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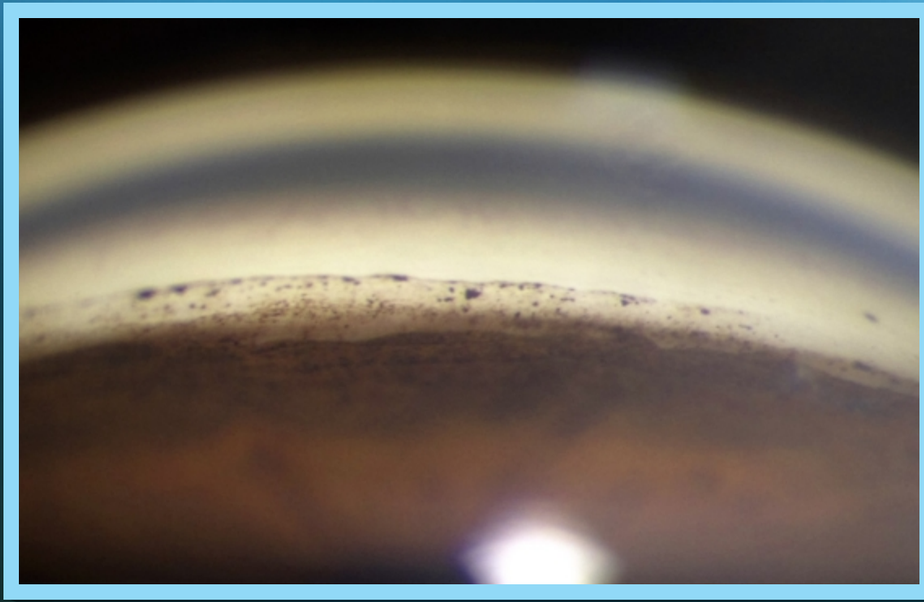
Diagnosis and Management of

PRIMARY ANGLE CLOSURE DISEASE



All India Ophthalmological Society

Diagnosis and Management of PRIMARY ANGLE CLOSURE DISEASE

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PACG Expert Group Meeting

Editorial

Primary angle closure disease (PACD) is a group of disorders characterised by a shallow anterior chamber with predisposition to appositional or synechial closure of the anterior chamber angle leading to raised intraocular pressure (IOP) and subsequent optic neuropathy. PACD is a preventable cause of irreversible blindness which currently impacts nearly 27 million people in India, in its entire spectrum [Primary angle closure suspect (PACS), primary angle closure (PAC), primary angle closure glaucoma (PACG)]. It is estimated that nearly 3 million people suffer from PACG and 10% are blind due to it. Population based studies in India have revealed that that nearly 90% of glaucoma remains undiagnosed and out of the diagnosed cases, majority of patients labelled as open angle are actually having angle closure disease. There is an unmet need to improve our detection rates for PACD and initiate correct laser/medical/surgical therapy at an early stage to prevent visual loss. The management of eyes with PACD is a complex issue and there may not be one well-defined algorithm which "best fits" to manage all the cases. Surgical decisions have to be taken on a case to case basis and are dependant on multiple factors including stage of disease, patient income (affordability to long-term medical therapy) and feasibility of regular follow-up, surgeon training and availability of tertiary care ophthalmology set-up to manage associated vitreo-retinal and corneal complications. This expert consensus publication provides a comprehensive outlay of the disease epidemiology, presentation and management strategies along with case studies, to impart practical knowledge to the reader regarding the diagnosis and treatment of PACD. A video assisted skill transfer has been included in the manual, incorporating diagnostic tests like gonioscopy, laser procedures (peripheral iridotomy) and surgical techniques (trabeculectomy and its complications). We hope that this module will aid in training ophthalmologists in diagnosing and treating PACD and ultimately improve the standard of care for patients with PACD in India leading to reduction in the disease burden and blindness caused by primary angle closure glaucoma.

Editorial Committee

Video Assisted Skill Transfer

Video	Topic
1	How to Perform Applanation Tonometry
2	Gonioscopy - Wide Open Angle with Ciliary Body Band Visible
3	Gonioscopy - Occludable Angle Opening on Manipulation In PACS
4	Gonioscopy - Occludable Angle with Blotchy Pigments on TM in PAC
5	Gonioscopy - Occludable Angle with Goniosynechiae in PAC
6	Gonioscopy - Completely Closed Angle in PACG
7	Optic Disc Evaluation Through Gonioscope - Inferior NRR Thinning
8	Optic Disc Evaluation With 90D - Baring of Circumlinear Vessel
9	How to Do Nd-YAG Laser Iridotomy
10	How Can You Make Trabeculectomy Safe and Effective
11	Trabeculectomy In Primary Angle Closure Glaucoma
12	How to Put Releasable Sutures – Technique I
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14	How to Perform Phaco-Trabeculectomy in PACG
15	Manual Small Incision Cataract Surgery with Trabeculectomy I
16	Manual Small Incision Cataract Surgery with Trabeculectomy II
17	Phacoemulsification in Post-Trabeculectomy Eye
18	Malignant Glaucoma - Can It Be Benign?
19	Malignant Glaucoma – Pars Plana Vitrectomy + Zonulo Hyloidectomy – In A Phakic Eye
20	Malignant Glaucoma – Pars Plana Vitrectomy + Zonulo Hyloidectomy – In A Pseudophakic Eye
21	Technique for Sclerectomy In Nanophthalmos
22	How to Perform Safe Phacoemulsification in Nanophthalmos
23	How to Prevent Uveal Effusion in Nanophthalmos
24	How to Perform Diode Laser Cyclo-Photocoagulation

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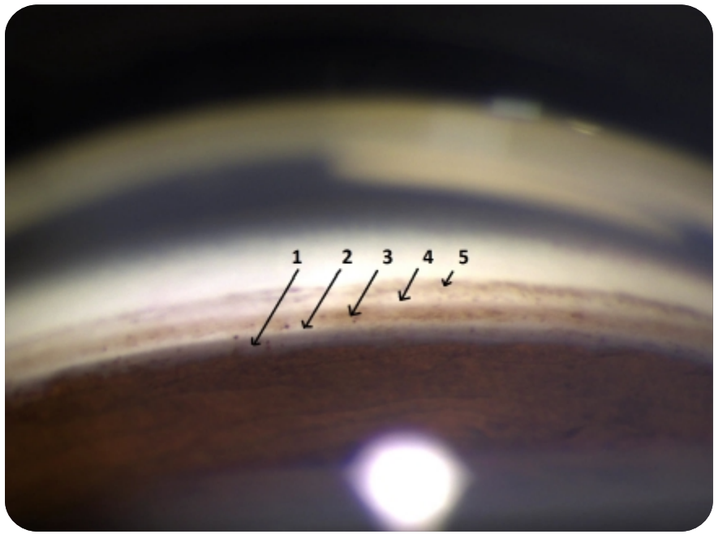
AIOS Expert Guidelines on Primary Angle Closure Disease

CONTENT

Normal angle structures as seen on gonioscopy in an open angle



Wide open angle showing angle structures;

1. Ciliary body band
2. Scleral spur
3. Posterior pigmented trabecular meshwork
4. Anterior non-pigmented trabecular meshwork
5. Schwalbe's line

1 What is Primary Angle Closure Disease?

In a normal eye with an open angle (Figure 1.1a-c), the aqueous humour drains via the trabecular meshwork. Primary Angle Closure Disease (PACD) is characterised by contact between the iris and the trabecular meshwork (Figure 1.2a,b and 1.3) leading to mechanical blockage of aqueous outflow with consequent rise in intraocular pressure (IOP) and damage to the optic nerve. Eyes with smaller anterior segment dimensions, especially shallow limbal and axial anterior chamber depth are predisposed to develop PACD.[1]

The contact between the iris and trabecular meshwork may be appositional (transient) or synechial (permanent). The diagnosis of PACD and the distinction between appositional versus synechial closure is established by viewing the anterior chamber angle anatomy through a gonioscope. Non-visibility of the posterior (pigmented) trabecular meshwork in 180 degrees or more of the angle circumference in the primary position on gonioscopy (performed in a dark room with a narrow 1 mm slit beam) is taken as the definition of an "occludable" angle and the patient is labelled as having PACD.[2]

The closure of the angle occurs due to various mechanisms, most common being a relative pupillary block. In predisposed eyes, especially in a mid-dilated pupil state, there occurs an apposition/contact of the lens and posterior iris surface, which leads to blockage of circulation of aqueous humour from the posterior chamber to the anterior chamber. This causes an increase in pressure in the posterior chamber and, thereby, causes the peripheral iris to bow forward and come in contact with the trabecular meshwork, thereby causing angle closure. Initially the irido-trabecular contact is appositional and can reverse on its own; however, prolonged contact leads to formation of peripheral anterior synechiae (PAS) with permanent closure of the trabecular meshwork. Other non-pupillary block mechanisms that can contribute to PACD include:

- A thick and anteriorly placed crystalline lens
- Plateau iris configuration
- High iris insertion or a thick peripheral iris
- Suprachoroidal effusion/ ciliary block

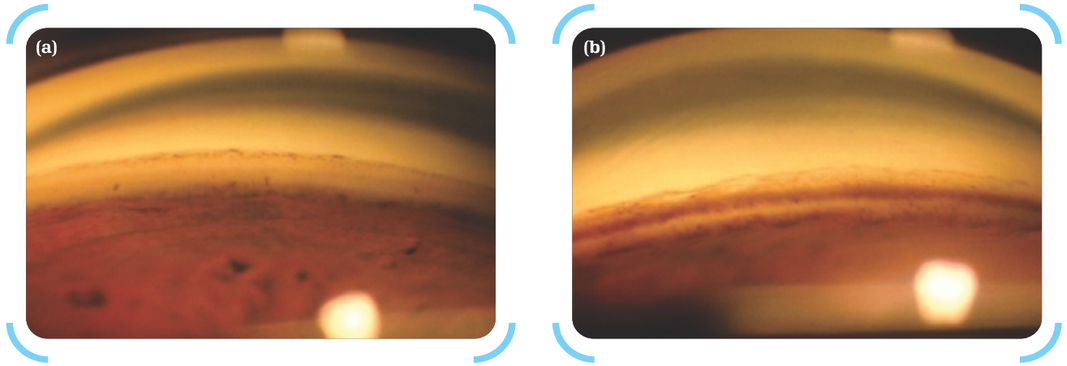


Figure 1.1 a,b: Wide open angle on gonioscopy and anterior segment optical coherence tomography (OCT) showing wide open angle.

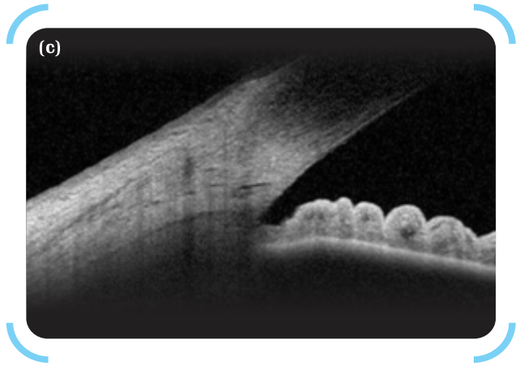


Figure 1.1 c: Wide open angle on gonioscopy and anterior segment optical coherence tomography (OCT) showing wide open angle.

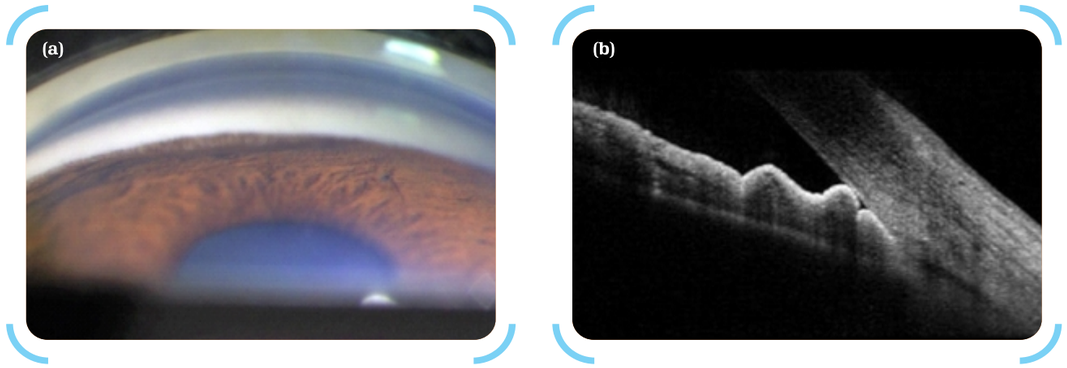


Figure 1.2 a and b: Convex iris configuration with a closed angle on gonioscopy and anterior segment OCT (ASOCT) showing apposition of the iris to the angle structures.

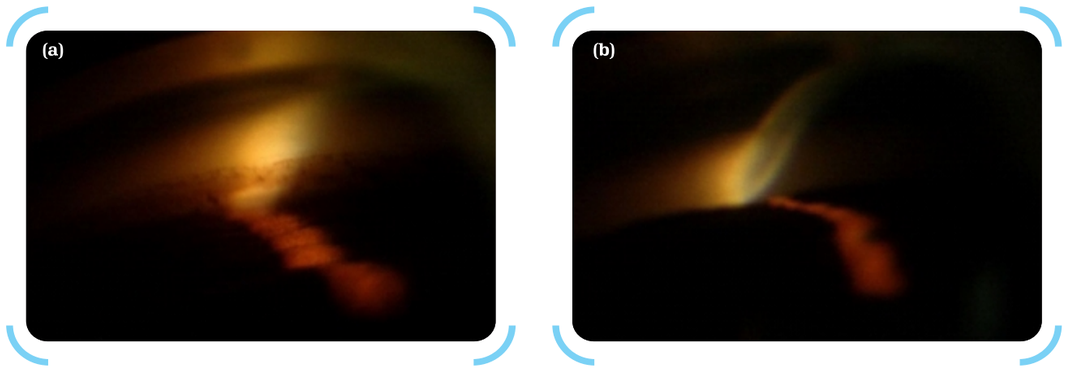


Figure 1.3: Open versus closed angle on primary gaze in gonioscopy. (a) Wide open angle with flat iris configuration. (b)- Angle structures not visible beyond corneal wedge, convex iris configuration.

2 What is the magnitude of PACD & blindness caused by it in India?

Glaucoma is the leading cause of irreversible vision loss both globally and in India. It is estimated that more than 15 million people suffer from PACD worldwide and this will increase to 21 million by 2020.

Information regarding the prevalence of glaucoma in India comes from seven population based studies conducted in India over the past 25 years. These include the Vellore Eye Survey (VES), Andhra Pradesh Eye Diseases Survey (APEDS), Aravind Comprehensive Eye Survey (ACES), Chennai Glaucoma Study (CGS), West Bengal Glaucoma Study (WBGS), Central India Eye and Medical Study (CIEMS), and the Hoogly River Glaucoma Study (HRGS).[3–6]

These studies have reported varying prevalence of PACG, ranging from 0.5% in the ACES to 4.32% for the VES in those aged 40 years and older. Part of the reason for these wide variations in disease prevalence are the different definitions used by these studies. However, PACG accounts for a significant proportion of all primary glaucoma. All forms of PACD including primary angle closure suspect (PACS), primary angle closure (PAC) and PACG are seen in upto 11% of the population aged 40 years or older.

In all population-based studies, the majority of those diagnosed with glaucoma were previously undetected, with previous diagnosis rates ranging from 6% to 17%. These rates are far below the 40% to 60% of previously diagnosed glaucoma from Western studies.[3]

The estimated number of persons with PACG was calculated based on published age and gender-specific data.[6] It was estimated that 2.54 (95% CI: 1.88 to 4.28) million persons have PACG in India and 0.25 million are blind due to it.[6] Those with some pathology secondary to angle closure (PAC and PACG: raised IOP with narrow angles or peripheral anterior synechiae (PAS) or glaucomatous optic disc or visual field changes) comprise 6.62 (95% CI: 4.78 to 9.41) million persons. The estimated total number of persons with PACD (including PACS) is 27.66 (95% CI: 24.00 to 30.92) million.[3,6]

Glaucoma is the second leading cause of blindness in the adult population in India. While there are more persons with primary open angle glaucoma (POAG) in the population, PACG causes blindness in a greater proportion of affected individuals than open angle glaucoma. From the Indian studies that reported blindness rates for both POAG and PACG, the rates of bilaterally blind because of POAG in the APEDS, ACES, CGS (rural), CGS (Urban) and WBGS were 11.1%, 1.6%, 3.2%, 1.5% and 5.2%, respectively. For angle closure glaucoma, the blindness rates for APEDS, CGS (rural) and CGS (Urban) were 16.6%, 2.9% and 5.9%, respectively.[3] PACG caused two-times the proportion of bilateral blindness than POAG (Table 2.1)

Challenges in the detection of angle closure disease

The main challenge in detecting angle closure disease is probably the limited gonioscopy training that an average ophthalmology resident receives. This is apparent from both the Chennai Glaucoma Study and the APEDS, both of which reported that among those persons in the population diagnosed with PACG and were already on glaucoma treatment, two-thirds were being treated as POAG because gonioscopy had not been performed or inappropriately done.[3] While there is a learning curve associated with gonioscopy, as with any other procedure, it is easily learnt. By performing gonioscopy on all patients (unless contraindicated), one develops familiarity with the technique and also with the normal variants seen in clinical practice. Only by actively looking for angle closure will we detect it in the clinic and prevent blindness caused by it.

Study	Year of study	Urban/ Rural	Age group	PACS (Prevalence, 95% CI)	PAC (Prevalence, 95% CI)	PACG (Prevalence, 95% CI)
VES	1994	Urban	30-60 years	4.3%		0.5%
APEDS	1996-2000	Both	>40 years	Urban-3.5% Rural-1.5%	Urban-0.8% Rural-0.2%	Urban-1.8% Rural -0.7%
WBGS	1998-1999	Rural	>50 years			0.81%
CGS	2001-2004	Both	>40 years			0.23%
Glaucoma survey-Chhattisgarh	2001	Both	>35 years			0.24%
CIEMS	2006-2008	Rural	>30 years	Urban-7.24% Rural-6.3%	Urban-2.75% Rural-0.71 %	Urban-0.88% Rural-0.88%

Table 2.1: Prevalence of PACD in various population based studies (VES- Vellore Eye Study, APEDS- Andhra Pradesh Eye disease Study, CGS- Chennai Glaucoma Study; CIEMS- Central India Eye and Medical Study; WBGS- West Bengal Glaucoma Study)

3 How to classify patients with PACD?

Patients with PACD were earlier classified into 3 clinical stages, based primarily on the symptoms – acute, intermittent/sub-acute and chronic stages of angle closure glaucoma regardless of the presence or absence of optic neuropathy. PACD can also be classified based on the anatomic level primarily contributing to trabecular obstruction into 4 levels: iris (pupillary block), ciliary body (plateau iris), lens-induced mechanism and forces posterior to the lens (malignant glaucoma).

The current classification of PACD adopted in most clinical and epidemiological studies is based on an International Society for Geographical and Epidemiological Ophthalmology (ISGEO) definition[7] and is based on the natural history of the disease. The new classification includes a description of PACS, PAC and PACG as follows:

1. **PACS -** An eye with occludable angle in which the peripheral iris is in appositional contact with posterior trabecular meshwork in $\geq$180 degree of the angle with a normal IOP, optic disc and visual field (Figure 3.1 a-b).

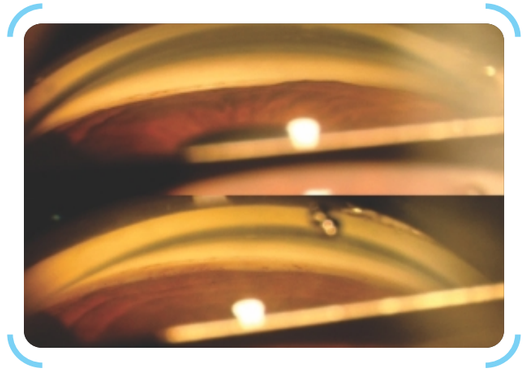


Figure 3.1a: Angle opening on manipulation in PACS.

Figure 3.1b: Trabecular meshwork not visible in primary gaze, opening completely on manipulation in PACS.

2. **PAC** – This is PACS with additional component of peripheral anterior synechiae (PAS) and/or elevated IOP, iris whorling or sectoral atrophy, and blotchy pigment deposition on the trabecular meshwork. PAC with elevated IOP (ocular hypertension) indicates trabecular outflow obstruction and has the highest risk for progression to PACG, and must be identified and treated as a separate entity (Figure 3.2a-d).

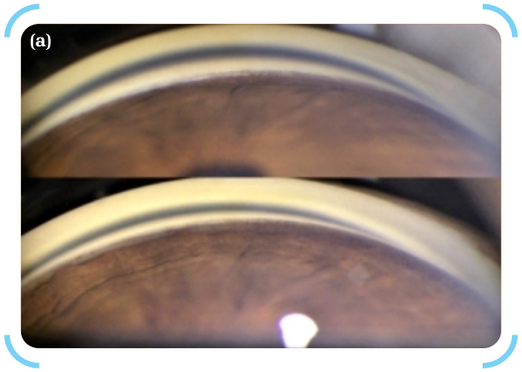


Figure 3.2a- Gonioscopy showing partial angle closure on primary gaze (above) and goniosynechiae seen on manipulation gonioscopy (below) in PAC

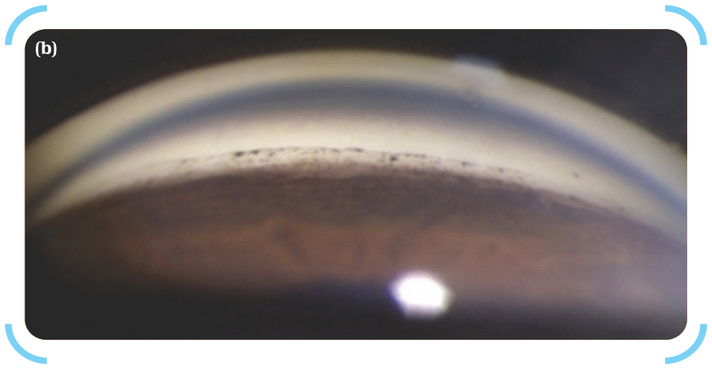


Figure 3.2b- Convex iris configuration with blotchy pigments anterior to trabecular meshwork and irido-trabecular contact with multiple goniosynechiae in PAC

Figure 3.2c – Goniosynechiae (hill-valley appearance) on manipulation gonioscopy in PAC

Figure 3.2d: Goniosynechiae in PAC (low and high magnification).

3. **PACG** - This is PAC with evidence of glaucomatous optic neuropathy - optic disc and field changes. The severity of glaucoma at this stage can be sub-classified into early/ moderate/ advanced based on the visual field damage using the Hodapp - Parrish - Anderson criteria or other classifications systems similar to POAG (Figure 3.3).

Figure 3.3- Completely closed angle not opening on manipulation in PACG

There may exist a subset of patients with glaucomatous optic neuropathy, raised IOP and narrow angles without PAS. These patients have components of both open- and closed-angle glaucoma and are classified as having a combined mechanism glaucoma.

Gonioscopy is mandatory for the diagnosis and categorisation of PACD as it ascertains if the trabecular meshwork is open or obstructed by the iris.

Risk of progression from PACS to PAC to PACG

Nearly 1 in 4 patients with PACS (22%) are likely to progress to PAC over 5 years, and a similar percentage are likely to progress from PAC to PACG (28.6%).[8] The risk of progression is higher in eyes with bilateral PAC, PAC with ocular hypertension, and if laser peripheral iridotomy (LPI) has not been performed.[9]

4 What are the important points in the history and examination of a patient?

Clinical evaluation should start with a history of the present illness, including symptoms, onset, duration and severity. The family history, medical history along with ongoing medical therapy for other systemic illness and evaluation of family income to establish affordability of long-term ocular hypotensive therapy are the important points to be noted.

History-taking in a patient with PACD

- Occurrence of unilateral headaches. Precipitating factors include dim illumination such as watching television in darkened room, adoption of a semi-prone position (reading) or acute emotional stress[10]
- Past history of similar attacks
- Coloured haloes around bright lights
- Blurred vision
- Severe eye pain with redness and associated nausea/vomiting (acute attack)
- Use of presbyopic glasses before 40 years of age
- Difficulty in climbing steps or bumping into furniture or people, suggestive of restriction of visual field
- Family history of glaucoma[11]
- Use of steroids in any form, including inhalers, dermatologic creams containing steroids or oral/injectable steroids
- Use of medications capable of precipitating angle closure in susceptible eyes.[12] The mechanism may be due to pupillary dilatation or swelling of the ciliary body, leading to irido-trabecular apposition and rise in IOP (Table 4.1 and 4.2)
- Associated systemic diseases – diabetes, hypertension, cardiovascular abnormalities, cerebrovascular accident, arthritis, chronic obstructive airway disease, constipation (Valsalva manoeuvre)
- Socioeconomic situation and geographic location to assess affordability with respect to medical therapy, compliance and ability to come for regular follow-ups.

Table 4.1: Common Drugs Predisposing Angle Closure (via Pupillary Dilatation)

Anticholinergics	Muscarinic Antagonists	Atropine, Botulinum Toxin A, Oxybutynin, Tropicamide, Ipratropium Bromide, Scopolamine, Hydroxizine, Pirenzepine, Diphenhydramine, Dicyclomine, Cyclopentolate
	Selective Serotonin Reuptake Inhibitors (SSRI)	Venlafaxine, Citalopram, Escitalopram, Fluoxetine, Paroxetine
	Tricyclic anti-depressants	Clopiramine, Imipramine, Trazadone, Amitryptyline
Sympatho-mimetics	beta2-agonists	Salbutamol, Ritodrine
	alpha adrenergic agonists	Phenylephrine
	Nasal Decongestants	Phenylpropanolamine
	Recreational Drugs	Cocaine

Table 4.2: Common Drugs Causing Idiosyncratic Reactions – Angle closure via forward movement of iris-lens diaphragm

Antihistamine	Promethazine
Amphetamine	Ecstasy
Anti-epileptic	Topiramate
Anti-depressant	Escitalopram
Anti-coagulants	Heparin
Sulpha-containing drugs	Sulphamethoxazole, Trimethoprim, Acetazolamide
Thiazide diuretic	Hydrochlorthiazide

Important points to be noted during examination

Anterior segment examination

Standard oblique flashlight test

A focused torch-light can be used parallel to the temporal limbus as a crude indicator of the anterior chamber depth, especially in the absence of slit lamp.[13,14] An anterior chamber of normal depth will have a flat iris that permits light to cross and illuminate the nasal limbus (Figure 4.1a). In a shallow anterior chamber, the anterior bowing or convexity of the iris will not allow the light across, and the nasal limbus will be shadowed (Figure 4.1b).

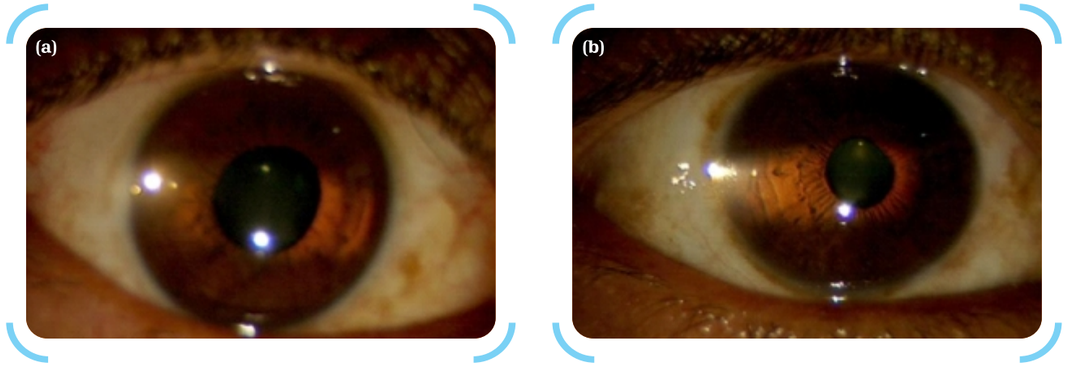


Figure 4.1a,b: Torch-light from the temporal side. (a) Normal eye; (b) Shallow anterior chamber (AC) showing nasal shadowing due to iris bombe.

Van Herick test

The Van Herick (VH) test is performed on a slit-lamp using a thin bright beam of light placed at the extreme periphery of the temporal AC, keeping the observation and illumination arms at 60 degrees (Figure 4.2, 4.3a-d). The peripheral anterior chamber depth is graded according to the ratio of the peripheral AC depth to the thickness of the peripheral cornea.

Grade 4: Peripheral anterior chamber depth > Corneal thickness (CT)

Grade 3: Peripheral anterior chamber depth 1/4 to 1/2 of CT

Grade 2: Peripheral anterior chamber depth 1/4 of CT

Grade 1: Peripheral anterior chamber depth <1/4 of CT

It is mandatory to perform gonioscopy in grades 1 to 3, as these can develop angle closure. Only VH grade-4 (peripheral AC depth $\geq$ corneal thickness) effectively rules out angle closure. Gonioscopy may not be performed in persons having VH grade 4 or in PACD eyes that are now pseudophakic. VH grading does not replace gonioscopy in persons having manifest or suspected glaucoma.

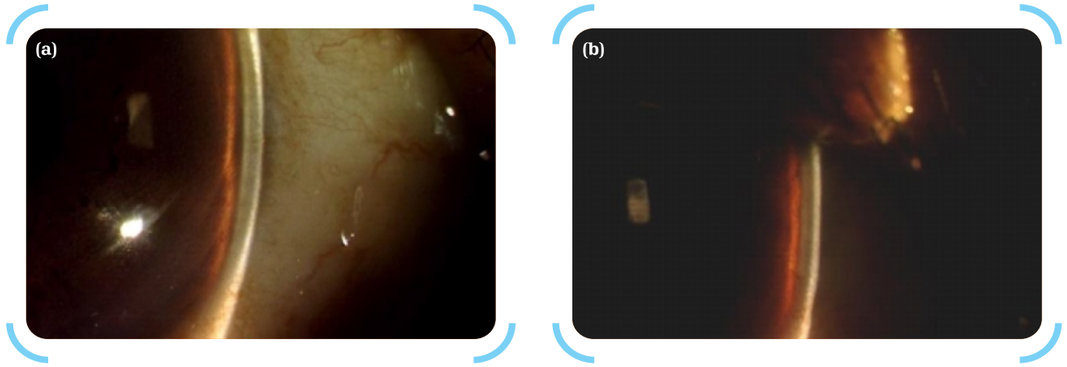


Figure 4.2a,b: Van Herick examination. (a) normal/shallow AC; (b) shallow AC. Shallow peripheral AC depth

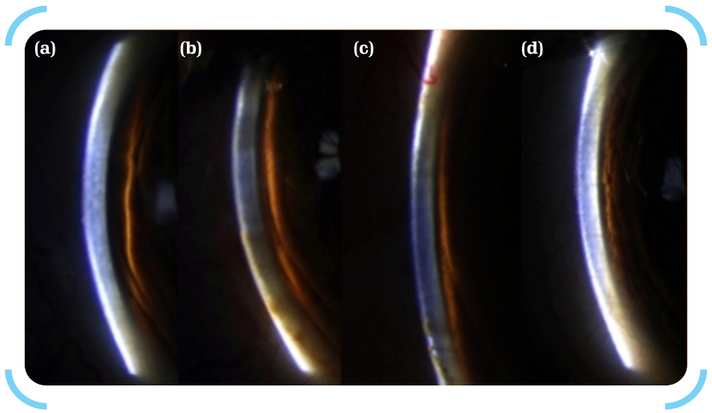


Figure 4.3 a,b,c,d: VH grade 4 (a), grade 3 (b), grade 2 (c) grade 1 (d)

Pupillary Reaction

A brisk direct and consensual reaction is seen if the optic nerve is functioning normally. In the presence of glaucomatous optic neuropathy, a sluggish reaction can be seen in the affected eye. One must document if Relative Afferent Pupillary Defect (RAPD) is present in a patient with glaucoma as it implies significant damage to the optic nerve head.

Iris Pattern

Iris undergoes ischemic changes due to recurrent angle-closure attacks. Early sphincter atrophy in PAC can be seen as a loss of the pupillary ruff (Figure 4.4a). The sphincter muscle atrophy may lead to a vertically oval pupil. After an acute angle-closure attack, sectoral atrophy with absence of radial folds on the iris in that sector may be seen (Figure 4.4b).

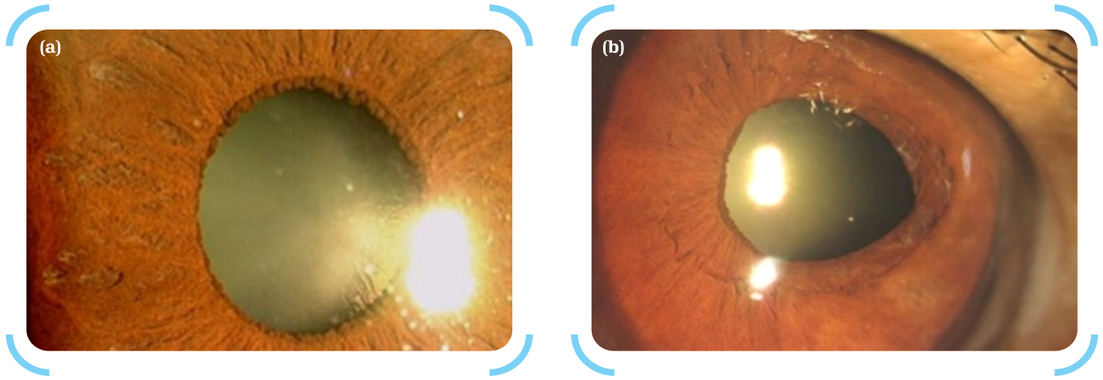


Figure 4.4 a,b: Pupillary ruff atrophy and sphincter atrophy with sectoral iris atrophy.

Lens status

The lens is usually thick or anteriorly placed in angle closure disease. Lens thickness should be documented in all the cases. Lens thickness may be considered excessive if it is >5 mm. Acute attacks may induce ischemic changes in the lens capsule denoted by the presence of Glaucomflecken (Spilt milk appearance) (Figure 4.5)

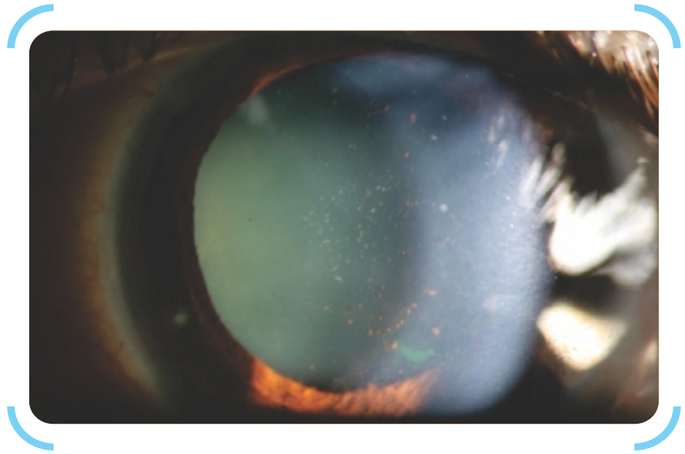


Figure 4.5: Glaucomflecken (Spilt milk appearance) - ischemic changes in the lens capsule seen as white flecks over the centre of the anterior capsule.

Optic Nerve Head Examination

Examination of the optic nerve head and retinal nerve fiber layer may allow diagnosis of glaucomatous damage before visual field loss. In PACD, there is generally a small optic nerve head, so that a cupping of even 0.4:1 becomes significant, and a careful examination of the neuro-retinal rim and retinal nerve fiber layer is mandatory. The degree of optic nerve damage helps to guide treatment goals, and this should be drawn/photographed at the time of glaucoma diagnosis (Figure 4.6).

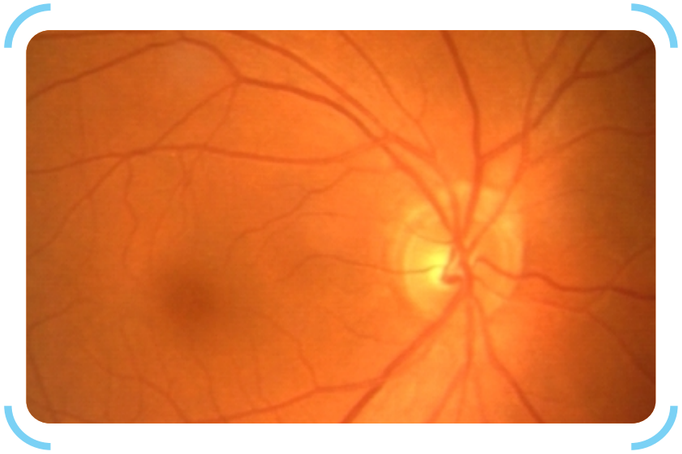


Figure 4.6: Normal Optic Nerve Head

Definite signs of glaucomatous optic neuropathy are:

1) Notching - a localised loss of the neuro-retinal rim (NRR) (Figure 4.7)
2) Retinal nerve fiber layer (RNFL) defect, as shown by the white arrow (Figure 4.7)
3) Asymmetry in the cup: disc ratio between the two eyes > 0.2

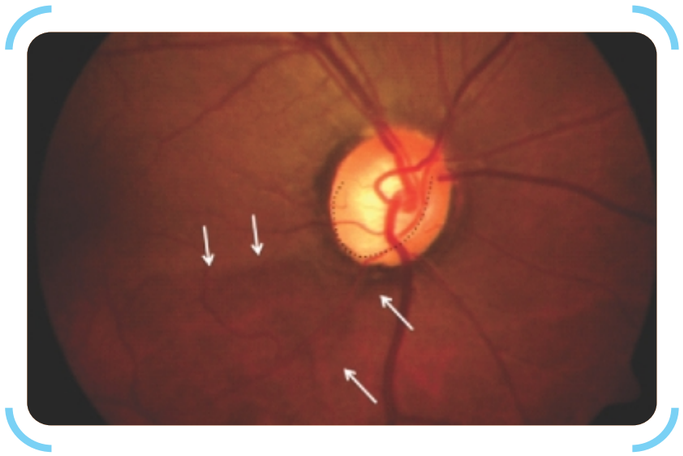


Figure 4.7: Notching and RNFL defect in early-to-moderate glaucoma. Cup boundary is marked by the black dotted line, showing inferior notch.

IOP measurements

Goldmann applanation tonometry is the current gold standard for measuring IOP. Applanation tonometry should be done at least 3 to 4 times, at different times of the day to diagnose a chronically raised IOP and to document diurnal pressure. Diurnal phasing should be done after 6 weeks of an iridotomy. If diurnal phasing is not possible, IOP may be taken at different times during multiple visits to determine the IOP fluctuation and the peak IOP. Early morning, afternoon and evening IOP readings should be noted to establish the baseline IOP.

Gonioscopy

Gonioscopy is the gold standard for examination of the anterior chamber angle. The most important sign to be picked up on gonioscopy is the presence of irido-trabecular contact and its reversibility (appositional versus synechial). The examination must be done in a dark room,

using a 1 mm narrow slit which should not cross the pupil, with the patient in primary gaze position. Indirect method of gonioscopy using Goldmann double mirror is recommended for most cases with manipulation to look over a steep iris configuration. If the trabecular meshwork is not visible after manipulation, indentation gonioscopy using a 4 mirror Posner/Sussman lens is required.

The main features of angle anatomy that need to be assessed and documented include the following:

- Structures seen in the primary gaze and on indentation/ manipulation
- Presence and extent of irido-trabecular contact/adhesion
- Width of the angle recess
- Configuration of the iris
- Level of insertion of the iris

Angle imaging using ASOCT cannot replace gonioscopic angle evaluation. Gonioscopic images of open-angle, occludable angle and peripheral anterior synechiae are shown in Figure 4.8a,b and c, respectively.

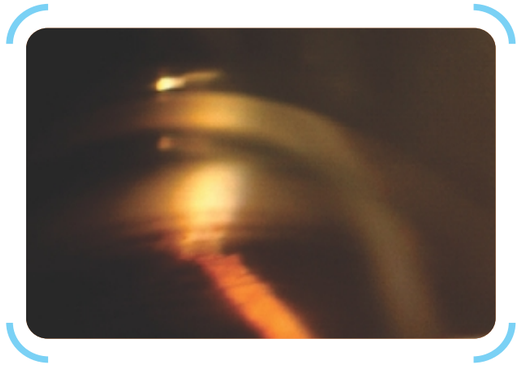


Figure 4.8a: Normal open angle, 35 degrees angle recess, with all structures – Schwalbe's line, trabecular meshwork, scleral spur & ciliary body band seen

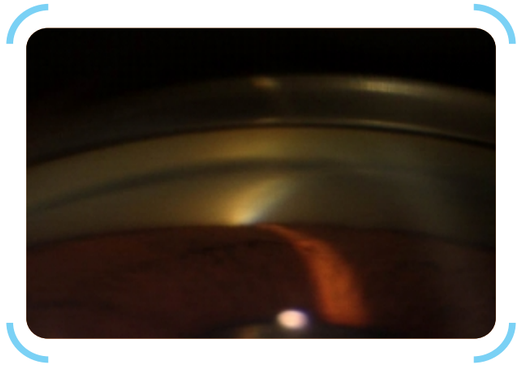


Figure 4.8b: Occludable angle, <10 degrees angle recess, with only corneal wedge seen. No angle structures visible

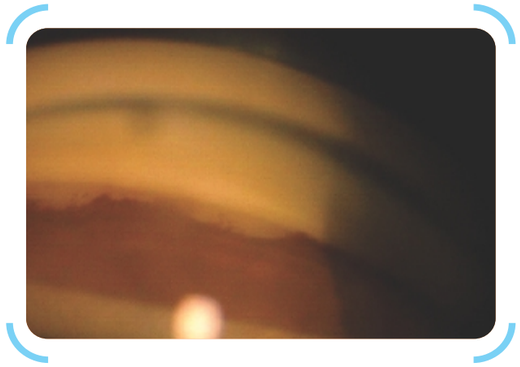


Figure 4.8c: 'PAS, iris seen attaching to trabecular meshwork and Schwalbe's line after manipulation

5 What are the risk factors?

The risk factors for PACD include anatomical and genetic predispositions. Eyes with PACD have one or more of the following biometric features:

- Smaller corneal diameter[15,16]
- Smaller radius of curvature
- Shallow central AC depth
- Smaller anterior chamber volume[17]
- Thicker peripheral iris
- Anterior insertion of peripheral iris[18]
- Thick lens
- Increased lens vault[19]
- Steeper curvature of anterior lens surface
- A more anterior lens position
- Prominent anteriorly displaced ciliary processes
- Increased lens thickness/axial length ratio
- Shorter axial length

Hyperopic eyes with a shallow anterior, chamber, increased lens thickness/vault and short axial length are the major risk factors for the development of PACD.

Other risk factors include

- Old age
- Female gender
- East Asian/ Inuit ethnicity
- Family history of glaucoma

6 What additional investigations may be required?

Although clinical examination including IOP evaluation, gonioscopy and stereoscopic optic nerve head evaluation is sufficient to diagnose a case of PACD, additional investigations such as anterior segment imaging may be useful in specific situations. Standard automated perimetry is mandatory in eyes with PACG to stage the disease severity. Ocular biometry with evaluation of the anterior chamber depth, lens thickness and axial length can additionally be performed in eyes with PACD. We do not advocate any provocative tests such as the dark room prone provocative test or phenylephrine test for the diagnosis of PACD.

Role of Anterior Segment Imaging: ASOCT and Ultrasound Biomicroscopy (UBM)

Both, ASOCT and UBM, can be used to image the anterior segment. ASOCT has the advantage of being a non-contact procedure and has a higher resolution; however, imaging posterior to the iris is not possible with ASOCT. UBM is required for confirming the diagnosis of posterior chamber/ retro-lenticular pathologies such as plateau iris or malignant glaucoma.

Indications

Plateau Iris configuration

A clinical suspicion of plateau iris configuration, with a flat iris configuration, a normal-looking central AC, and a sine-wave sign on gonioscopy (Figure 6.1a and b) should prompt one to confirm the clinical impression by doing a UBM. It should always be looked for, in case the IOP remains high after a patent iridotomy and no appreciable PAS is visible. A prominent anteriorly displaced ciliary body supporting the iris with obliteration of the irido-ciliary sulcus and irido-trabecular contact confirms the diagnosis.

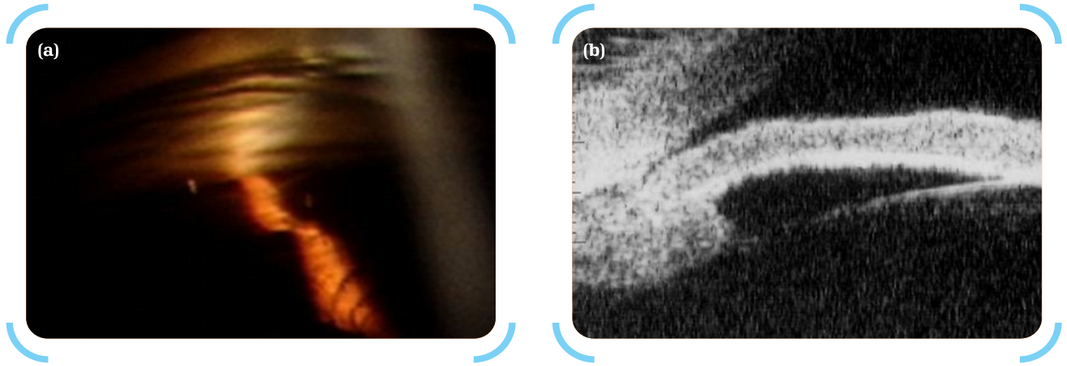


Figure 6.1 (a) Double hump sign seen on indentation gonioscopy in plateau iris; (b) obliterated irido-ciliary sulcus in plateau iris on UBM.

Irregular iris configuration suggestive of iris/ ciliary body cysts causing secondary angle-closure

In eyes with segmental angle-closure restricted to one/two quadrants, a space occupying lesion posterior to the iris should be suspected. These conditions can be easily diagnosed by performing a UBM. The main advantage of the UBM is its ability to view structures behind the iris, such as the ciliary body, the lens zonules and the anterior choroid. It is useful to investigate the pathogenesis of angle closure and the role of anterior rotation of the ciliary body in plateau iris, malignant glaucoma, the presence of iridociliary cysts (Figure 6.2), ciliary body tumours, choroidal effusions, or the presence of a tilted or subluxated lens (especially in pseudoexfoliation).

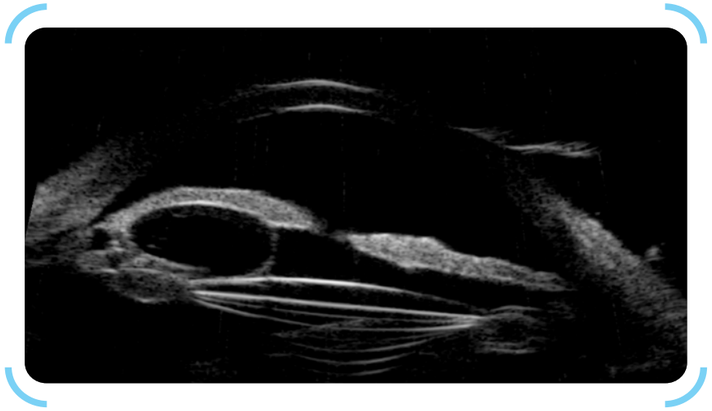


Figure 6.2- UBM showing fluid-filled cyst causing localized anterior chamber shallowing in a pseudophakic patient

Drug-induced glaucoma

Ciliary body swelling or effusion may be induced as a reaction to systemic drugs (such as topiramate) being taken by the patient. The patients present with myopia and an acute attack of angle-closure due to cilio-choroidal effusion and forward shift of the iris lens diaphragm. UBM is useful for detecting this condition.

Post Trabeculectomy Complications

Early complications of trabeculectomy such as blockage of the ostium by the iris can be imaged by ASOCT in the early post-operative period when contact investigations are not possible. UBM is useful for diagnosing malignant glaucoma to document the forward shift of ciliary processes. Bleb imaging with ASOCT can also performed for diagnostic and therapeutic purposes to manage late complications of trabeculectomy and bleb failure.

Thick/anteriorly displaced lens in a small eye

Clinic based studies have suggested that eyes with occludable angles and angle closure glaucoma have a shorter axial length, shallower anterior chamber, and a thicker lens.[20] (Figure 6.3)

The following parameters have been found to be useful for the evaluation of angle closure:

- Lens thickness/ lens vault measurement: Progressive increase in lens thickness with age results in greater shallowing of the anterior chamber.[21,22]

- Lens vault (LV) is a novel parameter independently associated with angle closure after adjusting for age, gender, anterior chamber depth (ACD), and LT. It is measured as the perpendicular distance between the horizontal line joining the two scleral spurs and the anterior pole of the crystalline lens, on horizontal AS-OCT scans. (Figures 6.4, 6.5).[19,23]
- Lens thickness/ axial length ratio: The LT/AL ratio has been found to be greater than normal for most age groups with angle-closure glaucoma.[24]

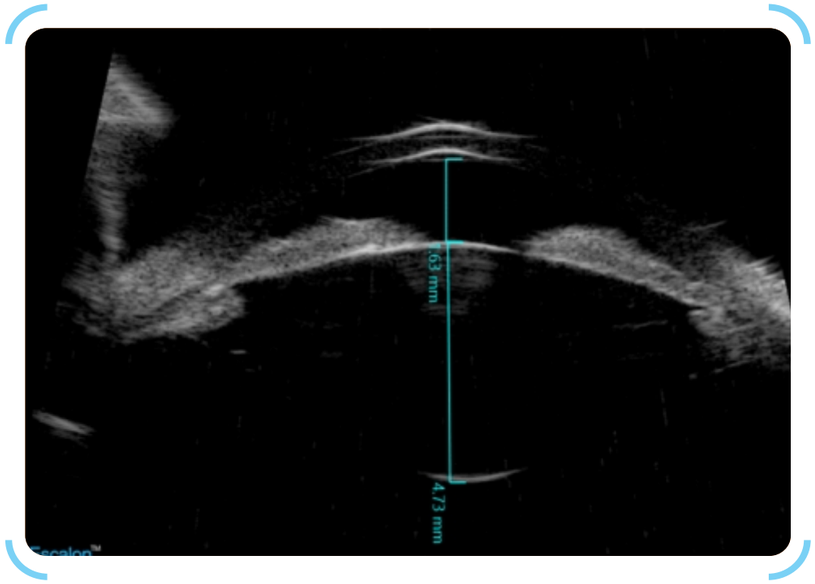


Figure 6.3- Shallow anterior chamber with very thick lens with iris draping the anterior lens surface in a young PACG (UBM)

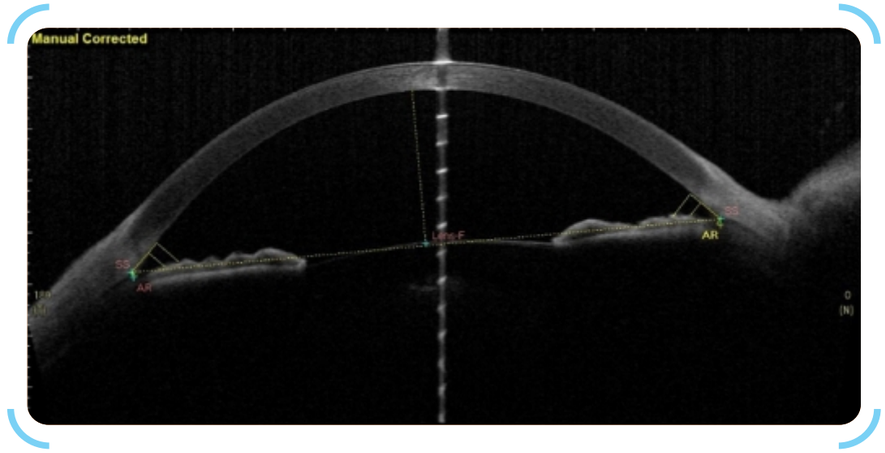


Figure 6.4: Normal Lens vault with an open angle. ACD=2.9mm; Lens Vault=0.04mm

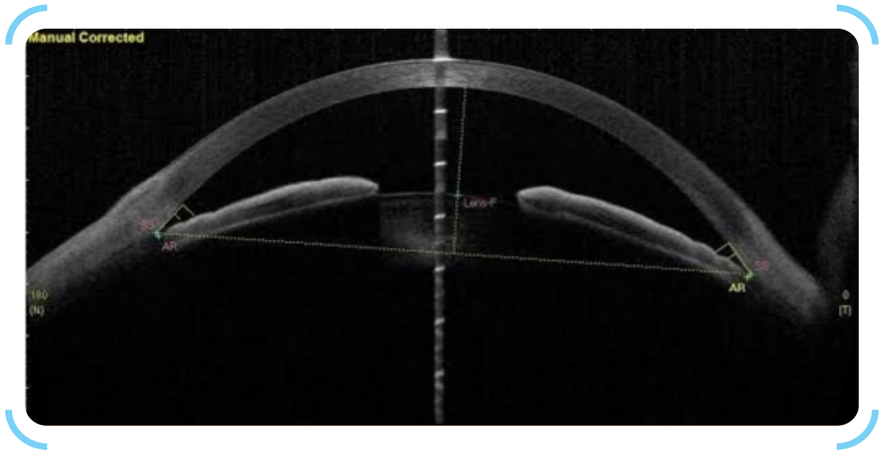


Figure 6.5: High lens vault (1.131mm) in a patient with narrow angles. ACD=2.07mm

Lens induced glaucoma with secondary angle closure

Eyes with spherophakia, post-traumatic lens subluxation/ dislocation, etc., may require imaging to confirm the diagnosis.

7 Management of Primary Angle Closure Suspects (PACS)

In eyes with an occludable-angle labelled as PACS, the patient should be counselled regarding the risk of developing an acute attack, the symptoms associated with increased IOP, risk of progression to PACG, the risk factors that can precipitate acute angle-closure, advantages/ side-effects of laser iridotomy, role of cataract surgery (if patient presents with a visually significant cataract), and the role of pilocarpine eye drops in case of an emergency where immediate medical care is not accessible.

In general, eyes with PACS do not require any medical/laser therapy and may be asked to come for follow-up at least once in two years to look for disease progression. A laser iridotomy may be required in certain situations that puts the patient in a higher risk category for developing an acute attack of angle closure and these patients require an annual review.

Which PACS patients should undergo a peripheral laser iridotomy?

- Fellow eyes of patients with either PAC or PACG[25] or one eyed patients
- Need for repeated dilatation due to posterior segment pathology (e.g. NIDDM)
- Use of systemic medications that may provoke pupillary block (Table 4.1 and 4.2)
- Patients having symptoms suggestive of prior acute or intermittent angle-closure[26]
- Family history of PACG
- Eyes with progressive narrowing of angles on follow-up
- Patients whose health/ occupation/ geographical location makes it difficult to access immediate ophthalmic care in the event of an acute attack of angle-closure
- Patients who understand the risk of progression and want a laser iridotomy
- Patients who cannot come for regular follow-up[27]

If the patient is at high risk and does not consent to a peripheral laser iridotomy or cannot undergo the procedure due to medical reasons, 2% pilocarpine eye drops may be prescribed to the patients. These drops are to be instilled only in case of symptoms suggestive of an acute attack of angle-closure, especially if there is a delay in contact with an eye care facility.

In case the patient has a visually significant cataract in one eye, cataract surgery may be advised in that eye, with laser iridotomy being performed in the other eye. Once pseudophakic, PACS eyes do not require any medical/laser therapy.

Follow-up protocol

PACS patients should be evaluated atleast once in every 2 years, if there are no associated risk factors, with a complete ophthalmic examination including gonioscopy and optic nerve evaluation with manual drawing or photographic documentation of both angle structures and optic nerve head (preferred).

PACS patients who undergo laser iridotomy can be evaluated after 24 hours, 6 weeks and then once a year.[28] It is important to counsel the patients about the risk of progression of disease, even after a laser iridotomy has been performed, and the need for regular follow-up. The risk of progression of cataract after LPI should also be explained.

Case study 1

Presentation

A 37-year-old female experienced acute-onset blurry vision and ocular pain in both eyes four days after beginning Tab Topiramate for suspected migraine.

Examination and investigations

On examination, her uncorrected visual acuity (UCVA) was 21 ETDRS letters (OD) and 19 ETDRS letters (OS). The manifest refraction was -3.0D/ -1.0D × 170° OD and −2.5D/-1.0D × 20° OS. IOP measured was 42 mmHg OD and 34 mmHg OS with corneal edema and Van Herick Grade I (OU).

The patient was treated with oral acetazolamide (1 tab 4 times/day), topical timolol/dorzolamide combination, along with discontinuation of Tab Topiramate. After 24 hours, her best-corrected visual acuity (BCVA) was 76 ETDRS letters (OD) and 72 ETDRS letters (OS). IOP was 18 34 mmHg OD and 16 34 mmHg in OS. Slit-lamp examination revealed disappearance of corneal oedema and markedly shallow anterior chambers without inflammation in both eyes. Gonioscopy revealed grade 0 angles on Shaffer classification OU. Undilated fundus examination revealed that maculae and optic discs were unremarkable. However, UBM confirmed a 360° ciliochoroidal effusion and anterior rotation of the ciliary processes with forward displacement of the lens-iris diaphragm in both eyes (Figure CS 1.1).

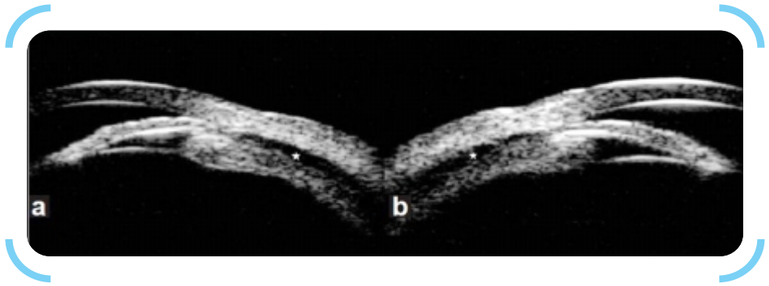


Figure CS 1.1: Ultrasound biomicroscopy on day 1 showed ciliochoroidal effusion (star) and an anterior rotation of the ciliary processes in the right (a) and left eyes (b)

Diagnosis

A diagnosis of topiramate-induced acute angle-closure was established. She was treated with topical prednisolone acetate and cycloplegic agents. Three days later, the UCVA was 88 ETDRS letters (OU) with deep anterior chambers and IOP was measured to be 16 mmHg OU. The spherical equivalent improved from −3.5 to −0.25D (OD) and from −3.0 to −0.5D (OS). Gonioscopy revealed grade 3 angles on Shaffer classification (OU). UBM revealed that the annular ciliochoroidal effusion had resolved in both eyes. Anti-glaucoma medications, prednisolone acetate, and cycloplegic eye drops were subsequently discontinued (Figure CS 1.2).

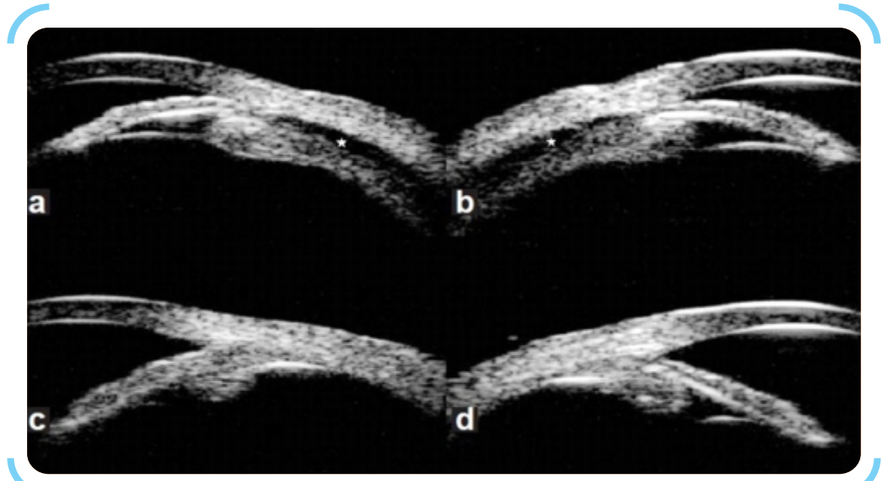


Figure CS 1.2: Ultrasound biomicroscopy on day 1 showed ciliochoroidal effusion (star) from the scleral spur, detachment of the ciliary body and an anterior rotation of the ciliary processes in the right (a) and left eyes (b), Ultrasound biomicroscopy on day 15 showed resolution of ciliochoroidal effusion and deepening of the anterior chamber in the right eye (c) and left eye (d)

Discussion:

Bilateral acute angle-closure glaucoma (AACG), ciliochoroidal effusion and transient myopia may be induced by drugs, such as oseltamivir,[29] topiramate,[30]methylenedioxymethamphetamine (ecstasy),[31] cabergoline,[32] acetazolamide,[33] "anorexiant mix," bupropion, cabergoline, escitalopram, flucloxacillin, hydrochlorothiazide, mefenamic acid, methazolamide.[34]

8 Management of Primary Angle Closure (PAC)

At this stage, there is both structural and functional damage to the trabecular meshwork manifesting as irido-trabecular adhesions (goniosynechiae, pigment clumps on trabecular meshwork) with /without an elevated IOP, iris whorling or sectoral atrophy, and blotchy pigment deposition on the trabecular meshwork. Eyes with PAC and ocular hypertension are at an increased risk to progress to PACG (optic neuropathy).[9] The main goal of treatment of these patients is to reverse the angle-closure process by doing a LPI and prevent damage to the optic nerve by lowering IOP using topical medications, if needed. Removal of a visually significant cataract in eyes with PAC is very useful.[35]

The comprehensive workup of a PAC should include a careful history about systemic medications and diseases, a baseline IOP, gonioscopy documentation and a baseline visual field examination. LPI should be performed in both eyes in all patients with PAC. If there is an anticipated delay in LPI, pilocarpine eye drops can be prescribed to prevent development of an acute attack, till the time LPI is performed. The IOP should ideally be checked at atleast 3 time points (morning, afternoon, evening); 6 weeks after LPI. The peak IOP and fluctuations in IOP should be noted in the records. The target IOP for PAC should be kept $\leq$ 21 mmHg. Visual field examination may be repeated after 1 year. Baseline biometry should be noted in patients as age related changes occur in lens thickness, leading to further shallowing of the anterior chamber and progression of angle closure despite a patent iridotomy.

Glaucoma medications should be started if the IOP is more than 21 mmHg after LPI. Prostaglandin analogues ([PGA]; latanoprost, travoprost, bimatoprst, tafluprost) may be given as the first choice in bilateral cases and aqueous suppressants may be given as the first choice in unilateral cases (eg. If the other eye is pseudophakic). Unilateral therapy with PGA is not desirable due to cosmetic side-effects of prostaglandins, if used only in one eye - (such as eyelash growth, periocular and iris pigmentation, etc.).[36] Some patients may not respond well to a particular prostaglandin, in which case an intra-class switch can be done such as substituting travoprost for latanoprost. If the IOP is not controlled on a single PGA, beta blockers/ alpha agonists/ carbonic anhydrase inhibitors can be added. If 3 drugs are required, combination therapy should be used to reduce frequency of eye drop instillation. If the IOP is not controlled medically, a short course of pilocarpine can be tried. Some elderly patients respond well to topical pilocarpine therapy and surgery can be deferred. A 24-hour drug holiday with pupillary dilatation and fundus evaluation should be done once in 3-6 months, if the patient is on long-term pilocarpine therapy. Eyes with PAC and ocular hypertension need frequent monitoring at least once in 6 months as they have a high risk for progression to PACG.

If the angle opens after LPI and the IOP is elevated, Selective Laser Trabeculoplasty (SLT) is an option (avoid areas of PAS formation) although IOP lowering capability is moderate, with reduction in effect over time.

Lens extraction

Temporal clear corneal phacoemulsification can be performed at an early stage in eyes with PAC with a visually significant cataract without performing a LPI. In eyes with no visually significant cataract and 20/20 vision, LPI should be performed as the first-stage procedure and topical medications added, if the IOP is not controlled. Performing a clear lens extraction in eyes with PAC is controversial and a high-risk surgery with potential sight-threatening complications. It may be considered in cases where the IOP is not controlled on 3 topical medications with a patent iridotomy.[37] At this time the choice is between doing a trabeculectomy versus a lens extraction, both being high-risk procedures. This decision should be taken by a glaucoma specialist and surgery should only be performed by an experienced surgeon at a tertiary eye care facility, where the facilities for corneal endothelial surgery (DSAEK/DMEK) and vitreo-retinal surgery are available in the event of complications. In eyes with a very shallow anterior chamber ($<$ 2 mm), a thick lens ($>$ 5 mm) and a high lens vault (ASOCT $>$ 1 mm), primary lens extraction may be considered as there is a high risk of trabeculectomy complications in such eyes due to pre-existing shallow AC.

Trabeculectomy may be considered in PAC patients having uncontrolled IOP despite medical therapy after lens extraction.

Hydrophobic square-edge monofocal IOLs are preferred in eyes undergoing lens extraction.

Follow-up

Biannual follow-up (6-monthly) should be done in eyes with PAC with ocular hypertension. Applanation IOP measurements, disc photography, RNFL OCT and visual field charting should be performed. In eyes with PAC post iridotomy with normal IOPs, annual follow-up is adequate. Patency of the LPI must be checked by looking at the lens capsule through the PI orifice on slit-lamp biomicroscopy.

Case study 2

Presentation

A 42-year old female presented to the OPD as a diagnosed case of bilateral angle-closure glaucoma. She had undergone bilateral LPI around 6 weeks back. She was on eye drops timolol BD, brimonidine BD and latanoprost HS in both eyes.

Examination and investigations

On examination, her best corrected visual acuity (BCVA) was 20/20 (OU). IOP was 24 mmHg (OD) and 14 mmHg (OS) on the aforementioned three topical medications. Central corneal thickness was 510 microns (OD) and 514 microns (OS). On slit-lamp examination, anterior chamber depth was Van Herick grade-1 bilaterally (Figure CS 2.1). On gonioscopic examination, forward iris bowing and narrow angle recess was confirmed (Figure CS 2.2). No angle structure was visible with gonioscopy without manipulation. On 90 D examination, vertical cup:disc ratio was 0.5:1 in both the eyes (Figure CS 2.3).

The axial length was 20.54 mm (OD) and 20.67 mm (OS). Anterior segment OCT revealed high lens vault with value being 1.5 (OD) and 1.05 (OS) (Figure CS 2.4).

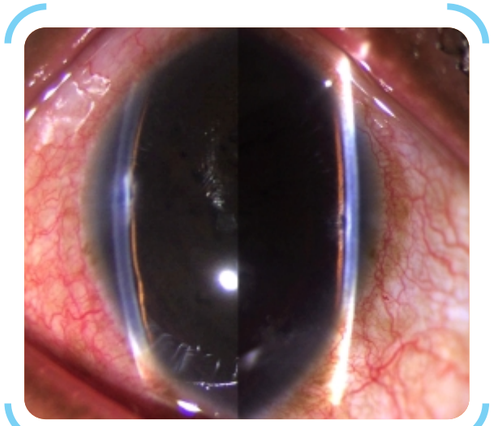


Figure CS2.1: Van Herick grade of the patient's anterior chamber.

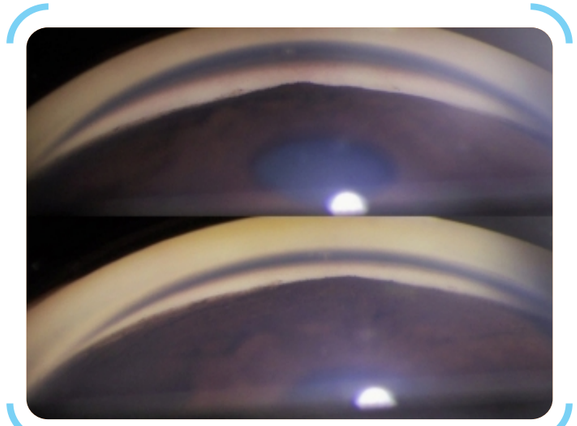


Figure CS2.2: Gonioscopy in primary gaze (above) shows no angle structures and on manipulation (below), trabecular meshwork is visible on the left.

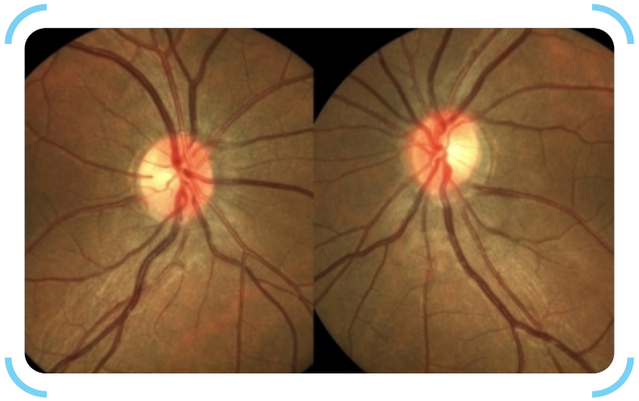


Figure CS2.3: Small optic nerve with cup:disc ratio of 0.5:1.

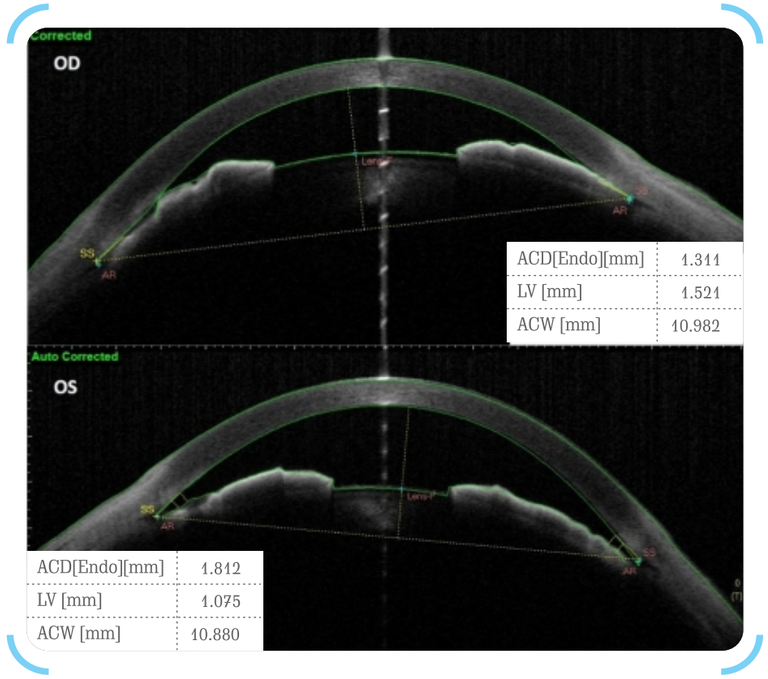


Figure CS2.4: High lens vault seen in both eyes.

Management

The patient was diagnosed to have bilateral Primary Angle Closure. Both iridotomies were patent. In view of high lens vault and uncontrolled IOP in the right eye, we decided to go ahead with the right eye lens extraction. Prostaglandin was stopped in the operated eye to reduce the chances of inflammation. The left eye was maintained at 3 medications. At 4 weeks of follow-up, IOP in the right eye was 14 mmHg with timolol 0.5% drops twice daily.

Discussion

This patient has a very shallow ACD and there was a very high lens vault pushing the iris forward with irido-trabecular contact, thus compromising outflow facility causing glaucoma. This can be effectively diagnosed with ultrasound biomicroscopy and ASOCT[38]. Lens extraction with an IOL opens the angle and prevents further closure and progression in such cases [39]. If IOP remains high on topical medications post lens extraction, a trabeculectomy can be performed.

9 Management of Primary Angle Closure Glaucoma (PACG)

Glaucomatous optic neuropathy in PACD is a sight-threatening situation and warrants immediate control of the IOP to maintain the optimal vision-related quality of life of the patient. The goal of management in PACG is to achieve a target IOP that prevents further damage to the optic nerve head.

PACG patients present with high baseline IOP, high inter-visit IOP fluctuations and are at a much higher risk for blindness as compared to POAG patients.[40] It is important to identify and note the baseline IOP, gonioscopy documentation of the amount of angle-closure, and severity of disc damage. The disc diagram may be drawn in the record sheet or an optic disc photograph should be taken. Once the diagnosis of PACG has been confirmed, the patient should be started on 2% pilocarpine eye drops TDS and LPI should be done in all the PACG eyes as soon as possible after lowering the IOP with topical and systemic glaucoma medications. After LPI has been performed, the medical therapy is on the same lines as POAG with PGA therapy as first-line, followed by beta-blockers, alpha agonists and topical carbonic anhydrase inhibitors. If the IOP is not controlled medically with these drugs, a trial of pilocarpine may be given before subjecting the patient to filtering surgery.[41]

Diurnal IOP fluctuation on medication after LPI and peak IOP along with the timings should be noted. Visual field examination should be repeated after every 4-6 months according to the severity of glaucomatous damage. The grading of glaucoma may be done according to the Hodapp- Parrish-Anderson (HPA) criteria (Table 9.1). Target IOP should be set according to the damage at presentation. The risk factors for progression include- thin central corneal thickness (CCT), family history, biometry, especially ACD, and lens thickness.[2]

Table 9.1: Hodapp-Parrish-Anderson (HPA) criteria

Minimum criteria for diagnosing acquired glaucomatous damage

A Glaucoma Hemifield Test outside normal limits on at least two fields; OR

A cluster of three or more non-edge points in a location for glaucoma, all of which are depressed on the pattern deviation plot at a $p<5\%$ level and of which is depressed at a $p<1\%$ level on two consecutive fields; OR

A corrected pattern standard deviation that occurs in less than 5% of normal fields on two consecutive fields

Classification of defects

Early defect:

- MD less than-6 dB
- Less than 25% of the points (18) are depressed below the 5% level and less than 10 points are depressed below the 1% level on the pattern deviation plot
- All point in the central 5° must have a sensitivity of at least 15 dB

Moderate defect:

- MD less than-12 dB
- Less than 50% of the points (37) are depressed below the 5% level and less than 20 points are depressed below the 1% level on the pattern deviation plot
- No points in the central 5° can have a sensitivity of 0 dB
- Only one hemifield may have a point with sensitivity of <15 dB within 5° of fixation

Severe defect (any of the following results):

- MD greater than-12 dB
- More than 50% of the points (37) are depressed below the 5% level or more than 20 points are depressed below the 1% level on the pattern deviation plot
- At least one point in the central 5° has a sensitivity of 0 dB
- Points within the central 5° with sensitivity <15 dB in both hemifields

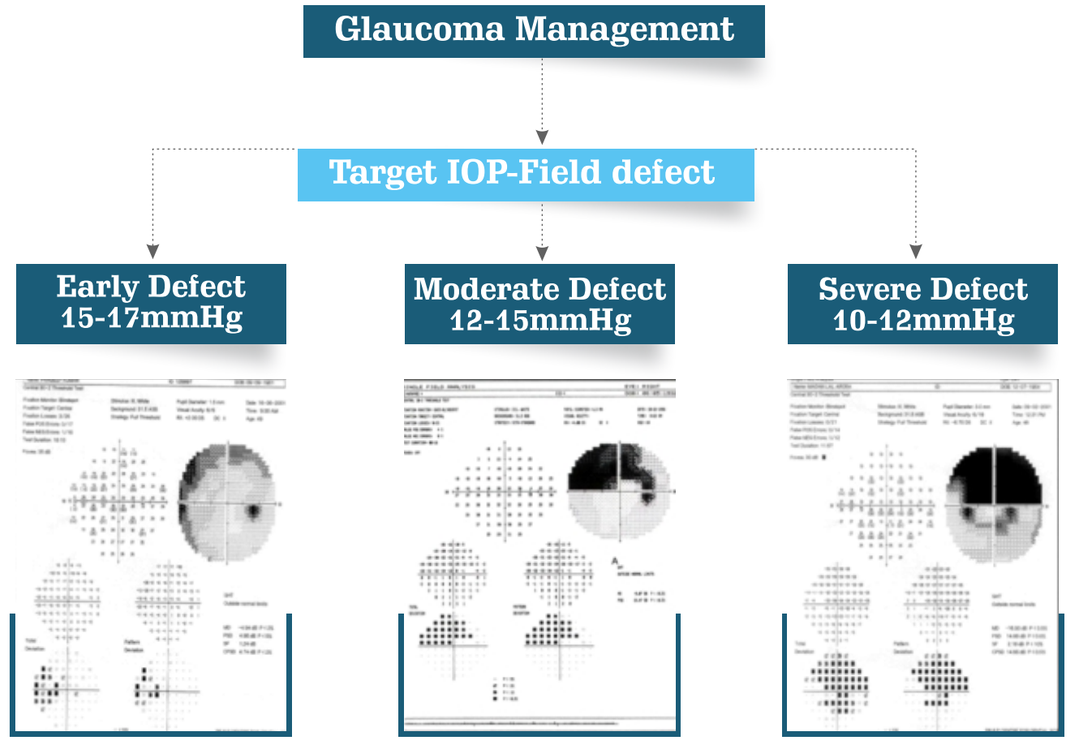


Figure 9.1: Deciding target IOP in Glaucoma.

Target IOP in PACG[42]

The target IOP should be set based on the severity of visual field damage and should be achieved with minimal medications at an affordable cost (Figure 9.1). In general, the visual field defect and an appropriate target IOP may be classified as -

Early defect - Target IOP < 18 mmHg

Moderate defect - Target IOP < 15 mmHg

Advanced defect - Target IOP <12 mmHg

In eyes with an early visual field defect, PGA/ beta-blocker/ alpha-agonist therapy can be started; however, in eyes with moderate/advanced damage, PGA therapy is mandatory and additional drugs can be added, if required. Further management can be done based on the control of IOP, degree of visual field damage, presence/absence of visually significant cataract, and patient affordability with respect to long-term medical therapy.

IOP on target with topical therapy and a visually significant cataract

In eyes with early/moderate disease, one can proceed with clear corneal phacoemulsification. In eyes with advanced disease, combined phaco - trabeculectomy can be considered to prevent the risk of post-operative IOP spikes and further deterioration of visual field.[43]

IOP not controlled on topical therapy and a visually significant cataract

If the IOP is not controlled despite using three to four topical medications, a combined phaco-trabeculectomy or manual SICS-trabeculectomy is the best option.[44] The IOP should be lowered by using hyperosmotics and systemic acetazolamide prior to surgery. If the angle is closed more than 180 degrees by PAS, filtering surgery must be added to cataract extraction.

IOP on target with topical therapy and no visually significant cataract

The patient can be asked to continue topical therapy and come for follow-up atleast once in 6 months for early/moderate disease and every 3-4 months for advanced disease. If the patient is not able to afford medications or there is an allergy to some glaucoma medications, lens extraction can be performed in eyes with an early disease however, a trabeculectomy is recommended in eyes with moderate – advanced disease. In eyes with very shallow anterior chamber depth (central corneal endothelium to lens capsule less than 2 mm), lens extraction should be a component of surgery as there is a high risk of trabeculectomy complications.

IOP not controlled on topical therapy and no visually significant cataract

If IOP is not controlled to target on topical therapy or there is progression in glaucoma, a trial of pilocarpine may be given in addition to the earlier medications. If pilocarpine trial also fails or if the patient is not able to tolerate it, lens extraction and/or filtering surgery - Trabeculectomy may be considered, depending upon the patient profile.

In a patient with an **early/ moderate visual field defect**, trabeculectomy augmented with mitomycin-C may be considered in expert hands and eyes with adequate anterior chamber depth (ACD).

In eyes with a critically shallow anterior chamber (ACD less than 2 mm), a thick lens (lens thickness more than 5 mm), or an anteriorly displaced lens (lens vault more than 1 mm), trabeculectomy is associated with a high complication rate and it would be prudent to do a two-stage surgery – lens extraction first, followed by trabeculectomy (if IOP is not controlled) or do a combined phaco-trabeculectomy, if the patient cannot come for follow-up, rather than a trabeculectomy alone. In PACG eyes with a very hard cataract where manual SICS is to be performed or if elective manual SICS surgery has been planned – it is better to do a combined manual SICS-Trabeculectomy in the same sitting.

In **moderate glaucoma** cases who are unable to follow-up or non-compliant or cannot afford medications or have drug allergy, trabeculectomy is a safer option.

In **severe or advanced glaucoma cases**, trabeculectomy augmented with mitomycin-C may be considered in expert hands and eyes with adequate ACD.

It is important to understand that performing cataract surgery in an eye with PACG post-trabeculectomy is quite difficult due to a very shallow anterior chamber and carries a high risk of corneal endothelial damage along with the risk of failure of the filtering bleb. On the other hand, performing a trabeculectomy in a pseudophakic PACG eye is an easier alternative and less prone to complications as compared to the phakic PACG eye.

This decision regarding lens extraction/ combined surgery should be taken by a glaucoma specialist and surgery should only be performed by an experienced surgeon at a tertiary eye care facility, where facilities for corneal endothelial surgery (DSAEK/DMEK) and vitreo-retinal surgery are available in the event of complications. Multifocal IOLs should not be used in eyes with PACG. Patients should be given information regarding the loss of uncorrected near vision immediately after surgery in eyes undergoing early lens extraction.

Currently, we do not recommend performing gonio-synechiolysis in the management of PACG.

Role of LPI

LPI should be done in all cases of PACG as it relieves the pupillary block, reduces IOP fluctuations and may have a role in the prevention of malignant glaucoma during trabeculectomy. In eyes with advanced PACG with complete synechial closure, there is a risk of significant IOP spike after LPI and systemic acetazolamide should be given along with topical ocular hypotensive medications.

Case study 3

Presentation

A 60-year old male patient presented to our OPD for a check-up. He was diagnosed to have bilateral angle-closure glaucoma 2 years ago, after which he underwent bilateral peripheral iridotomy and was started on latanoprost eye drops HS, brimonidine eye drops BD and timolol eye drops BD (OU). There was no history of any systemic illness. The patient presented with a recent complaint of progressive vision loss.

Examination and investigations

The BCVA in the right eye was 20/20 and in the left eye was 20/40. A thorough slit-lamp examination revealed a shallow anterior chamber with a clear lens in both eyes. Iridotomy was patent in both eyes. IOP was 18 mmHg (OD) and 26 mmHg (OS) on three topical medications. Central corneal thickness was 511 microns (OD) and 514 microns (OS). On gonioscopic examination, there was blotchy pigmentation and trabecular meshwork was not visible in 360 degrees (OU). The 90D examination revealed a vertical cup:disc ratio of 0.8-0.9 in the right eye and near total in the left eye (Figures CS 3.1a,b and 3.2).

The ACD in the right eye was 2.4 mm and left eye was 2.5 mm. The lens thickness in right eye was 4.35 mm and left eye was 4.39 mm.The lens vault in the right eye was 0.55 mm and left eye was 0.6 mm.

Diagnosis and Management

The diagnosis of bilateral PACG was made. Patient was started on timolol+brimonidine combination eye drops BD, travoprost eye drops HS and dorzolamide eye drops BD in both eyes. After 2 weeks, the IOP was 12 mmHg (OD) and 22 mmHg (OS). In view of uncontrolled IOP, adequate ACD and low lens vault, left eye trabeculectomy was performed.

Discussion

Here, we discuss a case of bilateral advanced PACG where IOP was high in spite of patent iridotomy in both eyes. Both eyes had above-target IOP and, therefore, modification of the drug therapy was tried. In the left eye, target IOP was not reached in spite of maximum tolerable medical therapy and, hence, a decision to perform a trabeculectomy was made in view of the adequate ACD and a clear lens.

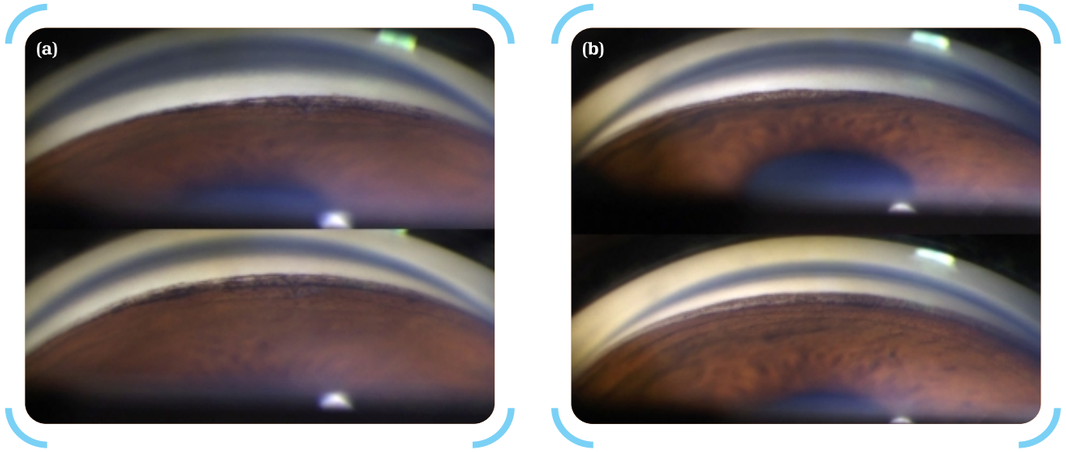


Figure CS3.1a and b: Gonioscopy in right eye and left eye shows blotchy pigmentation of the right and left eye with 360° angle closure.

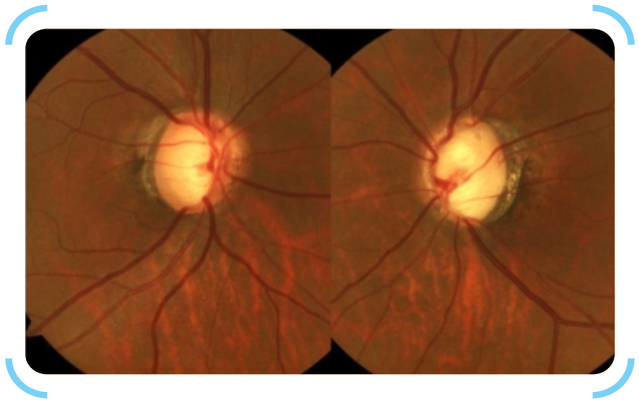


Figure CS3.2: Vertical cup:disc ratio of 0.8-0.9 in right eye and near total in left eye.

Case study 4

Presentation

A 54-year old man presented to our OPD with a complaint of occasional pain and frequent change in the power of his spectacles in both eyes. There was no history of any systemic illness.

Examination and investigations

Visual acuity was 20/32 (OD) and 20/200 (OS). On examination, anterior chamber was shallow with Van Herick Grading being 2 (OD) and 1 (OS) (Figures CS 4.1 and 4.2).

There was no history of any glaucoma medication use in past and baseline IOP was 26 mmHg (OD) and 38 mmHg (OS). Vertical cup:disc ratio was 0.7:1 (OD) and 0.9:1 (OS), and the central corneal thickness was 490 microns (OD) and 494 microns (OS). Gonioscopy revealed the presence of blotchy pigmentation and broad-based goniosynechiae with trabecular meshwork not being visible in both eyes (Figures CS4.2-4.4).

The axial length in the right eye was 22.02 mm and left eye was 21.82 mm. Examination with ASOCT revealed occludable angles with lens vault of 0.65 mm (OD) and 1.24 mm (OS). The anterior chamber depth was 2.4 and 1.86 in the right and left eye respectively.

Diagnosis and management plan

Diagnosis of PACG was made. The patient did not achieve the target IOP in the left eye with maximum tolerable medication. In view of high lens vault and diminished anterior chamber depth, the decision of combined phaco - trabeculectomy was made for the left eye.

Discussion

Lens vault is defined as maximum distance between horizontal line joining scleral spur and anterior surface of the lens. A high lens vault points towards an increased tendency of iridocorneal apposition due to forward push of the lens.[47] In such a case, a trabeculectomy alone may not be a good option due to increased risk of complications (shallow AC) and lens extraction is often necessary along with filtering surgery. Relative lens vault is another parameter that can further refine our decision making. [48]

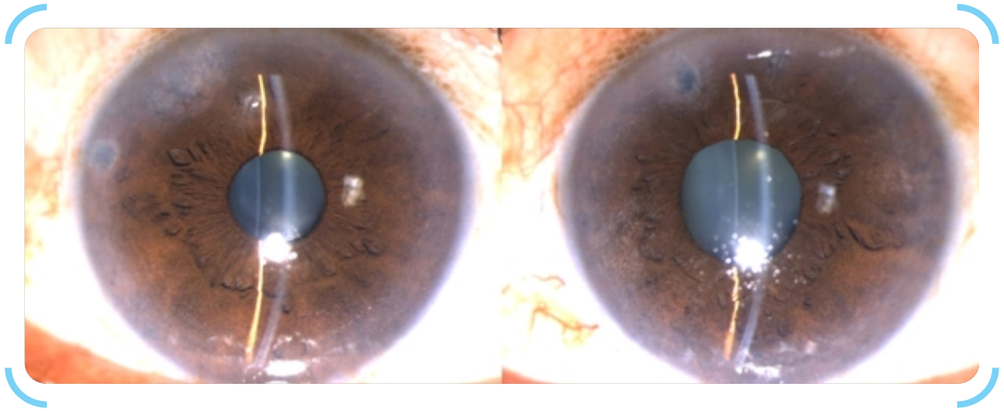


Figure CS4.1: Anterior segment photograph of right and left eye.

Figure CS4.2: Completely closed-angles in right eye (above) and left eye (below).

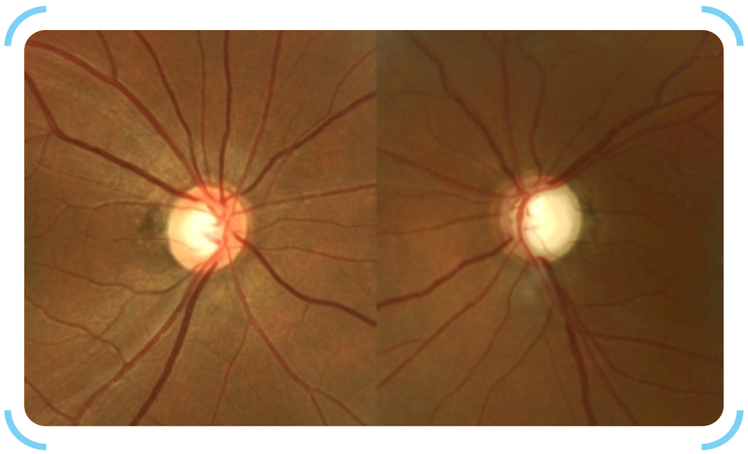


Figure CS4.3: Disc photograph of the patient showing right eye vertical cup:disc ratio of 07:1 and left eye near total cup.

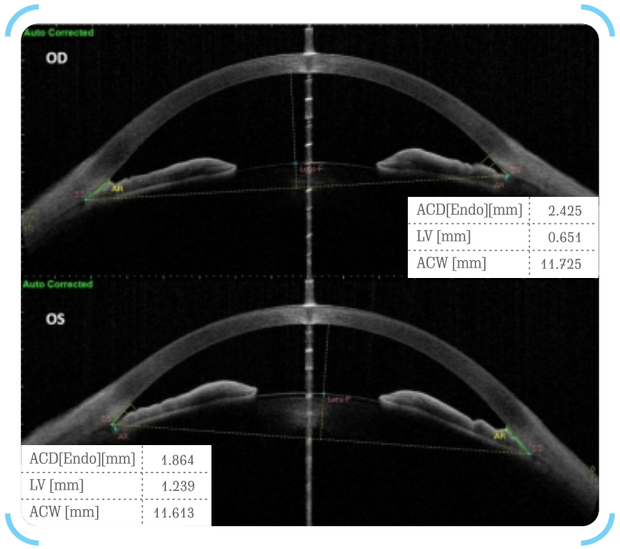


Figure CS4.4: Asymmetric lens vault in the right eye-0.65mm and left eye- 1.24mm.

10 Management of Acute Primary Angle Closure (APAC)

An acute attack of angle closure (AAAC), resulting from a complete or near complete closure of the anterior chamber angle, is characterised by a sudden high elevation in the IOP with associated decreased vision, severe pain in the eye, headache, corneal edema, ciliary congestion and a mid-dilated pupil. This is a sight-threatening emergency which requires immediate medical therapy to lower IOP followed by laser iridotomy in both eyes. (Figures 10.1 to 10.7).

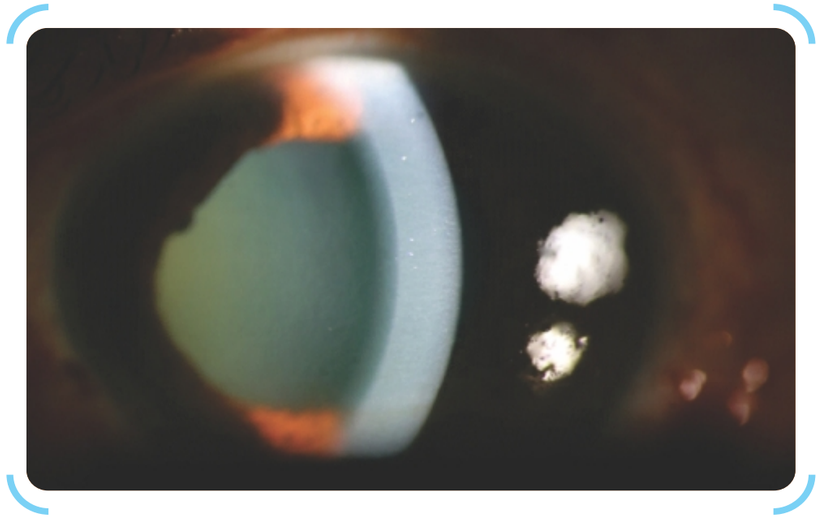


Figure 10.1: Corneal edema and mid-dilated pupil in an eye with APAC

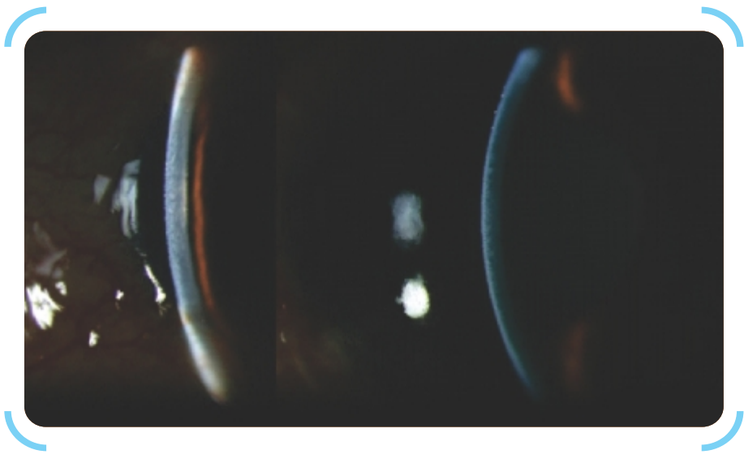


Figure 10.2: Shallow anterior chamber and iris convexity in APAC

Figure 10.3: Showing glaucomflecken (anterior subcapsular or capsular opacities of the lens, associated with focal epithelial infarct) following APAC

It is important to break the angle-closure attack as early as possible. Initial management is aimed at reducing the acutely elevated IOP in the eye, relieving the patient's discomfort and preventing optic nerve damage. The duration of the acute attack is significantly associated with lower corneal endothelial cell density in eyes with acute angle closure attack.[49] The treatment is initiated with medical therapy but definitive treatment in the form of LPI, done as soon as possible. Some of these patients would eventually require surgical intervention. The fellow eye must be evaluated and LPI performed as it is also at a high risk for developing an acute attack.

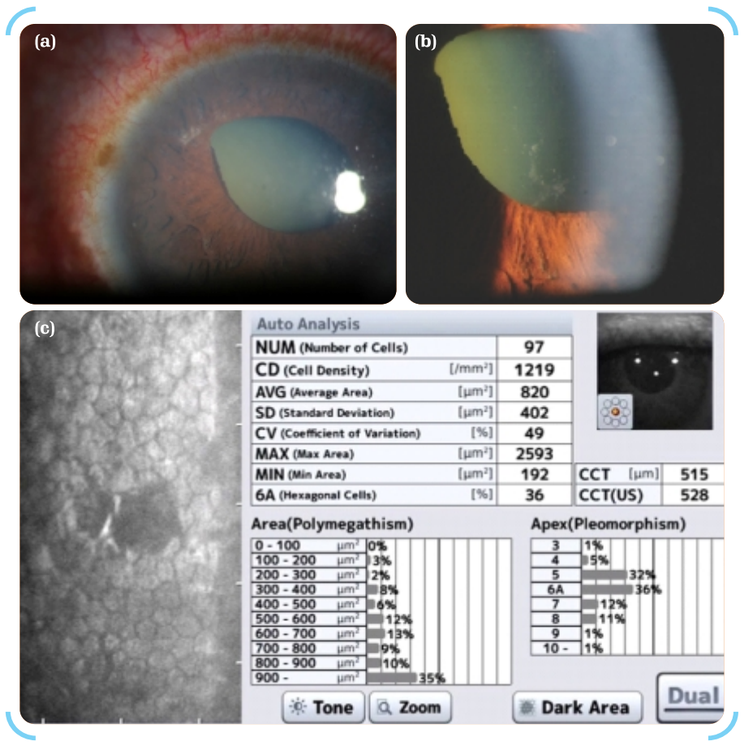


Figure 10.4 (a) Sphincter atrophy following an acute attack; (b): Glaucomflecken in the same eye; (c): Specular microscopy showing reduced corneal endothelial cell density following late presentation and delayed resolution of an acute attack.

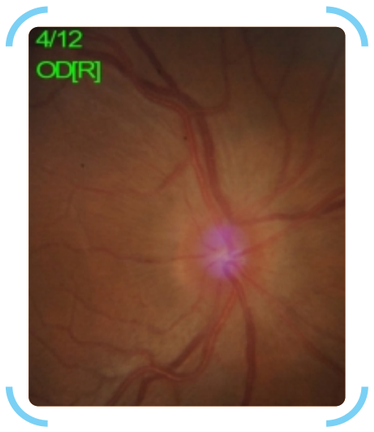


Figure 10.5: Disc edema in APAC

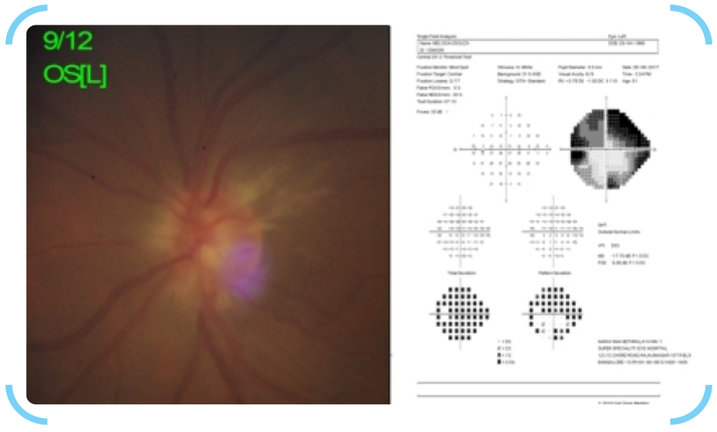


Figure 10.6: Disc cupping and field changes following APAC

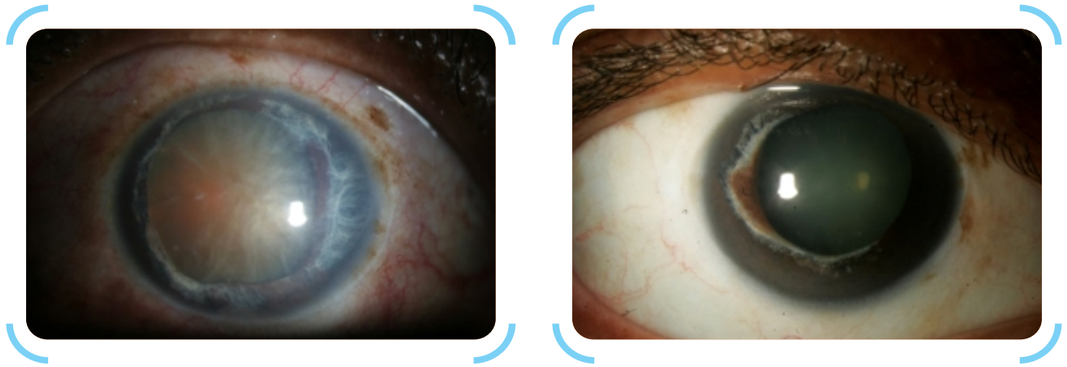


Figure 10.7: Iris atrophy and fixed, dilated pupil suggestive of APAC in the past

Medical Management

Medical therapy should be initiated after a brief history and examination to evaluate any contraindications to the use of hyperosmotic agents (mannitol and glycerol) and systemic acetazolamide (eg. heart disease, chronic kidney disease, diabetes, sulpha allergy etc). Initial therapy includes systemic medications such as oral carbonic anhydrase inhibitors (acetazolamide, 5–10 mg/kg) and hyperosmotic agents such as intravenous 20% mannitol (1–2 g/kg) and oral 50% glycerol (1–1.5 g/kg). After lowering IOP to below 30 mmHg, topical medications such as beta-blockers, carbonic anhydrase inhibitors and alpha-agonists can be added. Prostaglandin analogues may be avoided initially due to ongoing inflammation in the eye. Topical, direct-acting cholinergics such as pilocarpine should be started once the IOP has been lowered and the ischemic iris has been re-perfused. In addition to IOP lowering drugs, topical steroids help in controlling the inflammation in the eye. Systemic analgesics and anti-emetics may be prescribed to alleviate the patient's symptoms.

An attempt may be made to break the acute attack by indentation with a 4-mirror gonioscope or a swab stick pressed on the central cornea (risk of corneal epithelial injury/defect).

We do not recommend performing an AC paracentesis as there is a high risk of complications.

LPI

Lowering the IOP results in resolution of the corneal oedema, enabling a LPI to be performed in both eyes after pilocarpine is used to constrict the pupil. LPI alleviates the pupillary block and

results in resolution of the acute attack. Sequential use of argon and Nd:YAG is effective in these eyes with mid-dilated pupils and thick, dark irides; Argon laser burns thin the iris and YAG laser helps to penetrate it. Upto two-thirds of patients presenting with an acute attack can be satisfactorily treated with laser iridotomy alone and 60–75% of such patients recover without optic disc or visual field damage, if the IOP is promptly controlled.[26,50]

Following LPI, adequacy of IOP control is assessed and angles are re-evaluated by gonioscopy, to determine the extent of synechial closure. Disc and visual field evaluation aid in determining the extent of glaucomatous damage, guiding further management.

A prolonged duration of attack, presence of extensive areas of peripheral anterior synechiae (PAS), a higher IOP and a larger cup:disc ratio at presentation are other predictors that additional treatment in the form of medications or incisional surgery would be required even after the LPI.[51]

Argon Laser Peripheral Iridoplasty (ALPI)

In certain situations LPI cannot be performed due to a hazy cornea and the IOP remains elevated as the iris tissue plugs the trabecular meshwork. In this situation, ALPI can be used to cause peripheral burns on the iris surface which causes the iris to contract away from the trabecular meshwork and helps in resumption of aqueous flow through the trabecular meshwork.

Surgical Management of APAC

If IOP remains uncontrolled despite laser and medical treatment, surgical management may be needed. If a laser machine is not available, surgical iridectomy can be performed after the IOP is controlled medically. Performing a trabeculectomy to lower IOP in acute angle closure is not recommended[52] due to high risk of complications such as shallow AC and malignant glaucoma. Phaco-trabeculectomy should be performed, especially if associated with PAS> 180^{o} on indentation gonioscopy, disc or visual field damage. Eyes with a healthy optic disc and less than 180 degrees of PAS can be planned for a phacoemulsification lens extraction alone[53] with additional goniosynechiolysis to be performed by an expert surgeon at a tertiary eye care facility with facility for corneal transplantation and vitreo-retinal surgery to deal with the complications of surgery.

Follow-up

These patients require frequent follow-up and should be reviewed at 48 to 72 hours, 1 week, 4 weeks, 12 weeks and then every 6 months.

11 Principles of Medical Management in PACD

Principles of treatment

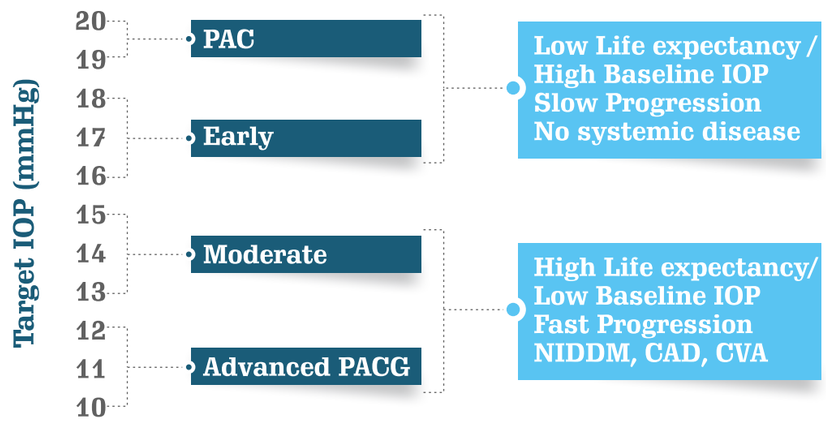
- Establishing a diagnosis
- Note the baseline IOP
- Setting of target IOP
- Start glaucoma therapy
- Halt glaucoma progression and maintain visual function
- Ensure no adverse effects on quality of life of the patient

Target IOP

Target IOP is defined as the mean IOP that prevents further progression of glaucoma. The baseline IOP and IOP post-medical therapy should be measured by Goldmann Applanation Tonometry preferably at multiple time-points in a given day to assess peak IOP and IOP fluctuation. It is important to note the central corneal thickness as an overestimation of IOP may occur with a thick cornea and underestimation of IOP with a thin cornea (especially post-refractive surgery). The target IOP is indivisualised based upon the glaucoma severity, baseline IOP, rate of progression, associated systemic disease, age and life expectancy of the patient (Figure 11.1).

- No glaucomatous optic neuropathy (PAC): target IOP18-20 mmHg
- Mild glaucomatous damage: target IOP 16-18 mmHg
- Moderate glaucomatous damage: target IOP 13-15mmHg
- Severe/ advanced glaucomatous damage: target IOP $\leq$12 mmHg

Figure 11.1 Setting Target IOP



How to start therapy?

Therapy should be started to achieve a target IOP with minimum possible medications at an affordable cost based upon the requirements of the patient. In eyes with pre-existing ocular surface disease, benzalkonium chloride (BAK)- free medications should be used. There are five classes of drugs currently available in India - prostaglandin analogues, beta-blockers, alpha-2 adrenergic agonists, carbonic anhydrase inhibitors and cholinergics. The drug of choice depends upon the baseline IOP of the patient, affordability and efficacy of the drug. If the baseline IOP is $\leq$30 mmHg, one medication should be started, if IOP is $>$30mmHg- two topical medications can be started to lower the IOP. Drugs should be added one at a time to evaluate the IOP lowering efficacy and this also helps to pinpoint causative agent for a drug allergy.

What therapy to start?

The first-line drug is usually a prostaglandin analogue or a beta-blocker in PAC and early PACG and prostaglandin analogue in eyes with moderate to advanced PACG. Systemic contraindications to beta-blockers must be ruled out. Alpha-2-agonists and carbonic anhydrase inhibitors are used as second-line drugs. Pilocarpine can be prescribed in PAC and PACG prior to laser therapy. If IOP is not controlled on maximal medical therapy post-LPI, a trial of pilocarpine may help to reduce IOP and prevent the need for surgery. However it leads to constriction of the visual field, therefore, may be avoided in moderate to severe glaucoma cases on a long-term basis. Drug holiday may be required in beta-blockers to avoid tolerance; and in patients on pilocarpine (dilate and do a fundus examination atleast once in 6 months) due to side-effects like posterior synechiae formation, iris cysts and retinal complications.

After starting therapy, the IOP should be re-evaluated after 4- 6 weeks ideally by doing a diurnal phasing (early morning to late evening). Another option, especially in eyes with asymmetric disease, is to start therapy only in one eye (worse disease). The response should be noted by measuring the difference in IOP between the two eyes (monocular therapeutic trial).

When to add or switch therapy?

If the first-line drug lowers IOP by atleast 20%, but the target IOP is not achieved, add a second medication. If the first drug lowers IOP by less than 20% (poor efficacy), switch to a different class of drug. Intra-class switch can be done between prostaglandin analogues. Combination therapy should be given to patients who require more than two drugs.

Patient education

All the patients on medical treatment for glaucoma should be educated about the disease, and the importance of putting life-long medication to prevent blindness and not as a cure. The side-effects of medications, interaction with systemic medications and signs of drug allergy should be explained. The method of instilling eye drops should be taught to minimise the side-effects, especially tear duct occlusion. They should be taught about methods to increase adherence such as putting daily alarms, etc.

12 Laser Therapy

Laser Peripheral Iridotomy

Purpose	To create an alternate pathway for aqueous to flow from posterior to anterior chamber, reducing the pressure gradient from the posterior to the anterior chamber and allowing the iris to fall back, thus reducing iridotrabecular contact.[40] It is relatively non-invasive and preferable to surgical PI. Earlier, argon laser peripheral iridotomy was used but nowadays, Nd:YAG (Neodymium Yttrium Aluminum Garnet) is the preferred modality.[54]
Indications of YAG PI	1- Acute primary angle closure glaucoma (APAC)- IOP is first lowered by topical and systemic medications. PI is done when oedema has subsided and cornea is sufficiently clear. 2- PACS- LPI can be done in high risk situations **Indications of PI in PACS are** Absolute indication- a- PAC or PACG in other eye / one eyed patient Relative indications- a- Positive family history of PACG b- Positive provocative test c- Patients on anticholinergic drugs d- Require frequent pupillary dilation for retinal examination e- Patient who cannot come for regular follow-up 3- All PAC eyes- Around 22% of these patients will develop PACG in 5 years, so PI is very useful in delaying progression of the disease. 4- All PACG eyes- PI can be helpful in avoiding acute IOP rise but additional medication or surgical intervention is mostly required. 5- Secondary angle closure with pupillary block
Contraindications	1- Uncooperative patient 2- Poor visualisation of iris- due to corneal oedema or opacity 3- Flat anterior chamber- attempt of PI in a flat anterior chamber can result in corneal burn 4- Absence of pupillary block- e.g. ciliary body rotation, neovascular glaucoma, ICE syndrome, uveitic glaucoma, etc.
Pre-laser evaluation	1- Carefully look for corneal oedema 2- IOP should be controlled. If not controlled on topical medication, oral acetazolamide or intravenous mannitol can be used. 3- Check if the patient is on some anticoagulant, especially in one eyed patients and they should be stopped at least 5 days before laser.
Pre-laser preparation	1- Procedure, expected outcome and potential complications should be explained to the patient in detail. 2- A written consent should be taken. 3- 1-4% pilocarpine should be instilled in the eye to be lasered. This stretches the iris and facilitates laser penetration. 4- Eye drops 1% apraclonidine, 0.2% brimonidine are applied to reduce post-operative IOP spike and inflammation. 5- 0.5% proparacaine or 4% xylocaine eye drops is used to anaesthetize the ocular surface.

	6- Abraham contact lens, +66D is most commonly used. It stabilises the eye, keeps the lid apart and provides a magnified image of the iris. It also concentrates the energy on a small area. 7- Site- A crypt is identified in the superior part of iris as the site for iridotomy. An iridotomy formed at this site will be covered by upper lid hence avoiding post-procedure diplopia or glare [55].
Lasers used for PI	1- Nd:YAG - 1064 nm 2- Argon green laser- 514 nm 3- Sequential: Argon laser to stretch and thin out the iris followed by Nd:YAG application. It is useful in thick iris. Also, coagulation with argon reduces chances post procedure bleed.
Parameters for Nd - YAG application	1- Energy setting- 3-7 millijoules/shot. Higher energy required for thick and heavily pigmented iris. 2- Number of shots- one to three pulses per burst. 3- Spot size- It is fixed at 50 microns. 4- Focus- beam should be focused slightly posterior to anterior surface of iris 5- End point- gush of fluid/pigments from posterior to anterior chamber and deepening of anterior chamber 6- In case of a lamellar PI, a repeat procedure at same or different site can be done after 1 week.
Post-laser management	1- Topical steroid or NSAID 4 to 6 times per day for 1 week to reduce inflammation. 2- IOP should be rechecked after 1 to 2 hours of procedure for any spike. Topical or systemic glaucoma medications can be used for control [56] of IOP 3- Stop pilocarpine; glaucoma medications should be titrated according to response in the post-operative period. 4- Verify patency of iridotomy with visibility of anterior lens capsule/ transillumination after 1 week 5- Pupillary dilation can be done to break suspected posterior synechiae, once PI is patent.
Complications, prophylaxis and management	1- Hemorrhage- commonest side-effect sometimes may be severe enough to cause significant hyphema. Gentle pressure on the globe with contact-lens can reduce bleed. 2- IOP elevation- due to pigments released and inflammation during procedure. It is generally transient but may require scaling-up medical treatment for a short duration. 3- Uveitis and post-operative inflammation- generally mild and subsides spontaneously 4- Posterior synechiae 5- Temporary blurring of vision 6- Damage to cornea, retina and lens- generally not significant to warrant any treatment [57] 7- Temporary visual disturbance 8- Closure of iridotomy- this occurs due to excessive healing response and requires a repeat procedure 9- Dysphotopsia

Argon Laser Peripheral Iridoplasty (ALPI)

Purpose	Krasnov first used laser spots placed near the root of the iris, to open up the angle.[58] ALPI involves placement of low energy spots near the root of the iris, causing contraction of the iris stroma and subsequent opening of the iris apposing trabecular meshwork along with deepening of the angle.
Indications	It is used in conditions where peripheral iridotomy has either failed, not possible or not effective. 1- Plateau iris syndrome 2- AACG with thick/ boggy iris where PI is not possible or failed 3- Angle closure due to thick or anteriorly positioned lens 4- Angle closure due to anterior rotation of ciliary body 5- As an adjunct or to facilitate laser trabeculoplasty
Contraindications	1- Corneal opacity or oedema obscuring visibility 2- Flat anterior chamber
Pre-laser preparation	1- Explain to the patient about possible outcomes and complications 2- Obtain a written explained consent 3- Constrict the pupil with 1-4% pilocarpine to facilitate peripheral laser application 4- Instill topical 0.5% proparacaine or 4% xylocaine for anesthesia 5- Lens: Abraham iridotomy (+66D button) contact lens is commonly used 6- Eye drops 1% apraclonidine, 0.2% brimonidine are applied to reduce post-operative IOP spike and inflammation.
Lasers used for iridoplasty	Different lasers which can be used are: 1- Argon laser- 514 nm most commonly used 2- Diode laser- 810 nm 3- Frequency double Nd:YAG laser- 532 nm
Parameters for APLI and its application	1- Energy- start with 240 Mw and increase power for darker iris 2- Spot size- 500 microns 3- Duration- 0.5-0.7 second 4- Location of spot- 10 to 40 applications over 180 to 360 degree; target the spot at the most peripheral location possible. There should be a gap of 1 to 2 spot diameter between spots [59]. 5- If the peripheral chamber too shallow, then mid-peripheral laser spots can be placed to make space 6- End point- there is contraction of iris stroma with deepening of anterior chamber. Reduce power if a "pop sound" is heard or bubbles are seen.[60]
Post-laser management	1- Topical steroid or NSAID 4 to 6 times per day for 1 week to reduce inflammation. 2- IOP measurement after 1 or 2 hours can be done to check any IOP spike. Titrate anti-glaucoma medications for adequate control of IOP 3- Gonioscopic examination should be done in follow-up to look for any developing synechiae and ascertain need of any additional procedure
Complications, Prophylaxis and management	1- Inflammation and iritis- post-operative steroids and NSAIDs are given prophylactically 2- Increase in IOP- brimonidine or apraclonidine before the procedure decreases the chances of IOP spike 3- Mydriasis 4- Corneal endothelial burns 5- Iris atrophy- over treatment should be avoided 6- Pre-existing peripheral anterior synechiae don't open up and additional PAS may form.

13 What are the special considerations for performing trabeculectomy in PACG?

When to do trabeculectomy in PACG?

- Surgery is indicated only when the threat to vision due to glaucoma is greater than the risk of the selected surgical procedure.

What are the indications of trabeculectomy in PACG?

- Target pressure not reached with maximum tolerable medical therapy
- Progressive glaucomatous damage despite reaching target
- Allergy to glaucoma medications
- Non-compliant patient
- Non-affordability

What precautions should be taken while performing trabeculectomy in PACG?

Pre-operative

- Good preoperative IOP control is mandatory- with systemic and topical glaucoma medications
- Pilocarpine may be stopped in pre-operative period to avoid further shallowing of the anterior chamber
- Peri-bulbar block followed by ocular massage ensures appropriate hypotony

Intra-operative

- Slow decompression of the anterior chamber to avoid malignant glaucoma
- Anterior chamber maintainer/ visco-cohesive viscoelastic may be used to avoid collapse of the anterior chamber
- Preplaced sutures over the scleral flap for immediate closure may be applied
- Releasable sutures over the scleral flap may be applied to avoid shallowing of the anterior chamber post-trabeculectomy
- Subconjunctival injection of atropine for good cycloplegia may assist in deepening of the anterior chamber

Post-operative

- Good pupillary dilation with the use of mydriatics and cyloplegics to avoid a shallow anterior chamber
- Releasable sutures should be removed only after 1st week of surgery
- Post-operative steroids should continue for 6-8weeks, antibiotics and mydriatics from 2-4 weeks of surgery

14 What are the special considerations for performing a lens extraction in PAC and PACG?

The surgical management of cataract in PACG presents the surgeon with numerous challenges. A sound knowledge of the anatomy of small eyes and pathophysiology of angle-closure disease, combined with thorough preoperative evaluation and tailoring each surgical step according to each individual case can enable the surgeon to achieve a good post-operative outcome.

1. **Preoperative workup should stress on the following:** (a) status of corneal endothelium- specular count; (b) AC depth; (c) IOP; (d) pupil dilation, extent of posterior synechiae; (e) gonioscopy for extent of synechiae in angle; (f) cataract grade; (g) subluxation of lens, if any (h) status of optic disc and retina; (i) previous history of malignant glaucoma or suprachoroidal haemorrhage in the fellow eye, if operated and (j) review of current anti-glaucoma prescription.

2. **Preoperative counselling:** It is important to explain to patient the nature of the disease, possibility of intra/postoperative complications and visual prognosis. A written consent is essential.

3. **Biometry and IOL power selection:** Since the eye is likely to be hyperopic with shallow AC and short axial length, special care is require to perform biometry and for IOL power selection. Hoffer Q, Holladay 2 and Barrett Universal II IOL power calculation formulae are preferred. The results obtained by all the three are taken into consideration to arrive at the required IOL power. However, IOL power calculation is tricky in such eyes and postoperative refractive surprise may occur.

4. **Surgical challenges:** An eye with PACG disease behaves differently from a normal eye. The surgeon must be aware of these challenges and the means to tackle them.

   a. Adequate steps must be taken to reduce IOP before surgery. Aqueous suppressants should be continued on the day of surgery. IV mannitol 30-60 minutes before surgery (unless contraindicated) helps to lower IOP, shrink vitreous and increase working space in the AC. The dose is 1.5-2g/kg given IV over 30-60 minutes before surgery. Sufficient lowering of IOP before surgery is important to avoid the complication of suprachoroidal haemorrhage.

   b. Miotics and prostaglandin analogues cause breakdown of blood-aqueous-barrier (BAB) and should preferably be stopped prior to the surgery - atleast 7 days.

   c. Oral anticoagulants such as warfarin and aspirin should be discontinued, if possible, after consultation with physician. These drugs can lead to hyphema, in case of intraoperative iris manipulation.

   d. Topical or subtenon's anaesthesia is preferable to peribulbar/ retrobulbar block. Retrobulbar block, vigorous digital massage and application of super-pinky are best avoided.

   e. Lid speculum should be applied in such a way that it should not exert pressure on the globe.

   f. Patient should be dilated carefully immediately before commencement of surgery in order to avoid precipitating an ACG attack.

g. A limited pars-plana-vitrectomy may be performed initially to remove a small volume of vitreous especially if surgery is being performed in an acute angle closure attack.[61]

h. Proper wound construction is important. Clear cornea incision (CCI), just enough in length to provide a snug fit for phaco-tip, is preferred. A small and anteriorly placed CCI helps to prevent wound leak and iris prolapse respectively. This is especially important since iris in such eyes is more likely to be atonic and floppy.

i. AC must be decompressed gradually, to allow choroidal vessels time to adapt to the new pressure. Sudden decompression may cause dilation of choroidal vascular bed, rupture of choroidal vessels, and may even lead to suprachoroidal haemorrhage.

j. A high molecular-weight viscoelastic (OVD) such as Healon 5 is preferred to create space for intraocular manoeuvres and to protect endothelium. Viscoelastics also aid in mydriasis and are required for synechiolysis, if required. Soft-shell technique is preferred. First a dispersive OVD is injected followed by a cohesive device. Dispersive OVD helps to protect the endothelium while the cohesive OVD helps to maintain the AC and capsular bag.

k. PACG eyes often have small pupils due to chronic miotic therapy, atrophy of dilator pupillae and the presence of posterior synechiae. Pupillary dilation may be achieved by visco-mydriasis, mechanical release of adhesions (synechiolysis), making a series of mini-sphincterotomies by Vannas scissors (not much preferred) or use of pupil expansion devices such as Malyugin ring and iris hooks. In the scenario of a very shallow AC, iris hooks may be preferred over other pupil dilating devices since the latter may cause more endothelial trauma.

l. Intraoperative shallowing of the AC, should be avoided. Elevation of bottle height and decreasing flow rate may be considered to further stabilize the AC.

m. Sufficiently large capsulorhexis should be made to enable easy removal of large bulky nucleus.

n. Phacoemulsification method of cataract surgery is preferred because of the small wound and closed-chamber technique. An initial trenching may be done to debulk the nucleus and increase working space in the AC. In-the-bag phacoemulsification is preferred, away from corneal endothelium. Chopping of fragments should be done slowly. There should be a low threshold for injecting OVD repeatedly. The rest of the surgery is similar to routine cases. However, phaco parameters and fluidics should be adjusted to control intraoperative IOP rise which could prove deleterious in a patient of glaucoma with an already compromised optic nerve head circulation.

o. The site of LPI or surgical iridectomy may harbour areas of zonular weakness. Even minimal surgical manipulation in such areas may lead to zonular dehiscence and vitreous prolapse.

p. Excessive fluid injection during hydrodissection or during irrigation-aspiration of cortical remnants must be avoided, since this can lead to acute intraoperative IOP rise with shallowing of the AC. This is called "infusion misdirection syndrome" and occurs due to the migration of irrigating fluid into the posterior segment via zonular fibers.[62]

q. IOL should be placed in the bag. Hydrophobic square edge acrylic "foldable" IOL should be preferred - this may reduce PCO formation. Though the required IOL power may be high (sometimes +35D or higher), a single IOL implant is preferred over multiple implants.

r. Goniosynechiolysis or surgical PI may be combined with cataract surgery.[63]

s. Thorough removal of OVD from AC and behind IOL is essential to prevent post-operative IOP spike due to retained OVD.

5. **Post-operative management:** Patients are placed on steroid-antibiotic and mydriatic eyedrop regimen. Steroids are used frequently to control inflammation. A topical NSAID eyedrop may be used to prevent post-operative cystoid macular oedema (CME). Careful monitoring of IOP in the post-operative period is essential. However, despite an uneventful surgery, PACG eyes are at a risk of developing "aqueous misdirection syndrome" post-operative. Patients should be followed-up for pain with shallowing of AC. Initial treatment is with topical and oral aqueous suppressants and vigorous cycloplegia. A prophylactic irido-zonulo-hyaloidotomy (IZH) at the completion of cataract surgery in the second eye may be done if there is a history of aqueous misdirection in the operated fellow eye.[64]

15 What are the special considerations for performing cataract surgery after trabeculectomy in PACG?

Performing cataract surgery in a PACG eye with an existing trabeculectomy bleb poses unique challenges for the anterior segment surgeon. The data from the Advanced Glaucoma Intervention Study (AGIS) showed that trabeculectomy, whether performed as primary procedure or after institution of medical therapy, increases the risk of cataract formation by as much as 78%.[65] AGIS data also showed that postoperatively a flat AC and exaggerated inflammation further increased the risk for developing cataract in such patients by 14%.

The aim of surgery is to restore visual function without compromising bleb function. Phacoemulsification technique is preferred because it does not involve any conjunctival handling. Manual small incision cataract surgery (SICS) may also be performed. Surgery can have an adverse effect on long-term bleb survival. When possible, cataract surgery should be done 6 months after trabeculectomy. Chen et al. suggested that optimum time for cataract surgery is atleast 6 months after trabeculectomy in order to increase the chances of bleb survival.[66] Seah and colleagues also suggested that the longer the time between trabeculectomy and cataract surgery, better the IOP control.[67] The challenges in such cases are similar to those in PACG eyes:

1) Limited space superiorly because of presence of bleb
2) Shallow AC, hypotony
3) Pre-existing zonular weakness
4) Chances of increased postoperative inflammation
5) Post-operative bleb failure

Pre-operative workup is similar to cataract surgery in PACG. Patient should be counselled about intraoperative complications and possibility of failure of glaucoma surgery post-operatively. Additionally, evaluation of IOP and bleb anatomy and function via slit-lamp and ASOCT (if available) should be done. Gonioscopy should also be done to check patency of internal ostium.

If IOP is high pre-operatively, a decision to revise filtration bleb (bleb needling) at the time of surgery with or without antimetabolite may be made. In case of a previous failed trabeculectomy, trabeculectomy at a different site or placement of a tube-shunt can be done at the same time as cataract surgery. If IOP is normal, vigorous anti-inflammatory measures must be instituted after surgery to prevent bleb fibrosis/ failure after surgery. If, however, IOP is low, certain steps of cataract surgery such as paracentesis, capsulorhexis and nuclear cracking may become difficult. IOL power calculation also becomes difficult in such eyes. Therefore pre-operative assessment is very important and a sound surgical plan must be in place before proceeding to cataract surgery. Precautions are similar to surgery in PACG eyes.

Few modifications in the surgical technique may be required:

a) Speculum is applied gently to avoid trauma to the bleb area. Rough application of the speculum may cause de-roofing or even rupture of the bleb. The speculum should not exert undue pressure on the bleb area.

b) Desiccation of the bleb can be avoided by coating the bleb area with a dispersive viscoelastic agent (such as Viscoat). This manoeuvre may be repeated several times during the surgery to protect the bleb.

c) A temporal CCI away from the bleb area is preferred. Side-port incisions should also be away from the bleb.

d) Endothelial protection is achieved with the use of dispersive viscoelastic agent.

e) Plugging of the internal ostium in order to prevent excess flow of fluid into the bleb is achieved by first injecting a dispersive OVD at the site of the ostium, followed by injection of a cohesive agent.

f) Iris hooks, when used, should be placed away from the bleb, so that they don't exert pressure on the bleb. Unnecessary iris handling should be avoided as this can lead to increased post-operative inflammation and thus higher risk of the bleb failure.

g) Sudden AC shallowing must be avoided. Surgery is performed as described in the previous section.

h) Trypan blue, used for anterior capsular staining, can be used to observe the bleb function. A well-functioning trabeculectomy bleb stains diffusely with the dye.

I) Meticulous care should be taken to prevent PCR or iatrogenic zonular dehiscence. However, in such an event, complete anterior vitrectomy (using intracameral triamcinolone, if required) should be performed since residual vitreous in the AC can block trabeculectomy ostium and lead to filter failure.

j) PCIOL should be placed in the bag. Angle-supported anterior chamber IOLs should be avoided, as the IOL footplate may disrupt and block the sclerostomy, causing inflammation and, ultimately, bleb failure.

k) Complete removal of viscoelastics is done to prevent post-operative IOP spike.

l) At conclusion of the surgery, bleb function must be tested by injecting balanced salt solution (BSS) into the AC. This will cause bleb to balloon up. If not, an internal bleb revision may be done by passing a cyclodialysis spatula though wound/ paracentesis port to the sclerostomy fistula and underneath the scleral flap. Seidel's test can be performed to confirm that there is no leak in the bleb area.

m) If bleb is assessed to be at a risk of bleb failure, subconjunctival (S/c) injection of 5-fluorouracil (5FU) 5mg/0.1ml may be given after the eye has been made water-tight.[68] 5FU should preferably be injected by a 30G needle. Any leakage should be soaked up by a cotton swab /Merocel sponge placed on the injection site. Precaution must be taken to prevent the entry of 5FU into the AC. Mitomycin-C (0.2-0.4 mg/ml) may also be used transconjunctivally.[69]

Post-operative management

Close follow-up in the post-operative period is required to monitor IOP and signs of the bleb failure. Topical steroids must be given frequently to decrease postoperative inflammation. In case of IOP spikes, anti-glaucoma therapy must be instituted. The bleb deserves special attention. Presence of corkscrew vessels and bleb encapsulation may point towards scar formation and subsequent bleb failure. S/c 5FU can be given in such a case.

Thorough pre-operative workup, careful surgical planning and preparation on the part of surgeon for intra/postoperative adjustments and interventions can lead to better visual rehabilitation and long-term IOP control in patients with PACG disease.

16 How to diagnose and treat malignant glaucoma post-trabeculectomy for PACG?

Malignant glaucoma is a condition with uniform shallowing of the anterior chamber in the presence of a patent iridotomy, with elevated or normal IOP.[70]

2-4% of eyes with ACG develop vitreous block after trabeculectomy.[71] It can also occur after LPI, laser iridotomy, Laser capsulotomy, cataract surgery, glaucoma implant surgery, deep sclerectomy, vitrectomy, laser suture lysis after trabeculectomy, and trans-scleral ciliary ablation and rarely spontaneously. [72]

Though the exact mechanism is unclear, there is, essentially, a forward rotation of the ciliary processes, with or without a supraciliary effusion and choroidal expansion, which causes an altered relationship between the ciliary processes, lens and the vitreous. A forward displacement of the lens- iris diaphragm and a predominant posterior flow of aqueous into the vitreous body occurs.[73,74] A patent iridotomy is essential to rule out pupillary block.

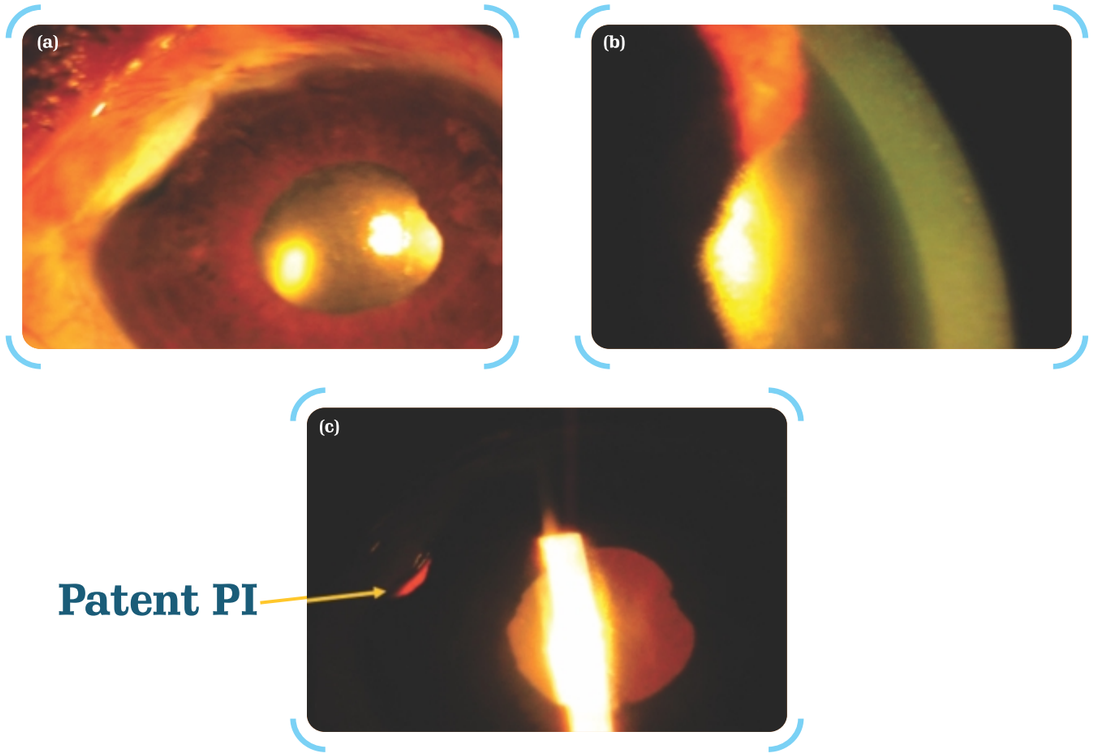
Risk factors

The risk factors are as follows

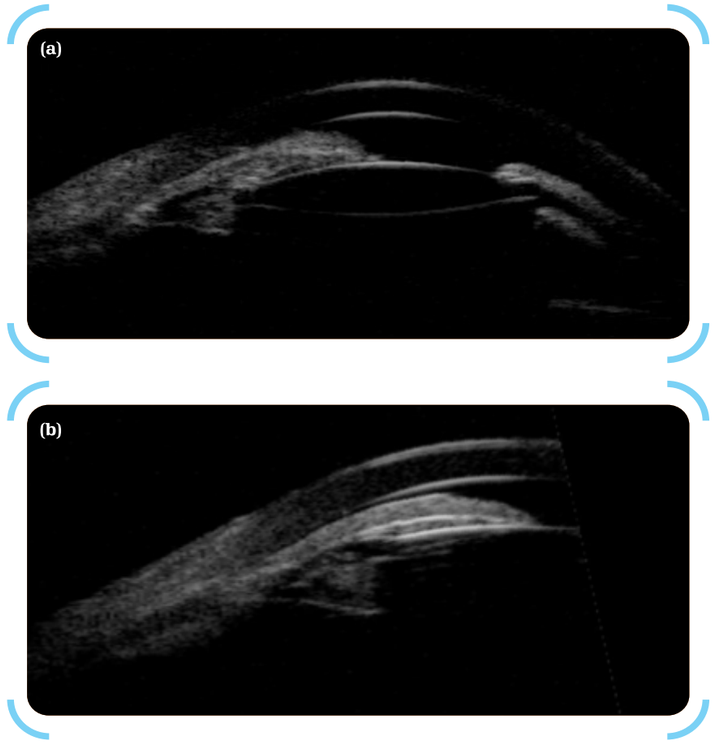
- Plateau iris
- Vitreous block in the fellow eye
- Nanophthalmos
- Zonular laxity
- Pre-operative shallow AC
- Pre-operative high IOP
- Sudden shallowing of the anterior chamber in the perioperative or post-operative period.

Clinical features

Vitreous block is to be considered, when there is a uniform shallowing of the AC with high or normal IOP, and with a patent PI, in the absence of a choroidal haemorrhage. Vitreous block can occur anytime from the first post-operative day till years after surgery.[71] It may occur in phakic, aphakic or peudophakic eyes ((Figures 16.1a,b and c). Patients present with a red, painful eye and blurred vision. Associated headache and nausea/ vomiting may be present, depending on the level of IOP. A myopic shift in refraction may occur due to the anterior movement of the lens.[75]



Figures 16.1a,b,c: Anterior segment photograph of pseudophakic eye post-trabeculectomy with raised IOP, shallow AC and patent PI- malignant glaucoma



Figures 16.2a,b: UBM of eye with malignant glaucoma showing flat AC with anterior rotation of ciliary processes.

Imaging

UBM is an important tool. UBM can demonstrate the uniform shallowing of AC, anterior rotation of ciliary body, and a patent PI (Figures16.2a and b). Small supra-choroidal effusions can be demonstrated in some eyes. These effusions contribute to the anterior rotation of the ciliary processes.[76] Quantitative measurements show that the angle between the posterior corneal surface and the anterior ciliary body in affected eyes is less than half of that in the unaffected eye. The posterior chamber is unrecognisable by UBM in these eyes and reappears after successful management.

Management

The aim is to restore the normal flow of aqueous from the posterior chamber into the anterior chamber and out through the trabecular meshwork/ internal ostium.

A sequential management approach is recommended. Initial medical management includes aqueous suppressants, namely acetazolamide, and hyperosmotics (mannitol IV). They help in shrinking the vitreous gel, and relieving the block between the ciliary body and the lens/ vitreous face. Topical cycloplegic-mydriatic like atropine 1%, tightens the lens zonules by relaxing the ciliary body, pulling the lens-iris diaphragm posteriorly, helping to break the ciliary block. If good response is seen with progressive deepening of the AC and lowering of the IOP, medical management can be continued up to 3-5 days. Some patients may need long-term topical cycloplegic- mydriatics to prevent recurrence. Medical treatment should be continued only if successful, else one should promptly proceed to other modalities. Presence of lens cornea touch mandates surgical intervention within 24 hours.

Laser: Laser photocoagulation of the ciliary processes either through slit lamp delivery, if they are visible through an iridectomy in aphakic eyes, or by trans-scleral cyclophotocoagulation can cause shrinkage of the ciliary processes and can break the block in some eyes. YAG laser capsulotomy and hyaloidotomy in aphakic eyes have also been reported to help in resolution, but are associated with recurrences.

Surgery : The definitive management of vitreous block is surgical. The procedure of irido-zonulo-capsulo-hyaloido-vitrectomy, where a communication is established between the anterior chamber and posterior segment, is the most effective surgery.[77] Essentially, the eye is made unicameral. (Figure 16.3)

Anterior approach: This can be done by the anterior segment surgeon using a vitrector through the surgical iridectomy, cutting the zonules, posterior capsule, and the anterior hyaloid face, and the underlying vitreous.[78] An immediate deepening of the anterior chamber is seen. If the patient is phakic, phacoemulsification is to be considered in addition to the above surgery. As the anterior chamber can be very shallow and positive pressure from the vitreous gel can make phacoemulsification difficult, initial decompression by limited vitrectomy can help in deepening of the AC, and after phacoemulsification, the vitrectomy can be completed. Recurrences have been reported after the anterior approach.

Posterior Approach: The other option is a complete three-port pars plana vitrectomy, with a zonulohyaloido iridectomy from posterior to anterior, using the vitrector. Pars plana vitrectomy alone may not be effective and recurrences are reported. The establishment of a communication between the posterior and anterior segment is key to resolution of the condition.[79]

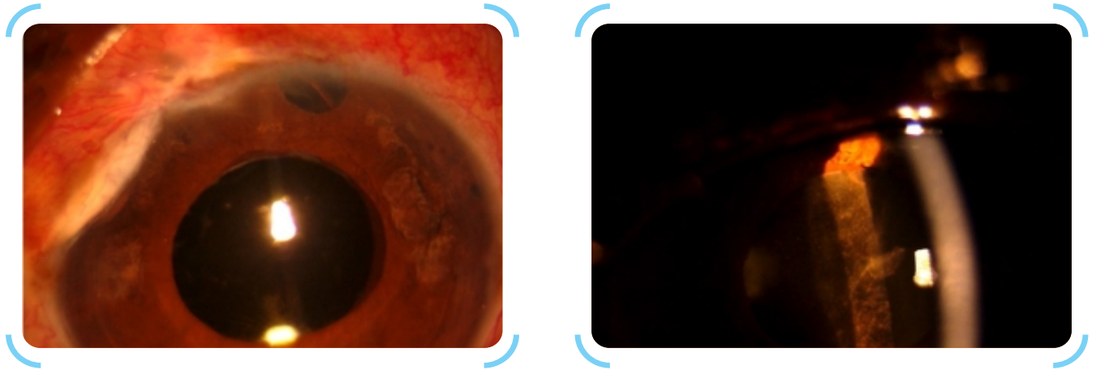


Figure 16.3: Post-operative anterior segment photograph showing resolution of corneal edema and deep AC.

Prognosis

Recurrence is common after medical and laser management. Definitive surgical management has very low recurrence rates, and if performed early enough, carries a good prognosis.

Prevention

- Laser iridotomy in all eyes with ACG prior to trabeculectomy
- Lower IOP medically before surgery
- Avoid sudden lowering of IOP, and shallowing of AC by using releasable sutures
- Use cycloplegic-mydriatic on the table post-operatively, and in the post-operative period

17 How to diagnose and manage plateau iris configuration and syndrome?

Plateau iris configuration (PIC) refers to eyes that continue to have iridotrabecular contact even after iridotomy, with occludable angles on gonioscopy and normal IOP.[25] (Figure17.1a, b). Eyes which show a high IOP with PIC are categorised as plateau iris syndrome (PIS).

Plateau iris occurs because of anteriorly-rotated ciliary processes, which pushes the peripheral iris to occlude the angle and can be seen in upto 30% of eyes in atleast 1 quadrant among PACG patients in India.

PIC may be either

- Complete closure to the level of Schwalbe's line, resulting in high IOP; or
- Incomplete closure to a lower level on the trabecular meshwork so that IOP does not rise much, but peripheral anterior synechiae (PAS) may develop over time.

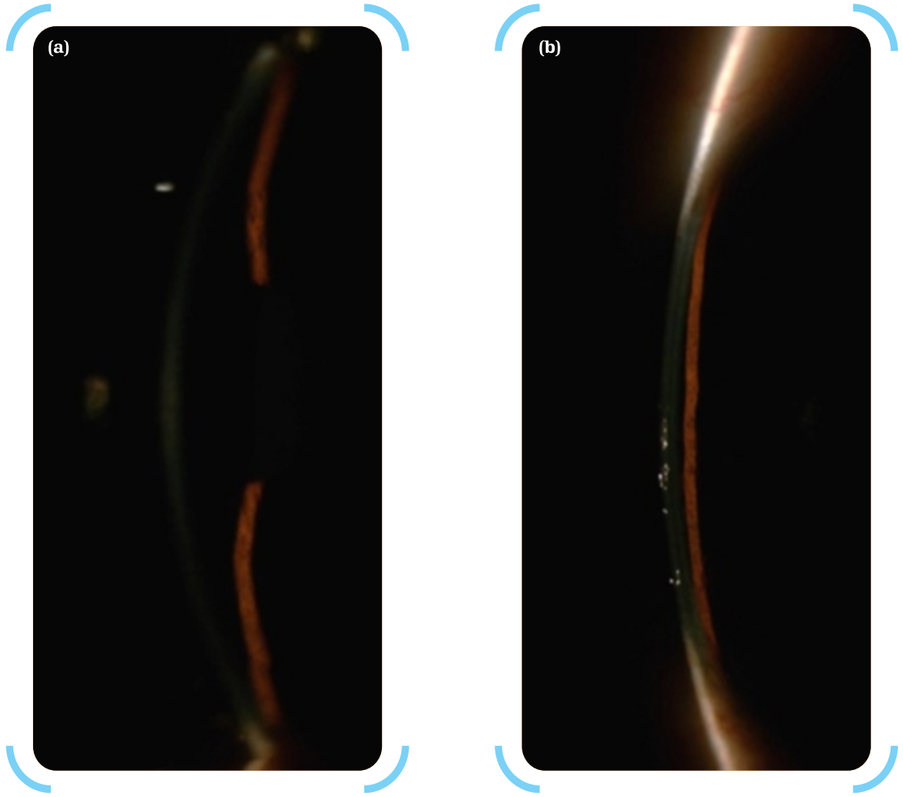


Figure 17.1(a)- Central AC showing normal depth; (b) - Peripheral AC shallow

Diagnosis of Plateau iris configuration and syndrome

Gonioscopy

On indentation gonioscopy, a characteristic double hump sign or sine wave sign is observed where the peripheral hump of the iris is caused by ciliary body propping up the iris root and the central hump represents the iris resting over the surface of the lens. The double hump sign is a soft sign and has not been validated. Anteriorly positioned ciliary body restricts mobility of iris, making it difficult to open on indentation as compared to primary angle closure (Figures 17.2a and 17.2b). Flat anterior iris insertion on ciliary body is common.[80]

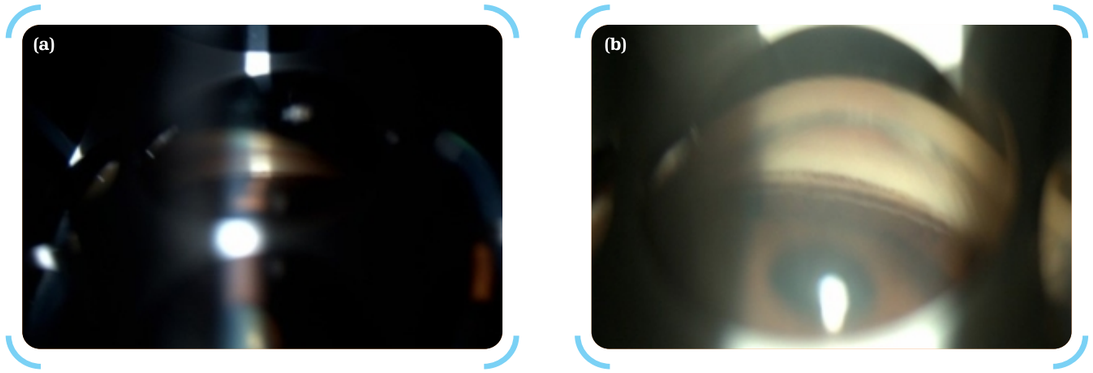


Figure17.2(a) - Gonioscopy with 4 mirror without indentation showing no angle structures; (b) - With indentation showing open angles up to scleral spur with double hump sign

Ultrasound Biomicroscopy (UBM)

UBM offers an objective way of assessing the relationship of the peripheral iris to the ciliary processes and plays a fundamental role in the diagnosis of PIC/PIS. The characteristic features seen are larger or anteriorly-rotated ciliary processes and the absence of ciliary sulcus (Figures 17.3 and 17.4). It can also identify ciliary body (CB) cysts and differentiate plateau iris from pseudo plateau iris. Combining gonioscopy and UBM appears to be more helpful in diagnosing plateau iris.[81]

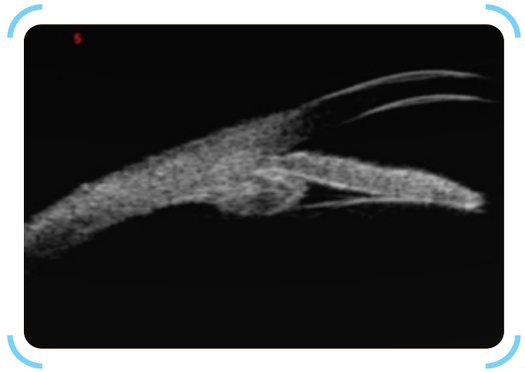


Figure 17.3 - UBM showing anteriorly rotated ciliary body in plateau iris with absent ciliary sulcus

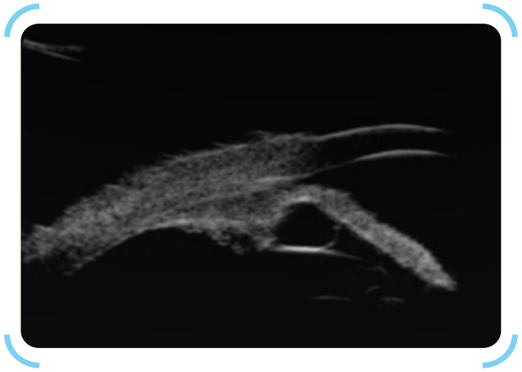


Figure 17.4 - UBM showing narrowing of the angle due to CB cyst - pseudo-plateau iris

Management

Peripheral iridotomy

A peripheral iridotomy (PI) (Figures 17.5a and b) must be performed as the initial management as pupillary block component may coexist. Gonioscopy is mandatory to evaluate residual angle closure persisting after PI. Iridotomy helps to diagnose PIS which is recurrent angle-closure after iridotomy. These patients must be regularly followed-up to look for IOP spike and progression of angle narrowing.

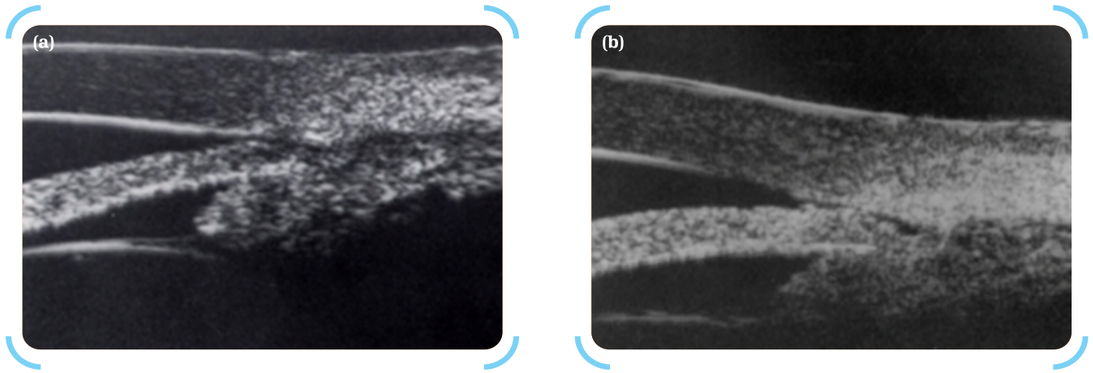


Figure 17.5(a) -Pre YAG PI UBM showing plateau iris; (b) - Post YAG PI UBM showing minimal angle opening

Argon Laser Peripheral Iridoplasty (ALPI)

Iridoplasty is considered in PIS patients who have uncontrolled IOP after PI. It involves applying low power burns of large size in the peripheral iris, for long duration, causing the iris to shrink and pull away from the angle, thereby effectively eliminating appositional residual closure. Ritch et al[82] proposed that ALPI was highly effective in keeping the angles open in approximately 87% of plateau iris angles at 6 years and few required retreatment. (Figure 17.6)

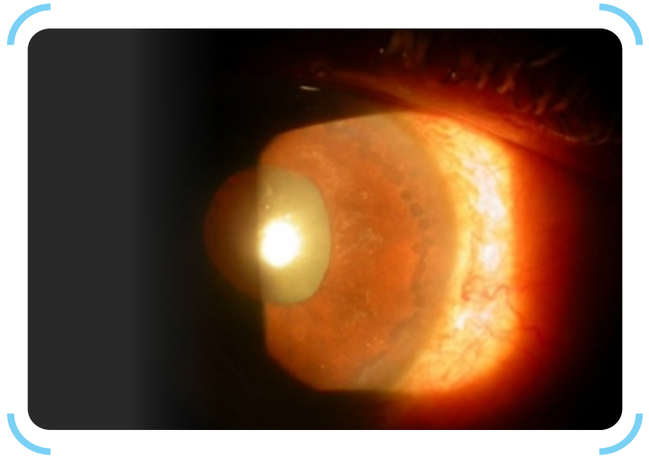


Figure 17.6 - Contraction burns in the peripheral iris seen after ALPI

Pilocarpine

Pilocarpine 1-2%, 2-4 times a day can be considered to minimise pupillary dilatation in PIS patients who refuse ALPI. They act by causing pupillary constriction, mechanically pulling the iris away from the trabecular meshwork, opening the angle and prevent formation of PAS.

Lens extraction

Phacoemulsification alone has also been shown to be successful in the treatment of plateau iris compared to ALPI in a recent study by Peterson et al.[83] Removal of the crystalline lens combined with goniosynechiolysis may also help when the angle closure continues despite iridoplasty.[7] However, iridociliary apposition and narrow angle can persist even after cataract extraction.

Combined phacoemulsification and trabeculectomy

In patients with uncontrolled glaucoma or with advanced glaucomatous damage, a combined cataract surgery and trabeculectomy must be considered. It must be remembered that these eyes are at high risk of aqueous misdirection and appropriate care has to be taken to avoid it.

18 How to diagnose and manage Nanophthalmos?

Nanophthalmos is a rare congenital high hyperopia with short axial length (<20.5 mm i.e. <2 standard deviation from the population mean) and a small eye with narrow palpebral fissure and deep set eyes Figures 18.1a and b). It is characterised by a small/ normal corneal diameter (Figure 18.2), a shallow anterior chamber, narrow angles, thick convex iris, high lens to eye volume ratio (Figure 18.3a and b), and thick sclera or retino-choroido-scleral (RCS) complex (Figure 18.4). There may be family history of blindness from angle-closure glaucoma or history of prior surgical complications in the fellow eye.

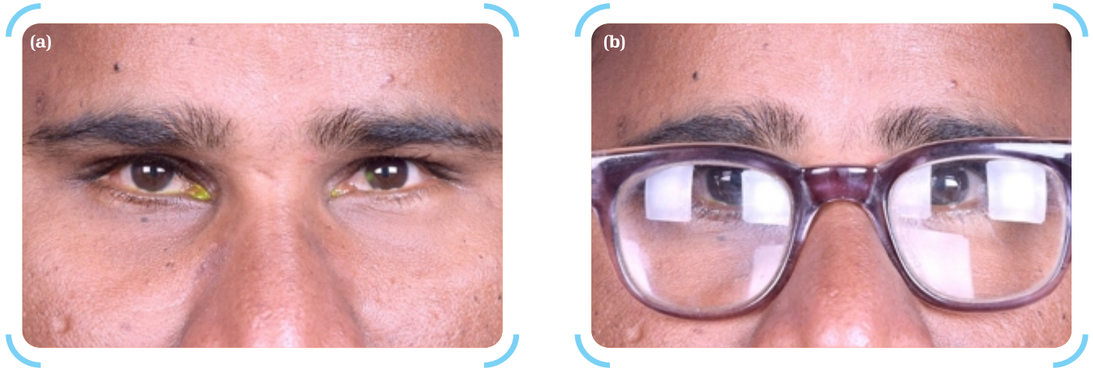


Figure 18.1(a)- Small eyes with narrow palpebral apertures in Nanophthalmos; (b)- High hyperopic glasses in nanophthalmos

These eyes have a predisposition to pupillary block and angle-closure disease in addition to predisposition to massive uveal effusion[84] due to sudden decompression of the globe, which may lead to secondary retinal detachment, intraocular haemorrhage, malignant glaucoma and loss of vision intra- or post-operatively after any intraocular surgery.

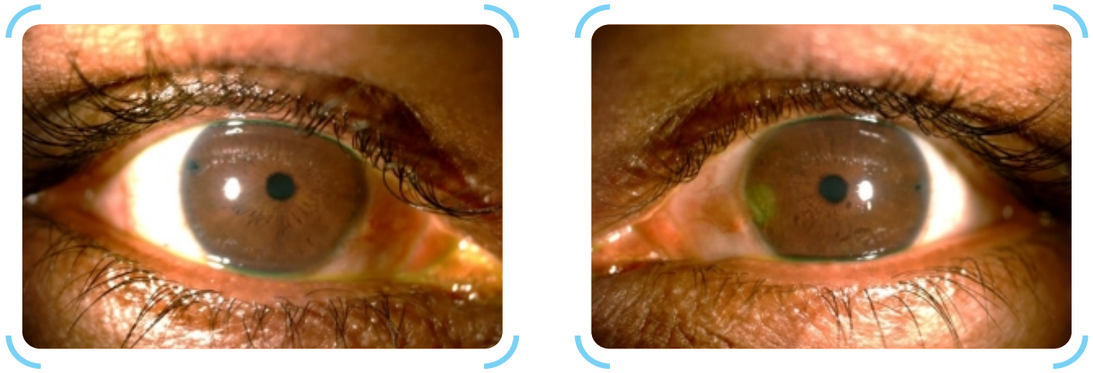


Figure 18.2-Small corneal diameters with patent PI in nanophthalmos

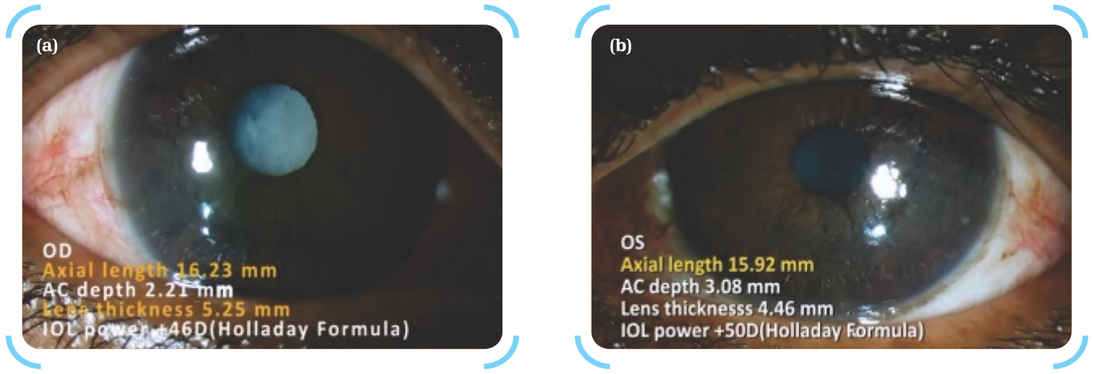
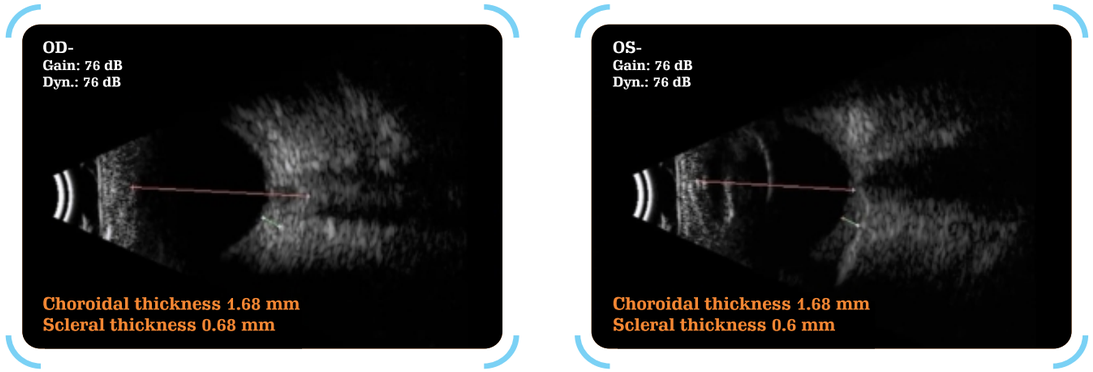


Figure 18.3(a)- Total cataract in a patient of Nanophthalmos; b- Shallow AC in a very small eye with high IOL power.



Parameter	OD	OS
Retino-choroidal thickness in mm	1.68	1.64
Scleral thickness in mm	0.68	0.6

Figure 18.4- High retinochoroidal thickness in nanophthalmos

Management of Nanophthalmos

Nanophthalmos should be managed at a tertiary care centre by glaucoma specialists, where the facility for retina surgery is available.

Treatment of elevated IOP in Nanophthalmos

Response to medical treatment is usually poor once the IOP is elevated. Miotics may worsen the condition by relaxing the lens zonules, further shallowing the anterior chamber. If angles open after laser iridotomy, cataract surgery alone with adequate preoperative and intraoperative precautions may help. If there is synechial angle closure with uncontrolled IOP, trabeculectomy with sclerotomies and releasable sutures can help.

Laser PI considerations in Nanophthalmos

Laser PI should be done early in cases of nanophthalmos due to very narrow angles with high risk of angle closure. Argon laser application prior to YAG PI thins the iris by shrinking the tissue and deepens the peripheral anterior chamber.

Cataract surgery in Nanophthalmos

Strategies to decrease IOP and, thus, reduce positive vitreous pressure:

1. **Pre-operative precautions**
   - Peripheral iridotomy (if occludable angle/ angle closure)
   - Preoperative topical steroids (in case of previous uveal effusion)
   - Oral Acetazolamide
   - Intravenous mannitol
2. **Surgery under general anesthesia (GA) or minimal local anesthesia with good ocular massage**

   GA is preferred
   - To avoid retrobulbar anesthesia induced increased intraorbital pressure
   - Decrease patient related anxiety
   - Hypotensive anesthesia possible under GA
3. **Intra-operative precautions**
   - Intra-operative use of the anterior chamber maintainer to avoid fluctuations in IOP
   - Intraocular injection of dispersive visco-elastic before withdrawal of any instrument
   - 1 or 2 prophylactic sclerotomy in the inferior quadrant
   - Adjunctive pars plana vitrectomy when the depth of the anterior chamber is very shallow
   - If choroidal effusion develops, it should be drained through the sclerotomies

In a randomized controlled trial done by Rajendrababu et al.[85], comparing cataract surgery with and without prophylactic posterior sclerotomy, they found higher complication rate in eyes without prophylactic sclerotomy compared to those with sclerotomy, in eyes that underwent small incision cataract surgery compared to phacoemulsification and in eyes with higher preoperative IOP (Figures 18.5 and 18.6).

Trabeculectomy

Combined procedure is usually not preferred unless it is absolutely indicated.

Major modifications in trabeculectomy procedure are as follows:

1. Preplaced scleral flap sutures
2. Avoiding intraoperative and postoperative hypotony
3. Tight suture closure
4. Keep anterior chamber formed
5. Performing inferior sclerotomies at one or two sites in temporo-inferior and naso-inferior quadrants

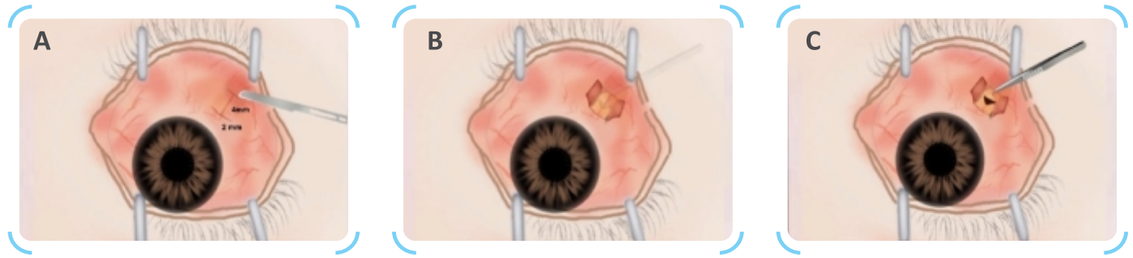


Figure 18.5: Schematic diagram demonstrating technique of prophylactic sclerotomy: 4 mm long scleral incisions starting 2 mm from limbus (A), creation of trap-door scleral flaps (B) and V-shaped sclerotomy (C)

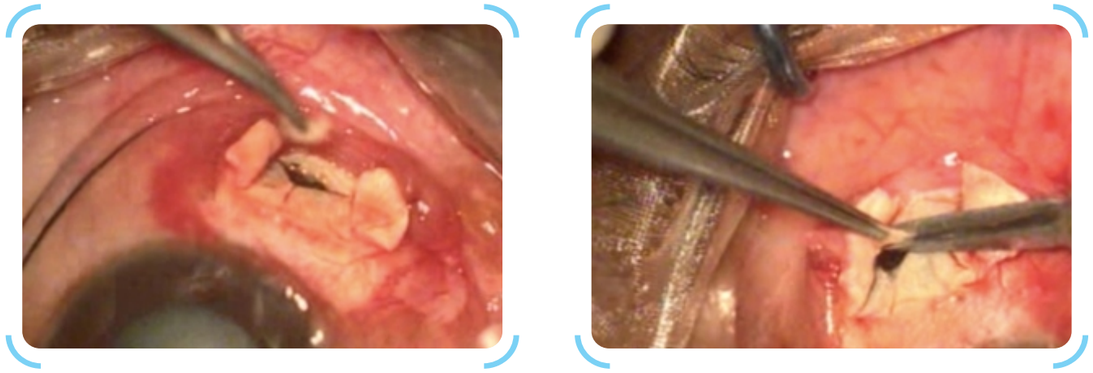


Figure 18.6- Technique of sclerotomy 4 mm behind the limbus in Nanophthalmos.

Singh et al.[86] reported poor IOP control and poor visual outcomes following glaucoma filtration surgery in eyes with nanophthalmos (60% of the 15 patients who had filtration surgery for glaucoma failed to achieve control and 86.6% suffered visual loss). Yalvac et al reported the cumulative probability of success of trabeculectomy was 85% at 1 year and 47% at 5 years after surgery. Visual acuity decreased in 13 (65%) patients but no eye lost vision.[87] There was 50% (10 eyes) incidence of choroidal effusion despite prophylactic sclerotomy.

A thorough preoperative workup to measure the axial length, scleral thickness and retino-choroidal thickness would help to plan appropriate prophylactic interventions and procedure.

19 Conclusion

Primary angle closure disease is an important public health problem in India which leads to irreversible blindness. It is important to increase public awareness about this disease, get family screening[88] done for any detected case, perform gonioscopy[89] in every case at diagnosis so that angle closure is not treated as an open angle, perform laser iridotomy in high risk eyes , consider early lens extraction in appropriate cases where the angle is partially open and that has an early optic nerve damage, perform filtering surgery in eyes with advanced damage and follow-up these patients life-long on a regular basis to prevent the blindness caused by this disease. In patients who present with severe degrees of visual impairment, appropriate strategies for visual rehabilitation should be adopted, certification done for degree of visual disability, counselling done for community based rehabilitation and information given regarding help available from Ministry of Social Justice and Empowerment, Govt. of India[90] (www. socialjustice.nic.in) for differently abled persons.

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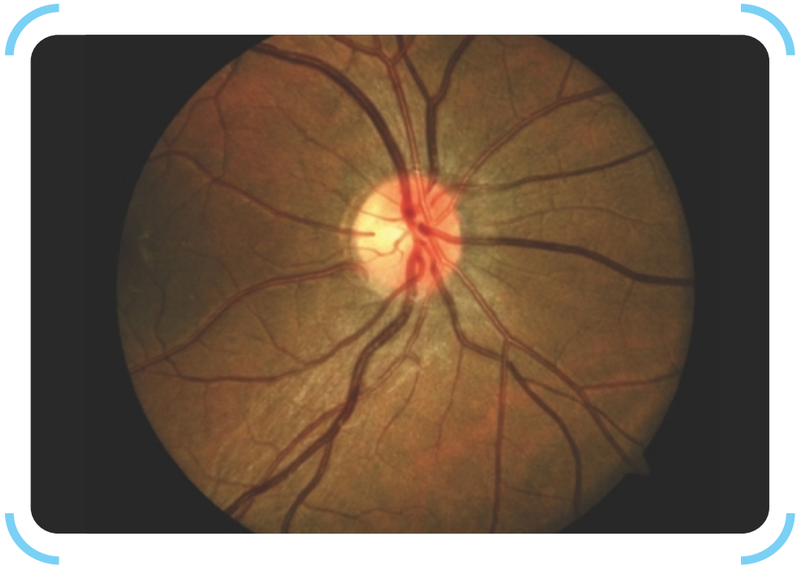
MANAGEMENT ALGORITHMS

Primary Angle Closure Disease
(PACS, PAC, PACG, APAC)

Primary Angle Closure Suspect (PACS)



Occludable angles – Pigmented part of Trabecular meshwork not visible in the primary position in more than 180 degress but angle opens on manipulation. No synechiae, healthy optic disc.

Primary Angle Closure Suspect

Observe once in 2 years*

Eyes at a high risk for developing an acute attack or progression of disease

- Fellow eyes of patients with either Primary Angle Closure (PAC) or PACG / one eyed patients
- Need for repeated dilatation due to posterior segment pathology (e.g. diabetes mellitus)
- Use of systemic medications which may provoke pupillary block
- Patients with symptoms suggestive of prior acute or intermittent angle closure
- Family history of PACG
- Patients whose health/ occupation/ geographical location makes it difficult to access immediate ophthalmic care in the event of an acute attack of angle closure
- Patients who cannot come for regular follow-up
- Patients who understand the risk of progression and want a laser iridotomy

All patients should understand the risk and give consent for laser iridotomy

If visually significant cataract

Clear corneal phacoemulsification**

Nd:YAG Laser Iridotomy
Follow-up once a year

If Gonioscopy reveals persistent angle closure - rule out plateau iris

* The actual frequency of follow-up depends on associated ocular and systemic risk factors and may be every 6 months or annual in patients who seem to be at a higher risk.

**Manual SICS may be used for lens extraction in all spectra of PACD, temporal approach is preferred so as to leave the superonasal conjunctiva intact for a future trabeculectomy.

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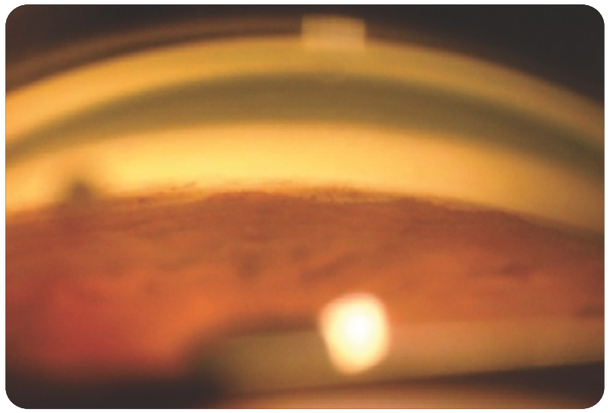
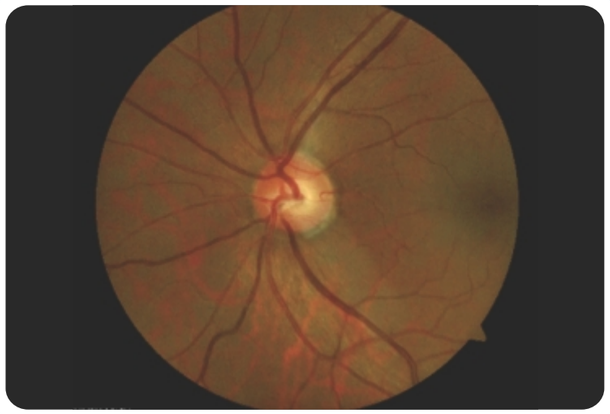
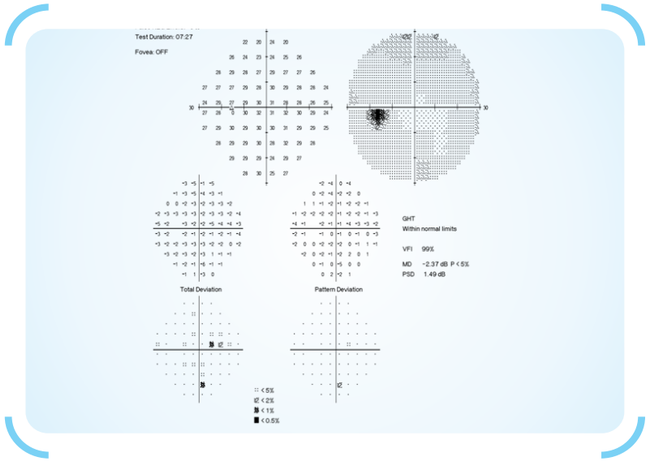
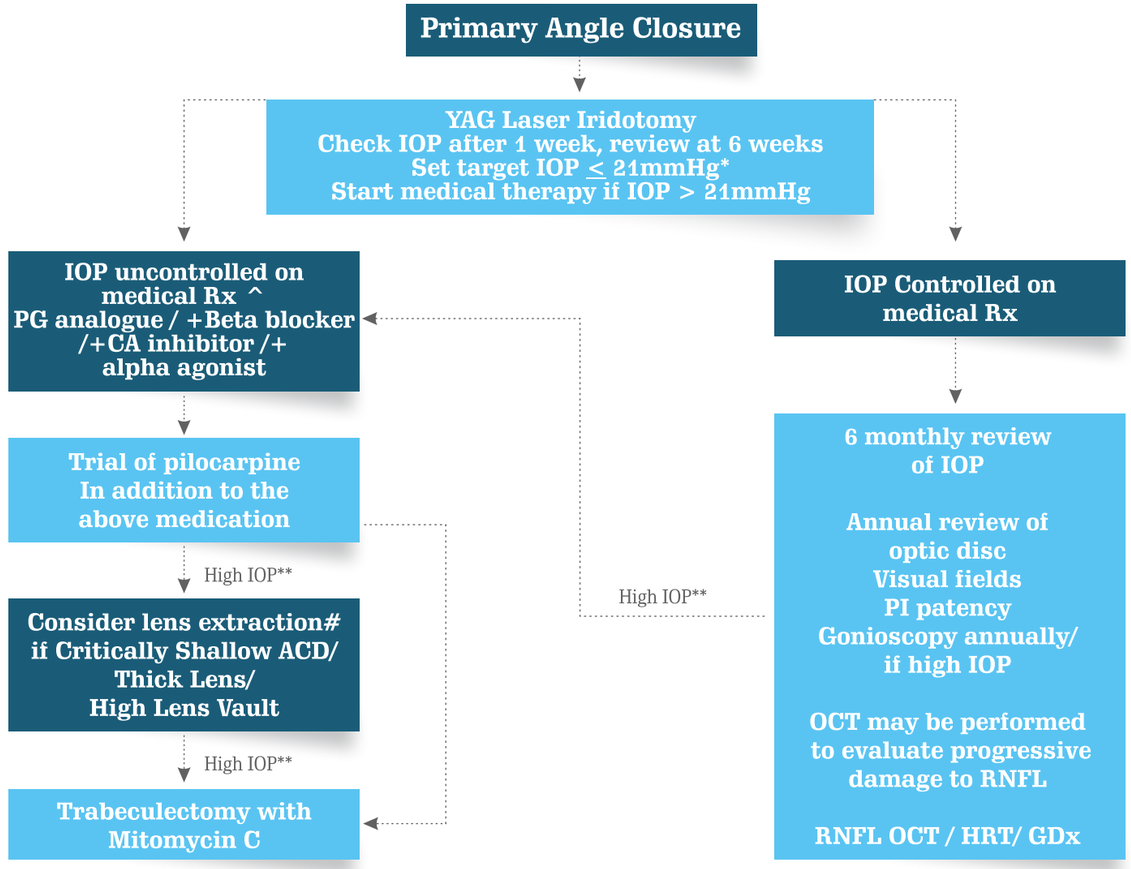
Primary Angle Closure (PAC)







Peripheral anterior synechiae on Gonioscopy. No disc or field changes.



*The Target IOP in PAC is arbitrarily taken as 21 mmHg as it is the higher range of normal IOP in our population. It may be lower or higher depending upon the corneal thickness, risk factors and progression on follow-up.

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** High IOP = IOP more than the target.

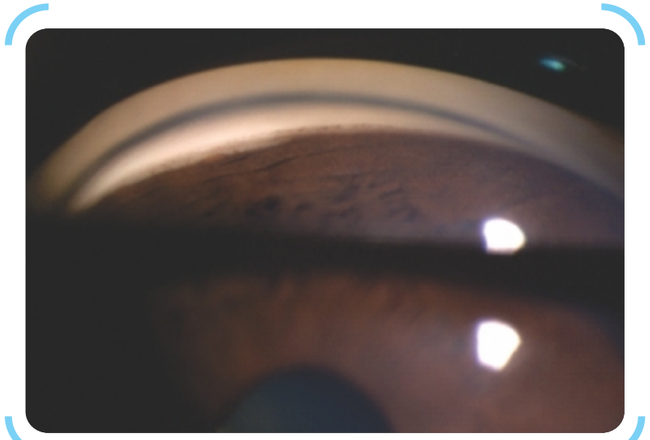
^ If IOP is high after laser PI and the angle remains occludable, a careful gonioscopy should be done to rule out plateau iris syndrome - confirmed by UBM and such cases respond well to 1 - 2% pilocarpine eye drops (BD).

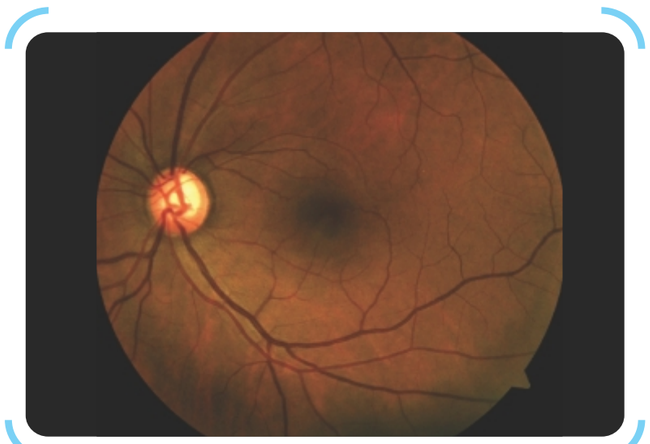
#Consider lens extraction in the presence of critically shallow anterior chamber with a thick lens and a high lens vault. There are no objective cut-off values which can be used for this purpose based on evidence from the published literature. However as a consensus from the AIOS expert group meeting - eyes with ACD < 2 mm, LT > 5 mm and LV > 1 mm may have a higher risk of complication post-trabeculectomy, and lens extraction may be an option in such eyes. It is important to understand that lens extraction in these eyes is technically challenging because of the crowded anterior segment and prone to complications. Surgery should be performed after informed consent by an expert at a hospital, which has the facility for glaucoma surgery, corneal endothelial transplantation and vitreo-retinal surgery, to take care of any complications arising from this procedure.

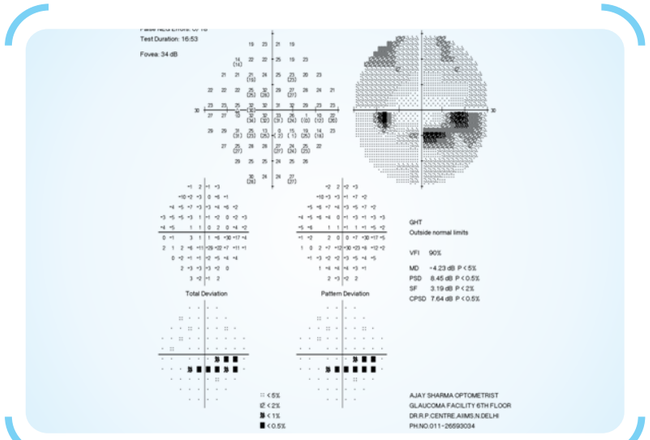
References:
Dada T, Rathi A, Angmo D, Agarwal T, Vanathi M, Khokhar SK, Vajpayee RB. Clinical outcomes of clear lens extraction in eyes with primary angle closure. J Cataract Refract Surg. 2015 Jul;41(7):1470-7.

Sihota R. An Indian perspective on primary angle closure and glaucoma. Indian J Ophthalmol. 2011 Jan; 59; Supplement S 76-81

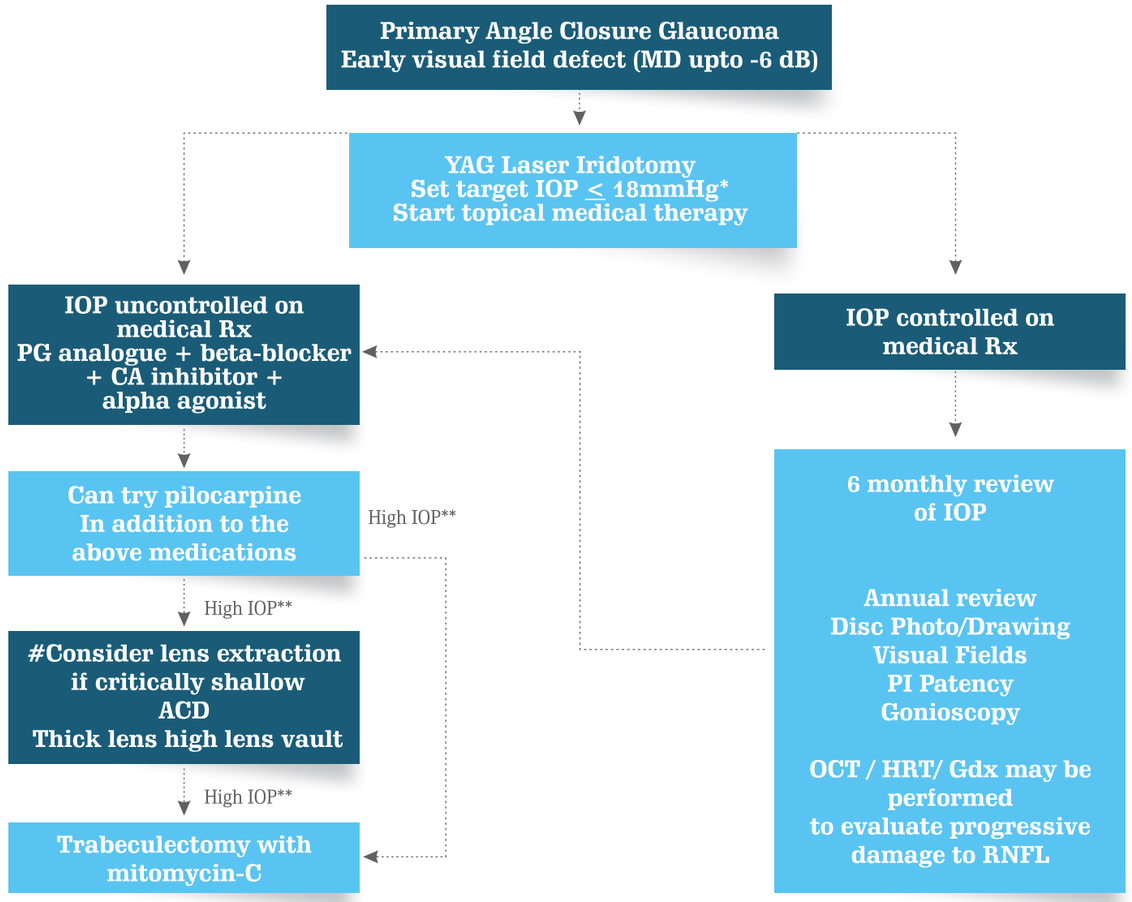
Primary Angle Closure Glaucoma (PACG)







PAS on gonioscopy with superior notching of NRR and early inferior visual field defect



*The target IOP's for all categories of PACG (early, moderate, advanced) are a suggestions and could be higher or lower based on the IOP at which the damage occurred, CCT, age, life expectancy, associated systemic disease, optic nerve head status and visual field damage (slow versus fast progression).

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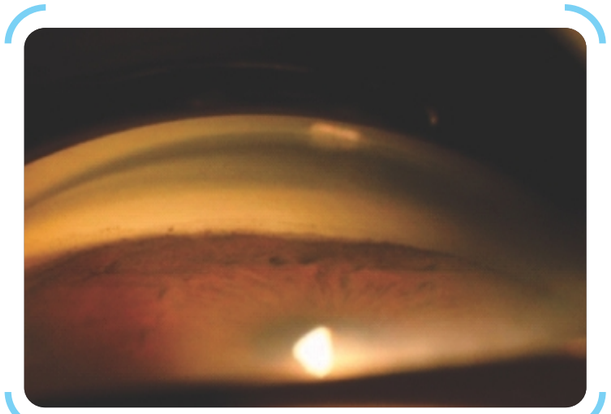
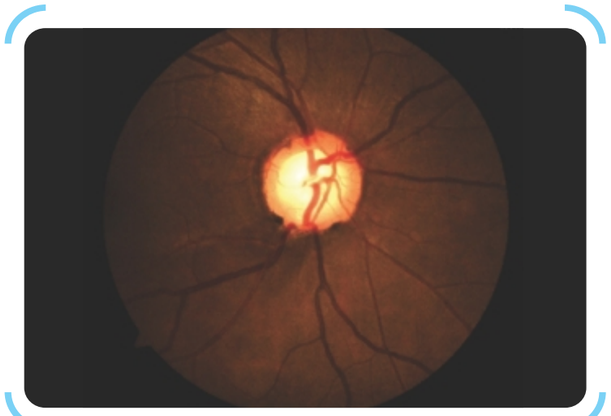
Consider lens extraction in the presence of critically shallow anterior chamber with a thick lens and a high lens vault. There are no objective cut off values which can be used for this purpose based on evidence from published literature. However, as a consensus from the AIOS expert group meeting - eyes with ACD < 2 mm, LT > 5 mm and LV > 1 mm may have a higher risk of complication post-trabeculectomy, and lens extraction may be an option in such eyes. It is important to understand that lens extraction in these eyes is technically challenging because of the crowded anterior segment and prone to complications. Surgery should be performed after informed consent by an expert at a hospital, which has the facility for glaucoma surgery, corneal endothelial transplantation and vitreo-retinal surgery, to take care of any complications arising from this procedure.

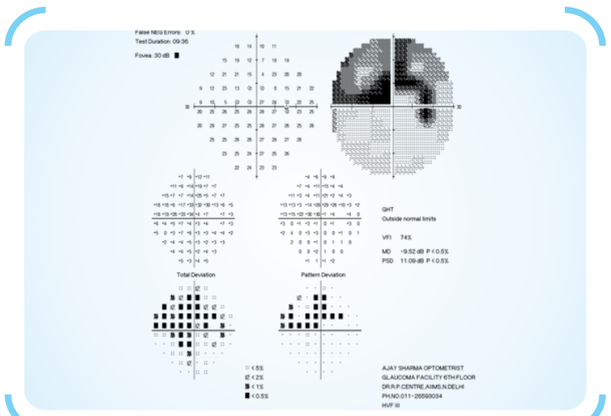
References:
Thomas R, Walland MJ. Management algorithms for primary angle closure disease. Clin Exp Ophthalmol. 2013 Apr;41(3):282-92.

Gazzard G, Foster PJ, Devereux JG, Oen F, Chew P, Khaw PT, Seah S. Intraocular pressure and visual field loss in primary angle closure and primary open angle glaucomas. Br J Ophthalmol. 2003 Jun;87(6):720-5

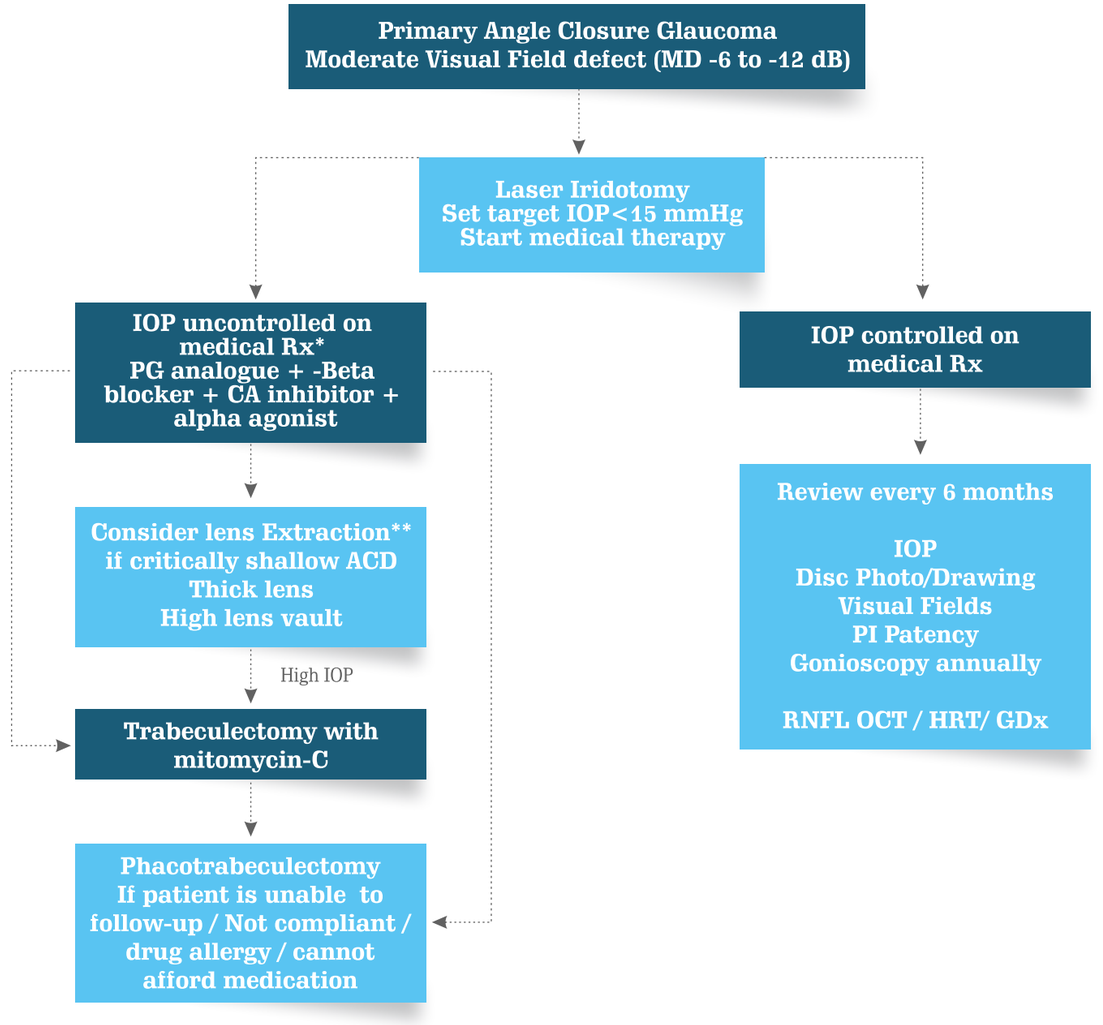
Primary Angle Closure Glaucoma (PACG)







PACG with PAS on gonioscopy, inferior notching of NRR and moderate visual field defect



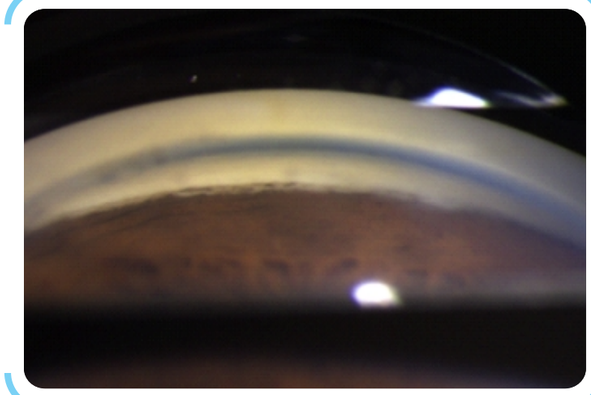
*In eyes with a persistent irido-trabecular contact post-laser iridotomy, plateau iris syndrome should be ruled out. Gonioscopy reveals a typical double hump/sine wave configuration and UBM confirms the diagnosis. Such cases respond well to low dose pilocarpine therapy (1-2 % BD). Can be given as combination with beta blocker.

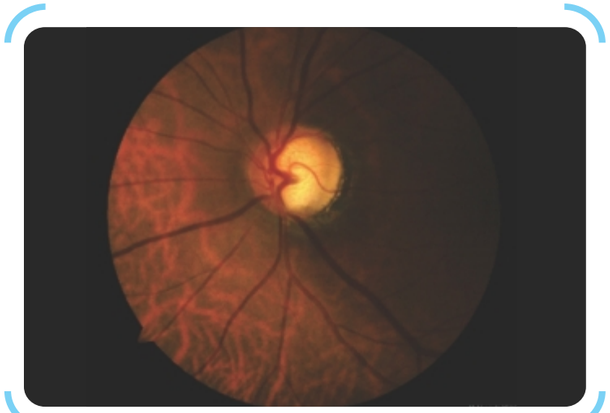
**See foot note in early PACG for Lens Extraction

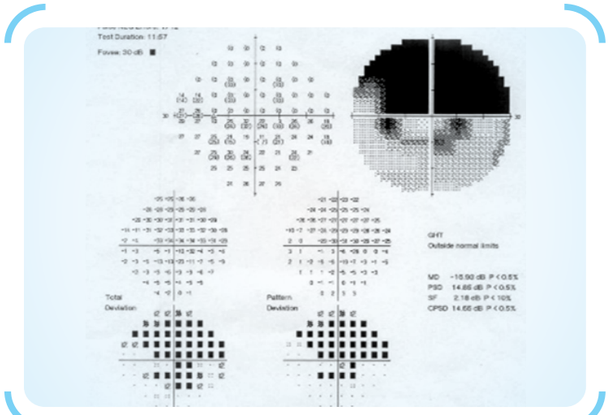
References:
Chen PP, Lin SC, Junk AK, Radhakrishnan S, Singh K, Chen TC. The Effect of Phacoemulsification on Intraocular Pressure in Glaucoma Patients: A Report by the American Academy of Ophthalmology. Ophthalmology. 2015 Jul; 122(7):1294-307

Thomas R, Walland M, Thomas A, Mengersen K. Lowering of Intraocular Pressure Following Phacoemulsification in Primary Open Angle and Angle Closure Glaucoma: A Bayesian Analysis. Asia Pacific Journal of Ophthalmology 2016; 5:79-84.

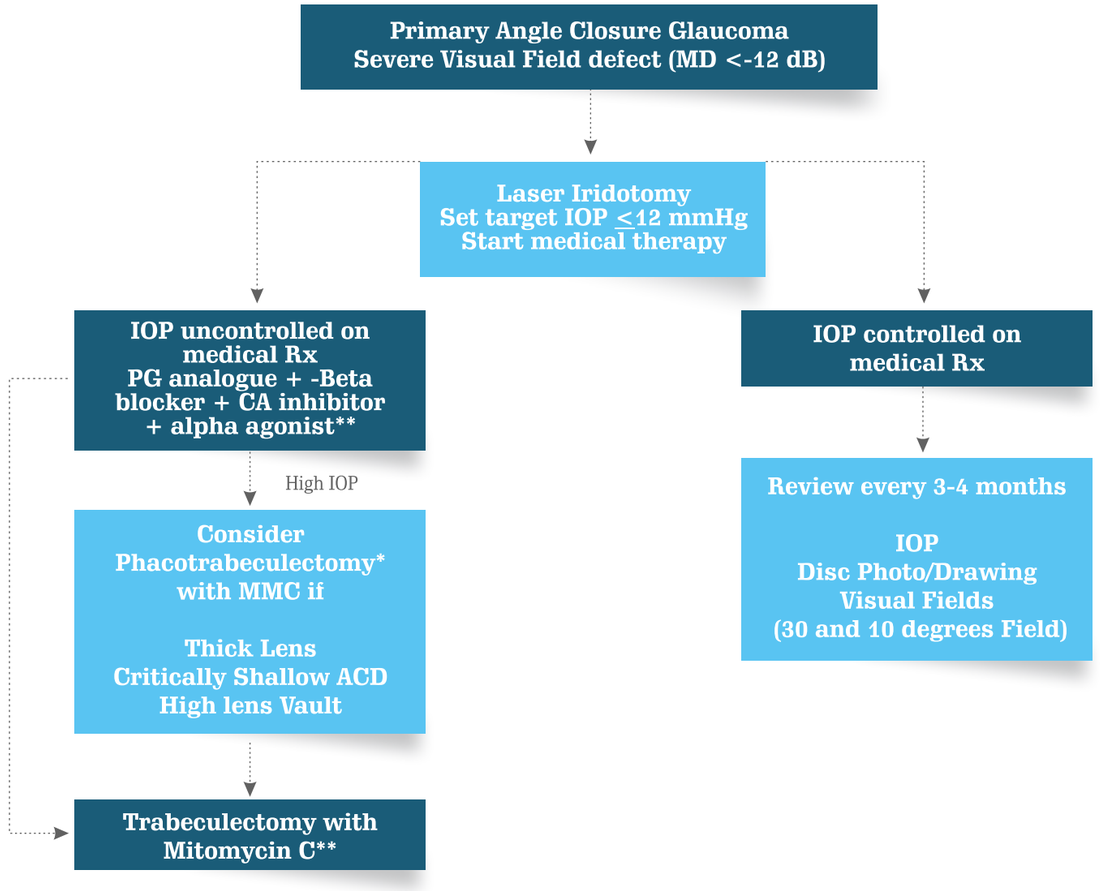
Primary Angle Closure Glaucoma (PACG)







Advanced PACG with extensive PAS, glaucomatous optic neuropathy and visual field defect involving central 10 degrees



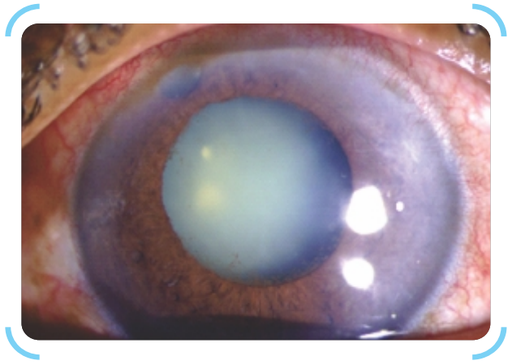
* Consider phaco-trabeculectomy in the presence of critically shallow anterior chamber with a thick lens and a high lens vault. There are no objective cut-off values which can be used for this purpose based on evidence from published literature. However as a consensus from the AIOS expert meeting - eyes with ACD < 2 mm, LT > 5 mm and LV > 1 mm may have a higher risk of complication post-trabeculectomy and combined surgery should be considered. Surgery in such cases is technically challenging should be performed after informed consent by an expert at a hospital, which has the facility for glaucoma surgery, corneal endothelial transplantation and vitreo-retinal surgery, to take care of any complications arising from this procedure.

** In eyes with a total glaucomatous optic neuropathy and absent light perception, 270 degrees of cyclophotocoagulation / cyclo-cryotherapy can be performed to alleviate symptoms of pain.

References:
Masis M, Mineault PJ, Phan E, Lin SC. The role of phacoemulsification in glaucoma therapy: A systematic review and meta-analysis. SurvOphthalmol. 2017 Sep 6.

Chan PP, Li EY, Tsoi KKF, Kwong YY, Tham CC. Cost-effectiveness ofPhacoemulsification Versus Combined Phacotrabeculectomy for Treating Primary Angle Closure Glaucoma. J Glaucoma. 2017 Oct;26(10):911-922

Primary Angle Closure Glaucoma (PACG) with age-related cataract



Primary Angle Closure Glaucoma with Visually Significant Cataract

Early visual field defect* or VCDR 0.6 - 0.7* with RNFL defect with thinning of NRR	Moderate visual field defect or VCDR 0.7 - 0.9* with loss of NRR in 1 vertical quadrant	Severe visual field defect or VCDR 0.9 to near total* cupping with loss of NRR in both superior and inferior quadrants
Phacoemulsification	Phacoemulsification	Phaco- Trabeculectomy
	2nd stage Trabeculectomy, if IOP is not controlled	

If the visual field is not possible, ascertain severity of disease by optic nerve head evaluation and amount of synechial angle closure. Eyes with more than 270 degrees of synechial closure should be treated as severe disease and filtering surgery must be a component of therapy.

Consider Phacotrabeculectomy with Mitomycin C* (0.1-0.2 mg/ml for 1-2 minutes) if :

1. Patient is unable to follow-up
2. Not adherent with medical therapy
3. Drug allergy
4. Socioeconomic indications
5. Wants to be off medications and understands risks of surgery
6. IOP not controlled despite maximal topical medications (3/4)

* Approx. vertical cup:disc diameter ratio for average size discs

References:

Sihota R, Angmo D, Chandra A et al. Evaluating the long-term efficacy of short-duration 0.1 mg/ml and 0.2 mg/ml MMC in primary trabeculectomy. Graefes Arch Clin Exp Ophthalmol. 2015 jul;253(7):1153-9.

Tham CC, Kwong YY, Leung DY et al. Phacoemulsification versus combined phacotrabeculectomy in medically uncontrolled chronic angle closure glaucoma with cataracts. Ophthalmology. 2009;116(4):725-31.

Acute Primary Angle Closure (APAC)

Immediate Treatment - Lower IOP medically
I.V Mannitol (20%) 1-2g/kgBW / Tablet Acetazolamide 5-15 mg/kgBW tid/ BOTH
Syrup Glycerol (50%) 1g/kgBW tid
Eye drops Timolol 0.5% BD, Brimonidine 0.2% BD,
Eye drops Pilocarpine 2-4% QID
Eyedrops 1 % Prednisolone QID

Rule out secondary cause
Phacomorphic glaucoma
Ciliary effusion
Drugs/Inflammation

Treat pathology

LPI not successful**
- **Thick boggy iris**
- **Inflammation**

Continue Medical management
·Increase topical steroids
·Try again after 24 hours
·ALPI if possible

LPI not possible

Surgical iridectomy

Schedule LPI in fellow eye

Acute Primary Angle Closure
- **Usually unilateral attack**
- **Peripheral AC usually flat; Central shallow**
- **Other eye gonioscopy occludable angles**

IOP uncontrolled

IOP control

Both eyes LPI urgently

-Compression centrally with an Indentation gonioscope to displace aqueous and open angle
-Attempt ALPI if possible

LPI successful; Patent iridotomy

Assess IOP

IOP Controlled on 2/3 topical medications

Follow-up
1 week, 1 month, 3 months, every 6 months
- **Gonioscopy to assess extent of angle opening**
- **Optic disc**
- **Visual Field**
- **Patency of PI**

IOP uncontrolled on topical medications
- Ascertain patency of LPI
- Gonioscopy for synechial angle closure
- Plan surgical treatment

If 180 degrees angle opens on indentation gonioscopy:
Can consider lens extraction

If < 180 degrees angle opens on indentation:
Phaco–Trabeculectomy*
4-6 weeks after an acute attack

*High risk of complications if trabeculectomy is alone done for Acute Attack of Angle Closure (AAAC)

** If IOP is controlled on medications, LPI can be done after inflammation has subsided.

Reference:

Trikha S, Perera SA, Husain R, Aung T. The role of lens extraction in thecurrent management of primary angle-closure glaucoma. CurrOpinOphthalmol. 2015 Mar;26(2):128-34.

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In POAG and OHT patients

Eye drops

optipres

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The only adjunct with OD dosing

- **Triple mechanism of action[1]**
- **18% to 23% IOP reduction as an adjunct[2]**
- **24-hours IOP lowering efficacy[3]**
- **Increases OBF and RGC survival[4]**
- **Convenient OD dosing[1]**

POAG: Primary open angle glaucoma; OHT: Ocular hypertension; IOP: Intraocular pressure; OD: Once daily; OBF: Ocular blood flow; RGC: Retinal ganglion cell

References:

1. Batra M, Gupta S, Nair AB, Dhanawat M, Sandal S, Morsy MA. Netarsudil: A new ophthalmic drug in the treatment of chronic primary open angle glaucoma and ocular hypertension. Eur J Ophthalmol. 2021 Sep;31(5):2237-2244.
2. Zaman F, Gieser SC, Schwartz GF, Swan C, Williams JM. A multicenter, open-label study of netarsudil for the reduction of elevated intraocular pressure in patients with open-angle glaucoma or ocular hypertension in a real-world setting. Curr Med Res Opin. 2021 Jun;37(6):1011-1020.
3. Peace JH, McKee HJ, Kopczynski CC. A Randomized, Phase 2 Study of 24-h Efficacy and Tolerability of Netarsudil in Ocular Hypertension and Open-Angle Glaucoma. Ophthalmol Ther. 2021 Mar;10(1):89-100.
4. Saha BC, Kumari R, Kushumesh R, Ambasta A, Sinha BP. Status of Rho kinase inhibitors in glaucoma therapeutics-an overview. Int Ophthalmol. 2022 Jan;42(1):281-294.

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