DOI: http://doi.org/10.4038/jcosl.v27i2.44

Review

Uveitis-Glaucoma-Hyphaema Syndrome

Nuwan Niyadurupola[1]

The Journal of the College of Ophthalmologists of Sri Lanka 2021; 27: 33-35

Uveitis-Glaucoma-Hyphaema (UGH) syndrome was first described in 1978 by Ellingsen[1]. Ellingson noticed that some anterior chamber intraocular lenses (IOLs), particularly those with a small size and warped footplates, had excessive IOL movement within the eye which caused mechanical irritation of angle structures and the iris. The mechanical trauma of uveal tissue, particularly the iris, ciliary body and angle structures, caused chronic inflammation, elevated intraocular pressure (IOP) and hyphaema; hence giving the name UGH syndrome. Although UGH syndrome was initially described and most commonly seen with anterior chamber IOLs, it can occur with any other types of IOLs. The UGH syndrome has been described in iris-plane IOLs, sulcus-placed IOLs (particularly single-piece IOLs that are not designed to be placed in the sulcus), IOLs that have been placed within the bag but have haptics popping out of the bag lying in the ciliary sulcus or IOLs placed within an unstable bag (such that the IOL-bag complex can move against the iris structures), and with scleral-fixated IOLs.

Vaulting, decentration or excessive movement of IOLs causes chafing of the delicate uveal tissue. Breakdown of the blood-aqueous barrier causes anterior uveitis and liberation of red blood cells from the uveal tissue leading to microhyphaema or hyphaema. Pigment and red blood cells released from uveal tissue can find their way into the trabecular meshwork and obstruct trabecular meshwork outflow. Macrophages and trabecular meshwork cells phagocytose the pigment and red blood cells. Swelling of the trabecular meshwork cells with the phagocytosed debris reduces the pore size within the trabecular meshwork, thereby increasing resistance to outflow. In addition, direct damage from the IOL haptic against the angle structures can cause scarring of the trabecular meshwork outflow pathway and thus the IOP can increase.

Patients often report intermittent blurred vision and they can also describe photophobia, redness and pain. The ocular discomfort is often out of proportion to the signs that are seen when the eye is examined. Anterior chamber cells or flare are often present, there may be a microhyphaema or hyphaema and the patient usually has an elevated IOP. Not all three signs are always evident and a microhyphaema may be difficult to spot in the presence of anterior chamber cells. If the condition is left for a long period of time iris neovascularisation can develop. Iris transillumination defects are quite characteristic and can actually point to the cause of the condition, for example, iris transillumination defects in the pattern of the IOL haptic can suggest that the IOL haptic is not located fully in the bag and is likely to be located in the ciliary sulcus by mistake. Friction of the IOL against the iris can cause pigment dispersion and signs similar to pigment dispersion syndrome, such as, pigment on the endothelial surface of the cornea and pigment dispersed on the anterior surface of the iris stroma, and in the trabecular meshwork. Where a haptic is impinging on the iris, the pupil can become distorted and peaked in the direction of the impingement. Intraocular lens-iris contact, a dislocated or malpositioned IOL, or a misplaced haptic may be evident on examination of the eye. Cystoid macular oedema can develop after prolonged inflammation. Although most patients have an elevated IOP, not all cases develop glaucomatous optic neuropathy. This is the problem that I have with the term UGH syndrome because most patients just have elevated IOP and not glaucoma. A more accurate term should be 'Uveitis-Hypertensive-Hyphaema Syndrome'. However, if this condition is left untreated for a long period of time or if there is damage to the angle structures then glaucomatous optic neuropathy can develop. Vitreous haemorrhage may be evident where there is a communication between anterior and posterior segments of the eye, perhaps through a ruptured posterior capsule (termed Uveitis-Glaucoma-Hyphaema Syndrome Plus). During examination, gonioscopy is helpful since blood

[1]*Consultant Ophthalmic Surgeon, Norfolk and Norwich University Hospital, United Kingdom, Lead for Ophthalmology and Honorary Senior Lecturer, Norwich Medical School, United Kingdom, Regional Glaucoma Lead for Ophthalmic Practitioner Training, Health Education England, United Kingdom.*

may be found in the drainage angle where hyphaema or microhyphaema is not easily detected. Dispersion of iris pigment may cause increased pigmentation of the angle and evidence of a haptic in the angle may be seen with gonioscopy.

Ultrasound biomicroscopy is the best investigation in cases of UGH since a malpositioned IOL can be visualised, the location of haptics can be checked and it can be determined whether there is any IOL/haptic contact with any uveal tissue. Anterior segment ocular coherence tomography can also be used, but it has the disadvantage that it cannot image behind the iris.

Management of UGH syndrome includes control of the inflammation with topical steroids and control of IOP with topical antiglaucoma medication (pilocarpine should be avoided due to miosis and possible increased risk of chafing). The hyphaema should be managed by limiting activity, keeping the head elevated and use of cycloplegics. However, this management only controls the symptoms and signs for a period of time and one must deal with the cause of the UGH condition: the IOL itself. Surgery needs to be performed to reposition the IOL, explant the IOL and leave the patient aphakic, or explant the IOL and replace it with another IOL. At Norfolk and Norwich University Hospital we favour explanting the offending IOL and using a scleral-fixated IOL to treat UGH syndrome.

In terms of preventing UGH syndrome, one must use the correct IOL for the intended position of placement of the IOL and the IOL needs to be placed correctly. A single-piece IOL can be placed in the bag but one must ensure the haptics are located entirely within the capsular bag. If the capsular bag is unstable, a capsule tension ring should be used to stabilise the IOL and the bag within the eye. If there is a posterior capsule rupture and enough anterior capsule support, then placement of the IOL in the sulcus is possible. A single-piece IOL should never be placed within the sulcus since they are too small and not designed for sulcus placement. A three-piece IOL should be used for sulcus placement. Three-piece IOLs are vaulted and need to be correctly orientated such that the optic lies posterior to the location of the haptics. The three-piece IOL for sulcus placement needs to have a haptic-haptic length of 13mm or more and a large optic diameter of at least 6mm. A sulcus positioned three-piece IOL can be stabilised within the bag by performing optic capture if there is sufficient support from the anterior capsulorrhexis. With optic capture, the haptics of the IOL are located within the ciliary sulcus, but the optic has been placed in the bag through the anterior capsulorrhexis edge. If there isn't sufficient capsular support for placement of a sulcus located IOL, then an anterior chamber IOL can be considered. The correct size of anterior chamber IOL is very important since an incorrectly sized anterior chamber IOL can move around too much within the eye causing the UGH syndrome. The traditional method to measure the correct size of an anterior chamber IOL is to measure the horizontal white-to-white of the eye and add one millimetre. Being a glaucoma surgeon, I personally don't like IOLs in the anterior chamber due to the risk of causing damage to the angular structures and increased risk of corneal decompensation. In the situation of there being insufficient anterior capsular support to place an IOL in the ciliary sulcus, a scleral-fixated IOL is often a better approach to reduce the risk of subsequent UGH syndrome and reduce the risk of damage to angular structures and corneal decompensation.

In summary, cataract surgery is generally beneficial to glaucoma patients. Large scale studies have shown that cataract surgery can help lower IOP in open angle glaucoma[2] and in primary angle closure and primary angle closure glaucoma, cataract surgery is generally beneficial compared to the conventional treatment of peripheral iridotomy[3]. Cataract surgery can also be combined with minimally invasive glaucoma surgeries. However, cataract surgery can also cause problems in glaucoma patients. We have previously shown that 39% of patients who had functioning trabeculectomies had failure of their trabeculectomy following cataract surgery despite intensive management of the trabeculectomy bleb[4]. Normal uncomplicated cataract surgery can cause IOP spikes and this can be dangerous for patients who have severe glaucoma with fixation threatening visual field defects. Complications during cataract surgery, such as iris trauma, which can liberate pigment, or posterior capsule rupture with vitreous loss or dropped nucleus, can lead to elevated IOP. Incorrect IOL placement or an unstable IOL can cause UGH syndrome. It is important that UGH syndrome is prevented by choosing the correct lens for the intended placement of the IOL. If sulcus placement of an IOL is needed, ensure that it is a three-piece IOL and not a single-piece IOL and if possible perform optic capture to stabilise the IOL. If UGH syndrome develops, then the symptoms and signs need to be treated but eventually the IOL will need to be repositioned or explanted and exchanged for another IOL.

References

1. Ellingson FT. The uveitis-glaucoma-hyphema syndrome associated with the Mark VIII anterior chamber lens implant. *Am Intraocular Implant Soc J*. 1978; **4**: 50-3.
2. Poley BJ, Lindstrom RL, Samuelson TW et al. Intraocular pressure reduction after phacoemulsification with intraocular lens implantation in glaucomatous and nonglaucomatous eyes: evaluation of a causal relationship between the natural lens and open-angle glaucoma. *J Cat Refract Surg*. 2009; **35**: 1946-55.
3. Azuara-Blanco A, Burr J, Ramsay C et al. Effectiveness of early lens extraction for the treatment of primary angle-closure glaucoma (EAGLE): a randomised controlled trial. *Lancet* 2016; **388**: 1389-97.
4. Nguyen DQ, Niyadurupola N, Tapp RJ et al. Effect of phacoemulsification on trabeculectomy function. *Clin Exp Ophthalmol*. 2014; **42**: 433-9.