OPHTHALMOLOGY CLINICS-I

for Postgraduates

History taking
Bedside examination
Long and short cases
Viva voce

Editors
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2nd Edition

JAYPEE

Contents

CHAPTER

3

Glaucoma

LONG CASES

PRIMARY ANGLE CLOSURE GLAUCOMA

Vaishali Ghanshyam Rai, Talvir Sidhu, Dewang Angmo

INTRODUCTION

Primary angle closure glaucoma (PACG) is usually allotted in practical examination as a long case. Primary angle closure (PAC) is appositional or synechial closure of the anterior chamber angle caused by pupillary block. The angle closure may or may not be associated with elevated intraocular pressure (IOP) or glaucomatous optic neuropathy and may occur in either an acute or chronic form. This entity does not include secondary forms of angle closure induced by other causes, for example, subluxed lens.

HISTORY

Chief Complaints

A case of PACG can present in following ways:

- Majority of patients with ACG are asymptomatic
- Blurred vision or smoke-filled room
- Some patients present acutely with color halos around lights due to corneal edema, aching eye, or brow pain, and/or eye redness
- Patients with acute angle closure attack present usually with unilateral diminution of vision with redness in the eye with severe eye pain associated with ipsilateral headache with nausea and vomiting.

History of Present Illness

Following points must be enquired:

- History of symptoms precipitated by watching television, darkened room, reading, and pharmacological mydriasis
- Brief history of patient's previous records to get baseline IOP
- Patient who is already on treatment for glaucoma is important to know how many medications he/she is using and whether this treatment has sufficiently controlled IOP and visual field loss
- Patient's previous visual fields record should be checked to know the progression of glaucoma.

History of Past Illness

Ask for previous use of glasses. Hypermetropic patients are at a higher risk to develop PACG.

History of Systemic Illness

- Diabetic or hypertensive patients who need frequent dilated fundus examination need special attention to rule out PAC or PACG as shallow anterior chamber depth may develop acute angle closer attack on dilation of the pupil.

- It is very important to ask about cardiovascular disease, renal diseases, and bronchial asthma before deciding on antiglaucoma treatment.

Family History

Relatives of PACG patients are at a higher risk for developing glaucoma. The severity and outcome of glaucoma in family members, including history of visual loss from glaucoma is also important.

Drug History

- History of use of ocular and systemic medications
- Known local or systemic intolerance to ocular or systemic medications
- History of drugs that induce angle closure attack needs to be asked. Such drugs include the following:
  - Anticholinergic agents (topical, e.g., atropine, cyclopentolate, and tropicamide; or systemic, e.g., antihistamine, antipsychotic, especially antidepressants, anti-Parkinsonian, atropine, and gastrointestinal spasmolytic drugs)
  - Adrenergic agents (topical, e.g., epinephrine and phenylephrine; or systemic, e.g., vasoconstrictors, central nervous system stimulants, bronchodilators, appetite depressants, and hallucinogenic agents)
- Specific questioning includes asking about the use of topical or systemic medication (e.g., sulfonamides, topiramate, phenothiazines) that may induce angle narrowing and subsequent symptoms that suggest intermittent angle closure attacks should be enquired.

Surgical History

History of previous ocular surgery such as trabeculectomy or any eye laser like iridotomy must be asked for.

EXAMINATION

General examination/specific systemic examination is carried out to look for any contraindication to antiglaucoma medications.

Ocular Examination

- *Eyeball:* Usually looks normal; small eyeball in case of hypermetropia
- *Lids:* Usually normal; in a patient who is already on antiglaucoma medications such as prostaglandin analogs, look for hyperpigmentation of lid margin and long eyelashes.
- *Conjunctiva:*
  - In case of acute angle closure attack, marked circumciliary congestion can be noted.
  - Patients of chronic ACG who are already on antiglaucoma treatment, always look for conjunctival congestion as all antiglaucoma medications can cause some form of conjunctival toxicity. This congestion is more in inferior quadrant.
- *Cornea:* Following signs can be seen—
  - In acute angle closure attack, unilateral epithelial and stromal cornea edema due to raised intraocular pressure (IOP)
  - In chronic cases, Krukenberg spindle (pigment distribution over the inferior corneal endothelium)
- *Anterior chamber:*
  - Anterior chamber is shallow **(Fig. 1)**. The Van Herick technique is useful for estimating the peripheral anterior chamber depth. In PACG cases, peripheral anterior chamber is shallow that can be graded by Van Herick technique (**Flowchart 1**) (discussed later in viva questions). When the peripheral anterior

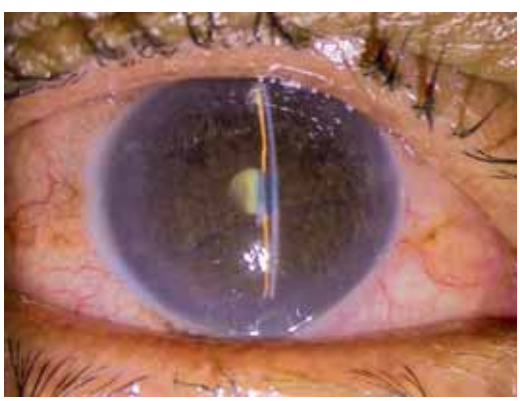
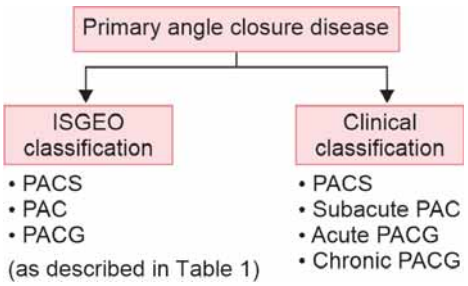


Fig. 1: Shallow anterior chamber.

Flowchart 1: Classification of PACD.



(PAC: primary angle closure; PACD: primary angle closure disease; PACG: primary angle closure glaucoma; PACS: primary angle closure suspect; ISGEO: International Society of Geographical and Epidemiological Ophthalmology)

chamber depth is less than one-fourth of the corneal thickness, the anterior chamber angle may be potentially occludable.
  - Anterior chamber inflammation suggestive of a recent or current attack.
- *Iris:* Following points must be looked for—
  - In acute attack, iris bombe is usually present due to pupillary block.
  - In recent PACG attack, iris whorling (sectoral infarction of the iris sphincter) or patchy iris stromal atrophy is usually present **(Fig. 2)**.
  - Mid-dilated pupil is common in acute or recent PACG attack.
  - Signs of previous angle closure attacks are peripheral anterior synechiae (PAS), segmental iris atrophy **(Fig. 3)**, posterior synechiae, and irregular pupil.
- *Lens:* In previous angle closure attack, look for Glaukomflecken [small gray-white anterior subcapsular or capsular opacities **(Fig. 4)** in the pupillary zone, due to infarction of lens fibers]. Lens thickness might be increased. An intumescent lens can be there in angle closure attack. Vogt's triad: Sectoral iris atrophy, Krukenberg spindle, and Glaucomflecken are characteristic of PACG.
- *Intraocular pressure:*
  - IOP is measured in each eye, preferably using a contact applanation method (typically a Goldmann tonometer) before performing gonioscopy.
  - During an acute attack of PACG, avoid measuring the IOP by Goldmann

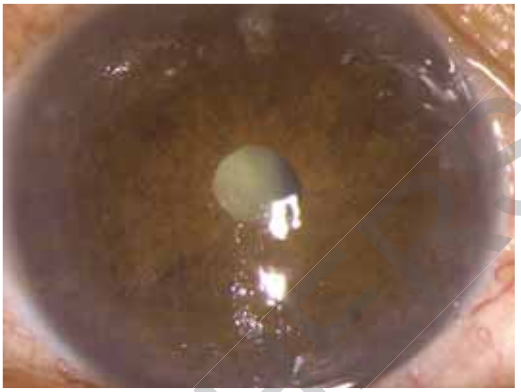


Fig. 2: Patchy iris stromal atrophy.

Fig. 3: Segmental iris atrophy.

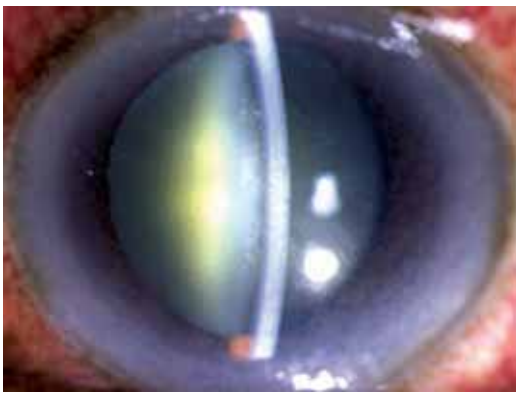


Fig. 4: Glaukomflecken.

applanation tonometer as due to raised IOP, corneal edema is present and it can cause fallacious recording of IOP due to increased corneal thickness along with rupture of bullae.
  - In acute attack, IOP is usually very high (50–100 mm Hg).
  - In chronic PACG, IOP elevation may be intermittent.
- *Gonioscopy:*
  - Gonioscopy of both eyes should be performed on all patients in whom angle closure is suspected.
  - This is best performed using first a two-mirror Gonio lens (e.g., Goldmann) to avoid artifactual distortion of the angle caused by inadvertent pressure on the cornea.
  - This is required to evaluate the angle anatomy, appositional closure, and presence of PAC (PAS) **(Fig. 5)**.
  - Compression (indentation) gonioscopy with a four-mirror or similar lens is particularly helpful to evaluate for appositional closure versus synechial angle closure and for extent of PAS.
  - Various grading systems including Scheie, Shaffer, and Spaeth have been proposed for the recording of gonioscopic findings. These gonioscopic grades provide an index of the likelihood of angle closure.
- *Fundus examination:*
  - For patients with PAC or narrow angle who are not in an acute attack, pupil dilation is contraindicated until iridotomies have been performed.
  - Although a dilated examination may not be advisable in patients with anatomic narrow angles or angle closure, an attempt should be made to evaluate the fundus and optic nerve using the direct ophthalmoscope or biomicroscope with +78D or +90D.
  - In acute attack, optic nerve head may look hyperemic and edematous in early stage, the disc then became pale and glaucomatous cupping can be observed after 9 to 10 days.

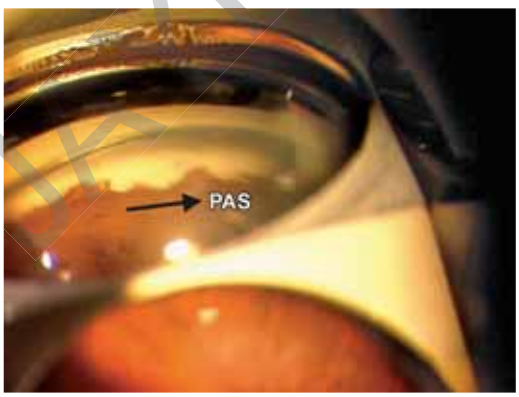


Fig. 5: Gonioscopy shows presence of PAS.

DIFFERENTIAL DIAGNOSIS

As primary narrow angles and PAC tend to be bilateral, the observation of a wide-open angle in the fellow eye suggests a diagnosis other than PAC.

- *Plateau iris syndrome:*
  - The peripheral iris is forced into the angle by anterior rotation of the ciliary body or anteriorly positioned ciliary processes.
  - Angle closure attack may be precipitated after dilatation of pupil even in presence of patent peripheral iridotomy.
- *Neovascular glaucoma:*
  - Neovascularization of iris or angles is a hallmark sign
- Inflammatory causes of angle closure (e.g., posterior synechiae, iris bombe)
- Iridocorneal endothelial (ICE) syndrome
- Ciliary body engorgement or suprachoroidal effusion caused by systemic medications (e.g., topiramate, sulfonamides, phenothiazines)
- Ciliary body engorgement associated with retinal vascular occlusion or scatter (panretinal) photocoagulation
- Anterior suprachoroidal effusions (e.g., congestion, edema, displacement)
- Aqueous misdirection (ciliary block) syndrome after incisional or laser surgery (e.g., following peripheral iridectomy):
  - May be differentiated clinically with acute attack of PACG
  - In aqueous misdirection, both central and peripheral shallowing of anterior chamber are seen. (Must rule out choroidal detachment or suprachoroidal

hemorrhage as a cause for this kind of shallowing of anterior chamber.)
  - Whereas in acute attack of PACG, typically there is peripheral shallowing of anterior chamber due to iris bombe.
- Lens-induced angle closure (e.g., phacomorphic or subluxed):
  - Will have intumescent lens causing pupillary block (phacomorphic)
  - Will have anterior subluxation or complete dislocation of lens causing blockage of anterior chamber angles (phacotopic glaucoma)
- Developmental disorders (e.g., nanophthalmos, retinopathy of prematurity, persistent hyperplastic primary vitreous)
- Iris or ciliary body mass lesions or cysts.
  - Can be visualized using ultrasound biomicroscopy (UBM).

CLASSIFICATION

Recently angle closure glaucoma has been classified into three categories by the International Society of Geographical and Epidemiological Ophthalmology (ISGEO)**(Table 1)**: (1) PAC; (2) PACG; (3) PACS; depending upon the presence of following:

- Iridotrabecular contact (>180°)
- Elevated IOP
- Peripheral anterior synechiae (PAS)
- Glaucomatous optic neuropathy
- *ISGEO classification:* The classification is described in **Table 1**.
- *Clinical classification:*
  - *PACS:* Appositional angle closure with normal gonioscopy and optic nerve (Similar to PACS as in ISGEO classification)
  - *Subacute PAC:* A rise in IOP occurs for a short period of time like minutes to some hours followed by spontaneous resolution of pupillary block.
    - Patient can have unilateral headache with blurring of vision with colored halos during the subacute attack.
  - *Acute ACG:* Acute/congestive ACG caused by sudden rise of high IOP most likely due to sudden occlusion of the angle.
    - Patient will complain of severe diminution of vision, headache with nausea and vomiting, pain, and redness of eye.
    - On clinical examination, typically there will be corneal edema, iris bombe,

Table 1: Classification and management of angle closure glaucoma.

Characteristics	***PACS***	***PAC****	***PACG†***
Iridotrabecular contact (>180°)	+	+	+
Elevated IOP	–	+/–	+/–
PAS	–	+/–	+/–
Glaucomatous optic neuropathy	–	–	+
Treatment	Close observation with serial gonioscopy or LPI‡	LPI+ medical or surgical therapies to control IOP	LPI+ medical or surgical therapies to control IOP

*For a diagnosis PAC either elevated IOP or PAS, one of them must be positive, along with iridotrabecular contact (>180°).
†For a diagnosis of PACG either elevated IOP or PAS, one of them must be positive, along with iridotrabecular contact (>180°) and optic nerve damage.
‡LPI should be considered taken into consideration of symptoms suggestive of intermittent angle closure, systemic medications that may predispose to pupillary block, need for frequent pupillary dilation or lack of reliable access to healthcare.
(IOP: intraocular pressure; LPI: laser peripheral iridotomy; PAC: primary angle closure; PACG: primary angle closure glaucoma; PACS: primary angle closure suspect; PAS: peripheral anterior synechiae)

ciliary and conjunctival congestion, and vertically oval mid-dilated pupil.

- *Chronic PACG:* IOP is chronically raised leading to synechial angle closure.
  - Patients may be asymptomatic and have gradual diminution of vision.

INVESTIGATION

- Visual field analysis by automated perimetry to document visual field loss.
- *UBM:* It can help to elucidate the underlying mechanism of angle closure in most cases, including plateau iris syndrome and iridociliary cysts, thereby allowing the appropriate treatment to be given.
  - Can especially assess the angle structures in cases of corneal edema where gonioscopy and anterior segment optical coherence tomography (AS-OCT) are not useful.
- *AS-OCT:* It can detect angle closure but has a high rate of false positives when assessed against gonioscopy. ASOCT has not replaced gonioscopy yet.

MANAGEMENT

Medical Management

Management of acute angle closure attack includes the following:

- Acetazolamide (250–500 mg) oral stat, then 125–250 mg tid/qid until symptoms subside or IV mannitol 1.5 g/kg of body weight of 20% 200 mL over 30 minutes, followed by oral acetazolamide 250 mg qid
- Topical pilocarpine 2% stat, then qid can be given but after inflammation control
- Analgesics and antiemetic as required
- Topical beta-blockers eye drop BD
- Topical steroids qid
- Preventive laser peripheral iridotomy (LPI) should be done in fellow eye as early as possible
- Once IOP is controlled and corneal edema clears, an LPI should be performed.

Management of Primary Angle Closure Suspect

Management options include LPI or close observations with regular follow-ups for IOP checking, gonioscopy, and disc evaluation. Indication for LPI in PACS includes the following:

- Patients who needs frequent dilatation, for example, diabetics, hypertensive age-related macular degeneration (ARMD)
- Patients who had previous angle closure attack in one eye. A prophylactic peripheral iridotomy (PI) for fellow eye is advised.
- Hyperopic patient
- Patient who is unlikely to come for regular follow-up
- Family history of glaucoma.

Surgical Treatment

A patient whose IOP does not come under control with PI and medications, and those with fairly advanced disease will require filtering surgery. The application of antifibrotic agents such as 5-fluorouracil (5-FU) and mitomycin C (MMC) results in greater success and lowers IOP following trabeculectomy.

VIVA QUESTIONS

Q.1. How do you grade peripheral anterior chamber depth (PACD) on slit lamp?

Ans. *By Van Herick method:*

- *Grade 0:* Iridocorneal contact
- *Grade 1:* PACD <1/4 corneal thickness
- *Grade 2:* PACD = 1/4 corneal thickness
- *Grade 3:* PAC = 1/4–1/2 corneal thickness
- *Grade 4:* PACD ≥1 corneal thickness

Grade 0, 1, and 2—suspicious of angle closure

Q.2. Which are the risk factors for developing PACG?

Ans. *Following are risk factors for PACG:*

- *Patient factors:*
  - Advancing age
  - Female gender
  - Asian or Inuit descent
  - Family history of angle closure
- *Ocular factors:*
  - Shallow anterior chamber
  - Narrow angle
  - Relative anterior location of iris-lens diaphragm
  - Hyperopia [increased lens thickness, small corneal diameters, and short axial length (AL)]

Q.3. What are the mechanisms of angle closure?

Ans.
- Pupil block (most common)
- *Abnormalities anterior to iris:*
  - PAS
  - ICE syndrome
  - Neovascular glaucoma
- *Abnormalities of iris and ciliary body:*
  - Thick peripheral iris, cysts of iris, or ciliary body
  - Peripheral iris roll
- *Abnormalities of lens:*
  - Thick intumescent lens
  - Subluxated lens
- *Abnormalities posterior to lens:* Malignant glaucoma

Q.4. What are the indications for laser peripheral iridotomy in PACS?

Ans.
- Patient who needs frequent dilatation, for example, diabetic, hypertensive, ARMD
- Patient who had previous angle closure attack in one eye, prophylactic PI for fellow eye
- Hyperopic patient
- Patient who is unlikely to come for regular follow-up
- Patient who has a family history of glaucoma

Q.5. Explain the technique to perform LPI.

Ans. The procedure of LPI involves the following:
- *Before LPI:* Pilocarpine (1%) eyedrops, then anesthetize the eye with 0.5% proparacaine
- *LPI:* Abraham's type of contact lens is applied. This lens has a +55 D, peripheral button over a routine contact lens. This lens helps in the following ways:
  - It stabilizes the eye and prevents undue movements.
  - It helps to open the eye and keep the lids retracted during the procedure.
  - It smoothens out the corneal surface.
  - It provides peripheral view, which is highly magnified.
  - It helps to reduce the axial expansion of plasma, which reduces the unnecessary spread of the damage.
  - It increases the power density of the spot.
  - Gives pressure to prevent the bleed from increasing.
- *Site of LPI:* The iridotomy site should be in the peripheral third of the iris, just anterior to the arcus. A crypt or a thinned area of the iris is recommended. Most ophthalmologists place the iridotomy between 11 o'clock and 1 o'clock, where the lids superiorly cover it.
- *Size of LPI:* Iridotomy be at least 200 μm in size. The preferable size is 500 μm in diameter.
- *End point*: Once the iridotomy is complete, one can notice a sudden gush of aqueous or outflowing of the pigment from the posterior to the anterior chamber along with sudden deepening of the anterior chamber. (The presence of retroillumination may be looked for after a few weeks of laser iridotomy; however, it is not a sure sign of total penetration. Visualization of the anterior lens capsule confirms LPI. AS-OCT can also be performed on follow up to look for patency of iridotomy).
- *Parameters for LPI*: In Indian patients with brown irides, LPI can be performed with a neodymium: yttrium-aluminum-garnet (Nd:YAG) laser, using the following settings:
  - *Power:* 4–8 mJ
  - *Pulses/burst:* 1–3
  - *Spot size:* Fixed
- *Monitoring and follow-up post LPI:* At 1 hour after completion of LPI, the IOP should be checked to make sure that it did not increase significantly (i.e., IOP has not increased by ≥8 mm Hg and that IOP does not exceed 30 mm Hg). Topical prednisolone acetate 1% is given four times a day for 5–7 days. Topical beta-blocker is added in cases of PACS or continue antiglaucoma medications if patient is already on antiglaucoma medications. At 1 week, the patient is seen to monitor IOP, to confirm the patency of the iridotomy site, and to check for any significant intraocular inflammation.

Q.6. What are the complications of LPI?

Ans. *Common complications are as follows:*

- Postoperative intraocular pressure spike
- Anterior uveitis
- Iris bleeding and hyphema
- Focal cataract
- Posterior synechiae
- Visual symptoms
- Corneal decompensation

Rare complications of LPI are as follows:

- Aqueous misdirection
- Recurrent herpetic keratouveitis
- Retinal and subhyaloid hemorrhage
- Choroidal and retinal detachment after argon LPI
- Stage I macular hole

Q.7. Risk of progression of PACS to PAC/PACG.

Ans. Over 5 years, PACS has 6% risk of progression to PAC/PACG.

Q.8. The International Society of Geographical and Epidemiology of Ophthalmology classification of ACG.

Ans. The International Society of Geographical and Epidemiology of Ophthalmology classified PAC disease into PACS, PAC, and PACG based on IOP, gonioscopy findings, and disc and visual field examination.

Q.9. Is there a role of LPI in PAC disease in a pseudophakic patient?

Ans. No, in case the patient is pseudophakic, there is no role of LPI in PAC disease.

Q.10. Methods to prevent or manage bleeding during LPI procedure.

Ans.

- With the use of Abraham's lens during the procedure, can apply pressure on the eye for some time till bleeding stops.
- Immediately abandon the procedure in case of bleeding causing media haze and poor visibility.
- Always ask history of the patient taking anticoagulants, as this can lead to severe bleeding during iridotomy. This can be easily prevented by adjusting the dose of anticoagulants before LPI.

Q.11. Points to note while performing gonioscopy.

Ans.

- Posterior most structure visible
- Angle structure visible after manipulative gonioscopy
- Pigmentation of angle
- Insertion of iris
- Presence of iris processes, gonio-synechiae, PAS
- Presence of neovascularization
- Presence of blood in Schelmm's canal

Q.12. What are the inclination of mirrors for Goldmann gonioprisms?

Ans.

- *Goldmann single mirror:* Mirror is inclined at 62° for gonioscopy
- *Goldmann three mirror:* One mirror inclined at 59° for visualizing the angle, the other mirror is inclined at 67° to visualize the ora serrata, and the third is inclined at 73° to visualize the equator.

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OPHTHALMOLOGY CLINICS-I
for Postgraduates

Ophthalmology Clinics is a book primarily dealing with "How to approach a case in clinical practice". Students doing MD/MS/DNB/Diploma/Fellowship all over India and abroad have to appear in various examinations for degree as well as fellowship. In examinations, the cases are to be presented in a manner, which is never given in any standard textbook. In addition, while dealing with cases in clinic, the clinicians have to approach systematically taking into consideration all the clinical findings. Standard textbooks never describe the topics in the way a clinician has to approach. This book presents the cases in a manner that will help the students and the clinicians to understand how to approach a case and arrive at a clinical diagnosis.

The book contains 32 long cases and 48 short/spot cases, commonly asked in postgraduate examinations. In addition, it includes a chapter on instruments that is often a universal component of all postgraduate viva voce examinations. We expect our effort would ease the pressure of postgraduate students significantly.

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