

## **What is Behçet's syndrome?**

Behçet's syndrome is a form of vasculitis. Vasculitis is a group of rare disorders characterized by inflammation of the blood vessels. Inflammation of blood vessels may result in aneurysms or narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues. Vasculitis is also classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissue.

## **Causes**

The cause of Behçet's syndrome is not fully understood and it 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 cause this disease.

## **Who Gets Behçet's syndrome?**

Behçet's mainly affects people in their 20s and 30s, but the syndrome can occur in individuals of all ages. While Behçet'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.

## **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.

The most common symptoms of Behçet'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

Less common, but serious symptoms include:

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

Most people have periods of relapse and remission, with symptoms often showing up in different parts of the body—sometimes years later.

Call your doctor if you have the symptoms listed above or other symptoms you are concerned about.

## Complications

Some complications of Behçet's are serious. For example:

- 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 Behçet's patients.
- Inflammation in the membranes of the **brain and spinal cord (meningitis)** can lead to significant disability if untreated.

## Diagnosis

There is no single test for diagnosing Behçet's syndrome. Your doctor will obtain a medical history and perform a **physical exam**, as well as:

- **Laboratory tests** to rule out other conditions, such as other forms of vasculitis, and to determine if the genetic marker HLA-B51 is present, however it is not a diagnostic test.
- **Imaging studies**, including x-rays and computed tomographic (CT scan).
- **Skin biopsy**, called pathergy test, 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 depends on the severity of disease and organ system involvement.

- **Topical corticosteroids**, in mild cases, may be applied to affected areas.
- **Oral corticosteroids** such as prednisone may be used for more serious disease.

- **Other drugs (some are immunosuppressive)** may be used with oral corticosteroids, these include colchicine, methotrexate, azathioprine, mycophenolate mofetil, apremilast and others.
- **Biologic agents** such as infliximab or rituximab and others may be prescribed. They target certain parts of the immune system to control inflammation.

Behçet's is a chronic disease with periods of relapse for some patients, so ongoing medical care may be necessary.

## 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
- Potential bone loss (osteoporosis), among others

It is 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 Behçet'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 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 (urinary system)
- Dermatologist (skin)

- Ophthalmologist (eyes)
- Neurologist (brain and nervous system)
- Pulmonologist (lungs)
- Gastroenterologist (digestive system)
- Others as needed

The best way to manage your disease is to actively partner with your health care providers and 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.

## **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.

## **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.

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