Spherophakia

Small Lens, Big Consequences.

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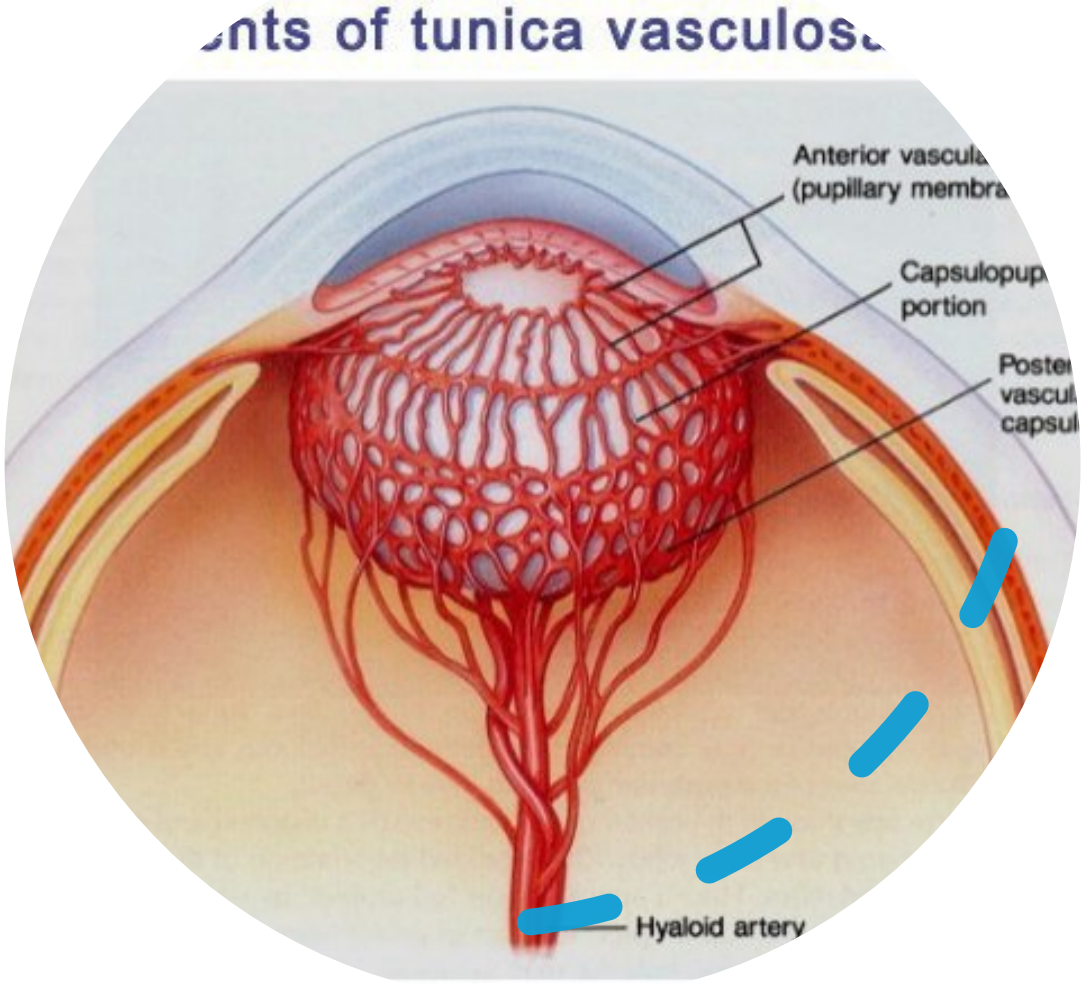
In collaboration with: MEACO MIDDLE EAST AFRICA COUNCIL OF OPHTHALMOLOGY

Spherophakia = Microspherophakia

- Rare bilateral congenital disorder
- longer anteroposterior distance (**4–6.75 mm**)
- smaller equatorial diameter (ED) (**6.5-8.0 mm)**
- Weak zonules, resulting in a triad of
  - ✓lenticular myopia
  - ✓shallow anterior chamber
  - ✓and angle closure glaucoma.



Spherophakia

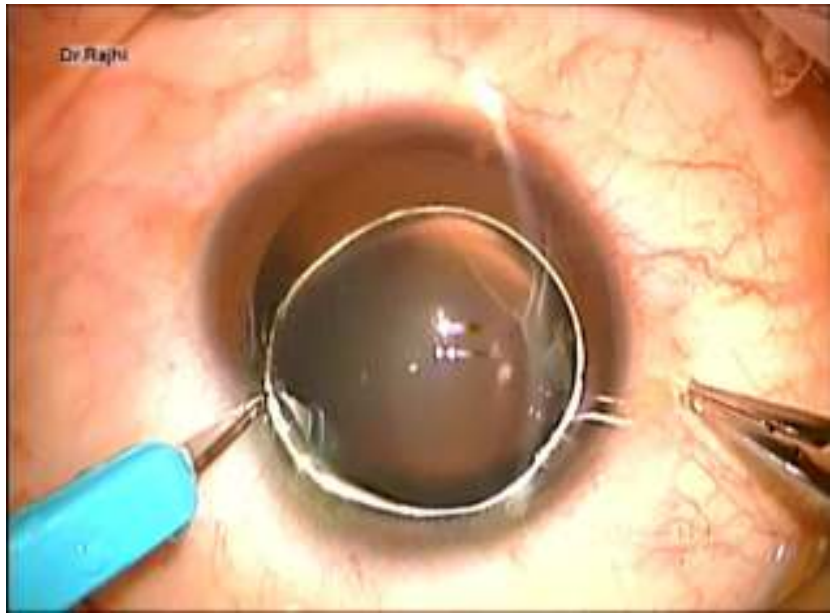
Unknown, but the prevalence of WMS is estimated at 1:100 000 (74-94% incidence of spherophakia).

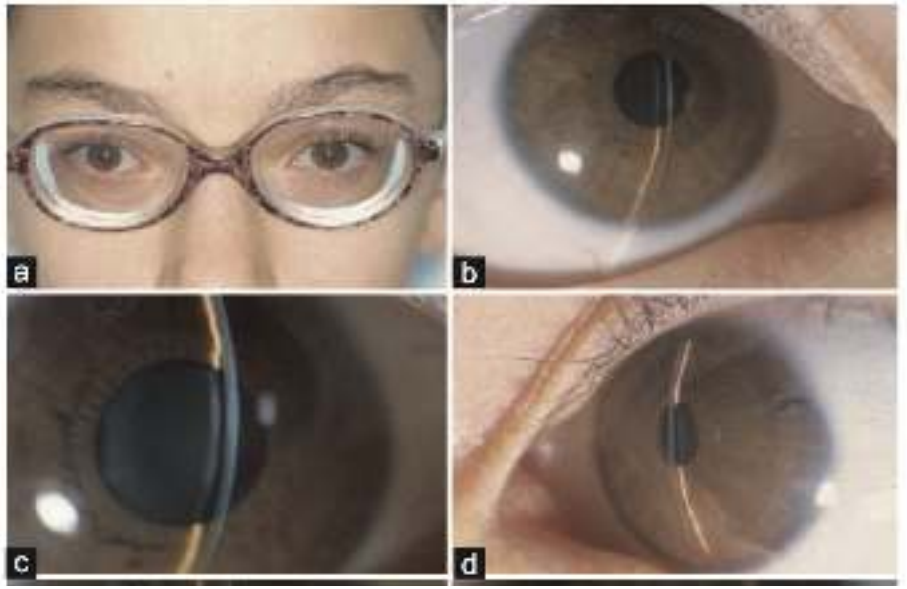
Secondary glaucoma is the main reason for severe permanent loss of vision, and it may be present in up to 51% of MSP eyes.

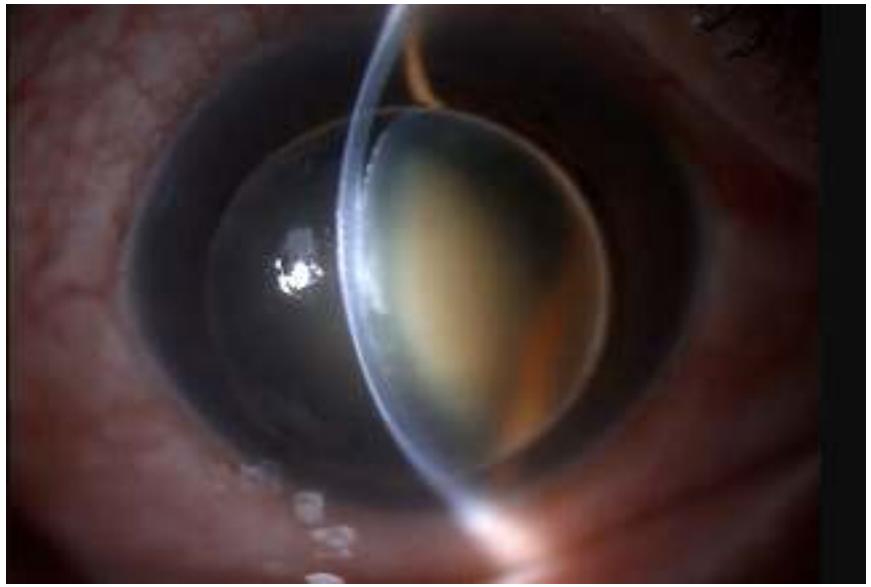
There **is no consensus** in the literature on the definitive treatment of MSP due to the intra-operative surgical difficulties, variable zonular laxity, and the progressive nature of MSP and only collective case series reported short-term outcomes.

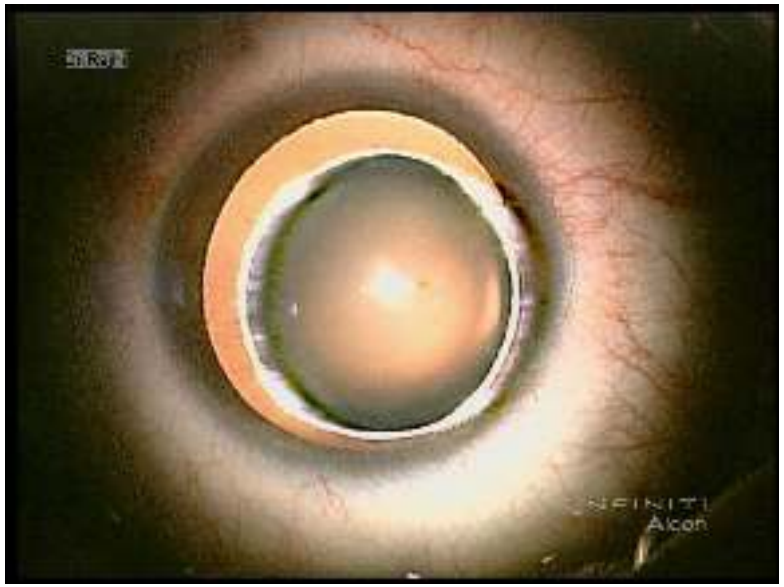
- Désir J, Sznajer Y, Depasse F, Roulez F, Schrooyen M, Meire F, et al. LTBP2 null mutations in an autosomal recessive ocular syndrome with megalocornea, spherophakia, and secondary glaucoma. Eur J Hum Genet. 2010; 18: 761-767.
- Khokhar S, Agarwal T, Kumar G, Kushmesh R, Tejwani LK. Lenticular abnormalities in children. J Pediatr Ophthalmol Strabismus. 2012; 49:32-37.
- Bi Bitar MS, Farooq AV, Abbasian J (2016) Challenges in Diagnosing Microspherophakia in a Pediatric Patient. JSM Ophthalmol 4(1): 1040.

Key features









Courtesy of Almazyad, Alrajhi

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Mechanism of glaucoma in spherophakia

- Pupillary Block (common)
- Lens Crowding
- Chronic Pupillary Block/PAS
- **Angle Abnormalities**

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

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Affiliations + expand

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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.

Methodology

This current study is the first and largest series to report on the long-term outcome and the magnitude of early lens aspiration with the implementation of CTR + PC IOL in MSP with secondary glaucoma.

A retrospective chart review 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 (Ali Al-Rajhi).

Baseline, long-term postoperative outcomes and complications were documented.

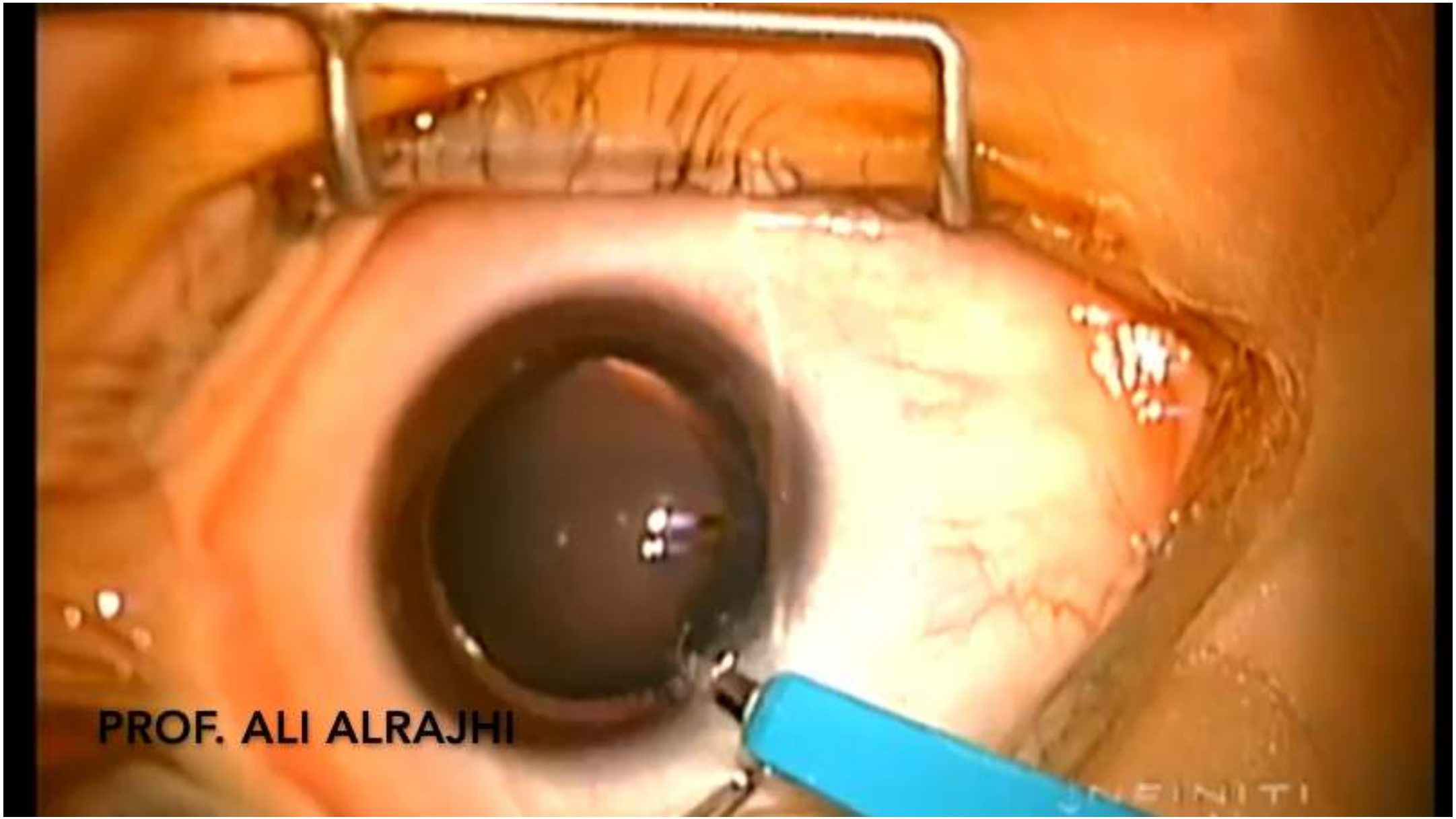
Results

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

✓There was a strong family history of 90%. (4 with a complete picture of WMS, 4 were a partial picture of WMS & 2 were unknown). Genetic mapping displayed *ADAMTS 17* in 3 patients, *ADAMTS 10* in 4 patients, and 1 was unknown.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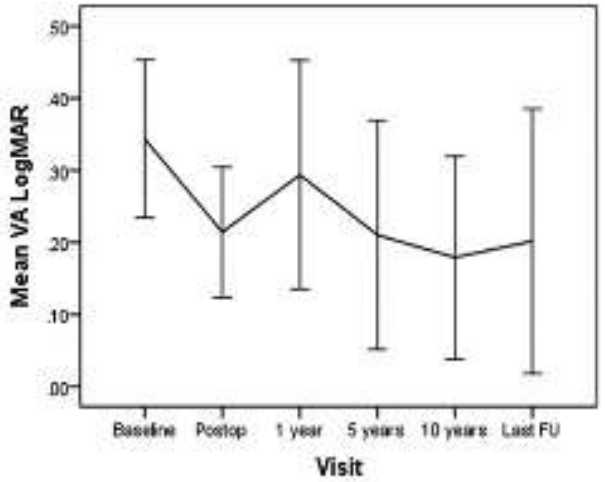
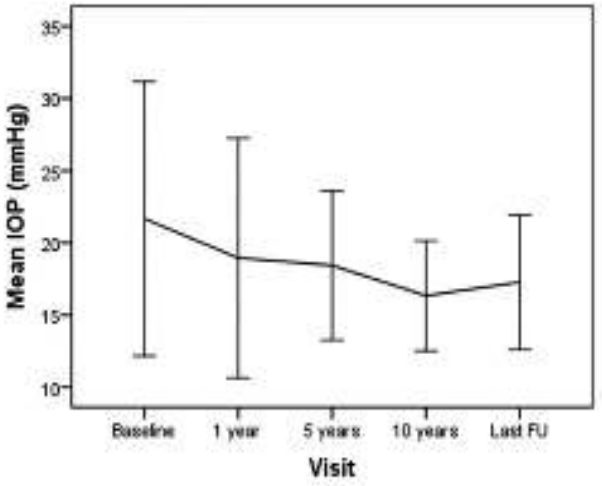
Results





BCVA (LogMar) showed a significant improvement over 1, 5, and 10 years + the last follow-up (mean = 8.8 ± 3.9 years, *P* = 0.006)when comparing BCVA of the last follow-up to baseline.

Likewise, there was an improvement in IOP. However, it showed no clinical significance when comparing IOP of the last follow-up to baseline (*P*-value = 0.131).

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).

Results

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

Results

5/18 eyes did not require antiglaucoma drops (neither pre-operation nor post-operation).

Two eyes remained using the same number of antiglaucoma drops.

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).

>>>3/8 eyes became completely independent from antiglaucoma drops.

Finally, only three eyes needed additional drops (*9 and 5 years after primary procedure*).

Results

Table 2: Preoperative and postoperative details of microspherophakia cases

Case	Age (years)	Prior intervention	Lensectomy to glaucoma surgery (years)	Glaucoma surgery after lensectomy
1	22	Yag PI		-
2	14	Yag PI	11	CPC
3	14	Yag PI	-	-
4	35	Yag PI, CPC	-	-
5	35	Yag PI	-	-
6	17	Yag PI	-	-
7	17	Yag PI	-	-
8	15	Yag PI	-	-
9	15	Yag PI	-	
10	12	Yag PI Trabeculectomy + MMC	2 11 11	Ahmad implant + pericardial patch, Then revision encapsulated bleb Then CPC
11	12	Yag PI CPC twice	1	Ahmad implant + pericardial patch
12	11	Yag PI CPC	1	Ahmad valve implant
13	11	Yag PI Trabeculectomy + MMC	4	Ahmad valve implant
14	17	Yag PI Trabeculectomy + MMC Then AC formation	1 2	CPC Then repeat CPC
15	9	Yag PI	-	-
16	9	Yag PI	-	-
17	12	Yag PI	-	-
18	12	Yag PI	-	

Six eyes had uncontrolled IOP post-primary procedure despite maximum antiglaucoma medications.

Discussion

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	7[*]	3	0.44±0.38	15.86±0.79	−1.25±0.78	3.16±0.52[**]	0.71±0.49

✓Compared to Yang et al , our study showed more clinically significant visual outcome.

✓Moreover, our study subject's age at intervention was much younger , emphasizing the importance of early lens aspiration + CTR + PC IOL in MSP.

Yang J, Fan Q, Chen J, Wang A, Cai L, Sheng H, *et al*. The efficacy of lens removal plus IOL implantation for the treatment of spherophakia with secondary glaucoma. Br J Ophthalmol 2016;100:1087-92.

Discussion

- 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
- Rao et al. articulated that lensectomy is crucial in relieving anterior chamber crowding and secondary glaucoma.
- *This was 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)*

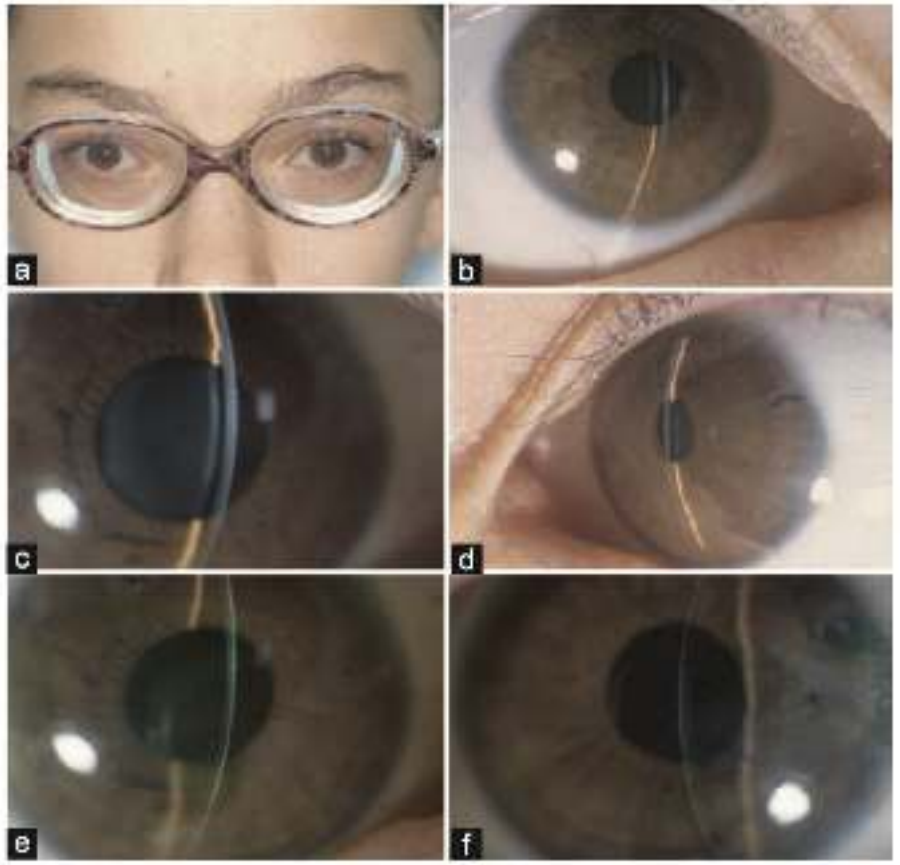


Figure 2: Micropsherophakia with high myopia (a) and iris corneal touch with peripheral iridotomy in OD (b and c), OS (d) Preoperatively and deepening of anterior chamber, clear cornea with intraocular lens in good position in both eyes (e and f) postoperatively

.Yang J, Fan Q, Chen J, Wang A, Cai L, Sheng H, et al. The efficacy of lens removal plus IOL implantation for the treatment of spherophakia with secondary glaucoma. Br J Ophthalmol 2016;100:1087-92.
Rao DP, John PJ, Ali MH, Kekunnaya R, Jalali S, Garudadri CS, et al. Outcomes of lensectomy and risk factors for failure in spherophakic eyes with secondary glaucoma. Br J Ophthalmol 2018;102:790-5.

Discussion

- 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

.

- Rao DP, John PJ, Ali MH, Kekunnaya R, Jalali S, Garudadri CS, et al. Outcomes of lensectomy and risk factors for failure in spherophakic eyes with secondary glaucoma. Br J Ophthalmol 2018;102:790□5.

Discussion

- All of our patients underwent Nd: Yag laser PI to primarily lower IOP following the recommendation in the literature.
- Nevertheless, PI was helpful only partially as many eyes progressed to PAS, shallow AC, and needed medical or surgical glaucoma intervention.

- Yu X, Chen W, Xu W. Diagnosis and treatment of microspherophakia. Cataract Refract Surg 2020;46:1674□9.
- Rao DP, John PJ, Ali MH, Kekunnaya R, Jalali S, Garudadri CS, et al. Outcomes of lensectomy and risk factors for failure in spherophakic eyes with secondary glaucoma. Br J Ophthalmol 2018;102:790□5.
- Senthil S, Rao HL, Hoang NT, Jonnadula GB, Addepalli UK, Mandal AK, et al. Glaucoma in microspherophakia: Presenting features and treatment outcomes. Glaucoma 2014;23:262□7.

Discussion

- 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.
- 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.

- .Yang J, Fan Q, Chen J, Wang A, Cai L, Sheng H, et al. The efficacy of lens removal plus IOL implantation for the treatment of spherophakia with secondary glaucoma. Br J Ophthalmol 2016;100:1087□92.
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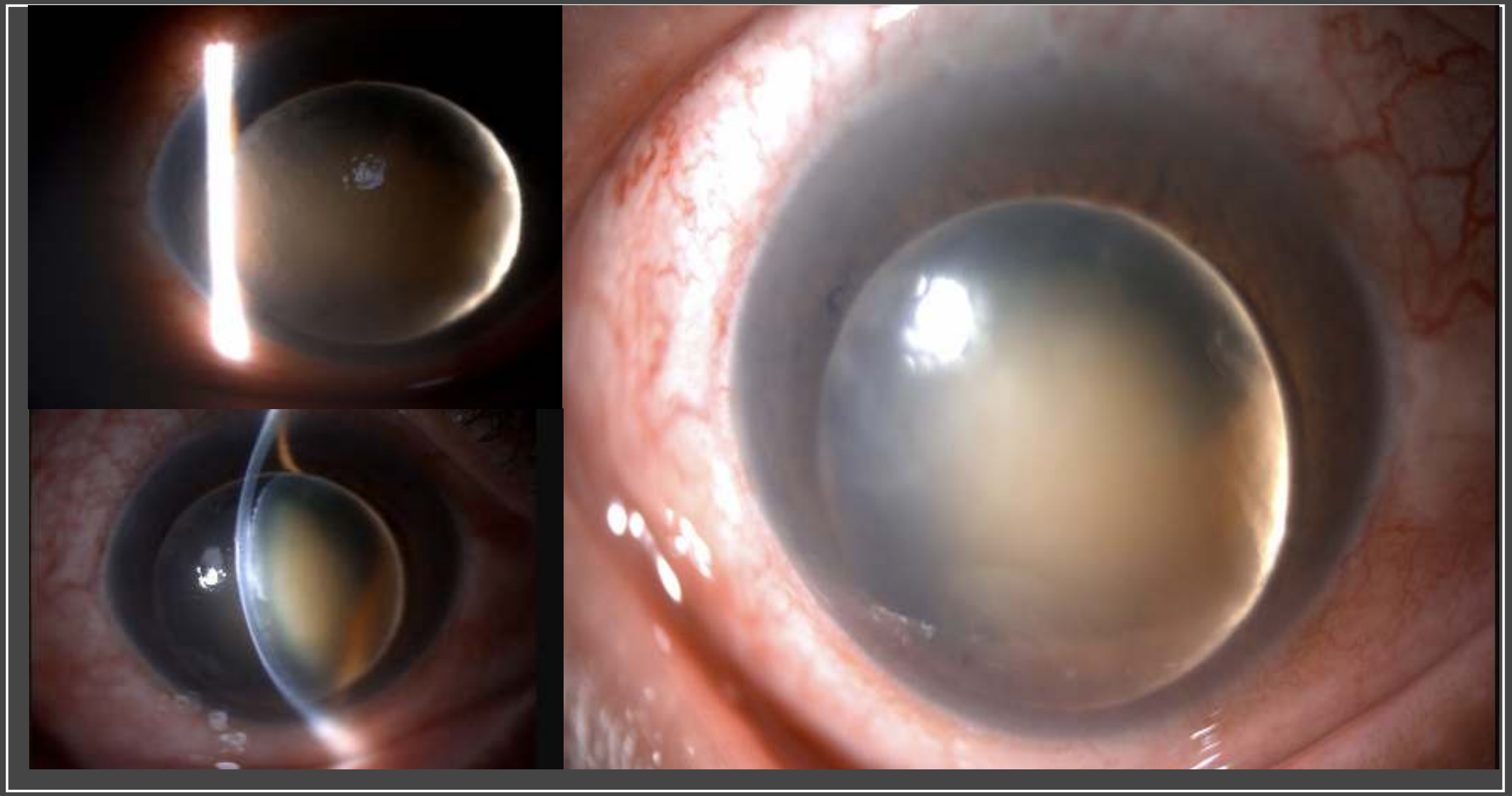
- Rao DP, John PJ, Ali MH, Kekunnaya R, Jalali S, Garudadri CS, et al. Outcomes of lensectomy and risk factors for failure in spherophakic eyes with secondary glaucoma. Br J Ophthalmol 2018;102:790□5.

Late presentation/Complications



Courtesy of: Enmar Almazyad

ENMAR ALMAZYAD , MD

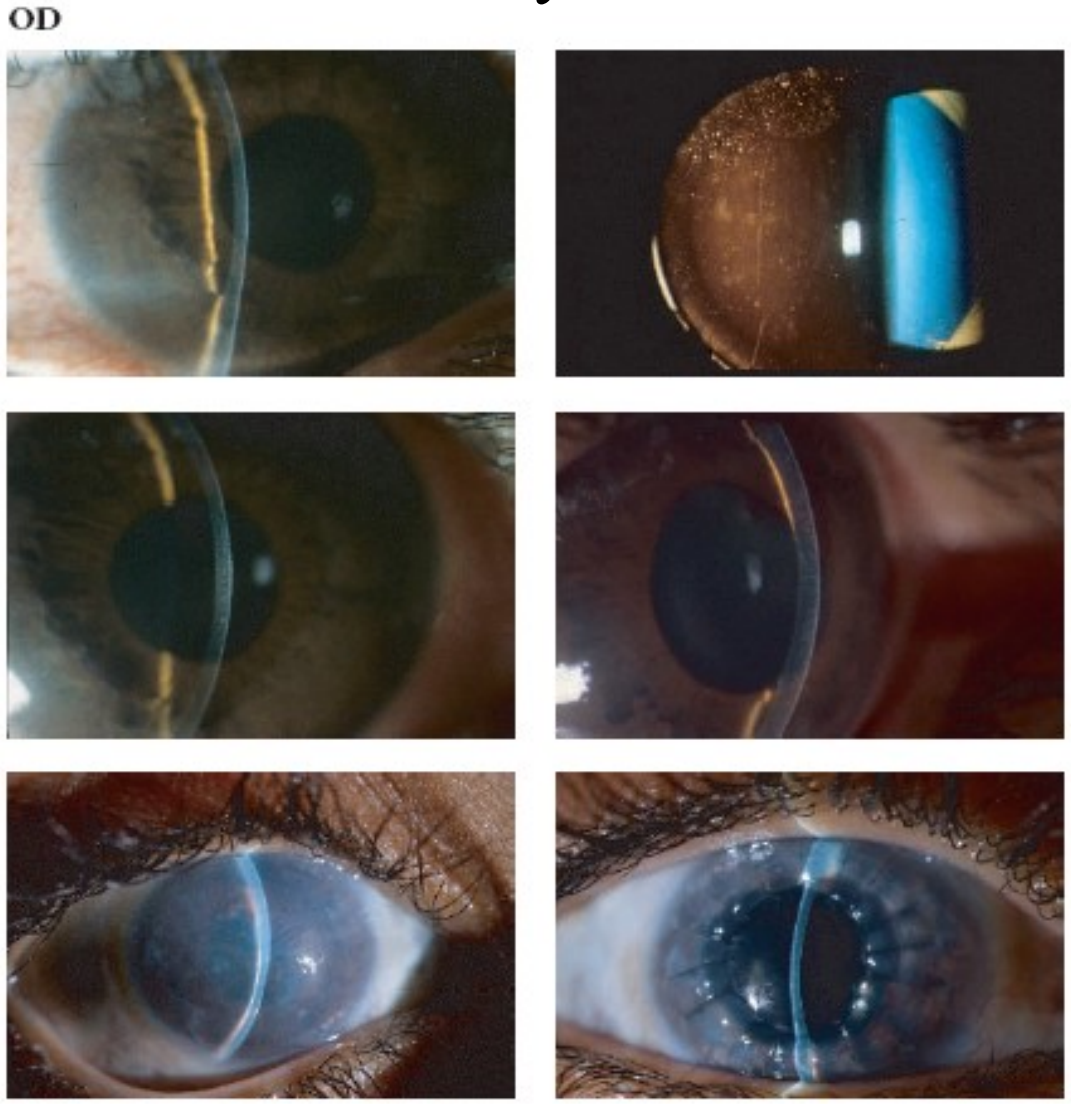
OD PKP+lensectomy and sutured IOL

Postop Va

OD 20/40,

OS 20/100,

Gl procedures for IOP control with total iris corneal adhesions



Courtesy of: Prof. Ali Alrajhi

Summary

- Microspherophakia is a rare disease with a variety of associated systemic syndromes and varied clinical presentations.
- Early identification of the disease, timely visual rehabilitation, and appropriate management of the lens and glaucoma can help us prevent blindness from this condition.
- Current treatment protocols for this condition are mainly based on case reports and retrospective studies with shorter follow-up. Due to the rarity of this disease, designing a large randomized controlled trial to identify the merits and demerits of each management strategy is challenging.
- Multidisciplinary care with lifelong follow-up is recommended, as this typically affects the younger population.

Recommendations

✓We recommend early prophylactic lens aspiration, CTR + PC IOL for the following reasons;

1. *Long□term follow□up proved to have stabilized capsular IOL complex.*
2. *Better visual outcome (better uncorrected vision and less ametropia) and overall quality of life.*
3. *Prevent undesirable complications in spherorphakic eyes such as secondary glaucoma, PAS (chronic glaucoma), progressive shallowing of AC with iris/lens corneal touch, and corneal decompensation.*
4. *Preserving the posterior capsule to maintain the separation between anterior and posterior compartments of the eye is critical to avoid the risk of vitreous loss, cystoid macular edema (CME), and retinal detachment (RD) in these susceptible individuals*
5. *Reduced hospital clinic/ emergency visits and admissions.*

Thank you

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