CASE STUDY

Restrictive pericarditis

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Abstract

***Background*—Pericardial thickening is an uncommon complication of cardiac surgery.**

***Objectives*—To study pericardial thickening as the cause of severe postoperative venous congestion.**

***Subjects*—Two men, one with severe aortic stenosis and single coronary artery disease, and one with coronary artery disease after an old inferior infarction. Both had coronary artery bypass grafting surgery.**

***Methods*—Magnetic resonance imaging (MRI), Doppler echocardiography, and cardiac catheterisation.**

***Results*—Venous pressure was raised in both patients. MRI showed mildly thickened pericardium, and cardiac catheterisation indicated diastolic equalisation of pressures in the four chambers. Jugular venous pulse showed a dominant "Y" descent coinciding with early diastolic flow in the superior vena cava, and mitral and tricuspid Doppler forward flow proved restrictive physiology. The clinical background suggested pericardial disease so both patients had pericardiectomy. This proved the pericardium to be thickened; the extent of fibrosis also involved the epicardium.**

***Conclusions*—Although rare, restrictive pericarditis (restrictive ventricular physiology resulting from pericardial disease) should be considered to be a separate diagnostic entity because its pathological basis and treatment are different from intrinsic myocardial disease. This diagnosis may be confirmed by standard investigational techniques or may require diagnostic thoracotomy.**

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Keywords: restrictive pericarditis; pericardial disease

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In 1935, White defined constrictive pericarditis as a chronic fibrous thickening of the wall of the pericardial sac that is so contracted that normal diastolic filling of the heart is prevented.[1] This definition supports conventional understanding of the pathophysiological consequence of chronic inflammatory and fibrotic pericardial disease.[2 3] Traditionally, the clinical challenge has been to distinguish constrictive pericarditis from its most important differential diagnosis, restrictive cardiomyopathy, as both conditions may display similar physical signs and haemodynamics.[4] The pericardium has a fibrous outer layer and an inner serous membrane composed of a single layer of mesothelial cells. The inner serous layer consists of parietal and visceral layers; the visceral pericardium is closely attached to the surface of the heart (the epicardium) and epicardial fat; the parietal layer is near the fibrous pericardium, and the pericardial cavity lies between the two. Pericardial disease, which affects the fibrous pericardium, may cause constriction by limiting expansion of the minor axes of both ventricles, and may be either localised or symmetrical in distribution. However, it does not interfere with long axis expansion which is controlled by the motion of the two atrioventricular rings. In contrast, if this process significantly involves the visceral pericardium (epicardium), both major and minor axes are likely to be affected. Thus, the physiological signs may mimic restrictive cardiomyopathy by causing a restrictive picture rather than classical constrictive pericarditis. Here, we report two patients with pericardial disease following cardiac surgery who displayed striking evidence of restrictive physiology on echocardiography, but in whom the underlying condition was pericardial disease. The presence of significant visceral pericardial (epicardial) involvement may explain the patients' poor response to conventional pericardectomy and worse overall outcome.

Case 1

A 66 year old man with severe calcific aortic stenosis and single vessel coronary artery disease was referred for elective surgery. He had aortic valve replacement with a 23 mm Medtronic Hall tilting disc valve and a left internal mammary graft to the left anterior descending artery. His pericardium was not closed and his postoperative course was unremarkable except for a sternal wound infection. Two months later he developed a recurrent left pleural effusion, which was treated by multiple aspirations and tetracycline pleurodesis. However, the pleural effusion persisted and six months after the operation he began to complain of shortness of breath on exertion, and facial congestion. Clinical examination revealed resting tachycardia and very high jugular venous pressure (+15 cm) with

Table 1 M mode echocardiographic values

	Case 1	Case 2
LV end diastolic dimension (cm)	4.4	4.4
LV end systolic dimension (cm)	3	3.6
Estimated ejection fraction (%)	49	45
LV long axis excursion (cm)		
Left	1	1
Septal	1.2	1
RV long axis excursion (cm)	1.4	1.6
Left atrial diameter (cm)	4.2	5.0

LV, left ventricle; RV, right ventricle.

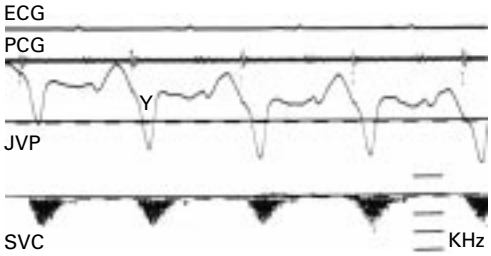

Figure 1 Jugular venous pulse and superior vena caval flow recordings from patient 1 showing an isolated deep "Y" descent and early diastolic flow.

prominent "Y" descent. Blood pressure was 140/80 mm Hg and normal prosthetic aortic sounds were present. There was dullness at the left base, abdominal distension, and ascites.

ECG showed sinus rhythm of 80 beats/min, normal QRS axis, and inferolateral T wave inversion. Chest radiography showed a normal cardiothoracic ratio and a left pleural effusion. The jugular venous pressure was very high. Pulse recording showed a dominant "Y" descent and no perceptible "X" descent (fig 1). Forward superior vena caval flow, measured by pulsed wave Doppler, was confined to early diastole; an echocardiogram showed a normal functioning aortic valve prothesis with no evidence of aortic regurgitation; and left and right ventricular cavity sizes were normal (table 1). No pericardial fluid collection was seen. Left ventricular isovolumic relaxation time was 40 ms and peak E wave velocity was 1.3 m/s with a deceleration time of 100 ms. Peak A wave velocity was 0.7 m/s giving an E/A ratio of 1.7. On the right side of the heart peak early diastolic tricuspid velocity was 0.7 m/s with a deceleration time of 100 ms. No flow was detected across the tricuspid valve with right atrial systole and none of the values on the left or right side of the heart showed perceptible variation with respiration (fig 2). Cardiac catheterisation showed diastolic equalisation of pressures in all four chambers but with evidence of significantly high pulmonary artery pressure (fig 3). Magnetic resonance imaging showed a mildly thickened pericardium (< 5 mm) and only mild impairment of filling at end diastole.

The patient continued to deteriorate with worsening venous congestion, peripheral oedema, ascites, and pleural effusion, despite high doses of intravenous diuretics. A clinical diagnosis of significant restrictive pericardial disease was made although the investigative findings were inconsistent with this diagnosis; the patient thus had surgical exploration with anterior pericardectomy. The pericardium was found to be diseased and thickened to at least 1 cm in both diaphragmatic and inferior areas, with constriction of the inferior and superior venae cavae. It was not possible to define any plane of cleavage between epicardium and myocardium. The postoperative course was complicated by hypotension, sepsis, renal, and hepatic failure with evidence of persistently raised right sided pressures. The patient had an asystolic arrest on the 13th day after operation. Postmortem pathology revealed left ventricular hypertrophy with a normal prosthetic aortic valve. The pericardium surrounding the right and left ventricles was thickened and stuck to the diaphragmatic surface leading to fusion of the fibrous pericardium with the diaphragm. Histology revealed fibrotic pericardium adherent to the pleura and diaphragm, with predominant epicardial involvement (fig 4). There was no evidence of myocardial disease.

Case 2

A 53 year old man had coronary artery bypass grafting after an inferior myocardial infarction with an uneventful postoperative recovery. Three years later he developed intermittent unexplained episodes of weight gain, breathlessness on exertion, abdominal distension, and peripheral oedema. Clinical examination was

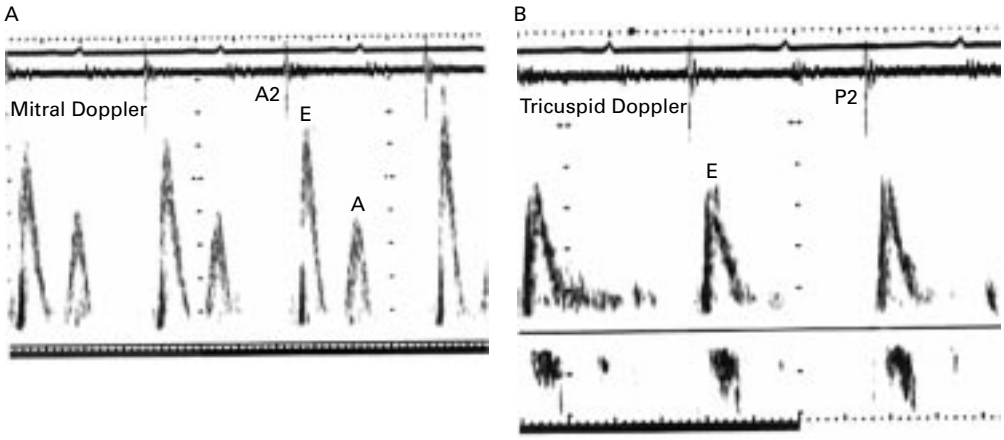

Figure 2 (Left) Transmitral Doppler flow velocities showing typical restrictive filling pattern. (Right) Transtricuspid Doppler flow velocities showing restrictive filling pattern. This did not alter significantly with respiration.

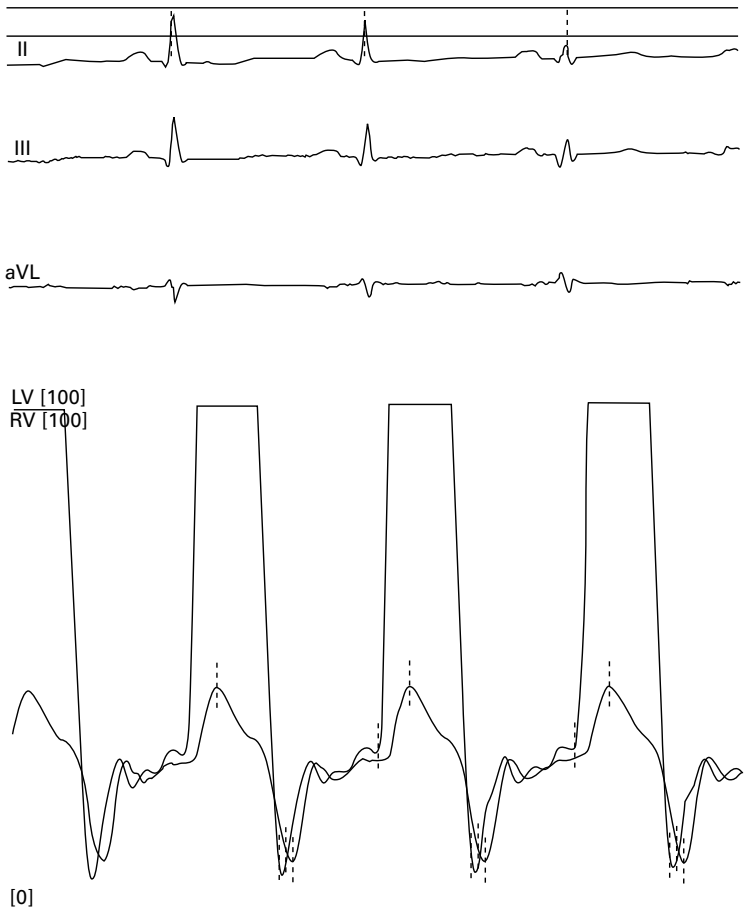

Figure 3 Left and right ventricular pressure recordings showing equalisation of diastolic pressures in the two chambers.

unremarkable although venous pressure was consistently high despite treatment with diuretics.

ECG showed sinus rhythm, normal QRS axis, and inferior T wave inversion. Chest radiography showed a normal cardiothoracic ratio with bilateral small pleural effusions. Echocardiography showed normal left ventricular cavity dimensions and no regional wall motion abnormality (table 1); no pericardial collection was seen. Left ventricular isovolumic relaxation time was 40 ms; peak transmitral E wave velocity was 0.9 m/s, and deceleration time was 80 ms. E/A ratio was 2.7. On the right side of the heart, right ventricular isovolumic relaxation time was 30 ms, peak transtricuspid E wave velocity 0.8 m/s, and deceleration time 90 ms. E/A ratio was again 2.7. Cardiac catheterisation showed evidence of diastolic equalisation of pressures in all four chambers with mildly raised pulmonary artery pressure (table 2). MRI showed a significantly thickened pericardium (6–8 mm), and there was abnormal diastolic filling with an abrupt stop which was suggestive of constrictive pericardial disease.

Table 2 Haemodynamic data (mm Hg)

	Case 1	*Case 2*
Right atrium (mean)	30/40 (28)	26/27 (22)
Right ventricle	52/5	43/5
Right ventricular EDP	24	27
Pulmonary artery (mean)	52/22 (33)	39/24 (28)
Pulmonary capillary wedge (mean)	25/37 (26)	34/28 (30)
Left ventricle	131/8	130/4
Left ventricular EDP	26	29
(LV−RV) EDP	+2	+2
RVEDP/RV systolic pressure	0.46	0.63

EDP, end diastolic pressure; LV, left ventricle; RV, right ventricle.

The patient had pericardectomy and, at operation, an abnormally thickened parietal and visceral pericardium was found which completely encased the heart; this was successfully removed. The patient had an uneventful postoperative course and currently remains clinically well.

Discussion

The differential diagnosis of pericardial constriction from restrictive myocardial disease is an important one (table 3). Usually, the diagnosis is made on the basis of cardiac catheterisation, as, in pericardial constriction, this will show equal or near equal diastolic pressures in both ventricles, without significant pulmonary hypertension.[5] A recent analysis has shown that unless three criteria are all satisfied—that is, a difference of <5 mm Hg between directly measured end diastolic pressures in the two ventricles, a peak right ventricular pressure of less than 50 mm Hg, and a ratio of right ventricular end diastolic to peak pressure of more than 0.33, haemodynamic assessment is not discriminating (table 3).[6] Computed tomography (CT) or magnetic resonance imaging (MRI) can show a thick

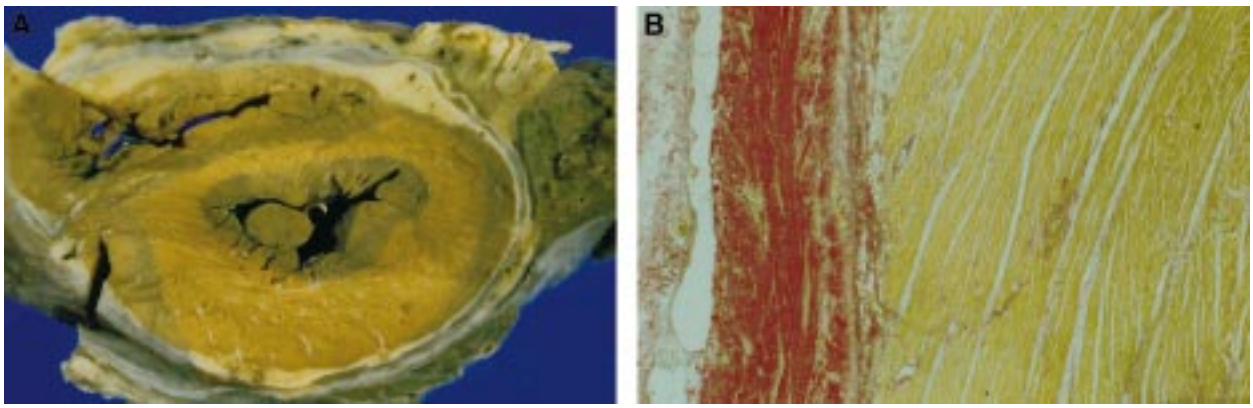

Figure 4 (A) Transverse section of the right and left ventricle showing the thickened pericardium adherent to the epicardium (arrow) and to the adjacent pleura. (B) Histology showing dense fibrosis (red) extending to the epicardium (yellow) elastic on Geison stain.

Table 3 Summary of diagnostic criteria

	Restrictive cardiomyopathy	*Constrictive pericarditis*	*Restrictive pericarditis*
Non-invasive			
Short IVRT	+	–	+
Sensitivity to respiration	–	+	–
Short E wave deceleration time	+	–	+
E/A ratio > 1.0	+	±	+
Jugular venous pulse	Y descent	X descent	Y descent
Invasive			
Ventricular ED pressures within 6 mmHg	–	+	+
Peak pressure < 50 mmHg	–	+	–
RV ED/peak pressure > 0.33	+	–	+

IVRT, isovolumic relaxation time; ED, end diastolic; RV, right ventricle.

pericardium,[7] but cannot predict whether any physiological disturbance is due to the pericardium. Echocardiographic criteria for constrictive pericarditis have been described. Early M mode echocardiographic evidence of constriction included an early diastolic posterior dip in the septum in constriction,[8] and a reduction in posterior wall thinning rate (table 1).[9] Unfortunately, neither of these has proved very reliable, and more recently, there has been increasing interest in the use of Doppler criteria based on respiratory variation in transmitral flow velocities.[10 11] In constriction, characteristically, there is more than 10% variation in peak mitral E wave velocity, with an increase during expiration along with a corresponding fall in the A wave. Analogous changes also occur on the right side of the heart, though tricuspid E wave velocity falls rather than increases with expiration.

Our results showed that left ventricular cavity size was within normal limits in the two patients, though ejection fraction was slightly depressed. Transmitral and transtricuspid Doppler estimates of flow velocities and their respective deceleration times indicated severe restriction.[12] No significant respiratory variation could be consistently detected for any Doppler variable. Long axis excursion was reduced though minor axis dimensions were maintained. Finally, the jugular venous pressure was greatly raised with a marked "Y" descent, and the early diastolic component of the vena caval flow was preponderant. Thus, all the Doppler criteria for restrictive myocardial disease were satisfied. CT and MRI were unhelpful in both cases as they showed only borderline values for the extent of pericardial thickening. The results of cardiac catheterisation were also equivocal, showing very high pulmonary artery pressures in case 1, and a wide difference between pulmonary artery diastolic and left ventricular end diastolic pressure in case 2.

In both cases, the diagnosis of pericardial disease was confirmed at surgery. The clinical condition was cured by pericardectomy in one patient and in the other patient histology showed no evidence of myocardial disease. We believe that the reason for these unexpected results was the significant involvement of the epicardium as well as the fibrous pericardium in the disease process. Subendocardial thickening, as in eosinophilic heart disease, is often taken as a paradigm of restriction,[13] and we suggest that the two cases studied here should be considered in the same light. In both cases, the diagnosis was clinically apparent from their postoperative state and both had additional clinical features suggestive of postcardiotomy syndrome, which may have been the cause of the pericarditis. We do not believe that surgery is the only cause, as it is possible that patients in whom additional diagnostic clues are absent might be missed and, thus, considered to have restrictive myocardial disease, particularly when cardiac catheterisation findings are inconclusive. We believe that there is a case for considering restrictive pericarditis as a separate diagnostic entity, since its pathological basis and treatment are different from intrinsic myocardial disease, even though the pathophysiology is similar. Therefore, a diagnostic thoracotomy in such cases cannot be regarded as an obsolete procedure.

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1 Killian DM, Furiasse JG, Scanlon PF, *et al*. Constrictive pericarditis after cardiac surgery. *Am Heart J* 1989;**118**:563–8.
2 Cimino JJ, Kogan AD. Constrictive pericarditis after cardiac surgery: report of 3 cases and review of the literature. *Am Heart J* 1989;**118**:1292–301.
3 Wood P. Chronic constrictive pericarditis. *Am J Cardiol* 1961;7:48–61.
4 Goodwin JF. The frontiers of cardiomyopathy: the clinical syndrome of restrictive cardiomyopathy mimics constrictive pericarditis. *Br Heart J* 1982;**48**:1–18.
5 Yu PN, Lovejoy FW, Joos HA, *et al*. Right auricular and ventricular pressure patterns in constrictive pericarditis. *Circulation* 1953;**7**:102–7.
6 Vaitkus PT, Kussmaul WG. Constrictive pericarditis versus restrictive cardiomyopathy: a reappraisal and update of diagnostic criteria. *Am Heart J* 1991;**122**:1431–41.
7 Hoit BD. Imaging the pericardium. In: Shabetai R, ed. *Diseases of the pericardium. Cardiology Clinics 8 (4)*. Philadelphia: WB Saunders, 1990:587–600.
8 Tei C, Child JS, Tanaka H, *et al*. Atrial systolic notch on the interventricular septal echogram: an echocardiographic sign of constrictive pericarditis. *J Am Coll Cardiol* 1983;**1**:907–12.
9 Morgan JM, Raposo L, Clague JC, *et al*. Restrictive cardiomyopathy and constrictive pericarditis: non-invasive distinction by digitised M mode echocardiography. *Br Heart J* 1989;**61**:29–37.
10 Hatle LK, Appleton CP, Popp RL. Differentiation of constrictive pericarditis and restrictive cardiomyopathy by Doppler echocardiography. *Circulation* 1989;**79**:357–70.
11 Garcia MJ, Rodriguez L, Ares M, *et al*. Differentiation of constrictive pericarditis from restrictive cardiomyopathy: assessment of left ventricular diastolic velocities in longitudinal axis by Doppler tissue imaging. *J Am Coll Cardiol* 1996;**27**:108–14.
12 Appleton CP, Hatle LK, Popp RL. Demonstration of restrictive physiology by Doppler echocardiography. *J Am Coll Cardiol* 1988;**11**:757–68.
13 Poole-Wilson PA. Dilated and restrictive cardiomyopathy. In: Julian DG, Camm AJ, Fox KM, *et al*, eds. *Diseases of the heart*; 2nd ed. London: WB Saunders, 1996:482–8.