

THE OUTCOME OF SURGERIES PERFORMED FOR RETINAL DETACHMENT ASSOCIATED WITH CHORIORETINAL COLOBOMA

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ABSTRACT

OBJECTIVES

To study the outcome of surgeries performed for retinal detachments associated with chorioretinal colobomas.

METHODOLOGY

This retrospective study enrolled 16 eyes of 16 patients with retinal detachments associated with chorioretinal colobomas. Standard 3-ports 23-gauge Pars plana vitrectomy (PPV) with silicon oil tamponade and endo-laser was performed along the rim of colobomas in all cases. In 4 (25.0%) eyes, buckling was performed with 240 silicon bands and vitrectomy to support the vitreous base. In 12 eyes (75.0%), the natural lens was removed with phaco / lensectomy and intra-ocular lens implantation.

RESULTS

Out of 16 eyes, the retina was re-attached in 15 eyes (93.75%) after the first operation. One eye needed a second surgery, which was successful anatomically after the second surgery. Mean pre-operative best-corrected visual acuity (BCVA) was 3.36 (± 0.52), while 2-weeks post-operative BCVA was 1.92 (± 0.88), which was statistically significant ($P < .001$) improvement. Increased intra-ocular pressure (IOP), silicone oil in the anterior chamber (AC), recurrent retinal detachment, and epiretinal membrane (ERM) formation were some of the complications observed post-operatively.

CONCLUSION

PPV with endo-laser photocoagulation along the rim of colobomas and silicon oil tamponade is an effective surgical procedure with a statistically significant retinal reattachment ratio and best corrected visual acuity improvement in cases of rhegmatogenous retinal detachment (RRD) associated with chorioretinal colobomas.

KEYWORDS: Rhegmatogenous Retinal Detachment, Chorioretinal Coloboma, Pars Plana Vitrectomy

INTRODUCTION

Chorio-retinal coloboma is a rare congenital anomaly of the posterior segment of the eye. It occurs in one per 10,000 live births. Chorio-retinal colobomas account for about 0.1% of the total general population.^{1,2} It occurs as a consequence of improper closure of the embryonic fissure. The Chorio-retinal coloboma can be sporadic. Also, it can be transmitted as an autosomal dominant, autosomal recessive, or X-linked recessive.^{3,4} Chorio-retinal coloboma can be unilateral or bilateral and is typically located in the inferotemporal area but may extend to involve the macula. It may be associated with the coloboma of other ocular structures along the embryonic fissure, such as the optic nerve, iris, or lid. Coloboma is associated with a higher incidence of retinal detachment. Almost half of the patients develop retinal detachment at some stage. This detachment is always rhegmatogenous and is caused by breaks outside the coloboma (Type-1) or within the coloboma (Type-2). Type-1 detachments are rare and can be treated by

scleral buckling alone. Type-2 detachments need vitrectomy with internal tamponade and are treatment-wise challenging as retinopexy will not be effective because of the absence of choroid and Retinal Pigment Epithelium (RPE) within the coloboma.^{5,6} It is recommended to perform a two or three-row prophylactic laser along the rim of the coloboma to create adhesions along the margins to reduce the chances of future retinal detachment. The chances of retinal detachment are ten to twelvefold higher in eyes that did not get prophylactic barricade laser compared to those that received laser. The laser will cause loss of the nerve fibre layer and, thus, a permanent visual loss if performed in the macular area or the area between the disc and fovea, even though it might prevent future retinal detachment.

METHODOLOGY

In this study, All the surgeries were performed at Hayatabad Medical Complex and Retina Eye Centre

between January 2015 and July 2023. 16 patients (8 women and 8 men) were selected, fulfilling the inclusion criteria, and were operated for coloboma-related retinal detachments. All the medical records were reviewed. A proforma was generated, and the recorded variables included personal details, pre-operative data, steps of the procedure, and post-operative details, including details of follow-up visits. Inclusion criteria included eyes with rhegmatogenous retinal detachments caused by breaks lying outside the coloboma. Both LogMAR and Snellen's visual acuities (VA) were recorded. Both pre and post-operative fundus examinations were performed using a slit-lamp with a 78D and 90D Volk condensing lens. The standard Pars Plana Vitrectomy (PPV) procedure with three ports and 23 gauge was performed in all cases. In 12 (75%) eyes, the lens was removed by phaco or lensectomy because some lens opacities hindered the excellent view during surgery. In 4 (25%), encircling of silicone band (No. 240) with silicone sleeve (No.270) was performed while using a non-absorbable 5/0 Ethibond suture (Ethicon). The buckle was performed to deal with the vitreo-reinal traction at the vitreous base. Endo-laser retinopexy was performed in all patients along the rim of the coloboma and at 360 degrees anterior to the equator. Areas near the disc and papilla-macular bundle were avoided while doing the endo-laser. Perfluorocarbon Liquid (PFCL) was used to achieve per operative flat retina and identify residual traction. After the fluid air exchange (FAX), 1000 CS silicone oil was used as intraocular tamponade in all patients. Retinal reattachment was achieved in 15 (93.75%) eyes after the primary procedure. In some cases, where needed, an indirect ophthalmoscope was also used for more details, especially in the retinal periphery. In all cases, a pre-operative retinal drawing was drawn, and details of the retinal detachments were highlighted.

RESULTS

The study was conducted on a total of 16 eyes of sixteen patients. Gender-wise, there were 8 women and 8 men, which indicated no predilection for any gender. All the patients were followed for a mean duration of 12 months with a minimum follow-up of 6 months and a maximum of 24 months. The mean age of all the patients was 30 years (ranging from 15 to 60 years). Pre-op LogMAR VA ranged from the perception of light (PL+ve) to 1 (6/60), with a mean of 1.40 (3/60). Intra-ocular pressure (IOP) pre-operatively, measured with Goldmann's tonometer, ranged from 12 to 18 mmHg with a mean of 15 mmHg. Six (37.5%) patients had bilateral choroidal colobomas, while the remaining 10 (62.5%) patients had unilateral disease. In 8 (50%)

patients, Iris coloboma accompanied chorioretinal coloboma. Micro-cornea was associated in 4 (25%) eyes, and 4 (25%) patients were associated with nystagmus. Associated grade-C PVR was present in 10 (62.5%) eyes, while the remaining 6 (37.5%) patients had a grade-B proliferative vitreo-retinopathy. None of the patients had undergone any previous retinal surgery. Pre-operative mean visual acuity was 1.4 (± 0.40), which improved to 0.5 (± 0.70) ($P < 0.001$). IOP was noted to be increased from a pre-operative mean of 15 (± 3) to 18 (± 2) mmHg, though this change was not statistically significant ($P = 0.075$). In one eye, inferior re-detachment was seen, managed with silicone oil removal, re-do surgery with the internal limiting membrane (ILM) peeling, endo laser enhancement, and re-oil injection. The retina was found flat after this second surgery attempt. Retina reattachment was found in 15 (93.75%) eyes after one operation. Mean visual acuity was found to have improved from light perception to 0.5, with a mean of 1.4 in the Log MAR system.

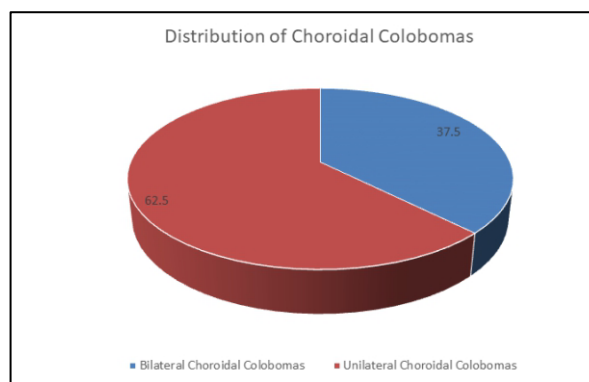


Figure 1: Distribution of Choroidal Colobomas

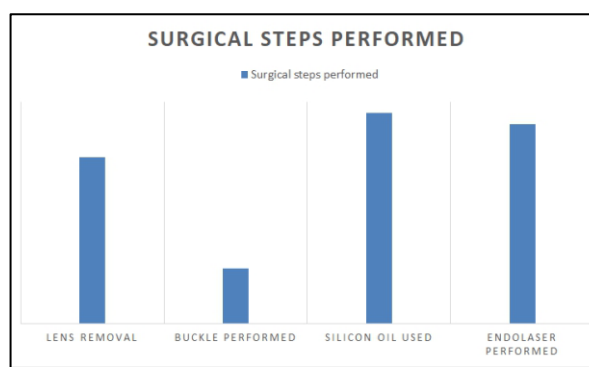


Figure 2: The Surgical Steps Performed with the Patients

DISCUSSION

Retinal detachment associated with chorio-retinal coloboma is amongst the rare fundus pathologies. Previously, such types of retinal detachments were

treated with the external approach by scleral buckling techniques, but the overall success rate was not satisfactory because of the relatively greater difficulty in finding and closing the causative breaks with this approach.^{1,2} Schepens et al. tried these coloboma cases by draining the sub-retinal fluid (SRF), followed by cryopexy, photocoagulation, or both, but still, the overall success rate was not great.^{3,4} A radial sponge was also tried with somehow better results but inducing very high astigmatism.⁵ Wang advocated pars plana vitrectomy with gas tamponade and additional band buckle to achieve up to 43 % success rate in cases of retinal detachments associated with chorioretinal colobomas.⁶ Gonver et al. achieved a high success rate while using silicone oil in these cases of coloboma RDs after undergoing a vitrectomy.⁷ Some researchers, in limited cases, performed barricade laser to wall off the retinal detachment borders, but doing laser in the papillo-macular bundle area was a risk and challenge.^{8,9,10} The success of surgical treatment in colobomatous eyes with RDs also depends on where the causative breaks are located. In fact, in coloboma, the retina splits into two layers, usually at the level of the inner nuclear layer, and mostly detachment occurs when there are breaks in the intercalary membrane. These detachments usually have already extended beyond the margins of the coloboma when the symptoms develop, and the patient approaches the ophthalmologist.^{11,12} In this study, we excluded colobomatous eyes with retinal detachments in which the primary or causative break was situated outside the boundaries of the coloboma. We achieved primary anatomical success (complete retina reattachment at the final follow-up visit) in 15 patients (93.5%). This success rate is well in the range of other studies performed in the same cases with the same surgical techniques at other centres worldwide.^{13,14,15,16,17,18} Different researchers have concluded that recurrent retinal detachment rates are 25- 40 % after initially successful reattachment in surgeries performed for coloboma-related retinal detachments.¹³ In our cases, only one eye had a re-detachment. Mean intraocular pressure (IOP) was elevated in our cases, although not statistically significant, indicating the significance of regular monitoring and timely treatment, if needed, at follow-up visits. In 4 (25%) of our cases, we augmented the encirclement band in addition to pars plana vitrectomy. We could not confirm its additional beneficial role in the anatomical success of the surgery, final best-corrected visual acuity, and Intra-ocular pressure rise. To our knowledge, the role of augmented buckling in elevating post-op IOP in eyes operated for coloboma-associated retinal detachments hasn't been addressed in previous studies. Also, studies have shown no altered anatomical or physiological outcome in

patients operated for coloboma-related retinal detachments in which additional lensectomy or phaco was performed. On the other hand, removing the natural lens facilitates meticulous vitreous base shaving and increased visibility of anterior PVR membranes. In our cases, gas tamponade was not used, so no statistical comparison between the gas and silicon tamponade groups can be made.

LIMITATIONS

The study's limitations were its retrospective nature, small number of patients as this disease is rare, and relatively short follow-up duration.

CONCLUSIONS

Retinal detachments associated with chorio-retinal colobomas are management-wise challenging cases. Pars plana vitrectomy with internal tamponade and endo-laser retinopexy along the rim of coloboma is a considerably effective procedure in anatomically re-attaching the retina and ultimately improving the final best-corrected visual acuity. Some patients may encounter intra-ocular pressure rise, usually transient and manageable with topical pressure-reducing medications.

CONFLICT OF INTEREST: None

FUNDING SOURCES: None

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CONTRIBUTORS

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2. **Farooq Khan** - Data Acquisition; Data Analysis/Interpretation; Drafting Manuscript
3. **Irfan Aslam** - Critical Revision



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