



Behçet's Syndrome Society

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Behçet's Syndrome and Blood Vessels

What is the effect of Behçet's syndrome on blood vessels?

One important feature of Behçet's Syndrome is inflammation in the walls of blood vessels (*vasculitis*). The underlying cause is not known. Behçet's syndrome is unusual in that it can cause inflammation in both arteries and veins and can involve vessels of any size.

Inflammation in a vessel means that many white blood cells, designed to fight infection, accumulate within the thickness of the vessel wall. This can lead to several outcomes:

- The lining of the vessel becomes more 'sticky' and there is therefore a risk of blood clot forming within the vessel and blocking it (*thrombosis*)
- Sometimes the vessel wall slowly becomes thicker as the cells accumulate, and the inside of the vessel, the lumen, becomes narrower so less blood can flow through it (*stenosis*)
- The wall may also become weaker as the white blood cells can disrupt the small layer of muscle which keeps the strength of the vessel wall. This can result in widening of the vessel (*aneurysm*).
- In rare cases the vessel wall of an aneurysm can tear, causing profuse bleeding (*rupture*)

Inflammation in Veins

About 25% of patients with Behçet's syndrome suffer from inflammation in a vein in the leg at some point. This is usually due to inflammation and occlusion of a superficial vein (ie near the skin surface), causing painful thickening of vessels visible just under the skin (*superficial thrombophlebitis*).

In about 5% of patients, a blood clot forms in one of the deep veins of the leg (*deep vein thrombosis*). This is usually treated with a blood thinner, warfarin. Rarely such a blood clot can dislodge and travel in the blood to the lung (*pulmonary embolus*). The symptoms of this are usually shortness of breath and/or chest pain.

Very occasionally larger central veins in the abdomen and chest become inflamed and occlude. This may manifest as swelling in the head and neck (*superior vena cava obstruction*) or legs (*inferior vena cava obstruction*).

Inflammation in small veins can cause some of the unusual rashes that are seen in Behçet's disease, including skin ulcers (*vasculitic ulcers*) and tender red nodules that are usually seen on both shins (*erythema nodosum*).

Inflammation in Arteries

Involvement of arteries is much less common than in veins but is often more serious. Involvement of arteries in the arms or legs can lead to ballooning (*aneurysm*) or occlusion (*arterial thrombosis*). Occlusion of the femoral arteries which supply the legs can cause a pale, suddenly painful leg.

The pulmonary artery, which takes blood from the heart to the lungs, can be narrowed due to inflammation leading to high pressures in this artery (*pulmonary hypertension*). This is an important cause of shortness of breath on exertion and sometimes causes a cough with small amounts of blood (*haemoptysis*).

What is the treatment when a complication arises?

When a complication occurs, it usually signifies vascular inflammation and the treatment is to suppress the inflammation using medication such as steroids and immunosuppressive drugs. The intensity of the treatment is tailored to the severity of the problem. If a blood clot has formed this is usually treated with warfarin. How long warfarin needs to be continued varies between individuals.

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