Behçet's Factsheet 5
The Eyes

How are the eyes affected by Behçet's?

Up to 70% of patients with Behçet's syndrome (or 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 although with newer treatments this is becoming less common. Therefore, the detection and treatment of eye disease in a timely manner is vital, and patients experiencing problems with their eyes are advised to see an ophthalmologist (an eye doctor) at the earliest opportunity.

Even though mouth and genital ulcers are very common in Behçet's disease, ulceration of the mucous membrane covering the eye, the conjunctiva, is extremely rare; the reason for this is not understood. The main eye problem in Behçet's disease is inflammation inside eye, known as uveitis. Not everybody gets uveitis, and it is impossible to predict who will. Many studies have shown that men may be affected more than women. One or both eyes can become inflamed. It appears the longer you have BD without eye involvement, for example more than 10 years, the less likely you are to get it. Also, it is very unusual to get the eye inflammation starting or to get a flare-up of uveitis over the age of 50 years.

How will I know if I have uveitis?

If you have no problems with your eyes then you are extremely unlikely to be suffering from uveitis, as an attack invariably causes symptoms.

A common scenario is of a sudden, often severe, loss of vision usually in one eye. This may occur in the morning on waking. The loss of vision may be so severe that you can only see shadows with this eye. You may also see grey/black objects floating in front of you (this is due to inflammation in the vitreous jelly). Seeing 'floaters' especially on bright, sunny days is normal for many people so they do not always imply the person has uveitis. Often the front of the eye is also inflamed, and the eye may be red, painful, and unusually sensitive to light. If the inflammation in the front of the eye is very severe there may be so many white blood (inflammatory) cells in the front chamber of the eye that they settle to the bottom forming a white sediment. It may even be possible to see this in a magnifying mirror.

If you experience any of these symptoms, you must see an ophthalmologist immediately. Do not drive to see the ophthalmologist, as you will have drops put in your eyes to enlarge the pupils and these will blur your vision.

What are the complications of having uveitis?

Inflammation at the front of eye normally improves quickly. 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 affect the back of the eye, and this often results in damage to the vision. The inflammation affects the retina, (the light sensitive inner layer at the back of the eye) causing leakage and blockage of retinal blood vessels (retinal veins). The inflamed veins leak fluid into the retina causing it to become waterlogged. If this occurs in the macula (the central part of the retina responsible for detailed reading and writing vision), then central, straight-ahead vision will be affected. Inflammatory cells also leak from the veins into the normally clear vitreous jelly of the eye causing floaters which, if dense, can also significantly reduce vision. The main nerve at the back of the eye, the optic nerve (that transmits information from the eye to the brain), can also be affected by the inflammation further damaging the vision.

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Repeated attacks of uveitis not only cause blockage of the retinal veins but also the retinal arterioles (blood vessels that provide blood and oxygen to the retina) leading to parts of the retina dying, as they are not receiving any oxygen. After many attacks, the entire retina may be lost resulting in complete and permanent loss of sight.

Other complications of the inflammation and/or its treatment (corticosteroids) may arise. These include cataract (clouding of the normally clear lens of the eye blocking light getting to the back of the eye), glaucoma (a rise in the pressure inside the eye which, if not controlled, leads to irreversible damage in the optic nerve and blindness), and the formation of new, abnormal blood vessels in the retina that tend to bleed and cloud the sight.

What happens when I see an ophthalmologist?

Your vision will be checked on the standard eye chart, and the ophthalmologist will examine the front of your eyes using a machine called a slit-lamp microscope. This is a special type of microscope that gives a magnified view of the eyes. The eye pressures are measured using a small device with a blue light after putting in some eye drops (a combination of an anaesthetic drop and an orange drop called fluorescein). This usually only takes several seconds to do. Then you will have more eye drops to enlarge the pupils. It normally takes about 30 minutes for these drops to work, and they will blur your vision (especially for reading) for a few hours. You should not drive after having your pupils enlarged. Making the pupils larger allows the ophthalmologist to examine the back of the eyes. This is usually done at the slit-lamp and the ophthalmologist looks through a small lens held in front of the eye to see the back of the eye. Sometimes another piece of equipment is used where you lie on your back on a couch and the ophthalmologist wears a headlamp and shines a very bright light through a lens that is held in front of the eye.

To get a better idea of what is happening to the back of the eye the ophthalmologist may need to do a few tests. A common procedure is the injection of a orange 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 retinal blood vessels and can also show any swelling in the retina. Another test commonly performed is an optical coherence tomography (OCT) scan, where you sit in front of the scanner that scans the back of the eye giving a three-dimensional picture of the retina. It is painless and takes only a number of seconds. This can show if there is any swelling in the macular (central) part of the retina and is also very useful in judging how well treatment is working.

What treatment might I receive?

If there is inflammation in the front of the eye then you will be given corticosteroid eye drops and drops to make the pupil larger, usually just for a few weeks as the inflammation in the front of the eye settles quite quickly. For inflammation at the back of the eye you will need something stronger than eye drops, as they do not penetrate through the eye to get to the back. Most ophthalmologists will start with high dose corticosteroid tablets (or intravenous corticosteroid). One may be on corticosteroid tablets for a long period of time, and these will inevitably cause side effects, and the likelihood of side effects increases the longer one 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 used immunosuppressive drugs include azathioprine, ciclosporin, methotrexate, mycophenolate mofetil, and tacrolimus. It is hoped that these drugs will reduce the frequency of ocular attacks and to preserve vision in the long term. Patients are often put on biologic drugs. For example, anti-TNF drugs, such as infliximab (given by intravenous infusion) or adalimumab (given by subcutaneous injection) and they have been very useful in treating a flare-up of the uveitis and preventing further attacks. Alpha-interferon is

another drug that has been used with success but now has been discontinued by the manufacturer. All these drugs have potential side effects so regular monitoring with blood tests is mandatory (essential). Colchicine (a drug often used in Behçet's disease) is not usually prescribed for eye disease as it does not seem to be helpful.

Treatment of Behçet's disease affecting the eye usually goes on for a long time and requires regular follow-ups with an ophthalmologist. However, the natural history of the disease is to burn out with time, although this may take 20–30 years. As there is no 'cure' for Behçet's disease, the role of the ophthalmologist is to keep the patient seeing as well as possible until burnout occurs without causing intolerable side effects from the drugs used.

If I have lost vision, will it come back?

Unfortunately, no guarantee can be given that this will happen. Prompt treatment may allow the vision to return but repeated attacks of uveitis are likely to lead to permanent loss of vision.

Can I drive a car if I've already lost vision in one eye?

If the other eye is healthy then the answer is yes. If in doubt, please ask your ophthalmologist.

Is there anything I can do to prevent getting uveitis?

Like many of the other problems with Behçet's disease, one cannot predict when an attack will happen. It is hoped that if you are already on treatment that some of drugs will prevent the eyes being affected or prevent getting a flare-up of the eye inflammation. As smoking is also associated with blocking off blood vessels, then it would seem logical and sensible to stop smoking.

What should I do if I am experiencing eye symptoms?

In summary, eye involvement in Behçet's disease can be very serious.

It always causes symptoms, so if the eye becomes red and painful and unusually sensitive to light, or the vision suddenly becomes blurred, then an urgent review by an ophthalmologist is recommended. This may require reporting to an Eye A&E Department or if there is not one close by then a General A&E Department and requesting a consultation with the duty ophthalmologist (and perhaps armed with this Factsheet). Treated appropriately in a timely manner hopefully will avoid further complications and any potential reduction in vision.

With newer drugs coming onto the market, it is hoped that permanent eye damage will be a thing of the past.

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