I. Arteritic Anterior Ischemic Optic Neuropathy (A-AION)
   a. Giant Cell Arteritis (GCA) is an Immune-Mediated Vasculitis
   b. Affects Medium and Large Arteries
      i. In eye mostly Posterior Ciliary Artery (PCA), then Central Retinal Artery (CRA), rarely Ophthalmic Artery (OA)
      ii. Explain why GCA affects eyes
      iii. Called GCA because of granulomatous inflammation. Arteries lined with multinucleated giant cells: macrophages, lymphocytes, and fibroblasts
   c. Incidence of 2.3/100,000 in 6th decade. Increases with Age.
   d. Happens mostly to those ≥ 50 years old
      i. Age range
   e. Predilection for North European, Scandinavian descent. Typically Caucasian though can happen in other races
   f. Why is it important?
      i. Profound vision loss
         1. Causes A-AION
         2. Can become bilateral in 14 days in 1/3 if untreated
         3. Treatable
   g. CRAO and Cilioretinal artery occlusion can occur
      i. CRAO occurs XXXX%
   h. A-AION
      i. Sudden painless vision loss
      ii. Unilateral Optic Disc Edema
      iii. 1/10 patients (≥ 50 years old) with optic disc edema are A-AION. Other 9/10 NA-AION
   i. Systemic Symptoms
      i. Jaw Claudication (Odds Ratio 9.0)
      ii. Neck Pain (Odds Ratio 3.4)
      iii. Anorexia
      iv. Less predictable symptoms
         1. Headache
         2. Fever
         3. Scalp Tenderness
         4. Malaise
   j. Visual Symptoms/Signs
i. Sudden, painless vision loss
ii. 31% had amaurosis fugax 1-2 weeks before
   1. This can be misleading. Not saying 31% of patients with Amaurosis Fugax have GCA
iii. 6% diplopia
iv. 8% ocular pain
v. Optic Disc Edema “chalky white pallor”

k. Lab Testing
i. ESR – 86% sensitive
ii. CRP – 100% sensitive, 82% specific
iii. Combined ESR and CRP – 99% sensitive, 97% specific
iv. CBC with differential looking for disease that can affect Red Blood Cells (i.e. Anemia, Polycythemia Vera)

l. Other Diagnostic Testing
i. Fluorescein Angiography (FA) – characteristic late filling of choroid in the 2 weeks after Optic Disc Edema starts
ii. Ultrasound, PET, MRI of limited benefit
iii. Temporal Artery Biopsy – especially indicated if clinical symptoms highly suggestive but either ESR or CRP are inconclusive

m. This is a diagnosis that can be made by Optometrist or Ophthalmologist with use of ocular signs and symptoms combined with systemic symptoms and lab testing. Additional testing often not needed.

n. Contralateral optic nerve can give us clues to A-AION vs. NA-AION
i. Go through Bayesian analysis
   1. If C/D is ≥ 0.4 1/5 have A-AION
   2. If C/D is ≤ 0.3 1/15 have A-AION

o. Some advocate American College of Rheumatology criteria
i. Need 3 of the following 5 criteria
   1. Over 50 years of age
   2. New onset of Headache
   3. Scalp tenderness or decreased temporal artery pulse
   4. ESR > 50 mm/h
   5. (+) Temporal Artery Biopsy
ii. Having 3 of these 5 gives 94% sensitivity and 91% specificity (not good enough)
p. Treatment
   i. Most studied and likely still most effective is oral steroid
      1. 80-100 mg/day until labs normalize
      2. Then taper for VERY long (years!)
      3. Evidence still lacks because not ethical to have a placebo group
   ii. No evidence that IV steroid in mega dose any more effective
   iii. Limited evidence for TNF blockers, Methotrexate and other immune modulators

q. Clinical Picture
   i. Unilateral Optic Nerve Edema
   ii. Systemic Symptoms
   iii. Possible Visual Symptoms
   iv. Lab Results
      1. ESR > 47 mm/h
      2. CRP > 2.45 mg/dl
   v. Optic Nerve in other eye > 0.4
   vi. Positive Temporal Artery Biopsy

II. NA-AION
   a. NA-AION is due to acute ischemia of ONH
      i. Primarily ischemia of PCA
      ii. Not Thromboembolic
         1. Proven by FA and transcranial Doppler
   b. Characterized by sudden, painless, unilateral vision loss
      i. 75% wake up with vision loss
   c. Happens to age 49-64
   d. Incidence of 2.3-10.2 per 100,000
   e. Systemic risk factors
      i. Hypertension
      ii. Diabetes Mellitus
      iii. Nocturnal hypotension
      iv. Hyperlipidemia
      v. Atherosclerosis
   f. Acuity is 20/20 in 1/3 of NA-AION. > 20/40 in 50%. < 20/200 in 20%
   g. No typically first visual field loss
h. More common in small optic cup
   i. $\leq 0.15$ in 33%
   ii. $\leq 0.3$ in 75%

i. **However, not all small cups are at risk. Incidence only 10.2 per 100,000 at most. Average cup 0.3-0.4. So, 66% of population has 0.3-0.4. Not all of them will get NA-AION.**

j. Small cup exacerbates NA-AION form but not primary factor
   i. ONH ischemia leads to axoplasmic flow stasis. In a small, crowded ONH this compresses surrounding tissue. This incites further swelling.
   ii. Nocturnal hypotension is likely inciting factor in most cases

k. Fellow eye
   i. 25% in 3 years
   ii. 17% in 5 years

l. Treatment
   i. 40% spontaneously resolve
   ii. ONH Decompression Trial
      1. 24% progressed with Decompression
      2. 12% progressed if left alone
   iii. ASA is no help
   iv. Corticosteroids may benefit
      1. 1 trial showed 70% in Tx group acuity improved compared to 40% Control
      2. VF improved 40% in Tx group vs. 25% Control
   v. Intravitreal Kenalog, Avastin – no evidence of benefit. Could exacerbate by increasing IOP

m. Amiodorone – no evidence that it causes
   i. Used to treat patients who likely have HTN, DM, and nocturnal hypotension

n. Viagra etc

o. Incipient NA-AION
   i. Not all eyes have poor acuity at onset
   ii. Frequently misdiagnosed as diabetic papillopathy

p. Clinical Picture
   i. Unilateral Optic Nerve Edema
   ii. Age $\geq 50$
iii. No systemic symptoms
iv. Typically Diabetic and/or Hypertensive
v. Optic Nerve in other eye $\leq 0.3$

III. Ophthalmic Sources
a. Drusen – B-Scan helps differentiate
b. CRVO
c. Hypotony
d. CRVO and Hypotony will be obvious upon examination. On the list to remind us that we don't need to search for other causes if these are present

IV. Inflammatory (Can be infectious or non-infectious)
a. Syphilis
b. TB
c. Lyme
d. Sarcoid
e. Uveitis

V. Optic Neuritis (this is a subset of Inflammatory)
a. Acute inflammatory optic neuropathy
   i. Result of demylenating process
b. Incidence between 1-5 per 100,000
c. Age: Majority between 18-46 (95%)
d. ¾ Women. 85% White
e. Symptoms
   i. Sudden unilateral vision loss (anywhere from 20/20 to NLP)
      1. In ONTT, 2/3 of contralateral eyes had some visual function loss
   ii. Pain – worse with eye movements
      1. 92% of patients in ONTT had eye pain
   iii. Vision loss lasts up to 4 weeks days before recovery
      1. May not recover full function
      2. Even if 20/20, patients say vision not as good as before
f. Signs
   i. 1/3 of patients manifest with swollen optic nerve (papillitis)
   ii. 2/3 retrobulbar – no swollen nerve
   iii. +APD
iv. Wide variety of VF defects – none distinguish Optic Neuritis from other Optic Neuropathies

v. Often impaired contrast sensitivity and/or color vision
   1. ONTT – 94% had color defects in acute phase
   2. ONTT – 40% had residual color vision defects
   3. Cannot help us differentiate between other optic neuopathies

   g. Systemic Associations
      i. Multiple Sclerosis (MS)
         1. Optic neuritis occurs in ½ of patients with MS
         2. Is initial event in ¼ patients with MS
         3. Symptoms
            a. Numbness, tingling, weakness
            b. Balance and gait disturbance
            c. Bladder dysfunction
            d. Uhthoff’s phenomenon – heat
            e. L’hermitte symptom

   4. Signs
      a. Oculomotor: nystagmus, INO, 6th nerve palsy
      b. Optic Neuritis

   5. Additional Testing
      a. Lab tests, MRI not needed for Dx of Optic Neuritis. MRI aids in prognosis and diagnosing MS

   6. Optic Neuritis Treatment Trial (ONTT)
      a. Evaluated use of steroids in treating acute optic neuritis
      b. 457 patients with unilateral optic neuritis
      c. Randomized to one of three groups
         i. Oral pred (1mg/kg/day) x 14 days
         ii. IV Methylprednisolone x 3 days then oral prednisone x 11 days
         iii. Oral placebo x 14 days
      d. 93% of all patients recovered to at least 20/40 at 1 year
      e. IV steroid followed by oral steroid speeded recovery (4 days compared to 15 days) and reduced
recurrence of developing Clinically Significant Multiple Sclerosis (CDMS) by 2 years
  i. About 1/3 overall had recurrence. In Oral Pred group, 44% recurrence
  ii. MS patients had more recurrence than non-MS patients
f. Oral steroid did not improve visual outcome and increased rate of new attack of optic neuritis
g. At 10 years:
  i. 91% were 20/40 or better
  ii. Overall rate of developing MS was 38%
    1. If no lesions on MRI, risk of MS was 20% at 10 years
    2. If 1 or more lesion, 56%
h. Clinical Picture
  i. Unilateral Optic Nerve Edema
  ii. Typically 18-45 years of age
  iii. Systemic Symptoms
    1. Possible L'Hermitte, Uthoff
    2. Numbness, tingling
  iv. Varying acuities
  v. +APD, reduced field, wide range of acuity, reduced color, reduced contrast

VI. Compressive
  a. Uncommon
  b. Most likely from Pituitary Tumor
    i. Can push forward and press on optic nerve
    ii. Junctional Scotoma
  c. Graves' Orbitopathy
**Main Points:**
GCA is a diagnosis that Optometrist can make.
  - No expensive testing
  - Very Serious!
Use Age
Use Symptoms
Use Other Eye
  - C/D
  - Bilateral ODE is different