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## 1. Behçet's - Overview

*Highlighted words are explained in Common Terms and Definitions sheet*

### **What is Behçet's syndrome?**

Behçet's syndrome (or Behçet's disease) 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 several European physicians in the late 19th and early 20th century.

Firstly, there is a debate about whether it should be called Behçet's disease or Behçet's syndrome. A syndrome is a collection of clinical features that may, or may not, be one condition and may be the presentation of several similar conditions. As there are differences in the [incidence](#) of Behçet's syndrome, and of its various [manifestations](#) in different countries, and since we do not yet know the exact cause, some physicians prefer to keep an open mind and use the term syndrome. Calling it Behçet's disease may suggest that we know that it is a single [entity](#) with a single cause in all cases.

Secondly, there is a debate about how one should diagnose the condition. Diagnosis in the individual patient depends on the clinical [acumen](#) and experience of the physician. Various schemes of diagnosis have been suggested over the years, none of which is perfect. There is no absolute diagnostic test for the condition. A test known as the pathergy test has been investigated over the years. It consists of pricking the skin of the forearm with a sterile needle and seeing if a red lump appears after 48 hours. This was thought to be specific for the condition but is not considered as such now since it is positive only in a minority of patients, the frequency of positive tests is falling over the years and some 'normal' patients may have a positive test.

Diagnosis, therefore, depends on a high index of suspicion ([See last section - When should Behçet's syndrome be suspected?](#)) and prompt referral of the patient to a physician who has considerable knowledge and experience of the condition. International Classification Criteria have been promulgated, but these are principally used for research purposes to ensure that studies of (for example) causation or treatment include patients who are comparable in different centres. These original criteria were not intended to be used for diagnosis in an individual patient, though

the 2013 International Criterion for Behçet's Disease proposes the criteria be adopted both as a guide for diagnosis and classification of Behçet's disease.

The condition is most frequent in Turkey, North Africa and the Middle East, and in South East Asia. While it is rare in Western Europe, it must not be overlooked. Although there are differences in various studies, it is generally found that women are more frequently affected in Europe, men are more frequently affected in the Middle East and South East Asia, and in all regions men have more severe manifestations.

The cause of the condition is not known. A major disturbance of the [immune system](#) has been demonstrated in laboratory tests, but this is not thought to be the underlying cause. Some authorities have suggested an infective cause, but this has never been convincingly confirmed. It is known that there is a strong genetic predisposition. Detection of some of the markers (like blood groups), or antigens, on the white cells in the blood, which are genetically transmitted, show a high incidence of one known as HLA-B51 compared with the general population. However, this is not the cause, as most people with this marker do not have Behçet's syndrome and, conversely, some patients with Behçet's syndrome do not possess the HLA-B51 antigen. Nevertheless, there is often a strong family history of the syndrome, or just a family history of mouth ulceration.

Studies of involved tissues in patients with Behçet's syndrome show white blood cells in and around blood vessels (i.e. inflammation) – and particularly around large and small veins. Large arteries may also be involved, but this is relatively rare. While some authorities have labelled this as a vasculitis, which usually means inflammation leading to injury or destruction of blood vessels, it is probably more accurate to call this a vasculopathy, which means involvement of blood vessels of all sizes for whatever cause. This all implies that continuing research into the causation of the condition is essential.

### **Most common features**

#### ***(Mucocutaneous)***

- Mouth ulceration – this is [recurrent](#) arbitrarily defined as occurring at least three times in any 12-month period, and occurs in 98% of patients. Ulcers may be multiple or single, are usually painful and, depending on their severity, heal in 7–21 days. Although this is the most frequent manifestation, it may not be the initial one, and other features may be present for a considerable time before the development of mouth ulcers. The 2% of patients who do not have mouth ulceration are, in all other respects, no different from those who do.
- Genital ulceration – again, this is recurrent and painful; it occurs on the scrotum, and less commonly on the penis, in males and on the vulva and in the vagina in females, in about 80% of patients.
- Skin lesions – these include acne-like lesions, red tender swellings known as erythema nodosum, and occasional ulceration.

## **Eye involvement**

Eye involvement occurs in up to 50% of patients. Inflammation of the front of the eye around the iris is referred to as anterior uveitis. The patient has a painful, red eye with blurring of vision. The patient feels uncomfortable looking at bright lights. This usually respond to eye drops Inflammation at the back of the iris is more serious and may lead to blindness. It is referred to as posterior uveitis. The patient suffers from blurred vision and floaters. This requires more systemic treatment. The blood vessels of the retina (retinal vasculitis) is another serious feature presenting with sudden loss of vision. Eye involvement occur usually in younger patients within a few years of disease onset.

## **Arthritis or arthralgia (joint pains)**

This occurs in about 50% of patients. Some patients may experience joint pains only, without any outward evidence of joint inflammation. However, when a true arthritis is present it is inflammatory, indicated by joint pain, stiffness, swelling, warmth and tenderness. The knees are most commonly affected, followed by the ankles, the small joints of the hands and wrists, and least commonly the shoulders and hips. The lining of the joints (synovium) has been shown to be inflamed (synovitis), but the appearance under the microscope is different from other inflammatory arthritides, ([other types of arthritis or arthritic conditions](#)) such as rheumatoid arthritis. Permanent bone damage within the joint may be seen both on X-rays and at surgery, but rarely.

There has been considerable debate over the years as to whether the joints of the spine are involved in the inflammatory process (spondylitis). Although some cases have been described, the consensus of opinion is now that spondylitis is not a part of the arthritis of Behçet's syndrome.

Joint pain (arthralgia) is more common than arthritis which is rarely deforming.

## **Other features**

- Thrombophlebitis – inflammation of veins, most frequently in the lower legs, resembling a deep vein thrombosis.
- Arteritis – inflammation of arteries, which may swell at points of weakness (aneurysms) or rupture causing bleeding into the tissues.
- Pulmonary lesions– arising from inflammation, and possible occlusion, of the pulmonary arteries.
- Central nervous system involvement – due to inflammation around arteries and veins in the brain and thrombosis of large veins (known as dural sinuses) inside the skull.
- Gastrointestinal ulceration– this is most frequent in patients in the Far East; the ulceration usually involves the colon (the large intestine), which may bleed or [perforate](#).

## **Treatment**

In many patients with less severe disease, treatment remains largely symptomatic and thus aimed at relieving the unpleasant symptoms of the condition. While there have been major advances in the effective treatment of the more serious manifestations of the condition, such as vasculitis (see above), especially with drugs which suppress or modify the inflammatory and immunological disturbances, no absolute cure is yet known. For further details on treatment, please refer to our Treatment factsheet.

## **When should Behçet's syndrome be suspected?**

A high index of suspicion will be raised if the patient presents with two or three possible manifestations, such as:

- Painful recurrent mouth ulcers and genital ulcers, or
- Painful recurrent mouth ulcers and an inflamed eye, or
- Painful recurrent mouth ulcers, genital ulcers and an inflamed eye, or
- Painful recurrent mouth ulcers, genital ulcers and inflamed joints, or
- Painful recurrent mouth ulcers, genital ulcers and skin lesions, or
- Inflamed eye(s) and joints and skin manifestations, or
- Inflamed eye(s), thrombophlebitis and skin manifestations, or
- Painful recurrent mouth ulcers, an inflamed eye and a positive family history.

These associations do not make the diagnosis, (having a mixture of these signs does not mean that the patient definitely has Behçet's) and many others could be listed. They are situations in which the diagnosis should be suspected and further advice should be sought.