

## Retinal Vasculitis

A Patient Education Monograph prepared for the American Uveitis Society

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by Lynn K. Gordon, M.D., Ph.D.

Jules Stein Eye Institute

University of California, Los Angeles

Los Angeles, CA, USA

**NOTE:** The opinions expressed in this monograph are those of the author(s) and not necessarily those of the membership of the American Uveitis Society, its leadership, or the Editorial Board of UveitisSociety.org. All medical decisions should be made in consultation with one's personal physician.

### Introduction

Retinal vasculitis is an [inflammatory](#) disease of the blood vessels of the [retina](#) that may be associated with primary ocular conditions or with inflammatory or infectious diseases in other parts of the body (systemic diseases). The most common systemic diseases associated with retinal vasculitis are Behçet's disease, sarcoidosis, and multiple sclerosis. Associations have also been noted with Wegener's granulomatosis, systemic lupus erythematosus, polyarteritis nodosa and other rheumatologic conditions. Infectious agents may produce a retinal vasculitis, including bacteria (for example syphilis or tuberculosis), viruses (for example herpes family viruses), and parasites (including *Toxocara canis*). Primary ocular causes of retinal vasculitis include Eales disease, pars planitis, birdshot retinochoroidopathy, and Fuchs uveitis syndrome.

### Course of Disease

The clinical symptoms of retinal vasculitis include blurred vision, altered color perception, metamorphopsia (distortion of images, especially straight lines), floaters, and scotomas (blind spots). Some cases may occur without visual symptoms, for example in multiple sclerosis the vasculitis may be discovered as an incidental finding on examination.

Retinal examination typically reveals sheathing (a whitish-yellow cuff of material surrounding the blood vessel) of the affected retinal vasculature associated with variable vitritis (inflammatory cells behind the lens in the vitreous body). Narrowing of the retinal blood vessels, bleeding in the back of the eye, and new blood vessel growth are commonly observed. A mild anterior uveitis may be seen in patients with retinal vasculitis. Choroidal inflammation, when present, may suggest a diagnosis of sarcoidosis or a specific uveitis syndrome such as birdshot retinochoroidopathy or presumed ocular histoplasmosis syndrome.

### Diagnosis and Testing

The majority of patients with isolated retinal vasculitis do not have an associated serious systemic disease. However, it is critical to fully evaluate each patient. A careful history and

physical examination followed by targeted laboratory and radiological testing are performed. Most evaluations will include blood tests, urinalysis, chest x-ray, and possibly fluorescein angiography, which often reveals leakage of the affected retinal blood vessels and may further demonstrate areas of discrete occlusion of the retinal vessels, retinal edema, neovascularization, and choroidal involvement.

### **Therapy**

Therapy is dictated by the patient's signs and symptoms. If inflammation is mild, vision is good, and there is no evidence by fluorescein angiography for extensive retinal involvement, then observation, corticosteroid eye drops, or periocular corticosteroid injection may be indicated. Systemic therapy (oral or intravenous medication) is indicated in severe cases, when there is loss of visual function, evidence of macular edema, or widespread aggressive retinal disease. Once control of the disease is achieved, a gradual taper of the corticosteroid is initiated. Corticosteroid therapy is discussed in detail elsewhere on this website. Other agents such as azathioprine, methotrexate, and cyclosporine can be used in refractory patients or as steroid-sparing agents. Careful monitoring of associated complications of this therapy must be performed.

Surgical therapies such as laser photocoagulation or vitrectomy are generally not indicated except in the management of persistent neovascularization (new vessel growth) or in cases of bleeding into the vitreous or glaucoma.

### **Prognosis**

The prognosis for patients with retinal vasculitis is variable. Many patients may undergo a relatively benign course or will have a dramatic response to systemic immunosuppressive therapy and will have both preservation of vision and visual function. Other patients are more resistant to therapy and despite the use of aggressive treatment will have permanent loss of function. It is difficult to predict the clinical course for an individual patient at the time of disease onset.

### **Research and Future Outlook**

There is significant interest in the potential utility of biologic agents in the therapy of immune-mediated diseases and there is significant hope for future novel therapies for retinal vasculitis.