

SWOLLEN OPTIC NERVES: NOW WHAT?

Nate Lighthizer, O.D.

1

Disclosures

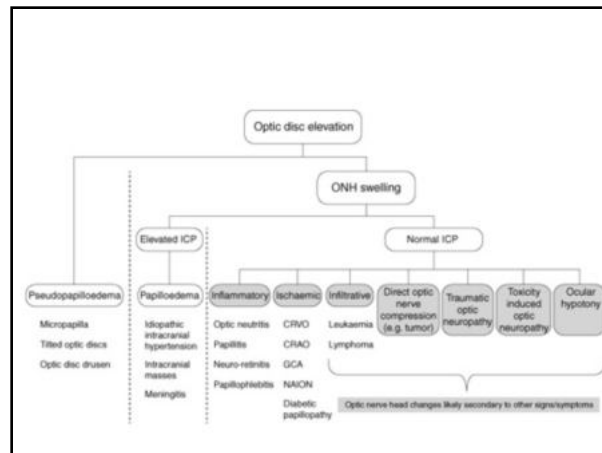
- ▣ Aerie Pharmaceuticals
- ▣ Biotissue
- ▣ Diopsys
- ▣ Ellex
- ▣ EyePromise
- ▣ Ivantis
- ▣ Lumenis
- ▣ Maculogix
- ▣ Nidek
- ▣ Nova Ocular
- ▣ Novartis
- ▣ Optovue
- ▣ Quantel
- ▣ Reichert
- ▣ RevolutionEHR
- ▣ Sight Sciences
- ▣ Shire
- ▣ Sun Pharma

2

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.

3



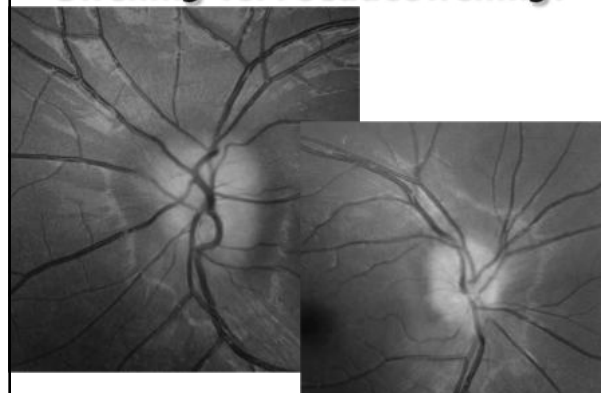
4

SWELLING VS. PSEUDOSWELLING

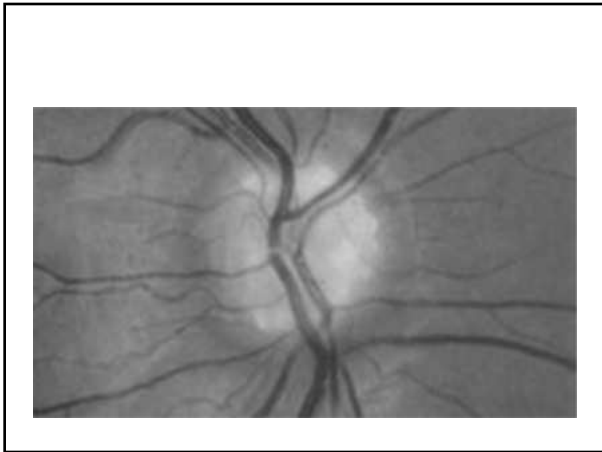
- ▣ Ways to differentiate:
 1. Direct viewing of the ONH
 - ▣ Are the vessels blurred as they cross the disc margin?
 - ▣ Is there SVP?

5

Swelling vs. Pseudoswelling?



6



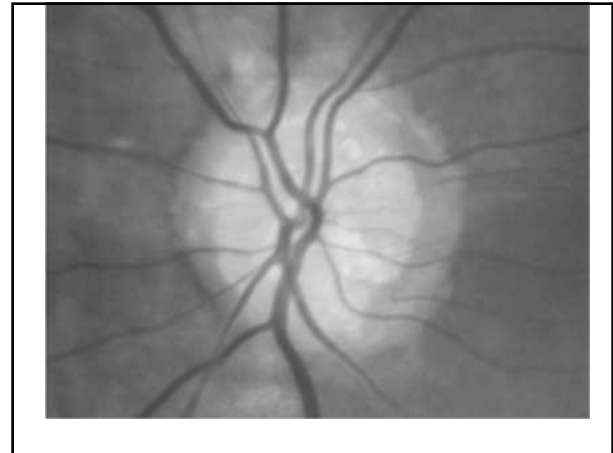
7



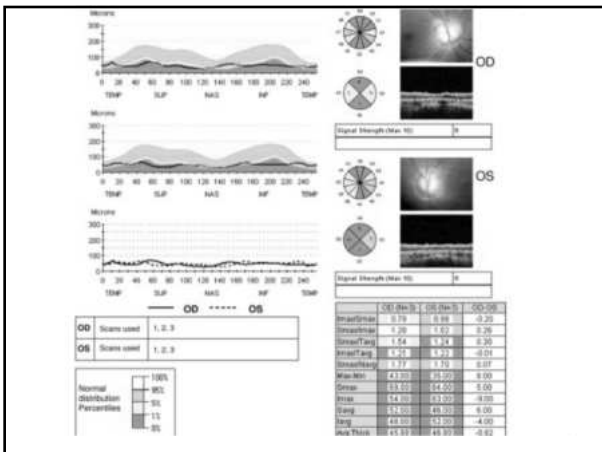
8



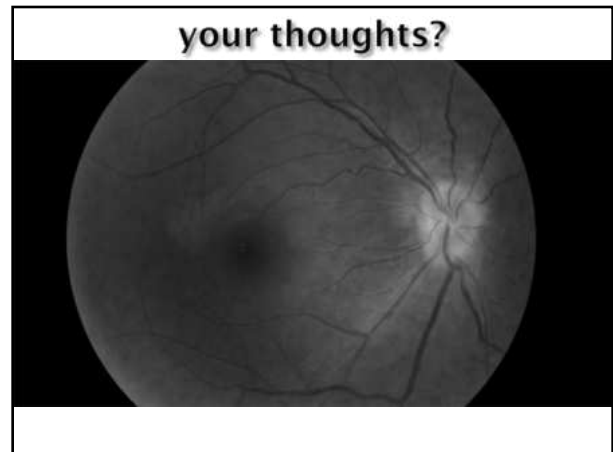
9



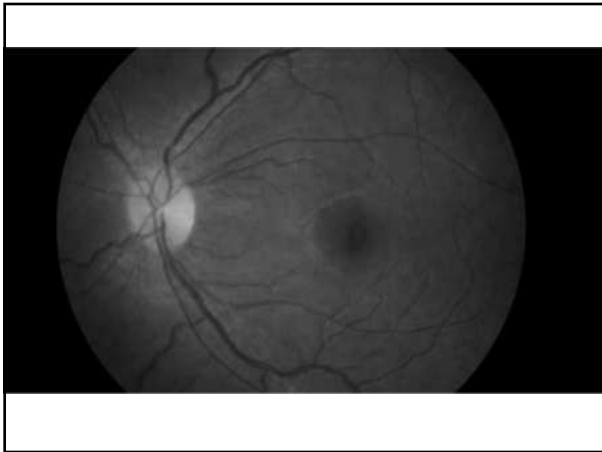
10



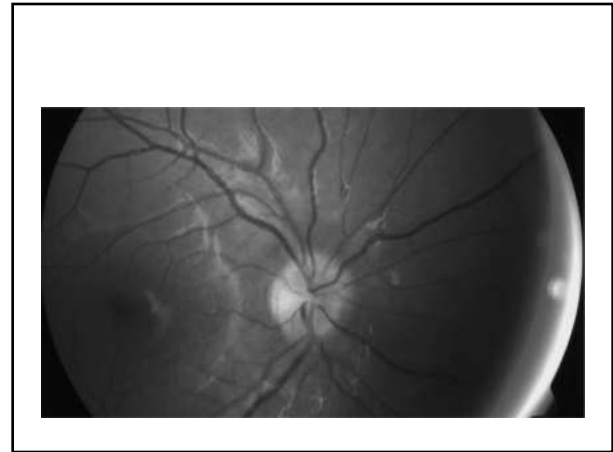
11



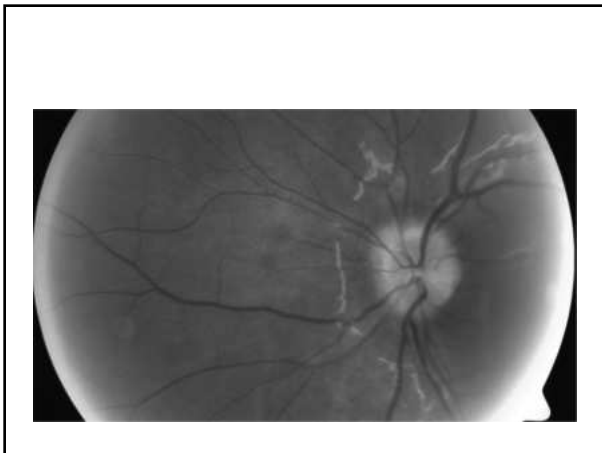
13



14



15



16

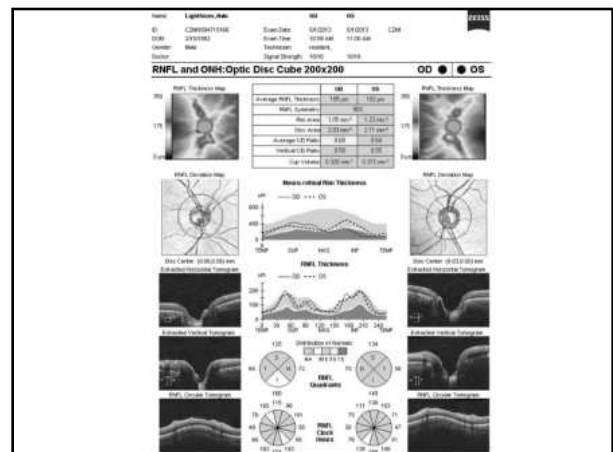


17

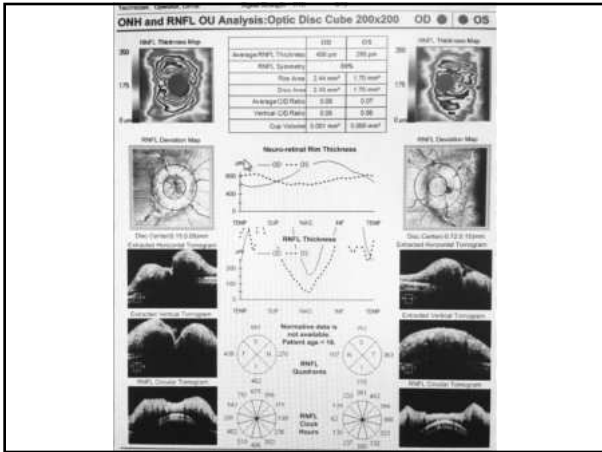
SWELLING VS. PSEUDOSWELLING

- Ways to differentiate:
 1. Direct viewing of the ONH
 - Are the vessels blurred as they cross the disc margin?
 - Is there SVP?
 2. OCT
 - rNFL thickness - normal or elevated or thin?
 - Is there a splitting of retinal layers deep in the retina?

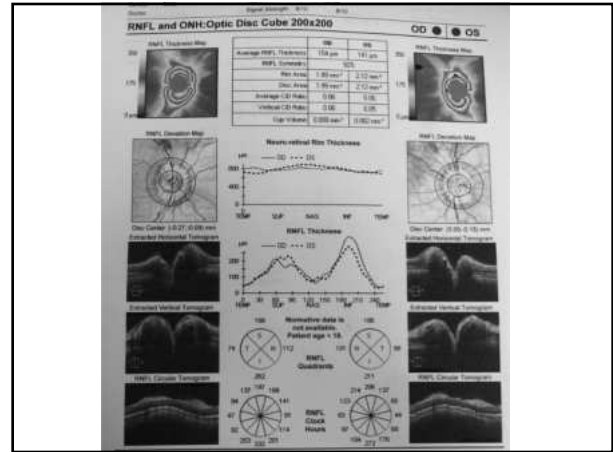
18



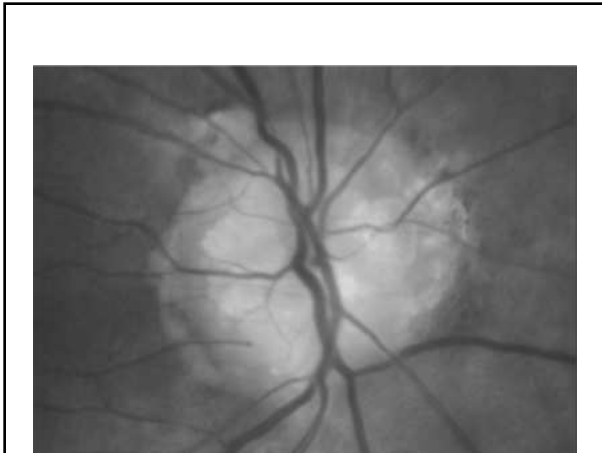
19



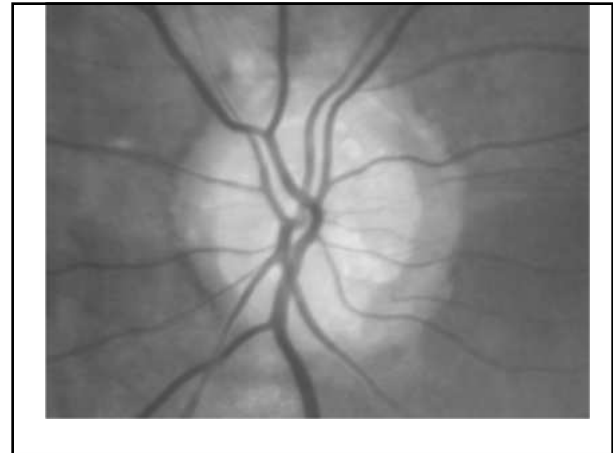
20



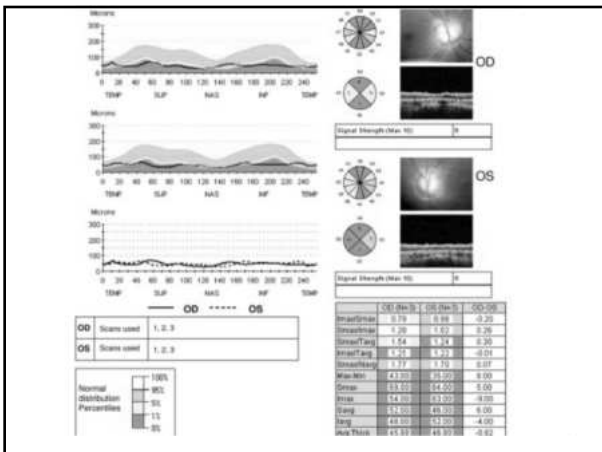
21



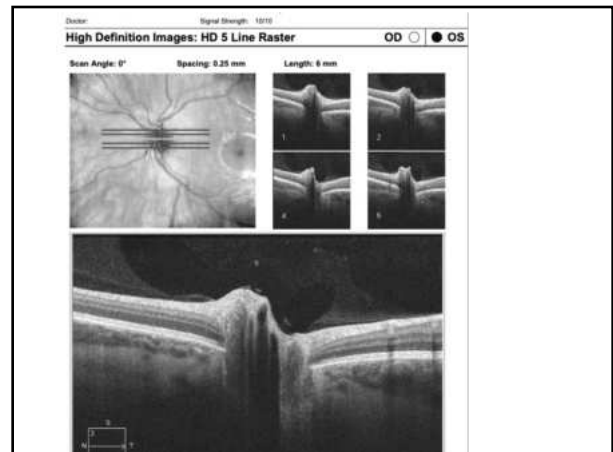
22



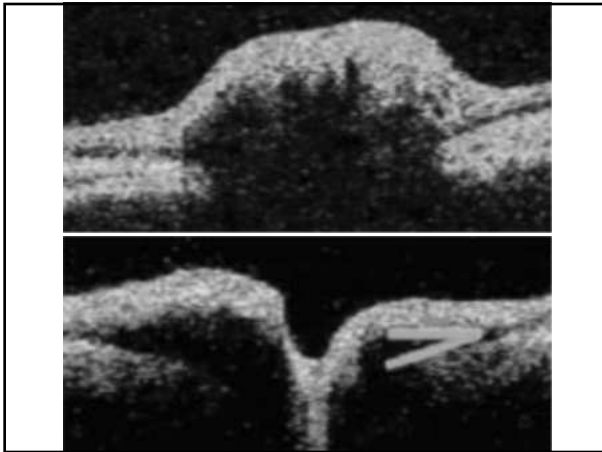
23



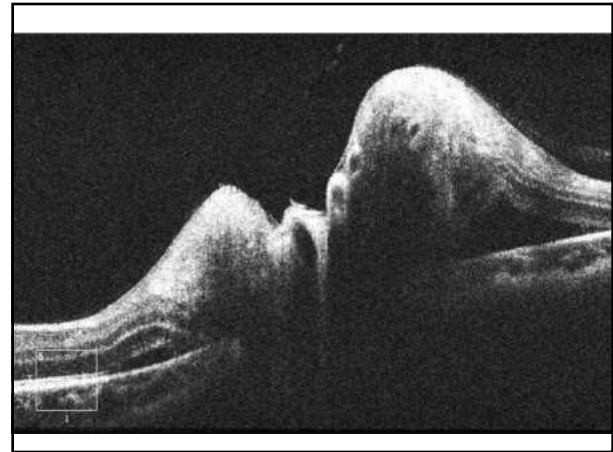
24



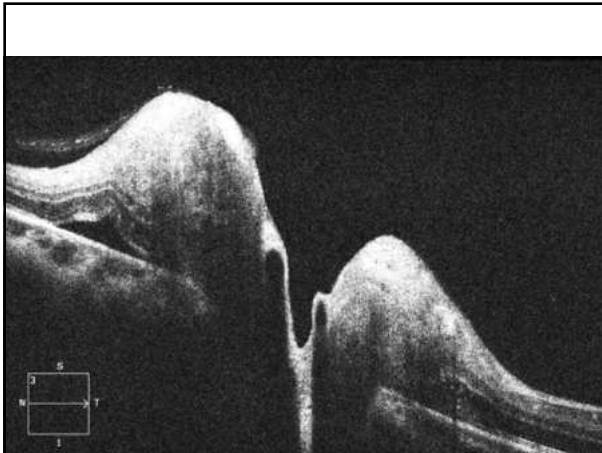
25



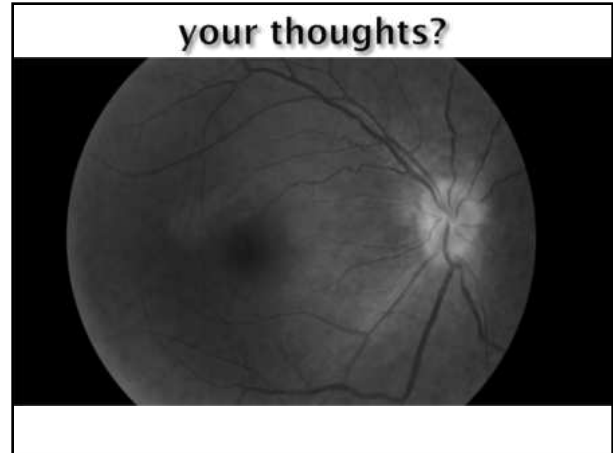
26



27



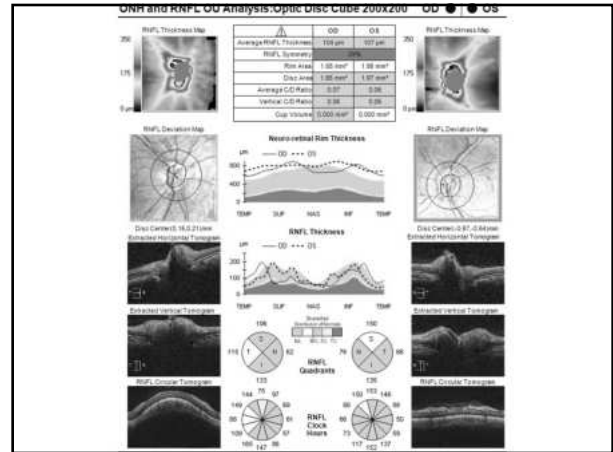
28



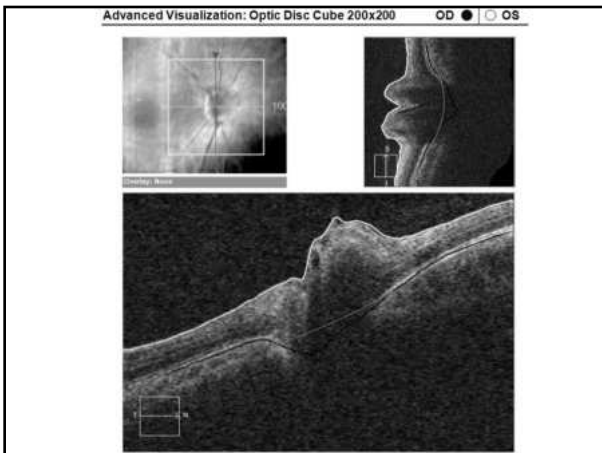
29



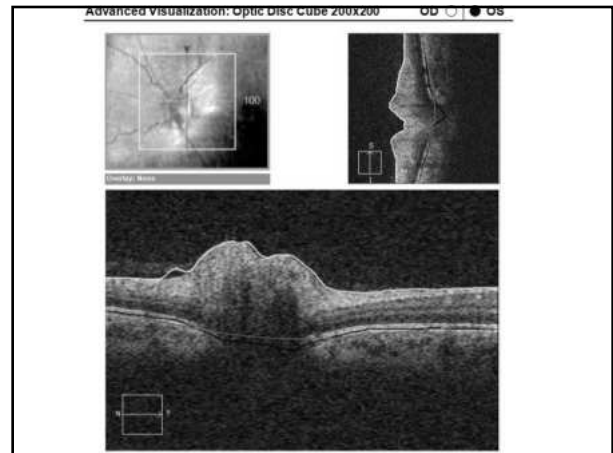
30



31



32



33

SWELLING VS. PSEUDOSWELLING

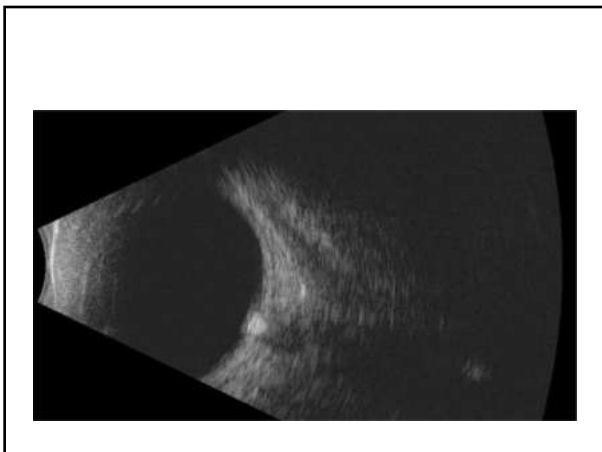
□ Ways to differentiate:

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

34

Swelling vs. Pseudoswelling?

35



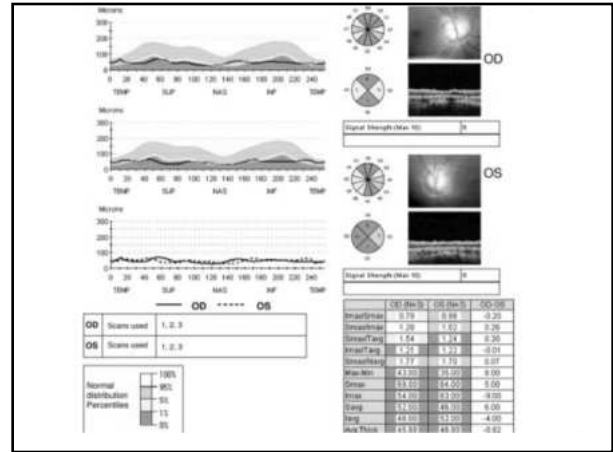
36



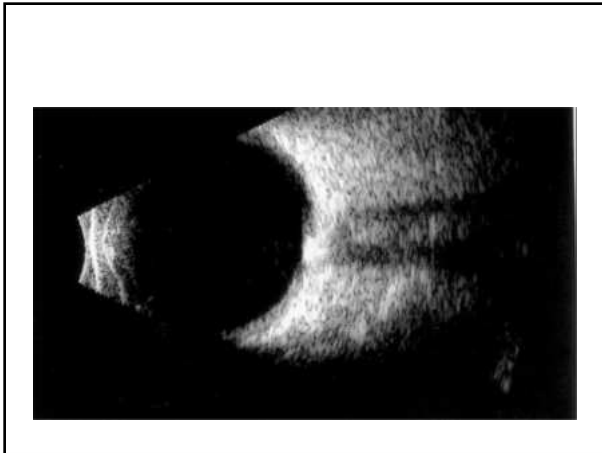
37



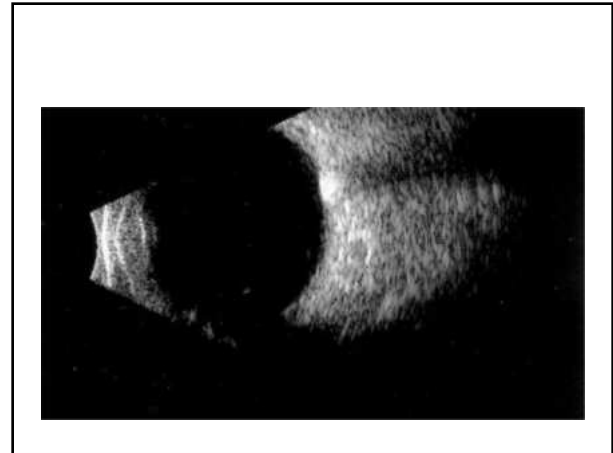
38



39



40



41

True swelling vs. Pseudoswelling 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

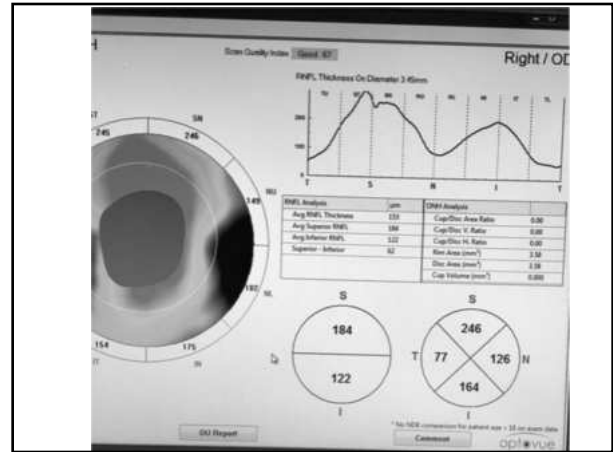
42



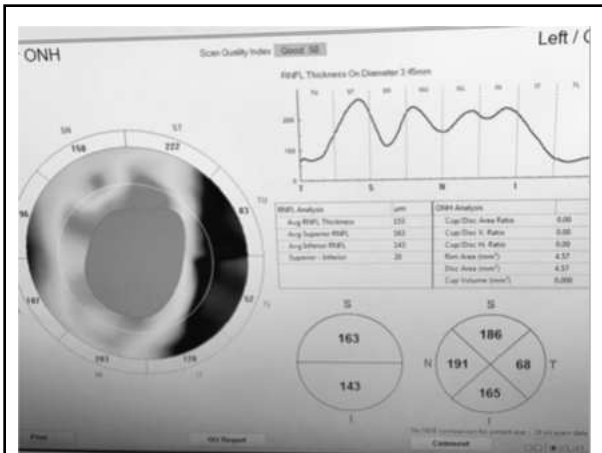
43



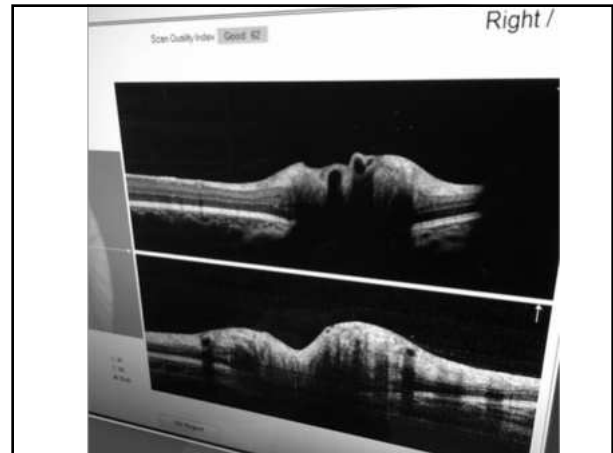
44



45



46



47



48

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

A. Pseudoswelling
B. True swelling

49

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

50

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?
 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???

51

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

52

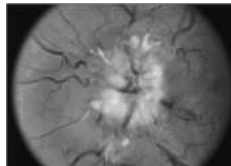
Pseudotumor Cerebri

- ▣ 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

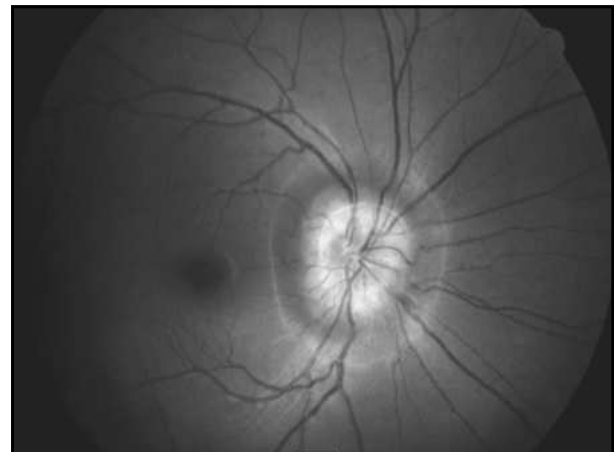
53

Pseudotumor Cerebri

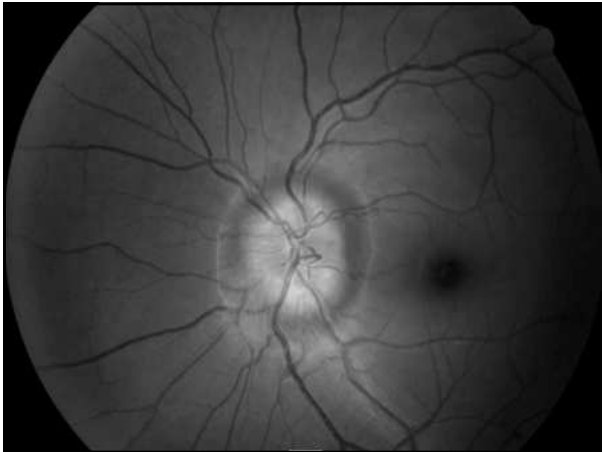
- ▣ Signs
 - Papilledema - hallmark sign of PTC
 - Increased intracranial pressure -> slowing axonal transport -> accumulation of axonal contents in the NFL -> elevated ONH's
 - Bilateral disc edema
 - Blurred disc margins
 - Obscuration of blood vessels*
 - Hyperemia of the disc
 - Venous dilation
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



54



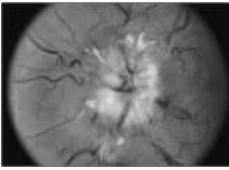
55



56

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



57

Pseudotumor Cerebri

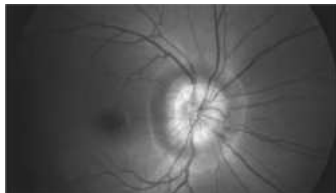
- ▣ Other signs
 - Enlarged blind spot
 - 6th nerve palsy
 - Tends to subside as treatment is effective



58

Pseudotumor Cerebri



- ▣ Differential Diagnosis:
 - Intracranial tumor/mass
 - Intracranial bleed
 - Hydrocephalus
 - Venous sinus thrombosis
 - IIH



59

Pseudotumor Cerebri

- ▣ Diagnosis:
 - Clean MRI/MRV
 - Lumbar puncture
 - Elevated ICP > 250mmH₂O in an obese pt
 - > 200mmH₂O in a non-obese pt
 - Normal CSF composition
 - No other neurological findings
 - Exception -> 6th nerve palsy
 - SVP
 - Yes -> not Pseudotumor
 - No -> ?????

60

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

61

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

62

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"

63

Date	Time	BP	Pulse
11/20	11:30am	114/70	103
11/20	5:30pm	117/71	86
11/21	8:30am	115/75	85
11/21	1:30pm	115/75	78
11/21	10:00pm	120/73	80
11/22	6am	117/77	71
11/22	8pm	117/70	80
11/22	7pm	125/78	84
11/22	8pm	124/79	84
11/22	6:50pm	124/80	82
11/22	9pm	124/77	84
11/23	6:30am	102/69	84
11/23	7:00am	115/70	80
11/23	9:00am	112/72	84
11/23	7:30am	114/56	80
11/23	8am	117/71	88
11/23	7am	118/71	87
11/23	7am	116/73	81
11/23	9pm	101/68	77
11/23	8:30pm	101/65	77
11/24	10am	114/70	84
11/24	7:30pm	114/71	81

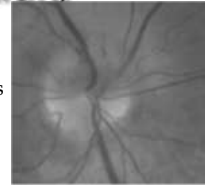
64

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?



65

Non-arteritic Ischemic Optic Neuropathy (NAION)

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

- ❑ **DIFFERENTIAL DIAGNOSIS:**
 - AAION

66

Non-arteritic Ischemic Optic Neuropathy (NAION)

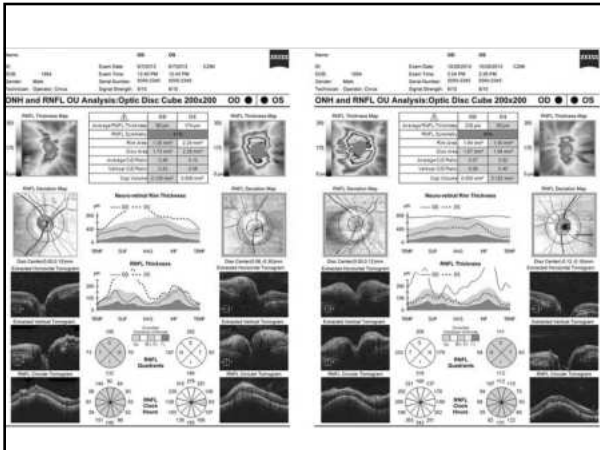
- ❑ **TREATMENT:**
 - No proven effective treatment

 - Options?
 - Aspirin
 - Lower IOP??
 - Intraocular VEGF treatment

- ❑ **Prognosis:**
 - unilateral.....
 - guarded.....but it depends on many factors



67



68

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

69

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

70

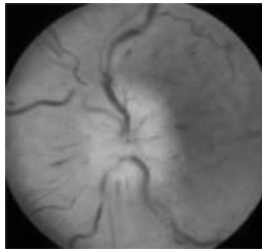
Giant Cell Arteritis

- 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

71

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



72

Giant Cell Arteritis

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



73

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

74

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

75

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

76

Optic Neuritis

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

77

Optic Neuritis

- ▣ **TREATMENT:**
 - MRI results? Already diagnosed with MS?
 - ONIT (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

78

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)

79

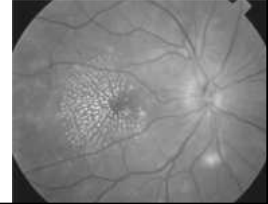
Neuroretinitis

- ▣ 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%)

80

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



81

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

82