

Behçet's Syndrome

What is Behçet's Syndrome?

Behçet's syndrome is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Behçet's affects blood vessels of all sizes and types, and can potentially involve any organ including the central nervous system. Treatment is essential in controlling symptoms and preventing serious complications such as blindness and stroke.

Common symptoms of Behçet's include painful mouth and genital sores, skin rashes, swollen joints, and eye inflammation. However, the disease also causes more serious symptoms such as vision loss, blood clots, gastrointestinal, brain and/or spinal cord problems.

Treatment depends on the severity of disease and organ system involvement. In mild cases, topical corticosteroids may be applied to affected areas. For more serious disease, medications that suppress the immune system are used. Behçet's is a chronic disease and relapses are common, thus ongoing medical care is important.

Causes

The cause of Behçet's is not fully understood. Vasculitis is classified as an autoimmune disorder—a disease that occurs when the body's natural defense system mistakenly attacks healthy tissue. Behçet's is one of the few forms of vasculitis where a specific gene – HLA-B51 – is a known risk factor for the syndrome. However, this gene is also seen in the general population, and not everyone who has it gets Behçet's. Therefore, it is believed that an infection and/or environmental factors may play a role in the onset of this disease.

Who Gets Behçet's Syndrome?

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Symptoms

The symptoms of Behçet's can vary greatly from person to person. Some may have milder disease, while others have severe, even potentially life-threatening symptoms. Most people have periods of relapse and remission, with symptoms often showing up in different parts of the body—sometimes years later.

The most common symptoms of Behçet's syndrome are:

- Painful ulcers, resembling canker sores, inside the mouth
- Painful, open genital sores
- Skin lesions resembling acne that can occur anywhere on the body
- Eye inflammation with symptoms of blurred vision (or blindness), redness and pain
- Joint swelling, pain, and stiffness, especially in the knees, ankles, elbows and wrists

Less common, but serious symptoms include:

- Blood clots
- Intestinal problems
- Inflammation of the brain and spinal cord, with severe headaches, stiff neck, and fever (meningitis)

Complications

Some complications of Behçet's are serious. Untreated eye inflammation can lead to decreased vision or even blindness. People with eye symptoms should have a full eye exam by an eye doctor (ophthalmologist). Blood clots that form in the extremities can lead to serious complications, as can inflammation of the blood vessels in the lung. Although rare, pulmonary artery aneurysm—an abnormal bulge in the artery wall that can burst—is the leading cause of death among Behçet's patients. Inflammation in the membranes of the brain and spinal cord can lead to significant disability if untreated.

Diagnosis

There is no single test for diagnosing Behçet's syndrome. Your doctor will consider a number of factors including a detailed medical history, a physical exam, laboratory tests, imaging studies, and possibly a skin biopsy. The presence of classic Behçet's symptoms, including recurrent mouth and genital sores, eye inflammation, and skin lesions, help confirm the diagnosis. The genetic marker HLA-B51 is more common among people with Behçet's, but the presence of the gene alone is not diagnostic.

Laboratory or imaging tests may be ordered to rule out diseases with similar symptoms such as other forms of vasculitis (granulomatosis with polyangiitis and polyarteritis nodosa), inflammatory bowel disease, systemic lupus erythematosus, rheumatoid arthritis, Lyme disease and others.

Your doctor may also order a pathergy test, a procedure in which a small, sterile needle is inserted into the skin of the forearm. After 24 to 48 hours, people with Behçet's can develop a lump or nodule at the needle insertion point, which indicates the immune system is overreacting to a minor injury. However, even a positive pathergy test is not conclusive.

Treatment

Treatment for Behçet's is aimed at reducing inflammation and preventing organ damage. Treatment depends on disease severity, symptoms, and organ involvement. The first line of treatment may include topical corticosteroids applied directly to the affected area such as skin creams, gels and ointments, eye drops and mouth rinses. Oral corticosteroids such as prednisone may also be prescribed to reduce inflammation. If topical or oral steroids aren't effective, your doctor may prescribe other medications to fight inflammation including colchicine, which is commonly used to treat gout.

More severe disease may require immunosuppressive drugs such as methotrexate, azathioprine, cyclosporine, apremilast, and cyclophosphamide.

Biologics are also an option when Behçet's is severe. Biologic medications are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation. Examples used to treat Behçet's include infliximab, etanercept, adalimumab, and interferon alpha.

Side Effects of Treatment

The medications used to treat Behçet's have potentially serious side effects such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also very important. Talk to your doctor about getting vaccines (e.g., flu shot, pneumonia and/or shingles vaccination), which can reduce your risk of infection.

Medical Follow-up/Relapse

Even with effective treatment, some individuals with CSVV will experience relapses. If your symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular checkups and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your Medical Team

Effective treatment for Behçet's may require the coordinated efforts and ongoing care of a team of providers and specialists. In addition to a primary care provider, Behçet's patients may need to see the following specialists:

- Rheumatologist (joints, muscles, and immune system)
- Gynecologist (female reproductive system)
- Urologist (male reproductive system and urinary system)
- Dermatologist (skin)
- Ophthalmologist (eyes)
- Neurologist (brain and nervous system)
- Pulmonologist (lungs)
- Gastroenterologist (digestive system) or others as needed

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications,

symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns about your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with Behçet's Syndrome

Living with Behçet's can be challenging at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting relationships, work, and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

There is no cure for Behçet's syndrome at this time, but treatment can relieve symptoms and prevent potentially serious complications such as blindness and stroke. Behçet's can be a chronic disorder with periods of remission and relapse, so most patients need to see a doctor on an ongoing basis.

In 2021 the American College of Rheumatology (ACR) published guidelines for the management of certain vasculitides, that were also endorsed by the Vasculitis Foundation (VF). Clinical practice guidelines are developed to reduce inappropriate care, minimize geographic variations in practice patterns, and enable effective use of health care resources. Guidelines and recommendations developed and/or endorsed by the ACR are intended to provide guidance for particular patterns of practice and not to dictate the care of a particular patient. The application of these guidelines should be made by the physician in light of each patient's individual circumstances. Guidelines and recommendations are subject to periodic revision as warranted by the evolution of medical knowledge, technology, and practice.

About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM disease (formerly Goodpasture's syndrome)
- Aortitis
- Behçet's syndrome
- Central nervous system vasculitis (CNSV)
- Cogan's syndrome
- Cryoglobulinemic vasculitis
- Cutaneous small-vessel vasculitis (CSVV) (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis (GCA)
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (formerly Henoch-Schönlein purpura)
- Kawasaki disease
- Microscopic polyangiitis (MPA)
- Polyarteritis nodosa (PAN)
- Polymyalgia rheumatica (PMR)
- Rheumatoid vasculitis
- Takayasu arteritis (TAK)
- Urticarial vasculitis
- (normocomplementemic or hypocomplementemic)

About the VF

The VF is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF is a 501(c)(3) nonprofit organization governed by a Board of Directors and advised on medical issues by a Medical and Scientific Advisory Board. VF's educational materials are not intended to replace the counsel of a physician. The VF does not endorse any medications, products, or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

The VF gratefully acknowledges Alexandra Villa-Forte, MD, Cleveland Clinic Center for Vasculitis Care and Research, for her expertise and contribution to this brochure.

To access additional VF support and educational resources, please scan the QR code below.

VF Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.



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