CHAPTER 16

Complications of Uveitis

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Highlights

- In uveitis, vision loss can result from multiple complications involving the cornea, lens, vitreous, retina, and optic nerve. Consequently, consultations with multiple ophthalmic subspecialists can help improve the management of challenging cases.
- When surgical intervention is required in patients with uveitis, strict perioperative control of inflammation is paramount.
- Uveitic macular edema, which is a common cause of vision loss, can be diagnosed with optical coherence tomography; treatment is typically focused on controlling inflammation, often with adjunctive local corticosteroids.

Calcific Band Keratopathy

Band keratopathy, or calcium deposition along the epithelial basement membrane and Bowman layer, may develop in patients with chronic uveitis, especially children. Most often seen in juvenile idiopathic arthritis–associated anterior uveitis or undifferentiated chronic anterior uveitis, band keratopathy may arise within months of uveitis onset. The deposition is typically located in the interpalpebral zone and becomes visually significant when it extends into the visual axis. It may also cause foreign body sensation. Because the calcium deposits are located beneath the corneal epithelium, their removal requires epithelial debridement followed by chelation with disodium EDTA. Recurrences may require repeated EDTA treatments. Photorefractive keratotomy may also be considered as an alternative to chelation with EDTA.

Cataracts

Chronic inflammation and/or long-term corticosteroid use can provoke cataract development. Indications for cataract surgery include functional impairment that interferes with activities of daily living, decreased vision, and amblyopia prevention. In uveitic eyes, an additional consideration for surgery is whether the cataract obscures the view of the fundus and interferes with monitoring of posterior segment inflammation. A careful preoperative evaluation will help determine whether the cataract is actually contributing to visual dysfunction.

Other potential causes of vision loss in uveitic eyes include corneal or vitreous opacity, macular edema, macular atrophy or fibrosis, and glaucoma.

Management

Uveitic eyes are at greater risk for complications after cataract surgery than are nonuveitic eyes. Thus, careful planning, such as preoperative medical management that includes control of inflammation and timing of the procedure, is key to a successful visual outcome. To decrease the risk of severe postoperative inflammation, the general guideline is to achieve well-controlled uveitis without flare-ups for at least 3 months before cataract surgery. However, this recommendation is based on retrospective clinical case series and clinical experience; no prospective or controlled trials have provided definitive data on the 3-month guideline. As such, exceptions to this guideline may be warranted, for example, in eyes with mild uveitis lacking sequelae, in patients with uveitic disorders that have a good surgical prognosis (eg, Fuchs uveitis syndrome), or in special circumstances such as lens-induced uveitis or when the posterior segment must be visible (eg, to repair a rhegmatogenous retinal detachment).

Of note, the "best possible control" of uveitis may not be achieved with corticosteroids alone. Before proceeding with surgery, the clinician should use all appropriate means for uveitis control, including systemic immunosuppression and/or referral to a specialist for help with systemic treatment.

Once long-term control of uveitis has been achieved, perioperative management may include oral corticosteroids (0.5–1.0 mg/kg/day, started 3 days before and subsequently tapered in the weeks after surgery) and/or intensive topical corticosteroids. Sub-Tenon or intravitreal corticosteroids may also be used. There are no prospective comparative data on optimal perioperative inflammatory control, so surgeons typically rely on preference and experience.

Patients with certain infectious uveitic entities (eg, those caused by *Toxoplasma gondii* infection and herpetic viral infections) may require perioperative prophylactic antimicrobial therapy to prevent surgically induced recurrence. Preoperative oral corticosteroids are usually not given to these patients.

In general, cataract surgery in uveitic eyes is more complex than in nonuveitic eyes because of possible sequelae of the disorder, including posterior synechiae, pupillary membranes, corneal edema or opacity, and hypotony. Entrance into the eye through a clear corneal approach is typical and may be particularly desirable in patients with scleritis to reduce the risk of postoperative scleral necrosis. Posterior synechiae and pupillary miosis may require mechanical or viscoelastic pupil stretching, sphincterotomies, or the use of flexible iris retractors (Video 16-1).

VIDEO 16-1 Synechiolysis, placement of iris dilator, and capsular staining in a patient with uveitis.
Courtesy of Russell W. Read, MD, PhD.

Although a curvilinear capsulorrhexis is preferred for uveitic eyes, a can-opener capsulotomy may be the only way to open a fibrotic anterior capsule. In eyes with zonular insufficiency, options include the use of a capsular tension ring or pars plana lensectomy and

vitrectomy with a 3-piece sulcus intraocular lens (IOL) or scleral-fixated IOL. In some situations, the eye may be left aphakic. Relative contraindications for IOL implantation in uveitic eyes include prior development of rubeosis, a history of extensive membrane formation, and hypotony; however, even in these circumstances, an IOL may be used in select cases if inflammation is well controlled before and after surgery. In patients with vision-limiting vitreous opacity or macular pathology, such as epiretinal membranes or macular hole, phacoemulsification with IOL implantation can also be done in conjunction with pars plana vitrectomy. Meticulous cortical cleanup is important to minimize proinflammatory material in the eye. For IOL choice, many surgeons prefer hydrophobic acrylic posterior chamber IOLs placed in the capsular bag. Studies have shown that phacoemulsification with implantation of a posterior chamber (in-the-bag) IOL effectively improves vision and is well tolerated in many eyes with uveitis, even over long periods. Silicone IOLs are rarely used in uveitic cataracts.

At the conclusion of surgery, periocular or intravitreal corticosteroids may be administered. Postoperatively, systemic immunomodulation is continued and supplemented with liberal use of topical corticosteroids, which are slowly tapered. Dosages of topical and/or oral corticosteroids may be tapered over weeks to months after surgery based on the severity of the preexisting uveitis and the postoperative inflammatory response. In patients who had extensive posterior synechiae and/or peripheral anterior synechiae before surgery, cycloplegic drops may be continued for a week or 2 after surgery to prevent re-formation of posterior synechiae to the anterior capsule or IOL.

In children with juvenile idiopathic arthritis–associated uveitic cataracts, the debate regarding IOL placement is ongoing. Recent studies have shown favorable outcomes of IOL placement with or without combined pars plana vitrectomy. Although avoiding aphakia in children is desirable, it may not always be in their best interest because of the potential complications of IOL placement in uveitic eyes. Choosing the proper IOL power, especially in children younger than 10 years, can also be challenging because of normal ocular/orbital growth. (For more information about IOL use in children, see BCSC Section 6, *Pediatric Ophthalmology and Strabismus*.) In-the-bag implantation of acrylic IOLs and primary posterior capsulorrhexis are generally preferred. Some surgeons may also perform a core anterior vitrectomy through the posterior capsulorrhexis before IOL placement. Regardless, the most important step in treating these children is stringent control of preoperative and postoperative intraocular inflammation with corticosteroids and immunomodulatory therapy (IMT). Administration of intraocular corticosteroids at the end of the procedure is extremely useful for controlling postoperative inflammation and uveitic macular edema (UME). When these methods are used, 75% of patients have obtained a visual acuity of better than 20/40.

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Complications

After cataract surgery, close monitoring of the patient for inflammation or complications is critical, as strict control reduces the risk of inflammatory debris and membrane accumulation on the surface of the IOL. In addition, aggressive dosing of topical and local corticosteroids as well as IMT are often necessary. In patients with major postoperative anterior segment inflammation, cycloplegic drops should be used to reduce the chance of developing posterior synechiae with iris adherence to either the IOL or the anterior capsule. Inflammatory cocooning of the IOL–lens capsule complex and uncontrolled inflammation necessitate IOL explantation in 5%–10% of patients.

Postoperative UME rates may be reduced by delaying surgery until the uveitis has been controlled for at least 3 months and through the use of perioperative corticosteroids. Postoperative use of topical nonsteroidal anti-inflammatory drugs (NSAIDs) has not been studied in uveitic eyes; however, similar to their use in nonuveitic eyes, topical NSAIDs may be employed to prevent postoperative macular edema. The incidence of posterior capsule opacification is higher in uveitic eyes than in nonuveitic eyes, leading to earlier use of Nd:YAG laser capsulotomy in this population. However, Nd:YAG laser capsulotomy may exacerbate uveitis, so patients should be monitored carefully after the procedure.

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Sen HN, Abreu FM, Louis TA, et al; Multicenter Uveitis Steroid Treatment (MUST) Trial and Follow-up Study Research Group. Cataract surgery outcomes in uveitis: the Multicenter Uveitis Steroid Treatment trial. *Ophthalmology*. 2016;123(1):183–190.

Glaucoma

In uveitic eyes, elevated intraocular pressure (IOP) may be acute, chronic, or recurrent. Multiple mechanisms may contribute to uveitic glaucoma, including mechanical factors (obstruction of the angle with peripheral anterior synechiae or inflammatory debris) and biochemical changes. Active uveitis alone is not necessarily the cause of elevated IOP, especially in the case of posterior uveitis. Acute anterior uveitis can even be associated with a *decrease* in IOP due to inflammation of the ciliary body. In eyes with long-term ciliary body inflammation, the IOP may fluctuate between abnormally high and low values. When acute anterior uveitis is accompanied by acute ocular hypertension, a herpes virus–associated anterior uveitis should be suspected. Another major contributor to uveitic glaucoma is overuse of topical and/or regional corticosteroids. Uncontrollable or barely controlled corticosteroid-induced glaucoma can be an indication to add systemic IMT.

Assessment of patients with uveitis and elevated IOP should include the same measures used for other cases of ocular hypertension: slit-lamp and dilated fundus examination,

pachymetry, gonioscopy, evaluation of the optic nerve head with disc photographs and optical coherence tomography (OCT), and serial automated visual fields.

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Uveitic Ocular Hypertension

Unilateral uveitis of sudden onset with open angles and increased IOP may have an infectious origin, particularly a viral cause or *Toxoplasma* infection. Thus, when IOP elevation occurs early in the course of uveitis, clinicians should resist the urge to prematurely taper corticosteroids because of a fear of steroid-induced ocular hypertension (which rarely occurs before 3 weeks of corticosteroid therapy). In patients with unilateral uveitis, early IOP elevations with active anterior segment inflammation almost always require aggressive anti-inflammatory treatment in addition to IOP-lowering medication.

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Uveitic Glaucoma

Uveitic glaucoma is classified by morphologic changes in angle structure as either secondary angle-closure glaucoma or secondary open-angle glaucoma. These disorders can be further subdivided into acute and chronic types. In cases of chronic uveitic glaucoma, corticosteroid-induced ocular hypertension and glaucoma should also be addressed. See also BCSC Section 10, *Glaucoma.*

Secondary angle-closure glaucoma

Acute disease with central shallowing of the anterior chamber Acute secondary angle-closure glaucoma may occur when choroidal inflammation results in forward rotation of the ciliary body and lens–iris diaphragm. It may also be the presenting sign of Vogt-Koyanagi-Harada syndrome or sympathetic ophthalmia. Affected patients present with pain, elevated IOP, and no posterior synechiae. The diagnosis is confirmed by ultrasound biomicroscopy (UBM) or ultrasonography showing choroidal thickening and anterior rotation of the ciliary body. Treatment involves aggressive corticosteroid therapy, aqueous suppressants, and cycloplegia to induce a posterior rotation of the ciliary body. As the inflammation subsides, the anterior chamber deepens and the IOP normalizes. Peripheral laser iridotomy or surgical iridectomy is not useful in acute disease because the underlying cause is not pupillary block.

Acute disease without central shallowing of the anterior chamber Chronic or acute recurrent anterior segment inflammation may lead to the formation of circumferential posterior synechiae with pupillary block. This is typically due to seclusion of the pupil and resultant iris bombé, which produces secondary peripheral angle closure. Although synechiae usually form over time, iris bombé may be an acute event. Peripheral laser iridotomy or surgical iridectomy results in resolution of the bombé and angle closure if the procedure is performed

before peripheral anterior synechiae become permanent. If peripheral anterior synechiae have started to develop, the procedure may be supplemented with goniosynechialysis; however, this approach is controversial. Iridotomies should involve multiple holes that are as large as possible. Intensive topical corticosteroid and cycloplegic therapy is administered after laser iridotomy. In patients with brown irides, pretreatment of the iris with an argon laser before Nd:YAG laser use may lessen the chance of bleeding and facilitate a wider opening. Exacerbation of inflammation after the laser procedure may cause closure of the iridotomy and necessitate re-treatment or surgical iridectomy.

Chronic disease Chronic intraocular inflammation may cause posterior and peripheral anterior synechiae as well as chronic secondary angle-closure glaucoma. In these cases, chronic secondary open-angle glaucoma and corticosteroid-induced glaucoma are often superimposed. Topical aqueous suppressants may not prevent progression of optic nerve head damage, requiring goniosynechialysis and trabeculectomy with mitomycin C or placement of a glaucoma drainage device. See BCSC Section 10, *Glaucoma,* for more details on the surgical treatment of glaucoma.

Secondary open-angle glaucoma

Acute disease Inflammatory open-angle glaucoma occurs when the trabecular meshwork is inflamed (ie, trabeculitis). The trabeculitis commonly occurs with infectious causes of uveitis such as *Toxoplasma* retinochoroiditis, necrotizing herpetic retinitis, herpes simplex and varicella-zoster anterior uveitis, cytomegalovirus anterior uveitis (including the Posner-Schlossman type), and sarcoidosis-associated uveitis or when inflammatory debris clogs the angle. This type of glaucoma often responds to treatment targeting the infectious agent, supplemented by topical cycloplegics and corticosteroids.

Chronic disease Chronic outflow obstruction is caused by direct damage to the trabecular meshwork. The management of chronic secondary open-angle glaucoma is similar to that of primary open-angle glaucoma (see BCSC Section 10, *Glaucoma*), with the addition of IMT to strictly control intraocular inflammation.

Combined-mechanism uveitic glaucoma

As noted previously, multiple mechanisms may be responsible for elevated IOP in uveitic eyes. Thus, a multimodal treatment approach that incorporates both medical and surgical therapies aimed at the responsible mechanisms should be used to control inflammation and IOP.

Corticosteroid-Induced Ocular Hypertension and Glaucoma

Elevated IOP in patients with uveitis should prompt consideration of one of the aforementioned angle issues or a corticosteroid-induced disorder. In patients with uveitis, corticosteroids in any formulation—topical, periocular, intraocular (injection and sustained release), or systemic—may also induce an elevation of IOP that may be difficult to distinguish from other causes of ocular hypertension. Fluocinolone intraocular implants are associated with an eventual need for glaucoma surgery in approximately 3% to 40% of eyes depending on

the implant corticosteroid dose. Topical difluprednate also appears to be associated with substantial and sometimes very rapid increases in IOP. This IOP rise may be avoided by use of a less-potent topical corticosteroid preparation, a less-frequent administration schedule, or both. Although fluorometholone and loteprednol may be less likely to elevate IOP than other corticosteroids are, they are also less effective in controlling intraocular inflammation.

Management

Medical management of uveitic ocular hypertension and uveitic glaucoma requires aggressive control of both intraocular inflammation and IOP to prevent progressive glaucomatous optic nerve damage and visual field loss (see BCSC Section 10, *Glaucoma*). Aqueous suppressants are generally the first-line treatment. Prostaglandin analogues may also be used to treat uveitic ocular hypertension and glaucoma and generally do not exacerbate intraocular inflammation, especially when used concomitantly with IMT and corticosteroids. However, caution is important when prostaglandin analogues are used in eyes with herpetic uveitis, because the medication may lead to viral reactivation. Pilocarpine should be avoided in uveitis, as pilocarpine breaks down the blood–aqueous barrier, and posterior synechiae may be more likely to form in the immobile small pupil.

When medical management fails, glaucoma filtering surgery is indicated. Although standard trabeculectomy has an increased risk of failure in uveitic eyes, results may be improved by using mitomycin C with intensive topical corticosteroids. After surgery, IOP control with 0 or 1 medication is achieved in up to 90% of patients 1 year after surgery and in approximately 62% of patients 5 years after surgery. Surgical complications include cataract formation, early and late bleb leakage (with increased risk of endophthalmitis), and choroidal effusions.

Several alternatives to classic trabeculectomy have shown short-term success in treating uveitic glaucoma. Nonpenetrating deep sclerectomy with or without a drainage implant controlled IOP in up to 90% of uveitic eyes for 1 year after surgery. Viscocanalostomy has shown even higher success rates in a limited number of studies. Among pediatric patients with uveitis, goniotomy has up to a 75% chance of reducing IOP to 21 mm Hg or less after 2 operations. However, this procedure may be complicated by transient hyphema and worsening of the preexisting cataract. Trabeculodialysis and laser sclerostomy have high rates of failure in treating uveitic glaucoma because of recurrent postoperative inflammation. The role of minimally invasive glaucoma surgery in uveitis remains unclear, with isolated reports of inflammation induced by devices placed in the angle.

Most cases of uveitic glaucoma, especially in pseudophakic or aphakic eyes, require aqueous drainage devices. These devices may be tunneled into the anterior chamber or placed through the pars plana directly into the vitreous cavity after vitrectomy. Use of a unidirectional valve design (ie, valve implant) can prevent postoperative hypotony. Compared with trabeculectomy, these implants are more likely to successfully control IOP in the long term; in studies, they reduced preoperative IOP by up to 75% as well as controlled IOP with 0 or 1 medication in nearly 75% of patients after 4 years. Unlike trabeculectomy, the drainage devices continue to function despite chronic, recurrent inflammation.

Complications of glaucoma drainage device surgery (10% per patient-year) have included shallow anterior chamber; hypotony; suprachoroidal hemorrhage; and occlusion

of the drainage device by blood, fibrin, or iris. Long-term complications have included device erosion through the conjunctiva, valve migration, drainage device–cornea touch, corneal decompensation, and retinal detachment.

Cyclodestructive procedures to treat glaucoma may worsen ocular inflammation and lead to macular edema, hypotony, and phthisis bulbi. Although laser trabeculoplasty is generally thought to be ineffective in eyes with uveitis, it is considered a lower-risk first step in management than incisional surgery.

As with all surgical procedures in uveitic eyes, tight and meticulous control of perioperative inflammation not only helps ensure the success of glaucoma surgery but also limits sight-threatening complications such as UME and hypotony. However, when the need for glaucoma surgery is urgent, it should not be delayed because of the attempt to attain at least 3 months of controlled uveitis. Management of perioperative inflammation includes preoperative regimens similar to those applied before cataract surgery as well as use of immunomodulators and corticosteroids. See BCSC Section 10, *Glaucoma*, for additional discussion of uveitic glaucoma.

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Hypotony

Acute inflammation of the ciliary body may cause aqueous hyposecretion and low IOP. This reduction in IOP is reversible by control of intraocular inflammation. In contrast, chronic inflammation may lead to ciliary body damage and atrophy of the ciliary processes, resulting

in permanent hypotony. Low IOP may result in hypotony maculopathy, vision loss, and/or phthisis bulbi. The hypotony is often accompanied by serous choroidal detachment, which complicates management. Prolonged choroidal effusions may require surgical drainage. In some cases, chronic hypotony can be treated with long-term local corticosteroid administration. If ciliary processes are atrophic (as demonstrated on UBM), vitrectomy with intraocular silicone oil or viscoelastic may help maintain ocular anatomy and increase IOP. If the ciliary body processes are preserved and there is ciliary body traction from a cyclitic membrane, surgical removal of the membrane may be considered. In some cases, vision may improve after surgery, but these gains can be transient. In general, chronic hypotony often responds poorly to treatment, so prevention (through strict control of uveitis) is the best strategy.

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Moradi A, Stroh IG, Reddy AK, et al. Risk of hypotony in juvenile idiopathic arthritis-associated uveitis. *Am J Ophthalmol.* 2016;169:113–124.

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Uveitic Macular Edema

Uveitic macular edema (UME) is a common cause of vision loss in eyes with uveitis. The edema is usually due to active intraocular inflammation that causes retinal vascular leakage and retinal pigment epithelium dysfunction, although UME severity may not correspond to the level of inflammatory disease activity. UME appears to be mediated by the proinflammatory cytokines vascular endothelial growth factor (VEGF), tumor necrosis factor-α (TNF-α), interleukin 6, and interleukin 1. Patterns of UME on OCT include diffuse intraretinal edema, cystoid macular edema, and serous retinal detachment. Macular thickening due to mechanical vitreomacular traction is not considered UME. Fluorescein angiography should also be used to evaluate UME, as retinal vascular leakage may be more extensive than appreciated on examination and macular OCT. UME is often slow to respond to treatment and may persist even after other signs of active inflammation have resolved. Cigarette smoking appears to be associated with a greater prevalence of UME, especially in intermediate uveitis and panuveitis.

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Treatment

The initial approach to treatment of UME is to control intraocular inflammation. If UME persists after inflammation has been controlled, additional regional or systemic treatment is required. Ruling out infectious uveitis is crucial before administering local corticosteroids, especially intravitreal formulations. See Chapter 6 for additional information on the use of corticosteroids and systemic IMT to treat uveitis and UME.

Once infectious uveitis has been sufficiently investigated, periocular corticosteroid injections may be delivered into the orbital floor or the sub-Tenon space. Ultrasonography studies comparing periocular injection locations suggest that a superotemporal posterior sub-Tenon injection delivers juxtascleral corticosteroid closest to the macula. Periocular triamcinolone acetonide injections of 20–40 mg may be repeated monthly, but they are only rarely appropriate as monotherapy.

Intravitreal preservative-free triamcinolone acetonide (2–4 mg) can also be effective for reducing UME, particularly in nonvitrectomized eyes. (In vitrectomized eyes, the corticosteroid is eliminated more quickly from the vitreous cavity.) In the PeriOcular vs. INTravitreal Corticosteroids for Uveitic Macular Edema (POINT) Trial, intravitreal corticosteroids were superior to periocular sub-Tenon triamcinolone acetonide for treating UME. After intravitreal injection, visual improvement and UME reduction typically occur within 4 weeks. Risk factors for poor prognosis with intravitreal corticosteroids are chronic UME and worse visual acuity at the time of diagnosis. Intravitreal corticosteroid–induced IOP elevation may occur in up to 40% of patients, especially those younger than 40 years.

The use of implants for sustained delivery of intravitreal corticosteroid is also effective for UME, but they should be avoided in aphakic eyes. In the United States, implants include Retisert (Bausch + Lomb; fluocinolone 0.59 mg), Ozurdex (AbbVie; dexamethasone 0.7 mg), and Yutiq (EyePoint Pharmaceuticals Inc; fluocinolone 0.18 mg). In Europe, the Iluvien implant (Alimera Sciences Inc; fluocinolone 0.19 mg) is available. The risk of ocular hypertension is lower for the dexamethasone delivery system than for the fluocinolone 0.59 mg implant; in the other fluocinolone implants mentioned, lower concentrations of the corticosteroid may be associated with lower risk of IOP elevation as well.

Topical corticosteroids are usually insufficient for treating UME; however, topical difluprednate 0.05% emulsion reaches the retina and choroid at a higher concentration than topical prednisolone acetate. Small studies suggest that topical difluprednate can reduce UME; however, IOP should be monitored closely because of the risk of rapid and severe elevation.

Intravitreal anti-VEGF agents such as bevacizumab (1.25 mg in 0.05 mL) and ranibizumab (0.5 mg in 0.05 mL) are used primarily for neovascular complications of uveitis. These agents, as well as intravitreal methotrexate, may be considered as second-line treatment for UME when periocular or intravitreal corticosteroids are contraindicated or ineffective. However, a recent randomized controlled clinical trial found that the dexamethasone implant is a significantly more effective treatment for UME in comparison to intravitreal ranibizumab or intravitreal methotrexate.

Systemic IMT can be increased or added to treat UME. Agents that target specific cytokines or other proinflammatory molecules may be particularly effective. Options include TNF-α inhibitors (eg, adalimumab, infliximab), anti–interleukin-6 antibody (tocilizumab), and interferon alfa-2a/2b. See Chapter 6 for further discussion of systemic IMT.

Topical NSAIDs can be beneficial in treating pseudophakic macular edema, but their efficacy in UME has not been established. Oral acetazolamide, 500 mg once or twice daily, may also reduce UME, particularly when inflammation is otherwise well controlled.

Surgical therapy for UME is still controversial. When there is hyaloidal traction on the macula (best seen on OCT), pars plana vitrectomy may improve anatomy and vision. In the absence of vitreomacular traction, pars plana vitrectomy may be beneficial in managing recalcitrant UME; however, this application requires further investigation.

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Epiretinal Membrane and Macular Hole

Epiretinal membranes and macular holes can occur in patients with active or inactive uveitis and are often associated with substantial vision loss. In some cases, epiretinal membranes and macular holes have improved with medical management alone. Although pars plana vitrectomy and membrane peel may also benefit these patients, consensus is lacking on the optimal techniques, timing, and case selection for surgical therapy. In general, the posterior hyaloid and epiretinal membranes are more adherent and resistant to removal in uveitic eyes than in nonuveitic eyes. Standard vitreoretinal techniques are described in BCSC Section 12, *Retina and Vitreous.* Similar to the approach to uveitic cataract surgery, well-controlled preoperative and postoperative inflammation improves the chances of successful anatomical and visual outcomes with epiretinal membranes and macular holes.

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Vitreous Opacification and Vitritis

Even when uveitis activity is reduced or controlled with treatment, visually significant vitreous membranes may persist. In a small study of eyes with controlled inflammation, vitrectomy improved visual acuity in 69%. A standard small (23- to 27-gauge) 3-port pars plana vitrectomy is the preferred technique, with a few minor variations (see BCSC Section 12, *Retina and Vitreous*).

Bansal R, Dogra M, Chawla R, Kumar A. Pars plana vitrectomy in uveitis in the era of microincision vitreous surgery. *Indian J Ophthalmol.* 2020;68(9):1844–1851.
Henry CR, Becker MD, Yang Y, Davis JL. Pars plana vitrectomy for the treatment of uveitis. *Am J Ophthalmol.* 2018;190:142–149.

Rhegmatogenous Retinal Detachment

Rhegmatogenous retinal detachment (RRD) occurs in 3% of patients with uveitis. Panuveitis and infectious uveitis are most frequently associated with RRD, although pars planitis and posterior uveitis have also been associated with rhegmatogenous and tractional retinal detachments. Patients with RRD often still have active uveitis. Up to 30% may also have proliferative vitreoretinopathy on presentation, a rate that is significantly higher than in patients with primary RRD without uveitis. Repair is challenging and often complicated by preexisting proliferative vitreoretinopathy, vitreous membranes, and poor visualization. Aggressive control of inflammation is essential in the perioperative period.

Retinal detachments due to acute retinal necrosis and cytomegalovirus retinitis are also difficult to repair because of multiple, often occult, posterior retinal breaks. However, the benefits of prophylactic laser treatment in acute retinal necrosis and cytomegalovirus retinitis are unclear. Pars plana vitrectomy and endolaser treatment with internal silicone oil tamponade are most often required to repair these detachments.

De Hoog J, Ten Berge JC, Groen F, Rothova A. Rhegmatogenous retinal detachment in uveitis. *J Ophthalmic Inflamm Infect.* 2017;7(1):22. doi:10.1186/s12348-017-0140-5

Choroidal and Retinal Neovascularization

Choroidal neovascularization (CNV) complicates some uveitic entities (eg, multifocal choroiditis, punctate inner choroiditis, or serpiginous choroiditis), whereas retinal neovascularization is more likely to occur in other entities (eg, retinal vasculitis, including Eales disease). The prevalence of uveitic CNV varies; for example, it can occur in up to 10% of patients with Vogt-Koyanagi-Harada syndrome versus 69% in those with punctate inner choroiditis. Risk factors for CNV in uveitis include disruptions in the Bruch membrane from chorioretinal inflammation and the presence of inflammatory cytokines that promote angiogenesis. Symptoms include metamorphopsia and scotoma, and diagnosis is based on clinical, angiographic, and OCT findings. Treatment includes medication to reduce uveitis activity in combination with intravitreal anti-VEGF. Improved control of inflammation with local corticosteroids and/or systemic IMT may also reduce the risk of CNV recurrence and the need for repeated anti-VEGF injections.

Uveitis-associated retinal neovascularization results from chronic inflammation and/or capillary nonperfusion. Unless there is vision-obscuring vitreous hemorrhage or extensive angiographic ischemia, first-line treatment is usually focused on reducing inflammation. Treatment with panretinal photocoagulation is not necessary in all cases, especially when inflammation is controlled. For example, sarcoidosis-associated panuveitis may manifest as neovascularization of the optic disc that resolves completely with IMT and corticosteroids

alone. Intravitreal anti-VEGF injections may be used as a short-term adjunct to systemic or local treatment of inflammation and/or laser photocoagulation. Dramatic regression of neovascularization typically occurs after 1 or 2 intravitreal anti-VEGF injections. Uveitic retinal neovascularization may also resolve after posterior vitreous detachment, particularly in young patients.

Baxter SL, Pistilli M, Pujari SS, et al. Risk of choroidal neovascularization among the uveitides. *Am J Ophthalmol*. 2013;156(3):468–477.e2.

Patel AK, Newcomb CW, Liesegang TL, et al; Systemic Immunosuppressive Therapy for Eye Diseases Research Group. Risk of retinal neovascularization in cases of uveitis. *Ophthalmology.* 2016;123(3):646–654.

Woronkowicz M, Niederer R, Lightman S, Tomkins-Netzer O. Intravitreal antivascular endothelial growth factor treatment for inflammatory choroidal neovascularization in noninfectious uveitis. *Am J Ophthalmol.* 2022;236:281–287.

Vision Rehabilitation

Despite optimal treatment, inflammatory disorders of the eye may lead to vision loss. Worldwide, inflammatory disease is a major cause of blindness and low vision. In the United States, 10% of all blindness is attributed to uveitis. Clinicians can assist their patients by asking whether vision loss is affecting day-to-day functions, such as reading, or mobility. Referral to vision rehabilitation is recommended for patients with visual acuity less than 20/40 in the better eye, reduced contrast sensitivity, disabling glare, or central or peripheral visual field loss (see BCSC Section 3, *Clinical Optics and Vision Rehabilitation*). The low vision section of the American Academy of Ophthalmology website (aao.org/education/low-vision-and-vision-rehab) defines *low vision* and discusses associated symptoms, diagnosis, treatment, rehabilitation, vision aids, and how patients can identify vision rehabilitation resources in their community.

For parents of children with uveitis, clinicians are encouraged to provide information about rehabilitation to optimize functions at school and in other activities. A useful guide for teachers and parents can be found at www.uveitis.org/patients/education/patient-guides.

Fontenot JL, Bona MD, Kaleem MA, et al; American Academy of Ophthalmology Preferred Practice Pattern Vision Rehabilitation Committee. Vision Rehabilitation Preferred Practice Pattern. *Ophthalmology.* 2018;125(1):P228–P278. doi:10.1016/j.ophtha.2017.09.030