Clinical Evaluation of Optic Nerve Head in Glaucoma

Shibal Bhartiya, Ritu Gadia, Harinder S Sethi, Anita Panda
Glaucoma Services, Dr RP Center for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence: Shibal Bhartiya, Glaucoma Services, Dr RP Center for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India, e-mail: shibalbhartiya@gmail.com

ABSTRACT

Glaucomatous optic neuropathy is characterized by changes in the intrapapillary and parapapillary region of the optic nerve head, including excavation of the optic nerve head and consequent defects in retinal sensitivity with visual field defects and other psychophysical alterations. Clinical evaluation of the optic nerve head has been shown to have a high specificity and good precision for glaucoma diagnosis and an experienced observer may in fact be better in distinguishing between normal and glaucomatous disks in comparison with the HRT or OCT. It is important to differentiate normal physiologic variations in the optic cup, the neural rim, and the peripapillary retina, developmental anomalies and nonglaucomatous optic atrophies from glaucomatous optic nerve head changes.

This review attempts to elucidate the morphology and anatomy of the optic nerve head, and correlate the same to the pathophysiology of glaucomatous optic neuropathy.

Keywords: Optic nerve head morphology, Glaucomatous optic neuropathy, Optic disk evaluation.

INTRODUCTION

Glaucoma can be defined as syndrome of progressive optic neuropathy characterized by excavation of the optic nerve head and consequent defects in retinal sensitivity with visual field defects and other psychophysical alterations. Glaucomatous optic neuropathy is characterized by changes in the intrapapillary and parapapillary region of the optic nerve head.1,2 One of the most important tests for determining if a patient has glaucoma is the evaluation of the optic nerve head. Studies have shown that careful evaluation of the optic nerve head has high specificity and good precision for glaucoma diagnosis and an experienced observer may in fact be better in distinguishing between normal and glaucomatous disks as compared to any other technology, like the HRT or OCT.1-3 Normal physiologic variations in the optic cup, the neural rim, and the peripapillary retina, developmental anomalies and nonglaucomatous optic atrophies are the common entities that can be confused with a glaucomatous disk.3-5

In the past few years, significant changes have evolved in the conceptual understanding of the underlying pathogenic mechanisms of primary open-angle glaucoma. The glaucomatous optic neuropathy is thought of as an optic nerve disorder in which IOP is a major risk factor among several others. At one time theories of glaucomatous pathophysiology were considered either “mechanical” or “vasogenic” but glaucomatous optic neuropathy is caused by a combination of several factors, which include a combination of mechanical and vascular factors which set up a cascade of events finally leading to biochemical changes and apoptosis of retinal ganglion cells.

Glaucomatous atrophy is thus caused by a progressive death of retinal ganglion cells, which manifests as characteristic excavation of the optic nerve head with sequential visual field deterioration in characteristic patterns. An in-depth understanding of the anatomy of the optic nerve head (ONH) is crucial for better understanding of pathophysiology of glaucomatous optic neuropathy.

Anatomy of the Optic Nerve Head

Each part of the ONH is made up of axons (nerve fibers) of retinal ganglion cells grouped into bundles, blood vessels and supporting glial tissue. The optic nerve head (ONH) can be divided into four anatomic parts:

a. **Surface layer**: The superficial nerve fiber layer (SNFL) of the ONH has its most anterior limit at the point where the nerve contacts the vitreous. For histopathologic and clinical purposes, the peripheral edge of the nerve is defined by the anterior limits of the scleral ring. The posterior limit of the SNFL is recognized histologically as the point at which the axon bundles have completed their 90° turn from the plane of the retina and have reached the level of the choroids.

b. **Prelaminar part**: The prelaminar portion of the ONH is the indistinct segment of the axons surrounded by the outer retina, choriocapillaris, and choroid; structurally the astroglial component here is considerably increased compared with the SNFL.

c. **Laminar part**: Laminar part of the nerve is contained within the lamina cribrosa; here the glial wrapped axon bundles are confined in the relatively rigid pores of the specialized laminar scleral plates.

d. **Retrolaminar part**: Posterior to lamina cribrosa is the retrolaminar portion of the optic nerve, where its thickness is doubled by the presence of myelinating oligodendrocytes. In the human eye, the distribution of the nerve fibers from the peripheral retina towards the optic nerve is such that axons...
from peripheral ganglion cells remain peripheral as they enter the disk while the central fibers enter centrally, adjacent to the physiologic cup. This topographic arrangement correlates with the clinical progression of the glaucomatous visual field; paracentral scotomas appear early in the disease as the cup enlarges, and the peripheral field remains until the peripheral axons in the nerve are affected.

The arterial blood supply to the ONH may vary among individuals, but there is general agreement about its fundamental components. The central retinal artery (CRA) and the short posterior ciliary arteries (SPCAs) all contribute directly or indirectly to a capillary plexus that supplies the ONH. The venous drainage of the ONH is almost entirely through branches of the central retinal vein, although important choroidal collaterals exist; these collaterals may appear as retinociliary shunts in instances of disturbed retinal circulation. The branches of the CRA supply the SNFL. This is the network responsible for the flame/splinter disk hemorrhages seen clinically, and it is also the vascular bed that appears in fluorescein angiograms of the ONH. The prelaminar ONH is supplied by branches of the SPCAs, which enter the disk substance through the adjacent sclera and posterior to the choroidal bed. Most investigators maintain that vessels derived from the peripapillary choroid make only a minor contribution to the blood supply of anterior part of ONH. The laminar portion is vascularized primarily by centripetal SPCAs, although an anastomotic capillary bed may also contribute to the vascular supply. The anterior portion of the retrolaminar nerve has both centripetal vascular supply from the piameninges and a significant axial vasculature from branches of the CRA.

Techniques of Optic Nerve Head Evaluation

There are various techniques of examination of the optic nerve head like the direct ophthalmoscopy, the most commonly used, the indirect ophthalmoscopy for hazy media, the slit lamp Binocular indirect ophthalmoscopy using the noncontact method (+60, +70, +90 D lenses) or the contact method (Goldmann’s prism).

The identification of structural, contour and color changes of the disk is best done stereoscopically with a dilated pupil. The evaluation of the optic nerve head (ONH) and retinal nerve fiber layer (RNFL) may be divided into two parts:

Qualitative Evaluation
- Contour of the neuroretinal rim
- Optic disk hemorrhages
- Parapapillary atrophy
- Bared circumlinear vessels
- Appearance of the retinal nerve fiber layer.

Quantitative Evaluation
- Optic disk size (vertical disk diameter)
- Cup/disk ratio (vertical)

The ONH can be evaluated by following ways:

Direct ophthalmoscopy: This is the simplest and most commonly used technique for optic disk evaluation. It gives a magnified erect image and is very useful for a quick screening. It lacks stereopsis, which is of great importance in assessing the topography of the disk. The direct ophthalmoscope can give three-dimensional information using parallax movements. It is essential to select a spot size with a diameter smaller than the diameter of the disk. This is to avoid light spreading from the peripapillary retina altering the appearance of the rim. The size of the disk can also be estimated by comparison with the circular small light spot of the ophthalmoscope. The smallest 5° aperture projects to an area of 1.7 mm². 

Indirect Ophthalmoscopy: Though it gives a three-dimensional picture, the magnification is not sufficient for detailed evaluation of the disk specially the changes in the blood vessels, neural rim and disk hemorrhages. It also fails to provide an accurate assessment of disk pallor and not being helpful in constricted pupil. It is useful in an eye with a hazy media.

Slit-lamp biomicroscopy: Slit lamp biomicroscopy is one of the most useful tools to study the ONH. It allows for a time-efficient and detailed stereoscopic examination of the posterior pole by providing both good magnification and stereopsis. It can be done by two ways:
- Contact method with Goldman three mirror lens
- Noncontact method with a Hruby lens or a 78/90 D lens.

A + 78 D lens provides more magnification and a detail while a + 90 D give a wider field and is better in cases with small pupils.

Although a yellow colored lens may be helpful in increasing patient comfort, it may mask some early color changes found in the glaucomatous optic nerve head. For this reason, the high plus lens should be clear. To ensure that the maximum benefit is achieved from the use of the high plus lens, it is crucial that the biomicroscope is appropriately set for the examiner. Even a slight variation from the correct interpupillary distance setting can affect the examiner’s stereopsis. Additionally, the angle between the illumination system and microscope system should be no more than 10° to ensure stereopsis. It is important to remember that the image seen through a high plus lens is a virtual image, and will be inverted with the right on the left, and the top on the bottom.

Being familiar with the normal optic nerve head (Fig. 1) is essential in order to critically examine the nerve for the changes typical of glaucoma. Although there is considerable variability, the normal optic nerve contains approximately one million axons which leave the eye in multiple bundles through the lamina cribrosa. The convergence of the fibers creates a circular depression in the optic nerve head which is known as the cup. The size of the cup is compared to the size of the disk and is
dependent both on the number of nerve fibers leaving the eye, and the size of the disk. Patients with a decrease in the number of nerve fibers leaving the eye, as occurs in the generalized loss of axons in moderate to severe glaucoma; or a larger sized optic disk with all of the axons intact, will both have cup-to-disk (C/D) ratios larger than normal. Equally true, a small disk will have very little cupping even if a loss of axons has occurred. Because of the effect of the nerve size on the cup-to-disk ratio, it is very important to evaluate the optic nerve head size before commenting on the C/D ratio.8,9

Normal Optic Disk Morphology

Optic Nerve Head

The disk area varies from 0.80 to 6.00 mm².5,65,66,67 The optic nerve head is vertically oval with the vertical diameter being more than the horizontal diameter by 7 to 10%.5 The average vertical diameter varies from 1.85 to 1.95 mm (range 0.95-2.9 mm) and the average horizontal diameter varies from 1.70 to 1.80 mm (range 0.9-2.6 mm).10, 11 The ratio between horizontal and vertical disk diameter varies between 0.70 to 1.37. The ONH size is not constant. It shows high inter-individual variability with numerous influencing factors as age, sex, anthropometry, refractive error and race. The disk size becomes constant after 3 to 10 years of age.5,65,66,67 The mean optic disk area is 3.2% larger in males than females66 and the disk size increases by 0.02 mm² for 10 cm increase in body length.68 Recent studies have shown that increasingly elongated optic disks are associated with myopia >12 D,63 corneal astigmatism and amblyopia64 suggesting that if an abnormal disk shape is found in children, a skiascopy should be undertaken to prevent amblyopia. Previous studies had quoted that disk size is independent of refractive error in the range of −5 to +5 D of ametropia5,66 whereas recent investigations have revealed a linear increase in disk area of 1.2% for each 1 D shift towards myopia.67 The size of the disk varies considerably in the normal population and among different races. Africans and Asians have larger disk size as compared to Europeans.

The size of the optic disk can be estimated by using the formula: r/4 × horizontal diameter × vertical diameter; (r is the correction factor). The vertical diameter of the optic disk can be measured at the slit lamp using a contact or a condensing lens. The slit beam should be coaxial with the observation axis; a narrow beam is used to measure the disk height using the white scleral ring as a reference landmark. The magnification corrections needed vary with the optical dimensions of the eye and with the lens used for measurement12 (Table 1).

On the basis of Gaussian-like distribution curve of optic disk area, disks smaller than mean minus twofold standard deviations are classified microdisks (< 1.29 mm²) and optic disks larger than mean plus twofold standard deviation are classified macrodisks (> 4.2 mm²). Microdisks are seen in hyperopia, aniridia, optic nerve hypoplasia, nonarteritic anterior ischemic optic neuropathy, etc. Primary macrodisks are independent of age after first year of life and slightly influenced by refractive error. They may be asymptomatic, e.g. large physiological cup or symptomatic as in optic disk pit (Fig. 21) or Morning Glory disk (Fig. 22). Secondary macrodisks increase in size after birth as in primary and secondary high myopia. Studies have found disk size variability to be pathogenically important. The optic disk size is normal in cases of primary open-angle glaucoma,69,73 Juvenile onset open-angle glaucoma,74 pigmentary glaucoma71 and age related atrophic type glaucoma.72 A study has shown that size of optic disk is smaller in eyes of pseudoexfoliative glaucoma than in eyes of POAG.41 The disk size is significantly larger in eyes with normal pressure glaucoma70 and in glaucomatous eyes with high myopia.

### Table 1: Magnification correction factors for commonly available high plus lenses

<table>
<thead>
<tr>
<th>Type of lens</th>
<th>Magnification correction factors¹²</th>
<th>Manufacturer’s specifications</th>
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</thead>
<tbody>
<tr>
<td>Volk 60D</td>
<td>1.11</td>
<td>1.15</td>
</tr>
<tr>
<td>78D</td>
<td>1.33</td>
<td>1.39</td>
</tr>
<tr>
<td>90D</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nikon 60D</td>
<td>1.03</td>
<td>1.02</td>
</tr>
<tr>
<td>90D</td>
<td>1.63</td>
<td>1.54</td>
</tr>
<tr>
<td>Haag-Streit Goldmann</td>
<td>—</td>
<td>1.14</td>
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</table>
Optic Cup

The optic cup is the central excavation in the optic nerve head (Figs 2 to 4). It lies below the level of neural rim and its bottom is formed by lamina cribrosa. It is usually horizontally oval with a horizontal diameter of 0.83 mm (0-2.08 mm) and a vertical diameter of 0.77 mm (0-2.13 mm) and an area of 0.72 mm² (0-3.41 mm²). The border between the optic cup and the neuroretinal rim is determined by contour and not by pallor. The combination of the horizontally oval shape of the optic cup and the vertically oval shape of the optic disk explains the configuration of the normal neuroretinal rim, which has its broadest parts in the inferior and superior disk regions and its smallest parts in the nasal and temporal region of the optic disk.¹⁰,¹¹

For the evaluation of the optic cup, it is useful to clinically examine the optic nerve head by stereo-optic disk photography or stereoscopic slit-lamp examination. Especially in eyes with shallow disk cupping, such as in highly myopic eyes with glaucoma, location of the kinking of vessels can be helpful for the determination of the border of the optic cup (Fig. 5). In normal eyes, the areas of the optic disk and optic cup are correlated with each other, i.e. the larger the optic disk, the larger the optic cup and vice versa. This feature must be considered in the morphologic diagnosis of glaucoma.¹²,¹³ Early or moderately advanced glaucomatous optic nerve damage may erroneously be overlooked in small optic disks with relatively low cup-to-disk ratios, if one does not consider that small optic disks normally have no optic cup. In contrast, a large optic cup in a large optic disk should not lead to the diagnosis of glaucoma if the other intrapapillary variables are normal, mainly the configuration of the neuroretinal rim. One should see if the ISNT rule is violated or not. Larger cup size, can be physiological if it is bilaterally symmetrical or is associated with large disk size, or high myopia and follows the ISNT rule¹⁴ (Fig. 6).
In addition to its area, the optic cup is ophthalmoscopically described by its depth. In normal eyes, the optic cup depth depends on the cup area and indirectly on the disk size: The larger the optic cup, the deeper it is. In glaucoma, the optic cup deepens depending on the type of glaucoma and the level of IOP.\(^3,11\)

**Shape of Optic Cup**

Shape of optic cup can be of one of the following types (Elschnig)\(^15\):

- Type I: Small funnel shaped
- Type II: Temporal cylindrical
- Type III: Central trough-shaped
- Type IV: Temporal or central with steep nasal wall and sloping temporal margin
- Type V: Developmental anomalies.

**Cup/Disk Ratio**

The cup/disk ratio (CDR) is the decimal value obtained by dividing the cup diameter with the disk diameter. It normally ranges from 0.2 to 0.5. The closer the value is to 1, the worse the damage. The vertical cup/disk ratio is a better measure of deviation from normal than the horizontal ratio, because early neuroretinal rim loss occurs preferentially at the upper and lower poles of the disk. A difference in cup/disk ratio between eyes with equal overall optic disk size is suggestive of tissue loss and therefore is highly suspicious of acquired damage. Expressing the size of a cup as a cup/disk ratio (C/D or CDR) is of limited value unless the actual size of the disk is known.

Because of the vertically oval optic disk and the horizontally oval optic cup, the cup/disk ratios in normal eyes are significantly larger horizontally than vertically. The quotient of the horizontal to vertical cup/disk ratios is usually higher than 1.0. This is important for the diagnosis of glaucoma, in which, in the early to medium advanced stages, the vertical cup/disk diameter ratio increases faster than the horizontal one, leading to an increase of the quotient of horizontal to vertical cup/disk ratios to values lower than 1.0. The diagnosis of glaucoma should be strongly considered if the difference in the cup/disk ratio between the two eyes is more than 0.2 (seen only in 1% of the normal population)\(^10,11\) (Fig. 7) or if the CD ratio is 0.7 or more (seen only in 10% of population).
When determining the amount of cupping, it is very important to evaluate the contour and not the pallor of the cup. This is because the optic nerve head damaged by glaucoma typically has cupping which is larger than the pallor, whereas the normal eye has cupping equal to the area of pallor. Pallor more than the cupping should raise suspicion of a non-glaucomatous cause for optic atrophy (Fig. 8).

Neuroretinal Rim (NRR)

Neuroretinal rim is the area of the bending of the axons from the disk margins to the edge of the optic cup. The evaluation of NRR width is based on the mark of change in contour than on the mark of change in color. The average area of NRR is 1.4 to 2.0 mm² and may decline with age. The NRR is not interindividually constant. The neuroretinal rim size correlates with the optic disk area; the larger the disk, the larger the rim. The correlation between rim area and disk area corresponds with positive correlation between optic disk size, optic nerve fiber count, number and total area of lamina cribrosa pores. It points towards a greater anatomic reserve capacity in eyes with large optic disks as compared to eyes with smaller ones. Because it is recognized that a large cup/disk size is not definitive as a diagnosis of glaucoma, less attention is placed on the size of the cup, and it is more important to focus on the appearance and configuration of the neural rim tissue found between the cup and the edge of the disk. The rim tissue is often the first area to show changes in glaucoma, and must be examined very critically during an optic nerve head evaluation. The normal neuroretinal rim tissue is uniformly pink in color indicating good vascular perfusion (Figs 1 and 2). Because there is a round cup located in a vertically elongated oval optic disk, the width of the neural rim tissue varies by quadrant. In the normal eye, the inferior quadrant has the widest rim tissue with the superior portion second in width. The nasal tissue is slightly thinner than the superior tissue and the tissue in the temporal quadrant is the thinnest (ISNT rule as termed by Werner) (see Fig. 6). This variation in rim sizes causes large physiologic cups to appear elongated horizontally. The rim tissue will thin as nerve fibers atrophy and this result in pallor in the area of atrophy and a decrease in the size of the rim tissue over time. If the nerve fiber loss is generalized, the atrophy of nerve fibers will cause an overall decrease in the width of rim tissue and an increase in the size of the cup. This generalized atrophy is typical in moderate to advanced glaucoma, with corresponding visual field loss. Because these changes are obvious only in the later stages of the condition, the increase in cup size is not very helpful in making a diagnosis of glaucoma early in the disease process.

In glaucoma, neuroretinal rim is lost in all sectors of the optic disk with regional preferences, depending on the stage of the disease. In early glaucoma, the inferior rim is usually affected first, with the superior rim a close second. The next tissue to be damaged is typically the temporal rim, with the nasal rim being the last to be affected. Thinning in one focal area of the disk can cause a “notch” to develop in the rim tissue over time. Since the inferior and superior rim tissues are affected first, notching is typically seen in one of these quadrants (Figs 9 to 10B). When evaluating the optic nerve, it is helpful to have the results of a visual field test performed on the same day readily available. This allows the comparison of areas of potential visual field defects to the nerve fiber responsible for that area of the field. It is estimated that 20% of the nerve fibers must be atrophied to cause a visual field defect of 5 dB and 40% to cause a 10 dB loss. Because of this, visual field results are best interpreted when used in conjunction with the optic nerve head and nerve fiber layer evaluation.

An oblique insertion of optic nerve head as seen in myopia, and occasionally a gray crescent in the optic nerve area, may obscure the view of NRR thus falsely mimicking NRR thinning. Color of NRR can be misinterpreted due to presence of nuclear sclerosis and use of coated +90 D lens.

Fig. 8: Note that the pallor is out of proportion to the cupping suggesting a nonglaucomatous etiology

Fig. 9: The optic nerve head with CD ratio of 0.8:1, inferior notch with thinning of neuroretinal rim inferiorly, bayoneting, baring of the circumlinear vessels
Rim/Disk Ratio (RDR)

It is the fractional decimal value obtained by dividing the rim thickness by the disk diameter. The closer the value is to 1, the better the optic disk appearance. It can be calculated as vertical diameters as for the cup/disk ratio but obviously with the opposite meaning, as rim area/disk area ratio. This latter can also be calculated for each degree of the optic disk as a sector index of a healthy disk.

Peripapillary Region

Peripapillary region is divided into an outer alpha and an inner beta zone (Figs 12A and B).22,23

The alpha zone forms the outer crescent characterized by irregular hypo- and hyperpigmentation representing the alteration in the distribution of the melanin pigment in the RPE. It is common finding in normal eyes.

The beta zone lies adjacent to the disk and is characterized by visible sclera, choroidal vessels and a total loss of pigment epithelium. This zone is more common in eyes with glaucoma.

In normal eyes, both the alpha zone and beta zone are largest and most frequently located in the temporal horizontal sector, followed by the inferior temporal area and the superior temporal region. They are smallest and most rarely found in the nasal parapapillary area.
Both zones are significantly larger and the beta zone occurs more often in eyes with glaucomatous optic nerve atrophy than in normal eyes. Size of both zones and frequency of the beta zone is significantly correlated with variables indicating the severity of the glaucomatous optic nerve damage, such as neuroretinal rim loss, decrease of retinal vessel diameter, reduced visibility of the retinal nerve fiber bundles, and perimetric defects. In eyes with small cup-to-disk ratios, the appearance of peripapillary atrophy may be a more sensitive indicator of glaucomatous optic nerve damage than cup-to-disk ratios. The appearance of peripapillary atrophy should raise the suspicion of glaucoma, and be used in conjunction with other test results when making clinical decisions on the diagnosis and management of glaucoma.

Retinal Nerve Fiber Layer Height (RNFLH)

The thickness of the RNFL depends on disk area, age, stage of the glaucomatous damage. The vertical polar sectors were thicker than nasal and temporal sectors as the nerve fibers arcuate around the macula and are concentrated at the two poles.

Morphology of the Glaucomatous Optic Disk

Changes in Optic Cup: Glaucoma results in loss of the retinal nerve fibers which manifests as the changes in the optic cup.3,11,21-24

1. Increase in the size of the cup (see Figs 3A to 5)
2. Increase in CD ratio (see Figs 3A to 5)
3. Vertical enlargement of cup due to localized loss of nerve fibers at the superior and inferior poles. Focal loss of NRR results in formation of a polar or a focal notch seen more commonly at inferior than at superior pole (Fig. 9)
4. Asymmetry between the two eye of more than 0.2 in the CD ratio (see Fig. 7)
5. Increase in depth of the cup
6. Diskrepancy in pallor/cupping usually the pallor at the disk is confined to the area of the physiological cupping, but as the glaucoma advances the cupping may progress ahead of the area pallor. Enlargement of the cup in such cases is evident by the kinking of the vessels at the cup margin. Initial enlargement may lead to a shallow cupping with sloping margins extending (saucerization) up to the disk margins but the NRR in may retain its normal color (tinted hollow), thus the area of pallor appears smaller than the area of cupping.

Changes in Neuroretinal Rim

Loss of NRR and decrease in NRR area is seen in glaucoma.3,11,16-19 The loss of neural rim can be either localized or diffuse. Both of these precede development of visual field defects. Diffuse loss results in concentric increase in the cup size and is more common. Localized loss results in formation of a focal notch more common at the inferior than at the superior pole (see Fig. 9). Sometimes it may be confused with a congenital pit of the optic disk. In advanced cases total loss of NRR occurs with extreme posterior bowing of the lamina (bean pot sign) (see Fig. 10).

Vascular Changes

1. Nasalization of vessels: Normally the retinal vessels enter the eye along the nasal border of the disk and their branches run along the margin of disk and cup and emerge somewhat temporally. With enlargement of the optic cup the major vessels may show a further nasal shift (Fig. 13), although, this is not specific for glaucoma.

2. Bayoneting of vessels: With advancement of the cupping, the vessels emerge from the floor of the cup, ascend up the steep wall of the cup under the overhanging edge of the cup (at which time they are not visible to the observer) and then emerge again at the disk margin pass making a sharp bend (that resemble bayonet of a rifle), may disappear and then emerge again at the disk margin (Fig. 14). This is specific for glaucomatous cupping.3,11,28

3. Over pass cupping: Normally the vessels run over the surface of the disk and NRR and then come out. A loss of NRR takes away their posterior support and they appear to hang over the disk, and bridging the cup which is known as over pass cupping.3,11,28

4. Baring of circumlinear vessels: Circumlinear vessels are the small branches arising from retinal vessels, seen in 50% of the normal eyes. These vessels follow a curvilinear path and run along the superior and inferior margins of the optic

![Fig. 13: Advanced glaucomatous optic atrophy with nasalization of the vessels](image-url)
Disk Hemorrhages

Disk hemorrhages in glaucoma were reported by Drance and Begg. The prevalence of small hemorrhages related to the optic disk has been estimated at 0 to 0.21% in the normal population and 2.2 to 4.1% in glaucomatous patients; they may be more common in normal-tension glaucoma (up to 40%). Since the prevalence of disk hemorrhage is low in the normal population, their presence is very likely to be pathological, especially if recurring. It is a sign of local vascular damage. The characteristic features of these hemorrhages are:

1. Hemorrhages typically appear blot-like when located on the disk, and flame or splinter shaped if they are in close proximity to the disk in the nerve fiber layer. Splinter hemorrhages are more common than blot ones (Figs 16A and B).
2. Inferotemporal location is most common.
3. It is a sign of progressive disease, may lead to nerve fiber layer defect, focal notching of NRR and progression of visual field defect.
4. Splinter hemorrhages have been shown to precede nerve fiber layer and visual field changes in some patients.
5. Although the hemorrhages can resolve in as short as 2 weeks or as long as 35 weeks, the average time to resolution is 10 weeks.

Baring of circumlinear vessels occurs with enlargement of the cup as an area of pallor appears between the cup margin and these vessels (Figs 15A and B). As the rim narrows the loss of tissue leaves this vessel isolated or ‘bared’. It may then remain superficial or come to lie on the inner slope of the rim or on the cup floor. Acquired bearing of circumlinear vessels is an early sign of rim thinning and thus diagnostic of glaucoma.
6. When such a hemorrhage is found in a patient who has already been diagnosed, and is being treated for glaucoma, it indicates an unfavorable prognosis, and the need for more aggressive therapy with resetting of the target IOP to a lower level.

7. More common in patients with large IOP variations.

8. More common in diabetics and hypertensives.

9. Higher association with normal tension glaucoma

In all cases with disk hemorrhages, other causes leading to disk hemorrhages like systemic anticoagulants, blood disorders and microvascular disease should be ruled out.30-33

Retinal Arterioles/Vessel Changes

Narrowing of the retinal vessels (diffuse or focal) may occur secondary to the loss of nerve fiber layer in cases with glaucoma. This finding is also found in eyes with nonglaucomatous optic disk damage, such as nonarteritic anterior optic neuropathy and descending optic nerve atrophy indicating that reduction in vessel caliber is typical to optic nerve damage and not to glaucoma. Appearance of collateral vessels on disk may occur in glaucoma but it is not specific for glaucoma.3,11

Patterns of Optic Nerve Head Changes in Glaucoma

A variety of classification schemes have been proposed to clinically distinguish subtypes of glaucoma based on the appearance of the disk but they are not universally accepted. Also there is considerable overlap of features as they appear in clinical practice. The ONH glaucoma patterns has been classified into following five patterns34-45 (Table 2).

<table>
<thead>
<tr>
<th>High Myopia Disk Pattern</th>
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<tr>
<td>It is seen in cases with high myopia and open-angle glaucoma (Fig. 5). They have larger and often abnormally shaped optic disks and their diagnosis represents a special problem in the management of glaucoma. Many myopic eyes have lost considerable vision from primary or secondary glaucoma before the ophthalmologist becomes aware of the diagnosis. The reasons for this difficulty are:</td>
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<td>a. The distance between the level of the lamina cribrosa and the level of the retina is much less than in normal or hyperopic eyes. The average value of this distance in the normal eye is about 0.7 mm, whereas that of the myopic eye is between 0.2 and 0.5 mm. Therefore, a completely cupped disk in a myopic eye will have only half the depth of the usual glaucomatous cup and therefore such a shallow excavation is difficult to appreciate clinically.</td>
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<tr>
<td>b. The myopic ONH is masked by the usual myopic conus, tilting of the disk, and peripapillary atrophy. In such cases, disk photographs are superior to drawings for monitoring of the subtle progression of the shallow cup with associated shifts in vessels or changes in peripapillary area.</td>
</tr>
<tr>
<td>c. The ocular rigidity usually is lower than that of normal eyes. Therefore, Schiotz tensions, using the ordinary conversion tables, are lower than the actual IOP. Hence, applanation tonometry should be used in these cases. These patients may have associated thin corneas causing falsely lower applanation IOP readings.</td>
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<tr>
<td>d. Visual field interpretation is difficult due to presence of associated retinal pathology in some cases and the presence of refractive scotomas.</td>
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Table 2: Subtypes of glaucoma by optic nerve head appearance

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<thead>
<tr>
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<th>High Myopia Disk Pattern</th>
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<tr>
<td>High myope</td>
<td>Focal ischemic</td>
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<td>Age and Sex</td>
<td>Age related</td>
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<td>Optic disk shape and size</td>
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<td>Optic cupping</td>
<td>POAG</td>
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<tr>
<td>Optic cupping</td>
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<tr>
<td>Disk hemorrhages or rim notches</td>
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<tr>
<td>Disk hemorrhages or rim notches</td>
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<tr>
<td>Optic cupping</td>
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<td>Focal RNFL defects</td>
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<td>Focal RNFL defects</td>
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<td>Visual field changes</td>
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<td>Visual field changes</td>
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<td>Peripapillary changes</td>
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<td>Peripapillary changes</td>
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<tr>
<td>IOP</td>
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<tr>
<td>Associated systemic abnormalities</td>
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Focal Normal-Pressure Pattern (Focal Ischemic)

Eyes with the focal type of normal-pressure glaucoma have normally sized and shaped optic disks, but with characteristic cupping. Often there is a steep and distinct edge to the cup, with the deep cup remaining visible as it vertically progresses to manifest rim notches, disk hemorrhages, and focal RNFL wedge defects. Despite the polar notching, often the remainder of the rim tissue remains relatively intact. The clinical associations for this disk appearance include a higher frequency among women, scotomas near fixation in the superior visual field, and a positive history for migraine headaches. These cases have nonspecific circulatory abnormalities in orbital circulation on color Doppler analysis. The inclusion of the term ischemic is based on the clinical impression of frequent disk hemorrhages in these eyes, which rarely demonstrate highly elevated IOPs.

Age-Related Atrophic Primary Open-Angle Glaucoma Pattern (Senile Sclerotic)

Eyes with age-related atrophic primary open-angle glaucoma are often associated with diffuse fundus changes described as both choroidal sclerosis and tessellated fundus (Fig. 17). These findings in combination with the association with older age, render the term “age related” or “senile sclerotic.” The disks are of normal size and shape, but peripapillary atrophy is a prominent finding, so the edges of the ONH must be carefully distinguished. The cupping is described as shallow and saucerized, even moth-eaten. As the concentric cupping progresses, the neural rim may nevertheless retain a pale but almost normal color. This anomaly results in a diskrepancy between the “color-cup,” as estimated on the basis of pallor (often with the monocular view of the direct ophthalmoscope), and the actual geometric, larger shape of the cup, as determined by high-magnification stereoscopic disk evaluation. The clinical appreciation of such color/cup diskrepancy is important and often explains why visual field loss appears to be greater than the optic disk changes.

The changes, such as polar notching, disk hemorrhages or wedge RNFL defects, are uncommon in this form of age-related atrophic cupping. Often the fundus appearance is striking in its mosaic, or tessellated, pattern of prominent choroidal vessels and mottled orange background. Other clinical associations are a mild to moderately elevated IOP, more generalized vascular disease (systemic hypertension and ischemic heart disease) and visual field loss that was often diffuse.

Juvenile Open-Angle Glaucoma Pattern

Disk findings in younger patients with open angles, with or without discrete angle anomalies, share features in common with disks subjected to sustained elevated IOPs from secondary causes. The nerves are of normal size and shape, and thus differ from myopic disks. The cupping is often distinctively steep-edged and deep, exposing the laminar pores and struts, and it enlarges in a concentric pattern. Focal changes, such as disk hemorrhages, wedge RNFL defects, and signs of peripapillary atrophy are infrequent.

Primary Open-Angle Glaucoma Pattern (Generalized Enlargement)

A distinctive category of disk appearance that is commonly seen in POAG with high IOPs includes a disk of normal size
with diffusely enlarged round cups (Figs 18A and B). Localized rim defects are uncommon, so abnormality or progression of cup enlargement necessitates comparison with the fellow eye or with serial photographs or drawings. The cup increase is often biased toward the temporal rim, with gradual attenuation of the neural rim. Usually the secondary forms of open-angle glaucoma, such as pseudoexfoliation and pigmentary dispersion, manifest similar disk pattern changes.

**Retinal Nerve Fiber Layer**

The importance of Retinal nerve fiber layer defects was first reported in 1973 by Hoyt et al. Evaluation of the nerve fiber layer is another useful tool to aid in the early diagnosis of glaucoma. This is because nerve fiber layer defects can occur before disk changes and visual field changes are documented or found.\textsuperscript{46-49} Retinal nerve fiber layer (RNFL) loss is thus the earliest sign of glaucoma. RNFL thickness is 200 microns near the disk, 60 microns in the area of papillomacular bundle and 40 microns at rest of the places. The nerve fiber layer (NFL) is best seen with a 78 D or 90 D lens or a contact lens at the slit lamp. RNFL is best examined in a red free light, because this light does not penetrate beyond the RNFL and is reflected back (Figs 19A and B). In the areas of RFNL loss, the light get absorbed by the RPE, thus a contrast is created between the normal and the degenerated area (Fig. 20). Red free light, which is absorbed by the pigment of the retinal pigment epithelium and the choroid, is therefore used to provide a dark background. The normal nerve fiber layer reflects light and appears as a whitish haze over the darker underlying retinal structures. There will be a striated appearance to the nerve fibers (the fiber bundles are seen as silver striations), with thicker nerve fiber layers appearing brighter. From about two disks diameters from the disk, the NFL thins and feathers out. Slit-like, groove-like or spindle-shaped apparent defects, narrower than the retinal vessels, are seen in the normal fundus. The NFL becomes less visible with age, and is more difficult to see in lightly pigmented fundi. Because the nerve fiber layer is thickest in the superior and inferior arcades closest to the disk, this area should be the brightest portion of the view. There will be less brightness in the thinner papillomacular region and the nasal side of the disk. Symmetry between the reflections in the superior and inferior arcades and between each of the patient’s eyes is expected.

**Patterns of RFNL defect:** Experimental studies have shown that the defects can be picked if 50% or more of the RNFL is lost. This is due to the sandwich pattern of NFL bundles. The first glaucomatous axons to be lost are from the temporal raphe which are in the middle and deep layers of the retina. Following types of NFL defects may be seen:\textsuperscript{3,11,46-49}

1. **Slit defects:** Dark areas which are slightly larger than arterioles and reach the disk following the normal course of the nerve fiber layer are called slit defects. They represent retrograde degeneration of the axons due to focal damage
of the optic nerve at the lamina. These can occur in approximately 10% of normal patients. Slit-like defects are difficult to identify and may be confused with the normal healthy grooves seen in normal RNFL.

2. **Wedge defects**: Wedge defects are caused by atrophy of many ganglion cells in the same area of the optic nerve. These defects start at the disk as narrow lines and expand as they get further from the disk. Notching of the neural rim tissue, as well as a visual field defect are often associated with wedge defects. Wedge shaped defect are usually seen in superior and inferior poles. They are easily detectable as compared to slit defects and may be preceded by appearance of a splinter hemorrhage in the same site.

3. **Diffuse loss**: Diffuse loss is the commonest type of RFNL loss seen in glaucoma, but is difficult to pick up. The diffuse atrophy typically occurs in the superior and inferior arcades. The nerve fiber layer in these areas loses its consistency and looks like it has been combed or raked with darker and lighter areas.

4. **Nerve fiber layer reversal**: In severe cases nerve fiber layer reversal can occur in which the normal pattern of superior and inferior brightness with increasing dimness towards the papillomacular bundle is lost and the papillomacular area becomes the brightest structure. Nerve fiber layer reversal is associated with thinning of the neural rim and a diffuse depression or constriction of the visual field.

5. **A combination of localized and diffuse loss**: RNFL defects are best seen within two disk diameters of the disk. Wedge and slit defects (wider than retinal vessels) are more apparent in early disease, when there is little generalized thickening of the NFL, and are seen as dark bands extending from the optic disk. Generalized thickening of the NFL, with a loss of brightness and density of striations, is a difficult sign to confirm objectively. When the NFL is thinned out, the blood vessel walls are sharp and the vessel appear to stand out in relief against a matt background. The initial abnormality in glaucoma may be either diffuse thinning or localized defects. Since the prevalence of true NFL defects is < 3% in the normal population, their presence is very likely to be pathological.

The various changes in optic nerve head in cases of glaucoma are summarized in Table 3.

### Recording of ONH Features

Color disk photos are useful for patient documentation. Color photography with a 15° field gives optimal magnification. Stereoscopic photographs are the preferred method. Pseudo-stereoscopic photos are also acceptable. Drawings are better than nothing, if a fundus camera is not available.

### Recording of the Nerve Fiber Layer (NFL) Features

The photographic methods require specialized processing of film. Patients must have clear media. Photography of lightly colored fundi are more difficult. The technique is available in some centres, though their use in routine clinical work is limited.

New systems for ONH and NFL assessment, using alternative technologies, are being evaluated for reproducibility, specificity and sensitivity, although currently very few are available to the general ophthalmologist in developing countries due to their prohibitive cost. These include confocal scanning laser ophthalmoscopy (e.g. Heidelberg Retinal Tomograph), scanning laser polarimetry (e.g. GDx), optical coherence tomography (e.g. OCT) and retinal thickness analyzer (e.g. RTA).

### Early or Preperimetric Diagnosis of Glaucomatous Optic Nerve Damage

For the early detection of glaucomatous optic nerve damage in ocular hypertensive eyes before the development of visual field loss, the most important variables are shape of the neuroretinal rim, size of the optic cup in relation to the size of

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**Table 3: Summary of the optic nerve head and retinal nerve fiber changes in glaucoma**

<table>
<thead>
<tr>
<th>I. Alterations of the cup and neural rim</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Enlargement of the cup</td>
</tr>
<tr>
<td>1. Increased cup size (centric or focal)</td>
</tr>
<tr>
<td>2. Increased cup-disk ratio</td>
</tr>
<tr>
<td>3. Alteration in cup shape (vertical-horizontal disproportion)</td>
</tr>
<tr>
<td>4. Asymmetry of cup size between the two eyes</td>
</tr>
<tr>
<td>5. Changes in position and appearance of lamina cribrosa</td>
</tr>
<tr>
<td>a. Baring of the lamina</td>
</tr>
<tr>
<td>b. Backward bowing of the lamina</td>
</tr>
<tr>
<td>c. Slit-like laminar openings</td>
</tr>
<tr>
<td>B. Loss of neural rim</td>
</tr>
<tr>
<td>1. Localized</td>
</tr>
<tr>
<td>2. Diffuse</td>
</tr>
<tr>
<td>3. Change in normal topographic configuration</td>
</tr>
<tr>
<td>(selective narrowing in inferior and superior quadrants)</td>
</tr>
<tr>
<td>a. Increased central area of pallor</td>
</tr>
<tr>
<td>b. Pallor of the neural rim</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>II. Vascular alterations</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Changes in vessel configuration and caliber</td>
</tr>
<tr>
<td>1. Nasalization</td>
</tr>
<tr>
<td>2. Bayonetizing</td>
</tr>
<tr>
<td>3. Overpass vessel</td>
</tr>
<tr>
<td>4. Circumlinear and cilio-retinal vessel baring</td>
</tr>
<tr>
<td>5. Narrowing of retinal vessels</td>
</tr>
<tr>
<td>B. Disc hemorrhage</td>
</tr>
<tr>
<td>C. Collateral vessels on disk</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>III. Peripapillary atrophic changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Increased area of peripapillary atrophy</td>
</tr>
<tr>
<td>B. Increased frequency of more profound (zone beta) type of peripapillary atrophy</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>IV. Loss of the retinal nerve fiber layer</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Localized loss</td>
</tr>
<tr>
<td>B. Diffuse loss</td>
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</tbody>
</table>
the optic disk, diffusely or segmentally decreased visibility of the RNFL, and occurrence of localized RNFL defects and disk hemorrhages. If the rim is not markedly broader in the inferior and superior disk regions as compared with the temporal disk region, a glaucomatous loss of rim tissue may be suspected in the inferior and superior disk regions. In other words, if the neuroretinal rim is more or less even in width in all disk sectors, glaucomatous optic nerve damage can be suspected. In the evaluation of the shape of the neuroretinal rim in glaucomatous eyes, one must account for the fact that the rim configuration depends on the distance to the exit of the central retinal vessel trunk on the lamina cribrosa surface (in glaucomatous eyes with the vessel trunk abnormally exiting in the superotemporal quadrant, the neuroretinal rim is often smallest in the inferonasal region). In eyes with small disks, the neuroretinal rim cannot clearly be delineated from the optic cup, thus, the shape of the rim cannot be clearly determined. In these eyes, the variable “cup size in relation to disk size” is the most important intrapapillary factor to detect glaucomatous optic nerve damage cup.

Martus et al evaluated whether various types of chronic open-angle glucomas differ in predictive factors for progression of glaucomatous optic nerve damage. For patients with elevated intraocular pressure, significantly predictive factors for eventual progression were older age, advanced perimetric damage, smaller neuroretinal rim, and larger area of beta zone of parapapillary atrophy. In contrast, in the normal intraocular pressure group, a significant predictive factor was presence of disk hemorrhages at baseline.

The diagnostic power of a novel digital stereoscopic imaging system in the diagnosis of glaucomatous optic neuropathy was studied by Morgan et al. A prospective cross-sectional analysis of the diagnostic accuracy of digital stereoscopic optic disk analysis in the diagnosis of glaucomatous optic neuropathy exhibiting mild to moderate field loss was done by three observers. With subjective stereoscopic analysis, sensitivity for glaucoma detection among the three observers was 80.8, 76.9, and 90.4%, with respective specificities of 94.4, 79.6 and 79.6%. Regression analysis of the NRR in 30° segments gave sensitivities between 69.2 and 80.8% and specificities between 83.3 and 90.7%. A combination of the subjective and quantitative analysis did not significantly improve discrimination. According to them the subjective analysis of digital stereoscopic images provides a useful method for the discrimination of normal and glaucomatous optic nerves. Planimetric analysis does not significantly improve the diagnostic precision of this technique.

Jonas reviewed the clinical implications of peripapillary atrophy in glaucoma. Recent studies showed an association of peripapillary atrophy with glaucoma and the eventual development of glaucomatous disk hemorrhages independent of a small neuroretinal rim area, and an association between increasing peripapillary atrophy and progressive glaucoma. A ranking of optic disk parameters to detect glaucomatous damage revealed that the alpha and beta zones of peripapillary atrophy, compared with neuroretinal rim parameters, are less useful. Pseudoexfoliation syndrome without glaucoma is not a risk factor for peripapillary atrophy. In arteritic anterior ischemic optic neuropathy, peripapillary atrophy does not enlarge. He summarized that peripapillary chorioretinal atrophy is one among several morphologic variables to detect glaucomatous abnormalities. Ranking optic disk variables for the detection of glaucomatous optic nerve damage, peripapillary atrophy is a variable of second order. It is useful for the differentiation of various types of chronic open-angle glucomas. In contrast to glaucomatous eyes, eyes with nonglaucomatous optic nerve atrophy, including eyes after arteritic anterior ischemic optic neuropathy, do not show enlarged peripapillary atrophy.

**Predictive Value of Nerve Head Evaluation for Glaucoma**

If done properly, evaluation of the optic nerve head and nerve fiber layer are very valuable methods to aid clinician in early diagnosis of glaucoma. Tielsch reported the specificity and sensitivity of a vertical cup-disk ratio greater than 0.5 as 98% and 29%, respectively. Airaksinen et al found evaluation of nerve fiber layer photographs had a specificity of 83% and a sensitivity of 94% for glaucoma detection. Balazsi and Werner reported baring of a circumlinear vessel had a specificity of 94% and a sensitivity of 65% when the vessel was present.

More recently investigators have begun to evaluate the various features of the optic disk and nerve fiber layer in relation to each other to find which signs or combinations of signs best discriminate between glaucomatous and normal discs.

Jonas et al have studied the qualitative characteristics of certain optic disk features in normal and glaucomatous eyes. The features with the highest diagnostic accuracy were the narrowest neural rim outside the temporal sector, the area of cupping greater than the area of pallor, detectable retinal nerve fiber layer loss, and a large area of peripapillary atrophy. Certain features, such as disk hemorrhage, baring of a cilioretinal vessel, and the overlap vessel sign, which are occasionally seen in glaucoma but almost never seen in normal individuals, had, as expected, very high specificities but very low sensitivities.

In studies on very large and very small optic disks, where the size of the cup and the cup-disk ratio can be very misleading, the following features were most useful in distinguishing normal from glaucomatous disks: Vertically oval cup, selective thinning of the inferior neural rim (violation of the ISNT rule), large area of peripapillary atrophy with presence of visible inner zone beta, and nerve fiber layer dropout.

Caprioli et al have reported several studies using optic disk and nerve fiber layer analysis to distinguish normal from glaucomatous disks. When utilizing computerized quantitative image analysis techniques, analysis of features of the retinal nerve fiber layer generally gave the best overall levels of
specificity and sensitivity. Qualitative evaluation of the disk and nerve fiber layer by experienced examiners, however, outperformed the image analysis system.\textsuperscript{58-60}

Montgomery evaluated neural rim area and disk area using an ophthalmoscopic technique. When corrected for disk area, neural rim measurement achieved a specificity of 95\% and a sensitivity of 91\%.\textsuperscript{61}

**Staging and Quantification of Optic Nerve Head Damage in Glaucoma**

Staging and quantification is necessary once optic nerve head changes have been noticed. Categorizing patients according to severity is important in giving them prognosis, monitoring progress deciding management and counseling patients.\textsuperscript{62}

In 1960 Armaly devised the first methodology for quantitatively evaluating disk damage. The method received world wide acceptance and is still commonly used today.\textsuperscript{75,76} The examiner compared the cup diameter to the entire disk diameter in any axis and expressed it as a ratio.

**Read-Spaeth system:** The system was described in 1974 and was also based upon the cup/disk ratio. The severity of disk damage was classified into six stages.

**Richardson system:** The classification system included optic disk changes and visual fields.

*Stage 1a:* Low-risk subject; normal visual fields and cup/disk ratio ($<0.3$ and pink rim of uniform width without asymmetry).

*Stage 1b:* High-risk subject; 1a with family history of glaucoma, vascular disease, pseudoexfoliation, pigment dispersion or large cups.

*Stage 2:* Early glaucomatous damage; incomplete Bjerrum defect or nasal step with cup disk alterations (cup/disk ratio $>0.3$ with vertical widening of cup, asymmetry or neuroretinal rim, disk hemorrhage).

*Stage 3:* Late stage glaucoma; arcuate scotoma with cup/disk ratio $<0.8$, pale rim of uneven width.

*Stage 4:* End stage glaucoma; central or temporal visual island with narrow pale rim.

The above systems have two major shortcomings as cup/disk ratios are not highly valid indicators of health and disease of optic nerve. The systems assume that that cups start centrally and progress concentrically. Although this occurs in some cases, the nerve damage frequently occurs eccentrically. The second problem is that the above systems do not take into consideration the disk size. It is now well-known that the size of the cup varies with the size of the disk with lower cup/disk ratios still being significant in small sized disks.

Read and Spaeth for the first time brought attention towards measuring rim width; they noted that onset of visual field loss was related to remaining rim width. This study formed the basis for later staging systems like the Nesterov’s system, Jonas method and disk damage likelihood scale:

The scale is the latest entry to the list of methodologies for the staging of optic nerve damage. It was devised by Spaeth et al. The scale divides disk damage into 10 grades of severity. It is better able to monitor disease progression than the other scales. Disk drawings are made after a slit lamp biomicroscopic examination and the size of the disk is measured by comparing to the beam length. The DDLS score is derived from the DDLS chart.

DDLS scores of 1 through 3 are rarely associated with glaucomatous visual field loss. Some individuals are born with DDLS three optic disks, whereas others begin with DDLS one disk. For this reason, noting that a person has a DDLS three optic disks indicates that it is reasonably healthy and that there is no visual field loss. This score is not proof that the disk’s health has not worsened, however, because it could have been a stage 1 or 2 in the past. The DDLS allows you to quantify the amount of damage that the optic nerve has sustained. Visual field loss usually will not occur before stage 5. The differentiation between very early and no damage is important, because a neuroretinal rim that has already narrowed is likely to become narrower still, whereas an undamaged rim is far more likely to remain stable. Unless glaucomatous progression has stabilized (e.g. in cases of inactive glaucoma secondary to trauma or corticosteroids), a DDLS score of 6 through 10 strongly supports
aggressive treatment. The DDLS grading performs well compared to C/D ratio and HRT-II evaluation.\textsuperscript{77}

**DISK DAMAGE LIKELIHOOD SCALE**

| Narrowest width of rim (rim to disk ratio) |
|---|---|
| **DDLS stage** | For small disk < 1.50 mm | For average disk 1.50-2.00 mm | For large disk > 2.00 mm | **DDLS stage** |
| 1 | 0.5 or more | 0.4 or more | 0.3 or more | 0a |
| 2 | 0.4 to 0.49 | 0.3 to 0.39 | 0.2 to 0.29 | 0b |
| 3 | 0.3 to 0.39 | 0.2 to 0.29 | 0.1 to 0.19 | 1 |
| 4 | 0.2 to 0.29 | 0.1 to 0.19 | Less than 0.1 | 2 |
| 5 | 0.1 to 0.19 | Less than 0.1 | 0 for less than 45° | 3 |
| 6 | Less than 0.1 | 0 for less than 45° | 0 for 46° | 4 |
| 7 | 0 for less than 45° | 0 for 46° to 90° | 0 for 91° to 180° | 5 |
| 8 | 0 for 46° to 90° | 0 for 91° to 180° | 0 for more than 180° | 6 |
| 9 | 0 for 91° to 180° | 0 for more than 180° | 0 for more than 270° | 7a |
| 10 | 0 for more than 180° | 0 for more than 270° | Not suggestive | 7b |

**Table 4:** Differentiation of glaucomatous from nonglaucomatous optic atrophy in presence of optic disk cupping

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Glaucomatous optic nerve changes</th>
<th>Neurological optic nerve changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual acuity</td>
<td>Affected late</td>
<td>Affected early</td>
</tr>
<tr>
<td>Color vision defect</td>
<td>Normal</td>
<td>Abnormal</td>
</tr>
<tr>
<td>RAPD</td>
<td>Absent unless unilateral advanced involvement</td>
<td>Present in unilateral cases</td>
</tr>
<tr>
<td>Rim changes</td>
<td>Rim defects present</td>
<td>Rim pallor</td>
</tr>
<tr>
<td>Disk field match</td>
<td>Present</td>
<td>Disk field mismatch</td>
</tr>
<tr>
<td>Visual field changes</td>
<td>Respects horizontal meridian</td>
<td>Respects vertical meridian</td>
</tr>
<tr>
<td>Disk field match</td>
<td>Early cases – arcuate scotomas</td>
<td>Early cases – central scotomas</td>
</tr>
<tr>
<td>Peripapillary changes</td>
<td>Match with the disk</td>
<td>Do not match with the disk</td>
</tr>
<tr>
<td></td>
<td>Suggestive of glaucomatous damage be present</td>
<td>Not suggestive</td>
</tr>
</tbody>
</table>

**Differentiation of Glaucomatous vs Nonglaucomatous Optic Neuropathy**

Glaucomatous and nonglaucomatous optic neuropathy may be difficult to distinguish and both can be associated with cupped disks along with a decreased diameter of the retinal arterioles, focal arteriole narrowing, and a reduced visibility of the RNFL.\textsuperscript{3,11} Increasing excavation and enlargement of the optic cup occurs most commonly in glaucoma, but can occur in arteritic anterior ischemic optic neuropathy and compressive lesions on the optic nerve, such as sphenoid wing meningioma. However, in these last two cases, the neuroretinal rim typically will have pallor whereas glaucoma will not. Localized RNFL defects can be found in glaucoma and in many types of nonglaucomatous optic nerve damage, such as in optic disk drusen and longstanding papilledema. Compared with nonglaucomatous optic nerve atrophy, the optic cup enlarges and deepens in glaucomatous optic neuropathy, and, in a complementary manner, the neuroretinal rim decreases. In addition to glaucoma, an enlargement of the optic cup and a loss of neuroretinal rim may be found in patients after arteritic anterior ischemic optic neuropathy and in a few patients with intrasellar or suprasellar tumors. Because parapapillary atrophy does not usually occur in eyes with nonglaucomatous optic nerve damage, it is helpful for the differentiation of glaucomatous versus nonglaucomatous optic neuropathy (Table 4).

In conclusion, a detailed evaluation of the optic disk and retinal fiber layer by stereoscopic slit lamp biomicroscopic techniques provides the clinician with an excellent method for early detection of glaucoma and in monitoring its progression. Annual disk photographs (both color and red free) should be made the standard practice pattern for follow up of a glaucoma patient.

**REFERENCES**

Clinical Evaluation of Optic Nerve Head in Glaucoma


