

# An Evidence-Based Approach to Phacomorphic Glaucoma

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## Abstract

Phacomorphic angle-closure is a secondary angle-closure caused by a mature cataract. The definitive treatment is cataract extraction. The objective of this review is to highlight the epidemiology, risk factors, and diagnostic and treatment strategies to optimize final visual acuity.

## Background

Cataract maturation is associated with anteroposterior lens diameter increase which, in some patients, particularly those with anatomically narrow anterior chambers, can lead to pupillary block and peripheral iridotrabeular apposition [1,2]. In other patients with previously deep anterior chambers and wide open angles, the cataract may become so swollen that it physically pushes the iris anteriorly, leading to iridotrabeular apposition [3]. The role of pupillary block in the pathophysiology of phacomorphic angle-closure is still uncertain. Limited evidence suggests that early in the course of the disease, the lens swelling is only large enough to cause pupillary block [4]. Progressive enlargement of the lens may then lead directly to peripheral iridotrabeular apposition. When the iridotrabeular apposition raises the intraocular pressure (IOP) enough to cause the signs and symptoms of an acute attack of secondary angle-closure glaucoma, it is called acute phacomorphic angle-closure. Historically this entity has been labeled phacomorphic glaucoma. As pointed out by Tham, however, using the word glaucoma implies an optic neuropathy [5]. Since many patients (as will be analyzed below) do not show glaucomatous optic neuropathy following the resolution of the acute angle-closure, it may be more accurate to call this disease acute phacomorphic angle-closure. The dense white cataract labeled as a mature cataract is an advanced form of cortical cataract, with widely hydrated cortex [6]. If the lens continues to mature and the lens protein begins to denature, the resulting hyperosmolarity leads to continued hydration and the lens becomes a swollen or intumescent cataract [7]. The lens capsule is stretched by the enlarging lens, becoming calcified in some areas and flaccid in others. Another etiology of lens hydration is traumatic puncture of the lens capsule, which follows a much more rapid course. Phacomorphic angle-closure is an uncommon condition in Western countries, though it has been cited as the cause of 3.9% of cataract extractions done in India [8]. Prevalence and incidence data is sparse. One report from Nepal Eye Hospital detailed a 2 year prevalence of 0.01% [9]. It is almost always unilateral, although one report found bilateral (but asynchronous) presentation in 14% of 86 cases [8].

## Risk Factors

Reported risk factors are listed below in Table 1. Among reported risk factors for the development of phacomorphic angle-closure, the most commonly cited is age [3]. One small study (49 patients) found that patients over 60 years old had an increased risk (odds ratio 2.7) of developing phacomorphic angle-closure [10]. More mature cataracts develop over time, and age is also a risk factor for primary angle-closure glaucoma, placing these patients at increased risk from having crowded angles. A retrospective case-control study compared 100 patients with phacomorphic angle-closure (the average age was 73 years old) to age-and gender-matched control eyes with mature cataracts [3]. The

study showed that shortened axial length is a risk factor: the average axial length in their control group was 23.7 mm versus 23.1 mm in the phacomorphic angle-closure group. Having an axial length  $\leq$  23.7 mm increased the risk of phacomorphic angle-closure by a factor of 4.3. Although narrow anterior chamber depth has been previously cited as a risk factor for this secondary form of angle-closure glaucoma [4], this particular study did not find any statistically significant correlation. However, this study used anterior chamber depth measurements from the fellow eye because the attack of angle closure would be expected to narrow the chamber and so a measurement during an attack would not be representative of the depth of the anterior chamber prior to the attack. Studies to further evaluate this would need to prospectively measure these values in healthy patients and follow them to identify risk factors, but the ethics involved with observing the development of phacomorphic angle-closure without treatment precludes a fully prospective trial. Furthermore, sex may be a risk factor, with a female predominance [10,11] reported to be as high as 3:1 [12] but this has not been a consistent finding.

It should be noted that some phacomorphic angle-closure patients will not have any identifiable risk factors. Although not consistent with the historical definition involving a mature cataract, phacomorphic angle-closure of the same mechanism has been reported in myopic eyes. In at least one case, the patient had increased lens thickness (5.33 mm) which required a clear lens extraction [13]. A 40 year-old highly myopic male (-15 diopters in the right and -18 in the left)

Confirmed Risk Factors
Age over 60 [3,10]
Axial length $\leq$ 23.7 mm <sup>3</sup>
Possible Risk Factors
Narrow anterior chamber [4]
Female gender [10,12]

Table 1: Risk Factors for Phacomorphic Angle-Closure.

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had bilateral phacomorphic angle-closure secondary to isolated spherophakia confirmed with ultrasound-biomicroscopy (UBM) [14]. A 16 year-old female with poorly controlled type 1 diabetes was also reported to have bilateral phacomorphic angle-closure, likely from the acute hyperosmolar myopic shift that occurs with hyperglycemia [15]. Zonular laxity can lead to both anterior lens shift and increased anteroposterior lens diameter. Phacomorphic angle-closure from zonular laxity was reported in cases of Weill-Marchesani syndrome (despite a patent peripheral iridotomy) [16], as well as in patients with a familial Marfanoid appearance [17].

## Diagnosis and Work-up

Multiple publications [1,3,10,18] report using the same criteria for diagnosis of phacomorphic angle-closure. The common cutoff for IOP above 21 mmHg was used, though the average presenting IOP is often over 40 mmHg [1,3,19]. As this is a secondary form of angle-closure, findings include injection, corneal edema, mid-dilated pupil, shallow anterior chamber, and a mature cataract. Of 100 patients, 71% complained of eye pain while 16% described it as a headache [1]. Nausea was reported by 8%. A B-scan should be done given an inadequate view of the fundus. Phacomorphic angle-closure caused by a choroidal melanoma causing anterior lens shift has been reported [20]. Between the acute attack and the dense cataract, the presenting vision is often extremely poor, averaging light perception only [1,3]. One study included biometry as diagnostic criteria, requiring a lens thickness of at least 5 mm (the average lens thickness is 4.63 mm) [21], and an anterior chamber depth less than 2 mm [19].

A vital part of the exam is gonioscopy to confirm a closed angle. If the corneal edema is severe, this may not be possible in the involved eye. Indentation gonioscopy may relieve the pupillary block component of the attack. Since the main differential for phacomorphic angle-closure is acute angle-closure glaucoma, gonioscopy of the fellow eye should be done to ensure that it is not occludable. The angle in the fellow eye may also be narrow, as 60% of 49 patients with phacomorphic angle-closure had a moderately narrow angle with a maximum angle width of 20 degrees in the fellow eye [10]. The fellow eye should have less of a dense cataract with a normal IOP, although this can vary depending on how much time passes between the attack and when the patient is seen. In one study of 49 patients, 80% had an immature cataract in the fellow eye [10].

In examining the cataract, it is important to evaluate for zonular laxity since it is associated with both main causes of the intumescent cataract, trauma and aging. There is also a possible association with pseudoexfoliation syndrome, in which the zonular laxity can predispose the patient to anterior lens movement. Pseudoexfoliation syndrome was present in 50% of cases in a small case series [4]. Since the mature cataract may limit any assessment of visual acuity, ancillary testing may help evaluate postoperative potential [6]. Testing can range from Purkinje vascular entoptic testing to visual-evoked potentials.

## Treatment

The goal in treating phacomorphic angle-closure is to reduce the IOP before the onset of acute glaucomatous optic neuropathy. Longer duration of an attack has been correlated with a progressive increase in IOP [8]. The only definitive treatment is cataract extraction [18]. However, to avoid operating on an inflamed eye with high pressure (increasing the risk of suprachoroidal hemorrhage from rapid IOP fluctuations) [18,22], with a limited view from corneal edema and an extremely shallow chamber, the initial goal is to stabilize the eye by

breaking the acute attack and lowering the IOP using either medical or laser treatment.

## Medical treatment

The most common initial treatment is topical anti-glaucoma medications, including beta-blockers, alpha-agonists, and carbonic anhydrase inhibitors [23]. Several studies relied on a standard treatment algorithm; timolol, acetazolamide and intravenous mannitol were among the most common medications used [11,18]. Pilocarpine should be avoided because it causes a forward shift of the iris-lens diaphragm which would worsen the angle-closure, and can increase the amount of inflammation [5,24]. The use of prostaglandin analogues have not been widely reported, possibly to avoid their proinflammatory effects during an acute attack [25]. Topical treatment alone may be insufficient to break the attack. In one study, patients received a topical beta-blocker, oral acetazolamide and glycerol and only 60% of patients achieved an IOP under 30 mmHg [10]. The presence of at least 180 degrees of peripheral anterior synechiae (PAS) at presentation may be an indication that topical treatment will be insufficient to break the attack [12]. If topical treatment fails to bring the IOP into a tolerable range until cataract extraction can be performed, there are 3 options. The first is oral or intravenous medicine such as acetazolamide or mannitol. Argon laser peripheral iridoplasty (ALPI) has successfully been used to break the attack and can often be used even if corneal edema precludes a view clear enough for the third option, laser peripheral iridotomy [26]. All of these treatments are temporizing because none remove the actual etiology: the swollen lens pushing the angle closed. A relatively small number of cases report combining phacoemulsification with glaucoma procedures (trabectulotomy [27] or aqueous shunts [28,29]) for pressure control, like in 4 patients who had an uncontrollable IOP (value unspecified) despite IV mannitol at presentation. Adding operative procedures to these eyes increases the risk of complications such as prolonged uveitis in 3 of 9 patients treated with combined cataract extraction and aqueous shunts [8]. In that study the final visual acuity was worse in the combined cases than in the eyes treated only with cataract extraction and combining glaucoma surgery with cataract extraction is not currently recommended.

## Laser treatment

Argon Laser Peripheral Iridoplasty has been suggested as a way to avoid the systemic side effects that come with oral or intravenous medicine, such as metabolic acidosis with acetazolamide or congestive heart failure with hyperosmotics [5]. The settings used vary but Tham et al. averaged 46 shots around 360 degrees of peripheral iris, with a laser spot size of 500 microns, power set at 264 mW for 0.5 second duration [5]. Despite the fact that only two of ten eyes showed any open angle on gonioscopy following iridoplasty, the average IOP was reduced from 56 at baseline, to 45 at 15 minutes following the procedure, to 34 at 50 minutes, and finally to 14 mmHg at 24 hours. UBM, which may have revealed a change in angle configuration, was not done [30]. The authors postulated that the inflammatory effects caused by the laser on the uveoscleral outflow or ciliary body may be responsible for lowering the IOP in those cases. One of the ten patients failed to achieve adequate IOP control (the IOP was 40 after two hours) and so received intravenous acetazolamide. There are no available reports on the timing of IOP reduction with systemic medications specifically for phacomorphic angle closure. To give some comparison, a randomized control trial comparing medication (intravenous acetazolamide and in some cases mannitol) to ALPI for primary acute angle-closure found that the IOP decreased much more rapidly with ALPI [31]. The eyes

treated with ALPI reached an IOP of 31 after only 15 minutes, whereas it took 60 minutes following medication administration to reach the same IOP, suggesting ALPI may achieve lower IOP's faster than systemic medicine. However, even though this decreases the chance of PAS formation and glaucomatous optic neuropathy by shortening the duration of an angle-closure attack, there are no longitudinal studies confirming this. Another series using iridoplasty to break the acute attack had less successful results according to a stricter protocol [24]. Two hours following the procedure, 19% of 21 patients had an IOP greater than 30 and so were taken for emergent cataract extraction.

The role of pupillary block in the pathophysiology of phacomorphic angle-closure is not completely understood. It is important to note that iridoplasty alone does not relieve pupillary block, which still would require an iridotomy, best done within 2-3 days [26]. One possible reason to do a laser iridotomy even though cataract extraction is already planned is the report of a patient who had an acute attack successfully aborted with topical medication only to sustain a second angle-closure attack precipitated by the preoperative dilating drops [4]. The intraoperative IOP climbed to 46 mmHg. A laser iridotomy will not relieve angle-closure in advanced cases where the lens is large enough to directly push the peripheral iris into the trabecular meshwork [4]. Finally, neither laser is an option if the cornea is too edematous to visualize the iris despite glycerol, or if the anterior chamber appears so narrow that the laser will damage the corneal endothelium [5].

### Surgical technique

Cataract extraction of the intumescent lens necessitates preparation due to the increased risk of complications. Specific risks include higher positive pressure, risk of expulsive hemorrhage, and association with zonular dialysis [18]. Intravenous mannitol (1-2 g/kg) has been used to reduce positive pressure and IOP [10,18]. The two most common choices for removing the mature lens are extracapsular extraction or phacoemulsification [1]. Extra-capsular cataract extraction has been used successfully, especially Manual Small-Incision Cataract Surgery (SICS) [32]. In a prospective study of 74 patients by Ramakrishnan et al., only 7% of these dense lenses had a ruptured posterior capsule, and only 1% had zonular dialysis requiring a capsular tension ring [18]. A blinded, randomized parallel trial (SICS versus phacoemulsification) of white cataracts with 270 patients found that SICS was a faster procedure (9 minutes versus 12 minutes) [32]. Six weeks after surgery, the best corrected visual acuity was 20/60 or better in 99% of the phacoemulsification group and 98% of the SICS group, although the average astigmatism was marginally higher with SICS (1.2 versus 0.8 diopters). Furthermore, the number of patients who achieved 20/30 or better, corrected and uncorrected, were both nearly 10% higher with phacoemulsification. There was a statistically similar number of posterior capsule tears (1% with SICS versus 2% with phacoemulsification), and no other serious complications were reported in either group.

With extra-capsular extraction employing SICS, a 6.5-7 mm superior frown-shaped sclero-corneal tunnel is fashioned [32], followed by a can-opener or continuous curvilinear capsulorrhexis [33]. The lens can be extracted with various techniques including aggressive irrigation or prolapse with hooks (such as a Sinskey hook) or a loop, as pressure is exerted on the posterior lip of the wound. After insertion of a single-piece lens, manual aspiration of any free cortical matter is done with a Simcoe cannula [10,11,32]. Many cases do not require sutures.

For phacoemulsification, the shallow chamber complicates numerous steps. There may be iridocorneal apposition which can

interfere with making the incisions. Iris prolapse is much more likely, and the positive posterior pressure redirects forces during capsulorrhexis, increasing the risk of radicalization [22,34]. A narrow chamber also forces the phacoemulsification tip closer to the corneal endothelium. One definitive option to deal with increased positive posterior pressure is to do a partial vitrectomy prior to the cataract extraction, aspirating 0.2 cc of vitreous [35]. Dada et al. report on a blind, single-site 23 gauge partial-core pars plana vitrectomy prior to phacoemulsification [22]. At the same time, they injected viscoelastic into the anterior chamber through a paracentesis to deepen the chamber. Although this deepened the chamber and facilitated successful phacoemulsification, the sclerotomy carries its own risks of retinal dialysis, tear or detachment, necessitating a comprehensive retinal exam at post-op visits.

To achieve a continuous curvilinear capsulorrhexis, it will likely be necessary to improve visualization on the capsule by using a dye such as trypan blue which will also be of use throughout the case by keeping the edge of the capsulorrhexis in constant view. Another aid that has been used is oblique illumination held at the limbus [36]. Focal calcifications of the capsule may necessitate redirecting the capsulorrhexis around the plaque or using scissors to cut through it [37]. Since hydrated cortex may escape from the mature lens into the anterior chamber when the capsulorrhexis is started, one method to clear the view is to pierce the capsule with a bent 26 gauge needle to allow initial egress and decompression. The free cortex can be removed with a small syringe before capsulorrhexis is continued under high magnification. To reduce the intralenticular pressure to keep the tractional forces on the capsulorrhexis flat, a 30 gauge needle can be used to directly withdraw cortex from the lens [36]. Excess viscoelastic will raise the intraocular pressure even further, and if liquified cortex escapes and mixes with viscoelastic it will stay near the capsulotomy site, obscuring the view [38]. Since the cortex is already hydrated in an intumescent cataract, hydrodissection may not be useful and may make the lens excessively mobile.

Sufficient dispersive viscoelastic should be used to protect the corneal endothelium. This can be used in combination with a cohesive viscoelastic which helps negate the positive posterior pressure, helping to neutralize radialization forces in a soft-shell technique [39]. In this technique, a low-viscosity dispersive agent is injected over the lens, followed by a high-viscosity cohesive agent injected posteriorly, forcing the dispersive agent anteriorly to form a protective coating on the corneal endothelium. The soft-shell technique has been shown in a meta-analysis to reduce corneal endothelial cell loss [40]. Viscoadaptive agents may be utilized instead. Intraoperative miosis may be improved with pupil-stretching using two hooks [35].

A divide and conquer method would be suboptimal because cracking would be difficult with such a large lens in a weakened capsule with possible zonular laxity. Employing a stop and chop method will thin the nucleus and offer an improved chance of successful chopping. During sculpting, high power will likely be needed, with one suggested setting detailing power of 80%, aspiration flow rate of 25 cc per minute and vacuum of 140 mmHg [7]. During chopping, using a longer tip (1.75 mm) improves the chance of a successful chop. Another technique called cratering involves deep sculpting of the entire central core of the nucleus (leaving behind only a small posterior cushion), after which the remaining nuclear rim can be split apart with cracking or chopping [41]. Vanathi et al. applied cratering then chopping to 26 eyes with mature cataracts and there were no posterior capsule tears [41]. Only 3 patients had corneal edema on postoperative day one. Other helpful techniques include using a second instrument to physically break the dense strands connecting nuclear fragments during aspiration [6].

There is an increased risk of posterior capsule tear with mature lenses because the large, dense cataract physically stretches the capsule, which can lead to a thinning and slacking. Excess slack predisposes the capsule to come anteriorly toward the phacoemulsification tip. Finally, there is often no remaining epinucleus which normally acts as a cushion between the tip and the posterior bag. This can be prevented by injecting some viscoelastic underneath the lens as soon as some red reflex is visible after removal of the first part of the cataract, offering a cushion between the posterior capsules whilst raising the nucleus closer to the phacoemulsification tip [6].

On the day following surgery, common exam findings include severe iritis in up to 84% of patients [10], inflammatory fibrin precipitates (48%) and severe corneal edema (24%) [18]. These are expected to resolve following topical treatment. Descemet's folds lasted an average of 6 days in one study, and their presence did not correlate with final visual acuity [19]. Hyphema has been reported in 3% [18], - 8% [10] which resolved by postoperative week one.

### Prognosis

The visual prognosis in most cases will be defined by the final visual acuity following removal of the dense, large cataract. The other main risk factor for poor visual outcome is chronically elevated IOP. Studies have found that presenting IOP and duration of the attack may help predict final visual outcomes.

Two studies show that there is no statistically significant association between the preoperative IOP measurement and the final visual acuity [1,18]. It would seem that this one-time measurement of IOP prior to surgery does not correlate with visual outcome. Only one study found that presenting IOP over 40 mmHg [11] was a risk factor for poor visual outcome, but this study suffered from poor follow-up as only 30% of patients came for a second visit after surgery. Presenting visual acuity is also a poor indicator of final acuity: out of 18 patients who presented with light perception without projection, 83% improved to vision better than counting fingers and 17% improved to 20/40 or better [8]. The duration of symptoms prior to surgery (used as a surrogate for duration of the IOP rise since IOP was not measured in these studies prior to presentation) does appear to correlate with the final visual acuity [11]. A final visual acuity 1 year after surgery of 20/40 or better was achieved in 70% of patients who presented within 10 days of the onset of symptoms but in only 58% of patients who presented within 11-20 days [18]. This finding was confirmed in a second study which found a small but significant correlation ( $r^2=0.1$ ,  $p=0.001$ ) between a shorter delay from onset of symptoms to surgery and final visual acuity [1]. A third study showed that a delay of more than five days between the onset of pain and cataract extraction was a strong risk factor for poor final visual acuity (odds ratio 3.1) [10].

The length of the attack has also been correlated with optic nerve head changes (either cupping or pallor). In a group of patients who had an attack lasting 5 days or less, only 9% had signs of glaucomatous optic neuropathy, versus 25% in the group with an attack longer than 5 days [8]. There were 10 patients with an attack lasting 21 days or longer, and 8 of them developed glaucomatous optic neuropathy while the remaining 2 developed diffuse pallor. This corroborates the data suggesting that an attack lasting longer than 5 days is a significant risk factor for worse outcomes, in terms of both glaucoma status and final visual acuity.

### Long-Term Prognosis

The two most important factors to examine in patients with

phacomorphic angle-closure are final visual acuity and how many of these patients will go on to develop chronic glaucoma from peripheral anterior synechiae. The final visual acuity varies widely in the literature. Reports detailed visual acuity in various subsets, but in order to give an overview, reports with similar groupings are averaged in Table 2. There are case reports of patients who presented with hand motion vision achieving a final visual acuity of 20/20 [4]. In a study of 74 patients, visual acuity of 20/40 or better was achieved in 68% [18]. A retrospective study followed 82 cases of phacomorphic angle-closure for up to 9 years [1]. Only 82% of the patients had any improvement in visual acuity at final follow-up. On average, 19 months after cataract extraction, the visual acuity had improved from light perception to 20/250. Yet in the largest series of cases, only 40% of 298 cases achieved vision better than 20/200 [11]. Reasons for decreased vision include decreased corneal endothelial cell count, spherical aberration from a fixed pupil, glaucomatous optic neuropathy, and other ocular comorbidities such as macular degeneration, in this elderly population who frequently sought care only after a significant delay [19]. Corneal endothelial cell loss following an attack was reported to be 15% [12].

Since the attack of angle-closure is due to the swollen lens, if there was minimal formation of peripheral anterior synechiae, the patient may not develop chronic glaucoma. One study reported that 3 years after cataract extraction, there was an average of 100 degrees of PAS [1]. Surprisingly, the degree of PAS formation did not correlate to the time delay between symptom onset and surgery, the presenting IOP, or the final IOP. Unfortunately the mean duration of symptoms was not specified, and there were no details of the gonioscopic findings at presentation (which may not be possible with corneal edema) in the affected or fellow eye. In another study, with a mean follow-up of 2 years after cataract extraction, 0 of 21 patients showed any progression of PAS [24]. As expected, cataract extraction following phacomorphic angle-closure deepens the angle: 3 years following cataract extraction, the mean Shaffer grade of 100 eyes was 3 [1].

A review of the literature reveals only a few reports investigating the occurrence of chronic glaucoma. There are only two reports investigating longitudinal changes with conventional glaucoma testing other than IOP. Looking at IOP alone in the study with the longest average follow-up (4 years) [19], the final average IOP was only 13, and only a single patient (4%) was reported as having an a late IOP elevation following cataract extraction, which was treated with two topical medicines. In the retrospective study of 100 patients, the mean final IOP was 16 mmHg [1]. Five years following cataract extraction, 94% of patients had IOP  $\leq$  21, though some of these patients required IOP-lowering treatment. While 81% of these patients required no glaucoma treatment following cataract extraction, 15% needed pressure-lowering drops (an average of 1.6 medications), and a total of 3 patients (4%) needed a glaucoma procedure (one laser peripheral iridotomy and 2 trabeculectomies). Although in this study the average vertical cup-to-disc ratio was 0.2 higher in the affected eye compared to the fellow eye, vertical cup-to-disc ratios were identical between the two eyes in another study [42]. The same study analyzed general indices from visual fields in the affected eye, where the mean deviation was 5.2

Best Corrected Final Visual Acuity	Mean Percent of Patients
20/20-20/50	50
20/60-20/200	24
20/200-20/400	11
Worse than 20/400	15

Table 2: Average of reported final visual acuities following cataract extraction [10,11,18,19].

dB after 3 years. Since no comparison was given to the fellow eye, it is not known if this can be attributed solely to the phacomorphic angle-closure. In another report using visual fields, there was no statistically significant difference between the pattern standard deviation in the affected and unaffected eye [42].

The single study on Optical Coherence Tomography (OCT) analysis of the optic nerve head measured the Retinal Nerve Fiber Layer (RNFL) thickness in ten patients following an attack (no baseline measurements were available) [42]. Three months after the attack, the average RNFL thickness was statistically similar in the involved and fellow eyes. At 9 months significant thinning had occurred in the eye with phacomorphic angle-closure. The involved eye had an average RNFL thickness of 85 microns (a 9 micron decrease over the previous 6 months) versus 100 microns in the fellow eye, which was stable. The thinning was worse in the superior and inferior quadrants.

In planning long-term follow-up, because the first visit where any elevation in IOP was noted occurred at 18 months, Lee et al. recommend following patients with phacomorphic angle-closure for the development of glaucomatous changes for at least 2 years [1]. Although phacomorphic angle-closure itself should not recur after removing the cataract, aphakic pupillary block has been reported 17 months after cataract extraction (despite already having an iridectomy), providing another reason for long-term follow-up [4]. The fellow eye must receive periodic gonioscopy since up to 14% of cases were reported to have bilateral disease [8]. The duration between fellow eye involvements, however, averaged 10 years, suggesting that a prophylactic peripheral iridotomy in the fellow eye would not be needed. The fellow cataract should be aggressively monitored so that it can be removed before it can become intumescent.

## Conclusion

Phacomorphic angle-closure usually occurs in elderly patients with short axial length, as a mature lens swells and forces the peripheral iris into the trabecular meshwork. Increased IOP leads to the typical signs and symptoms of acute angle-closure. The definitive treatment is cataract extraction. In order to avoid operating on an inflamed eye with increased IOP, the attack is treated first with topical IOP-lowering medicines. If further acute treatment is needed, the next step is systemic medication, a laser iridotomy, or a laser iridoplasty if the view allows and the chamber is not flat. Surgical technique for removing the cataract must be adjusted to account for the large, dense lens and positive posterior pressure. Final visual outcome varies in the literature but 80-90% of patients are expected to have an improved visual outcome. At least 80% of patients have been found not to need long-term glaucoma treatment once the lens is removed but still require regular follow-up.

## References

1. Lee JW, Lai JS, Yick DW, Tse RK (2010) Retrospective case series on the long-term visual and intraocular pressure outcomes of phacomorphic glaucoma. *Eye (Lond)* 24: 1675-1680.
2. Ritch R, Shields MB, Krupin T (1996) *The glaucomas*. (2nd edn) Mosby, St. Louis.
3. Lee JW, Lai JS, Lam RF, Wong BK, Yick DW, et al. (2011) Retrospective analysis of the risk factors for developing phacomorphic glaucoma. *Indian J Ophthalmol* 59: 471-474.
4. Tomey KF, al-Rajhi AA (1992) Neodymium:YAG laser iridotomy in the initial management of phacomorphic glaucoma. *Ophthalmology* 99: 660-665.
5. Tham CC, Lai JS, Poon AS, Chan JC, Lam SW, et al. (2005) Immediate argon laser peripheral iridoplasty (ALPI) as initial treatment for acute phacomorphic angle-closure (phacomorphic glaucoma) before cataract extraction: a preliminary study. *Eye (Lond)* 19: 778-783.
6. Steinert RF (2010) *Cataract surgery: techniques, complications and management*. (3rd edn) Saunders, Philadelphia.
7. Buratto L, Osher RH, Masket S (2000) *Cataract surgery in complicated cases*. SLACK, Thorofare, NJ.
8. Jain IS, Gupta A, Dogra MR, Gangwar DN, Dhir SP (1983) Phacomorphic glaucoma--management and visual prognosis. *Indian J Ophthalmol* 31: 648-653.
9. Rijal AP (2005) Clinical analysis of glaucoma in hospital patients. *Kathmandu Univ Med J (KUMJ)* 3: 249-254.
10. Prajna NV, Ramakrishnan R, Krishnadas R, Manoharan N (1996) Lens induced glaucomas--visual results and risk factors for final visual acuity. *Indian J Ophthalmol* 44: 149-155.
11. Pradhan D, Hennig A, Kumar J, Foster A (2001) A prospective study of 413 cases of lens-induced glaucoma in Nepal. *Indian J Ophthalmol* 49: 103-107.
12. Angra SK, Pradhan R, Garg SP (1991) Cataract induced glaucoma--an insight into management. *Indian J Ophthalmol* 39: 97-101.
13. Jayaprakasam A, Martin KR, White AJ (2012) Phacomorphic intermittent angle closure in a patient with retinopathy of prematurity and lenticular high myopia. *Clin Experiment Ophthalmol* 40: 646-647.
14. Sowka J, Girgis N (2010) Bilateral phacomorphic angle-closure glaucoma in a highly myopic patient secondary to isolated spherophakia. *Optometry* 81: 432-436.
15. Skrabec V, Ivanisevic M, Stanic R, Unic I, Bucan K, et al. (2010) Acute bilateral cataract with phacomorphic glaucoma in a girl with newly diagnosed type 1 diabetes mellitus. *J Pediatr Ophthalmol Strabismus* 47 Online: e1-3.
16. Barkana Y, Shihadeh W, Oliveira C, Tello C, Liebmann JM, et al. (2006) Angle closure in highly myopic eyes. *Ophthalmology* 113: 247-254.
17. Dagi LR, Walton DS (2006) Anterior axial lens subluxation, progressive myopia, and angle-closure glaucoma: recognition and treatment of atypical presentation of ectopia lentis. *J AAPOS* 10: 345-350.
18. Ramakrishnan R, Maheshwari D, Kader MA, Singh R, Pawar N, et al. (2010) Visual prognosis, intraocular pressure control and complications in phacomorphic glaucoma following manual small incision cataract surgery. *Indian J Ophthalmol* 58: 303-306.
19. Lee SJ, Lee CK, Kim WS (2010) Long-term therapeutic efficacy of phacoemulsification with intraocular lens implantation in patients with phacomorphic glaucoma. *J Cataract Refract Surg* 36: 783-789.
20. Wanner JB, Pasquale LR (2006) Glaucomas secondary to intraocular melanomas. *Semin Ophthalmol* 21: 181-189.
21. Hoffer KJ (1993) Axial dimension of the human cataractous lens. *Arch Ophthalmol* 111: 914-918.
22. Dada T, Kumar S, Gadia R, Aggarwal A, Gupta V, et al. (2007) Sutureless single-port transconjunctival pars plana limited vitrectomy combined with phacoemulsification for management of phacomorphic glaucoma. *J Cataract Refract Surg* 33: 951-954.
23. Sowka J (2006) Phacomorphic glaucoma: case and review. *Optometry* 77: 586-589.
24. Yip PP, Leung WY, Hon CY, Ho CK (2005) Argon laser peripheral iridoplasty in the management of phacomorphic glaucoma. *Ophthalmic Surg Lasers Imaging* 36: 286-291.
25. Schumer RA, Camras CB, Mandahl AK (2002) Putative side effects of prostaglandin analogs. *Surv Ophthalmol* 47 Suppl 1: S219.
26. Ritch R, Tham CC, Lam DS (2007) Argon laser peripheral iridoplasty (ALPI): an update. *Surv Ophthalmol* 52: 279-288.
27. Rijal AP, Karki DB (2006) Visual outcome and IOP control after cataract surgery in lens induced glaucomas. *Kathmandu Univ Med J (KUMJ)* 4: 30-33.
28. Das JC, Chaudhuri Z, Bhomaj S, Sharma P, Gupta R, et al. (2002) Combined extracapsular cataract extraction with ahmed glaucoma valve implantation in phacomorphic glaucoma. *Indian J Ophthalmol* 50: 25-28.
29. Das JC, Chaudhuri Z, Bhomaj S, Sharma P, Gupta R, et al. (2002) Combined extracapsular cataract extraction with Ahmed Glaucoma Valve implantation in phacomorphic glaucoma. *Indian J Ophthalmol* 50: 25-28.

30. Thyagarajan S (2006) Immediate argon peripheral iridoplasty (ALPI) as initial treatment phacomorphic glaucoma: a safe and cost-effective treatment? Eye (Lond) 20: 1323.
31. Lam DS, Lai JS, Tham CC, Chua JK, Poon AS (2002) Argon laser peripheral iridoplasty versus conventional systemic medical therapy in treatment of acute primary angle-closure glaucoma : a prospective, randomized, controlled trial. Ophthalmology 109: 1591-1596.
32. Venkatesh R, Tan CS, Sengupta S, Ravindran RD, Krishnan KT, et al. (2010) Phacoemulsification versus manual small-incision cataract surgery for white cataract. J Cataract Refract Surg 36: 1849-1854.
33. Agarwal S, Agarwal A, Apple DJ, Buratto L, Alio JL, et al. (2002) Textbook of ophthalmology. (1st edn) Jaypee Brothers Medical Publishers, New Delhi, India.
34. Rao SK, Padmanabhan P (1998) Capsulorhexis in eyes with phacomorphic glaucoma. J Cataract Refract Surg 24: 882-884.
35. Chang DF (2001) Pars plana vitreous tap for phacoemulsification in the crowded eye. J Cataract Refract Surg 27: 1911-1914.
36. Bhattacharjee K, Bhattacharjee H, Goswami BJ, Sarma P (1999) Capsulorhexis in intumescent cataract. J Cataract Refract Surg 25: 1045-1047.
37. Vajpayee RB, Bansal A, Sharma N, Dada T, Dada VK (1999) Phacoemulsification of white hypermature cataract. J Cataract Refract Surg 25: 1157-1160.
38. Chan DD, Ng AC, Leung CK, Tse RK (2003) Continuous curvilinear capsulorhexis in intumescent or hypermature cataract with liquefied cortex. J Cataract Refract Surg 29: 431-434.
39. Arshinoff SA (1999) Dispersive-cohesive viscoelastic soft shell technique. J Cataract Refract Surg 25: 167-173.
40. Van den Bruel A, Gailly J, Devriese S, Welton NJ, Shortt AJ, et al. (2011) The protective effect of ophthalmic viscoelastic devices on endothelial cell loss during cataract surgery: a meta-analysis using mixed treatment comparisons. Br J Ophthalmol 95: 5-10.
41. Vanathi M, Vajpayee RB, Tandon R, Titiyal JS, Gupta V (2001) Crater-and-chop technique for phacoemulsification of hard cataracts. J Cataract Refract Surg 27: 659-661.
42. Lee JW, Lai JS, Yick DW, Yuen CY (2012) Prospective study on retinal nerve fibre layer changes after an acute episode of phacomorphic angle closure. Int Ophthalmol 32: 577-582.

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