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. 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). 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.

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.

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

**Figure 2:** Primary angle closure on gonioscopy.
(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.5 described this technique in a prospective case series of 10 eyes with AcACC. The mean IOP reduced from 66.6 ± 9.1 to 15.1 ± 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 synechial/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
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 phacoemulsification alone (Group I, 35 patients) or to combined PKE with trabeculectomy with adjunctive Mitomycin 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% CI −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 iridocorneal 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 framework 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.

References