ABSTRACT
BACKGROUND: Primary open angle glaucoma is characterized by quantifiable parameters including the intraocular pressure, the aqueous outflow facility, and geometric measurements of the optic disc and visual defects. Despite this, diagnosis remains controversial.

OBJECTIVE: To review the current concepts in the diagnosis of primary open angle glaucoma and adapt them to situations where high technology facilities are lacking.

DATA SOURCE: Information was obtained from journals/medline, Hinari, the American Academy of Ophthalmology preferred practice pattern CD, and reputable textbooks using publications from 1972 to 2007.

RESULTS: The Preferred Practice Pattern Committee of the American Academy of Ophthalmology recommends that the comprehensive initial glaucoma evaluation (history and physical examination) includes comprehensive adult eye evaluation with special attention to those factors that specifically bear on the diagnosis such as the optic disc, nerve fibre layer and visual field evaluation, open anterior chamber angles on gonioscopy and absence of secondary causes of glaucoma. Intraocular pressure is no longer relied on in the diagnosis of primary open angle glaucoma. Sequential evaluation of optic disc cup and size, neuroretinal rim size and shape, retinal nerve fiber layer, presence of peripapillary atrophy, and presence of retinal or optic disc haemorrhages enhance the ability to detect glaucomatous damages.

CONCLUSION: A simple systematic approach in examination of the optic discs and visual field will improve accurate diagnosis of glaucoma. WAJM 2009; 28(3): 141–147.

Key words: Glaucoma, diagnosis, optic disc, visual fields, review article.

RéSUMÉ
CONTEXTE: le glaucome de montage ouvert primaire est caractérisé par les paramètres faciles à évaluer en incluant la pression intraoculaire, la facilité d’écoulement aqueuse et les mesures géométriques du disque optique et des défauts visuels. En dépit de cela, le diagnostic reste controversé.

OBJECTIF : reconsidérer les concepts actuels dans le diagnostic de glaucome de montage ouvert primaire et les adapter aux situations où l’équipement de technologie de pointe manque.


RÉSULTATS: le Comité de Dessin de Pratique Préféré de l’Académie américaine d’Ophtalmologie recommande que l’évaluation de glaucome initiale complète (l’histoire et l’examen physique) inclut l’évaluation d’œil adulte complète avec l’attention spéciale à ces facteurs qui ont un effet spécifiquement sur le diagnostic tel que le disque optique, la couche de fibre de nerf et l’évaluation de terrain visuelle, ouvrent des angles de chambre antérieurs sur gonioscopy et absence de causes secondaires de glaucome. On ne compte plus sur la pression intraoculaire dans le diagnostic de glaucome de montage ouvert primaire. L’évaluation séquentielle de tasse de disque optique et de grandeur(taille), neuroretinal la grandeur(taille) de bord et la forme, retinal la couche de fibre de nerf, la présence d’atrophie de peripapillaire et la présence de retinal ou d’hémorragies de disque optiques améliore la capacité de découvrir des dommages de glaucomatous.


Mots clé: le Glaucome, le diagnostic, le disque optique, les champs visuels, reconsidère l’article.
INTRODUCTION

Glaucoma is amongst one of the most controversial subjects in medicine.1 Primary open angle glaucoma is characterized by the most quantifiable parameters of any ophthalmic entity. These include the intraocular pressure, the aqueous outflow facility, and a variety of geometric measurements of the optic disc, as well as, a large number of ways to test and describe the visual defects. Despite this, most aspects of glaucoma remain a matter of debate and research.1,2 The definition, etiology, diagnostic criteria and choice of therapy in primary open-angle glaucoma are still debatable.1,2 The diagnosis of this condition in particular may be difficult especially in early stages. Terminologies such as ‘low or normal tension glaucoma’ for glaucomatous optic nerve damage in the absence of increased intraocular pressure; ‘ocular hypertension’ for increased Intraocular pressure in the absence of glaucomatous optic nerve damage and ‘glaucoma suspect’ for individuals with clinical findings and/or a constellation of risk factors that indicate an increased likelihood of developing primary open angle glaucoma, only serve to show the diagnostic difficulties in glaucoma.3

Primary open angle glaucoma is a progressive, chronic optic neuropathy in adults where intraocular (IOP) and other currently unknown factors contribute to the damages and the absence of other identifiable causes. There is a characteristic acquired optic atrophy of the optic nerve and loss of retinal ganglion cells and their axons. These are associated chamber angle that is open by gonioscopic appearance.

In Nigeria, diagnosis is frequently easy because many patients present in the advanced stages of the disease with poor vision, marked optic disc cupping, atrophy and visual loss.3-4 Unfortunately, by the time characteristic visual field defects are demonstrable, a third or more of the optic nerve is already damaged and patients complains of significant field loss, probably 10% or fewer axons remain.2 Thus, early diagnosis is important in prevention of irreversible visual loss from glaucoma. It is in these early stages that there are diagnostic difficulties. This paper will concentrate on the current concepts in the diagnosis of glaucoma.

Comprehensive Initial Glaucoma Evaluation

The Preferred Practice Pattern Committee of the American Academy of Ophthalmology4 recommends that the comprehensive initial glaucoma evaluation (history and physical examination) includes all components of the comprehensive adult eye evaluation in addition to and with special attention to those factors that specifically bear upon the diagnosis, causes and treatment of primary open-angle glaucoma (POAG). Completion of the evaluation may require more than one visit. For instance, an individual might be identified as having glaucoma at one visit, but may return for further evaluation, including additional intraocular pressure (IOP) measurements, central corneal thickness determination, visual field assessment, and optic nerve head evaluation and documentation.

History and Physical Examination

This includes a review of ocular, family and systemic history. It also includes an assessment of the impact of visual function on daily living and activities. Review of pertinent records with particular reference to the status of the optic nerve, visual field, IOP; ocular surgery; the use of ocular, and systemic medications, are known local or systemic intolerance to glaucoma medications; family history of glaucoma, including history of visual loss from glaucoma.5 The pupil’s are examined for reactivity and an afferent pupillary defect. A slit-lamp biomicroscopic examination of the anterior segment can provide evidence of physical findings associated with narrow angles, corneal pathology, or a secondary mechanism for elevated IOP such as pseudo exfoliation, pigment dispersion, iris and angle neovascularization, or inflammation.6

Optic Nerve Head and Retinal Nerve Fiber Layer

Histological studies performed by Quigley ET al7 have shown that there can be a significant loss of ganglion cells before evidence of functional loss on conventional achromatic visual field testing. For this reason attention has been focused on alternative, more sensitive ways of detecting early ganglion cell damage that is possible with white-on-white perimetry. The newer psychophysical and electrophysiological techniques, has shown that abnormalities in the .appearance of the optic disc and retinal nerve fiber layer may precede visual field defects.10-17 These abnormalities include7 diffuse thinning, focal narrowing, or notching of the optic disc rim especially at the inferior or superior poles; documented progression of cupping of the optic disc; diffuse or localized abnormalities of the peripapillary retinal nerve fibre layer especially at the inferior and superior poles; disc rim or peripapillary retinal nerve fibre layer hemorrhages and optic disc neural rim asymmetry of the two eyes, consistent with loss of neural tissue. In a study performed to evaluate the diagnostic power of these optic nerve head variables, the best variables to separate between the normal subjects and the individuals with ocular hypertension who have nerve fiber layer defects were the vertical cup-to-disc diameter ratio corrected for optic disc size, the total neuroretinal rim area, the rim-to-disc area ratio corrected for optic disc size, and the cup-to-disc area ratio corrected for disc size.18 Interestingly, the vertical cup-to-disc diameter ratio corrected for disc size was one of the best variables. The cup-to-disc diameter ratios had partially lost their importance in the clinical diagnosis of glaucoma because it became apparent that the cup-to-disc diameter ratios depend on the disc size.19-21

A high cup-to-disc diameter ratio can be normal, if the optic disc is large,22 and a low cup-to-disc diameter ratio can be glaucomatous if the optic disc is small.23 The cup-to-disc diameter ratio in the clinical diagnosis of glaucoma is very useful if the dependence of the cup-to-disc diameter ratios on the disc size is taken into account.19 The clinical value of the vertical cup-to-disc diameter ratio corrected for disc size is further emphasized by the fact that the cup-to-disc diameter ratios and the disc size can be determined by a slit lamp examination, whereas neuroretinal rim area and the rim-to-disc area ratio as the two other
important optic disc variables have to be determined by time-consuming and sophisticated techniques such as morphometric evaluation of fundus photographs or co focal scanning laser tomography imaging of the optic nerve head. It means for the setting of a busy glaucoma practice, in which co focal tomography laser scanning systems have not been introduced, that the optic disc size can be measured ophthalmoscopically using a fundus viewing lens and a slit lamp, with which the length of the beam can be adjusted to the diameter of the optic disc.

After correction for the measured disc size, the vertical cup-to-disc diameter ratio is one of the most important variables to describe the status of the optic nerve in glaucomatous eyes.

For the clinical description of an optic nerve, it may thus be acceptable to give the vertical cup-to-disc diameter ratio in combination with the estimated disc size. The reason that the corrected vertical cup-to-disc ratio was one of the best variables for the early detection of glaucomatous optic nerve damage may be the pattern of glaucomatous loss of neuroretinal rim. Rim loss usually starts predominantly in the inferior and superior optic disc regions. It leads to a vertical elongation of the optic cup. It also explains why the vertical cup-to-disc diameter ratio is superior to the horizontal cup-to-disc diameter ratio in differentiating the preperimetric glaucoma patients from the normal control individuals.

**Optic Nerve Head and Retinal Nerve Fiber Layer Evaluation**

The preferred technique for optic nerve head and retinal nerve fiber layer evaluation involves magnified stereoscopic visualization (as with the slit-lamp biomicroscope), preferably through a dilated pupil. Direct ophthalmoscopy is useful in some cases to complement magnified stereoscopic visualization, providing more comprehensive information of optic nerve detail due to the greater magnification of the direct ophthalmoscope. Red-free illumination may aid in evaluating the retinal nerve fiber layer. Inability to dilate the pupil should be documented. Examination of the fundus, through a dilated pupil whenever feasible, includes a search for other abnormalities that might account for visual field defects (such as optic disc pallor, tilted disc, disc drusen, optic nerve pits, optic nerve hypoplasia, neurological disease, macular degeneration, and other retinal disease). Important points to note during fundoscopy include the following.

The hallmark of glaucomatous optic neuropathy is excavation of the neuroretinal rim (cupping and atrophy). Advanced glaucomatous cupping can result in a pale optic disc but disc pallor should raise a suspicion of another cause such as optic atrophy:

- Colour difference should not be used to distinguish the cup edge: change in direction of the blood vessels is a more reliable indicator; the size of the cup always appears smaller when viewed monoscopically than stereoscopically, a measurement of cup/disc ratio (CDR) alone is insufficient and may be misleading as small discs will have smaller cups and hence smaller CDR. It should be related to the disc size. The inferior rim is usually thicker than the superior rim, which is thicker than the nasal rim, and the temporal rim is the thinnest (this is known as the ‘ISNT’ rule). The glaucomatous optic disc does not obey the ‘ISNT’ rule.

- Diffuse or localized retinal nerve fibre dropout. Early localized damage is characterized by slit or wedge-shaped defects in the retinal nerve fibre layer. As glaucomatous damage progresses the defects become larger and at the end-stages glaucoma, there is total atrophy of the nerve fibre layer characterized by complete baring of the larger retinal blood vessels, which run in this layer. The atrophic area appears darker and mottled because of enhanced visualization of the retinal pigment epithelium.

- Glaucogmatous cups are usually larger than physiological cups, although a large cup is not necessarily pathological. Assessment of the symmetry, thickness and colour of the neuroretinal rim is more important.

- The spectrum of disc damage in glaucoma ranges from highly localized tissue loss with notching of the neuroretinal rim (polar notching) associated with small areas of parapapillary atrophy or choroidal sclerosis (type 1) to diffuse concentric enlargement of the cup with thinning of the neuroretinal rim (type 4). Within the spectrum is found the myopic glaucomatous disc (type 2) characterized by polar notching and a large temporal crescent in the absence of degenerative myopia and the senile sclerotic disc (type 3) characterized by a shallow, saucered cup and gently sloping neuroretinal rim, a ‘moth-eaten appearance’ and para papillary atrophy or choroidal sclerosis surrounding the circumference of the nerve.

There is a correlation between para papillary atrophy and glaucoma. Atrophy surrounding the optic disc is conceptualized as consisting of two zones: an inner ‘beta’ zone, bordering the disc margin, which in turn is concentrically surrounded by an outer ‘alpha’ zone. The alpha zone which displays variable irregular hyper- and hypo pigmentation of the retinal pigment epithelium is larger in patients with primary open-angle glaucoma but its frequency is similar in glaucomatous and normal subjects. However, the beta zone is not only larger but occurs more frequently in patients with primary open-angle glaucoma than in normal individuals. The beta zone exhibits chorioretinal atrophy with visibility of the sclera and large choroidal blood vessels.

**Optic Nerve Head Documentation**

Optic nerve head evaluation and documentation can be done by imaging, photography, or drawing. Colour stereo photography and computer-based image analysis of the optic nerve head and retinal nerve fiber layer are the best currently available methods to document optic disc morphology and should be performed. The quantitative 3-dimensional reconstruction of the optic nerve head is very useful for the diagnosis of glaucoma. Limitations in optic disc and retinal nerve fibre layer assessment have stimulated the development of imaging devices that measure either the optic disc cup and neuroretinal rim area or the retinal nerve fibre layer. These include scanning laser tomography and scanning laser polarimetry (retinal nerve fibre analyzer). They offer greater
objectivity but are limited by potential sources of error and so the results must still be interpreted in association with clinical findings. This quantitative imaging may be useful in early diagnosis before obvious visual field loss occurs and may allow increased sensitivity to detect progression of the condition.

Co-focal scanning laser ophthalmoscopy (CSLO), scanning laser polarimetry (SLP), and optical coherence tomography (OCT) have been shown to be able to discriminate eyes with ocular hypertension (OHT), glaucoma-suspect eyes (GSE) or early glaucomatous eyes (EGE) from normal eyes. In the absence of these technologies, a nonstereoscopic photograph or a detailed drawing of the optic nerve head should be recorded, but these are less desirable alternatives to stereo photography or computer-based imaging. However, in developing countries like Africa, a detailed drawing of the optic nerve head may be achieved with the aid of a slit lamp biomicroscope and a fundus viewing lens.

Evaluation of the Visual Field

Automated static threshold perimetry is the preferred technique for evaluating the visual field. Careful manual combined kinetic and static threshold testing is an acceptable alternative when patients cannot perform automated perimetry reliably or if it is not available. Glaucomatous visual field defects include, visual field defects consistent with retinal nerve fibre layer damage (nasal step, arcuate field defects, paracentral depression in clusters of test sites, etc); visual field loss in upper hemifield that is different compared with the lower hemifield ie across the horizontal midline (in early or moderate cases); they are reproducible and there are no other explanations for them.

New fast test visual field strategies, such as SITA (Swedish Interactive Threshold Algorithm), have become available which improve patient test compliance. Computerized programmer for serial visual field analysis (PROGRESSOR), which assess progression of disease by accounting for test variability are available. Other modes of testing which involve motion detection, frequency-doubling technology (FDT) and short-wavelength automated parameterizes (SWAP) detect visual field defects before standard automated perimetry in patients with preperimetric glaucoma and may enable earlier diagnosis. In a report by the American Academy of Ophthalmology, the four automated perimetry techniques that were assessed were short wavelength automated perimetry (SWAP), frequency doubling technology perimetry (FDT), high-pass resolution perimetry (HPRP), and motion automated perimetry (MAP).

The algorithms described were the Swedish interactive threshold algorithm (SITA) and SITA fast. With the exception of SWAP, these techniques and algorithms reduce testing time and inconsistent patient performance when compared with conventional full threshold testing. Short wavelength automated perimetry detected visual field loss earlier than standard threshold automated perimetry, with a sensitivity and specificity of about 88% and 92% respectively. However, it is a lengthy demanding test that is sensitive to media opacities, and has a greater magnitude of long-term fluctuation compared with standard threshold automated perimetry, which make it difficult to assess disease progression accurately. When compared to standard threshold automated perimetry, FDT perimetry showed sensitivity and specificity greater than 97% for detecting moderate and advanced glaucoma, and sensitivity of 85% and specificity of 90% for early glaucoma. As FDT perimetry has a short testing time and is resistant to blur and pupil size, it may be a useful screening tool.

When Fast Swedish interactive threshold algorithm (SITA) short-wavelength automated perimetry (SWAP), lengthier full-threshold SWAP, and standard automated perimetry (SAP) using the SITA Fast program were used to detect early glaucomatous visual field loss, the SITA SWAP identified at least as much glaucomatous visual field loss as the older full-threshold SWAP, although test time was considerably reduced. Conventional SAP using SITA Fast was not significantly less sensitive than either of the 2 SWAP programs. Causes of visual field loss other than glaucomatous optic neuropathy should be sought and assessed during the history review and physical examination. It is important to use a consistent examination strategy when visual field testing is repeated.

Intraocular Pressure

Glaucoma has traditionally been defined by the triad of increased intraocular pressure, optic disc changes, and visual field defects. Intraocular pressure (IOP) is now regarded as a causative risk factor, not a diagnostic factor since some patients with characteristic POAG will have normal pressures while some with elevated IOP will not have glaucomatous damage. IOP can be measured in a variety of ways including contact (indentation and applanation) and non-contact techniques. Various sources of error, including central corneal thickness (CCT) and structural corneal rigidity, have been proposed for Goldmann applanation tonometry. Corneal rigidity is a major source of error in indentation techniques (Schiotz Tonometer).

Goldmann applanation tonometry (GAT), first introduced in 1957, is still regarded as the gold standard for measuring IOP. However, its accuracy has been questioned in eyes with abnormal central corneal thickness and structural rigidity, and in those that have undergone corneal ablation with laser refractive surgery. Goldmann applanation tonometry was calibrated for a CCT of 520μm, and it underestimated IOP in eyes with thin corneas and overestimates IOP in eyes with thick corneas, with several clinical implications. The Pascal dynamic contour tonometer (DCT) is a novel device designed for IOP measurements assumed to be largely independent on CCT and corneal curvature. When Goldmann applanation tonometer was compared with the TonoPen and DCT, agreement with GAT measurements was higher for Pascal DCT than for TonoPen readings; however, Pascal DCT significantly overestimated IOP values compared with GAT. Measurements of IOP obtained with both Pascal DCT and the TonoPen appeared to be influenced by CCT, and this influence appeared to be greater for the latter. In structurally normal thin corneas DCT may give a more
accurate assessment of the true IOP but it does not appear to have any benefit over GAT in thick corneas. The corneas with steeper curvature also cause higher corneal rigidity and produce more overestimation of non-contact tonometer (NCT) measurement, while they have stronger capillary attraction of the precorneal tear film for the GAT tip and also produce overestimation of GAT measurement. As a result, [NCT/GAT] is believed not to be influenced by the corneal curvature. However, dynamic contour tonometry measurements in African Americans seem to provide an estimate of IOP that is less influenced by corneal properties (including corneal thickness and curvature) than those provided by GAT. IOP is measured in each eye, preferably using a contact application method (typically a Goldmann tonometer) before gonioscopy or dilation of the pupil. Time of day should be recorded because of diurnal variation. The assessment may benefit from determining diurnal IOP fluctuations, either on the same day or on different days, which may be indicated when disc damage exceeds the amount expected based on a single IOP measurement.

Central Corneal Thickness

Measurement of central corneal thickness (pachymetry) aids the interpretation of IOP measurement results and stratification of patient risk. Measurement methods include ultrasonic and optical pachymetry. Central corneal thickness influences, and is a source of error in IOP measurement. Patients with thicker corneas will have overestimated IOPs and may be misclassified as having ocular hypertension. By contrast, the relative under measurement of IOP by GAT in thinner corneas or in corneas following laser refractive surgery may delay the early detection and treatment of glaucoma.

The thick cornea has more influence on the measurement with non-contact tonometer than Goldmann applanation tonometry, because IOP is measured with NCT over a wider applanation area.

Direct and Indirect Gonioscopy

The diagnosis of POAG requires careful evaluation of the anterior-chamber angle to exclude angle closure or secondary causes of IOP elevation, such as angle recession, pigment dispersion, peripheral anterior synechiae, angle neo-vascularization, and trabecular precipitates.

Gonioscopy can be done by the aid of direct and indirect gonioscopes. Indirect gonioscopy is the preferred technique for diagnosis. There are two main types: the Zeiss 4 mirror contact lens and the Goldmann gonio lens which may be a single mirror lens or one of the lenses of the 3 mirror lens. The 4-mirror lens has the advantage that it can visualize the entire 360° of the anterior chamber angle with minimal rotation and it does not require a coupling agent.

Functional Acuity Contrast Test (FACT)

The FACT Chart (Figure 1) is an accurate and comprehensive sine-wave
grating chart that tests functional visual acuity. A recent study has been conducted to investigate spatial-contrast sensitivity (CS) assessment as a tool for diagnosis of early glaucoma in patients with good visual acuity. It was found that both the presence of a significant difference between the CS of glaucoma patients and control subjects and a high specificity of contrast sensitivity suggests that the FACT test may be used as a tool for diagnosis of patients with glaucoma, besides other methods such as short-wavelength automated perimetry (SWAP).

**CONCLUSION**

Ophthalmoscopical evaluation of the optic disc is a feasible and largely accessible method to diagnose glaucoma. Considering individual variations in the details of topography or tissue components damaged by the glaucomatous process, however, adequate identification of glaucomatous optic disc signs requires training and experience. Adequate guidelines of optic disc examination are required so that the ophthalmologist does not miss important aspects that could lead to adequate diagnosis or identification of progression in a patient with established glaucoma. Optic disc qualitative parameters are better than quantitative parameters in separating glaucomatous from normal eyes. The sequential evaluation of optic disc cup and size, neuroretinal rim size and shape, retinal nerve fiber layer, presence of peripapillary atrophy, and presence of retinal or optic disc hemorrhages enhances the ability to detect glaucomatous damage and its progression. Automated static threshold perimetry is the preferred technique for evaluating the visual field. Careful manual combined kinetic and static threshold testing is an acceptable alternative when patients cannot perform automated perimetry reliably or if it is not available. Newer techniques such as short-wavelength automated perimetry can diagnose glaucoma earlier than standard automated perimetry. Raised intraocular pressure should not be relied on as the only triggering factor in glaucoma investigations. A simple systematic approach may allow improved diagnosis and management of glaucoma.

**REFERENCES**


