Glaucoma and the Optic Nerve

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Learning objectives

• Review the anatomy of the optic nerve
• Recognize the appearance of the healthy optic nerve
• Ascertain the critical components of optic nerve assessment
• Identify possible glaucomatous changes of the optic nerve, peripapillary region, and RNFL
• Identify the cardinal features of glaucomatous optic neuropathy
• Analyze optic nerve images to solidify today’s discussion
What is glaucoma?

- A *progressive optic nerve disease* characterized by *retinal ganglion cell death* and resultant *axon loss* seen as *excavation of the optic nerve* head with consequent defects in retinal sensitivity that can be measured with visual field tests.
What is glaucoma?

- Optic neuropathy
- Axon loss
- Excavation of the optic nerve
- Resultant VF defects
Glaucomaticous damage may be due to

- Elevated IOP
- Poor perfusion pressure to the ONH
- Obstruction of axoplasmic flow within the ganglion cell axons
- Anatomic weakening of the lamina cribrosa
  - Myopia
  - Optic nerve pits
- Programmed cell death of the ganglion cell axons (apoptosis)
Pre-perimetric glaucoma?

20-40% of ganglion cells are lost before VF defects are detected on standard automated perimetry.

So what?

Assessment of the ONH is *critical* for early diagnosis and management to prevent VF defects before they occur.
Optic Nerve Head (ONH)

- Careful evaluation of the ONH has high specificity and good precision for glaucoma diagnosis
- It is one of the most important aspects of glaucoma assessment
Anatomy of the optic nerve

- Ganglion cell axons make up 90% of neuroretinal rim tissue of the optic disc
  - 1-1.5 million axons leave via the ONH through the scleral canal
  - Grouped into bundles by glial cells
- Remainder of neuroretinal rim is composed of capillaries and astrocytes
- Axons in superior and inferior poles have less structural support
Anatomy of the ONH

- Four distinct layers of the ONH
  - Surface layer
  - Prelaminar ONH
  - Laminar ONH
  - Retrolaminar ONH
Anatomy of the ONH

- **Surface layer**
  - Anterior limit of the ONH
  - Point of contact with the vitreous
  - Peripheral edge is defined by anterior limits of the scleral ring
  - Posterior limit: axonal bundles have completed 90 degree turn from the plane of the retina and reached the level of the choroid
Anatomy of the ONH

- **Prelaminar ONH**
  - Indistinct segment of axons surrounded by outer retina, choriocapillaris, and choroid
Anatomy of the ONH

- **Laminar ONH**
  - Ganglion cell axon bundles wrapped in glial cells and confined in rigid pores of the lamina cribrosa
Anatomy of the ONH

- **Retrolaminar ONH**
  - Posterior to lamina cribrosa
  - ONH thickness is doubled by presence of myelinating oligodendrocytes
RNFL distribution
Lamina cribrosa

- Composed of several sheets of connective tissue
  - Fenestrated to allow passage of nerve fiber bundles carrying ganglion cell axons
- Variable number of pores: 200-600
  - Larger pores at superior and inferior poles - may provide less support than smaller fenestrations in nasal and temporal regions, resulting in greater damage to RGC axons in these areas
- Laminar dots become more exposed and numerous with progressive axon loss
Lamina Cribrosa
Changes in the lamina cribrosa

- Normally pores are obscured by nerve fibers
- As nerve fibers undergo atrophy, pores become more visible
  - AKA laminar dot sign
  - Can be present in healthy eyes
- Thinning and backward bowing of lamina cribrosa occurs with deepening of cup
Thinning and backward bowing of LC

[Diagram showing normal optic nerve, early glaucoma, and advanced glaucoma with labels for optic disk, optic nerve, optic cup, early damage, and severe damage.]
Laminar dot sign
ONH Blood supply

- **Superficial ONH**
  - Branches from CRA
- **Pre-laminar ONH**
  - Short posterior ciliary arteries (SPCA)
- **Laminar ONH**
  - Circle of Zinn-Haller: anastomoses of adjacent SCPA’s
- **Retro-laminar ONH**
  - SPCA
  - Pial vascular plexus
  - Axial vasculature from CRA
Ocular perfusion pressure

Higher IOP Negatively Impacts Perfusion Pressure

Lower Diastolic, Systolic, or Mean Pressure Reduces Perfusion Pressure

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

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

- Via central retinal vein
- In chronic glaucoma, shunt vessels may appear due to disturbed retinal circulation
  - AKA optociliary shunt vessels
  - Pre-existing capillaries that become more visible as they dilate to re-route blood around an area of obstruction
Optociliary shunt vessels

- Differential diagnoses
  - CRVO
  - Optic nerve sheath meningioma
  - Chronic glaucoma
  - Chronic papilledema
- Different from neovascularization of the disc
  - Do not leak on FA
Optic cup

- Central excavation in the ONH
- Devoid of axons and capillaries
  - Pale due to visibility of collagenous lamina cribrosa
  - Size is dependent on number of nerve fibers leaving the eye and the size of the scleral canal
- Cup depth usually depends on cup size
  - Small cup = shallow
  - Large cup = deep
Optic cup

- High inter-individual variability
- Lies below the level of the neural rim
- Bottom is formed by the LC
- Border between cup and rim is determined by contour, not the color
  - Point of deviation of vessels on the surface of the ONH
  - Area of pallor of the cup usually corresponds to the borders of the cup
Contour vs. color
Contour < color
Contour = color
Contour vs. color
Optic cup variants
Optic cup variants
Optic cup variants
Changes in the optic cup

- Increased size
- Increased depth
- Visualization/increase in laminar dots
- Vertical enlargement
  - Localized neuroretinal rim loss at superior and inferior poles
- Asymmetry between two eyes greater than 0.2
  - In the absence of disc size asymmetry
Optic pit

- Localized weakening in the lamina cribrosa
- Usually located infero-temporally
- More prevalent in NTG
- Will have a corresponding but stable VF defect
Neuroretinal rim

- Point of exit of nerve fiber bundles from the eye through the scleral canal
- Healthy rim tissue should be pink due to presence of pre-laminar capillaries
- May be difficult to assess in high myopes, patients with tilted discs, and nerves with significant pallor
Peripapillary area

- Elschnig’s scleral ring
  - Thin white ring outside of disc margin
  - AKA scleral lip
  - Anterior extension of sclera between the choroid and the optic nerve – RPE and choroid stop short of the disc
Peripapillary area

- **Choroidal crescent**
  - RPE stops short of the disc
  - Underlying choroid visible
  - Usually slate-gray in color
Peripapillary area

- Peripapillary RPE hypertrophy
  - Darker than choroidal crescent
  - Increased amount of peripapillary RPE pigment
  - Normal variant
Peripapillary area

- Grey crescent: located within scleral lip on the neuroretinal rim
  - Caution: may lead to false interpretation of neuroretinal rim – may be interpreted as thinner than it truly is
  - May be pigmentation within the neural tissue cells – melanocytes, RPE cells, or free pigment granules
  - Normal finding
Peripapillary area

- Zone alpha
  - Found in normals
- Zone beta
  - More common in glaucoma patients
Peripapillary zones

- 1 = cup
- 2 = neuroretinal rim
- 3 = scleral crescent
- 4 = zone beta
- 5 = zone alpha
Optic nerve variants
Optic nerve variants
Caution: myopic nerves

- Certain features of highly myopic discs interfere with interpretation of the neuroretinal rim and amount of cupping
  - Large disc area
  - Oblique insertion of optic disc causing distorted view of the temporal rim
  - Tilt makes assessment of superior and inferior poles difficult
  - Shallow cupping makes C/D ratio difficult to assess
Caution: myopic nerves & oblique insertion
Caution: myopic nerves

- Wide temporal peripapillary crescent causes difficulty in assessing temporal rim
- In this case, look at asymmetry and the integrity of the nasal rim
Myopic nerves: pearls and pitfalls

• Photodocumentation is vital to evaluate for change
  ▫ Serial imaging with OCT to monitor for change is extremely beneficial
  ▫ Caution: normative databases on imaging technologies do not apply

• Any change in VF status is suspicious

• May have higher risk of converting into glaucoma due to weakened lamina cribrosa

• May be more vulnerable to even slight IOP increase due to longer globe, thinner LC, and thinner scleral wall
Optic disc evaluation in glaucoma
Goals of optic disc evaluation

- Diagnose: distinguish between normal and abnormal
- Quantify: how much damage has occurred
  - 20-40% of ganglion cell axons can be lost before reproducible VF loss appears on automated perimetry
  - Ganglion cells die at the level of the lamina cribrosa, with retrograde atrophy back to their cell bodies in the retina
- Monitor for change
  - Stable
  - Worsening
- Quantify rate of change: slow vs. rapid
Optic disc evaluation

- Slit lamp biomicroscopy: ideal
  - Stereoscopic view
  - Measuring optic disc size
- Direct ophthalmoscopy
  - Good magnification
  - No stereo
- Indirect ophthalmoscopy
  - Poor magnification and detail
- Optic disc photography
  - Great for documentation and monitoring for progression
  - Always taken at baseline, and usually every 2 years afterwards
What to look for?

- Disc: size and shape
- Neuroretinal rim: size, shape, color, localized defects (notching)
- Cup: size and shape *in relation* to the optic disc size
  - 0.7 C/D in a 1.8mm nerve – probably NOT ok
  - 0.7 C/D in a 2.7mm nerve – probable NOT glaucoma
- Optic disc hemorrhage: presence and location
- Nerve fiber layer defect
- Peripapillary atrophy
- Retinal arterial attenuation
Optic disc size

- Critical in distinguishing between physiologic and pathologic cupping
  - Scleral foramen/canal: 1-3mm
    - Large foramen = large disc = large cup
    - Small foramen = small disc = small cup
Optic disc size

- Measurement of vertical disc diameter
  - Length of vertical beam of slit lamp light
  - Multiplied by correction factor of condensing lens
    - Volk 60D: x 1.0
    - Volk 78D: x 1.1
    - Vold 90D: x 1.3
Optic disc size

- Average vertical diameter: 1.8-2.0mm
- Small optic nerve vertical diameter: ≤1.5 mm
- Large optic nerve vertical diameter: >2.2mm
Disc vs. cup size

- Larger discs = larger cups
  - Due to the size of the scleral canal
- Always determine the size of the disc
Large disc = large cup
Small disc = small cup
Early and moderate glaucomatous damage in small discs may be missed due to initial low C/D ratios.
C/D ratio

- Optic disc elongation
  - Vertically oval optic disc
  - Horizontally oval optic cup
- In normal eyes: horizontal C/D ratio > vertical C/D ratio
- In glaucomatous eyes: vertical C/D ratio > horizontal C/D ratio
- Documentation
  - Always include horizontal and vertical CD ratio
  - Stereophotographs of the ONH always beneficial
Neuroretinal rim

- Reflects selective loss of tissue
- It is the primary location of pathologic changes
- C/D ratio is often a poor indicator of early glaucoma
- Pay attention to the width and the health of the neuroretinal rim
- Look at the donut, not at the hole!
The neuroretinal rim

- Size
- Shape
- The ISNT rule
- Color
  - Glaucoma: cupping WITHOUT pallor
The ISNT rule
The ISNT rule
The neuroretinal rim

- Look for
  - Thinning
  - Notching: localized defect in the neuroretinal rim
  - Pallor: suspect a different or additional optic neuropathy
The neuroretinal rim

- Usual sequence of loss in glaucoma:
  - Inferotemporal/superotemporal
  - Temporal
  - Inferonasal/superonasal

- In non-glaucomatous optic nerve damage, the rim is not always affected, therefore its contour is maintained
Patterns of cupping

- Diffuse cupping
Patterns of cupping

- Focal atrophy: notching
Patterns of cupping

- **Bean-pot cupping**
  - Extreme posterior displacement of lamina cribrosa
Peripapillary chorioretinal atrophy

- Irregular pigmentation around the optic nerve
- Nonspecific finding
  - Seen in normals
  - Should raise suspicion for POAG and NTG
- Associated with acquired damage to the optic nerves from glaucoma
- Clinical appearance
  - Moth-eaten appearance of the RPE temporal to ONH
  - Adjacent to area of neuroretinal rim thinning
Peripapillary atrophy

- **Zone Alpha**
  - Hypo and hyper pigmented areas due to RPE irregularity
  - Nasally bounded by zone beta
  - Temporally bounded by normal retina
  - Present in normal eyes
  - Present in glaucomatous eyes
Peripapillary atrophy

- **Zone Beta**
  - Atrophy of the RPE and choriocapillaris
  - May be due to poor perfusion to the peripapillary area
  - Large choroidal vessels become visible
  - More common in glaucomatous eyes
Peripapillary atrophy

- Helps differentiate between glaucomatous and non-glaucomatous optic nerve damage
  - Beta zone larger and more frequent in glaucoma
  - Nasal PPA more frequent in glaucoma
- Width of beta zone inversely correlated with adjacent rim width
  - Larger beta zone → thinner neuroretinal rim
- Progression of beta zone associated with progression of glaucoma
Vascular changes

- Optic disc hemorrhages
- Baring of curcumlinear vessel
- Bayonetting of vessels
  - Advanced cupping causes vessels to emerge from floor of the cup, disappear as they ascend up the excavated wall of the cup, and emerge again at the disc margin
- Nasalization of vessels: major vessels show nasal shift
- Optic nerve shunts/collaterals
- Retinal artery attenuation
Optic disc hemorrhage

- Aka drance hemorrhage
- Splinter or flame shaped
- Located on the disc margin
- Hallmark of glaucomatous optic nerve damage
  - 4-10% of eyes with glaucoma
- Found in early and moderate stages, rare in advance stages
- Usually located on IT and ST disc margins
Optic disc hemorrhage

- Can resolve within 6-10 weeks of onset
  - Can take anywhere between 2-35 weeks
- Associated with localized RNFL defects and rim notching
- Suggests progression
  - Appearance may precede RNFL loss, notching, and VF defect
- More common in NTG
- Can be seen in PVD, BRVO, HTN retinopathy, and NAION
Baring of vessels
Bayonetting of vessels
Nasalization of vessels
Optic nerve shunts/collaterals
Retinal artery attenuation

- Diffuse narrowing
  - Decreasing neuroretinal rim
  - Increased RNFL loss
  - Increased VF defects

- Focal attenuation
  - More common in NTG
  - Degree of narrowing increases with amount of damage
Retinal nerve fiber layer (RNFL)

- RNFL: retinal ganglion cell axons covered by astrocytes and bundled by Muller cell processes
- Seen as bright fine striations fanning off the disc
- Best evaluated with red-free filter
- Can be difficult to appreciate in the blond fundus
- Most visible infero-temporally and supero-temporally
- Obscures details of underlying peripapillary retinal vascular walls
Clinical assessment of RNFL

- Requires
  - Bright light
  - Red-free filter
    - Green light produced by filter is absorbed by the RPE and choroid, creating a dark background
    - The RNFL reflects the green light and is contrasted against the dark background
Normal RNFL

- Bright, linear, striated appearance
- Coarse texture
- Casts white haze over underlying retinal structures and obscures smaller blood vessels
- Normal pattern: bright-dim-bright
  - Pattern should be symmetric between S/I bundles and between the two eyes
- Brightness depends on
  - Integrity of RNFL bundles
  - Amount of pigmentation in RPE and choroid – blonde fundi, dull RNFL
  - Media clarity
RNFL defects in glaucoma

- Selective damage to superior and inferior arcuate bundles
- Relative sparing of papillomacular and nasal bundles
- Defects appear as darker zones in areas of expected brightness
- Retinal vessels appear redder and darker
- Small vessels become more visible
RNFL defects

- Diffuse
  - Most common and most difficult to detect
  - Compare S/I and R/L striations: raked appearance and loss of brightness
  - Peripapillary vessels appear bare
  - Underlying choroidal vessels more clearly visible
RNFL defects: diffuse loss

- **Mild (D1)**
  - Striations are less bright and less coarse
  - Medium size vessels apparent
  - Small vessels still obscured
- **Moderate (D2)**
  - Striations even less prominent
  - Medium and small vessels clear
- **Severe (D3)**
  - Few striations visible
  - Deep retinal layers have grainy appearance
  - Pseudosheathing of blood vessels: collagen walls become more visible
RNFL: diffuse loss
RNFL: diffuse loss
RNFL defects: focal loss

- Slit or wedge
  - Easiest to identify
  - Less common
  - Usually associated with notch at disc or current/prior drance hemorrhage
RNFL: focal loss
RNFL defects: focal loss

- **Slit:**
  - Larger than an arteriole in width
  - Travels back to the ONH
- **Wedge:**
  - Expanding focal damage
  - Associated with notching and arcuate VF defect
Take-home points

- C/D ratio is NOT the only factor to consider when evaluating the ONH
- You MUST give due diligence to the neuroretinal rim
  - Focal defects
  - Generalized thinning
- You MUST evaluate any asymmetry in the superior and inferior poles of the same eye
- You MUST evaluate any asymmetry between the two eyes
- Always remember, glaucoma is cupping WITHOUT pallor
- Use imaging technologies and perimetry to evaluate suspicious nerves and high-risk patients
Optic nerve evaluation checklist

- Measure size and shape of the ONH
- Evaluate size and shape of the optic cup
- Determine the vertical and horizontal C/D ratio
- Compare the expected C/D ratio based on vertical disc diameter
- Neuroretinal rim integrity/thinning/notching/pallor
  - Superior vs inferior
  - OD vs OS
- Vascular changes: disc hemorrhages, nasalization of vessels, arteriole narrowing, optociliary shunt vessels, baring of vessels
- Peripapillary atrophy
- RNFL defects: diffuse/focal
Let us look at some nerves!
Thank you!