Surgical Management in Patient with Uveitis: A Literature Review

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ABSTRACT

Surgery in the treatment of uveitis can be classified according to the indication: therapeutic, diagnostic, or for the management of complications. Surgery for cataract removal, band keratopathy, corneal scarring, pupillary membrane removal, and removal of thick vitreous membranes are all examples of visual rehabilitation; Aqueous tap, vitreous biopsy, and tissue biopsy (of the iris or choroid) are used for diagnostic purposes. Complication care includes anti-glaucoma surgery, vitreous hemorrhage, retinal detachment, and persistent hypotonia. This review discusses the surgical approach for visual rehabilitation and the management of complications, including uveitis-related cataract surgery, glaucoma surgery, vitreo-retinal surgery, Pediatric uveitis surgery, Fuchs uveitis syndrome surgery, and uveitis caused by juvenile idiopathic arthritis.

Keywords: Uveitis, Surgery, Cataract Glaucoma, Vitrectomy, Children, Fuchs uveitis syndrome, Juvenile idiopathic arthritis.

INTRODUCTION

Uveitis is a condition characterized by inflammation of the uvea, which is the central pigmented layer of the eye that includes the iris, ciliary body, and choroid. In clinical practice, treating uveitis patients is difficult because patients frequently develop chronic, reoccurring inflammation that is resistant to therapy and can progress to vision-threatening consequences[1]. Clinicians can use the Standardization of Uveitis Nomenclature (SUN) standards to categorize uveitis for clinical and research purposes. With the SUN criteria, an anatomic foundation of categorization is used to split uveitis into anterior uveitis (iris and ciliary body), intermediate uveitis (vitreous), posterior uveitis (retina as well as choroid), and panuveitis (all anatomical structures are affected)[2].

The anatomic site of uveitis is critical to ascertain since the site of inflammation frequently dictates differential diagnosis and therapeutic therapy. In anterior uveitis, the anterior chamber is the predominant source of inflammation, which comprises inflammation of the iris alone (iritis), the anterior region of the ciliary body (anterior cyclitis), or both components (iridocyclitis)[3]. The vitreous is the primary location of inflammation in intermediate uveitis, which comprises the hyaloid membrane of the vitreous body (hyalitis) as well as the posterior section of the ciliary body (posterior cyclitis). Pars planitis refers to the production of a "snowbank" or "snowball" in the absence of infection or an accompanying systemic condition[4].

Inflammation of the retina, choroid, both the choroid and retina (chorio-retinitis), or the optic disc characterizes posterior uveitis (neuro-retinitis). Panuveitis is characterized by inflammation of all three components of the uvea: its anterior chamber, vitreous, choroid, and/or retina[5, 6]. The indications for surgery in the management of uveitis can be classified as:

Visual rehabilitation surgery includes cataract removal, band keratopathy, corneal scarring, pupillary membrane removal, and thick vitreous membrane removal.

Complication management includes anti-glaucoma operations, vitreous hemorrhage, detached retina, and persistent hypotonia[7].

Aqueous taps, vitreous biopsies, and tissue biopsies are all diagnostic procedures.

In uveitis, the rule for uveitis surgery is to do the procedure with a calm eye, especially if it is an elective procedure like a surgery for visual rehabilitation. At least three months must have passed since the previous episode of the disease being active before acting. While complete absence of the anterior chamber as well as vitreous cells is crucial to enhancing outcomes, this may not be really necessary or attainable in some cases of uveitis (like Fuch's uveitis syndrome). Preoperative and perioperative scaling up of corticosteroids is frequently needed, especially in more severe cases of uveitis, depending on the type[7].

Cataract Surgery for Uveitis: In individuals with chronic or recurrent uveitis, cataract is one of the most frequent consequences that results from intraocular inflammation and/or long-term use of topical and systemic corticosteroids[8]. According to the underlying etiology and site of intraocular inflammation, the risk of cataract in patients with uveitis varies; it ranges from 8.5 percent to 35 percent in a diverse set of uveitic entities[9-11].

Approximately 1.2 percent of all eyes having cataract surgery are uveitic. Cataracts are the most prevalent reason for surgical intervention in uveitic eyes and account for 40% of all occurrences of vision loss in uveitis patients[12]. Compared to non-uveitic senile cataract, the management of uveitic cataract is difficult and accompanied by less predictable surgical outcomes[13].

Cataract surgery is typically recommended for phacoantigenic uveitis in uveitis patients (regardless of visual potential and degree of inflammation); quiescent eyes with visually significant cataracts as well as good visual potential; cataracts obstructing visualization of the posterior segment for medical social management; and the potential to perform cataract surgery as part of other intraocular surgical operations, including pars plana vitrectomy or glaucoma procedures. The choice to operate is taken at the surgeon's discretion in eyes with low visual potential and a quite guarded prognosis[14]. Several studies have demonstrated that the uveitic eye remaining dormant for at least 3 months prior to surgery significantly lowers the risk of post-operative

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inflammation and cystoid macular edema, or "CME," and is therefore the most crucial factor for good visual results after cataract extraction[14, 15].

Although phacoemulsification is the preferred method due to its many benefits, extra-capsular cataract extraction as well as posterior chamber intraocular lens implantation (IOL) can occasionally be done without risk in situations where there are significant synechiae. In the majority of uveitis patients, phacoemulsification with intraocular lens implantation seems to be safe and successful with cautious patient selection, meticulous surgery, and proper postoperative care[7]. In a recent study, 67 percent of the 89 patients who had cataract extraction received extracapsular cataract surgery (ECCE) with posterior chamber intra-ocular lens implantation (PCIOL). Only 10.4 percent of patients with appropriate papillary dilatation underwent phacoemulsification (PE). There was no statistically significant difference in postoperative visual function six months after surgery in either group. Other research, however, has found that phacoemulsification outperforms ECCE. In our experience, the end outcome does not appear to differ in either procedure as long as the operative time and iris manipulations are kept to a minimum[16].

The surgeon should probably choose the procedure. In situations of idiopathic non-granulomatous anterior uveitis, posterior chamber intra-ocular lens implantation can be safely performed[17, 18]. In disorders such as Vogt-Koyanagi-Harada syndrome, autonomic ophthalmia, Behcet's disease, pars planitis, and any burnt-out or latent uveitis, moderate progress is expected. Most specialists believe IOL implantation to be contraindicated in children with juvenile idiopathic arthritis-associated uveitis because the IOL is thought to act as a scaffold for the creation of intraocular membranes, resulting in cyclitic membranes, hypotony, and eventually phthisis bulbi[19, 20].

Before intraocular lens implantation can be advised as a safe surgery in these circumstances, more experience and longer periods of follow-up are required. Bhargava and colleagues found that small incision cataract surgery (SICS) with posterior chamber intraocular lens implantation was tolerable in most uveitis cataracts and improved best corrected visual acuity (BCVA) at six months[21]. Uveitis cataract surgery problems include posterior capsular tear, zonular dehiscence, retained lens matter or nuclear fragment, hyphema, severe post-operative inflammation, cystoid macular edema, secondary glaucoma, uveitis flare-up, chronic inflammation, optic capture, and pupillary membranes, posterior capsular opacification, and hypotony[7].

Glaucoma Surgery in Uveitis: Uveitis-related increases in intraocular pressure (IOP) are known as uveitic glaucoma (UG), which causes optic nerve damage and glaucomatous loss of vision[22]. The development of posterior or peripheral anterior synechiae, the obstruction of trabecular outflow by protein, inflammatory cells, or other debris, and the use of corticosteroids to lessen inflammation are all indicators of glaucoma-causing uveitis[23]. Glaucoma in the context of uveitis is a clinical issue, both in terms of diagnosis and management. It is predicted that approximately 20% of uveitis patients may acquire secondary glaucoma. Medical therapy is beneficial but insufficient; even in such high-risk individuals, glaucoma surgery should be viewed as a vision-saving technique rather than an end-of-life procedure[7, 24].

Secondary glaucoma has multiple mechanisms:

Inflammation of the anterior segment is severe, resulting in reduced trabecular filtration[7]
Glaucoma with Pupillary Blockage (Posterior Synechia)
Angle closure because of anterior synechia on the periphery
Steroid reaction.

In the case of chronic inflammation as well as posterior synechia, the iris is really very shallow and peripheral, making pupillary block glaucoma difficult to diagnose. In such circumstances, laser iridotomy serves as both a diagnostic and therapeutic therapy[7]. Approximately 30% of uveitic glaucoma patients require surgical intervention. Glaucoma surgery success rates in uveitis patients vary. Trabeculectomy success rates in uveitic glaucoma vary from 50 to 100 percent, as do glaucoma drainage devices (GDD) success rates, which range from 57 to 94 percent[25-27].

Several factors contribute to the variety in success rates shown in prior studies. First, anti-metabolites such as 5-fluorouracil (5FU) and mitomycin C (MMC) are used[27-29]. The second aspect is the length of follow-up as well as the distinction between early and late success. Although glaucoma surgical failures can appear as early as six months and as late as five years, there is no established time frame to distinguish between the two time intervals[28]. Furthermore, several studies have shown that the success rate of filtering surgery has decreased from 80 percent on average at 2 years to 30 percent on average at 5 years, owing primarily to the patient's younger age, inflammation-mediated fibrous tissue growth, and bleb closure[27, 30].

Chawla et al. recently published a retrospective study that exhibited good long-term survival rates with 5-FU augmented trabeculectomy in patients with uveitic glaucoma and concluded that for patients over the age of 30, 5-FU enhanced trabeculectomy is recommended as first-line surgery, while a glaucoma drainage implant must be employed as the primary operation[31]. The current surgical strategy involves the use of such aqueous drainage devices as the Ahmed glaucoma valve (AGV). By channeling aqueous from the anterior chamber through such a lengthy tube to an equatorial plate, which thus promotes bleb growth, these devices provide alternate aqueous pathways[30].

In 21 uveitic glaucoma eyes, Da Mata et al. examined the safety and effectiveness of the Ahmed glaucoma valve. Among the uveitic entities examined were juvenile rheumatoid arthritis, idiopathic anterior uveitis, herpes simplex virus, HLA-B27-associated uveitis, sarcoidosis, and congenital syphilis. Surgery was performed while the uveitis was under control, despite the fact that full quiescence had been obtained[26]. Prior to surgery, oral prednisolone 1 mg/kg/day was administered until the uveitis stopped acting. Oral steroids were reduced during a 4-week period after surgery. No eyes had worsened in vision after two years, and the average intraocular pressure was 11.6 3.8 mmHg. Although anterior uveitis was present in the majority of eyes (80.9%), the high prevalence of subsequent operations (52%), which was employed in their research, may help to explain the aggressive uveitis therapy strategy that produced positive visual and clinical results[26].

Over a 4-year period, Papadaki et al. examined the long-term impact of AGV in 60 eyes with UG. Anterior uveitis affected 60% of the included eyes, panuveitis affected 35%, intermediate uveitis affected 3%, and posterior uveitis affected 1%. As necessary, the perioperative uveitis control (PUC) regimen was used. Three days before surgery, 1 mg/kg/day of oral prednisolone was tapered over 30 days postoperatively. Before surgery, 1% prednisolone acetate was applied to the skin eight times per day for two days. A combined glaucoma operation with pars plana vitrectomy, penetrating keratoplasty, intraocular lens explantation, or anterior vitrectomy was required in 43 percent of the eyes[32]. At 1 and 4 years, the control of intraocular pressure without severe complications was 57% and 39%, respectively. The study

concluded that the Ahmed glaucoma valve was relatively effective in uveitic glaucoma and that the use of glaucoma medications improved long-term success rates. Given the high rate of panuveitis and combined surgical interventions among operated eyes, tighter and longer-term uveitis control could have improved success rates[32].

Trabeculectomy with intraoperative 5-fluorouracil or mitomycin-C applications is a relatively effective therapy for patients with secondary glaucoma who have failed medical management[33]. Several procedures have been performed on uveitic patients with persistent glaucoma, including trabeculectomy, glaucoma drainage implantable devices, ciliary body destructive operations, and angle surgeries[34, 35].

Vitreo-retinal surgery in uveitis: Posterior uveitis can be focal, multifocal, or diffuse, affecting structures in the posterior segment such as the choroid, retina, retinal blood vessels, and optic nerve head. In terms of etiology, the entities can be infective or non-infective. The differential diagnosis of posterior uveitis is broad. Once the diagnosis of posterior uveitis has been confirmed, the next critical step is to determine the extent of the inflammation's involvement. A thorough diagnostic work-up is necessary, with the patient's signs and symptoms, the history of the presenting complaints, and a clinical examination serving as guides. Pars plana vitrectomy (PPV) has long been utilized to manage various types of uveitis because it allows for detailed fundus evaluation during surgery[36, 37].

In the treatment of patients with uveitis, pars plana vitrectomy is increasingly used for both diagnostic and therapeutic purposes. The indications can be divided into three categories: diagnostic pars plana vitrectomy, therapeutic pars plana vitrectomy, and both indications combined[38]. Even though epiretinal membrane (ERM) is common within uveitis, the membrane seen on ultrasound was only in the vitreous cavity due to the vitreous's dense opacity and did not involve the retina. They were unable to perform optical coherence tomography (OCT) due to the dense vitreous opacity in Muthie et al.'s case report. Optical coherence tomography can help confirm the presence of epiretinal membranes and distinguish inflammatory macular edema from that brought on by vitreomacular traction, identifying cases that are more likely to respond to surgical treatment[39].

Epiretinal membranes associated with uveitis appear to differ in cellular composition from idiopathic ERM, implying that they could emerge through a distinct pathogenic mechanism. In the case report by Muthie et al., spectral domain OCT revealed a normal retinal surface on the right eye[39, 40]. Vitrectomy is the best choice for uveitis with unusual clinical manifestations. It is also performed for uveitis that does not respond to empirical treatment with corticosteroids or immunosuppressants, as well as for rapidly progressing disease with an inconclusive non-invasive work-up or if there is a strong suspicion of malignancy[7].

To obtain the best visualization of the fundus, the vitrectomy technique employs standard three-port vitrectomy, which is preferable when performing combined therapeutic and diagnostic vitrectomy. This method also allows for better intraocular medication diffusion and yields more vitreous samples for analysis[39, 41]. Previous research has shown that vitrectomy without the 3-month inactivity rule can be successful. The use of adjunctive anti-metabolites intra-operatively as well as proper perioperative uveitis control are two ways to improve the chances of long-term success for uveitis patients undergoing surgery without the need for a 3-month inactivity[27, 29, 42, 43]. In the presence of medial opacity or hypotony, preoperative ancillary assessment may also include B-scan ultrasound imaging or ultrasonic biomicroscopy to detect underlying chorio-retinal pathological changes, including exudative retinal or choroidal disassociation, and cyclitic membranes, which may influence the surgical plan[7, 41].

Surgical Management of Uveitis in Children: Geographic, environmental, and socioeconomic variables all play a role in the etiology of uveitis in children. Certain endemic locations have documented a specific type of granulomatous anterior uveitis with a white anterior chamber granuloma. After being exposed to canal water, it usually affects youngsters in rural regions[44]. Many efforts have been made to determine the cause of such lesions. In recent investigations, molecular evidence of a digenetic trematode has been described as a source of such unusual inflammation[45, 46].

Uveitis in children is frequently misdiagnosed during standard pediatric physical examinations and is difficult to identify and treat because children are often unwilling to cooperate or disclose symptoms[47]. Some disorders, as with juvenile idiopathic arthritis (JIA)-associated uveitis, may not appear with symptoms common in adults, such as discomfort, photophobia, or redness, but rather with a white, quiet eye[1].

Complications may occur and result in visual impairment caused by structural complications in up to 34 percent of cases at the time of the first diagnosis only at the initial eye examination, with the risk of complications increasing to 69 percent of the overall cases six months after diagnosis, given the possibility for children to have a lengthy period of illness prior to the recognition of uveitis[48, 49]. Cataracts are a frequent consequence of juvenile uveitis and can impair vision because intraocular inflammation as well as topical corticosteroid usage can aggravate lens clouding. Another common anterior segment issue is band keratopathy (calcium deposits upon that cornea), which can span the central visual axis and cause vision impairment[49].

Synechiae can also arise because of ocular inflammation as peripheral anterior synechiae (adhesions from the iris to the trabecular meshwork) or posterior synechiae (adhesions from the iris to the posterior lens capsule), potentially resulting in secondary angle closure glaucoma[50]. Continuous monitoring of intraocular pressure is essential in all uveitic patients because they are predisposed to secondary glaucoma due to inflammation and persistent corticosteroid treatment. On the other hand, ciliary body shutdown can cause ocular hypotony (low intraocular pressure), resulting in poor visual results. Cystoid macular edema is a frequent problem in the posterior region that can result in diminished central visual acuity[1, 50].

When specific problems arise, medical treatment is ineffective and surgical intervention is necessary. Individuals with severe cataracts have the option of cataract surgery. However, deciding whether or not to implant an intraocular lens is a difficult decision because intraocular lens insertion may be associated with additional posterior synechiae formation, intraocular lens subluxation with pupillary seclusion, chronic inflammation, and pain if the patient's inflammation is not treated perioperatively. If cataract surgery is necessary, ophthalmologists should not proceed until at least three months have passed since intraocular inflammation has been shown to be at rest[1, 51].

In pediatric uveitis and glaucoma, many surgical treatments are reported. Angle operations, fistulizing surgical procedures, glaucoma drainage implants (GDI), as well as cyclodestructive and cyclophotocoagulation (CPC) operations, are the four types of surgical interventions. The purpose of an angle operation is to allow aqueous fluid to enter Schlemm's canal more easily. The first chamber angle surgical procedure reported is trabeculodialysis, which is conducted at the level of the Schwalbe's line[52]. At the time of goniotomy, the anterior trabecular meshwork is incised right below Schwalbe's line

with a knife or needle, while the chamber angle is directly seen with a gonioscopy lens. A goniotomy is typically conducted over 4 to 5 hours, and it can be redone (extended to more hours)[53].

Surgery is difficult due to the patients' young age and the accompanying uveitis. Due to a varied response to surgery and medicine, the requirement for general anesthesia, and the frequently difficult cooperation during pre- and post-op examinations, the young age itself is a crucial component of care. Furthermore, it is critical to manage the underlying uveitis as well as possible, as active uveitis may result in early surgery failure[54]. The compromised trabecular meshwork, along with ciliary body dysfunction induced by chronic uveitis and/or past intraocular pressure-lowering therapies, exacerbates the delicate balance between ocular hypertension and hypotony[52].

In difficult instances with a miotic pupil, the presence of cyclitic membranes, and extensive vitreous opacities, surgical procedures such as pars plana vitrectomy plus lensectomy may be explored[55]. Pars planitis may necessitate surgical intervention by pars plana vitrectomy for epiretinal membrane development, vitreous opacity, and tractional retinal detachment[56]. If medication therapy fails to manage juvenile uveitic glaucoma, goniosurgery and glaucoma draining device implantation (e.g., tube shunt) may be necessary to establish long-term intraocular pressure control and avoid glaucomatous visual neuropathy[57].

Surgical Management of Fuchs Uveitis Syndrome: Fuchs uveitis syndrome (FUS) is a persistent, generally unilateral, mild inflammatory condition that mostly affects the anterior uvea with vitreous[58]. The majority of FUS patients are diagnosed during a normal ophthalmological examination in their 3rd or 4th decade of life, regardless of gender. Fuchs uveitis syndrome often affects one eye, resulting in iris heterochromia but no ocular discomfort or pain[59-61].

Cataract and glaucoma are the most common sequelae of FUS; however, because of the success of cataract surgery, glaucoma is regarded as the more dangerous condition. In the early stages, intraocular pressure increases in these individuals are frequently intermittent and subacute, and they may react to topical corticosteroids[62, 63]. Cataract is the most prevalent complication in Fuchs uveitis eyes and has been documented to occur at significant rates of up to 90.7 percent. The most commonly used surgical treatments for uveitis patients are phacoemulsification and foldable intraocular lens implantation[64].

The correct intraocular lens materials and design are also critical factors in cataract surgery results. Several studies have been conducted to explore the biocompatibility of various intraocular lens designs and materials in uveitis patients. An implanted intraocular lens has a biological influence on the uveal and capsular regions[65, 66]. Later, Bhargava and colleagues examined the efficacy and safety of phacoemulsification as well as small incisional manual cataract surgery 'SICS' to treat cataract in Fuchs uveitis syndrome patients and discovered that both methods achieved satisfactory visual results with a low risk of complications. These studies found that SICS was a viable and cost-effective procedure for subcontinental nations with restricted access to phacoemulsification since it reduced instrumentation and surgical time[21, 67].

The Fuchs uveitis syndrome causes glaucoma, which is frequently unresponsive to medical care, laser therapy, and surgical procedures. Trabeculectomy with surgical mitomycin C is the preferred surgical method. Ahmed glaucoma valve drainage devices perform better than trabeculectomy in the treatment of glaucoma due to Fuchs uveitis syndrome[68, 69]. Several possible causes for the rise in intraocular pressure have been proposed, including recurrent hyphema, neovascularization of the angle, marginal anterior synechiae, trabeculitis, trabecular sclerosis, the breakdown of the Schlemm's canal, steroid treatment, and cataract extraction, but no single and obvious cause can usually be found[62, 70].

Few studies have described the treatment of glaucoma in Fuchs uveitis syndrome and found that medical care was insufficient. According to Liesegang's research, 66 percent of individuals with secondary glaucoma needed surgical intervention. La Hey and colleagues, as well as Jones, revealed in Fuchs uveitis syndrome that medical therapy failed in 73% and 55.5 percent of the patients, respectively[71-74].

Melamed et al. examined uveitis patient records and discovered that Fuchs uveitis syndrome was responsible for 19% of subsequent glaucoma cases. Despite medicinal and surgical treatment, one-third of patients had progressive field of vision loss as well as optic nerve damage, indicating that glaucoma is an underestimated, vision-threatening consequence in individuals with uveitis. Another study found that trabeculectomy without adjuvant antimetabolites had a 54% success rate in patients with uveitis after five years[75, 76]. Ab externo trabeculotomy has been shown to be beneficial in lowering intraocular pressure in uveitic glaucoma; nevertheless, the failure rate in individuals with FUS was much greater than in other uveitides[77].

In Towler's study, with extra intraoperative 5-fluorouracil, effective filtration was accomplished in 67 percent of eyes 5 years following trabeculectomy. According to those study findings, the cumulative likelihood of success in FUS patients who had trabeculectomy with mitomycin C was 90.9 percent in one year and 62.3 percent in four years[78, 79]. Similarly, Ye and Jiang reported 92.3 percent success in mitomycin C trabeculectomy for FUS and subsequent glaucoma; nevertheless, no evident benefits of mitomycin C over 5-fluorouracil appear. Esfandiari and colleagues examined the outcomes of Ahmed glaucoma valve implantation and primary mitomycin C in a cohort of patients with FUS-related glaucoma[80, 81].

They determined that the Ahmed glaucoma valve had a considerably greater rate of success than trabeculectomy, and that patients required fewer drugs to treat their FUS-associated glaucoma. A retrospective cohort study, on the other hand, found that trabeculectomy outperformed the Ahmed glaucoma valve in the surgical therapy of secondary glaucoma in Fuchs uveitis syndrome patients. To gain a better understanding of the operative outcome of glaucoma surgery, randomized control studies with a longer length of follow-up are required[62]. As previous research has shown, glaucoma with FUS is frequently resistant to medicinal therapy, necessitating surgical intervention to reduce the IOP. Liesegang reported adequate IOP control after one operation (usual trabeculectomy without mitomycin C) in 12 of 21 patients (57 percent)[71, 74].

La Hey and colleagues observed that after a mean follow-up of 26 months (about 2 years), surgical intervention (mainly trabeculectomies) was able to reduce intraocular pressure in 72 percent of patients[71, 72, 74].

Surgical Management of Juvenile Idiopathic Arthritis-Associated Uveitis: Juvenile idiopathic arthritis (JIA) is by far the most prevalent chronic inflammatory rheumatic disease in children and adolescence, as well as the most often associated systemic illness in pediatric uveitis[82-84]. It is a clinically separate category of persistent non-infectious idiopathic arthritides that begin before the age of 16 and continue for at least 6 weeks (about 1 and a half months)[85, 86]. Uveitis is the most common extra-articular symptom of juvenile idiopathic arthritis and has the potential to be one of the most destructive. Juvenile

idiopathic arthritis-associated uveitis (JIA-U) generally presents like an asymptomatic bilateral anterior non-granulomatous chronic uveitis of rapid onset[87-89].

However, the most prevalent sign of enthesitis-related arthritis (ERA), a subgroup of juvenile idiopathic arthritis, is acute anterior solitary or alternating uveitis with a red and sore eye[85]. In most situations, arthritis comes before uveitis, although in around 10% of cases, uveitis comes first. This condition is a significant burden in ophthalmological practice because up to 80 percent of pediatric uveitides are related to JIA, and all juvenile idiopathic arthritis patients without uveitis need frequent ophthalmological monitoring for many years[90]. Younger age upon juvenile idiopathic arthritis diagnosis, shorter period of illness, female gender, oligoarticular subtype of disease, and anti-nuclear antibody 'ANA' positivity are known risk factors for uveitis in juvenile idiopathic arthritis patients[83, 91, 92].

A recent study found that an increased erythrocyte sedimentation rate predicts the development of uveitis. In terms of the clinical history, male gender, uveitis anterior to arthritis, and the existence of ocular structural problems during the first ophthalmological screening have all been recognized as risk factors for a poor prognosis[93, 94]. In addition to medication therapy, surgical care is required to predict possible structural issues such as cataracts and glaucoma. These guidelines, however, are confined to screening, follow-up techniques, and medical care. There are very few recommendations for pediatric non-infectious non-juvenile idiopathic arthritis-associated uveitis[87, 95]. When ocular surgery is required (for example, cataract surgery, glaucoma surgery, or others), peri-operative extra therapy (usually steroids to be supplied before, during, and/or after surgery) must be carefully managed between the ophthalmologist and pediatric rheumatologist. Furthermore, systemic immunosuppression, including biologics, should not be discontinued prior to eye surgery[83].

SUMMARY

It is conceivable, and even anticipated, to obtain successful visual results after different uveitis procedures. Predictability has improved, mostly due to doctors' increased understanding of uveitis. Pre-operative variables include effective patient selection and counseling, as well as pre-operative inflammation management. Chronic inflammation, even at modest levels, can cause irreversible damage to the retina and optic nerve, making inflammatory management both pre-operatively and post-operatively essential. Cataract is a typical issue in uveitis that needs specific attention and treatment. Its therapy in uveitic eyes necessitates a thorough preoperative examination, meticulous surgical technique, and close attention to postoperative problems. After cataract extraction, good results can be obtained if peri-operative inflammation is effectively treated and close postoperative surveillance is maintained.

When compared to individuals with primary open-angle glaucoma, uveitic glaucoma patients are younger, have higher intraocular pressure levels, and may have a systemic etiology. An underlying systemic etiology should be addressed in patients with recurrent or chronic uveitis. A diagnosis requires a direct history, physical findings, and further testing. All uveitic glaucoma patients should have a gonioscopy, full blood work, and syphilis serology done, as well as a thorough eye examination. When medical therapy fails, surgical intervention is required. Age, past ocular operations, inflammatory conditions, conjunctival scars, and post-operative intraocular pressure objectives all influence surgical options. While most instances of uveitis may be treated medically, prompt referral for surgical surgery is necessary since glaucoma is the most visually damaging consequence of uveitis.

Vitrectomy is an effective treatment for vitreoretinal problems caused by uveitis; in this case, vitrectomy was advantageous for both diagnostic purposes and visual rehabilitation. The diagnosis and therapy of juvenile uveitis are complicated, and the doctor has several problems. Children might appear with no symptoms and frequently have a history of chronic and recurring illnesses.

To minimize long-term problems resulting in vision impairment, prompt identification and transfer to steroid-free immunosuppressive medication are crucial. In monitoring juvenile patients through the care of such a vision-threatening ocular condition, a multidisciplinary approach combining the ophthalmologist and rheumatologist is crucial. Early management of postoperative problems, including inflammation and glaucoma, has also led to better results. Several problems remain unsolved, particularly in the domain of pediatric uveitis with cataract, which continues to challenge ophthalmologists to perfect surgical technique and seek novel treatments.

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REFERENCES

1. Kim L, Li A, Angeles-Han S, et al. Update on the Management of Uveitis in Children: An Overview for the Clinician. Expert Rev Ophthalmol 2019; 14(4-5): 211-8.
2. Jabs DA, Nussenblatt RB , Rosenbaum JT. Standardization of Uveitis Nomenclature for Reporting Clinical Data. Results of the First International Workshop. Am J Ophthalmol 2005; 140(3): 509-16.
3. Agrawal RV, Murthy S, Sangwan V, et al. Current Approach in Diagnosis and Management of Anterior Uveitis. Indian J Ophthalmol 2010; 58(1): 9-11.
4. Babu BM , Rathinam SR. Intermediate Uveitis. Indian J Ophthalmol 2010; 58(1): 21-7.
5. Purvin V, Sundaram S , Kawasaki A. Neuroretinitis: Review of the Literature and New Observations. J Neuroophthalmol 2011; 31(1): 58-68.
6. Bansal R, Gupta V , Gupta A. Current Approach in the Diagnosis and Management of Panuveitis. Indian J Ophthalmol 2010; 58(1): 45-54.
7. Murthy SI, Pappuru RR, Latha KM, et al. Surgical Management in Patient with Uveitis. Indian J Ophthalmol 2013; 61(6): 284-90.
8. Mehta S, Linton MM , Kempen JH. Outcomes of Cataract Surgery in Patients with Uveitis: A Systematic Review and Meta-Analysis. Am J Ophthalmol 2014; 158(4): 676-92.
9. AlBloushi AF, Alfawaz AM, Al-Dahmash SA, et al. Incidence, Risk Factors and Surgical Outcomes of Cataract among Patients with Uveitis in a University Referral Hospital in Riyadh, Saudi Arabia. Ocul Immunol Inflamm 2019; 27(7): 1105-13.
10. Al Dhibi HA, Al Shamsi HN, Al-Mahmood AM, et al. Patterns of Uveitis in a Tertiary Care Referral Institute in Saudi Arabia. Ocul Immunol Inflamm 2017; 25(3): 388-95.

11. Jones NP. The Manchester Uveitis Clinic: The First 3000 Patients, 2: Uveitis Manifestations, Complications, Medical and Surgical Management. Ocul Immunol Inflamm 2015; 23(2): 127-34.
12. Chu CJ, Dick AD, Johnston RL, et al. Cataract Surgery in Uveitis: A Multicentre Database Study. Br J Ophthalmol 2017; 101(8): 1132-7.
13. Kim WS , Kim KH. Challenges in Cataract Surgery. 1 ed. Berlin, Heidelberg: Springer, 2016
14. Rojas B, Zafirakis P , Foster CS. Cataract Surgery in Patients with Uveitis. Curr Opin Ophthalmol 1997; 8(1): 6-12.
15. Bélair ML, Kim SJ, Thorne JE, et al. Incidence of Cystoid Macular Edema after Cataract Surgery in Patients with and without Uveitis Using Optical Coherence Tomography. Am J Ophthalmol 2009; 148(1): 128-35.
16. Hazari A , Sangwan VS. Cataract Surgery in Uveitis. Indian J Ophthalmol 2002; 50(2): 103-7.
17. Javadi MA, Jafarinasab MR, Araghi AA, et al. Outcomes of Phacoemulsification and in-the-Bag Intraocular Lens Implantation in Fuchs' Heterochromic Iridocyclitis. J Cataract Refract Surg 2005; 31(5): 997-1001.
18. Harper SL , Foster CS. Intraocular Lens Explantation in Uveitis. Int Ophthalmol Clin 2000; 40(1): 107-16.
19. Ganesh SK, Babu K , Biswas J. Phacoemulsification with Intraocular Lens Implantation in Cases of Pars Planitis. J Cataract Refract Surg 2004; 30(10): 2072-6.
20. BenEzra D , Cohen E. Cataract Surgery in Children with Chronic Uveitis. Ophthalmology 2000; 107(7): 1255-60.
21. Bhargava R, Kumar P, Bashir H, et al. Manual Suture Less Small Incision Cataract Surgery in Patients with Uveitic Cataract. Middle East Afr J Ophthalmol 2014; 21(1): 77-82.
22. Neri P, Azuara-Blanco A , Forrester JV. Incidence of Glaucoma in Patients with Uveitis. J Glaucoma 2004; 13(6): 461-5.
23. Kalogeropoulos D , Sung VC. Pathogenesis of Uveitic Glaucoma. J Curr Glaucoma Pract 2018; 12(3): 125-38.
24. Takahashi T, Ohtani S, Miyata K, et al. A Clinical Evaluation of Uveitis-Associated Secondary Glaucoma. Jpn J Ophthalmol 2002; 46(5): 556-62.
25. Kalin-Hajdu E, Hammamji K, Gagné S, et al. Outcome of Viscodilation and Tensioning of Schlemm's Canal for Uveitic Glaucoma. Can J Ophthalmol 2014; 49(5): 414-9.
26. Da Mata A, Burk SE, Netland PA, et al. Management of Uveitic Glaucoma with Ahmed Glaucoma Valve Implantation. Ophthalmology 1999; 106(11): 2168-72.
27. Towler HM, Bates AK, Broadway DC, et al. Primary Trabeculectomy with 5-Fluorouracil for Glaucoma Secondary to Uveitis. Ocul Immunol Inflamm 1995; 3(3): 163-70.
28. Magliyah MS, Badawi AH, Alshamrani AA, et al. The Effect of Perioperative Uveitis Control on the Success of Glaucoma Surgery in Uveitic Glaucoma. Clin Ophthalmol 2021; 15: 1465-75.
29. Patitsas CJ, Rockwood EJ, Meisler DM, et al. Glaucoma Filtering Surgery with Postoperative 5-Fluorouracil in Patients with Intraocular Inflammatory Disease. Ophthalmology 1992; 99(4): 594-9.
30. Rachmiel R, Trope GE, Buys YM, et al. Ahmed Glaucoma Valve Implantation in Uveitic Glaucoma Versus Open-Angle Glaucoma Patients. Can J Ophthalmol 2008; 43(4): 462-7.
31. Chawla A, Mercieca K, Fenerty C, et al. Outcomes and Complications of Trabeculectomy Enhanced with 5-Fluorouracil in Adults with Glaucoma Secondary to Uveitis. J Glaucoma 2013; 22(8): 663-6.
32. Papadaki TG, Zacharopoulos IP, Pasquale LR, et al. Long-Term Results of Ahmed Glaucoma Valve Implantation for Uveitic Glaucoma. Am J Ophthalmol 2007; 144(1): 62-9.
33. Jad A, Céline T, Bahram B, et al. Fuchs' Heterochromic Cyclitis: A Post-Infectious Manifestation of Ocular Toxoplasmosis? Int Ophthalmol 2013; 33(2): 189-94.
34. Miserocchi E, Carassa RG, Bettin P, et al. Viscocanalostomy in Patients with Glaucoma Secondary to Uveitis: Preliminary Report. J Cataract Refract Surg 2004; 30(3): 566-70.
35. Ceballos EM, Parrish RK, 2nd , Schiffman JC. Outcome of Baerveldt Glaucoma Drainage Implants for the Treatment of Uveitic Glaucoma. Ophthalmology 2002; 109(12): 2256-60.
36. Sato T, Kinoshita R, Taguchi M, et al. Assessment of Diagnostic and Therapeutic Vitrectomy for Vitreous Opacity Associated with Uveitis with Various Etiologies. Medicine (Baltimore) 2018; 97(2):1-9.
37. Kaza H, Modi R, Rana R, et al. Effect of Adjunctive Pars Plana Vitrectomy on Focal Posterior Segment Inflammation: A Case-Control Study in Tuberculosis-Associated Uveitis. Ophthalmol Retina 2018; 2(11): 1163-9.
38. Gupta A, Bansal R, Gupta V, et al. Ocular Signs Predictive of Tubercular Uveitis. Am J Ophthalmol 2010; 149(4): 562-70.
39. Muthie FA, Widjaja SA, Sasono W, et al. Surgical Management on Posterior Uveitis with Vitreous Opacity Suspected Retinal Detachment. International Journal of Retina 2020; 3(1):1-9.
40. Nicholson BP, Zhou M, Rostamizadeh M, et al. Epidemiology of Epiretinal Membrane in a Large Cohort of Patients with Uveitis. Ophthalmology 2014; 121(12): 2393-8.
41. Vitale AT. Vitrectomy in Patients with Uveitis: Review of opthlamology; 2006 [Available from: https://www.reviewofophthalmology.com/article/vitrectomy-in-patients-with-uveitis.
42. Quinones K, Choi JY, Yilmaz T, et al. Pars Plana Vitrectomy Versus Immunomodulatory Therapy for Intermediate Uveitis: A Prospective, Randomized Pilot Study. Ocul Immunol Inflamm 2010; 18(5): 411-7.
43. Giuliari GP, Chang PY, Thakuria P, et al. Pars Plana Vitrectomy in the Management of Paediatric Uveitis: The Massachusetts Eye Research and Surgery Institution Experience. Eye (Lond) 2010; 24(1): 7-13.
44. Amin RM, Goweida M, Bedda A, et al. Clinical Patterns and Causes of Intraocular Inflammation in a Uveitis Patient Cohort from Egypt. Ocul Immunol Inflamm 2019; 27(6): 859-67.
45. Amin RM, Radwan AE, Goweida MB, et al. Management of Presumed Trematode Induced Granulomatous Uveitis in Pediatric Patients. Jpn J Ophthalmol 2019; 63(1): 119-25.
46. Amin RM, Goweida MB, El Goweini HF, et al. Trematodal Granulomatous Uveitis in Paediatric Egyptian Patients: A Case Series. Br J Ophthalmol 2017; 101(8): 999-1002.
47. Wentworth BA, Freitas-Neto CA , Foster CS. Management of Pediatric Uveitis. F1000Prime Rep 2014; 6:1-4.
48. Majumder PD , Biswas J. Pediatric Uveitis: An Update. Oman J Ophthalmol 2013; 6(3): 140-50.
49. Rosenberg KD, Feuer WJ , Davis JL. Ocular Complications of Pediatric Uveitis. Ophthalmology 2004; 111(12): 2299-306.
50. Smith JA, Mackensen F, Sen HN, et al. Epidemiology and Course of Disease in Childhood Uveitis. Ophthalmology 2009; 116(8): 1544-51
51. Quiñones K, Cervantes-Castañeda RA, Hynes AY, et al. Outcomes of Cataract Surgery in Children with Chronic Uveitis. J Cataract Refract Surg 2009; 35(4): 725-31.
52. van Meerwijk C, Jansonius NM , Los LI. Uveitic Glaucoma in Children: A Systematic Review on Surgical Outcomes. J Ophthalmic Inflamm Infect 2022; 12(1):1-5.
53. Freedman SF, Rodriguez-Rosa RE, Rojas MC, et al. Goniotomy for Glaucoma Secondary to Chronic Childhood Uveitis. Am J Ophthalmol 2002; 133(5): 617-21.

54. Chaku M, Bajwa A, Lee JK, et al. Treatment of Uveitis and Outcomes of Glaucoma Drainage Implant Surgery: A Meta-Analysis. Ocul Immunol Inflamm 2020; 28(5): 833-8.
55. Yangzes S, Seth NG, Singh R, et al. Long-Term Outcomes of Cataract Surgery in Children with Uveitis. Indian J Ophthalmol 2019; 67(4): 490-5.
56. Kurokawa T, Kikuchi T, Ohta K, et al. Ocular Manifestations in Blau Syndrome Associated with a Card15/Nod2 Mutation. Ophthalmology 2003; 110(10): 2040-4.
57. Bohnsack BL , Freedman SF. Surgical Outcomes in Childhood Uveitic Glaucoma. Am J Ophthalmol 2013; 155(1): 134-42.
58. Kreps EO, Derveaux T, De Keyser F, et al. Fuchs' Uveitis Syndrome: No Longer a Syndrome? Ocul Immunol Inflamm 2016; 24(3): 348-57.
59. Accorinti M, Spinucci G, Pirraglia MP, et al. Fuchs' Heterochromic Iridocyclitis in an Italian Tertiary Referral Centre: Epidemiology, Clinical Features, and Prognosis. J Ophthalmol 2016; 2016:1-4.
60. Cunningham ET, Jr. , Baglivo E. Fuchs Heterochromic Iridocyclitis--Syndrome, Disease, or Both? Am J Ophthalmol 2009; 148(4): 479-81.
61. Norrsell K , Sjödell L. Fuchs' Heterochromic Uveitis: A Longitudinal Clinical Study. Acta Ophthalmol 2008; 86(1): 58-64.
62. Nilforushan N, Yadgari M , Alemzadeh SA. Surgical Management of Glaucoma in Fuchs Uveitis Syndrome: Trabeculectomy or Ahmed Glaucoma Valve. J Curr Ophthalmol 2019; 31(1): 24-30.
63. La Hey E, de Jong PT , Kijlstra A. Fuchs' Heterochromic Cyclitis: Review of the Literature on the Pathogenetic Mechanisms. Br J Ophthalmol 1994; 78(4): 307-12.
64. Tugal-Tutkun I, Güney-Tefekli E, Kamaci-Duman F, et al. A Cross-Sectional and Longitudinal Study of Fuchs Uveitis Syndrome in Turkish Patients. Am J Ophthalmol 2009; 148(4): 510-5.
65. Özdamar Erol Y, Özdemir Yalçınsoy K , Özdal P. The Outcomes of Cataract Surgery in Eyes with Fuchs Uveitis. J Ophthalmic Inflamm Infect 2023; 13(1): 1-4.
66. Abela-Formanek C, Amon M, Schauersberger J, et al. Results of Hydrophilic Acrylic, Hydrophobic Acrylic, and Silicone Intraocular Lenses in Uveitic Eyes with Cataract: Comparison to a Control Group. J Cataract Refract Surg 2002; 28(7): 1141-52.
67. Bhargava R, Kumar P, Sharma SK, et al. Phacoemulsification Versus Manual Small Incision Cataract Surgery in Patients with Fuchs Heterochromic Iridocyclitis. Asia Pac J Ophthalmol (Phila) 2016; 5(5): 330-4.
68. Voykov B, Doycheva D, Deuter C, et al. Outcomes of Ahmed Glaucoma Valve Implantation for Glaucoma Secondary to Fuchs Uveitis Syndrome. Ocul Immunol Inflamm 2017; 25(6): 760-6.
69. Kok H , Barton K. Uveitic Glaucoma. Ophthalmol Clin North Am 2002; 15(3): 375-87.
70. Velilla S, Dios E, Herreras JM, et al. Fuchs' Heterochromic Iridocyclitis: A Review of 26 Cases. Ocul Immunol Inflamm 2001; 9(3): 169-75.
71. Muñoz-Negrete FJ, Moreno-Montañés J, Hernández-Martínez P, et al. Current Approach in the Diagnosis and Management of Uveitic Glaucoma. Biomed Res Int 2015; 2015: 1-9.
72. La Hey E, de Vries J, Langerhorst CT, et al. Treatment and Prognosis of Secondary Glaucoma in Fuchs' Heterochromic Iridocyclitis. Am J Ophthalmol 1993; 116(3): 327-40.
73. Jones NP. Glaucoma in Fuchs' Heterochromic Uveitis: Aetiology, Management and Outcome. Eye (Lond) 1991; 5 (Pt 6): 662-7.
74. Liesegang TJ. Clinical Features and Prognosis in Fuchs' Uveitis Syndrome. Arch Ophthalmol 1982; 100(10): 1622-6.
75. Stavrou P , Murray PI. Long-Term Follow-up of Trabeculectomy without Antimetabolites in Patients with Uveitis. Am J Ophthalmol 1999; 128(4): 434-9.
76. Melamed S, Lahav M, Sandbank U, et al. Fuch's Heterochromic Iridocyclitis: An Electron Microscopic Study of the Iris. Invest Ophthalmol Vis Sci 1978; 17(12): 1193-9.
77. William A, Spitzer MS, Doycheva D, et al. Comparison of Ab Externo Trabeculotomy in Primary Open-Angle Glaucoma and Uveitic Glaucoma: Long-Term Outcomes. Clin Ophthalmol 2016; 10: 929-34.
78. You YA, Wu Y , Hu S. Surgical Management of Secondary Glaucoma in Fuchs' Heterochromic Iridocyclitis. Graefes Arch Clin Exp Ophthalmol 2013; 251(7): 1785-90.
79. Towler HM, McCluskey P, Shaer B, et al. Long-Term Follow-up of Trabeculectomy with Intraoperative 5-Fluorouracil for Uveitis-Related Glaucoma. Ophthalmology 2000; 107(10): 1822-8.
80. Esfandiari H, Loewen NA, Hassanpour K, et al. Fuchs Heterochromic Iridocyclitis-Associated Glaucoma: A Retrospective Comparison of Primary Ahmed Glaucoma Valve Implantation and Trabeculectomy with Mitomycin C. F1000Res 2018; 7:1-8.
81. Ye CH , Jiang YQ. Long-Term Effects of Trabeculectomy on Fuchs' Heterochromic Uveitis with Secondary Glaucoma. Chin J Pract Ophthalmol 2000; 18: 548-50.
82. Jari M, Shiari R, Salehpour O, et al. Epidemiological and Advanced Therapeutic Approaches to Treatment of Uveitis in Pediatric Rheumatic Diseases: A Systematic Review and Meta-Analysis. Orphanet J Rare Dis 2020; 15(1):1-8.
83. Heiligenhaus A, Minden K, Tappeiner C, et al. Update of the Evidence Based, Interdisciplinary Guideline for Anti-Inflammatory Treatment of Uveitis Associated with Juvenile Idiopathic Arthritis. Semin Arthritis Rheum 2019; 49(1): 43-55.
84. Mastrangelo G, Foeldvari I, Anton J, et al. Defining Outcome Measures in Juvenile Idiopathic Arthritis Associated Uveitis by a Systematic Review Analysis: Do We Need a Consensus? Pediatr Rheumatol Online J 2019; 17(1):1-11.
85. Gueudry J, Touhami S, Quartier P, et al. Therapeutic Advances in Juvenile Idiopathic Arthritis - Associated Uveitis. Curr Opin Ophthalmol 2019; 30(3): 179-86.
86. Ferrara G, Mastrangelo G, Barone P, et al. Methotrexate in Juvenile Idiopathic Arthritis: Advice and Recommendations from the Marajia Expert Consensus Meeting. Pediatr Rheumatol Online J 2018; 16(1): 1-6.
87. Asproudis I, Katsanos A, Kozeis N, et al. Update on the Treatment of Uveitis in Patients with Juvenile Idiopathic Arthritis: A Review. Adv Ther 2017; 34(12): 2558-65.
88. Okamoto N, Yokota S, Takei S, et al. Clinical Practice Guidance for Juvenile Idiopathic Arthritis (Jia) 2018. Mod Rheumatol 2019; 29(1): 41-59.
89. Angeles-Han ST, Lo MS, Henderson LA, et al. Childhood Arthritis and Rheumatology Research Alliance Consensus Treatment Plans for Juvenile Idiopathic Arthritis-Associated and Idiopathic Chronic Anterior Uveitis. Arthritis Care Res (Hoboken) 2019; 71(4): 482-91.
90. Hawkins MJ, Dick AD, Lee RJ, et al. Managing Juvenile Idiopathic Arthritis-Associated Uveitis. Surv Ophthalmol 2016; 61(2): 197-210.
91. Choi J, Hawley DP, Ashworth J, et al. An Update on the Modern Management of Paediatric Uveitis. Br J Ophthalmol 2019; 103(12): 1685-9.
92. Campanilho-Marques R, Bogas M, Ramos F, et al. Prognostic Value of Antinuclear Antibodies in Juvenile Idiopathic Arthritis and Anterior Uveitis. Results from a Systematic Literature Review. Acta Reumatol Port 2014; 39(2): 116-22.
93. Tappeiner C, Klotsche J, Sengler C, et al. Risk Factors and Biomarkers for the Occurrence of Uveitis in Juvenile Idiopathic Arthritis: Data from the Inception Cohort of Newly Diagnosed Patients with Juvenile Idiopathic Arthritis Study. Arthritis Rheumatol 2018; 70(10): 1685-94.

94. Angeles-Han ST , Rabinovich CE. Uveitis in Children. Curr Opin Rheumatol 2016; 28(5): 544-9.

95. Wells JM , Smith JR. Uveitis in Juvenile Idiopathic Arthritis: Recent Therapeutic Advances. Ophthalmic Res 2015; 54(3): 124-7.