Chapter 10
Optic Nerve Head Visualization

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Optic Nerve Head Visualization [1–3]

Clinical Optic Nerve Head Visualization Techniques

The optic nerve head (ONH), or optic disc, clinically can be examined using a direct ophthalmoscope, an indirect ophthalmoscope, or a slit-lamp biomicroscope with a posterior pole lens. As the examination is uncomfortable for the patient and the time for the ONH evaluation is often short, it is essential for the examiner to have an examination strategy in mind to answer some key observations. Dilating the pupil facilitates and improves the accuracy of the examination with all instruments [4].

- ***The direct ophthalmoscope*** is cheap and portable device that gives magnified view and, when used with red-free filter, enhances detection of the nerve fiber layer of the posterior pole. However no stereo view is available and subtle changes in optic disc topography may not be evaluated sufficiently.
- ***The indirect ophthalmoscope*** is portable device that can be adjusted in specific cases as young children or uncooperative patients, patients with high myopia or media opacities. However, the magnification is limited for detailed examination, as optic nerve cupping and pallor appears less pronounced than using slit-lamp. Hereby, this method is not recommended for routine ONH examination.
- ***The slit-lamp biomicroscopy*** combined with posterior pole contact lenses ensure proper evaluation of the ONH in glaucoma patients as it provides high

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Table 10.1 Magnification of the lens and its correction factor

Lens	Correction factor (magnification)
Volk 60 D	0.88–1.0
Nikon 60 D	~1.0
Volk 90 D	1.3
Volk 78 D	1.1
Superfield NC Volk	1.5
Goldman 3-mirror	1.27

magnification, excellent illumination and a stereoscopic view. The disc diameter can be calculated by adjusting the height of the slit beam. The disc is viewed through the handheld convex lens, till the height of narrow slit beam is the same as vertical diameter of the disc, using the inner margin of the white Elschnig's ring as the reference (not the scleral rim surrounding the neural tissue). The magnification of the lens has to be noted and correction factor applied (Table 10.1). The vertical diameter of the disc, measured with the 60D lens is almost equal the height in millimeters set on the scale, as the correction factor is nearly 1 [1, 2, 5].

Non-clinical (Research) Optic Nerve Head Evaluation Techniques

With clinical ONH evaluation techniques the ONH measurement reads depends on instrument and technique used as well as eye magnification properties (corneal curvature, axial length, and refractive error). ONH can be measured independently from magnification correction error, directly during vitreoretinal surgery or histologically [6].

- ***Histomorphometry*** of histological preparations ensures irreplaceable data on structural peculiarities of the ONH, this type of research technique is complicated, time-consuming and expensive, requires specialized equipment and depends on changes in the tissue postmortem, the fixation method or the histological techniques. Measurements using this techniques has been performed only by a few researchers and ONH size ranged from 2.57 to 2.81 mm^2 in normal eyes [6].
- ***Planimetry*** is a research technique that provides quantitative manual or computerized measurements of ONH stereophotographs plotted on paper. Mean disk area ranged from 1.70 to 2.89 mm^2 in normal eyes using this technique. This method was used in large population studies and decent inter- and intra-observer agreement for experienced observers using this method was shown. However, it is a time-consuming and subjective technique as the edge of the optic disk is marked manually and the usage of refraction as the correction factor for magnification may underestimate the size of the ONH in large eyes and overestimate it in small ones [6–9].

Optic Nerve Head Clinical Examination [2, 3, 6]

During purposeful ophthalmoscopic assessment of ONH, peripapillary area and retinal nerve fiber layer possible glaucomatous damage can be detected. However, in many cases it is difficult to decide whether it is glaucomatous change or normal individual ONH appearance.

Ophthalmoscopic signs of glaucoma can be classified as generalized, focal and less specific. *Generalized signs* are described as changes of the cup (large optic cup, asymmetry of the cups and/or progressive enlargement of the cup; *focal signs* are defined as narrowing (notcing) of the rim, vertical elongation of the cup, cupping of the rim margin, nerve fiber layer haemorrhage, nerve fiber layer loss; exposed lamina cribrosa, nasal displacement of vessels, baring of circumlinear vessels, peripapillary crescent are assigned to the *less specific signs* [4].

ONH: Disc Size, Color, Tilted Appearance

The ONH size varies in population and approximately is 1.5–2.2 mm in diameter. The size of the ONH varies between races as well; on average, larger disc areas and larger cup–disc ratios are found in nonglaucomatous black individuals than in whites, although a substantial overlap exists. Refractive error has impact on ONH size as, on average, people with myopia have larger eyes and ONH and cups than do those with emmetropia or with hyperopia. The ONH size in normal tension glaucoma (NTG) eyes has particular interest considering the chance of incorrect diagnosing of NTG that may be initiated in a large disk and the cup/disc ratio patients than in those with a small ones. ONH size varies between NTG and primary open angle glaucoma patients (POAG) as some studies assessed it to be similar or larger in NTG patients compared to POAG [2, 6, 9].

Normally ONH is pink. Glaucomatous neuropathy results in pallor within the cup. Pallor of remaining tissue should alert about other nonglaucomatous optic atrophy. However, ONH pallor can occur due to previous IOP elevation.

Tilted appearance due to the oblong insertion of the optic nerve into the globe, as well ONH with optic disc drusen or coloboma, requires special considerations while assessing possible glaucoma damage. As these conditions can present visual field loss mimicking glaucomatous changes.

Neuroretinal Rim, ISNT Rule

Normally the shape of ONH is round or slightly oval with a greater vertical diameter and contains a central cup. The tissue between the cup and the disc margin is called the neural rim or neuroretinal rim (NRR).

The width of neuroretinal rim typically follows the ISNT (Inferior-Superior-Nasal-Temporal) rule: it is widest in infero-temporal sector followed by supero-temporal, nasal and then temporal sectors. However, in many eyes rim can be wider superiorly than inferiorly. This rule can be inapplicable in ONH size variations too as NRR in large ONH is relatively narrow due to more even distribution and can be misinterpreted as glaucomatous while in small OND cupping may not be evident at first and glaucoma associated changes may result in ONH flattening instead of cupping formation.

The ISNT rule can be applied in the early stage of glaucoma, as rim gets thinner in temporal-inferior or temporal-superior regions and the width of the rim in those regions can become equal with the rim width in temporal region.

Cup, Cup-Disc Ratio

The size of the physiologic cup is related to the size of the disc as for a certain amount of nerve fibers, the larger the overall disc area, the larger the cup and vice versa. Consequently, the cup-disc ratio (CDR) depends on the ONH size. The vertical cup–disc ratio is usually between 0.1 and 0.4, although nearly 5% of normal individuals have cup–disc ratio larger than 0.6. A large CDR, for example 0.7, in a large optic disc may be erroneously considered as glaucomatous instead of normal, whereas a 0.3 ratio in a very small disc may be uninterpreted as pathologic. Cup-disc ratio (CDR) alone is not adequate as a sign of possible glaucomatous damage.

Cup size between two eyes tends to be symmetrical in healthy patients. Asymmetry of the CDR of more than 0.2 occurs in less than 1% of normal individuals and it may be coherent to the ONH size asymmetry. Therefore, a difference in CDR between eyes with equal ONH size should be considered acquired damage and rise suspect of glaucoma.

Physiologically increased size of the cup may be inherited, determinated in high myopia eyes, or influenced by age, as it may increase slightly.

Differentiating physiologic cupping from acquired glaucomatous cupping can be difficult due to broad spectrum of signs of disc damage. The glaucomatous changes of the cup varies from concentric enlargement to localized tissue loss with notching of the rim, as well as other more subtle changes.

Generalized enlargement of the cup and uniform RNN thinning may be the earliest change in glaucoma, verified by serial monitoring and documenting ONH appearance by photographs or diagrams.

Focal enlargement of the cup appears as localized notching or narrowing of the NRR typically at inferior-temporal and superior-temporal poles of the ONH in early glaucomatous optic neuropathy (*ISNT rule* may be useful). If the rim widths do not follow ISNT rule, the possible existence of focal loss of rim tissue should be evaluated.

Elongation of the cup.

Following the thinning of the NRR rim at superior or inferior (or both) quadrants, the cup becomes vertically oval.

Lamina cribrosa dot sign. With glaucomatous optic neuropathy the underlying lamina becomes more evident and lamina dot sign may be an indicator of advanced glaucoma. However, this sign is not specific, as lamina pores may be seen as grayish dots in the base of the physiologic cup.

ONH vessels changes. Cup enlargement is often followed by retinal blood vessels changes as bending, bayoneting or baring of circumlinear vessels, and the central retinal artery and vein may pass more nasally. Positional vessels changes can be used in determination of progression when they are documented at the baseline and during the course of disease.

ONH haemorrhages appears as a liner red streaks (spliner shape) in nerve fiber layer on or bordering the ONH that typically clear over several weeks to months and often are followed by localized notching of the reuroretinal rim and corresponding visual field loss. ONH haemorrgahes or even repeated episodes of them occurs in one third of glaucoma patients at some time during the course of the disease, more often in patients with normal tension glaucoma. ONH haemorrhages are estimated as a risk factor for glaucoma or a marker of inadequate control, associated with disease progression and development or progression of visual field loss. Therefore patients with a splinter hemorrhage requires detailed evaluation and follow-up, most eligible with photographs.

Only 0.2% of normal population is estimated to have ONH haemorrhages that are accompanied general conditions like arterial hypertension, diabetes, usage of antiplatelet agents.

Retinal nerve fiber layer (RNFL) evaluation. In the healthy eyes, the nerve fiber layer (RNFL) surrounding peripapillary retina appears as fine silver striations and have a plush, refractile appearance. RNFL becomes less visible with age and in less pigmented fundi as well as in progressive glaucomatous neuropathy. Glaucomatous RNFL defects are best seen within two disc diameters from the ONH and can be localized to specific bundles wedge-shaped defects or diffuse (generalized) defects that are larger and have no distinct borders. Focal (wedge and slit like grooves) defects are seen as dark bands extending from the ONH margin and wider than retinal vessels (unless they are obscured by vessels) and more easily seen than generalized thinning of the RNFL. However, early wedge defects sometimes can be visible only at the distance from the ONH margin and slit-like, groove-like, or spindle-shaped focal defects narrower than retinal vessels can be seen in non-glaucomatous patients. Diffuse RNFL thinning, seen as loss of brightness and density of striations, is more common for glaucoma.

The RNFL defects clinically can be evaluated with the red-free filter and wide slit beam or a short, narrow, bright white light slit beam at high magnification to assess the peripapillary region. However, more accurate current assessment RNFL loss even in early disease stage and progression estimation are achieved with high-contrast red-free photographs.

In the early stages of nerve fiber loss, often before enlargement of the cup, NRR can be observed to become more translucent. This sign can be observed during slit-lamp examination employing a thin slit beam and confining it to the disc surface.

Peripapillary atrophy, which occurs in 2 types, may be significant in glaucoma and predict early damage in patients with ocular hypertension. Both zones usually are located in the temporal ONH side (more inferotemporally than superotemporally).

Alpha-zone peripapillary atrophy (outer) is superficial retinal pigment epithelial changes (hyperpigmentation and hypopigmentation) is presented in almost any eye and has no known impact on glaucoma.

Beta (inner) zone is characterized by visible sclera and large choroidal vessels regarding chorioretinal atrophy, is presented in approx. 25% of normal eyes and in significantly higher ranges in glaucomatous eyes as well as the higher risk of further glaucoma progression. The location of this atrophy often corresponds the defects in the visual field tests. However, according European Glaucoma Society Guidelines, in everyday clinical practice, a large Beta zone (in particular in non myopic eyes) should not be assessed as definite sign of glaucoma, simply as an extra-clue.

Manual ophthalmoscopic ONH examination remains a mainstay in the evaluation of a glaucoma patient, however more sophisticated devices such as scanning laser polarimetry, confocal scanning laser ophthalmoscopy, and ocular coherence tomography are used to complement the clinical examination with the quantitative measurements.

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