Don't Let Swollen Optic Nerves Make You Nervous

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Financial disclosures

- No financial disclosures

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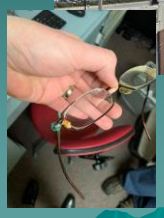
Examination Techniques

- Stereoscopic viewing essential
- VA and VF: SVP
- Pupil testing and color vision
- Brightness comparison and red cap test



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Papilledema

- Bilateral (but can be sequential with one nerve becoming swollen before the other, thus unilateral at presentation) optic nerve head swelling secondary to increased ICP
- Swollen, blurred margins with splinter hemorrhages and exudates as well as nerve fiber layer edema. Patton's folds may be seen

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Papilledema

- May be asymmetric
- VA varies but typically mild reduction only or no loss at all
- May get diplopia secondary to abducens nerve compression
- With increased ICP, can get choroidal folds only (before papilledema) at lower pressure levels

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Papilledema

- VF usually shows enlarged blind spot
- No pupillary defect. Normal color vision
- SVP absent with obliterated cup

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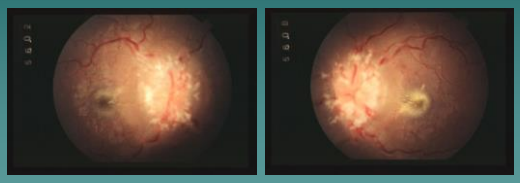
Papilledema (IIH)



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Papilledema IIH age 15

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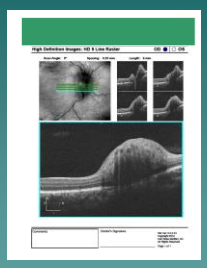
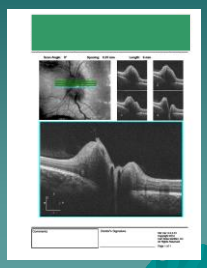
OCT





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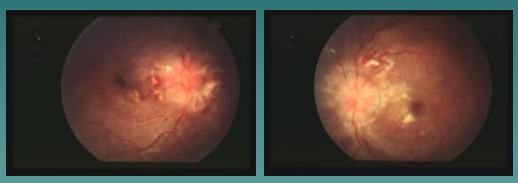
Papilledema (HTN)



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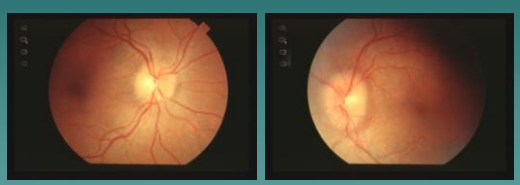
Papilledema (tumor)



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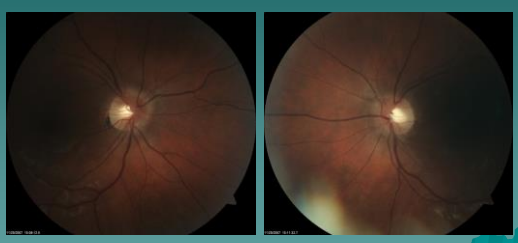
Subtle papilledema (IIH)



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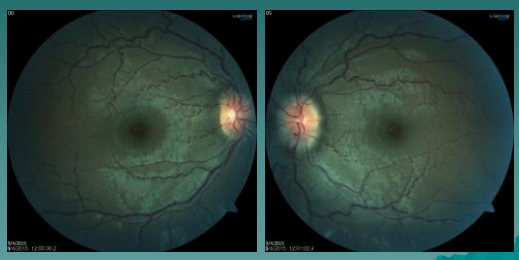
Papilledema IIH



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Papilledema IIH

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Papilledema IIH

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Papilledema with Patton's folds

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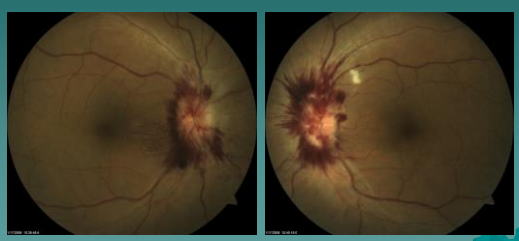
Terson's and papilledema



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Papilledema progression

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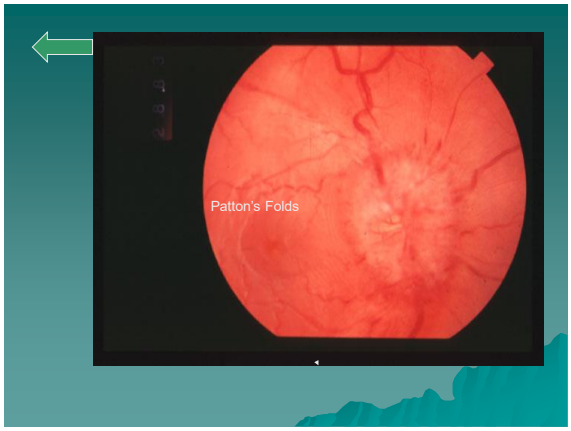

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Patton's folds: RNFL thickness 231in OD, 295 in OS

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Patton's folds: now you see them……

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Back then in 2007 you did not…

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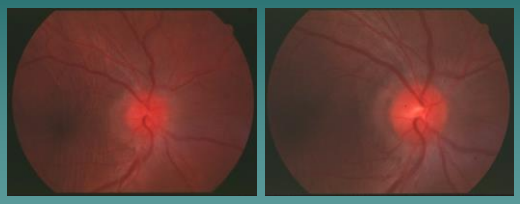
Patton's folds



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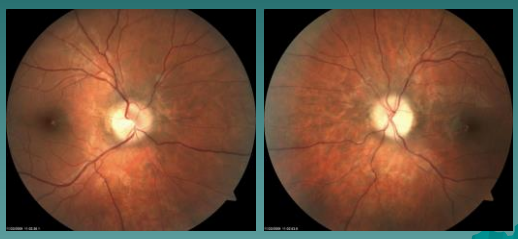
Longstanding papilledema with optic atrophy (IIH)



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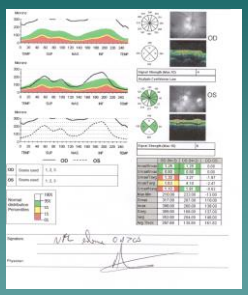
Papilledema OCT NFL



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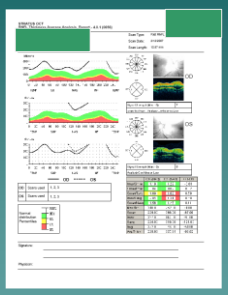
NFL edema



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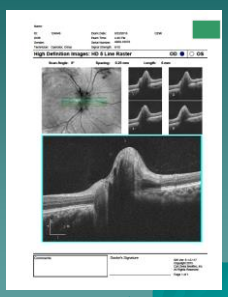
Papilledema OCT



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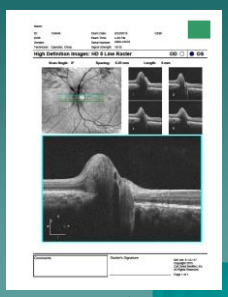
Papilledema OCT



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Increased ICP

- Variations are due to anatomical considerations
- If the channels connecting the central cavity and optic nerve sheath allow equal flow on both sides and in both directions papilledema will occur and will improve with decreased ICP

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Increased ICP

- If there is a difference in the communications then the edema will be asymmetric. Usually the result of a smaller bony canal opening on one side limiting the swelling.
- If the valves are one-way then the swelling will not improve rapidly with Tx

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Increased ICP

- An acute rise in ICP that resolves rapidly is not typically associated with papilledema. Elevation must be chronic
- Increased pressure is transmitted from the sub-arachnoid space to the optic nerve head via the nerve sheath. Venous pressure in CRV increases
- Disruption in axoplasmic flow at lamina cribosa leads to swelling

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Increased ICP

- Studies show that ONH swelling as measured by OCT can decrease (but not instantly resolve) immediately after lumbar puncture
- Measured in lateral decubitus position with OCT sideways!
- Shows that reduction of ONH compression is very rapid
- Shows that pressure in spinal column is associated with pressure at ONH

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Etiologies of Increased ICP

- Space occupying lesion ; must always be ruled out!
- Infection or anatomical abnormality
- Malignant hypertension
- IIH
- Certain medications
- ? Sleep apnea (obesity): ICP may be elevated only at night! Men especially
- Must order MRI in all cases

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Idiopathic Intracranial Hypertension (IIH)

- Older term is "pseudotumor cerebri"
- Young overweight females (F 8X M)
- 1/ 100,000 in population as a whole ; 20 / 100,000 in 20 to 44-year-old women 10% over ideal weight
- May be related to medications including TCN (minocycline especially), HRT, lithium, high dose Vitamin A supplementation, steroid withdrawal
- Emerging evidence that elevated testosterone / androgen levels may be the cause
- Sleep apnea link
- Can affect children, often overlooked
- Doubles cardiovascular risk in females

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IIH

- Symptoms of transient blur, diplopia , tinnitus (intracranial noises, not just ringing) , headaches , etc.
- ICP usually severely elevated ; normal is 50 – 200 mmH20. Over 25 cm (250 mm) considered definitively abnormal. Single measurement can be misleading : levels can vary over 24 hours
- Very rare variant of normal pressure IIH. S/S, but repeatedly normal ICP

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IIH more rare over age 50

- Less often female
- Fewer headache complaints
- More frequently discovered incidentally due to papilledema with no symptoms
- Lower opening CSF
- More likely to have concomitant medical conditions
- Less likely to use tetracycline family antibiotics

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IIH

- Diagnosis involves normal MRI / MRV and CSF studies with elevated ICP
- Watch for spinal chord tumors
- Differential: Cerebral Venous Sinus Thrombosis
- MRV

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CVST(cerebral venous sinus thrombosis)

- Young women and some men
- Often not overweight
- Can be life threatening
- Treat with blood thinners, Diamox
- Can be seen with MRI, but potentially missed if MRV not performed
- Stenosis may be secondary to IIH

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Optic atrophy post CVST induced papilledema

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IIH Management

- Refer to a neurologist
- Medical management includes Diamox , Lasix, Topamax
- Weight loss

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IIH Management

- If recalcitrant….
  - Repeated lumbar taps (ugh!)
  - Lumbo-peritoneal shunt
  - Ventricular shunt

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IIH Management

- If progressive changes in visual acuity or visual field occur , consider an optic nerve sheath decompression
- Several small fenestrations in the optic nerve sheath are created to allow room for expansion
- Performed by a neuro-ophthalmologist. Often do worse eye only because 50% get improvement in the fellow eye

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Chronic IIH induced edema leading to atrophy: S/P decompression

22 year old AA F

Light perception 10/700

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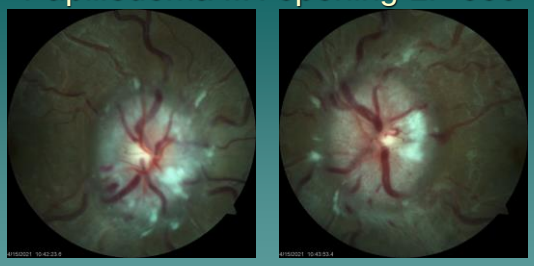
Papilledema IIH opening LP 550



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After 3 weeks on Diamox

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Side by side comparison

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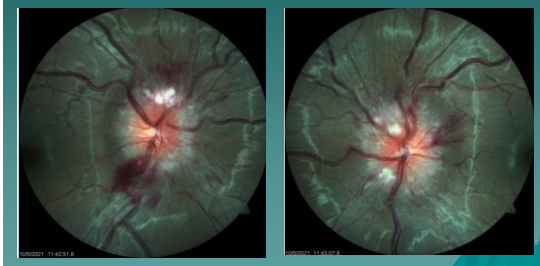
Minocycline induced elevated ICP papilledema



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Foster Kennedy Syndrome

- Swollen optic nerve on one side , advanced optic atrophy on the other
- Advanced optic atrophy prevents swelling making a bilateral problem appear to be unilateral
- Often seen in chiasmal tumors

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Compressive Optic Neuropathy

- Compression leads to axoplasmic stasis and retrograde death of nerve fibers
- Pale, choked, swollen nerve
- Rarely see hemes; + APD

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Compressive Optic Neuropathy

- Optic atrophy and severe vision loss with time
- MRI with and without contrast: neurosurgery referral
- Possibly endoscopic optic nerve decompression

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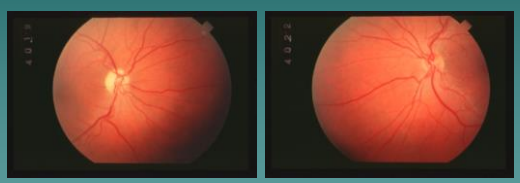
Pituitary tumor



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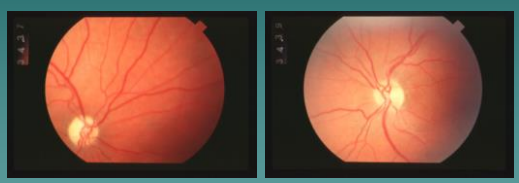
Pituitary tumor post surgery



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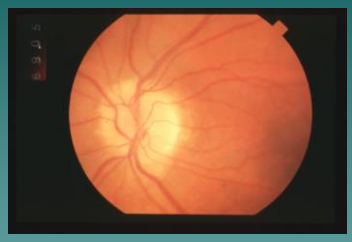
Sphenoid wing meningioma



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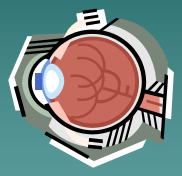
ION

- Nonarteritic
- Arteritic



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Nonarteritic ION

- Swollen , hyperemic nerve with splinter hemes and exudates
- Often sectoral
- Ischemic / hyoperfusion event caused by interruption of micro-vascular circulation, often at night.
- Highly associated with sleep apnea (75-90% in several studies)
- NAION has 5x risk of sleep apnea, 8x risk in women

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NAION

- No systemic symptoms; normal ESR / CRP
- Most common cause of ONH swelling over the age of 55 (2-10 cases per 100,000 per year)
- 45-60 year-olds most commonly (any age possible) with no sex predilection)

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Nonarteritic Etiologies

- 1) Sleep apnea! Up to 90%
- 2) Hypertension (med related?)
- 3) Idiopathic
- 4) Diabetes
- 5) Atherosclerosis
- 6) Migraine
- 7) Increased Homocysteine / Decreased vitamin B6
- 8) HIV infection

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Nonarteritic ION

- Typically seen in "disc at risk" patients with very small cups. Therefore 70 + % Caucasian
- Approximately 15% of cases will involve the fellow eye in 5 years (more common with VA < 20/200 in first eye, diabetics, and platelet polymorphisms). Repeat attacks in same eye < 5%

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NAION

- VA varies widely from normal to severe loss: 45% 20/40 or better but 33% 20/200 or worse
- VA loss progresses over 2-4 weeks
- VA improves by up to three lines at six months in 40%
- In patients under 50 years of age, there is a higher rate of bilateral involvement and more visual recovery

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Nonarteritic ION

- Often APD , color vision usually normal
- Most frequent visual field defect is inferior nasal / partial altitudinal but may get essentially any type. FDT may be more sensitive and often shows spillover of loss in to "non-affected" hemifield
- After swelling resolves the nerve is pale but often not cupped-cupping may occur, however
- Why does area of swelling not always match VF defect?

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NAION 2 weeks after onset of symptoms

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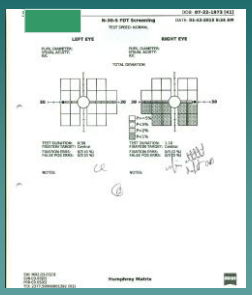
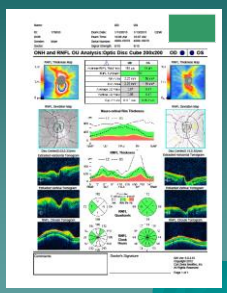
NAION





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Nonarteritic ION Treatment

- No treatment other than managing the underlying cause has proven to be consistently effective
- Blood thinners may debatably protect the fellow eye but will not alter the course of recovery.
- Order CBC , ESR and CRP , lipid profile , hemoglobin A1C. Check BP
- Check for sleep apnea!

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Steroids?:

- SS Hayreh: 2008 study utilizing oral steroids….
- If VA 20/70 or worse,oral prednisone resulted in VA improvement (3 or more lines) in 70% of treated patients, only 40% of untreated
- Beginning dose of 80mg for 2 weeks with slow taper.
- Not commonly offered, no definitive evidence of benefit

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Incipient ION

- Early swelling, but no impact yet on VA or VF
- May resolve without loss of vision or VF, may become full blown NAION with loss
- Can only impact by treating underlying condition

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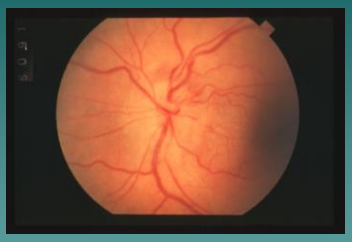
NAION



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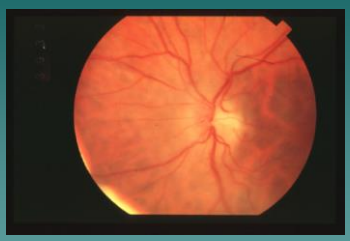
NAION



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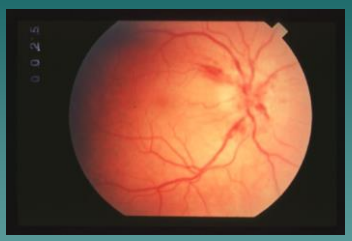
NAION



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NAION

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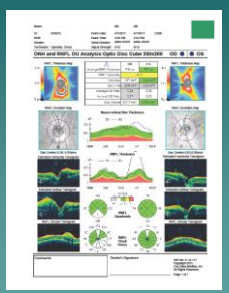
NAION secondary to OSA



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NAION OD secondary to HIV

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Another HIV induced optic neuropathy

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Old NAION OD

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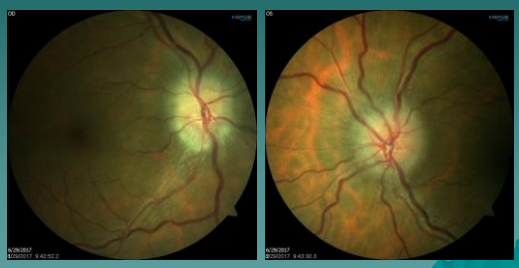
Bilateral NAION secondary to OSA (40% blood oxygen level)



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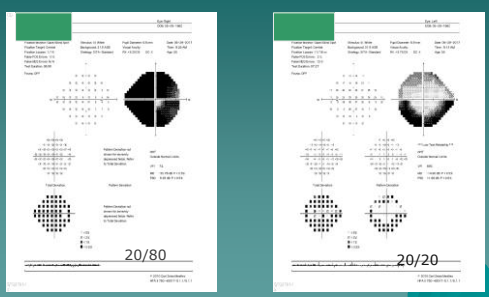
Accompanying VF



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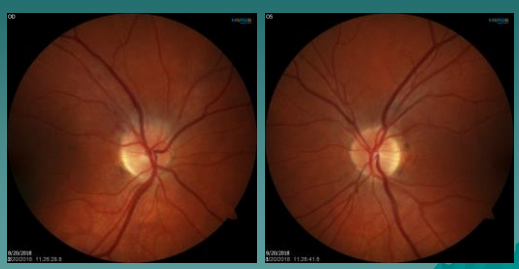
NAION OD and fellow eye



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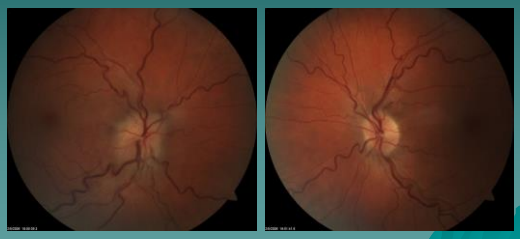
NAION OD: The Beginning

NAION **Fellow Eye**



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Optic atrophy / incipient ION

Optic Atrophy **Incipient NAION**

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NAION OS

Optic Atrophy **NAION OS**

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Optic atrophy OU

Post NAION **Post NAION**

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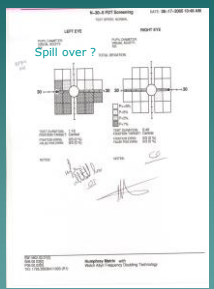
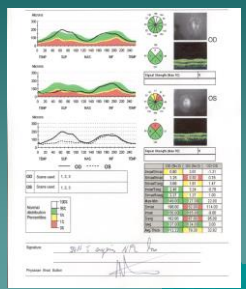
ION OS with matching VF / NFL loss





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Arteritic ION

- Pale disc swelling with splinter hemorrhages
- Average age 76 (80% over 70), F>M 3:1
- Increased ESR,C-Reactive protein, platelet
- ESR normal in about 25%!
- VA 20/200 or worse in 60% of cases
- Traditional thinking from past studies of a high predilection for Caucasians, but a large 2019 study showed only a slight predilection for Caucasians over African Americans.

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Arteritic ION

- Sudden, painless loss of vision with APD
- Altitudinal VF loss most common, others possible
- Symptoms of GCA but about 1/3 are symptom free
- Very high five-year mortality rate

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Giant Cell Arteritis

- GCA is a disease of unknown etiology (emerging evidence that zoster may be involved, but other studies have refuted this) affecting the large and medium arteries including the temporal , ophthalmic , and posterior ciliary arteries
- Symptoms include HA , scalp tenderness , jaw claudication , malaise , fever , and fatigue

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GCA

- May also see CWS, CRAO, and amaurosis fugax
- 20% of cases with ocular involvement are CRAO, 80% ION
- Obtain stat Westergren ESR, CRP, CBC with platelets

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Giant Cell Testing

- Normal ESR is age/2 for men and age +10/2 for women
- C-Reactive protein testing is not specific for GCA but it is nearly 100% sensitive so very useful test
- Temporal artery biopsy when indicated

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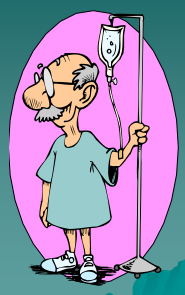
Giant Cell Arteritis

- 25% of untreated patients develop AION
- 2/3 will develop in the second eye within weeks if not treated, up to 50% within one week
- Rheumatology referral



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Giant Cell Treatment

- IV hydrocortisone followed by long term oral prednisone. Maintenance dose of 10mg daily for years. Follow ESR, other markers
- Average cumulative steroid dose over course of treatment…….. …….over 5000 mg of prednisone!

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Temporal (Giant Cell) Arteritis

- Newly FDA approved treatment
- Subcutaneous Tocilizumab (Actemra)
- Used with steroids (not in place of): makes steroid dose much lower
- Immunosuppressant
- Risk of infections, no live vaccines
- Delivered IV
- Also used with RA and other forms of arthritis

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Amiodarone induced optic neuropathy

- Mimics NAION in nerve appearance but bilateral instead of unilateral
- Afflicts 2% of patients taking it
- Slow, insidious onset of visual loss
- Slow, complete recovery over many months after medication is discontinued (very long half-life)

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Viagra / Cialis / Levitra and NAION

- 553 cases officially reported to the FDA by the end of 2014. 443 were Viagra
- ? Under reported
- These medications also occasionally used for pulmonary HTN
- Visual loss most often noted upon awakening the morning after use
- Is the association real or coincidence?
- Likely the "straw that broke the camel's back" in those with risk factors. But……..

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ED drugs and NAION

- Very interestingly, has been reported in a 7 month-old infant, 28 year old, and 33 year old, presumably all taking them for pulmonary HTN
- At those young ages, not as likely to have other NAION risk factors
- Also, 2 reported cases of PION with Sildenafil, one in a 39 YO female with pulmonary HTN

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Viagra / Cialis

- What is the proposed mechanism? Nitrous oxide release actually dilates vessels….but drops blood pressure.
- Do ION patients have faulty autoregulation?
- Ask all males with NAION about ED drug use. D/C if using to protect fellow eye.

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Optic Neuritis

- Unilateral (usually) swollen nerve. Often retrobulbar (2/3) with no visible abnormality. Hemorrhages uncommon
- Diffuse visual field loss or enlarged blind spot. Subtle defects often present in the fellow eye
- Centro-cecal defect with Goldmann perimetry
- About 5% in US bilateral, but 30% in Asia

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Optic Neurits

- Younger patients (20-40 peak), F > M: more common in Caucasians
- APD, wide range of VA loss, decreased color vision; pain on eye movement

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Optic Neuritis

- Often associated with post viral syndromes or demyelinating diseases such as MS (initial symptom in 20% of cases-usually retrobulbar)
- VA recovers over weeks to months to near baseline level but often seems dim or washed out to the patient
- Get MRI in most cases
- May represent form fruste MS
- Several cases reported linked with use of TNF (tumor necrosis factor). Used for RA & JA: etanercept, infliximab, etc.

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Optic neuritis associated with MS

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Optic Neuritis Treatment Trial

- 457 patients in three treatment groups 1) oral steroids (1mg / kg / day X 14 days), 2)IV steroids (250mg Q 6h X 3 days) followed by orals (as above for 11 days), 3) placebo
- Orals followed by short taper of 20 mg on day 15 and 10 mg on days 16 and 18
- Hospitalized while on IV methylprednisone
- Traditional treatment of oral steroids proved to be the least effective of the three! Actually increased recurrence rate

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ONTT

- IV followed by orals hastens VA recovery by about 2 weeks but does not improve end result
- Delays the onset of MS symptoms up to 2-3 years: no benefit at 5 years

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ONTT 15-year F /U

- 294 patients seen 15 years out
- 15-year risk of developing MS was 50% (6% had known MS entering the trial)
- 72% if lesions on original MRI, 25% without
- VA 20/20 or better in 72%
- Factors indicating a lesser chance of developing MS include: 1) male gender, 2) optic disc swelling, 3) peripapillary hemorrhages and exudates, 4) no pain on eye movement, 5) NLP vision

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MS lesions

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Optic Nerve Head Drusen

- Increased prevalence in small nerves with small cups. Therefore, more common in whites than in AA. Higher incidence in patients with RP (10%)
- Compression of axons leads to stasis of axoplasmic flow and hyaline is excreted then calcifies over time, leading to the formation of drusen
- Nerve appears elevated but no splinter hemes or exudates and the margins are distinct.
- Abnormal vessel branching

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Optic Nerve Head Drusen

- Not always visible! Buried early in life but become visible with time. Creation of more drusen push some forward to the surface of the nerve
- Can cause decreased vision and variable visual field defects. More loss with visible drusen
- Common and under diagnosed

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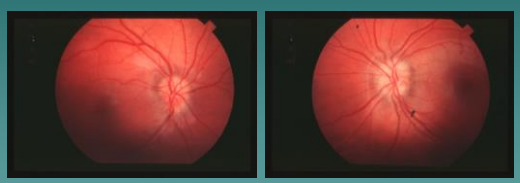
Optic Nerve Drusen

- SVP/EVP not affected: APD and color vision loss rare but possible
- Change with time
- Use B-scan or OCT to detect buried drusen
- Also seen with CAT scan, MRI, IVFA, and FAF

106

ONH Drusen

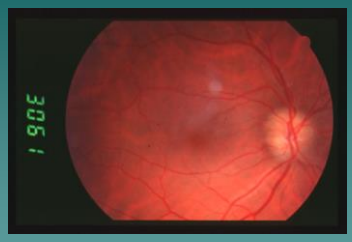


107

ONH Drusen

108

ONH Drusen



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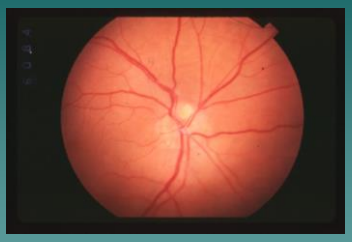
ONH Drusen



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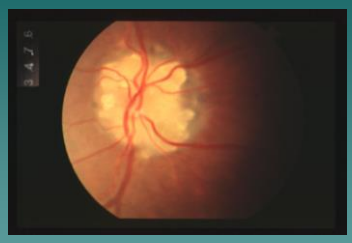
ONH Drusen



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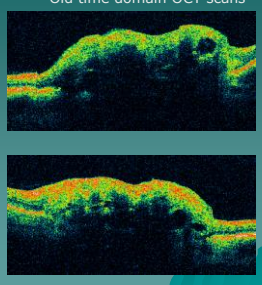
ONH drusen

Old time domain OCT scans



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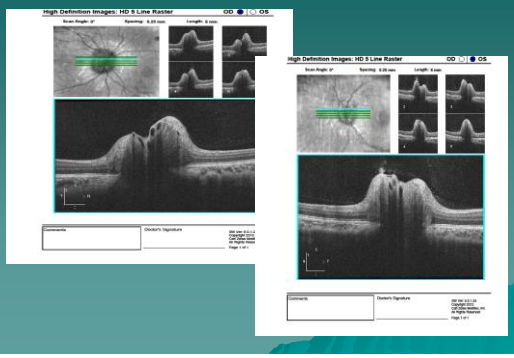
ONH DRUSEN SD-OCT



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ONH drusen detection with OCT

- Optic Disc Drusen Consortium Consensus……
- Always use EDI
- Blood vessels are more solid, cast a shadow, and can show as figure 8
- Drusen always prelaminar
- Drusen always hyporeflective
- Drusen often have a hyper-reflective border, especially superiorly

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ONH drusen detection with OCT

- Drusen can conglomerate, and these areas can have some internal reflectivity from borders
- The old concept of a hypo-reflective fluid wedge at the edge of the nerve in true papilledema DOES NOT APPLY with SD-OCT. Was a time domain OCT artifact.

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Peripapillary Hyper-reflective Ovoid Mass-like structures (PHOMS)

- "Fomms"
- Seen best with EDI
- Only seen with OCT, nothing else
- Circular innertube like structure around the disc above Bruch's membrane
- Herniated optic nerve fibers
- Seen in any condition that leads to nerve swelling or congestion
- ION, papilledema, disc drusen

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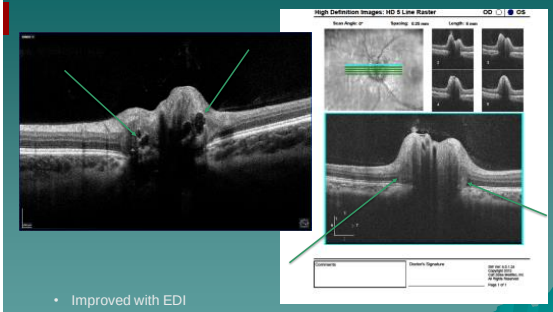
ONH DRUSEN



- Improved with EDI

PHOMS

SCHOOL OF OPTOMETRY

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FAF ONH Drusen

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FAF ONH Drusen

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NFL loss with ONH drusen

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Longstanding ONH drusen OU & new cat scratch disease OS

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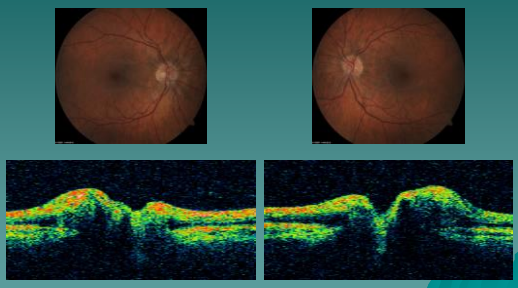
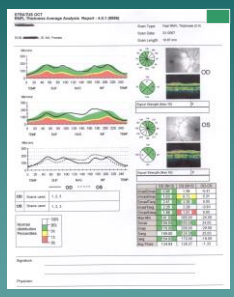
IIH with ONHD and papilledema



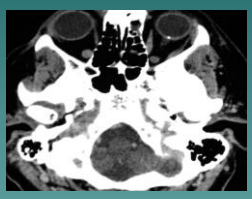
122

IIH with ONHD and papilledema



123

ONH drusen MRI



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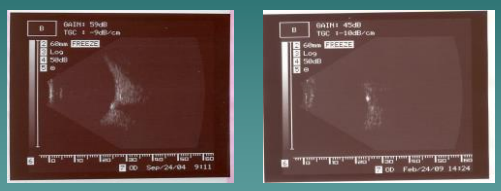
ONH drusen B-scan



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Papillophlebitis (optic disc vasculitis)

- An inflammatory variant of CRVO striking otherwise healthy, young adults (f 2x m)
- Disc edema out of proportion with retinal hemorrhaging
- Usually mild VA reduction to around the 20/30 level but can be worse

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Papillophlebitis

- Vague prodrome of scintillating, colored lights with visual disturbances
- Enlarged blind spot on the visual field
- Dilated and tortuous veins
- Condition is self limiting over the course of several months and a complete recovery is the norm
- Separate entity? Systemic work-up? Are we looking for the wrong things? Antiphospholipid antibody syndrome (APA)

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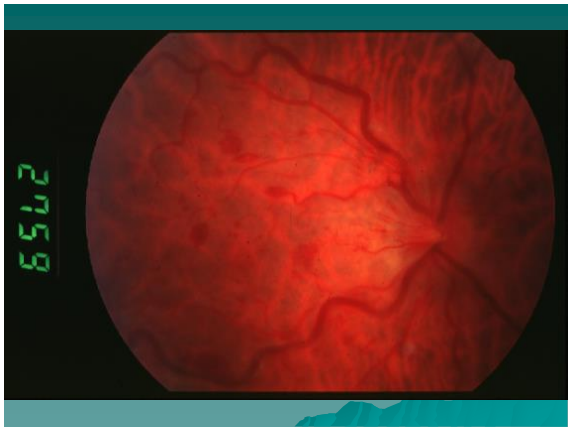

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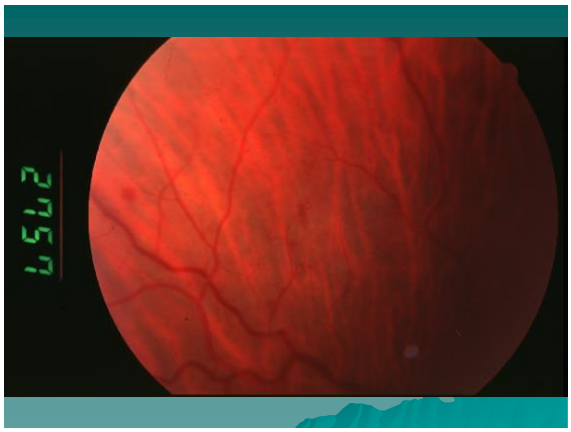

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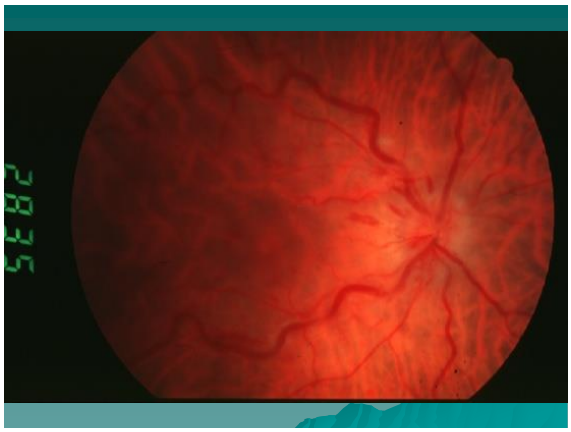

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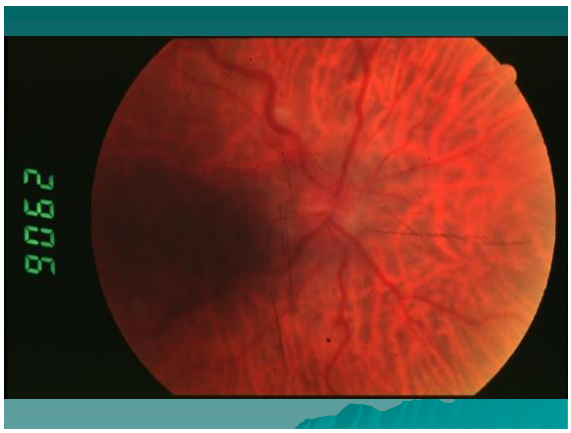

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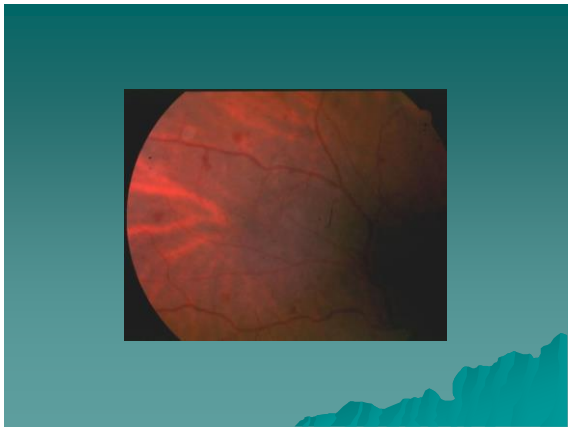

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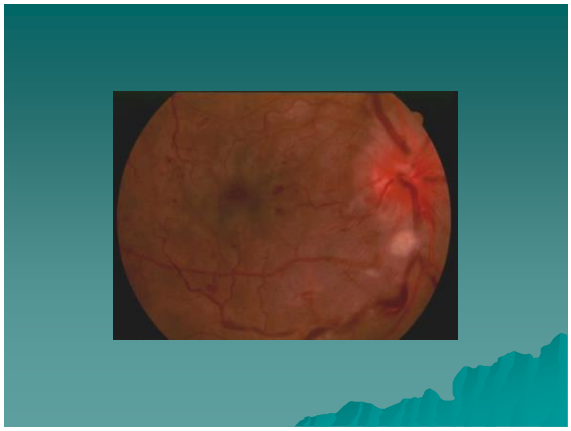

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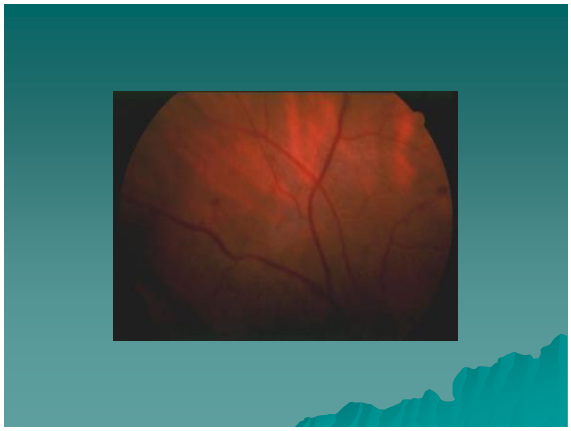

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Papillophlebitis

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Papillophlebitis

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Diabetic Papillitis

- More common in young, type I diabetics but can also be seen in adults with type II
- Diffuse ONH edema that may be unilateral or bilateral
- Relatively mild vision loss
- No altitudinal defect on VF; various patterns of mild loss seen

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Diabetic Papillitis

- Slow resolution of ONH edema but complete or nearly complete recovery of vision is the norm
- Like NAION, more prominent in nerves with small cups
- Is it real……..or just a variant of NAION?

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Diabetic Papillitis

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Grave's disease

- Remember No SPECS…….
- Soft tissue edema
- Proptosis
- EOM involvement
- Corneal involvement from exposure
- Sight threatening complications
- Hyper (most common), hypo, or euthyroid

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Grave's disease

- The sight threatening complication is optic neuropathy from compression at the muscle cone
- Requires oral steroids and / or orbital decompression. Also Tepezza infusion (possible issues with hearing loss / elevated blood sugar)
- Type II Grave's patients
- 75-80% of Grave's patients are smokers!

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The end!

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