

Long-term follow-up of atrial myxoma

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Experience of 12 patients with atrial myxoma over a period of 10 years is reviewed. Eleven patients had a left atrial tumour and one a right atrial tumour.

Successful surgical removal was carried out in 9 (8 left, and 1 right). Three patients died before operation, the diagnosis having been made in life in 2 of them.

Seven patients with left atrial myxoma have been followed for 5 to 10 years after operation. Two died after 4 and 6 years, respectively, from probable neoplasia of the lungs without evidence of a recurrence of the myxoma.

All the remaining 5 patients are well and without any evidence of recurrence, apparently having been cured of the condition.

Angiocardiography and cardiac catheterization in 4 patients revealed relief of left atrial and pulmonary arterial hypertension. There was no evidence of a space-occupying lesion in the left atrium.

The importance of early diagnosis and surgical treatment is stressed, and the advisability of removing part of the atrial septum surrounding the tumour is commented upon. The value of echocardiography in establishing the diagnosis, and thus avoiding the necessity for angiography, is discussed. It is concluded that though recurrence, which has been reported in 2 cases, is rare, regular follow-up is advisable.

Atrial myxoma is the commonest primary cardiac tumour (Leach, 1947). The features of left atrial obstruction, systemic embolism, and systemic illness are now well known and the profile of the disorder was reviewed by Greenwood (1968).

In 1962, we (Goodwin *et al.*) reported our initial experience in the diagnosis and management of 4 patients with left atrial myxoma, 3 of whom came to successful surgical treatment.

We now review the fate of 12 patients with atrial myxoma over a period of 11 years, with emphasis on the long-term results of surgical treatment.

Patients

Three patients died before operation. In the first the diagnosis was not made until necropsy, the significance of the association of severe pulmonary hypertension, signs suggesting mitral valve disease, haemolytic anaemia, and fever not having been appreciated (McGregor and Cullen, 1959). The second patient who was reported in 1962

(Goodwin *et al.*) was diagnosed, but died before operation. The third patient was admitted *in extremis* as a result of left atrial obstruction, and died after diagnostic investigation.

Of the remaining 9 patients, 8 had a left and one a right, atrial myxoma. The tumour was successfully removed in all. The patient with the right atrial myxoma (Case 6) was a girl of 17 years, diagnosed and referred for operation by Dr. Ian Gray of Coventry. The clinical and angiographic features were characteristic (Barlow, Fuller, and Denny, 1962) and the postoperative course has been uneventful. There are now no abnormal signs, and angiography is normal.

Surgical technique

The technique of the operation has remained constant (Goodwin *et al.*, 1962). All patients have been operated upon under cardiopulmonary bypass, the left atrium being approached through the right atrium and atrial septum. The tumour is gently delivered and the tumour pedicle and the surrounding portion of atrial septum completely excised. Aortic occlusion during manipulation of the tumour and careful inspection of the chambers of the heart, and lavage, is essential before closure. A special sucker is used, the contents of which are discarded.

TABLE I *Atrial myxoma: long-term follow-up*

Case No.	Sex	Age	Symptoms and duration (yr)	Operation	Result	Postoperative study
1	F	24	Dyspnoea 2	1960	Clinical cure	—
2	F	47	Dyspnoea; syncope 1	1961	Clinical cure	9 yr; all normal
3	F	44	Dyspnoea 1	1963	Clinical cure	7 yr; all normal
4	M	42	Embolism 1	1963	Clinical cure	Declined
5	F	45	Dyspnoea; palpitations 2	1965	Well; jugular venous pressure raised; systolic murmur	5 yr; right ventricular end-diastolic pressure raised
6	F	17	Tiredness; dyspnoea 6 mth	1968 (right atrial)	Clinical cure	1 yr; all normal
7	F	47	Syncope; dyspnoea 3	1969	Clinical cure	2 wk; all normal

Immediate post-operative results

In all patients improvement was dramatic. Raised left atrial and pulmonary artery pressure fell, as reported in our first 4 patients (Goodwin *et al.*, 1962). Symptoms of a raised left atrial pressure (dyspnoea, cough, haemoptysis) disappeared and the increase in cardiac output resulted in improvement in well-being. Embolism, fever, and other constitutional features resolved, and the serum proteins returned towards normal (Goodwin *et al.*, 1962). The cachectic features resolved, and patients who had lost weight rapidly regained it. Abnormal auscultatory features disappeared in all save one patient.

Long-term results

Five of our patients with left atrial myxoma have been followed for from 5 to 10 years after operation (Table 1). All remained well, and with no symptoms or signs to suggest recurrence of the tumour, but 2 have died, 4 and 6 years after operation, respectively. Both were men, aged 56 and 68 years, respectively, heavy smokers, who developed signs and

symptoms of bronchial neoplasia, which was confirmed in one by bronchoscopy. Unfortunately, necropsies were not obtained, as the patients died elsewhere and outside hospital, but there was no evidence of recurrence of the atrial myxoma. The remaining 5 patients are healthy and symptom free. One, a girl of 24 years at the time of her operation in 1960, has not been seen since 1965, when she was well, and free from symptoms and abnormal physical signs (Patient 3 in our original report) (Goodwin *et al.*, 1962). The other 4 patients have been observed for periods of 9, 7, 7, and 5 years, respectively. All are well and without symptoms. None has signs of mitral disease, except for one (Case 5) a woman of 45 years who has a soft apical systolic murmur, and slight rise in the jugular venous pressure. Clubbing of the fingers has disappeared, and evidence of systemic illness has almost entirely resolved (Table 2).

Erythrocyte sedimentation rate, serum proteins, and haemoglobin results The results are shown in Table 2 which also includes our most recent patient (Case 7) who

TABLE 2 *Pre- and postoperative erythrocyte sedimentation rate (ESR), haemoglobin, and protein electrophoresis in 7 patients with left atrial myxoma*

Case No.	ESR (mm/hr)		Serum protein electrophoresis		Hb (g/100 ml)		
	Preop	Postop	Preop	Postop	Preop	Postop	
3	13-23	10	γ ↑	Normal	12.4	13.6	7 yr
5	44	6	α_2 ↑	Normal	12.1	13.9	7 yr
2	15	28	α_2 ↑	Normal	11.6	12.2	9 yr
8	8, 15, 36, 27	14, 10	α_2 ↑	↑ Slightly	14.1	14.9	5 yr
7	10	15	Normal	↑ Slightly	15.2	14.5	2 mth
9	75	7	α ↑	Normal	11.8	13.2	Died 4 yr; cancer of bronchus
10	10, 34, 60	10, 112	γ ↑	Normal	11.6	13.1	Died 6 yr; cancer of bronchus

Case 1, who was well when last seen in 1965, has been omitted from the Table as no recent data are available.

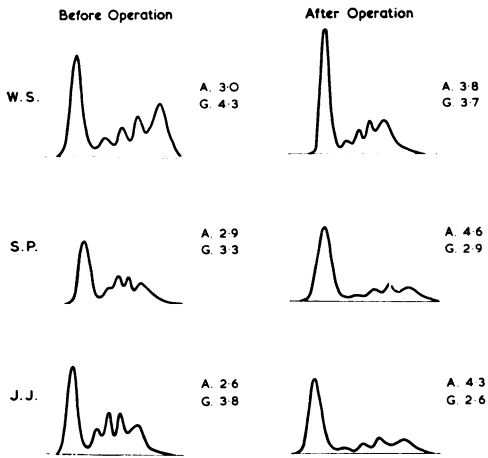


FIG. 1 Serum proteins before and shortly after operation in Case 10 (W.S.), Case 1 (S.P.), and Case 2 (J.J.); showing normal albumin/globulin ratio after operation. (Reproduced from Goodwin et al. (1962) by permission of the Editor and publishers.) (A, albumin g/100 ml; G, globulin g/100 ml).

was operated on 6 months ago. The erythrocyte sedimentation rate was raised before operation in all save this patient, and became normal in all except Case 2.

Serum protein electrophoresis showed increased gamma-globulin in 2, and alpha-globulin in 4 patients; only Case 7 showing a normal pattern before operation. Fig. 1 shows the serum protein pattern before and shortly after operation in 3 of our first 4 patients who have been previously reported (Goodwin et al., 1962). The protein pattern became completely normal in 5 patients, slight residual abnormalities being present in 3.

Haemodynamic studies In view of the reports of recurrence of atrial myxoma (Bahl et al., 1969) 4 of the 5 available patients have been studied after removal of the tumour at 9, 7, 5 years, and 3 weeks, respectively. The fifth

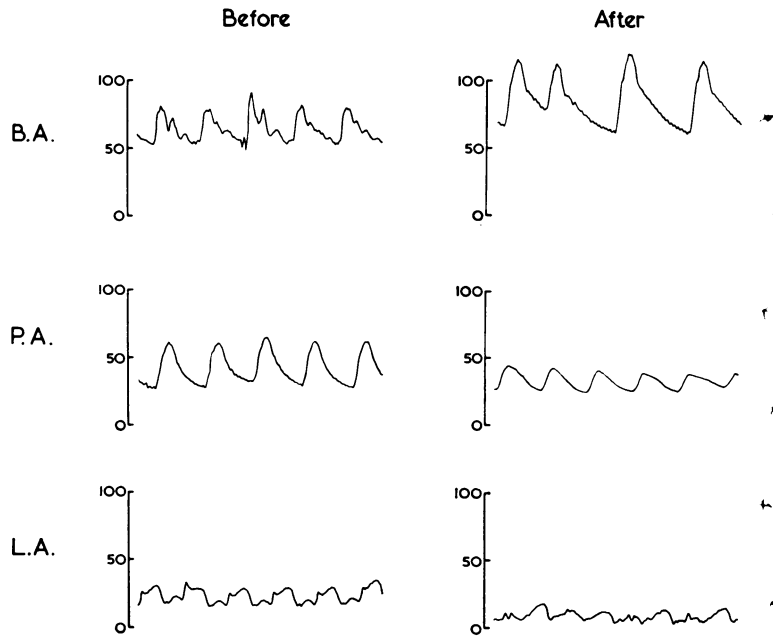


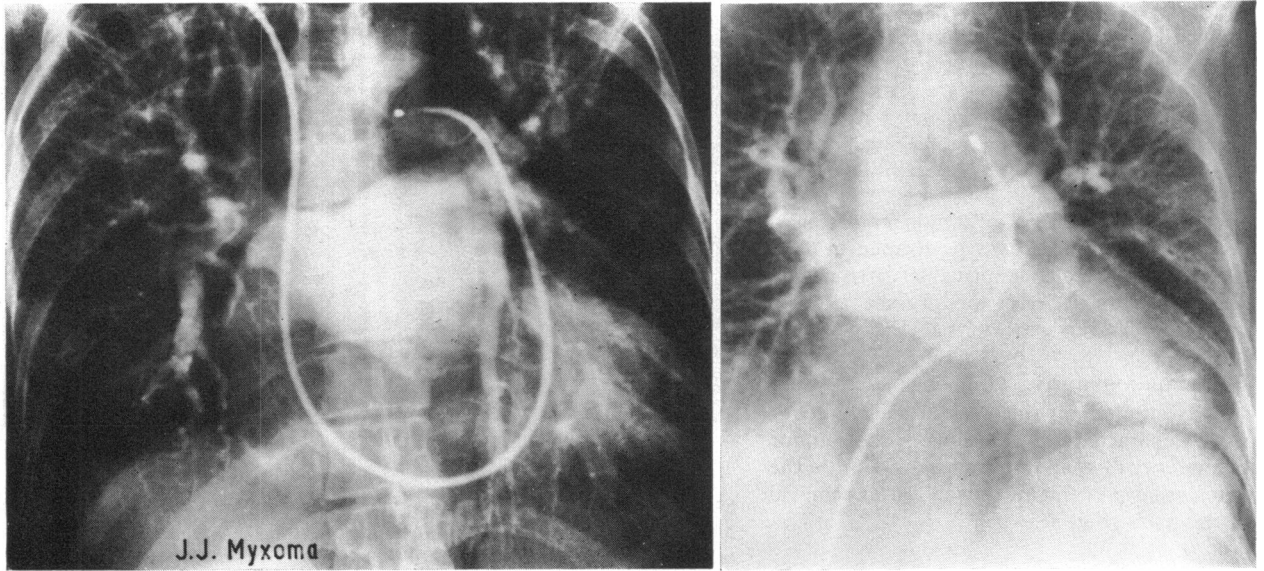
FIG. 2 Brachial artery (BA), pulmonary artery (PA), and left atrial (LA) pressures before and immediately after removal of left atrial myxoma in Case 2, showing an immediate rise in brachial artery pressure, and fall in left atrial and pulmonary artery pressures. (Reproduced from Goodwin et al. (1962) by permission of the Editor and Publisher.)

patient (Case 4) declined investigation. The results of cardiac catheterization are shown in Table 3. In all patients the left atrial pressure, which was greatly increased before operation showed a fall to 25-30 per cent of the pre-operative level. Likewise, the pulmonary artery pressure fell, and the cardiac index rose in 3 patients.

The haemodynamic improvement tended to occur immediately the tumour was removed as shown in Case 2, our original patient No. 4 (Fig. 2). Our most recent patient (Case 7),

TABLE 3 Pre- and postoperative haemodynamic data in 4 patients with left atrial myxoma

Case No.	Time from operation of postop study	Pulmonary arterial pressure (mmHg) Mean		Indirect 'Left atrial' pressure (mmHg)		Cardiac index (l./min/m ²)	
		Preop	Postop	Preop	Postop	Preop	Postop
3	7 yr	44	15	38	10	2.2	3.0
8	5 yr	55	17	30	10	2.8	2.5
2	9 yr	50	15	25	8	2.1	3.2
7	3 wk	75	40	27	10	1.8	2.3



(a)

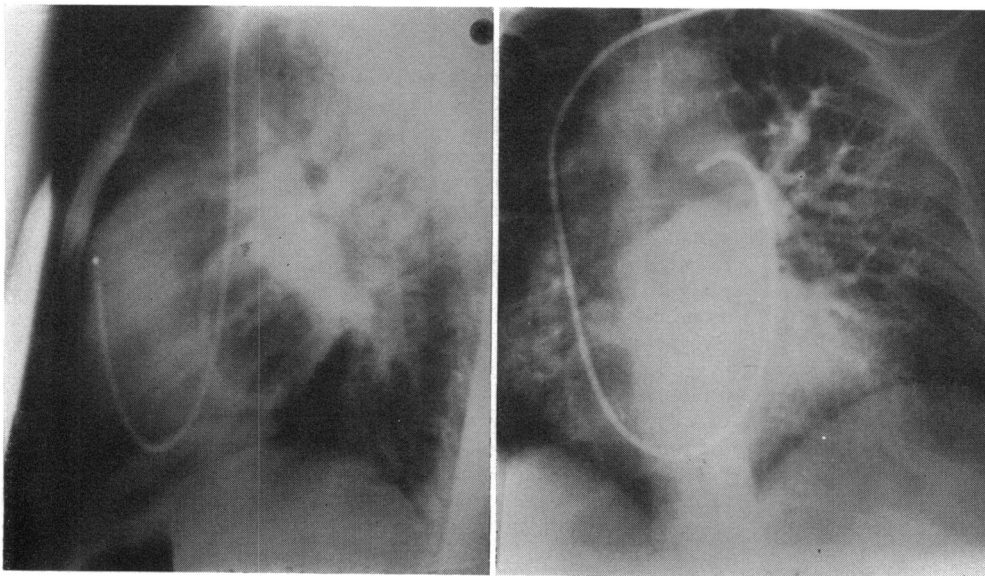
(b)

FIG. 3 Pulmonary arteriogram (anteroposterior projection), in Case 2. (a) Before removal of myxoma in 1961 showing filling defect between left atrium and left ventricle due to the myxoma. Contrast medium can be seen remaining in the medium-sized pulmonary arteries which show irregularity and tortuosity, with reduction in calibre at the bases indicating pulmonary hypertension due to a raised left atrial pressure. (Mean pulmonary artery pressure, 50 mmHg; mean left atrial pressure, 25 mmHg.) (b) In 1970 after removal of the tumour, showing a normal left atrium and left ventricle: no contrast medium remains in the pulmonary arteries, and the pulmonary vascular pattern is normal (Table 3).

FIG. 4 (a) Pulmonary arteriogram, lateral projection, in Case 3 before operation in 1963, showing filling defect in the left atrium due to myxoma. (b) Frontal projection showing normal left atrium and ventricle, in 1970.

(a)

(b)



who was studied 3 weeks after operation showed equally impressive figures. These results suggest that the disproportionately high pulmonary artery pressures are maintained by vasoconstriction which is released when the pulmonary venous pressure has been effectively reduced.

Angiocardiography Pulmonary arteriography in two planes was performed in all 4 patients and revealed a normal left atrium. No pericardial filling defect was present, and the pulmonary vessels were normal (Fig. 3 and 4).

Echocardiography Only one patient (Case 7) was studied by this technique which revealed characteristic echoes from the tumour before operation and normal movement of the anterior cusp of the mitral valve after removal of the mass (Fig. 5).

Residual cardiovascular signs All the abnormal cardiac features of left atrial myxoma, such as the widely split first heart sound due to delayed mitral valve closure, and the dull, long, early diastolic sound due to the tumour entering the left ventricle, disappeared in all patients.

Diastolic murmurs due to interference with mitral valve function and obstruction to left ventricular inflow disappeared in all patients. Systolic murmurs due to interference with mitral valve closure by the tumour and mitral regurgitation disappeared in all but one patient (Case 5) in whom a soft systolic murmur remained, which was associated with raised right ventricular end-diastolic pressure and jugular venous pressure. No evidence of recurrence of the myxoma was seen on angiography and no signs of obstruction to the tricuspid valve or of a right atrial tumour were found, but right atrial angiography was not performed. The mean left atrial pressure was 10 mmHg and the mean pulmonary artery pressure was 17 mmHg after operation. It is probable that the murmur represents minimal mitral regurgitation due to slight residual deformity of the mitral valve caused by the tumour. The raised right ventricular end-diastolic pressure is unexplained. It is of interest that in our second patient, who died before operation, the mitral valve cusps were found to be abraded and deformed by the tumour and two chordae tendineae were severed. A systolic murmur had been heard (Goodwin *et al.*, 1962).

Electrocardiography In all patients signs of right ventricular hypertrophy due to pulmonary hypertension disappeared.

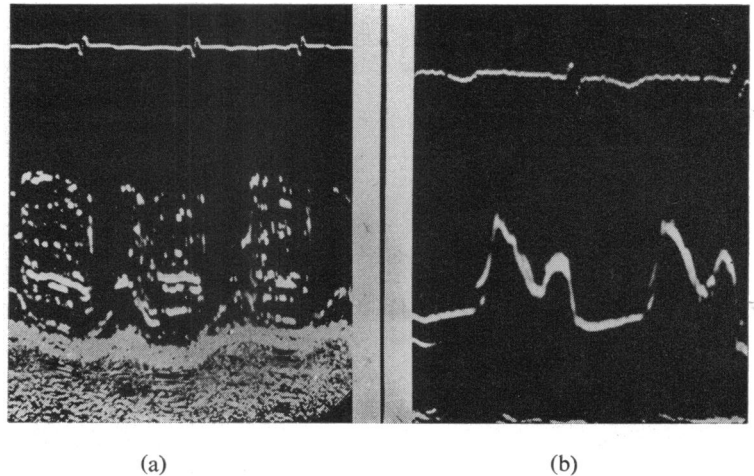


FIG. 5 Echogram of mitral valve and left atrium in Case 7. (a) Before operation showing massive echoes from the tumour. (b) After removal showing normal thickness, amplitude of movement, and type of movement of the anterior cusp of the mitral valve.

Chest radiography The vascular patterns of raised pulmonary venous pressure and interstitial pulmonary oedema, when present, reverted to normal.

Discussion

Following the first report of successful removal of left atrial myxoma (Crafoord, 1955) there have been numerous reports of successful removal without recurrence (Newman, Cordell, and Prichard, 1966; Firor, Aldridge, and Bigelow, 1966) and this seems to be the usual expectation. However, 2 recurrences have been reported, one 4 years after (Gerbode, Kerth, and Hill, 1967) and the other 6 years after removal (Bahl *et al.*, 1969). Gerbode *et al.* (1967) advocate removal of the entire atrial septum and replacement with a Dacron patch, and employed this technique in 4 of their 7 patients. Bahl *et al.* (1969), who reported the recurrence in the other patient, excised the endocardium underlying the tumour but left the atrial septum intact. Our own practice has been to excise a portion of the atrial septum around the base of the tumour.

Apart from the possibility of recurrence of the tumour, the question of malignancy must be considered. The pathological features of atrial myxoma suggest that it is a non-malignant tumour, and no malignant or invasive features have so far been reported in the left atrium or in tumour emboli lodged in peri-

pheral arteries. However, the report by Heath and Mackinnon (1964) of invasion of the wall of pulmonary arteries by emboli from a right atrial myxoma is disquieting, though they regarded this limited invasion as merely further evidence that myxomata are true neoplasms rather than degenerating thrombi.

Interference with the mitral valve by the myxoma, which damages the cusps and may rupture chordae, can result in permanent mitral valve abnormality, but this has not been a problem, though our second patient had two ruptured chordae, and one of our successfully operated patients has evidence suggestive of mild residual mitral regurgitation. Damage to the tricuspid valve from right atrial myxoma seems to be more common than damage to the mitral valve in left atrial myxoma, perhaps because right atrial myxomata appear to calcify more readily than left atrial myxomata, and complete destruction of the tricuspid valve, requiring valve replacement, has been reported by Proctor Harvey (1968).

The available evidence suggests that recurrence of atrial myxoma is improbable but that excision of the atrial septum around the pedicle is a wise precaution.

Regular follow-up by clinical examination, estimation of haemoglobin, erythrocyte sedimentation rate, and serum proteins is advisable. Echocardiography has already been shown to be of value in the detection of left atrial tumours (Wolfe, Popp, and Feigenbaum, 1969), though it may not reveal a small tumour if the myxoma is not in contact with the mitral valve. The technique should also provide a valuable assessment of postoperative mitral valve function.

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Addendum

Case 7 was reported to the Clinical Section of the Royal Society of Medicine in 1970, and remains well and without abnormal signs 15 months after the operation.

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