

Cataract Management-“A Surgical Challenge in Typical Iris Colobomas”

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ABSTRACT

This prospective study analysed 15 patients aged 18 years and above of congenital typical iris coloboma with cataract. In these patients iris coloboma was present in isolation or associated with other ocular structures, (lens, choroid, retina, and optic nerve) with or without ocular abnormalities (predominantly microphthalmia, microcornea, strabismus etc. Detailed evaluation of patients was performed on slit lamp biomicroscopy. Best corrected visual acuity, intra ocular pressure, vertical and horizontal corneal diameters and curvature, grade of cataract and subluxation, type of coloboma, Axial length and IOL powers were recorded. Cataract surgery was performed by either phacoemulsification or small incision cataract surgery. Intraoperative challenges handled in patients with shallow anterior chamber, small pupil, microcornea and microphthalmia. Challenges became more profound in zonular dialysis extension and vitreous herniation into anterior chamber. Anterior vitrectomy was performed meticulously in such cases and intraocular lens were placed if possible. 1 month follow up was done to determine post surgical visual outcome.

Eyes with microcornea (33.33%), microphthalmia (26.66%), small pupil (13.33%), shallow anterior chamber (59.99%) with irido fundal colobomas (73.33%) posed various surgical challenges which were appropriately managed to avoid post operative complications and as per the situation decision to implant IOL (66.66%) or not (33.33%), 1 month post operative visual acuity in these colobomatous eyes was satisfactory (66.66%).

Cataract in coloboma patients is the only treatable cause of visual disability hence, careful pre operative evaluation with modifications of the intra operative challenges are necessary to avoid post operative complications.

Key words: Micro cornea, Micro ophthalmos, Typical iris coloboma, Fundal coloboma, Cataract.

INTRODUCTION

Colobomas are congenital defects consistent with failure of closure of the fetal fissure. They may involve any of the ocular structures, including the iris, lens, choroid, retina, and optic nerve⁽¹⁾ and can occur in isolation or in association with other ocular abnormalities (predominantly microphthalmia (<20mm), nystagmus, strabismus, anophthalmia)

Colobomas are commonly accompanied with formation of cataract which is the only important treatable cause of visual disability. The incidence of nuclear sclerosis is much higher

in the cases of iris coloboma with microcornea and irido-fundal colobomas compared to the general population.[1] The presence of microcornea (horizontal diameter <10mm), 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 retino choroidal coloboma and optic disc abnormality also affects the final functional outcomes. (2)

The use of standard techniques for cataract management like phacoemulsification (PE), small incision cataract surgery (SICS), extra capsular cataract extraction (ECCE) in these small eyes are challenging due to the risk of serious

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DOI: 10.33309/2639-8788.040104

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intraoperative and postoperative complications. Our purpose is to study surgical difficulties during cataract management in colobomatous eyes.

MATERIALS AND METHODS

It is a prospective study conducted on patients of congenital coloboma associated with cataract in an ophthalmic department of a tertiary care hospital of central India.

INCLUSION CRITERIA

- 1) All patients above 18 years of age having cataract with coloboma.
- 2) Patients having congenital colobomas with cataract and associated other ocular co morbidity like microphthalmos, strabismus, nystagmus, anophthalmos.

EXCLUSION CRITERIA

- 1) Patients below 18yrs of age.
- 2) Patients with history of previous intraocular surgeries
- 3) Patients having uncontrolled diabetes and hypertension.

Detailed evaluation of patients of congenital colobomas who presented in our OPD was done on slit lamp biomicroscopy (SLM) .The best corrected visual acuity (BCVA), intra ocular pressure measurement by goldmann applanation tonometry,vertical and horizontal corneal diameters, grade of cataract, type of coloboma, Keratometry, Axial length of eye and the SRK-T formula was used to calculate the IOL power. Presence of nystagmus and strabismus were also documented. The Ultrasound B scan was performed in dense cataract. After primary preoperative evaluations and cataract surgery was performed by single surgeon. The method of cataract extraction was decided based upon the density of cataract and associated features like corneal diameter, anterior chamber depth, size of pupil, zonular weakness, axial length and visual outcome of the eye at the surgeon’s discretion. The choice between Phacoemulsification (PE) and Small incision cataract surgery (SICS) was guided by severity of microcornea, hardness of cataract and zonular instability. Criteria for preferring PE were eyes with **a)** Corneal diameter >10 mm with soft cataract. **b)** Corneal diameter between 8 and 10 mm in the presence of hard cataract **c)** Eyes with no zonular instability.

In eyes with microcornea of 6- 8 mm or in the presence of hard cataract, deep set eyes, shallow anterior chamber and zonular loss , SICS was the preferred modality.

All patients were followed-up for at least 1 month to determine the post surgical visual outcome. Significant improvement in visual acuity was noted after the surgery without major complications. (Figure 1 and Figure 2)



Figure 1. shows microcornea(6-8mm)



Figure 2. shows complicated cataract

RESULTS

On gender distribution in coloboma patients (66.66%) male preponderance was seen in our study as compared to the minor female population which was (33.33%).(Table 1)

Table 1. Sex Distribution In Coloboma Patients

No. of patients	Sex Distribution
10	Male
5	Female

On age distribution in these coloboma patients, maximum patients(46.66%) in our study belonged to the age group of 18-30 years, (40%) in the age group of 31-50 years and (13.33%) in the age group of 51-70 years.(Table 2)

Table 2. Age distribution in coloboma patients

No. of Patients	Age distribution(in years)
7	18-30
6	31-50
2	51-70

The grading of cataract in our study played an important role as to the choice of surgery to be performed as shown in (Table 4) which in our study (46.66%) had hard brown cataract, (40%) had mature cataract, (6.6%) were senile immature and complicated cataract respectively.

On comparing the different types of colobomas in our study,(Table 4)(73.33%)patients had colobomas involving the iris and choroid both(irido fundal colobomas),while only (26.66%) had coloboma involving the iris alone(iris colobomas).The extent and the type of colobomas were related with the visual outcome in these patients.(Figure 3 and Figure 4)

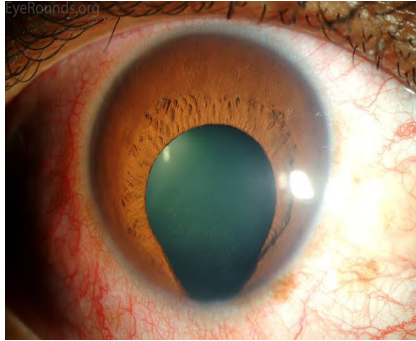


Figure 3. shows typical iris coloboma

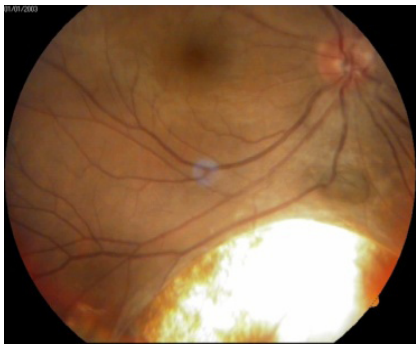


Figure 4. shows type 3 fundal coloboma

Table 3. Grading of Cataract in Coloboma Patients

No. of Patients	Grading of Cataract
1	Immature Senile Cataract
6	Mature cataract
7	Hard brown cataract
1	Complicated cataract

Table 4. Types of colobomas

No. Of patients	Types of coloboma
4	Only typical iris coloboma
11	Typical iris with fundal coloboma

Coloboma has associations with various ocular anomalies which were present in our study as well.(33.33%)micro cornea and micro ophthalmos both,(26.66%)had micro ophthalmos alone,(13.33%) had micro cornea alone while only a minority (6.6%) had nystagmus and strabismus with associated corneal opacity, as shown in (Table 5)

The pre operative vision in coloboma patients varied significantly due to associated ocular anomalies as shown in (Table 6),(73.33%) patients in our study had a pre operative vision of finger counting or hand movements due to dense cataract with associated irido fundal colobomas, while (13.33%) had a pre operative vision of PL+ who had grade 5 coloboma according to the Ida Mann classification and remaining (13.33%) had PL + with PR inaccurate due to extensive colobomas involving peripheral retinal thinning.

Table 5. Associated ocular anomalies in coloboma patients

No. of patients	Associated ocular anomaly
4	Micro ophthalmos alone
2	Only micro cornea
5	Micro cornea and micro ophthalmos
1	Nystagmus with strabismus
2	Strabismus Alone
1	Corneal opacity

Table 6. Pre operative vision in coloboma patients

No. of patients	Pre operative vision
2	PL+
2	PL+,PR inaccurate
11	Hand movement/finger counting

Colobomatous eyes posed us with various intra operative challenges.(73.33%)patients presented with poor fundal glow,(33.33%) had micro ophthalmos and microcornea posing challenges with surgical incision, intra cameral manipulation and IOL implantation,(26.66%) had micro ophthalmos alone,while (13.33%) presented with small pupils with zonular weakness and phacodonesis.(Table 7)

Table 7. Intra operative challenges in coloboma patients

No. of patients	Intra operative challenges
4	Micro ophthalmos alone
2	Only micro cornea
5	Micro cornea and micro ophthalmos
2	Small pupil
2	Phacodonesis with zonular weakness
11	Poor fundal glow

On comparing the intra operative complications in our study (Table 8), (33.33%)had posterior capsular rent,(20%) had vitreous loss, while (13.33%) had zonular weakness.

Various intra operative challenges(micro ophthalmos,micro cornea,fundal colobomas) and intra operative complications

were the deciding factor for IOL implantation. (66.66%) patients were pseudophakic while (33.33%) were left aphakic in our study.(Table 9)

Table 8. Intra operative complications in coloboma patients

No. of patients	Intra operative complications
5	PCR
2	Zonular weakness
3	Vitreous loss

Table 9. IOL implantation in coloboma patients

No. of patients	Iol implantation
5	Aphakia
10	Pseudophakia

On comparing the 1 week post operative BCVA in these colobomatous eyes post cataract extraction as shown in (Table 10),(40%) patients had vision of finger counting-hand movement,(26.6%)had vision in the range of 6/24-6/60,(20%) vision in the range of 6/6-6/18, while (13.33%) had a vision of 5/60-1/60.

Table 10. Post operative(1 week) BCVA in coloboma patients

No. of patients	Post operative-1 week bcva
3	6/6-6/18
4	6/24-6/60
2	5/60-1/60
6	Finger counting-hand movement.

The post operative(1 month)BCVA in these coloboma patients were satisfactory in which ,(40%) had vision in the range of 6/24-6/60,(26.66%) had vision of 6/6-6/18,(20%) had 5/60-1/60 vision while (13.33%)were finger counting-hand movement.(Table 11)

Table 11. Post operative(1 month) bcva in coloboma patients

No. of patients	Post operative-1 month bcva
4	6/6-6/18
6	6/24-6/60
3	5/60-1/60
2	Finger counting-hand movement/PL+

DISCUSSION

Cataract surgery in eyes with coloboma presented with various intra operative challenges and possessed higher risk of complications because of the associated ocular malformations. Most studies reported significant cataract formation at a

young age as compared with the age-related cataract; a similar observation was noted in our study. The mean age of presentation in our study was 40 years in which (46.66%)were in 18-30 years of age group and the remaining (53.44%)were in the elder age group age with a male preponderance(66.66%) similar to other retrospective series[7] [2]

Additionally we observed majority of the cases(46.66%) patients had hard brown cataract,(40%) had mature cataract and the remaining (13.33%) had complicated and immature senile cataracts which is one of the important factor for selecting the surgical approach {phacoemulsification or SICS},(60%) patients in our study underwent phacoemulsification when other factors also came into play, similar observations were seen in studies by Sahay *et al* [9] in their retrospective case series(2).

Coloboma can occur in isolation or in association with other ocular anomalies, as in our study (73.33%)patients had irido fundal colobomas and (26.66%)had only typical iris colobomas associated with ocular anomlies like micro cornea(horizontal diameter<10mm) , microphthalmia (<20mm), nystagmus, strabismus along with corneal opacity . these imposed challenges during wound creation, intra cameral manipulation, capsulorhexis formations, iol implantation etc. similar to retrospective series by Khokhar *et al.*[3.]

Pre operative vision of finger counting or hand movement were noticed in (73.33%) patients due to significant cataract and fundal colobomas,(13.33%) patients had a vision of PL+ with inaccurate PR which was thought to be related to colobomas involving the macula/optic nerve or in choroidal colobomas associated with complications(Retinal Detachment).similar observations were seen in retrospective series of Chaurasia *et al*(7)

Intra-operative challenges/complications.

The colobomatous malformation was frequently associated with other structural defects, all of which collectively posed surgical challenges. In our series, microcornea was the most common association (48%), followed by micro ophthalmia in (26.6%). These conditions imposed challenges with wound creation, and intra cameral manipulations due to their association with shallow anterior chambers which were managed by increasing the infusion bottle height and by using high viscosity visco elastics (dispersives).

Poor pupillary dilatation(13.33%) was another difficult situation we encountered , as capsulorhexis and nucleus delivery became challenging , in these cases Pupillary diameters of 2mm to 3mm were managed by using kuglen’s iris hooks(7.9%) and in 3mm to 4mm multiple sphincterotomies were performed.(5.4%)Capsulorhexis were performed with 26 gauge cystitome, in shallow anterior chambers rhexis was

performed by increasing the infusion bottle height and by using high viscosity visco elastics (dispersives). Smaller eyes with a larger and harder nucleus had difficulty in nucleus prolapse or nucleus delivery. In 46.6% patients of hard brown cataracts and 40% mature cataracts the preferred choice of surgery was SICS although soft mature cataracts and immature cataracts were managed with PE. (13.33%) of patients had zonular weakness which was managed by using capsular tension rings when the dialysis was upto 4 clock hours, cionni rings with Ior 2 eyelets were used for ,4-6 clock hour dialysis

(33.3%) patients during cataract surgery had posterior capsular rent were managed by lowering the parameters of phaco emulsification, low infusion bottle height and anterior vitrectomy.

Cases associated with microcornea with microphthalmia (33.33%) and having large retino choroidal coloboma were left Aphakic (33.34%),as placement of an intraocular lens was not feasible in these cases due to large size of the intraocular lens and almost nil visual prognosis due to large retino choroidal coloboma.(Figure 5-Figure 9)



Figure 5. shows capsular staining in manual small incision cataract surgery

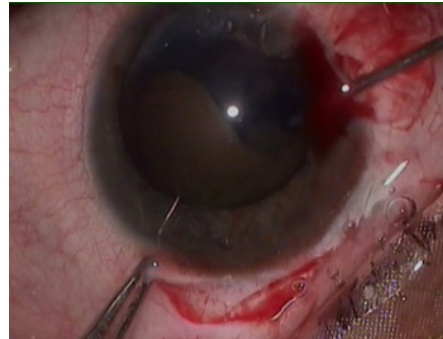


Figure 6. shows continuous curvilinear capsulorhexis in manual small incision cataract surgery.



Figure 7 shows nucleus delivery in manual small incision cataract surgery

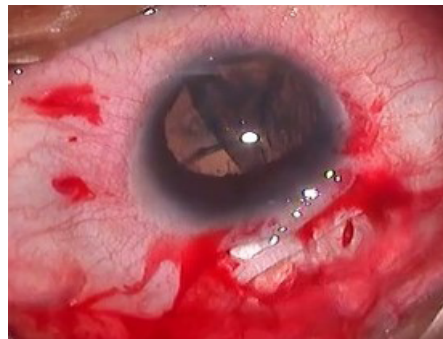


Figure 8. shows increasing zonular dialysis after nucleus extraction

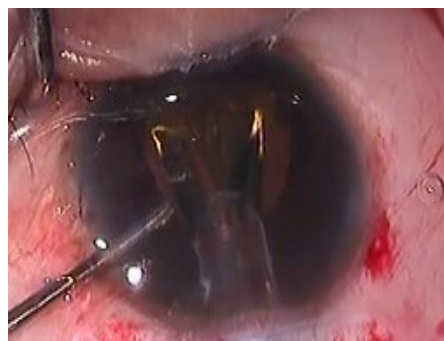


Figure 9. shows foldable intra ocular lens implantation in phacoemulsification surgery.

Post operative visual outcome

Irido fundal colobomas had a significant impact on the post operative visual status of coloboma patients undergoing cataract surgeries and it depends on the extent of coloboma. The **1 month** post operative visual acuity in these colobomatous eyes was satisfactory, where (40%) patients had vision in the

range of 6/24-6/60,(26.66%)had vision in the range of 6/6-6/18,(20%) had vision of 5/60-1/60 and remaining (13.33%) had vision of hand movement or perception, similar results were shown by Gaurav-et-al,Tanya-et-al in their retrospective series.

CONCLUSION

Phacoemulsification or SICS both surgeries were undertaken depending on various factors and both had good visual outcomes with almost similar intra operative risks and gains. Cataract in coloboma patients is the only treatable cause of visual disability hence; it is recommended that cataract surgery should be planned in the early stages of nuclear sclerosis, as postponement would only lead to a more difficult surgical situation. Appropriate and timely intervention for cataract in eyes with congenital coloboma will result in improved surgical prognosis. Microcornea, microphthalmos, macular and Chorio retinal colobomas along with intraoperative complications were the important factors leading to poor functional outcome.

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How to cite this article: Thatte S, Prakash P, Choudhary R, Batra I. Cataract Management-“A Surgical Challenge in Typical Iris Colobomas”. *Clin Res Ophthalmol* 2022;4(1):22-27.
DOI: 10.33309/2639-8788.040104