The Optic Nerve and Glaucoma

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Abstract:
Glaucomatous optic neuropathy is classified by morphologic changes to the optic nerve head and the retinal nerve fiber layer. These changes can be evaluated by careful clinical assessment of the neuroretinal rim, optic nerve size, vascular signs, parapapillary chorioretinal atrophy and the appearance of the retinal nerve fiber layer. The current level of understanding of the presence and significance of these signs will be presented with respect to the normal optic nerve and the early diagnosis of glaucoma. The 5-Rs a simplified grading system that incorporates these basic principles, will be presented.

1. Systematic Assessment

The 5-Rs (Weinreb, Medeiros, Susanna, 2005)

- Observe the scleral Ring to identify the limits of the optic disc and its size
- Identify the size of the Rim
- Examine the Retinal nerve fiber layer
- Examine the Region of parapapillary atrophy
- Look for Retinal and optic disc hemorrhages

2.1 Optic Disc Size

- 0.80 mm^2 to 6.00 mm^2 (1:7 in a normal Caucasian population)
- Largely independent of the refractive error between -5 and +5 D.
  - 1.2% ± 0.15% for each diopter increase towards myopia.
  - >+5 D the optic disc is significantly smaller.
  - <-8D the optic disc is significantly larger than in emmetropic eyes.
- Race dependent
  - Caucasians < Hispanics < Asians < Afro-Americans.
- Correlated with the size of the optic cup and neuroretinal rim
  - A large cup in a large optic disc can, therefore, be normal, while a small optic cup in a very small optic disc suggests glaucomatous optic nerve damage.
- Mean Caucasian optic disc area non-highly myopic s examined in various studies ranged between 2.1 mm^2 and about 2.8 mm^2

2.2 Optic Disc Shape

- Vertically oval (7-10%)
- Min-to-max disc diameter ratio 0.64 and 0.98 (1:1.53)
- No difference between normal (<-8D.) and glaucoma eyes
- Glaucoma susceptibility is mostly independent of the shape of the optic disc
3.1 Neuroretinal Rim Size
- Correlated with the optic disc area: the larger the disc, the larger the rim
- Infero-temporal and supero-temporal disc sectors have the highest predictive power

3.2 Neuroretinal Rim Shape
- ISN'T rule: Neuroretinal rim broadest Inferior, followed by the Superior, Nasal and Temporal
- Neuroretinal rim is lost in all sectors of the optic disc with regional preferences depending on the stage of the disease
  - Early glaucoma, rim loss predominantly in the inferotemporal and superotemporal disc regions
  - Moderate glaucoma, the temporal horizontal disc region
  - Advanced glaucoma, the rim remnants are located mainly in the nasal disc sector
- This sequence of disc sectors (inferotemporal - superotemporal - temporal horizontal - nasal inferior - nasal superior) correlates with the progression of visual field defects

3.3 Neuroretinal Rim Pallor
- Increasing pallor of the optic disc and especially of the neuroretinal rim is a typical sign of optic nerve damage
- More marked in eyes with non-glaucomatous optic neuropathy
- Mis-match between pallor and neuroretinal rim margin can be sign of early glaucoma

4.1 Optic Cup Size
- High interindividual variability
- The area of the optic disc and cup are correlated in normal eyes
  - The larger the optic disc, the larger the optic cup
- In contrast to glaucoma, the optic cup does not markedly enlarge in eyes with non-glaucomatous optic nerve damage

4.2 Optic Cup Shape and Depth
- In normal eyes, the shape of the optic cup is horizontally oval with the horizontal diameter being about 8% bigger than the vertical diameter
- The optic cup depth depends on the cup area in normal eyes,
  - The larger the cup, the deeper the cup
- In glaucoma, the optic cup deepens dependent on the type of glaucoma and the level of intraocular pressure
- Saucerization, a diffuse, shallow cupping that extends to the disc margin, is an early sign of glaucoma

5. Cup/Disc Ratios
- In normal eyes, larger horizontally than vertically
• Range in a normal population between 0.0 and almost 0.9
• Eyes with physiologically high cup/disc ratios and large discs should not be overdiagnosed as glaucomatous, and eyes with increased intraocular pressure, small optic nerve heads, and average or low cup/disc ratios should not be underdiagnosed
• Cup/disc asymmetry of >0.2 can be an early sign of glaucoma

6. Parapapillary Chorioretinal Atrophy
• Central beta zone (bordering the optic disc)
  - Characterized by visible sclera, visible large choroidal vessels, RPE atrophy and absolute scotoma
• Peripheral alpha zone
  - Characterized by irregular pigmentation, apparent thinning of the chorioretinal tissue and relative scotoma
• Alpha zone is present in almost all normal eyes and is thus more common than beta zone (found in 15%-20%)
• The myopic scleral crescent differs histologically from the glaucomatous beta zone
  - In myopic crescent, only the inner limiting membrane and underlying retinal nerve fiber layer or its remnants cover the sclera
  - In the glaucomatous beta zone, Bruch’s membrane and the choroid are interposed between the remnants of the retina and the sclera
• A large beta zone is associated with fundus tessellation, shallow cupping, a low frequency of disc hemorrhages, localized nfl defects, concentric loss of neuroretinal rim, and normal IOP

7. Position of the Central Retinal Vessel Trunk
• The longer the distance to the central retinal vessel trunk exit, the more marked the neuroretinal rim and visual field loss
• Glaucoma eyes with a temporal cilioretinal artery retained central visual field (and temporal neuroretinal rim)

8. Optic Disc Hemorrhages
• Splinter-shaped or flame-shaped hemorrhages at the border of the optic disc are a hallmark of glaucomatous optic neuropathy however:
  - Low sensitivity 4% to 7%
• Rarely found in normal
  - High specificity 1%
• In early glaucoma, usually located inferotemporal or superotemporal
• Associated with localized RNFL defects, neuroretinal rim notches and visual field loss
• Sign of progressive disease
• Within two months of detection can develop RNFL and visual field defect
• Most often in patients with focal normal-pressure glaucoma

9. Diameter of Retinal Arterioles
• Vessel diameter reduces with decreasing area of neuroretinal rim, diminishing visibility of the RNFL and increasing visual field defects
• Secondary to a reduced demand in the superficial layers of the retina.
• Focal narrowing in optic nerve damage such as nonarteritic anterior ischemic, optic neuropathy and glaucoma

10. Retinal Nerve Fiber Layer
• Most visible in the temporal inferior sector, followed by the temporal superior, the nasal superior and the nasal inferior sector (correlates with the sectors' sequence with respect to rim configuration and retinal artery caliber).
• Visibility of the RNFL decreases with age, correlates with age-related reduction of the optic nerve fiber count with an annual loss of c.4,000 to 5,000 fibers/year
• Defects typically occur in c.20% of all glaucoma eyes
• Also found in optic nerve atrophy
• Rarely present in normal eyes
• Important sign for “pre-perimetric” glaucoma but not pathognomonic of glaucoma
• Frequency increases significantly from early to moderate disease
• In severe disease no longer detectable due to the pronounced loss in all sectors.
• More often in eyes with the focal type normal-pressure glaucoma
• Localized defects are often found 6 to 8 weeks after optic disc hemorrhage
• Most often located in the temporal inferior sector followed by the temporal superior sector, rarely in nasal region

11. Diagnosis of Early or "Preperimetric" Glaucomatous Optic Nerve Damage
• Carefully examine:
  - Neuroretinal rim for characteristic ISN’T rule
  - Optic disc size versus cup size
  - Optic disc hemorrhages
  - Localized RNFL defects
  - Diffuse RNFL
  - The presence of zone beta or extensive zone alpha parapapillary atrophy