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Major review

The Ocular Immunology and Uveitis Foundation preferred practice patterns of uveitis management

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ABSTRACT

Ocular inflammatory disease is a leading cause of vision loss worldwide. Uveitis encompasses a wide spectrum of pathology, both with respect to its etiology and the anatomic location within the eye. Inflammation can be confined to the eye and may also be seen systemically. The cornerstone of management of ocular inflammatory disease historically has been corticosteroids, which are invaluable in the immediate control of inflammation; however, corticosteroids are inappropriate for long-term use as they are associated with a wide array of toxic side effects. As we continue to learn more about the various etiologies and elucidate the basic science pathways and mechanisms of action that cause intraocular inflammation, new therapeutic approaches have evolved. They include employment of immunomodulatory agents (corticosteroid-sparing therapies) that have expanded our treatment options for these vision-threatening diseases. These pharmacologics provide therapy for ocular and systemic inflammation in an individualized, patient-tailored, stepladder approach with the ultimate goal of durable, corticosteroid-free remission. We review the preferred practice patterns of a tertiary care center specializing in ocular inflammatory disease.

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The protocols presented in the above tables (Table 8, specifically) are specific to the practice of Dr. C. Stephen Foster and are not intended to suggest that they represent protocols in common usage.

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1. Global impact on vision

Intraocular inflammatory disorders encompass a broad spectrum of disease in which the eye or its various parts may be attacked by the immune system, leading to severe visual impairment. Uveitis is the third leading cause of worldwide blindness and currently accounts for approximately 10% of preventable vision loss in the US and up to 15% worldwide.[10,21,24,47,63] In the US alone, uveitis has an estimated prevalence of about 38 cases per 100,000 and an incidence of 15 cases per 100,000.[24] An estimated more than 2 million people worldwide have uveitis. Uveitis may affect individuals of any age, from infancy to adulthood. A large, US population-based study, the Northern California Epidemiology of Uveitis Study (NCEUS)[24] reported a 3-fold increase in the incidence of uveitis, as compared to previous estimates, in addition to an increased incidence due to an overall aging of the population. There is a higher incidence of uveitis in women than men, and the largest differences were seen in older age groups.[24]

2. Uveitis etiology: Noninfectious and infectious

Several uveitis classifications schemes exist. They vary based on the anatomic location of inflammation, clinical course, etiology, and histopathology. The Standardization of Uveitis Nomenclature (SUN) Working Group developed an anatomic classification system in 2005 that arguably serves as the most widely used today.[28] The following are types of uveitis based on anatomic location (Table 1).

(1) Anterior uveitis
(2) Intermediate uveitis
(3) Posterior uveitis
(4) Panuveitis

Anterior uveitis has the highest prevalence, followed by panuveitis, then posterior and intermediate uveitis. In addition, the SUN Working Group recommended consistent terminology for grading uveitic activity, whereby anterior chamber cell, and flare, as well as vitreous haze are graded on an escalating 0 to 4 severity scale (Tables 2–7). Although a consensus has not been reached on the inclusion of vitreous cell or retinal vasculitis in the classification system or individual grading schemes, most uveitis experts will describe the degree to which these features are seen in the same fashion.

In the posterior segment, uveitis, macular edema, retinal vasculitis, retinal detachments, and optic neuropathy contribute to loss of vision. Intermediate uveitis, posterior uveitis, and panuveitis are responsible for most visual disability in patients with ocular inflammatory disease. The other sight threatening complications of uveitis include phthisis bulbi, hypotony,[8,48] band keratopathy, glaucoma, and retinopathy. The etiologic distribution of uveitis varies around the globe. In general, anterior uveitis is most often idiopathic, whereas an infectious etiology is more common among patients with posterior uveitis. Generally, infectious entities of uveitis carry a poorer overall prognosis than the noninfectious posterior uveitides.[54]

Table 1 – SUN Working Group anatomic classification of uveitis[6]

Type	Primary site of inflammation	Includes
Anterior	Anterior chamber	Iritis Iridocyclitis Anterior cyclitis
Intermediate	Vitreous	Pars planitis Posterior cyclitis Hyalitis
Posterior	Retina or choroid	Focal, multifocal, or diffuse choroiditis Chorioretinitis Retinochoroiditis Retinitis Neuroretinitis
Panuveitis	Anterior chamber, vitreous, and retina or choroid	

Table 2 – SUN Working Group grading scheme for anterior chamber cells[6]

Grade	Cells in field[a]
0	<1
0.5+	1–5
1+	6–15
2+	16–25
3+	26–50
4+	>50

a Field size is a 1 mm by 1 mm slit beam.

3. Medical management of uveitis

Therapy for uveitis may appropriately involve medical or surgical intervention. The preferred practice pattern that we advocate in the care of patients with uveitis is aimed at cure. The first stage in the successful pursuit of cure is the induction of durable, corticosteroid-free remission—and doing whatever it takes to accomplish that goal—whereas at the same time doing no harm, not producing quality of life-altering side effects or complications from the strategies used in that quest for durable remission.

Table 3 – SUN Working Group grading scheme for anterior chamber flare[6]

Grade	Description
0	None
1+	Faint
2+	Moderate (iris and lens details clear)
3+	Marked (iris and lens details hazy)
4+	Intense (fibrin or plastic aqueous)

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