# Behçet's syndrome

## 1. Behçet's syndrome

Behçet's syndrome, also known as Behçet's disease, is a rare multi-system inflammatory condition. In the UK, the most common presentation is with skin inflammation and mucous membrane ulceration, but the condition has significant geographic variation both in incidence and in the type of presentation. It is a complex disease, and management will be led by teams in secondary care. In primary care, we can help by being aware of when to suspect and refer, and by having some understanding of the associated risks to watch out for.

This article uses information from a 2024 British Association of Dermatology (BAD) and British Society of Rheumatology (BSR) joint guideline, and also from reviews in the Lancet and the New England Journal of Medicine (BJD 2024;191:e8, Lancet 2024;403:1093, NEJM 2024;340:640).

*This article was updated in December 2024.*

## 1.1. Incidence and epidemiology

Peak incidence of Behçet's is between 20 and 40 years of age, and presentation in childhood or past the age of 50 is rare (Lancet 2024;403:1093). If children do develop Behçet's, the presenting symptoms are often musculoskeletal or gastrointestinal rather than mucocutaneous, making diagnosis a challenge.

It is more common in men than women.

Worldwide prevalence is estimated at around 10/100 000 people (Lancet 2024;403:1093), but there is high geographic variability. There are around 1000 people in the UK living with the condition (BAD Patient Hub - Behçets).

## 1.2. Pathogenesis

The pathogenesis of Behçet's is not fully understood, but seems to be multi-factorial with a genetic predisposition, followed by exposure to environmental triggers such as infection, gut and oral microbiome imbalance, stress, poor oral hygiene and a diet high in histamine-containing foods (NEJM 2024;340:640). It shows features of both autoimmune and auto-inflammatory pathology.

## 1.3. Diagnosis

Diagnosis is clinical, with no universally-accepted diagnostic criteria and no validated biomarker we can use for confirmation (Lancet 2024;403:1093).

The BAD/BSR guideline recommends using one of 2 diagnostic tools: the International Study Group 1990 tool or the International Criteria for Behçet's disease, updated in 2024.

| **International Study Group Criteria 1990** | **International Criteria for Behçet's 2024** |
|---|---|
| • Recurrent oral ulceration (major, minor or herpetiform)<br>• Plus:<br>  • At least 2 other recurrent lesions affecting skin, genitals or eyes<br>  • OR a positive pathergy test. | • Score 2 points each for any of:<br>  • Recurrent oral or genital ulcers, or eye lesions.<br>• Score 1 point for any of:<br>  • Skin, neurological or vascular signs.<br>  • A positive pathergy test.<br>• A score of 4 is needed for diagnosis. |

*The pathergy test: a non-specific skin hyperreactivity response is seen after an intradermal skin prick (the guidelines do not discuss using this test in primary care).*

| **Presentation by body system** *(NEJM 2024;340:640 unless otherwise indicated)* |
|---|
| Brain (affects 10–30% of people):<br>• More common in men. |

- Acute presentation may be encephalitis, meningitis, myelitis.
- More chronic presentations include ataxia, dementia, confusion and incontinence (Lancet 2024;403:1093).

Eyes (affects 50% of people):

- Panuveitis.
- Retinal vasculitis.
- Hypopyon.
- Previously a significant cause of vision loss in affected individuals (10–20%), but this has improved with newer therapies.

Skin and mucosa:

- Oral ulcers affect 98% of people.
    - Recurrent, painful, round or oval mucosal erosions, mainly on the lips, cheeks and tongue (Lancet 2024;403:1093).
- Genital ulcers affect 60–65% of people.
    - Tend to be larger than oral ulcers and take longer to heal. Usually affect scrotum or labia (Lancet 2024;403:1093).
- Skin lesions affect around 75% of people.
    - Pseudofolliculitis.
    - Papulopustular lesions (acne-like) on face, chest and shoulders.
    - Erythema nodosum.
- Pathergy response (a non-specific skin hyperreactivity response is seen after an intradermal skin prick such as a vaccination).

Vascular (affects up to 40% of people):

- Arterial:
    - Arterial aneurysm.
    - Arterial stenosis or thrombosis.
- Heart:

- Valvulitis.
- Myopericarditis.
- Coronary arteritis.

- Venous:
  - VTE (DVT, PE, suprahepatic embolism).
  - Superficial thrombophlebitis.

GI tract (40–60% of people in the UK will have GI involvement (Lancet 2024;403:1093). Worldwide rates are generally lower (NEJM 2024;390:640)):

- Ulcers commonly found in terminal ileo-caecal or rectal bowel, but can affect any part of the GI tract.
- Can result in perforation and/or haemorrhage.

Musculoskeletal (affects 50% of people):

- Recurrent asymmetric monoarthritis or oligoarthritis, usually affecting knees, ankles, wrists or elbows.
- Not usually associated with (Lancet 2024;403:1093):
  - Joint deformity.
  - Erosions on imaging.
  - Spinal involvement.

## 1.4. Management

If we suspect Behçet's, we should refer to secondary care for MDT review and diagnostic assessment (BJD 2024;191:e8). The summary below is taken from the BAD/BSR joint guideline, which goes into detail on the secondary care management of the different disease manifestations.

### In secondary care

At MDT review, the specialist teams should use a disease-specific tool to assess disease activity and quality of life.

Treatment depends on the manifestation of the disease, but will usually involve potent topical corticosteroids followed by systemic steroids (oral or IM), with disease-modifying steroid-sparing drugs once the condition is stabilised. Combinations of DMARDs, anti-TNF drugs and biologics are often used. Musculoskeletal symptoms may respond to colchicine.

## 1.5. Prognosis

Behçet's shows a variable course, with a relapsing and remitting pattern (NEJM 2024;390:640). Prognosis is worse in those diagnosed at a younger age and in those with higher degree of disease activity. Males are 5x more likely to die of the condition. Vascular disease carries the highest mortality. Overall mortality is estimated at almost 10% over the 20 years after diagnosis.

Regardless of which body systems are impacted, there is a significant adverse effect on quality of life, affecting both physical and psychological wellbeing.

**Behçet's syndrome**

- A rare multisystem inflammatory condition which usually presents with skin inflammation and mucous membrane ulceration.
- Refer any suspected case to secondary care for diagnosis and management with DMARDs and corticosteroids.
- Can affect multiple organ systems, including GI, eye and musculoskeletal presentations.
- Prognosis is worse in males, those diagnosed at a younger age or those with higher disease burden.

**Useful resources:**

*Websites (all resources are hyperlinked for ease of use in Red Whale Knowledge)*

- BAD Patient Hub - Behçets

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