THE SYNDROME OF ACUTE MITRAL REGURGITATION — CLINICAL RECOGNITION, INVESTIGATION AND MANAGEMENT

B L Chia
B Ee
A Tan
M Choo
L Tan
N C Tan
B Singh
E P K Mah
S Quek

University Department of Medicine (II)
Singapore General Hospital
Outram Road
Singapore 0316

B L Chia, MBBS, FRACP, FACC
Professor and Head

A Tan, MBBS, AM, FRACP
Senior Lecturer

M Choo, MBBS, MRCP, M MED
Senior Lecturer

University Department of Medicine (I)
Singapore General Hospital

B Ee, MBBS, AM, MRCP, M MED
Senior Lecturer

Department of Diagnostic Radiology
Singapore General Hospital

L Tan, MBBS, AM, FRCR
Senior Radiologist

Department of Cardiothoracic Surgery
Singapore General Hospital

N C Tan, MBBS, FRACS, AM, FACS, FACC, FICA
Senior Surgeon and Head

Department of Cardiology
Singapore General Hospital

S Quek, MBBS, AM, M MED
Senior Registrar

Youngberg Memorial Adventist Hospital

B Singh, MBBS, M MED, MRCP
Chief of Staff

Department of Medicine, Alexandra Hospital

E P K Mah, MBBS, AM, FRACP
Consultant Physician

SYNOPSIS

The syndrome of acute mitral regurgitation due to either chordal or papillary muscle rupture usually presents dramatically with acute pulmonary oedema and requires early diagnosis and surgical treatment. This paper describes our experience with 9 patients. Five patients (group I cases) with no known previous cardiac disease presented with sudden cardiac failure. Clinical examination revealed a loud apical pansystolic murmur with a fourth heart sound. The ECG's of these 5 patients were normal and their chest xrays showed either a normal sized heart or minimal cardiomegaly. In contrast, the 4 patients in group II all had previous cardiac disease. They also presented with sudden onset of cardiac failure and apical systolic murmurs. However, unlike the group I cases, considerable cardiomegaly detected clinically and radiologically was present in these patients. In 3 of the 9 patients in both group I and II, the chordal rupture was due to infective endocarditis. Two dimensional echocardiography is an excellent tool for confirming chordal rupture and was positive in 4 out of the 5 patients studied. Left ventricular angiography showed severe mitral regurgitation in all 9 patients. All the patients could be stabilised on vigorous medical therapy and subsequently underwent surgery. Rupture of the chordae tendinae to the anterior and posterior mitral valve leaflet was seen in 4 patients each. In the last patient, rupture of both papillary muscles resulted in flail anterior and posterior valve leaflets. Valvuloplasty was carried out in 2 patients and valvular replacement in the remaining 7. All 9 patients improved significantly after operation attesting to the excellent results which can be expected in this condition if surgery is not unduely delayed.

INTRODUCTION

The mitral valve apparatus is a complex structure consisting of the valve leaflets, the annulus, the chordae tendinae and the papillary muscles. Competency of the mitral valve is critically dependent on all these components in addition to the posterior and lateral wall of the left atrium and the free wall of the left ventricle (1). It has been traditionally assumed in the past that mitral regurgitation (MR) is most frequently due to previous rheumatic fever resulting in damage to the valve leaflets. In this condition, circulatory adjustments, particularly enlargement of the left atrium, evolve over a long period of time. Consequently, the patient is usually asymptomatic even though the MR is severe until late in the course of his illness. In recent years, it has become apparent that MR is frequently due to non-rheumatic causes e.g. prolapsing mitral valve syndrome, papillary muscle dysfunction, rupture of the chordae tendinae, mitral annular calcification etc. Mitral regurgitation due to chordal or papillary muscle rupture, unlike rheumatic MR, presents dramatically, appearing with striking suddenness as a result of an abrupt disruption of the valve apparatus. The syndrome that is produced is classical and requires recognition because surgery is necessary

in most instances (2). We describe in this paper 9 cases of acute MR which we encountered in the past few years.

PATIENTS AND RESULTS

Table 1 classifies the patients, their clinical presentation and physical findings. These patients can be divided into 2 groups. In group I, there is no known cardiac disease in the past. All the patients in group II have had some form of previous cardiac disease or disability. For example, cases 6, 8 and 9 were known to have a cardiac murmur for many years. Case 7 had been treated for hypertension for 10 years and had suffered acute myocardial infarction recently.

The ages of the patients ranged from 13 to 59 years (mean 39). There were 8 male and 1 female and 8 Chinese and 1 Malay patients. None of them had a previous history of rheumatic fever.

Table 2 summarises the results of the laboratory investigations. Table 3 summarises the medical and surgical therapy and the long term follow up of the patients.

DISCUSSION

Clinical Presentation

Acute mitral regurgitation presents characteristically with a sudden onset of left heart failure or pulmonary oedema and is usually due to rupture of either the chordae tendinae or the papillary muscle of the mitral valve apparatus. It can occur in patients with no known cardiac disease or disability in the past and in this situation the syndrome of acute MR is seen in its most acute and purest expression. In this present series, 5 patients (Cases 1 to 5) presented in this manner and all were found subsequently to have ruptured chordae tendinae. In Cases 1 and 3, the rupture was most probably due to infective endocarditis, whereas in Cases 2, 4 and 5 no apparent cause could be found. All the group II cases (Cases 6 to 9) had a history of previous cardiac disease and rupture of the chordae tendinae or papillary muscle had occurred as a complicating event in the patients' illnesses. The presentation of these patients in terms of heart failure was sudden but somewhat less acute than the group I cases. In Case 6, infective endocarditis was documented, and Case 9 had chronic rheumatic mitral valve disease. Case 7 suffered acute myocardial infarction accompanied by rupture of his papillary muscles. Other causes of ruptured chordae tendinae which have been described but which were not seen in this series include mitral valve prolapse syndrome, and blunt chest trauma.

Physical Examination

The bedside clinical findings of this syndrome are rather distinctive and permit a diagnosis to be made in many instances particularly in patients with no previous cardiac disease. A loud apical pansystolic murmur is usually heard (Fig. 5) and this may radiate either medially (usually when the chordae tendinae to the posterior mitral leaflet are ruptured) or posteriorly (usually in anterior chordal rupture). In the former situation, the murmur frequently radiates to the base of the heart and up the neck and may spuriously suggest the co-existence of aortic stenosis. Because the left atrium is usually of normal size or only minimally dilated and is contracting well, a fourth heart

TABLE 1

Case No	Race	Age	Sex	Presentation	Physical Finding
Group 1					
1	Ch	20	M	Fever and dyspnoea x 2/12	Apical PSM 4/6 → left axilla. S4.
2	Ch	45	M	Sudden dyspnoea	Apical PSM 4/6 → medially. S4.
3	Ch	23	F	Fever, sudden dyspnoea	Apical PSM 4/6 → left axilla. S4.
4	Ch	44	M	Sudden dyspnoea	Apical PSM 4/6 → medially. S4.
5	Ch	42	M	Sudden dyspnoea	Apical PSM 5/6 → medially. S3.
Group 2					
6	Mal	13	M	Recurrent fever and CCF x 5 months	Apical PSM 5/6 → left axilla. S3.
7	Ch	59	M	AMI - 4/12 ago. Since then severe angina and dyspnoea	Apical PSM 5/6 → left axilla. S3.
8	Ch	50	M	Sudden dyspnoea	Apical PSM 4/6 → left axilla. S3.
9	Ch	55	M	Sudden dyspnoea	Apical PSM 4/6 → medially. S3.

Ch = Chinese. Mal = Malay. CCF = Congestive Cardiac Failure.
AMI = Acute Myocardial Infarction. PSM = Pansystolic Murmur.

sound is frequently heard. In the later stages of the disease or when the chordal rupture complicates previous cardiac disease, the clinical findings may be less distinctive. The left ventricle and the left atrium may be moderately dilated and a third instead of a fourth heart sound may be heard. In this series, all patients had loud apical pansystolic murmurs. In 4 patients (all of whom had posterior leaflet chordal rupture) the murmurs radiated medially. In the other 5 patients (4 with anterior chordal and 1 with both anterior as well as posterior chordal rupture) the murmurs radiated posteriorly. A fourth heart sound was heard in 4 and a third heart sound was heard in 5 patients (Table 1).

Laboratory Investigations

Laboratory investigations showed leucocytosis in Cases 1, 3 and 6. All these 3 patients presented with fever and were diagnosed clinically as having infective endocarditis. Blood cultures were negative for organisms in Cases 1 and 3 and grew alpha haemolytic streptococci in Case 6.

Of considerable importance was the finding that the heart size in the initial chest x'rays were either normal or only mildly enlarged in Cases 1 to 5 (all in group 1) (Fig. 1 & 2). Since Cases 4 and 5 underwent operation only 1 year later, an opportunity was present to examine their serial chest xrays which showed progressive cardiomegaly. However radiological signs of pulmonary oedema at some time or other were seen in every patient in this group. This contrasts with chronic rheumatic MR where considerable cardiomegaly is invariably present when the patient becomes symptomatic. All the 4 patients in group II had moderate to severe cardiomegaly (Fig. 3). These 4 patients also showed radiological evidence of left heart failure or pulmonary oedema.

All group I patients had normal ECG's and were in sinus rhythm (Fig. 4). Again this contrasts with chronic rheumatic MR where the ECG in the later stages usually shows atrial fibrillation with evidence of left ventricular hypertrophy. In the group II patients, evidence of anterior and inferior transmural myocardial infarction was seen in Case 7, atrial flutter in Case 8 and left ventricular hypertrophy in Case 9.

Echocardiographic Abnormalities

Five patients had both M mode as well as 2 dimen-

TABLE 2

Case No	TW	Blood Cultures	CXR	ECG	M Mode Echoe	2D Echoe	Haemodynamic Pressures in mmHg		LV Angio
							PCW	V Wave	
Group I									
1	30,600	Neg x 6	Heart size - N. Pulmonary oedema.	N. SR	MV veg.	Flail MV ant leaflet with veg.	NO	NO	Gd IV MR
2	N	ND	Heart size - N. Pulmonary oedema.	N. SR	ND	ND	22	30	Gd IV MR
3	12,000	Neg x 3	Heart size - N. CCF.	N. SR	Flail MV ant leaflet.	Flail MV ant leaflet with veg.	24	35	Gd IV MR
4	N	ND	Heart size - N. CCF.	N. SR	ND	ND	18	25	Gd IV MR CA - N.
5	N	ND	Heart size - N.	N. SR	Flail MV post leaflet.	Flail MV post leaflet.	NO	NO	Gd IV MR
Group II									
6	13,000	A. haem strept	Heart size CCF.	N. SR	Veg on MV leaflets.	Flail MV ant leaflet.	32	68	Gd IV MR
7	N	ND	Heart size CCF.	Ant and inf MI. SR.	ND	ND	NO	NO	Gd IV MR. Akinesia of antero-lat. and apical segments of LV. LMCA = 75%. LAD, Cx and mid RCA - total occlusion.
8	N	ND	Heart size CCF.	Atrial flutter.	Flail MV. LA + + LV + +	ND	NO	NO	Gd IV MR. Diag. A = 60%
9	N	ND	Heart size CCF.	LVH. SR.	Rheumatic MR.	Rheumatic MV.			Gd IV MR. CA - N

TW = Total White Count. Neg = Negative. A. Haem. Strept. = Alpha Haemolytic Streptococcus. ND = Not done. N = Normal. CXR = Chest Xray. CCF = Congestive Cardiac Failure. SR = Sinus Rhythm. Ant = Anterior. Post = Posterior. Inf = Inferior. MI = Myocardial Infarction. LVH = Left Ventricular Hypertrophy. MV = Mitral Valve. Veg = Vegetation. LA = Left Atrium. LV = Left Ventricle. PCW = Pulmonary Capillary Wedge. Angio = Angiography. MR = Mitral Regurgitation. LMCA = Left Main Coronary Artery. LAD = Left Anterior Descending Artery. Cx = Circumflex. RCA = Right Coronary Artery. Diag = Diagonal. CA = Coronary Arteries. NO = Not Obtained.

sional echocardiographic examinations, whilst 1 patient had only M mode echocardiographic studies. Abnormalities due to both infective endocarditis and chordal rupture seen in both M mode as well as 2 dimensional echocardiographic studies have been well described in the past (3, 4, 5). It is generally accepted that both these techniques are useful for the detection of vegetative masses, but 2 dimensional echocardiography is clearly superior for diagnosing chordal rupture (6). The M-mode echocardiographic abnormalities of chordal rupture are (a) chaotic diastolic fluttering of either the anterior or posterior mitral leaflets depending on whether posterior or anterior chordal rupture has occurred (b) delayed anterior movement of the posterior leaflet in diastole in posterior chordal rupture (c) systolic fluttering and prolapse of the valve leaflets and (d) the presence of echoes in the left atrial cavity in systole. However none of these M mode echocardiographic findings are specific for chordal rupture but their presence (particularly if more than one abnormality is present) in the correct clinical setting should suggest this possibility. The hallmark of 2 dimensional echocardiographic abnormality of chordal rupture is an overshoot of the leaflet tip into the left atrium during systole due to lack of leaflet support. This is a reasonably sensitive and specific finding in chordal rupture and is a very useful aid in the diagnosis of this condition (6). In 4 patients (Cases 1, 3, 5 and 6) 2 dimensional echocardiography clearly showed chordal rupture (Fig. 6 and 7) but M mode echocardiography suggested this condition in only 2 patients (Fig. 5). In 1 patient (Case 8) who had only M mode echocardiography done, findings suggesting posterior chordal rupture were present. In one patient with chronic rheumatic mitral valve disease (Case 9) both M mode and 2 dimensional

TABLE 3

Case No	Medical T/T	Surgical Findings	Operative Procedure	Long Term Follow Up
Group I				
1	Antibiotics x 6 weeks. CCF T/T.	Flail ant. MV leaflet due to RCT.	MVR with S/E valve.	II NYHA
2	CCF T/T.	Flail post. MV leaflet due to RCT.	MVR with S/E valve.	II NYHA
3	Antibiotics x 1/52. CCF T/T.	Flail ant. MV leaflet due to RCT with veg.	MVR with Hancock valve.	II NYHA
4	CCF T/T.	Flail post. MV leaflet due to RCT.	Valvuloplasty.	II NYHA. Residual loud apical PSM.
5	CCF T/T.	Flail post. MV leaflet due to RCT.	MVR with S/E valve.	II NYHA
Group II				
6	Repeated courses of antibiotics CCF T/T.	Flail ant. MV leaflet due to RCT.	MVR with S/E valve.	II NYHA
7	Nitrates. CCF T/T.	Both papillary muscles infarcted — flail ant. and post. MV leaflets.	MVR with Hancock valve. CABG to post desc. A, Diag. A, LAD and postero-lat. branch of Cx A.	III NYHA
8	D.C. shock. CCF T/T and Quinidine.	Flail ant. MV leaflet due to RCT.	Valvuloplasty. CABG to Diag. A.	II NYHA
9	CCF T/T.	Flail post. MV leaflet due to RCT.	MVR with S/E valve	II NYHA

CCF = Congestive Cardiac Failure. T/T = Treatment. Ant. = Anterior. Post. = Posterior. MV = Mitral Valve. RCT = Ruptured Chordae Tendinae. Veg. = Vegetation. MVR = Mitral Valve Replacement. S/E = Starr Edwards. CABG = Coronary Artery Bypass Grafting. Post. Desc. = Posterior Descending. Diag. = Diagonal. LAD = Left Anterior Descending. Post. Lat. = Postero-lateral. Cx = Circumflex. A = Artery. PSM = Pan Systolic Murmur. NYHA = New York Heart Association Functional Status.

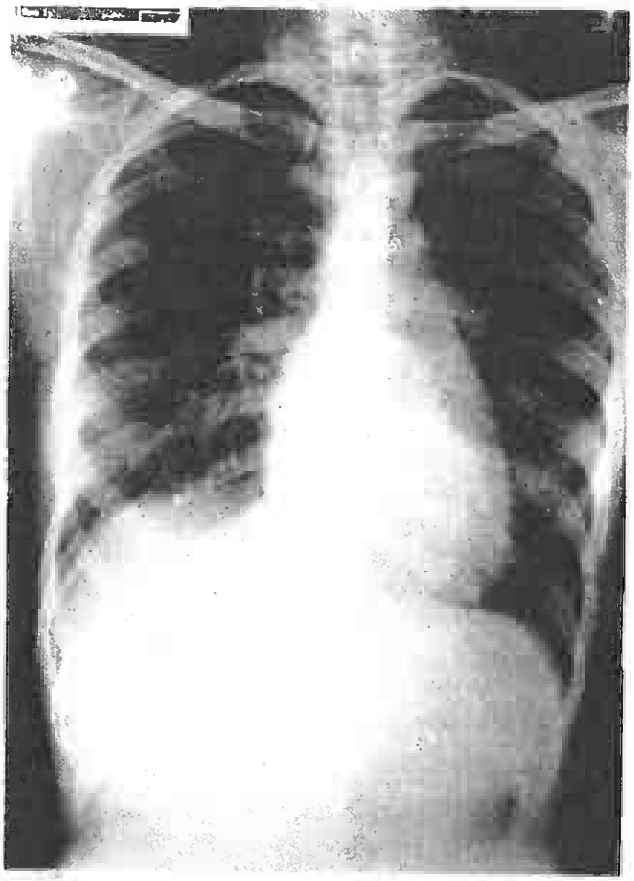

Fig. 1 Chest Xray of Case 3 taken on admission. The heart size is minimally enlarged and changes of left heart failure are present.

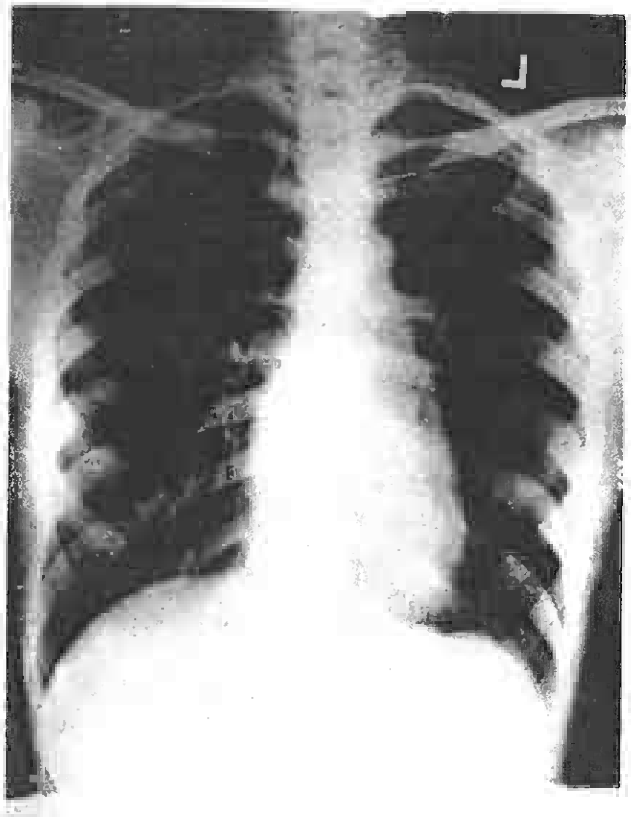

Fig. 2 Chest Xray of Case 3 taken 2 weeks after admission following vigorous treatment for heart failure. The heart size is now normal and the lung fields are clear.

echocardiographic findings showed rheumatic mitral valve involvement and chordal rupture was unsuspected pre-operatively.

Cardiac Catheterization and Angiography

Cardiac catheterization and left ventricular angiography were performed in all the 9 patients. The pulmonary artery pressure was elevated in every patient. However the most distinctive finding was an elevated wedge pressure with a very tall V wave and this was seen in 4 patients where these pressures were able to be obtained (Fig. 8). Left ventricular angiogram showed severe MR (grade IV/IV) in all the 9 patients with regurgitation of the dye into the pulmonary veins. All the group I patients had normal size left ventricles which contracted excessively and normal size left atria (Fig. 9 and 10). Cases 6, 7, 8 and 9 in group II had considerable left ventricular and left atrial enlargement. In addition, Case 7 showed akinesia of the anterolateral and apical segments of the left ventricle due to his previous myocardial infarction. Selective coronary angiography in this patient showed a 75% stenosis of the left main coronary artery. The left anterior descending artery, the circumflex artery and the middle portion of the right coronary artery were all totally occluded. Selective coronary angiography in Case 8 showed a 60% stenosis of the diagonal artery and normal coronary arteries in Cases 4 and 9.

All the 9 cases were given vigorous medical treatment consisting essentially of anti-heart failure therapy such as digoxin, frusemide, and hydrallazine for periods of time varying from a few weeks to several

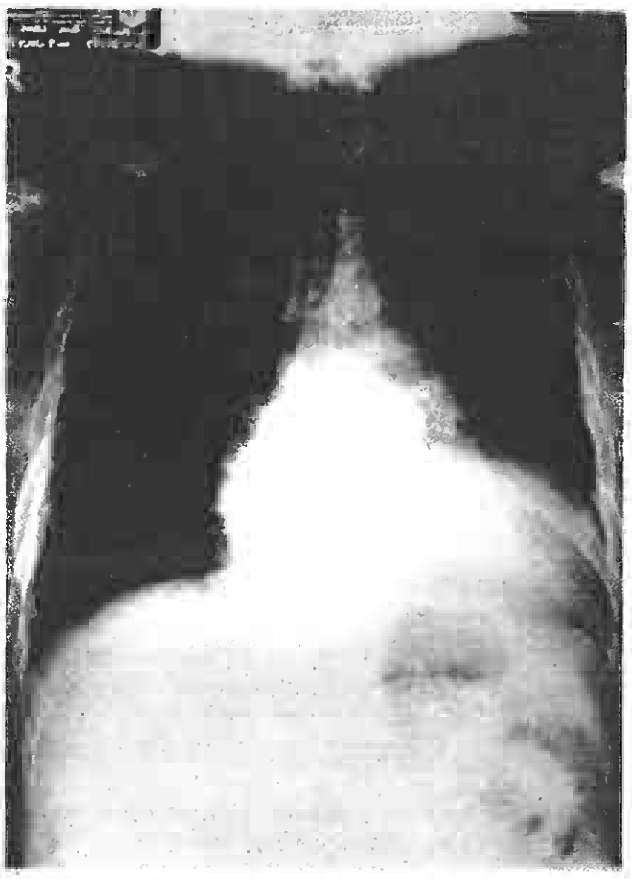

Fig. 3 Chest Xray of Case 9 showing considerable cardiomegaly.

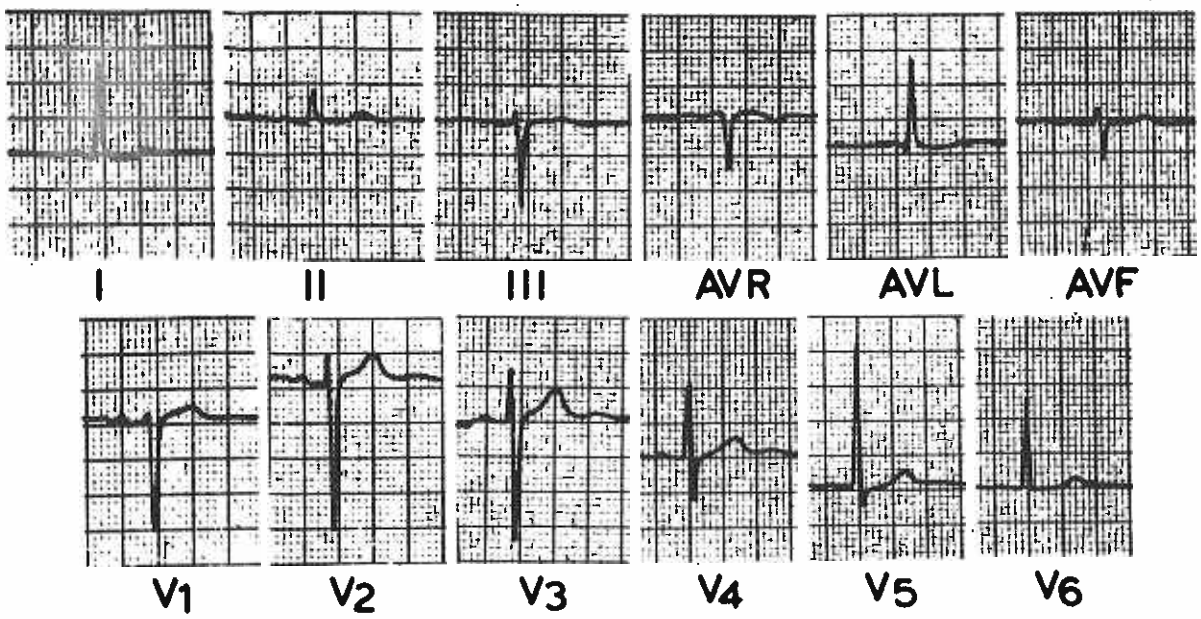

Fig. 4 12 lead ECG of Case 3 showing a normal pattern.

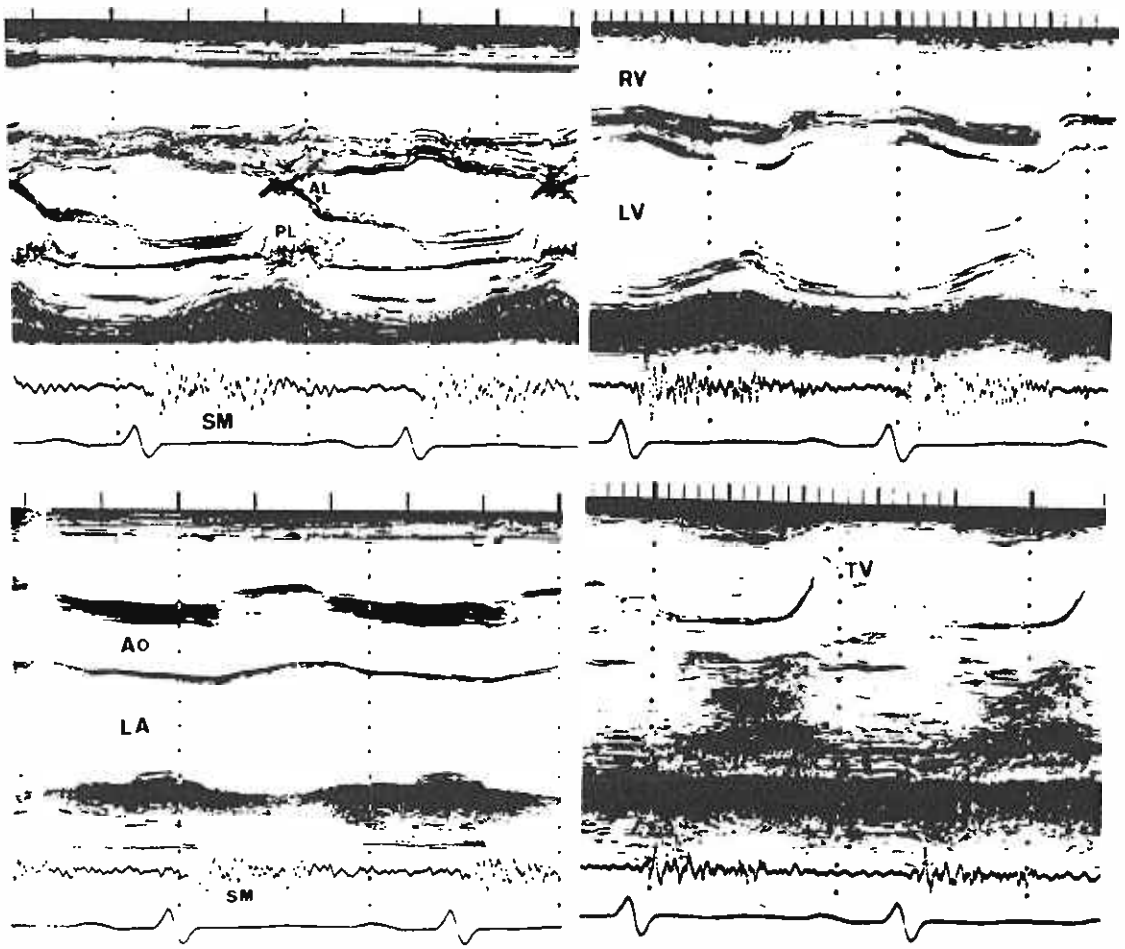

Fig. 5 Echophonocardiographic examination of Case 5. Top right hand panel shows M mode echocardiographic recording of mitral valve. The posterior valve leaflet movement is delayed and anterior. The phonocardiogram is recorded at the apex and shows a pansystolic murmur. Top left hand panel shows left ventricular study. The left ventricle is slightly dilated and contracts well. Bottom right hand panel shows normal sized aortic root and left atrium. The phonocardiogram is recorded at the fifth left intercostal space at the left sternal edge and shows a loud pansystolic murmur. The lower left hand panel shows incomplete recording of the tricuspid valve. AL = anterior leaflet of mitral valve. PL posterior leaflet of mitral valve. SM = Pansystolic murmur. Ao = Aorta. LA = left atrium. RV = right ventricle. LV = left ventricle. TV = tricuspid valve.

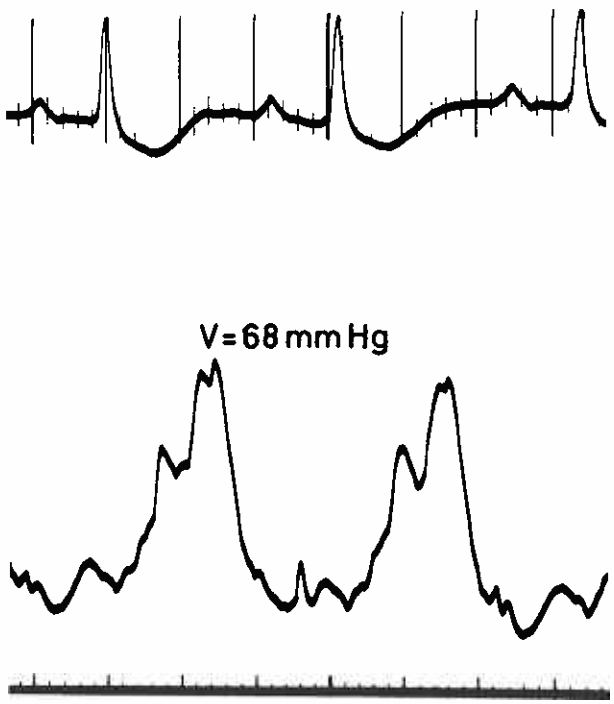

Fig. 8 Left atrial pressure tracing of Case 6 showing a grossly elevated V wave of 68 mmHg.

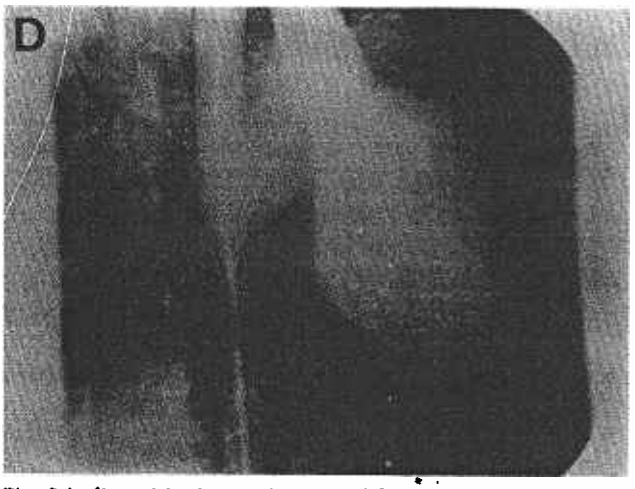

Fig. 9 Left ventricular angiogram of Case 3 in diastole showing normal sized left ventricular cavity.

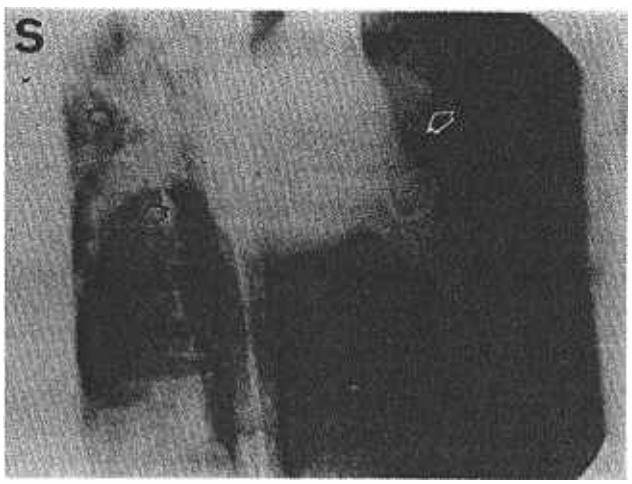

Fig. 10 Left ventricular angiogram of Case 3 in systole showing severe mitral regurgitation with reflux of dye into the pulmonary veins (arrow). The left atrial size is normal.

anterior and posterior mitral valve leaflets. Case 2 and 8 underwent valvuloplasty. All the other cases had mitral valve replacement - 5 with a Starr-Edwards prosthetic valve and 2 with a Hancock bioprosthesis. In addition, Case 7 had saphenous vein bypass grafts to his posterior descending artery, diagonal artery, left anterior descending artery and postero-lateral branch of his circumflex artery. Case 7 had a similar graft to his diagonal artery.

All the 9 patients survived their operation. The follow up period of these patients ranged from 1 to 36 months (mean 11 months). All the patients improved significantly following surgery and are in Class II (NYHA) functional status except for Case 7. This patient although angina free still has some degree of effort intolerance (Class III NYHA) although he too has improved markedly. The heart size of Cases 1, 2 and 3, as assessed radiologically, is at present normal. Cases 4 to 9 all have residual cardiomegaly. The pan-systolic murmur present preoperatively in Case 4 who had valvuloplasty has persisted after the operation. These results confirm previous reports and attest to be the excellent results which can be obtained in patients with chordal rupture if surgery is not too unduly prolonged and emphasise once again the importance of recognition and diagnosing of this syndrome.

ACKNOWLEDGEMENTS

Case 8 has been included in this series by courtesy of Dr Charles Toh. Case 6 was referred for investigation and treatment by Dr Khoo Kah Lin from Kuala Lumpur and Case 9 by Dr Chee Kim Hoe from Singapore.

Cases 2 and 7 were operated by Dr Victor Chang from St Vincent's Hospital, Sydney; Cases 3 and 4 by Sir Brian Barratt Boyes from Green Lane Hospital, Auckland and Case 8 by Dr J. Kay from St Vincent's Hospital, Los Angeles.

REFERENCES

1. Roberts WC, Perloff JK: Mitral valvular disease. Ann Intern Med 1972; 77: 939-75.
2. Selzer A, Kelly J, Vannitamby M et al. The syndrome of mitral insufficiency due to isolated rupture of the chordae tendinae. Amer J Med 1967; 43: 822-35.
3. Mintz G, Kotler M, Segal B, Parry W: Two dimensional echocardiographic recognition of ruptured chordae tendinae. Circulation 1978; 57: 244-50.
4. Chia BL, Ee B: Echocardiographic abnormalities in acute rupture of the aortic valve due to infective endocarditis. Angiology 1981; 50: 846-51.
5. Martin R, Meltzer R, Chia BL, Stinson E, Rakowski and Popp R: Clinical utility of 2 dimensional echocardiography in infective endocarditis. Amer J Cardiol 1980; 46: 379-85.
6. Mintz G, Kotler M, Wayne P, Segal B: Statistical comparison of M mode and two dimensional echocardiographic diagnosis of flail mitral leaflets. Amer J Cardiol 1980; 45: 253-9.
7. Simpson P, Bristow JD: Recognition and management of emergencies in valvular heart disease. Clin N Amer 1979; 63: 155-72.