



# Behçet's Syndrome Society

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## The Eye in Behçet's disease

Eye involvement in Behçet's disease has been recognised since the time of Hippocrates. Estimates of the prevalence vary in different countries and may represent differences in the disease, in access to health care and in available treatments. Up to 70% of patients with the disease will get eye involvement and current estimates from around the world indicate that severe visual impairment occurs in 25% of involved eyes. Accordingly the detection and treatment of eye involvement is vital.

Despite the fact that ulceration of mucous membranes in the mouth and elsewhere are so common, ulceration of the mucous membrane covering the eye, the conjunctiva, is extremely rare and the reasons for this are not understood. The main ocular involvement in Behçet's disease is where the inflammatory process spreads inside the eye, causing uveitis. This may affect the front part of the eye causing symptoms of redness, pain and sensitivity to light. Sometimes the inflammation may be quite severe and give rise to a fluid level of pus in the eye called a hypopyon. This may be observed as a thin white line in the bottom half of the iris and may occur without the eye being red. If only the front part of the eye is affected by inflammation then the outlook for vision is good with 90% of patients retaining good vision at 5 years. Unfortunately, the usual scenario is for the inflammation to spread to the back of the eye. Here it may affect the jelly of the eye (the vitreous) and the presence of inflammatory cells in the jelly gives rise to little black specks clouding the vision (floaters) which, if dense, can significantly reduce vision. In addition the inflammation affects the blood vessels that supply the retina (the light sensing tissue lining the back of the eye). As a result, the blood vessels may leak more fluid than normal giving rise to water logging of the retina and, if the central part of the retina (the macula) is affected, a reduction in central vision. Alternatively the blood vessels close off due to thrombosis. The consequence here is that the bit of retina supplied by the blood vessel dies and if this happens recurrently, eventually the entire retina is lost, with loss of sight.

Other complications of the inflammation or its treatment may arise. These include cataract (clouding of the lens of the eye), glaucoma (a rise in the pressure inside the eye which if not controlled leads to irreversible changes in the optic nerve and blindness), and the formation of new abnormal blood vessels in the retina and iris (which have a propensity to bleed and thus cloud the sight). Late changes include detachment of the retina, low pressure in the

eye and eventual shrinkage of the eye but by this time the eye has usually lost all useful vision.

Treatment of Behçet's disease in the eye usually goes on for a long time but the natural history of the disease is to burn out with time although this may take 20-30 years. Accordingly, as there is no "cure" for Behçet's, the role of the eye doctor is to keep the patient seeing as well as possible until burnout occurs without causing intolerable side effects from the drugs used. Some doctors feel that meticulous control of mouth ulcers reduces the severity and frequency of eye attacks but this has not been proven.

If the inflammation only affects the front of the eye treatment is usually with steroid and dilating drops. Occasionally, no treatment is given as the condition is often self-limiting and painless. If the back of the eye is affected then there are a number of options but most eye specialists will start with steroid tablets in sufficient dosage to control the inflammation. These inevitably will cause side effects and the likelihood of side effects increases the longer the patient is on the drug. If the side effects are intolerable or the disease cannot be kept under control with a small enough dose then second line drugs called immunosuppressives are added. The most commonly used is a drug called azathioprine which has a proven ability to reduce the frequency of ocular attacks and to preserve vision in the long term. Otherwise, drugs such as cyclosporin, mycophenolate and methotrexate may be given but there is less scientific evidence to support their use in eye disease. All these drugs affect the ability of the body's bone marrow to produce white cells which fight off infection and they may also affect the liver and kidneys. Regular monitoring with blood tests is therefore mandatory. Colchicine is not usually used in eye disease.

There are a number of new drugs on the market for the treatment of severe eye disease which broadly fall into two classes. Interferon alpha is a protein produced naturally by the body to fight viral infections but has shown remarkable properties in controlling Behçet's disease in the eye. In Germany cases have been reported where it appears to eliminate disease for long periods of time. The second class is a group of drugs that are widely used by rheumatologists called TNF alpha blockers (such as Infliximab and Etanercept). These are usually given by infusion but have a very rapid and powerful effect on eye inflammation. It is uncertain whether they can be used longterm.

In summary, eye involvement in Behçet's disease can be very serious. It always causes symptoms so if the eye becomes red and painful or the vision becomes blurred then urgent review by a specialist is recommended. Hopefully with the newer drugs coming onto the market, permanent eye damage will be a thing of the past.

***Prof Miles Stanford, May 2008***