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Pupillary block glaucoma in child with persistent hyperplastic primary vitreus – case report

Blok źreniczny u dziecka z pierwotnie przetrwałym hiperplastycznym ciałem szklistym – opis przypadku

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Summary: Purpose: The purpose of this paper is to present the case of a patient with PHPV, in whom the complication in a form of pupillary block glaucoma was observed as a result of idiopathic and complete lens dislocation into the anterior chamber.
Material and methods: Examination was performed on 3 months old boy with grey pupilla reflex, noted from the birth, in the right eye. Microphthalmia, subcapsular cataract and anterior-posterior form of PHPV was found in that eye. The patient was qualified to surgical treatment. Before the appointed time of operation the increased intraocular pressure and significant globe enlargement were noted. The reason of that was pupillary block caused by lens dislocation into the anterior chamber. One-step surgical procedure: trabeculectomy, lensectomy and cutting out the retrolenticular fibrous membrane, was performed in urgent course.
Results: Performed complicated and difficult surgical treatment resulted in normalizing IOP and created good conditions for vision rehabilitation for the child. Postoperatively the detachment of the choroid was noted as a transient complication.
Conclusions: In this case luxated lens and pupillary block was caused by constriction of retrolenticular fibrous membrane. Early surgical intervention is necessary to prevent progressive pathologic changes in eyes with this developmental disorder and to obtain the best possible visual results.

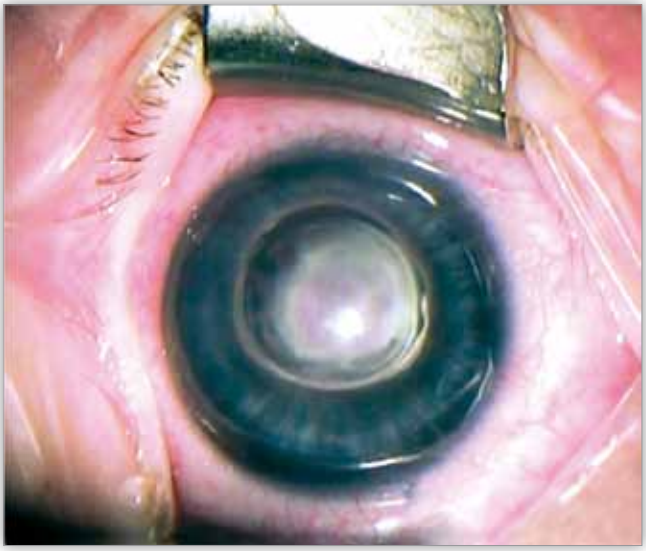
Słowa klucze: pierwotne hiperplastyczne przetrwałe ciało szkliste (PHPV), podwichnięcie soczewki, jaskra, lensektomia, trabekulektomia, pozasoczewkowa błona włóknista.
Key words: persistent hyperplastic primary vitreus (PHPV), lens luxation, glaucoma, lensectomy, trabeculectomy, retrolental fibrous membrane.

The recommendations for surgical treatment of persistent hyperplastic primary vitreous have changed in the last years simultaneously with increasing knowledge on the subject of PHPV and significant progress in surgical instruments (1,2). According to early reports on PHPV, it was recommended to perform surgery at as early an age as possible in order to prevent later complications, which may lead to eye pain and its atrophy (3). Particular stress is laid nowadays on the possibilities of improvement of the infant vision after their surgical treatment. Unfortunately many controversies are still present among which, the most important, surrounds the problem what kind of a patient should be treated. In the opinion of many authors of presented reports only eyes with anterior form of PHPV have good prognosis to obtain satisfying visual acuity after surgical intervention (4). Good functional results that might be expected are limited by changes in posterior pole and anomalies of retina and optic nerve. Early performed surgical treatment should be optimistically evaluated since it may prevent pathological changes induced by PHPV, from becoming worse and gives hope for positive visual results.

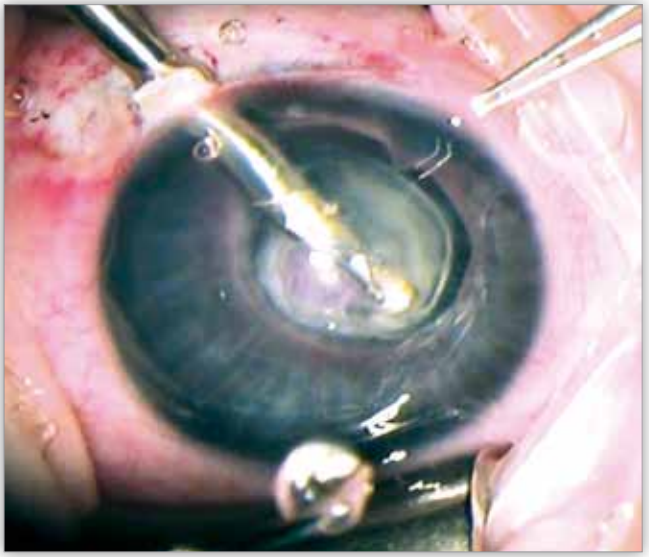
The purpose of this paper is to present the case of a child with persistent hyperplastic primary vitreous and with rapidly increasing buphthalmia caused by complete lens dislocation into the anterior chamber.

Case description

Parents with 2 months old infant were seen in the Department of Ophthalmology of children's Memorial Health Institute, because of white pupilla reflex noted from the birth in their child's right eye. Ophthalmological examination showed light reactions of both pupillas as normal. Corneal diameter of the right eye was 10.5 mm and left 11.5 mm. In the left eye no abnormalities were noted, both in the anterior and posterior segment. Slit lamp examination of the right eye revealed shallow anterior chamber and central subcapsular lens opacification that was in conjunction with fibrous vascularized membrane pulling ciliary processes. In biometry measure-



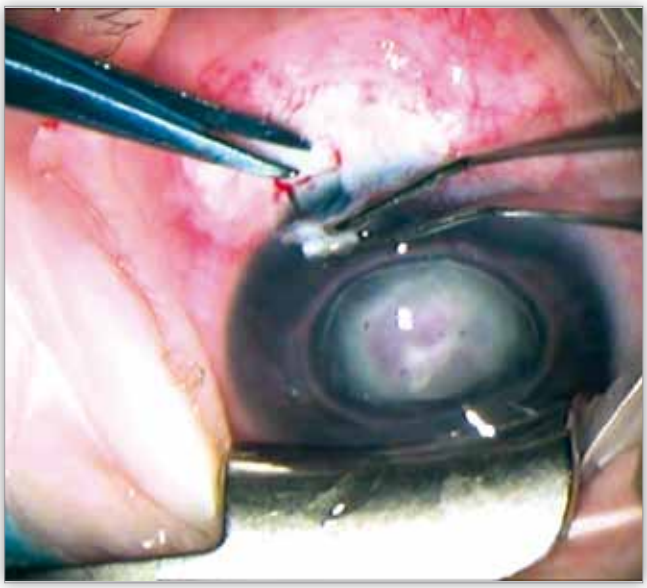
Ryc. 1. Soczewka zwichnięta do komory przedniej.
Fig. 1. Dislocated lens into the anterior chamber.



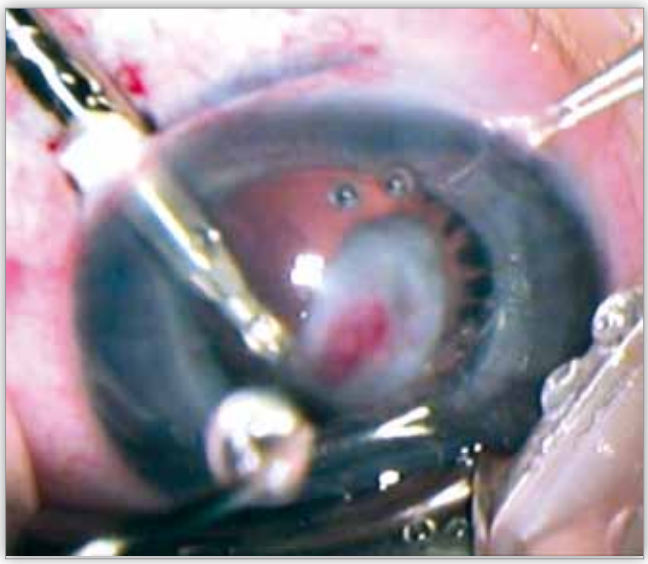
Ryc. 3. Lensektomia przez otwór trabekulektomii.
Fig. 3. Lensectomy through trabeculectomy.

ments length of the right eye was 15.5 mm and the left 17.5 mm. Ultrasonography examination showed the funiculus coming from retrolenticular membrane to optic nerve disc. Clinical picture was consistent with the diagnosis of developmental disorder, which was anterior-posterior form of persistent hyperplastic primary vitreous. The child was qualified to planned surgical treatment including: lens extraction, cutting off fibrous cord and excision of postlenticular membrane, in order to release ciliary processes from being tense. The assumption of surgical treatment was to restore the anatomy of eyeball and to make conditions for infant's visual rehabilitation. In ten days after the examination and before the planned date of surgery the parents came back to the clinic because of rapidly increasing buphtalmia of the right eye. It was caused by complete lens dislocation into the anterior chamber with pupillary block (Fig. 1). Intraocular pressure in the right eye increased to the level of 42 mmHg whereas in the left eye equals 15 mmHg. The right eye length prolonged to 19.5 mm and the left did not change (17.5 mmHg). Antiglaucomatous drug treatment such as eye drops and mannitol (i.v.), were given to a child as preparation to surgical intervention. The child was qualified to multiprocedural operation because of observed complications.

Trabeculectomy was performed under a scleral flap after conjunctiva dissection and exposing the surgical limbus (Fig. 2). Next the anterior chamber was inflated with viscoelastic agent and the anterior capsule was separated from endhothelium. After that the vitrectom was installed through sclerectomy and the lens was removed from the anterior chamber (Fig. 3). The retrolenticular fibrous membrane was cut off with vitrectom and microscissors relasing the ciliary processes. Fibrovascular cord connective with optic disc was severed after being coagulated; its stump over optic disc was left (Fig. 4, 5, 6). Finally two sutures were placed on scleral flap and conjunctiva was closed.



Ryc. 2. Trabekulektomia.
Ryc. 2. Trabeculectomy



Ryc. 4. Wycięcie włóknistej błony zasoczewkowej pociągającej wyrostki rzęskowe.
Fig. 4. Cutting off the retrolenticular fibrous membrane pulling ciliary processes.

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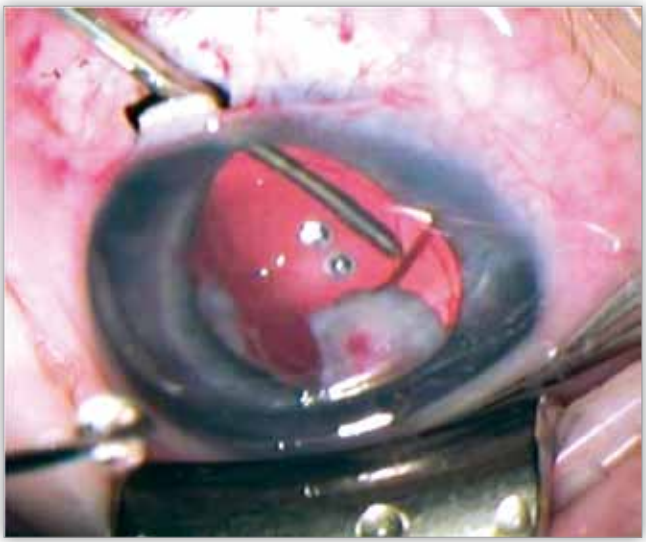
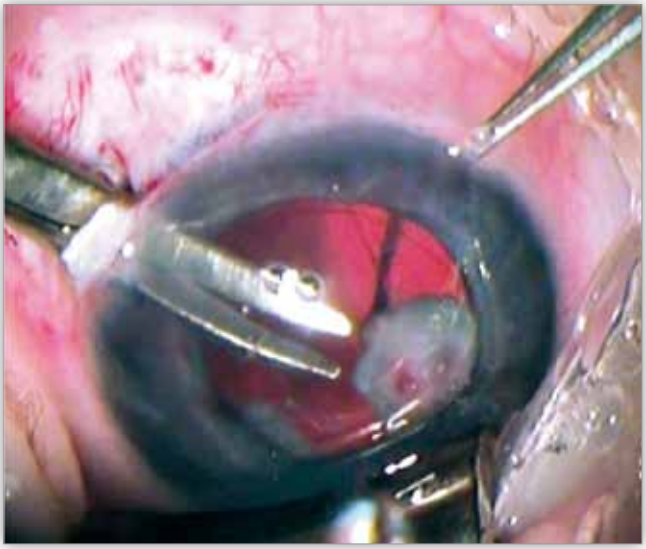


Ryc. 5. Diatermokoagulacja włóknistonaczyniowego powrózka połączonego z tarczą n. wzrokowego.
Fig. 5. Diathermocoagulation of the fibrovascular cord connected with optic disc.

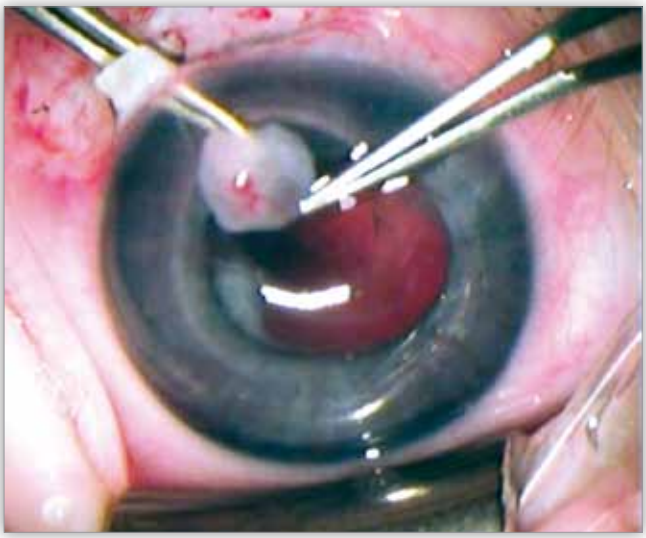


Ryc. 6. Wycięcie błony włóknistonaczyniowej.
Fig. 6. Excisio of the fibrovascular membrane.

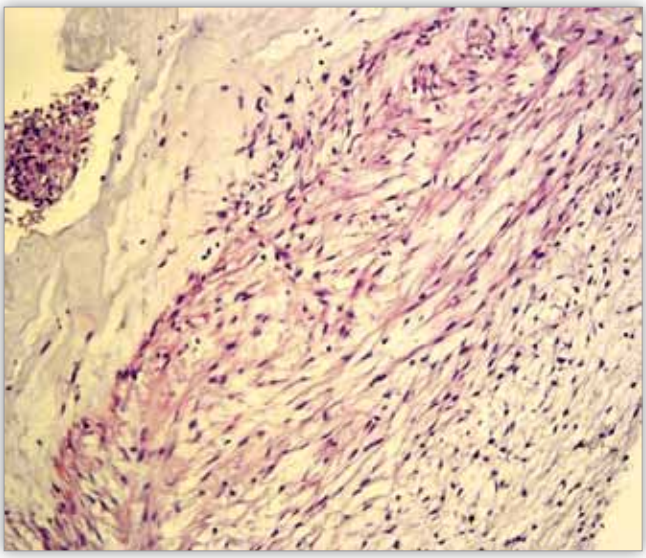
Postoperatively two serious, but temporary, complications were noted despite the fact that we used the closed-chamber technique. The first one, cornea decompensation, was found in 24 hours after the surgery. It was caused by contact with dislocated lens and surgical manipulation in the anterior chamber. The second complication was hemorrhagic detachment of the choroid induced by rapid decrease of intraocular pressure to 8 mmHg. During hospitalization gradual improvement of clinical state was observed. Postoperatively antiglaucomatous drops were given to maintain IOP on the level of 16 mmHg, and after three months it was still on this level without any drops.

Discussion

Most authors in their reports about coexistence of lens opacification and anterior-posterior form of PHPV suggest the necessity of surgical treatment (5). Decision of no treatment may lead to many complications such as closure-angle glaucoma, vitreous hemorrhage, retinal detachment and even eyeball atrophy. Lens dislocation in the eye with no treated PHPV has been not described in literature, so far. However, surgical treatment of eyes with developmental abnormalities seem to be quite a difficult procedure burdened with many possible complications. It is demanded to be performed by an experienced surgeon in a specialized center where additional studies (USG, UBM) can be done to let the surgeon decide on the most satisfying surgical technique. Our gain in those cases is to minimize the risk of intra- and postoperative complications and to choose the least invasive surgical technique. The reasons of soft lens dislocation into anterior chamber in children are different. The most common are posttraumatic and caused by congenital predispositions. In our case we suspected that the lens dislocation resulted from rentrolenticular membrane contraction, what caused "pushing" the lens into anterior chamber. Those assumptions were confirmed by histopathological examination of excised membrane and the cord adjoining it to optic disc (Fig. 7).



Ryc. 7. Pobieranie wyciętej błony w celu badania histopatologicznego.
Fig. 7. Uptaking of the cut out membrane for histopathological examination.



Ryc. 8. Obraz mikroskopowy błony hialinowej i fibroblastów.
Fig. 8. Microscopical image of fibroblasts and hyaline membrane.

Microscopical image showed contracting fibroblasts and and hyalinizating membrane (Fig 8). Acute angle-closure glaucoma induced by retrolenticular membrane contraction in a 30 years old female is also presented by Sawada et al in their report (6). In our case lens dislocation was the reason of disturbances of aqueous circulation, high increase of intraocular pressure and eye ball enlargement. Decision of necessity of surgical intervention concerned not only removing the reason of the complications – dislocated lens and retrolenticular membrane, but also glaucoma. Taking into account possible lesions in the filtration angle, which might have appeared in microphthalmia with buphthalmus, we decided to perform trabeculectomy in one-step surgery. Other activities to decrease intraocular pressure are presented by Khan who describes the case of 4 months child with glaucoma (40 mmHg), PHPV and buphtalmus, but without lens dislocation (7). In that case surgery included lensectomy and membranectomy. Intraocular pressure was to be decreased by diode laser cyclodestruction of the inferior hemisphere. Postoperatively no serious complications were noted by the author. In one year after surgery IOP was on the level of 15 mmHg on dorzolamid 2% and timolol 0.5%. In our case at early postoperative stage we found cornea haziness with Descemets' folds and detachment of the choroids in the second 24 hours. These complications were caused by surgical removal of pupillar block with rapid IOP growth followed by its sudden reduction. It appeared that despite performing only one ora serrata 1.5 mm cut during surgery and closed-chamber technique, which is recommended by many authors for such cases, we noted a few complications. Our gain in qualification the patient to surgical treatment was to give him a chance for visual rehabilitation and to obtain useful visual acuity. Later complications that occurred such as lens dislocation followed by pupillary block glaucoma made the surgical procedure much more difficult and induced serious potential postoperative complications.

The conclusions of summing up the analyzed case are the following:

1. In the presented case of anterior-posterior form of PHPV lens dislocation followed by pupillary block glaucoma was caused by contraction of retrolenticular fibrous membrane.
2. Early surgical intervention is necessary to prevent progressive pathologic changes in eyes with this congenital disorder and to create conditions for rehabilitation and visual development for a child.

References:

1. Kanigowska K, Grałek M, Klimczak-Ślączka D, Seroczyńska M: *Pierwotnie przetrwałe ciało szkliste – wada rozwojowa gałki ocznej u dzieci*. Klin Oczna 2006, 108, 225-227.
2. Dass AB, Trese MT: *Surgical results of persistent hyperplastic primary vitreus.* Ophthalmology 1999, 106, 280-284.
3. Anteby I, Cohen E, Karshai I, BenEzra D: *Unilateral persistent hyperplastic primary vitreus: Course and Outcome*. J AA.POS 2002, 6, 92-99.
4. Pollard ZF: *Results of treatment of persistent hyperplastic primary vitreus.* Ophthalmic Surg 1991, 22, 48-52.
5. Kanigowska K, Grałek M, Chipczyńska B, Hautz W: *Problemy w leczeniu chirurgicznym pierwotnie przetrwałego ciała szklistego u dzieci.* Klin Oczna 2006, 108, 51-55.
6. Sawada H, Fukuchi T, Ohta A, Suda K, Togano T, Nakatsue T, Funaki S, Hara H, Shirakashi M, Abe H: *Persistent hyperplastic primary vitreous – a case report of adult onset acute angle-closure glaucoma.* Nippon Ganka Gakkai Zasshi 2001, 105, 711-715.
7. Khan A.O.: *Buphthalmos in the setting of persistent hyperplastic primary vitreous cataract.* Am. J. Ophthalmol. 2003, 136, 945-947.

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