Pericardial constriction: diagnosis and management

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The diagnosis of pericardial constriction is challenging and elusive. It is a post-inflammatory condition that occurs when a thickened, fibrotic, scarred and sometimes calcified pericardium firmly encases the cardiac chambers and restricts filling of the heart, causing venous overload and diminished cardiac output. This review includes the diagnosis and management of this condition.

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The first observation of the pathology of pericardial constriction was made in the second century AD, when Galen noted a thickening of the pericardium occurring in animals and hypothesized that the same process could affect humans. In 1669, Richard Lower described features of pericardial constriction (concretio cordis). Two scientists, Corrigan (the pericardial knock or *bruit de frappement*) and Kussmaul (pulsus paradoxus), gathered the characteristic features of pericardial constriction during the 1800s. Lancisi first described the classical features of the syndrome in 1842. However, pericardial constriction syndrome was credited to Pick (1896), who published data on a series of patients with ascites without peripheral oedema – Pick's syndrome.

STRUCTURE AND FUNCTION OF THE PERICARDIUM

The pericardium consists of two layers – the visceral (fibrous) and parietal (serous) pericardium. Up to 50 ml of fluid lie between the two layers for lubrication of the pericardial space. The pericardium serves to fix the heart in its correct anatomical location and to prevent excessive movement and dilatation, and friction between the heart and its neighbouring organs. It also provides a physical barrier to the extension of infection and the spread of malignant disease. By providing a relatively rigid casing to the heart, the pericardium assists in the distribution of hydrostatic forces during the cardiac cycle.

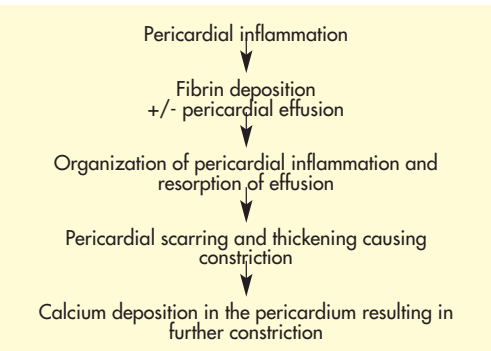

Figure 1. Development of pericardial constriction

PATHOLOGY

The initial pericardial insult may be subclinical and triggers the deposition of fibrin in both layers of the pericardium – visceral and parietal. This is often accompanied by an effusion. The next stage is organization and resorption of the pericardial effusion. Scarring and thickening of the pericardium, followed by obliteration of the pericardial space, eventually limit cardiac chamber filling (*Figure 1*). The process may extend into the myocardium and result in myocyte atrophy and fibrosis, and it is important to recognize that the process may involve the cardiac muscle as well as the pericardium.

Ultimately, calcification of the pericardium encases the heart in a thickened, non-pliable casing. Calcification of the pericardium is most commonly found over the right ventricle (Masui et al, 1992). The left atrium is unlikely to be affected in pericardial constriction, as it is typically an extrapericardial structure. Constriction involving the atrioventricular groove results in left atrial outflow obstruction. Inadequate release of this constriction at the time of operation may fail to provide symptomatic relief (Somerville, 1968).

Constriction usually occurs in a symmetrical distribution, resulting in a uniform restriction of every cardiac chamber. However, localized constriction caused by bands are sometimes found in the atrioventricular groove, aortic groove, right ventricular outflow tract and the vena cavae.

PHYSIOLOGICAL EFFECTS OF PERICARDIAL CONSTRICTION

As the pericardium becomes stiff and non-compliant, it acts as a box compressing the cardiac chambers and distorting normal cardiac function with three major physiological effects, described by Shabetai et al (1970) and Santamore et al (1986).

Impaired left ventricular filling

During inspiration in normal subjects, there is a decrease in intrathoracic pressure, which is transmitted to all cardiac chambers and the pulmonary veins, allowing blood to drain into the left ventricle from the pulmonary veins. The process of pericardial constriction isolates the cardiac chambers from intrathoracic pressure changes, eventually resulting in a reduction of the gradient between the pulmonary veins and the left ventricle. This causes a reduction in flow velocity in the pulmonary veins and subsequently impairs filling of the left ventricle. The impaired filling of the left side of the heart transmits pressure and volume overload to the right side of the heart.

Elevated end-diastolic pressures in both right and left ventricles

As the constricting pericardium limits distension of both ventricles, diastole (the relaxation and filling phase) is impaired, resulting in elevated end-diastolic pressures in right and left ventricles.

Impaired diastolic filling and increased heart rate

An estimated 75% of ventricular filling occurs during the second phase of diastole. In pericardial constriction, early diastolic filling is relatively unimpaired, and conversely as atrial pressures are elevated, blood is forced to enter the ventricles at a marginally increased velocity. However, this rapid filling comes to an abrupt halt in mid diastole, at the point where the non-compliant pericardium limits further ventricular distension. The reduction in stroke volume triggers an increase in heart rate to maintain cardiac output.

AETIOLOGY

Constrictive pericarditis is now an uncommon disorder, most often idiopathic in origin. Tuberculosis (TB) infection remains the leading cause of pericardial constriction worldwide. With the control of TB and the availability of effective antituberculous therapy in the West, this is currently an uncommon aetiology. However, the resurgence of human immunodeficiency virus-related TB infection and multi-drug resistant strains may result in an increased incidence of tuberculous pericardial constriction in the future.

Pericardial constriction may also occur after cardiothoracic surgery, implantation of devices such as pacemakers and implantable cardioverter defibrillators, radiation treatment for malignancy and a variety of infections. Case reports of miscellaneous causes include constriction following self-mutilation by insertion of sewing needles

TABLE 1.
Aetiologies of pericardial constriction

Idiopathic		
Infective	Mycobacterial	Tuberculosis
	Viral	Coxsackie
	Fungal	Histoplasmosis
		Coccidiomycosis
	Parasitic	Amoebiasis
		Echinococcus
Malignant disease	Lymphoma	
	Lung carcinoma	
	Breast carcinoma	
	Hodgkin's disease	
	Melanoma	
	Primary mesothelioma	
Post-thoracotomy	Coronary artery bypass surgery	
	Valve replacement/repair	
	Cardiac transplantation	
	Trauma	
Drug-related	Hydralazine	
	Procainamide	
	Penicillins	
	Isoniazid	
	Minoxidil	
	Phenylbutazone	
Post-radiation therapy		
Connective-tissue disease	Rheumatoid arthritis	
	Systemic lupus erythematosus	
	Scleroderma	
	Dermatomyositis	
Renal failure		
Post-myocardial infarction		
Post-pacemaker/implantable cardioverter defibrillator insertion		
Sarcoidosis		
Amyloidosis		
Carcinoid syndrome		
Whipple's disease		
Asbestosis		
Porphyria cutanea tarda		
Miscellanea	Sewing needles	
	Toothpicks	

into the chest cavity (Keogh et al, 1981). Aetiologies are shown in *Table 1*.

CLINICAL PRESENTATION

The diagnosis of constrictive pericarditis is often reached after a prolonged course of investigation (*Table 2*). The presentation is insidious, non-specific and chronic, and may occur weeks or years after the initial pericardial insult, which may have been asymptomatic. One of the principal reasons for misdiagnosis is failure to detect elevation of the jugular venous pressure (JVP), particularly when elevated above the angle of the jaw. This often leads the clinician to diagnose liver disease and to request inappropriate invasive investigations, such as liver biopsy.

TABLE 2.
Differential diagnosis

Primary restrictive cardiomyopathy	
Restrictive cardiomyopathy secondary to	Amyloidosis
	Sarcoidosis
	Radiation injury
	Haemochromatosis
	Hypereosinophilic syndrome
Tricuspid stenosis	
Tricuspid regurgitation	
Hypertrophic cardiomyopathy	
Right atrial myxoma	
Superior vena cava obstruction	
Nephrotic syndrome	
Liver disease	
Intra-abdominal malignancy	
Pregnancy	

Previous research (Cordell and Beason, 1968; Wychulis et al, 1971; Robertson and Mulder, 1984; De Valeria et al, 1991; Arsan et al, 1994; Tirilomis et al, 1994; Trotter et al, 1996) has shown a male:female preponderance of the disease varying between 2:1 and 4:1, although Wood (1961) found a male:female preponderance of 2:7 (*Table 3*). Constriction has been reported in ages ranging between 7–75 years.

Symptoms

Early complaints of anorexia and abdominal distension are usually a result of hepatic congestion. Peripheral oedema, attributable to elevated right heart pressures, may also be present. Subsequently, patients may complain of exertional dyspnoea, cough, orthopnea and, ultimately, platypnoea (dyspnoea while in the upright position) as a result of pulmonary venous congestion and pleural effusions.

In a small percentage of patients, compression of the coronary arteries by the pericardium or underperfusion of the myocardium may precipitate ischaemic chest pains.

End-stage disease presents with fatigue, weight loss and cachexia as a reflection of impaired cardiac output.

Signs

History and physical examination frequently suggest congestive cardiac failure, in which features of right-sided failure predominate. However, preliminary investigations may fail to demonstrate significant impairment of ventricular function.

The elevated JVP is often undetected during normal examination, and the patient may need to be examined sitting upright at 90°. The normal x descent indicative of atrial relaxation is preserved, but the y descent is sharp, reflecting the

TABLE 3.
Clinical features of pericardial constriction

Reference (no of patients)	M:F	Raised JVP	Peripheral oedema	Hepatomegaly	Ascites	S3	Pulsus paradoxus	Dyspnoea
Wood (1961) (*n*=40)	2:7	100	68		55		24	
Cordell and Beason (1968) (*n*=14)	2.5:1	100	57	71	64			100
Wychulis et al (1971) (*n*=137)	2.7:1	100	61	98	77	15	29	90
Das et al (1973) (*n*=77)	1.7:1	96	68	92	71		68	
Robertson and Mulder (1984) (*n*=31)		93	61	61		67	48	90
Bashi et al (1988) (*n*=118)	2.7:1	100	84	100	90		84	
De Valeria et al (1991) (*n*=36)	1.7:1		81		22			100
Arsan et al (1994) (*n*=82)	2:1	55	55	55	55	70		91
Tirilomis et al (1994) (*n*=71)	2.5:1	51	59	68	37			97
Trotter et al (1996) (*n*=21)	4:1		62					48

JVP = jugular venous pressure; S3 = third heart sound

rapid ventricular filling abruptly halted by the stiff pericardial casing (Friedrich's sign), characteristic of pericardial constriction. The elevated right atrial pressure impedes right atrial filling, leading to an elevation of JVP with inspiration (Kussmaul's sign) (*Figures 2* and *3*).

Venous engorgement as a result of elevated right heart pressures leads to hepatomegaly and peripheral oedema. Other features of hepatic dysfunction, such as spider naevi, palmar erythema and jaundice, are sometimes evident. Cases of protein-losing enteropathy in association with constriction have been previously reported (Wilkinson et al, 1965).

TABLE 4.
Signs and symptoms of pericardial constriction

Elevated jugular venous pressure with prominent x and y descent
Peripheral oedema
Pulsatile hepatomegaly
Anorexia
Ascites
Absent apex pulse
Tachycardia
Pericardial knock
Pulsus paradoxus
Angina
Cachexia

TABLE 5.
Investigation results

Electrocardiography	Tachycardia
	Atrial fibrillation in less than 50%
	Low-voltage QRS complexes
	P mitrale
Chest X-ray	Lateral chest X-ray
	Calcified pericardium
	Heart size may be reduced/normal/enlarged
Echocardiography	Thickened pericardium
	Normal left ventricular function
	Enlarged left atrium
	Dilated hepatic veins and inferior vena cava
	Increased E:A ratio
	Decreased inspiratory flow reduction in hepatic veins
CT/MRI	Thickened pericardium
	Dilatation of inferior vena cava/hepatic veins
Right and left heart catheterization	Pressure tracings

CT = computed tomography; E:A ratio = velocity of mitral inflow, specifically early rapid ventricular filling (E) and late filling with atrial contraction (A); MRI = magnetic resonance imaging

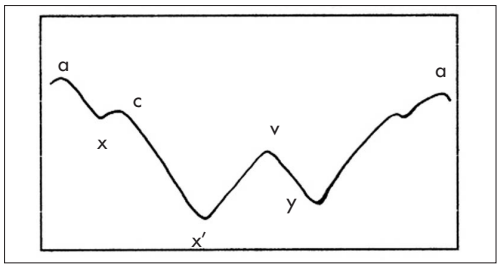

Figure 2. Normal jugular venous pressure waveform.

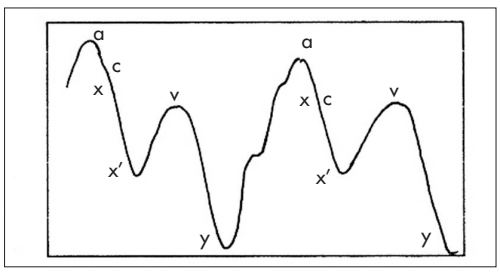

Figure 3. Jugular venous pressure in pericardial constriction.

The apex pulse is diffuse and impalpable. A compensatory tachycardia maintains cardiac output. Abrupt termination of rapid ventricular filling by the constricted pericardium causes the 'pericardial knock', an abnormal, early, diastolic sound or third heart sound, best heard at the left sternal edge. Pulsus paradoxus is an invariable finding in cardiac tamponade. It is rarely found in pericardial constriction, but if found, it is usually mild, as the reductions in arterial pressure are usually <10 mmHg unless pericardial pressures are elevated themselves by a tense effusion (*Table 4*).

RESULTS OF INVESTIGATIONS

The investigations described are rarely definitive in isolation. If clinical suspicion is high, different imaging modalities must be used to confirm the diagnosis (*Table 5* summarizes investigations).

Electrocardiography

The electrocardiogram (ECG) is non-diagnostic. The changes commonly found are low-voltage QRS complexes, generalized T-wave inversion and tachycardia. Left atrial enlargement is characterized by P mitrale. Atrial fibrillation occurs in less than 50% of cases from previous studies (Lorell, 1997). Localized constriction of the epicardial coronary arteries may produce changes consistent with infarction, and right ventricular constriction may produce changes of right ventricular hypertrophy and strain pattern (Chesler et al, 1976).

Chest X-ray

X-ray findings may be variable and are not diagnostic. The cardiac silhouette may appear reduced, normal or enlarged, depending on the extent of

constriction, presence of coexistent effusion, thickness of the pericardium and the condition of the ventricle before the onset of constriction.

The classic sign of pericardial constriction is calcification of the pericardium (*Figure 4*). However, even if pericardial calcification is shown, constriction might not always be indicated. Lateral views are helpful in demarcating the extent and location of calcification.

Left atrial enlargement may be present as well as prominent pulmonary vessels as a result of venous engorgement. Pleural effusions are found in up to 60% of patients with constriction. Pulmonary oedema is rare.

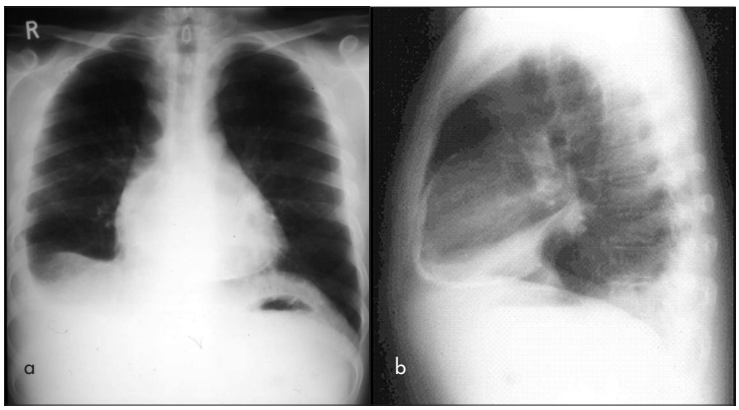

Figure 4. Pericardial calcification on chest X-ray. a. Posterior–anterior. b. Lateral.

Echocardiography

Two-dimensional echocardiogram: On the two-dimensional echocardiogram, the pericardium appears bright and thickened, although this method is unreliable for assessing pericardial thickness. The septum displays sudden posterior movement during early diastole, and the inferior vena cava and hepatic veins appear dilated with diminished respiratory variation.

M-mode echocardiogram: The M-mode echocardiogram confirms the presence of a thickened pericardium, abnormal septal motion, left atrial dilatation and normal left ventricular size and function. Normal ventricular function strongly supports a diagnosis of constrictive pericarditis as opposed to restrictive cardiomyopathy.

Doppler studies

Doppler studies, used in conjunction with respiratory recordings, are highly sensitive for diagnosing constriction and guiding further management (Hatle et al, 1989; Oh et al, 1994). The Doppler velocity of mitral inflow, specifically early rapid ventricular filling (E) and late filling with atrial contraction (A), was measured with flow velocities in the hepatic vein or superior vena cava during inspiration and expiration. Constriction was associated with an increase (>25%) in the E velocity and an increased diastolic flow reversal with expiration in the hepatic veins.

Computed tomography and magnetic resonance imaging

Both imaging modalities are helpful in differentiating pericardial constriction from restrictive cardiomyopathy by defining the presence of a thickened pericardium (*Figure 5a, b*). Clinical and echocardiographic features in these conditions may be very similar. Masui et al (1992) demonstrated that the diagnostic accuracy of magnetic resonance imaging (MRI) for diagnosing pericardial constriction was 93%. Detection of pericardial thickness of >4 mm distinguished pericardial constriction from restrictive cardiomyopathy – an important differentiation, as therapeutic interventions which influence the survival rates are possible for pericardial constriction (Smith et al, 2001).

The classic changes visible on MRI or computed tomography scanning that are suggestive of a constrictive process include a thickened pericardium, small tubular shaped ventricles, a flattened or sigmoid-shaped septum and a dilated right atrium and inferior vena cava.

Right and left heart catheterization

Haemodynamic changes characteristic of constriction are obtained during right and left heart catheterization.

In *Figure 6a*, the lower trace represents the pressure in the right atrium during the procedure;

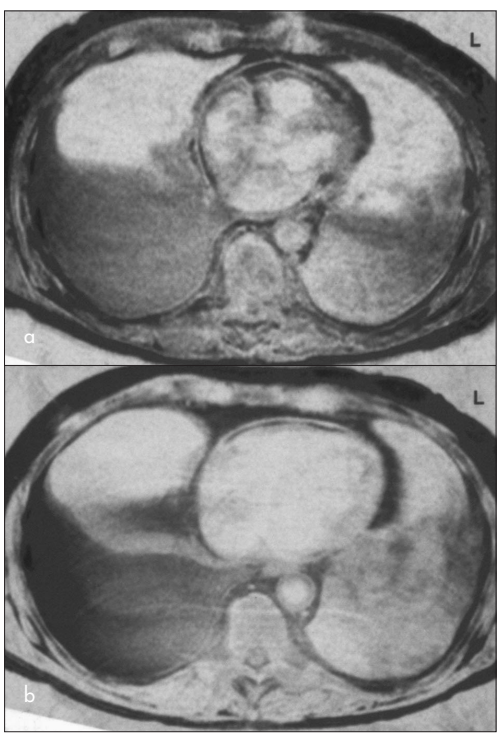

Figure 5. a. T1 weighted and (b) T2 weighted axial images show diffusely thickened pericardium encasing the heart. From Smith et al (2001).

the other trace is the left ventricular pressure. The normal right atrium trace is usually low pressure (0–5 mmHg) and follows a similar pattern to JVP variations with respiration. In this example, right atrial pressures range from 15–25 mmHg.

The constrictive process results in an increase in right atrial pressure (Kussmaul's sign). The elevated right atrial pressures and impaired ventricular filling enhance the normal variation seen with inspiration and expiration to produce an 'M' or 'W' type configuration (boxed area, *Figure 6a*).

The lower trace in *Figure 6b* shows right ventricular pressures and the other trace pressures in the left ventricle. Rapid ventricular filling produces a steep y descent that is abruptly curtailed by the constricting pericardium limiting ventricular expansion. This is manifested on the simultaneous right and left ventricular pressure recordings by the 'dip and plateau' waveform or 'square root sign' (shaded area, *Figure 6b*). During diastole, right and left ventricular pressures are equalized.

Vaitkus and Kussmaul (1991) assessed the reliability of haemodynamic criteria for differentiating between the diagnoses of pericardial constriction and restrictive cardiomyopathy. They identified three characteristics that are classic for pericardial constriction:

1. A difference between right and left ventricular end diastolic pressures of <5 mmHg
2. Right ventricular systolic pressures <50 mmHg
3. Right ventricular end diastolic pressure:right ventricular end systolic pressure of >1:3.

Endomyocardial biopsy

Endomyocardial biopsy is not essential for diagnosis. Biopsy may help identify the specific cause of restrictive cardiomyopathy and identify myocyte atrophy in cases of pericardial constriction.

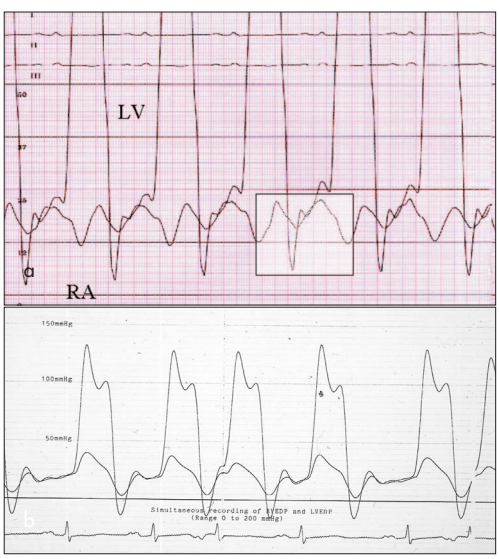

Figure 6. a. Simultaneous pressure recordings of the left ventricle (upper trace) and right atrium (lower trace) in pericardial constriction. b. Simultaneous pressure recordings of the left (upper trace) and right (lower trace) ventricles in pericardial constriction.

MANAGEMENT

Initial management with low-sodium diet, diuretics and aspiration of pleural and ascitic fluid may temporarily palliate the patient. However, the disease will recur and progress. Negatively inotropic drugs should be avoided, as tachycardia is a compensatory response to maintain cardiac output.

Therapy for tuberculous pericarditis should include steroids, as these reduce the progression to constriction. However, in patients with coexistent constriction and active tubercle infection, management should be directed towards surgery in conjunction with anti-tuberculous therapy, and pericardiectomy should not be delayed for fear of TB-related complications.

Surgical outcomes

The definitive treatment is extensive and complete pericardiectomy. Recurrence of the disease process is likely if resection is incomplete. Early in-hospital mortality figures vary from 0–18% (*Table 6*). Without surgery, myocyte atrophy continues to impair ventricular function and the outlook is very poor. The patient becomes rapidly disabled and dependent (Somerville, 1968).

Longer term follow-up confirms the beneficial effect of pericardiectomy (72–96% survival).

The results of varying surgical management of patients with pericardial constriction were compared in a contemporary cohort between 1995–2000 at the Middlesex Hospital, London. Thirty-six cases of pericardial constriction were identified over a 5-year period, of which 47% (n=17) were female. Mean age at presentation was 47 years (range 18–73 years). Mean length of follow-up was 12.5 months (range 0–47 months). Aetiology was variable and included malignant disease (n=21), post-cardiotomy (n=5), tuberculous disease (n=3) and others including connective tissue disease (n=1), idiopathic (n=4), traumatic (n=1) and post-infection (n=1). Preferred treatment was a pericardial window in 23/36 patients (64%); four patients (11%) underwent partial pericardiectomy through a median sternotomy, and nine patients had a total pericardiectomy (25%).

Patients with malignant disease with (n=8) or without (n=13) mediastinal irradiation fared worst, irrespective of age (mean 39.3 years) or method of pericardiectomy: pericardial window (14/15 (93%), mean survival 5.6 months), total pericardiectomy (3/4 (75%), mean survival 12.3 months), partial pericardiectomy (1/2 (50%), mean survival 4 months). Patients undergoing procedures for constriction post-cardiotomy were older (mean age 51 years *vs* 33.2 years) and yielded more favourable results (mortality 20%, mean survival time 29.8 months). Patients with TB and

other causes of pericardial constriction displayed the most favourable outcomes (mortality 10%, follow-up 14.3 months to date). Outcome was associated with diagnosis at presentation. Similar results were found at the Mayo Clinic (Ling, 1999).

Pericardiectomy carries little benefit for patients with malignant disease, irrespective of treatment with or without radiation or the prognosis from the malignancy itself.

CONCLUSIONS

A diagnosis of constrictive pericarditis is often reached after a prolonged course of investigation and is difficult and challenging. The aetiology is most often idiopathic in the West, although TB is the most common cause worldwide. Clinical signs of elevated JVP, tachycardia, ascites, hepatomegaly and peripheral oedema with preserved ventricular systolic function should lead to the diagnosis. Investigations should be extensive, including ECG, chest X-ray, echocardiography, right and left heart catheterization and computed tomography or MRI. Treatment with extensive pericardiectomy provides best results. HM

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Conflict of interest: none.

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TABLE 6.
Early operative mortality and long-term survival in pericardial constriction

Reference (*n*)	Early mortality (<30 days) (%)	Long-term survival (%)	Follow-up range (mean)
Wood (1961) (*n*=40)	15	82	4 years (N/A)
Cordell and Beason (1968) (*n*=14)	14	86	8 months–17years (7.8 years)
Wychulis et al (1971) (*n*=137)	14	72	1.3–25.3 years (10.6 years)
Das et al (1973) (*n*=77)	18	87	5 months–17 years (N/A)
Robertson and Mulder (1984) (*n*=31)	10	96	7 months–5 years (2.6 years)
Arsan et al (1994) (*n*=105)	10.5	88	1 year–11 years (5.8+/-2.1 years)
Bashi et al (1988) (*n*=118)	16	92	2 years–30 years (9 years)
De Valeria et al (1991) (*n*=36)	5.6	77	N/A (4.8 months)
Tirilomis et al (1994) (*n*=71)	6	80	1 month–13.25 years (11+/-5.8 years)
Trotter et al (1996) (*n*=21)	0	N/A	N/A

N/A = not available

KEY POINTS

- The aetiology of pericardial constriction is most commonly idiopathic. Worldwide, tuberculosis is the commonest cause.
- Clinical signs of right ventricular overload include elevated jugular venous pressure, tachycardia, ascites, hepatomegaly, peripheral oedema and preserved ventricular systolic function.
- Investigations should be extensive and include electrocardiography, chest X-ray, echocardiography, right and left heart catheterization, computed tomography and magnetic resonance imaging.
- Treatment with complete pericardiectomy provides best results.