

# SWOLLEN OPTIC NERVES: NOW WHAT?

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## Disclosures

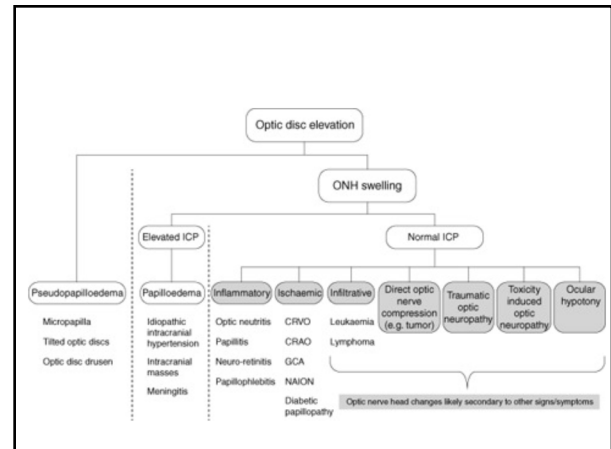
- |                         |                  |
|-------------------------|------------------|
| ▣ Aerie Pharmaceuticals | ▣ Nova Ocular    |
| ▣ Biotissue             | ▣ Novartis       |
| ▣ Diopsys               | ▣ Optovue        |
| ▣ Ellex                 | ▣ Quantel        |
| ▣ EyePromise            | ▣ Reichert       |
| ▣ Ivantis               | ▣ RevolutionEHR  |
| ▣ Lumenis               | ▣ Sight Sciences |
| ▣ Maculogix             | ▣ Shire          |
| ▣ Nidek                 | ▣ Sun Pharma     |

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## Expected Learning Objectives

- ▣ To enable the ON to increase their comfort level in managing swollen optic nerves
- ▣ At end of session, attendees should be able to:
  - To become familiar with the key signs to help differentiate pseudoswelling of the ONH with true swelling of the ONH.
  - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for optic neuritis and MS.
  - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for non-arteritic anterior ischemic optic neuropathy
  - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for arteritic ischemic optic neuropathy
  - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for papilledema/pseudotumor cerebri.
  - To become more familiar and update the signs, symptoms, differential diagnosis and treatment for neuroretinitis.

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## SWELLING VS. PSEUDOSWELLING

- ▣ Ways to differentiate:
  1. Direct viewing of the ONH
    - ▣ Are the vessels blurred as they cross the disc margin?
    - ▣ Is there SVP?

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## Swelling vs. Pseudoswelling?



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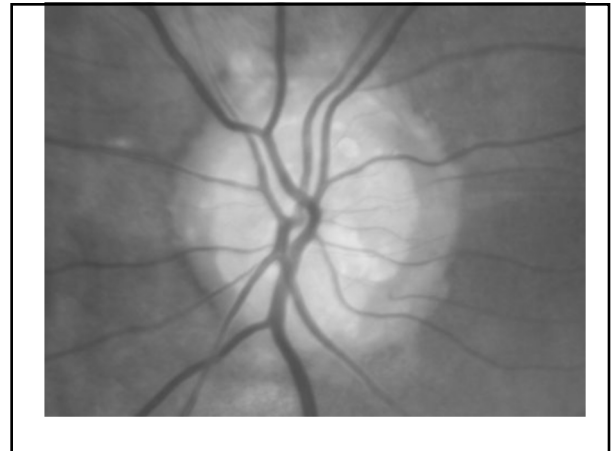
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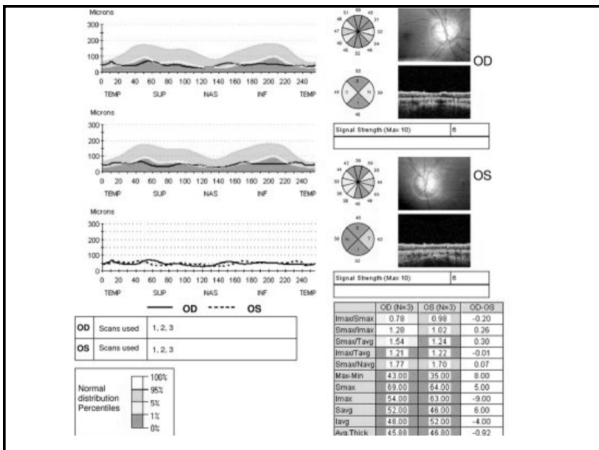
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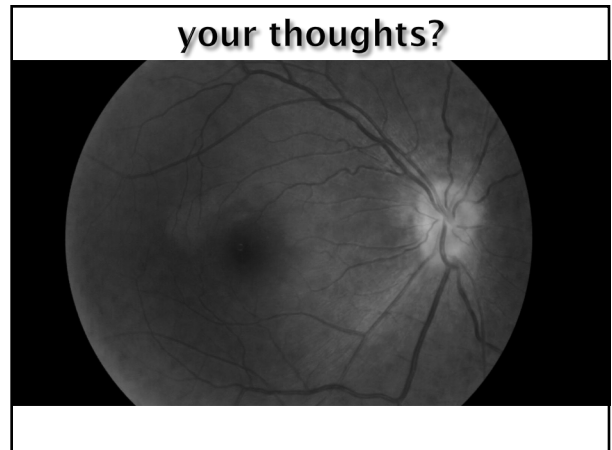
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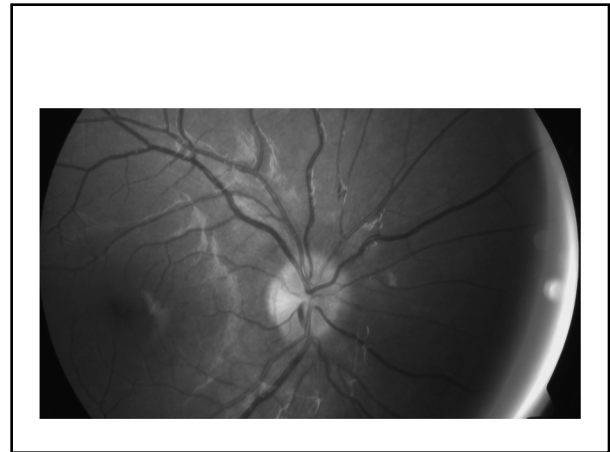
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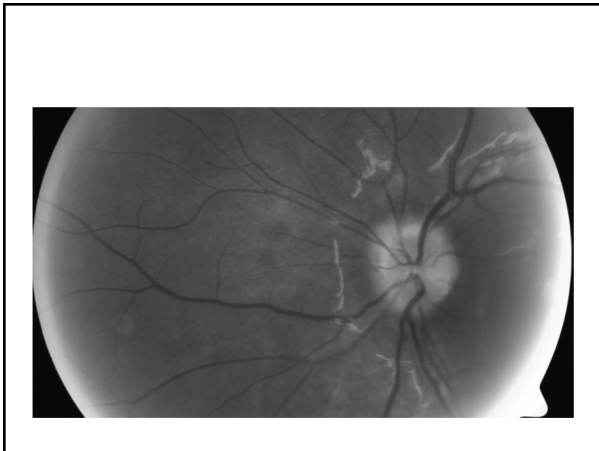
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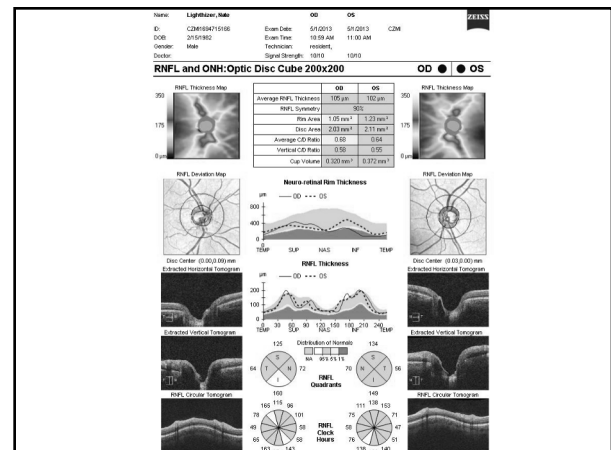
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## SWELLING VS. PSEUDOSWELLING

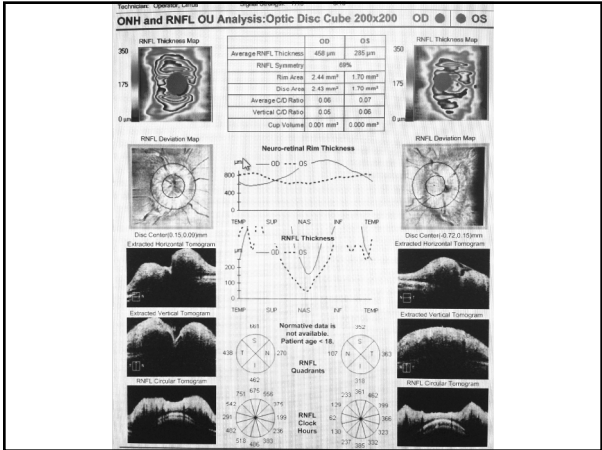
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2. OCT
  - rNFL thickness – normal or elevated or thin?
  - Is there a splitting of retinal layers deep in the retina?

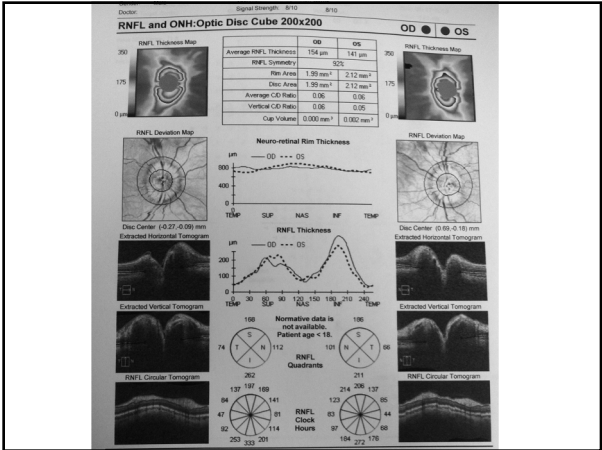
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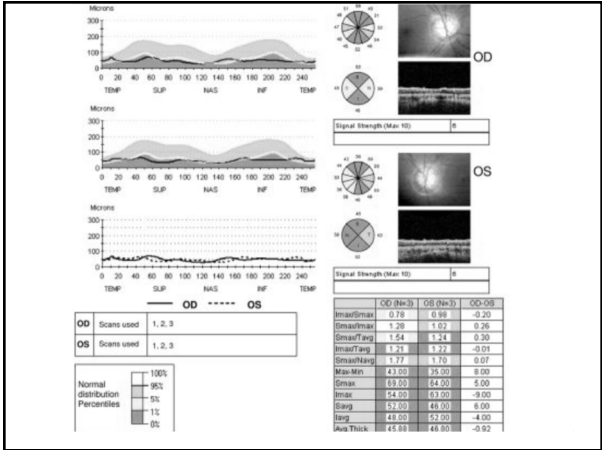
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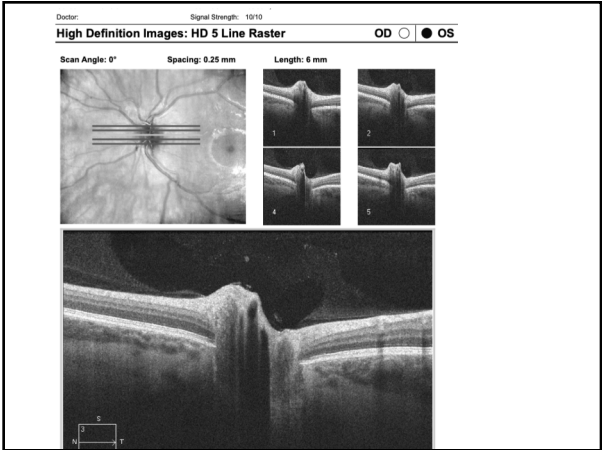
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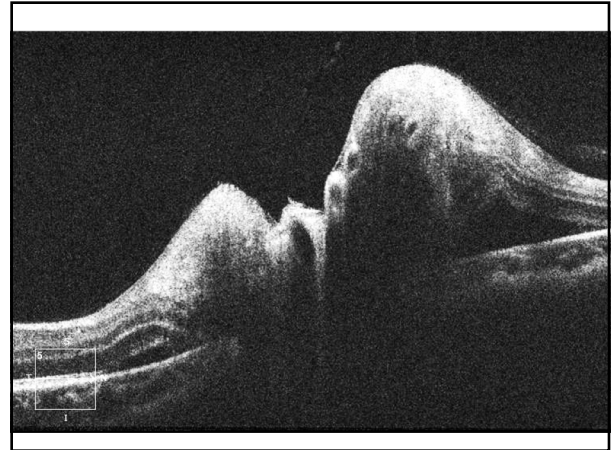


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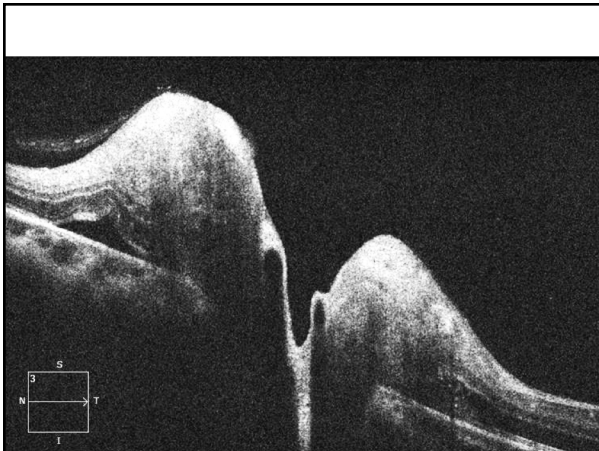




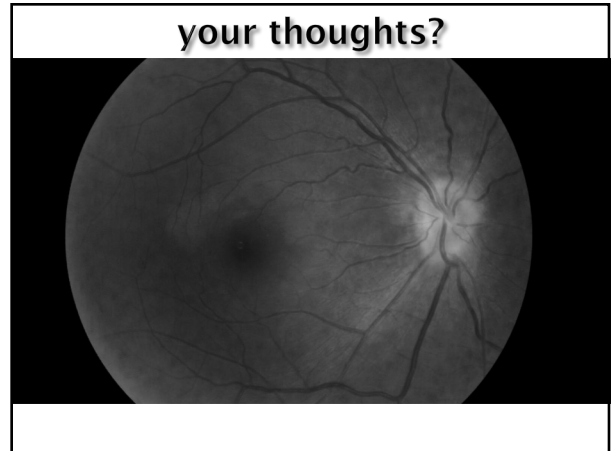
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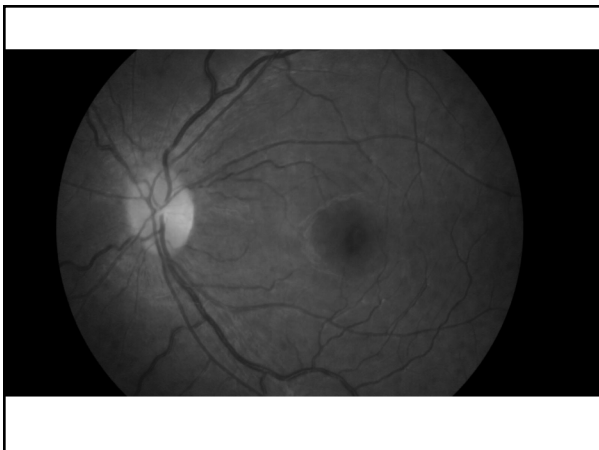
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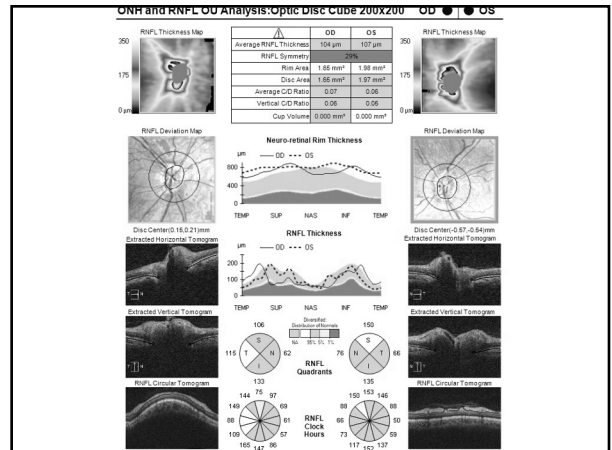
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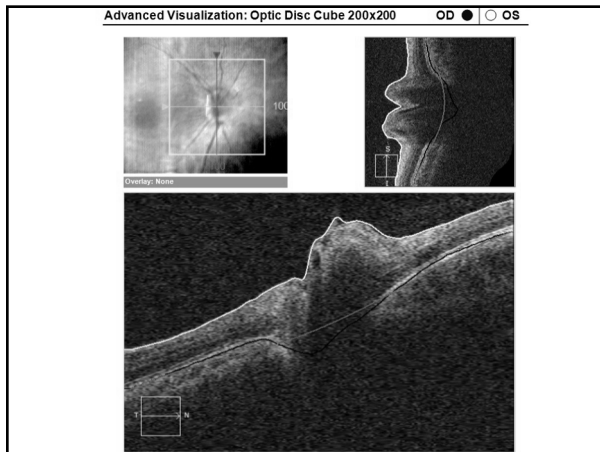
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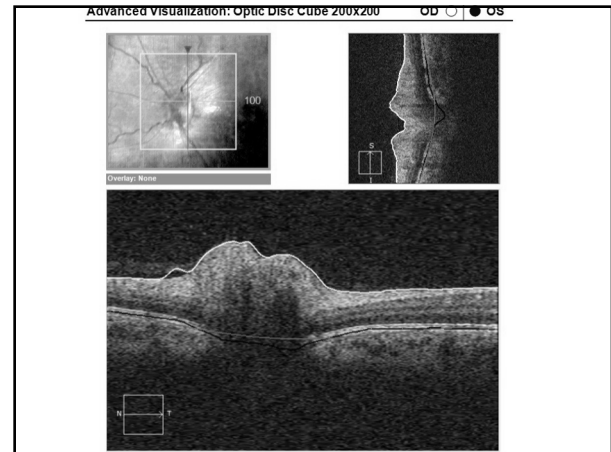
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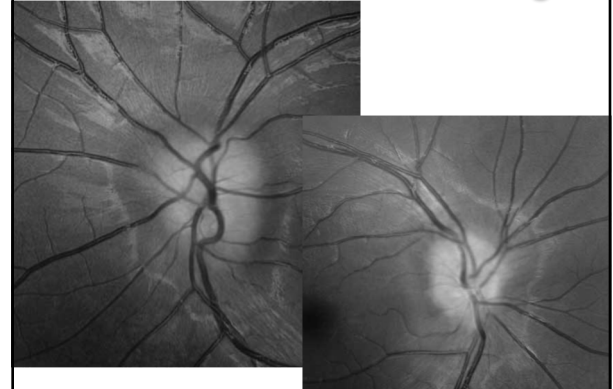
## SWELLING VS. PSEUDOSWELLING

### □ Ways to differentiate:

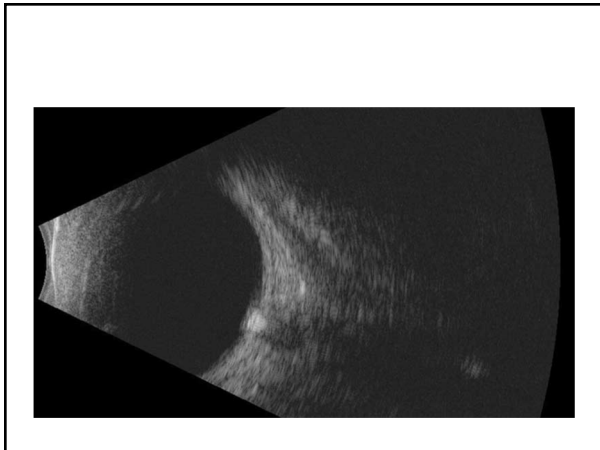
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  - Is there SVP?
2. OCT
  - rNFL thickness - normal or elevated?
  - Is there a splitting of retinal layers deep in the retina?
3. Symptoms?
4. History?
5. B-scan
  - Drusen???

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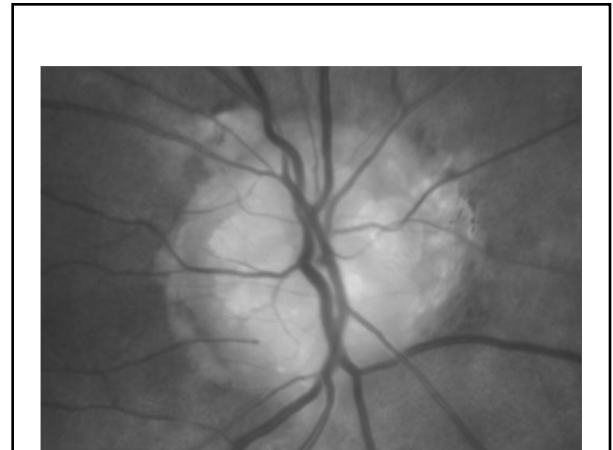
## Swelling vs. Pseudoswelling?



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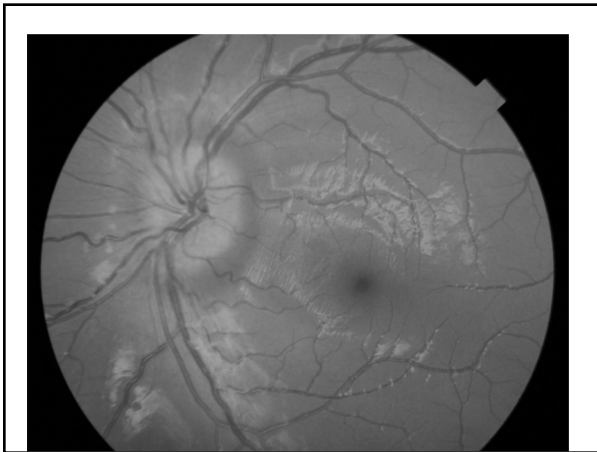
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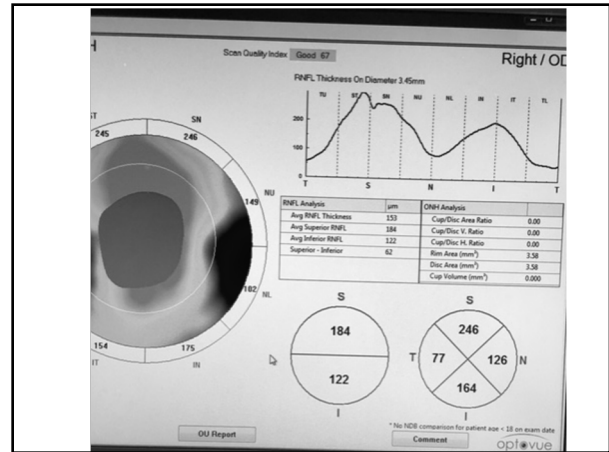
**True swelling vs.  
Psuedoswelling case????**

- ☐ 12 yoM
- ☐ "In for annual eye exam". No complains, concerns or symptoms
- ☐ Ocular Hx:
  - Longstanding alternating esotropia
  - +3.25 with mild astigmatism OU
- ☐ VA:
  - OD - 20/20
  - OS - 20/20

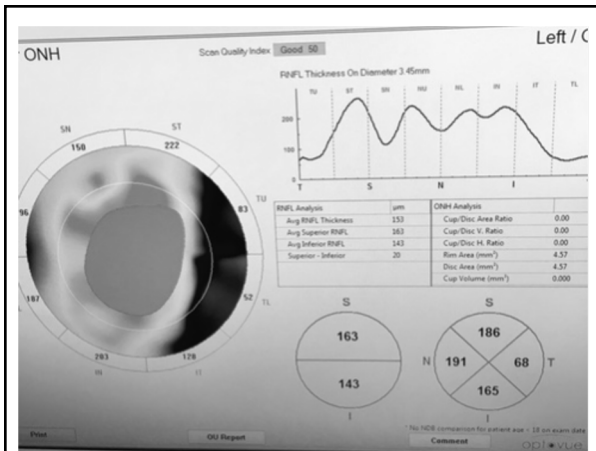
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**What do you think?**  
**Pseudoswelling vs true swelling?**

- A. Pseudoswelling
- B. True swelling

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## True swelling vs. Pseudoswelling case????

- ▣ My recommendation:
  - see a pediatric or neuro-ophthalmologist for a second opinion
  - Not overly concerned
- ▣ Pediatric ophthalmologist:
  - Diagnosis:
    - Pseudopapilledema
    - Monitor & see back in 4-6 weeks to monitor for stability

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    - Drusen???

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## Pseudotumor Cerebri

- ▣ AKA
  - Idiopathic intracranial hypertension
- ▣ Elevated intracranial pressure
  - Not caused by tumor, infection, or obstruction of the ventricular system
  - Increased production vs. decreased absorption
- ▣ Etiology:
  - Idiopathic (young, obese females)
  - Medications
    - Oral contraceptives, Tetracyclines, too much vitamin A
  - Trauma

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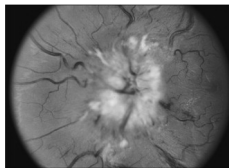
## Pseudotumor Cerebri

- ▣ Symptoms:
  - HA's (90-98%)
  - Visual disturbances (72%)
    - Transient visual obscurations (TVO's)
  - Tinnitus (20-60%)
  - N&V (30-40%)
  - Diplopia (20-30%)
  - Blurred vision
  - Abnormal color vision - rare

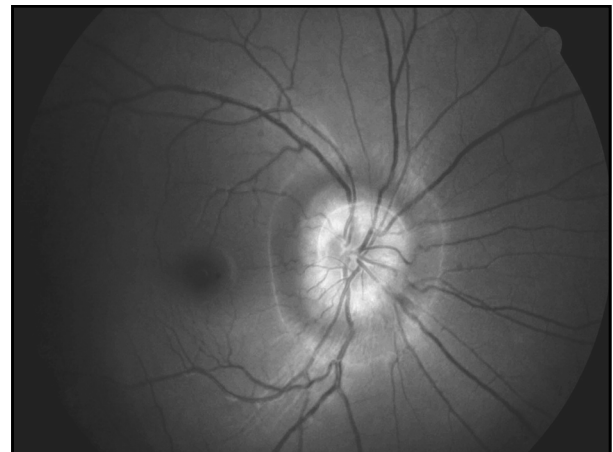
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## Pseudotumor Cerebri

- ▣ Signs
  - Papilledema – hallmark sign of PTC
    - Increased intracranial pressure -> slowing axonal transport -> accumulation of axonal contents in the NFL -> elevated ONH's
    - Bilateral disc edema
    - Blurred disc margins
    - Obscuration of blood vessels\*
    - Hyperemia of the disc
    - Venous dilation
    - Peripapillary hemorrhages & CWS
    - Paton's lines



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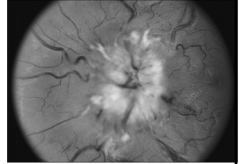


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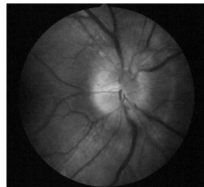


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## Pseudotumor Cerebri

### □ Other signs

- Enlarged blind spot
- 6<sup>th</sup> nerve palsy
  - Tends to subside as treatment is effective

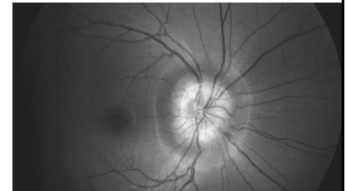


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## Pseudotumor Cerebri

### □ Differential Diagnosis:

- Intracranial tumor/mass
- Intracranial bleed
- Hydrocephalus
- Venous sinus thrombosis
- IIH

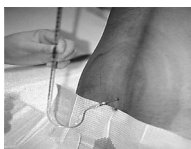


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## Pseudotumor Cerebri

### □ Diagnosis:

- Clean MRI/MRV
- Lumbar puncture
  - Elevated ICP > 250mmH<sub>2</sub>O in an obese pt
  - > 200mmH<sub>2</sub>O in a non-obese pt
  - Normal CSF composition
- No other neurological findings
  - Exception -> 6<sup>th</sup> nerve palsy
- SVP
  - Yes -> not Pseudotumor
  - No -> ????



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## Pseudotumor Cerebri

### □ Treatment:

- Weight Loss\*
  - Papilledema resolution with weight loss of 6% of total body weight
- Diamox (acetazolamide)
  - 500 mg Sequels BID-QID
  - Taper as the sx's stabilize
- Lumbar-peritoneal shunt (CSF shunting)
- Optic nerve sheath fenestration/decompression

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## Non-arteritic Ischemic Optic Neuropathy (NAION)

- Lack of perfusion to the ONH or embolic disease that affects the arteries/arterioles that supply the ONH
- Mean age of onset = 61-66 years old
- Associated risk factors:
  - HTN, atherosclerosis, DM, nocturnal hypotension, sleep apnea

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## Non-arteritic Ischemic Optic Neuropathy (NAION)

- SYMPTOMS:
  - Sudden, unilateral, painless loss of vision
  - "I woke up and I can't see out of this one eye"

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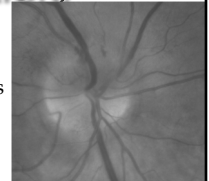
For Dr. Kote

Date	Time	Visual Acuity	Visual Field
10/5	11:30 am	114/70	153
10/6	8:30 am	117/71	86
	8:30 p	115/75	85
10/7	7:30 am	115/75	78
	10 p	120/73	80
10/8	8 am	119/77	71
	8 p	111/70	86
10/9	7 am	125/78	
10/10	8 am	123/79	84
10/11	6:50 p	126/80	82
	9 p	115/74	76
10/12	8:30 p	108/69	
10/14	7 p	115/70	
10/15	9:30 p	112/72	84
10/16	7:30 am	114/56	80
	8 p	117/71	88
10/17	8 am	118/71	87
10/18	7 am	116/73	81
	9 p	109/68	79
10/19	8:30 p	107/65	
10/21	10 am	119/70	84
10/23	7:30 p	114/71	81

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## Non-arteritic Ischemic Optic Neuropathy (NAION)

- SIGNS:
  - Diffuse or segmental disc edema
  - Peripapillary flame-shaped hemes
  - Retinal arterial attenuation
  - (+) APD
  - VF defect - often inferior altitudinal
  - What does the other eye look like?
    - Small nerve?
    - Small cup?



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## Non-arteritic Ischemic Optic Neuropathy (NAION)

- DIAGNOSIS:
  - Normal ESR & CRP
  - (-) symptoms of GCA

- DIFFERENTIAL DIAGNOSIS:
  - AAION

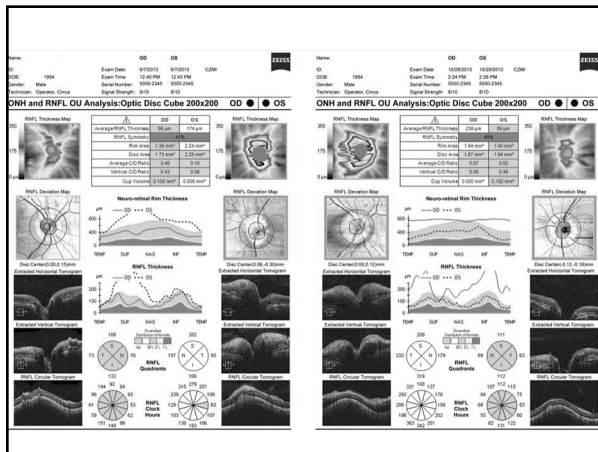
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## Non-arteritic Ischemic Optic Neuropathy (NAION)

- TREATMENT:
  - No proven effective treatment
  - Options?
    - Aspirin
    - Lower IOP??
    - Intravitreal VEGF treatment
  - Prognosis:
    - unilateral.....
    - guarded.....but it depends on many factors



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## Non-arteritic Ischemic Optic Neuropathy (NAION)

### TREATMENT:

- No proven effective treatment

### Options?

- Aspirin
- Lower IOP??
- Intraocular VEGF treatment

### Prognosis:

- unilateral.....
- guarded.....but it depends on many factors

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## Giant Cell Arteritis

- Chronic inflammatory disorder affecting the medium-large sized cranial blood vessels
- Inflammatory mediators cause:
  - proliferation, thickening, and fibrosis of vessel walls
  - > inflammatory occlusion
- Risk factors:
  - Age
  - Females
  - Scandinavian
- Accounts for 6% of ischemic optic neuropathy cases

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## Giant Cell Arteritis

### Symptoms:

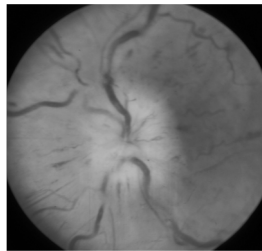
- New onset HA
- Jaw claudication
- Scalp tenderness/pain
- Flu-like sx's/weight loss
- Pain and stiffness in the shoulders, hips, torso
  - Polymyalgia Rheumatica (PMR)
- Sudden, severe, painless vision loss
  - Usually unilateral
- Diplopia

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## Giant Cell Arteritis

### Signs:

- Sudden, severe, painless vision loss
- (+) APD
- Pale, swollen optic disc
  - Flame shaped hemes
  - CWS's
- CRAO
- Ocular ischemic syndrome
- EOM problems



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## Giant Cell Arteritis

### Diagnosis:

- Clinical symptoms
- Prominent temporal artery
- Lack of temporal artery pulsation
- CBC with differential & platelets
- ESR males =  $\text{age}/2$  females =  $(\text{age}+10)/2$
- CRP
- Platelets
- Temporal artery biopsy



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## Giant Cell Arteritis

- ▣ Treatment:
  - Refer
  - IV and/or oral steroids
    - IV 250 mg i.v. q6h (1g/ day) for 3 days and/or
    - Oral 1-2mg/kg/day
  - Baby aspirin
- ▣ Prognosis:
  - Extremely poor

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## Optic Neuritis

- ▣ Patient is typically < 45 years old
- ▣ Females > males
- ▣ **SYMPTOMS:**
  - Acute vision loss – most often unilateral
  - Eye pain in/behind the eye (80-90%)
    - worsens with eye movements

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## Optic Neuritis

- ▣ **SIGNS:**
  - Visible ONH swelling (33%)
  - (+) APD
  - Color vision abnormalities
    - red cap test
  - Brightness reduction
    - brightness comparison test
  - Visual field defect – often central
- ONH pallor – 4-12 weeks after onset of symptoms

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## Optic Neuritis

- ▣ **DIAGNOSIS:**
  - MRI with gadolinium

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## Optic Neuritis

- ▣ **TREATMENT:**
  - MRI results? Already diagnosed with MS?
  - ONTT (Optic Neuritis Treatment Trial)
    - No oral steroids
    - IV methylprednisolone (1g/day) X 3 days
      - oral steroids (1mg/kg/day) X 10-14 days
      - Taper oral steroids over 4-7 days

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## Optic Neuritis

- ▣ **TREATMENT:**
  - MRI results? Already diagnosed with MS?
  - Controlled High-Risk Subjects Avonex MS Prevention Study (CHAMPS)
    - IV methylprednisolone (1g/day) X 3 days
    - Avonex (interferon beta-1a)

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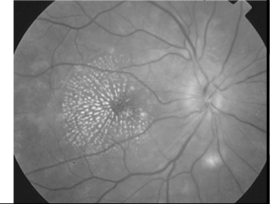
## Neuroretinitis

- ▣ Unilateral vision loss in the presence of an optic neuritis and macular star
- ▣ Etiology:
  - Idiopathic (25%)
  - Cat-scratch disease (60%)
    - *Bartonella henselae*
  - Syphilis, Lyme disease, Sarcoid, Toxo, TB
- ▣ Affects all ages, 10-40 year olds most affected
- ▣ Symptoms:
  - Painless, usually unilateral visual loss
    - Starts gradual
    - Becomes more severe after about 1 week
  - Prior viral-like illness (50%)

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## Neuroretinitis

- ▣ Signs:
  - Usually unilateral:
    - Papillitis with peripapillary and macular edema
    - Macular star develops as the disc edema resolves
    - Other inflammatory signs (cell & flare, vitreous cells)
  - Parinaud's oculoglandular syndrome



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## Neuroretinitis

- ▣ Diagnosis:
  - Clinical picture
  - History of cat scratch/bite/lick
  - Cat-scratch serology ELISA – very sensitive and specific
  - FTA-ABS, VDRL, Lyme titer, Toxo titer, ACE, ANA
- ▣ Treatment
  - Usually self limiting condition in immunocompetent individuals
  - Azithromycin 500 mg p.o. for 1 day, 250 mg/day X 4 days
  - Doxycycline 100 mg p.o. BID
  - Bactrim

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