February 2017 Volume XXXV No 1

# insight

Scientific Journal of

## MEDICAL & VISION RESEARCH FOUNDATIONS

www.sankaranethralaya.org

Editor: Parthopratim Dutta Majumder

insight

Scientific Journal of **Medical & Vision Research Foundations**

Year: 2017

Issue: Vol. XXXV | No. 1 | February 2017 | Pages 1–33

Typeset at: Nova Techset (P) Ltd., Chennai, India

Printed at: Gnanodaya Press, Chennai, India



**Sankara Nethralaya – The Temple of the Eye.**

It was in 1976 when addressing a group of doctors, His Holiness Sri Jayendra Saraswathi, the Sankaracharya of the Kanchi Kamakoti Peetam spoke of the need to create a hospital with a missionary spirit. His words marked the beginning of a long journey to do God's own work. On the command of His Holiness, **Dr. Sengamedu Srinivasa Badrinath**, along with a group of philanthropists founded a charitable not-for-profit eye hospital.

Sankara Nethralaya today has grown into a super specialty institution for ophthalmic care and receives patients from all over the country and abroad. It has gained international excellence and is acclaimed for its quality care and compassion. The Sankara Nethralaya family today has over 1400 individuals with one vision – to propagate the Nethralaya philosophy; the place of our work is an Alaya and Work will be our worship, which we shall do with sincerity, dedication and utmost love with a missionary spirit.

# Contents

# Foreword

## Lingam Vijaya

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Glaucoma is the leading cause for irreversible blindness across the world; half of the cases blinded by glaucoma are due to primary angle-closure glaucoma (PACG). PACG is considered to be a more visually destructive form of disease than primary open-angle glaucoma.[1] The prevalence of the disease varies significantly among different ethnicities and is the most common one among Inuits. Asian ethnicity, older age and female gender are considered to be risk factors for the disease.

Primary angle-closure disease (PACD) is a spectrum of disease with various stages in its natural history, the initial stage being primary angle-closure suspect (PACS) followed by primary angle-closure (PAC) and PACG. Defining the various stages is essential for determining the appropriate treatment strategy. The initiating event is prolonged and/or repeated irido-trabecular contact that leads to defective aqueous filtration and formation of peripheral anterior synechiae (PAS). Pathogenesis involves a combination of predisposing anterior segment anatomy that involves axial length, iris volume, angle configuration, ciliary body size and position and lens thickness with exaggerated physiological responses. Pupillary block is a major component of the disease process in at least 75% of eyes and these eyes respond well to the laser iridotomy (LPI). Non-pupillary mechanisms such as plateau iris configuration and syndrome play a role in the rest. In view of the complexity of its pathogenesis, the natural history of the disease is still evolving.

Scientific research in the field of PACD has provided valuable information for disease management. One area that deserves special recognition is improvement in using imaging technology for eye diseases. Both ultrasound biomicroscopy (UBM) and anterior segment optical coherent biomicroscopy (AS-OCT) have contributed significantly in understanding the pathophysiology of PACD.[2, 3] UBM imaging of the iridociliary apparatus and the lens equatorial sulcus have shown that the plateau iris phenomenon is due to anteriorly positioned ciliary processes that causes angulated peripheral iris and narrow angle. Similarly, imaging the structures posterior to the lens–iris diaphragm has helped us to understand the causes for angle-closure disease in younger adults. UBM examination has revealed that angle closure in younger patients is most often due to secondary causes such as ciliary body cysts or abnormal morphology of the crystalline lens. Studies involving AS-OCT has helped us to understand the importance of increased iris thickness, area and curvature in the pathogenesis of PACD. Imaging the iris with AS-OCT has revealed an increase in iris volume after pupil dilatation in eyes with angle closure in comparison to open angles.[4] The clinical implications of these findings are still not clear. However, the use of newer ASOCT technology such as swept-source OCT may help us to understand this phenomenon better and probably may allow us to take it further for clinical use. Among the anatomic risk factors, lens thickness and position are important. Recently, AS-OCT studies have shown that increased lens vault (LV) contributes to the pathophysiology of PACD. The LV is defined as the perpendicular distance between the horizontal line joining the two scleral spurs and the anterior pole of the crystalline lens and represents the anterior portion of the lens on AS-OCT. An increased LV indicates increased lens thickness and/or bulk anterior to the scleral spur plane, which subsequently increases the risk of angle closure. However, further studies have suggested that relative LV in relation to anterior chamber depth is more important than absolute LV.[5] At present, it appears that we need to fine-tune this information with further studies to understand the actual relationships.

Information from epidemiological population-based studies has given insights into the natural history of the disease.[1] There seems to be a large proportion of people with PACS; progression to PAC or PACG is very slow and chronic and not all progress to the next stage. These findings have strengthened the recent shift in clinical practice of not treating all PACS and having a selective approach in performing laser peripheral iridotomy (LPI) for PACS.

There seems to be a relationship between cataract and PACD. In eyes with significant cataract and PACS, removal of cataract will serve as an alternative to LPI. Eyes with PAC or PACG require definite treatment with either LPI or cataract extraction if there is an associated cataract.

Why cataract surgery for PACD? The debate on offering cataract surgery as a treatment modality for PACD was started many years back. Improvements in cataract surgery techniques have made cataract surgery a safer, faster, and more affordable procedure; in view of this, its role in the treatment of angle closure is coming to the forefront. A recent multicenter randomized control

study (EAGLE) has evaluated LPI versus clear lens extraction for eyes with PAC and PACG, and concluded that clear lens extraction was found to be economical and efficient.[6] One should realize the removal of cataract is not equal to clear lens extraction; eyes with PACD possess a challenge for safer surgery in view of their anatomically small configuration. One randomized study data cannot be extrapolated to the whole population at large. Till we gather sufficient information, the practice of offering cataract surgery for those eyes with significant cataract seems to be a better option.

Significant research has been done in the field of PACD; however, a lot more remains to be done to understand the complexities of the disease.

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**How to cite this article** Vijaya L. Foreword, *Sci J Med & Vis Res Foun* 2017;**XXXV**:1–2.

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# Classification and management of primary angle closure disease

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It is estimated that almost 15 million people worldwide have been affected by angle closure disease in 2010 and the number is expected to increase to 21 million by 2020.[1] Angle closure disease is responsible for nearly half of the world's blindness due to glaucoma. The sheer magnitude of the disease and its amenability to treatment, if detected early, makes it imperative for all ophthalmologists to follow evidence-based protocols for the management of the disease.

The current classification of primary angle closure disease (PACD) is based on the definition proposed by the International Society for Geographical and Epidemiological Ophthalmology (ISGEO).[2] The classification was primarily described for epidemiological research but has today become an integral part of our day-to-day practice.

## Classification of PACD

**(1) Primary angle closure suspect (PACS)**

An eye in which there is irido-trabecular contact for at least 270° on gonioscopy with the eye in the primary position, without compression, using appropriate illumination, with normal intraocular pressure (IOP), optic disc and visual fields (Fig. 1).

**(2) Primary angle closure (PAC)**

The presence of irido-trabecular contact for at least 270°, with either raised IOP and/or peripheral anterior synechiae (PAS), but with normal optic disc and visual fields (Fig. 2).

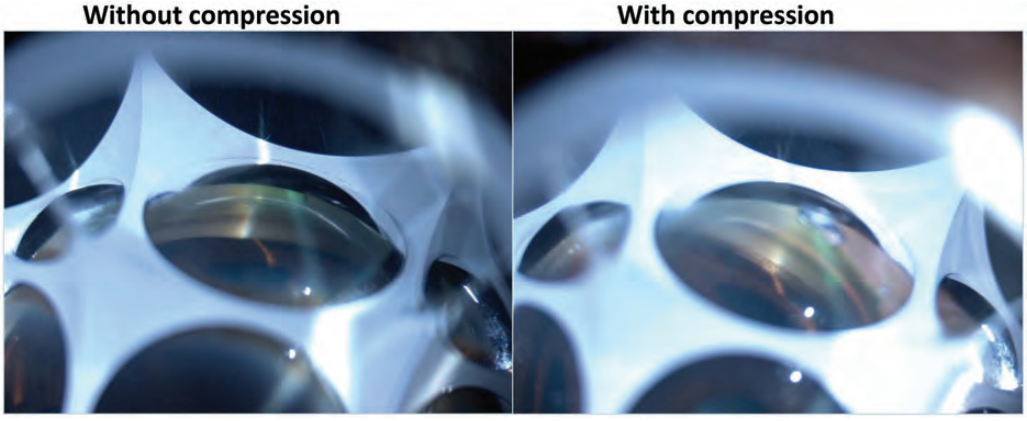


**Figure 1:** Primary angle closure suspect on gonioscopy.

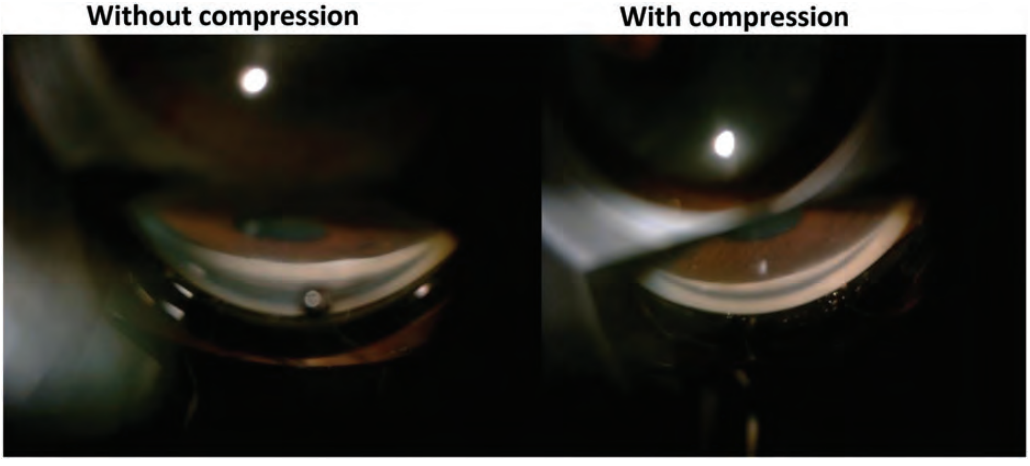


**Figure 2:** Primary angle closure on gonioscopy.

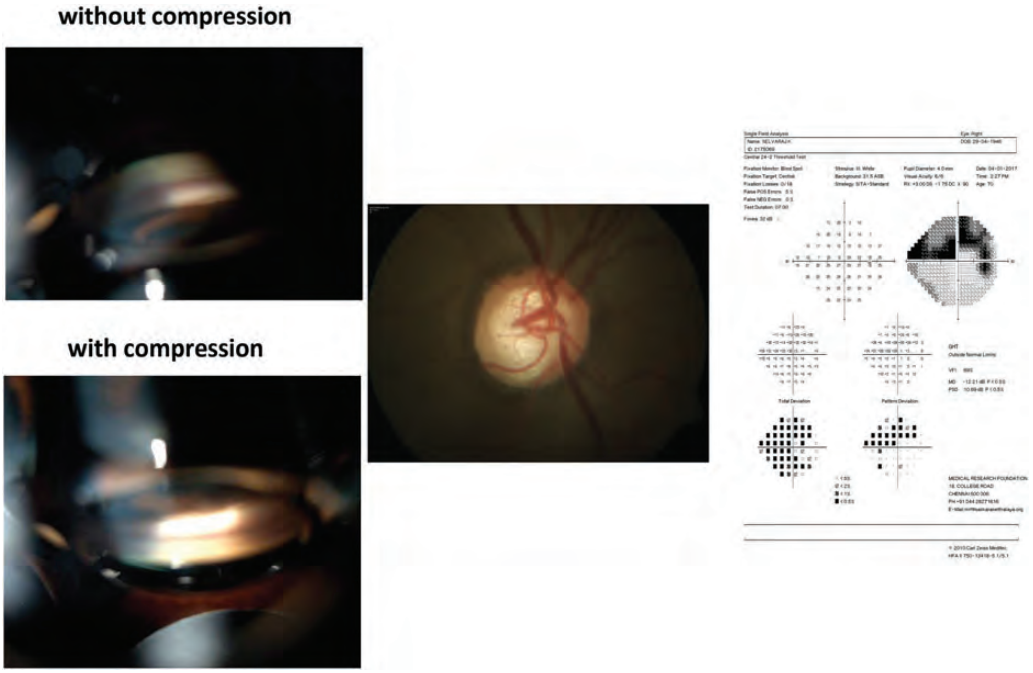


**Figure 3:** Primary angle closure glaucoma.

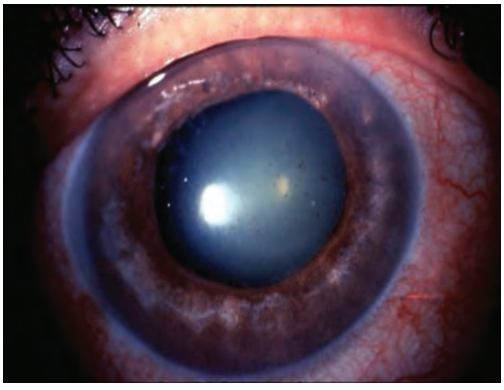

**Figure 4:** Acute angle closure crisis.

**(3) Primary angle closure glaucoma (PACG)**
PAC with evidence of glaucoma (optic disc/field changes) (Fig. 3).

**(4) Acute angle closure crisis**
Symptoms of pain, either ocular or periocular, often accompanied by headache, nausea or vomiting, presenting with an IOP of >21 mmHg, with signs such as circumcorneal congestion, corneal edema, mid-dilated pupil, and a shallow anterior chamber (Fig. 4).

## Management of angle closure disease

Various treatment algorithms have been described for the management of PACD. A number of randomized control trials (RCTs) have been performed to offer us some evidence on which to base these treatment protocols and these will be described under the following headings:

1 Management of an acute attack of angle closure (acute angle closure crisis or AcACC)

2 Management of PAC

3 Management of PACG

4 Management of PACS

The rationale of treatment, once angle closure has been diagnosed, is to

1 eliminate pupillary block;

2 assess the extent of residual angle closure; either appositional or synechial;

3 establish baseline features following iridotomy in terms of goniosopic, optic disc, and visual field assessment;

4 follow up in terms of progression of angle closure, IOP elevation, disc or field changes.

**Management of acute angle closure crisis (AcACC)**
The mainstay of treatment is to lower the IOP initially, in order to alleviate the patient's symptoms, to use topical steroids to reduce inflammation, and to relieve the pupillary block by performing a laser peripheral iridotomy (LPI).[3]

The initial treatment may involve the use of one or more of the following: topical aqueous suppressants, oral carbonic anhydrase inhibitor (acetazolamide), oral hyperosmotic agent (glycerol), intravenous use of acetazolamide, if available, and intravenous hyperosmotic agent such as mannitol. Topical steroids may also be used to control inflammation. Topical pilocarpine may be

used after IOP has been sufficiently lowered since it does not constrict the pupil in the presence of sphincter ischemia.

Once adequate lowering of IOP has been achieved and intraocular inflammation has subsided, LPI can be performed.

In case of poor visibility due to corneal edema and failure to control IOP by medical methods, the following treatment options can be attempted.

#### (a) Role of Argon laser iridoplasty in acute angle closure attack

Argon laser iridoplasty helps us to mechanically pull the peripheral iris away from the trabecular meshwork by placing contracture burns in the peripheral iris, thereby opening the angle and lowering the IOP.[4] Once the IOP has been lowered, inflammation has subsided and the cornea becomes sufficiently clear, a laser iridotomy may be safely performed.

A randomized control trial in which 73 eyes of 64 consecutive patients who presented with acute angle closure crisis was conducted in which patients were randomized after receiving topical pilocarpine 4% and topical timolol 0.5% to one of the two treatment groups; one arm received systemic IOP lowering therapy and other arm was assigned to immediate argon laser peripheral iridoplasty (ALPI).[5] The ALPI group had lower IOP levels than the medical treatment group up to 2 hours after the start of the treatment. The difference was insignificant thereafter.

#### (b) Role of paracentesis

In cases of poor visibility precluding laser therapy, paracentesis is a useful option, providing immediate lowering of IOP and relief of symptoms. Lam et al.[6] described this technique in a prospective case series of 10 eyes with AcACC. The mean IOP reduced from $66.6 \pm 9.1$ to $15.1 \pm 3.5$ mmHg immediately after the procedure and remained <21 mmHg after 2 hours and beyond. There were no complications reported. However, the technique may be difficult to perform if the patient is very symptomatic or has a very shallow anterior chamber.

#### (c) Trabeculectomy

A substantial percentage of cases of AcACC remains unresponsive to medical treatment, especially if residual PAS extending to >180° is present. Laser therapy in the setting of an acute attack may not be possible or may be too risky to perform. In such situation, trabeculectomy may provide an alternative solution.

In a retrospective analysis of 56 patients with AcACC, who underwent trabeculectomy without antimetabolites, two groups were compared; Group A; medical failure group and Group B; medical success group (IOP <22 mmHg but had evidence of chronic AC).[7] Successful control of IOP was achieved in 65.6% patients in Group A and 87.5% of patients in Group B. Early postoperative complications occurred more frequently in Group A; 31.3% vs 16.7% in Groups A and B, respectively. The authors concluded that in the face of high failure rates in terms of IOP control, trabeculectomy may not be the best option available.

#### (d) Role of lens extraction

Clear lens extraction in AcACC.

A second option in the face of AcACC which is unresponsive to conventional treatment due to the formation of extensive PAS is to perform clear lens extraction. The rationale of performing this procedure is that increasing lens thickness, relative anterior lens position and increasing lens vault may be responsible for the crowding of the angle, and this may be ameliorated by removing the crystalline lens. Lam et al.[8] reported the results of an RCT in which 62 eyes of 62 Chinese patients were randomized into two groups; early phacoemulsification (PKE) and LPI. The prevalence of IOP elevation at 18 months following treatment was 3.3% in the PKE group and 46.7% in the LPI group. The number of IOP lowering medications was significantly less in the PKE group ($p < 0.001$). The angle was significantly more open in the PKE group compared with the LPI group at all time points ($p < 0.001$). There were no serious adverse events in either group. All complications were managed with conservative measures.

#### Treatment of the fellow eye

The risk of the fellow eye suffering an attack is high; hence, it is essential to do a peripheral iridotomy for the fellow eye.[9]

A long-term follow-up in terms of IOP control, progression of angle closure, and optic disc and visual field changes is necessary following management of the initial attack in all eyes with AcACC.

#### Management of PAC and PACG

The initial treatment consists of medical treatment of elevated IOP followed by laser iridotomy. After sufficient time has elapsed for treatment effects such as inflammation, and IOP spikes following treatment to subside, re-assessment needs to be done. The extent of residual syncechial/appositional closure needs to be established. A long-term follow-up is required because >40% ultimately require surgical therapy for the management of elevated IOP.

There is no clear cut evidence as to which algorithm to follow depending on the extent of PAS formation. Most algorithms use a cut-off of 180° based more on intuition than science at present. It the extent of PAS is <180°, iridoplasty alone, in the presence of a clear crystalline lens, or lens

extraction alone, if there is a significant cataract, are the options available. Elevated IOP following either of these procedures is managed with medical therapy, although one study did report a favorable response using selective laser trabeculoplasty in the management of residual glaucoma following laser iridotomy.[10]

If the extent of PAS is >180°, the options are to consider trabeculectomy alone; if the IOP remains, elevated despite topical medications; and if there are significant lens changes, combined surgery is the option.

Two RCTs have been performed by Tham et al.[11, 12] to study the effects of lens extraction alone versus either combined surgery or trabeculectomy: one in eyes with medically controlled chronic angle closure glaucoma (CACG) following iridotomy and one in eyes with medically uncontrolled CACG following LPI.

The first trial consisted of randomizing 72 medically controlled patients with PACG with cataract to either phacoemuslification alone (Group I, 35 patients) or to combined PKE with trabeculectomy with adjunctive Mitimycin C (Group II (37 patients)). At the end of 24 months, although the number of IOP-lowering medications required was less in Group II ($p<0.001$), the IOP control and rates of progression were similar in both groups. There were 14 postoperative complications in Group II and one postoperative complication in Group I ($p<0.001$).

The second trial consisted of 50 medically uncontrolled PACG eyes without cataract which were randomized either to PKE alone (26 eyes) or to trabeculectomy with adjunctive Mitomycin C (24 eyes). IOP reduction at the end of 24 months was similar for the two groups, 34% for the PKE group, and 36% for the trabeculectomy group. The number of postoperative complications was higher in the trabeculectomy group; 4 vs 46 % for the PKE group versus the trabeculectomy group, respectively, $p=0.001$.

Some of the limitations of these trials include the small sample sizes in each group, as well as the lack of information as to the extent of synechial closure in each group prior to the surgical therapy.

Role of clear lens extraction in the management of PAC and PACG. *The EAGLE study.*

Azuor-Blanco et al.[13] recently published the results of their RCT using clear lens extraction for the management of PAC and PACG. Of the 419 patients enrolled, 155 had PAC and 263 had PACG. A total of 208 were assigned to clear lens extraction and 211 were assigned to standard care, i.e. LPI with medical therapy. The main outcome measures included patient-reported quality of health status, IOP and cost-effectiveness gained after 36 months of follow-up. The mean health status score was 0.052 higher and the mean IOP was −1.18 mmHg lower (95% C.I −1.99, −0.38) after clear lens extraction compared to standard of care. Irreversible loss of vision occurred in one patient in the clear lens extraction group and in three patients who received standard of care.

There are several reasons for which the study results cannot be extrapolated to the treatment of PAC or PACG in the clinical scenario. Younger patients, i.e. <50 years of age, will lose their ability to accommodate when clear lens extraction is performed. The extent of glaucomatous damage was limited to very early glaucoma; the range of moderate glaucoma varied from −3 dB to −7.2 dB, which is much lower than the extent of damage seen in our population. Gonioscopy data were missing in 247 (58.9%) patients. The difference in IOP between the two groups was very minimal. The quality-of-life measure would have reflected the correction of refractive error in the clear lens extraction group.

#### Management of PACS

Although LPI is recommended as the initial treatment of choice in angle closure disease, it is not mandatory to perform an LPI on all primary angle course suspects (PACS). Progression from PACSs to primary angle closure was reported by Thomas et al.;[14] 82 of 118 persons identified to be PACSs in 1995 were invited for a follow-up examination in the year 2000, along with 110 normal persons. Of the 50 persons who presented for examination, 11 (22%, 95% CI 1.8 to 34.2) developed primary angle closure based on the development of raised IOP or synechiae. All of them were bilateral PACS. The relative risk of progression to primary angle closure was calculated as 24 (95% CI 3.2, 182.4). None proved to have any biometric risk factors.

There is no current evidence which establishes the role of LPI in preventing progression to PAC or PACG in angle closure suspects.

However, laser iridotomy is performed in the following situations.

Fellow eyes of patients with either PAC or PACG would definitely require prophylactic LPI. Other factors to be considered include the presence of patchy pigmentation of the posterior trabecular meshwork suggestive of prolonged iridotrabecular contact, symptoms suggestive of intermittent closure, or in those requiring repeated pharmacological dilatation such as patients with diabetes mellitus. Relative indications include an inability to follow-up on a regular basis and family history of blindness due to angle closure glaucoma.

## Summary

A frame work for the management of PACD throughout its spectrum has been established, as far as possible, based on evidence. Although the

mainstay of treatment consists of laser iridotomy with medical therapy, it is very clear that long-term follow-up is required since a large percentage of patients, especially those with chronic disease, continue to progress. The role of cataract extraction in the presence of PACD has been quite clearly established; however, the role of clear lens extraction in the Indian scenario has to be carefully considered on an individual basis.

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**How to cite this article** Shantha B. and David R.L. Classification and management of primary angle closure disease, *Sci J Med & Vis Res Foun* 2017;**XXXV**:3–7.

# Anterior segment imaging in angle-closure disease

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## Introduction

Angle-closure disease (both primary and secondary) is a spectrum of disorders characterized by irido-trabecular contact obstructing aqueous outflow. Anterior segment imaging plays an important role in understanding the pathophysiology of angle closure, diagnosis, and monitoring the treatment, especially in challenging cases.

Indirect gonioscopy, the gold standard for visualizing the anterior chamber angle (ACA) is a subjective technique with moderate agreement among experts and inability to visualize structures posterior to the iris. It is affected by room illumination, pressure on the globe, clinicians' skill, and patient cooperation. With advances in technology, many instruments are available for imaging the anterior segment. Each comes with its own advantages and limitations which the clinician must consider while making decisions.

## Ultrasound biomicroscopy

Ultrasound biomicroscopy (UBM), developed by Pavlin et al.,[1] provides high-frequency (50–100 MHz) in vivo B-scan ultrasonography images of the anterior segment of the eye up to a depth of 4 mm with 25 µm axial and a 50 µm lateral resolution. In angle-closure disease, where more than one mechanism often coexists, UBM helps us to understand the dynamics and mechanism involved. Sound penetrates both opaque media and iris. Thus, retro-iridal structures like the ciliary body can be imaged. Images can also be obtained in the presence of corneal scars, hyphema, or corneal oedema providing invaluable information in secondary angle-closure disease.

In pupillary block glaucoma, UBM demonstrates a peripherally shallow anterior chamber, iridotrabecular contact, convex iris configuration (Fig. 1) and a well-formed posterior chamber.[1] Pavlin et al. described reproducible quantitative information in the form of indices like angle opening distance (AOD), trabecular-ciliary process area, and iris thickness at specific positions (Fig. 2). However, this assumes the iris surface to be a straight line. To overcome this, Ishikawa and Schuman[2] defined angle recess area (ARA), taking into account the iris irregularities (Table 1). They developed a semi-automated software to provide these parameters after identification of the scleral spur (SS). These parameters document the change with time or with treatment and help us to classify angle-closure disease into subtypes. Marchini et al. found a shorter axial length, a shallower anterior chamber depth (ACD), a thicker lens, more anteriorly located lens, a narrower ACA, a shorter trabecular-ciliary process distance (TCPD), and a smaller AOD500 in eyes with PACG as compared to normal.[3] Guzzard et al.[4] showed peripheral ACA widening and a reduction in iris convexity following laser peripheral iridotomy (LPI) (Fig. 3). Nonaka et al.[5] reported angle widening and alteration in ciliary process configuration following cataract surgery. UBM can also be used to assess the adequacy and patency of LPI. Ramani et al.[6] predicted the risk of development of PAC in eyes post-LPI. They found that 28% of patients who progressed to PAC had smaller ARA. UBM demonstrated a shallower ACD, narrow chamber angle, greater LV and a more anteriorly rotated ciliary body in eyes with acute primary angle closure (APAC). UBM can show residual appositional angle-closure post-LPI due to a more anteriorly positioned ciliary body, larger lens, and a thicker peripheral iris predisposing to progressive angle closure. UBM is the preferred imaging modality in plateau iris where it demonstrates a flat central iris plane, a steep rise in iris root from the point of insertion, an anteriorly positioned ciliary body (Fig. 4), absence of ciliary sulcus, large and long ciliary processes causing iridotrabecular contact impairing aqueous outflow despite patent LPI.[7] It also helps in identifying pseudo-plateau iris produced by irido-ciliary cysts (Fig. 5), ciliary body edema, tumours or infiltration. UBM can also show iris flattening and subsequent angle opening post laser iridoplasty.

In aqueous misdirection syndrome or malignant glaucoma, UBM shows swelling or anterior rotation of the ciliary body with forward movement of the lens-iris diaphragm, uniform shallowing of AC and angle closure by the iris pushing against the trabecular meshwork (Fig. 6). UBM has been used to differentiate malignant glaucoma

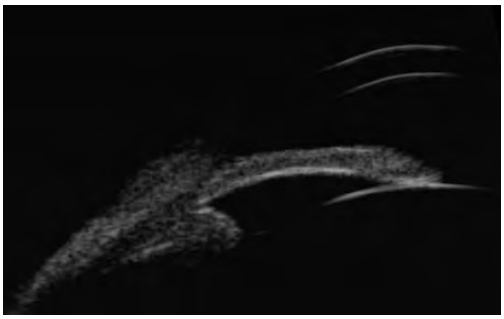

**Figure 1:** UBM image showing appositional angle closure due to convexity and forward bowing of the iris.

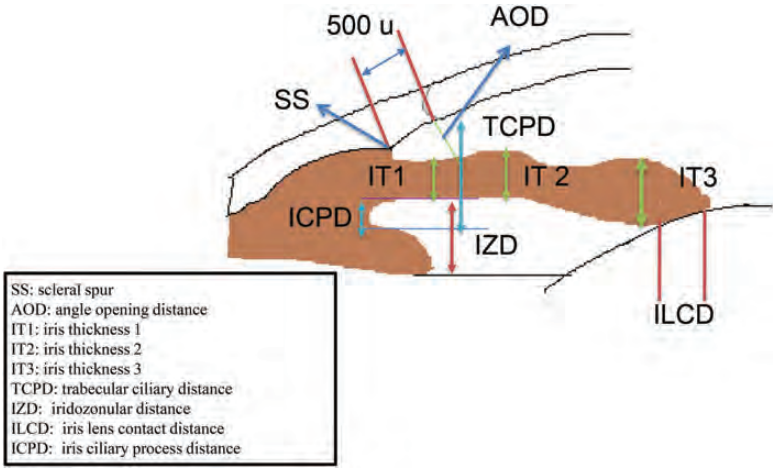


**Figure 2:** Diagram showing some of the quantitative parameters used for objective measurement of the anterior segment.

**Table 1** Parameters for quantitative measurement of anterior segment with UBM and ASOCT.

| | |
|---|---|
| Angle opening distance (AOD 500/750) | Perpendicular distance from the trabecular meshwork at a specified distance (500 microns or 750 microns), anterior to SS to anterior iris surface |
| Trabecular iris surface area (TISA) | Area bounded anteriorly by the AOD, posteriorly by a line drawn from the SS perpendicular to the plane of inner scleral wall to the opposing iris, superiorly by the inner corneoscleral wall and inferiorly by the iris surface |
| Trabecular iris angle (TIA) | Defined in degrees as the angle formed from angle recess to points 500 micm from SS on trabecular meshwork and perpendicular on the surface of iris |
| Iris thickness 1 (IT 1) | Measured along the line extending from corneal endothelium at 500 micm from SS perpendicular through the iris |
| Iris thickness 2 (IT2) | Iris thickness at 2 mm from the iris root |
| Iris thickness 3 (IT 3) | Maximum iris thickness near the pupillary margin |
| Trabecular iris contact length (TICL) | Linear distance of contact between iris and cornea / sclera beginning at the SS |
| ARA | The area of triangle between angle recess and iris and cornea 500 / 750 micm from the SS |
| AC depth (ACD) | Distance from corneal endothelium to anterior surface of the lens |
| AC width (ACW) | Distance of a horizontal line joining the two SSs |
| Iris cross-sectional area (ICSA) | The average of cross-sectional area of both nasal and temporal and nasal sides |
| Iris curvature (ICurv) | Maximum perpendicular distance between iris pigment epithelium and line connecting most peripheral to most central point of epithelium |
| Scleral thickness (ST) | Measured perpendicular from the scleral spur to the episcleral surface |
| Lens vault (LV) | Perpendicular distance between anterior pole of crystalline lens and line joining the two scleral spurs |
| TCPD* | Distance between trabecular meshwork and ciliary process 500 microns anterior to the SS. |
| ICPD* | Distance between iris and ciliary process along the line of ICPD |
| IZD* | Distance between iris and zonules along the line of ICPD |

*Can be measured with UBM only.

from pupillary block glaucoma and to classify it into two groups (with and without supraciliary effusion) and planning the clinical management. It demonstrates a shallow AC and thicker lens in eyes with pseudo-exfoliation presumably due to zonular weakness. It can also be used to determine the mechanism of unilateral or secondary angle-closure glaucoma, e.g. pupillary block in pseudophakic eyes, nanophthalmos, uveal effusion syndrome, Vogt Koyanagi Harada syndrome, and drug-induced uveal effusion (Fig. 7). UBM can be used to demonstrate a large, intumescent lens with forward movement leading to due narrowing of ACA in phacomorphic glaucoma.

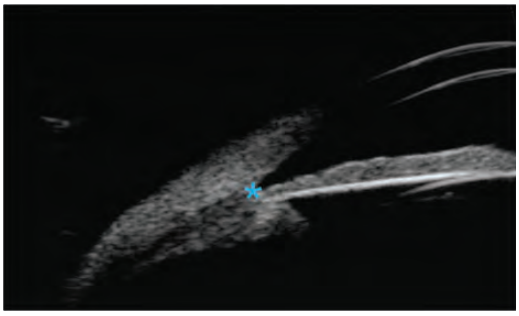

**Figure 3:** UBM images of the angle of anterior chamber before and after LPI showing flattening of the iris and widening of the ACA.

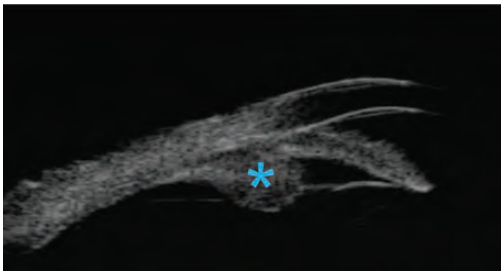

**Figure 4:** UBM image showing a narrow ACA with an anteriorly directed ciliary process (blue asterisk), absent ciliary sulcus in an eye with plateau iris.

Mansouri et al. showed a significant correlation of UBM with ASOCT in the measurement of ACA in eyes with angle-closure disease but poor agreement.[8] Kaushik et al.[9] and Radhakrishnan et al.[10] found angle widening following LPI to be better appreciated by UBM as compared to gonioscopy. More than 90% agreement with gonioscopy when done in a dark room has been reported.[11]

Continuing advances in technology aim at providing better speed, sensitivity, and depth of focus. High-frequency annular arrays, linear arrays for imaging without probe movement, are under investigation. An 80-MHz UBM (iScience, iUltrasound) has been used to image the Schlemm's canal.

## Challenges

UBM is a contact procedure, requires an eyecup with a coupling medium (saline or methylcellulose) and supine positioning of the patient. This can influence the angle configuration and AC depth. Inability to indent makes it difficult to differentiate between appositional and synechial angle closure. To overcome these limitations, some investigators have used special eyecups which allow corneal compression and probes with bag/balloon covers or bubble tips enabling UBM in sitting position without a waterbath. Despite this,

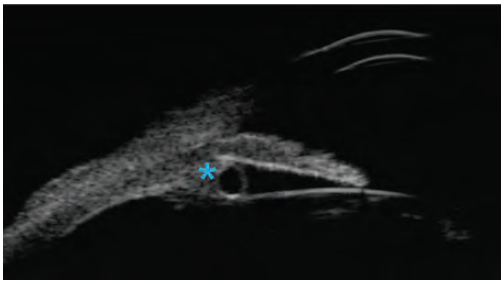

**Figure 5:** UBM image showing ciliary body cyst (blue asterisk) causing pseudo-plateau iris.

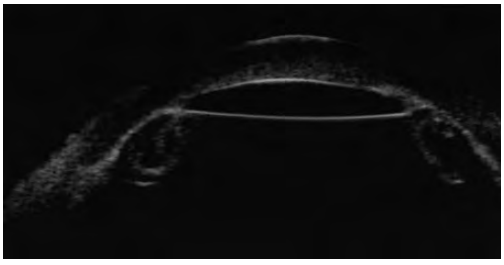

**Figure 6:** UBM image in aqueous misdirection syndrome showing flat anterior chamber, the supraciliary fluid, and anterior rotation of the ciliary body.

the procedure is more time consuming and requires a skilled operator. Moreover, the SS used as a reference for quantitative parameters needs to be marked manually. Good intraobserver but moderate interobserver agreement has been reported. Lin et al.[12] found increased interobserver variation with respect to UBM parameters involving ciliary processes and recommended measurement by the same observer. Radhakrishnan et al.[13] reported good correlation, similar reproducibility and sensitivity with UBM and ASOCT in eyes with narrow angles.

The UBM's ability to visualize the ciliary body, zonules and posterior chamber, regardless of media opacities, good agreement with gonioscopy outweigh its limitations. It provides qualitative and quantitative information regarding the pathophysiology of angle-closure disease which supplements clinical examination.

## Anterior segment optical coherance tomography

Anterior segment optical coherance tomography (ASOCT), introduced in 2003, is a non-contact method which uses a 1310 nm diode laser to provide cross-sectional, three-dimensional, high-resolution images (18 µm) images of the anterior chamber.[14] It allows qualitative and quantitative assessment of the anterior segment structures involved in the pathogenesis of glaucoma. The main advantage over UBM is the fact that it is a

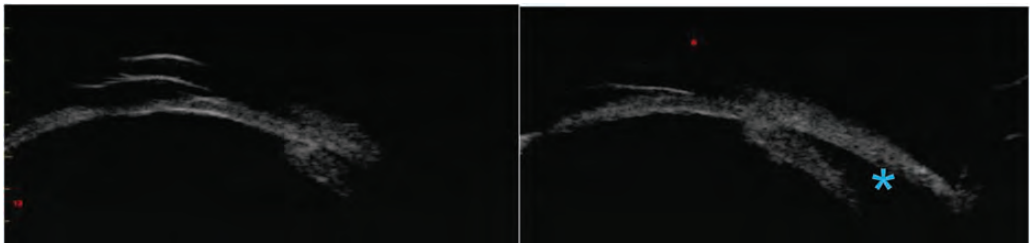

**Figure 7:** UBM image showing secondary angle closure in an eye with drug-induced uveal effusion (blue asterisk).

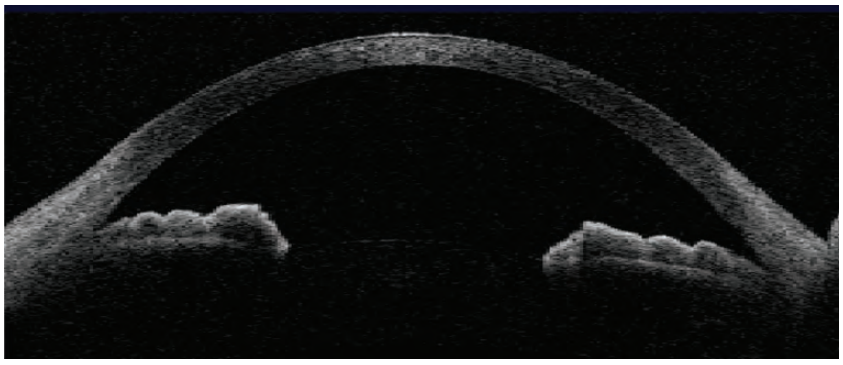

**Figure 8:** Depicts the normal anterior segment as imaged with ASOCT.

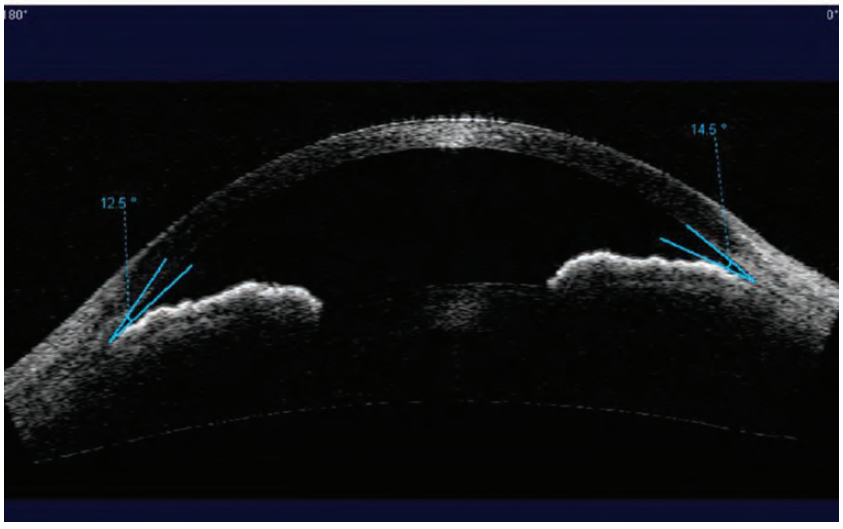


**Figure 9:** ASOCT picture of PACS showing narrow angle.

non-contact procedure that can be performed in a sitting position. High-definition and three-dimensional imaging of anterior segment structures with SD OCT provides a definition of ocular tissues comparable to histology.

ASOCT permits optimal visualization of angle structures, including the iris root, angle recess, anterior ciliary body, scleral spur and sometimes the Schlemms'canal (Fig. 8). Quantitative data provided by ASOCT (Fig. 9) can determine the mechanism of angle-closure disease by revealing the relationship between peripheral iris and trabecular meshwork, configuration of peripheral iris and its level of insertion. Polarization-sensitive OCT with its additional tissue-specific contrast has been used to visualize trabecular meshwork.[15, 16] A variety of ASOCT machines are currently available (Table 2). The scleral spur, used as a landmark, may not always be visible. Sakata et al.[17] reported that scleral spur was detected in 72% of the ASOCT images with difficulty in localization in the superior, inferior quadrants and in quadrants with a gonioscopically closed angle. Algorithms to detect angle parameters independent of scleral

**Table 2** Commercially available OCT systems used for anterior segment imaging.

| | Stratus OCT | Visante OCT | RT vue OCT | SL OCT | Cirrus OCT |
|---|---|---|---|---|---|
| OCT platform | Time domain | Time domain | Spectral domain | Time domain | Spectral domain |
| Light source | Superluminescent diode 820 nm | Superluminescent diode 1310 nm | Superluminescent diode 1310 nm | Superluminescent diode 840 nm | Superluminescent diode 840 nm |
| Scan speed | 400 A scans/ second | 2000 A scans/ second | 200 A scans/ second | 26000 A scans/ second | 27,000 A scans/ second |
| Axial resolution | 10 µm | 18 µm | Less than 25 µm | 5 µm | Less than 10 µm |
| Scan size (width × depth) | 6 mm × 2 mm | 16 mm × 6 mm | 15 mm × 7 mm | 2 mm × 2 mm | 3 mm × 1 mm |

spur have been developed. Various parameters available to quantify the iridocorneal angle are listed in Table 1.

Narayanaswamy et al.[14] assessed the diagnostic performance of angle measurements using ASOCT for eyes with narrow angles and found the AOD at 750 µm to be the most useful parameter. Studies have shown good repeatability and reproducibility for measurement of AOD, TISA, ARA and TIA with ASOCT. Apart from AC depth, ASOCT parameters associated with angle closure include decreased ACD, smaller AC width, ACA, ACV, larger lens vault, greater iris thickness, curvature and area. Angle parameters in fellow normal eyes of unilateral APAC had significantly shallower AC, smaller angles, shorter AOD, marked iris root curvature and a greater degree of closure. Nongpuir et al.[14] found eyes with angle closure to have thicker lenses with a greater lens vault. Aptel et al.[14] reported an increase in iris volume after pharmacological mydriasis in eyes with narrow angles. A reduction in AOD and TISA with physiological mydriasis under low-light conditions has been seen with ASOCT, especially in eyes with narrow ACD.

ASOCT has shown a change in angle parameters following LPI, the patency of LPI, and the detection of extent and location of peripheral anterior synechiae.[14] It can document secondary angle closure from a subluxated or dislocated lens. Phacomorphic glaucomas have shallower anterior chamber in the periphery than centrally, with iridotrabecular contact associated with the cataractous lens.

In malignant glaucoma, a uniform shallowing of anterior chamber, marked displacement of anterior segment structures with peripheral iridocorneal touch and forward displacement of lens beneath the iris can be visualized. ASOCT can demonstrate ciliochoroidal detachment and anterior rotation of ciliary body in drug-induced bilateral secondary angle closure. It can image PAS and iridocorneal adhesions in eyes post penetrating keratoplasty with corneal scars.

Radhakrishnan et al. has reported excellent reproducibility of anterior segment parameters with repeated images obtained in the same session with greatest reproducibility for ACD (intraclass correlation coefficient 0.93) in nasal and temporal quadrants (ICC 0.67–0.90). Inferior quadrant showed lower values, whereas the superior quadrant was not imaged. The long-term reproducibility (measured 24 hours apart) showed very good to excellent reproducibility (ICC 0.56–0.93).

Sakata et al.[18] revealed a fair agreement (Kappa 0.40, 95% CI 0.35–0.45), whereas Park et al. showed good agreement between van Herrick's and gonioscopy, but poor agreement with ASOCT. Lavanya et al. using gonioscopy as the reference standard demonstrated a sensitivity and specificity of 0.88 and 0.63, respectively. The low specificity has raised concerns in screening for PAC. Nolan et al.[19] showed that gonioscopy detected 44.4% of eyes with angle closure, whereas ASOCT detected 66.7% eyes. High percentage detected by ASOCT could be related to the ability of ASOCT to evaluate angles in dark.

## Swept source OCT

Swept source OCT (SSOCT) uses a monochromatic tunable fast scanning laser (1310 nm) with scan dimensions of 16 mm × 16 mm × 6 mm achieving high-resolution images of 10 µm (axial) by 30 µm (transverse). With an improved high-speed scanning of 30,000 A scans/second the ACA can be imaged 360° in 128 cross sections (each with 512 A scans) in 2.4 seconds. A three-dimensional display of iris and ACA can be generated with a three-dimensional reconstruction of individual frames.

It measures biometric parameters similar to those measured using ASOCT (Fig. 10). It can visualize both the SS and Schwalbe's line in a high-resolution scan mode improving the precision of measurements and detection of angle closure. TISA was found clinically superior to AOD and ARA, as it includes the entire iris contour and excludes area posterior to SS, thus representing the angle filtering area. The iridotrabecular contact index (ITC) is a quantitative measure of the extent of angle closure across 360° of the

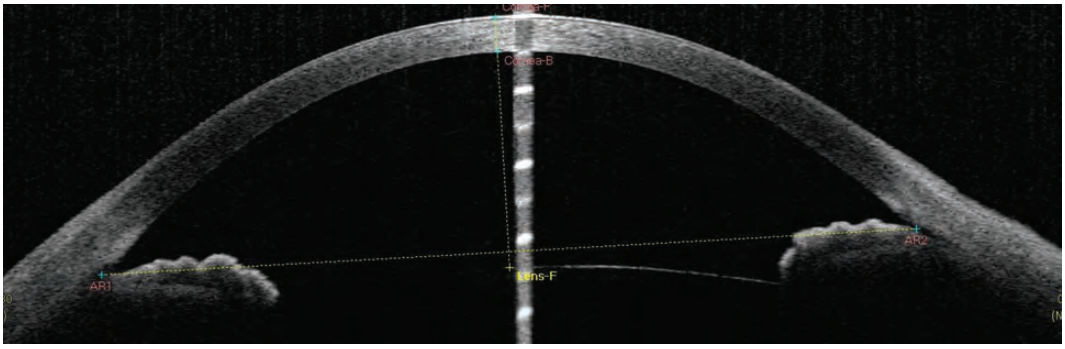


**Figure 10:** SSOCT image showing normal angle structures.

**Table 3** Major Differences between UBM and ASOCT

| | UBM | ASOCT |
|---|---|---|
| Principle | Ultrasound based | Near infra-red light based |
| Technique | Contact procedure | Non-contact procedure |
| Patient posture | Supine position | Sitting position |
| Axial resolution | 25 μm | 18 μm |
| Operator | Requires skilled operator | Easier to use |
| Depth of imaging | Images retro iridial structures like ciliary body, zonules, pars plana, peripheral retina | Unable to image retro iridial structures |
| Imaging | All quadrants can be easily imaged | Difficulty in imaging superior and inferior angles due to eyelids |
| Artefacts | Artefacts due to pressure from eye cup | Artefacts due to light scattering/ refraction, lids |
| Time | Time consuming | Rapid image acquisition |
| Opaque media | Can image in the presence of corneal haze | Better images in eyes with corneal scars, hyphema |

angle expressed as a percentage.[20] It has a moderate agreement and good diagnostic performance for angle closure with gonioscopy as reference.

## Challenges

Inability of the ASOCT to visualize ciliary sulcus and posterior border of ciliary body limits its use in plateau iris and opaque media. Poor identification of SS has been noted in 25% of images. ASOCT is less likely to obtain a good image of superior and inferior angles due to eyelids. Differentiation between appositional and synechial angle closure is not possible due to the inability to perform indentation. Advantages include non-contact technique, higher image resolution, and more precise localization of the position of interest for evaluation compared with UBM. The major differences between the two imaging modalities are highlighted in Table 3.

## Goniophotography

The advantage of gonioscopy includes low cost, visualization of the angle details, ability to indent to differentiate appositional from synechial closure. Goniophotography using a slit lamp-mounted camera can be used to obtain direct images of the angle. However, it is challenging to obtain good-quality images. Gonioscopes with automated 360° gonoiphotography are under development.

## EyeCam

EyeCam provides high-resolution colour images of the ACA using 120° and 130° wide-field lenses. It has a good degree of agreement with clinical gonioscopy, with moderate sensitivity and specificity for detecting angle closure.[21]

Limitations include cost, inability to perform indentation, effect of illumination, non-visualization of corneal wedge or lightly pigmented trabecular meshwork, learning curve and gravity due to supine position. It has moderate to poor reproducibility. Multiphoton lasers and Axicon system (bessel beam microscopy) are under investigation and have been used in porcine eyes to obtain high-resolution images of the ACA.

## Scanning peripheral ACD analyser

Scanning peripheral ACD analyser (SPAC), using slit lamp-based photography, measures ACD at various points in 0.67 seconds at 0.4 mm intervals with a reported sensitivity and specificity of 0.89

and 0.80, respectively, for identifying PAC and PACS in a population-based screening. Measurements correlate strongly with angle assessment by modified van Herrick (sensitivity and specificity of 0.85 and 0.73), UBM and ASOCT for identifying narrow angles. It has a similar sensitivity and specificity as slit lamp-based ASOCT for identifying subjects at risk for PAC in a hospital-based population.[22, 23]

However, it provides information only about peripheral ACD.

## Pentacam

Pentacam allows non-contact quantification of the AC parameters using a rotating Scheimpflug camera to create a three-dimensional image. It can measure ACD (corneal endothelium to anterior lens surface), ACA, ACD and ACV, respectively, with a correlation coefficients of 0.65, 0.85, and 0.81 with gonioscopy. However, this was less than correlation of UBM with gonioscopy.[24, 25] Limitations include inability to identify the SS in many cases due to light scattering affecting angle reproducibility, no direct visualization of ACA, and difficulty in obtaining images in children, elderly, and patients with nystagmus.

## Conclusion

Advances in imaging technology provide invaluable information for the assessment of the anterior segment of the eye aiding in both diagnosis and management of angle-closure disease. However, neither technique is a substitute for detailed clinical examination but a useful adjunct providing information which complements gonioscopy.

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**How to cite this article** Khurana M. and Sushmitha S. Anterior segment imaging in angle-closure disease, *Sci J Med & Vis Res Foun* 2017;**XXXV**:8–14.

# Management of secondary angle closure glaucoma

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## Introduction

Secondary angle closure glaucomas are a separate entity from primary angle closure glaucoma. Secondary angle closure glaucoma is usually caused by multiple factors, so identification of the primary cause is important for appropriate management.

(Table 1)[1]

## Aqueous misdirection syndrome (AMS)

Aqueous misdirection syndrome is a post-ocular surgery secondary angle closure glaucoma characterized by raised intraocular pressure (IOP) with shallowing of central and peripheral anterior chamber (AC) in the presence of patent peripheral iridotomy (PI).

## Clinical features

Aqueous misdirection has been reported post trabeculectomy, post cataract/combined surgery, and post glaucoma valve surgery, following laser peripheral iridotomy (LPI), Argon laser suture lysis, and diode cyclophotocoagulation (CPC). Risk factors include a diagnosis of primary angle closure disease, postoperative shallowing of the AC, and the use of miotic therapy. AMS occurs in 2–4% of eyes undergoing surgery for angle closure glaucoma[2]. Proposed theories include misdirection of aqueous into the vitreous cavity, poor fluid conduction through the vitreous, decreased permeability of the anterior vitreous face, and choroidal expansion.

AMS can occur anytime from first postop day to months after surgery, characterized by elevated IOP, shallow AC, patent PI, normal fundus, and B-scan ultrasonography. Ultrasound biomicroscopy (UBM) shows anteriorly rotated ciliary processes (Fig. 1). IOP may also be normal in patients with functional bleb. It is important to confirm PI patency to rule out pupillary block.

## Management

Medical management consists of hyperosmotic agents, aqueous suppressants, and cycloplegics to reduce vitreous volume and to increase permeability of the vitreous. In pseudophakic and aphakic eyes, YAG laser disruption of the posterior capsule/anterior hyaloid is performed to break the anterior hyaloid face which allows percolation of aqueous through the vitreous. Surgery is necessary if medical treatment fails to reverse the aqueous misdirection within 4–5 days[3].

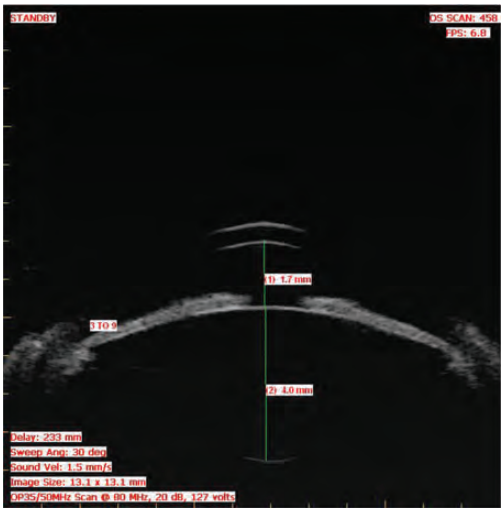


**Figure 1:** UBM showing shallow anterior chamber with anterior rotation of ciliary processes.

The goal of surgery is to establish direct communication between the anterior and vitreous cavity by disturbing the anterior hyaloid face. Various studies have reported resolution of AMS with vitrectomy, hyaloidozonulectomy, iridectomy, and addition of phacoemulsification in phakics[4, 5]. Recurrences occur due to incomplete removal of anterior hyaloid and zonular fibres.

Diode CPC has also been described at an early stage in the treatment of AMS, prior to any surgical intervention in cases where medical treatment has been insufficient to control the IOP[6].

Prophylactic measures include prolonged use of atropine after trabeculectomy surgery (with careful monitoring after this is stopped), avoidance of AC shallowing in the postoperative period (using tight scleral flap suturing) and avoiding use of miotics[7].

## Neovascular glaucoma

This is a severe form of secondary glaucoma characterized by proliferation of fibrovascular tissue in the AC angle which develops secondary to retinal ischaemia[8].

## Clinical features

NVG is commonly associated with proliferative diabetic retinopathy (PDR), central retinal vein occlusion (CRVO), and ocular ischemic syndrome. In response to tissue hypoxia, endothelial cells secrete pro-angiogenic factors such as vascular

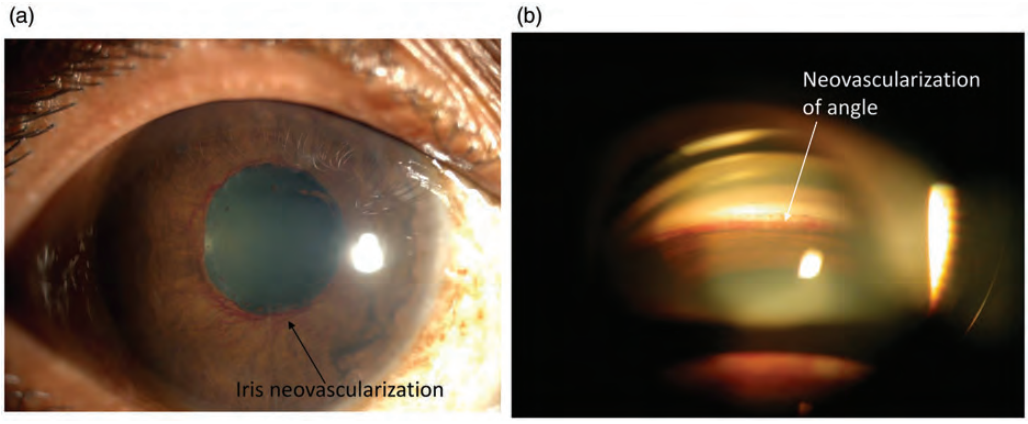


**Figure 2:** (a) Iris neovascularization. (b) Angle neovascularization.

endothelial growth factor (VEGF), basic fibroblast growth factor[9]. These angiogenic factors activate endothelial cells which proliferate and migrate with the formation of new, leaky, fragile blood vessels. Neovascularization may involve the iris (NVI) (Fig. 2A), the angle (NVA) (Fig. 2B) or both, causing formation of the fibrovascular membrane obstructing the aqueous outflow through the trabecular meshwork and resulting in open-angle glaucoma. As the disease progresses, the fibrovascular membrane contracts, leading to ectropion uveae and synechial angle closure.

The clinicopathologic course and treatment may be described in the following stages.

Rubeosis stage—rubeosis of iris usually begins at pupillary margin with IOP being normal at this stage. Gonioscopy should be done to look for NVA, since it can precede NVI. Pan retinal photocoagulation (PRP) decreases the metabolic oxygen demand of retina, thereby reducing the stimulus for release of angiogenesis factors[10].

Open-angle glaucoma stage—neovascularization involves iris stroma and the angle, associated with normal or elevated IOP.

Angle closure glaucoma stage—overlying fibrovascular membrane contracts resulting in synechial closure of the angle and ectropion uveae leading to intractable elevation of IOP and damage to the optic nerve.

## Management

Medical management includes reduction of IOP, treating the underlying cause, and control of inflammation. Prostaglandin (PG) analogues and miotics are avoided as they can worsen inflammation.

Filtration surgery has limited success with risk of intraoperative and postoperative intraocular haemorrhage. Progressive fibrovascular membrane can block the trabeculectomy stoma. PRP performed preoperatively minimizes the risk of these complications, by reducing active neovascularization. Takihara et al.[11] reported success rate of trabeculectomy with mitomycin C (MMC) in NVG of 62.6% at 1 year declining to 51.7% by 5 years. Role of bevacizumab in glaucoma surgery has been studied by several authors[13, 14]. Anti-VEGF agents are proposed to cause reduction of inflammation, vascular permeability, regression of NVI, and NVA. Kobayashi et al.[15] studied the long-term outcome of pre-operative intravitreal bevacizumab for trabeculectomy with MMC in NVG eyes and reported a cumulative surgical success rate of 83.3% at 1 and 3 years in 12 eyes. PRP was found to be an important factor to delay the need for surgery[16].

Glaucoma drainage devices (GDDs) reported a better outcome as the tube evades the fibrovascular membrane. Park et al.[17] have reported success rates of 83.8% at 1 year and 68.5% at 3 years in 31 eyes of NVG following treatment of PDR who underwent GDD surgery. Comparable success rates were noted for non-valved implants.

Ablation of the ciliary body (CB) is reserved for symptomatic poor visual potential eyes.

## Iridocorneal endothelial syndromes (ICE)

ICE is a spectrum of disorders associated with corneal endothelial abnormalities with beaten metal appearance of endothelium and iris abnormalities. Proliferation of abnormal endothelial membrane (ICE membrane) over the corneal endothelium, angle and iris leads to contraction of the membrane causing secondary synechial closure.

## Clinical features

Prevalence of glaucoma associated with ICE ranges from 46 to 82%. It is subdivided into three clinical variants. Essential iris atrophy (Fig. 3A) is characterized by iris atrophy with pupillary stretching and polycoria, Chandler syndrome (Fig. 3B) with severe corneal oedema, mild iris atrophy, and Cogan–Reese syndrome with pigmented nodules over the iris (Fig. 3C). The fine, beaten metal appearance of the cornea and PAS occur in various degrees in all the subgroups.

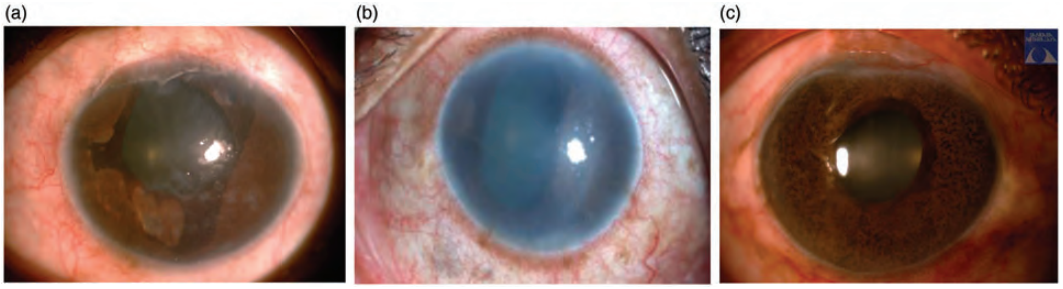


**Figure 3:** (a) Essential Iris atrophy. (b) Chandler's syndrome. (c) Cogan-Reese syndrome.

ICE is a unilateral condition seen most commonly in middle-aged women with IOP spike secondary to synechial angle closure and corneal oedema which later leads to corneal decompensation.

## Management

ICE responds poorly to medication and surgery is eventually indicated in most cases[18]. Trabeculectomy yields 3- and 5-year survival rates of only 44% and 29%, respectively, compared with 71% and 53% with glaucoma drainage implants as noted by Doe et al.[19]. Blockage of the trabeculectomy stoma or tube lumen due to ICE membrane results in surgery failure. Revision surgery is required to relieve the tube block in almost 30% of GDD surgery[20]. Repeated Nd: YAG laser of the tube ostium can help to maintain the tube patency over time. Corneal transplant after glaucoma drainage implant may be required in 30–50% of ICE patients[21].

The extent of corneal involvement and the presence of secondary glaucoma decide the prognosis.

## Inflammatory glaucoma

Glaucoma is one of the serious complications of uveitis which can affect the visual outcome.

## Clinical features

It is seen in ~20% of these patients[22]. Various mechanisms can cause glaucoma such as inflammation leading into 360° posterior synechiae causing iris bombe (pupillary block) (Fig. 4), PAS, and less commonly, non-pupillary block angle closure glaucoma occurs due to CB oedema.

Due to multifactorial aetiology of ocular inflammatory disease, identifying the cause for angle closure is required for further management. Uveitis and IOP elevation needs to be treated together. Corticosteroids are necessary to control the inflammation and aqueous suppressants to control IOP. PG analogues are avoided as it can worsen inflammation and increase the risk of CME.

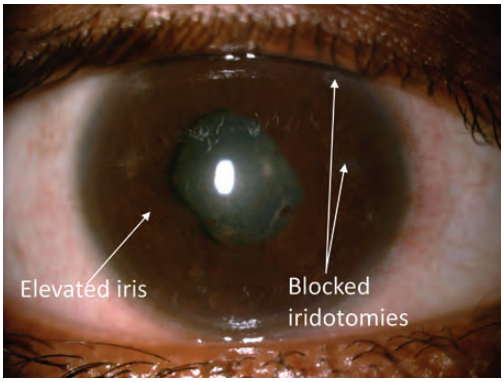


**Figure 4:** Iris bombe with blocked iridotomies.

## Management

Iris bombe needs YAG PI, although fibrin can close small iridotomies in an inflamed eye. In case laser iridotomy is unsuccessful, surgical iridectomy may help if <75% of angle is closed.

If the above measures fail, then surgical intervention is required. Aggressive anti-inflammatory therapy should be given before and after trabeculectomy. Trabeculectomy with MMC has reported survival rates of 90% at 1 year and 79% at 2 years as noted by Nobel et al.[23] compared with AGV's success rate of 77% and 50% at 1 and 4 years, respectively reported by Papadaki et al.[24].

Diode CPC is considered in eyes with low visual potential; however risk of hypotony in uveitic eyes can be high.

## CB cysts

Cysts of the CB neuroepithelium cause anterior displacement of the peripheral iris and may result in angle closure (Fig. 5A). It can cause either acute or chronic angle closure glaucoma, also called as pseudo-plateau iris. Diagnosis and the extent of cysts can be confirmed on UBM[25] (Fig. 5B).

## Management

Laser iridotomy, laser iridoplasty[26], and laser cyclostomy can be used. Therapeutic trial of pilocarpine therapy has also been described.

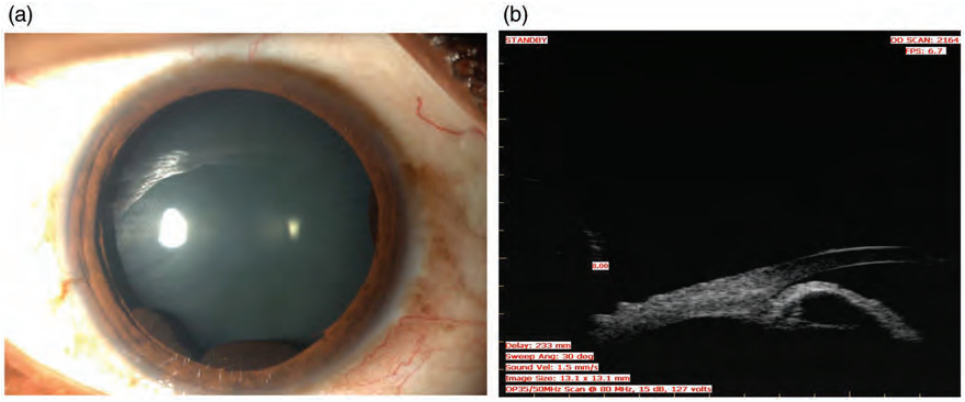


**Figure 5:** (a) Ciliary body cysts seen better with dilation. (b) Ciliary body cysts seen in UBM.

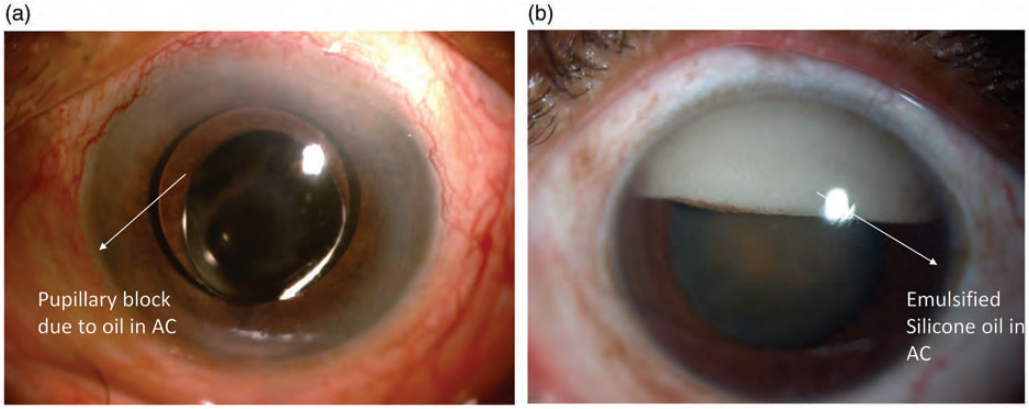


**Figure 6:** (a) Pupillary block due to Silicone oil in anterior chamber. (b) Emulsified Silicone oil in anterior chamber.

## Glaucoma following scleral buckling procedures

Vortex vein compression due to scleral buckle can cause CB swelling which presents as angle closure glaucoma.

## Clinical features-

Anteriorly placed buckle, pre-existing narrow angle, high myopia, old age, and the use of encircling band are predisposing risk factors. Incidence of angle closure post buckle ranges from 1.4 to 4.4%. It usually resolves spontaneously over a period of days or weeks.

## Management

Aqueous suppressants, topical steroids, and cycloplegics form the mainstay of treatment. Cycloplegics help to pull iris lens diaphragm posteriorly and prevent synechiae formation. Miotics are not helpful and YAG PI is contraindicated in these eyes.

Trabeculectomy is difficult due to conjunctival scarring secondary to previous surgery. Nikhil et al.[27] noted a significant reduction in postoperative IOP and number of antiglaucoma medications in patients who underwent AGV post scleral buckle. Diode CPC results are unpredictable.

## Intraocular gas

Air or long-acting gases are injected into the vitreous cavity as their surface tension exerts tamponading effect on retinal breaks. The expansion of these gases during the early postoperative period can lead to IOP elevation

## Clinical features

Incidence of IOP rise following intravitreal SF6 range from 6.1% to 67% and C3F8 18% to 59%[28]. SF6 expands to twice its volume within 24–48 h and stays for 10–14 days whereas C3F8 expands to four times its volume in 48–72 h and stays for 55–65 days[29]. IOP spike is transient, common during gas expansion phase causing angle closure secondary to anterior pushing of iris lens diaphragm.

## Management

Emphasis should be laid to maintain prone position and to abstain from air travel until complete reabsorption of intraocular gas.

If the IOP spike is severe, gas aspiration from the vitreous cavity (via the pars plana) may be indicated. Laser PI is helpful in cases with pupillary block.

Glaucoma drainage device is considered if trabeculectomy is not possible due to conjunctival scarring.

### Glaucoma after silicone oil injection

Silicone oil is used as a vitreous substitute for retinal tamponade. It can produce glaucoma by pupillary block (Fig. 6A), migration of oil into AC (Fig. 6B), inflammation, synechial closure, or by open-angle mechanism. The incidence reported is 6–30%[30].

### Management

Avoiding overfilling of oil and creating inferior prophylactic iridectomy helps in preventing pupillary block in pseudophakic and aphakic eyes. Zonule-lens barrier prevents anterior migration of the silicone oil in phakic eyes.

Medical therapy includes aqueous suppressants, cycloplegics, and corticosteroids. Laser PI is attempted for blocked surgical PI secondary to fibrinous reaction.

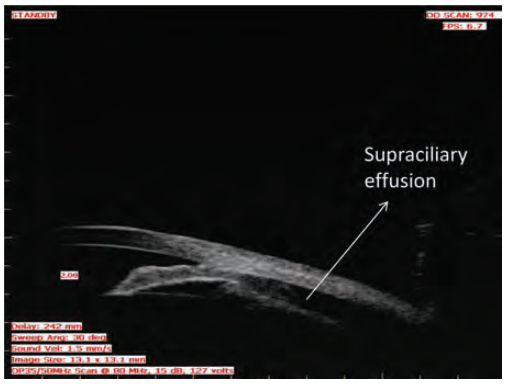


**Figure 7:** UBM showing supraciliary effusion following Topiramate intake causing secondary angle closure.

If silicone oil removal is planned then it is combined with glaucoma surgery. Silicone oil removal carries some risk of retinal detachment[31]. The GDD should be positioned in the inferior quadrant in silicone oil-filled eyes[32].

Diode CPC or endocyclophotocoagulation (ECP) can be used in refractory cases.

### Glaucoma after laser photocoagulation

An elevated IOP may follow extensive laser photocoagulation of the retina.

### Clinical features

Angle closure was observed within hours after PRP in 44% of patients who underwent PRP[32]. It is postulated to be due to anterior displacement of the iris secondary to CB swelling. The incidence of ciliochoroidal effusion following PRP was noted to be 90% on UBM study[33].

### Management

In the majority of cases, the angle closure and resultant IOP rise is transient and asymptomatic. IOP spike usually resolves within 1 month and can be managed with topical medications.

### Drug-induced acute angle closure

Sulfa-based drugs acetazolamide[34], topiramate[35], hydrochlorothiazide, and cotrimoxazole are known to cause secondary angle closure (Fig. 7). Patients present with acquired myopia, uniform shallowing of AC, and raised IOP.

### Clinical features

CB effusion secondary to idiosyncratic response to drug causes anterior displacement of the lens–iris complex leading to secondary angle closure. UBM helps to diagnose CB effusion.

### Management

Management requires discontinuation of drug, reduction of IOP with aqueous suppressants, and cycloplegics. PI and miotics have no role.

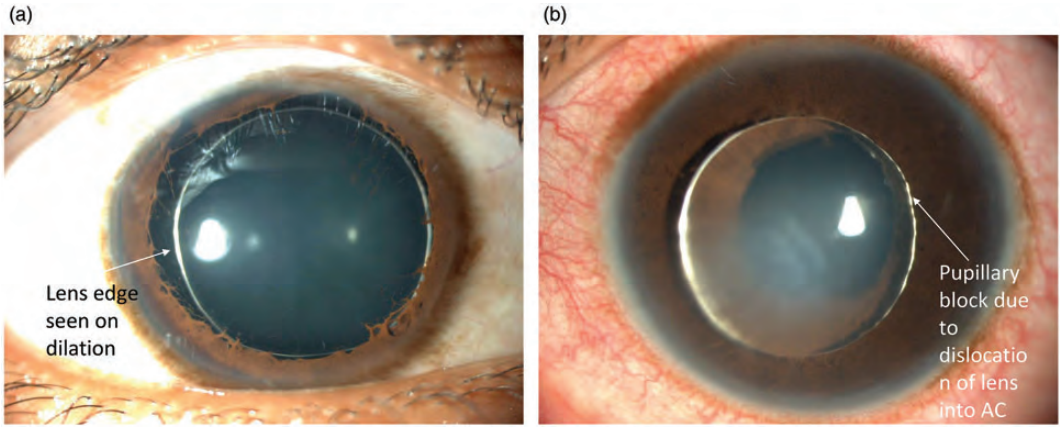


**Figure 8:** (a) Microspherophakia seen on dilation. (b) Microspherophakia causing pupillary block due to dislocation into anterior chamber.

Systemic and topical carbonic anhydrase inhibitors should be avoided.

## Nanophthalmos

It is a condition in which the eye ball is of normal shape but small in size. Angle closure glaucoma is usually seen in the middle age. (Dealt in detail in case reports).

## Microspherophakia

Lens is usually smaller in size, spherical in shape with increased anteroposterior diameter[36]. When angle closure is associated with high myopia, possibility of microspherophakia should be kept in mind. With dilation, the pupil entire edge of the lens can be visualized at the slit lamp (Fig. 8A). (Dealt in detail in case reports)

## Phacomorphic glaucoma

In phacomorphic glaucoma, angle closure occurs due to swelling of the lens causing intumescent cataract. After IOP control and reduction of inflammation, cataract extraction is the definitive treatment as it removes the cause for angle closure glaucoma; however, it is combined with trabeculectomy if the duration of the attack lasts for more than 3 weeks. There is increased risk of expulsive haemorrhage, positive pressure, and zonular dialysis due to high IOP in these eyes. Manual small incision surgery has been studied to provide effective IOP control and good visual recovery in phacomorphic glaucoma eyes[37].

## Conclusion

A careful history, astute clinical examination, and when necessary anterior segment imaging such as UBM aid in recognizing the aetiology for secondary angle closure. Identifying the cause early and timely institution of appropriate therapy helps in improving the visual outcome and reducing the ocular morbidity.

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**How to cite this article** Parivadhini A. Management of secondary angle closure glaucoma, *Sci J Med & Vis Res Foun* 2017;**XXXV**:15–21.

# Microspherophakia with secondary glaucoma

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## Introduction

Microspherophakia is characterized by an increased anteroposterior diameter and a reduced equatorial diameter of the crystalline lens leading to a spherical configuration. It is seen either as an isolated anomaly or along with systemic disorders like Weill–Marchesani syndrome, Marfans disorder, Alports syndrome, Klinefelter syndrome, and Mandibulofacial dystostosis.[1] We report a case of isolated microspherophakia with secondary angle closure glaucoma.

## Case

A 23-year-old woman presented with complaints of gradually progressive diminution of vision in the left eye since 1 year. It was associated with occasional headache. She was on topical intraocular pressure (IOP) lowering agents for the past 5 months.

The best corrected visual acuity in the right eye was 6/18 [−6.00 diopter sphere/−1.00 cylinder at 40] and 6/24 [−6.00 diopter sphere/−1.25 cylinder at 70°] in the left eye. IOP recorded with Goldmann applanation tonometer was 20 and 40 mmHg for the right and left eye, respectively.

Slit lamp examination showed a shallow anterior chamber [Van Herick grade 2] in both eyes. Gonioscopy revealed 360° of angle closure in both eyes (Fig 1.1). Relative afferent pupillary defect was noted in the left eye. Ultrasound biomicroscopy (Fig 1.2) revealed a decreased anterior chamber depth [1.3 mm OD, 1.5 mm OS], an increased lens thickness [4.3 mm OU], and a reduced equatorial lens diameter [6.3 mm OD, 6.2 mm OS] suggestive of microspherophakia.[2] Peripheral laser iridotomy

**Figure 1.1:** (a) Gonioscopy showing synechial angle closure. (b) Slit-lamp photo showing shallow central anterior chamber.

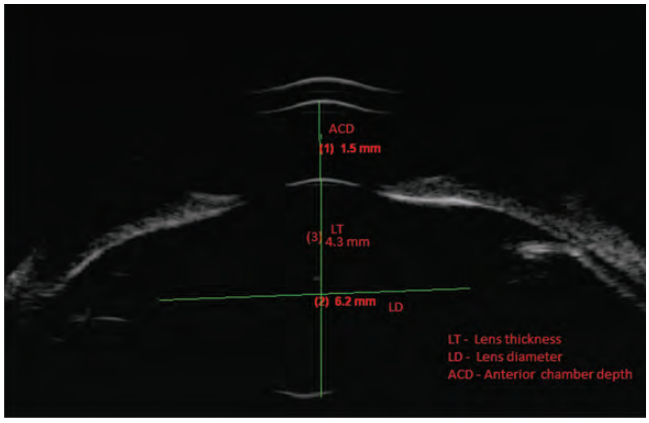


**Figure 1.2:** UBM of left eye showing reduced equatorial lens diameter and shallow anterior chamber.

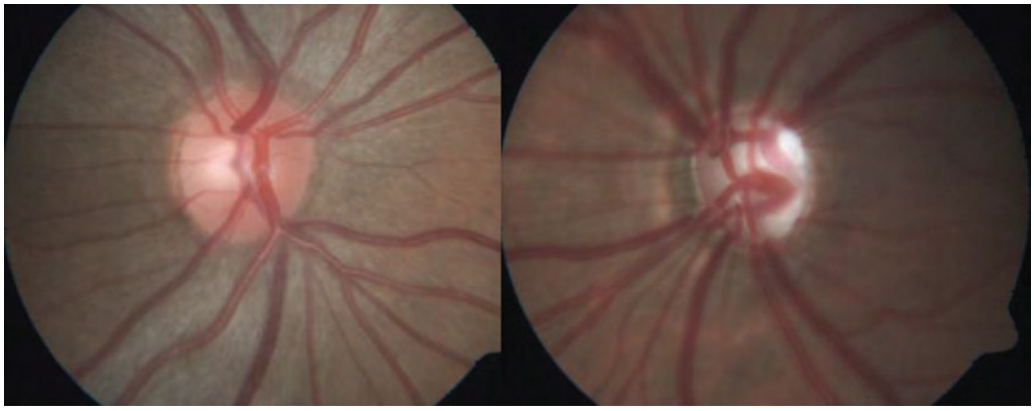

**Figure 1.3:** Optic nerve head (a) Right eye with a healthy disc. (b) Left eye showing bipolar rim thinning.

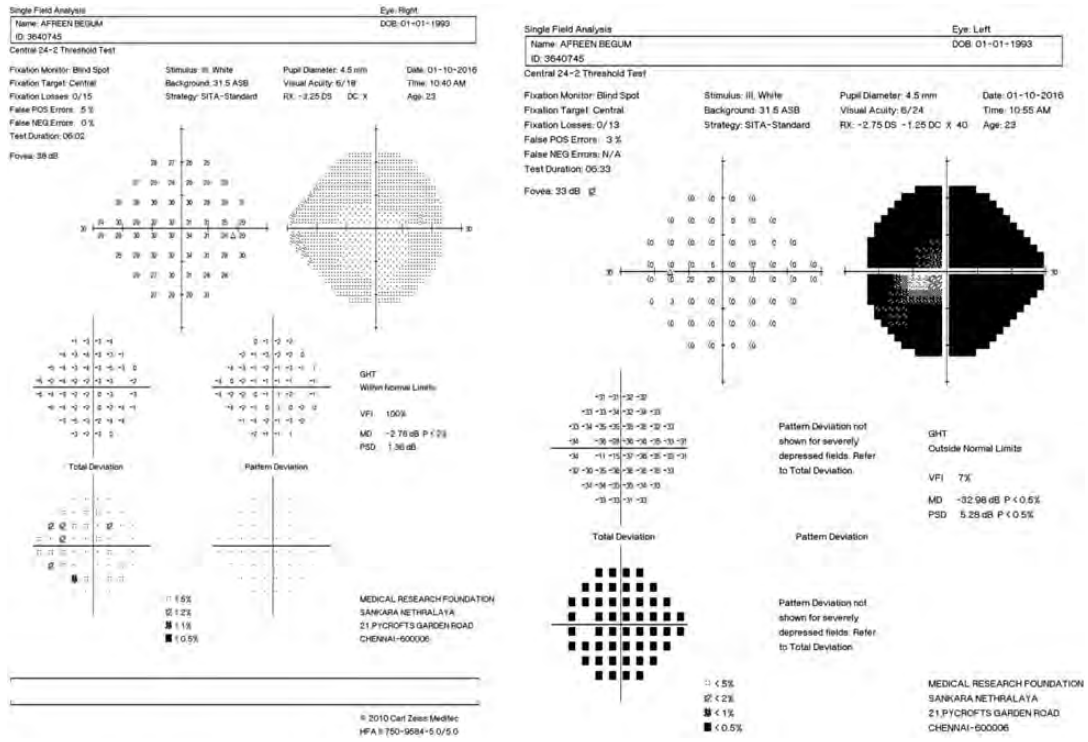


**Figure 1.4:** Humphrey's visual field.

was performed in both eyes. The optic nerve head was healthy in the right whereas there was advanced cupping in the left eye (Fig 1.3). There was advanced field loss in the left eye (Fig 1.4). Since IOP was not under medical control she was advised surgical intervention in the left eye. Post-dilatation, lens edge was visible all around (Fig 1.5)

## Discussion

Microspherophakia is clinically characterized by a triad of angle-closure glaucoma, small spherical crystalline lens, and lenticular myopia. Visual compromise in patients with microspherophakia is attributed to refractive error or secondary glaucoma. Mechanism of glaucoma is due to pupillary

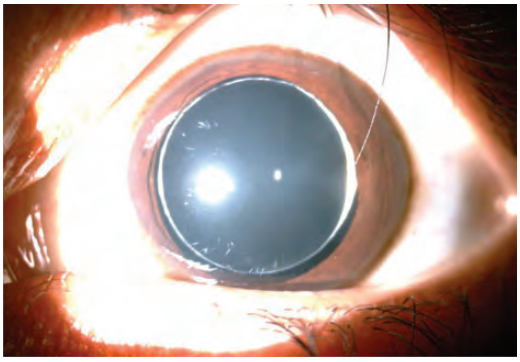

**Figure 1.5:** Lens edge visible all around after dilation.

block and/or peripheral anterior synechiae. Acute angle closure can result from pupillary block due to anterior dislocation of the crystalline lens when associated with weak zonules. Such recurring pupillary block attacks results in synechial angle closure resulting in chronic angle closure. Initial treatment consists of relieving the pupillary block component by laser iridotomy followed by medical management of elevated IOP. Indications for surgery include poor control of IOP and/or lens dislocation. Scleral fixation of the intraocular lens may be required[3, 4].

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**How to cite this article** Sankaranarayanan N and Ganesh N. Microspherophakia with secondary glaucoma, *Sci J Med & Vis Res Foun* 2017;**XXXV**:22–24.

# Epidemiology of primary angle closure disease—the chennai glaucoma study and the chennai eye diseases incidence study

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## Introduction

Glaucoma is considered to be the leading cause of irreversible blindness worldwide. Primary angle closure disease (PACD) is a subtype of glaucoma that predominantly affects Asian ethnicity more than Caucasians and Africans.[1–3] Estimating the prevalence and identifying the risk factors for glaucoma are the goals of population-based studies. The information from these studies is used to address treatment protocols and health-care policies. The Chennai Glaucoma Study (CGS) was one such population-based cross-sectional survey designed to estimate the prevalence of glaucoma in a rural and an urban population aged 40 years and above from south India. A sample size of 4758 was arrived for an 85% response rate for an assumed 3% population prevalence of glaucoma with a relative precision of 25% and a design effect of 2. The CGS was conducted between June 2001 and May 2004 by enumerating a cohort of 9600 (4800 each from rural and urban arms) individuals. The rural study arm consisted of residents from 32 consecutive neighbouring villages spread over two districts of the state of Tamil Nadu, whereas urban study population consisted of residents from Chennai city using a multistage random cluster sampling procedure and 960 subjects from five divisions were enumerated. During enumeration demographic information was collected by household questionnaire and the eligible subjects were invited to come to the base hospital that was created exclusively for the study of detailed ophthalmic examination.[4] From this cohort, 7774 subjects (rural:urban—3924:3850) were examined. Incidence studies provide information on the true risk of developing new disease over a period of time. It also helps in exploring associations between preexisting factors at baseline and the development of the disease. The Chennai Eye Disease Incidence Study (CEDIS) was conducted to find the incidence of eye disease in the same cohort, 6 years from the baseline visit (2007–2010). Five thousand four hundred and thirty-two subjects were eligible; out of this 4421 (rural:urban 2510:1911) subjects were examined at the base hospital.

Glaucoma was diagnosed based on structural and functional evidence of glaucomatous damage as per the International Society Geographical and Epidemiologic Ophthalmology (ISGEO).[5] Glaucoma was classified into three categories based on three levels of evidence. In category 1, diagnosis was based on structural and functional evidence which required a cup disc ratio (CDR) or a CDR asymmetry equal to or greater than the 97.5th percentile for the normal population or a neuroretinal rim width reduced to 0.1 CDR (between 10 and 1 o'clock or 5 and 7 o'clock) with a definite visual field defect consistent with glaucoma. A glaucomatous field defect was diagnosed on threshold visual field examination (SITA standard 24–2) of the central 24°, if the glaucoma hemifield test was outside normal limits and three or more abnormal contiguous non-edge points (except the nasal horizontal meridian) were depressed to the P5% level. Reliability criteria were as recommended by the instrument's algorithm (fixation losses 20%; false positive and false negative, 33%). Category 2 was based on advanced structural damage with unproved field loss. This included those subjects in whom visual fields could not be done or were unreliable, with CDR or CDR asymmetry equal to or greater than the 99.5th percentile for the normal population. Lastly, category 3 consisted of persons with an intraocular pressure (IOP) greater than the 99.5th percentile for the normal population, whose optic discs could not be examined because of media opacities. Angle closure disease was sub-classified into primary angle closure suspect (PACS), primary angle closure (PAC), and primary angle closure glaucoma (PACG) based on structural and functional evidence of glaucomatous damage as per ISGEO. PACS was defined as an eye in which the posterior trabecular meshwork was not visible for >180° on gonioscopy; PAC, an eye with PACS along with peripheral anterior synechiae and/or elevated IOP without optic neuropathy; and PACG, the presence of PACS combined with clinical evidence of glaucoma.

## Prevalence of PACD

The prevalence of PACS[6, 7] (6.3%, 95%CI, 5.51–7.03 versus 7.24%, 95%CI, 6.38–8.02) and PACG (0.87%, 95% CI 0.58 to 1.16 versus 0.88%, 95%CI, 0.60–1.16) was similar in the rural and urban populations. However, PAC was found to be significantly higher in the urban population (2.75%,

95%CI, 2.01–3.49 versus 0.71%, 95%CI, 0.45–0.98). One possible reason for this difference could be earlier cataract surgery in the rural population, which either masks PAC or prevents progression from PACS to PAC. The disease was found to be silent and chronic and none had any evidence of acute attack. Age was found to be a significant risk factor for PACD in both populations, with increasing age the prevalence of the disease increased across both populations. Female gender is considered to be a risk factor for PACD, our rural population had female preponderance whereas in our urban study population this trend was seen only in PACS and PAC groups. One possible reason could be due to the small numbers. Diabetes mellitus and hyperopia were associated with PAC and PACG only in our urban population. Association of diabetes in the urban population can be attributable to the higher prevalence of diabetes in our urban population. We found a definite association between nuclear sclerosis and myopia in the rural population; this secondary myopia could have masked a hyperopic refractive error and resulted in lack of association of hyperopia with angle closure in the rural population. PACG-related blindness was more common among urban subjects (5.9%) than rural subjects (2.9%). None of the PACG subjects in the rural population were aware of their disease, whereas in the urban population 14.7% (5 out of 34 subjects) were diagnosed to have glaucoma earlier. Among the five subjects with PACG 2, 40% were diagnosed to have open-angle glaucoma, this possibly can be explained by failure to do gonioscopy as part of glaucoma clinical evaluation. Using the biometry we found that eyes with PACD seem to be associated with shallower anterior chambers, thicker crystalline lenses, and shorter axial lengths.[8] In eyes with pseudoexfoliation deposits of PEX material on the zonules results in zonular weakness. This can lead to an anterior shift of the lens and occludable angle. In the CGS we observed 8.3% of eyes with pseudoexfoliation having occludable angles.[9]

## Incidence of PACD

We reported the 6-year incidence of PACD from the CEDIS; the incidence of PACD was defined as the development of PACD during the follow up in subjects without PACD at baseline in a phakic eye.[10] Out of 4421 subjects were evaluated in the CEDIS,134 subjects (M:F 62:72, rural: urban 82:52) were diagnosed to have any form of PACD, incidence of 4.0% (95% CI, 3.3 to 4.7, rural 2.5%, 95%CI, 1.8 to 3.2,urban 1.6%, 95%CI, 0.9 to 2.2). Assuming a linear incidence of PACD the annual incidence was 0.7%. The incidence of the three sub-types of PACD is as follows: PACS—88 subjects (2.6%, 95%CI, 2.1–3.2, M:F 36:52, rural: urban 56:32), PAC—37 subjects (1.1%, 95%CI, 0.7–1.5, M:F 24:13,rural: urban 21:16) and PACG —9 subjects (0.3%, 95%CI, 0.1–0.4, M:F 2:7, rural: urban 5:4). In the nine subjects with PACG, the diagnosis was based on category 1 in five subjects, category 2 in three subjects, and category 3 in one subject. Three subjects had bilateral disease and none were blind.

Higher intraocular pressure, increased lens thickness, shorter axial length, shallow anterior chamber depth, an anteriorly positioned lens and hyperopia were found to be the baseline risk factors for PACD in 6 years. There was an inverse relationship between the incidence of PACD and the cataract surgery rates. The incidence of PACD peaked in the 50–59 years age group and declined after 60 years of age, whereas the cataract surgery rate increased exponentially after the age of 60 years. In the past, a national survey was conducted in 15 states of India that includes the state of Tamil Nadu where our study was carried out, and it was found that the state of Tamil Nadu has a cataract surgery rate of 14.7% and a surgical coverage of 82.8%. In terms of actual number of surgeries Tamil Nadu is listed among the top five states. The decline in PACD incidence after 60 years of age in our study is mainly due to this increase in cataract surgery rates in our study population and due to the successful cataract blindness program.

The incidence of PACD increased in eyes shorter than 21 mm, those with an ACD <2.5 mm and a lens thickness of >5.5 mm. The lens plays an important role in pathogenesis of angle closure disease either because of increased thickness or due to a more anterior position. Both these factors can cause crowding of the anterior chamber angle in eyes with a smaller anterior segment and result in greater predisposition to PACD. But the exact relationship between angle closure disease and lens thickness or lens position is not very clear. Theoretically, the thickness and position of the lens should alter the angle recess width, but the association was found to be equivocal. One possible explanation proposed for this inconsistency is the inability to control the accommodation while measuring the lens thickness. In spite of the limitation we suggest that our biometry data can be used as a risk factors guide for PACD development. We also reported the risk of cataract progression among PACS subjects 6 years after they underwent laser peripheral iridotomy (LPI).[11] One hundred ninety subjects who had LPI for PACS at baseline were examined in the CEDIS. We found there was a significant cortical cataract progression in 6 years following LPI for PACS (OR 1.6, 95%CI 1.1–2.3, p = 0.007).

## Implications of study findings

There is significant PACD in the population above 40 years of age. Like open angle glaucoma PACD

also is silent and chronic. Majority of it is undiagnosed glaucoma. This suggests that a comprehensive eye exam is essential for diagnosis of glaucoma that includes appropriate application of gonioscopy to diagnose angle closure. Older age and shorter eyes are the risk factors for the disease and needs closer watch for development of angle closure in those eyes. Laser iridotomy for PACS can cause cortical cataract with time, risk, and benefit of the LPI should be discussed in offering LPI for asymptomatic PACS. Cataract surgery seems to have an effect on incidence of PACD, with improvements in cataract surgical techniques and outcomes probably eyes that have PACS and PAC with significant cataract will benefit from cataract surgery.

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**How to cite this article** Vijaya L., Asokan R., Najiya Sundus K.M. and Mazumdar D. Epidemiology of primary angle closure disease—the chennai glaucoma study and the chennai eye diseases incidence study, *Sci J Med & Vis Res Foun* 2017;**XXXV**:25–27.

# Genetics of angle closure disease

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Primary angle closure glaucoma (PACG) has long been suspected to have a significant genetic risk because of its associations with race, gender, and in families.[1] This was first reported from Eskimos among whom the presence of PACG in an individual conferred a 3.5 times greater risk among first degree relatives. From a study in South Indian siblings of angle closure patients had more than 33% having PAC and siblings of those with PAC/PACG patients had a >10% risk of prevalent PAC/PACG. [2]Genetic analysis in a large family with nanophthalmos, an extreme variant of angle closure, resulted in the identification of the gene nanophthalmos 1 (NNO1) on chromosome 11.[3] In spite of the high heritability of PACG this traditional approach of looking for genetic mutations associated with PACG in families was unsuccessful suggesting that PACG was a complex disease with multiple factors playing a role in determining whether a person would develop PACG. When associations with the known genes for primary open-angle glaucoma were studied, no association was found with MYOC in multiple studies. A positive association was seen with CYPB1 (commonly associated with congenital glaucoma) in patients of Chinese, Indian, and Canadian origin.[4–6]

With the completion of the human genome project a comprehensive map of the human genome became available and this enabled the use of genome-wide association studies (GWAS) in medicine. No significant genetic markers for angle closure disease were identified using relatively small sample sizes. However, two multicentre multinational studies that recruited a large number of persons with angle closure disease were successful in identifying eight new loci for PACG. The first included 1854 PACG cases and 9608 controls across five sample collections in Asia. The replication cohort consisted of 1917 PACG cases and 8943 controls collected from a further six sample collections. Significant associations were found for three new loci: rs11024102 in PLEKHA7 (per-allele odds ratio (OR) = 1.22; $P = 5.33 \times 10(-12)$), rs3753841 in COL11A1 (per-allele OR = 1.20; $P = 9.22 \times 10\ (-10)$), and rs1015213 located between PCMTD1 and ST18 on chromosome 8q (per-allele OR = 1.50; $P = 3.29 \times 10(-9)$).[7]

In a combined dataset of patients recruited from Nepal and Australia, COL11A1 rs3753841 and rs1015213 showed significant associations with $p$-values of 0.009 and 0.004, respectively. Single-nucleotide polymorphism (SNP) PLEKHA7 rs11024102 showed suggestive association with PACG ($p$-value 0.035) and no association was found with rs3788317. The association of rs1015213 (PCMTD1-ST18) as a risk factor for PAC was further replicated in another South Indian population.[1]

In a more recent publication of 10,503 PACG cases and 29,567 controls across Asia, Australia, Europe, North America, and South America the earlier three loci were confirmed and an additional five new genetic loci at EPDR1 rs3816415, CHAT rs1258267, GLIS3 rs736893, FERMT2 rs7494379, and DPM2-FAM102A rs3739821 were identified.[8]

Biometric factors such as shorter axial length and anterior chamber depth are known to be risk factors for PACG.[9] Some of the genetic associations are probably associated with biometric parameters that influence disease risk. Collagen has been reported to be potentially involved in glaucoma pathogenesis. Alterations in collagen deposition can influence the biomechanical and remodelling abilities of the sclera resulting in an effect on axial length a known risk factor for PACG. In addition to this it could impact the biomechanical properties of the scleral ring which can influence the optic nerve head's biomechanical responses to intraocular pressure (IOP). COL11A1 encodes one of the two α chains of type XI collagen, which is highly expressed in the scleral tissue and is one of the genes associated with PACG.[1] From the EPIC-Norfolk Eye Study cohort from UK an association was found between SNP rs1015213 (PCMTD1-ST18) rs1015213 and anterior chamber depth. In a larger cohort of over 7000 eyes there was nominal evidence of association and a shallower anterior chamber depth (ACD) when all data were meta-analyzed ($\beta = -0.033$, $P = 0.021$). However, when multiple testing was considered, the observation was non-significant.[10] In another recent GWAS study from a larger Asian cohort ABCC5 was found to influence ACD and increase the risk of PACG development.[11] The list of genes associated with PACG and other biometric factors are shown in Table 1.

The extracellular composition of the sclera is modified and influenced by intraocular pressure fluctuations. Enzymes such as the matrix metalloproteinases participate in this tissue remodelling response. Matrix metalloproteinase-9 (MMP 9) has been reported to be associated with a GWAS from a Chinese population as well as an Australian Caucasian population for PACG and with acute anterior chamber in a Taiwanese and Pakistani cohort.[12–14] However, no association was reported for Singaporean Chinese.[15]

**Table 1:** List of genes associated with angle closure disease and nanophthalmos

| Gene | PACG | Nanophthalmos | Axial length | ACD | Others |
|---|---|---|---|---|---|
| *NNO1* | | Yes | | | |
| *PLEKHA7, COL11A1, PCMTD1, ST18, EDPR1, CHAT, GLIS3, FERMT2 and DPM2-FAM102A* | Yes | | | | |
| *MMP9* | Yes | | | | |
| *MTHFR* | Yes | | | | ECM remodelling |
| *MFRP* | Yes | | | | |
| *MFRP* | | Yes (AR) | Yes (short) | | |
| *CHX10* | Yes | | | | |
| *HGF* | Yes | | | | |
| *PRSS56* | Yes | | | | Posterior microphthalmia |
| *ABCC5* | Yes | | | Yes | |
| *MYOC* | Yes | | | | |
| *CYP1B1* | Yes | | | | |
| *eNOS* | Yes | | | Yes | |
| *HSP70* | | | | | |
| *SPARC* | Yes | | | | IOP regulation |
| *CALCRL* | Yes (acute) | | | | |

Changes in iris characteristics have also been implicated in angle closure disease. The iris is known to lose less volume while dilating in PACG as opposed to normal eyes.

In two Chinese patient cohorts CALCRL (calcitonin receptor-like) polymorphisms have been associated with acute angle closure but not chronic angle closure disease.[16] CALCRL belongs to a group of receptors mediated by G proteins that activate adenyl cyclase. Overexpression of this gene results in relaxation of the sphincter pupillae, closure of the anterior chamber angle leading to obstruction of the aqueous outflow.[17–19] This is inherited through mitochondria. Since each cell contains several genetically heterogeneous mitochondria, which are susceptible to age and stress induced mutations, mitochondria in ocular and blood lymphocytes (used for genetic analysis) may show different characteristics.[1, 20]

The newer genetics findings have confirmed the heritable nature of angle closure disease and have led to molecular insights into clinical parameters associated with angle closure disease. However, with the exception of risk counselling for families of patients with angle closure glaucoma there are no genetic tests that can provide disease risk currently.

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**How to cite this article** George R. Genetics of angle closure disease, *Sci J Med & Vis Res Foun* 2017;**XXXV**: 28–30.

# Nanophthalmos - Preparing for the challenge

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Nanophthalmos, a rare condition, is an important cause of secondary angle closure, especially in young adults. Cataract surgery in a nanophthalmic eye is challenging. Here we highlight the meticulous planning that is imperative to an uncomplicated outcome.

This is a report of a 40-year-old lady who came with complaints of decreased vision in both eyes since 1 year. She gave history of wearing thick glasses since childhood. On examination, she had small eyes and visual acuity was hand movements close to face. She had shallow anterior chamber with dense brunescent cataracts in both eyes precluding fundal view. The intraocular pressure (IOP) was 12 mmHg by Goldmann applanation tonometry. Gonioscopy showed appositional closure. Ultrasound Bscan showed increased retinochoroidal scleral thickness. Biometry findings are summarized in Table 1. Ultrasound biomicroscopy showed anteriorly placed ciliary body and no supraciliary effusion. Findings were suggestive of nanophthalmos. The patient underwent Laser peripheral iridotomy bilaterally followed by phacoemulsification with single-piece hydrophobic acrylic intraocular lens (40D) implantation under local anaesthesia, with prophylactic anterior lamellar sclerectomy with sclerotomy in two quadrants. Intraoperative and postoperative period was uneventful.

**Table 1**

| | OD | OS |
|---|---|---|
| Bscan axial length | 15.7 mm | 15.1 mm |
| Bscan RCS thickness | 2.7 mm | 2.7 mm |
| UBM lens thickness | 5.1 mm | 5.3 mm |
| UBM ACD | 2.1 mm | 1.8 mm |
| IOL master axial length | 15.77 mm | 15.83 mm |
| Hoffer Q (emmetropia) IOL power, 118.4 A constant | +59 D | +58 D |

## DISCUSSION

It is important to categorize the 'Small eye' phenotype as each has different implications—clinical and surgical

(a) *Simple microphthalmos:* Short axial length (>2 SD smaller than age-based normative) and no other ocular malformations.[1] (b) *Relative anterior microphthalmos:* Normal axial length with disproportionately small anterior segment.[1] (c) *Nanophthalmos:* Short axial length (<20.5 mm), shallow anterior chamber depth <2.2 mm, normal or increased lens thickness, thickened sclera, choroid >1.7 mm posteriorly with an increased predisposition for uveal effusion.[1]

*Pathophysiology* of effusion: Thickened sclera compresses vortex vein, impeding normal choroidal venous drainage.[2] Transcleral protein egress is hampered and lack of lymphatic drainage results in suprachoroidal fluid retention.[3]

*Common complications*: Posterior capsular rupture (4–11.7%), aqueous misdirection (0–25%), suprachoroidal haemorrhage(0–2.7%), prolonged anterior uveitis (2.3–11.8%), and uveal effusion (9.3%).[4, 5] The variation in range is probably due to differences in the axial length considered by different authors for their definition of nanophthalmos.

*Preoperative considerations:* Axial length measurement by partial coherence interferometry/optical low coherence reflectometry is preferred over ultrasound biometry. Hoffer *Q* formula is more accurate for axial length <22 mm[6]. Peripheral iridotomy is warranted in the case of

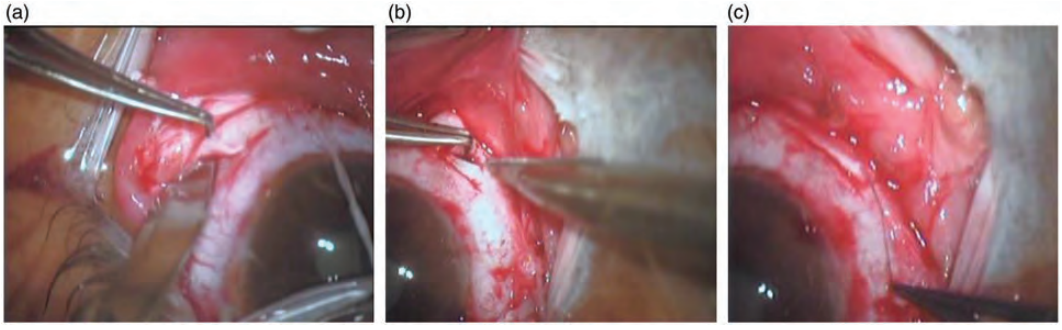


**Fig. 1** (a) 4–5 mm from the limbus, a 5 mm partial thickness rectangular scleral flap is raised. (b) V-shaped full-thickness scleral cut down is made under the flap to expose the suprachoroidal space. (c) The flap is then replaced and sutured with 10-0 nylon.

narrow angles and/or elevated IOP. Intravenous mannitol helps reduce vitreous pressure.

*Intraoperative considerations:* General anaesthesia is preferred, as it does not increase orbital volume and adequately relaxes rectus muscle tone. High-molecular-weight OVDs aid capsulorrhexis. Many surgeons do two/four quadrant prophylactic sclerotomies which serves to drain uveal exudation and relaxes scleral tension, indirectly decompressing the vortex veins. Flap suturing helps reinforce globe integrity.[7]

*Postoperative care:* Postoperative concerns include correction of residual refractive error by glasses and contact lenses, inflammation control and prevention of aqueous misdirection with use of strong cycloplegics.

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**How to cite this article** Kadambi S.V. and Krishnamoorthy S. Nanophthalmos - Preparing for the challenge, *Sci J Med & Vis Res Foun* 2017;**XXXV**:31–32.

# Angle closure disease

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There has been a huge increase in the number of publications regarding angle closure glaucoma. Approximately half the 5700 pubmed indexed publications on angle closure glaucoma have occurred in the last decade.

This is partly because of the realization of the large numbers of persons with primary angle closure disease (PACD) worldwide and the understanding that primary angle closure glaucoma (PACG) is much more likely to cause blindness compared with primary open-angle glaucoma.[1] The modified definitions of angle closure disease have resulted in the standardization of diagnostic criteria and have made studies across the globe comparable.[2] While this simplified classification has put less emphasis on the mechanism of angle closure thus resulting in potentially grouping diseases with dissimilar etiologies the benefits of having a simple less subjective classification outweigh the limitations.

Along with the realization of the burden of disease came the revolution in genetic technologies that have for the first time resulted in identifying some genetic risk factors for PACD. While these do not explain a huge proportion of angle closure disease they do help in the understanding of the pathophysiology of disease which could potentially result in biomarkers or therapeutic targets being identified.

The other major explosion in recent year has been in imaging of the anterior segment. The UBM still remains the Gold Standard for understanding the pathophysiology of angle closure disease and improved our understanding of the lens–ciliary body complex and its role in disease. While the OCT has improved our understanding of the RNFL in glaucoma manifold, the anterior segment OCT has not been as effective. The potential for over diagnosis with the ASOCT may be addressed with newer versions of the technology; the humble gonioscope remains the most important tool for the glaucoma specialist.

The influence of changing environmental factors on angle closure disease is something that we are still trying to assess. A study from Singapore reported that the risk of acute angle closure was highest when ambient temperatures were high.[3] This contradicted a study from Finland that reported that highest rates of angle closure were noted in winter and autumn and that the number of hours without sunshine was positively associated with the incidence of acute closed angle glaucoma.[4]If one assumes that people are more likely to spend time indoors when the ambient temperatures are hot then the potential mydriaisis indoors or in the darker skies at higher latitudes is the common thread in all these environments. With angle closure predominantly affecting less developed economies that are now in a phase of rapid economic growth with increasing numbers of person working indoors changes in the rates of acute closure are possible.

Height is known to be associated with axial length, with taller persons having longer eyes.[5] Improved nutrition and increasing GDP is also associated with increases in height.[6] The impact of these changes on angle closure disease are yet unknown. [7]Would it result in a decreased risk of angle closure (which is associated with shorter eyes) or would the crowded anterior segment remain unaltered? The other factor is increased near work in childhood. This is a known risk factor for myopia again associated with longer axial lengths. With more children undergoing schooling and longer hours spent indoors or with personal electronic devices would angle closure spare the next generation.[8]

The high rates of cataract extraction will also work to decrease the risk of angle closure in eyes with shallow anterior chambers. However, even with recent evidence there is still no justification for clear lens extraction in all eyes with angle closure/PACG.

With large proportions of the populace at different stages of economic growth it is unlikely that we may see dramatic changes in the rates of angle closure in the short term during which disease burden will continue to grow. In the long term while patterns of disease may change, the effect of the myriad changing environmental parameters is difficult to predict.

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**How to cite this article** George R. Angle closure disease, *Sci J Med & Vis Res Foun* 2017;**XXXV**: 33–34.

SANKARA NETHRALAYA

# SANGAM 2017

## A SANKARA NETHRALAYA GLAUCOMA MEET

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