OPTIC NERVE DISEASE ACROSS AGES

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OBJECTIVES

- Provide an overview of optic neuropathies that present in different age groups.
- Review how to assess a patient with suspected optic neuropathy
- Describe the different characteristics of the optic neuropathies and key features to recognize
- Discuss the different modalities for these conditions of the optic nerve

CASE PRESENTATION

- 56 year old African American female presents with decrease vision/dimming in her left eye for 4-5 days. She reports no other associated symptoms.

- Ocular hx is unremarkable
- Medical hx is positive for diabetes (x 10 yr.s) and hyperlipidemia
- Medications for diabetes and hyperlipidemia

CASE PRESENTATION

- VA OD = 20/20
  OS = 20/40- (PHNI)

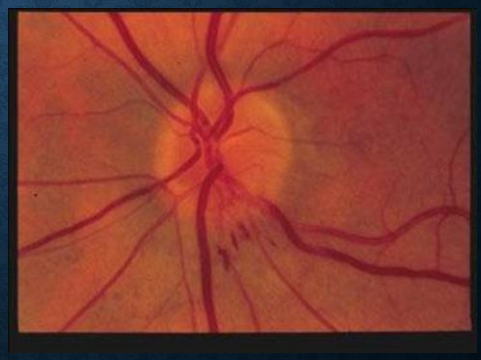
- Pupils were round/reactive to light (OD>OS) with a 1+ APD noted in the left eye
- EOM's were unremarkable
- SLE and IOP's were unremarkable

- Other testing to consider that you can do at that moment?

CASE PRESENTATION

- Right eye – unremarkable

Left eye



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CASE PRESENTATION

- What are your differentials??

- How would you manage this patient?

CASE PRESENTATION

- 56 year old African American female presents with a gradual decrease vision/dimming in both eyes for 4-5 month. She reports no other associated symptoms.

- Ocular hx is unremarkable
- Medical hx is positive for diabetes (x 10 yr.s) and hyperlipidemia
- Medications for diabetes and hyperlipidemia

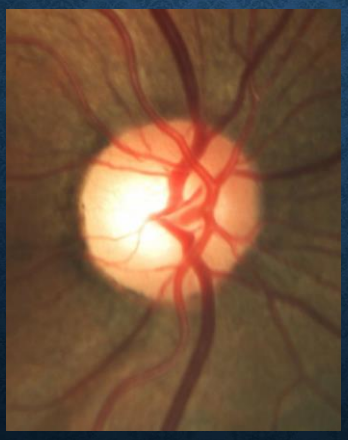
CASE PRESENTATION

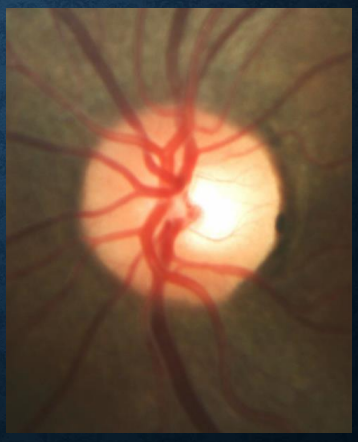
- VA OD = 20/40- (PHNI)
  OS = 20/40- (PHNI)

- Pupils were round/reactive to light (OD=OS) with negative APD noted
- EOM's were unremarkable
- SLE was unremarkable and IOP's (20mmhg OU)
- Other testing to consider that you can do at that moment?

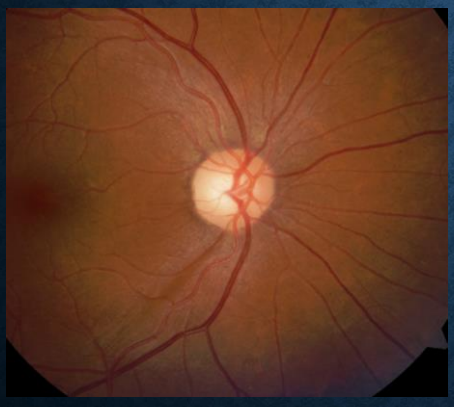
CASE PRESENTATION

- Posterior Segment

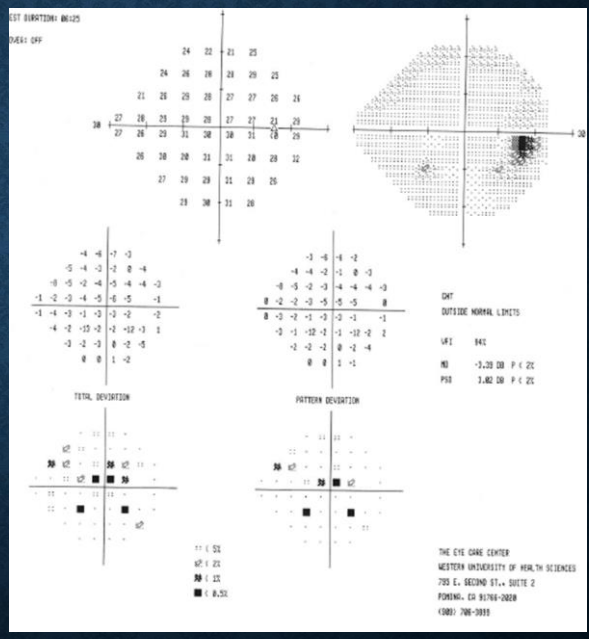


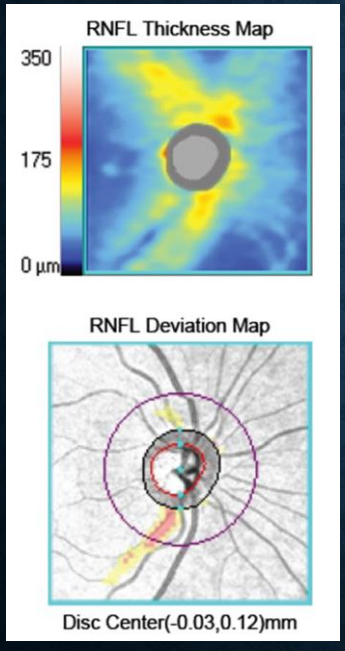


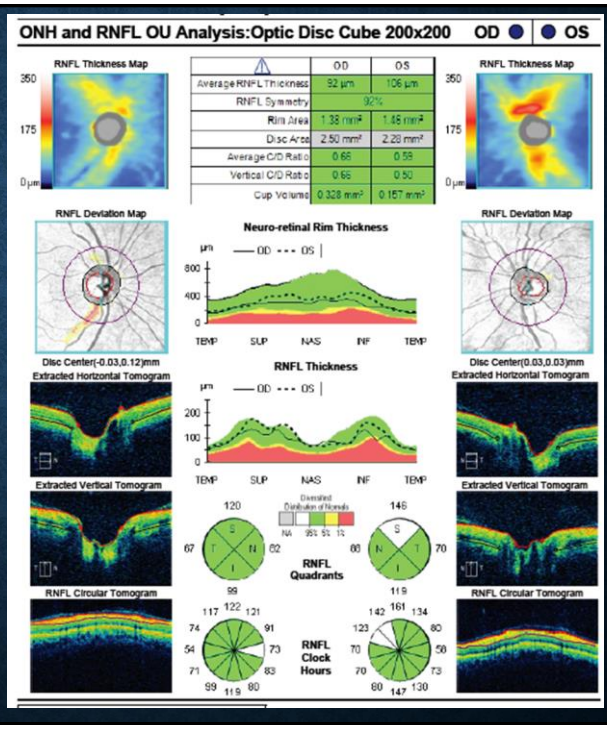
CASE PRESENTATION



Visual Fields
SITA STD 24-2
OD good reliability with
paracentral defects
MD = -3.39 PSD = 3.02
OS good reliability / unremarkable

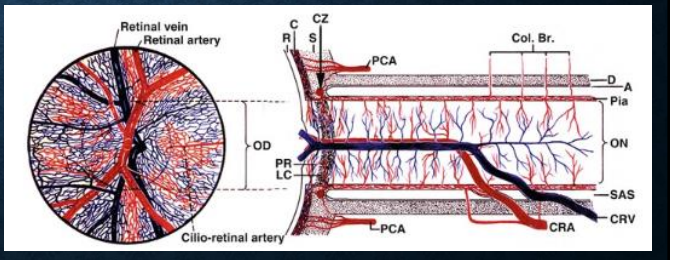






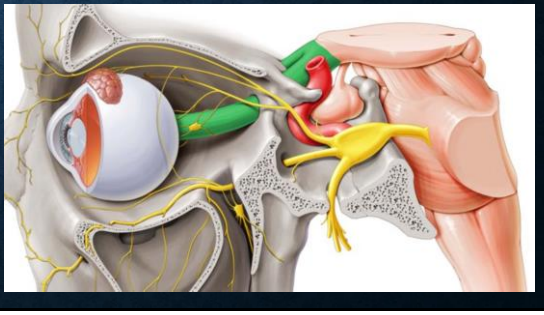
ANATOMY OVERVIEW

- The optic nerve can be subdivided into 4 main parts:
  - Intraocular (optic nerve head) – measures ~1mm and is located within the eye where it is approximately 1.5mm horizontally x 1.8mm vertically.
    - Blood supply = typically supplied by the retinal arterioles.
  - Intraorbital – is the ~25mm of optic nerve travelling from the posterior segment to the intraorbital opening of the optic canal.
    - Main blood supply – peripapillary choroid (segmental)
    - Surrounded by the meningeal layers: dura, arachnoid, and pia mater



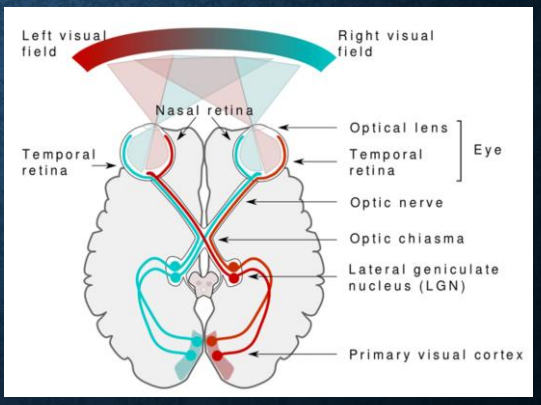
ANATOMY OVERVIEW

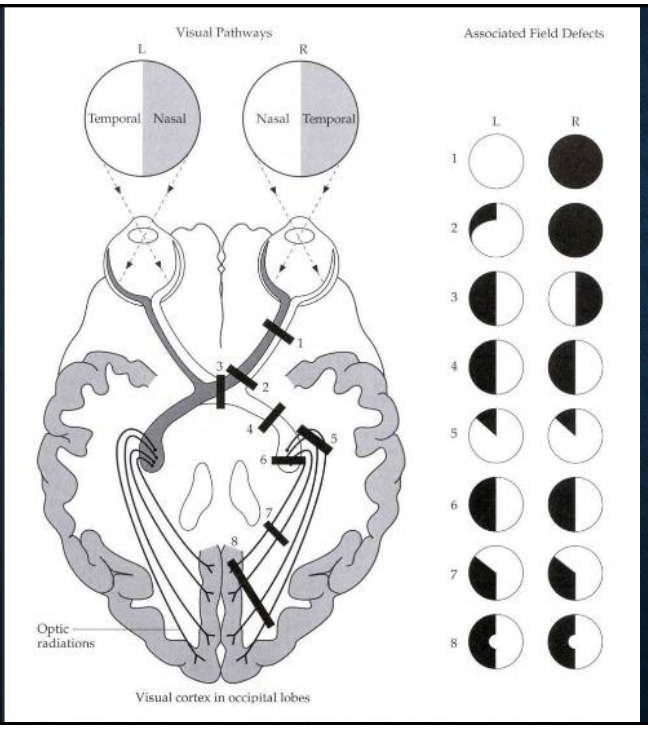
- Intracanalicular – lies within the optic canal, which is formed by the lesser wing of the sphenoid bone. The length is variable (between 4-10mm).
- Intracranial – the last 10mm of the optic nerve. The optic nerve unites with its contralateral optic nerve to form the optic chiasm. Key areas it passes prior to this: above the diaphragm sellae before passing above the suprasellar part of the cavernous sinus.



ANATOMY OVERVIEW

- At the optic chiasm, the optic nerve fibers originating from the temporal side of the retina of the right eye continue in the right optic tract (post chiasmatic part of the optic nerve). At the point of the decussation, the fibers that originated from the nasal field of the left eye, cross over and enter the right optic tract. Therefore, visual input from the left visual field travels in the right optic tract. A similar decussation occurs with fibers arising in the nasal side of the contralateral eye.





OPTIC NERVE ASSESSMENT

- Case Hx – as with any disease process it is important to ascertain time of onset, duration, & resolution of symptoms.
- Entrance Testing – VA symptoms tend to be described as dimming/graying of vision rather than reduced acuity/metamorphopsia. Other tests: Pupils and CV are remarkable with optic nerve disorders
- Ocular Health Assessent
  - Cupping vs Pallor
- Visual Field Testing – variable with optic nerve disease

OPTIC NERVE ASSESSMENT

ONH Disease

- Pupil testing *
- Color vision *
- Red cap desaturation
- Pulfrich Phenomena
- Contrast Sensitivity

Macular Disease

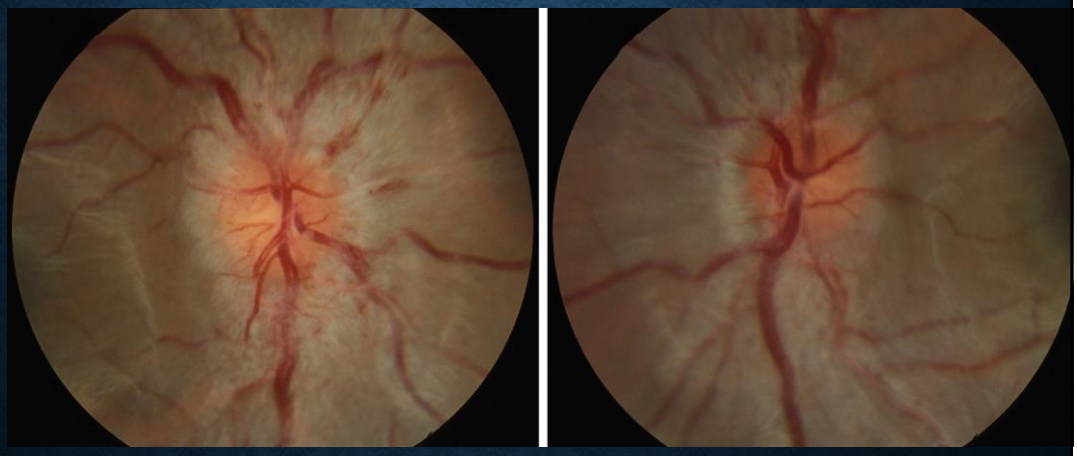
- Amsler Grid
- Photostress Test

ONH DISORDERS: < 20YO

- Not including developmental disorders (optic nerve hypoplasia, myelinated RNFL, Morning Glory syndrome, Leber's Hereditary Optic Neuropathy)
- Papillitis – swelling of the optic nerve due to inflammation
  - Over 75% of papillitis in children is preceded by an episode of fever (viral) or related with immunizations (hepB and influenza vaccines).
    - MOA is unknown but is hypothesized to be a delayed type IV hypersensitivity reaction.

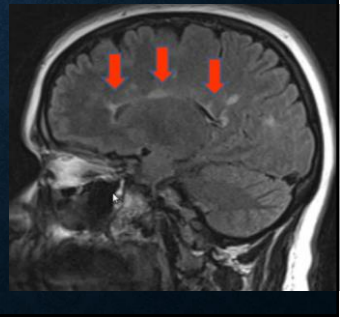
ONH DISORDERS: < 20YO

- Symptoms – vision loss (*bilateral), pain behind the eyes (not a consistent feature), and an alteration in color vision.
- Ocular health – optic nerve heads appear swollen
- Treatment is conservative unless involvement is bilateral and VA is dramatically reduced.
- Prognosis – Good (better than 20/40 in 80% of cases)



ONH DISORDERS: 20 - 40YO

- Optic Neuritis
  - Is inflammation of the optic nerve that mainly occurs due to demyelinating event (most commonly multiple sclerosis).
  - Demyelinating optic neuropathy / Retrobulbar optic neuritis presents as unilateral vision loss mainly observed in female patients (77%).
  - Symptoms: pain upon eye movement, RAPD, decreased CV, VF defects, and vision changes.
  - Often there are no ophthalmoscopic signs of inflammation (2/3)



ONH DISORDERS: 20 - 40YO

- Optic Neuritis (cont'd)
  - Uhthoff's phenomenon – transient worsening of visual symptoms with increase body temp.
  - L'Hermitte's sign – electronic "shock-like sensation" that runs down the spine and into the upper extremities with forward lexion of the neck.

ONH DISORDERS: 20 - 40YO

- Visual Field defects can be variable. . .

ONH DISORDERS: 20 - 40YO

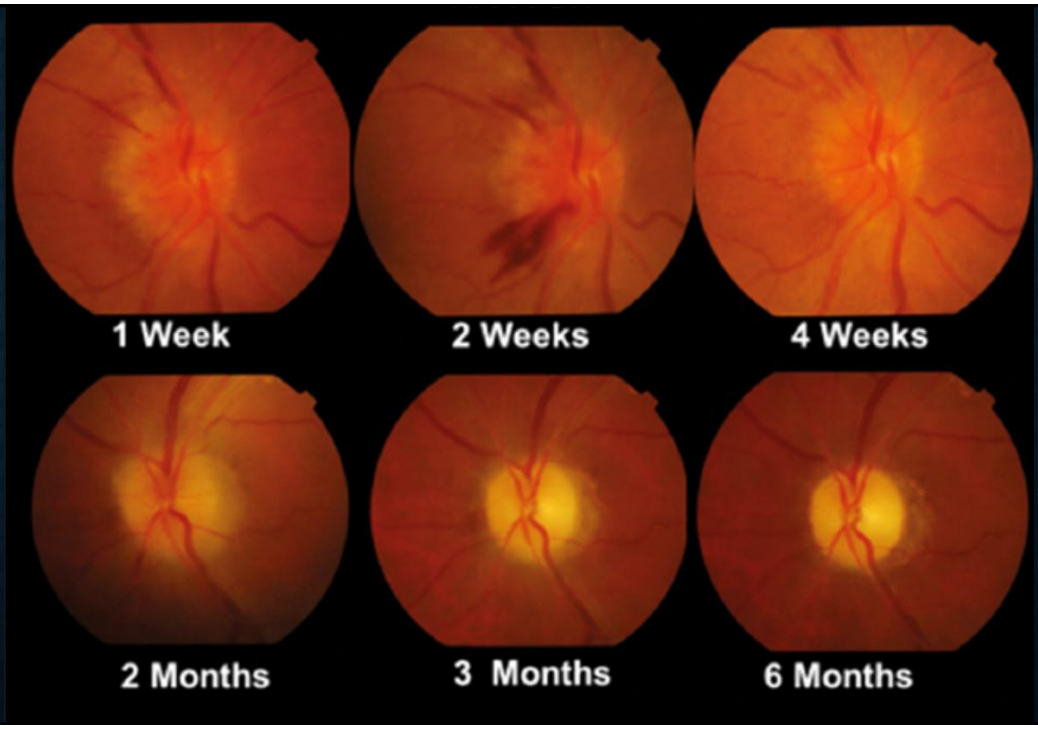
- Optic Neuritis (cont'd)
  - Is the 1st presenting symptom in 20% of patients who subsequently are diagnosed with MS and it is seen in nearly 75% of MS patients at some point. Most useful predictor of clinically definite multiple sclerosis (CDMS) in a patient with optic neuritis = presence of white matter lesions on MRI.
  - Optic Neuritis Treatment Trial (ONTT) – risk of CDMS with one or more lesions on MRI was 56% at 10 years and only 22% in patients who had normal MRI scans. Key finding: pt's with two or more lesions on MRI who were treated with high-dose IV methylprednisone had a 50% less chance of developing CDMS vs those treated with placebo or oral prednisone. BUT offers only short-term reduction – by 3 yr.s post-treatment the protective benefits ceased.
  - **Oral prednisone actually was found to increase the risk of recurrent optic neuritis in the same or fellow sys.**

ONH DISORDERS: 20 - 40YO

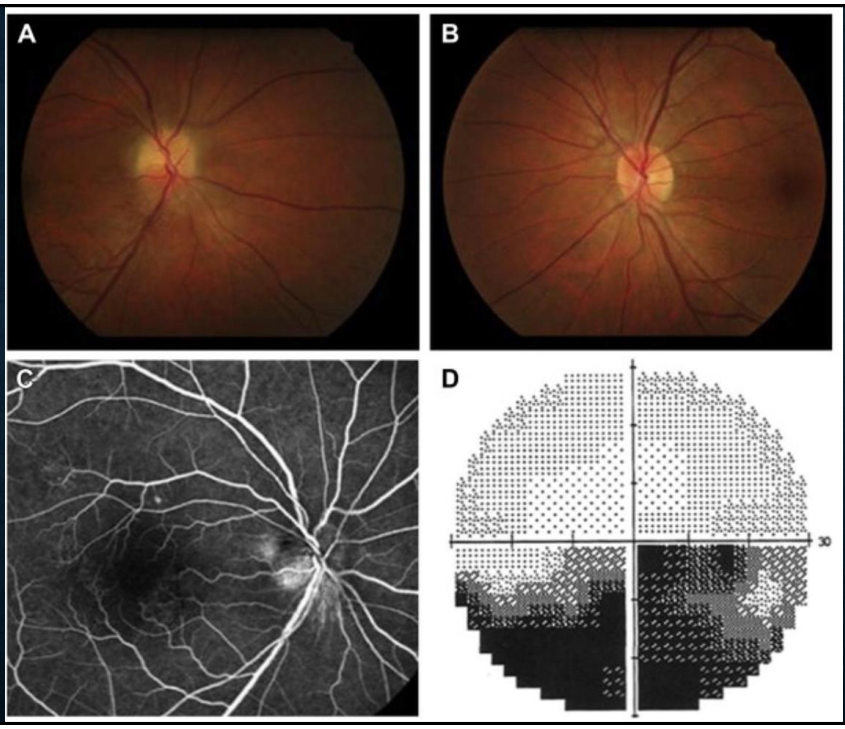
- Optic Neuritis (cont'd)
  - Controlled High Risk Subjects Avonex Multiple Sclerosis Prevention Study (CHAMPS) – demonstrated that the risk of developing CDMS was further reduced from 50% in the placebo group to 35% in the treatment group when the patient was treated with weekly injections of Avonex (interferon-beta1a, Biogen Idec), an immunomodulating agent. There was also a reduction in the # of lesions detected by MRI at 18 months.
  - BENEFIT Study (Betaferon/Betaseron in Newly Emerging MS for Initial Treatement) - provide further evidence supporting early initiation of treatment with interferon beta -1b in patients with a frist event suggestive of MS.
  - Visual prognosis is good in optic neuritis with most patients recovering to 20/20 – 20/40, but most patients will experience ecrease in visual function due to changes to contrast sensitivity, color saturation and visual field.

ONH DISORDERS: 40 - 60YO

- Glaucoma
- Non-arteritic anterior ischemic optic neuropathy (NAAION)
  - Pt's typically >50
  - Ischemic process affecting the SPCA
  - Typically presents as sudden, painless loss of vision with vision worsening over the next 2 weeks (45%) and up to 1 month (29%) with VA typically better than 20/60 in 50% of cases.
  - ONH appearance – edema and flame hemorrhages
  - Visual field defects tend to be altitudinal
  - Risk factors: small disc, microvascular disease
  - Fellow eye involvement – 18%



CullenJ. Non-arteritic anterior ischaemic neuropathy(NA_AION) : a review. EyeNewsUK.com

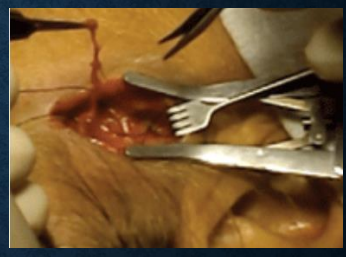


Tzu-LunH, Kung-Hung L, Rong-KungT.Tzu. Treatment strategy for non-arteritic anterior ischemic optic neuropathy. Chi Medical Journal Jan-2013)

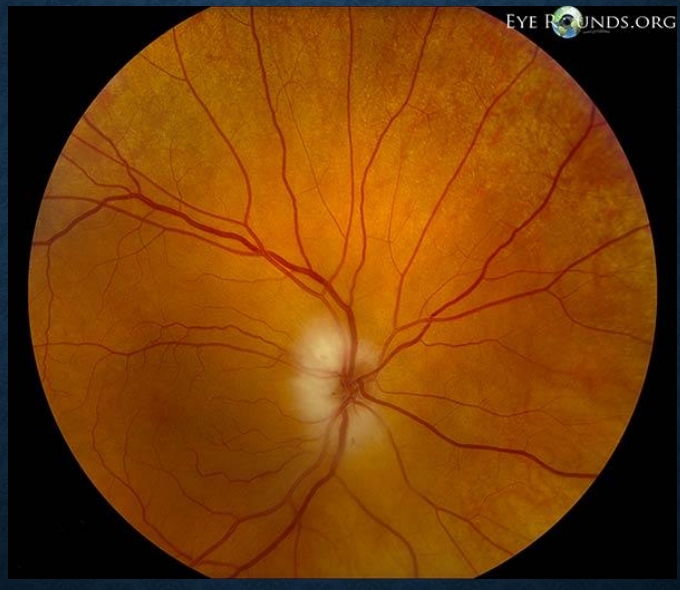
ONH DISORDERS: > 60YO

- Arteritic Anterior Ischemic Optic Neuropathy (AAION)
  - Occurs mainly in women with the avg age of onset being 75yo
  - Is an inflammatory disorder of the arteries that supply the optic nerve. Strongly associated with Giant Cell Arteritis (systemic inflammation of the elastic tissue of arterial walls)
  - Vision loss is severe, sudden, and rapidly progressive with most patients with VA of 20/200 to NLP.
  - Onset is typically unilateral but fellow eye can be involved in the next 10 days (up to 75% of untreated patients)
  - Symptoms: temporal HA's, jaw claudication, neck pain/stiffness, scalp tenderness, anorexia, myalgia (polymyalgia rheumatica), malaise, weakness and/or fever.
  - TMB 2-19% / Oc. Motility concerns 2-43% (CN III) / CRA 5-10%

ONH DISORDERS: > 60YO

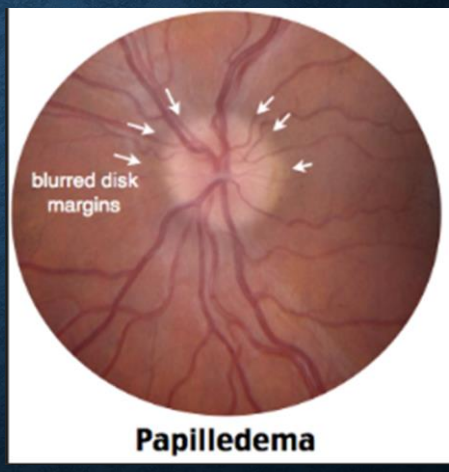


- AAION
  - Laboratory Testing: Westergren erythrocyte sedimentation rate / C-reactive protein
    - Both are indicators of acute inflammatory activity.
    - Together they have a 97% sensitivity for diagnosing GCA
  - Temporal artery biopsy
- Treatment
  - In pt's over 60yo assume GCA and treatment should be initiated until prove otherwise.
  - High-dose corticosteroid therapy – typically IV or methylprednisolone 250mg q6hr.s for 12 doses with oral maintenance doses until blood work is reduced.



ODDS AND ENDS

- Papilledema
  - Optic disc swelling and elevation secondary to increased intracranial pressure.
  - Typically bilateral with vessels getting obscured in the acute phase.
  - Paton's lines – swelling that is visualized as retinal folds/lines circumferentially around the optic nerve.



Papilledema

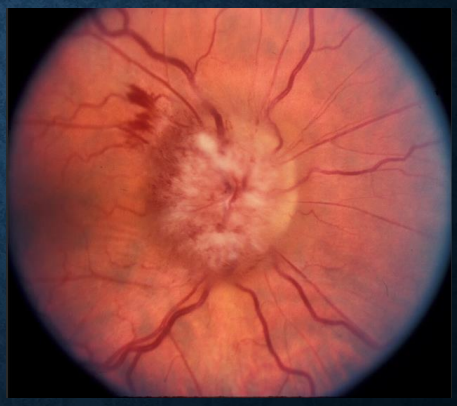
Table 1. Modified Frisén Scale

Papilledema Grade
0 (Normal Optic Disc)
Prominence of the retinal nerve fiber layer at the nasal, superior, and inferior poles in inverse proportion to disc diameter
Radial nerve fiber layer striations, without tortuosity
1 (Minimal Degree of Edema)
C-shaped halo that is subtle and grayish with a temporal gap; obscures underlying retinal details[a]
Disruption of normal radial nerve fiber layer arrangement striations
Temporal disc margin normal
2 (Low Degree of Edema)
Circumferential halo[a]
Elevation (nasal border)
No major vessel obscuration
3 (Moderate Degree of Edema)
Obscuration of ≥1 segment of major blood vessels leaving disc[a]
Circumferential halo
Elevation (all borders)
Halo (irregular outer fringe with finger-like extensions)
4 (Marked Degree of Edema)
Total obscuration on the disc of a segment of a major blood vessel on the disc[a]
Elevation (whole nerve head, including the cup)
Border obscuration (complete)
Halo (complete)
Grade 5 (Severe Degree of Edema)
Obscuration of all vessels on the disc and leaving the disc[a]

[a] Key features (major findings) for each grade.

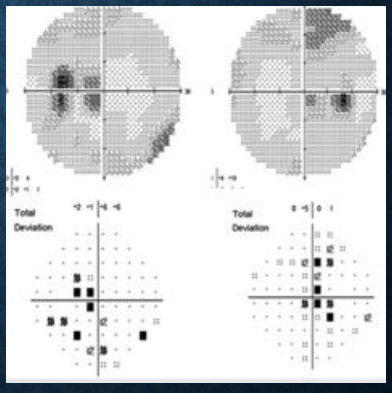
ODDS AND ENDS

- Symptoms: transient vision obscurations (prodrome), HA's (worsen with postural changes), transient horizontal diplopia, mild neck discomfort, tinnitus, nausea, vomiting, and dizziness.
- Vision is minimally affected until edema progresses to optic atrophy.
- Causes: intracranial or spinal tumor, cerebral edema or encephalitis, epidural or subdural hematoma, subarachnoid hemorrhage, meningitis, venous sinus thrombosis (VST), drug or vitamin toxicity, or idiopathic intracranial hypertension.



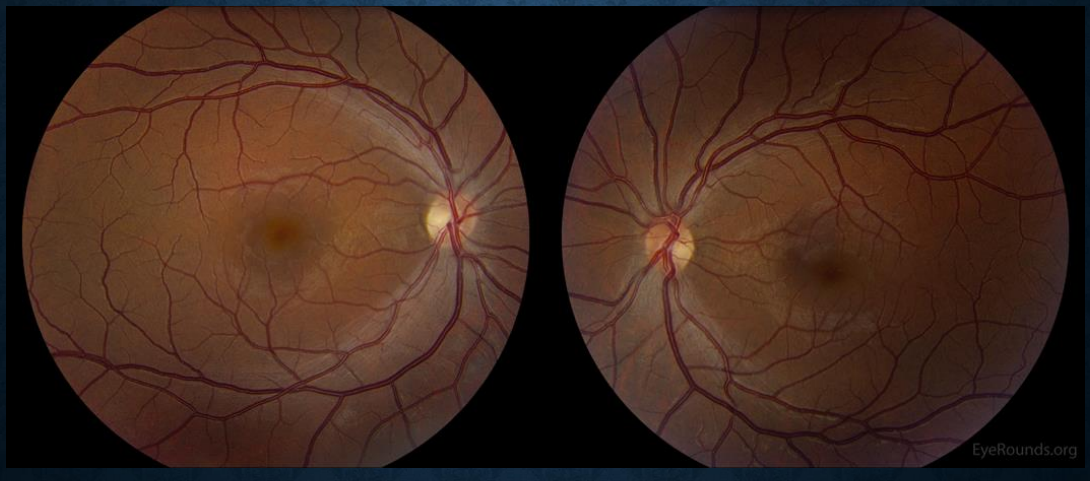
ODDS AND ENDS

- Toxic/Nutritional Optic Neuropathy
  - Mainly associated with patients with personal histories of alcohol and tobacco abuse.
  - Symptoms: gradual blurring/dimming of vision. Vision loss is gradual, painless and typically symmetric with VA ranging between 20/50 to 20/200.
  - Visual Fields – reveal central or cecocentral scotomas.

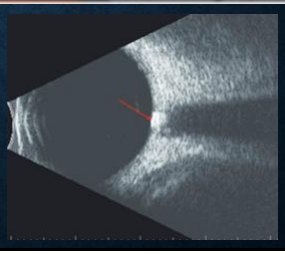


ODDS AND ENDS

- Toxic/Nutritional Optic Neuropathy
  - Signs: ONH appearance can range from unremarkable to edematous with peripapillary hemorrhages (acute cases) and temporal pallor (chronic cases) with RNFL loss greatest in the papillomacular bundle.
  - Vitamin B12 deficiency common due to absorption being affected in alcoholics and nutritional/dietary intake being poor in these patients.



ODDS AND ENDS



ANY QUESTIONS?

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