

## Mitral valve replacement for mitral stenosis caused by Libman-Sacks endocarditis

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**SUMMARY** A woman who developed mitral stenosis from Libman-Sacks endocarditis is described. The mitral valve was replaced by a Starr-Edwards prosthesis. One year later, despite her being maintained on steroids and azathioprine, the verrucous endocarditis progressed to cause sudden, severe dysfunction of the prosthetic valve.

A non-bacterial verrucous endocarditis which was later shown to be associated with systemic lupus erythematosus was described by Libman and Sacks in 1924. Since then endocardial involvement found at necropsy in patients with systemic lupus erythematosus has been reported in 13 to 63 per cent of cases (Kong *et al.*, 1962; Heijtmancik *et al.*, 1964; Bulkley and Roberts, 1975a). Valvular involvement rarely produces significant haemodynamic dysfunction (Kong *et al.*, 1962; Heijtmancik *et al.*, 1964), and only 4 instances of valve replacement have been reported. Shulman and Christian (1969) described 3 patients with aortic regurgitation, 1 of whom required valve replacement and a further case was added by Oh *et al.* (1974). Mitral valve replacement has been described by Murray *et al.* (1975) and 1 has been reported from the National Institute of Health, Bethesda, Maryland (Myerowitz *et al.*, 1974; Paget *et al.*, 1975). We report a patient with systemic lupus erythematosus who presented with obstruction of the mitral valve and after mitral valve replacement proceeded to obstruction of the prosthesis within one year of operation. As far as we know, no case has been previously described in which active verrucous endocarditis has progressed to cause serious dysfunction of a prosthetic valve.

### Case report

A 36-year-old housewife who had no past history of rheumatic fever was admitted in January 1975. For 5 months she had suffered from a febrile illness associated with myalgia and painful swollen knee and ankle joints. One month before admission she experienced 3 episodes of pleuritic chest pain. Two weeks later she noted shortness of breath on one flight of stairs and developed a dry cough, trouble-

some at night. On examination she was normotensive and in sinus rhythm. There were no signs of congestive cardiac failure. The pulmonary component of her second heart sound was loud and a short soft diastolic murmur was heard at the apex. The rest of her physical examination was normal. Investigations showed her haemoglobin to be 12.1 g/dl, white cell count 11 000/mm<sup>3</sup> with a normal differential, ESR 55 mm/h, blood urea 6 mmol/l. Antinuclear antibody was negative but lupus erythematosus cells were present. Blood cultures were consistently negative. Electrocardiogram showed sinus rhythm and left atrial hypertrophy. Echocardiography indicated mitral valve disease with thickening and rigidity of the valve cusps. The intracardiac pressures found at cardiac catheterisation were as follows: right atrium: mean 9 mmHg; right ventricle: 50/0-10 mmHg; pulmonary artery: 50/24 mmHg; pulmonary wedge pressure: mean 31 mmHg; left ventricle: 125/0-13 mmHg; aorta: 125/80 (mean 95) mmHg. A left ventricular angiogram showed a filling defect arising from the interventricular septum. This was thought to be either a thickened papillary muscle or vegetations associated with the mitral valve (Fig. 1).

A diagnosis of systemic lupus erythematosus was made and it was thought the patient had endocardial involvement affecting the mitral valve. The patient was treated with frusemide, prednisone 30 mg daily, and azathioprine 100 mg daily and her condition improved.

In March 1976 she was readmitted with increasing shortness of breath and ankle oedema. She was afebrile. Her jugular venous pressure was raised 2 cm and she had fine crepitations at both bases. The apical diastolic murmur was still present. Electrocardiogram showed sinus rhythm and there was pulmonary venous congestion on the

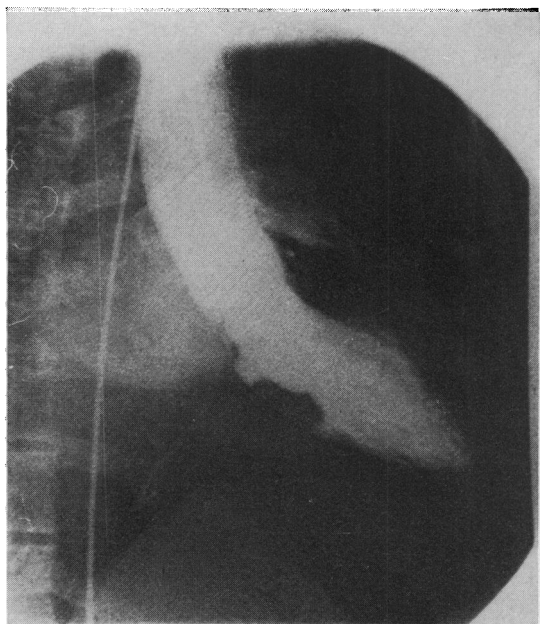


Fig. 1 Left ventriculogram showing a filling defect which was thought to be vegetations associated with the mitral valve.

chest x-ray film. Her haemoglobin was 10.3 g/dl, white cell count 12 000/mm<sup>3</sup>, with a normal differential and ESR 74 mm/hour.

In view of her deteriorating clinical condition and the presence of mitral stenosis it was decided to explore her mitral valve. At operation the orifice of the mitral valve was almost completely obscured by a plaque of red granulation tissue resembling clot. A tiny aperture at the anterolateral commissure was the only communication to the left ventricle. The tissue extended onto the left atrial wall and measured 5 cm in diameter by 0.5 cm in thickness. It was so adherent to the valve cusp that it could not be separated. The valve was, therefore, excised with it and the affected left atrial wall was scraped clean. The valve was replaced by a No. 3M Model 6120 Starr-Edwards prosthesis without complication. There was no evidence of disease in the left ventricle.

Examination of the valve showed it to be thickened and fibrotic with increased vascularity. A large fibrinous vegetation was attached to its upper surface. Areas of fibrinoid degeneration were present within the valve and the histological appearances were considered to be consistent with the diagnosis of Libman-Sacks endocarditis. No infective agent was found (Fig. 2).

The patient made an uncomplicated recovery and was discharged on prednisone 10 mg daily, azathio-

prine 100 mg daily, warfarin, and frusemide.

She remained well on the above regimen until January 1977 when she developed more discomfort and swelling in the knees and ankles and her ESR was found to have risen from 42 to 62 mm/hour. Prednisone was increased to 15 mg daily. In February 1977 she was readmitted as an emergency with a 3-day history of increasing shortness of breath, haemoptysis, and paroxysmal nocturnal dyspnoea. On examination she was cold and clammy. Her pulse rate was 100 per minute, blood pressure 90/60 mmHg, and there were signs of severe pulmonary oedema. A loud pansystolic murmur was heard at the apex and radiated to the axilla. It was thought she had developed acute mitral regurgitation with resultant cardiac failure. Her condition deteriorated rapidly after admission and she had a cardiac arrest. She was immediately resuscitated, taken to theatre and put on cardiopulmonary bypass. At operation the ball of the mitral prosthesis was found to be fixed in the open position. Three-quarters of the orifice below the valve seating was obstructed by friable, acellular tissue leaving a small fixed opening which was both obstructive and regurgitant. When the prosthesis was removed the left ventricular cavity was found to contain a large mass of similar tissue attached to the posterior wall. This was removed with the prosthesis and a new No. 4M Model 6120 Starr-Edwards valve was inserted.

Postoperatively the patient remained critically ill. She was unconscious, unresponsive, and developed epileptic fits. She became anuric and required peritoneal dialysis. In spite of all supportive measures her condition deteriorated and she died 4 days later.

Histological examination of the tissue removed from around the prosthetic valve showed it to consist predominately of collagen, areas of which were necrotic. It was relatively acellular but occasional plasma cells were present. On the surface were both organised and unorganised thrombus. Necropsy showed severe bilateral pulmonary oedema and hepatic venous congestion. There was a right-sided cerebral infarction. Both kidneys showed the gross appearances of acute tubular necrosis and on histological examination the characteristic lesions of systemic lupus erythematosus were seen.

## Discussion

The initial diagnosis of systemic lupus erythematosus in this patient was made on the history of arthritis without deformity, the evidence of pleural involvement, a raised sedimentation rate, and the presence of lupus erythematosus cells. When a

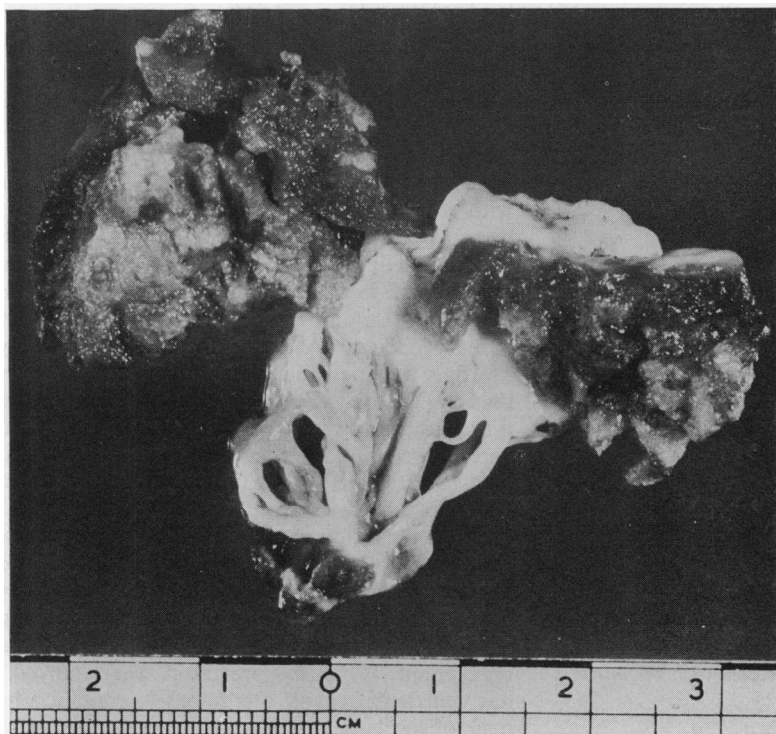


Fig. 2 Excised mitral valve with adherent vegetations.

diastolic murmur at the apex was heard the possibility of Libman-Sacks endocarditis was raised. A murmur is the commonest initial cardiovascular manifestation (Kong *et al.*, 1962). There is, however, a poor correlation between the appearance of a murmur and the presence of Libman-Sacks valvulitis at necropsy (Shearn, 1959; Kong *et al.*, 1962). Mitral valve involvement was demonstrated in this case by echocardiography and cardiac catheterisation.

The left ventricular filling defect seen at cardiac catheterisation (Fig. 1) was thought to be a mass of verrucous material associated with the mitral valve. A similar appearance has been reported by Seningen *et al.* (1974). At the initial operation, however, no mass was seen and the left ventricle was grossly free of disease. At the second operation a large mass of tissue was found attached to the posterior wall of the left ventricle and this was the cause of the prosthetic valve dysfunction.

The pathology of the endocarditis has been well described (Gross, 1940; Klemperer *et al.*, 1941). Histological examination of the tissue removed from this patient did not show all the classical features but the absence of lupus erythematosus cells and haematoxyphil bodies has been reported in other cases (Oh *et al.*, 1974) and was attributed to the

effect of steroid therapy. Recently Bulkley and Roberts (1975a) have also drawn attention to the alteration to pathology induced by corticosteroid therapy. They postulate that 'healing' of Libman-Sacks endocarditis may occur by fibrosis and calcification. This was rare before the advent of steroids but now, either as a direct effect of the drug or as a result of increased longevity of the patients, this 'healing' process is seen more commonly. One consequence of this could be to increase the incidence of valve dysfunction by scarring (Bulkley and Roberts, 1975a).

The distribution of the endocardial lesions also has been modified by steroid therapy. Previously all four valves were commonly involved but now there is a preponderance of left-sided lesions affecting particularly the mitral valve (Heijtmancik *et al.*, 1964; Bulkley and Roberts, 1975b). In the two previously reported cases of mitral valve replacement in this condition the lesions had produced mitral regurgitation. It is of interest that in this patient the vegetations produced predominant mitral stenosis.

Our patient had an initially successful mitral valve replacement but over the course of a year the endocarditis progressed to produce malfunction of the prosthesis, which was ultimately fatal. During this period therapy with prednisone and azathiop-

prine was maintained and though her sedimentation rate had remained moderately raised at about 42 mm/hour she had been asymptomatic. One month before her final admission she developed a recurrence of joint pains and the dose of prednisone was increased. Perhaps, in spite of being asymptomatic, the therapy was inadequate and she should have been on higher doses. However, Heijtmancik *et al.* (1964) question the value of steroids in preventing Libman-Sacks endocarditis and the history of this patient may provide further evidence to support this view.

We, therefore, offer a warning that the endocardial manifestations of the disease can progress despite therapy, and relatively silently, to cause sudden and severe dysfunction of a prosthetic valve. In addition, we feel that the disease behaves like a malignant growth with local extensions, and perhaps the affected portion of left atrial wall, found at the initial operation, should have been excised.

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