Overview of Behçet’s Disease

Behçet’s disease is a chronic condition that causes mouth or genital sores, and inflammation in parts of the eye. In some people, the disease also produces arthritis (swollen, painful, stiff joints), skin problems, and inflammation of the digestive tract, brain, or spinal cord.

The disease is named after the doctor who first described it, Dr. Hulusi Behçet.

Prognosis

Doctors can use many medicines to relieve pain, treat symptoms, and prevent complications. You will probably enter a period of remission (a disappearance of symptoms). In some people, treatment does not relieve symptoms, and gradually more serious symptoms, such as eye disease, may occur. Serious symptoms may appear months or years after the first signs of Behçet’s disease.

Who Gets Behçet’s Disease?

Anyone can develop Behçet’s disease, but it is more common among:

- People in the Middle East, Asia, and Japan, as compared with the United States.
- Women than men in the United States.
- Men than women in Middle Eastern and Asian countries.

Behçet’s disease tends to develop in people in their twenties or thirties, but people of all ages can develop this disease.

Symptoms of Behçet’s Disease

Behçet’s disease affects each person differently. You may have only mild symptoms, such as sores in the mouth or on the genitals. Fever, stiff neck, and headaches could be signs of a more severe issue: meningitis (inflammation of the membranes that cover the brain and spinal cord). These more severe symptoms usually appear months or years after the first signs of Behçet’s disease.
Symptoms can last for a long time or may come and go in a few weeks. Symptoms usually appear, disappear, and then reappear. You are going through a “flare” if you are having symptoms. Each flare could have different symptoms. You should keep a record of which symptoms occur and when. This will help your doctor diagnose and follow your disease, especially since many conditions may look like Behçet’s disease.

Common symptoms of Behçet’s disease include the following:

- **Mouth sores** affect almost all people with Behçet’s disease. They are probably the first symptom that you noticed and may occur long before any other symptoms appear. The sores usually have a red border and several may appear at the same time. They may be painful and can make eating difficult. Mouth sores go away in 10 to 14 days but often come back. Small sores usually heal without scarring, but larger sores may scar.
- **Genital sores** affect more than half of all people with Behçet’s disease. They usually appear on the scrotum in men and vulva in women. The sores look similar to the mouth sores and may be painful. They may cause scarring after several outbreaks.
- **Skin problems** are a common symptom of Behçet’s disease. The sores are red and raised, and may look like a pus-filled bump or bruise. They typically appear on the legs and on the upper torso.
- **Inflammation of the middle or back part of the eye** (the uvea) and the iris happens in more than half of all people with Behçet’s disease. This is more common among men than women and typically begins within 2 years of the first symptoms. Eye inflammation can cause blurred vision. In rare cases it causes pain and redness. You should immediately tell your doctor if you have these symptoms, since continued eye inflammation can cause vision loss.
- **Arthritis** occurs in over half of all people with Behçet’s disease. Arthritis causes pain, swelling, and stiffness in the joints, especially in the knees, ankles, wrists, and elbows. The arthritis usually lasts a few weeks and does not cause permanent joint damage.
- **Blood clots** sometimes form in inflamed veins, usually the legs, of some people with Behçet’s disease. The affected area may be painful, swollen, and warm. Blood clots can cause serious health issues. So, be sure to report any of these symptoms to your doctor right away. Some people may have artery problems such as aneurysms (balloon-like swelling of the artery wall).
- **Brain and spinal cord problems** occur in about 1 in 4 people with the disease in the United States. Behçet’s disease can cause inflammation of the brain and the thin membrane that covers and protects the brain and spinal cord. Symptoms can include fever, headache, stiff neck, and difficulty coordinating movement. You should immediately report any of these symptoms to your doctor, as a stroke can result if the condition is not treated.
- **Digestive tract inflammation and sores** occur in rare cases. Symptoms can include stomach pain, diarrhea, and/or bleeding. Your doctor will need to rule out other diseases that
have similar symptoms, such as ulcerative colitis and Crohn’s disease.

Causes of Behçet’s Disease
No one knows what causes Behçet’s disease, and it is not contagious. Researchers think that one or more genes produce an abnormality in the immune system, making you more at risk for the disease. This abnormality may make your body’s immune system mistakenly attack its own blood vessels, instead of attacking foreign substances such as germs. Researchers don’t know what triggers this attack, although it could be something in the environment such as bacteria or a virus. The blood vessels become inflamed, producing the symptoms of the disease.

Diagnosis of Behçet’s Disease
There is no specific test for Behçet’s disease, which makes it hard to diagnose. Symptoms that help your doctor diagnose the disease include:

- Mouth sores at least three times in 12 months.
- Any two of the following symptoms:
  - Genital sores that keep coming back.
  - Skin sores, or sores that develop after your doctor pricks your skin with a needle. Only up to half of people with the disease will see a bump appear about 1 to 2 days later.
  - Eye inflammation with loss of vision.

Your doctor will need to rule out other conditions that have similar symptoms, such as Crohn’s disease and reactive arthritis. Diagnosis may take some time, as some symptoms may take years to appear.

You may even see different doctors before you are diagnosed. An eye specialist will help identify health issues related to eye inflammation. A dermatologist may examine your mouth, genital, or skin sores to help rule out other disorders.

Treatment of Behçet’s Disease
Although there is no cure for Behçet’s disease, you can control symptoms with proper medication, rest, exercise, and a healthy lifestyle. Treatments make you more comfortable and prevent serious health effects, such as disabling arthritis and blindness. You may need a combination of treatments for your specific symptoms. Make sure you tell all of your doctors about medications you take so your doctors can coordinate treatment.
Medicines

One or more medicines may help treat the various symptoms of Behçet’s disease:

- **Rinses, gels, ointments, or creams** are applied directly to sores to relieve pain and swelling.
- **Corticosteroids** are prescribed to reduce pain and inflammation from skin sores or of the joints, eye, or brain and spinal cord. It is important to see your doctor on a regular basis to watch for any side effects from these drugs.
- **Immunosuppressive drugs** help control an overactive immune system, reduce inflammation, and lessen the number of disease flares. These medicines are very strong and can have serious side effects, so it’s important to see your doctor regularly if you take these drugs.

Rest and Exercise

Rest is important during when you are having symptoms from Behçet’s disease. Moderate exercise, such as swimming or walking, are recommended when the symptoms have improved or disappeared. This can help keep your joints strong and flexible.

Who Treats Behçet’s Disease?

Because the disease affects different parts of your body, you will probably see several different doctors. You may find it helpful to coordinate your care through one doctor, who can watch for any side effects from the different medicines you take. Care is often managed by a rheumatologist, a doctor specializing in arthritis and other inflammatory disorders.

The following specialists also treat other symptoms:

- Dermatologists treat skin sores.
- Urologists treat genital sores.
- Gynecologists treat genital sores for women.
- Ophthalmologists treat eye inflammation.
- Gastroenterologists treat digestive tract symptoms.
- Hematologists treat disorders of the blood.
- Neurologists treat disorders of the brain and spinal cord.

Living With Behçet’s Disease

Most people with Behçet’s disease can lead productive lives and control symptoms with proper medicine, rest, and exercise. Rest is important when you are having symptoms from Behçet’s disease. Moderate exercise, such as swimming or walking, are recommended when the
symptoms have improved or disappeared. This can help keep your joints strong and flexible.

Research Progress Related to Behçet’s Disease

Researchers are exploring possible causes and treatments of Behçet’s disease. Genetic studies show strong association of the gene HLA-B51 with the disease, but the exact role of this gene in Behçet’s is uncertain. Other genes have also been identified that appear to be associated with Behçet’s disease. Studies of these genes and how they work may provide new understanding of the disease and possibly new treatments.

Researchers are also investigating factors in the environment, such as bacteria or viruses, that may trigger Behçet’s disease. Several microbes are being studied.

Finally, researchers are identifying other medicines to better treat Behçet’s disease. TNF inhibitors reduce joint inflammation and have shown some promise in treating Behçet’s disease, although this medicine does have serious side effects. Interferon alpha, a protein that helps fight infection, has also shown promise in treating Behçet’s disease.

For More Info

U.S. Food and Drug Administration
Toll free: 888-INFO-FDA (888-463-6332)
Website: https://www.fda.gov

Drugs@FDA at https://www.accessdata.fda.gov/scripts/cder/daf. Drugs@FDA is a searchable catalog of FDA-approved drug products.

Centers for Disease Control and Prevention, National Center for Health Statistics
Website: https://www.cdc.gov/nchs

National Institute of Dental and Craniofacial Research
Website: https://www.nidcr.nih.gov

National Institute of Diabetes and Digestive and Kidney Diseases
Website: https://www.niddk.nih.gov

National Eye Institute
Website: https://www.nei.nih.gov
National Institute of Neurological Disorders and Stroke  
Website: https://www.ninds.nih.gov

American College of Rheumatology  
Website: https://www.rheumatology.org

American Behçet's Disease Association  
Website: http://www.behcets.com

American Skin Association  
Website: http://www.americanskin.org

Autoinflammatory Alliance  
Website: http://www.autoinflammatory.org

If you need more information about available resources in your language or other languages, please visit our webpages below or contact the NIAMS Information Clearinghouse at NIAMSInfo@mail.nih.gov.

- Asian Language Health Information
- Spanish Language Health Information