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
- Neurotoxic Processes
  - Release of excitotoxins
  - Blockage of neurotrophic growth factors
  - Programmed cell death, “Apoptosis”
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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
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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

VF s #2
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RNFL OU Analysis

- **RNFL THICKNESS MAP**: Shows the patterns and thickness of the nerve fiber layer within the full 6mm x 6mm area.
- **RNFL DEVIATION MAP**: Overlaid on the OCT fundus image, it precisely localizes RNFL thickness deviation 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 fenestrated 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 fenestrations 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.
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Lower Diastolic, Systolic, or Mean Pressure Reduces Perfusion Pressure

Higher IOP Negatively Impacts Perfusion Pressure

Lower Perfusion Pressure Is Associated with Increased Risk for Open Angle Glaucoma

Hayreh SS, Trans Am Acad Ophthalmol 1974;70,943-54

Ocular Perfusion Pressure

Perfusion Pressure Is a Result of A Delicate Balance Between IOP and Blood Pressure

CSF Pressure

Intraconal CSF

Trans-lamina cribrosa

Pressure Gradient

Progression of Excavation

Normal optic nerve

Early glaucoma

Advanced glaucoma

Cerebrospinal Fluid Pressure


- 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 I C and peripapillary sclera, the retroorbital 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 DP, the CSF pressure and the IOP
- Low CSF pressure may play a role in the pathogenesis of NTG

Low OPP = Higher Risk

- May be due to:
  - High IOP
  - Low BP
- Physiological
- Over treatment of systemic HTN
- Nocturnal Hypotension
Gluomatos 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.
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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 describe what you are seeing

- Photography (stereo) = Also a standard.

Physiological Optic Nerve

- Inner Cup Margin
- Disc Margin
- Neural Retina Rim

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° 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]
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
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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 atrophy (PPA)
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The myopic optic disc

- Sloped and tilted contour
- Very difficult to evaluate
  » Very high myopia (>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

Grouping Disc Appearances

- Contour, Shape & Slope of Cup

Different Types of Glaucomatous Discs

- Focal enlargement - Notching NRR
  » easier to detect
- Concentric enlargement (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
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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=.48, 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.

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

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Early Cupping

- Focal Notch
- Cup/Pallor

Notching

- PN

Vertical Elongation

ProgressIVE Excavation

- Thinning of NRR, exposure of laminar dots
Assessing the Glaucomatous Optic Nerve

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 circumpolar vessel
- Bayonetting
  » Very advanced stage change

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)

Optic Disc Hemorrhage

Drance Heme and Progression

Drance/Disc hemorrhage
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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
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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 Nerve Evaluation in Glaucoma - Summary
Clinical stereoscopic observation is the mainstay of diagnosis / prospective evaluation
- Expansion of zones $\alpha$ and $\beta$
- 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
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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 earliest 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°]

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 retinal 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 to indicate safe.

Case EM
Assessing the Glaucomatous Optic Nerve

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 shallow cupping
» peripapillary atrophy (PPA)
» more focal/sectoral damage than generalized
» Drance (disc) hemorrhages

Shallow Cupping w/ PPA

Non-Glaucomatous Disc

Shows pallor and atrophy ACROSS 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

AION:
- Flat Disc,
- 4+ Pallor
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