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Core Messages

- Optic disc evaluation is of fundamental importance in the management of glaucoma.
- Clinical examination of the optic disc is best performed with slit lamp biomicroscopy, utilizing contact or handheld lenses.
- Subjective assessment or measurement of optic disc size is paramount, as there is a strong correlation between optic disc size and optic cup size.
- Great attention should be paid to neuroretinal rim contour, as well as the presence of retinal nerve fiber layer defects and optic disc hemorrhages, which can easily be missed.
- Over time, disc changes are better identified with optic disc photographs or automated devices. The rate of disc changes in glaucoma is quite variable in different individuals and depends upon the stage of the disease, among other things.

- A large proportion of individuals with optic disc hemorrhages will present with progressive changes in the optic nerve fiber layer or optic disc within 2 years of hemorrhage, and these individuals should be monitored closely.

2.1 How Should I Examine the Optic Nerve?

Optic nerve-head examination is probably the most important step in the diagnosis of glaucoma and is also extremely important in monitoring patients with established glaucoma. There are several ways to clinically examine the optic nerve head, including direct ophthalmoscopy, indirect ophthalmoscopy, and slit lamp biomicroscopy with contact lenses (such as a Goldman lens), handheld lenses (such as a 78- or 90-diopter lens), or the Hruby lens. The advantages of slit lamp biomicroscopy, the preferred method for optic nerve evaluation, over the other methods mentioned are the quality of the stereopsis and magnification provided. Although slit lamp biomicroscopy with handheld lenses can be performed through an undilated pupil, a stereoscopic view may be possible only if the pupil is dilated.

In addition to slit lamp examination, optic disc stereophotography provides complimentary clinical information. For example, data from the

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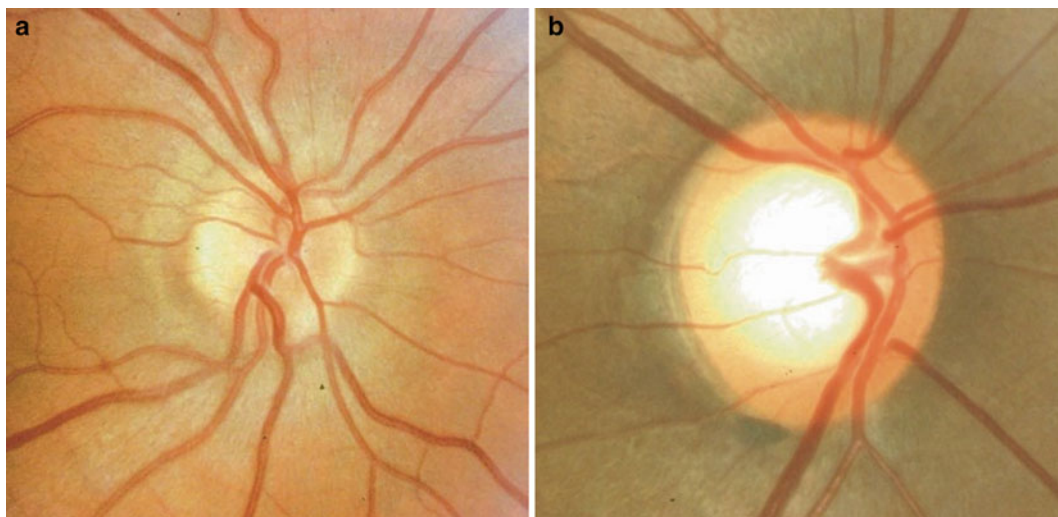


Fig. 2.1 Examples of a small optic disc (a) and a large optic disc (b). The disc and rim areas, measured with confocal scanning laser tomography, were 1.3 and 1.1 mm² in

disc (a) and 3.5 and 1.6 mm² in disc (b). Note that the whole disc of example (a) is smaller than just the rim area alone in example (b)

Fig. 2.2 Optic disc with infero-temporal disc hemorrhage, associated with thinning of the neuroretinal rim in the same location. The inferior circumferential blood vessels exhibit beading



Ocular Hypertension Treatment Study (OHTS) show that 84 % of 128 cases of optic disc hemorrhages were detected on disc photographs but not on the clinical exam [1].

Clinical examination of the optic nerve should be performed with similar methodology each and every time it is executed, in order not to miss important aspects of the examination. In my view, examination of the optic nerve head should start with an evaluation of optic disc size since disc size is extremely important in the interpreta-

tion of other optic nerve findings (see Fig. 2.1). Even a simple subjective assessment, without specific measurements, of whether the disc is small, large, or average in size can be of value. The exam should then proceed to a careful assessment of the neuroretinal rim, looking for areas of thinning, notching, nasal cupping, and vessel abnormalities. There is a helpful rule for examining the contour of the neuroretinal rim (the ISNT mnemonic), which states that in normal discs the inferior neuroretinal rim is thickest, followed in

decreasing order by the superior, nasal, and temporal neuroretinal rims [2, 3]. The optic nerve in Fig. 2.1b follows the ISNT rule, while the nerve in Fig. 2.2 does not. After the disc size is estimated and the neuroretinal rim has been examined, one should examine the peripapillary area carefully, paying great attention to the presence of optic disc hemorrhages and retinal nerve fiber layer defects (both diffuse and localized), and, to a lesser degree, to the presence and location of peripapillary atrophy [4–9].

Summary for the Clinician

- Examination of the optic nerve is critical for the diagnosis of glaucoma and its progression.
- Slit lamp biomicroscopy with handheld lenses is the best method of optic nerve examination since it provides good stereopsis and magnification.
- Optic disc stereophotographs are complementary to slit lamp examination and may pick up findings missed on the clinical exam.
- Optic nerve examination should be systematic.

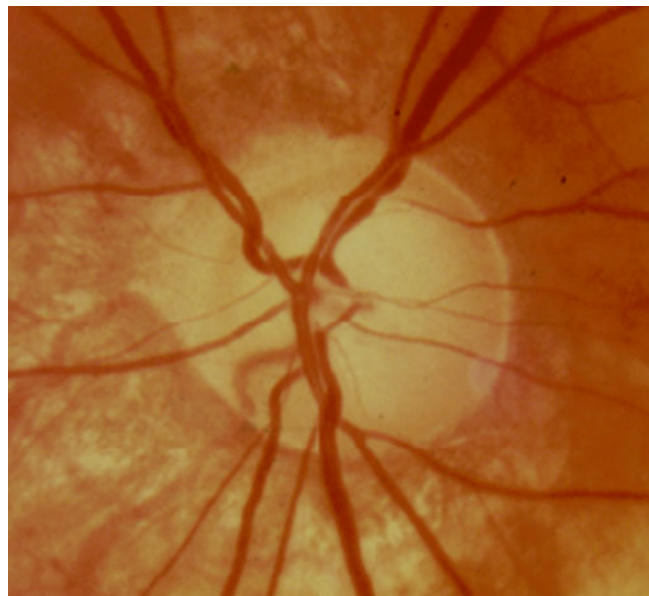
- Disc size (small, average, large) should be estimated first.
- The neuroretinal rim should be examined for diffuse and focal changes. The ISNT rule is helpful.
- Disc hemorrhages, nerve fiber layer defects, and peripapillary atrophy should also be noted.

2.2 How Does One Establish the Borders of the Nerve and Follow the Neuroretinal Rim Contour?

In clinical studies, the disc margin is determined as the internal edge of the scleral ring. In most cases, identification of the white scleral ring is relatively easy although it might not be clearly visible all the way around the optic disc, especially in the nasal area (Fig. 2.3). Establishing the borders of the optic nerve can be very challenging in cases of tilted discs, crowded discs, or highly myopic eyes with significant peripapillary atrophy.

The neuroretinal rim is identified by its normally pink color and/or the change of contour from the rim to the cup, which is best determined

Fig. 2.3 Example of an optic disc with a sclerotic appearance. The internal borders of the neuroretinal rim are usually difficult to determine in these discs with a saucerized type of cupping



by following the trajectory of the blood vessels within the optic disc. Determining and describing the internal borders of the neuroretinal rim (or the limits of its excavation) is sometimes difficult. The size of the optic cup varies significantly in normal eyes, and it is strongly correlated with the size of optic disc [3]. Normally, circumlinear blood vessels rest on neuroretinal rim. Therefore, in most cases, the boundaries of the optic disc cup are best determined by following the trajectory of these vessels inside the optic disc. As neuroretinal rim disappears underneath the blood vessels, various terms are used to describe the appearance of the unsupported blood vessels. “Bayonetting,” a term borrowed from the shape of bayonet guns, refers to the sharp 90° turn (or occasionally more than 90° turn) a blood vessel develops as it dips into an acquired pit of neuroretinal rim loss and then emerges out onto the disc edge (see Fig. 2.2). “Baring” of circumlinear vessels refers to the unsupported appearance vessels have when there is no neuroretinal rim directly in contact with them (see Fig. 2.2). “Nasalization” of blood vessels occurs as increased cupping causes a nasal shift of the major blood vessels emerging from the nerve. Blood vessels can also narrow as glaucoma develops. In the so-called sloped or saucerized cups, oftentimes present in sclerotic optic discs, the precise determination of the borders of the cup is more difficult and subjective, and a good stereoscopic view of the optic nerve is extremely helpful in those situations (Fig. 2.3) [10].

Recent studies with Spectral Domain OCT (SDOCT) provide new insights on the anatomy of the optic disc head. Reis et al. [11] have shown that the clinically defined disc margin does not have one unique anatomic correlate on OCT images, but might rather co-localize to the ending of Bruch’s membrane or other aspects of the border tissue of Elschnig, which varies between individuals and between regions of a single eye. In addition, the geometrical orientation used to measure the neuroretinal rim has been evaluated and a new minimum rim width presented better diagnostic ability than the traditional horizontal plane following the back of the eye [12]. Figure 2.4 illustrates how information from SDOCT can be incorporated into the clinical evaluation of a suspect optic disc.

Summary for the Clinician

- Correctly identify the edge of the disc/scleral ring as the first step of evaluation.
- Follow the trajectory of the vessels on the optic disc to assess the contour of the neuroretinal rim. Look for bayonetting, baring, nasalization, and narrowing of the blood vessels.
- New analysis of optic disc OCT might complement the clinical examination.

2.3 How Does One Avoid Misinterpreting Rim Loss?

The inherent variability in size and shape of the optic disc among normal individuals and among patients with glaucoma hampers the clinician’s ability to determine rim loss with high accuracy. Detection of rim loss over time can have higher specificity than cross-sectional detection of glaucoma, since detection over time does not depend on the interindividual variability of optic disc appearance. Nevertheless, certain steps should be taken to avoid misinterpreting rim loss.

The first step for a correct interpretation of rim loss is factoring in the assessment of optic disc size, as mentioned earlier. Optic disc size can influence the interpretation of rim loss in two ways: (1) a large optic disc might appear to be glaucomatous because large discs normally have large cups and apparently thin neuroretinal rim, although if one measures the total area of the neuroretinal rim it is usually larger in large discs; (2) a small optic disc might “hide” neuroretinal rim loss, as sometimes even a small cup in a small disc is abnormal (Fig. 2.1) Another step to avoid misinterpretation of rim loss is careful observation of rim contour as opposed to cup size, which can lead one to miss subtle changes of the neuroretinal rim. Looking for matching clues between the inside and outside of the optic disc is also useful, such as confirming the presence of an RNFL defect or hemorrhage in an area where the neuroretinal rim is suspicious.

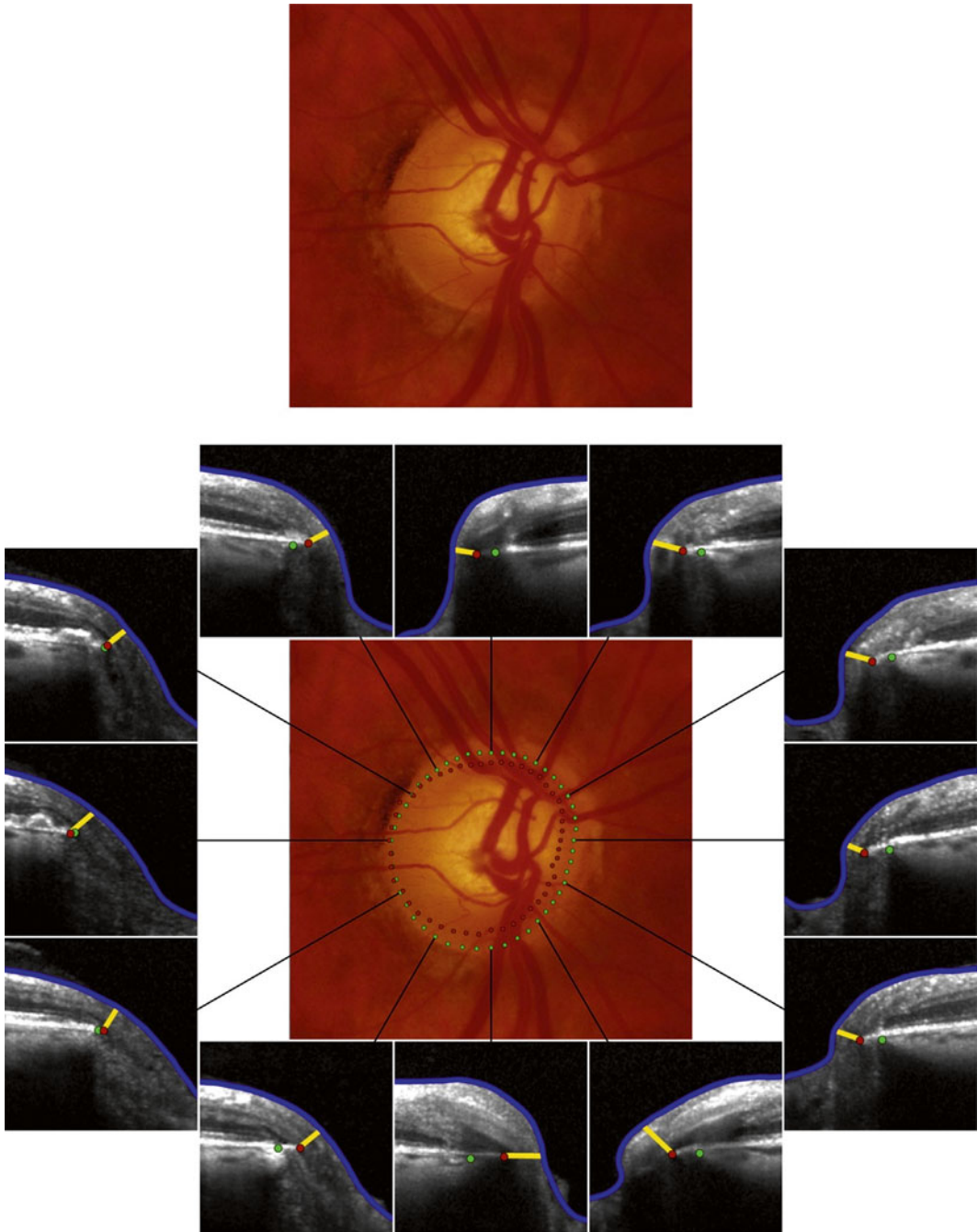


Fig. 2.4 Example of spectral-domain optical coherence tomography (SD OCT) evaluation of a right eye suspected of glaucoma (*Top* photo). In this case, SD OCT with analysis of Bruch's membrane opening minimum rim width (BMO-MRW) by clock hour (*Bottom*) provides valuable additional information. There is a considerable mismatch between the clinically visible disc margin (*green dots*) and BMO (*red dots*), indicating an invisible extension of

Bruch's membrane inside the clinically determined disc margin, particularly inferiorly and nasally (note the mismatch between the red and green dots in the three inferior OCT scans). In these locations, as well as in the superior sector, which is the most suspicious, the neuroretinal rim is considerably thinner than the clinician would estimate from a disc margin-based evaluation (reproduced with license from [13] © Elsevier)

The color of the rim should also be evaluated. Even though some non-glaucomatous optic neuropathy may show optic disc cupping, a mismatch between the amount of pallor and the rim loss increases the likelihood that a non-glaucomatous optic neuropathy is present [14].

One should acknowledge that interpretation of optic disc findings is subjective and agreement is less than perfect, even among fellowship-trained glaucoma subspecialists. Most studies report only moderate agreement among specialists in detecting glaucomatous abnormality [15, 16]. Similarly, agreement in determining progressive rim loss from serial optic disc photographs has been less than ideal, with most studies showing moderate agreement [17–20]. The Glaucomatous Optic Neuropathy Evaluation Project (gone-project.com) is an interesting free online tool to exercise optic disc evaluation [21].

tion study showed that cup to disc asymmetry is significantly associated with optic disc size asymmetry and that asymmetry alone was not useful in identifying patients with glaucoma (in fact, at all levels of asymmetry [0.2, 0.3, 0.4, etc.] individuals were more likely to be normal than to have glaucoma) [23]. Therefore, when assessing asymmetry of cup or neuroretinal rim between eyes it is important to examine whether or not the optic disc size and shape are symmetrical. It is also advisable to correlate the asymmetric disc findings with other findings such as intraocular pressure (IOP) asymmetry or visual field asymmetry, even very subtle asymmetry. In our experience, the vast majority of cases referred to me as glaucoma suspects solely on the basis of optic disc cup asymmetry, without other significant findings suggestive of glaucoma, turn out to have optic disc size asymmetry accounting for the cup asymmetry.

Summary for the Clinician

- Pay attention to rim contour rather than to cup size.
- Pay attention to disc size, as it affects the apparent amount of neuroretinal rim.
- Pay attention to rim color.
- Look for corroborating findings between the rim and the nerve fiber layer.
- Acknowledge that interpretation of optic disc findings is subjective. Agreement is not perfect even among experienced fellowship-trained glaucoma specialists.

Summary for the Clinician

- Cup to disc ratio asymmetry of 0.2 or greater is part of the classic definition of glaucoma.
- Asymmetry of optic disc size and shape can give the appearance of cup to disc ratio asymmetry.
- Asymmetry of cup to disc ratios should be correlated to asymmetry in other parts of the clinical examination (i.e., IOP, visual field sensitivity, quantitative measurements of the optic nerve or RNFL).

2.4 How Much Asymmetry Between Neuroretinal Rims and Nerves Is Important?

Cup to disc ratio asymmetry of 0.2 or greater has long been held to be suggestive of glaucoma. In a variety of research studies, the definition of a glaucomatous optic disc has included asymmetry of 0.2 or greater between fellow eyes [22]. However, data from the Blue Mountains popula-

2.5 How Can I Estimate Disc Size and Compare Disc Size Between the Two Eyes?

Disc size can be estimated by a variety of methods. During clinical examination, disc size can be estimated with the direct ophthalmoscope in a technique described by Gross. The 5° aperture of the Welch-Allyn ophthalmoscope produces a

circular spot with a diameter of 1.5 mm and an area of 1.77 mm², which is slightly smaller than an average-sized optic disc, which has an approximate area of 2.1–2.7 mm² [24]. Another option, which is easier in our opinion, is to adjust the height of the slit lamp beam to coincide with the edges of the optic disc while performing biomicroscopy with handheld lenses such as the 90-diopter or contact lenses. The height of the slit beam can then be read off the scale [25, 26]. Disc size comparisons between eyes can easily be done with either one of the methods described above.

The use of automated optic disc technology, such as confocal scanning laser tomography (clinical instrument is the Heidelberg retinal tomograph—HRT), also allows for a fairly accurate, easy assessment of optic disc size and comparisons between the two eyes, however the contour line has to be correctly marked.

Summary for the Clinician

- The 5° aperture on the direct Welch-Allyn ophthalmoscope is just slightly smaller than an average-sized optic nerve head and can be used to approximate optic nerve-head size.
- During slit lamp biomicroscopy with a handheld lens, the slit beam can be adjusted to measure the height of the optic nerve heads.
- Optic nerve-head size can be easily measured with confocal scanning laser tomography.

because of the generally slow nature of the disease (which means studies require very long follow-up time), the lack of universally accepted methods to assess change (different criteria will lead to different “rates of progression”), and the fact that we cannot pinpoint the “beginning of the glaucomatous process” (therefore, any given study will contain individuals who are in different stages of their disease and probably are already undergoing change) [27].

Methods to assess change of the optic disc over time include the use of optic disc drawing comparisons, sequential optic disc photographs (mono or stereo), and quantitative and qualitative parameters on automated devices, such as confocal scanning ophthalmoscopy. In our opinion, subjective drawings are not very useful, and therefore, disc photographs or automated devices are the best options in assessing structural change in glaucoma.

Optic disc changes are more easily observed in early cases of glaucoma when the dynamic range for change is greater. In more advanced cases, the optic disc may be too damaged to appreciably note further thinning of the neuroretinal rim, and at this point in the disease it is easier to follow progression of the visual field. Data from randomized clinical trials of ocular hypertensive individuals has provided information regarding rate of optic disc change in these individuals. In the observation group of the OHTS, the cumulative probability of conversion to glaucoma over 60 months was 9.5 and 67 % of these individuals converted to glaucoma on the basis of optic disc change alone. In the European Glaucoma Prevention Study (EGPS), the cumulative probability of conversion to glaucoma in the placebo group after 60 months was 14 %, but only 37 % of the conversions occurred on the basis of optic disc changes [28]. The difference in optic disc progression rate between the OHTS and the EGPS highlights how different criteria can lead to different progression rates.

Possible glaucomatous changes over time that can occur on the optic disc include diffuse or focal thinning of the neuroretinal rim, widening or appearing of a retinal nerve fiber layer defect, and enlargement of beta-zone peripapillary atrophy. In addition to that, detection of a new optic

2.6 How Can I Look for Optic Nerve Change Over Time?

The rate of optic nerve change, similar to the rate of visual field change, is extremely variable among different patients, even in patients with similar IOP levels. It is always difficult to define rates of change

disc hemorrhage is a significant finding, probably the most significant predictor of visual field progression [29].

Summary for the Clinician

- The rate of optic nerve change is variable from one individual to the next.
- There are many barriers to detecting optic nerve change.
- Probably the best way(s) to monitor for optic nerve change is to use photo documentation and/or automated devices.
- Changes in the optic nerve are more easily detected when significant rim is available to observe the change.

2.7 If I See a Disc Hemorrhage on Healthy Appearing Neuroretinal Rim, How Soon Can I Expect to See a Change in the Rim?

In the OHTS, progressive changes occurred in only 14 % of patients with ocular hypertension who had at least one disc hemorrhage [1]. Data from the Blue Mountain study also have shown that despite a strong association between the presence of optic disc hemorrhage and established glaucoma (with visual field defect), the majority of disc hemorrhages (70 %) were found in individuals without definite signs of glaucoma [30]. Unfortunately, very few studies to date have reported on the follow-up of these “normal” individuals with disc hemorrhages. In repeated glaucoma surveys performed in the population of Dalby, disc hemorrhages were found in 28 out of 3819 individuals without glaucoma (prevalence of 0.7 %). Five out of ten of these individuals who were followed developed glaucoma with a visual field defect 2–7 years after the disc hemorrhage was noted [31].

A more common situation is the occurrence of a disc hemorrhage on a healthy appearing area of the neuroretinal rim in a glaucomatous disc. Disc hemorrhages usually occur at the infero-temporal or supero-temporal areas of the rim. Often they recur in the same area until a notch is formed, and then will start occurring at the opposite side of the same disc where the rim is still normal [6, 32–34]. Studies have shown that optic disc progression occurs in 50–80 % of patients with glaucoma following an optic disc hemorrhage, with median follow-up of 2–3 years [1, 35, 36].

Besides being a risk factor for future visual field progression, disc hemorrhages were detected more frequently on locations corresponding to the visual field sector with fastest pre-hemorrhage progression rates [37], suggesting that the hemorrhages are indicators of active ongoing glaucomatous damage. Therefore, it is important for the clinician to carefully monitor glaucoma patients after optic disc hemorrhages.

Summary for the Clinician

- Disc hemorrhages occur in non-glaucomatous eyes.
- In initially non-glaucomatous eyes, it is unclear what percent of nerves and over what period of time glaucomatous change of the optic nerve occurs after disc hemorrhage. One study found a visual field defect to occur after 2–7 years in 5 of 10 eyes that were followed.
- In optic nerves with established glaucoma, disc hemorrhages are more common.
- Disc hemorrhages are typically found in the infero-temporal or supero-temporal regions of the optic nerve.
- 50–80 % of patients with glaucoma and disc hemorrhages have been found to progress after 2–3 years of follow-up.
- 14 % of patients in the OHTS study with disc hemorrhages showed progressive neuroretinal rim loss after median follow-up of 13 months.

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