# Cataract surgery in eyes with associated coloboma: Predictors of outcome and safety of different surgical techniques

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**Purpose:** The aim of this study was to report the outcome of cataract surgery with different surgical techniques in eyes with coexisting coloboma and to define factors of prognostic importance. **Methods:** Retrospective case sheet review of patients presenting between January 2016 and December 2018, who underwent cataract surgery in eyes with coexisting coloboma. **Results:** Of the 3,30,231 cases operated during the study period, 280 eyes of 276 patients had associated colobomatous malformation. The prevalence of coloboma in eyes undergoing cataract surgery was 0.085%. The mean age of the patients was 46.4 years (range 19 -88 years). Phacoemulsification (PE) was performed in 130 eyes (46.4%), manual small incision cataract surgery (M-SICS) was done in 115 eyes (41.1%), and 35 eyes (12.5%) underwent intra capsular cataract extraction. Intra-operative complications were noted in 26 (9%) eyes. Incidence of intra-operative and post-operative complications was comparable between PE and M-SICS groups ($p = 0.94$). The mean corrected distance visual acuity (CDVA) improved from logMAR $1.71 \pm 0.62$ to $0.87 \pm 0.61$ ($p = 0.00009$). On multivariate analysis, microcornea ($p = 0.002$), type 1 and 2 coloboma ($p < 0.001$), and intraoperative complications ($p = 0.001$) were associated with poor visual outcome. **Conclusion:** Favorable functional outcomes can be achieved with phacoemulsification in eyes with softer cataract and corneal diameter >8 mm and with M-SICS in eyes with hard cataracts and corneal diameter of 6–8 mm. PE should be considered as the primary choice whenever permissible by the corneal diameter and severity of nuclear sclerosis. Poor functional outcomes were seen in eyes with smaller corneal diameter, extensive chorioretinal coloboma, and intraoperative complications.

**Key words:** Cataract, coloboma, MSICS, phacoemulsification

Coloboma of the iris, choroid, and retina is a rare congenital anomaly which results from failure of closure of embryonic fissure. Patients with coloboma develop cataract at a much earlier age as compared to a normal population.[1-6] The presence of microcornea, non-dilating pupils, absence of zonules or lens coloboma, and other structural anomalies make cataract surgery more challenging and fraught with complications in these eyes. Additionally, the degree of retinal choroidal coloboma and optic disc abnormality also affects the final functional outcomes.

Several studies have reported outcomes of cataract surgery in eyes with coloboma using different techniques which include intra-capsular cataract extraction (ICCE), extracapsular cataract extraction (ECCE), manual small incision cataract surgery (M-SICS), and phacoemulsification (PE).[2,3,7,8]

Irrespective of the technique used, functional outcomes have been inconsistent.[7-9] Herein, we report the outcomes of cataract surgery in a large cohort of colobomatous eyes from a tertiary center in India, and describe the predictors of the outcomes of different surgical techniques.

## Methods

In this study, we retrospectively analyzed the clinical records of all consecutive patients with ocular coloboma who underwent cataract surgery between January 2016 and December 2018 at a tertiary eye care facility in central India. Only patients with a minimum follow-up of 6 weeks were included in the study. We excluded patients with history of previous intraocular surgeries. The study was approved by the Institutional Review Board and adhered to the tenets of the Declaration of Helsinki.

### Pre/intra operative assessment

Case sheets were reviewed for basic demographic characteristics, pre-operative and postoperative corrected distance visual acuity (CDVA), intraocular pressure (IOP) along with detailed ocular examination. All patients underwent anterior as well as posterior segment examination to look for existing comorbidities. Slit-lamp examination included grade of cataract, microcornea, and zonular loss/phacodonesis. Presence

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of nystagmus and strabismus were also documented. The diagnosis of amblyopia and retinal pathologies was challenging in many cases due to presence of dense cataract. Ultrasound B scan was performed to pick up presence of intercalary membrane defect (ICMD) in doubtful cases. Laser barrage was done pre-operatively or post-operatively to the coloboma margin sparring the macula for eyes with evidence of ICMD clinically or on B- scan. Intra-operative details pertaining to the type of cataract surgery, intra-operative complications, and their management were retrieved from the case sheets.

CDVA was measured using Snellen vision chart, which was converted to logMAR values. The patients were categorized according to the six grades of visual impairment scale. To better elucidate the visual gain, the logMAR values obtained were superimposed on to a standardized international visual impairment scale by the international council of ophthalmology (ICO),[10] This helped us in grading the functional impairment and highlighting achieved change in the grade of blindness.

The maximum horizontal corneal diameter was measured intra-operatively using a caliper. Based on this, the eyes were divided into three categories; grade 1- severe microcornea (<8 mm), grade 2- mild-to-moderate microcornea (8–10 mm), and grade 3- normal (>10 mm). Cataracts were graded according to Lens Opacity Classification System III.[11] The 'Nucleus colour' (NC) was used to define the hardness of the cataract. NC from 1 to 3 was considered 'Soft' cataracts, while categories 4 to 6 were included in 'Hard' cataracts. The chorioretinal coloboma was graded according to Ida Mann Classification.[12]

### Surgical technique and post-operative evaluations

We used the immersion technique to determine the axial length and the SRK-T formula to calculate the IOL power. A mean of 3 readings was used for estimating the axial length. The surgeries were done by multiple surgeons with similar surgical experience of more than five years. The choice of the surgical procedure was at the surgeon's discretion. The choice between PE and M-SICS was guided by severity of microcornea, hardness of cataract and zonular instability. PE was the preferred choice in eyes with corneal diameter >10 mm and in eyes with soft cataract. For eyes with corneal diameter between 8 and 10 mm in the presence of hard cataract, PE was preferred in eyes with no zonular instability. In eyes with microcornea of 6- 8 mm or in the presence of hard cataract, M-SICS was the preferred modality when PE was not permissible due to deep set eyes, shallow anterior chamber and zonular loss [Fig. 1].

Surgical technique required several modifications depending upon the case. PE through sclerocorneal tunnel was preferred in eyes with microcornea and peripheral corneal scarring. Iris hooks were used in semi dilated or poorly dilated pupils. Capsular tension ring (CTR)/cionni ring was used in cases with significant zonular instability. The timing of CTR implantation depended on the severity of bag laxity. CTR was implanted soon after making capsulorhexis whenever laxity was severe. In cases having lesser laxity, CTR implantation was delayed as much as possible till the removal of the cortical matter. During PE, fluid misdirection through colobomatous region may lead to hydration of vitreous leading to vitreous upthrust and shallow anterior chamber. High molecular weight dispersive viscoelastic substance was used over

colobomatous area to prevent misdirection of fluid during emulsification [Fig 2].

Being a retrospective study, no specific universal protocol could be followed. Patients were examined at least on the first postoperative day and then at six weeks with variable visits in between, depending upon the healing course and complication related to the individual needs. Complete ocular examination was done in each visit. Corneal edema was graded according to the Oxford Cataract Treatment and Evaluation Team (OCTET).[13,14] The SUN grading system was used to measure anterior chamber inflammation.[15]

### Statistical analysis

Data analysis was done using STATA 11.2 (College Station, TX, USA). Binary logistic regression analysis was performed to predict the factors associated with poor visual outcome. Chi-square test for goodness of fit was used to measure the association between cataract, microcornea, colobomas, complications and functional outcome with type of procedure, and it was expressed as frequency and percentage. For continuous variables; mean, standard deviation, and ranges were calculated. The mean values were compared using Student's $t$ test or Mann Whitney test. A value of $P < 0.05$ was considered as statistically significant.

## Results

### Demographic details [Table 1]

Out of the 3,30,231 cases operated between 2016 and 2018, 280 eyes had an accompanying colobomatous malformation. The prevalence of coloboma in eyes undergoing cataract surgery was 0.085%. The study cohort consisted of 140 males and 136 females, with a mean age of 46.4 years (range 19-88 years). Bilateral coloboma was present in 143 patients while 97 had unilateral presentation. Thirty-six patients had only one functioning eye.

### Baseline clinical characteristics and associated findings [Tables 1 and 2]

The iridofundal coloboma was the most common presentation (n = 264, 94.3%). Type 3 fundus coloboma was the most common type of chorioretinal (CR) coloboma noted in 145 (53%) eyes. Microcornea was seen in 134 eyes (48%). Hard cataracts (118 eyes, 42%) were also of common occurrence. Zonular weakness and/or visible segmental zonular loss (lens coloboma) was noted in 67 eyes (24%). Other preoperative complicating conditions are listed in Tables 1 and 2.

### Surgical procedures [Table 3]

PE was performed in 130 eyes (46.4%), M-SICS was done in 115 eyes (41.1%), and ICCE was done in 35 eyes (12.5%). Several differences were noticed while comparing pre-operative characteristics between PE and M-SICS groups. Patients who underwent PE were significantly younger in age ($p = 0.0006$), had better pre-operative visual acuity ($p = 0.00002$), and had lesser hard cataracts ($p = <0.00001$). Distribution of macula involving CR coloboma nearly attained statistical significance ($p = 0.052$) with more eyes in M-SICS group. Considering the overall distribution of cases in respect to the cataract density, PE was the preferred choice in 60% (98/162) of the eyes with soft cataracts, while M-SICS was the preferred choice for 53.4% (63/118) of the eyes with hard cataract.

## Flow chart for choosing a surgical technique in coloboma
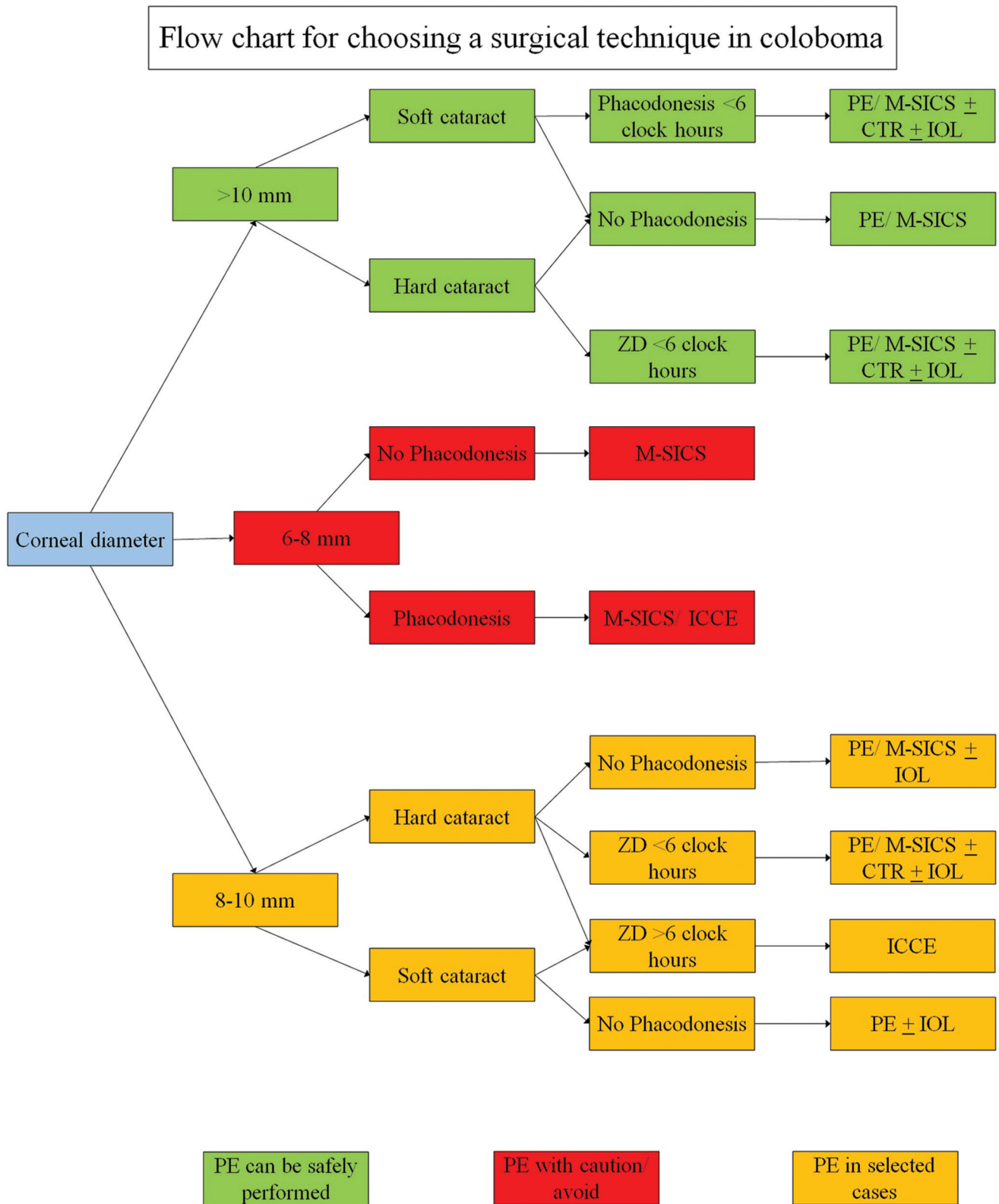
**Figure 1:** Algorithm to guide choosing a surgical technique based on corneal diameter, hardness of cataract and zonular stability. ZD- zonular dialysis, PE- phacoemulsification, M-SICS- manual small incision cataract surgery, CTR- capsular tension ring, IOL- intraocular lens, ICCE- intra-capsular cataract extraction

The majority of eyes in the ICCE group (23/35, 65.7%) had hard cataracts. Microcornea was also a common association seen in 28 (80%) eyes, out of which 12 eyes had severe microcornea. Macular involving coloboma was found in 24 eyes (68%). ICCE

**Figure 2:** Various scenarios and surgical modifications. (a-c) CTR implantation after removal of the cortical matter in phacoemulsification. One end of the CTR is grasped by a non-traumatic forceps and pushed in the capsular bag while another 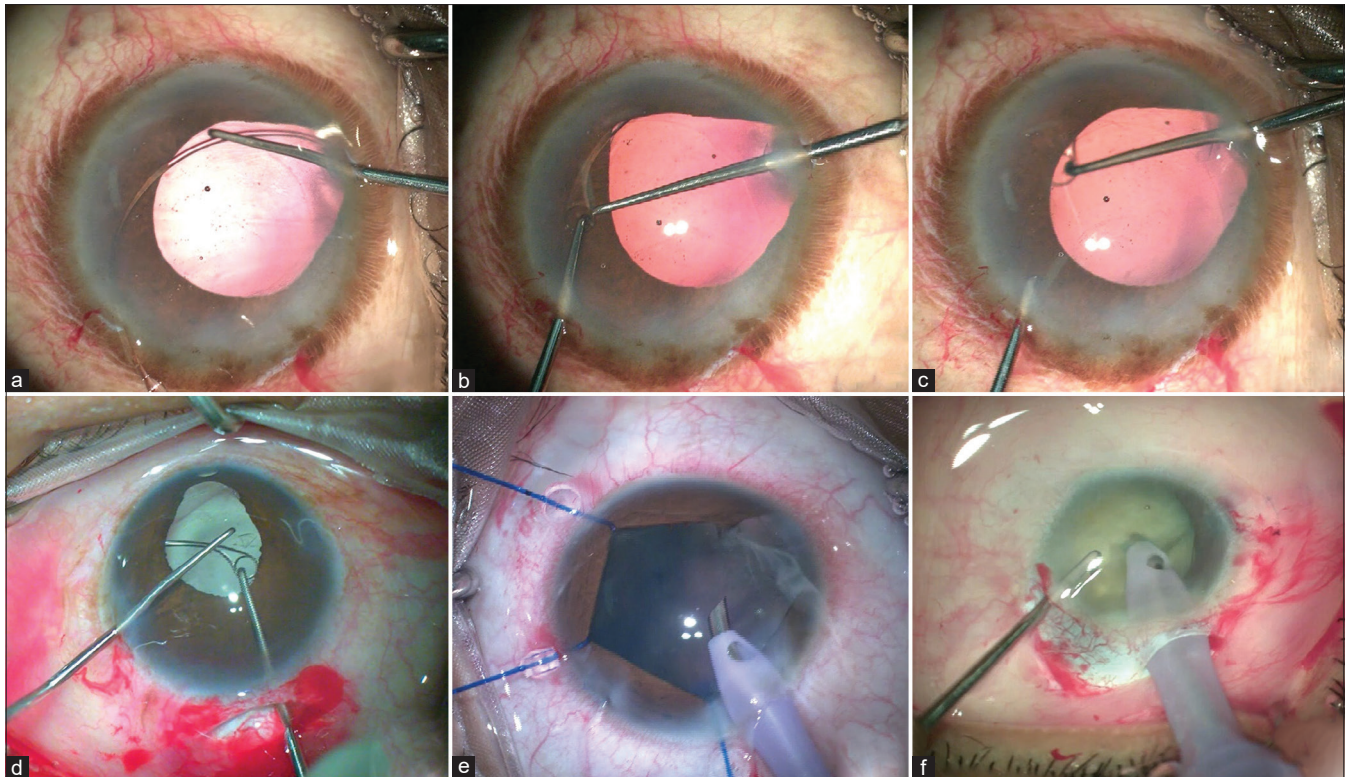blunt instrument (sinskey hook) supports and guides it into proper position. Then sinskey hook holds the second end of the CTR and carefully places it in the bag. (d) CTR implantation using the same approach in M-SICS. (e) Use of 3 iris hooks to improve visualization in poorly dilated pupil. (f) Phacoemulsification using sclerocorneal tunnel in microcornea

had to be performed due to combination of various challenging factors like pre-existing subluxation (*n* = 4), extensive zonular loss (*n* = 16), severe microcornea, and hard cataract.

### Intra-operative complications [Tables 2 and 4]

Complications were noted in 26 (9%) eyes, of which capsulorhexis extension was the most frequent complication in both PE (*n*= 6, 4.6%) and in M-SICS (*n* = 5, 4.3%) procedures. Posterior capsular rent (PCR) was the second most common complication, with similar prevalence in the eyes undergoing PE (*n* = 3, 2.3%) and M-SICS (*n* = 4, 3.4%). Intra-operative zonular dialysis (ZD) was noted in two eyes which were initially scheduled for M-SICS and required conversion to ICCE. Both the eyes had hard cataracts (NC-6).

Descemet membrane detachment (DMD) was noted in 6 (2.1%) eyes, four in M-SICS and two eyes in ICCE group. In 2 out of 6 eyes, DM reposition was not possible due to complete loss of the detached DM flap. The DMD was noted during capsulorhexis in two eyes and after nucleus delivery in four eyes. All these six eyes had microcornea and hard cataracts.

Planned posterior chamber intraocular lens (IOL) implantation was possible in 243/245 eyes (99%) in the PE and M-SICS groups. IOL implantation was not done in 35 eyes that underwent a planned ICCE, while 2 eyes were left aphakic due to ZD in the M-SICS group.

### Post-operative outcome [Tables 2, 3 and Fig. 3]

The incidence of intra-operative and post-operative complications was comparable between the PE and M-SICS
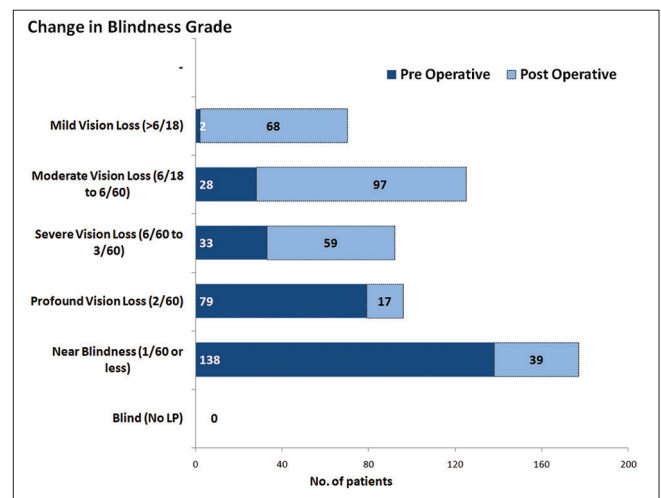


**Figure 3:** This chart represents the different grades of blindness as per the ICD 9 classification and depicts the change in the blindness grade following surgery, among the study cohort

group. Corneal edema of more than grade 2 was the most common observed condition in 29 eyes (10.3%). Repeat intervention was required due to vitreous prolapse into the anterior chamber in 1 aphakic eye.

The mean CDVA for the study cohort was logMAR 1.71 ± 0.62 which improved to 0.87 ± 0.61 following surgery (*p* < 0.0009). At presentation, 217 (77.5%) eyes had

## Table 1: Demography and pre-operative baseline characteristics

| Parameter | Number (%) |
|---|---|
| Mean age (years) | 46.4 (range 19-88) |
| Gender | |
|   Male:Female | 140 (50.7):136 (49.3) |
| Mean preoperative CDVA (logMAR) | 1.71±0.62 |
| Microcornea total | 134 (47.8) |
|   Grade 1, ≤8 mm | 20 (7.1) |
|   Grade 2, >8 - ≤10 | 114 (40.7) |
| Associated ocular pathology | |
|   Nystagmus | 44 (15.7) |
|   Strabismus | 15 (5.3) |
|   Cornea (scar, dystrophy) | 3 (1.1) |
|   Retina (ERM) | 2 (0.7) |
| Cataract density: | |
|   Soft (NC-1,2,3) | 162 (57.9) |
|   Hard (NC-4,5,6) | 118 (42.1) |
| Keratometry: | |
|   Mean K1 | 44.05±2.40 |
|   Mean K2 | 44.92±3.59 |
| Axial length: | |
|   <21 mm | 24 (8.5) |
|   21-25 mm | 154 (55) |
|   >25 mm | 102 (36.4) |
| Mean IOL power calculation (D) | 17.07±6.39 |
| Type of colobomas: | |
|   Mixed Iridofundal | 264 (94.3) |
|   Iris only | 3 (1) |
|   Chorioretinal only | 10 (3.6) |
|   Disc only | 3 (1) |
| Type of CR colobomas (*n*=274) | |
|   1 | 74 (27) |
|   2 | 43 (15.7) |
|   3 | 145 (52.9) |
|   4 | 7 (2.5) |
|   5 | 3 (1.1) |
|   6 | 2 (0.7) |

CDVA- corrected distant visual acuity, NC- nucleus colour, RD- retinal detachment, ERM- epiretinal membrane, IOL- intraocular lens, CR- chorioretinal

blindness grade of ≥3. Following surgery, only 56 (20%) eyes remained in the category ≥3. Overall, 165 eyes (60%) were shifted out of the economic blindness.

### Factors associated with poor visual acuity [Table 5]

Bivariate analysis revealed association of microcornea (p<0.001), macula involving coloboma (*p* < 0.001), intraoperative complications (*p* = 0.009), nystagmus (*p* < 0.001) and male gender (*p* = 0.005) with poor visual outcome. However, male gender and nystagmus were not significant factors in the multivariate analysis. Other significant factors like microcornea and macula-involving coloboma were more common in males than females, which may explain spurious association of male gender with poor functional outcomes in bivariate analysis. Odds of having poor visual outcomes for multiple significant risk factors was 6.2 for microcornea + macula involving coloboma, 9.38 for microcornea + intra-operative complications, 5.02 for macula involving coloboma + intra-operative complications.

## Discussion

In eyes with coloboma, the interplay of various associated anatomical defects collectively poses challenges for cataract extraction. Currently the evidence pertaining to the procedure of choice, risk factors, and outcomes for cataract surgery in eyes with coloboma remains conjectured around small observational series.[3,7-9] In our study, both PE and M-SICS were performed in almost equal proportion. The mean corrected distance visual acuity (CDVA) improved from logMAR 1.71 ± 0.62 to 0.87 ± 0.61 (*p* = 0.00009). On multivariate analysis, microcornea (*p* = 0.002), type 1 and 2 coloboma (*p* < 0.001), and intraoperative complications (*p* = 0.001) were associated with poor visual outcome. Other expected risk factors like nystagmus, strabismus or hard cataracts were not found to play a significant role for poor functional recovery.

Comparison of our findings with previous studies is enlisted in Table 6.

The mean age of presentation for our cases was 46.4 years. This was in keeping with the past observations of early development of cataract in eyes having coloboma.[7,16] Iridofundal coloboma along with macula sparring type 3 CR coloboma was the most common presenting type in our series; an observation similar to the retrospective series by Khokhar *et al.*[3] Disc and macula sparring coloboma was more common in a series of 26 eyes by Chaurasiya *et al.*[7] In a series of 39 eyes by Sahay *et al.*, type 1 and 2 CR coloboma were most commonly seen.[9] We additionally observed frequent occurrence of harder cataracts (42.1%) in our series. Similar incidence of 43.6% was also noted by Sahay *et al.*[9] Since our facility is a tertiary referral centre, it is important to consider that the estimation of the prevalence of cataract with coloboma and other associated defects may be an exaggerated reflection of the true population.

Mohamed *et al.* described a unique morphological type of cataract in uveal coloboma which they named 'coloboma cataract'. Coloboma cataract was not an uncommon finding (29%) in their series which presented in the form of linear lenticular opacity in the colobomatous region. Many patients in our series had much denser cataract with nuclear sclerosis due to late presentation. Hence, we used LOCS 3 grading to classify the degree of nuclear sclerosis to get an estimation of the cataract density and its effect on the decision making while choosing the surgical technique.

### Intra-operative challenges/complications

The colobomatous malformation was not infrequently associated with other structural defects, all of which collectively posed surgical challenges. The risk of incurring complications in such eyes is driven by the predicament of the demanding and complex scenarios which makes atraumatic manipulations challenging. In our series, microcornea was the most common association (48%), the severity of which imposed challenges with wound creation, intra cameral manipulation and served as potential bedrock for incurring complications. Visible segmental zonular loss was the second most frequent association seen in 67/280 (23.9%) eyes of which 45 eyes required a capsular tension ring implantation.

We noted similar incidence of intra-operative complication between M-SICS and PE group. However, the M-SICS group

was more prone for DMD which can be attributed to smaller corneas, shallow anterior chamber, and harder nucleus. The above finding should be interpreted with caution, taking into consideration the unequal distribution of complex cases amongst the groups. The surgical technique of M-SICS and ECCE require prolapsing of the nucleus into the anterior chamber risking contact and trauma to the endothelium. The risk is more in smaller eyes with a larger and harder nucleus

### Table 2: Issues before, during, and after cataract surgeries

| Difficulties/Complications | Number (%) |
|---|---|
| Pre-existing/non iatrogenic conditions | |
|   Segmental zonular loss/subluxated lens | 67 (23.9) |
|   CTR/cionni implanted in pre-existing weak zonules | 45 (16.1) |
|   Small pupil required iris hooks | 36 (12.9) |
|   Adherent leukoma | 1 (0.4) |
|   Vitreous prolapse through colobomas | 4 (1.4) |
| Iatrogenic intraoperative complications (n=26, 9.3%) | |
|   Capsulorhexis extension | 11 (3.9) |
|   Posterior capsule rent/zonular dialysis | 9 (3.2) |
|   DMD* | 4 (1.4) |
|   DM loss | 2 (0.7) |
| Post-operative considerations (n=56, 20%) | |
|   Corneal edema $\geq$ grade 3 | 29 (10.3) |
|   Anterior chamber reaction grade $\geq$ 3 | 18 (6.4) |
|   Hyphaema | 2 (0.7) |
|   Vitreous prolapse requiring vitrectomy | 1 (0.35) |
|   Immediate raised IOP | 6 (2.1) |

CTR- capsular tension ring, DMD- Descemet membrane detachment, IOP- intraocular pressure

and can occur at various steps like wound construction, capsulorhexis, nucleus prolapse or nucleus delivery.

The incidence of complications in our study (9.3%) was lower than the previous series by Khokhar *et al.* (44%), Chaurasiya *et al.* (42%) and Sahay *et al.* (28%).[3,7,9] Other authors predominantly performed PE. PE is more challenging to perform in dense coloboma cataracts and carries higher risk of incurring complications. In M-SICS, manipulations in the bag and related to the nucleus are less. The lower complication rate in our series probably reflects a tailored approach with consideration of M-SICS in more complex cases. Contrary to us, Chaurasiya *et al.* noticed significant complications only in M-SICS/ECCE group.[7] They noted capsulorhexis extension into PCR while performing nucleus delivery or IOL implantation. Again this reflects a selection bias with more complex cases undergoing M-SICS or ECCE compared to PE. Hence, creating large capsulorhexis is important while doing M-SICS.[7]

Such comparison of incidence of complications may be unfair due to key differences in the baseline features. In a study by Sahay *et al.*,[9] microcornea was more prevalent than our series. Smaller number of cases in other studies could also have exaggerated the overall incidence.

### Post-operative visual outcomes and predictors of poor outcomes

Improvement in vision was seen in 248 (88.6%) eyes. Visual gain was achieved in >90% of the cases in the PE and M-SICS group. More importantly, there was a drastic decrease in the burden of cataract related blindness. Out of the 217 eyes with $\geq$ grade 3 blindness pre-operatively, 161 (74%) eyes improved out of the blindness category. The subgroup of eyes not showing improvement was higher in eyes undergoing

### Table 3: Comparison of variables between different surgical procedures: M-SICS versus PE

| Parameter | M-SICS, n (%) | PE, n (%) | P |
|---|---|---|---|
| Total eyes | 115 (41) | 130 (46) | - |
| Mean Age (years) | 49.3 | 43.8 | 0.0006 |
| Mean preoperative CDVA | 1.80 | 1.49 | 0.00002 |
| Cataract | | | |
|   Soft (NC 1,2,3) | 52 (45) | 98 (75.3) | <0.00001 |
|   Hard (NC 4,5,6) | 63 (55) | 32 (24.6) | |
| Microcornea | | | |
|   Grade 1 | 4 (3.4) | 4 (3) | 0.78 |
|   Grade 2 | 44 (38.3) | 54 (41.5) | |
| Type 1 or 2 CR colobomas | 51 (44.3) | 42 (32.3) | 0.052 |
| Pre-existing complications (eyes) | 41 (35.6) | 34 (26.1) | 0.10 |
| Iatrogenic complications (total) | 15 (13) | 9 (6.9) | 0.11 |
| Capsulorhexis runaway | 5 (4.3) | 6 (4.6) | |
| Large ZD - aphakia | 2 (1.7) | 0 | |
| PCR | 4 (3.4) | 3 (2.3) | |
| DMD | 3 (2.6) | 0 | |
| DM loss | 1 (0.9) | 0 | |
| Early Post-operative complications* | 20 (17.4) | 22 (17) | 0.94 |
| Mean post-operative CDVA (logMAR) | 0.86 | 0.65 | 0.00009 |
| Mean post-operative CDVA gain | 0.94 | 0.84 | 0.13 |

M-SICS- manual small incision cataract surgery, PE- phacoemulsification, CDVA- corrected distant visual acuity, NC- nucleus colour, CR- chorioretinal, ZD- zonular dialysis, PCR- posterior capsular rent, DMD- Descemet membrane detachment. *Includes corneal edema $\geq$ grade 3, anterior chamber reaction $\geq$3, hyphema, raised intraocular pressure, vitreous prolapse

**Table 4: Profile of patients who had important intra-operative complications**

| Age/Sex | Pre op CDVA | Type of CR Coloboma | Micro-cornea grade | Cataract grade | Pre or intra operative difficulties | Complication | Surgery | Post op CDVA |
|---|---|---|---|---|---|---|---|---|
| 50/F | 1.78 | 3 | 2 | 3 | | PCR | PE | 0.18 |
| 58/F | 1.30 | 3 | 3 | 3 | | PCR | PE | 0.48 |
| 48/F | 2.0 | 3 | 2 | 3 | Small pupil- iris hooks used, Nystagmus | PCR | PE | 1.0 |
| 51/F | 2.0 | 2 | 2 | 2 | | PCR | MSICS | 1.0 |
| 38/M | 2.6 | 1 | 2 | 5 | Nystagmus, Weak zonules | PCR | MSICS | 1.78 |
| 45/F | 2.6 | 2 | 1 | 5 | Iris hooks used | PCR | MSICS | 2.0 |
| 24/M | 2.0 | 1 | 2 | 1 | | PCR | MSICS | 2.0 |
| 56/F | 1.78 | 1 | 3 | 3 | | ZD | MSICS | 0.78 |
| 45/M | 2.0 | 3 | 3 | 3 | | ZD | MSICS | 1.78 |
| 25/F | 2.6 | 1 | 1 | 4 | Nystagmus | DM loss | ICCE | 2.6 |
| 50/M | 2 | 3 | 2 | 5 | Nystagmus, iris hooks used | DMD | MSICS | 0.78 |
| 45/M | 2 | 3 | 2 | 5 | Subluxation | DMD | ICCE | 1.78 |
| 41/M | 2 | 3 | 2 | 5 | Iris hooks used | DMD | MSICS | 1.48 |
| 25/F | 2.6 | 3 | 1 | 4 | Nystagmus | DM loss | ICCE | 2.6 |
| 75/F | 2.6 | 3 | 2 | 6 | | DMD | MSICS | 2.6 |

CDVA- corrected distant visual acuity, CR- chorioretinal, M-male, F-female, PCR- posterior capsule rent, ZD- zonular dialysis, DMD- Descemet membrane detachment, PE- phacoemulsification, M-SICS- manual small incision cataract surgery, ICCE- intra-capsular cataract extraction

**Table 5: Analysis of factors associated with poor visual outcome after cataract surgery**

| Variables | Bivariate analysis | | | Multivariate analysis | | |
|---|---|---|---|---|---|---|
| | OR | 95% C. I | *P* | OR | 95% C. I | *P* |
| Age | 0.98 | 0.96-1.01 | 0.138 | 0.99 | 0.98-1.02 | 0.969 |
| Male gender | 2.12 | 1.24-3.61 | 0.005 | 1.72 | 0.95-3.14 | 0.07 |
| Microcornea | 3.36 | 1.94-5.85 | <0.001 | 2.73 | 1.42-5.25 | 0.002 |
| Grade 1 or 2CR Colobomas | 3.98 | 2.30-6.90 | <0.001 | 3.76 | 2.03-6.96 | <0.001 |
| Hard cataracts | 1.94 | 1.14-3.27 | 0.013 | 1.69 | 0.92-3.10 | 0.091 |
| Strabismus | 1.72 | 0.59-5.00 | 0.319 | 1.69 | 0.50-5.67 | 0.397 |
| Nystagmus | 3.44 | 1.77-6.67 | <0.001 | 1.50 | 0.69-3.25 | 0.309 |
| Pre-operative Complication | 1.71 | 0.99-2.95 | 0.052 | 1.17 | 0.63-2.18 | 0.625 |
| Intra-operative Complication | 3.04 | 1.32-6.99 | 0.009 | 4.98 | 1.89-13.16 | 0.001 |

CR- chorioretinal

ICCE (37%). This could be attributed to presence of multiple factors in ICCE cases like, extensive posterior pole involving coloboma (69%), microcornea (80%), postoperative corneal edema and inflammation (40%), an observation also found in the series by Sen *et al*.[17] The visual outcomes in our study were not influenced by the presence of preoperative complications, grade of cataract and the choice of surgery but remained influenced by the presence of microcornea, macula involving coloboma, and intraoperative complications. Khokhar *et al*. also noted poor visual outcomes in the fovea involving coloboma.[3]

The postoperative vision (0.87 logMAR) in our series was similar to that attained by Khokhar *et al*. (0.96 log MAR) but better than Sahay *et al*. (1.64).[3,9] Poor vision in the later study may be attributed to higher incidence of microcornea (74.3%) and macula involving coloboma (46%).

### The preferred surgical technique: Choosing between PE/M-SICS/ICCE

Even though PE is accepted as the current gold standard for the management of cataracts, its safety profile in comparison to M-SICS in eyes with coloboma remains questionable due to lack of comparative studies. The concerns of incision size, surgery-induced astigmatism and good IOL centration, depict the advantages of undertaking a PE procedure, but little do these benefits affect the outcomes of eyes already compromised by severe microcornea and extensive coloboma of the fundus. Amidst the limited evidence, the natural selection in our series was guided by surgeons' preferences, severity of microcornea, hardness of cataract and other pre-operative complicating situations.

The selection procedure for the surgical technique in our study was similar to that by Chaurasiya *et al*.[7] In their series, M-SICS and ECCE were the preferred choice in eyes with harder cataracts and/or phacodonesis, while PE was done in soft cataracts. Due to obvious selection bias created by asymmetrical distribution of cases between the PE and M-SICS groups, it remains only logical to assume that the two procedures are similar in their outcomes but under different scenarios. Cataract surgery in eyes with coloboma is fraught with complications and the aim should be to choose a pragmatically simpler approach which requires less intraocular manipulation.

**Table 6: Comparison of studies of cataract surgeries in coloboma**

| Parameter | Chaurasia et al.[7] | Khokhar et al.[3] | Sahay et al.[9] | Current study |
|---|---|---|---|---|
| Cases (n) | 26 | 22 | 39 | 280 |
| Mean age of patients (years) | 37.6 | 27.7 | 36.7 | 46.4 |
| Mean preoperative CDVA (logMAR) | NA | 1.3±0.53 | 1.83±0.31 | 1.71±0.62 |
| Microcornea, n (%) | 12 (46) | 6 (27.3) | 29 (74.35) | 134 (47.8) |
| Axial length (mm) | NA | NA | 23.52 | 23.9 |
| Macular involvement of coloboma, n (%) | 8 (30.7) | 5 (22.7) | 18 (46.15) | 117 (42.7) |
| Grade of cataract, n (%) | | | | |
| Soft | 16 (62) | 15 (68) | NA | 162 (57.9) |
| Hard | 10 (38) | 7 (32) | NA | 118 (42.1) |
| Type of surgery | | | | |
| PE | 16 (61.5) | 22 (100) | 22 (56.4) | 130 (46.4) |
| M-SICS | 0 | 0 | 5 (12.8) | 115 (41.1) |
| Lens aspiration | 6 (23) | 0 | 2 (6.7) | 0 |
| ECCE | 3 (13) | 0 | 5 (12.8) | 0 |
| ICCE | 1 (3.8) | 0 | 4 (10.3) | 35 (12.5) |
| PPL | 0 | 0 | 1 (2.6) | 0 |
| Intraoperative complications, n (%) | | | | |
| Capsulorhexis extension | 5 (19.2) | 0 | 1 (2.6) | 11 (3.9) |
| PCR | 6 (23) | 1 (4) | 3 (7.7) | 7 (2.5) |
| ZD/bag dialysis | 0 | 9 (40) | 7 (17.9) | 2 (0.7) |
| DMD | 0 | 0 | 0 | 6 (2.1) |
| Use of CTR, n (%) | 7 (26.9) | 9 (40) | 6 (15.3) | 45 (16.1) |
| IOL implantation, n (%) | 21 (72.4) | 19 (86) | 27 (69.2) | 243 (86.8) |
| Immediate Postoperative complications, n (%) | | | | |
| Raised IOP | NA | 0 | 11 (28.2) | 6 (2.1) |
| Corneal edema | NA | 4 (18) | 11 (28.2) | 29 (10.3) |
| Hyphaema | NA | 0 | 0 | 2 |
| Follow-up mean duration (month) | 7.4 | NA | 5.9 | 1.5 |
| Mean post-op CDVA | NA | 0.96±0.55 | 1.64±0.51 | 0.87±0.61 |

CDVA- corrected distant visual acuity, PE- phacoemulsification, M-SICS- manual small incision cataract surgery, ECCE- extracapsular cataract extraction, ICCE- intra-capsular cataract extraction, PPL- pars plana lensectomy, PCR- posterior capsular rent, ZD- zonular dialysis, DMD- Descemet membrane detachment, IOP- intraocular pressure, NA- not available

Final visual acuity was better in PE group than in M-SICS group. However, mean visual gain was found to be comparable between both groups. The eyes in the PE group had better pre-operative vision, lesser challenges in terms of hard cataracts and pre-operative complications, and less severe colobomatous defect, which explain the achievement of better CDVA.

Our observations highlight the safety and efficacy of M-SICS in eyes with harder and PE in eyes with softer cataracts. Even though we don't intend to condemn the use of PE in eyes with hard cataracts, we propose caution to be exercised when considering a procedure in the setting of severe microcornea, shallow anterior chamber, and hard cataract. The data pertaining to safety of M-SICS in eyes with severe microcornea is currently limited and needs further comparative trials for conclusive deductions.

A planned ICCE through a self-sealing sclero-corneal tunnel was done in 35 eyes with severe grade of microcornea and/or existing severe zonular weakness, wherein PE or M-SICS was deemed not possible by the operating surgeon.

The safety of cataract surgery in eyes with smaller corneas of less than 6 mm has not been dealt with in our report. Various modifications for PE have been described to reduce the intra- operative struggle for eyes with small corneas. Khokhar

et al. studied the safety of PE in corneas of less than 9 mm using modified scleral tunnels.[8] In their series of eight eyes, three eyes had a corneal diameter of even <6.5 mm. Posterior incision helped minimizing the risk of port-site Descemet detachment, while imparting greater wound strength. A pars plana approach for phacofragmentation has been described by Sen et al. with successful outcomes in such eyes. Anterior approach was deferred in anticipation of sight threatening complication.[17]

### Limitations

Being a retrospective series, there are pertinent concerns due to selection bias and standardization of the extracted data. Since the study involved multiple surgeons, it may have affected the procedural choices due to individualistic preferences and inhibitions, similarly affecting the outcomes also. What appears as a selection bias in choosing a particular surgical technique, reflects the inclination of surgeons in choosing a pragmatically safe technique for the given operative scenario. Considering the low prevalence of coloboma, majority of the existing studies on coloboma are retrospective in design with data spanning over years. This also explains why it is practically difficult to involve only one surgeon or only one investigator for doing the evaluation. Even though multiple surgeons were involved in the study, they all shared a common surgical experience

for operating in eyes with coloboma or microcornea and were equally proficient in both PE and M-SICS.

The data on anterior chamber depth and its association with the outcomes remains under evaluated in our series. Subgroup analysis of the eyes with severe microcornea undergoing PE was not possible due to the small number of cases; this limits us from elucidating the safety of PE for eyes with extreme grades of microcornea. Long term post-operative and visual outcomes were not studied in the present study. We emphasized on the intra-operative safety of the different surgical techniques along with their effect on the immediate post-operative recovery and visual rehabilitation.

### Strengths

Most of the literature on the surgical outcomes of coloboma has been evaluated through small retrospective series, the limitations of which are many. We present a robust data of 280 eyes with comparative evaluation of PE and M-SICS procedures. The retrospective evaluation of our series provides us insight into the patient related factors dictating the surgeon's choices in eyes with coloboma.

## Conclusion

Both PE and M-SICS provide good post-operative outcomes with similar intra-operative risks and gains. PE should be considered as the primary choice whenever permissible by the corneal diameter and severity of nuclear sclerosis. Microcornea, macula involving CR coloboma, and intraoperative complications were important factors leading to poor functional outcomes.

### Financial support and sponsorship
Nil.

### Conflicts of interest
There are no conflicts of interest.

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