Int Ophthalmol (2019) 39:1607–1612
https://doi.org/10.1007/s10792-018-0972-5

CASE REPORT

Novel microsurgical management of uveitis-glaucoma-hyphema syndrome

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Received: 15 February 2018 / Accepted: 16 June 2018 / Published online: 26 June 2018

Abstract
Purpose To report a series of cases and microsurgical management of rarely occurring uveitis-glaucoma-hyphema (UGH) syndrome in patients with single-piece acrylic intraocular lens (IOL) placed in the capsular bag.
Methods It was a series of patients with UGH syndrome induced by posterior chamber IOL/capsular bag complex instability (pseudophakodonesis), who underwent IOL fixation to the iris. Visual acuity, intraocular pressure, number of glaucoma medications and IOL status (position) were recorded by the same protocol before and 6 months after the surgical treatment.
Results The case series presents three patients with UGH syndrome caused by single-piece acrylic IOL placed in-the-bag. Each patient had uneventful phacoemulsification with posterior chamber IOL implantation few years ago and pseudophakodonesis caused by weakened zonules from pseudoexfoliation with subsequent development of UGH syndrome. IOL fixation to the iris with satisfactory postoperative results was performed due to the development of UGH. Signs of syndrome did not recur 6 months after the operation.
Conclusion UGH syndrome can be induced by unstable in-the-bag IOL due to zonular laxity. Depending on the severity of the syndrome, this condition can be fought by applying a minimally invasive approach—IOL suturing to the iris with direct observation under the surgical microscope precisely in the anticipated location with no or minimal pupil deformation. Symptoms of UGH did not recur due to increased stability of the IOL and, as a result of this, declined irritation of the uveal tissue.

Keywords Uveitis-glaucoma-hyphema syndrome · Acrylic single-piece intraocular lens · Pseudophakodonesis · Intraocular lens fixation to the iris

Poster: The XXXV Congress of the European Society of Cataract and Refractive Surgeons, October 2017, Lisbon, Portugal.

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Introduction

Uveitis-glaucoma-hyphema (UGH) syndrome is a rare, but potentially severe cataract surgery complication [1], first reported in 1978 by Ellingson and known as chafing of anterior segment structures—

mechanical iris trauma—by first-generation anterior chamber intraocular lenses (IOLs) [1, 2]. Although UGH syndrome is classically associated with anterior chamber IOL, reported cases of decentred or dislocated posterior chamber IOL/capsular bag complex have also been described as possible triggers [1, 3].

The incidence of this syndrome has declined from a mean of 2.2–3 to 0.4–1.2% thanks to improved IOL design, surgical techniques and increased usage of posterior chamber IOL [4]. A more recent study revealed that UGH syndrome was one of the most frequent indications for IOL exchange (11.9% from 109 cases over a period of 5 years) [5]. However, this complication is still relevant nowadays due to a growing number of people with a pseudophakic eye.

A typical cause is weakened zonules [often from pseudoexfoliation (PXF)] that allow pseudophakodonesis, which can irritate uveal tissue and cause development of UGH syndrome. Treatment of this condition depends on the severity of UGH syndrome and involves a conservative (anti-inflammatory and IOP lowering drugs) or a surgical approach.

In cases when UGH syndrome is strongly expressed, it may make sense to have a vitreoretinal surgery to remove the IOL. However, if the syndrome is mild and IOL is slightly decentred or pseudophakodonesis is observed, then an anterior approach is often successful.

We reported a series of three cases of UGH syndrome in patients with single-piece acrylic IOL placed in the capsular bag with pseudophakodonesis who did not respond sufficiently to conservative treatment and therefore have undergone a minimally invasive surgery (IOL suturing to the iris) to fix the problem.

Materials and methods

All procedures involving human participants were carried out according to the Declaration of Helsinki, and the study protocol was approved by the Lithuanian University of Health Sciences Review Board. Written informed consent was obtained from all the patients.

It was a series of patients with UGH syndrome induced by posterior chamber IOL/capsular bag complex instability (pseudophakodonesis), who underwent IOL fixation to the iris, performed by one surgeon (V.J.) in 2017. Patients were examined by the same protocol before and 6 months after the surgical treatment. Demographic information (age and gender), data of original cataract surgery, other previous operations or laser procedures, ocular comorbidity, visual acuity, intraocular pressure (IOP), number of glaucoma medications and IOL status (position) were recorded.

Surgical technique

Two stab incisions in the clear cornea are performed along the anticipated path of the needle's entry and exit. 10-0 Polypropylene suture with a long curved needle (PC 10, ALCON) is used. The needle is passed through the first stab incision (Fig. 1a-1) and the pupil beneath the haptics and guided into the anterior chamber through the iris in a matched place for IOL fixation. IOL/capsular bag complex can be temporarily stabilized by planting a spatula behind it to facilitate puncture of a fibrosed capsular bag and to prevent subsequent damaging residual Zinn's zonules. A 27-gauge cannula is introduced through the distal paracentesis (Fig. 1a-2), and the needle is docked to facilitate exit. The needle is passed back into the eye through the same stab incision, iris (close to the first perforation), above IOL haptics, through the pupil into the anterior chamber and out of the eye through the first stab incision (Fig. 1b, c). At this moment, IOL haptic is encircled by the suture. With a microhook (Fig. 1d-3) which is introduced through the second stab incision, both sutures are hooked and pulled out of the eye. This is performed in the following manner: one suture (short) (Fig. 1e-4) is pulled out of the eye completely, while the second (long) one (Fig. 1e-5) is pulled out just to form a loop for making a throw. The first double throw is performed by wrapping a short suture end around the medial part of the loop (Fig. 1f). Both ends of the thread are pulled outside the eye, while the knot is formed inside the eye and beneath the iris (Fig. 1g). The long suture is repeatedly drawn through the second stab incision with a microhook to form a loop (Fig. 1h, i) and second double throw is performed (Fig. 1j). The third single throw is performed in the same manner. The ends of the suture are pulled away, bringing the third throw snugly into position without twisting (Fig. 1k). The ends of the suture are cut-off with vitrectomy scissors or knife

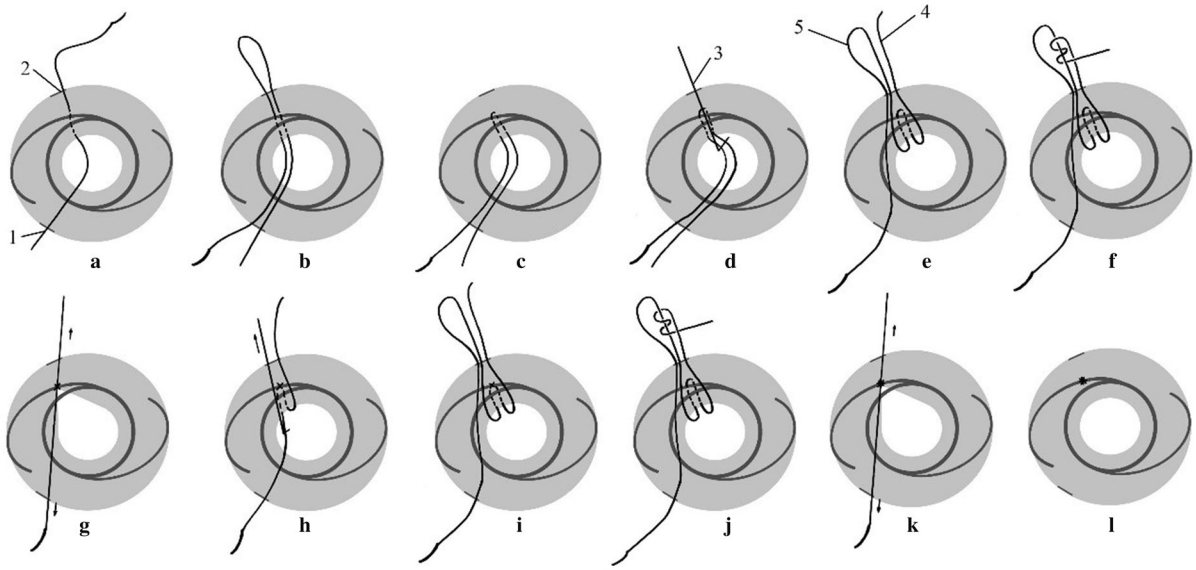

Fig. 1 Suture fixation of the haptics to the iris. **a** The needle is passed through the first stab incision and the pupil beneath the haptics and guided into anterior chamber through the iris in a matched place for IOL fixation. Then the needle is pulled out through the distal paracentesis. **b**, **c** The needle is passed back into the eye through the same stab incision, iris (close to the first perforation), above IOL haptics, through the pupil into anterior chamber and out of the eye through the first stab incision. **d** With a microhook which is introduced through second stab incision both sutures are hooked and pulled out of the eye. **e** One suture (short) is pulled out of the eye completely, while the second (long) one is pulled out just to form a loop for making a throw. **f** First double throw is performed by wrapping short suture end around medial part of the loop. **g** Both ends of thread are pulled outside the eye, while the knot is formed inside the eye and beneath the iris. **h–j** The long suture is repeatedly drawn through second stab incision with a microhook to form a loop and second double throw is performed. **k** The ends of suture are pulled away, bringing the third throw snugly into position without twisting. **l** The ends of suture are cut-off with Ong scissors or knife inside the eye, and the knot is buried behind the iris

inside the eye and the knot is buried behind the iris. The pupil remains circular after the procedure.

Results

Case 1

A 72-year-old man was treated for blurring vision and ocular pain in the left eye. Such short-term episodes recurred six times in 8 months. There was no trauma history. Patient had uneventful phacoemulsification with posterior chamber IOL (Restor) implantation under topical anaesthesia in the left eye in 2008. During the examinations throughout the UGH syndrome episodes, his uncorrected visual acuity fluctuated from hand motion to 0.1. Intraocular pressure was observed from 20 to 40 mmHg with medication.

Anterior segment examination of the left eye revealed microhyphema, pseudoexfoliation on iris papillary border, pseudophakodonesis, IOL optics well covered by the capsulorrhexis margin and transillumination defect on the iris temporal side (Fig. 2a).

Despite conservative treatment, UGH syndrome episodes recurred. Therefore, it was decided to perform fixation of IOL to the iris.

Six months after the surgery, symptoms of UGH syndrome did not recur, best corrected visual acuity at the last examination was 1.0, and intraocular pressure was 12 mmHg without antiglaucoma treatment. Anterior segment examination of the left eye revealed stable, slightly decentred posterior chamber IOL with temporal iris transillumination and small indentation of the iris at the site of fixation (Fig. 2a).

Case 2

A 67-year-old man was treated for UGH syndrome of the left eye. There was no trauma history. He had

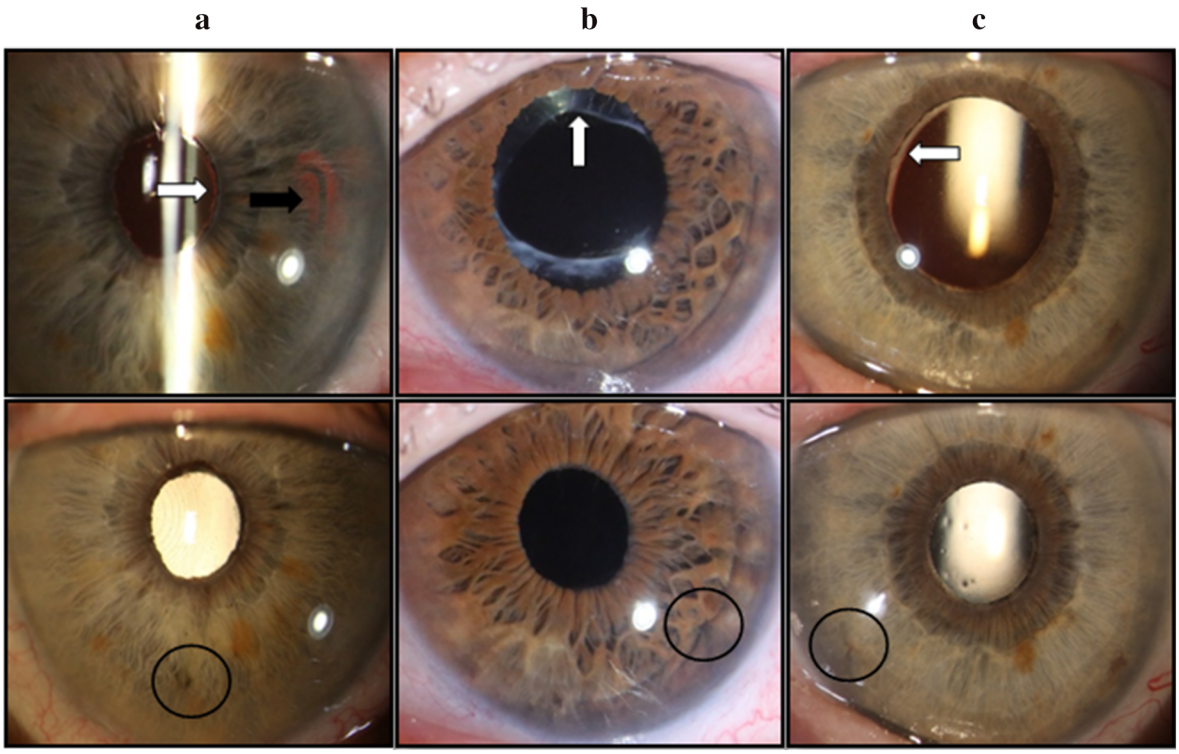

Fig. 2 Patients' eyes before (at the top) and after (at the bottom) intraocular lens fixation to the iris. Anterior segment photography demonstrates capsulorrhexis margin (white arrow), transillumination defect of the iris (black arrow) and indentation of the iris in the site of fixation (black circle)

undergone uneventful phacoemulsification with posterior chamber IOL (AcrySof Natural) implantation under topical anaesthesia in the left eye in 2013. During the examinations throughout the UGH syndrome episode, his uncorrected visual acuity was hand motion, while intraocular pressure was 40 mmHg with medication.

Anterior segment examination of the left eye revealed hyphema and pseudophakodonesis of the posterior chamber IOL placed in the capsular bag and well covered by the capsulorrhexis margin (Fig. 2b).

Hyphema disappeared after the conservative treatment. However, due to the expressed pseudophakodonesis, it was decided to perform fixation of IOL to the iris.

Six months after the surgery, symptoms of UGH syndrome did not recur, best corrected visual acuity at the last examination was 1.0, and intraocular pressure was 18 mmHg without antiglaucoma treatment. Anterior segment examination of the left eye revealed stable perfect centred posterior chamber IOL with small indentation of the iris at the site of fixation (Fig. 2b).

Case 3

A 72-year-old woman treated for UGH syndrome of the right eye. Short-term UGH episodes were assessed three times during 1 year. There was no trauma history. She had undergone uneventful phacoemulsification with posterior chamber IOL (AcrySof Natural) and capsule tension ring implantation under topical anaesthesia in the right eye at 2013. During the examinations throughout the episode, her uncorrected visual acuity fluctuated from 0.1 to 0.3. Intraocular pressure was 30 mmHg with antiglaucoma treatment.

Anterior segment examination of the left eye revealed microhyphema, pseudophakodonesis, posterior chamber IOL well covered by the capsulorrhexis margin (Fig. 2c).

Despite conservative treatment, UGH syndrome episodes recurred and a decision was made to suture IOL to the iris.

Six months after the surgery, symptoms of UGH syndrome did not recur, best corrected visual acuity at the last examination was 0.8, and intraocular pressure was 17 mmHg with antiglaucoma treatment (beta blockers two times per day). Anterior segment examination of the right eye revealed stable, perfectly centred posterior chamber IOL with small indentation of the iris at the site of fixation (Fig. 2c).

Discussion

Although single-piece acrylic IOL placed in-the-bag rarely cause UGH, we report a series of three similar cases. For these patients, IOL fixation to the iris with satisfactory postoperative results was performed. UGH syndrome did not recur 6 months after the surgery. We assume that it was achieved due to the increased stability of the IOL and the resulting disappearance of the irritation of the uveal tissue. However, before considering a treatment method, it is important to clarify the causes of UGH syndrome.

The main cause of UGH syndrome is mechanical chafing of uveal structures with IOL or artificial iris implants [6], leading to the breakdown of the blood-aqueous barrier and empowers cytokines to trigger an inflammatory cascade, causing a chronic inflammation as well as recurrent hyphema or microhyphemas and glaucoma [1]. Moreover, UGH syndrome is now known to be caused by the placement of a single-piece IOL in the sulcus. Single-piece IOL haptics is thick and massive, and the optic edges are square and abrasive, which allows contact and chafing with the posterior iris surface [1, 7]. In such cases, the best treatment option would be to remove or exchange the IOL.

Different causes with in-the-bag placement of a single-piece IOL linked to UGH syndrome development include: chafing of the iris by wrong position of IOL optic or haptic (e.g. IOL bending during loading) [8], iridociliary cyst [9], displacement of an IOL haptic caused by capsular bag contraction [10] or IOL-capsular bag complex dislocation in case of PXF syndrome [3]; chafing of the iris in relation to capsular bag fibrosis (especially combined with plateau iris configuration) [11] or formation of a Soemmering ring [1]. An increasing cause of UGH is pseudophakodonesis from zonular laxity caused by PXF.

In such cases, various treatment options are available. Explantation or exchange of the implanted IOL has been the traditional surgical approach to UGH syndrome treatment [12]. However, the harm–benefit ratio and risk of potential complications of new surgery should be evaluated [13]. In cases where surgical treatment has to be postponed or cannot be performed, conservative first-line medical treatment methods include: topical corticosteroids to control anterior inflammation for uveitis; IOP lowering agents, such as beta-adrenergic antagonists, alpha-adrenergic agonists and carbonic anhydrase inhibitors for glaucoma medical management; cycloplegics and methods of head elevation or limited physical activity for hyphema [14].

As the studies suggest, there are more alternative methods to managing UGH syndrome other than IOL explantation, such as cyclophotocoagulation of the involved ciliary process or placement of a capsular tension ring to redistribute zonular tension [11]. Walland described in a case report that local laser iridoplasty was an option for the treatment of UGH syndrome, wherein solid-state (‘argon’) laser in two wedge patterns was applied to the iris on either side of haptic contact to avoid removal of the IOL. In that case, the surgical intervention was an attractive option, given excellent visual acuity and risks of surgical complications [15]. In another case report, Rech et al. used serial intracameral injections of anti-vascular endothelial growth factor agent bevacizumab to reduce inflammation and neovascularization to control UGH syndrome, because, despite its potential side effects, bevacizumab may benefit selected patients and offer a temporizing or long-term option to high-risk IOL manipulation cases [16].

We present the minimally invasive surgical treatment of UGH syndrome by fixing the IOL to the iris. This technique has advantages, such as IOL haptics (with surrounding capsule) being sutured to the iris under direct observation under the microscope, precisely in the anticipated location with no or minimal pupil deformation (Fig. 2) with the knot tied behind the iris. Fixation of dislocated IOL to the iris generally is performed in a ‘blind’ manner (when the needle is behind the iris). This may cause an improper localization of the haptic and suture to the iris in the unintended place. Furthermore, it is challenging to

penetrate the iris from behind close to the first prick. Also, the presence of fibrosed capsular bag makes the procedure more risky due to IOL luxation into the vitreous. The described surgical technique of additional stabilization of capsular bag/IOL complex with an instrument (spatula) in a surgery prevents damage to the remaining Zinn's zonules.

In conclusion, UGH syndrome can be induced by unstable in-the-bag IOL due to zonular laxity. Depending on the severity of the syndrome, this condition can be fought by applying minimally invasive approach—IOL suturing to the iris with direct observation under a surgical microscope, precisely in the anticipated location with no or minimal pupil deformation. Symptoms of UGH did not recur thanks to increased stability of the IOL and, as a result, declined irritation of the uveal tissue in our series.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent Informed consent and consent form for case reports were obtained from all individual participants.

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