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Behçet's disease

Behçet's disease (also known as Behçet's syndrome) is a rare condition which causes frequent ulcers in your mouth and genital area and can affect different parts of your body. It most commonly affects your eyes and skin. It is more common in people from Mediterranean countries and also those from the Middle East and Eastern Asia. Various treatments are available which help reduce long-term complications.

Who develops Behçet's disease?

Behçet's disease is not commonly seen in Northern Europeans. It most commonly affects people from the Middle East, Mediterranean and Eastern Asia. It usually affects people aged 20-30 years and affects men and women equally. Although it is not known to be a genetic condition, it can run in families.

The actual cause of this condition is still not completely understood, although there is plenty of research being done in this area.

What are the common symptoms?

Behçet's disease can affect different systems of the body. These include:

Skin

Painful ulcers in the inside of your mouth are very common and these are usually very painful. They can last for around three weeks. Many people experience frequent ulcers in their mouths. Ulcers can also occur in your genital region and are usually painful. However, these are not sexually transmitted.

Some people develop red swellings on their legs, known as erythema nodosum. These are usually painful. You may also develop acne-like spots on your arms and/or your legs.

Eyes

Eye symptoms are very common in people with Behçet's disease. The most common eye condition is uveitis which is inflammation of the middle layer of your eye. Either the front or back of your eye can be affected by uveitis. Less commonly, other parts of your eyes can be affected. This can lead to clouding of the lens in your eye (a cataract) or raised pressure in your eye (glaucoma) developing.

Joints

Around half of people with Behçet's disease develop pain in their joints, most commonly the knees or ankles. This pain can be associated with swelling and increased warmth. Sometimes the small joints in your hands, your wrist, shoulders and hips can be affected in a similar way. However, this does not usually cause a type of arthritis that damages your joints.

Blood vessels

Behçet's disease can cause inflammation of different blood vessels in your body. Inflammation of the veins in your legs can lead to a condition called thrombophlebitis. Inflammation of the lining of your arteries can lead to a swelling occurring in your arteries, called an aneurysm.

Other systems in your body can sometimes also be affected. This can lead to memory problems and can also affect your balance and speech in some cases. The gut (gastrointestinal tract) can be affected in some people, causing ulcers in the lining of the bowel. This can lead to symptoms of cramping abdominal pain and diarrhoea. Some people may even experience more severe symptoms, including bleeding from the back passage (rectum).

How is the diagnosis made?

There is no specific test to diagnose Behçet's disease. Your doctor may often be able to diagnose this condition by your symptoms alone.

Your doctor may arrange further tests. These are usually performed to exclude any other conditions which may cause similar symptoms to Behçet's disease. These may include blood tests and scans. Your doctor may perform a pathergy test, which measures the sensitivity of your skin. You will be given a small pinprick or injection into your skin; if a characteristic red spot appears, the result is positive. However, this is not a very sensitive test. So, if your result is positive this does not mean that you definitely do have Behçet's disease.

What is the treatment for Behçet's disease?

The treatment you receive will depend on the symptoms you are experiencing and also which part of your body is affected by this condition. Your doctor will be able to give you specific information about your individual case. The treatment is given to try to prevent any long-term complications from developing.

If your symptoms are mild then you may only need treatment when you have a flare-up. For example, if you are experiencing repeated mouth and/or genital ulcers your doctor may give you some rub-on (topical) treatment (usually steroid cream) to use on the ulcers as and when they occur. Steroid eye drops are given if you have inflammation of the middle layer of your eye (uveitis). Some people also need medication in the form of tablets in addition to eye drops.

Simple painkillers such as paracetamol or ibuprofen may improve any painful joints. These should be taken regularly. They are available either over the counter from the chemist or from your doctor.

However, if you are experiencing more severe symptoms then medication is usually given to try to suppress the disease. This is usually in the form of steroid tablets and also another medication to suppress your immune system, which then suppresses the activity of the disease in your body. Examples of these medications include azathioprine and cyclophosphamide.

More recently other medications are being given which work to reduce inflammation in your body. These are usually given by injection. An example of these medications is infliximab.

What is the outlook (prognosis)?

Behcet's disease usually becomes worse for a time (a flare-up) and then gets better. During flare-ups, the symptoms you already have may become more of a problem or you may present with new ones. It is at this time that your medication may be increased or you may even be prescribed different medication.

People with Behçet's disease usually have a normal life expectancy. It is important to have regular reviews with your doctor.

Further reading

- EULAR recommendations for the management of Behçet disease; European League Against Rheumatism (January 2008)
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