

## 5. Behçet's syndrome and the Eye

### **How are the eyes affected by Behçet's disease?**

Eye involvement in Behçet's disease has been recognised since the time of Hippocrates. Estimates of the prevalence ([how many people are affected by it](#)), vary in different countries and may represent differences in the disease, in access to healthcare and in available treatments. Up to 70% of patients with the Behçet's 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; 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. ([Uveitis is swelling or pain within the eye](#))

### **Complications**

Uveitis 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, the outlook for vision is good; 90% of patients retain 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 waterlogging 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. ([when clots form in blood vessels](#)) The consequence here is that the part of retina supplied by the blood vessel dies; if this happens recurrently ([again and again](#)), 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.

### **Investigations**

As part of the investigation of why the eye has been affected, several tests are done in the eye clinic. These include tests to check the pressure of the eye and tests to see how much inflammation is affecting the retinal blood vessels. A common procedure here is the injection of a yellow dye into an arm vein, followed by a series of pictures being taken of the back of the eye using a blue flash. This test (the fluorescein angiogram) is very useful in detecting leaky and blocked blood vessels. Another common test is optical coherence tomography (OCT), in which the patient is asked to sit in front of a camera which then scans the back of the eye and gives a three-dimensional picture of the retina. This is very useful in judging how well treatment is working.

### **Treatment**

Treatment of Behçet's disease in the eye usually goes on for a long time. However, 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 disease, 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. There are a number of options if the back of the eye is affected, but most eye specialists will start with steroid tablets in sufficient dosage to control the inflammation. These will inevitably 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, 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 cyclosporine, 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 the white cells that fight off infection, and they may also affect the liver and kidneys. Regular monitoring with blood tests is therefore mandatory (**essential**). Colchicine is not usually used in eye disease.

There are several 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 it has shown remarkable properties in controlling Behçet's disease in the eye.

In Germany, cases have been reported in which it appears to eliminate disease for long periods of time. The second class is a group of drugs that are widely used by rheumatologists. These TNF-alpha blockers (such as infliximab and etanercept) are usually given by infusion but have a very rapid and powerful effect on eye inflammation. It is uncertain whether they can be used long term. 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.