# Decision-making and management of uveitic cataract

### Nicole Shu-Wen Chan, Seng-Ei Ti, Soon-Phaik Chee

The visual outcome of uveitic cataract surgery depends on the underlying uveitic diagnosis, the presence of vision-limiting pathology and perioperative optimization of disease control. A comprehensive preoperative ophthalmic assessment for the presence of concomitant ocular pathology, with particular emphasis on macula and optic nerve involvement, is essential to determine which patients will benefit from improved vision after cataract surgery. Meticulous examination in conjunction with adjunct investigations can help in preoperative surgical planning and in determining the need for combined or staged procedures. The eye should be quiescent for a minimum of 3 months before cataract surgery. Perioperative corticosteroid prophylaxis is important to reduce the risk of cystoid macular edema and recurrence of the uveitis. Antimicrobial prophylaxis may also reduce the risk of reactivation in eyes with infectious uveitis. Uveitic cataracts may be surgically demanding due to the presence of synechiae, membranes, and pupil abnormalities that limit access to the cataract. This can be overcome by manual stretching, multiple sphincterotomies or mechanical dilation with pupil dilation devices. In patients <2 years of age and in eyes where the inflammation is poorly controlled, intraocular lens implantation should be deferred. Intensive local and/or oral steroid prophylaxis should be given postoperatively if indicated. Patients must be monitored closely for disease recurrence, excessive inflammation, raised intraocular pressure, hypotony, and other complications. Complications must be treated aggressively to improve visual rehabilitation. With proper patient selection, improved surgical techniques and optimization of peri- and post-operative care, patients with uveitic cataracts can achieve good visual outcomes.



Key words: Cataract surgery, steroid prophylaxis, synechiolysis, uveitic cataract, uveitis

Cataract is a common complication of chronic or recurrent uveitis and is a sequelae to chronic intraocular inflammation and chronic systemic and/or topical corticosteroid therapy.<sup>[1,2]</sup> It accounts for up to 40% of the visual loss in patients with uveitis<sup>[3]</sup> and is the most common indication for surgical intervention in uveitis.<sup>[4]</sup>

Uveitic cataracts comprise approximately 1.2% of all cataract surgeries and are more surgically demanding<sup>[5]</sup> with far less predictable postoperative outcomes. This is largely due to the inflammatory sequelae, structural abnormalities, and higher rates of copathology, many of which limit visual outcome.

Before the introduction of corticosteroids, cataract surgery in eyes with uncontrolled uveitis had a high incidence of severe complications<sup>[6-8]</sup> resulting in markedly worse vision or even loss of the eye.<sup>[4]</sup> In recent years, proper patient selection, availability of biocompatible intraocular lenses (IOLs) and improved surgical techniques have considerably decreased the incidence of complications in uveitic cataract extraction.<sup>[6-10]</sup> Together with improved perioperative control of inflammation and anticipatory management of postoperative complications, this has improved visual outcomes, with 59.3% to 82% of eyes achieving postoperative best corrected visual acuity (BCVA) of 20/40 or better.<sup>[5,11-14]</sup>

The visual outcome depends on the type of uveitis. Eyes with anterior uveitis have the best outcome, with

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91.1% to 94.5% achieving BCVA of 20/40 or better.<sup>[11,15]</sup> Eyes with posterior or panuveitis (57.4%),<sup>[11]</sup> Vogt–Koyanagi– Harada (VKH) disease (67.1%–68%),<sup>[11,16]</sup> Behcet's disease (12.2%–16.7%),<sup>[11,17]</sup> and sympathetic ophthalmia have substantially worse visual outcomes,<sup>[18]</sup> likely due to concomitant chorioretinal injury.

Although postoperative BCVA in uveitic eyes is between one and two lines worse than in nonuveitic eyes, cataract surgery nevertheless offers a high degree of visual improvement proportionate to the preoperative visual acuity as in nonuveitic eyes. The worse preoperative VA in uveitic eyes likely represents a more advanced cataract with higher rates of visually significant copathology.<sup>[5]</sup>

# Visual Outcomes of Cataract Surgery in Different Types of Uveitis

The uveitis diagnosis provides an indication as to the medical and surgical treatment response, the incidence and severity of postoperative complications and the prognosis.

#### Fuchs uveitis syndrome

Blurring of vision secondary to cataract is one of the most common presentations of Fuchs uveitis

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syndrome (FUS).<sup>[19,20]</sup> The average incidence of cataract in FUS is around 50% (15%–75%).<sup>[4,6,19-22]</sup>

Eyes with FUS have better visual prognosis compared to other types of uveitis, with 92% achieving BCVA of 20/40 or better after cataract surgery if the inflammation is well controlled.<sup>[18]</sup> This is comparable to that for nonuveitic cataracts (96%).<sup>[23]</sup> Vitreous haze was the major cause of postoperative BCVA of <20/20.<sup>[19]</sup>

The good visual outcomes in FUS can be attributed to the low-grade inflammation that predominantly affects the anterior segment. As posterior synechiae and posterior segment complications such as macular edema are uncommon, there is less preexisting vision-limiting pathology and/or postoperative inflammation compared to other types of uveitis.<sup>[18]</sup>

The incidence of postoperative complications is low,<sup>[24]</sup> although the risk of postoperative inflammation is higher in FUS eyes with glaucoma, severe iris atrophy and severe abnormalities in iris vasculature.<sup>[25]</sup> Common complications, in order of frequency, are hyphema, progressive vitreous opacification, glaucoma, and spontaneously resolving vitreous hemorrhage. Glaucoma is the most visually significant complication, where permanent elevation of IOP develops in 3%–35%. Although control of IOP may initially be achieved by medication alone, up to 70% of these patients will eventually require filtering surgery.<sup>[22]</sup> Less commonly reported complications include retinal detachment, extensive synechiae formation, and corneal edema.<sup>[19-21]</sup>

#### Juvenile idiopathic arthritis-associated uveitis

Juvenile idiopathic arthritis (JIA)-associated uveitis presents in younger patients. Unlike in FUS, the cataract extraction is associated with more severe complications and persistent intraocular inflammation.<sup>[26]</sup>

As eyes with JIA-associated uveitis have a marked tendency to form synechiae,<sup>[27]</sup> implantation of a posterior chamber IOL at the time of cataract extraction often results in posterior synechiae reforming if the uveitis is not adequately controlled. The development of membranes over the IOL leads to cocooning of the IOL,<sup>[26,28]</sup> pupil distortion, and IOL displacement.<sup>[27,29]</sup> Seclusio or occlusio pupillae may cause malignant glaucoma, in which a surgical peripheral iridectomy and capsulectomy are needed to relieve the pupil block.<sup>[28]</sup> As the mere presence of an IOL may cause persistent postoperative inflammation and poorer vision requiring subsequent IOL explantation,<sup>[30]</sup> it has been recommended that IOLs be avoided in patients with poorly controlled JIA-associated uveitis.<sup>[27,29]</sup>

Although glaucoma occurs in about 25% of JIA-associated uveitis, some eyes develop hypotony due to the traction of the ciliary body and its processes by cyclitic membranes.<sup>[27,31]</sup> In addition to intraoperative technical difficulties, hypotony is associated with an increased risk of postoperative choroidal effusion, macular edema, and phthisis bulbi.

Even with successful cataract surgery, the outcome of cataract surgery in JIA patients is often limited by optic nerve and macula involvement and may be compromised by the presence of band keratopathy.<sup>[27,28]</sup>

Only 22% of eyes with active JIA-associated uveitis achieve BCVA of 20/40 or better after cataract surgery.<sup>[18]</sup>

However, since the introduction of the concept of adequate immunosuppression aimed at zero tolerance of inflammation, the outcomes of cataract surgery in JIA-associated uveitis have improved.<sup>[27,32]</sup> When the uveitis is controlled preoperatively, visual outcomes improve significantly to 71% of eyes attaining BCVA of 20/40 or better.<sup>[18]</sup> It is therefore recommended that the anterior chamber (AC) is free of inflammatory cells for at least 3 months before cataract surgery<sup>[33]</sup> and that these eyes be treated with perioperative steroids.<sup>[27]</sup>

Several studies have reported successful postoperative outcomes with limited complications with peri- and post-operative immunosuppression.<sup>[27]</sup> Grajewski *et al.*<sup>[34]</sup> recommended a perioperative intravitreal injection of triamcinolone acetonide (TA) in addition to a preoperative well-controlled uveitis for good visual outcomes in JIA-associated uveitis. Children with recalcitrant uveitis uncontrolled on maximum medical therapy also benefit from the use of intravitreal steroid implants such as Retisert (fluocinolone acetonide)<sup>[35]</sup> and Ozurdex (dexamethasone).<sup>[36]</sup>

As most JIA patients are in the amblyogenic age when they develop cataracts, achieving quiescence before cataract extraction should be balanced against the benefits of early surgery.

#### **Behcet's disease**

Cataract occurs in 17%–36% of ocular Behcet's disease.<sup>[2,37]</sup> Postoperative visual acuity depends on the preoperative status of the posterior segment. The visual prognosis is significantly worse in eyes with Behcet's disease<sup>[38]</sup> compared to other types of uveitis also due to severe posterior segment complications, mainly optic atrophy, inflammatory macular degeneration, and macular edema.<sup>[38-41]</sup>

A key element to successful outcomes is control of inflammation. Kadayifçilar *et al.* showed that visual acuity after cataract surgery improved to 0.1 or better in 75.8% and 0.5 or better in 42.4%<sup>[38]</sup> if ocular inflammation was suppressed for at least 3 months preoperatively in ocular Behcet's disease. Other authors have suggested that eyes should be quiescent for a minimum of 6 months before cataract surgery to reduce the risk of postoperative attacks.<sup>[42,43]</sup>

The most frequent complication is posterior capsular opacification (PCO) (37.5%). Other complications include severe inflammation leading to posterior synechiae, and posterior segment complications such as epiretinal membrane formation, cystoid macular edema (CME), and optic atrophy.<sup>[44]</sup>

#### Vogt–Koyanagi–Harada syndrome

Cataract is the most common complication of VKH syndrome with a prevalence of about  $40\%^{[45]}$  and accounts for 25% of the visual loss.<sup>[46]</sup>

Patients with VKH syndrome achieve good visual outcomes with cataract surgery. In a series of 50 eyes by Quek *et al.*, 82% improved by 2 or more Snellen lines, with 68% achieving a BCVA of 6/12.<sup>[16]</sup> Ganesh *et al.*<sup>[47]</sup> similarly reported that the visual acuity 80% of eyes with VKH syndrome improved by 1 or more Snellen lines. The median VA in Moorthy *et al.*'s study significantly improved from 6/120 to 6/12.<sup>[46]</sup>

The main vision-limiting factors in VKH syndrome are subretinal gliosis and optic atrophy.<sup>[47]</sup> Macula pigmentary disturbance was identified as the most common reason for BCVA poorer than 6/12 in Moorthy *et al.*'s study.<sup>[46]</sup>

CMO is uncommon in VKH syndrome<sup>[48]</sup> and is possibly a complication of cataract surgery rather than of VKH syndrome, especially if the disease was quiescent preoperatively.<sup>[16]</sup> This is supported by studies that show the similar incidence of CMO in eyes with and without uveitis.<sup>[16,33]</sup> The incidence of PCO (58%–76%, of which 42%–45% require Nd:YAG capsulotomy)<sup>[16,47]</sup> is also comparable to that in age-matched nonuveitic eyes.<sup>[49]</sup>

#### Sympathetic ophthalmia

Six sympathizing eyes of patients with sympathetic ophthalmia underwent cataract extraction in Reynard and Minckler's study.<sup>[50]</sup> Although all eyes had minimal inflammation at the time of surgery, 50% had severe uncontrolled postoperative inflammation despite corticosteroid therapy and developed cyclitic membranes or phthisis, with only light perception vision. Two eyes achieved VA better than 20/40.

#### **Ocular toxoplasmosis**

Cataract extraction has been reported to significantly increase the risk of reactivation (36%) in eyes with ocular toxoplasmosis compared to controls (2%).<sup>[51]</sup>

## **Planning for Surgery**

Initial assessment involves evaluating if the eye is quiescent, ocular comorbidities and the grade of cataract, to ascertain if the cataract is the main reason for the patient's poor visual acuity. This is essential as visual rehabilitation also depends on the degree of preoperative permanent vision-limiting pathology.

#### **Indications for surgery**

Cataract surgery is indicated in the following circumstances:<sup>[52]</sup> first, phacoantigenic uveitis, which is an absolute indication for cataract extraction; second, visually significant cataract in an eye with good expected visual potential and well-controlled inflammation; third, cataract that impairs fundus assessment in a patient with suspected posterior segment pathology; and fourth, cataract that impedes adequate visualization of the posterior segment in a patient undergoing posterior segment surgery such as a pars plana vitrectomy.

#### Evaluating causes of the poor visual acuity

The eye in which visual loss is mainly attributable to cataract is most likely to benefit from cataract surgery. A thorough preoperative ophthalmic assessment for any concomitant permanent structural damage, especially those affecting the macula and optic nerve, should be done. Chronic macular edema, macular ischemia, atrophy, hole or scar, as well as optic atrophy and glaucomatous cupping of the disc are poor prognostic factors.<sup>[16,53-57]</sup>

The retina should also be examined for ischemia, detachment, and epiretinal membranes. In presence of dense lens opacity, B scan ultrasonography should be done to rule out retinal detachment.

Other abnormalities such as corneal scars, band keratopathy, posterior synechiae, severe iris atrophy, hypotony, cyclitic membranes, and vitreous opacities may also compromise the visual outcome.<sup>[54,56]</sup>

Diseases that spare the posterior segment generally have a better prognosis than those involving the macula, retina, and/or optic nerve. The decision for cataract surgery in eyes with suboptimal predicted visual outcomes is at the surgeon's discretion, except in phacoantigenic uveitis where early cataract extraction is indicated irrespective of the visual prognosis or degree of inflammation.

#### **Timing of cataract surgery**

For successful uveitic cataract surgery, the eye needs to be quiescent for at least 3 months before surgery and the disease well-controlled throughout the postoperative period.<sup>[4,10,33,38,55]</sup> This has been shown to reduce the risk of postoperative CME<sup>[33]</sup> and ocular damage from postoperative inflammation. In pediatric patients, the risk of amblyopia needs to be considered when cataract surgery is delayed.

However, as the risk of postoperative recurrence in Behcet's disease is significantly related to the frequency of ocular attacks in the 12 months before cataract surgery, the disease should, therefore, be inactive for at least 6 months preoperatively to reduce the chance of postoperative attacks.<sup>[42,43]</sup>

It has been proposed that uveitic cataracts should be done early for several reasons. First, mature cataracts may interfere with timely detection of posterior segment complications or disease relapse, of which delayed treatment might result in irreversible functional damage. Second, advanced cataracts pose greater surgical difficulties and higher surgical risk.

#### Evaluating the risk of postoperative hypotony

Eyes with poorly controlled uveitis are at high risk of severe postoperative inflammation and hypotony or even phthisis bulbi.<sup>[57]</sup> Poor prognostic signs for postoperative hypotony include a low IOP (especially readings of 6 mmHg or less) even when quiescent; seclusio pupillae with a normal IOP; apparent phacodonesis without evident zonulysis; or a diffusely thickened choroid or choroidal effusion on B-scan ultrasonography.

#### Investigations

Investigations are directed toward identifying coexisting ocular pathologies that may contribute to surgical difficulties or warrant combined or staged procedures.

Ultrasound biomicroscopy must be conducted in eyes with relative hypotony to assess the state of the ciliary body and its processes, the zonular support and the posterior capsule for preoperative surgical planning.<sup>[58]</sup> An atrophied ciliary body has a high risk of hypotony. If cyclitic membranes are causing traction on the ciliary body processes with resultant ciliary body detachment, cataract surgery should be combined with vitrectomy, trimming of the ciliary membrane aided by scleral indentation and silicone oil filling to relieve ciliary body traction and restore normal IOP.<sup>[59]</sup>

Other investigations include optical coherence tomography of the macula to assess the macula thickness and presence of epiretinal membranes and a visual field assessment in optic nerve pathology. A fundus fluorescein angiogram to look for macular ischemia or edema, retinal ischemia and posterior segment disease activity including disc leakage may also be performed.<sup>[60]</sup>

# Counseling the Patient for Uveitic Cataract Surgery

Managing patient's expectations with regards to the visual prognosis are paramount, especially if the disease involves the posterior segment. In addition to the general risks of cataract surgery, it is necessary to emphasize that surgery could be more complicated with prolonged surgical duration due to abnormal anatomy. The patient should also be aware of the possibility of significant postoperative inflammation, delayed visual recovery, and the need for compliance with immunosuppressive medications and frequent follow-up. The loss of accommodation in uveitis with the need to use reading glasses after cataract surgery should not be overlooked. The decision as to whether an IOL should be implanted, and the type and design of the implant should also be discussed with the patient. As a general principle, multifocal lens implants are to be avoided.

# **Perioperative Optimization and Postoperative Care**

#### Control of perioperative inflammation

Cataract surgery success is critically related to careful control of inflammation, both pre- and post-operatively.<sup>[15,33,54,57,61,62]</sup>

#### Pre- and post-operative steroids

In cases where surgery is required urgently, but the intraocular inflammation is active despite heavy immunosuppression, such as in an intumescent cataract with progressive shallowing of the AC, preoperative intravenous high-dose corticosteroids are recommended.

Patients with no inflammatory activity should continue with their existing maintenance immunosuppressive regime. Patients on chronic oral corticosteroids should be prescribed a stress dose on the day of surgery and into the immediate postoperative period, which is then tapered accordingly.

Perioperative steroid prophylaxis should be given to patients who are at risk of developing postoperative macular edema or recurrence of uveitis.<sup>[63]</sup> Although several studies have reported the efficacy of various routes of perioperative steroid administration, no standard protocol is available at present.

Postoperatively, the patient should be treated with intensive topical and/or oral steroids and topical NSAIDs.[64] Oral corticosteroid prophylaxis is indicated for eyes with severe and difficult to control inflammation, especially eyes with panuveitis, such as VKH disease and Behcet's disease and perhaps also in one-eved patients especially if they had lost the other eye through uncontrolled inflammation. This is typically prescribed 3 days before surgery starting at a dose of 0.5 mg/kg of oral prednisolone in addition to the ongoing immunosuppression, and slowly tapered by 5 mg/week until 20 mg daily when it can be tapered by 5 or 2.5 mg daily every 1-2 weekly depending on the uveitis activity. After a dose of prednisolone 10 mg daily is reached, the taper can be even more gradual by 2.5 mg or 1 mg 2-4 weekly and finally stopped or until it reaches the presurgery dose. The use of a laser flare meter is extremely useful for monitoring the disease activity when tapering immunosuppression to avoid excessively rapid taper, which may result in a recurrence of uveitis. In a publication on 50 eyes with VKH undergoing cataract surgery, the authors found that only recurrent inflammation was associated with a greater likelihood of having poor vision at 6 and 12 months. No association was reported between the use of corticosteroid prophylaxis or the initial steroid dose and poor vision.<sup>[16]</sup>

Today, with the availability of stronger topical steroids such as difluprednate, intravitreal TA and Ozurdex, oral steroids may be replaced by local therapy described above or below. A study comparing the efficacy of intravitreal TA prophylaxis found no difference in the visual outcome, AC reaction, central macular thickness, or recurrence of uveitis compared to postoperative oral steroid prophylaxis.<sup>[65]</sup>

#### Intraoperative steroids

An intravitreal injection of preservative-free TA 4 mg in 0.1 mL at the end of cataract surgery<sup>[65-67]</sup> has been shown to be as effective as perioperative systemic steroids.<sup>[65]</sup> An intravitreal dexamethasone implant (Ozurdex<sup>®</sup>, Allergan Inc) also helps to prevent the recurrence or worsening of CME in uveitic patients with a history of CME who undergo phacoemulsification.<sup>[68]</sup> This may be given to eyes with increased risk of CME and recurrence that are nonsteroid responders and have infectious uveitis. Eyes that received the dexamethasone implant within 4 weeks before cataract surgery showed improvement in CME.

An alternative for patients in whom high doses of oral steroid should be avoided such as poorly controlled diabetics is an orbital floor or subtenon's injection of depot steroid. A single-dose intraoperative orbital floor injection of TA 40 mg/1 mL is as effective as a 4-week course of postoperative oral prednisolone in terms of reducing postoperative inflammation, macular edema, and improving the visual outcome.<sup>[69]</sup>

Topical steroids such as prednisolone acetate 1% or disodium dexamethasone 0.15% can also be given together with oral and topical nonsteroidal anti-inflammatory drugs (NSAIDs). An intensive perioperative topical steroid regimen alone was statistically comparable to oral steroids in preventing postoperative uveitis relapse in a small nonmasked randomized controlled trial. However, the recurrence rate was higher.<sup>[70]</sup>

#### Antimicrobial prophylaxis

Preoperative prophylaxis should be considered in eyes with infectious uveitis that may reactivate after surgery.

#### Ocular toxoplasmosis

The role of prophylactic anti-toxoplasma therapy for eyes with ocular toxoplasmosis undergoing cataract surgery is controversial. Bosch-Driessen *et al.* reported a 36% risk of reactivation of toxoplasmic retinochoroiditis following cataract surgery<sup>[51]</sup> and therefore suggested the use of prophylactic antiparasitic drugs during and after cataract surgery in patients at risk of visual loss due to lesions near the macula or the optic nerve. However, a more recent retrospective analysis by Heringer *et al.* found that intraocular surgery in the absence of preoperative antitoxoplasmic prophylaxis did not result in a significant reactivation rate of toxoplasmic retinochoroiditis.<sup>[71]</sup>

#### Herpes simplex uveitis

Oral acyclovir 400 mg twice daily<sup>[72]</sup> or valacyclovir 0.5 g four times daily preoperatively and for 2–3 weeks postoperatively

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may help prevent recurrence of herpes simplex uveitis after surgery. Postoperatively, topical NSAIDs in combination with lower doses of topical corticosteroids is useful, as NSAIDs are associated with less risk for herpetic corneal recurrence compared to topical corticosteroids.<sup>[73]</sup>

# **Uveitic Cataract Surgery**

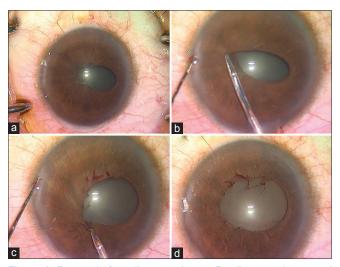
#### **General considerations**

Surgery is challenging because of the patient and ocular factors. Successful surgery aims to reduce the occurrence of commonly cited postoperative complications such as fibrinous reaction in the AC, peripheral anterior synechiae (PAS), iris bombe, anterior capsular fibrosis, IOL deposits, glaucoma, hypotony, CME, PCO, and recurrent uveitis.<sup>[15,56,61]</sup>

If the patient with ankylosing spondylitis has postural issues such as kyphosis, the bed may have to be tilted in a Trendelenburg position.

Recurrent anterior segment inflammation often leaves the sequalae of small pupil which may be associated with posterior synechiae and pupillary membrane [Fig. 1]. In addition, surgical visibility may be reduced due to cornea scars, band keratopathy, and cornea haze and vascularization secondary to chronic ocular surface inflammation. Capsular dyes such as trypan blue stain provide contrast and facilitate visualization during the capsulorrhexis. The use of 23G optic fiber xenon illumination device placed externally near the cornea or into the AC greatly facilitates visualization by counteracting the light scatter from the operating microscope and improving contrast and depth perception.

Regional anesthesia such as a peribulbar block should be considered for patient comfort to avoid pain during iris manipulation. The uveitic iris may be atrophic and floppy. It is important to minimize iris trauma during surgery as this increases intraoperative floppiness which can lead to iris prolapse and tattering of iris. The incisions thus need



**Figure 1:** Removal of pupillary membrane. Pupillary membrane and posterior synechiae prevent pharmacological pupil dilation (a). The micrograsper is used to remove the pupillary membrane by carefully grasping the membrane edge (b) and freeing the membrane from the pupil margin using a capsulorhexis maneuver (c), thereby achieving a round pupil (d)

to be snug and not too short. An injection of intracameral dexamethasone (0.4 mg/0.1 ml)<sup>[74]</sup> at the conclusion of surgery may help to control early postoperative inflammation and reduce fibrinous reaction, especially if the pupil was manipulated.

#### Small pupil scenarios

#### No pupil synechiae or peripheral anterior synechiae

The small pupil makes access to the cataract difficult. The pupil may be dilated pharmacologically, using manual stretching or mechanical dilation with devices. The first step of using balanced salt solution with adrenaline (1ug/ml; 0.5 ml of 1:1000 adrenaline in 500 ml BSS) through the paracentesis site may have some effect on the pupil but is usually inadequate to dilate the pupil adequately.

Dispersive or even more effective, viscoadaptive ophthalmic viscoelastic devices (OVD) sometimes may be enough to push the pupil wider. However, if the pupil does not dilate with OVD, it can be mechanically stretched using 2 Kuglen hooks 180° apart (push and pull technique) [Fig. 2] to widen it in the horizontal and vertical meridians. However, if the iris is thin and atrophic, it is advisable to avoid stretching the pupil forcefully as it can rip.

Pupil dilation devices include self-retaining iris hooks (4 hooks placed through 4 sub-1 mm paracentesis ports in a diamond pattern) [Fig. 3], or pupil expanders such as the Malyugin ring (MST, Redmond, Wash) [Figs. 4 and 5]. Iris hooks are very useful as it is possible to adjust the amount of tension applied on each hook for adequate pupil opening size. Placing the iris hook under the main phaco incision also helps to keep the iris away from the incision and phaco probe.

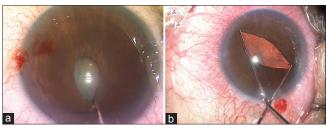
#### Partial pupil synechiae with peripheral anterior synechiae

If both PAS and posterior synechiae are present, one should release the PAS and open the angles first, with visco-dissection before releasing posterior synechiae. The OVD cannula is directed into the angle space with a gentle sweeping action while simultaneously injecting OVD to lyse the PAS circumferentially, taking care not to detach Descemet's membrane.

If small sections of pupil synechiae are present, release the adhesions with dispersive OVD and Kuglen hook, which facilitates separation of the adherent iris from anterior capsule.

#### Total membrane occluding pupil

With OVD in the AC, a 23 gauge microforceps (e.g. Kawai [Kenji Kawai capsulorrhexis forceps; ASICO, LLC]) is used to grasp



**Figure 2:** Pupil enlargement with Kuglen hooks. The Kuglen hook is used in a pull movement (a) to release the posterior synechiae. The pair of angled Kuglen hooks stretches the pupil through a simultaneous push and pull of the iris (b). This is repeated at an axis perpendicular to the first stretch

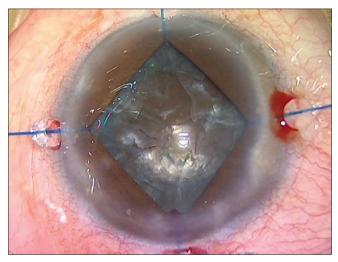
the pupil edge to initiate pupil edge membrane peeling. Stripping of the fibrotic tissue band around the pupil margin allows for immediate dilation of the pupil. If mild bleeding occurs, keeping the eye pressurized helps to prevent further bleeding.

If the pupil size is still <4 mm, multiple small sphincterotomies [Fig. 6] may be performed; earlier mentioned options include bimanual stretching with Kuglen Hooks, iris hooks or subsequent use of a pupil expansion device (e.g. Beehler pupil dilator<sup>[75]</sup> [Fig. 7] or Malyugin ring). Iris hooks are suitable for eyes with a small pupil and shallow AC. For a deep AC and with a pupil that is not scarred or atrophic, the Malyugin ring or Beehler pupil dilator can be considered. Made of 5/0 prolene (instead of 4/0 prolene of the first version), the recent Malyugin ring 2.0 is softer, more elastic and designed with a new injector to enter through a 2 mm clear cornea incision.

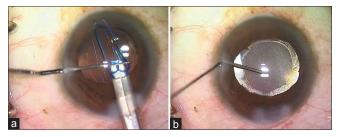
The pupil size should be maximized to enable achievement of a "well-sized" capsulorrhexis of approximately 5 mm, as this helps to reduce the postoperative incidence of progressive anterior capsular phimosis and recurrent synechiae formation.

#### Phacoemulsification

During the phacoemulsification stage, care should be taken to observe and maintain the AC fluid dynamics. The vertical



**Figure 3:** Pupil enlargement with iris hooks. Adjustable pupil expansion is achieved using self-retaining iris hooks which are placed in a diamond configuration, retracting the iris in the sub-incisional location



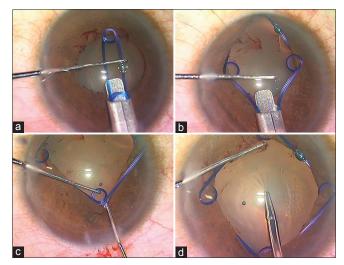
**Figure 5:** Removal of the Malyugin ring. As the ring is retracted into the injector at the conclusion of the surgery, the 2 lateral rings are compressed by a Sinskey hook (a). The result is a round reactive pupil (b)

phaco-chop technique is safer when the pupil is small as the instruments are kept in the center of the pupil.

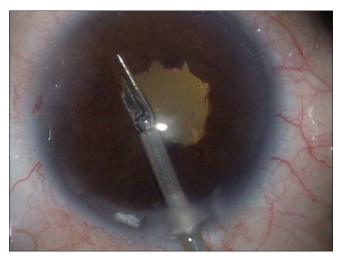
#### Role of femtosecond laser cataract surgery

The choice of femtosecond laser cataract surgery (FLACS) is surgeon-dependent. In the presence of dense nuclear opacity, particularly if the endothelial cell count is low (<1000 cell/mm<sup>2</sup>), FLACS may be safer as it has been shown to be superior to conventional phacoemulsification with less postoperative endothelial cell reduction.<sup>[76]</sup> If the pupil is small, the laser capsulotomy and fragmentation steps may be performed after pupil synechiolysis or with pupil expanders in place (e.g. Malyugin ring). The incisions are sutured after the pupil ring is inserted to avoid any wound leakage during the laser suction procedure. This is followed by the phacoemulsification procedure.

To date, no randomized study has compared the outcomes between conventional and FLACS cases in uveitic eyes.



**Figure 4:** Pupil enlargement with the Malyugin ring. The leading scroll is inserted (a). After viscoelastic injection under the iris, the two side scrolls are manipulated (b). A Kuglen hook is used to gently stretch the pupil to tuck the distal scroll into position (c), creating a fixed round pupil (d)



**Figure 6:** Multiple small sphincterotomies. A small pupil may be enlarged by making multiple small sphincterotomies

It is known that the FLACS capsulotomy step triggers prostaglandins release and secondary miosis.<sup>[77]</sup> Therefore, as in nonuveitic eyes, topical NSAIDS should be prescribed preoperatively and continued postoperatively for 6 weeks, to reduce the effect of CME.<sup>[78]</sup>

#### Intraocular lens implant

In eyes with poorly controlled uveitis, IOL implantation should be deferred.

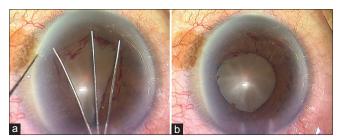
In terms of IOL types, a Cochrane Database review  $2014^{[79]}$  on adult uveitic cataracts had reported results of 4 randomized controlled trials (RCTs) comparing up to 4 different IOL types. In the largest RCT (n = 140, 1 year follow-up), fewer eyes with hydrophobic acrylic IOLs had posterior synechiae compared to eyes receiving silicone IOLs; however, there was no significant correlation to IOL types for the risk for PCO, corneal edema, CME, or mild IOL decentration. In another RCT of 60 eyes, no statistical or clinical difference was found between hydrophobic and hydrophilic acrylic IOL types (3-month follow-up) in the proportions of patients with two or more Snellen lines of visual improvement. The rates of PCO at 6-month follow-up with hydrophobic and hydrophilic acrylic IOLs were similar. Therefore, it was concluded that there was no definite evidence as to which IOL type provided the best visual outcomes.

If a complication such as posterior capsule rupture occurs during surgery, and there is inadequate anterior capsule rim support for a sulcus placed IOL, an AC IOL should be avoided. The patient should be left aphakic and scleral fixation of a 3-piece IOL performed by a cataract specialist at a later date when the postoperative inflammation has settled.

## The Pediatric Eye

Issues are specific to pediatric cataracts include biometry calculations, axial length changes after cataract surgery, increased rate of PCO,<sup>[80]</sup> increased postoperative inflammation, secondary glaucoma and amblyopia management. If acute-on-chronic inflammatory changes develop, severe band keratopathy, fibrin and synechiae formation may ensue.<sup>[62]</sup> Children with JIA-associated uveitis are likely to have a more relentless postsurgical course compared with uveitis secondary to other causes.<sup>[26,32]</sup>

Intraoperatively, besides the small pupil, the anterior capsule is more elastic, and there is general agreement that primary posterior capsulotomy with anterior vitrectomy [Fig. 8] for children <8 years of age is required because of the imminent PCO soon after surgery. The use of TA as an anti-inflammatory and vitreous visualizer has been explored by many authors. In a



**Figure 7:** Pupil enlargement with the Beehler pupil dilator. A 3-pronged Beehler pupil dilator is inserted to enlarge a small pupil (a), creating a round dilated pupil (b)

prospective case series of 43 eyes (mean 14.8 months old), Praveen *et al.*<sup>[81]</sup> showed that preservative-free TA improved visualization of the vitreous during pediatric cataract surgery, thereby ensuring thorough and complete anterior vitrectomy. They reported no adverse effects. Dixit *et al.*<sup>[82]</sup> and Gupta *et al.*<sup>[83]</sup> have advocated intraoperative preservative-free TA as it significantly reduced postoperative anterior segment inflammation with less pigment deposits on the IOL surface while affording enhanced visualization of vitreous during the surgery itself.

The choice of IOL remains a contentious issue; most surgeons would avoid an IOL in uveitic eyes of below 2 years of age. In older children, most studies document the use of PMMA or acrylic IOLs.<sup>[84,85]</sup>

In pediatric uveitis cases, a multistage surgery may be the safer approach so that various complications are addressed at different sittings.<sup>[86]</sup>

## **Postoperative Complications**

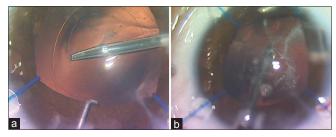
Cataract surgery in uveitic eyes is associated with significantly higher rates of both intra- and post-operative complications.<sup>[5]</sup>

# Excessive postoperative inflammation and cystoid macular edema

Uveitic eyes typically have an unusually exuberant postoperative inflammatory response.<sup>[55,87]</sup> The severity or duration of inflammation varies and can lead to posterior synechiae and IOL capture, CME<sup>[14,61,88]</sup> (incidence of 33-56%<sup>[89,90]</sup> and 12-59%<sup>[61]</sup> following extracapsular cataract surgery and phacoemulsification, respectively) epiretinal membranes,<sup>[89]</sup> glaucoma, pupillary or ciliary membrane formation<sup>[91]</sup> and even phthisis if uncontrolled. Eyes at risk of CME include those with previous CME with each recurrence, or those with intermediate, posterior, and panuveitis.

Complications should be treated aggressively. The dose of oral steroid prophylaxis should be increased to combat any inflammation. If no prophylactic oral steroids had been given, a pulse of oral steroids or periocular steroid injection should be administered. Alternatively, to circumvent the need to adjust the systemic immunosuppression, an intravitreal injection of TA can be given if not previously given intraoperatively.<sup>[92]</sup>

A stepwise approach to the treatment of postoperative CME is as follows. The first line of treatment is intensive topical prednisolone acetate or difluprednate in combination with topical NSAIDs if not already given prophylactically. If this is inadequate to control the CME, in the absence of associated severe intraocular inflammation, periocular steroid



**Figure 8:** Additional steps required in the pediatric cataract. In the pediatric cataract, other steps include a posterior capsulorhexis using microcapsulorhexis forceps (a) and a limited 23 Gauge anterior vitrectomy performed with triamcinolone staining of vitreous (b)

injection of TA 4 mg/1 ml can be given transeptally to the orbital floor or subtenon route.<sup>[93]</sup> Intravitreal TA 0.4 mg/ml or dexamethasone (Ozurdex) is also an option. Intravenous 'pulse' methylprednisolone is indicated in eyes where the CME fails to respond to the conventional therapy. A study of 14 eyes with persistent chronic inflammatory CME despite corticosteroid or immunosuppressive therapy had rapid resolution of CME within 3 days.<sup>[94]</sup>

Although the abovementioned treatment options have a relatively transient effect, they are effective as a bridge to definitive therapy when used concomitantly with steroid-sparing immunosuppressants. If CME persists in spite of the above treatment options, it may be treated with intravitreal antiangiogenic agents, systemic carbonic anhydrase inhibitors, interferon  $\alpha$ 2a subcutaneous injections<sup>[95]</sup> and rarely, somatostatin analogues such as octreotide.

The response of CME to treatment should be monitored clinically and with postoperative OCT scans of the macula.

#### **Raised intraocular pressure**

Postoperatively, ocular hypertension may be related to the increase in inflammatory debris and retained ophthalmic viscoelastic substance, and when the presentation is delayed, may be due to steroid response. On the first few days following surgery, a spike of IOP may be managed by carefully releasing aqueous at the slit lamp. This is done by slowly and gently depressing the posterior lip of the main incision, which is often sited temporally. Concurrently, antiglaucoma medications should be given, avoiding antiprostaglandin analogs to reduce the risk of postoperative macular edema, especially if the eye is aphakic and the posterior capsule has been opened. Systemic carbonic anhydrase inhibitors may be administered intravenously or systemically, depending on the urgency of IOP lowering required. When steroid response ocular hypertension is suspected, the topical steroid frequency should be tapered, and if the inflammation persists, oral immunosuppression should be increased. Rarely, glaucoma filtration with anti-metabolites or set on surgery may be indicated if the IOP is recalcitrant to therapy and there is progressive optic disc cupping and visual field loss.

#### Hypotony

In the event of postoperative hypotony, after wound leakage has been ruled out, both topical and systemic anti-inflammatory therapy should be sharply increased to raise the IOP. Eyes in which hypotony recurs on tapering or discontinuation of the topical steroids may require long-term topical steroids to maintain the IOP. An intraocular injection of sodium hyaluronate through a limbal paracentesis has been shown to stabilize the IOP in nonuveitic eyes,<sup>[96]</sup> although this has not been studied in uveitic eyes.

#### **Recurrence of uveitis**

A laser flare meter can be used to monitor the degree of intraocular inflammation to detect signs of disease recurrence. As the recurrence rate is as high as 51%,<sup>[14]</sup> stepping up the immunosuppression for the long term may be necessary to prevent further recurrence.

#### **Delayed complications**

#### Posterior capsular opacification

PCO is the most common late postoperative complication following any type of cataract surgery. The risk is even higher

in uveitic eyes<sup>[87,90,97]</sup> especially in younger patients.<sup>[49,98]</sup> The incidence of PCO in uveitic eyes is 34.2-81.7%<sup>[96,97,99]</sup> and 3.6-32.2% require Nd:YAG capsulotomy.<sup>[96,97,99]</sup> Nd:YAG capsulotomy is indicated when the visual acuity is reduced or when visibility of the posterior segment is compromised due to PCO. This should be delayed until the postoperative inflammation is under control and preferably 6-month postsurgery to reduce the risk of a retinal tear and detachment associated with the possible induction of posterior vitreous detachment unless the latter is already preexisting.

#### Capsular phimosis and membrane formation

There is a greater tendency to capsular phimosis [Fig. 9] in uveitic eyes due to the increased postoperative inflammatory and scarring response. This may be avoided by enlarging the capsulorhexis at the end of surgery if it too small. In addition, a capsular tension device should be inserted in eyes with weak zonules, which occurs frequently when the uveitis has been chronic. This will help to resist the contracture forces and reduce the risk of capsular phimosis.

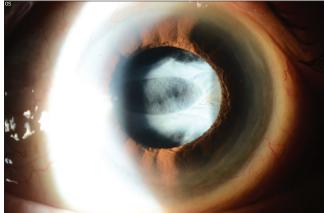
Severe capsular phimosis may impair vision. This can be managed by cutting the fibrotic capsulorhexis rim with the YAG laser, placing four spaced out radial incisions and taking care to avoid damaging the IOL.

Any associated membrane formation [Fig. 9] may also be handled with the YAG laser. However, failure to remove a membrane that may be adherent to the IOL surface or to open the phimotic capsule should be managed by surgical means. Intraocular forceps and scissors are used to cut the membrane and enlarge the capsule opening. Intracameral dexamethasone 0.4 mg/0.1 ml should be administered at the end of surgery followed by aggressive postoperative regimen of topical steroids to avoid recurrence of membrane formation.

#### Intraocular lens giant cell and pigment deposits

This may occur following surgery with chronic postoperative inflammation. When the uveitis is under control, these deposits on the anterior surface of the IOL which cause light scattering and visual impairment may be blasted off by careful YAG laser with the anteriorly positioned focus beam and low power, taking care not to pit the IOL.

Figure 9: Capsular phimosis and membrane formation. Capsular phimosis and membrane formation over the intraocular lens that impair vision can be treated with a YAG laser



## Intraocular lens intolerance

Removal of IOL in uveitic eyes is rarely necessary unless serious IOL-related complications develop.<sup>[100,101]</sup> These include chronic low-grade inflammation unresponsive to anti-inflammatory treatment, the formation of perilenticular membranes, cyclitic membranes resulting in hypotony and maculopathy. Foster *et al.* proposed that these complications were secondary to undetected chronic subclinical inflammation present after surgery. Eyes with the inflammation centered on the pars plana region such as in uveitis associated with sarcoidosis, JIA, and pars planitis, especially intermediate or panuveitis, are at higher risk of IOL intolerance.<sup>[102]</sup> Lenses thought to be contributing to complications are often explanted earlier in the postoperative course.<sup>[100]</sup>

# Conclusion

A thorough ophthalmic assessment is needed to identify uveitic patients who will benefit from cataract surgery. The disease should be quiescent for a minimum of 3 months, and the patient should be given adequate perioperative prophylactic anti-inflammatory therapy. Although uveitic cataracts are more surgically demanding due to pupil abnormalities that limit access to the cataract, various surgical techniques can be employed to ensure successful cataract surgery. The eye should be monitored closely for postoperative complications and treated aggressively should they occur. Good visual outcomes can be achieved after uveitic cataract surgery if there is minimal macula or optic nerve pathology and the inflammation is well controlled.

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#### **Conflicts of interest**

There are no conflicts of interest.

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