

Behçet's Disease and the Heart

Since 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 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, and how much is incidental. Problems with the heart may be caused by thrombosis (blockage due to clotting) of large blood vessels, or to 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 which ensures that the chambers of the heart contract in a co-ordinated manner.

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:

- acute myocardial infarction (heart attack)
- ventricular thrombosis (blood clot in the 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 the 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 legs)

More recently, techniques such as electrocardiography (ECG), echocardiography (ultrasound scanning of the heart) and Doppler tissue imaging (a form of echocardiography giving more information about muscle contraction and blood flow) performed in healthy volunteers and people with Behçet's 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 (main artery as it leaves the heart)

- Blood clots 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 medications which may affect the heart

Tacrolimus (FK506)

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). Should be avoided in people with preexisting moderate to severe heart failure and stopped if symptoms of heart failure develop.

Please note; this is not a comprehensive list of drugs which 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.

Possible symptoms of heart involvement

Pain on exertion, breathlessness, faints or palpitations should be reported to your doctor as further investigation may be required.

Investigation of suspected heart involvement

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

Prevention and treatment

General measures: important for everyone but even more so for people with Behçet's.

- A healthy diet with not too much animal fat, regular consumption of oily fish once or twice a week, five portions of fruit or vegetables per 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 (75mg daily) can help 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, corticosteroids (prednisolone) and/or immunosuppressive drugs may be necessary.

Dr Hilary Longhurst, April 2008