

Optic nerve evaluation



❧ Glaucoma is defined as *heterogeneous group of disorders which manifests as chronic progressive optic neuropathy characterized by specific morphological changes at optic nerve head and retinal nerve fibre layer with resultant loss of retinal ganglion cells which results in loss of visual fields.*

GLAUCOMA

NO PAIN. NO SYMPTOMS. NO WONDER IT'S CALLED
THE SILENT THIEF OF SIGHT.



For many years , the diagnosis of glaucoma was based on :

1. raised intraocular pressure
2. visual field changes
3. optic nerve head evaluation



Intraocular
pressure

Optic disc
evaluation

Glaucoma
diagnosis

Field defects



- ❧ Raised IOP is not a diagnostic criteria but a risk factor for glaucoma and patients with normal IOP may develop glaucomatous disc changes.
- ❧ It has been proved that 20-40% of ganglion cells are lost before visual field defects develop in patients of glaucoma on standard automated perimetry.

Pre perimetric glaucoma



- ❧ Pre perimetric glaucoma is a new terminology to describe glaucomatous optic neuropathy before development of visual field defects.
- ❧ It is detected by evaluating and documenting the optic nerve head changes and retinal nerve fibre layer defects.
- ❧ In one study fifty five percent of patients of ocular hypertension converted to treatment group based on optic disc changes without field changes.
- ❧ Another study showed that sixty percent of ocular hypertensives have RNFL defects up to 6 years before they developed visual field defects on standard automated perimetry.

Normal optic nerve head



Neuroretinal rim

The neuro retinal rim (NRR) is the tissue between the outer edge of the cup and the optic disc margin.

The normal rim has an orange or pink colour and a characteristic configuration in most healthy eyes: the inferior rim is the broadest followed by the superior, nasal and temporal (the 'ISNT' rule).

The cardinal feature of glaucomatous optic neuropathy is a loss of NRR from the inner edge of the rim.

'ISNT' rule

- The neuro-retinal rim is widest in the Inferior disc region → Superior disc region → Nasal disc area → Temporal disc area

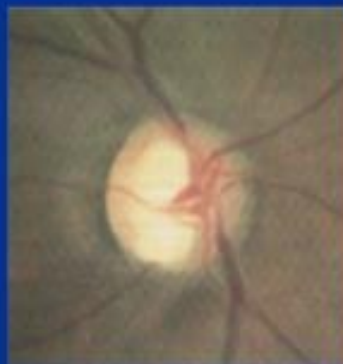


- In glaucoma, the neuro-retinal rim is lost in all sections of the of the optic disc, depending upon the stage of the disease..... →

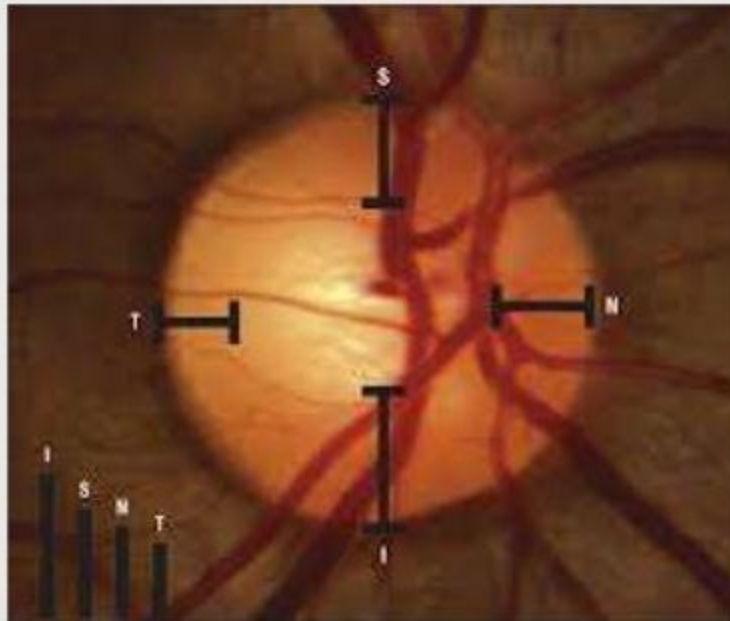
Infero-temporal → *Supero-temporal* →

Temporal horizontal → *Infero-nasal* →

Supero-nasal



I>S>N>T





Features that should raise suspicion that glaucomatous damage has already occurred include:

1. notching of the rim
2. hemorrhage crossing the rim
3. undercutting of rim
4. asymmetry of rim width between eyes in absence of asymmetry of disc size
5. an abnormally thin rim in one or two sectors.



- ⌘ An approximate rule is that a vertical cup-disc ratio of >0.7 or loss of rim to the disc margin anywhere outside the temporal sector strongly suggest glaucoma . This rule may not apply if the disc is extremely large or very tilted.

Normal optic disc



Optic disc size



- ❧ Optic disc size is important in deciding if a cup-disc (C/D) ratio is normal.
- ❧ Normal median vertical diameter for non-glaucomatous discs is 1.50 mm in a Caucasian population.

Cup-disc ratio



- ❧ The C/D ratio indicates the diameter of the cup expressed as a fraction of the diameter of the disc; the vertical rather than the horizontal ratio is generally used in clinical practice.
- ❧ The NRR occupies a relatively similar cross-sectional area in different eyes.
 1. Small discs have small cups with a median C/D ratio of about 0.35
 2. Large discs have large cups with a median C/D ratio of about 0.55
 3. Only 2% of the population have a C/D ratio greater than 0.7.
 4. In any individual, asymmetry of 0.2 or more between the eyes should also be regarded with suspicion, though it is critical to exclude a difference in overall disc size.

Normal vertical cup-disc ratio for vertical disc diameter



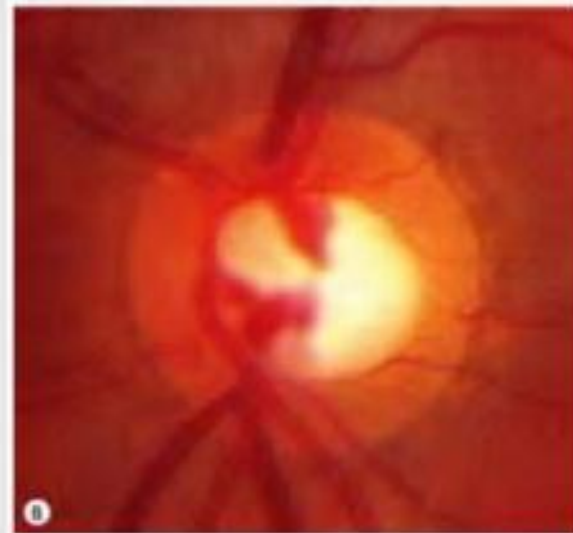
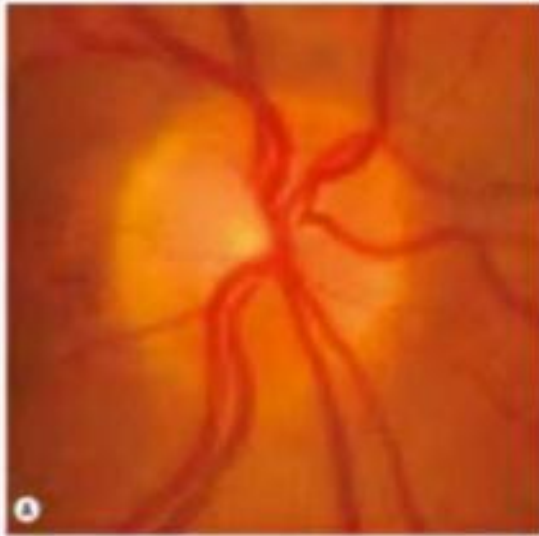
Disc diameter(in mm)	Mean cup-disc ratio
<1.0	0.26
1.2	0.33
1.4	0.39
1.6	0.45
1.8	0.5
>2.0	0.55



- ❧ Disc size is extremely variable. It can be measured by using the small size spot of a direct ophthalmoscope. This spot size can be used to estimate whether a disc is large or smaller than average.
- ❧ Disc size can also be evaluated using conventional photographic means with overlay grid as well as optic nerve head analysis.

Small optic disc with small cup

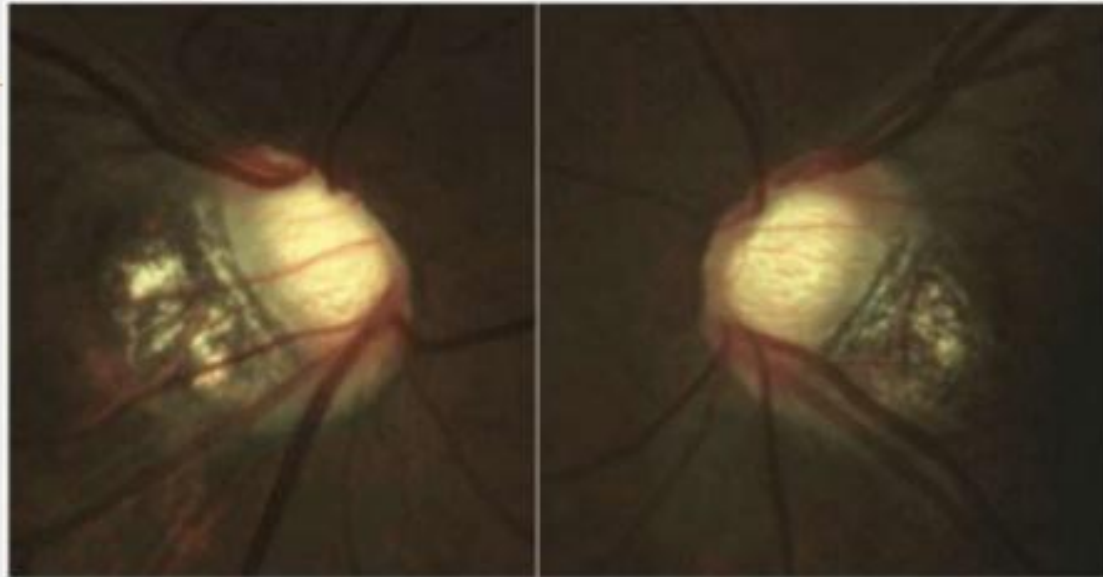
Large optic disc with large cup



Optic nerve head in myopia

Certain features of highly myopic discs interfere with interpretation of NRR and cupping:

- ❧ Relatively large disc area
- ❧ Oblique insertion of optic disc which distorts the view of temporal rim.
- ❧ A temporal gray crescent which could result in physiologic rim being misinterpreted as pathologically thin in that area.
- ❧ A shallow cupping which masks the deepening of cup in glaucoma.



Oblique insertion of optic nerve heads in myopic eyes can obfuscate the interpretation of the neuroretinal rim and creates a wide temporal peripapillary crescent. In this case, the asymmetry and loss of the superonasal rim of the right eye corresponds to glaucomatous damage

Peripapillary retina



- ∞ Retinal nerve fiber layer(RNFL) is seen as striations in the light reflexes from the bundles of nerve fiber commonly seen with red free light either on direct ophthalmoscope or on fundus camera.
- ∞ RNFL is seen better at the posterior pole in the peripapillary region where they reach a critical thickness especially at the vertical poles of optic nerve head.

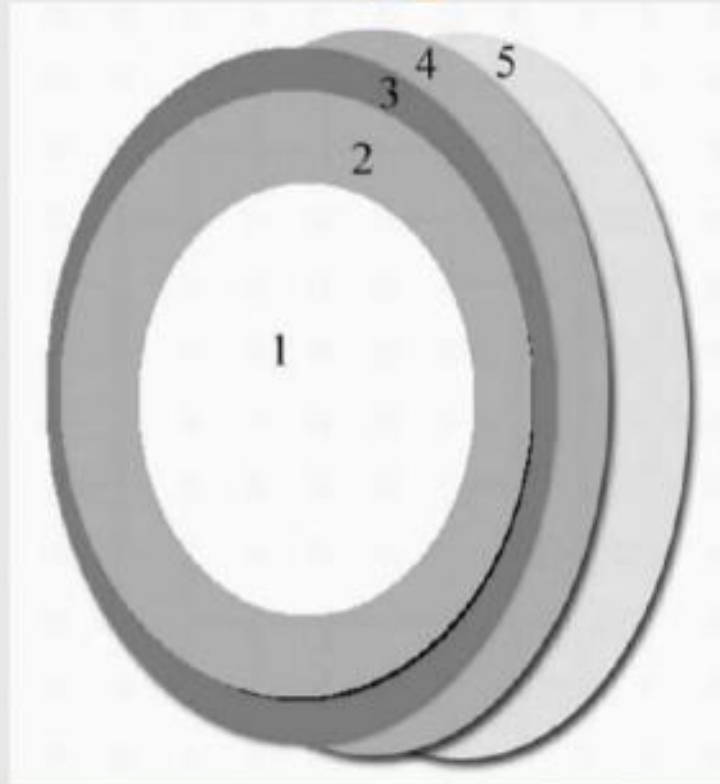
Peripapillary pigmentation

- ❧ Scleral lip
- ❧ It is seen as thin even white rim outside disc margin
- ❧ It represents anterior extension of sclera between the choroid and the optic nerve.

Scleral lip



1=cup;2=NRR;3=scleral
lip;4=zone beta;5=zone alpha



Grey crescent

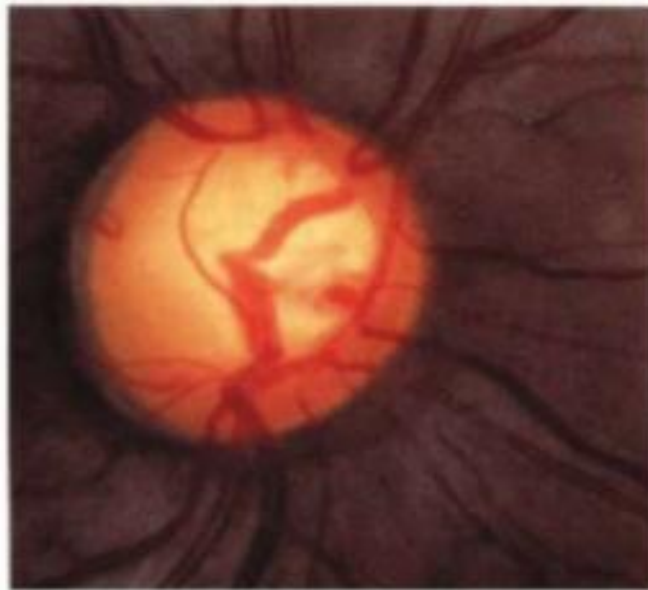


- ❧ it is seen within peripheral tissues of optic nerve head
- ❧ Scleral lip is seen peripheral to the grey crescent.
- ❧ It is often bilateral and it is usually located along temporal or infero-temporal disc margin.
- ❧ Grey crescent is due to internal extension of Bruch's membrane in the peripapillary scleral ring.
- ❧ At times in evaluation of optic nerve head ,the grey crescent can be taken as peripapillary tissue and not NRR resulting in falsely small optic disc and small NRR area.

Temporal grey crescent

❧

❧



A



B

Figure 4.10 Gray crescents in the optic nerve head of a patient with large physiologic cups. The thin crescent is seen just inside the scleral lip in the temporal quadrant of the right eye (A) and the inferotemporal quadrant of the left (B).

Zone beta



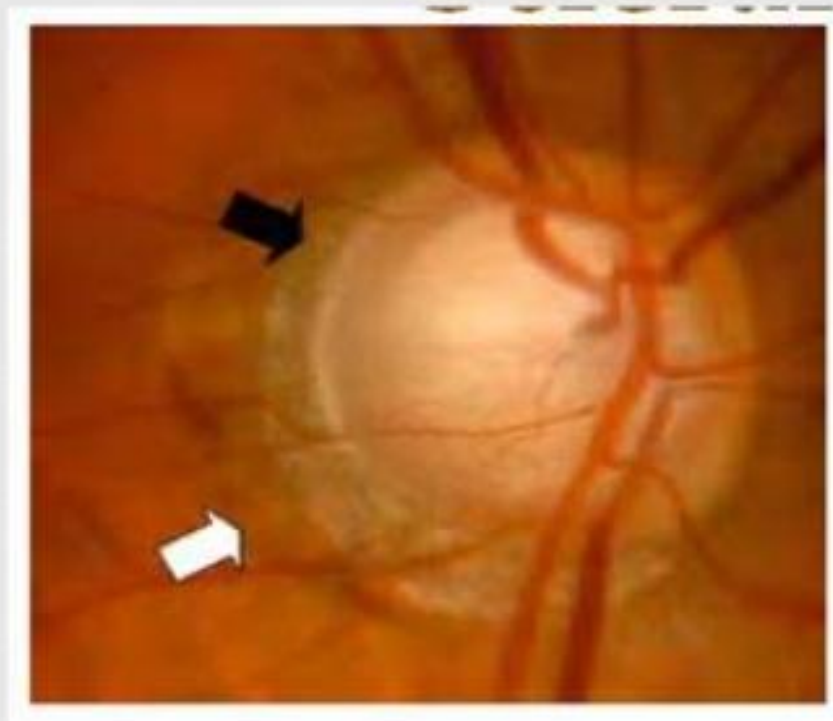
- ❧ It is seen between peripheral NRR and zone alpha.
- ❧ It represents a retraction of retinal pigment epithelium from the disc margin due to atrophy of RPE.
- ❧ Sclera and large choroidal vessels are visible due to RPE atrophy.
- ❧ It is more frequent and more extensive in patients with POAG and NTG.
- ❧ Location and extent of zone beta atrophy correlates with visual field loss.
- ❧ It indicates that the area has poor perfusion.

Zone alpha



- ❧ It is better detected at the temporal disc margin
- ❧ On the inner side ,it is bounded by zone beta or peripapillary scleral ring and on the outer side by retina.
- ❧ It is seen as irregular hyperpigmentation and hypopigmentation.
- ❧ It is due to parapapillary crescent of RPE irregularity close to the margin of Bruch's membrane.
- ❧ It is present in almost all normal eyes.

Zone beta atrophy (black arrow)
zone alpha atrophy (white arrow)



The optic nerve head changes in glaucoma



- ∞ The spectrum of disc damage in glaucoma ranges from highly localized tissue loss with notching of the NRR to diffuse concentric enlargement of the cup.

Morphology of glaucomatous optic atrophy



∞ Glaucomatous damage results in characteristic signs involving

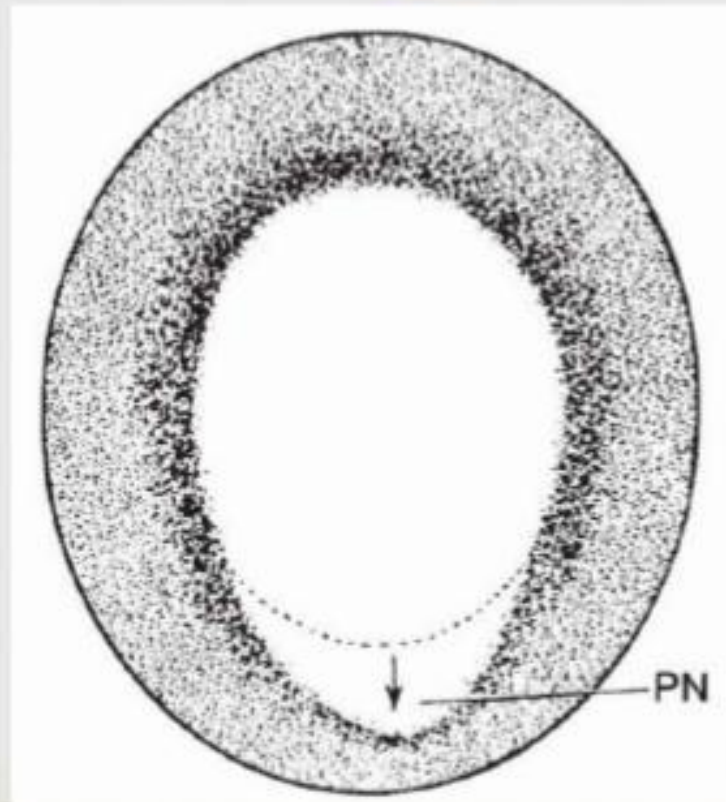
- (a) *the optic nerve head*
- (b) *the peripapillary area*
- (c) *the retinal nerve fibre layer*

Focal atrophy



- ✧ Selective loss of neural rim tissue in glaucoma occurs primarily in the inferotemporal region of optic disc and less commonly in superotemporal sector in early glaucoma resulting in enlargement of cup in vertical or oblique direction.
- ✧ In early glaucomatous eyes, the inferotemporal neural rim area is smaller than superotemporal area so that the vertical cup disc ratio is more than horizontal cup disc ratio.

Focal atrophy with polar notching



Focal atrophy with polar notching





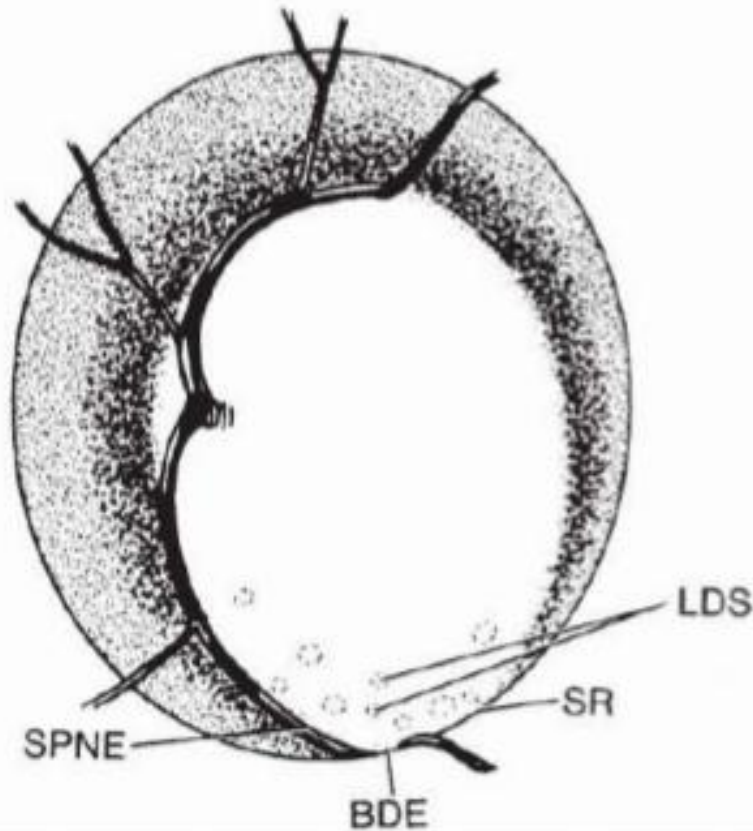
- ❧ As the glaucomatous process continues, temporal neural rim is involved after vertical poles, with nasal quadrant being the last to be destroyed.
- ❧ The focal atrophy of the neural rim begins as small discrete defect , usually in inferotemporal quadrant, which is known as **polar notching or focal notching or pit-like changes.**

Bayoneting



- When the local thinning of neural rim tissue reaches the disc margin, a **sharpened rim** is produced.
- If a retinal vessel crosses the sharpened rim, it will bend sharply at the edge of the disc creating **bayoneting** at the disc edge.

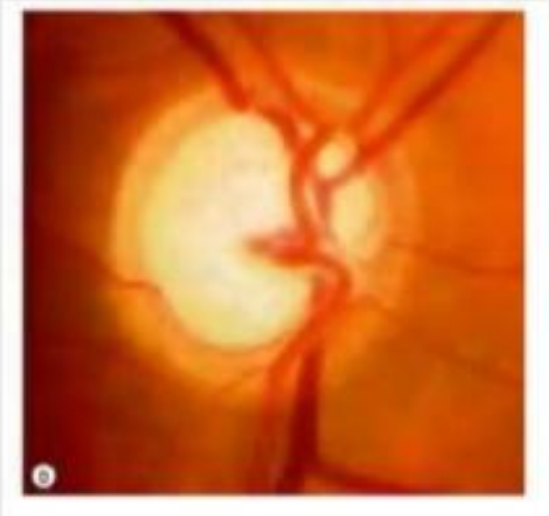




Inferotemporal loss of neural rim in glaucomatous optic atrophy, creating a sharpened rim (SR) at the disc margin, a sharpened polar nasal edge (SPNE) along the cup margin, bayoneting at the disc edge (BDE) where the vessels cross the sharpened rim, and laminar dot sign (LDS) due to exposure of fenestrae in lamina cribrosa.

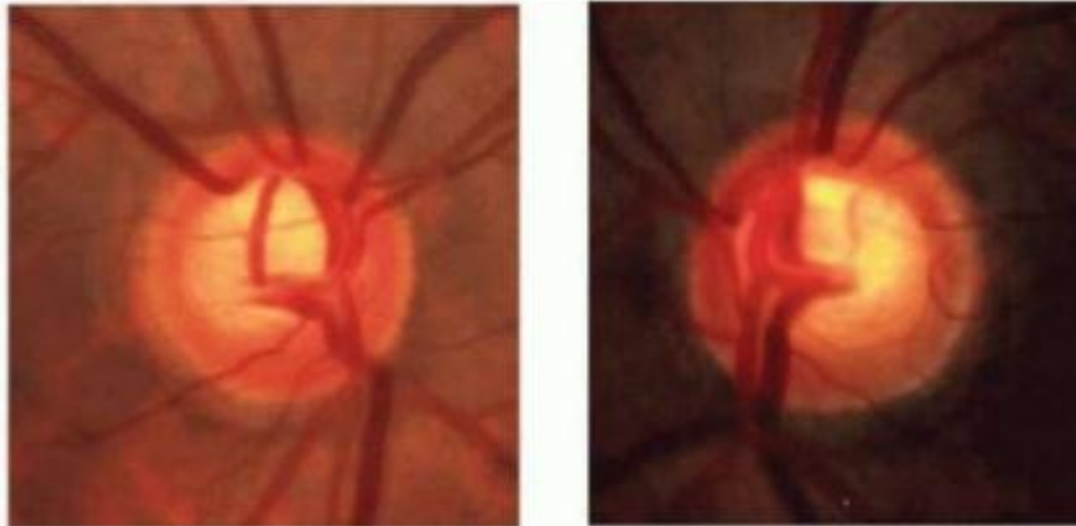
Concentric atrophy

In some cases the loss of neural rim begins temporally and then progresses circumferentially towards the poles resulting in enlargement of cup in concentric circles with retention of round shape of cup. This type of glaucomatous disc damage is referred to as **concentric atrophy** and the phenomenon involved is known as **temporal unfolding**.





- ✧ It is difficult to distinguish concentric atrophy from physiologic cup ,hence it is important
 - to compare the cup in fellow eye
 - to study serial photographs for evidence of progressive change.



Large physiologic optic nerve head cups that are symmetrical and intact. The shape of the physiologic cup is roughly correlated with the shape of the disc, which means that the margins of cup and disc tend to run more or less parallel. The cup has a horizontally oval shape in most normal eyes; thus, a vertical CDR greater than the horizontal CDR should be considered suspicious.



- ☞ Sometimes a thinning of neural rim may be seen as **crescentic shadow** adjacent to the disc margin on direct ophthalmoscopy.
- ☞ It is a sign of early glaucomatous disc damage and it should not be confused with gray crescent in optic nerve head.

Deepening of cup



- ❧ In some cases deepening of cup is predominant pattern of early glaucomatous disc damage .
- ❧ It occurs only if the lamina cribrosa is not initially exposed and it produces **overpass cupping**, in which the blood vessels initially bridge the deepened cup and later collapse into it.

☞ Continued deepening of cup causes exposure of underlying lamina cribrosa, which is often recognized by gray fenestrae of lamina, seen on direct ophthalmoscope as gray dots. This has been referred to as **laminar dot sign**.



Pallor cup discrepancy



- ❧ In early stages of glaucomatous optic atrophy enlargement of cup progresses ahead of area of pallor.
- ❧ This pattern differs from other types of optic atrophy in which area of pallor is more than the enlargement of the cup.
- ❧ It is important to differentiate area of cupping from area of pallor either by examining the optic disc with **stereoscopic techniques** or by seeing the **kinking of blood vessels** at the cup margin.

Cup margin shown by kinking of blood vessel





- ❧ Pallor-cup discrepancy may occur with focal or diffuse enlargement of cup.
- ❧ **Saucerization** refers to a pattern of early glaucomatous change in which diffuse shallow cupping extends to the disc margin with retention of a central pale cup.
- ❧ **Focal saucerization** refers to more localized ,shallow sloping cup, usually in inferotemporal quadrant.
- ❧ The retention of normal neural rim color in the area of focal saucerization is known as **tinted hollow**. As the glaucomatous process continues, this is replaced either by grayish hue known as **shadow sign** or by laminar dot sign.

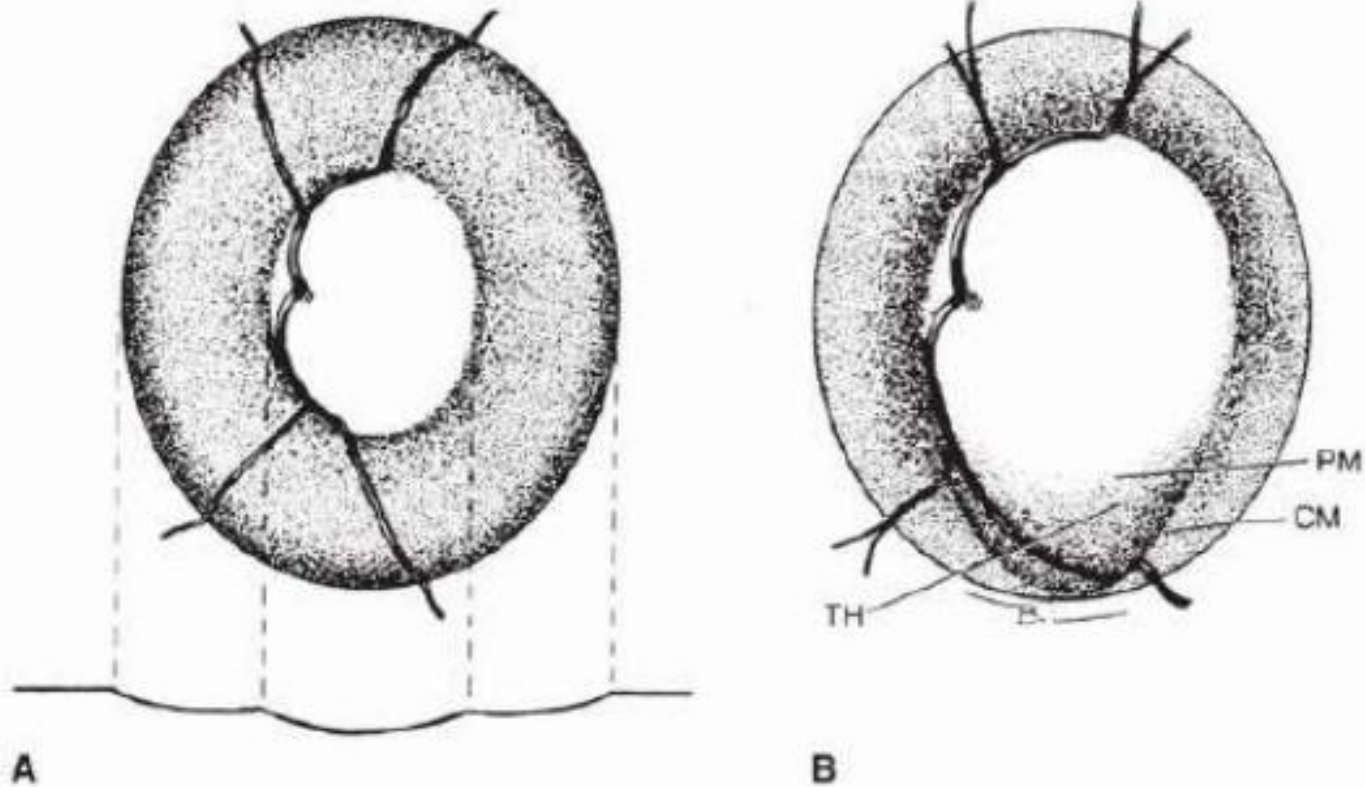


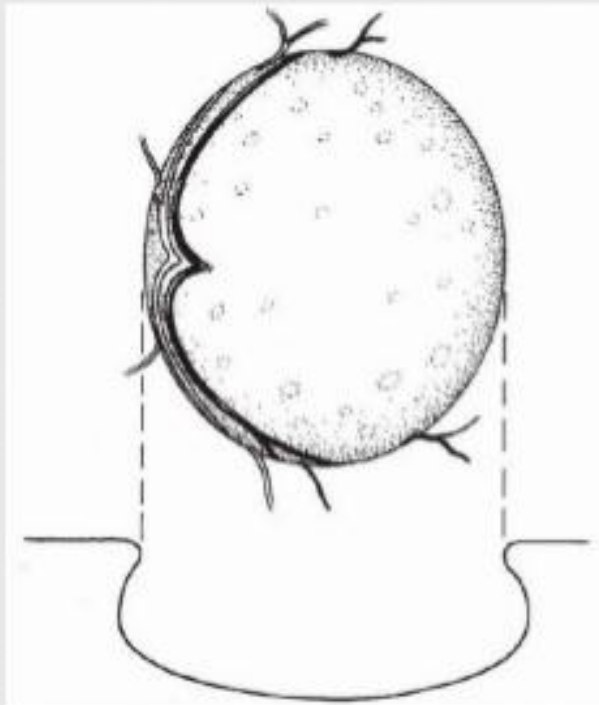
Figure 4.15 Glaucomatous optic atrophy. Pallor-cup discrepancy. A: Saucerization with corresponding cross-sectional view. B: Focal saucerization with tinted hollow (TH) between pallor margin (PM) and cup margin (CM). Note kinking of vessels in both cases.

Advanced glaucomatous cupping



- ⌘ Eventual loss of all neural rim tissue results in **total cupping** which is characterized by white disc with bending of all blood vessels at disc margin.
- ⌘ This is also known as **bean-pot cupping** because the cross section of a histologic section reveals *extreme posterior displacement of lamina cribrosa* and *undermining of disc margin*.

Advanced glaucomatous optic atrophy with bean-pot cupping



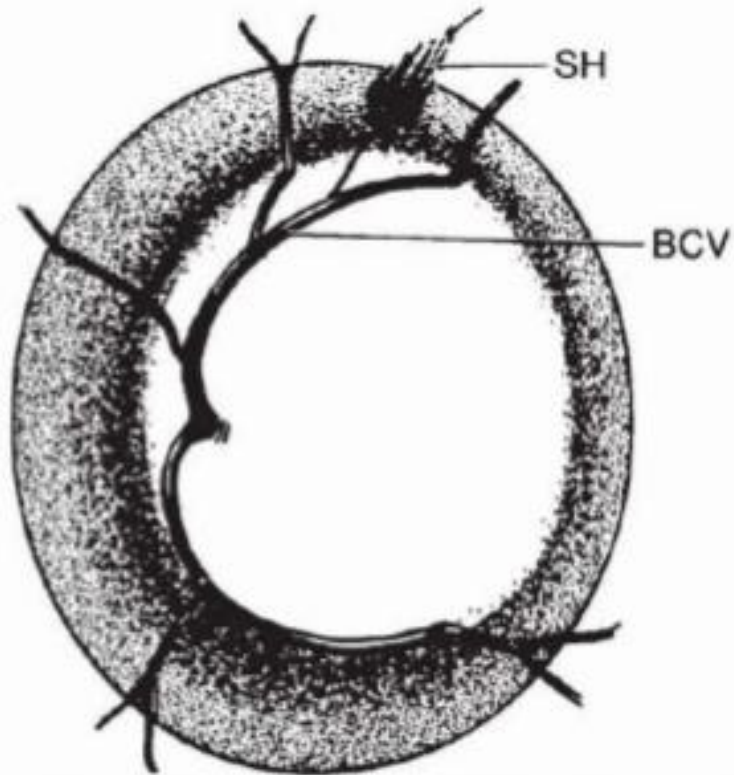
Vascular signs of glaucomatous optic atrophy



Optic disc hemorrhages:

- ☞ Also referred to as **splinter hemorrhages** or **drance hemorrhages** .
- ☞ They are seen more commonly in patients of normal tension glaucoma than COAG.
- ☞ They tend to come and go and reappear at same site or different site.
- ☞ They typically cross the disc margin but during resorption, the papillary portion may disappear first so that an extra papillary hemorrhage remains.

Splinter hemorrhages





- ❧ The most common location of splinter hemorrhage is the inferior quadrant.
- ❧ They most often occur in early or middle stages of glaucomatous optic atrophy and disappear in advanced stage of glaucomatous optic atrophy with absent neural rim.
- ❧ Although not pathognomonic sign of glaucoma, splinter hemorrhages are a significant finding as they often precede RNFL defects ,polar notching and visual field defects.



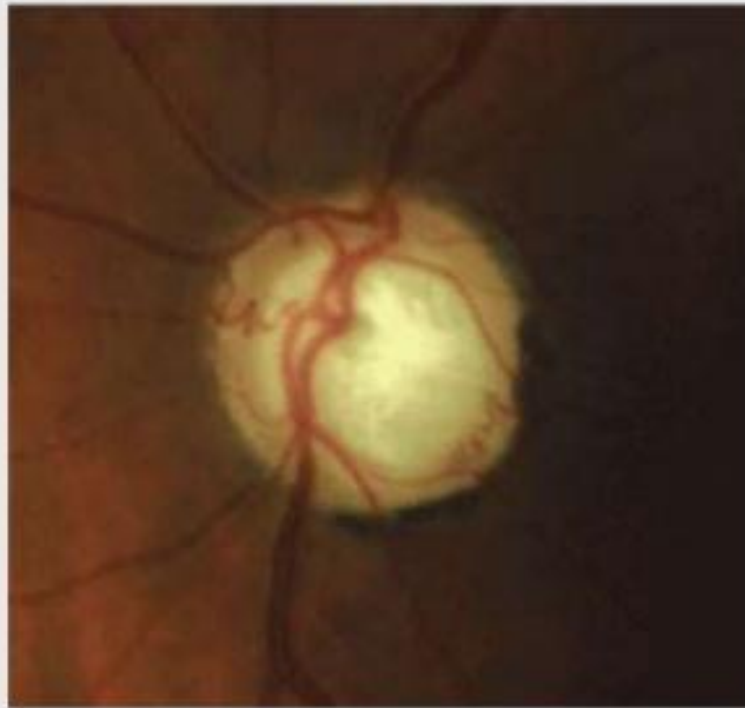
- ❧ They are especially suggestive of glaucoma when associated with high IOP.
- ❧ They are more commonly seen in diabetic patients with glaucoma than non-diabetic patients with glaucoma.
- ❧ Disc hemorrhages are more commonly associated with progressive visual field changes and are suggestive that glaucoma may be out of control.

Tortuosity of retinal blood vessels



- ❧ Tortuosity of retinal blood vessels may be seen in advanced glaucomatous disc damage and they represent loops of collateral blood vessels in response to chronic retinal vessel occlusion.
- ❧ Venovenous anastomoses associated with chronic branch retinal vessel occlusion and massive flame shaped hemorrhages also occur with increased frequency in glaucomatous eyes.

Tortuosity of retinal blood vessels



Cilioretinal arteries



- ❧ One study suggested that presence of cilioretinal arteries in glaucomatous eyes was associated with larger cup-disc ratio and progressive visual field defects.
- ❧ Another study suggested that presence of cilioretinal arteries was associated with retention of central visual field for longer duration.

Location of retinal blood vessel in relation to cup



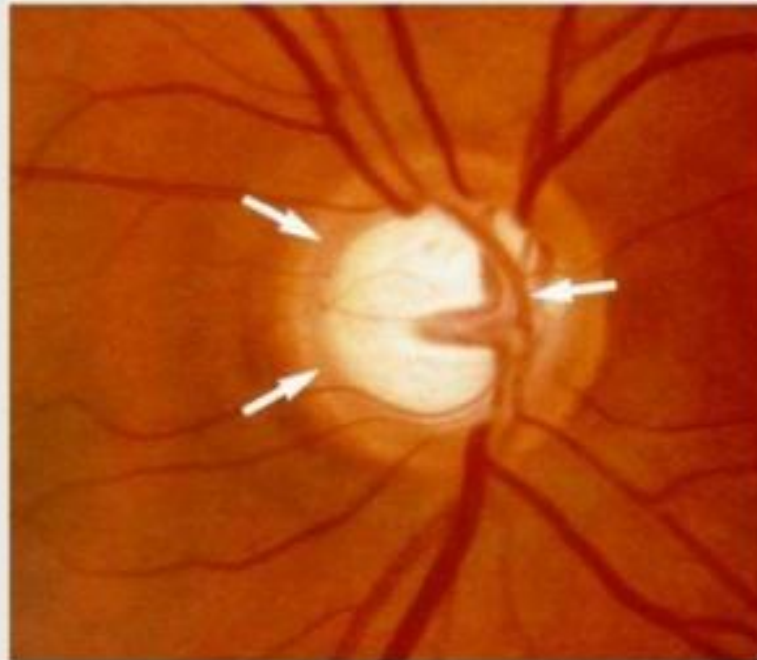
- ❧ Overpass cupping
- ❧ Baring of circumlinear vessel
- ❧ Nasal displacement of blood vessels
- ❧ Vertical eccentricity of central retinal vessel trunk
- ❧ Generalized arterial narrowing

❧ **Overpass cupping** is due to deepening of cup where a blood vessel bridges the cup and then collapse into it.

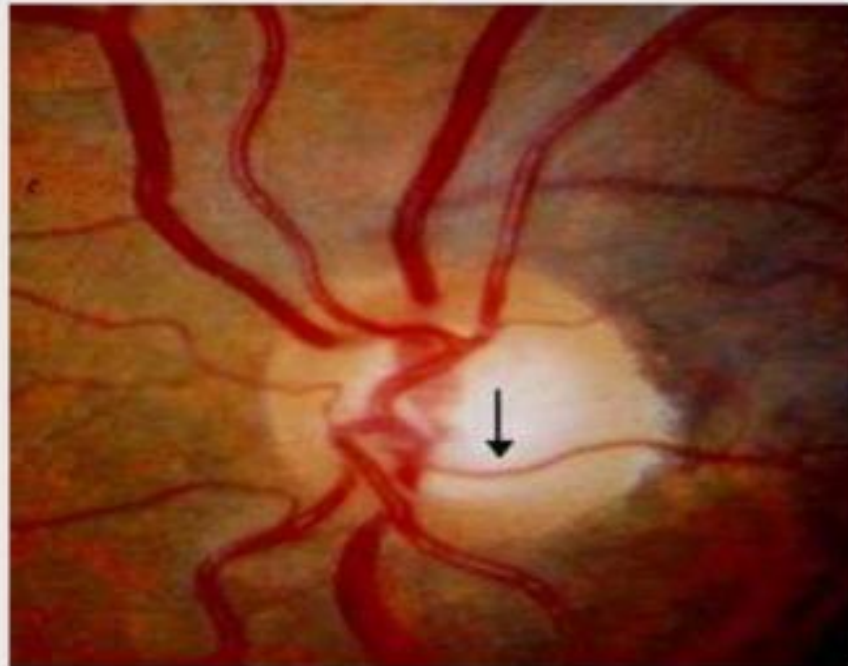
❧ **Baring of circumlinear vessel:**

- a. In many normal optic discs, one or two blood vessels curve the outline of a portion of physiologic cup. With glaucomatous enlargement of the cup, these circumlinear blood vessel may be bared from the margin of the cup.
- b. This sign is not pathognomonic of glaucoma but it's presence in glaucoma suspect patients is associated with visual field loss.

Overpass cupping with sharpened rim



Baring of circumlinear vessel





- ❧ Earlier thought to be related to glaucomatous disc changes, **nasal displacement of blood vessels** in now no longer considered diagnostic of glaucoma.
- ❧ However **vertical eccentricity** of central retinal vessel trunk may be related to course of glaucomatous optic atrophy.
- ❧ Neural rim loss was more likely to occur in the vertical quadrant further away from the central retinal vessel trunk.



❧ **Generalized arterial narrowing** outside optic nerve head is seen in some patients of chronic open angle glaucoma but it is a nonspecific finding also seen in patients of anterior ischemic optic neuropathy.

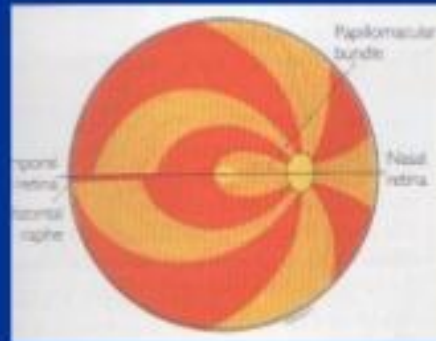
Peripapillary changes associated with glaucomatous optic atrophy



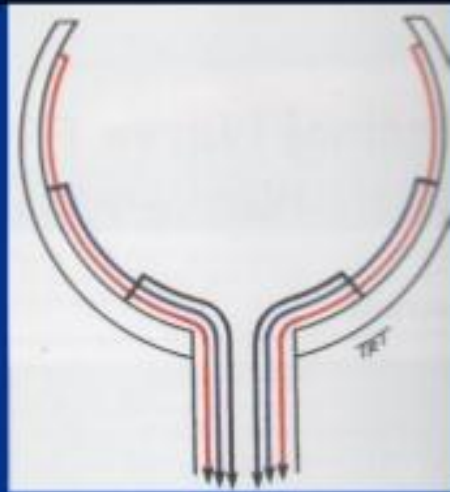
- ❧ Retinal nerve fibre bundle defects
- ❧ Peripapillary pigmentary changes

Retinal nerve fibers

- There are 1.2 million RNFs which form the optic nerve head(optic disc)
- Within the retina -



- Within the optic nerve head –



Retinal nerve fibre bundle defects



- The loss of axonal bundles in glaucoma lead to
 - ❧ loss of neural rim tissue
 - ❧ visible defects in retinal nerve fibre layer.
- ❧ These defects appear as
 - a. Dark stripes or wedge shaped defects in peripapillary region
 - b. Diffuse loss of retinal striations.
- ❧ They often follow disc hemorrhages and correlate highly with visual field defects and lost neural rim tissue.

☞ RNFL defects are also seen in

- many neurological disorders
- ocular hypertension
- healthy eyes.

☞ Diffuse RNFL loss occurs in following order of decreasing frequency:

COAG > ocular hypertension > normotensive individuals.

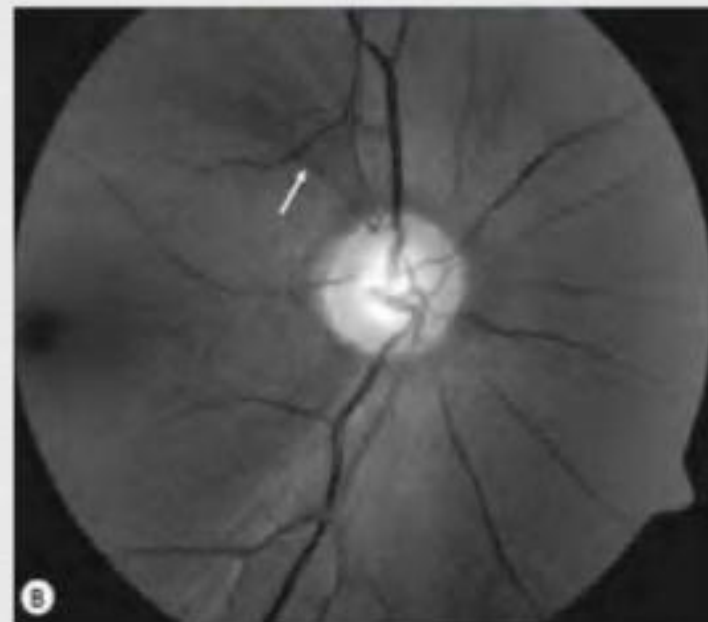
☞ RNFL defects either localized or diffuse may be the initial sign of glaucomatous disc damage

Retinal nerve fibre bundle defects



Superotemporal wedge shaped defect

Same defect in red free light

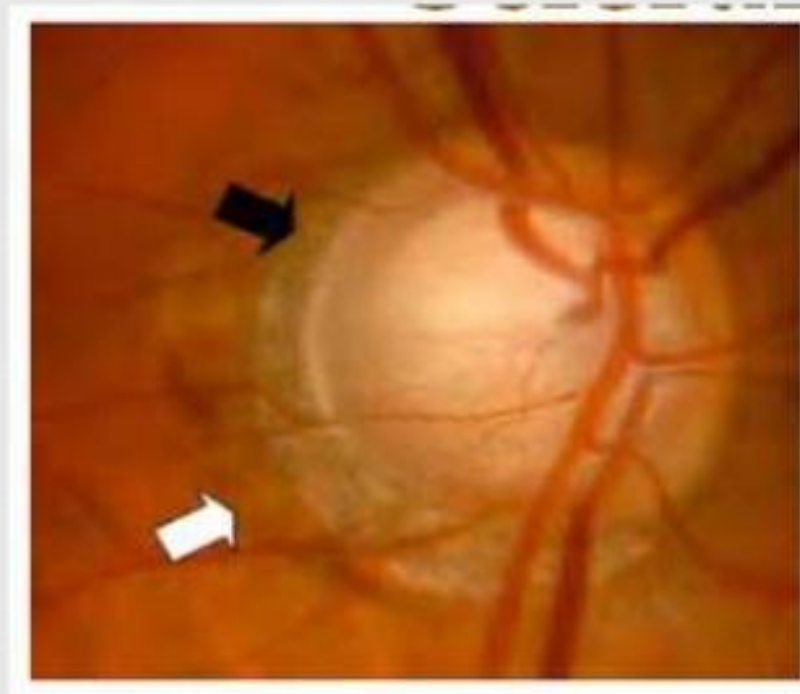


Peripapillary pigmentary changes



- ❧ These changes are nonspecific for glaucoma as they also occur in myopia and ageing.
- ❧ Peripapillary atrophy occur more frequently and it is larger in glaucomatous eyes than normal eyes.
- ❧ It enlarges with progression of glaucoma and correlate with the quadrant of greater neural rim loss.
- ❧ Absence of peripapillary atrophy may be associated with decreased progression of glaucomatous damage in patients of ocular hypertension.

Zone beta atrophy (black arrow)
zone alpha atrophy (white arrow)

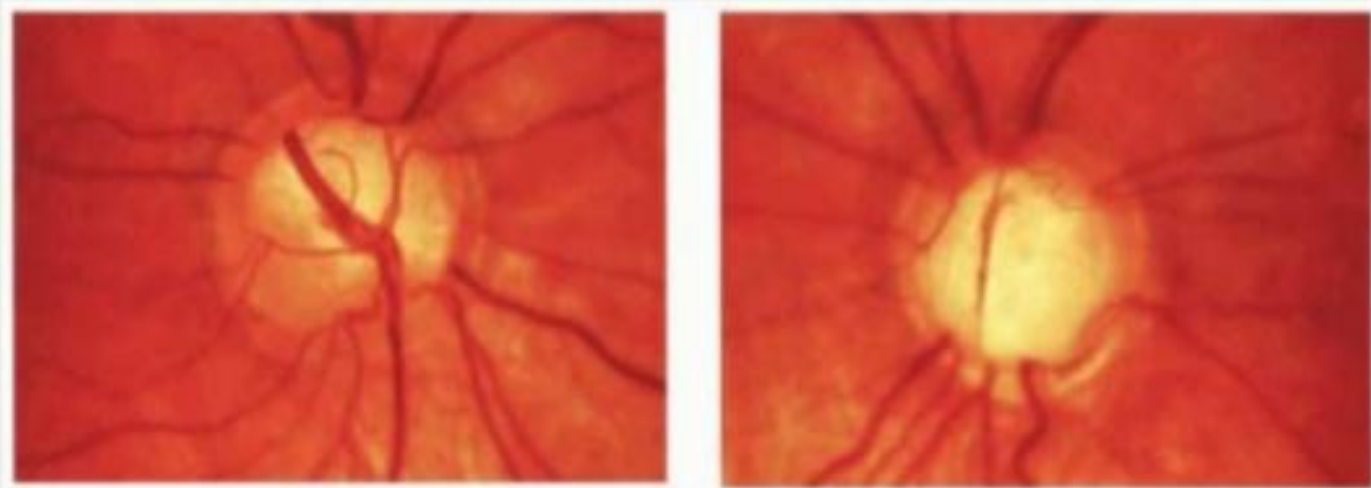


Differential diagnosis of glaucomatous optic atrophy



- ❧ 1. Normal variation in physiological cup
- ❧ 2. Optic disc coloboma
- ❧ 3. Optic pit
- ❧ 4. Morning glory syndrome
- ❧ 5. Tilted disc syndrome
- ❧ 6. Optic disc hypoplasia
- ❧ 7. Arteritic type of anterior ischemic optic neuropathy
- ❧ 8. Intra cranial aneurysms

Optic disc coloboma



- Colobomas of the optic nerve heads can simulate glaucomatous cupping. This patient would appear to have nearly total cupping and pallor, and yet the IOP was low normal and the visual fields were full with normal central vision.

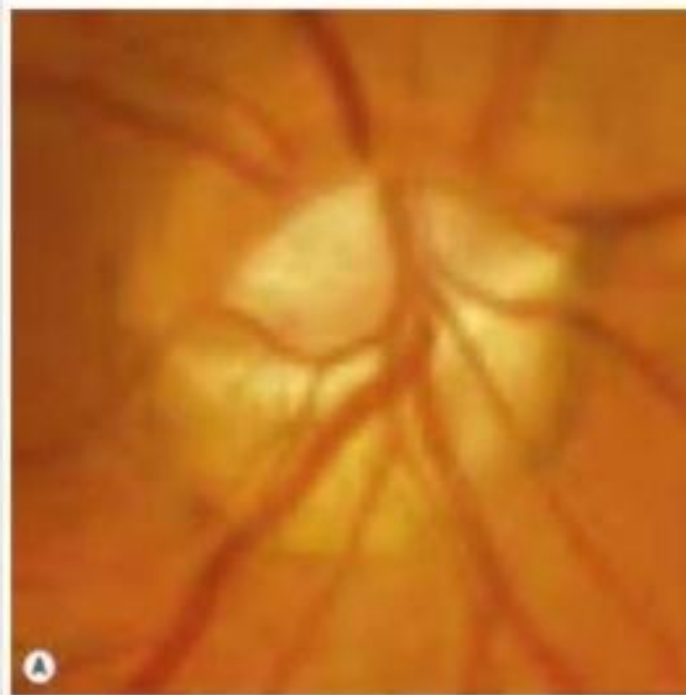
Optic pit



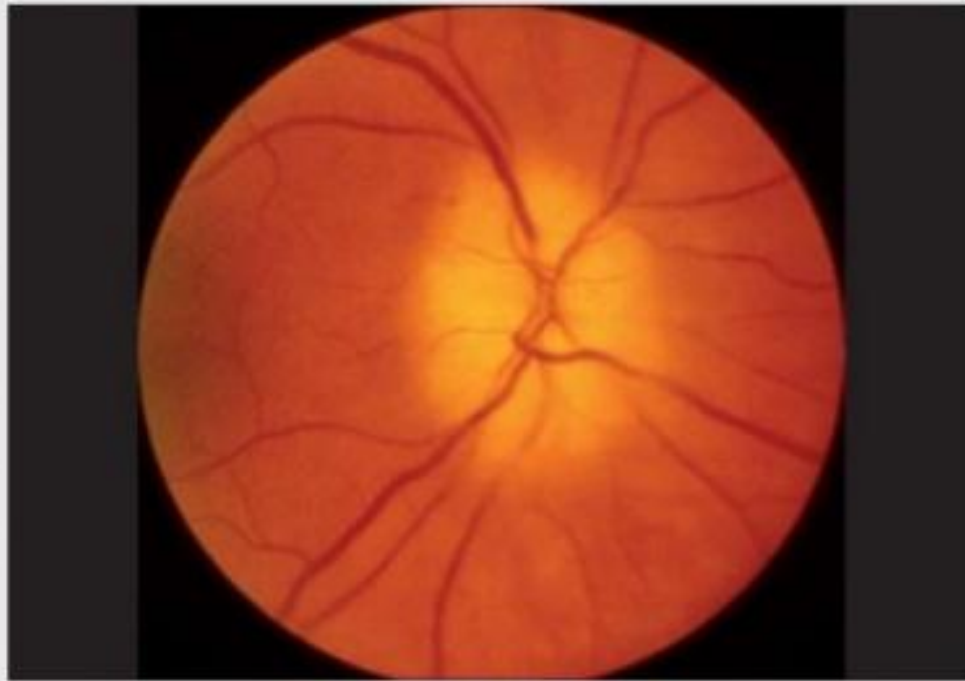
Morning glory syndrome



Tilted disc syndrome



Arteritic type of anterior ischemic optic neuropathy



Optic disc hypoplasia



Various techniques of optic disc evaluation



Qualitative assessment

1. Direct ophthalmoscopy
2. Indirect ophthalmoscopy
3. Slit lamp biomicroscopy using
 - 90 d lens
 - 78 d lens
 - 60 d lens
4. Fundus camera photography



Quantitative assessment

1. Confocal scanning laser tomography
2. Scanning laser polarimetry
3. Optical coherence tomography

Slit lamp biomicroscopy



Optic disc can be assessed clinically as follows:

1. A narrow slit beam is focussed on the disc using a fundus biomicroscopy lens
2. The height of the beam is adjusted until it matches the distance between the superior and inferior limits of the NRR (not the scleral rim surrounding the neural tissue) and the diameter in mm read from the slit-lamp graticule.
3. A correction factor may be necessary, dependent on the lens used. Refractive error affects measurement only minimally, although myopia above -8 dioptres may distort the result.

Correction factors for estimating optic disc diameter



Lens Correction factor

∞ Volk 60 D × 0.88-1.0

∞ Nikon 60 D Around 1.0

∞ Volk 90 D ×1.3

∞ Volk 78 D ×1.1

∞ Goldman 3-mirror ×1.27

Disc recording



Careful detailed drawing of the optic disc should be done on every visit.

Following structures should be always documented

1. Size and shape of the optic nerve head.
2. Size and shape of the cup.
3. Neuroretinal rim thinning at the vertical cup
4. Disc hemorrhages
5. Position of blood vessel as they cross the cup margin.
6. Scleral crescent
7. Peripapillary atrophy
8. Nerve fiber layer defects
9. Tortuosity of blood vessel
10. Arteriolar narrowing



THANK YOU