BEHCET'S SYNDROME

What is Behcet's syndrome?

Behcet'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. Behcet's affects blood vessels of all sizes and types, and can potentially involve any organ, including the brain and spinal cord. Treatment is essential in controlling symptoms and preventing serious complications such as blindness and stroke.

Common symptoms of Behcet's syndrome 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, intestinal problems, and brain and spinal cord inflammation.

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, oral corticosteroids such as prednisone may be combined with medications that suppress the immune system. Behcet's is a chronic disease with periods of relapse for some patients, so ongoing medical care may be necessary.

Causes

The cause of Behcet's syndrome is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissue. Behcet'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 Behcet's. Therefore, it is believed that an infection and/or environmental factors may play a role in the onset of this disease.

Who gets Behcet's syndrome?

Behcet's mainly affects people in their 20s and 30s, but the syndrome can occur in individuals of all ages. While Behcet's occurs in both men and women, the disease is usually more severe in males.

People from the Mediterranean, the Middle East and Asia are more likely to get this disease; it is rare in the United States. Prevalence is estimated at 3 to 5 per 100,000 people in the U.S. Turkey has the highest prevalence, with approximately 400 cases per 100,000 people.

Symptoms

The symptoms of Behcet'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 Behcet's syndrome are:

- Painful ulcers, resembling canker sores, inside the mouth
- Painful, open sores on the genitals
- 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

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Less common, but serious symptoms include:

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

Complications

Some complications of Behcet's are serious. Untreated eye inflammation can lead to decreased vision or even blindness. People with eye symptoms should schedule an appointment with an eye doctor. 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 Behcet's patients. Inflammation in the membranes of the brain and spinal cord (meningitis) can lead to significant disability if untreated. Call your doctor if you have the symptoms listed above or other symptoms you are concerned about.

Diagnosis

There is no single test for diagnosing Behcet's syndrome. Your doctor will consider a number of factors, including a physical exam, medical history, laboratory tests, imaging studies, and possibly a skin biopsy. The presence of classic Behcet'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 Behcet'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 Behcet'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 Behcet'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 immunosuppressant drugs, such as methotrexate, azathioprine, cyclosporine, apremilast, and cyclophosphamide.

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Biologics are also an option when Behcet'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 Behcet's include infliximab, etanercept, adalimumab, and interferon alpha.

Side effects

The medications used to treat Behcet'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 important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment and periods of remission, some individuals will experience relapse of Behcet's—sometimes months or even years after the original symptoms subside. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your medical team

Effective treatment for Behcet's may require the coordinated efforts and ongoing care of a team of providers and specialists. In addition to a primary care provider, Behcet's patients may need to see the following specialists: rheumatologist (joints, muscles, and immune system); gynecologist (female reproductive system); urologist (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 with 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 Behcet's syndrome

Living with Behcet'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.

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Outlook

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

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

For more information on vasculitis research, visit: www.vasculitisfoundation.org/research

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 (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

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About the Vasculitis Foundation

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

Connect with the VF on Social Media

- Instagram: vasculitisfoundation
- Twitter: @VasculitisFound
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation 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.

Join the VPPRN!

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

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