Rhegmatogenous Retinal Detachment in Childhood

Chrapek Oldřich[1,2], Šuráňová Markéta[1,2], Chovancová Veronika[2], Sičová Kristína[2], Březík Michal[2], Vysloužilová Daniela[1,2], Matušková Veronika[1,2]

[1]Eye Clinic, University Hospital, Brno, Czech republic
[2]Department of Ophthalmology, Faculty of Medicine, Masaryk University, Brno, Czech Republic

Doc. MUDr. Oldřich Chrapek, Ph.D.

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Correspondence address:
Slunečná 820/1D
779 00 Olomouc
Česká republika
E-mail: chrapek.oldrich@fnbrno.cz

SUMMARY

Aims: To retrospectively evaluate the anatomical and functional success of surgical treatment for rhegmatogenous retinal detachment (RRD) in pediatric patients aged 18 years and younger.

Material and methods: The study includes 14 eyes of 14 patients, 9 (64%) of whom were males, with an average age of 12 years. All patients underwent surgery for rhegmatogenous retinal detachment (RRD) performed by a single surgeon at the Department of Ophthalmology, University Hospital Brno, and Masaryk University, from July 1, 2019, to June 30, 2024. The surgical techniques used included cryosurgery and 25G+ pars plana vitrectomy (PPV). In 3 patients a 25G+ PPV was combined with pre-equatorial scleral buckling. A history of blunt ocular trauma was permissible. The cause of RRD was retinal tear(s), regardless of their number or location. The transparency of the anterior segment enabled reliable visualization of the posterior segment. Preoperative proliferative vitreoretinopathy (PVR) grades A-D2 were acceptable. Patients with a history of penetrating ocular trauma were excluded. Anatomical success was defined as complete retinal reattachment. Each patient's final visual acuity (VA) was assessed using a Snellen chart. Numerical outcomes were expressed as arithmetic means and percentages. Since no comparative analysis was conducted between different groups, statistical tests were not required.

Results: In 13 patients (93%), complete retinal reattachment was achieved. In 1 patient (7%), the retina became detached again after silicone oil removal, with rapid progression of PVR, leading to anatomical treatment failure due to inoperability. A total of 11 patients (78%) achieved visual acuity (VA) of ≥ 4/40.

Conclusion: We consider cryosurgical techniques utilizing episclerally fixed encircling bands and buckles, 25G+ PPV, and potentially a combination thereof as appropriate methods for managing RRD in children.

Key words: rhegmatogenous retinal detachment, visual acuity

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INTRODUCTION

Rhegmatogenous retinal detachment (RRD) is the separation of the neurosensory retina in the presence of a tear or hole from the retinal pigment epithelium, upon which fluid penetrates through this tear or hole into the subretinal space, resulting in a mutual separation of both retinal layers. The incidence of RRD in adult patients is associated with changes of the vitreous body most commonly upon a background of patient age, in which liquefaction of the vitreous occurs with its detachment from the retina and collapse into the space behind the lens of the eye. Movements of the eye and head then cause traction in the region of the posterior edge of the base of the vitreous, with potential onset of a tear of the retinal neuroepithelium, infiltration of fluid into the subretinal space and the progression of retinal detachment. However, this pathogenic development of RRD does not apply in the case of children. In children RRD is generally associated with a non-ablated vitreous, and tears or holes in the neuroepithelium most commonly occur in connection with risk states, which were defined in the 1960s and 70s when large cohorts of RRD in child patients were presented and the risk factors were defined as trauma, myopia, aphakia and retinopathy of prematurity [1–5].

However, the latest studies remind us also of the connection between RRD in childhood and pseudophakia (usually due to a congenital cataract), developmental defects of the type of iris coloboma, genetically conditioned disorders such as Stickler syndrome, Marfan syndrome, genetically conditioned vitreoretinopathies such as Wagner syndrome and X-linked juvenile retinoschisis. If none of the risk factors is present, in children the condition is referred to as idiopathic retinal detachment [6].

The procedures usually used today for the treatment of RRD in childhood are cryosurgical techniques or pars plana vitrectomy (PPV). In the 1950s Schepens referred to the use of cryosurgical techniques with the aid of episclerally fixed cerclage bands and plombs [7,8]. At the turn of the 1970s Machemer introduced PPV [9].

The aim of this study is to conduct a retrospective evaluation of the anatomical success and functional results of the surgical treatment of RRD in pediatric patients, i.e. aged 18 years and younger, who were operated on at the Clinic of Eye Treatments at the University Hospital Brno in the period from July 1, 2019 to June 30, 2024. The first objective is to assess the success rate of the performed primary operations, the second objective is to evaluate the success of definitive reattachment of the retina. The third objective is to evaluate the resulting postoperative visual acuity (VA) of the patients.

COHORT AND METHOD

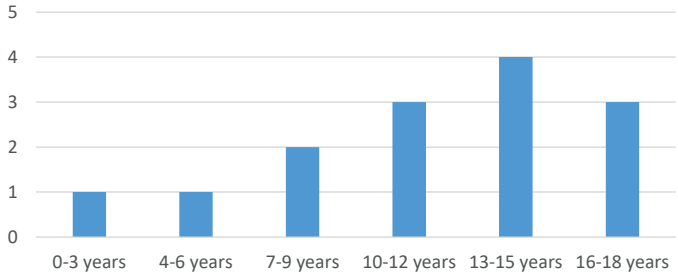
Patients with RRD were included in the evaluated cohort, in which blunt ocular trauma in the patient's medical history was permissible. The cause of retinal detachment was retinal tear(s), regardless of their number and localization. Transparency of the cornea and anterior chamber enabled reliable visualization of the posterior segment of the eye. Patients in whom proliferative vitreoretinopathy (PVR) was diagnosed before surgery within the grades of A-D2 were also included in the evaluated cohort. Patients who had experienced penetrating ocular trauma were excluded from the study.

Surgical treatment of RRD on the patients in the cohort consisted in the use of a cryosurgical technique or 25G+ PPV with intraocular tamponade using silicone oil (SO). Pneumatic retinopexy was not performed on any of the patients.

The basis of cryosurgical treatment was perilimbal peritomy, localization of the retinal tear(s) and exocryocoagulation of the edges of the retinal tear(s) with the use of an indirect ophthalmoscope and episcleral plombage or cerclage.

The basis of three-port 25G+ PPV was removal of the vitreous, identification of the retinal tears, according to requirement peeling of the epi/subretinal membranes, temporary perioperative stabilization of the detached retina with air or liquid perfluorocarbon, treatment of the retinal tears with endolaser photocoagulation or exocryocoagulation, and intraocular tamponade with SO. The most frequently used was SO 1300 cts. In the case of complicated findings, pre-equatorial cerclage was also selected during the course of 25G+ PPV, with the suturing of a cerclage band with a width of 2 mm into the pre-equatorial position with the anterior edge of the cerclage band 11 mm from the corneal limbus.

The patients were evaluated in the early postoperative period, i.e. 1 to 3 months after surgery, but we also took into account the long-term results usually associated with the moment of evacuation of the silicone oil. Examination of the ocular fundus was performed on the patients biomicroscopically and by indirect ophthalmoscopy. The operation was evaluated as anatomically successful if the retina was reattached in its full scope, i.e. if contact of the neuroepithelium and the pigment epithelium was restored on the posterior pole and throughout the entire periphery. The operation was evaluated as anatomically unsuccessful if the retina was judged to be detached, i.e. if persistent separation of the neuroepithelium and pigment epithelium was found, at least in part, on the posterior pole or in the retinal periphery. Resulting (postoperative) VA was assessed in each patient, examined as natural VA or VA with glasses correction. If natural VA or VA with glasses correction did not appear to be satisfactory in the postoperative period, for example after implantation of SO, glasses correction according to a current measurement on an autorefractometer was selected in order to determine the resulting VA. Examination of VA was performed on a Snellen chart. We used the best VA recorded within the postoperative period as the resulting VA.



Graph 1. Age of patients in the cohort. Frequency distribution of operated children across different age groups

The arithmetical average was used for numerical expression of the attained results, and the numerical values were also expressed in percentages. Since there was no mutual comparison of different groups, it was not necessary to use a statistical test.

The cohort comprised 14 eyes of 14 patients, of whom 9 (64%) are boys and 5 (36%) girls. The average age of the patients in the cohort was 12 years (youngest 3 years, oldest 18 years), and the age distribution of the patients in the cohort is illustrated in Graph 1. All the patients were operated on by a single surgeon at the Clinic of Eye Treatments at the University Hospital Brno in the period from July 1, 2019 to June 30, 2024. The right eye was afflicted in 8 cases (57%) and the left eye in 6 cases (43%). In 13 eyes of 13 patients the finding concerned newly diagnosed, untreated RRD. In the case of 1 eye of 1 patient, the operating surgeon took into our care a patient who had originally been unsuccessfully operated on by another surgeon for newly diagnosed RRD. All the patients were operated on under general anesthesia.

RESULTS

A cryosurgical procedure was performed on 5 (36%) patients, with suturing of an episcleral plomb or cerclage band, while 25G+ PPV with SO tamponade was performed on 9 (64%) patients, in 3 of whom 25G+ PPV was combined with suturing of a pre-equatorial cerclage. 25G+ PPV was

not closed with gas tamponade in any of the patients. The frequency and type of performed procedure on the patients in the cohort is expressed in Graph 2.

A single operation was sufficient in order to achieve retinal reattachment in 12 (86%) patients. In 2 (14%) patients the retina was not reattached after a cryosurgical procedure, and both these patients required reoperation. For one of these patients a cryosurgical procedure was again selected for reoperation, in which pre-equatorial cerclage in the place of the tear was reinforced with symmetrical accenting of the cerclage, while for the second patient 25G+ PPV was chosen as the reoperation procedure with implantation of SO. Both reoperations were successful. Overall, in the early postoperative period we can state that out of 14 patients we achieved retinal reattachment within the full scope in 14 eyes (100%), and therefore definitive anatomical success, even if at the price that SO was present inside the eye postoperatively in 10 (71%) patients (9x after primary operation, 1x after reoperation). In order to achieve this result it was not necessary to operate on any of the pediatric patients more than 2x.

In the period of 3–28 months after the primary operation we proceeded to evacuation of SO in 8 patients. The time in months from surgery until the moment of evacuation of SO for each individual pediatric patient is expressed in Table 1.

At the end of the observed and evaluated period (30. 6. 2024) SO had been successfully evacuated in 7 patients, and the retina remained attached following evacuation of SO within the full scope. In 2 (14%) patients anatomical success was achieved at the end of the observation period, though this was at the price that SO remained present inside the eye. In 1 patient in whom evacuation was performed 8 months after the primary operation, the retina once again became completely detached, with rapid development of PVR and inoperability of the finding. In the case of this 1 (8%) patient we therefore state anatomical failure of the treatment.

The resulting level of VA in the postoperative period is expressed in Graph 3. Only 1 (8%) patient achieved VA of ≥ 4/8. In the case of 11 (78%) patients we can state VA of ≥4/40. For the 1 patient in whom we did not achieve anatomical success, the resulting VA was 0.

DISCUSSION

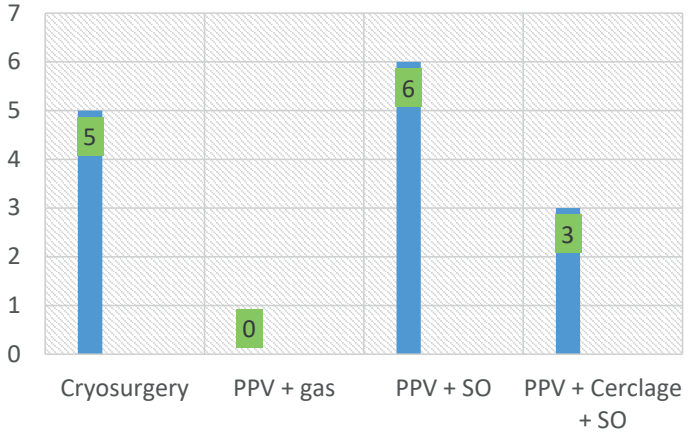
Numerous studies point to the fact that the etiopathogenesis of RRD in children and adults is different. The authors of these studies mention that the risk factors of RRD in childhood include among others retinopathy of prematurity, myopia and blunt ocular trauma [1–5]. We also observed these 3 risk factors in our own cohort, in which 3 pediatric patients had been treated since early childhood for stage ROP 3+ with cryoretinopexy, 4 patients were monitored for myopia gravis, of whom 2 had undergone scleroplasty, 1 patient was monitored for myopia modica and 1 for myopia levis. In 2 patients there was an ocular anamnesis of blunt ocular trauma, in 1 child blunt head trauma after falling off a horse. In only 2 (14%) of the children in our cohort we stated an entirely negative ocular and personal anamnesis, and RRD in these children could be indicated as idiopathic. Our finding here corresponds with the observation of Lee et al., who stated idiopathic RRD in 19% of children in their cohort [6].

Three therapeutic procedures are generally considered in the treatment of RRD at present: pneumatic retinopexy, cryosurgical techniques and PPV. For children in the case of RRD we decided between a cryosurgical solution and PPV, in which we prefer a cryosurgical approach and use it in all cases where the retinal finding so allows. We apply the criteria for indication of a cryosurgical procedure identically for children and adults. The condition must be uncomplicated RRD with PVR maximally of stage A or B. Blunt ocular trauma is permissible in the pa-

Table 1. Evacuation of silicone oil. Time period in months between the implantation of silicone oil and its final evacuation for each individual patient who underwent both SO implantation and subsequent evacuation

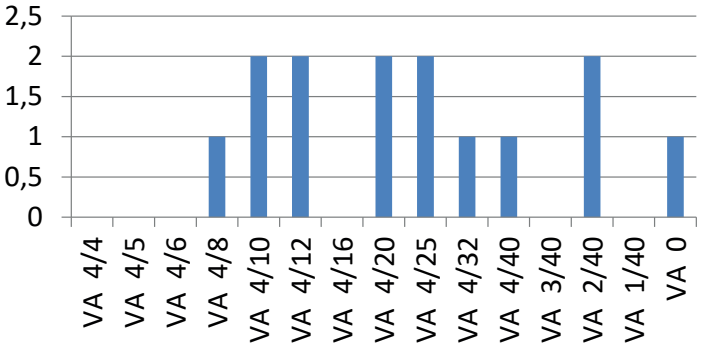
	Final Evacuation of Silicone Oil							
Patient Number	1	2	3	4	5	6	7	8
Number of Months	3	4	7	8	10	15	20	28

SO – Silicone oil



Graph 2. Surgical procedures. Frequency distribution of various types of surgical procedures performed in the studied cohort

PPV – Pars plana vitrektomy, SO – Silicone oil



Graph 3. Final visual acuity of operated eyes. Frequency distribution of different levels of final visual acuity among patients in the cohort

VA – Visual acuity

tient's medical history. The cause of retinal detachment is one pre-equatorial retinal tear, two pre-equatorial tears anywhere in the retinal periphery, or three adjacent pre-equatorial tears, in which all three are located within a range of maximally one quadrant (90 degrees of circumference). All the patients must have their own lens, transparency of the lens and all optic media must be problem-free in order to enable reliable visualization of the posterior segment of the eye. Our preference for this surgical procedure on children is documented by the fact that 5 (36%) children in this pediatric cohort were operated on using this cryosurgical technique.

25G+ PPV with intraocular SO tamponade was performed 9x, in which 6 of these procedures were simple 25G+ PPV and 3 procedures were 25G+ PPV combined with pre-equatorial cerclage, which we indicated in the case of RRD in an eye treated in childhood for ROP with cryoretinopexy, or if RRD was caused by a tear in the lower retinal periphery or if RRD at the moment of determination of the diagnosis was complicated by PVR ≥C1. In the indication of this procedure, which combines PPV with cerclage, we start out from our own positive experiences. We used this technique on 11 adult patients with RRD in the sole seeing eye, in all of whom we achieved complete retinal reattachment by means of the primary operation [10]. Other authors also mention the benefit of combining PPV and cryosurgical techniques in pediatric patients with RRD. Similarly to us, Smith also used a cryosurgical approach, PPV or PPV combined with buckling techniques for the treatment of RRD in 212 eyes of 191 patients aged under 18 years. He states that primary operation was successful in a total of 65% of eyes, while the best results were observed in children operated on with PPV in combination with cerclage (68%), while the cryosurgical approach was the second most successful (63%), and the poorest results were attained by PPV performed separately (51%) [11].

Of other studies, Al-Zaidi highlights the potential benefit of combining PPV with buckling techniques in order to improve the anatomical result of RRD surgery in children. Out of 166 eyes of 148 patients aged under 16 years he used a cryosurgical approach on 19 (11.5%) eyes, and indicated PPV as the primary operation in 141 (85%) eyes, though of these PPV was performed separately on only 27 (16.5%) eyes, while PPV combined with cerclage was performed on 114 (68%) eyes [12].

In our cohort 25G+ PPV was not closed with gas tamponade in any of the children, but SO was always implanted at the end of 25G+ PPV. The reasons mentioned in the literature for the preference for silicone oil in the treatment of RRD in childhood include problems with maintaining the positioning regimen in children in the case of gas tamponade [12], as well as PVR, whether this is present at the moment of determination of the diagnosis of RRD or complicating the postoperative course of healing. Al-Zaidi in his cohort of 166 eyes stated PVR in 55 (33%) eyes at the moment of determination of the diagnosis of RRD, in which 18 eyes had PVR grade C and worse, and 4 eyes had present PVR changes on the posterior pole. He closed PPV in 96 eyes (57.8%) with implantation of SO [12].

In our cohort the average resulting VA of the patients after surgery was 4/20, which corresponds to a value of 0.2. Smith attained resulting VA of 20/372 at 3 months after surgery, which corresponds to a value of 0.05 [11]. However, resulting VA depends to a very large extent on the nature of the initial finding and the complications associated with the condition of the operated eye.

CONCLUSION

We can state that we achieved anatomical success in the case of 11 (78%) patients in our cohort, in which no intraocular tamponade is now present in the operated eye. In anatomical terms these patients can be considered completely cured. In the case of 2 (14%) patients we can state that we achieved anatomical success, but only at the price that SO still remains present in their vitreous cavity. In the case of these patients we therefore cannot state definitive success in the treatment of RRD. Assessing the degree of anatomical success or failure in this small subgroup of patients requires further monitoring with evaluation of the applicable number of findings of reattached retina following the definitive drainage of SO in future. In 1 (7%) patient the retina again became detached after the evacuation of SO, without the possibility of further surgical correction. This fact is also linked to the functional result, in which the patient's resulting VA was 0. On the other hand, it is necessary to emphasize that 11 (79%) patients attained a resulting VA of 4/40 or better. Overall we therefore consider a cryosurgical technique with the use of episclerally fixed cerclage bands and plombs, 25G+ PPV and if applicable also their mutual combination to be appropriate methods for the treatment of RRD in childhood.

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