# The medical and surgical management of chronic intermediate uveitis (pars planitis)

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Inflammation of the intermediate uvea has been referred to by many different names — for example, pars planitis, chronic cyclitis and peripheral uveitis [1-3]. It is a disease which characteristically affects individuals of both sexes, frequently starting in the second and third decades.

Often, the earliest symptoms consist of floaters, blurred vision and frequent loss of accommodation. The effected eye is comfortable, without evidence of external inflammation, although a mild anterior iritis may be present. However, within the anterior vitreous body are characteristic inflammatory cellular clumps termed "snowballs". As the disease progresses, a gelatinous exudate is noted over the pars plana, with frequent invasion by neovascularization from the peripheral retina. The adjacent peripheral fundus may show perivascular sheathing, particularly of the retinal veins.

Histopathologic examination of intermediate uveitis has presented two contrasting patterns. Kimura and Hogan [4] observed the presence of chronic uveitis with an inflammatory infiltrate consisting primarily of lymphocytes and plasma cells. In contrast, Kenyon and colleagues [5] noted the presence of retinal phlebitis and fibroglial proliferation at the vitreous base. The clinical presentation of retinal periphlebitis and neovascularization extending into the pars plana exudate is certainly consistent with the latter observation.

The natural history of intermediate uveitis is resolution within five years in the majority of cases, although occasional persistence beyond ten years has been documented. The major vision-threatening complications from persistent inflammation are the development of cataract and macular degeneration from cystoid macular edema. Nevertheless, Smith and colleagues [6], in a review of patients with intermediate uveitis at the Proctor Foundation, commented that 75% of patients maintained good visual acuity 12 years after the onset of their disease. Despite this observation, extended duration of disease appears to be associated with a poorer prognosis for vision. Retinal complications associated with pars planitis, in addition to cystoid macular edema, are retinoschisis, retinal tear with or without vitreous hemorrhage and optic disc edema. Although unusual, retinal detachment, glaucoma and phthisis bulbi have been reported.

Since the disease is frequently limited in duration, primary attention must be given to prevention of visually disabling complications until a natural quiescence is encountered. A four-step scheme to treatment of patients with intermediate uveitis has been employed at Emory Clinical, with very satisfactory results (Figure 1).

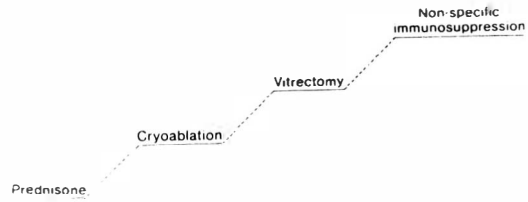


Figure 1. A four-step scheme to treat patients with intermediate uveitis with either a reduced visual acuity or the development of a secondary cataract.

The first step in the treatment of intermediate uveitis involves the use of systemic or periocular corticosteroids for immunosuppression. Although the disease may be present and mildly symptomatic, unless visual acuity is reduced to or less than 20/30 or a secondary cataract is noted, treatment is not instituted. If the disease has a symmetrical presentation, then systemic prednisone (80 mg) in four daily divided doses is started. High dosage systemic prednisone is continued for two weeks, at which time the dose is altered to a single morning administration. Subsequently, medication is given every other day and slowly tapered to maintain a favorable therapeutic response. If unilateral involvement is predominant, then periocular triamcinolone acetonide (40mg) is administered in lieu of systemic prednisone. Repeat periocular injections are given at two- or

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three-week intervals depending upon the therapeutic response.

If intolerance or unresponsiveness to corticosteroid therapy is encountered, the second step of the therapeutic scheme is performed, namely, cryoablation. Aaberg and colleagues [7] reported a favorable response to cryoablation in the area of involvement in intermediate uveitis. A single freeze-thaw technique is employed by us, with inclusion of the area of intermediate uveitis and one row of treatment anterior and posterior to the involved area. Repeated freeze-thaw applications can result in an exudative retinal detachment in response to treatment, but this appears to be minimized by the single freeze-thaw technique. A favorable response to treatment is seen in the majority of patients who undergo cryoablation. The pars plana exudate will resolve in the area of treatment, but may recur anterior or posterior to the cryotherapy scar. Unfortunately, the duration of response is frequently limited to a period of six months to two years. Thus, cryoablation represents a temporizing modality of treatment while awaiting natural regression of the disease.

With future recurrences of intermediates uveitis, or with complications secondary to peripheral vitreous traction (i.e., retinal tear and/or vitreous hemorrhage), pars plana vitrectomy is performed as a third step. Most patients with chronic inflammation have a total posterior vitreous detachment, so that a total vitrectomy can be performed without difficulty. The primary surgical objective is the release of peripheral vitreous traction to prevent the subsequent development of retinal tears and/or vitreous hemorrhage. However, for reasons which are not yet clear, the removal of inflamed vitreous in such eyes will frequently result in the resolution of visually disabling cystoid macular edema and a decrease in the severity and frequency of subsequent recurrent inflammation.

If pars plana vitrectomy is not successful in maintaining satisfactory visual acuity, the fourth and final step in the scheme of treatment is non-specific immunosuppression [8]. Until recently, a first generation immunosuppressor such as cyclophosphamide was the mainstay of treatment. However, the well-known bone marrow toxicity of such medication, as well as the possible increased incidence of subsequent malignant disease, has tempered our enthusiasm. Cyclosporine, a second generation antigen-nonspecific immunosuppressor, has recently been utilized to achieve similar objectives. Although this medication is not associated with bone marrow toxicity, it does have both renal and hepatic toxicity, which are reversible upon discontinuation of the medication. Furthermore, initial dosage regimens with cyclosporine were associated with an unacceptably high occurrence of Epstein-Barr virus (EBV)-induced lymphoma. Subsequent decrease in the dosage has dramatically reduced the incidence of such bone marrow neoplasms. Nevertheless, impressive experimental evidence exists to suggest that cyclosporine interferes with the host's natural immunity to EBV infection. Thus, the possible effects of chronic treatment with reduced levels of this medication will warrant careful observation.

With a four-step scheme of treatment as outlined, the overwhelming majority of patients with intermediate uveitis can retain good visual acuity until there is a natural resolution of their disease. It is imperative that recurrent episodes of inflammation not be allowed to cause permanent visual damage. Thus, frequent and diligent attention to patients with this disease is required to insure their eventual good prognosis.

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