Behçet's Syndrome Society

Registered Charity No: 326679
Caring for those with a rare, complex and lifelong disease www.behcets.org.uk

2. Behçet's syndrome: A Patient's Guide by

Highlighted words are explained at the end

So you have Behçet's syndrome?

So you've just received a diagnosis or a 'potential diagnosis' of Behçet's syndrome? You're probably feeling a mixture of emotions now, but this will include relief at receiving a diagnosis and also fear about what this illness is and how it's going to affect you and potentially your family.

Firstly, congratulations on finding us. We are the UK Charity supporting people with Behçet's syndrome and their families. We were established in 1983 and have a great deal of experience in helping people with this condition. By finding us, you've also found comprehensive medical information checked by our medical panel.

Also, receiving a diagnosis or a suspected diagnosis of Behçet's syndrome is a real step forwards. It means you must have found a medical person who could recognise the symptoms and was aware of this rare illness. This is not something to take lightly, as many of our members have waited several years to obtain a diagnosis. However, rest assured that we have in England three national Centres of Excellence for Behcet's (in London, Birmingham and Liverpool) where you can choose to be treated. If you live in Wales, Northern Ireland or Scotland, you can access the centres but this needs prior approval (and funding) from local health boards.

What is Behçet's syndrome and how is it pronounced?

You will hear many different pronunciations of the illness, but Hulushi Behçet who gave his name to the illness pronounced his name "Betjet". To add further complication, it is also referred to as both Behçet's syndrome and Behçet's disease. The two are the same and are used interchangeably.

The syndrome is named after a Turkish Professor of dermatology who described the **main** features of the condition in 1937. It had previously been described as long ago as the 5th century BC by Hippocrates, by a Chinese physician in the 5th century AD, and by a number of European physicians in the late 19th and early 20th century.

The cause of the condition is not known. It's been proved that there is a disturbance of the immune system, but this isn't thought to cause the illness. Current theories are that it is potentially a genetic weakness that is triggered by an infection or

virus, but this has still to be proved. It tends to be diagnosed when patients are in their 20s or 30s, but it can be diagnosed outside of these age brackets.
It is known that the illness is most prevalent in Turkey, North Africa, the Middle East and South East Asia – the old Silk Route, which gives the condition its other name – Silk Route disease. It is rare in Western Europe, with an estimated 1000 people in the UK having a diagnosis. It seems to be more common among women in Europe, but men are more frequently affected in other regions, and men also tend to have more severe disease activity.

How is it diagnosed?

There is no diagnostic test for Behçet's syndrome, which is partially the reason for the elongated diagnosis period. A clinician who has experience of Behçet's syndrome would be able to make a diagnosis based on a collection of the following symptoms presenting themselves (other clinicians may have a high index of suspicion and refer you to an expert):

- Ulcers – in both the mouth and genital region.
- Skin lesions – acne-like or red tender swellings known as erythema nodosum.
- Eye involvement – including inflammation of the front or back of the eye (uveitis) or around the retina (retinal vasculitis).
- Arthritis or arthralgia (joint pain) – particularly in the knees, hands and wrists.
- Thrombophlebitis – inflammation of veins, usually in the lower legs, resembling deep vein thrombosis.
- Pulmonary lesions – arising from inflammation around the heart.
- Central nervous system involvement – arising from inflammation around the brain.
- Gastrointestinal ulceration – affecting the small intestine or the colon.
- Pathergy reaction – a red lump appearing 48 hours following a sterile needle pricking the skin. (Not everyone with Behcet's will have this reaction.)
- Chronic fatigue.

This list is long and affects different areas of the body, but you would be very unlucky to have all of these symptoms. Behçet's syndrome seems to affect patients differently both in the symptoms you first present with and also in what may develop later. It is best to be aware of the various areas it can affect so that you can discuss any concerns you have with your consultant.

Appointments and your doctors

It may well have been that up to the symptoms of Behçet's syndrome presenting themselves you didn't have a need to visit your GP and the hospital very much. Unfortunately, that is likely to change and you will need to build relationships with the medical professionals who are providing your care.

The Society was successful in 2012 in working with a medical team to establish Centres of Excellence for Behçet's Syndrome in Barts & the London, Birmingham City Hospital and Aintree University Hospital Liverpool. You can choose to enrol at one of these Centres where you'll be seen by a team of consultants including a

rheumatologist, ophthalmologist and oral specialist, with other specialists being available at specific clinics.
You can ask to be referred to one of the centres in England if you are a patient in Scotland, Wales and Northern Ireland, but your doctor will need to get approval first (and funding) from your countries NHS.
At each of these Centres, there is also a support worker available who can help with non-medical matters such as giving benefits advice and help on how to tell friends, family and work about the illness. The advantage of these Centres is that all the consultants have a specific interest in Behçet's syndrome and they provide holistic care with all the specialists discussing your care together and sharing their expertise. It will also reduce the number of hospital appointments you'll need to attend.

You can choose to remain with your current consultant in a local hospital, or alternatively the Society can help you to find a consultant with experience in Behçet's syndrome close to you, although we cannot make a recommendation. With this arrangement, depending on what symptoms you have, you may just be seeing one consultant and your GP or you may have several consultants concentrating on different areas of your illness. The regular medical appointments you will need can become exhausting, particularly when you're unwell, but it is important that you attend all of them or let the clinic know if you are too unwell.

Whichever treatment route you choose, when your symptoms flare, it is very important to seek prompt medical advice. The Centres have a phone number you can ring for advice - **if you are registered with them**. For local care, a call to your consultant's secretary or your GP may be required. If you have built up a relationship with your clinician, this is far easier.

The future

Behçet's syndrome is currently incurable, but that doesn't mean it is untreatable. Many drugs are on the market which, although not licensed specifically for Behçet's syndrome, can have a positive effect on controlling the symptoms.

The course of the illness tends to be that it 'flares up' and then subsides. During these flare-ups, the symptoms you already have may become more of a problem or you may present with new ones. It is at this point that your medication may be increased or new drugs added to your prescription. However, during times when your disease activity is minimal, you will still need to take this medication to prevent any inflammation.

There are different drugs you may be prescribed depending on your symptoms. Steroids are often used to dampen down the immune system, but immunosuppressant's may also be required, together with topical treatments for other symptoms.

Unfortunately, Behçet's syndrome doesn't qualify you for free prescriptions from the NHS. If you do not qualify for free prescriptions due to another condition or

your financial circumstances, it may be beneficial to consider a Prepayment Certificate for your prescriptions.
These can be bought quarterly or annually and can work out more economical depending on how many medicines you take regularly. Different arrangements currently apply to prescription charges in Scotland, Wales and Northern Ireland.

What can the Society do for you?

The Behçet's Syndrome Society offers a large number of services, including:

- Helpline (0345 130 7329) or helpline@behcetsdisease.org.uk – operated by volunteers who either have Behçet's syndrome themselves or have a relative with it. They offer a confidential service and will listen to your concerns and do their best to answer any questions you have.
- Website (www.behcets.org.uk) – our website provides a wealth of information for both patients and medical professionals. It has a chat forum available for members to share tips and seek help from other members. It is also used to keep our members informed of what the Society is doing and the events you may be interested in.
- Factsheets – these provide medical information on all the different areas affected by Behçet's syndrome. They are written by our medical experts and are aimed at both patients and their medical carers.
- Support groups – having a rare illness can be isolating, and support groups help to overcome that feeling. We are currently developing a network of groups throughout the country. Please get in touch to see if there's one in your area.
- Newsletter – our members receive a quarterly newsletter keeping them informed of both what the Society is doing on their behalf and also what research is taking place.
- Annual Conference – this is an opportunity for our members to meet other people with Behçet's syndrome. We also invite prominent medical experts to discuss advances in the treatment of the illness.
- Carry card – our members can receive a carry card which is the size of a credit card. This concertinas to contain valuable information about your medical carers, the medicines you take and the illness in general should you need it in an emergency.
- Benefits help – we can signpost to up to date and relevant information.
- Small personal Grant awards – for more information, please contact us and ask for a copy of our guidance notes for applicants.
- Supporting research – as a small charity we are unable to conduct research ourselves, but we provide support where we can to research projects that are aiming to benefit Behçet's syndrome patients.

Can you help the Society?

You may think that you're new to this illness and that you have nothing to offer the Society, but you will be best placed to let us know what services are most needed. A fresh viewpoint is always welcomed.

If you're refreshing your knowledge with this factsheet, maybe you're at a stage now when you could bring your experience to the Society?

You can help in many ways, whether this be joining the Board or volunteering for the Helpline or whether you'd like to help with fundraising. This can involve selling our Christmas cards or encouraging friends and family to help. Maybe they'd like to jump out of a plane, run a marathon or even undertake a challenge event such as walking the Great Wall of China! Contact info@behcetsdisease.org.uk for more information.

Explanation of highlighted words in the order they appear in the factsheet.

Syndrome:	a set of symptoms associated with a particular disease or diseases
Dermatology:	study of the skin and diseases of the skin
Immune system:	the body's defence system. Full definition in useful words (see separate sheet)
Genetic weakness or susceptibility:	this means the genes you inherit from your parents may make you more likely to have a particular condition
Prevalent:	how many people are affected by the illness in that area
Vasculitis:	inflammation in the walls of veins and arteries
DVT:	when a blood clot forms in one of the deep veins in the leg
Gastrointestinal:	about the stomach and the intestines(guts)
Rheumatologist:	a doctor with specialist training in the diagnosis and treatment of patients with diseases of the joints and muscles.
Ophthalmologist:	a doctor who specializes in studying and treating diseases of the eye
Holistic:	treating the whole person and all aspects of the disease not just one part
"Flare Up":	a relapse after a period of being well, with little or no symptoms, or when the condition gets worse
Immunosuppressant:	drugs that reduce inflammation over longer periods of time
Topical medicine:	that is usually put directly onto the skin in the area that is affected