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Received: 2020.04.18 Accepted: 2020.06.16 Available online: 2020.07.21 Published: 2020.08.28		.18 .16 .21 .28	Iris-Claw Lens Implantation in a Patient with Iridoschisis	
Authors' Contribution: ABCDEF Study Design A ABCDEF Data Collection B ABCCE Statistical Analysis C ABCE Data Interpretation D BE Manuscript Preparation E BE Literature Search F DEG Funds Collection G DEG		ABCDEF ABCDEF ABCE BE BE DEG	Barbara Pieklarz Emil T. Grochowski Diana A. Dmuchowska Emil Saeed Patryk Sidorczuk Zofia Mariak	Department of Ophthalmology, University Teaching Hospital of Białystok, Białystok, Poland
Corresponding Author: Conflict of interest:		ng Author: of interest:	Barbara Pieklarz, e-mail: barbara.pieklarz@gmail.com None declared Male, 47-year-old Bilateral iridoschisis Visual loss — Pars plana vitrectomy • lensectomy • artificial lens implantation Ophthalmology	
	Patient: Final Diagnosis: Symptoms: Medication: Clinical Procedure: Specialty:			
Objective:		Objective:	Rare co-existance of disease or pathology	

propose an alternative approach to aphakia correction.

aphakia by implantation of scleral fixated lenses, rather than iris-claw lenses.

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Aphakia • Glaucoma • Hyphema • Iris Diseases • Uveitis, Anterior

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syndrome, have not been reported previously.



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Background:

Case Report:

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Iridoschisis is a rare condition defined as a separation of anterior iris stroma from the posterior stroma and muscle layers. The etiopathogenesis of iridoschisis is not fully understood. We report the case of uveitis-glaucoma-hyphema (UGH) syndrome related to the iris-claw lens implantation in a patient with iridoschisis, and

A 47-year-old male was referred to our department with bilateral iridoschisis, associated lens subluxation,

mature cataract, and secondary glaucoma. The patient underwent bilateral surgery, with entirely different anterior segment results depending on the method of artificial lens implantation. To the best of our knowledge, iris-claw implantation in iridoschisis and the potential association of iridoschisis with increased risk of UGH

Due to the possibly increased risk of UGH syndrome in patients with iridoschisis, one may consider treating

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Background

Iridoschisis is a rare condition defined as a separation of anterior iris stroma from the posterior stroma and muscle layers. The iris strands float in the aqueous humor [1]. The etiopathogenesis of iridoschisis is not fully understood. The age of onset is typically between 60 and 70 years. Iridoschisis is usually found in the inferior quadrants of the iris [2]. We report the case of a patient with iridoschisis who developed complications related to iris-claw lens implantation, and propose an alternative approach to this condition. To the best of our knowledge, neither the iris-claw implantation in iridoschisis nor the potential association of iridoschisis with increased risk of uveitis-glaucoma-hyphema (UGH) syndrome have been reported thus far.

Case Report

A 47-year-old Caucasian male was referred to the Department of Ophthalmology, Medical University of Bialystok, with a history

of significant bilateral visual loss in both eyes lasting for a few weeks. The patient had no previous history of an ocular disease but presented with the symptoms of moderate intellectual disability and according to his family, experienced headbanging episodes in the past. As the patient's cooperation was poor, it was difficult to assess the visual acuity accurately and to perform some other diagnostic tests. On ophthalmological examination, visual acuity was "hand motion" bilaterally. Intraocular pressures (IOP) in the right eye (RE) and left eye (LE) were 34 mmHg and 30 mmHg, respectively. Slit-lamp examination of both eyes revealed superotemporal iridoschisis (Figure 1A, 1B), shallowed anterior chambers, especially temporally, and iridodonesis. The pupils were round with normal light reactions. The presence of bilateral mature cataract hindered the visualization of the fundus. Bilateral temporal lens subluxation was visible on mydriasis (Figure 2A, 2B). Endothelial cell counts were 2412 cells/mm² and 3189 cells/mm², the axial length was 22.76 mm and 22.68 mm for the RE and LE, respectively. The keratometry readings equaled 7.92 and 7.77 mm for the RE, and 7.97 and 7.91 mm for the LE. Pre-operative gonioscopy, ultrabiomicroscopy or anterior segment optical coherence



Figure 1. (A, B) Preoperative right eye (RE): superotemporal iridoschisis - "shredded" appearance of the iris and mature cataract.



Figure 2. (A, B) Preoperative left eye (LE): mydriasis, mature cataract, temporal lens subluxation.



Figure 3. (A, B) Postoperative left eye (LE): superotemporal iridoschisis and round residual inflammatory deposits on the iris-claw lens surface.



Figure 4. (A) Postoperative left eye (LE): gonioscopic view of open angle, excess trabecular meshwork pigmentation. (B) Optic nerve head: advanced glaucomatous optic neuropathy, c/d=0.9.

tomography (OCT) were not performed. After the introduction of an antiglaucoma treatment, the IOP in the RE and LE dropped down to 16 mmHg and 26 mmHg, respectively.

The LE underwent pars plana vitrectomy and lensectomy. Posterior chamber iris-claw lens was implanted at 10 and 4 o'clock position to omit the iridoschisis. In our Department, an iris-claw lens is the first-line treatment for aphakia. The procedure to obtain the intraocular lens for scleral fixation takes about 4 weeks. In this case, we aimed to do surgery without delay. We preferred a combined surgery, as the patient was operated on under general anesthesia. This was due to the moderate intellectual disability and the resultant poor cooperation. On the first postoperative day, visual acuity was "counting fingers", and IOP equaled 13 mmHg. Anterior chamber depth was normal. Aqueous humor contained dispersed erythrocytes and the lens was stabilized well. The pupil appeared slightly irregular. The cup to disc (c/d) ratio was 0.9 with otherwise normal fundus. Five days after the procedure, visual acuity was 20/500, and IOP equaled 10 mmHg. A 3 mm hyphema and subtle fibrin deposits on the lens surface were found on slit-lamp examination (no photos available). These findings raised suspicion of UGH syndrome. Antiglaucoma therapy was discontinued.

Another 5 days later, dense round inflammatory deposits on the lens surface were observed on slit-lamp examination (no photos available). No evidence of hyphema was found, but IOP in the LE increased again, up to 29 mmHg, and hence, topical anti-glaucoma treatment was reintroduced. The final postoperative outcome was satisfactory (Figure 3A, 3B). No significant vision improvement in the LE was expected, given the presence of advanced glaucoma (Figure 4A, 4B).

Surgery on the RE was performed 1 month after the first procedure. Both pars plana vitrectomy with lensectomy and intrascleral sutureless intraocular lens fixation (Yamane technique) were uneventful [3]. Two weeks after the surgery, visual acuity was at least 20/200 (poor cooperation), with IOP of 17 mmHg,

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Figure 5. (A) Postoperative right eye (RE): superotemporal iridoschisis, quiet anterior chamber and intrascleral sutureless intraocular lens fixation. (B) Anterior segment optical coherence tomography of the right eye (RE) showing disorganization of the iris stroma – iridoschisis.

quiet anterior chamber and good lens stabilization found on slit-lamp examination (Figure 5A, 5B). Fundus examination of the RE showed c/d=0.6 and otherwise normal results. Further follow-up of both eyes confirmed that the results were stable. The macula OCT scan of the RE did not reveal any pathology. Subtle parafoveal intraretinal cystoid spaces were shown in the LE. Since the patient presented with physical features characteristic for Marfan syndrome, a genetic consultation was recommended. At the time of submission of this manuscript, the results were yet unknown.

Discussion

We report the case of a patient with bilateral iridoschisis, associated lens subluxation, mature cataract, and secondary glaucoma complicated by postoperative UGH syndrome in the LE.

Iridoschisis co-existing with presenile cataract has been well described in the literature [4]. However, we found only a few reports documenting rare co-existence of iridoschisis with lens (sub)luxation [5–7]. In our patient, lens subluxation might be either trauma-related (a consequence of head-banging episodes) or associated with Marfan syndrome. While we did not have access to the results of any previous ophthalmological examination, based on the information from the patient and his relatives, the deterioration of visual acuity progressed rapidly.

Glaucoma is found in more than two-thirds of patients with iridoschisis [1]. Severe optic disc damage found in our patient, especially in the LE, was suggestive of pre-existing glaucoma, which might be a consequence of iridoschisis or lens subluxation.

UGH syndrome can be triggered by any type of pseudophakic lenses – mainly rigid anterior chamber lenses, but also posterior-chamber and iris-supported ones [8]. The syndrome may develop immediately after cataract surgery or within several years, and may be recurrent [9]. We assumed that in our case iridoschisis might have further increased the risk of UGH syndrome, in addition to the iris-claw lens. Should UGH syndrome relapse, one may consider the iris-claw lens removal and scleral fixated lens implantation.

In this patient's case, while mild anterior uveitis preceded hyphema development, the full-blown inflammation, another component of the triad, developed a few days later, in conjunction with elevated IOP. It is unclear if the elevated IOP was a consequence of the syndrome or pre-existing glaucoma. Hyphema could be due to various causes, including blunt trauma or surgery itself. Taking into account the patient history and clinical characteristics, the diagnosis of UGH syndrome seemed most probable.

Duchêne et al. [10] analyzed retrospectively the risk factors for UGH, among these risks was the type of posterior chamber lens. According to these authors, UGH was present in 50% of the eyes with iris-sutured lenses and only 13% of the eyes with scleral fixated lenses [10]. Our observations are consistent with those findings, as the anterior segment results in our patient differed depending on the method of artificial lens implantation. Various approaches can be used to deal with aphakia. In the case of the RE of our patient, intraocular lens fixation with Yamane technique was not associated with either intraor postoperative complications. Our study had a limitation of a relatively short follow-up of 3 months. This was due to the change in the place of residence of the patient. We assume that other scleral fixation procedures could also be applicable in similar cases. None of the previously published reports documented iris-claw lens or scleral fixated lens implantation for aphakia correction in iridoschisis. It needs to be stressed that patients with iridoschisis require special attention.

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Conclusions

Due to the possibly increased risk of UGH syndrome in patients with iridoschisis, one may consider treating aphakia by implantation of scleral fixated lenses, rather than iris-claw lenses.

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Conflict of interest

None.

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