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Original article

Outcomes of surgery for retinal detachment in eyes having choroidal coloboma

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ABSTRACT

Purpose: To study the anatomical and visual outcomes of surgery for retinal detachment (RD) in patients with choroidal coloboma.

Methods: Forty-six eyes of 46 patients underwent surgery for RD with choroidal coloboma. Patients were divided into three groups, according to the extent of choroidal coloboma: Coloboma not involving the disc and macula (Group A), coloboma involving the disc (Group B), and coloboma involving the disc and macula (Group C). Thirty-eight eyes underwent pars plana vitrectomy with silicone oil (SO) (Group 1) and eight eyes underwent scleral buckling surgery (Group 2). SO removal was carried out in 18 eyes at final follow up.

Results: After a mean follow-up of 16.2 months, 40 eyes (86.9%) had an attached retina and the best corrected visual acuity (VA) improved from a preoperative median of 20/800 to 20/250 ($p = 0.001$). Thirty-four eyes (74%) recovered ≥20/400 VA. The final median VA in Group A (20/200) was better than in Group B (20/800) and Group C (light perception and hand movement) which was statistically significant ($p = 0.01$). Redetachment occurred in 10 eyes (21.8%), of which 7 eyes (18.91%) belonged to Group 1 and 3 eyes (37.5%) belonged to Group 2, and all of these eyes underwent resurgery. Of these 10 eyes, 9 had an attached retina at final follow up.

Conclusion: Postoperative visual recovery in RD with choroidal coloboma is dependent upon the involvement of the disc and macula with the colobomatous area. Resurgery following redetachment improves the final anatomical outcomes.

1. Introduction

A coloboma of the choroid is characterized by the congenital absence of a portion of the normal retina, retinal pigment epithelium and choroid.[1,2]

It is recognized clinically as a prominent white zone of the fundus, which is usually located inferiorly and slightly to the nasal side. This anomaly is due to a defect in the normal fusion of the fetal fissure of the optic cup. It may occur as an isolated finding, but is frequently associated with other anomalies such as iris coloboma, microphthalmos, microcornea, cataract, lens coloboma, and optic pit. It may also be associated with various systemic birth defects.[3,4]

Choroidal coloboma is an infrequent finding and is found in only 0.14% of a large series of eye patients.[1] The prevalence of the retinal detachment (RD) in eyes with choroidal coloboma has been reported to be from 8.1% to 42%.[2–6]

If RD is secondary to retinal breaks outside the colobomatous area, it can be treated by scleral buckling surgery, however, when the coloboma is etiologically responsible for RD, management options are different.[7,8] Anatomical alterations in colobomatous eyes, such as retinal thinning, staphylomatous and thinned sclera, and involvement of the disc and macula, as well as lack of choroid and retinal pigment epithelium, make the operation technically difficult and hence the prognosis less satisfactory.

In this study, we report our data on anatomical and visual outcomes of choroidal coloboma with RD.

2. Materials and methods

Forty six eyes of 46 consecutive patients who had choroidal coloboma with RD were included in this study. Patients were divided in three groups according to the extent of choroidal coloboma: Coloboma not involving the disc and macula (Group A) (Fig. 1A and B), coloboma involving the disc (Group B) (Fig. 2), and coloboma involving the disc and macula (Group C) (Fig. 3). Thirty-eight eyes underwent pars plana vitrectomy with silicon oil (Group 1) and eight eyes underwent scleral buckling surgery

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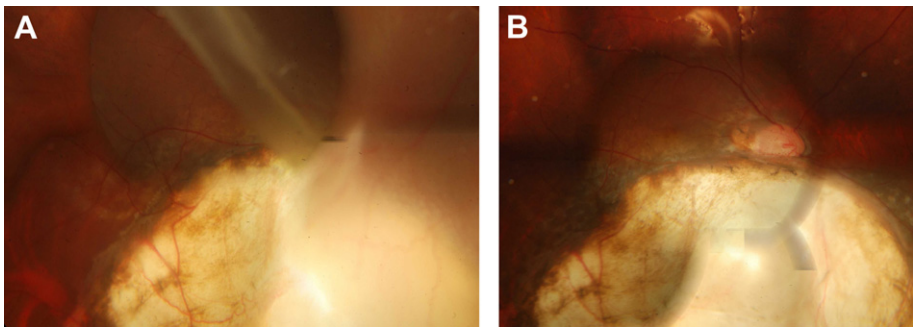

Fig. 1. (A and B) Pre and Post operative photo of Group A case (Coloboma not involving disc and macula).

(Group 2). Patients with peripheral retinal breaks were treated with conventional scleral buckling surgery. Those eyes with posterior holes and rhegmatogenous RD with no holes were considered candidates for vitrectomy procedures.

Patient demographics, pre- and postoperative examinations, type of surgery, best corrected Snellen VA (BCVA) and postoperative complications were recorded. Ophthalmic examinations included slit lamp and indirect ophthalmoscopic examinations. Intraocular pressure (IOP) was measured in all eyes. Anatomical success was defined as retinal reattachment posterior to the band or buckle. Fundus drawings and photographs were taken in each follow-up.

Statistical analysis was performed using the Wilcoxon Signed Ranks Test. Qualitative variables were described using percentages; quantitative data were defined using means and standard deviations.

Surgical procedures: The surgical management involved pars plana vitrectomy and scleral buckling surgery after informed consent. Thirty-eight eyes (82.60%) underwent pars plana vitrectomy. Vitrectomies were performed in a standard fashion employing a three-port 20G pars plana approach. Scleral buckling with an encircling band (No. 240) was done in 28 cases with proliferative vitreo retinopathy (PVR) grade C or more. The lens was removed in 10 eyes (26.31%) due to existing cataracts. A meticulous search for the retinal breaks in the periphery was done with the use of a wide angle viewing system (Volk HRX VIT SSV, VOLK(r) Mentor, Ohio, USA). Then, simultaneous air-fluid exchange and internal drainage was performed through the retinal break at the margin of, or within, the choroidal coloboma. If this maneuver did not result in total retinal attachment, endodiathermy was performed to create another retinotomy, through which subretinal fluid was drained to totally reattach the retina, which was performed in eight eyes. All eyes underwent intraoperative laser photocoagulation therapy (two rows of diode endolaser) around the colobomatous area and retinal breaks followed by air/fluid exchange. The areas of the macula, disc and papillomacular bundles were spared from laser. Cryotherapy was used for the anterior margins of coloboma, whenever laser manifestation was difficult to assess in the periphery. Internal tamponade was performed with silicone oil (SO) (1000 centistokes) in all of the eyes.

Retinal reattachment was achieved in all eyes at the end of the procedures and in the early postoperative period of 6 weeks (Figs. 4 and 5). Patients were placed in the prone position for 1 week postoperatively.

Eight eyes (17.39%) underwent primary scleral buckling surgery, with a circumferential silicone sponge (No. 506, LABTICIAN Ophthalmics, Inc. Canada). Cryopexy was done to suspicious areas and breaks. Subretinal fluid was drained through a tiny hole in the sclera at the maximum height of the detached retina. In four eyes, intravitreal air was injected, due to hypotony after subretinal fluid drain. Three of these eight eyes experienced retinal redetachment secondary to PVR in two eyes and new breaks outside the coloboma in one eye; they underwent pars plana vitrectomy with SO tamponade. Three eyes underwent sponge removal due to infection and extrusion of the sponge. A microphthalmic eye with a small orbit is possibly predisposed to the higher rate of sponge extrusion.

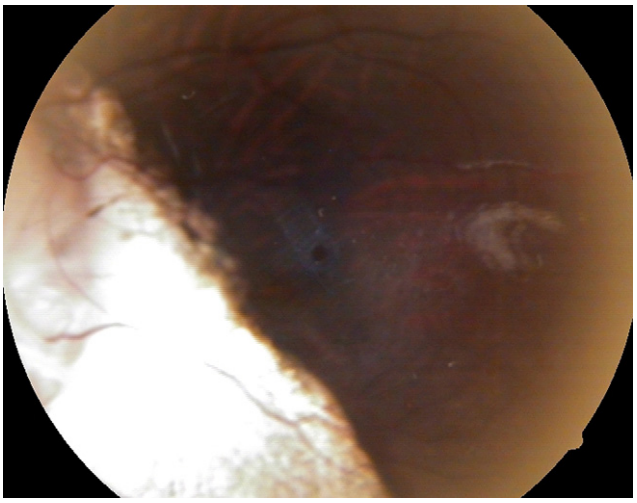

Fig. 2. Postoperative photo of Group B case (Coloboma involving Disc but not macula).

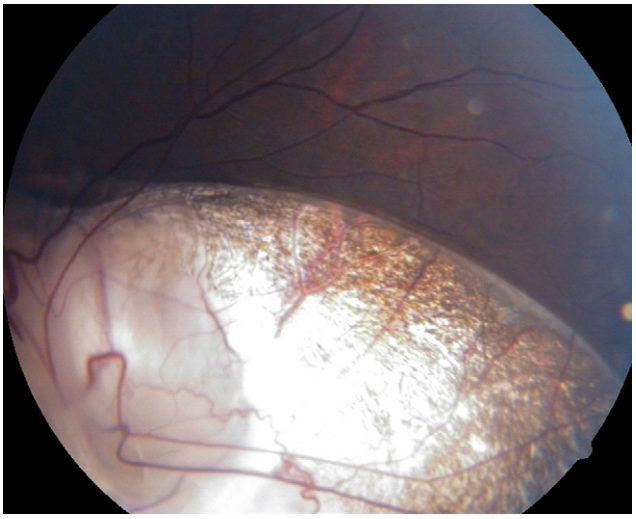

Fig. 3. Postoperative photo of Group C case (Coloboma involving Disc and Macula).

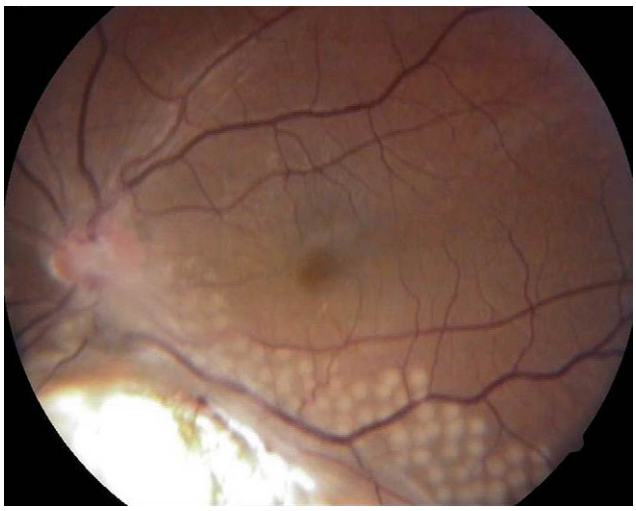

Fig. 4. Postoperative photo of Group A case.

Silicone oil removal: SO was removed in 18/41 eyes (43.9%). The lens was removed simultaneously in 6 eyes (37.5%), due to the development of cataracts. The interval between SO injection and removal ranged from 3 months to 24 months (mean = 6.4 months). Of the 18 eyes which underwent silicone removal, 6 had redetachment of the retina, which underwent revision surgery.

3. Results

The demographic details, as well as the patients' characteristics, are depicted in Table 1.

Nystagmus was present in 11 (23.9%) eyes, iris coloboma in 32 (69.5%) eyes, microphthalmos in 7 (15.21%) eyes, microcornea in 9 (19.5%) eyes, and lenticular opacity in 10 (21.7%) eyes. Esotropia was present in 6 (13%) eyes and 4 (8.69%) eyes had exotropia.

Overall, the mean number of procedures (including SO removal) was 1.58 for each eye; 27 eyes (58.69%) underwent surgery once, 14 eyes (30.43%) were operated on twice, three eyes (6.52%) required surgery three times, and one eye underwent surgery four times.

Reattachment was achieved in all eyes at the end of the procedures and in the early postoperative period of 6 weeks.

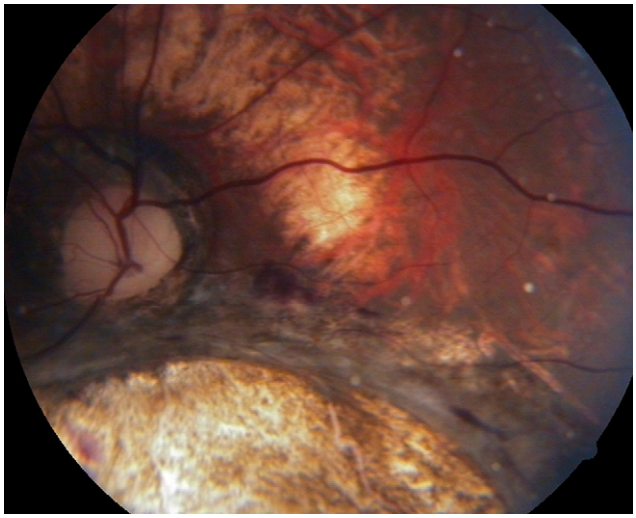

Fig. 5. Postoperative photo of Group A case.

Table 1
Patient characteristics ($n = 46$).

	No.	%
Age (y)		
Mean	21.9	
Range	7–40	
Sex		
Male	31	67.4
Female	15	32.6
Colobomatous eye		
RE	6	13
LE	6	13
Both	34	74
Colobomatous eye with RD		
RE	22	47.8
LE	18	39.2
Both	6	13

LE = left eye; RE = right eye.

Redetachment occurred in 10 eyes (21.8%), of which seven eyes (18.91%) belonged to Group 1 and three eyes (37.5%) belonged to Group 2. The recurrence was caused by PVR changes in six cases and new breaks outside the coloboma in four cases. Revision surgery involved membrane peeling and injection of silicone if the first surgery was with scleral buckling and removal of membranes under SO in eyes with SO. Retreatment of the coloboma border was done with laser during reoperation. After a mean follow-up of 16.2 months, the retina remained attached in nine eyes (90%) in which revision surgery was done. The cause of redetachment in the remaining one eye was due to a new break formation. Of the 18 eyes which underwent silicone removal, six had redetachment of the retina and eventually underwent revision surgery. Of these six eyes, four had PVR changes, while in the remaining two eyes, no obvious break was noted in the non colobomatous retina. The total follow-up was at least 6 months to 28 months (mean = 16.2 months) among the eyes that underwent SO removal, and the mean post SO removal follow-up was 12 months (range = 4 months to 18 months). The interval between SO injection and removal ranged from 3 months to 24 months (mean = 6.4 months).

The distribution of Snellen VA based on the site and extent of the coloboma is presented in Table 2. The final median VA in Group A (20/200) was better than in Group B (20/800) and Group C (light perception and hand movement), which was statistically significant ($p = 0.01$).

After a mean follow-up of 16.2 months, the best corrected VA improved from a preoperative median of 20/800 to 20/250 ($p = 0.001$). Thirty-four eyes (74%) recovered ≥20/400 VA (Table 3). The retina remained attached in 40 eyes (86.9%). Out of six eyes with a detached retina, one was operated on previously for redetachment. The recurrent retinal detachment was caused by severe PVR changes in five eyes and new breaks outside the coloboma in one eye.

Table2
Final Snellen visual acuity in 46 cases following colobomatous retinal detachment repair based on the extent of the coloboma.

	Without disc or macular coloboma (Group A)	Only disc coloboma (Group B)	Both disc and macular coloboma (Group C)
n	31	9	6
Median	20/200	20/800	HM

HM = hand movement.

Table 3
Final visual acuity ($n = 46$).

Best corrected visual acuity	No. of patients	%
20/20–20/40	1	2.1
20/60–20/120	6	13.0
20/200–10/200	27	58.9
20/800–counting finger	5	10.9
Hand movement–perception of light	7	15.1
	46	100

The postoperative complications included raised IOP in five (10.8%), hazy media in two (4.3%) and epithelial defect in two (4.3%) cases, and were managed with medical treatment, without necessitating early SO removal.

4. Discussion

The mean follow-up in this study (16.2 months) was longer than that of some other large studies (13.4 months[9] and 14months[10]) reporting outcomes of retinal detachment surgery for colobomatous RD. The rate of final retinal reattachment in our study was 86.9%, achieved with a mean number of 1.58 operations per eye, which compares favorably with other reports at 81.2%,[9] 81.8%,[8] 87.5%,[10] and 100%.[12–14] These studies, however, included only vitrectomized colobomatous eyes, while excluding eyes undergoing scleral buckling (SB). Only one study included vitrectomy and sclera buckling procedures with 40 months of follow up.[15] The present study demonstrated a good anatomical success rate and significant visual improvement. VA $\geq$ 20/400 was achieved in 74% of operated eyes in the current series, however, this level of vision was reported in 69.9%,[9] 78.4%[10] and 35.7%[15] cases in three large studies reporting on 85, 42 and 28 cases, respectively. Colobomatous involvement of the disc and/or macula is a crucial prognostic factor in terms of functional outcomes.[15] In the current study, the final median VA was significantly better in eyes without disc and macular involvement.[15] The final median VA in our study was counting fingers at 3 meters (10/200), which was comparable to that reported by Ramezani et al[15] (cf 2 meter). Although SB can be considered as a treatment modality in eyes with colobomatous RD,[10] reported anatomical outcomes in the literature (35% to 57%) have not been satisfactory. This is due to the difficulty in locating the retinal breaks, their posterior location, and an inability to create adequate chorioretinal adhesions around the coloboma by cryo or laser. Scleral buckling has been performed using an encircling element, or with two radial buckles around the colobomatous area, as described by Wang and Hilton,[16] with a success rate of only 35%. Jesberg and Schepens[17] believed that colobomatous eyes are not suitable for SB surgery. Most reports on SB surgery in colobomatous eyes date back to decades ago, e.g., 1961,[17] 1981,[18] and 1985.[16] There has been a decline in the trend toward SB operations in colobomatous eyes; however, in the case of a definite peripheral break, SB may still be considered as the first choice.[10] Encircling SB was the preferred method in only eight out of 46 eyes (17.4%) in our study.

The preferred surgical procedure in our study was vitrectomy in 82.6% of eyes. The possibility of finding breaks is higher with vitrectomy. Vitrectomy with SO tamponade was successfully employed for colobomatous RD for the first time in 1983.[19] Since then, it has frequently been used by different experts.[8,11–13] Twenty-eight eyes (73.68%) in our study received an encircling band at the time of vitrectomy. In the study by Gopal et al,[8] 82.4% of vitrectomies included an encircling element. Without referring to any statistics, the authors concluded that placing an encircling band had no beneficial effect. In the report by Pal et al,[10] only 38% of eyes received an encircling element.

Revision surgery was successfully employed for redetachment in 10 eyes after primary procedures. It involved membrane peeling and injection of silicone if the first surgery was with sclera buckling and removal of membranes under SO in eyes with SO. After a mean follow-up of 16.2 months, the retina remained attached in nine eyes (90%) in which revision surgery was done. The cause of redetachment in the remaining one eye was due to new break formation.

SO has been preferred for tamponade in colobomatous RD repair by most surgeons.[8,9,11–13] In the study by Gopal et al,[9] it was used in 80/85 eyes (94%); three of the five eyes which received gas at the end of the operation, experienced a retinal redetachment. The authors concluded that gas is not a suitable tamponade, because these eyes require a long effect over the entire edge of the coloboma. However, it should be noted that neither study had enough data to compare the use of SO versus gas during vitrectomy for colobomatous RD.

SO removal was performed in nearly half (43.9%) of our patients, which is comparable to other studies.[9,10,15] Retinal redetachment occurred in 6/18 (33.3%) eyes undergoing SO removal. In other reports, corresponding rates have been 27.3%,[15] 10%[10] and 15.6%.[9]

Increased IOP was another common complication in our patients; it occurred in five (10.8%) of vitrectomized eyes and was controlled with medication. This difference may be due to different timing for SO removal, because SO emulsification has been considered as an important risk factor for increased IOP. The shortcoming of this study is its retrospective nature. In addition, we were unable to compare vitrectomy with sclera buckling, and SO versus gas in the management of colobomatous RD. This retrospective study adds more data to the literature regarding the preferred surgical options, as well as visual and anatomical outcomes of RD repair in eyes with chorioretinal coloboma. Based on this report and similar studies, vitrectomy with SO tamponade may be a good choice for the management of colobomatous RD. Despite an acceptable anatomical result, the functional outcome is rather poor as compared to the anatomical outcome, which reflects involvement of vital structures in the eye as well as amblyopia.

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