
Behçet's Disease

What are the aims of this leaflet?

This leaflet has been written to help you understand more about Behçet's disease (also referred to as Behçet's Syndrome). It tells you what it is, what may cause it, how it affects the mouth and the rest of the body, what can be done about it and where you can find out more about it.

What is Behçet's disease?

Behçet's disease is a rare condition that causes the tissues of the body to become inflamed, particularly small blood vessels. This can affect many different parts of the body and can give rise to a number of symptoms. It can affect both men and women of all age groups, but often develops in young adults aged 20-30. The prevalence of Behçet's disease in the UK is less than 1 per 100,000 and it is more common in those from the Mediterranean, Middle East, Japan and South-East Asia.

What causes Behçet's disease?

It is not known what causes Behçet's disease but there is evidence to suggest that it may have more than one cause. The condition is often described as an autoinflammatory disease in which the body's immune system mistakenly attacks its own, healthy cells. Behçet's disease is not contagious and cannot be passed from one person to another.

Is Behçet's Syndrome hereditary?

There is no evidence that Behçet's disease is hereditary. However, it is recognised that some patients may have a genetic predisposition to developing the disease and therefore a genetic test (HLA B*51) may be carried out. It is important to note, however, that this test may also be positive in people not suffering from Behçet's disease.

What are the symptoms of Behçet's disease?

Behçet's disease can have a number of symptoms and patients will often experience episodes of active disease followed by periods of remission. All patients develop ulcers in the mouth. Genital ulceration, inflammation of the eyes, skin problems, excessive tiredness, joint pains, bowel problems and headaches are also symptoms of Behçet's syndrome. Behçet's syndrome also increases the risk of developing a blood clot. It is important to note that not all symptoms occur in every patient.

What does Behçet's disease look like?

- In the mouth, ulcers can vary in size from a millimetre to greater than one centimetre and can be single or multiple. They have a grey/yellow base with an area of surrounding redness. They may heal with scarring. For more information on ulcers please refer to the patient information leaflet on Recurrent Aphthous Stomatitis (RAS).
- Genital ulcers affect the vulva and vagina of women and the scrotum and, less commonly, the penis of men.
- Ulceration around the anus can affect some patients.
- Inflammation of the eye can cause blurred vision, redness, pain and 'floaters' (dark specks that float across your field of vision). If you develop any of these symptoms it is important that you contact your doctor immediately as severe cases can lead to loss of vision.
- A number of skin problems may affect those with Behçet's disease, including red, raised lumps under the skin, acne-like spots, boils or ulcers.

How is Behçet's disease diagnosed?

There is no specific test for Behçet's disease. Doctors therefore base their diagnosis on a 'high index of suspicion' with patients experiencing the following:

- Recurrent mouth ulcers (more than 3 in a 12 month period)
- Plus 2 of the following:
 - Genital ulcers

- Skin problems typical of Behçet's disease
- Eye inflammation typical of Behçet's disease
- Positive pathergy test (red, raised spot appearing after minor injury to the skin)

The symptoms of Behçet's disease can often be confused with other illnesses so blood tests are often arranged to exclude an alternative cause. Sometimes it is necessary to take a small sample (biopsy) from an affected area inside the mouth for microscopic examination to rule out any causes of mouth ulceration. A local anaesthetic injection to 'numb' the biopsy site is necessary for this procedure.

Can Behçet's disease be cured?

There is currently no cure for Behçet's disease, although it can become less active over time. Treatment aims to reduce the symptoms associated with the condition and reduce inflammation.

How can Behçet's disease be treated?

Treatment aims to increase your quality of life by relieving discomfort, reducing inflammation and encouraging healing.

- Anaesthetic/analgesic mouthwashes can be used if your mouth becomes sore and can be particularly helpful before meals.
- Covering agents form a barrier against secondary infection and irritation. They are available as pastes or gels.
- Topical corticosteroids are very helpful for most patients and are available as mouthwashes, inhalers, sprays and dissolvable pellets.
- Systemic corticosteroids (tablets) may be required during a flare-up of the condition. These are not regularly used on a long term basis and will often be substituted for alternative systemic medications (for example azathioprine) that work by suppressing the body's immune system. Your doctor will discuss these with you and you will be required to have regular blood tests.

- Other medications may be used if recommended by a specialist (e.g. colchicine, ciclosporin, infliximab, adalimumab, thalidomide). You will need to have regular blood and other tests while taking these drugs.

What can I do?

There are a number of ways in which you can help to reduce the impact of Behçet's disease on your daily life:

- Maintain a healthy, balanced diet, which includes plenty of fresh fruit and vegetables. Avoid spicy or acidic food if they make your symptoms worse.
- Take regular periods of rest and exercise – It is important to pace yourself and plan your day to incorporate regular periods of rest before and after activities
- Keep your teeth clean. You may wish to enrol the help of a hygienist.
- Some tooth pastes may aggravate your mouth ulcers and it may be helpful to use a mild toothpaste with a minimum of flavourings and other ingredients.
- Continue to visit your dentist regularly

Where can I get more information about Behçet's disease?

The Behçet's Syndrome Society: www.bhecets.org.uk/

Arthritis Research UK: www.arthritisresearchuk.org/

This leaflet has been prepared by the British Society for Oral Medicine (BSOM). It is reviewed periodically to reflect relevant advances and improved understanding. Not all the information will be relevant to all patients. For individual advice please see your Oral Medicine specialist. The BSOM is not responsible for information on web sites where links are provided.

This leaflet is available online at www.bsom.org.uk, review date April 2019