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Optic Neuropathy Pattern Recognition Table

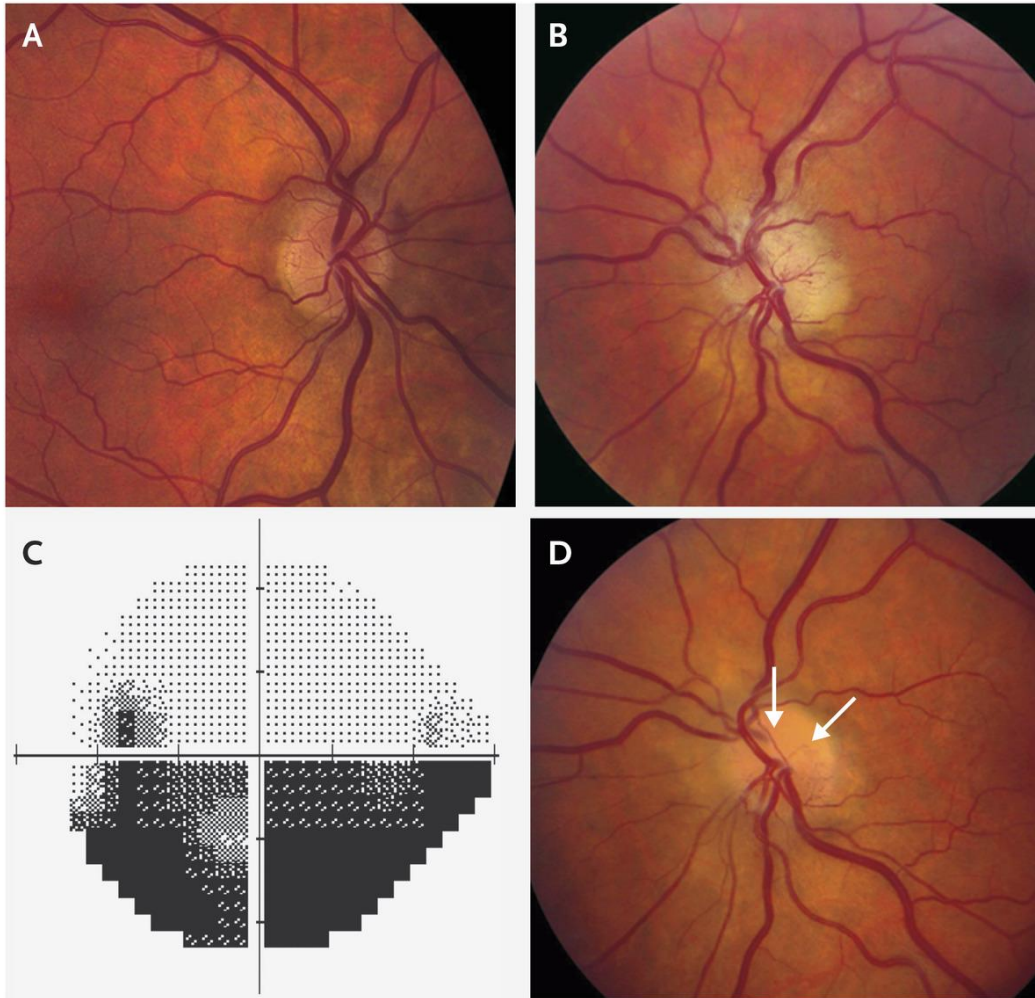
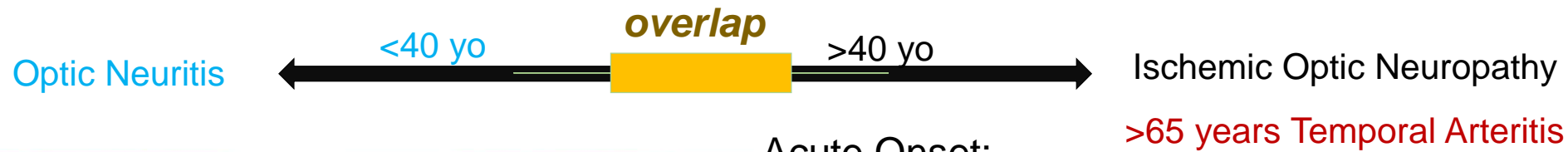
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| Optic Neuropathy Pattern Recognition Table |   |
|--|---|
| Change in Vision                           | ✓ |
| Dyschromatopsia                            | ✓ |
| RAPD +                                     | ✓ |
| Optic Disc: normal edema, pale             | ✓ |
| VF defect respects the horizontal meridian |   |

Optic Neuritis

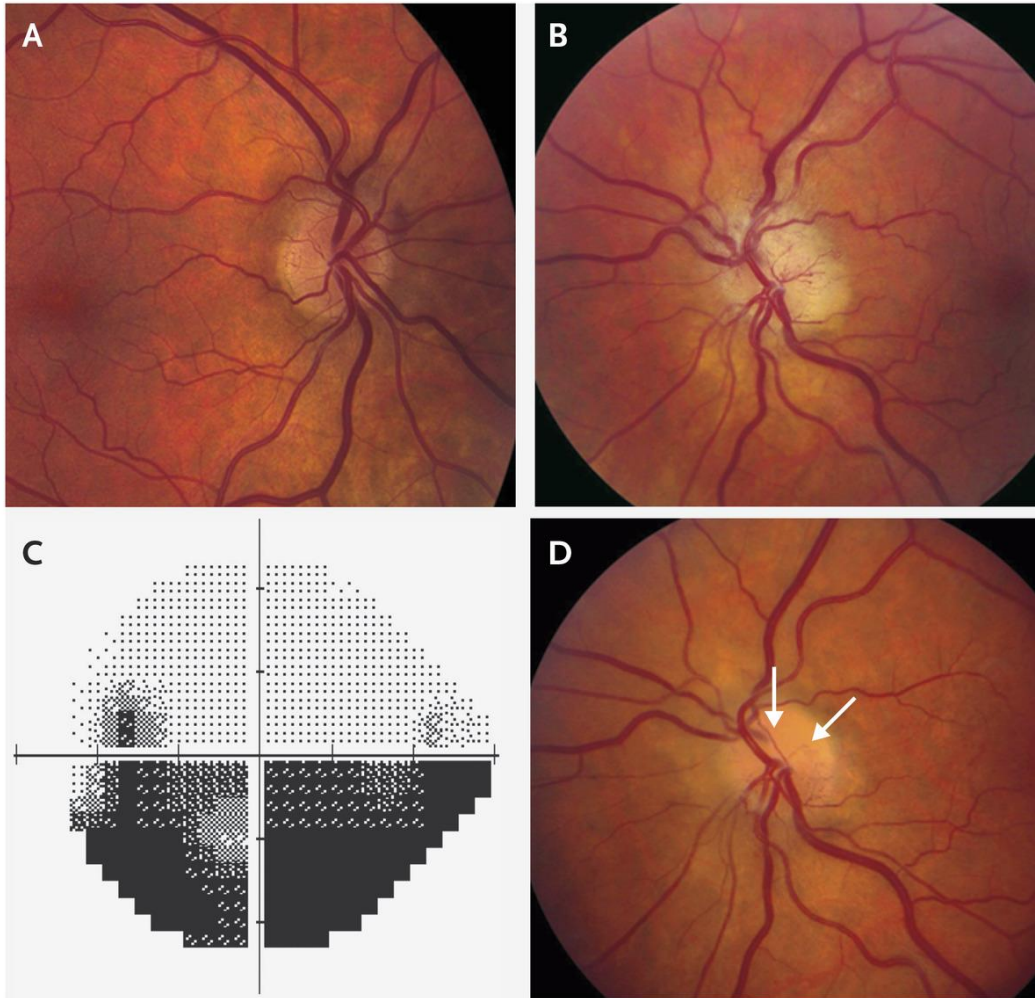
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Ischemic Optic Neuropathy

>65 years Temporal Arteritis



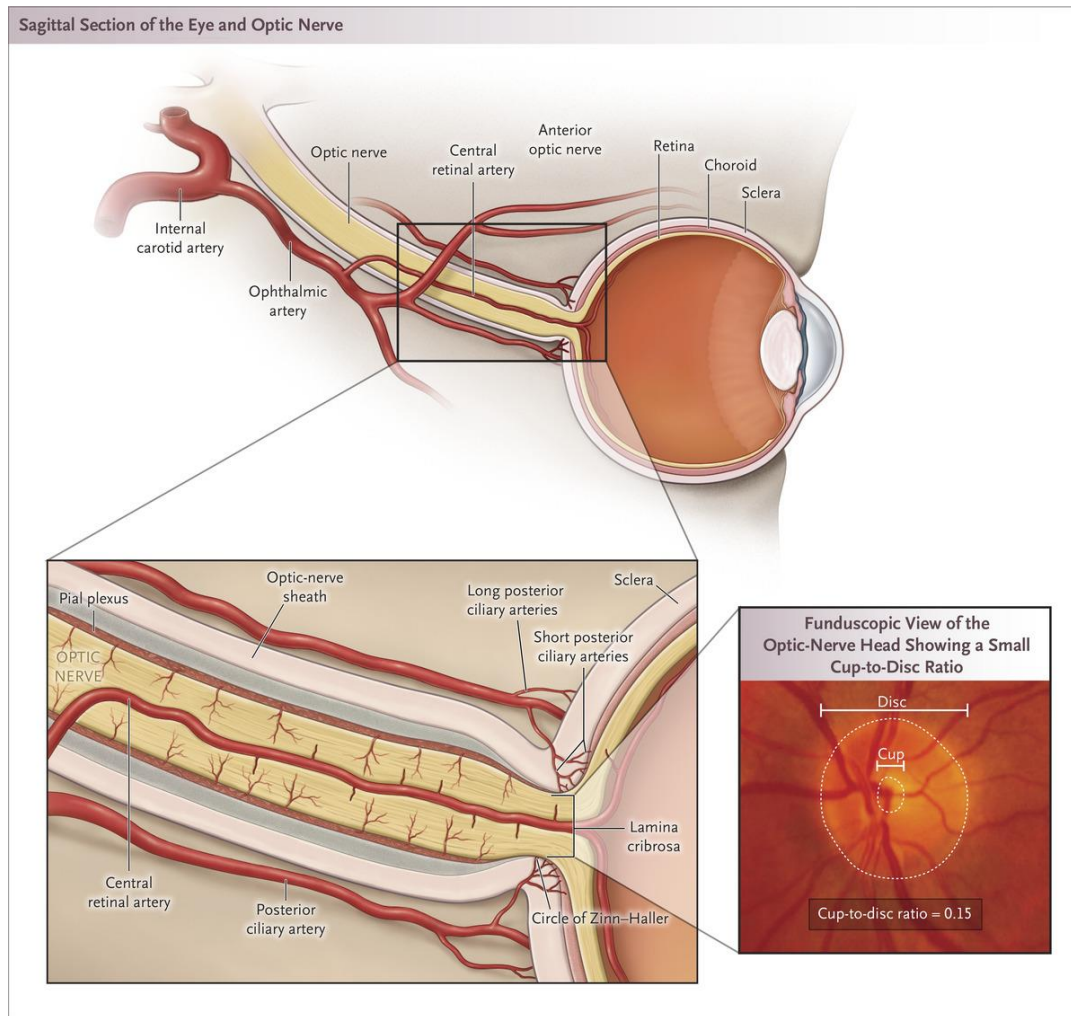
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| Optic Neuropathy Pattern Recognition Table |   |
|--|---|
| Change in Vision                           | ✓ |
| Dyschromatopsia                            | ✓ |
| RAPD +                                     | ✓ |
| Optic Disc: normal edema, pale             | ✓ |
| VF defect respects the horizontal meridian | ✓ |

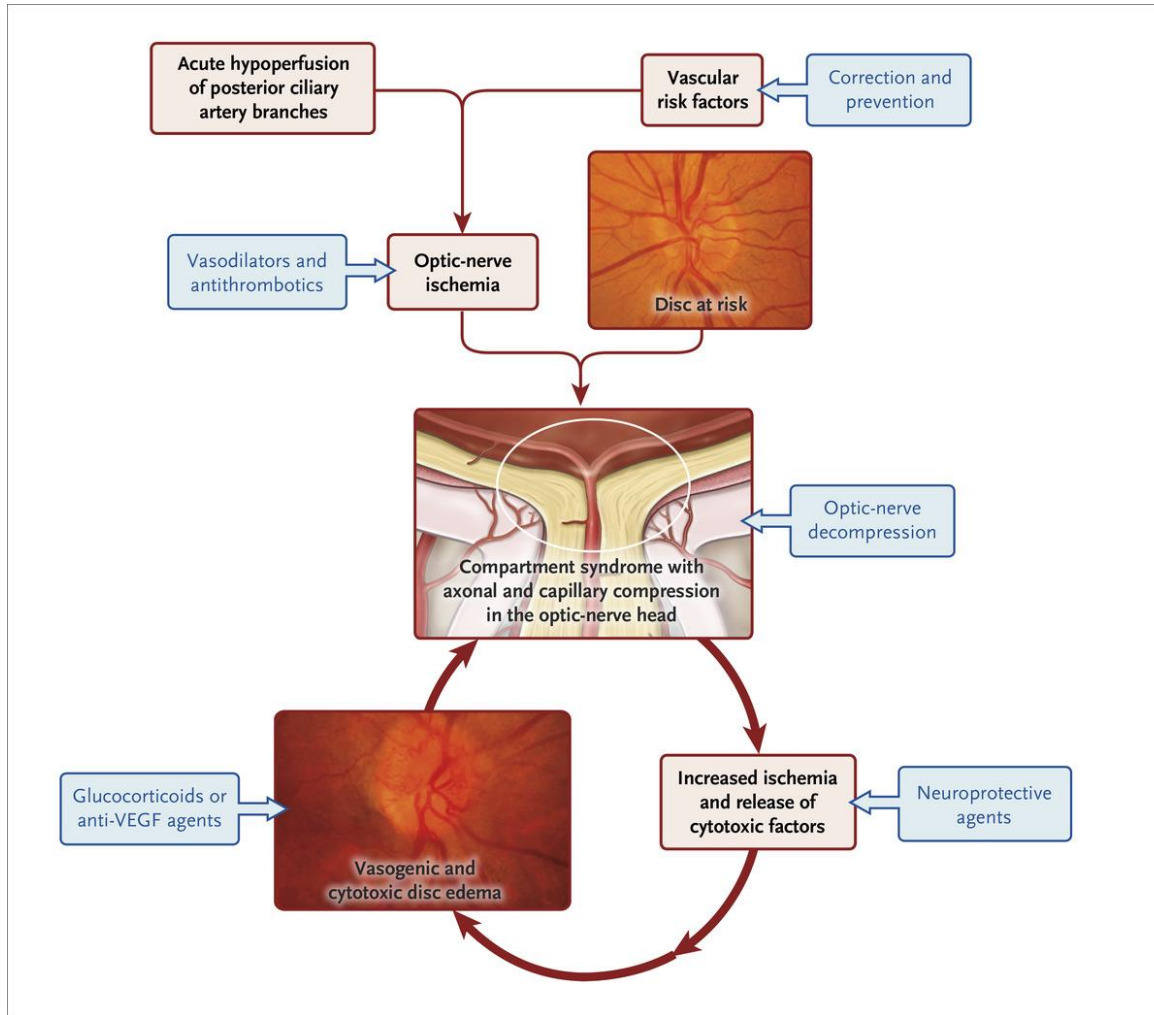
# Non-arteritic Anterior Ischemic Optic Neuropathy (NAION)

## Blood Supply to the Optic Nerve and Anatomy of the Optic-Nerve Head



- The ophthalmic artery is the first branch of the internal carotid artery.
- Central retinal artery → inferior and superior arcade → inner retina
- Choroid arteries come from posterior ciliary artery → outer retina
- Hypoperfusion in the territory of the ophthalmic artery and its branches → vascular ischemia

# Non-arteritic Anterior Ischemic Optic Neuropathy (NAION)



- Accounts for 90% - 95% of cases of AION.
- Visual loss usually present upon awakening.
- Structural “crowding” of the optic nerves (“disc at risk”).
- 5-year risk of contralateral involvement is 14.7 to 24%.
- Risk of same eye recurrence is about 1-5%.



# Summary of Non-Arteritic Ischemic Optic Neuropathy

**Increased risk of NAION in patients with hypertension especially if they take their ANTI-HYPERTENSIVES AT NIGHT → nocturnal hypotension.**

|                     |   |                 |  |
|---------------------|---|-----------------|--|
| Demographic         | Mean, 60 years, F = M   |                 |  |
| Symptoms            | Smudge, sudden blur of vision, progress to blurred inferior vision  |                 |  |
| Associated symptoms | Usually none, no headache, no stigmata of GCA   |                 |  |
| Risk factors        | Systemic: Diabetes * (debated), hypertension, hyperlipidemia, sleep apnea<br>Ophthalmic risk: disc at risk (small cup to dis), optic disc drusen  | Natural history | 31% improve, (30-30-30 rule)<br>30% - stay the same in 3 – 6 months<br>30% - worsen<br>30% - improves 1-2 lines of vision<br>Rare but can be simultaneously involved<br>24-25 % in other eye within 5 years. |
| Visual acuity       | <ul style="list-style-type: none"> <li>➤ Central vision preserved</li> <li>➤ 20/200 in &gt; 60% of cases</li> <li>➤ Some cases are worse if there is macular involvement such as macular edema</li> </ul> | Work up         | Careful history<br>Inflammatory markers: ESR, CRP, Platelets<br>No MRI or CT scans needed<br>Clinical diagnosis  |
| Disc/fundus         | Small cup in fellow eye<br>Optic disc edema :<br>Sectorial disc edema with peripapillary flame shape hemes  | Treatment       | Steroids? – NO, controversial<br>Mitigate risk factors<br>No treatment   |
| Visual field defect | Inferior arcuate or inferior altitudinal defect   |                 |  |

# Arteritic vs Non-Arteritic Ischemic Optic

| Characteristic      | Arteritic   | Non-arteritic   |
|---------------------|---|---|
| Demographics        | Mean, 70 years<br>F > M   | Mean, 60 years<br>F = M   |
| Symptoms            | Sudden transient loss of vision like a curtain in vision .<br>Sudden darkening and loss of vision. Black out or white out of vision, especially with demand | Usually none, no headache, no stigmata of GCA   |
| Associated symptoms | Headache, tender scalp, jaw claudication, transient visual loss, double vision  | Usually none  |
| Risk factors        | Can be associated to PMR  | Systemic: Diabetes * (debated), hypertension, hyperlipidemia, sleep apnea<br>Ophthalmic risk: disc at risk (small cup to dis), optic disc drusen  |
| Visual acuity       | < 20/200 in > 60% of cases<br><br>Quickly progresses to LP or NLP vision without treatment  | Central vision preserved<br><br>20/200 in > 60% of cases<br><br>Some cases are worse if there is macular involvement such as macular edema  |
| Disc/fundus         | Pallid disc edema (acute), cotton-wool spots.<br><br>Diffuse optic disc edema with whitening of the optic disc<br><br>Hemes may be present                  | Small cup in fellow eye<br>Small cup in fellow eye<br>Optic disc edema :<br>Sectorial disc edema with peripapillary flame shape hemes<br><br>Then resolution over 4 to 12 weeks leading to sectorial pallor |

# Arteritic vs Non-Arteritic Ischemic Optic Neuropathy

| Characteristic  | Arteritic   | Non-arteritic  |
|---|---|--|
| <p><b>Increased risk of NAION in patients with hypertension especially if they take their ANTI-HYPERTENSIVES AT NIGHT resulting in nocturnal hypotension.</b></p> |   |  |
| Natural history   | <p>Rarely improve, fellow eye involvement common<br/>Severe vision loss if not treated</p>  | <p>31% improve, (30-30-30 rule)<br/>30% - stay the same in 3 – 6 months<br/>30% - worsen<br/>30% - improves 1-2 lines of vision<br/>Rare but can be simultaneously involved<br/>24-25 % in other eye within 5 years.</p> |
| Work up   | <p>Elevated inflammatory markers: ESR, CRP, Platelets<br/>MRI brain and orbit with and without contrast<br/>Ultrasound (temporal artery)<br/>Temporal artery biopsy</p> | <p>Careful history<br/>Inflammatory markers: ESR, CRP, Platelets<br/>No MRI or CT scans needed<br/>Clinical diagnosis</p>  |
| Treatment   | <p>Systemic steroids</p>  | <p>Steroids? – NO, controversial<br/>Mitigate risk factors<br/>No treatment</p>  |

Optic Neuritis

<40 yo

**overlap**

>40 yo

Ischemic Optic Neuropathy  
>65 years Temporal Arteritis

### Optic Neuropathy Pattern Recognition Table

Change in Vision

Dyschromatopsia

RAPD +

Optic Disc: normal  
edema, pale

VF defect respects the  
horizontal meridian

## Case 1

Optic Neuritis

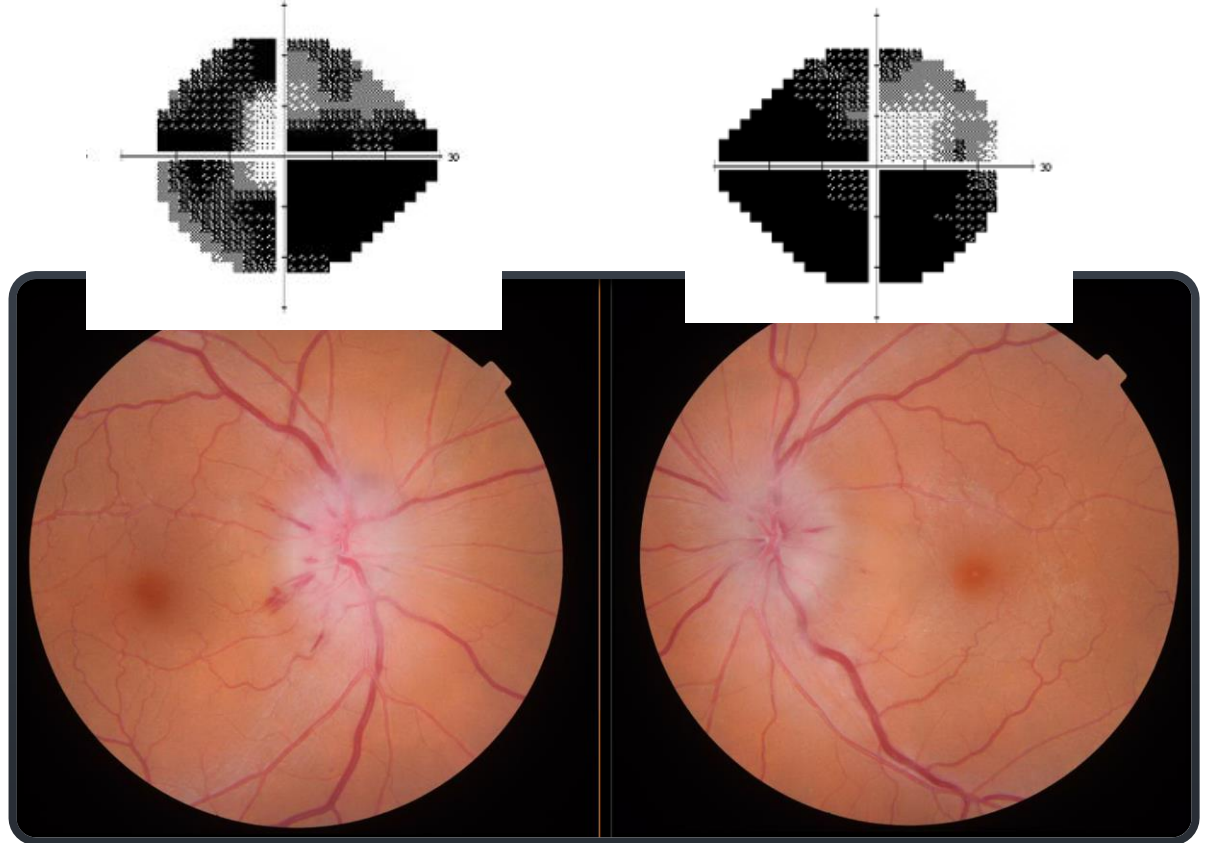


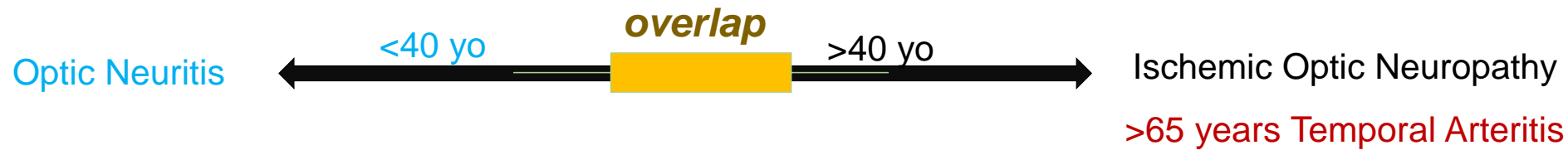
Ischemic Optic Neuropathy  
>65 years Temporal Arteritis

72 year old woman with sudden blurry vision, right eye upon awakening in the morning

- 2-3 weeks prior she developed 'dirty glasses' with shadows but did not have floaters, no headache or eye pain. No headache, scalp tenderness, jaw pain, weight loss, or fever.

| Optic Neuropathy Pattern Recognition Table |   |
|--|---|
| Vision                                     | Right eye: 20/100 pin hole without improvement<br>Left eye: 20/20   |
| Dyschromatopsia                            | Right eye: 0 color plates<br>Left eye: Normal   |
| RAPD ?                                     | Right eye grade 3+ relative afferent pupillary defect   |
| VF defect respects the horizontal meridian | <div style="display: flex; justify-content: space-around;"> <div style="text-align: center;"> <p>Left</p> </div> <div style="text-align: center;"> <p>Right</p> </div> </div> |



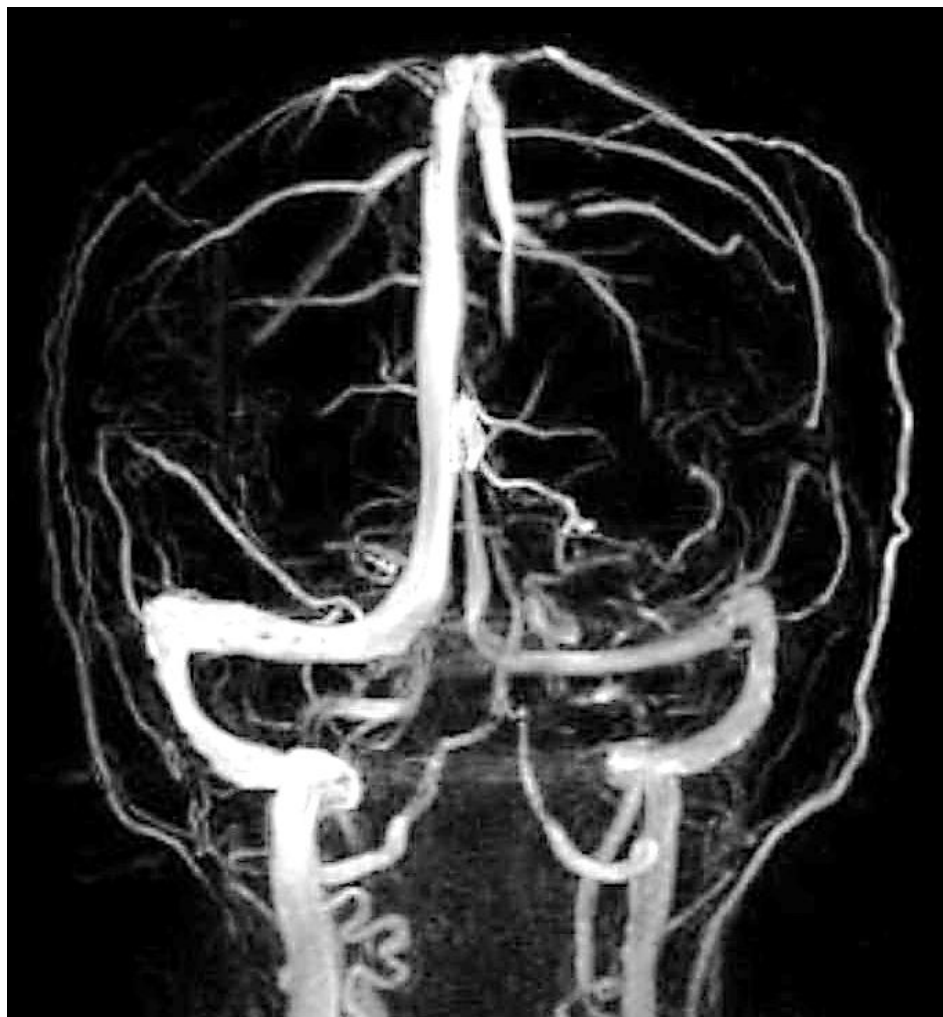


| Optic Neuropathy Pattern Recognition Table |   |
|--|---|
| Change in Vision                           | ✓ 20/100 phni right eye   |
| Dyschromatopsia                            | ✓ No color vision in right eye                                      |
| RAPD +                                     | ✓ rAPD right eye  |
| Optic Disc: normal edema, pale             | ✓ Bilateral optic disc edema with hemes                             |
| VF defect respects the horizontal meridian | ✓ bilateral visual field defect dense inferior – altitudinal defect |

Can you clarify some points?

1. Why does she not have color vision in the right eye?
2. Both eyes appear affected why is there a rAPD? How much of a difference do you start to notice a rAPD?
3. Both optic nerves are affected, how do we know this isn't papilledema?
4. The visual field defect affects both upper and lower halves of vision, why?

What should a neurologist do?



CBC/BMP MCV elevated at 93.8 (81-91), otherwise WNL

ACE 22 (8-53)

CRP/ESR 2.4/10

NMO negative

MOG negative

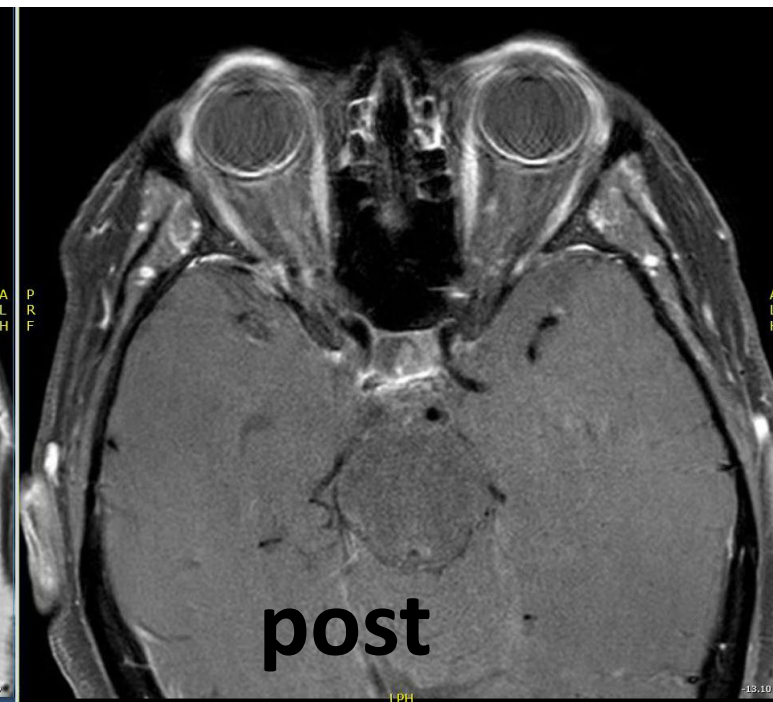
Bartonella, treponema, lyme antibodies negative

SPEP, IgG, IgA, SSA/B, RF, within normal limits

Speckled ANA pattern, 1:40

ANCA pending

**LP:** Opening pressure (cm H<sub>2</sub>O): **14 cm of water**  
Protein: 42, Glucose 93 (H)  
CSF 3 cells, normal protein and glucose, cytology

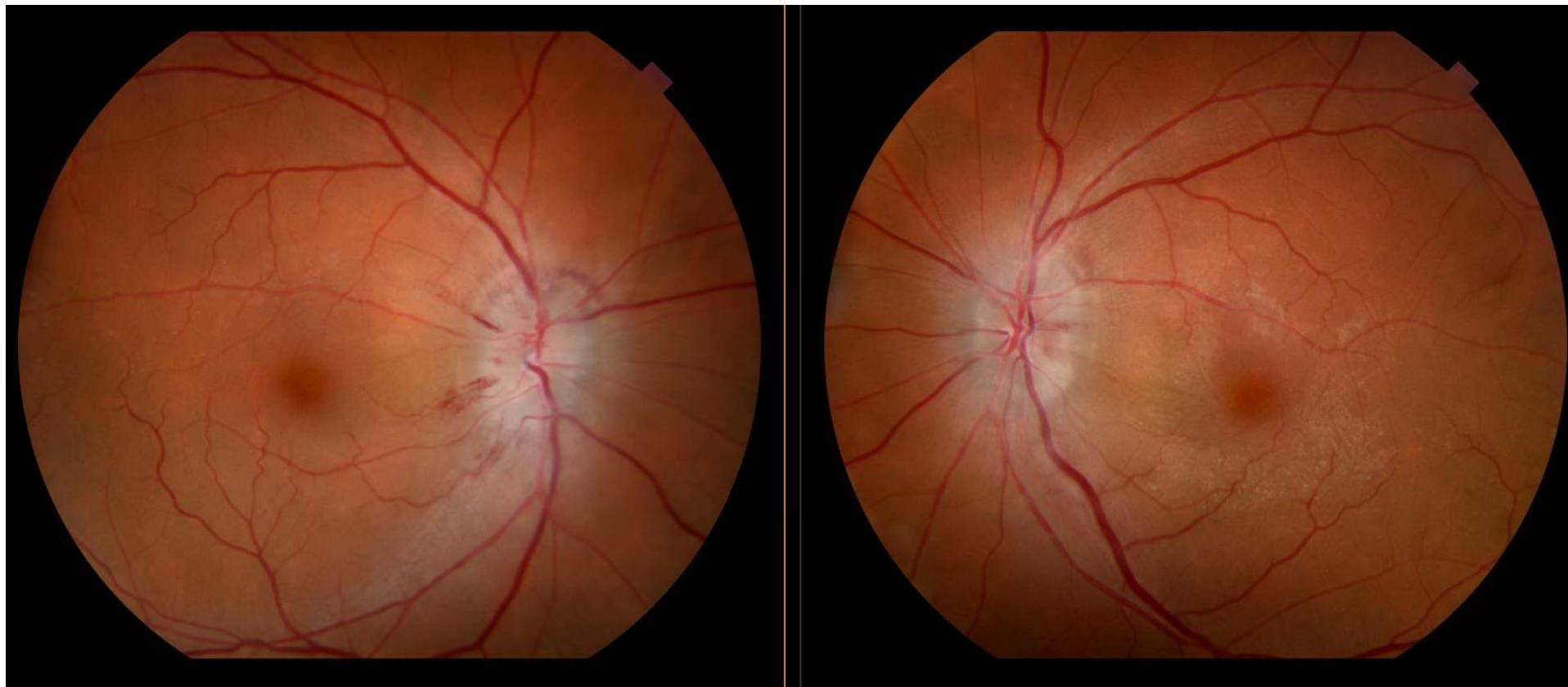


IMPRESSION:

\*----- Impression -----\*

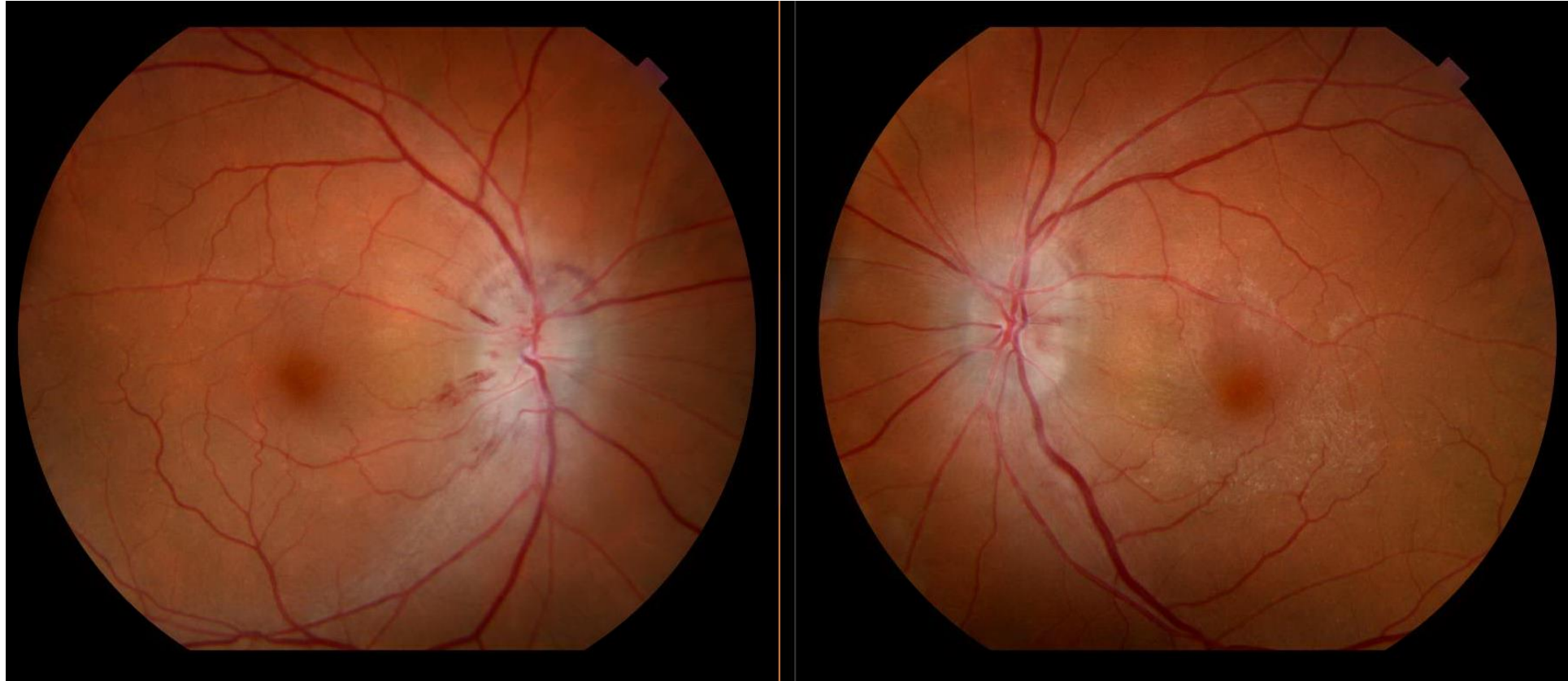
1. NO ACUTE INFARCTION, HEMORRHAGE, MASS LESION, OR ABNORMAL ENHANCEMENT. NO ABNORMAL SIGNAL OR ENHANCEMENT OF THE OPTIC NERVES.
2. NO EVIDENCE OF DURAL VENOUS SINUS STENOSIS OR THROMBOSIS.

10 days later





10 days later



Bilateral Sequential painless loss of vision with normal work up  
Non-Arteritic Ischemic Optic Neuropathy

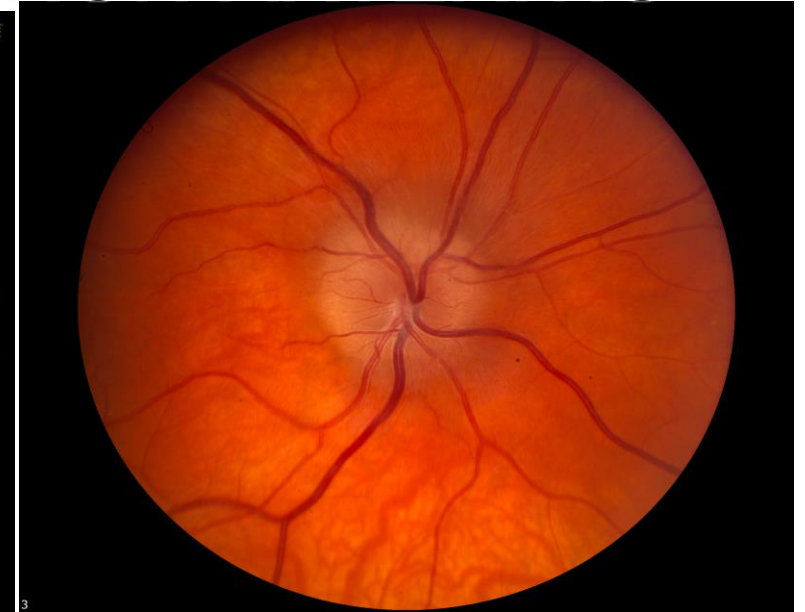
# Anterior ischemic optic neuropathy (AION)

- **The stakes are high.**
- Inflammatory or thrombotic occlusion of the **short posterior ciliary arteries**
- Severe visual loss (CF or worse)
- Retinal ischemia (cotton-wool spots, artery occlusion)
- Problems in choroidal filling

## ARTERITIC VS. NON-ARTERITIC



Chalky white disc edema



Hyperemic in NAION

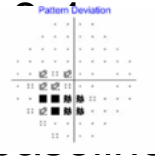
Jaw claudication is the most specific symptom and may predict a worse visual outcome.

Occult GCA may occur in 20% of patients.

**Check ESR, CRP, and platelets**

# Case 2

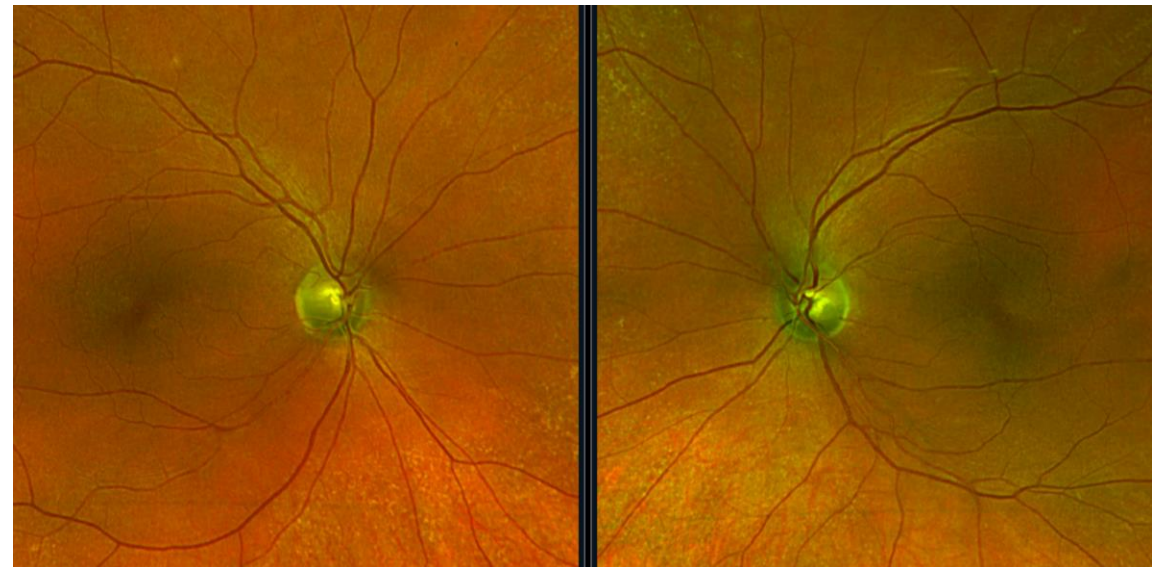
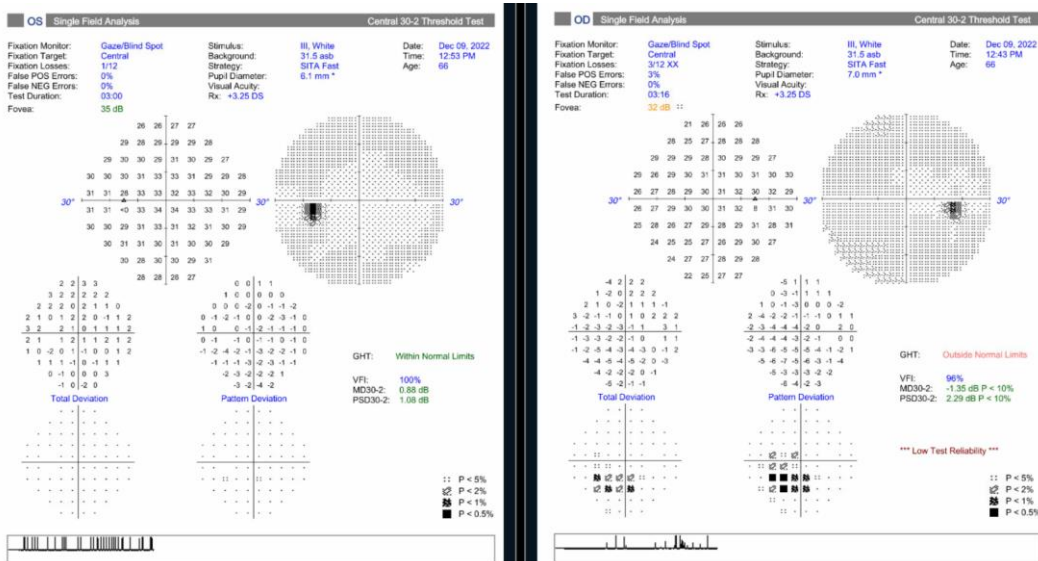
- 66 YO woman 7 months prior to presentation: acute onset intermittent severe right sided headache, intense including the orbit radiating to the right side and shoulder.
- Chronically fatigue (feeling poorly)
- 4 months prior to clinic visit she describes transient complete vision in loss of the right eye lasting minutes when outdoors golfing.
- Mild shoulder and hip girdle pain worse than from rheumatoid arthritis
- No fever, chills, weight loss, jaw claudication
- Initially thought due to chronic migraine with recurrence of headaches on the right side.
- She had been followed by her rheumatologist for rheumatoid arthritis and started on steroids, with significant improvement



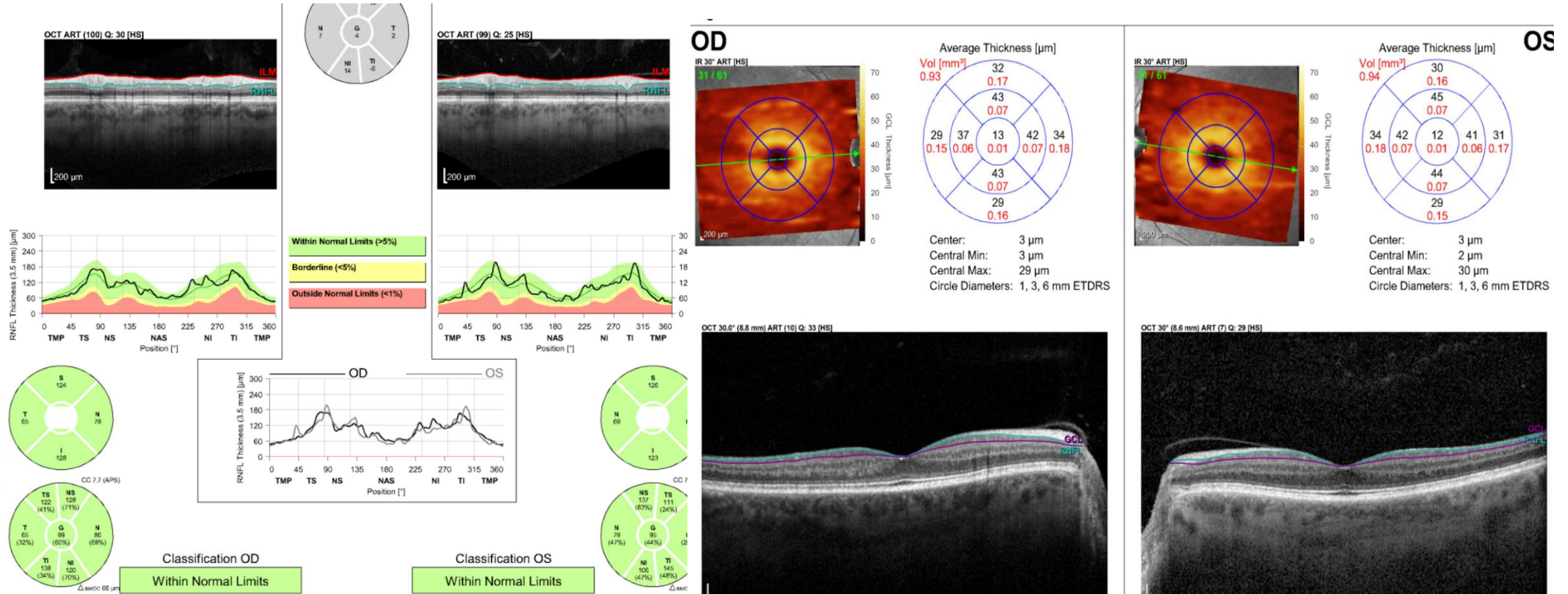
| Optic Neuropathy Pattern Recognition Table |  |
|--|--|
| Vision                                     | Right eye 20/25+, PHNI<br>Left eye 20/20 |
| Dyschromatopsia                            | Right eye: Normal<br>Left eye: Normal    |
| RAPD ?                                     | 0.9 log units left eye                   |
| VF defect respects the horizontal meridian | Full, no visual field defects            |
| Optic Nerve appearance?                    |  |

# Case 2

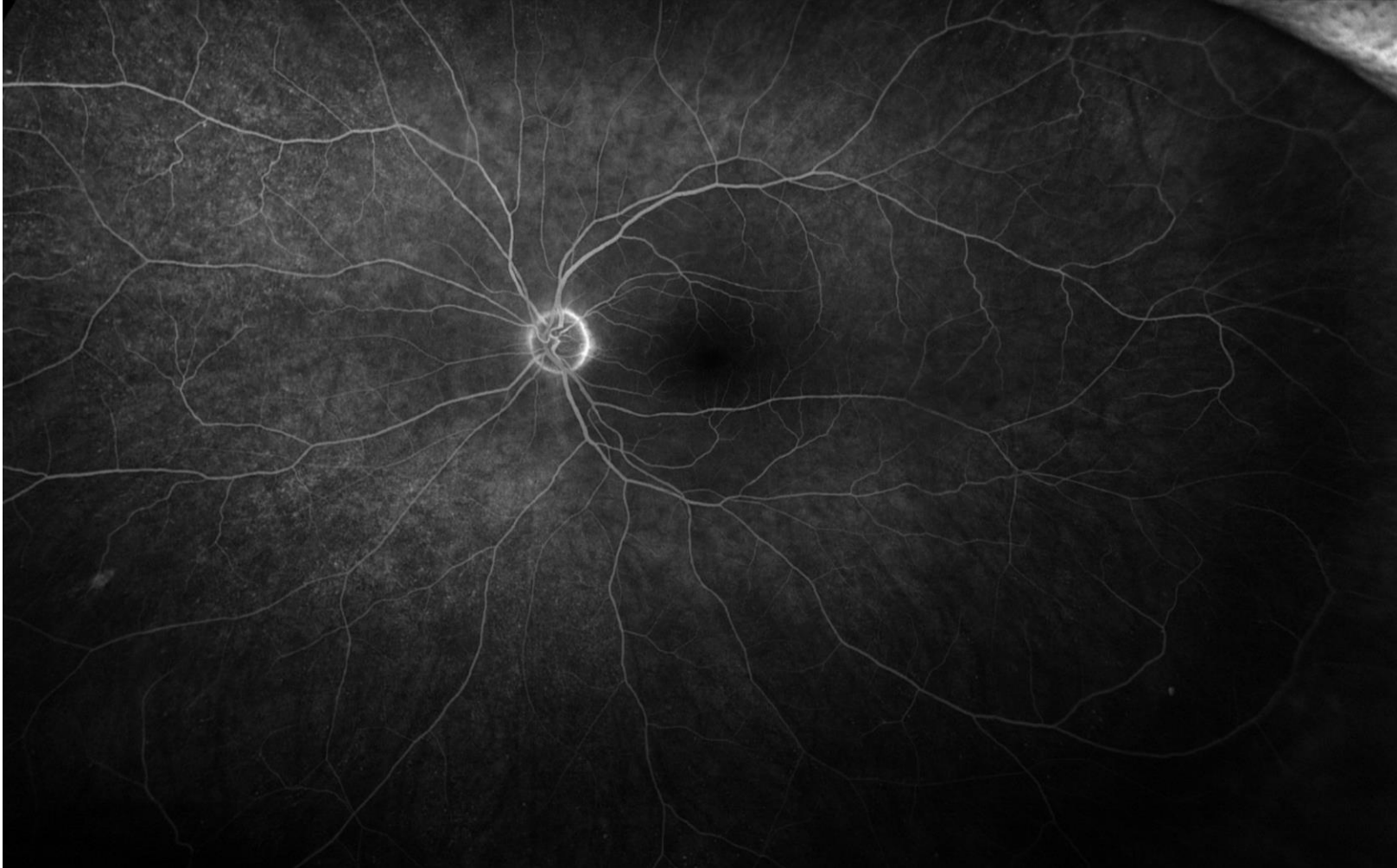
- Work up: ESR 42 (mild elevation) mm / Hg CRP 3.9 (within normal)



# Optical Coherence Tomography



# Fluorescein Angiogram



- A. Temporal artery, right side (biopsy):**
- Negative for active or healed arteritis
  - Intimal hyperplasia
  - 719 sections examined
  - Elastic stain supports the above interpretation.

# Does this patient have an optic neuropathy?

- 66 YO woman 7 months prior to presentation: acute onset intermittent severe right sided headache, intense including the orbit radiating to the right side and shoulder.
- Chronically fatigue (feeling poorly)
- **4 months prior to clinic visit she describes transient complete vision in loss of the right eye lasting 3-4 minutes when outdoors golfing.**
- Mild shoulder and hip girdle pain worse than baseline from rheumatoid arthritis
- No fever, chills, weight loss, jaw claudication
- Initially thought due to chronic migraine with recurrence of headaches on the right side.
- She had been followed by her rheumatologist for rheumatoid arthritis and started on steroids, with significant improvement

| Optic Neuropathy Pattern Recognition Table |  |
|--|--|
| Vision                                     | Right eye 20/25+, PHNI<br>Left eye 20/20<br>✓ transient complete vision in loss of the right eye lasting 3-4 minutes when outdoors golfing |
| Dyschromatopsia                            | Right eye: Normal<br>Left eye: Normal  |
| RAPD ?                                     | ✓ 0.9 log units left eye   |
| VF defect respects the horizontal meridian | Full on confrontation  |
| Optic nerve appearance                     | Normal   |

# Is this GCA/ Temporal Arteritis?

| Optic Neuropathy Pattern Recognition Table |  |
|--|--|
| Vision                                     | Right eye 20/25+, PHNI<br>Left eye 20/20<br>✓ transient complete vision in loss of the right eye lasting 3-4 minutes when outdoors golfing |
| Dyschromatopsia                            | Right eye: Normal<br>Left eye: Normal  |
| RAPD ?                                     | ✓ 0.9 log units left eye   |
| VF defect respects the horizontal meridian | Full on confrontation  |
| Optic nerve appearance                     | Normal   |

| Other findings          |   |
|-------------------------|---|
| Normal                  | Retinal nerve fiber and ganglion cell layer measurements on OCT |
| Fluorescein angiography | Normal  |
| Temporal artery biopsy  | Negative  |

This patient is clinically diagnosed with temporal arteritis and maintained on steroids. Plan to transition to actemra.



# Anterior ischemic optic neuropathy (AION)

- **The stakes are high.**

- Inflammatory or thrombotic occlusion of the **short posterior ciliary arteries**
- Severe visual loss (CF or worse)
- Retinal ischemia (cotton-wool spots, artery occlusion)
- Problems in choroidal filling

## ARTERITIC VS. NON-ARTERITIC



Chalky white disc edema



Hyperemic in NAION

Jaw claudication is the most specific symptom and may predict a worse visual outcome.

Occult GCA may occur in 20% of patients.

**Check ESR, CRP, and platelets**

# Case 3

- 76 year old right handed woman with history of diabetes
- Three weeks prior to seeing PCP she developed bifrontal headaches and fever; elevated inflammatory markers were noted but she did not get steroids.
- Temporal artery biopsy was negative
- MRI brain and orbit showed microvascular ischemic changes
- Vision worsened to bare light perception and was admitted to the hospital for IV steroids.

## Optic Neuropathy Pattern Recognition Table

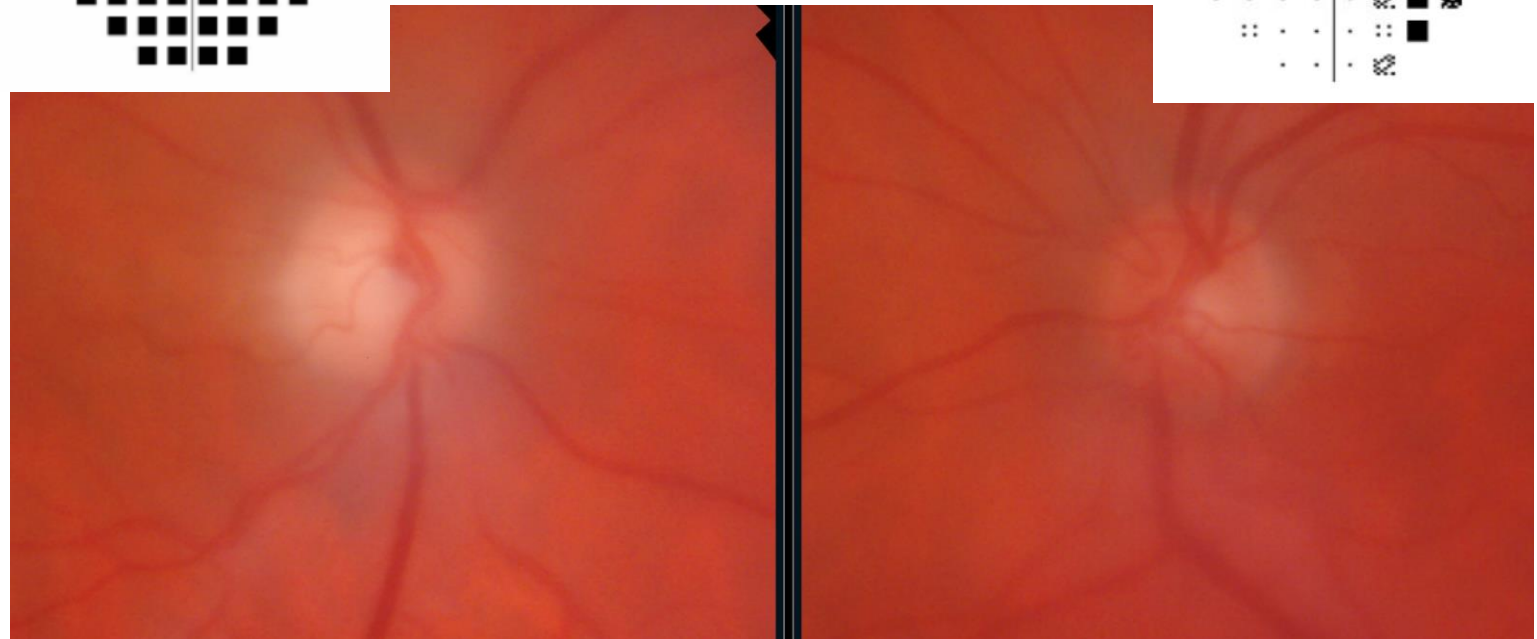
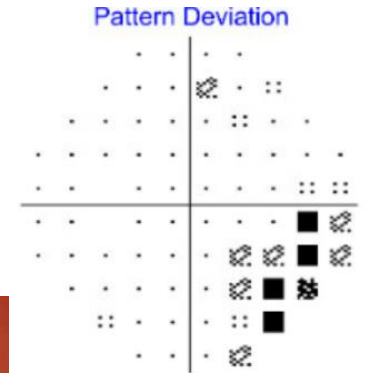
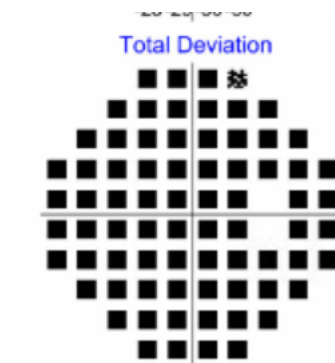
Vision ✓ NLP, right eye  
Normal 20/20 left eye

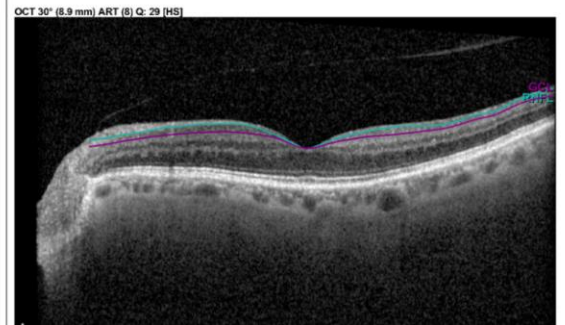
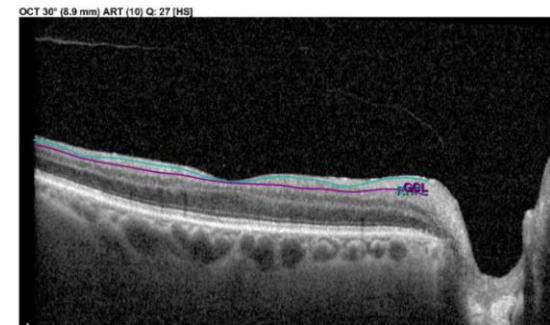
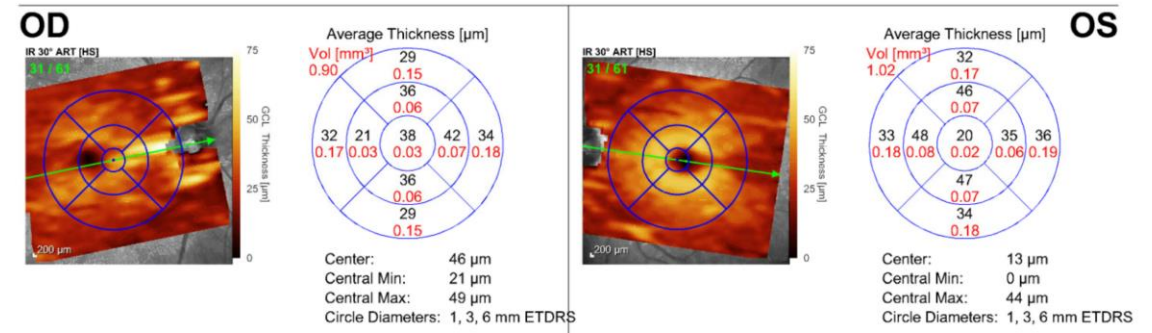
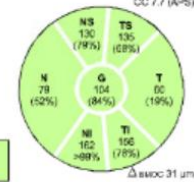
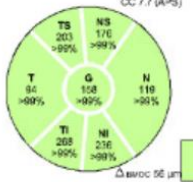
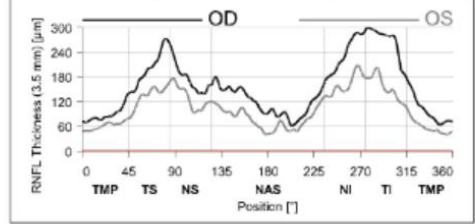
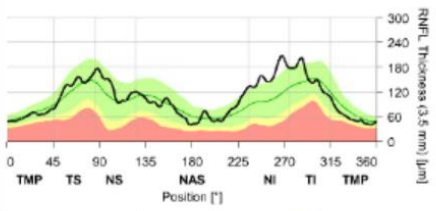
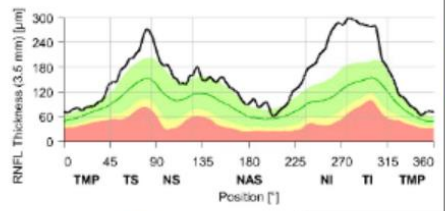
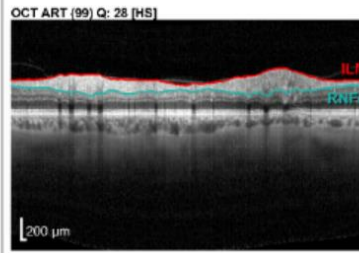
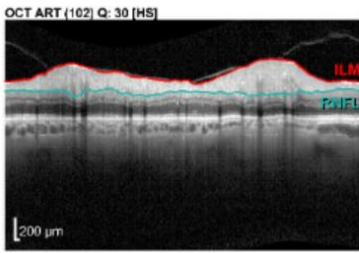
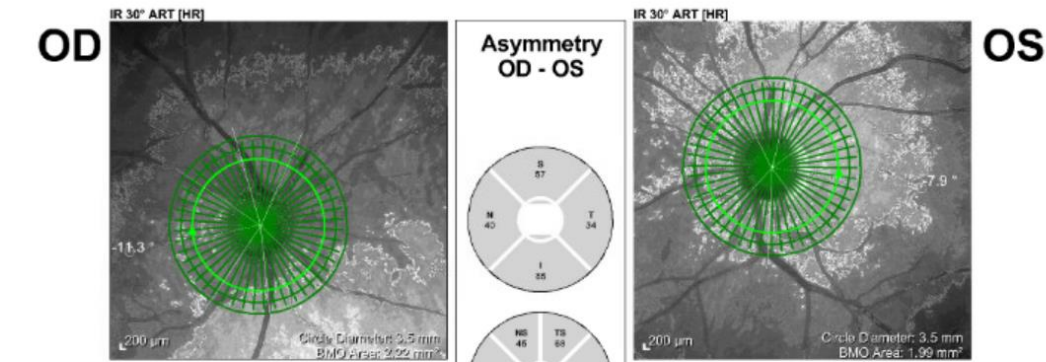
Dyschromatopsia ✓ 0 right eye  
Normal left eye

RAPD ? ✓ 3.0 log units left eye

VF defect respects the horizontal meridian ✓ Complete defect

Optic nerve appearance ✓ Pallid edema





**A. Temporal artery, right side (biopsy):**

- Negative for active or healed arteritis
- Intimal hyperplasia
- Elastic stain supports above interpretation
- 225 sections examined

# References