



THE OCULAR IMMUNOLOGY  
AND UVEITIS FOUNDATION  
*Dedicated to Eye Disease Cure and Education*

## **Ocular Autoimmune Disease: An Introduction**

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The immune system, which ordinarily functions to protect the body from harm, including infections and cancer cells, can become dysregulated, resulting in an immune attack against the body's own tissues. This state of affairs is termed autoimmunity, or immune attack against self.

There are numerous autoimmune diseases, among the most well-known being rheumatoid arthritis. In rheumatoid arthritis the white blood cells of the immune system become dysregulated or "confused" and begin to attack the individual's joints. A number of autoimmune diseases exist in which various parts of the eye may be attacked by the white blood cells. More commonly, the autoimmune disease is systemic; that is, a variety of organs throughout the body are affected by the immune attack. Examples of such diseases include rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa, relapsing polychondritis, granulomatosis with polyangiitis (formerly called Wegener's granulomatosis), scleroderma, Behçet disease, reactive arthritis (formerly called Reiter syndrome), inflammatory bowel disease (ulcerative colitis and Crohn disease), and ankylosing spondylitis. The eye may be affected as a target of immune system attack in any of these diseases. The eye may, however, in certain instances, be the specific and only organ affected by certain autoimmune diseases. Examples include ocular cicatricial pemphigoid, Mooren ulcer, and various forms of uveitis, such as birdshot chorioretinopathy.

Regardless of the form of autoimmunity, any autoimmune disease affecting the eye will require systemic (e.g., oral as opposed to local, topical, ocular) therapy; the components of the immune system reside not in the eye, but rather are systemic, and therefore, regulation of those components will require systemic therapy. Such therapy is typically designed to suppress the overly aggressive immune system, allowing the body to eventually re-regulate itself, with the result often being that after the patient has been kept

on systemic medications to suppress the inappropriate immune response for a finite length of time (for example, one year), medication can then be tapered and stopped without recurrence of the autoimmune attack. Sometimes resumption of the attack does occur, in which case the patient must be re-treated.

Ophthalmologists, in general, are not accustomed to treating patients systemically, and in particular, are not trained to use immunosuppressive drugs in order to control autoimmune phenomena. Many ophthalmologists, however, realize that such treatment is appropriate and indicated for the aforementioned problems, and therefore, the ophthalmologist will collaborate with a rheumatologist who will take responsibility for monitoring and managing the patient's systemic therapy, while the ophthalmologist monitors the progress of the ocular manifestation of the autoimmune attack (inflammation). This collaboration between the ophthalmologist and rheumatologist works very well, and our experience in helping ophthalmologists establish such collaborations and to effectively treat patients with autoimmune diseases affecting the eye has been gratifyingly successful in almost every country around the globe. This represents a major change from years ago, when many patients still lost all use of one or both eyes from the ravages of improperly treated autoimmune disease affecting the eye.

The hope for the future is for more selective treatment strategies for specific autoimmune diseases, for example, cloning the causative gene for that protein, which we could use as a strategy to re-regulate patient's immune systems to that protein without the use of immunosuppressant drugs. It is entirely possible that similar strategies can be applied effectively in all autoimmune ocular diseases and studies are underway.