

## CASE REPORT

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## Aorto–atrial fistula without aneurysm formation in Behçet's disease

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### Abstract

Cardiac disease is known to occur in patients with the multisystemic inflammatory disorder Behçet's disease. An aorto–atrial fistula secondary to Behçet's disease without a sinus of Valsalva aneurysm has not been previously reported. A 30 year old man with a four year history of symptoms and signs consistent with Behçet's disease presented with an aorto–

atrial fistula and a large left to right shunt. At operation the defect was repaired with a bovine pericardial patch. There was no evidence of aneurysm formation. The patient made a good recovery and was well at one year follow up.

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Keywords: aorto–atrial fistula; Behçet's disease.

Behçet's disease is characterised by aphthous stomatitis, genital ulcerations, and eye disease.<sup>1</sup> The disease has been confirmed as a chronic, relapsing, multisystemic inflammatory disorder characterised by widespread vasculitis of both the arterial and venous side of the circulation.<sup>2</sup> Important cardiac disease may co-exist with Behçet's disease.<sup>3</sup> The pathogenesis remains unknown although it is likely that infection could act as a trigger in genetically susceptible individuals.<sup>2</sup>

### Case report

A 30 year old man with a four year history of episodic thrombophlebitis, mouth and genital ulceration, weight loss, night sweats, arthralgia, pustulous skin lesions, gastrointestinal bleeding (clinical manifestations of Behçet's disease), and a recently developed loud machinery-type heart murmur was referred for further investigation.

Transoesophageal echocardiography showed flow from the non-coronary sinus of the aorta to the right atrium and right ventricle. Cardiac catheterisation confirmed an aorto–right atrial fistula with a Qp/Qs of 1.5:1. In view of associated moderate cardiomegaly early elective surgical closure was planned. At operation the fistula was found to extend from the non-coronary sinus directly through the annulus of the tricuspid valve into the right atrium (figs 1 and 2). There was no evidence of an aneurysm of the sinus of Valsalva.

To facilitate exposure, the base of the septal leaflet of the tricuspid valve was disconnected and the atrial side of the fistula was repaired by incorporating the disconnected septal leaflet on to the right atrial wall. The aortotomy was extended into the non-coronary sinus and a bovine pericardial patch sutured up from the base of the non-coronary sinus to close both the defect and the aorta. Biopsy specimens were taken from the thymus, pericardium, fistula, and

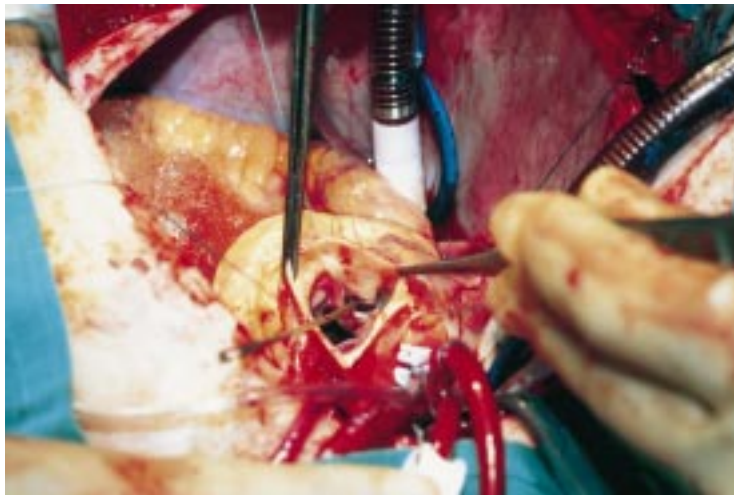


Figure 1 Operative view looking down into the aorta. The probe is passed into the fistula arising from the non-coronary sinus. Note the absence of any aneurysm.

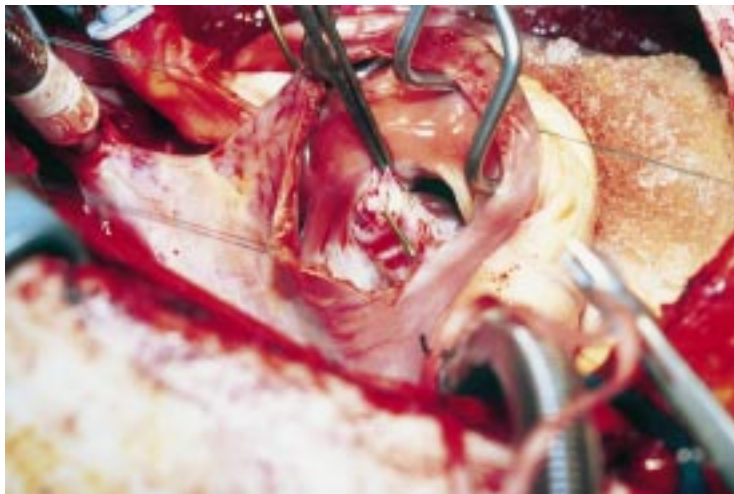


Figure 2 Operative view from the right atrium showing the probe passed through the fistula from the aortic side through the tricuspid valve and into the right atrium.

pericardial fluid. The patient had an uneventful recovery and was discharged home six days after the operation. Postoperative echocardiography showed no residual shunt and the patient remains well one year after surgery. Histology did not reveal any evidence of an inflammatory reaction or acute vasculitis.

### Discussion

Surgically important cardiac disease (aortic valve regurgitation, aneurysm of the sinus of Valsalva, pulmonary artery aneurysm, myocardial infarction, and endocarditis) may be the presenting feature of Behçet's disease.<sup>4-6</sup> Development of an aneurysm of the non-coronary aortic sinus with rupture into the right atrium and successful surgical repair has previously been reported.<sup>6</sup> However, in our case, we found no evidence of aneurysm formation either at surgery or at subsequent histological examin-

ation. We feel that the cause of the fistula in this case was vasculitis of the aortic wall with adherence and fistula formation to the right atrium. This type of aorto-atrial fistula without an aneurysm has been described in patients with rheumatoid arthritis but not previously in Behçet's disease.<sup>7</sup>

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