Mid-Term Outcome of Phacoemulsification in Congenital Iris Coloboma

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Abstract

***Purpose*:** We describe our experience on safety and effectiveness of phacoemulsification in cataract and congenital iris coloboma and point out some specific surgical recommendations aimed to minimize its complications.

***Methods*:** A prospective case series study was conducted on nineteen consecutive patients with cataract and congenital iris coloboma referred to the Farabi Eye Hospital in Tehran. After primary preoperative evaluations, cataract surgery was performed for each patient. All patients were followed-up for at least six months to determine surgical mid-term outcome.

***Results*:** Mean preoperative best corrected visual acuity (BCVA) in the participants was 1.99±0.70 logMAR which was improved to 0.82±0.61 logMAR postoperatively ($p<0.001$). Mean cell area was increased from 419.0±103.9 μm^2 to 656.8±281.6 μm^2 after surgery ($p=0.001$), while endothelial cell density was decreased from 2313.6±474.2 cell/mm^2 before surgery to 1361.2±448.2 cell/mm^2 after the operation ($p<0.001$). None of the patients developed corneal decompensation within the follow-up period. Regarding postprocedure complications, vitreous loss was observed in three patients, followed by penetration of dye to vitreous and remnant of the posterior capsule. None of the patients also experienced glare or photophobia and all of them were satisfied with the cosmetic result of their pupils.

***Conclusion*:** Based on our experience phacoemulsification with cosmetic repair of the coloboma can be a useful and safe procedure in patients with cataract and congenital iris coloboma.

***Keywords*:** Cataract, Surgery, Iris, Coloboma, Phacoemulsification

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Introduction

Iris coloboma is characterized as a notch, fissure, or hole in iris, which can be originated congenitally or acquired. Defective closure of embryonic fissure during the embryonic period is known as a main cause of congenital coloboma.[1-3] The overall prevalence of congenital iris coloboma has been estimated about 4.89 in 100,000 normal births (2.4 to 10 in 100.00 normal births).[4,5] The iris defect can increase the risk of cataract surgery lead to some complications such as vitreous loss and retinal detachment.[2-4] Therefore, most surgeons postpone the operations and this delay might lead to a vicious cycle causing more difficulties due to the maturity of cataract.[2]

There are a few studies reporting surgical methods and outcomes of extracapsular cataract extraction in such patients concerning the results of phacoemulsification with or without insertion of capsular tension ring (CTR) or pupiloplasty.[5-12] Different methods have been recommended to improve surgical outcome in affected patients. According to the study of Cionni et al, some patients with iris coloboma develop glare and photophobia after cataract surgery, which could be prevented with performance of pupiloplasty.[13]

Here, we described our experience on safety and effectiveness of this procedure in patients with cataract and congenital iris coloboma and finally point out some specific surgical recommendations aimed to minimize its complications.

Methods

A prospective case series study was conducted on nineteen consecutive patients with cataract and congenital iris coloboma referred to Farabi Eye Hospital in Tehran from March 2007 to December 2009. Farabi eye hospital is the oldest and largest eye referral hospital in Iran that admits the patients from all provinces of the country. Inclusion criteria were presence of iris coloboma, visually significant cataract, visual acuity (VA) more than light perception, absence of retinal detachment on funduscopy and sonography, and endothelial cell count>1000 cell/mm^2. The study protocol was approved by ethics committee of the Tehran University of Medical Sciences and the tenets of declaration of Helsinki were observed.

Best corrected visual acuity (BCVA), slit-lamp examination, funduscopy, specular microscopy, A- and B-scan echography were performed for all the patients. The severity of the cataract was graded according to Lens Opacities Classification System 3 (LOCS 3).[12] Posterior segments were evaluated by using B-scan echography. Biomtery was done and according to Hoffer Q and SRK T formula we select intraocular lens (IOL) power and the target refraction was -0.5 diopter. All operations were done by a single surgeon (S.B). The patients' pupils were dilated preoperatively by cocktail of tropicamide1%, cyclopentolate 1%, and phenylephrine 10% three times every five minutes. The procedures were performed under general anesthesia. In 17 patients (except patients No. 14, 15), after temporal clear corneal incision by microkeratome 3.2 mm (Sharpoint, IQ Geometry slit knife angled, Bevel Up), two paracenteses (Sharpoint stab Knife straight) were made. If the pupils were not dilated adequately, interacameral 1/10000 epinephrine was injected. If it was not suitable, disposable iris retractors were inserted. Ophteisbio 1.6% intraocular gel (sodium hyaluronate) was injected and continuous curvilinear capsulorrhexis (CCC) was performed by cystotome or capsulorrhexis forceps. Morcher capsular tension ring type 14 MR-1400 was inserted in a bag via main incision (except in patients No 2, 4, 14, 15, 16). Horizontal chop technique with vacuum 250 mmHg, air pressure 80 mmHg and power 30% by phaco machine (Pharo ARC laser AG Schweiz) was done.

One-piece acrylic posterior chamber intraocular lens (PC-IOL) (Alcon SN60WF) was inserted (Duckworth & Kent injector and Monarch D Cartridge) into the capsular bag except (in patients No 2, 4, 8, 14, 15). Then, the iris retractors were removed and intracameral carbachol (Optichol) was injected. Pupiloplasty with the technique described by Cionni et al[13] was done as follows: two radial incisions in the nasal and temporal colobomatous defect toward the iris root were created by intraocular scissor. Pupiloplasty was done by two peripheral corneal incisions (inferonasal and inferotemporal) with CIF-4 needle (Ethicon) 10-0 Prolene suture. The needle entered the

anterior chamber through inferotemporal grasped edge of the central iris leaflet and exited from the inferonasal incision. The suture was retrieved from the anterior chamber by a Sinskey hook from an inferior corneal incision and was tied up by 5 throws in a single knot.

All the patients received 1 mg/0.1 cc intracameral cefuroxime and 1 cc betamethasone LA subtenon at the end of the operation. Follow-up visits were performed at one day, one week, one month, and six months after the operation. BCVA, slit-lamp examination, and funduscopy were performed for all patients in all follow-up visits. Specular microscopy was done (Topcon SP-2000P; Topcon America Corp, Paramus, NJ, America) after six months and thus the patients outcome were evaluated within a follow-up time at least six months.

For statistical analysis, Snellen BCVA measurements were converted to logarithm of the minimal angle of resolution units (logMAR) to allow averaging and statistical analysis. Results were presented as mean ± standard deviation (SD) for quantitative variables and were summarized by absolute frequencies and percentages for categorical variables. Changes in study variables after the operation were assessed using paired *t* test or Wilcoxon test. Statistical significance was determined as a p value of ≥0.05. All statistical analysis was performed using SPSS software (version 16.0, SPSS Inc., Chicago, Illinois).

Results

Nineteen eyes of 19 patients (11 right and eight left eyes) were evaluated. The demographic and preoperative data are presented in table 1. Eight patients (42%) were males and 11 (58%) were females with the mean age of 51 years (ranged 23 to74 years). Sixteen patients had bilateral iris coloboma. Moreover, 17 eyes had lens coloboma and missing zonules, four had evident lens subluxation, and 2 had phacodonesis. Subsequently, in postoperative funduscopy, 10 patients had retinal coloboma and six had macular and optic nerve coloboma. According to LOCS3 classification, all participants had nuclear color 6, nuclear opalescence 6 (NC6/NP6). Mean preoperative BCVA was significantly improved from 1.99±0.70 logMAR to 0.82±0.61 logMAR, postoperatively (p<0.001). Preoperative refraction could not be measured due to the density of cataract and postoperative refractions are indicated in table 2. Mean corneal diameter was 9.78±1.65 mm, being in the category of microcornea. Four patients developed posterior capsular rupture and were left aphakic (Table 1). One of the patients presented severe microphthalmos, so prophylactic sclerotomy from 3.5 mm of the limbus with 3 mm radial incision was performed for him. Because of the posterior capsular rupture and vitreous loss in this patient, anterior vitrectomy was done and the patient remained aphakic. One patient suffered from severe phacodonesis and the fellow eye was aphakic, so intracapsular cataract extraction with anterior vitrectomy was planned and the patient remained aphakic. Trypan blue dye leaked to the vitreous cavity in a patient and intraocular inflammation was noticed at the first postoperative day, which resolved after one week. In that patient, final BCVA remained 0.09 logMAR (Table 2). Specular microscopic indices were significantly changed after the operation (p<0.001), while none of the patients developed corneal edema during 6 months follow-up period (Tables 1 and 2). Mean cell area was increased from 419.0±103.9 μm^2 to 656.8±281.6 μm^2 after surgery (p=0.001) and endothelial cell density was also significantly decreased from 2313±6 cell/mm^2 before surgery to 1361.2±448.2 cell/mm^2 after the operation (p<0.001) (Tables 3 and 4). Regarding postprocedure complications, vitreous loss appeared in three patients, followed by penetration of dye to vitreous and remnant of posterior capsule. None of the patients developed glare or photophobia and all of them were satisfied with cosmetic result of their pupils.

Table 1. Baseline characteristics of the study patients

Patient no.	Eye	Age (yr)	Sex	Diagnosis	Corneal Diameter (mm)	Fundoscopy
1	Left	45	F	IC+LC+Cat	12	WNL
2	Left	60	F	IC+LC+Cat	10	Macular optic nerve coloboma
3	Left	43	F	IC+LC+Cat	9	Inferior retinal coloboma
4	Right	61	M	IC+LC+Cat+previous SCC of conjunctive	10	Myopic change
5	Right	61	M	IC+LC+Cat	12	WNL
6	Left	52	F	IC+LC+Cat	10	Inferior retinal coloboma
7	left	46	M	IC+LC+Cat	9	WNL
8	Right	74	M	IC+LC+Cat	11	WNL
9	Right	43	M	IC+LC+Cat	9	WNL
10	Left	40	F	IC+LC+Cat	6	Tilted disc and macular coloboma
11	Right	43	F	IC+LC+Cat+ET	10	Inferior and macular coloboma
12	Left	52	M	IC+LC+Cat	10	Macular coloboma
13	Right	43	F	IC+LC+Cat	9	WNL
14	Left	23	M	IC+LC+Cat+nystagmus+microphthalmus	6	Inferior retinal coloboma
15	Left	64	M	IC+LC+Cat	11	Hypoplastic disc
16	Left	51	F	IC+LC+Cat	10	Optic disc coloboma
17	Right	51	F	IC+LC+Cat	12	WNL
18	Right	60	F	IC+LC+Cat	10	Inferior macular coloboma
19	Left	57	F	IC+LC+Cat	10	Inferior retinal coloboma

IC: Iris coloboma, LC: Lens coloboma, Cat: Cataract, WNL: Within normal limit, ET: Esotropia, SCC: Squamous cell carcinoma

Table 2. Postoperative data of the study patients

Patient no.	Procedure	Complications	Refraction
1	PE+CTR+IOL+Pup	-	-2.00, -0.75×90º
2	PE+Ant vit	VL	-4.00, -2.00×180º
3	PE+CTR+IOL+Pup	-	-4.00, -1.00×70º
4	PE+Ant vit+Pup	VL+penetration of dye to vitreous	+11.00
5	PE+CTR+IOL+Pup	-	-0.50, - 3.00×120º
6	PE+CTR+Sph	-	+0.50, -1.00×80º
7	PE+CTR+IOL+Pup	-	+1.00, -1.00×180º
8	PE+Ant vit+CTR	VL+remnant of posterior capsule	+13.00, -1.00×40º
9	PE+CTR+IOL+Pup	-	-2.75, -3.00×170º
10	PE+CTR+IOL+Pup	-	+0.50, -1.00×90º
11	PE+CTR+IOL+Pup	-	+1.50, -0.5×70º
12	PE+CTR+IOL+Pup	-	+1.75, -1.5×180º
13	PE+CTR+IOL+Pup	-	+1.50, -0.75×120º
14	PE+Ant vit+Sclerotomy	-	+3.00, -1.5×70º
15	ICCE+Ant vit	-	+5.00, -1.0×180º
16	PE+IOL	-	+1.50, -2.0×180º
17	PE+CTR+IOL+Pup	-	+2.50, -1.0×90º
18	PE+CTR+IOL+Pup	-	-1.00
19	PE+CTR+IOL+Pup	-	-1.25

PE: Phacoemulsification, CTR: Capsular tension ring, Ant Vitx: Anterior vitrectomy, IOL: Intraocular lens, Pup: Pupiloplasty, ICCE: Intracapsular cataract extraction, VL: Vitreous loss, Sph: Sphincterotomy

Table 3. Changes in best corrected visual acuity and specular microscopy parameters after operation in each patients

Patient no.	Procedure	BCVA logMAR		Min (μm^2)		Max (μm^2)		Mean (μm^2)		SD (μm^2)		CV (%)		CD ($/mm^2$)	
		Before	After	Before	After	Before	After	Before	After	Before	After	Before	After	Before	After
1	PE+CTR+IOL+Pup	2.00	0.15	265	345	506	1148	739	739	92	246	23	33	2522	1353
2	PE+Ant vit	2.69	1.69	216	232	783	941	505	505	161	146	33	28	2002	1979
3	PE+CTR+IOL+Pup	2.00	1.0	268	298	736	778	547	547	124	130	27	23	2179	1828
4	PE+Ant vit+Pup	1.69	0.09	208	431	805	1591	843	843	148	527	20	46	2100	861
5	PE+CTR+IOL+Pup	1.69	0.2	361	280	1107	1455	876	876	204	332	28	37	1401	1141
6	PE+CTR+Sph	1.39	0.3	220	388	397	431	519	519	50	83	15	15	3186	1926
7	PE+CTR+IOL+ Pup+Sph	1.30	0.3	208	470	1051	1561	1062	1062	232	304	40	28	1760	939
8	PE+Ant vit+CTR	1.30	0.5	221	272	565	1305	827	827	93	287	24	34	2500	1208
9	PE+CTR+IOL+Pup	2.00	0.3	178	298	504	751	516	516	156	113	35	21	2262	1937
10	PE+CTR+IOL+Pup	2.18	1.52	210	229	633	618	889	889	121	369	32	41	2662	1129
11	PE+CTR+IOL+Pup	1.69	1.52	239	300	649	900	550	550	119	140	29	38	2455	1110
12	PE+CTR+IOL+Pup	1.69	1.0	234	358	471	1691	980	980	61	346	17	35	2930	1020
13	PE+CTR+IOL+Pup	3.69	1.39	186	346	524	1755	272	272	102	382	25	39	2471	1028
14	PE+Ant vit+ Sclerotomy	3.69	1.0	201	276	612	458	372	372	104	47	21	12	2020	1685
15	ICCE+Ant vit	2.00	0.5	302	503	868	1100	750	750	161	200	28	38	1782	1000
16	PE+IOL	1.69	1.39	170	192	420	309	273	273	201	232	31	37	2111	1700
17	PE+CTR+IOL+Pup	1.39	0.5	201	219	475	405	301	301	65	47	20	26	1813	1002
18	PE+CTR+IOL+Pup	2.39	2.0	171	243	515	1018	451	451	80	181	22	41	2834	2215
19	PE+CTR+IOL+Pup	1.52	0.3	199	226	453	1816	1246	1246	68	319	20	25	2968	802

BCVA: Best corrected visual acuity, log MAR: Logarithm of minimal angle of resolution, Min: Minimum size, Max: Maximum size, SD: Standard deviation of size, CV: Coefficient of variation of size

Table 4. Total postoperative changes in best corrected visual acuity and specular microscopy parameters in comparison with preoperative status

Item	Before surgery (n=19)	After surgery (n=19)	p
Mean cell area (μm^2)	419.0±103.9	658.8±281.6	0.001
CV (%)	25.8±6.5	31.4±9.3	0.034
CD ($/mm^2$)	2313.6±474.2	1361.2±448.2	<0.001
BCVA (logMAR)	1.99±0.70	0.82±0.61	<0.001

BCVA: Best corrected visual acuity, logMAR: Logarithm of minimal angle of resolution
SD: Standard deviation of size, CV: Coefficient of variation of size

Discussion

Herein, we evaluated the results of cataract surgery in 19 patients with coloboma. The mean age of our studied patients was 51 years at the time of operation which is comparable to other reports.[5,12,14] This is relatively low age for cataract surgery in regards to general population. In a study by Nordlund et al[5] on 7 patients with coloboma, it was shown that although cataract surgery in eyes with congenital cataract and coloboma was associated with many pre- and postoperative challenges but it showed to be a safe procedure.

Considering the very advanced stage of cataract NC6/NP6 according to LOCS3 classification and the existence of iris coloboma some unfavorable results were expected. Moreover, similar to other reports,[5] the involvement of retina and macula, as was expected in such patients caused the final visual acuity improve but not significantly.

CTR expands the capsular fornix, reinforces the area of zonular weakness and spreads the zonular tension equally around the capsular equator that causes intact zonules to extend their strength to zonulolysis areas. CTR can be implanted at any time after

CCC, but in colobomatous patients, CTR insertion is preferred to be applied before phacoemulsifications to prevent the escape of fluid into the vitreous body and hydration of vitreous, which consecutively leads to anterior chamber shallowness. CTR prevents IOL decentration, capsular fornix aspiration and consecutive extension of zonular dialysis, and penetration of fluid behind the posterior capsule.[11,13,15]

The exposed IOL optic edge through the corectopia is associated with the location of the iris coloboma can cause photophobia, glare and monocular diplopia. Pupiloplasty made a nearly well centered pupil in our patients and prevented those complications. But it is no obligatory. Unfortunately, specular microscopy revealed significant loss of endothelial cells after cataract surgery in our patients, which might be caused by the high density of cataract, microcornea, and the low anterior chamber volume in a very dense cataract it is logical to perform extracapsular cataract extraction technique.

Some limitations of this study are relatively short follow-up time (six months) and positive points are nearly large number of cases and phacoemulsification and CTR insertion and pupiloplasty in most cases.

According to the study findings, some recommendations may decrease the incidence of complications of cataract surgery in patients with coloboma: 1) if the pupil dilation is not adequate, mechanical pupil stretching, sphincterotomy or iris retractor is recommended; 2) during CCC, extension of the capsulorrhexis edge very close to the colobomatous side of the anterior capsule should be avoided; 3) phacochop technique by an expert surgeon with minimum phaco time, power and low fluidic currents would be preferred; 4) in patients with congenital iris coloboma and cataract phacoemulsification with PC-IOL, CTR insertion and pupiloplasty is a safe and useful approach, besides the cosmetic effects, pupiloplasty can prevent the glare, and monocular diplopia; 5) IOL with a large optical diameter to occupy most of the capsular bag is recommended; 6) Since patients have higher rates of retinal detachment, regular posterior segment examination after operation is suggested; 7) phacoemulsification of colobomatous eyes must be done by expert surgeons.

Conclusion

Our results confirmed that phacoemulsification is a safe method to remove cataract in eyes with congenital coloboma.

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References

1. Jesberg DO, Schepens CL. Retinal detachment associated with coloboma of the choroid. Arch Ophthalmol 1961;65:163-73.
2. Vaughn LW, Schepens CL. Progressive lenticular astigmatism associated with nuclear sclerosis and coloboma of the iris, lens, and choroid: case report. Ann Ophthalmol 1981;13(1):25-7.
3. Elder MJ. Aetiology of severe visual impairment and blindness in microphthalmos. Br J Ophthalmol 1994;78(5):332-4.
4. Onwochei BC, Simon JW, Bateman JB, Couture KC, Mir E. Ocular colobomata. Surv Ophthalmol 2000;45(3):175-94.
5. Nordlund ML, Sugar A, Moroi SE. Phacoemulsification and intraocular lens placement in eyes with cataract and congenital coloboma: visual acuity and complications. J Cataract Refract Surg 2000;26(7):1035-40.
6. Völcker HE, Tetz MR, Daus W. Cataract surgery in eyes with colobomas. Dev Ophthalmol 1991;22:94-100.
7. Nixeaman DH. Cataract extraction in a case of congenital coloboma of the iris. Br J Ophthalmol 1968;52(8):625-7.

8. Jaffe NS, Clayman HM. Cataract extraction in eyes with congenital coloboma. J Cataract Refract Surg 1987;13(1):54-8.
9. Watt RH. Inferior congenital iris coloboma and IOL implantation. J Cataract Refract Surg 1993;19(5):669-71.
10. Rapaullha P, Sudhir R, Mathew M, et al. Cataract surgery in eyes with iris coloboma; risk factors, complications and results. All India Ophthalmological Society Annual Conference. Available at: http://www.aios.org/proceedings.cfm?pid=146&show=y. Assessed August 15, 2006.
11. Mizuno H, Yamada J, Nishiura M, Takahashi H, Hino Y, Miyatani H. Capsular tension ring use in a patient with congenital coloboma of the lens. J Cataract Refract Surg 2004;30(2):503-6.
12. Chylack LT Jr, Wolfe JK, Singer DM, Leske MC, Bullimore MA, Bailey IL, et al. The Lens Opacities Classification System III. The Longitudinal Study of Cataract Study Group. Arch Ophthalmol 1993;111(6):831-6.
13. Cionni RJ, Karatza EC, Osher RH, Shah M. Surgical technique for congenital iris coloboma repair. J Cataract Refract Surg 2006;32(11):1913-6.
14. Klein BE, Klein R, Linton KL. Prevalence of age-related lens opacities in a population. The Beaver Dam Eye Study. Ophthalmol 1992;99(4):546-52.
15. Kim JH, Kang MH, Kang SM, Song BJ. A modified iris repair technique and capsular tension ring insertion in a patient with coloboma with cataracts. Korean J Ophthalmol 2006;20(4):246-9.