

# Neuro-optometric Grand Rounds

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# Learning Objectives

Review the normal anatomy of the visual pathway.

Discuss the appropriate workup for suspected neuro-ophthalmic disease.

Discuss neuro-ophthalmic case reports presenting to a primary eye care clinic.

Identify the optometrist's role in managing patients with neuro-ophthalmic disease.

## **Optic Nerve Anatomy**

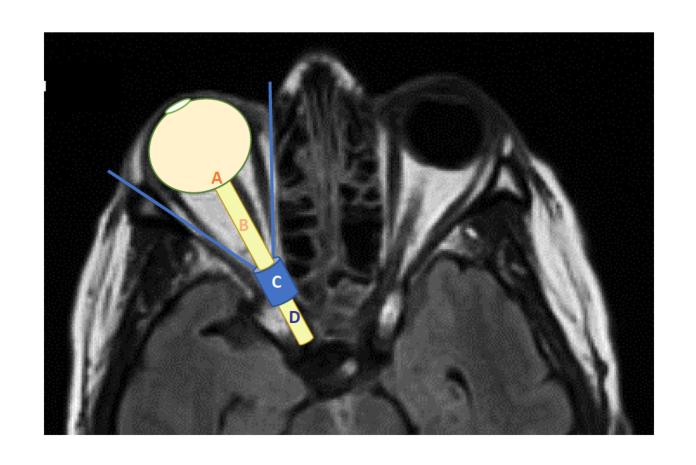


Intraocular: 1 mm

Intraorbital: 25-30 mm

Intracanalicular: 6-9 mm

Intracranial: 10 mm



## **Optic Nerve Vascular Supply**



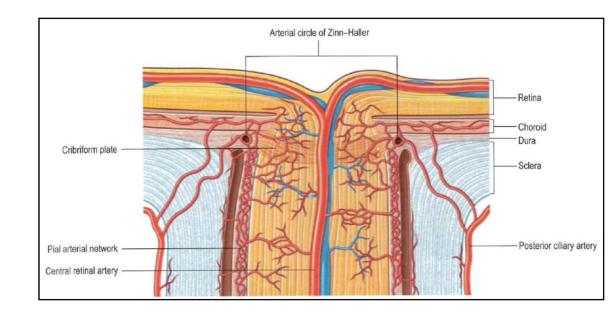
**Surface RNFL**: retinal arterioles

**Prelaminar**: branches of Posterior Ciliary Artery (PCA)

Lamina cribrosa: short PCA's (Circle of Zinn Haller)

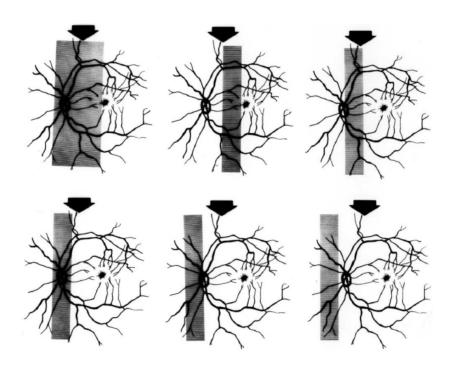
**Retrolaminar**: Pial vessels, branches from ophthalmic

artery, branches from CRA



## **Watershed Zones**





Border between the territories of any two end-arteries

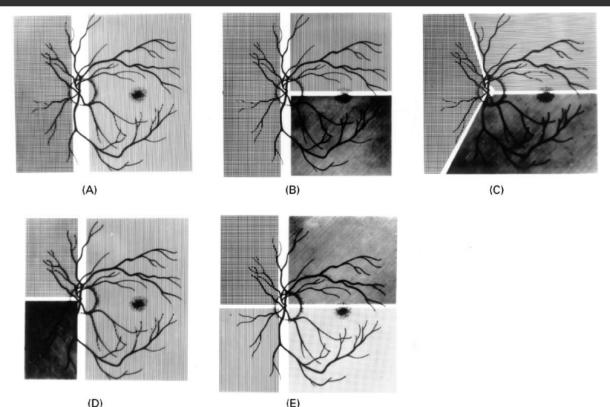
If perfusion pressure drops in the vascular bed of an end-artery, the watershed zone is the most vulnerable to ischemia

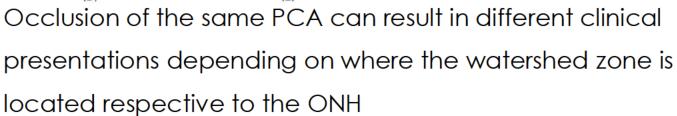
The watershed zone passes through the ONH in most individuals

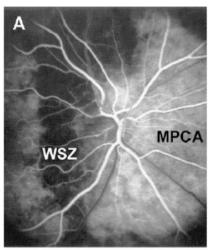
There is significant variability in its location, which results in different clinical presentations of the same pathologic process

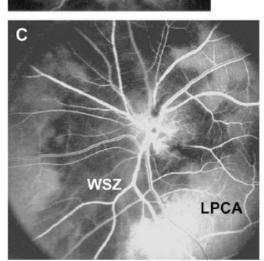
## **Watershed Zones**

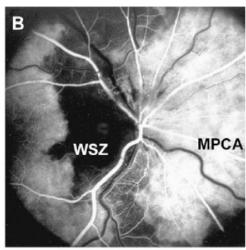


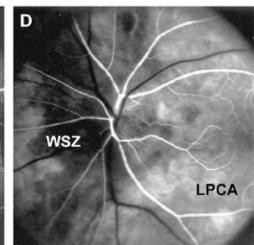
















**VA** loss

VF loss

Dyschromatopsia

Reduced contrast sensitivity

**RAPD** 



# **Clinical Cases**



## 64-year-old Caucasian male

CC: gradual blur in the right eye over a one-month period

POH: longstanding stable vision blur OS, pseudophakia OU

PMH: hypertension, hypercholesterolemia

Medications: Lisinopril, Simvastatin, Viagra

Social Hx: 1-2 glasses of wine each week, 1 cigar on Sunday nights



BCVA: OD 20/60 OS: 20/30

Refraction: OD: +0.75 OS: +1.00

EOMs: Full & smooth OD & OS

Pupils: PERRL (-) APD

CVF: Constricted to finger count 360 OD & OS



Slit Lamp: Normal findings for age OD & OS

Tonometry: OD: 15mmHg OS: 15mmHg, time: 1:30 PM

DFE: Refer to photos

Threshold VF: Refer to photos

BP: 140/95, RAS at 2:00 PM



**Assessment:** Optic nerve edema OD, optic nerve atrophy OS. Likely due to sequential NAION, however must rule out space-occupying lesion.

**Plan:** MRI of brain and orbits with and without gadolinium contrast to rule out space-occupying mass, with special attention to the left frontal lobe. Order bloodwork: CBC with differential, ESR, CRP, RPR-VDRL. RTC 1 week for neuroimaging and bloodwork review.



Concerning element: self-reported gradual vision blur OD

#### Differential diagnoses

Compressive optic neuropathy (Foster-Kennedy Syndrome)

Non-arteritic anterior ischemic optic neuropathy (N-AION)

Arteritic anterior ischemic optic neuropathy (A-AION)

Inflammatory optic neuropathy

Infectious optic neuropathy

## **Bloodwork Review**



High-yield tests

CBC with differential

**ESR** 

**CRP** 

RPR-VDRL

ACE



Results: MRI non-contributory. Bloodwork non-contributory.

**Assessment:** Sequential NAION.

**Plan:** Patient educated about guarded potential for visual recovery. Discussed potential risk factors for additional ischemic events (nighttime use of HTN medication, Viagra, smoking) and communicated the same concerns to patient's PCP. Monitor in 6 weeks.

## **NAION**



#### Risk factors

- Disc at risk
- Glaucoma
- HTN
- DM
- Sleep apnea

- Atherosclerosis
- Arteriosclerosis
- Nocturnal arterial hypotension
- Elevated cholesterol
- Ischemic heart disease
- Age > 55 years

## **NAION**



#### Features

- Males > Females
- Caucasian
- VA varies from 20/20 20/200
- Inferior altitudinal VF defect most common
- VA and VF can improve up to 6 months, but minimal change after that
- PDE5 inhibitors can lead to NAION in patients with predisposing risk factors

#### **NAION Treatment**



Reduction of risk factors

Systemic corticosteroids

- Hayreh and Zimmerman (2008): 364 treated eyes in acute phase showed more VA improvement than non-treated eyes<sup>7</sup>
- Saxena et al. (2018): steroids improved resolution of ON edema in acute phase, but did not show significant improvement in VA at 6 months compared to controls<sup>8</sup>

Optic nerve sheath decompression

Aspirin

Intravitreal steroids and anti-VEGF

## **Don't forget Arteritic AION**



#### Features

- Systemic vasculitis: T-cell dependent disease with predilection for medium and large sized arteries
- Predilection for PCA, resulting in thrombotic occlusion and severe ischemia
- 50+ years of age
- 71% F, 29% M
- Predilection for Caucasian patients
- Amaurosis fugax: sign of impending vision loss in GCA
- Occult GCA: 21% of patients have vision loss without any systemic symptoms

## **Don't forget Arteritic AION**



Systemic symptoms: anorexia, weight loss, headache, jaw claudication, scalp tenderness, neck pain, myalgia, fatigue, anemia

Bloodwork: CBC with differential (includes platelets), ESR, CRP

Temporal artery biopsy: at least 1 inch long

#### GCA treatment:

- High-dose corticosteroids with slow taper
- Tocilizumab (Actemra) Interleukin-6 receptor inhibitor maintains sustained remission after a 26week steroid taper



# CASE 2



## 34-year-old Latino male

CC: sudden onset blur in the right eye

HPI: patient noticed vision loss 1 day ago when he closed his left eye at the shooting range

POH: unremarkable prior to today's visit

PMH: non-secreting pituitary macroadenoma 8 years prior to current presentation

Medications: none

Social Hx: patient denies history of alcohol or recreational drug use



BCVA: OD: CF @ 1 ft. OS: 20/20

Refraction: OD: plano OS: plano

EOMs: Full & smooth OD & OS

Pupils: PERRL (+) Grade 4 APD OD

CVF: OD: severe constriction 360 OS: FTFC



Slit Lamp: Normal findings for age OD & OS

Tonometry: OD: 14mmHg

OS: 14mmHg, time: 11:00 AM

DFE: Refer to photos

Threshold VF: Refer to photos

Ishihara Color Vision: OD: 1/14 plates seen OS: 14/14 plates seen

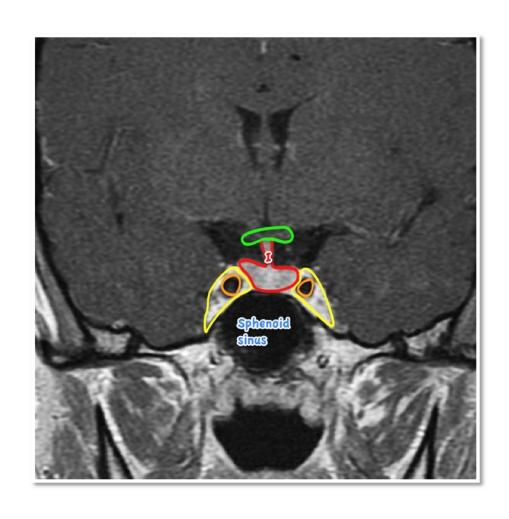


**Assessment:** Optic nerve atrophy OD with significant VA and VF loss, likely due to pituitary adenoma recurrence. Early temporal hemianopic VF defect OS, likely due to pituitary adenoma recurrence.

**Plan:** STAT referral to neurosurgery for MRI of brain with attention to parasellar region. RTC to clinic after full evaluation and any necessary intervention by neurosurgery.

## **Normal Parasellar Anatomy**





## **Case 2: Considerations**



Compressive optic neuropathy typically has a gradual onset and progression.

Clinicians must always have a high suspicion of a tumor recurrence in a patient with optic neuropathy who has a history of a prior tumor.

Rapid identification of a visual pathway tumor leads to earlier treatment and better prognosis for visual recovery.

## **Pituitary Adenoma**



Most common benign tumor of the sellar region

F > M

4<sup>th</sup> - 5<sup>th</sup> decades

10-15% of all intracranial neoplasms

Usually a slow-growing epithelial tumor, but aggressive growth is possible

Affects 0.1–0.2 % of the population

Hormonal changes may result due to overproduction or underproduction of hormones

## Pituitary Adenoma: good prognostic factors



Tumor size < 3 cm

Younger age

Shorter duration of symptoms

Less pre-operative VA and VF loss

Normal color vision

Minimal optic nerve pallor

Normal ERG

Less RNFL thinning on OCT

Normal ganglion cell complex on OCT

## Pituitary Adenoma: Optometrist's Role



Make the appropriate referrals

Neurology/neurosurgery

Endocrinology

- Frequent follow-up after initial diagnosis, then longer intervals after findings have stabilized
- Communicate any changes in VA and/or VF to neurosurgery and endocrinology teams immediately

## **Approaching Optic Neuropathies**



Rule out the most life-threatening and vision-threatening etiologies first

Malignant hypertension

Arteritic AION

Intracranial mass

Order targeted bloodwork and neuroimaging based on the clinical presentation and case history

MRI is more sensitive than CT for neurologic disease

MRI of orbits is indicated when optic nerve disease is present

Gadolinium contrast: highlights pathologic lesions with blood brain barrier breakdown

Consider rare etiologies when the pieces don't fit

