Angle-closure glaucomas — posterior (pushing) mechanisms other than pupillary block

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Abstract

Angle-closure is often erroneously considered synonymous with pupillary block, the most common mechanism leading to acute or chronic iridocorneal apposition. However, abnormalities exist in structures and/or forces other than those related to pupillary block and may cause apposition despite a patent iridotomy. A 4-point classification of these mechanisms has been described, and includes mechanisms originating in the ciliary body, lens, or posterior segment. This review outlines the characteristics of angle-closure specific to each of these 3 levels.

Key words: Ciliary body, Glaucoma, angle-closure, Lens, crystalline, Pupil disorders

Introduction

Angle-closure is often erroneously considered synonymous with pupillary block, the most common mechanism leading to acute or chronic iridocorneal apposition. Other abnormalities exist in structures and/or forces other than those related to pupillary block and may cause apposition despite a patent iridotomy.1-4 Ritch et al5 and Tello et al6 have described a 4-point classification of these mechanisms and, in addition to pupillary block occurring at the level of the iris, have included mechanisms originating in the ciliary body, lens, or posterior segment. Each mechanism is characterized by specific clinical signs, the recognition of which permits specific treatment directed at the underlying pathophysiology. In this review, we outline the characteristics of angle-closure specific to each of the 3 levels, aware that various mechanisms for angle closure may coexist, complicating the diagnosis.

Accurate diagnosis of the specific type of angle closure relies on 2 modalities. The first is darkroom indentation gonioscopy. If discernable angle widening occurs after pressure is applied to the central cornea during gonioscopy, reversible iridocorneal apposition is present. In addition, the behavior of the iris during indentation gonioscopy is specific to the level at which angle-closure originates, as detailed below.

To correctly discern a narrow, but open, angle from a closed one, gonioscopy must be performed in a completely darkened room, as light reflex–induced miosis may prevent apposition of the iris to the angle wall (Figure 1).7,8 For this reason, the smallest slit beam should be used, ensuring that it does not project into the pupil. The narrowest quadrant is usually the superior angle, so it is logical to begin the examination at this location.5 The quadrant is first examined without pressure on the cornea and with the patient looking toward the mirror, allowing the examiner to see as deeply
into the angle as possible. Corneal indentation is subsequently performed and any changes in angle and iris configuration are noted.

The other modality relates to objective quantification of angle and anterior chamber anatomy. High-frequency ultrasound biomicroscopy (UBM) is currently the most established device, providing objective high-resolution images of angle structures and visualization of structures in the posterior chamber that are otherwise hidden from clinical observation. Anterior segment optical coherence tomography (AS-OCT) is another option, and advantages over UBM include its non-contact nature, with the ability to image the eye in the sitting position. Due to its reliance on light rather than sound waves, AS-OCT cannot image structures posterior to the pigment epithelium of the iris and ciliary body. High-frequency UBM and AS-OCT imaging are used as adjunctive tools for establishing the diagnosis of angle-closure in posterior pushing mechanisms. However, an important aspect of the diagnosis and management of these mechanisms is dark room gonioscopic evaluation of the angle following laser iridotomy, which must be performed for all patients.

**Angle-closure originating at the level of the ciliary body**

Angle-closure at the level of the ciliary body is caused mainly by plateau iris syndrome, first described by Tornquist in 1958. Plateau iris configuration denotes an iris root that is sharply angulated forward and then centrally. The iris surface appears flat (hence the term 'plateau'), and the anterior chamber is of relatively normal depth. Primary plateau iris is associated with a large or anteriorly positioned ciliary body that physically supports the iris root against the trabecular meshwork (Figure 2) and, during dynamic gonioscopy, no posterior movement of the mid-peripheral iris occurs. Gonioscopy reveals the ‘double hump’ sign (Figure 3), or the slit beam follows the convex curvature of the iris to its deepest point at the equator of the lens, and then rises again over the ciliary process before sharply dropping peripherally. This anatomic configuration prevents posterior fall of the iris following iridotomy, and can persist even after lens extraction.

The term plateau iris syndrome was coined by Wand et al in 1977 and refers to the development of angle-closure, either spontaneously or after pupillary dilatation, in an eye with plateau iris configuration despite the presence of a patent...
iridotomy (Figure 2). Greater force is needed with gonioscopy to displace the ciliary processes and open the appositionally closed angle than in pupillary block.

As plateau iris syndrome results in angle-closure, further treatment needs to be considered, whereas eyes with plateau configuration only require follow-up. In eyes with plateau iris syndrome, whether or not the intraocular pressure (IOP) rises during angle-closure depends on the level of iridocorneal apposition with respect to the angle structures, or the ‘height’ to which the plateau rises.16 If the angle closes completely to the upper trabecular meshwork or Schwalbe’s line, the IOP will rise, whereas if the angle closes partially, leaving the upper portion of the filtering meshwork open, the IOP will not rise. The latter situation is far more common and is clinically significant because these patients may develop peripheral anterior synechiae (PAS) years after a successful iridotomy produces what appears to be a well-opened angle. Although infrequent, some patients with plateau iris syndrome may develop acute angle closure that persists despite, or subsequent to, iridotomy.3,17-19

Patients with plateau iris tend to be female, young, and less hyperopic than those with relative pupillary block, and often have a family history of angle-closure glaucoma. In a series of 67 patients with angle-closure developing at the age of 40 years or younger, 35 had plateau iris syndrome.20 Of these, one had high axial myopia, a finding that should not rule out angle-closure and routine gonioscopy.21 Progressive enlargement of the lens with age can cause further narrowing of the angle, reflecting a greater component of pupillary block causing the iris to be less ‘plateau’ and more convex. Periodic gonioscopy is indicated for these patients.

The goal for eyes with iridocorneal apposition secondary to plateau iris syndrome is to prevent the development of PAS, both chronic and acute angle-closure, and intermittent IOP elevations occurring in the dark. Iridotomy may result in an open angle, but if apposition persists despite a patent iridotomy (Figure 2), argon laser peripheral iridoplasty (ALPI) is indicated. This procedure, which has good long-term results, compacts and contracts the peripheral iris stroma and opens the angle (Figure 2).22-25 In a study of ALPI, 20 of

23 eyes (87%) had open angles throughout the entire follow-up period of 72 to 188 months after a single treatment. In the 3 remaining eyes, the angles were readily re-opened and maintained open by a single repeat treatment. No filtration surgery or progression of PAS was documented in any eye.

Pilocarpine can be used to thin the iris and widen the angle, an effect visualized well with gonioscopy. The lowest effective concentration of pilocarpine should be chosen and consideration should be given to using it only at night. Long-term non-occludability of the angle with a single treatment of ALPI is preferable, since it produces a more stable angle and avoids the side effects of long-term use of pilocarpine, particularly in younger individuals.

Secondary, or pseudoplateau, iris is caused by lesions such as iris or ciliary body cysts, leading to enlargement of the ciliary body.26 The clinical appearance of the iris and angle is similar to that seen in plateau iris syndrome, with the exception that these lesions typically involve only part of the angle circumference (Figure 4).20,27 Solid lesions of the ciliary body such as tumor or inflammation can also produce a pseudo-plateau iris configuration, especially when there is homogenous full-circumference infiltration. Although these lesions can sometimes be seen through the well-dilated pupil, UBM demonstrates the discrete lesion and is extremely helpful for making a definite diagnosis.

Angle-closure originating at the level of the lens

When the lens physically compresses the iris and ciliary body against the trabecular meshwork, lens-related angle-closure, or phacomorphic angle-closure or glaucoma, is said to occur (Figure 5). This is distinct from open-angle lens-related increases in IOP or ‘phacolytic’ or ‘phacoanaphylactic’ glaucoma.

Most commonly, phacomorphic angle-closure occurs secondary to progressive age-related increases in lens size.
In these eyes, a component of pupillary block that increases with progressive lens thickening may be present, requiring smaller degrees of peripheral iris bowing to close the angle. Early management with laser iridotomy allows for better evaluation of the lens-induced component. When indentation gonioscopy is performed in an eye with mainly pupillary block, the iris convexity is easily flattened and the irido-trabecular apposition relieved. However, in a lens-induced mechanism, the lens physically supports the iris. Hence, the iris can be seen to closely drape over the lens in a volcano-like configuration, with the pupil as the crater (Figure 6). Greater force is needed during indentation not only to force aqueous into and open the angle, but also to displace the lens posteriorly.

Swelling of the lens may cause acute angle-closure with markedly increased IOP. Again, in these cases, pupillary block may be an associated factor and LPI may help to control IOP, at least initially.

The identification of a phacomorphic component to angle-closure has specific implications for treatment. Pilocarpine should be used with extreme caution, as it contracts the longitudinal ciliary muscle and causes the lens to become thicker axially and also to move anteriorly, even in elderly patients. We have described a highly myopic eye in which pilocarpine caused reversible angle closure. If pilocarpine is used in these eyes, IOP and angle width should be assessed after a single application. If angle narrowing and/or increase in IOP are observed, pilocarpine should be discontinued. In patients for whom surgery is not immediately planned, cycloplegics (preferably cyclopentolate) may bring about angle widening.

If lens-related appositional angle-closure persists after laser iridotomy, ALPI (if no visually significant lens opacification is present) or lens extraction may lead to widening of the angle and lowering of IOP. With complete or near-complete synechial angle-closure, angle width or IOP will not be affected. If IOP is uncontrolled by medications, trabeculectomy or combined cataract extraction and trabeculectomy is indicated. In cases where synechial closure is of recent onset and short duration, goniosynechialysis may be attempted. In combination with lens extraction, goniosynechialysis is effective in patients with acute or subacute primary angle-closure uncontrolled by medical and laser treatment.

In the management of lens-related acute angle closure, ALPI may be effective in lowering IOP prior to iridotomy and lens extraction. The latter may be performed without prior iridotomy.

**Angle-closure originating posterior to the lens**

This category encompasses a group of conditions that are less common and pathophysiological understood, and more therapeutically challenging. These conditions are characterized by a shallow anterior chamber, angle-closure, and increased IOP persisting after initial medical therapy and iridotomy. First coined in 1869 by von Graefe, these conditions are commonly referred to as ‘malignant glaucoma’, (Figure 7); however, this term needs review as several discrete conditions may cause this clinical picture and are treated differently.
The majority of these cases have been observed after filtration surgery in eyes with angle-closure glaucoma. The exact mechanism remains unclear, but Shaffer suggests that a pressure differential is created between the posterior and anterior segments, and the iris-lens diaphragm and ciliary body are pushed anteriorly, resulting in shallowing of the anterior chamber and angle closure.\textsuperscript{58}

Initial medical therapy includes lowering IOP with aqueous suppressants, shrinking the vitreous with hyperosmotic agents, and posterior displacement of the lens-iris diaphragm with cycloplegics. If unsuccessful or if lens-cornea contact occurs, Nd:YAG laser disruption of the anterior vitreous face is warranted, after confirmation of a patent iridotomy. The theory of aqueous misdirection is often confirmed by immediate deepening of the anterior chamber and a decrease in IOP, with establishment of direct vitreo-aqueous communication.\textsuperscript{59-63} When laser hyaloidotomy is not possible or unsuccessful, vitrectomy should be performed. Vitrectomy alone is often less successful than its use combined with excision of part of the lens capsule or zonules. The latter usually establishes vitreo-aqueous communication, further supporting the proposed mechanism.\textsuperscript{64-68}

UBM has led to the description of clinical cases indistinguishable from malignant glaucoma. These cases, in which the accumulation of supraciliary fluid leads to the detachment and anterior rotation of the ciliary body and consequent angle-closure (Figure 8), may be part of diffuse choroidal detachment observable with B-scan ultrasound,\textsuperscript{69} or a subtle finding observable only with UBM.\textsuperscript{70} Medical therapy only may be indicated for these patients, highlighting the need for careful diagnosis.

Ciliary body swelling and/or anterior suprachoroidal effusion can be a manifestation of well-defined clinical conditions including posterior scleritis,\textsuperscript{71} central vein occlusion,\textsuperscript{72-74} and following panretinal photocoagulation.\textsuperscript{75} Infrequently, these conditions lead to angle-narrowing or angle-closure.

Bilateral anterior chamber shallowing and angle-closure has been associated with idiosyncratic drug reactions and systemic inflammatory conditions.\textsuperscript{76-79} UBM has enabled the identification of ciliary body detachment as the underlying mechanism.\textsuperscript{80,81}

Angle closure has been reported after scleral buckling.\textsuperscript{21,82} It has been proposed that when pupillary block is absent, the underlying mechanism is detachment and/or edema of the ciliary body resulting in anterior displacement and angle-closure. If diagnosed early in the postoperative period, PAS and chronic angle-closure can be prevented with ALPI.\textsuperscript{83}

Rarely, tumors in the posterior segment can present with angle-closure.\textsuperscript{84,85} A tumor should be suspected when the fellow eye has a wide open angle and the 2 eyes have similar refraction. If the posterior segment cannot be visualized, imaging with ultrasound is required.

References

8. Gazzard G, Foster PJ, Friedman DS, Khaw PT, Seah SK.


47. Lai JS, Tham CC, Chan JC. The clinical outcomes of cataract extraction by phacoemulsification in eyes with primary angle-closure glaucoma (primary angle-closure glaucoma) and co-existing cataract: a prospective case series. J Glaucoma. 2006;15:47-52.


