Early lens aspiration with posterior chamber intraocular lens and capsular tension ring in microspherophakia to avoid lens-induced complications

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Abstract:

PURPOSE: To report the surgical outcome of early lens aspiration, posterior chamber intraocular lens (PC IOL), and capsular tension ring (CTR) in a case series of microspherophakia (MSP) and secondary glaucoma.

METHODS: Case series of 18 eyes of MSP cases presented with lenticular myopia and secondary glaucoma that underwent early lens aspiration, PC IOL and CTR by one ophthalmologist. Baseline, long-term postoperative outcomes and complications were documented.

RESULTS: All cases underwent successful surgery with lens aspiration PC IOL implantation and CTR insertion without intraoperative complications. One of the 18 cases was a delayed referral which had broad anterior synechiae and following lens aspiration developed corneal decompensation. In one eye, CTR implantation was not possible hence, lens aspiration with scleral fixation (SF) of 3 piece IOL was performed (excluded from the analysis). Overall there was an improvement in visual acuity (from 0.3 ± 0.1 to 0.2 ± 0.2 LogMar, $P = 0.006$), intraocular pressure (IOP), and most notably, deepening of the anterior chamber. Some cases required subsequent glaucoma surgery to control IOP. After a long duration of follow-up, all cases had stable capsular lens complex and no capsular phimosis.

CONCLUSION: Early Lens aspiration with CTR and PCIOL alone in MSP with lens subluxation has a significant impact on the patient's quality of vision, deepening the anterior chamber and preventing complications or poor outcomes. In addition, good capsular-lens complex stability and absence of capsular phimosis or phacodonesis on long-term follow-up were obtained.

Keywords:

Capsular tension ring, glaucoma, lens aspiration, lens-induced myopia, spherophakia

Introduction

Microspherophakia (MSP) is a rare bilateral congenital disorder, in which the crystalline lens develops a spherical form with a longer anteroposterior distance and a smaller equatorial diameter (ED) secondary to compromised nutritional support of the tunica vasculosa lentis during the 5th and 6th months of intrauterine life.[1]

MSP can be found in familial anomalies that are autosomal dominant (AD) or autosomal recessive (AR), and in isolation. However, it typically presents in association with systemic disorders such as WeillMarchesani syndrome (WMS) (most commonly),[2] Homocystinuria, Marfan syndrome, Alport syndrome, Klinefelter syndrome, Lowe syndrome, Peter's anomaly, and Cri-du-chat syndrome.[1]

Typical lens morphological features in MSP include 4–6.75 mm anteroposterior distance, between 6.5 and 8.0 mm for equatorial distance, and weak zonules, which are responsible for the typical common complications present in MSP: Lenticular myopia and subsequent pupillary block glaucoma. Secondary glaucoma is the main reason for severe permanent loss of vision, and it may be present in up to 51% of MSP eyes.[1]

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There is no consensus in the literature on the definitive treatment of MSP. However, conservative management such as antiglaucoma medications, spectacles, and a contact lens is usually futile in this condition. Therefore, surgical interventions aiming to counteract pupillary block glaucoma and lenticular myopia have been favored in MSP. However, due to the intra-operative surgical difficulties, variable zonular laxity, and the progressive nature of MSP, determining a universal surgical approach is challenging.[1,3]

Although lens aspiration is consensual in the literature in relieving anterior chamber crowding and secondary glaucoma, successful surgical outcomes in intraocular pressure (IOP) control and visual acuity improvement in MSP cases depend on many factors. These include disease duration, anterior chamber angle at presentation, glaucomatous disc damage, degree of zonular laxity, lens subluxation/dislocation, and amblyopia therapy.[1,4-8]

In general, only a few collective case series and individual case reports on lens aspiration with CTR and posterior chamber intraocular lens (PC IOL) have been described with relatively short-term outcomes. However, the long-term outcome of early intervention is lacking. This current study is originated to determine evidence-based practice on the long-term outcome and the magnitude of early lens aspiration with the implementation of CTR and PC IOL in MSP with secondary glaucoma. This may change our practice to a simpler technique with long-term stability.

Methods

Retrospective chart review of ten consecutive MSP cases (18 eyes) at King Khaled Eye Specialist Hospital (KKESH). Eighteen eyes who underwent primarily lens aspiration with CTR and PC IOL by one ophthalmologist were included in this study. The Institutional Review Board approved the study in KKESH.

Patients

All cases had clinical features of MSP including; increased anteroposterior lenticular distance, decreased lens equatorial diameter, lens-induced myopia, secondary glaucoma, peripheral anterior synechiae (PAS) or iridocorneal touch, and a history of high IOP and peripheral iridotomy (PI). In addition, genetic study mapping was done for all except for two patients.

The thickness of the lenses and anterior chamber depth (ACD) were determined using a scheimpflug imaging (Pentacam® HR; OCULUS Optikgerate, Wetzlar, Germany) and Ultrasound biomicroscopy for axial length (AL) and lens diameter.

Surgical procedures

The study included 10 MSP patients; 19 eyes underwent lens aspiration with CTR and PC IOL (18 eyes were included in the study. One eye had significant severe zonular laxity, attempting CTR was unsuccessful and underwent scleral fixated IOL, not included in the study). In addition, six eyes required the release of anterior synechiae intraoperatively, and four eyes needed pars plana dry anterior vitrectomy to deepen the anterior chamber. CTR size 12/10 was used for all. One ophthalmologist (AAR) performed all procedures [Supplementary Video 1].

Outcome measures

Both baseline and postoperative measurements of the following were collected; uncorrected visual acuity; best-corrected visual acuity (BCVA) in manifest refraction; IOP; spherical equivalent (SE); lens thickness and ACD (using Pentacam); the number of antiglaucoma medications. BCVA and IOP measurements were performed at baseline, 1, 5, 10 years, and last follow-up. Complications were documented as well. Not all eyes had all parameters measured, some are missing due to the nature of the retrospective study.

Statistical analysis

Data were collected, managed, and coded using Microsoft Excel (Excel 2010; Microsoft Corp., Redmond, WA). Data analysis was performed using SPSS® version 21.0 (IBM Inc., Chicago, Illinois, USA). Descriptive analysis was primarily done, where variables were presented in the form of mean ± standard deviation, minimum-maximum, range, median, and mode. Paired samples *t*-test was used to compare preoperative variables with postoperative variables or variables at the last follow-up. Any output with a $P < 0.05$ was interpreted as an indicator of statistical significance.

Results

The demographics and characteristics of the total ten MSP patients (18 eyes) were documented and described in Table 1. Three were males, and 7 were females out of 6 families. There was a strong family history of 90%. Four patients presented with a complete picture of WMS, four were a partial picture of WMS, and two were unknown. Genetic mapping was performed for all except two, and it displayed ADAM Metallopeptidase With Thrombospondin Type 1 Motif 17 (*ADAMTS 17*) in three patients, A disintegrin and metalloproteinase with thrombospondin type 1 motif 10 (*ADAMTS 10*) in four patients, and 1 was unknown.

All cases had a clinical picture of increased MSP with increased anterior-posterior lens distance and reduced lens equatorial diameter under slit-lamp examination. The mean of lens thickness was 4.43 ± 0.47 mm (measured in 6 eyes), and equatorial lens diameter was documented in two eyes as 7.42 mm and 7.84 mm. All patients had lenticular myopia (baseline SE with mean of − 12.8 ± 3.5 D) with mildly subluxated lens and secondary glaucoma (IOP mean = 21.7 ± 9.5 mmHg). All cases presented with PAS, shallow anterior chamber (mean = 1.4 ± 0.6 mm, measured in 11 eyes), one eye was unmeasurable due to lenticular corneal touch and absent anterior chamber [Table 1].

Glaucoma management was primary Nd: YAG laser PI for all eyes, as well as additional antiglaucoma drops, except five eyes that did not require any drops. Three eyes required cyclophotocoagulation (CPC), 3 required

Table 1: Demographics and characteristics of microspherophakia cases (n=18 eyes)

Variable	Mean±SD	Minimum-maximum	Range	Median	Mode
Age (years)	16.4±7.3	9-35	26	14.5	12
Axial length (mm)	21.9±1.2	20-24	4	21.7	20.8
Lens thickness (mm)*,†	4.43±0.47	4-5.18	1.18	4.21	4
Preoperative ACD (mm)*,†	1.4±0.6	0.3-2	1.8	1.6	0.3
Preoperative (baseline) SE (D)†	−12.8±3.5	−8-−20.6	12.6	−11.8	−14.5
Preoperative (baseline) BCVA (LogMar)	0.3±0.1	0.2-0.5	0.4	0.3	0.3
Preoperative (baseline) IOP (mmHg)†	21.7±9.5	8-40	32	19.5	12
Duration of follow up (years)	8.8±3.9	0.7-13.3	12.7	10.5	5

*Scheimpflug imaging (Pentacam HR; OCULUS Optikgerate, Wetzlar, Germany). †Missing data anterior chamber depth. BCVA: Best-corrected visual acuity, LogMar: Logarithm of the Minimum Angle of Resolution, IOP: Intraocular pressure, SE: Spherical equivalent, ACD: Anterior chamber depth, SD: Standard deviation

trabeculectomy + MMC (one of those who had an absent anterior chamber, required anterior chamber reformation). There was a tendency of certain patients/eyes to present or continue to have severe glaucoma that requires preoperative and postoperative glaucoma procedures [Table 2: Cases no. 10–14]

Eighteen eyes that met inclusion criteria underwent lens aspiration with CTR and PC IOL. One patient with a delayed referral underwent lens aspiration with CTR and PC IOL, developed corneal decompensation 1 year later, then underwent penetrating keratoplasty with IOL replacement with scleral fixated PC IOL in the same eye. The other eye underwent immediate penetrating keratoplasty and lensectomy with SF PC IOL due to delayed referral and development of corneal decompensation secondary to iris lens corneal touch (not included in the study). CTR size 12/10 was used for all. The mean of IOL power used was 23 ± 3.8 with minimal power of 15 and a maximum power of 31 (documented in operative notes in 16 eyes, Alcon single piece).

The visual outcome measured by BCVA (LogMar) showed a significant improvement over 1, 5, and 10 years of follow-up as well as the last follow-up (follow-up mean = 8.8 ± 3.9 years) with a significant $P = 0.006$ when comparing BCVA of the last follow-up to baseline. Moreover, the mean value of SE at the last follow-up was − 1.5 ± 1.6 D (ranging from 2 D to − 4.5 D) [Table 3 and Figure 1].

Likewise, there was an improvement in IOP in the same duration of follow-up. However, it showed no clinical significance when comparing IOP of the last follow-up to baseline (P-value = 0.131) [Table 3 and Figure 1]. In addition, the difference of ACD was significant when comparing baseline to postoperative documentation with P value < 0.001 (baseline ACD was 1.4 ± 0.6 mm [min 0.3-max 2 mm] and postoperatively was 3.3 ± 0.5 mm [min 2.5– max 4.1 mm] showing 1.9 mm difference).

Overall, all cases had Stable PC IOL-CTR capsular complex without any subluxation or dislocation, and none developed capsular phimosis. However, subsequently, six eyes developed posterior capsular opacity and underwent laser YAG capsulotomy.

Five out of eighteen eyes did not require antiglaucoma drops, neither preoperation nor postoperation. Two eyes remained using the same number of antiglaucoma drops. Nevertheless, there was a decrease in the number of antiglaucoma drops in eight eyes from baseline (preoperative) mean of 1.8 ± 1.3 drops to 1.2 ± 1.2 drops postoperatively at the last follow-up (P-value = 0.076). Furthermore, three of those eight eyes became completely independent from antiglaucoma drops. Finally, only three eyes needed additional drops (9 and 5 years after primary procedure) [Table 2].

Furthermore, six eyes had uncontrolled IOP postprimary procedure despite maximum antiglaucoma medications. Accordingly, two eyes underwent CPC (one of them was 11 years postprimary procedure, and the other was 1-year postprimary procedure then repeated a year later), and 4 had Ahmad valve implant (ranging 1–4 years postprimary procedure, and one of them had encapsulated bleb revision 11 years later then CPC was performed) [Table 2].

Discussion

MSP is a rare ocular developmental disorder. Its rarity is recognized in the literature due to the scarce reported relevant data. Although it is most commonly reported in Asians and North Africans, its prevalence is yet to be determined. However, the prevalence of WMS is estimated at 1:100,000.[3]

A recently published bibliographic study on the prevalence of rare diseases worldwide published a rough estimation of a total of 22 reported cases of MSP/spherophakia with variable features and presentations.[9] Similarly, in a specialized lens clinic in India, only 1.2% of children presented for lens abnormalities reported MSP.[10] Similarly, in the Arab world, there was a paucity of reported cases on WMS testifying to the rare presentation of MSP.[11]

MSP is reported as AD, AR familial traits, as well as in isolation. Most commonly, it is associated with systemic disorders such as WMS.[1,2] In this study, 90% of our patients had a strong family history and the most common systemic association was WMS.

The crystalline lens adapts a spherical shape in MSP, leading to high lenticular myopia, lens dislocation or subluxation, and

Table 2: Preoperative and postoperative details of microspherophakia cases

Case	Age (years)	Prior intervention	Lensectomy to glaucoma surgery (years)	Glaucoma surgery after lensectomy	BCVA preoperative*	BCVA postoperative*	SE preoperative	SE postoperative
1	22	Yag PI		-	20/70	20/30	–19.6	–2.00
2	14	Yag PI	11	CPC	20/40	20/30	–11.6	–4.5
3	14	Yag PI	-	-	20/30	20/25	–11.75	–4.00
4	35	Yag PI, CPC	-	-	20/40	20/30	–	–0.50
5	35	Yag PI	-	-	20/40	20/25	–8.00	–0.25
6	17	Yag PI	-	-	20/40	20/20	–17.00	–2.25
7	17	Yag PI	-	-	20/40	20/20	–14.00	–2.50
8	15	Yag PI	-	-	20/40	20/30	–10.00	–1.50
9	15	Yag PI	-	-	20/40	20/30	–10.00	–1.00
10	12	Yag PI Trabeculectomy + MMC	2 11 11	Ahmad implant + pericardial patch, Then revision encapsulated bleb Then CPC	20/70	20/30	–9.00	–1.00
11	12	Yag PI CPC twice	1	Ahmad implant + pericardial patch	20/50	20/25	–8.5	–1.75
12	11	Yag PI CPC	1	Ahmad valve implant	20/50	20/20	–10.5	–2.00
13	11	Yag PI Trabeculectomy + MMC	4	Ahmad valve implant	20/50	20/40	–13.00	–0.60
14	17	Yag PI Trabeculectomy + MMC Then AC formation	1 2	CPC Then repeat CPC	20/30	20/40	–10.75	-
15	9	Yag PI	-	-	20/40	20/30	–14.50	-
16	9	Yag PI	-	-	20/40	20/25	–14.50	-
17	12	Yag PI	-	-	20/60	20/25	–15.50	+ 2.00
18	12	Yag PI	-	-	20/60	20/25	–13.25	+ 0.12

Case	IOP								Antiglaucoma drops		Number of antiglaucoma drops	
	Prelensectomy	1 month	3 months	6 months	1 year	5 years	10 years	Last	Preoperative	Postoperative	Preoperative	Postoperative
1	17	16	13	15	18	21	11	11	None	None	0	0
2	24	19	22	24	23	29	20	16	Xalatan	Lumigan	1	1
3	21	17	15	14	16	12	15	15	Xolamol	Timilol , Azopt	2	2
4	40	8	25	15	19	12	-	13	Alphagan Xolamol	None	3	0
5	18	-	-	-	15	15	-	16	Alphagan Xolamol	None	3	0
6	23	20	-	-	19	26	19	22	Xola	Xolamol	1	2
7	18	-	-	-	19	22	19	20	Xola	Xolamol	1	2
8	15	18	15	16	12	16	17	17	None	None	0	0
9	14	15	13	15	13	13	13	15	None	None	0	0
10	12	14	18	-	46	16	22	22	Maximum	Xolamol	4	2
11	36	31	7.5	8	22	17	10	17	Apraclonadine + Xolamol	Xolamol	3	2
12	30	-	-	-	12	18	20	18	Bromidine.Xolamol	Lumigan, Alphagan, Xolamol	3	4
13	8	-	-	-	17	26	16	17	Bromidine.Xolamol	Xolamol	3	2

Contd...

Table 2: Contd...

Case	IOP								Antiglaucoma drops		Number of antiglaucoma drops	
	Prelensectomy	1 month	3 months	6 months	1 year	5 years	10 years	Last	Preoperative	Postoperative	Preoperative	Postoperative
14	12	-	-	-	32	-	-	32	Diamox, Timilol	None	2	0
15	-	-	-	-	-	-	-	-	None	None	0	0
16	-	-	-	-	-	-	-	-	None	None	0	0
17	31	-	-	-	13	17	-	17	Timilol, Alphagan	Betagan	2	1
18	36	-	-	-	15	15	-	15	Timilol, Alphagan, Truspot	Betagan	3	1

*BCVA measured in Snellen chart. Yag PI: Peripheral iridotomy, CPC: Cyclophotocoagulation, BCVA: Best-corrected visual acuity, SE: Spherical equivalent, IOP: Intraocular pressure, AC: Anterior chamber, MMC: Mitomycin C

secondary glaucoma. In the existing studies, the incidence of lens subluxation is 44.4%, while the incidence of glaucoma ranges from 44.4% to 51%.[12,13] These reported features were coherent with our study subjects; however, our cases had more prevalent glaucoma as all of our patients underwent Nd: Yag laser PI to primarily lower IOP following the recommendation in the literature [Tables 1, 2 and Figure 2].[1,5,13]

Nevertheless, PI was helpful only partially or temporally as many eyes progressed to PAS, shallow AC, and needed medical or surgical glaucoma intervention. Eyes with severe glaucoma upon presentation (such as five eyes of 3 of our patients) were more likely to require preoperative and postoperative glaucoma procedures and continued to have antiglaucoma medications in the long run after the primary procedure. On the other hand, those with milder glaucoma upon presentation had their IOP controlled with one or a few topical medications [Table 2]. Such observation should be brought to our attention to emphasize the importance of early intervention and to follow them up in the long term.

Such unique morphological characteristics and complex complications resulted in various challenges in the management of MSP. Therefore, there is no consensus in the literature regarding the surgical management of patients with MSP. However, lensectomy/lens aspiration is widely accepted as several studies have advocated early lens aspiration in association with CTR to stabilize capsule-lens complex to prevent subsequent glaucomatous damage, capsular phimosis, and IOL decentration.[1,8,14,15]

Furthermore, variable modalities of IOL implantation have been reported for visual rehabilitation; ranging from in-the-bag posterior chamber IOL with and without CTR,[14] with scleral fixed CTS/Cionni-MCTR,[8] and scleral fixated IOL,[6,7] all of which depends on the degree of zonular laxity at presentation or intra-operatively.[1]

Yang *et al*. described different lens management with or without preserving the capsular bag in two interventional groups (group 1 managed by phacoemulsification + CTR + PC IOL, and Group 2 managed by pars plana lensectomy with SF IOL). These groups were divided according to the extent of zonule laxity, and both procedures were effective in correcting spherophakia, and both gave positive results in visual and IOP outcomes in 3 years duration of follow-up. However, the increase in BCVA in group 1 was not as clinically significant as Group 2.[4]

It is important to mention that, compared to Yang *et al*. short follow-up of 3 years of their phacoemulsification + CTR + PC IOL in a small group of 7 eyes, our study has long term follow-up of 8.8 ± 3.9 years and larger case series (18 eyes) which showed more clinically significant visual outcome. To illustrate, Yang *et al*. BCVA improved from baseline 0.79 ± 0.36 to 0.44 ± 0.3 LogMar at the last follow-up, but it was not clinically significant ($P = 0.11$). On the other hand, our current study established a clinically significant BCVA in the long run,

Table 3: Best-corrected visual acuity and intraocular pressure obtained postoperatively during follow-up

Variable	Mean±SD (minimum-maximum), range					
Timeline (years)	Preoperative (baseline)	1 year[†]	5 years[†]	10 years[†]	Last follow up[§]	P[*]
BCVA (LogMAR)	0.3±0.1 (0.2-0.5), 0.4	0.3±0.2 (0-0.8), 0.8	0.2±0.2 (0-0.5), 0.5	0.2±0.1 (0-0.4), 0.4	0.2±0.2 (0-0.8), 0.8	0.006[**]
IOP (mmHg)	21.7±9.5 (8-40), 32[†]	18.9±8.3 (12-46), 34	18.4±5.2 (12-29), 17	16.3±3.8 (10-22), 12	17.3±4.7 (11-32), 21	0.131

*P value comparing last follow-up to baseline.**Statistically significant at 5% level of significance, [§]8.8±3.9 (0.7-13.3) years, [†]Missing data. BCVA: Best-corrected visual acuity, LogMar: Logarithm of the Minimum Angle of Resolution, IOP: Intraocular pressure, SD: Standard deviation

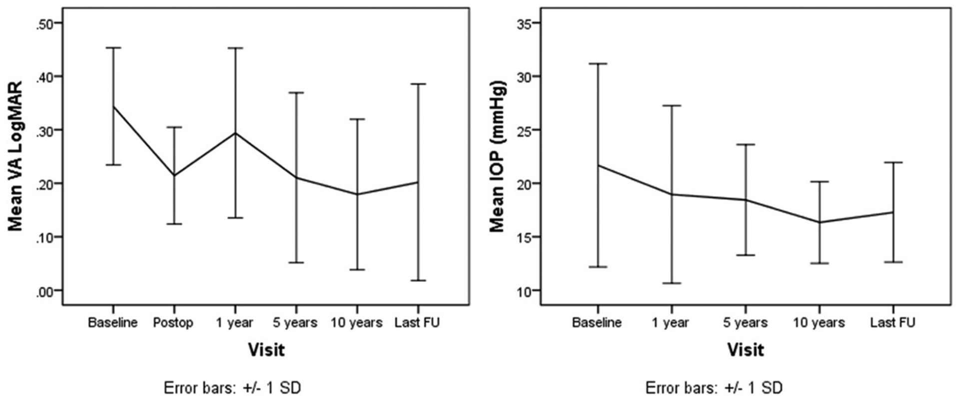

Figure 1: Best-corrected visual acuity and intraocular pressure obtained postoperatively during follow-up

whereas our BCVA has improved from baseline 0.3 ± 0.1 to 0.2 ± 0.2 LogMar at the last follow-up (significant $P = 0.006$). Of note, our refractive outcome at the last follow-up in terms of SE was − 1.5 ± 1.6 D, which is comparable to − 1.25 ± 0.78 D SE outcome reported by Yang *et al*. [Table 4].[4]

Yang *et al*. elaborated in their paper that the nonsignificant change in BCVA outcome in group 1 might be rationalized by a better BCVA at presentation due to lower lens decentration. Also, 5 out of 7 eyes developed posterior capsule opacification (which underwent YAG capsulotomy), and some eyes had a slight IOL decentration which they believe it might all be played a role in the decrease of the improvement in the BCVA. While this may be speculatively accurate, in the current study, although six eyes developed posterior capsular opacity and underwent laser YAG capsulotomy, we still achieved clinically significant visual outcomes with no long-term decentration of IOL. Mind you, our baseline BCVA (0.3 ± 0.1 LogMar) is comparable to Yang *et al*.'s initial BCVA (0.79 ± 0.36 LogMar). However, our study subject's age at intervention was much younger (18 eyes, 16.4 ± 7.3 years) in comparison to Yang *et al*.'s (7 eyes, 40.28 ± 24.22 years) [Table 4].[4] Accomplishing that, not only does the current study build on existing studies with our long-term evidence on the efficacy of lens aspiration + CTR + PC IOL in MSP management, but it also reinforces and emphasizes the importance of early management and intervention in such cases. Furthermore, our study also confirmed the stability of the capsular PC IOL complex after Yag posterior capsulotomy.

Moreover, in a reported case by Khokhar *et al*., authors described the additional use of scleral fixated CTS to stabilize the capsular-lens complex and reported 1-month postoperative successful outcomes and capsule-lens stability. They speculated possible capsule-lens complex dislocation in the long term due to the progressive nature of the disease.[8] Contrary to this hypothesis, our study has successfully reported long-term stable PC IOL + CTR capsular complex without the need of suturing of CTR or CTS, and no one developed capsular phimosis or phacodonesis in the approximately 9 years of follow-up. Except for one eye (which was excluded from the study), CTR could not be inserted in the capsular bag as zonules are very weak, and lens aspiration with SF IOL was performed instead. It could have been more successful if a smaller size (8–10 mm) custom-made CTR was available to decrease the stress on the zonules.

Yang *et al*. reported an increase in the ACD from baseline 1.21 ± 0.55 to 3.16 ± 0.52 mm at their last follow-up [Table 4].[4] Rao *et al*. articulated that lensectomy is crucial in relieving anterior chamber crowding and secondary glaucoma.[5] with their approach of lensectomy and limited anterior vitrectomy (pars plana or limbal approach) and in line with our outcomes of obtaining a significant AC deepening when compared to preoperative data (from baseline ACD 1.4 ± 0.6 mm to 3.3 ± 0.5 mm postoperatively, $P < 0.001$) [Table 4].

Rao *et al*. also found that lensectomy alone was effective in controlling IOP without antiglaucoma medications in 69% of eyes with spherophakia and secondary glaucoma at 1 year and 51% at 5 years postoperatively. The rest, 40% of eyes at the last follow-up, needed antiglaucoma medications, and 7.7% of eyes need glaucoma surgery for IOP control postoperatively.[5] In the current study, all 18 eyes underwent Nd: Yag PI upon presentation. However, it was partially helpful as 13 out of 18 eyes (75%–80%) needed additional antiglaucoma drops to control IOP before the primary procedure.

Table 4: Comparison between baseline and postoperative outcomes

Study	Eyes	Preoperative (baseline) mean±SD (minimum–maximum)					
		Age (years)	BCVA (LogMar)	IOP (mmHg)	SE (D)	ACD (mm)	Antiglaucoma medications
Current study (retrospective case-series)	18	16.4±7.3 (9-35)	0.3±0.1 (0.2-0.5)	21.7±9.5 (8-40)	−12.8±3.5 (−8–20.6)	1.4±0.6 (0.3-2)[§]	1.8±1.3 (0-4)
Yang *et al.*[4]/2016 (prospective case series)	7*	40.28±24.22 (13-67)	0.79±0.36 (0.15-1.3)	28.84±5.36 (20-36)	−11.85±3.71 (−7–18)	1.21±0.55**	4.14±0.9
Study	**Eyes**	**Last follow-up mean±SD (minimum-maximum)**					
		Duration of follow up (years)	BCVA (LogMar)	IOP (mmHg)	SE (D)	ACD (mm)	Antiglaucoma medications
Current study (retrospective case-series)	18	8.8±3.9 (0.7-13.3)	0.2±0.2 (0-0.8)	17.3±4.7 (11-32)	−1.5±1.6 (2–4.5)	3.3±0.5 (2.5-4.1)[§]	1.2±1.2 (0-4)
Yang *et al.*[4]/2015 (prospective case series)	7*	3	0.44±0.38	15.86±0.79	−1.25±0.78	3.16±0.52**	0.71±0.49

*Compared to group 1 in the study (phacoemulsification + CTR + PC IOL), **Ultrasound biomicroscopy the AOD at 500 mm anterior to the scleral spur (AOD 500), [§]Scheimpflug imaging (Pentacam HR; OCULUS Optikgerate, Wetzlar, Germany). BCVA: Best-corrected visual acuity, LogMar: Logarithm of the Minimum Angle of Resolution, IOP: Intraocular pressure, SE: Spherical equivalent, ACD: Anterior chamber depth, CTR: Capsular tension ring, PC IOL: Posterior chamber intraocular lens, AOD Angle opening distance

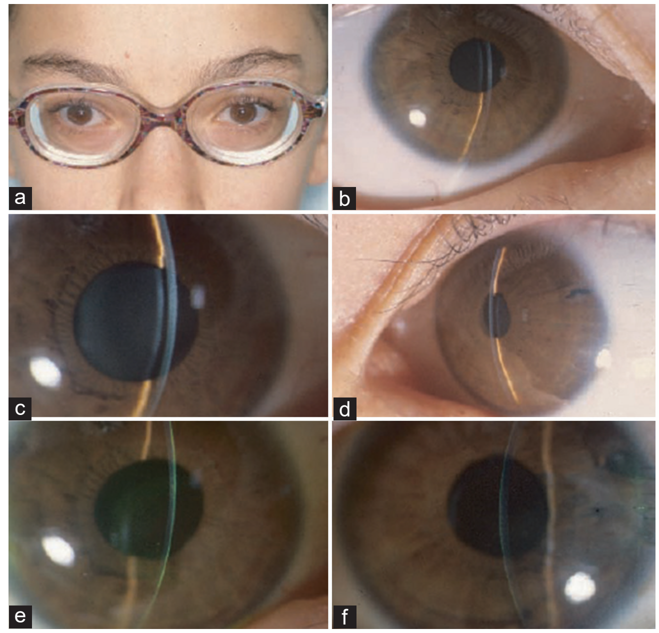

Figure 2: Micropsherophakia with high myopia (a) and iris corneal touch with peripheral iridotomy in OD (b and c), OS (d) Preoperatively and deepening of anterior chamber, clear cornea with intraocular lens in good position in both eyes (e and f) postoperatively

Furthermore, seven eyes (38%) required glaucoma management either before or after primary procedure. Specifically, six eyes (30%) required additional glaucoma surgery postprimary procedure despite maximum antiglaucoma drops. In detail, 2 underwent CPC (CPC = cyclophotocoagulation, one of them was 11 years postprimary procedure, the other was 1-year postprimary procedure then repeated a year later), and 4 had Ahmad valve implant (ranging 1–4 years postprimary procedure, one of them had encapsulated bleb revision 11 years later then CPC) [Table 2]. Likewise, Yang *et al*. reported that two out of seven patients who underwent Phaco + CTR + IOL required additional Ex-press shunts to control, and most patients required additional antiglaucoma drops.[4]

Thus, despite all efforts, lensectomy/lens aspiration alone might not be adequate in controlling IOP and may fail to effectively lower IOP in late presented cases with a synechial angle or eyes with possible associated angle anomaly that existed in the re-opened anterior angle.[4,5,7,16,17] Rao *et al*. also noted that younger age, higher IOP, and larger cup to disc ratio at presentation were risk factors for poor glaucoma control after lensectomy.[5]

Consequently, although our linear graph analysis of IOP showed a favorable decrease over the years, it was not statistically significant [Table 3 and Figure 1]. Similarly, when comparing the last visit to baseline, the decrease in antiglaucoma drops from baseline (preoperative) mean of 1.8 ± 1.3 drops to 1.2 ± 1.2 drops postoperatively at last, follow-up, was not clinically significant as well ($P = 0.076$).

One of the limitations of our retrospective study is its inherent challenges of variable follow-up and some missing data. However, a rare condition such as spherophakia with scarce experience in the literature highlights the value and impact of our reported long-term outcomes and lens capsule complex stability in such challenging cases. Therefore, we also encourage physicians to include gonioscopy, ocular coherence tomography, pentacam, equatorial, and anteroposterior diameter measurements in all spherophakia cases work up as it might help in correlating preoperative findings with outcome or developments of complications.

Conclusion

This is the largest case series and the longest follow-up of lens aspiration + CTR + PC IOL in spherophakic eyes with significant visual improvement and a successful capsular-lens complex stability. This procedure alone might not have a direct impact on IOP or glaucoma progression in such cases. In addition to lens position and shape abnormality factor contributing to glaucoma development, late presented eyes with PAS development and possible underlying angle dysgenesis in some spherophakic eyes might also play a role in uncontrolled IOP postlens aspiration. We believe that early recognition and diagnosis will allow better visual outcome (better uncorrected vision and less ametropia) and prevent undesirable complications in spherorphakic eyes such as secondary glaucoma, progressive shallowing of AC with iris/lens corneal touch, and corneal decompensation. We also recommend early prophylactic lens aspiration, CTR + PC IOL for the following reasons; long-term follow-up proved to have stabilized capsular IOL complex and reduced hospital clinic/ emergency visits and admissions.

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Conflicts of interest

There are no conflicts of interest.

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