

## RESEARCH ARTICLES

# MANAGEMENT OF RETINAL DETACHMENTS IN EYES WITH CHOROIDAL COLOBOMA

## KOROİDAL KOLOBOMLU GÖZLERDE GELİŞEN RETİNA DEKOLMANLARININ TEDAVİSİ

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### ABSTRACT

**Purpose:** To present the results of surgical management of retinal detachment in eyes with choroidal coloboma. **Methods:** 12 eyes of 10 consecutive patients with retinal detachment and choroidal coloboma were involved in the study. Retinal detachments associated with the colobomatous area were treated with pars plana vitrectomy, endolaser and silicon oil tamponade, while those not associated with coloboma were treated with scleral buckling, cryocoagulation of the retinal breaks followed by drainage of the subretinal fluid. **Results:** The mean follow-up time was 17 months. Retinal detachment was related to coloboma in all but 4 eyes. The retina was successfully reattached in all cases. Recurrence of detachment limited to the area of choroidal coloboma was detected in 5 eyes (41.6%). **Conclusion:** The management plan in these anomalous eyes is of special concern and must be individualized for each patient according to the character of retinal detachment associated with coloboma or not.

**Key Words:** Retinal Detachment, Choroidal Coloboma, Surgical Treatment.

### INTRODUCTION

Choroidal coloboma is a congenital defect caused by incomplete closure of the embryonic fissure of the eye during the sixth and seventh weeks of fetal development. It is characterized by the absence of normal retina, retina pigment epithelium and choroid. This anomaly is frequently located in the inferonasal region and is often associated with several other ocular abnormalities, including iris coloboma, loss of inferior lens zonules, inferior lens opacities,

### ÖZET

**Amaç:** Koroidal kolobom ile birlikte retina dekolmanı olan gözlerde cerrahi tedavi sonuçlarını sunmak. **Gereç ve Yöntem:** Koroidal kolobom ile birlikte retina dekolmanı olan 10 hastanın 12 gözü çalışmaya dahil edildi. Retina dekolmanı kolobomatöz alanla birlikte olan gözlere pars plana vitrektomi, endolaser ve silikon yağ tamponadı uygulanırken, dekolman alan kolobomatöz alandan ayrı olan gözlerde skleral çökertme, kriyo ve subretinal sıvı drenajı uygulandı. **Bulgular:** Olgular ortalama 17 ay takip edildiler. Gözlerin 4'ü hariç tümünde retina dekolmanı kolobomla ilişkili idi. Tüm vakalarda retina yapıştı. Beş gözde (%41.6) sadece kolobomatöz alana sınırlı bir retina dekolmanı mevcuttu. **Sonuç:** Koroidal kolobomlu gözlerde tedavi planı bazı özellikler taşımaktadır ve dekolmanın kolobomatöz alanla ilişkisine göre her hastada kişiye göre ayrı tedavi planı yapılmalıdır.

**Anahtar Kelimeler:** Retina Dekolmanı, Koroidal Kolobom, Cerrahi Tedavi.

microcornea, microphthalmos and optic disc pathologies. Chorioretinal colobomas can occur in isolation or in association with systemic abnormalities like CHARGE syndrome (Coloboma of the uvea, congenital Heart disease, Atresia of the choanae, Retardation of growth and mental development, Genital anomalies, Ear malformations), cleft lip, cleft palate and growth retardation.

The prevalence of retinal detachment in eyes with choroidal colobomas has been reported

to be 8-40% (1-2). Schubert has described the histopathology of the choroidal coloboma and has shown that the retina splits into two layers near the margin of the coloboma; the inner neuroblastic layer shows central continuation as the intercalary membrane to the coloboma while the outer neuroblastic layer turns back, become disorganized, and fuse with the retinal pigmentary epithelium. The choroid is terminated as a distinct pigmented layer peripheral to this point of reversal. The schisis-like configuration at this reversal point has been termed "locus minoris resistentiae". The intercalary membrane progressively becomes thinner as it is traced centrally. The locus minoris resistentiae and the intercalary membrane are reported to be the most common areas for the breaks to occur (3).

There are different treatment strategies for retinal detachments in eyes with choroidal coloboma. Eyes with retinal breaks and retinal detachment outside the area of the coloboma are successfully managed with conventional scleral buckling techniques. The treatment of retinal detachment associated with choroidal coloboma with external scleral buckling, on the other hand, is reported to be only rarely successful because of the difficulty in identifying the breaks in the colobomatous area, the impossibility of creating chorioretinal adhesion around the breaks due to the absence of the choroid and the retinal pigmentary epithelium, and the posterior location of the breaks (1,4,5). Pars plana vitrectomy has opened up a new approach to the management of retinal detachment associated with choroidal coloboma by enabling the identification of the breaks in the intercalary membrane, allowing direct closure of the breaks with cyanoacrylate glue (6,7), peroperative isolation of the coloboma with endolaser, and use of intraocular tamponades.

We present herein the results of the surgical management of retinal detachment in 12 eyes with choroidal coloboma.

#### MATERIALS AND METHODS

We reviewed the medical records of 12 eyes of 10 consecutive patients with retinal detachment and choroidal coloboma who had been surgically managed between 1995 and 2002 (Table 1). There were 3 females and 7 males and their ages ranging from 6 to 55 years. Anterior segment examination revealed iris and lens

coloboma together with some inferior lens opacities in all eyes. Two patients had bilateral retinal detachment and 4 patients had high myopia.

Retinal detachment was related with the coloboma in all but 4 eyes (Fig. 1). All of the cases with retinal detachment that were not associated with coloboma had high myopia and peripheral retinal breaks localized out of the colobomatous area. These cases, except for one (case 5), were treated with circumferential scleral buckling, cryocoagulation of the retinal breaks followed by drainage of the subretinal fluid. Case 5 also had a retinal break outside the coloboma; however, it was a giant tear on the nasal retina associated with proliferative vitreoretinopathy that led us to perform a primary pars plana vitrectomy (PPV) and apply a gas tamponade. Retinal detachment was localized to the nasal retina so that the macula was flat in this case. Vitreous surgery was applied in all the other cases. A standard three-port PPV was followed by a search for a retinal break at the intercalary membrane or locus minoris resistentiae, which could be identified by observation of the schieren sign during simultaneous air-fluid exchange and endodrainage. If a retinal break could not be found, a small retinotomy was created with diathermy for the drainage of subretinal fluid. We stripped the epiretinal membranes and performed relaxant retinotomies when it was necessary to flatten the retina in cases with advanced proliferative

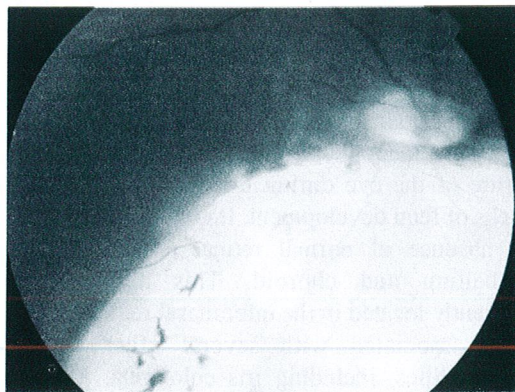


Fig. 1: Preoperative fundus photography of case 2. Note that the retinal detachment is associated with choroidal coloboma.

Table-1: Characteristics of the patients

Case	Age/Sex/Eye	Preop. VA	Characteristics of the eye	Surgery	Postop. VA	Anatomic Result	Follow-up (months)
1	55 / M / R	CF 2m	-RD and peripheral retinal breaks outside the colobomatous area -High myopia	-Scleral buckling Cryocoagulation -Drainage of SRF	CF 5m	Attached	42
2	36 / M / R	HM	-RD associated with the coloboma -High myopia -Fellow eye phthisis	-PPV -Endolaser -Silicon oil tamponade	CF 3m	Attached	19
3	49 / F / L	CF 1m	-RD associated with the coloboma Microphthalmus, Amblyopia, Nistagmus	-PPV -Endolaser -Silicon oil tamponade	CF 1m	Attached	27
4	18 / M / R	CF 1m	-RD associated with the coloboma -Fellow eye phthisis -Glaucoma	-PPV -Endolaser -Silicon oil tamponade	CF 1m	Attached	8
5	30 / M / R	20/20	-Break outside coloboma -High myopia -PVR -Fellow eye optic atrophy	-PPV -Endolaser -C <sup>2</sup> F <sup>6</sup> gas tamponade	20/20	Attached	12
6	30 / F / R	CF 3m	-RD associated with the coloboma -Fellow eye phthisis	-PPV -Endolaser -Silicon oil tamponade	20/200	Attached	10
7	17 / M / R	HM	-RD associated with the coloboma -Previous failed surgery, -PVR, cataract	-PPV -Retinotomy, -Endolaser -Silicon oil tamponade	CF 2m	Attached	23
8	6 / M / R	CF 4m	-Previous failed surgery, -Break near foveal border of coloboma	-PPV -Endolaser -Silicon oil tamponade	CF 4m	Attached	14
8	6 / M / L	CF 3m	-Previous failed surgery, -Macular pucker, -Cataract	-PPV, -Endolaser -Silicon oil tamponade	CF 1m	Attached	9
9	19 / M / R	HM	-RD and peripheral retinal breaks outside the colobomatous area -High myopia	-Scleral buckling Cryocoagulation -Drainage of SRF	CF 2m	Attached	17
9	19 / M / L	CF 1m	-RD associated with the coloboma	-PPV -Endolaser -silicon oil tamponade	CF 4m	Attached	11
10	52 / F / R	HM	-RD and peripheral retinal breaks outside the colobomatous area -High myopia -Fellow eye phthisis	-Scleral buckling Cryocoagulation -Drainage of SRF	CF 3m	Attached	12

VA: Visual Acuity  
CF: Counting finger  
RD: Retinal detachment

SRF: Subretinal fluid  
HM: Hand motions  
PVR: Proliferative vitreoretinopathy

PPL: Pars plana lensectomy  
PPV: Pars plana vitrectomy

vitreoretinopathy (PVR). Once the retina was flattened, an endolaser was applied around the drainage hole and along the borders of the coloboma except the papillomacular bundle area. Silicone oil (1000cs) was used as a long-acting intraocular tamponade and the patients were positioned face-down for a week. Inferior iridectomy was felt to be unnecessary because all of the operated eyes had inferior iris colobomas.

Lensectomy (phacoemulsification or pars plana approach) was performed in eyes with lenticular opacities that prevented good visualization of the retina.

## RESULTS

Patients were followed for a mean of 17 months (8-42 months). The retina was successfully reattached in all eyes after the first

or second surgery. Recurrence of detachment limited to the area of choroidal coloboma was detected in 5 eyes (41.6%). Normally developed retinal areas remained flat in all eyes at the final visit. Postoperative visual acuity improved in 7, worsened in 1 and did not change in 4 eyes.

We used silicone oil as the internal tamponading agent in 8 eyes, and perfluoropropane in 1 eye (case 5). Silicone oil was felt to be mandatory because of the possibility of unidentified retinal breaks along locus minoris resistentiae in cases with retinal detachment which are associated with the colobomatous area.

One eye (case 7) was referred to our clinic because of recurrent retinal detachment, which underwent vitreous surgery at another center and had received a silicone oil tamponade already. There was an advanced PVR, the retina was shortened at the inferior part and it became flat only after 180 degrees of relaxant inferior peripheral retinotomy. An argon laser was applied to the borders of the coloboma and retinotomy, and silicone oil was again selected as the tamponading agent. Case 8 had a bilateral large inferior choroidal coloboma extending from the optic disc to the periphery through the macula which was associated with retinal detachment bilaterally. As no retinal tear was observed pre- or postoperatively, both eyes were treated with vitrectomy and silicone oil tamponade. An endolaser was applied to the coloboma border, sparing the papillomacular bundle (Fig. 2A-B).

Both the right and left eyes of this case developed recurrent detachment during follow-up; right eye had a break located at the coloboma margin just under the fovea. We performed subretinal fluid drainage in the right eye in the postoperative third month and it was necessary to treat the papillomacular bundle with an endolaser as the break was the cause of detachment in this situation. The left eye developed a posterior subcapsular cataract that was detected during follow-up. Retinal detachment recurred following lensectomy and silicone oil removal at the postoperative sixth month because of proliferative membranes in the macular area of the left eye and was managed with vitreous surgery together with membrane peeling and silicone oil replacement. The foveal break (located outside the coloboma) was surrounded by a good endolaser application, which resulted in anatomical success, but visual acuity failed to improve.

Seven eyes were aphakic in the last examination. Lensectomy was performed in the same session with vitrectomy in 3 eyes, 2 eyes were already aphakic and cataract developed postoperatively in 2 eyes (Case 7 and 8-L). Lens extraction was performed during silicone oil removal during the 19th and sixth postoperative months respectively in these cases.

One eye developed open angle glaucoma in the first postoperative month, and was treated with the use of timolol. Three of the fellow eyes had developed phthisis and one underwent

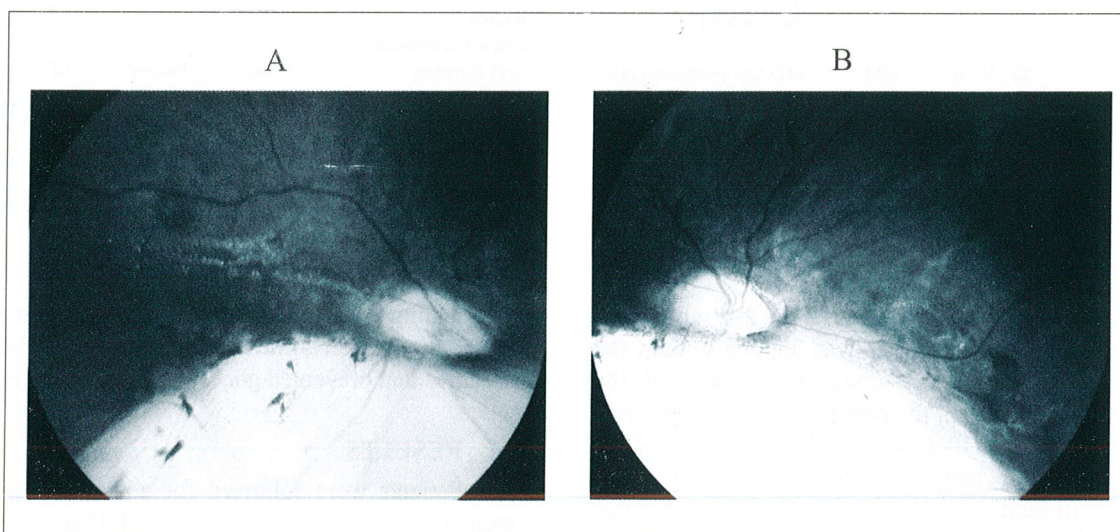


Fig. 2: Postoperative fundus photography of case 2: Temporal retina (A). Nasal retina (B). Note that retina is attached and there are laser scars along the borders of the colobomatous area.

vitreous surgery at another clinic, which resulted in anatomical success but optic atrophy.

#### DISCUSSION

Eyes with retinal breaks and retinal detachment outside the area of the coloboma can be managed successfully with conventional scleral buckling techniques, as was the case in 25% of our cases. However, in cases with coloboma-associated retinal detachment, external scleral buckling yields poor surgical results because of the posterior location of the breaks and absence of the choroid and retinal pigment epithelium, which makes identifying breaks difficult and creating chorioretinal adhesion impossible. On the other hand, some of these cases have high myopia and peripheral retinal degenerations causing rhegmatogenous retinal detachment not associated with choroidal coloboma. Such cases have all been successfully treated with circumferential scleral buckling surgery in the presented case series. Gonvers was the first to use pars plana vitrectomy and silicone oil tamponade to manage a recurrent retinal detachment in an eye with choroidal coloboma, and this approach has become the treatment of choice in these complicated cases (6-11).

The largest series in the literature belongs to Gopal et al. (8), who presented the results of the surgical treatment of 85 eyes with retinal detachment associated with coloboma of the choroid. Recurrent retinal detachment was reported to occur in 16% of silicone-used eyes and 60% of gas-filled eyes and they suggested that retinal detachment secondary to coloboma of the choroid was best treated by pars plana vitrectomy along with silicone oil tamponade. However it was difficult to compare the results of these two tamponading agents because there were only 5 eyes in the gas-filled group. The major disadvantages of silicone oil, i.e. the need for a second operation for the removal, secondary glaucoma, cataract formation and corneal side effects especially in aphakic cases, create the need to avoid the use of silicone oil whenever possible. McDonald et al. used sulphhexafluoride (SF<sub>6</sub>) or perfluoropropane (C<sub>3</sub>F<sub>8</sub>) and reported anatomic success in 6 of 7 eyes; over an average follow-up of 10 months, only one eye had undergone second surgery after vitrectomy because of recurrence in this series (9). We have used C<sub>3</sub>F<sub>8</sub> gas in cases with retinal detachment not

associated with the colobomatous area, and preferred silicone oil in cases with coloboma-associated retinal detachment, advanced PVR and recurrent detachments.

On the other hand, when the retinal break is located inferiorly, as in retinal detachment associated with coloboma, the ability of both gas and silicone oil to tamponade the break is compromised. Lee et al. reported a case in which a recurrent retinal detachment associated with coloboma following vitreous surgery with SF<sub>6</sub> gas was repaired successfully by using short-term intravitreal perfluoroperhydrophenanthrene (10), but this approach has not been performed by many because of the potential complications of perfluoroperhydrophenanthrene in the retinal layers.

Isolation of the choroidal coloboma with endolaser photocoagulation or cryotherapy is an appropriate approach especially in those cases where the detachment is directly related to coloboma because identifying breaks at this rudimentary area is difficult and adhesion does not occur in the absence of choroid and retinal pigment epithelium. Restricting the anomalous retina becomes more important in these eyes as subclinical detachments and breaks limited to the intercalary membrane have been observed in a considerable number of cases. Although Gopal et al. reported only 5 cases (5.8%) with postoperative redetachment localized to the colobomatous area, this was the case in 41.6% of our cases (8). This result is considered successful in these eyes since there was a strong barrier between the normal retina and the rudimentary retina.

Endolaser photocoagulation treatment around the optic disc can result in nerve fiber layer damage in the papillomacular bundle (7, 8). McDonald et al. observed anatomical success but a lack of functional recovery in 2 cases that received intraoperative endolaser photocoagulation treatment of the papillomacular bundle and they suggested the application of a krypton laser postoperatively in such cases (9). Gopal et al. leave this small area untreated in the first instance and treat it only in the event of recurrence and use a diode laser treatment postoperatively as diode laser has deeper tissue penetration (8). We applied either a diode laser or cryocoagulation along the borders of coloboma in

all eyes of our series. Special attention was paid to spare the papillomacular bundle during the isolation of the coloboma by laser, and this area was treated only in cases with recurrent detachments. In the right eye of case 8, which had recurrence secondary to a break at the temporal border of the coloboma, we needed to treat the papillomacular bundle by laser, as this seemed to be the only way to create adhesion and isolate the fovea from the break, but this eye failed to improve in visual acuity. Cyanoacrylate glue, which was reported to form an immediate, long-lasting, strong adhesion between the retina and the pigment epithelium, may be a good alternative to an endolaser, especially in eyes with associated optic disc coloboma to possibly avoid destruction of the nerve fiber layer during laser treatment (6,9). This may be especially important in cases where the colobomatous area involves some part of the optic disc and macula.

PVR was shown to be the most frequent cause of recurrence after successful surgery in the management of retinal detachments associated with choroidal coloboma. Gopal et al. (8) observed PVR in 15% of eyes, Hanneken et al. reported recurrence secondary to PVR in 6 of 8 eyes (6); 2 cases had developed PVR in our study. Good surgical technique and the avoidance of excessive laser application can reduce the number of eyes that develop PVR postoperatively.

In conclusion, vitreous surgery involving treatment of the colobomatous border with an endolaser and using long-acting internal tamponades provides an effective treatment for retinal detachments associated with choroidal coloboma. Circumferential scleral buckling together with cryotherapy and subretinal fluid drainage may also result in retinal reattachment in eyes with retinal detachment not related with the coloboma but with a peripheral retinal tear. The management plan in these anomalous eyes raises special concerns and must be individualized for each patient.

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