

# Aortic valve replacement in rheumatoid aortic incompetence

A B DEVLIN, P GOLDSTRAW, AND P K CAVES

From the University Department of Cardiac Surgery, Royal Infirmary, Glasgow G4 0SF, UK

**Devlin, A B, Goldstraw, P, and Caves, P K (1978).** *Thorax*, 33, 612-615. **Aortic valve replacement in rheumatoid aortic incompetence.** Rheumatoid aortic valve disease is uncommon, and there are few reports of valve replacement in this condition. Aortic valve replacement and partial pericardiectomy was performed in a patient with acute rheumatoid aortitis and aortic incompetence.

Previous reports suggest that any patient with rheumatoid arthritis who develops cardiac symptoms should be carefully assessed for surgically treatable involvement of the pericardium or heart valves.

The commonest symptomatic cardiac valvar lesion in patients with rheumatoid arthritis is aortic incompetence, but aortic stenosis (Lassiter and Tassy, 1965) and mitral regurgitation (Carpenter *et al*, 1967) have been reported. Rupture of the sinus of Valsalva with complete heart block has also been described (Howell *et al*, 1972).

This clinical experience contrasts with post-mortem studies that show the similarity of rheumatoid and rheumatic cardiac disease with mitral, aortic, tricuspid, and pulmonary valvar involvement occurring in descending order of frequency. Aortic valve replacement (Roberts *et al*, 1968) for aortic incompetence is the only recorded cardiac operation for valvar disease in such patients. Six cases have been reported (Barker, 1971; Iveson *et al*, 1975; Yates and Scott, 1975), although Iveson knows of two others (personal communication from M W J Boyd, 1974). We report in detail a seventh successful case.

## Case report

A 56-year-old man had suffered from classical rheumatoid arthritis for eight years. There was no history of rheumatic fever. The disease was widespread, nodular, erosive, and seropositive (rheumatoid factor present in titre of 1/128). His drug treatment consisted of indomethacin and chloroquine. No previous symptoms or signs had been attributed to the cardiovascular system.

In November 1976 he was admitted to hospital with left ventricular failure. The peripheral pulses

were collapsing in character, and his blood pressure was 170/35 mmHg. Auscultation showed loud aortic systolic and diastolic murmurs. He had "pistol-shot" femoral pulses. There was rheumatoid arthritis of his elbow, wrist, knee, and ankle joints.

His plain chest radiograph showed cardiomegaly with pulmonary oedema. His electrocardiogram showed left ventricular hypertrophy with strain, the cardiac rhythm varying between sinus tachycardia and atrial fibrillation. He was apyrexial.

The results of full blood count and routine biochemical investigations were normal. His ESR was 87 mm in the first hour, and he had persistent microscopic haematuria. Blood cultures were repeatedly sterile.

Acute aortic incompetence with an aortic systolic flow murmur was diagnosed, but the aetiology of the valve lesion was not certain. Subacute bacterial endocarditis was thought to be the probable diagnosis.

Although the patient improved rapidly on bed rest, digoxin, and diuretics, he later suffered two further episodes of acute left ventricular failure. He was transferred to Glasgow Royal Infirmary for further cardiological assessment. Results of tests for syphilis, Q fever, and systemic lupus erythematosus were all negative. He remained afebrile, and blood cultures were again sterile.

Echocardiography showed normal mitral valve movement. Cardiac catheterisation showed gross aortic regurgitation with a left ventricular end-diastolic pressure of 25 cm of water. Coronary

arteriography was normal. An intravenous pyelogram showed a left-sided hydronephrosis with a calculus at the pelviureteric junction.

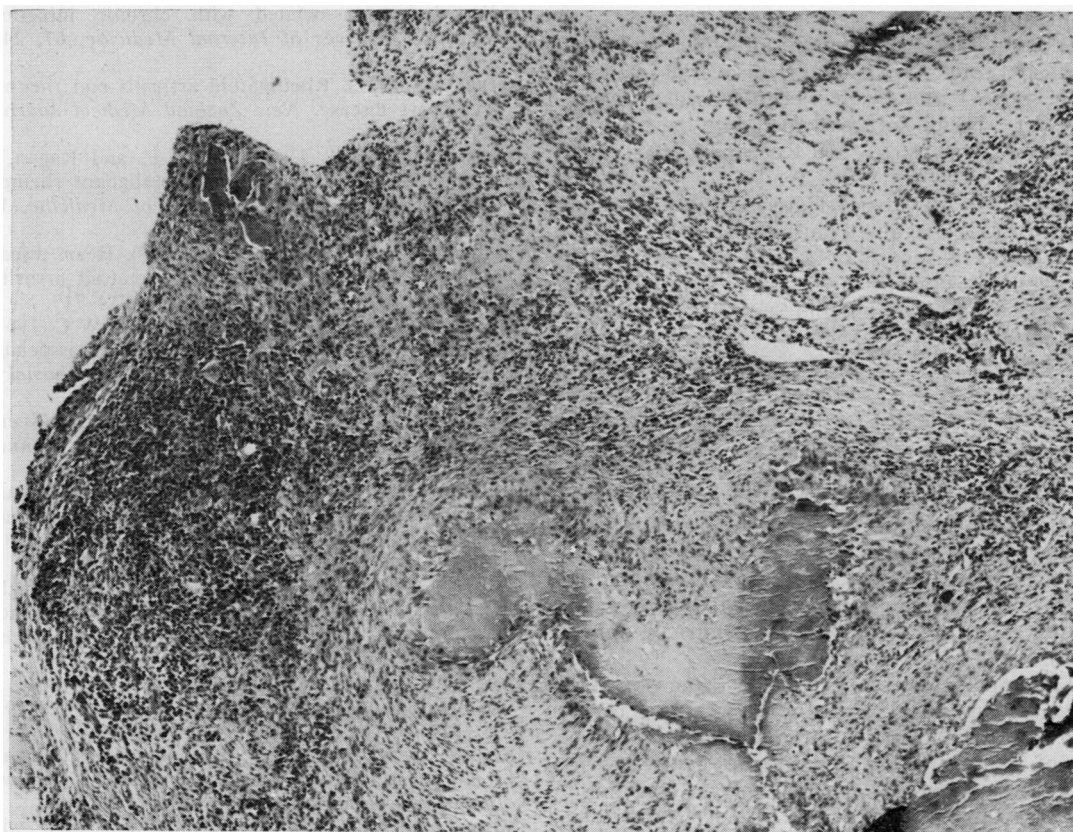
### **Operation**

On 6 January 1977, two months after the first cardiac symptoms, operation was performed through a median sternotomy. No evidence of pericarditis was seen. The aortic valve and aortic wall at the attachment of the leaflets appeared acutely inflamed. All three aortic leaflets were shortened and greatly thickened, especially at the annular attachment. That part of the mitral valve that could be seen appeared completely normal. The aortic valve leaflets were excised, their substance being almost fluid in places. A porcine xenograft (Hancock Laboratories) was sutured in place using interrupted horizontal mattress sutures.

The anterior pericardium was removed before closing the chest.

No organisms were grown from the excised valve leaflets. Histological sections showed that the normal structure of the valve was replaced by an inflammatory process, including foci of fibrinoid necrosis, which were surrounded by palisaded fibroblasts. Most of the inflammatory cells were of plasma cell type with only a few polymorphs. The appearances were considered diagnostic of rheumatoid nodules (see figure).

The patient recovered rapidly, and one year after his operation remains well. He has no cardiac symptoms and no signs of aortic incompetence. He remains in atrial fibrillation controlled by daily digoxin with a blood pressure of 120/70 mm Hg. His serial chest radiographs have shown considerable diminution in the size of the cardiac silhouette. His rheumatoid arthritis is easily managed with indomethacin.



**Figure** *Normal structure of valve is replaced by an inflammatory process including foci of fibrinoid necrosis of collagen, which are surrounded by palisaded fibroblasts. Most inflammatory cells are plasma cells with only a few polymorphs. Culture of valve did not result in growth of any pathogenic bacteria. Appearances are considered diagnostic for rheumatoid nodules (Haematoxylin and eosin  $\times 75$ ).*

## Discussion

Rheumatoid heart disease has been a well-recognised pathological entity and is now much more often diagnosed clinically. The commonest lesion seen is pericarditis with an incidence varying from 11–50% in different post-mortem studies (Fingerman and Andrus, 1943; Young and Schwedel, 1944). Careful clinical examination will detect the presence of pericardial effusion in 10% of patients with rheumatoid arthritis (Kirk and Cosh, 1969), but with ultrasound this figure rises to 34% with an even higher figure of 50% in the subgroup of patients with subcutaneous nodules (Bacon and Gibson, 1974). The vast majority of these pericardial lesions are clinically silent, but several case reports show that sudden death may occur from constrictive pericarditis or cardiac tamponade even in young patients with mild rheumatoid arthritis (Bevans *et al*, 1954; Stern and Sobel, 1961; Partridge and Duthie, 1963; Smyth, 1965; Latham, 1966).

Rheumatoid granulomata affecting the heart itself are much less common; an overall incidence of between 1 and 3% has been recorded in post-mortem studies of patients with rheumatoid arthritis (Baggenstoss and Rosenberg, 1941; Bonfiglio and Atwater, 1969).

Reports of valvar heart disease due to rheumatoid arthritis diagnosed during life are rare. We have found six reports of aortic valve replacement for rheumatoid aortic incompetence (Barker, 1971; Iveson *et al*, 1975; Yates and Scott, 1975), although the certainty of the diagnosis in one of Iveson's cases appears doubtful. Aortic valve replacement was successful in five patients, the sixth patient dying after operation from generalised vasculitis.

A notable clinical feature in our patient was the short period over which he developed severe aortic regurgitation. Because of this and the presence of a high ESR and haematuria, subacute bacterial endocarditis was diagnosed, despite the persistently negative blood cultures. Surgical intervention became urgent because of his progressive cardiac failure. The true pathology was confirmed only at operation.

The important technical point during operation in this condition is the soft, fleshy nature of the acutely inflamed aortic annulus and root. The use of interrupted sutures, supported if necessary with pledgets of felt, would appear essential if a paraprosthesis leak is to be avoided. Simultaneous pericardiectomy would also appear advisable in view of the risk of subsequent pericardial involvement in the rheumatoid process.

The occurrence of clinically significant heart

disease is not related in any reliable way to the severity of rheumatoid arthritis, although it tends to be commoner in patients with subcutaneous nodules. Thus any patient with rheumatoid arthritis who develops cardiac symptoms should be carefully assessed since surgery may be needed for persistent pericardial effusion or progressive valvar dysfunction.

We wish to thank Dr Ian Hutton, consultant cardiologist, for his referral of this patient and Dr H W Simpson, consultant pathologist, both of Glasgow Royal Infirmary, for the histological report on the valve and the illustration.

## References

- Bacon, P A, and Gibson, D G (1974). Cardiac involvement in rheumatoid arthritis—an echocardiographic study. *Annals of the Rheumatic Diseases*, **33**, 20–24.
- Baggenstoss, A H, and Rosenberg, E F (1941). Cardiac lesions associated with chronic infectious arthritis. *Archives of Internal Medicine*, **67**, 241–258.
- Barker, A (1971). Rheumatoid arthritis and rheumatoid heart disease. *New Zealand Medical Journal*, **73**, 14–17.
- Bevans, M, Nadell, J, Demartini, F, and Ragan, C (1954). The systemic lesions of malignant rheumatoid arthritis. *American Journal of Medicine*, **16**, 197–211.
- Bonfiglio, T, and Atwater, E C (1969). Heart disease in patients with seropositive rheumatoid arthritis. *Archives of Internal Medicine*, **124**, 714–719.
- Carpenter, D F, Golden, A, and Roberts, W C (1967). Quadriculvalvular rheumatoid heart disease associated with left bundle branch block. *American Journal of Medicine*, **43**, 922–929.
- Fingerman, D L, and Andrus, F C (1943). Visceral lesions associated with rheumatoid arthritis. *Annals of the Rheumatic Diseases*, **3**, 168–181.
- Howell, A, Say, J, Hedworth-Whitty, R (1972). Rupture of the sinus of Valsalva due to severe rheumatoid heart disease. *British Heart Journal*, **34**, 537–540.
- Iveson, J M I, Thadani, V, Ionescu, M, and Wright, V (1975). Aortic valve incompetence and replacement in rheumatoid arthritis. *Annals of the Rheumatic Diseases*, **34**, 312–320.
- Kirk, J, and Cosh, J (1969). The pericarditis of rheumatoid arthritis. *Quarterly Journal of Medicine*, **38**, 397–423.
- Lassiter, G S, and Tassy, F T (1965). Malignant rheumatoid disease with aortic stenosis. *Archives of Internal Medicine*, **116**, 930–936.
- Latham, B A (1966). Pericarditis associated with rheumatoid arthritis. *Annals of the Rheumatic Diseases*, **25**, 235–241.
- Partridge, R E M, and Duthie, J J R (1963). Rheumatoid pericarditis. *British Medical Journal*, **1**, 611.



- Roberts, W C, Kehoe, J A, Carpenter, D F, and Golden, A (1968). Cardiac valvular lesions in rheumatoid arthritis. *Archives of Internal Medicine*, **122**, 141–146.
- Smyth, H A (1965). Rheumatoid arthritis and constrictive pericarditis. (Discussion. Lange, R K, Weiss, T E, and Ochsner, J L). *Arthritis and Rheumatism*, **8**, 403–417.
- Stern, J B, and Sobel, H J (1961). Haemorrhagic rheumatoid pericarditis. *American Journal of Cardiology*, **8**, 670–673.
- Yates, D B, and Scott, J T (1975). Cardiac valvular disease in chronic inflammatory disorders of connective tissue—factors affecting survival after surgery. *Annals of the Rheumatic Diseases*, **34**, 321–325.
- Young, D, and Schwedel, J B (1944). The heart in rheumatoid arthritis. *American Heart Journal*, **28**, 1–23.

Requests for reprints to: Professor P K Caves, Department of Cardiac Surgery, Royal Infirmary, Glasgow G4 0SF.