

## 10. Behçet's Disease and the Heart

### How is the heart affected by Behçet's disease?

As Behçet's disease causes inflammation of blood vessels (vasculitis) and an increased tendency to blood clotting (thrombosis), it is reasonable to assume that the heart may sometimes be affected. Studies show that 7–46% of people with Behçet's disease will have involvement of the heart or blood vessels. However, heart disease is common even in apparently healthy people, so it is difficult to know how much of the problem is caused by the Behçet's disease and how much is incidental.

Problems with the heart may be caused by thrombosis (blockage due to clotting) of large blood vessels or by vasculitis (inflammation) of smaller blood vessels, such as the arteries providing the blood supply to the heart muscle. Vasculitis can cause inflammation or fibrosis (scarring) of the heart muscle and potentially leads to reduced pump function of the heart or abnormalities of the heart's conduction system that ensures the chambers of the heart contract in a coordinated manner. Possible symptoms of heart involvement include pain on exertion, breathlessness, faints and palpitations. Any such symptoms should be reported to your doctor, as further investigation may be required.

### Complications

Cases of people with Behçet's disease with the following cardiac complications have been reported, raising the possibility that these complications may be associated with Behçet's disease:

- Acute myocardial infarction (heart attack).
- Ventricular thrombosis (blood clot in one of the chambers of the heart).
- Aortic aneurysm (bulging of the main artery leading from the heart).
- Valvular dysfunction (leakiness or partial blockage of a valve in the heart).
- Endocarditis (inflammation of the lining of the heart).
- Myocarditis (inflammation of the heart muscle).
- Pericarditis (inflammation of the membrane covering the heart).
- Congestive cardiomyopathy (weakening of the heart muscle, causing fatigue, breathlessness and swelling of the legs).

More recently, techniques such as electrocardiography (ECG), echocardiography (ultrasound scanning of the heart) and Doppler tissue imaging (a form of echocardiography that gives more information about muscle contraction and blood flow) performed in healthy volunteers and people with Behçet's disease have suggested that there may be an increased incidence of the following complications:

- Aneurysms – bulging of the arteries, especially the aortic root and ascending aorta (the main artery as it leaves the heart).
- Blood clots, which can form in aneurysms, potentially causing further problems with interruption of blood supply to other organs (thrombosis or embolism).
- Heart rhythm problems (arrhythmias).
- Heart failure (left ventricular diastolic dysfunction).

### **Behçet’s disease medications that may affect the heart**

Tacrolimus may cause problems with heart muscle damage (cardiomyopathy) or enlargement (hypertrophy), heart rhythm problems or thrombosis (blood clots). People taking tacrolimus may need monitoring with echocardiography. Infliximab, etanercept and adalimumab may cause heart failure (weakening of the heart muscle leading to shortness of breath and poor exercise tolerance). They should be avoided in people with pre-existing moderate-to-severe heart failure and stopped if symptoms of heart failure develop. (Please note: this is not a comprehensive list of drugs that may affect the heart. Sometimes drugs with a risk of heart side effects may nevertheless be the best choice to treat Behçet’s-related heart disease.)

### **Investigations**

Initial investigations of suspected heart involvement may include ECG to check for rhythm problems and signs of heart strain. Further tests may include echocardiography, Doppler imaging or 24-hour ECG (monitoring of the heart rhythm for 24 hours while normal activities are carried out).

### **Prevention and treatment**

#### ***General measures***

The following are important for everyone, but even more so for people with Behçet’s disease:

- A healthy diet with not too much animal fat, regular consumption of oily fish once or twice a week, and five portions of fruit or vegetables a day.
- Daily gentle exercise such as walking.
- Avoidance of excess weight gain.
- Your doctor should ensure that your blood pressure remains within the normal range and should check you for raised cholesterol or diabetes, giving treatment with standard medications if necessary.

#### ***Specific measures***

Low-dose aspirin (75 mg daily) can help to protect against cardiovascular disease. Heart failure and heart rhythm problems should be treated with standard medications. If the heart disease is thought to be caused by the Behçet’s disease, corticosteroids (prednisolone) and/or [immunosuppressive drugs](#) may be necessary.

*Immunosuppressive drugs reduce inflammation over long periods of time (generally years rather than months)*