Retinal Detachment in the Eye with the Choroidal Coloboma

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SUMMARY
Introduction Choroidal coloboma is a congenital defect caused by an inadequate closure of embryonic fissure. About 40% of the eyes with the choroidal coloboma (CHC) develop the retinal detachment (RD). It is extremely difficult to manage these cases due to the lack of pigmentation at the site of choroidal coloboma.

Outline of Cases This is a case series of five patients with CHC and RD who were successfully operated using one of two different surgical techniques: pars plana vitrectomy (PPV) + silicone oil internal tamponade and/or scleral buckle with encircling band with laser photocoagulation (SB+EB+LPC) around the coloboma. The purpose of this paper is to present how to successfully handle patients with CHC, who have concurrent retinal detachment in the same eye and to compare two different techniques and indications for the predominant use of one of them in a specific case.

Conclusion Both surgical techniques can be applied with equal success in the operation of retinal detachment in eyes with the choroidal coloboma. Which one will be used depends only of the posterior segment of eye findings. We use scleral buckling in cases with RD accompanied by CHC when the peripheral break is evident and there are no breaks in the coloboma itself. We also perform, 2-3 days after surgery, laser burns around the coloboma, which is our modification of this technique. In all other cases it is indicated to perform PPV+silicon oil internal tamponade.

Keywords: choroidal coloboma; retinal detachment; scleral buckling; laser photocoagulation; vitrectomy

INTRODUCTION
Choroidal coloboma is a congenital defect caused by an inadequate closure of the embryonic fissure. About 40% of the eyes with the choroidal coloboma develop the retinal detachment. It occurs in 0.14% of the general population [1]. At the edge of the coloboma, the retina is divided into two layers: the inner and the outer, which is inverted and connected with the retinal pigment epithelium [2]. This spot of inversion of the retinal outer layer is the place of less resistance of retinal rupture [3, 4]. Retinal detachment with the choroidal coloboma is a surgical problem because of its specific pathomorphological substrate and difficulties to reattach retina.

CASE REPORTS
During the last 15 years 5 cases of the choroidal coloboma with retinal detachment due to retinal breaks have been operated at our Clinic (Table 1). During the follow-up period of the minimum 3 years in all of the five operated cases the retina had maintained reattached. Two of the patients had to undergo laser photo coagulation of retinal breaks which had developed after surgery on the different place then the previous ones. We used two different techniques to operate these five patients, scleral buckling in 2, and pars plana vitrectomy with silicon oil internal tamponade in 3 cases. The patients were relatively young when retinal detachment occurred, on average 24.4 years of age. Two of them had visual acuity slightly diminished before the event occurred. In three cases visual acuity was 1.0 on both eyes. All patients had bilateral chorioretinal colobomas, while two of them had accompanied iris and lens colobomas.

We are presenting a more detailed description of two cases from different groups in order to explain the technique used in a specific case.

A 25-year-old female patient presented at our hospital with the history of impaired visual acuity of formerly partially-sighted right eye 2 weeks ago. Personal and family histories were not relevant.

The patient underwent a detailed ophthalmological examination. The best corrected visual acuity of the right eye on admission was 1/60, and natively of the left eye was 3/60, and with the correction of 1.75, DSpH was 1.0. An intraocular pressure of the right and the left eye was 14 mmHg and 12 mmHg, respectively.

The examination of the ocular fundus of the right eye showed the choroidal coloboma below the optic disc, with the temporal bullous detachment involving the macula (Figure 1). Finding of the left ocular fundus was within physiological limits. Goldmann three mirror contact lens examinations showed 3 smaller breaks at 11 o’clock.
Upon appropriate preoperative preparation, the patient was operated by scleral buckling with the encircling band. On postoperative day 1, the control examination of the right eye fundus confirmed the complete reapplication of retina. On postoperative day 3, the patient underwent laser photocoagulation around the choroidal coloboma. The patient was discharged on postoperative day 4, with the right eye visual acuity of 0.50/60 and intraocular pressure of 20 mmHg.

At the nine-month control, the patient’s visual acuity was 3/60, and her retina maintained reattached (Figure 2).

The second patient was a 30-year-old male who was referred to our Clinic from a local health centre because of a sudden drop of visual acuity on the right, previously healthy eye. Upon admittance his visual acuity on the right eye was 0.10/60 with intraocular pressure 10 mmHg. The left eye visual acuity was 1.0 and IOP 13 mmHg. Examination of the anterior segment of the eye showed a bilateral coloboma of the iris at the 6 o’clock position along with the coloboma of the lens in the same meridian. The rest of the findings were within normal limits.

Examination of the eye fundus showed bilateral chorioretinal colobomas inferior to the papilla nervi optici, one smaller 1 diameter PNO in size, and second one bigger 3-4 diameter PNO in size, which corresponded to colobomas of the iris and lens. Changes were almost symmetrical on both eyes. Right eye fundus findings, besides the coloboma, showed a total bullose retinal detachment with giant posterior retinal tear from 8-12 o’clock (Figure 3). We decided to perform a combined operation of phacoemulsification of the lens with implantation of the posterior chamber lens and pars plana vitrectomy with internal silicon oil tamponade. Endolaser photocoagulation was performed during the operation around the colobomas and peripherally 360 degrees. Postoperatively the retina was reattached and visual acuity was, with correction, 3/60 three months after the operation silicon oil. The retina was attached in all parts including chorioretinal colobomas (Figure 4). The anterior segment showed stable posterior chamber intraocular lens and secondary cataract. His visual acuity was 0.5. During the follow-up period of 3 years his retina remained attached and visual acuity stable.

The remaining three patients had retinal detachment associated with retinal coloboma (Table 1). We used pars plana vitrectomy in two cases, scleral buckling with postoperative laser in the third patient.

Preoperative visual acuities were very low; half a meter in two cases and 4 meters in one case. Postoperatively the retina was reattached and the visual acuities improved to 3 m, 0.1 m and 0.2 m, respectively. Also, retina remained attached after silicone oil removal and in all three cases remained stable in the follow-up period of minimum three years.

### Table 1. Major patients’ data, treatment and results

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Clinical finding</th>
<th>Pre-op visual acuity</th>
<th>Treatment</th>
<th>Additional treatment</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>23</td>
<td>Female</td>
<td>Rhegmatogenous retinal detachment</td>
<td>0.50/60</td>
<td>PPV + silicone oil /</td>
<td>Retinal reattachment</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>25</td>
<td>Female</td>
<td>Rhegmatogenous retinal detachment</td>
<td>1/60</td>
<td>Scleral buckling</td>
<td>LFC barrage</td>
<td>Retinal reattachment</td>
</tr>
<tr>
<td>3</td>
<td>21</td>
<td>Female</td>
<td>Rhegmatogenous retinal detachment</td>
<td>0.50/60</td>
<td>PPV + silicone oil</td>
<td>/</td>
<td>Retinal reattachment</td>
</tr>
<tr>
<td>4</td>
<td>31</td>
<td>Male</td>
<td>Rhegmatogenous retinal detachment</td>
<td>0.10/60</td>
<td>PPV + silicone oil</td>
<td>/</td>
<td>Retinal reattachment</td>
</tr>
<tr>
<td>5</td>
<td>22</td>
<td>Male</td>
<td>Retinal detachment – gigant retinal tear</td>
<td>4/60</td>
<td>Scleral buckling</td>
<td>LFC barrage</td>
<td>Retinal reattachment</td>
</tr>
</tbody>
</table>

**Figure 1.** Choroidal coloboma and bullous retinal detachment  
**Figure 2.** Laser photocoagulation around the choroidal coloboma – retina is reattached nine months after operation
DISCUSSION

Detachment of the retina is frequent in the eyes with the choroidal coloboma, about 40% due to retinal break within the coloboma area, most often at the edge of colobomatous zone, or otherwise in the retinal area out of the coloboma [1].

This paper is an illustration of the application of two techniques combined together, scleral buckle (SB) with encircling band (EB) and laser photocoagulation around the zone of the coloboma.

We have 3 questions and will try to answer them.

1. How many techniques of the management of these problems are usually used?

Until now, two techniques of the management of these problems have been presented. The first and most frequently used technique is pars plana vitrectomy with the internal gas or silicone oil tamponade. The second technique is SB with EB.

2. What are advantages of these two techniques?

The advantages of the first technique are easier identification of the site of break, and the eye stabilization for subsequent breaks by an internal tamponade. Pars plana vitrectomy with the internal gas or silicone oil tamponade is most frequently used technique. In recent studies, this technique has been reported to produce very good results [5-8]. The problem with this technique is the application of retinopexy on the site of the intercalary membrane which has no pigmentation.

On the other hand, if it was possible to pinpoint exactly the peripheral breaks, the second technique SB+EB would be the technique of choice. The operation is simpler and recovery is faster and more comfortable to the patient. Surgical technique SB+EB with laser photocoagulation may be employed, especially in the event of no retinal break in the zone of the coloboma and without retinal detachment in this zone. This technique would be completely satisfactory if peripheral ruptures were predominant, without perioperative finding of retinal breaks on the intercalary membrane within the zone of choroidal coloboma, which is sometimes very difficult due to depigmentation and poor contrast at this area.

3. What is the third technique and when this technique may be employed?
The third technique is the combination of second technique (SB+EB) with the laser photocoagulation of the coloboma zone few days after operation. This surgical method is indicated for the treatment of retinal detachment in the eye with the choroidal coloboma, if no breaks in the choroidal coloboma zone were preoperatively found. Due to the big risk of breaks within the choroidal coloboma itself, we perform laser burns around the coloboma, which is our modification of this technique. With laser photocoagulation around the zone of the coloboma we can prevent retinal detachment due to formation of breaks at the intercalary membrane.

Our cases confirmed the findings presented in the literature. As shown, it is possible to identify breaks in the intercalary membrane pre- or intra-operatively. We use scleral buckling for the cases with RD accompanied with CHC when peripheral break is evident and there are no breaks in the coloboma itself. On the other hand breaks at the locus minoris resistentiae are not identifiable but can be expected along the coloboma border. Additionally secured coloboma zone by laser photocoagulation, with the SB+EB in the zone of peripheral breaks, would be, according to our experience, a completely satisfactory surgical method for the treatment of retinal detachment in the eye with the choroidal coloboma, if no breaks in the choroidal coloboma zone were preoperatively found.

REFERENCES

Аблација мрежњаче у оку с хороидалним колобомом

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КРАТАК САДРЖАЈ
Увод. Хоридални колобом је конгенитално оштећење које настaje услед неправилног затварања ембрионалне фишу-ре. У око 40% случајева конгенитални колобом развије се аблација мрежњаче. Извуцао се тешко решаваћи ове случајеве услед недостатка пигментације на самом колобому.
Приказ болесника. Приказана је серија од пет испитаника с хорошидалним колобомом и аблацијом мрежњаче који су успешно оперисани двајма различитим хируршким техникама: 1) витректомијом парс плана с унутрашњом тампонацијом силиконским улом; и 2) техником склералне пломбе са серијом и ласерском фоторокоагулацијом око колобома. Овим радом желили смо да покажемо како се могу успешно лечити болесници с хорошидалним колобомом и аблацијом мрежњаче, као и да упоредимо две различите хируршке технике и индикације за њихово коришћење у специфичним случајевима.
Закључак. Обе технике се могу применити с подједнаким успехом у хируршком лечењу аблације мрежњаче с хорошидалним колобомом. Која ће од њих бити коришћен зависи од налаза на задњем сегменту оке. Ми применујемо технику склералне пломбе у случајевима аблације мрежњаче удружене с хорошидалном колобомом када су јасне периферне рутуре, а не виде се рутуре на зони колобома. Такође, други-трети дан после операције вршимо ласерску барајку мрежњаче око колобома, што је наша модификација ове технике. У свим осталим случајевима индиковано је урадити витректомију с ендотампонадом силиконским улом.
Кључне речи: колобом хориоиде; аблација ретине; ла-серска фоторокоагулација; склерална пломба; витректомија

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