BEHÇET'S SYNDROME
(Also called Behçet's Disease)

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What is Behçet's Syndrome?

Behçet's Syndrome is a variable vessel vasculitis, meaning it can cause inflammation of arteries and veins of all sizes. This results in involvement of potentially any organ, though we still see certain patterns of symptom presentation.

Who is affected?

Behçet's Syndrome is a rare condition affecting 10 – 500 per million people. There is a wide ethnic variation, with those along the **Silk Road** affected the most (eg. **Turkey**, the Middle East, and oriental countries). In North America, it is estimated to affect 100-300 per million individuals. The condition normally starts in early adulthood (age 20-30s), with few cases presenting before puberty or past age 50.

What causes Behçet's Syndrome?

The exact cause is unknown – a combination of genetic, infectious and environmental factors likely contributes, in variable proportions, to its onset.

The **HLA-B51** gene has been linked to Behçet's Syndrome (particularly those of Turkish & Asian background), however having a positive HLA-B51 **does not** necessarily mean one has the condition, as it is present in up to 10% of the general population. Conversely, more than 40% of patients do not carry the HLA-B51 gene.

What are possible symptoms?

Patients can have a variety of findings (though not all of them will develop in every patient), including but not limited to:

- **Recurrent oral ulcers** – painful "canker sores" in the mouth that often appear in clusters. They affect virtually all patients and are often the 1st symptom.
- **Genital ulcers** – painful, round white sores that look similar to oral ulcers but on the genitals.
- **Skin rash** – several rashes and skin lesions can occur, the most common one being clusters of pustular acne-like lesions. Non-healing skin lesions or wounds from minor injuries / abrasions can occur.
- **Eye inflammation** – several types occur, the most common being uveitis which often affects both eyes. The eye can be red, painful or the vision may be altered, without any pain.
- **Brain involvement** (uncommon) – headache is the most common symptom, but seizures, strokes and other neurological symptoms are possible.
- **Vascular involvement** (uncommon) – includes blood clots and aneurysms (vessel wall outpouchings, particularly in the main lung arteries that can rupture and cause massive bleeding).
- **Joint involvement** – severe joint pain with or without joint inflammation (swelling of the joints) can occur.
- **Gastrointestinal tract involvement** – with abdominal discomfort and ulcerations. The pattern of ulcers can be hard to distinguish from that of inflammatory bowel disease.

How is Behçet's Syndrome diagnosed?

No tests can determine whether you have the condition, so your doctor will rely primarily on your signs and symptoms. Because nearly everyone with the condition develop recurrent oral ulcers (mouth sores), this is necessary for a diagnosis of Behçet's.

Other tests that your doctor may order to support the diagnosis include:

- **Blood tests** that demonstrate elevated inflammatory markers
- **Eye exam** that demonstrates inflammation
- **Biopsy** of certain skin rashes/lesions
- **Imaging** of specific body parts, depending on your symptoms
- **Skin pathergy test**, in which your doctor punctures your skin with a sterile needle and injects a small amount of saline, then examines the skin 1-2 days later. If the test is positive, a small red bump forms under your skin where the needle was inserted. This indicates your immune system is overreacting to minor injury.
- Lumbar puncture (spinal tap) to look for inflammation in the brain – only done if there are compatible neurological symptoms or abnormal findings on brain imaging.

Note that routine HLA-B51 testing is **not recommended** as it is only useful in certain ethnic contexts and can be positive in healthy individuals also. Many other more common conditions can mimic Behçet's (eg. IBD, celiac disease, lupus and chronic infections) and need to be ruled out during the investigatory phase.

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How is Behçet's Syndrome treated?

Treatment depends on the severity of symptoms and which organs are affected. The goal is to suppress inflammation, reduce the frequency of "flares", and prevent long-term organ damage.

For mild features such as oral ulcers, treatment typically includes:

- Topical steroid creams
- Colchicine or dapsone
- Hydroxychloroquine
- Apremilast
- Low-dose prednisone

For more severe manifestations affecting the eyes, gastrointestinal tract, blood vessels or central nervous system, stronger immunosuppressive drugs are often required, including:

- Methotrexate
- Azathioprine
- Mycophenolate mofetil
- Anti-TNFs (eg. infliximab, adalimumab, etanercept)
- High-dose prednisone (with taper)
- Cyclophosphamide

Combinations of medications are commonly used, especially when the condition does not respond to single-agent therapy, or if the condition is organ or life threatening.

How is Behçet's Syndrome monitored?

Since the presentation of the condition is so variable, monitoring of labs and other investigations depends heavily on which organs are affected.

Monitoring can range from an "as needed basis" to frequent blood tests and imaging in severe forms of the condition.

Common things that can be monitored include:

- Lab tests for blood counts, inflammatory markers (CRP and/or ESR), and liver & kidney function
- Brain imaging (CT scan or MRI) if there is suspected inflammation within the central nervous system
- Routine eye exams to check for uveitis and other forms of eye inflammation

Testing is typically more frequent at the beginning or when there is active inflammation, and less frequent once the condition is stable and in remission.

Depending on which organs have been affected, your rheumatologist may work with other specialists, including **ophthalmologists**, **dermatologists**, and **neurologists**, to help treat and monitor your condition.

Is Behçet's Syndrome dangerous?

There are many forms of the condition and everyone is affected differently. Patients may have a milder form, characterized mainly by recurrent oral ulcers with some genital ulcers or skin involvement. While these symptoms can be painful and frustrating to go through, they will not affect your life span.

Those that have the more serious forms, such as involvement of the eyes, brain, heart, or large vessels (eg. aneurysms or clots) unfortunately do worse and have a higher rate of complications. The key is working with your specialists to keep the condition and its inflammatory complications under control with medications.

What will happen to me? How long do I need to take medications?

Like other autoimmune conditions, Behçet's Syndrome can be treated but not yet cured. The optimal duration of treatment is unknown. Symptoms and features of the condition can return when medications are tapered or stopped completely, resulting in relapses or "flares".

Because of this, most individuals are kept on medications for several years, and even longer if they have had severe organ damage already.

BEHÇET'S SYNDROME SUMMARY TOOL

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My symptoms and features included:

- ☐ Oral ulcers
- ☐ Genital ulcers
- ☐ Papulopustular acne-like skin lesions (can look like clusters of small whiteheads)
- ☐ Erythema nodosum (painful raised, red nodules usually on the legs)
- ☐ Vessel dilation (aneurysms)
- ☐ Blood clots (thrombosis)
- ☐ Joint pain and/or joint swelling
- ☐ Abdominal pain and/or bloody stool
- ☐ Coughing up blood
- ☐ Headaches
- ☐ Aseptic meningitis
- ☐ Strokes (eg. facial droop, paralysis of arm or leg, unable to speak normally)
- ☐ Eye pain, redness and/or severe light sensitivity
- ☐ Vision loss
- ☐ Recurrent fevers
- ☐ Unusually severe fatigue
- ☐ Other: _____

Important medications for my Behçet's:

	Date started	Date stopped
Prednisone (Starting dose _____ mg)		
IV methylprednisolone _____ mg		
Colchicine _____ mg		
Dapsone _____ mg		
Hydroxychloroquine _____ mg		
Methotrexate _____ mg		
Folic acid (only if taking methotrexate)		
Azathioprine _____ mg		
Leflunomide _____ mg		
Mycophenolate mofetil _____ mg		
Apremilast _____ mg		
Cyclophosphamide _____ mg		
Adalimumab _____ mg		
Infliximab _____ mg		
Etanercept _____ mg		
Other: _____ _____ _____		

I have had the following investigations done:

- ☐ Blood tests checking for inflammation markers (ESR, and/or CRP)
- ☐ Skin pathergy (by formal testing)
- ☐ Full eye examination
- ☐ Lumbar puncture (spinal tap)
- ☐ Colonoscopy or upper endoscopy
- ☐ X-rays (circle all that apply): Chest | Abdomen | Joints
- ☐ CT scans (circle all that apply): Chest | Abdomen | Head |
- ☐ MRI (circle all that apply): Brain | Spine
- ☐ Biopsy (circle all that apply): Skin | Ulcer | GI tract
  Other: _____

What do I need to do?

- ✓ Attend **regular** follow-up visits with my rheumatologist and other specialists
- ✓ Have routine eye examinations at least once a year to assess for inflammation in my eyes
- ✓ **Do regular** monitoring blood tests
- ✓ Monitor for new symptoms such as prominent skin lesions, non-resolving headaches, or vision problems.
- ✓ **Call 911 ASAP if I develop new stroke-like symptoms or coughing up blood!**