Assessing the Glaucomatous Optic Nerve

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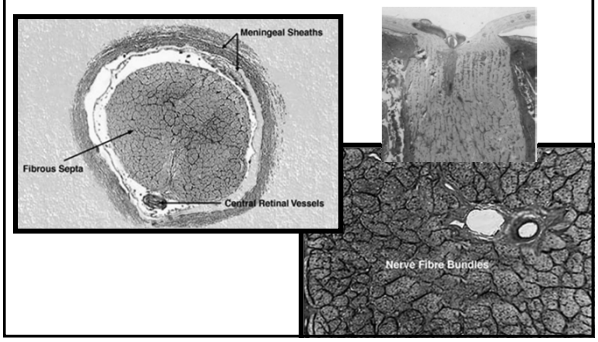
Disclosures

No disclosures related to the content of this presentation.

Learning Objectives:

1. **Learn to accurately and efficiently assess the optic nerve appearance in patients with glaucoma.**
2. **Learn a standardized stepwise approach of clinical examination.**
3. **Recognize characteristic glaucomatous changes in the optic nerve.**
4. **Be able to compare optic nerve photos to their OCT images.**
5. **Be able to compare optic nerve photos to their Visual Field results.**
6. **To review techniques for the determination of disease progression based upon optic nerve photos.**

Glaucomatous Optic Atrophy



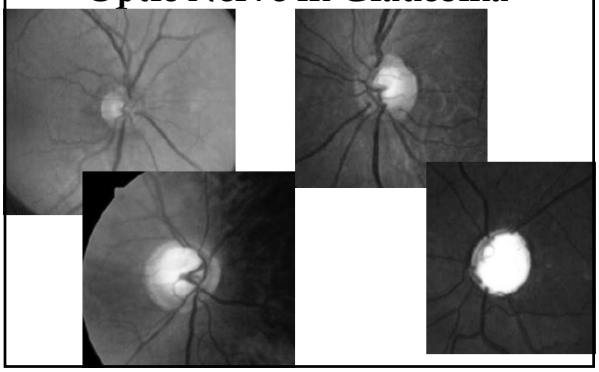
What we know so far:

- **Development of glaucoma is related to multiple risk factors**
  - **IOP is very important**
  - **Ocular Perfusion Pressure is very Important**
- **Glaucoma is an optic nerve disease that has RGC/Axon loss that leads to permanent vision loss**

Causes of Glaucomatous Damage

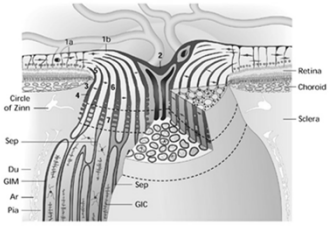
- **Elevated IOP**
- **Ischemia, Poor blood flow perfusion to ONH**
- **Compression of GCA**
- **Anatomic weakening of LC**
- **Faulty connective tissue support in LC**
- **<u>Neurotoxic Processes</u>**
  - **Release of excitotoxins**
  - **Blockage of neurotrophic growth factors**
  - **Programmed cell death, "Apoptosis"**

Optic Nerve in Glaucoma

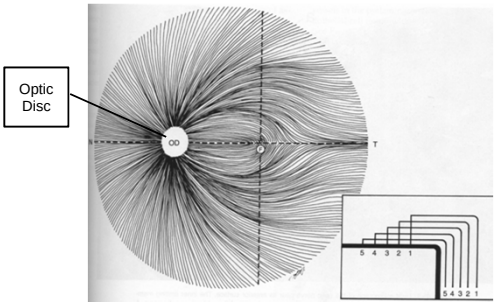


Optic Nerve Head

- In cross section:
- Surface Retinal NFL
- Prelaminar region
- Laminar cribrosa region
- Blood supply
- Retrolaminar region

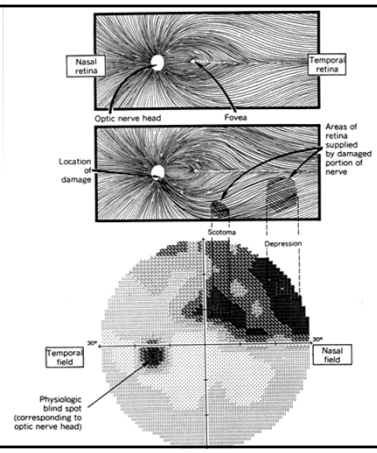


RNFL axon organization: OS

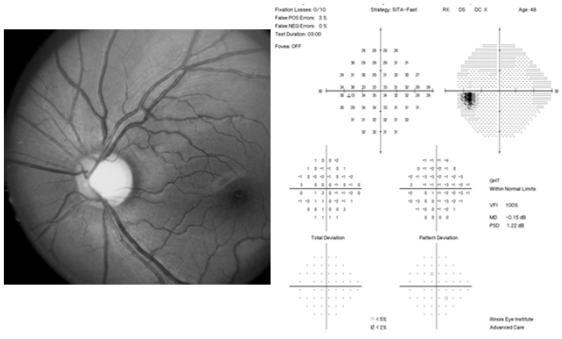


Nerve Damage and VF Loss

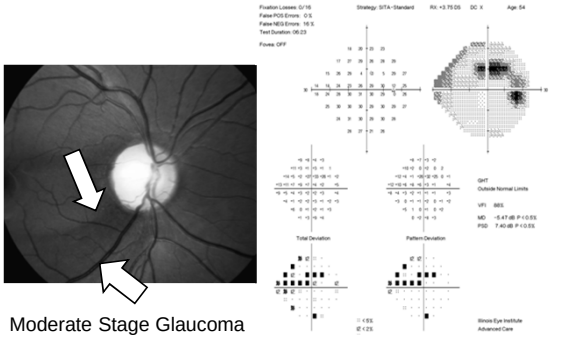
- Damage to the inferior temporal optic nerve head leads to superior nasal loss in the visual field due to the inverse projection on the retina
- Example: classical glaucoma damage



Normal: Optic Nerve, RNFL, VF

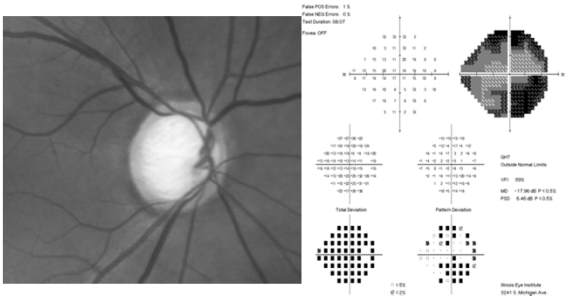


Nerve Fiber Layer Drop Out: focal



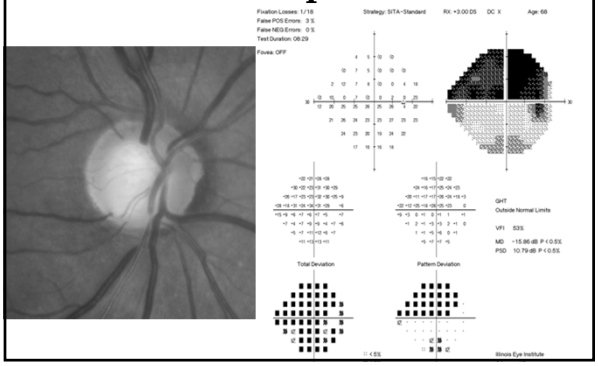
Moderate Stage Glaucoma

Diffuse Loss of RNFL



Advanced Stage Glaucoma

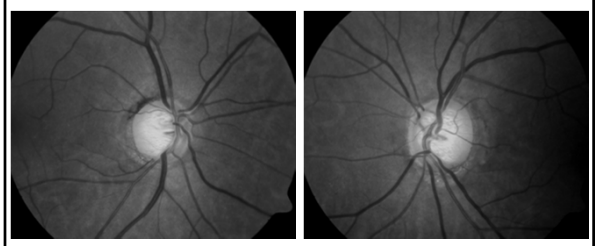
Inferior Temporal Notch



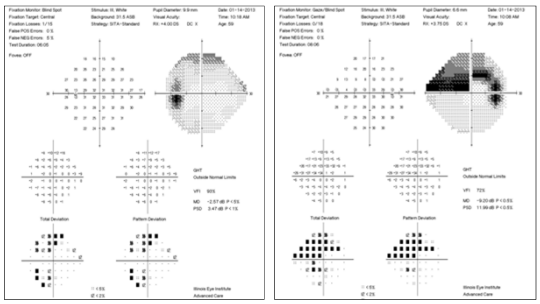
CASE SR

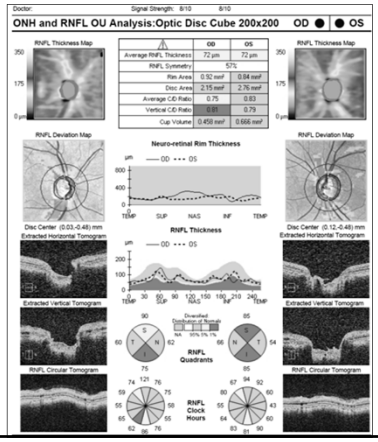
65 yo, diabtic
GAT = 19 OD, 18 OS
CCT = 505

Disc Photos

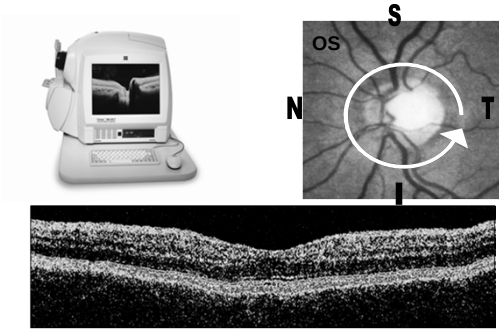


VFs #2





Cirrus OCT: RNFL Analysis



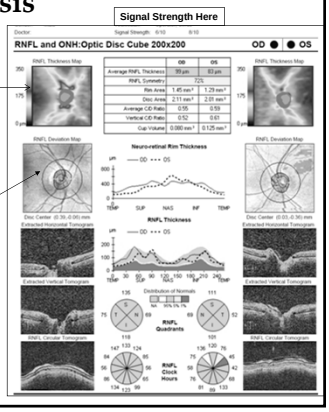
RNFL OU Analysis

RNFL THICKNESS MAP shows the patterns and thickness of the nerve fiber layer within the full 6mm x 6mm area

RNFL thickness and comparison to normative data is shown in circle, quadrants and clock hour display

RNFL DEVIATION MAP, overlaid on the OCT fundus image, illustrates precisely where RNFL thickness deviates from the normal range. Data points that are not within normal limits are indicated in red and yellow.

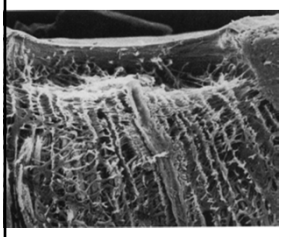
Quadrant and Sector MAPs in middle of page

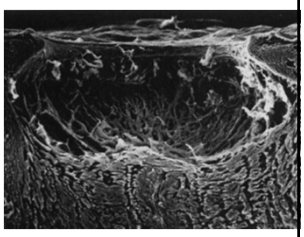


Lamina Cribosa:

- **Composed of ten lamellae (sheets) of connective tissue. They are fennestrated and organized to allow for the passage of nerve fiber bundles carrying the ganglion cell axons.**
- **There are 200 to 600 pores, varying in size, with the larger ones at the superior and inferior poles. These may provide LESS support than the smaller fennestrations in the nasal and temporal regions and allow greater damage to the RGC axons.**

Deepening of the Lamina

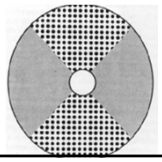


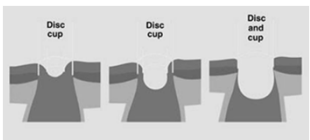


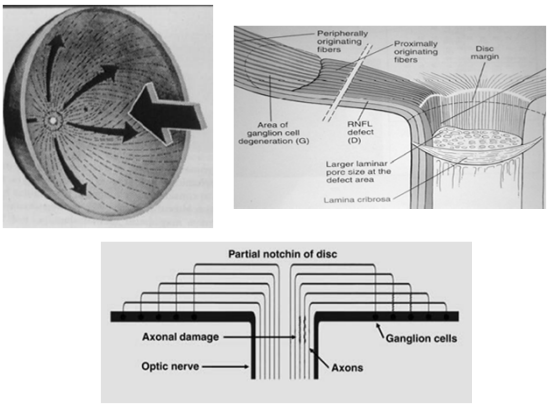
- **Seen in POAG**

Pathogenesis of ONH Excavation and "cupping"

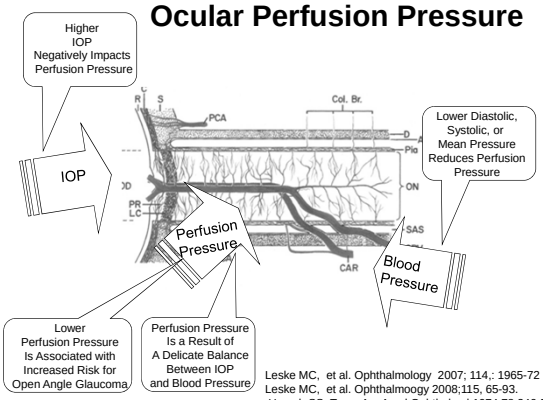
- **Damage occurs at the lamina cribosa**
- **Primarily involving bundles in the superior and inferior poles.**
- **Loss of axonal tissue results in "excavation" of the optic nerve.**







Ocular Perfusion Pressure



Leske MC, et al. Ophthalmology 2007; 114,: 1965-72
Leske MC, et al. Ophthalmoogy 2008;115, 65-93.
Hayreh SS. Trans Am Acad Ophthalmol 1974;78:240-54

Low OPP = Higher Risk

- May be due to:
  - –High IOP
  - –Low BP
    - Physiological
    - Over treatment of systemic HTN
    - Nocturnal Hypotension

CSF Pressure

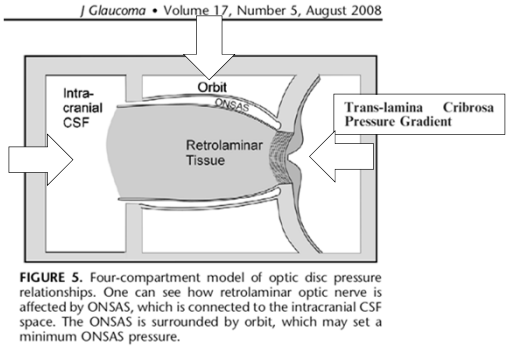
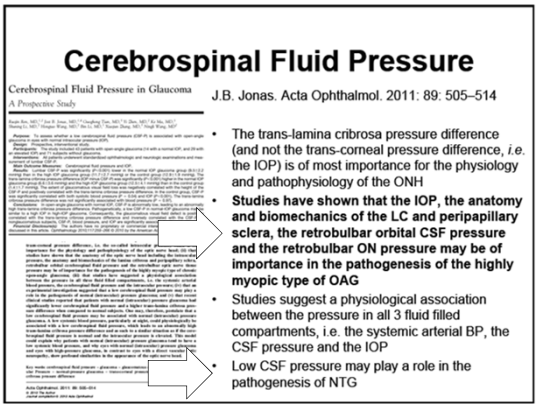


FIGURE 5. Four-compartment model of optic disc pressure relationships. One can see how retrolaminar optic nerve is affected by ONSAS, which is connected to the intracranial CSF space. The ONSAS is surrounded by orbit, which may set a minimum ONSAS pressure.

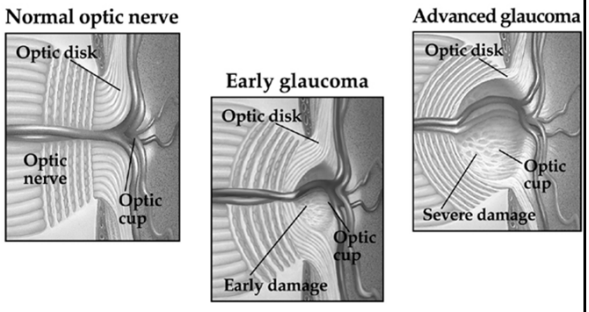
Cerebrospinal Fluid Pressure



J.B. Jonas. Acta Ophthalmol. 2011: 89: 505–514

- The trans-lamina cribrosa pressure difference (and not the trans-corneal pressure difference, *i.e.* the IOP) is of most importance for the physiology and pathophysiology of the ONH
- **Studies have shown that the IOP, the anatomy and biomechanics of the LC and peripapillary sclera, the retrobulbar orbital CSF pressure and the retrobulbar ON pressure may be of importance in the pathogenesis of the highly myopic type of OAG**
- Studies suggest a physiological association between the pressure in all 3 fluid filled compartments, i.e. the systemic arterial BP, the CSF pressure and the IOP
- Low CSF pressure may play a role in the pathogenesis of NTG

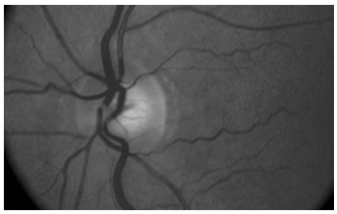
Progression of Excavation



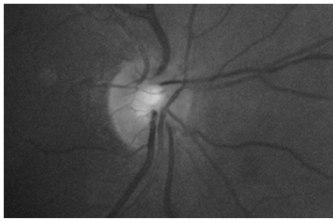
Glaucomatous Disc Features

Some of terms you will get to know :

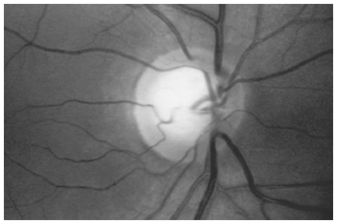
- ***increased*** **(meaning it changed) cup-to-disc ratio or significant cup asymmetry;**
- **decreased or documented change in neuroretinal rim area;**
- ***notch*** **of the neuroretinal rim;**
- ***saucerization*** **of neuroretinal rim;**
- **flame-shaped *disc hemorrhage*;**
- **nerve fiber layer loss;**
- **peripapillary atrophy.**



- **Normal cupping with healthy neuroretinal rim**

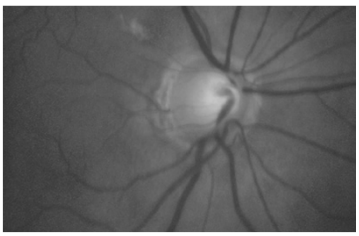


- **A small cup in a smaller optic nerve, again with healthy neuroretinal rim.**



- **A big nerve with a large cup in a patient without glaucoma.**

- **The horizontal cup is larger than the vertical cup in this normal rim.**
  - » **larger vertical cupping is more typical of glaucomatous damage**

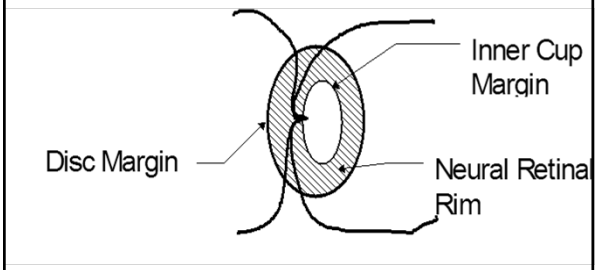


- **An optic nerve of a patient without glaucoma with a distinct peripapillary crescent,**
  - » **probably representing a misalignment of the retinal choroidal layers.**

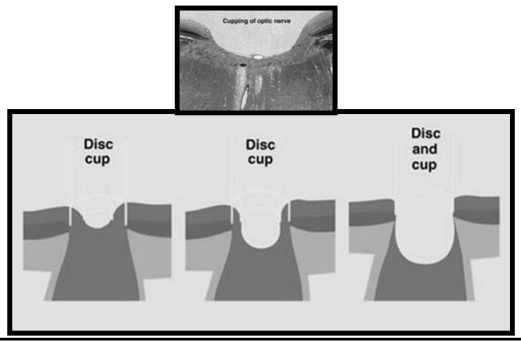
Recording Techniques:

- ***However*:**
  - » **Include horizontal and vertical C/D ratios across disc surface (for documentation purposes)**
- **Detailed drawings with descriptions !!!!!!!**
  - » **Best way to "force" yourself to decribe what you are seeing**
- **Photography (stereo) = Also a standard.**

Physiological Optic Nerve



Think 3-D !



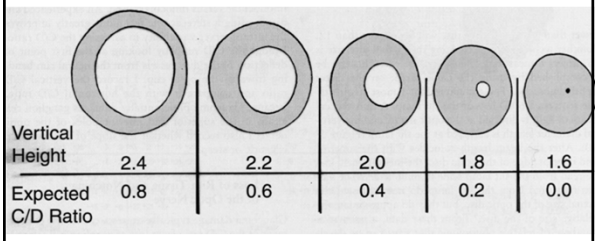
Disc Size vs. Cup Size

- **Larger discs will have larger cups, based on the size of the scleral canal.**
- **Determine the size of the disc:**
  - » **With direct ophthalmoscope use 5^0 aperture: normal disk 10-20% larger**
- **With nerve heads that are larger, you will expect to see a larger cup.**

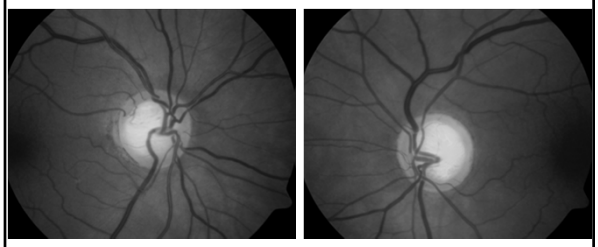
Optic disc size and shape

- **African-Americans have larger discs than Caucasians**
- **Diameter (DD) is 2.1 mm V X 2.8 mm H [average] (Caucasian)**
- **Generally circular; May appear oval due to oblique insertion and be normal**
- **Hyperopic discs are relatively smaller while myopic discs are relative larger**
  [Outside the range +5.00 D to -5.00D]

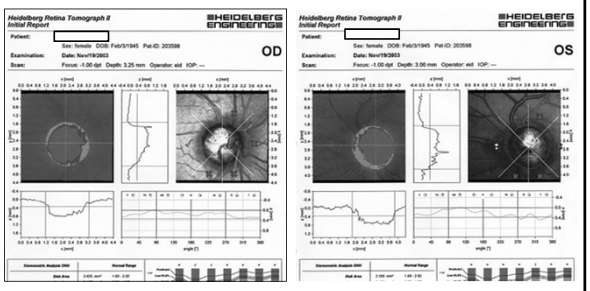
Disc Size vs. Cup Size

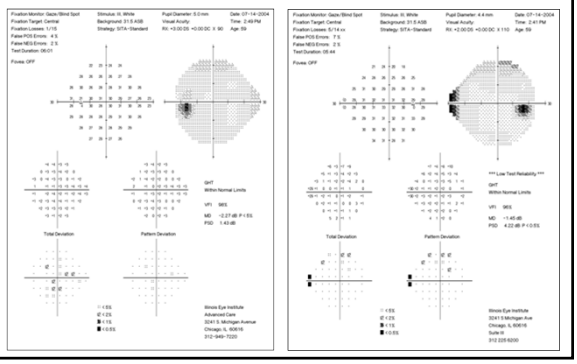


Large Disc / IOP = 18 mmHg

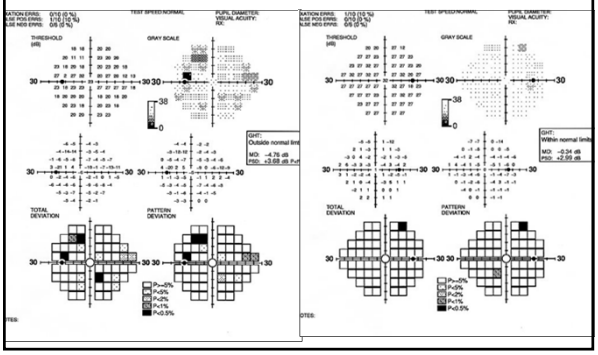


HRT



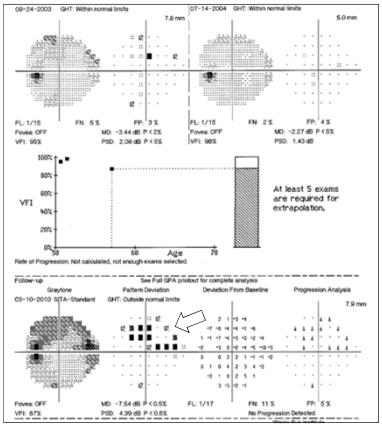


Left Matrix FDT VFs Right

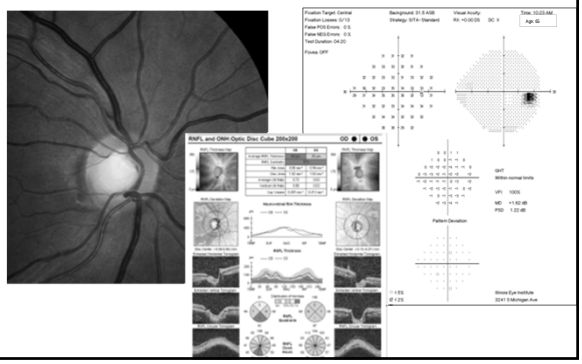


2010

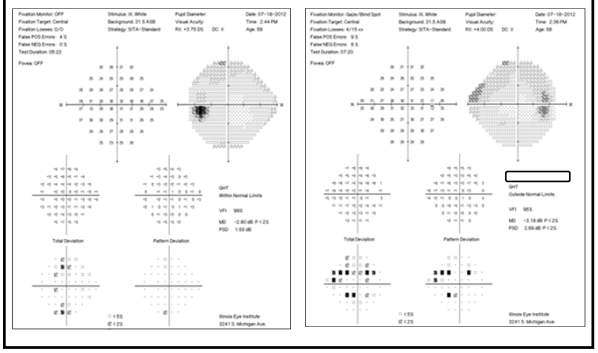
- **No change in vision or new complaints**
- **Not Taking Glaucoma Medications**
  - **GAT = 19 OD 20 OS**
- **Diagnosed with hypertension**
  - **BP = 110/80 w/ atenolol and nifidipine**
- **Now has Medical Insurance**



Pre-Perimetric Glaucoma



Visual Fields



Grouping Disc Appearances

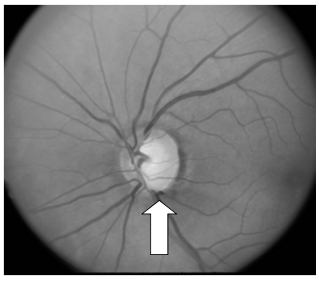
An introductory system used to begin to classify the widely variable appearance of glaucomatous optic nerves.

Grouping Disc Appearances: Types

- **Focal Glaucomatous Disc**
  - »**polar notching**
- **Myopic Glaucomatous Disc**
  - »**tilted insertion, temporal crescents**
- **Senile Sclerotic Disc**
  - »**shallow, sloping cup w/ PPA**
- **Generalized Enlargement**

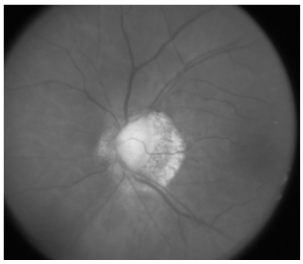
Grouping Disc Appearances

- **Focal Glaucomatous Disc**
  - »**polar notching**



Grouping Disc Appearances

- **Myopic Glaucomatous Disc**
  - »**tilted insertion**
  - »**Peripapillary atropy (PPA)**

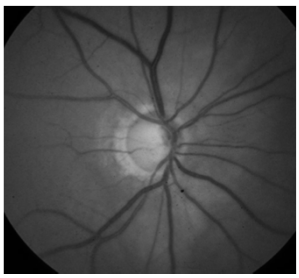


The myopic optic disc

- Sloped and tilted contour
- Very difficult to evaluate
  - » Very high mypoia (>15D) has very high risk
- Scanning laser tests won't help diagnosis but may help identify change
- May rely more heavily on functional visual field testing

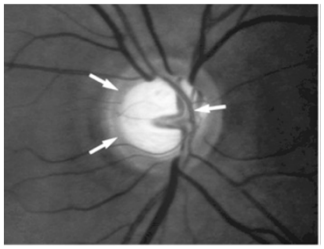
Grouping Disc Appearances

- **Senile Sclerotic Disc**
  - » Pale, shallow, sloping cup w/ PPA

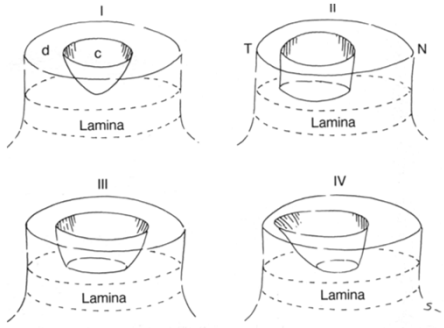


Grouping Disc Appearances

- **Generalized Enlargement**



Contour, Shape & Slope of Cup



Different Types of Glaucomatous Discs

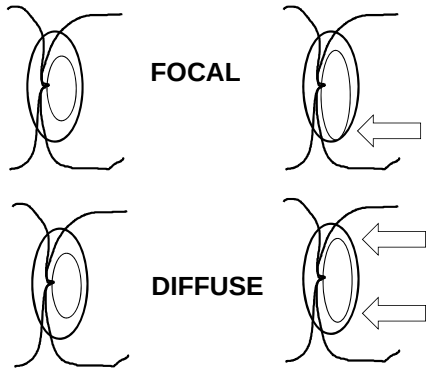
- <u>Focal enlargement</u> - Notching NRR
  - » easier to detect
- <u>Concentric enlargement</u> (diffuse)
  - » even thinning to the NRR
  - » occurs quite regularly
- Deepening of the cup
  - » laminar dots become visible

TIPS and PITFALLS

- Determine the size of the disk,
  - » larger disks will have larger cups.
- Evaluate symmetry between eyes

Coming Up:

- » Disc hemorrhages (NTG)
- » Baring of circumlinear vessel
- » Disk color/pallor; usually healthy

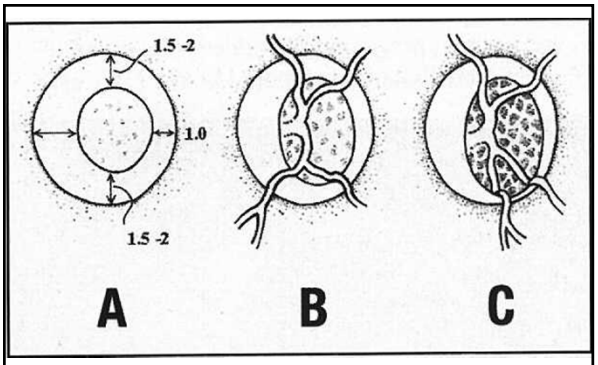


Neural Retinal Rim:

- **"Look at the donut, not at the hole".**
- **Is the PRIMARY location of pathologic changes.**
- **Thus a C/D ratio is often a poor indicator of early glaucoma.**
- **Pay attention to the width and health of the NRR.**

Glaucomatous Neural Rim:

- **Reflects selective loss of tissue, termed, "Notching".**
- **Descriptive terms:**
  - **Thinning of the NRR; Saucerization, Sloping**



Width of the NRR around the disc.

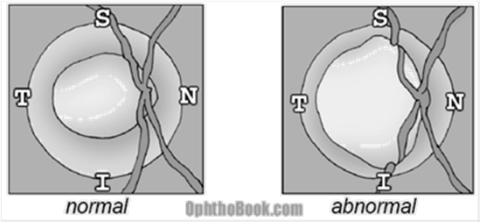
ISNT Rule of the NRR

- **Normal Nerve=**
  - **Inferior= broadest in width, then**
  - **Superior**
  - **Nasal**
  - **Temporal**

Generalizations: of Rim Changes

- **Early Glaucoma=**
  - **inferotemporal and superotemporal rims**
- **Moderate Glaucoma= temporal NRR**
- **Advanced Glaucoma= all around the Rim**

ISNT Rule



- **With glaucoma you begin to see vertical thinning, with atrophy along the inferior and superior rims.**
- **Thus, when optic nerves don't follow the ISNT rule, they may have had glaucomatous damage.**

Can the ISNT rule be applied to patients of African Ancestry?

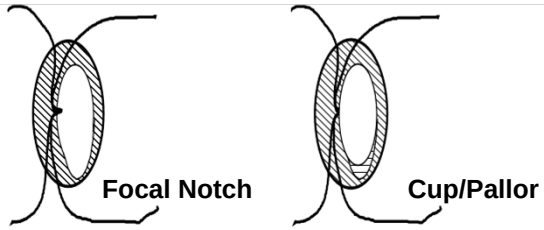
- A study conducted at the New York Eye and Ear Infirmary examined the validity of the ISNT rule (ie, the decreasing order of rim thickness width should be inferior, superior, nasal, temporal) in black and white subjects.
- The investigators evaluated 47 healthy subjects (24 blacks and 23 whites) and 48 OAG patients (18 blacks and 30 whites) by means of simultaneous stereo disc photos and optic nerve imaging using the HRT II and the Stratus OCT.
- Glaucoma diagnosis was based on the visual field and not on any optic nerve criteria. Clinical evaluation of disc photos revealed that the ISNT rule was applicable to 38 of the 47 (80.9%) normal eyes, with no significant difference between blacks and whites (P=.46, Fisher exact test).
- The investigators concluded that the ISNT rule is clinically applicable to healthy, black subjects but is disobeyed in glaucoma patients.
- They also observed that, likely due to the way the data are processed using current software, automated optic nerve topography using the HRT II and the Stratus OCT was not consistent with a clinical assessment of the ISNT rule in healthy subjects.

Glaucoma Today 2005 (presentad at ARVO 2005)

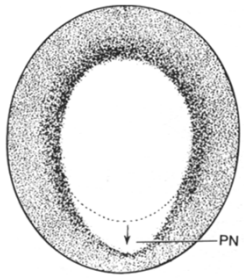
ISNT Rule Summary:

- **Is best used as a critical evaluation technique that "forces" you to spend sufficient time evaluating the the NRR all around the disc**
- **This may be best done on a stereo photograph following the clinical exam**

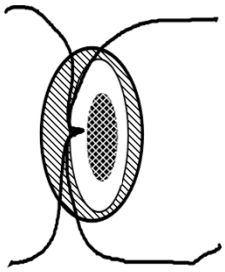
Early Cupping



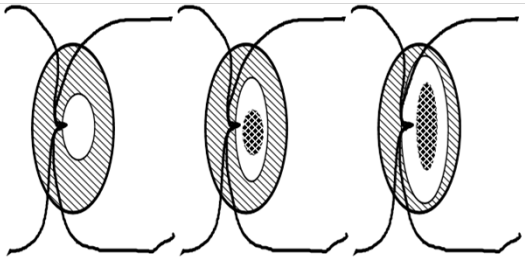
Notching



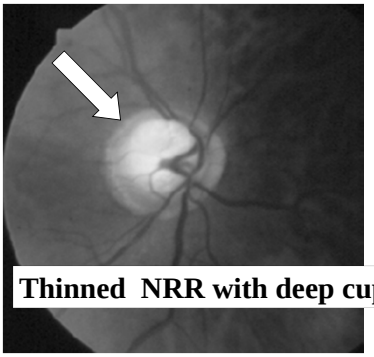
Vertical Elongation



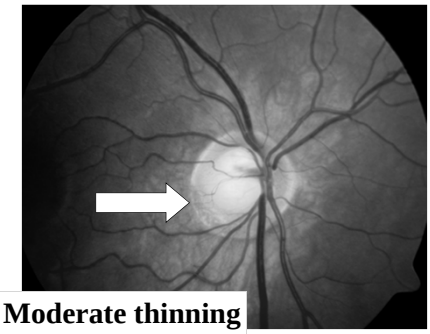
Progressive Excavation



Thinning of NRR, exposure of laminar dots



Thinned NRR with deep cup



Moderate thinning
? Disc Margin

Peripapillary Atrophy

- Irregular pigmentation around the nerve head.
- Non-specific, because also seen in normal eyes, but should raise your suspicion for POAG and NTG.
- Two zones
  - » Alpha
  - » Beta

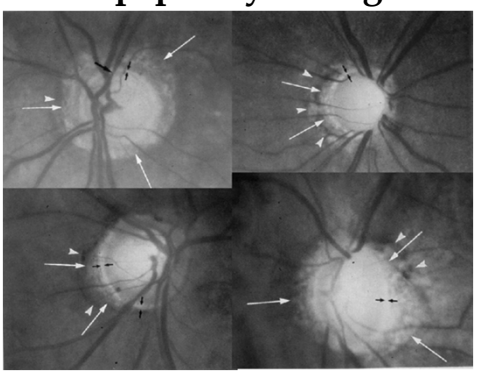
Peripapillary Atrophy

- Alpha Zone
  - » outermost zone appearing as irregular peripapillary pigmentation
- Beta Zone
  - » exposed choridal vessels and sclera
  - » Inside (on disc margin) and adjacent to alpha zones
- Often not able to distinguish the two

Peripapillary Atrophy

- Recent studies have shown that careful PPA evaluation can help in distinguishing glaucomatous nerves from normal nerves
  - » alpha is larger in normals
  - » beta is more frequent / larger in glaucoma
  - » nasal zones more frequent in glaucoma
  - » PPA more frequent in NTG

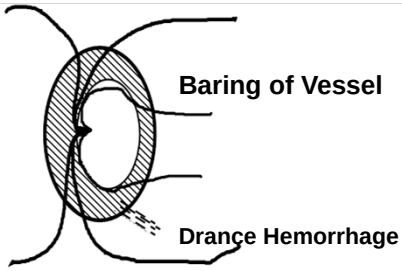
Peripapillary Changes



Vascular Signs:

- **Optic disc (Drance) hemorrhages**
- **Baring of circumlinear vessel**
- **Bayonetting**
  - **Very advanced stage change**

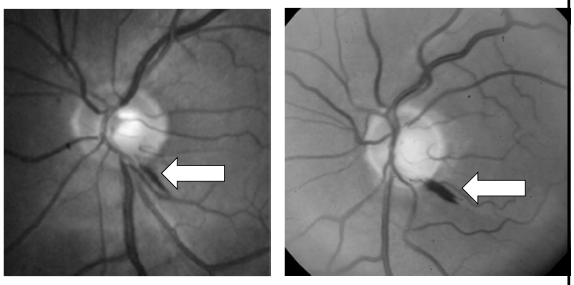
Vascular Signs



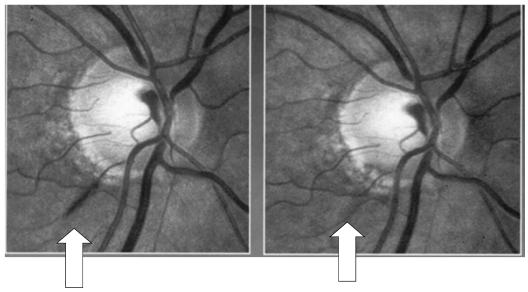
Optic disc hemorrhages

- **Appearance may precede NFL loss, notching, VF defect**
- ***Associated with progressive VF defects in glaucoma or OHT (up to 20X greater risk);* especially among females** [Drance et al. AJO 2001]
- **More frequent in NTG than COAG or OHT**
- **Also seen in PVD, RBVO, hypertensive retinopathy, NAION (< 2% of all ONH hemorrhages)**

Drance/Disc hemorrhage



Optic Disc Hemorrhage



Drance Heme and Progression

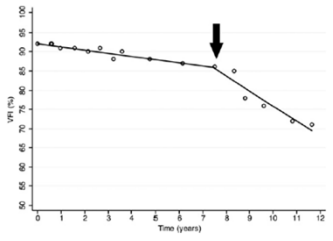
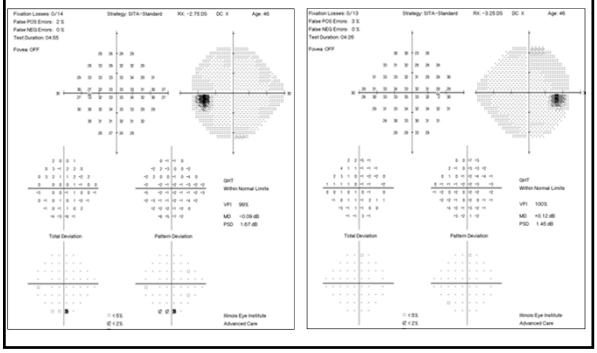


Figure 5. Example of an eye that was progressing at a rate of VFI of −0.86%/year before disc hemorrhage. This rate increased to −4.13%/year after the episode of hemorrhage (*arrow*). The eye had no substantial change in mean IOP levels after the episode of hemorrhage compared with before hemorrhage (15.6 mmHg vs. 15.8 mmHg). VFI = visual field index.

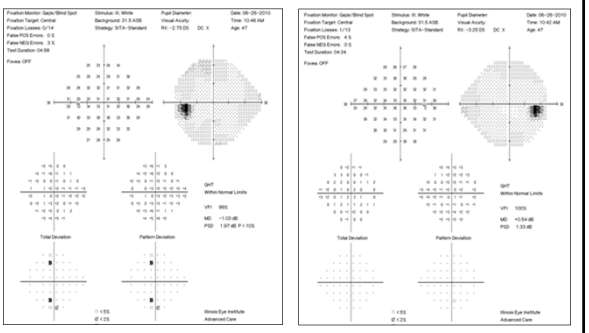
CASE ML

47 yrs old
GAT = ~ 20-21 OD and OS
Asymmetric Cupping
CCT= 525 OD OS
Referred for Treatment

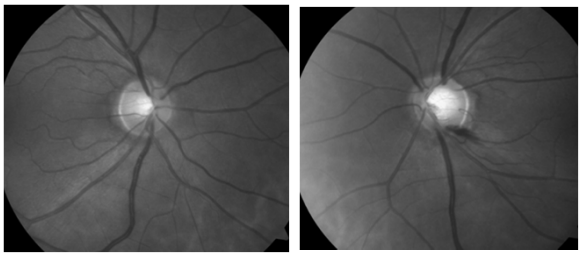
Baseline VFs #1



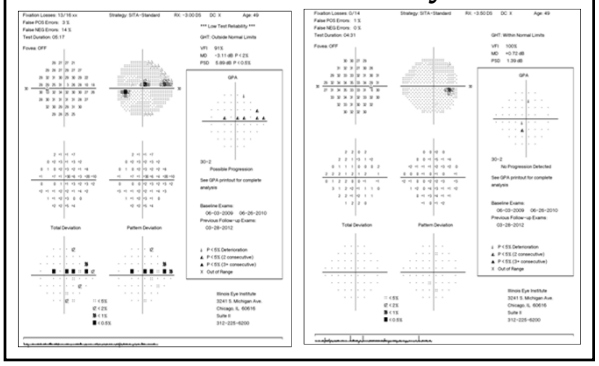
Baseline VFs #2



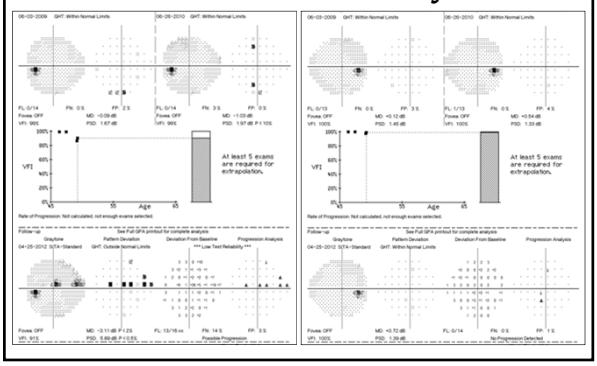
Photos 2 Years Later

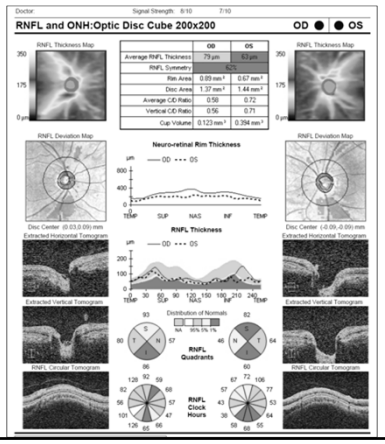


VF with Event Analysis



VF with Trend Analysis

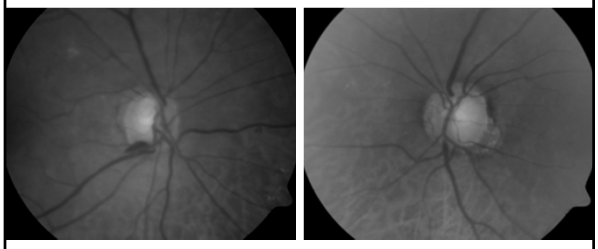




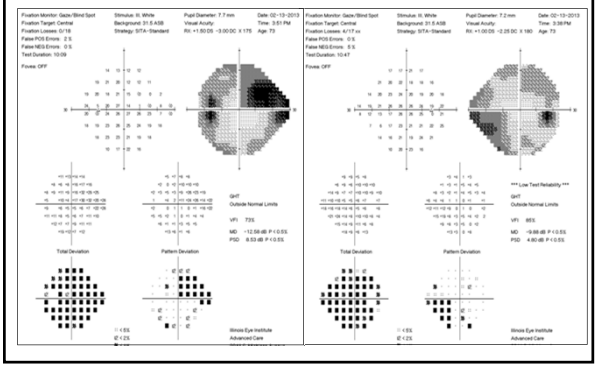
Case JP

- 73 yo, Hispanic woman, no complaints
- HTN, Diabetes
- +3.00D Hyperope
- Narrow Angles, Early Cataracts
  - » now S/P LPI OU
- IOP Max: 19 and 21 mmHg
- Current meds:
  - » Latanoprost qd OU 15, 16 OD OS
- Presents for post LPI follow up exam

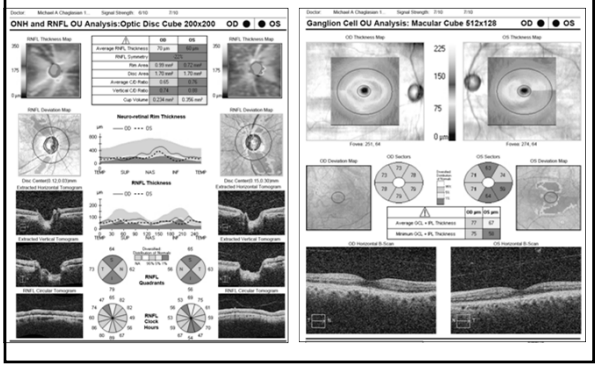
Optic Nerves



Visual Fields



Cirrus OCT



Optic Nerve Evaluation in Glaucoma - Summary

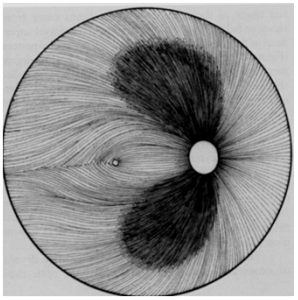
Clinical stereoscopic observation is the mainstay of diagnosis / prospective evaluation

- » ***Expansion of zones α and β***
- » ***Appearance or change in shape of laminar dots***
- » ***Discovery of a splinter hemorrhage at the ONH***
- » ***Digital means of examination may be more valuable for monitoring progression***

Optic nerve description is more than C/D

Nerve Fiber Layer Evaluation

- **Glaucoma evaluation is not only more than C/D . . .**
- **It also includes nerve fiber layer evaluation**



Nerve Fiber Layer Dropout

- **Under red-free filter examination dark slit-like defects may be noticed in patients with glaucoma.**
- **Indicates axonal death/loss.**
- **Perhaps the <u>earliest</u> of all objective signs, but only detectable with experience and optimal conditions.**
- **Thus, not a common clinical technique.**

Retinal NFL Defects

- **Diffuse defects**
  - **Most common of the retinal NFL defects but may be most difficult to identify**
  - **Compare S/I and R/L striations; Look for "raked" appearance/loss of brightness**

Retinal NFL Defects

- **Wedge defects**
  - **Represent territorial loss of NFL**
  - **Easiest to identify but least common**
  - **Usually associated with a notch at the disc and corresponding VF defect; But may "hide" between stimulus presentations [spaced @ 6^0]**

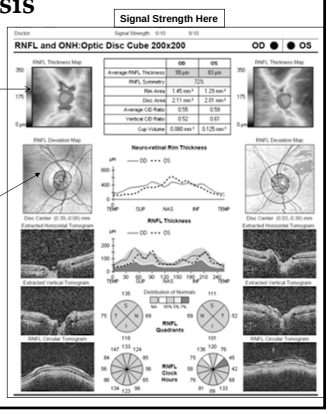
RNFL OU Analysis

<u>RNFL THICKNESS MAP</u> shows the patterns and thickness of the nerve fiber layer within the full 6mm x 6mm area

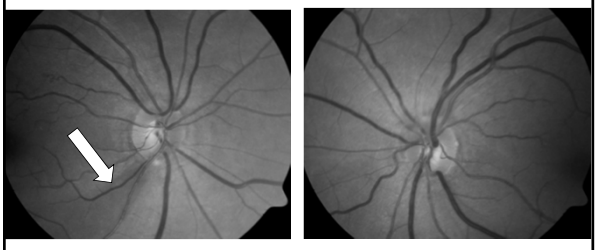
RNFL thickness and comparison to normative data is shown in circle, quadrants and clock hour display

<u>RNFL DEVIATION MAP</u>, overlaid on the OCT fundus image, illustrates precisely where RNFL thickness deviates from the normal range. Data points that are not within normal limits are indicated in red and yellow.

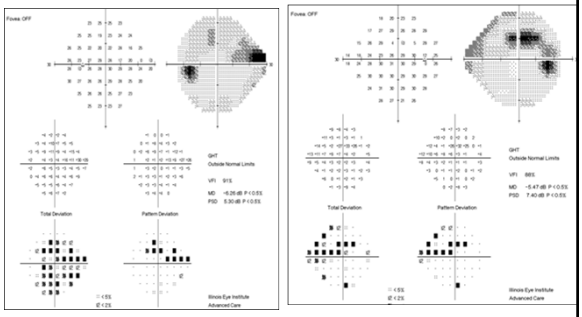
<u>Quadrant and Sector MAPs</u> in middle of page



Case EM

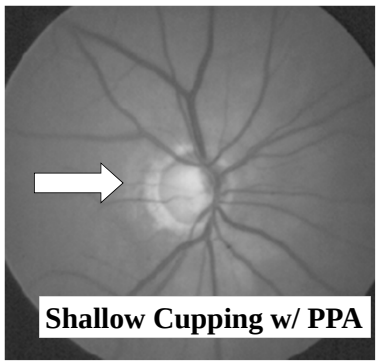


Visual Fields



Normal Tension: Optic Nerve

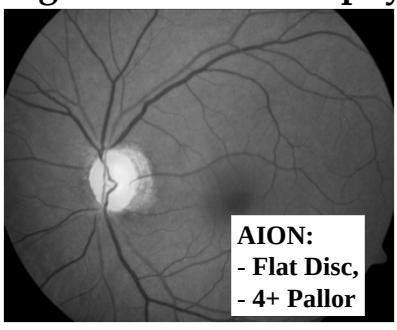
- **Some general characteristics as compared to POAG, but can see a wide spectrum of presentations:**
  - **overall larger and more <u>shallow</u> cupping**
  - **peripapillary atrophy (PPA)**
  - **more focal/sectoral damage than generalized**
  - **Drance (disc) hemorrhages**



Non-Glaucomatous Disc

- **Shows pallor and atrophy <u>ACROSS</u> the entire disc and NRR without significant excavation.**
- **Can result from many causes of optic neuropathy:**
  - **Optic Neuritis, Anterior Ischemic Optic Neuropathy, compressive lesions, chiasmal lesions, infections, inflammation**

Non-glaucomatous atrophy



TIPS and PITFALLS

- **Do not emphasize the C/D ratio**
- **Concentrate on the neural retinal rim**
- **Look for focal defects (notching) and and/or generalized thinning**
- **Gauge the depth of the cup**
- **Evaluate symmetry between eyes**

TIPS and PITFALLS

- **Peripapillary atrophy (NTG)**
- **Disc hemorrhages (NTG)**
- **Baring of circumlinear vessels**
  - **Loss of NRR tissue**
- **Disk color or amount of pallor**
- **Use imaging and perimetry to evaluate suspicious nerves and high risk patients**