Clin. Cardiol. 18, 341–350 (1995)

# Clinical Pathologic Correlations

*This section edited by Bruce Waller, M.D.*

# Constrictive Pericarditis: Its History and Current Status

NOBLE O. FOWLER, M.D.

Division of Cardiology, Department of Medicine, University of Cincinnati College of Medicine, Cincinnati, Ohio, USA

**Summary:** The diagnosis of constrictive pericarditis remains a challenge because it is often mimicked by restrictive cardiomyopathy. The last few years have seen numerous advances in our ability to differentiate between these two conditions which often have similar physical findings and hemodynamics. This review begins with a brief history of constrictive pericarditis; this is followed by an extensive discussion of newer etiologies, and then the classical clinical history and physical examination findings are described. Radiologic, electrocardiographic, and angiographic findings are discussed. The hemodynamics of constrictive pericarditis are reviewed. Recent results of echocardiographic and echo-Doppler investigations are presented. Emphasis is placed upon the limitations of M-mode echocardiography in the diagnosis of constrictive pericarditis. The value of echocardiographic Doppler studies of mitral and tricuspid flow velocity patterns, as well as of those in the pulmonary veins and hepatic veins, is described. Nuclear ventriculograms and angiocardiograms tend to show more rapid ventricular filling in constrictive pericarditis than in restrictive cardiomyopathy. Although only a small number of patients has been studied, these evaluations seem to have merit in separating restrictive cardiomyopathy from constrictive pericarditis. The role of computed tomography scanning and magnetic resonance imaging studies of pericardial thickness in confirming the presence of constrictive pericarditis is discussed. Abnormal pericardial thickening (> 3 mm) confirms the diagnosis of constrictive pericarditis, but only if the characteristic hemodynamic pattern is present. The usefulness of endomyocardial biopsy in recognizing specific varieties of restrictive cardiomyopathy is presented. The topic of occult constrictive pericardial disease is discussed briefly. A discussion of the timing of pericardial resection for the treatment of constrictive pericarditis ends the review.

**Key words:** constrictive pericarditis, pericardial disease

## Introduction and Historic Events

Constrictive pericarditis has been defined as a chronic fibrous thickening of the wall of the pericardial sac which is so contracted so that normal diastolic filling of the heart is prevented.[1]

The existence of constrictive pericarditis has been known for centuries. In 1669, Richard Lower wrote of dyspnea and intermittent pulse in a patient with constrictive pericarditis.[2] In 1842, Corrigan described the pericardial knock sound (bruit de frappement).[3] In 1873, Kussmaul described the paradoxical arterial pulse in mediastinopericarditis.[4] The eponym "Pick's disease" was given to constrictive pericarditis with ascites and hepatomegaly following Pick's description.[5] The first successful pericardiectomy in the U.S. was performed in 1929 by Churchill.[6] The modern era of diagnosis and treatment of this disease was signaled by Paul Dudley White's St. Cyre's Lecture in 1935.[1] This paper described 15 patients, 7 of whom were successfully operated upon at the Massachusetts General Hospital. Bloomfield[7] demonstrated elevated right atrial pressure and elevated right ventricular (RV) diastolic pressure with an early diastolic dip in a patient who had constrictive pericarditis. Hansen *et al.*'s paper in 1951 dealt with the RV dip and plateau pressure-pulse pattern.[8] A similar pressure-pulse pattern in restrictive cardiomyopathy was described 2 years later.[9] Hancock popularized the condition known as effusive-constrictive pericarditis.[10] Constrictive pericarditis as a complication of cardiac surgery was first reported in 1972.[11] The value of computed tomography (CT) scanning in constrictive pericarditis was discussed by Isner *et al.* in 1982,[12] and Soulen *et al.* wrote of magnetic resonance imaging (MRI) studies in this disease in 1984.[13]

Address for reprints:

Noble O. Fowler, M.D.
University of Cincinnati College of Medicine
Medical Sciences Building
231 Bethesda Avenue
M.L. #542
Cincinnati, OH 45267, USA

Received: May 17, 1994
Accepted: June 14, 1994

TABLE I Etiologic background of constrictive pericarditis

1. Unknown antecedent
2. Following idiopathic pericarditis
3. Specific infections
   - Bacterial
   - Tuberculosis
   - Fungal disease—e.g., histoplasmosis, coccidioidomycosis
   - Viral diseases, especially Coxsackie B
   - Parasitic disease: amebiasis, echinococcosis
4. Connective tissue disease: rheumatoid arthritis, lupus erythematosus, scleroderma
5. Neoplastic disease
   - Secondary to breast cancer, lung cancer, lymphoma, melanoma
   - Primary mesothelioma
6. Trauma
   - Nonpenetrating
   - Penetrating
7. Post cardiac surgical procedures (incidence 0.2–0.3%)
8. Radiation therapy
9. End-stage renal disease
10. Following cardiac pacemaker insertion
11. Following certain drugs—e.g., methysergide, procainamide-induced lupus syndrome
12. Hereditary: mulibrey nanism
13. Rare: sarcoidosis, asbestosis, postmyocardial infarction, amyloidosis

## Etiology

Most causes of acute pericarditis may also cause chronic constrictive pericarditis (Table I). Idiopathic pericarditis headed the list of known antecedents in some series[14, 15] but the most common group is that with no recognized antecedent.[15–19] In earlier series, tuberculosis was the most commonly recognizable cause[20, 21] In a 1990 series from the Mayo Clinic, comprising 313 patients operated upon since 1936, 6% had tuberculosis.[18] However, in nonindustrialized nations such as India, tuberculosis was found to cause 61% of 118 instances of constrictive pericarditis.[22]

Constrictive pericarditis may follow nonpenetrating[23] or penetrating trauma, including cardiac surgical procedures or the penetration of pacing catheters into the pericardial space.[24] Cardiac surgical procedures are estimated to be followed by constrictive pericarditis in 0.2 to 0.3% of instances; 158 cases were found in the world literature in 1989.[25] Another report in the same year described personal experience with 45 cases. The interval between the cardiac operation and the appearance of symptoms ranged from 1 to 204 months (mean 23.4 months). In this series, 62% had had the post-pericardiotomy syndrome.[26] An unusual instance of traumatic constrictive pericarditis resulted from self-mutilation by sewing needles inserted through the chest wall.[27]

Radiation therapy of mediastinal tumors is an important cause of pericardial disease, in addition to causing myocardial, valvular, and coronary artery disease.[28] Radiation therapy may be followed by acute pericarditis; clinical evidence of constrictive pericarditis may appear years later. Radiation therapy for Hodgkin's disease may be followed by some variety of pericarditis in 20% of instances.[29] Radiation therapy was a leading cause of constrictive pericarditis in 95 cases reported from Stanford University, in which 31% of instances followed radiotherapy.[17] On the other hand, radiation therapy was responsible in only 5% of 313 cases studied at the Mayo Clinic.[18] Radiation therapy was responsible in 2 of 27 cases in another series[19] and in none of 26 cases in another.[30]

Patients with end-stage renal disease who are treated by hemodialysis or renal transplantation may develop constrictive pericarditis in addition to acute pericarditis and cardiac tamponade.[31–34] We have seen one case recently.

### Connective Tissue Disease

Rheumatic fever generally is considered a rare cause of constrictive pericarditis, although Roberts and Spray found it to be a cause in 2 of 314 cases.[35] A Mayo Clinic study found rheumatic fever to be the cause in 4 of 231 cases of constrictive pericarditis.[36] Rheumatoid arthritis is a fairly common cause of constrictive pericarditis or effusive-constrictive pericarditis.[37–40] Lupus erythematosus may be followed by constrictive pericarditis. A total of seven cases, six in men, had been reported by 1988.[41] Rarely, constrictive pericarditis may follow a drug-induced lupus-like syndrome.[42, 43]

### Infectious Diseases

Many pericardial infections may be followed by constrictive pericarditis.[44–46] Infections caused 3% of constrictive pericarditis in cases reported in the Mayo Clinic series.[18] Tuberculosis was responsible for 0–8% of cases in recent studies in Western countries,[17, 30, 36] but causes the majority of cases in India[22] and in certain areas of Africa.[47] Viral pericarditis has been identified as a cause of constrictive pericarditis[15, 48, 49] or was suspected because the syndrome of constrictive pericarditis followed an epidemic of viral disease.[14] In endemic areas, histoplasmosis may be a relatively common cause of pericarditis and may be followed by constrictive pericarditis;[50] however, a follow-up study of 10 of 16 cases of acute histoplasma pericarditis found no instances of constrictive pericarditis 6 months to 10 years later.[51] Histoplasmosis may produce fibrosing mediastinitis, accompanied by both constrictive pericarditis and superior vena caval obstruction.[52] Recently reported cases of constrictive pericarditis caused by infections include *Legionella pneumophila,*[53] meningococcal infection,[54] Lassa fever,[55] Whipple's disease,[56] actinomycosis,[57] nocardia asteroides,[58] staphylococcal infection after cardiac surgery,[59] salmonella,[60] and *Streptococcus milleri.*[61]

### Neoplastic Disease

Metastatic neoplasm, most commonly from the lung, the breast, or one of the lymphoma group, may be responsible for the syndrome of constrictive pericarditis.[62] In one recent series,

4 of 27 cases were caused by neoplastic disease.[19] Occasional instances are caused by primary pericardial mesothelioma.[63]

### Rare and Uncommon Causes (Table II)

Myocardial infarction may be followed by constrictive pericarditis, but only rarely, with only a few instances having been reported.[19, 64–67] Some instances are associated with Dressler's postmyocardial infarction syndrome,[67] and others with hemopericardium complicating anticoagulant therapy.[65]

There appears to be an association between congenital atrial septal defect and constrictive pericarditis.[68–72] Just and Mattingly[73] reported 4 cases, and 63 from a literature review, of association between atrial septal defect and pericardial adhesion, effusion, or constriction. The reason for this association is unknown.

Constrictive pericarditis may have a hereditary background, for example, mulibrey nanism, reported principally from Finland.[74] These patients have skeletal muscle hypotonia, hepatic enlargement (? congestive), dilated cerebral ventricles, and retinal pigmentation hence mu for *mu*scle, li for *li*ver, br for *br*ain, ey for *ey*es. Nanism is from the Greek nanos, a dwarf. A few instances have been reported from the United States.[75]

Other uncommon causes of constrictive pericarditis include sarcoidosis,[76] primary chylopericardium,[77] dermatomyositis,[78] asbestosis,[79, 80] systemic amyloidosis,[81] and implantation of a cardioverter defibrillator.[82]

A few instances have been associated with drugs, such as procainamide-induced lupus syndrome,[42] methysergide therapy,[83, 84] and hydralazine-induced lupus syndrome.[43] Rarely, a transient hemodynamic pattern of constrictive pericarditis may be found in acute idiopathic pericarditis.[85]

## History

Many series have noted a male preponderance. In a series described by Wychulis *et al.*, 100 of 137 patients were male, ranging in age from 8 to 70 years.[15] The symptoms of constrictive pericarditis usually develop slowly over a period of years, but occasionally, especially when the cause is known, can be shown to develop within a few months after cardiac surgery or mediastinal irradiation. In one series of 45 patients, symptoms developed between 1 and 204 months following cardiac surgery.[26] Gimlette[86] reported that 28 patients developed constrictive pericarditis within 1 year after acute pericarditis. Wise and Conti[87] reported on more than 100 patients, among whom dyspnea, present in 78%, was the most common symptom; edema was present in 64%, abdominal swelling in 64%, abdominal discomfort in 32%, fatigue in 25%, and orthopnea in 22%. Abdominal discomfort, nausea, and vomiting may be due to hepatic or bowel congestion. Schiavone[88] reported dyspnea in each of 18 patients, edema in 13 of 18, and bloating in 12 of 18. Wychulis *et al.*[15] reported effort dyspnea in 90% of 137 cases. Chest pain, possibly due to active inflammation, was present in 24%. Right upper quadrant or abdominal pain was reported in 11%, and only three patients reported orthopnea or paroxysmal nocturnal dyspnea. Cameron *et al.*[17] reported exertional dyspnea in 56% of 95 cases; fatigue was present in 55%, increasing abdominal girth in 29%, and abdominal pain in 12%.

TABLE II Rare and uncommon causes of constrictive pericarditis (1988–1993)

| |
|---|
| Amyloidosis |
| Actinomycosis |
| Nocardia asteroides |
| Implantable cardioverter defibrillator infection |
| Myocardial infarction (Dressler's syndrome) |
| Asbestosis |
| Whipple's disease |
| Lassa fever |
| Hydralazine-induced lupus-like syndrome |
| Pericardial mesothelioma |
| Dermatomyositis |
| Self-mutilation with sewing needles |
| Sclerotherapy of esophageal varices |

## Physical Examination

General examination may show abdominal enlargement due to ascites. Schiavone reported ascites in 17 of 19 patients;[88] however, the Stanford group reported ascites in only 28% of 95 cases,[17] and Bashi *et al.*[22] in 90% of their 118 cases. Tuna and Danielson found ascites in 60% of the Mayo Clinic series.[18] Hepatomegaly is common, being found in 73% of one series,[18] and in 100% of another.[22] The combination of ascites and hepatomegaly may lead to a mistaken diagnosis of liver disease, as occurred initially in 10 of 95 patients reported from Stanford.[17] Splenomegaly due to portal hypertension is common[16, 21] However, the almost universal presence of elevated jugular venous pressure, 99% in one study[18] and 100% in two others,[22, 89] should eliminate liver cirrhosis as the cause of hepatomegaly and ascites. Peripheral edema was found in 64% of one series[87] and 70% of another.[36] Schiavone reported edema in 13 of 19 cases,[88] and edema was reported in 84% of another group.[22] The cardiac apical impulse is often absent; this was true in 90% of Wood's series.[21]

Many of the physical findings that are thought characteristic of constrictive pericarditis are, in fact, quite variable. Paradoxical arterial pulse is an inconsistent finding; it was found in 40% in one series,[15] 16% in another,[17] 14% in another,[89] but in 84% in one study.[22] A pericardial knock sound was described in 5%,[17] in 46% ($S_3$),[36] in 11 of 19 cases,[88] and 36%.[89] Kussmaul's sign (inspiratory swelling of the neck veins) is a rather uncommon finding, appearing in only 13% of 95 patients.[17] This sign is not specific; it also occurs with RV failure, restrictive cardiomyopathy, RV infarction, and in tricuspid stenosis.[90] The mechanism of Kussmaul's sign is uncertain.[91] Kussmaul ascribed this phenomenon to inspiratory

traction on the great veins in the mediastinum. Studies of patients with constrictive pericarditis show little respiratory variation in superior vena caval flow velocity.[92] One study ascribed the inspiratory swelling of the neck veins to transmission of the normal inspiratory increase of intra-abdominal pressure to a tense, overly filled systemic venous system.[90]

One physical finding is against the diagnosis. Cardiac murmurs usually are not found unless there is complicating valvular disease or a fibrous band constricting the RV outflow tract. Paut *et al.*[20] found murmurs in only 3 of 53 patients, one of whom had aortic stenosis. Schrire *et al.*[93] reported tricuspid diastolic murmurs in two cases, probably caused by constriction of the atrioventricular ring, producing tricuspid stenosis.

## Radiologic Studies

The typical chest radiogram shows a heart of normal size, with clear lung fields. However, the cardiopericardial silhouette may be enlarged, especially with effusive-constrictive pericarditis. Left atrial enlargement may occur.[94] Paul *et al.* found moderate or marked increase in heart size in 38% of patients of their series.[20] Radiologic evidence of pericardial calcification was found in 40% of 231 cases in the Mayo Clinic series,[36] but in only 5% of the Stanford series.[17] In the study by Bashi *et al.* of 118 patients, pericardial calcification was found in 21%[22] and in another by Oh *et al.*[89] it was found in 1 of 25 patients. Pleural effusions are found in ≥ 60% of patients, and pulmonary edema in 5–10%.[95]

Angiocardiographic studies tend to show loss of the normal outward convexity of the right atrial border, as well as evidence of pericardial thickening at the right atrial level.

## Electrocardiographic Studies

The electrocardiogram (ECG) is seldom normal in constrictive pericarditis. An intra-atrial conduction defect with "P mitrale" pattern is common.[96] P-wave changes suggestive of left atrial enlargement were found in 37% of 54 cases in one study,[97] in 19% of 47 cases in another,[98] an in 31% of 122 cases in another.[99] Low voltage of the QRS complex and atrial arrhythmias are frequent findings. Paul *et al.*[20] reported atrial fibrillation in 18 of 52 patients and atrial flutter in 5. Wood found atrial fibrillation in 35% and atrial flutter in 10%.[21] In his study, atrial fibrillation was more common when the process was of longer duration. Cameron *et al.*[17] reported atrial fibrillation in 13% of 95 patients. The study by McCaughan *et al.* of 231 patients reported low-voltage QRS in 40% and atrial arrhythmias in 29%.[36] Bashi *et al.*[22] found low-voltage QRS in 75% of 118 patients and atrial arrhythmias in 10%. Atrial flutter was found in 5 of 52 cases in one study,[20] in 3 of 78 cases in another,[97] and in 3 of 67 in another.[98] Patterns of bundle-branch block or ventricular hypertrophy are uncommon. Right ventricular hypertrophy, in some instances, produced by a fibrous band constricting the RV outflow tract, was reported in 6 of 122 cases.[99] Chesler *et al.* also reported a single case due to right ventricular outflow tract obstruction by a fibrous band.[100] We reported an instance of RV hypertrophy associated with constriction of the left atrioventricular groove in a 13-year-old boy.[94] Fukuda *et al.*[101] reported a case with ECG evidence of RV hypertrophy without outflow tract obstruction. Levine reported changes of left ventricular (LV) hypertrophy in 5 of 67 patients and a suggested pseudoinfarction pattern in 6 of the 67.[98]

## Echocardiograms and Echo-Doppler Studies

### M-Mode Echocardiograms

M-mode echocardiograms provide useful information in constrictive pericarditis, but are not diagnostic of the disease. Ventricular dimensions usually are normal and ventricular function is preserved. Pericardial thickening was recognized in only 38% of one series of 40 patients[102] and in 42% of another.[22] In the series of Engel *et al.*,[102] left atrial enlargement was present in 75% and premature pulmonary valve opening in 14% of cases. This last finding may be explained by the fact that the elevated RV diastolic pressure is equal to pulmonary artery diastolic pressure, or nearly so. Paradoxical septal motion is nearly always present. Diastolic flattening of the LV posterior wall is often present and was found in 85% of this series. An atrial systolic septal notch may be seen.[102]

### Two Dimensional Echocardiograms

Two-dimensional echocardiograms may offer some help in recognizing constrictive pericarditis but are more useful when supplemented by Doppler studies. Characteristically, there is biatrial enlargement, with normal ventricular dimensions. Ventricular ejection fraction is preserved. Diastolic septal bounce may be seen. Fast-speed echocardiographic studies may show evidence of rapid ventricular filling in early diastole.[103] D'Cruz *et al.*[104] described a decreased angle (<150°) between the posterior wall of the left atrium and that of the left ventricle in the parasternal long-axis view in five of seven patients with constrictive pericarditis. This was not seen in other forms of heart disease with left atrial enlargement, except in 1 of 16 patients with mitral stenosis. Inspiratory movement of the interventricular and interatrial septum toward the left has been described.[105]

*Inferior vena caval plethora:* The inferior vena cava usually is dilated in constrictive pericarditis, and its diameter shows little respiratory variation;[106] however, RV failure or cardiac tamponade may show the same findings.

*Superior vena caval flow velocity patterns:* Superior vena caval Doppler flow velocity patterns were studied in 14 patients with cardiac tamponade, 7 with constrictive pericarditis, and 8 normal subjects.[92] In six of seven patients with constrictive pericarditis, diastolic flow velocity exceeded systolic flow velocity. There was little respiratory variation in systolic flow velocity in normal subjects or in constrictive pericarditis. In tamponade there was little diastolic flow in the first expiratory heart beat, corresponding to a loss of the right atrial *Y* descent.

With tamponade, there was marked inspiratory augmentation of both systolic and diastolic flow velocity.

*Pulmonary venous flow:* Schiavone *et al.*[107, 108] in a study of four patients with constrictive pericarditis, found that both systolic and diastolic flow velocity increased during expiration in constrictive pericarditis, but only diastolic flow velocity showed an expiratory increase in four cases of restrictive cardiomyopathy due to amyloidosis. Klein *et al.*[109] studied 14 patients with constrictive pericarditis by Doppler transesophageal echocardiography. In inspiration, the pulmonary venous systolic/diastolic flow velocity ratio fell below 0.65 in constrictive pericarditis. Also, peak diastolic flow velocity fell 40% on average during inspiration. These two features separated constrictive pericarditis from restrictive cardiomyopathy.

*Hepatic vein flow velocity patterns:* Von Bibra and associates[110] studied 13 patients with constrictive pericarditis and 25 with RV pressure overload. Patients with constrictive pericarditis showed late systolic and late diastolic flow reversal; those with tricuspid regurgitation showed only systolic flow reversal. Oh and associates[89] found expiratory augmentation of diastolic flow reversal (≥ 25% of forward flow) in a study of 25 patients with constrictive pericarditis.

*Tricuspid and mitral valve flow velocity patterns:* Using Doppler echocardiography, Hatle *et al.*[111] studied mitral and tricuspid valve flow velocity patterns in 7 patients with constrictive pericarditis and in 12 with restrictive cardiomyopathy. Patients with constrictive pericarditis had marked inspiratory decrease in early mitral flow velocity and increase in early tricuspid flow velocity compared with normal controls and with patients with restrictive cardiomyopathy. Mancuso *et al.*[112] studied seven patients with constrictive pericarditis and six with restrictive cardiomyopathy. Patients with constrictive pericarditis showed higher diastolic mitral flow velocity patterns at the onset of expiration, with a decrease at the onset of inspiration. Reciprocal flow velocity changes with respiration were found across the tricuspid valve. Patients with restrictive cardiomyopathy showed little change in mitral and tricuspid flow velocity with respiration, but had moderate to severe mitral and tricuspid regurgitation. Trivial mitral and tricuspid regurgitation was found in only one patient with constrictive pericarditis. Oh and associates[89] found that there was < 10% respiratory variation in mitral valve early diastolic velocity in normal subjects and in those with restrictive cardiomyopathy, whereas with constrictive pericarditis there was a > 25% expiratory increase in mitral valve early diastolic velocity.

## Hemodynamics

The hemodynamics of constrictive pericarditis were reviewed by Shabetai *et al.*[113] Since, by definition, constrictive pericarditis impairs diastolic filling of the ventricles, elevation of both RV and LV end diastolic pressures is to be expected. This finding was reported in 100% of the Mayo Clinic series.[36] Right atrial pressure elevation with prominent *X* and *Y* descents ("W" wave form) was first reported by Bloomfield *et al.*[7] These authors also described the early diastolic dip and narrow pulse pressure in the RV pressure pulse. Hansen and co-workers[8] described a diastolic dip and plateau pattern in the right ventricle in six cases of constrictive pericarditis. Wood showed that LV end-diastolic pressure usually did not exceed RV end-diastolic pressure by more than 5 mmHg.[21] Yu *et al.* stated that RV systolic pressure usually did not exceed 50 mmHg and that RV diastolic pressure was characteristically more than one-third of RV systolic pressure in constrictive pericarditis.[114]

Wood pointed out that the cardiac output tends to be greater and the systemic arteriovenous oxygen difference smaller in constrictive pericarditis than in cardiomyopathy.[21] In his series, cardiac output averaged 4.7 l/min in constrictive pericarditis, and 3.5 l/min in cardiomyopathy. Arteriovenous oxygen difference averaged 51 ml/l in constrictive pericarditis, and 75 ml/l in cardiomyopathy. In early constrictive pericarditis, the cardiac output tends to be normal.[115] In a study of 10 patients with constrictive pericarditis, Reddy[116] reported mean right atrial pressures from 7 to 30 mmHg, and RV end-diastolic pressures from 9 to 32 mmHg. Left ventricular end-diastolic pressures were from 9 to 32 mmHg. The cardiac index ranged from 1.4 to 3.2 l/min/m$^2$, and the arteriovenous oxygen difference from 4.8 to 9.2 vol %. In seven patients, Tyberg *et al.* reported right atrial pressures from 14–24 mmHg and pulmonary wedge pressures 14–26 mmHg.[117] Cardiac index was 2.0–3.2 l/min/m$^2$. Left ventricular ejection fractions were normal in all. Reddy[116] reported that right atrial pressure tracings showed prominent *X* and *Y* descents, with little respiratory variation. Normally, the right atrial mean pressure falls several mmHg relative to intrathoracic pressure during inspiration. With constrictive pericarditis, because of the fibrotic shell surrounding the heart, inspiratory fall in intrathoracic pressure is not reflected in the right atrial pressure tracing, and the right atrial pressure tends to show no change or may actually rise during inspiration.

Cameron *et al.*[17] reported on 95 patients with constrictive pericarditis, 23 of whom had effusive-constrictive disease. Mean right atrial pressure was 16 ± 5 mmHg; mean RV end-diastolic pressure was 18 ± 6 mmHg; mean pulmonary capillary wedge pressure was 19 ± 5 mmHg; and mean LV end-diastolic pressure was 21 ± 5 mmHg. Mean cardiac index was 2.2 ± 0.7 l/min/m$^2$.

Occasional instances of localized cardiac constriction by fibrous bands are reported. Vallance *et al.*[118] described an instance of constriction of the RV outflow tract, with an RV pressure of 115/7–16 mmHg and a pulmonary arterial pressure of 30/11 mmHg. There was an ECG pattern of RV hypertrophy. Pulmonary trunk constriction by a fibrous or a fibro-calcific band has been reported.[100, 119] Fibrous bands constricting the atrioventricular grooves may produce tricuspid or mitral stenosis.[55, 93, 94] Some instances of RV outflow tract obstruction by fibro-calcific bands have followed previous surgery for constrictive pericarditis.[120]

Circulating atrial natriuretic factor tends to be low or normal in constrictive pericarditis, rising after pericardial resection.[121, 122] This suggests that atrial natriuretic factor release is more likely to be associated with atrial stretch than with in-

creased atrial pressure alone. Anand and associates studied 16 patients with constrictive pericarditis and compared them with a group with myocardial disease and edema.[123] Right atrial pressure tended to be higher and pulmonary arterial pressure lower in the group with constrictive pericarditis. Total body water, extracellular fluid volume, and exchangeable sodium were higher in constrictive pericarditis, and circulating atrial natriuretic peptide values were lower. Plasma norepinephrine, renin activity, and aldosterone were comparably elevated in the two groups.

## Effusive-Constrictive Pericarditis

This condition was mentioned by Wood[21] and by Spodick and Kumar[124] and was popularized by Hancock.[10] In this disease, in addition to pericardial thickening and diastolic cardiac constriction, there is a collection of fluid between the parietal and visceral pericardium (epicardium). As a result, the cardiopericardial silhouette may be larger on chest radiogram than is usually the case with purely constrictive pericarditis. In the Stanford series of 23 cases, 10 followed radiotherapy.[17] Other etiologies consisted of seven instances of idiopathic pericarditis, three of connective tissue disease, two that followed infections, and one neoplastic. I have seen instances due to rheumatoid disease, tuberculosis, and penetrating trauma. Some instances are associated with uremia.[10] Effusive-constrictive pericarditis was reported to follow Lassa fever,[55] salmonella infection,[60] and streptococcal infection.[61]

The hemodynamic features are characteristic. Right atrial, pulmonary wedge, and intrapericardial pressures are equally increased, and there is a prominent *X* descent and no prominent *Y* descent in the right atrial pressure trace and no prominent early diastolic dip in the RV pressure tracing. When all the pericardial fluid is removed by needle pericardiocentesis, intrapericardial pressure falls to near zero, but the right atrial, RV diastolic, and pulmonary wedge pressures remain elevated. In addition, a prominent *Y* descent appears in the right atrial pressure trace and a large early diastolic dip appears in the RV pressure record.

## Distinction between Constrictive Pericarditis and Restrictive Cardiomyopathy

Patients with constrictive pericarditis and restrictive cardiomyopathy may have similar clinical and hemodynamic patterns. Both may have persistent elevation of systemic venous pressure, a positive Kussmaul's sign, pulsus paradoxus, and a heart that is of normal size or slightly enlarged on chest radiogram. Both conditions often have a preserved LV ejection fraction of ≥ 0.50 and a similar hemodynamic pattern, with an early diastolic dip and plateau pattern in pressure records of both right and left ventricles. Right atrial pressure and pulmonary capillary wedge pressures usually are increased within the range of 12–32 mmHg. In constrictive pericarditis, the pulmonary wedge pressure typically does not exceed the right atrial pressure by more than 4–5 mmHg. Similarly, LV and RV diastolic pressures may be within 4–5 mmHg of each other and are equal to the right atrial and pulmonary capillary wedge pressures. Some instances of restrictive cardiomyopathy have this same equalization of RV and LV filling pressures. Several studies have described methods of distinguishing between these two clinically similar disorders. These include the rate of LV filling; patterns of diastolic flow across the mitral and tricuspid valves; superior vena caval, pulmonary venous, and hepatic venous flow velocity patterns; and MRI and CT scan studies of pericardial thickness. Studies of the relationship between RV and LV diastolic pressures may be helpful, but are seldom definitive. Endomyocardial biopsy may be helpful if a specific infiltrative cardiomyopathy is found. The following studies have been made in relatively small numbers of patients, especially those with restrictive cardiomyopathy, and need confirmation in larger series.

### Left Ventricular Filling Rate

Tyberg and associates, using angiocardiography, found LV diastolic filling to be more rapid in constrictive pericarditis (averaging 85% in the first half of diastole) than in normals (averaging 65% in the first half of diastole), or in amyloid restrictive cardiomyopathy (averaging 45% in the first half of diastole).[117] More rapid LV filling in constrictive pericarditis than in restrictive cardiomyopathy, using fast-speed echocardiography, was found in a study by Janos *et al.*[103] Gerson *et al.*[125] using nuclear ventriculography, found more rapid LV filling in constrictive pericarditis than in normals or in restrictive cardiomyopathy. Aroney *et al.*[126] found that LV diastolic filling was more rapid in constrictive pericarditis than in restrictive cardiomyopathy throughout the first 10–60% of the diastolic period.

### Mitral and Tricuspid Diastolic Flow Patterns

Hatle and associates,[111] using Doppler echocardiography, found that patients with constrictive pericarditis had a marked inspiratory decrease in early mitral flow velocity (≥ 25%), whereas this decrease was < 15% in normals and in patients with restrictive cardiomyopathy. Oh and associates[89] found similar respiratory variations in early mitral flow velocity in patients with constrictive pericarditis. It should be pointed out that similar flow patterns may be found with obstructive airway disease and cardiac tamponade. Mancuso *et al.*[112] found mitral and tricuspid regurgitation common in restrictive cardiomyopathy and uncommon in constrictive pericarditis.

### Pulmonary Venous Flow Velocity

In constrictive pericarditis, Doppler transesophageal echocardiography showed a peak diastolic flow velocity fall of > 40% on inspiration. This plus a systolic/diastolic flow ratio < 0.65 in inspiration demarcated constrictive pericarditis from restrictive cardiomyopathy.[109]

### Hepatic Vein Flow Velocity Patterns

Patients with constrictive pericarditis tend to show late systolic and diastolic flow reversal by Doppler echocardiography.[110] Patients with constrictive pericarditis were found to have expiratory augmentation of diastolic flow reversal.[89] With restrictive cardiomyopathy, there is inspiratory augmentation of flow reversal.[89]

### MRI and CT Scan Studies

This subject was reviewed by Hoit.[127] The normal pericardium is < 3 mm in thickness. If a patient has a hemodynamic pattern consistent with constrictive pericarditis or restrictive cardiomyopathy, the diagnosis of constrictive pericarditis can be made when pericardial thickness is found to be > 3 mm by CT scanning or MRI study. However, not all cases of constrictive pericarditis have such evidence of pericardial thickening. McCaughan *et al.* described pericardial thickening by CT scan in 13 of 16 cases (81%).[36] Masui *et al.* found pericardial thickening (≥ 4 mm) by MRI study in 15 of 17 patients with constrictive pericarditis.[128] Killian *et al.*[26] found the CT scan to show increased pericardial thickening in 23 of 29 post cardiac surgical cases of constrictive pericarditis. Cacoub *et al.*[19] found that only 6 of 16 patients with constrictive pericarditis had pericardial thickening on CT scan, and two were negative on MRI study. Oren *et al.*[129] found increased pericardial thickness in each of five cases of constrictive pericarditis, using cine computed tomography. Oh *et al.*[89] found increased pericardial thickening by CT scan in each of 21 patients with constrictive pericarditis.

### Hemodynamic Investigations

Both restrictive cardiomyopathy and constrictive pericarditis may produce equal elevations of RV and LV end-diastolic pressures to between 12 and 30 mmHg. Pressure values favoring constrictive pericarditis include an RV systolic pressure < 50 mmHg; LV diastolic pressure not exceeding RV diastolic pressure by more than 5 mmHg; RV diastolic pressure exceeding one-third of RV systolic pressure.[130] Vaitkus and Kussmaul's review found that of 70 patients meeting all three criteria, 91% had constrictive pericarditis. Of 18 satisfying one criterion or none, 17 (94%) had restrictive cardiomyopathy.[130]

## Endomyocardial Biopsy

Endomyocardial biopsy may be helpful in distinguishing between constrictive pericarditis and restrictive cardiomyopathy, especially when a specific diagnosis of restrictive disease can be made, for example, cardiac amyloidosis.[131] When the myocardial biopsy shows a normal pattern or a nonspecific pattern of myocardial cell hypertrophy or myocardial fibrosis, there is a 77% probability of constrictive pericarditis, given the characteristic hemodynamic pattern. A problem with regard to interpretation of myocardial biopsy results is that both myocardial and pericardial disease may be present, in particular with sarcoidosis,[76] radiation therapy,[126] pancarditis,[131] or connective tissue disease.

## Treatment

Although some improvement in pulmonary and systemic congestion can often be obtained by the use of diuretics, this is achieved at the expense of a reduction in cardiac output. Occasional cases of subacute constrictive pericarditis will respond to medical management, including adrenal steroids, if treated before the stage of pericardial fibrosis.[30] Thus, in symptomatic patients, the treatment is ordinarily that of pericardial resection. Because the mortality rate of this operation tends to be higher in patients with advanced symptoms, one should not wait until the patient is totally incapacitated. On the other hand, patients with few or no symptoms may remain stable for years, and one can safely defer operation in those who are in functional class I or early class II of the NYHA. Patients who are in late and progressive class II should be recommended for pericardial resection. Tuna and Danielson[18] reported an operative mortality rate of 1% for patients in functional classes I and II, 10% for those in class III, and 46% for those in class IV. The overall mortality rate for 313 patients operated upon at Mayo Clinic since 1936 was 14%.

The surgical mortality rate was 16% in 118 cases reported by Bashi *et al.*,[22] and 11% in 52 patients operated upon in the last 12 years of this series. The operative mortality was 12% in Cameron *et al.'s* report of 95 patients undergoing surgery at Stanford University.[17] In a review by Siefert *et al.*,[132] 80–90% of hospital survivors achieved NYHA class I or II functional status following pericardial resection.

### Occult Constrictive Pericardial Disease

Bush and associates[133] described 19 patients with occult constrictive pericarditis. These patients had normal or nearly normal right atrial pressures (1–8 mmHg), which rose abnormally and equilibrated with pulmonary wedge or LV end-diastolic pressures after infusion of one liter of normal saline within 6–8 min. A dip and plateau pattern in the RV pressure trace and a lack of respiratory variation in right atrial pressure also appeared after infusion. Eleven patients were operated upon and had improvement in fatigue and dyspnea; all had pericardial adhesions at operation. The place of this test in diagnosing and treating pericardial disease is uncertain. It is difficult to explain all the symptoms in these patients entirely on the basis of abnormal hemodynamics. Also, there may be some risk in saline infusion at this rate, and pulmonary wedge pressure should be carefully monitored if this test is carried out.

## References

1. White PD: Chronic constrictive pericarditis (Pick's disease). Treated by pericardial resection. Lancet 2, 539–548; 597–603 (1935)

2. Lower R: *Tractatus de corde.* J. Allestry, London, 1669. Cited by Shabetai R: In *The Pericardium.* Grune and Stratton (1981) 283
3. Connolly DC, Mann RJ, Cominic J: Corrigan (1802–1880) and his description of the pericardial knock. *Mayo Clin Proc* 55, 771–773 (1980)
4. Kussmaul A: Ueber schwielige Mediastino-Pericarditis und den parodoxen Puls. *Berl Klin Wochenschr* 10, 433–435 (1873)
5. Pick F: Ueber chronische, unter dem Bilde der Lebercirrhose verlaufende Pericarditis (pericarditische Pseudolebercirrhose). *Zeitschr Klin Med* 29, 385–410 (1896)
6. Churchill ED: Decortication of the heart (Delorme) for adhesive pericarditis. *Arch Surg* 19, 1457–1467 (1929)
7. Bloomfield RA, Lauson HD, Cournand A, Breed ES, Richards DW: Recording of right heart pressures in normal subjects and in patients with chronic pulmonary disease and various types of cardio-circulatory disease. *J Clin Invest* 25, 639–664 (1946)
8. Hansen AT, Eskildsen P, Götzsche H: Pressure curves from the right auricle and the right ventricle in chronic constrictive pericarditis. *Circulation* 3, 881–888 (1961)
9. Hetzel PS, Wood EH, Burchell HB: Pressure pulses in the right side of the heart in a case of amyloid disease and in a case of idiopathic heart failure. *Pros Staff Meet Mayo Clin* 28, 107–112 (1953)
10. Hancock EW: Subacute effusive constrictive pericarditis. *Circulation* 43, 183–192 (1971)
11. Kendall ME, Rhodes GR, Wolfe W: Cardiac constriction following aorta-to-coronary bypass surgery. *J Thorac Cardiovasc Surg* 64, 142–153 (1972)
12. Isner JM, Carter BL, Bankoff MS, Konstam MA, Salem DN: Computed tomography in the diagnosis of pericardial heart disease. *Ann Intern Med* 97, 473–479 (1982)
13. Soulen RL, Stark DD, Higgins CB: Magnetic resonance imaging of constrictive pericardial disease. *Am J Cardiol* 55, 480–484 (1985)
14. Robertson R, Arnold CR: Constrictive pericarditis with particular reference to etiology. *Circulation* 26, 525–529 (1962)
15. Wychulis AR, Connolly DC, McGoon DC: Surgical treatment of pericarditis. *J Thorac Cardiovasc Surg* 62, 608–617 (1971)
16. Hirschmann JV: Pericardial constriction. *Am Heart J* 96, 110–122 (1978)
17. Cameron J, Oesterle SN, Baldwin JC, Hancock EW: The etiologic spectrum of constrictive pericarditis. *Am Heart J* 113, 354–360 (1987)
18. Tuna IC, Danielson GK: Surgical management of pericardial diseases. In *Diseases of the Pericardium* (Ed. Shabetai R). W.B. Saunders, Philadelphia, Cardiology Clinics (1990) 8:4 683–696
19. Cacoub P, Wechsler B, Chapelon C, Gandjbakch I, Blétry O, Piette JC, Cabrol C, Godeau P: Chronic constrictive pericarditis, 27 cases. *Presse Med* 20, 2185–2190 (1991)
20. Paul O, Castleman B, White PD: Chronic constrictive pericarditis; a study of 53 cases. *Am J Med Sci* 216, 361–377 (1948)
21. Wood P: Chronic constrictive pericarditis. *Am J Cardiol* 7, 48–61 (1961)
22. Bashi VV, John S, Ravikumas E, Jairaj J, Shyamsunder K, Krishnaswami S : Early and late results of pericardiectomy in 118 cases of constrictive pericarditis. *Thorax* 43, 637–641 (1988)
23. Goldstein S, Yu P: Constrictive pericarditis after blunt chest trauma. *Am Heart J* 69, 544–549 (1965)
24. Schwartz DJ, Thanavaro S, Kleiger RE, Krone RJ, Connors SP, Oliver GC: Epicardial pacemaker complicated by cardiac tamponade and constrictive pericarditis. *Chest* 76, 226–227 (1979)
25. Cimino JJ, Kogan AD: Constrictive pericarditis after cardiac surgery: Report of 3 cases and review of the literature. *Am Heart J* 118, 1292–1301 (1989)
26. Killian DM, Furiasse JG, Scanlon PF, Loeb HS, Sullivan HJ: Constrictive pericarditis after cardiac surgery. *Am Heart J* 118, 563–568 (1989)
27. Keogh BE, Oakley CM, Taylor KM: Chronic constrictive pericarditis caused by self-mutilation with sewing needles. A case report and review of published reports. *Br Heart J* 59, 77–80 (1988)
28. Applefeld MM, Cole JF, Pollock SH, Sutton FJ, Slawson RG, Singleton RJ, Wiernik PH: The late appearance of chronic pericardial disease in patients treated by radiotherapy for Hodgkin's disease. *Ann Intern Med* 94, 338–341 (1981)
29. Coltart RS, Roberts JT, Thom CH, Petch MC: Severe constrictive pericarditis after single 16 MeV anterior mantle irradiation for Hodgkin's disease. *Lancet* 1, 488–489 (1985)
30. Miller JI, Mansour KA, Hatcher CR Jr: Pericardiectomy: Current indications, concepts and results in a university center. *Ann Thorac Surg* 34, 40–45 (1982)
31. Lindsay J Jr, Crawley IS, Callaway GM Jr: Chronic constrictive pericarditis following uremic hemopericardium. *Am Heart J* 79, 390–395 (1970)
32. Wolfe SA, Bailey GF, Collins JJ Jr: Constrictive pericarditis following uremic effusion. *J Thorac Cardiovasc Surg* 63, 540–544 (1972)
33. Weiss SW, Tau FL, Hutchins GM: Constrictive uremic pericarditis following hemodialysis for acute renal failure. *Johns Hopkins Med J* 132, 301–305 (1973)
34. Jenzer HR, Graedel E, Blumberg A: Diagnosis and successful surgical treatment of chronic constrictive uremic pericarditis. *Br Heart J* 38, 304–306 (1976)
35. Roberts Wl, Spray TL: Pericardial heart disease: A study of its causes, consequences and morphologic features. In *Cardiovascular Clinics*, Vol 7, No 3 (Ed. Spodick DH). F.A. Davis, Philadelphia (1976) 11–68
36. McCaughan BC, Schoff HV, Piehler JM, Danielson GK, Otszulak TA, Puga FJ, Pluth JR, Connolly DC, McGoon DC: Early and late results of pericardiectomy for constrictive pericarditis. *J Thorac Cardiovasc Surg* 89, 340–350 (1985)
37. Liss JP, Bachmann WT: Rheumatoid constrictive pericarditis treated by pericardiectomy. Report of a case and review of the literature. *Arth Rheum* 13, 869–876 (1970)
38. Arthur A, Oskvig R, Basta LL: Calcific rheumatoid constrictive pericarditis with cardiac failure treated by pericardiectomy. *Chest* 64, 769–771 (1973)
39. Thadani U, Iveson JMI, Wright V: Cardiac tamponade, constrictive pericarditis and pericardial resection in rheumatoid arthritis. *Medicine* (Baltimore) 54, 261–270 (1975)
40. John JT Jr, Hough A, Sergent JS: Pericardial disease in rheumatoid arthritis. *Am J Med* 66, 385–390 (1979)
41. Wolf RE, King JW, Brown TA: Antimyosin antibodies and constrictive pericarditis in lupus erythematosus. *J Rheumatol* 15, 1284–1287 (1988)
42. Sunder SK, Shah A: Constrictive pericarditis in procainamide-induced lupus erythematosus syndrome. *Am J Cardiol* 36, 960–962 (1975)
43. Richards FM, Fulkerson WJ Jr: Constrictive pericarditis due to hydralazine-induced lupus erythematosus. *Am J Med* 88, 56N–59N (1990)
44. Fowler NO, Manitsas GT: Infectious pericarditis. *Prog Cardiovasc Dis* 16, 323–336 (1973)
45. Spodick DH: Infective pericarditis. Etiologic and clinical spectra. In *Pericardial Disease* (Eds. Reddy PS, Leon DF, Shaver JA). Raven Press, New York (1982) 307–312
46. Fowler NO: Constrictive pericarditis: New concepts. *Am J Cardiol* 50, 1014–1017 (1982)
47. Fowler NO: Tuberculous pericarditis. *J Am Med Assoc* 266, 99–103 (1991)
48. Howard EJ, Maier HC: Constrictive pericarditis following acute Coxsackie viral pericarditis. *Am Heart J* 75, 247–250 (1968)
49. Matthews JD, Cameron SJ, George M: Constrictive pericarditis following Coxsackie virus. *Thorax* 25, 624–626 (1970)
50. Kilburn CD, McKinsey DS: Recurrent massive pleural effusion due to pleural, pericardial, and epicardial fibrosis in histoplasmosis. *Chest* 100, 1715–1719 (1991)

51. Picardi JL, Kauffman CA, Schwartz J, Holmes JC, Phair JP, Fowler NO: Pericarditis caused by histoplasma capsulatum. *Am J Cardiol* 37, 82–88
52. Dunn EJ, Ulicny KS, Wright CB, Gottesman L: Surgical implications of sclerosing mediastinitis. *Chest* 97, 338–346 (1990)
53. Nelson DP, Rensimer ER, Raffin TA: *Legionella pneumophila* pericarditis without pneumonia *Arch Intern Med* 145, 926 (1985)
54. Nichols DA, Peter RH: Constrictive pericarditis as a late complication of meningococcal pericarditis. *Am J Cardiol* 55, 1442–1443 (1985)
55. Yanase O, Motomiya T, Watanabe K, Tokuyasu Y, Sakurada H, Tejima T, Hiyoshi Y, Sugiura M, Yabata Y, Kitazumi H: Lassa fever associated with effusive-constrictive pericarditis and bilateral atrioventricular annular constriction: A case report. *J Cardiol* 19, 1147–1156 (1989)
56. Freychet L, Duboc D, Guérin F, Chomette G: Péricardite constrictive et maladie de Whipple. *Arch Mal Coeur* 83, 1729–1732 (1990)
57. O'Sullivan RA, Armstrong JG, Rivers RT, Mitchell CA: Pulmonary actinomycosis complicated by effusive-constrictive pericarditis. *Aust NZ J Med* 21, 879–880 (1991)
58. Kessler R, Follis F, Daube D, Wernly J: Constrictive pericarditis from nocardia asteroides infection. *Ann Thorac Surg* 52, 861–862 (1991)
59. D'Cruz IA, Pallas CW, Heck A: Echocardiographic diagnosis of effusive-constrictive pericarditis due to staphylococcal pericarditis after cardiac surgery. *South Med J* 84, 1375–1377 (1991)
60. Doig JC, Hilton CJ, Reid DS: Salmonella: A rare cause of subacute effusive-constrictive pericarditis. *Br Heart J* 65, 296–297 (1991)
61. Hirata K, Asato H, Maeshiro M: A case of effusive-constrictive pericarditis caused by streptococcus milleri. *Jpn Circ J* 55, 154–158 (1991)
62. Brockington GM, Zebede J, Pandian NG: Constrictive pericarditis. In *Diseases of the Pericardium* (Ed. Shabetai R). W.B. Saunders, Philadelphia, Cardiology Clinics Vol. 8, No. 4, (1990) 645–661
63. Llewellyn MJ, Atkinson MW, Fabri B: Pericardial constriction caused by primary mesothelioma. *Br Heart J* 54–57 (1987)
64. Beaufils Ph, Bardet J, Tembine J, Masquet Ch, Sebastian Ph, Zelasko Y: Syndrome de Dressler: Péricardite constrictive post-infarctus opérée avec succèss. *Arch Mal Coeur* 68, 651–656 (1975)
65. Karim AH, Salomon J: Constrictive pericarditis after myocardial infarction. Sequela of anticoagulant-induced hemopericardium. *Am J Med* 79, 389–390 (1985)
66. Ramsdale DR, Epstein EJ, Coulshed N: Constrictive pericarditis after myocardial infarction. *Br Heart J* 56, 476–478 (1986)
67. Cheung PK, Myers ML, Arnold JMO: Early constrictive pericarditis and anemia after Dressler's syndrome and inferior wall myocardial infarction. *Br Heart J* 65, 360–362 (1991)
68. Albers WH, Hugenholtz PG, Nadas AS: Constrictive pericarditis and atrial septal defect, secundum type. With special reference to left ventricular volumes and related hemodynamic findings. *Am J Cardiol* 23, 850–857 (1969)
69. Harada J, Seki I, Okuni M: Constrictive pericarditis with atrial septal defect in children. *Jpn Heart J* 19, 531–543 (1978)
70. Kumar S, Szatkowski J, Thomas W: Atrial septal defect complicated by constrictive pericarditis. *Chest* 60, 101–105 (1971)
71. Kouvaras G, Goudevenos J, Chronopoulos G, Sofranos G, Reed P, Adams P, Cokkinos DV: Atrial septal defect and constrictive pericarditis. An unusual combination. *Acta Cardiol* 44, 341–349 (1989)
72. Chou TM, Jue J, Merrick SH, Schiller NB, Foster E: Effusive constrictive pericarditis and atrial septal defect. *Am Heart J* 125, 1193–1195 (1993)
73. Just H, Mattingly TW: Interatrial septal defect and pericardial disease. Coincidence or casual relationship? *Am Heart J* 76, 157–167 (1968)
74. Tuuteri L, Perheentupa J, Rapola J: The cardiopathy of mulibrey nanism, a new inherited syndrome. *Chest* 65, 628–631 (1975)
75. Voorhess ML, Husson GS, Blackman MS: Growth failure with pericardial constriction. The syndrome of mulibrey nanism. *Am J Dis Child* 130, 1146–1148 (1976)
76. Garrett J, O'Neill M, Blake S: Constrictive pericarditis associated with sarcoidosis. *Am Heart J* 107, 394 (1984)
77. Morishita Y, Taira A, Furoi A, Arima S, Tanaka H: Constrictive pericarditis secondary to primary chylopericardium. *Am Heart J* 109, 373–375 (1985)
78. Tamir R, Pick AJ, Theodor E: Constrictive pericarditis complicating dermatomyositis. *Ann Rheum Dis* 47, 961–963 (1988)
79. Fischbein L, Namade M, Sachs RN, Robineau M, Lanfranchi J: Chronic constrictive pericarditis associated with asbestosis. *Chest*, 94, 646–647 (1988)
80. Davies D, Andrews MIJ, Jones JSP: Asbestos-induced pericardial effusion and constrictive pericarditis. *Thorax* 46, 429–432 (1991)
81. Daubert JP, Gaede J, Cohen HJ: A fatal case of constrictive pericarditis due to a marked selective pericardial accumulation of amyloid. *Am J Med* 94, 335–340 (1993)
82. Kassanoff AH, Levin CB, Wyndham CRC, Mills LJ: Implantable cardioverter defibrillator infection causing constrictive pericarditis. *Chest* 102, 960–963 (1992)
83. Muran MK, Ahmed AH, Parsons FM, Anderson CK: Constrictive pericarditis due to methysergide therapy. *S Afr Med J* 50, 1595–1597 (1976)
84. Orlando RC, Moyer P, Barnett TB: Methysergide therapy and constrictive pericarditis. *Ann Intern Med* 88, 213–214 (1978)
85. Sagristà-Sauleda J, Permanyer-Miralda G, Candell-Riera J, Angel J, Soler-Soler J: Transient cardiac constriction: An unrecognized pattern in effusive acute idiopathic pericarditis. *Am J Cardiol* 59, 961–966 (1987)
86. Gimlette TMD: Constrictive pericarditis. *Br Heart J* 21, 9–16 (1959)
87. Wise DE, Conti CR: Constrictive pericarditis. In *Pericardial Diseases* (Ed. Spodick DH). Cardiovascular Clinics Vol 7, No 3. F.A. Davis, Philadelphia (1976) 197–210
88. Schiavone WA: The changing etiology of constrictive pericarditis in a large referral center. *Am J Cardiol* 58, 373–375 (1986)
89. Oh JK, Hatle LK, Seward JB, Danielson GK, Schaff HV, Reeder GS, Tajik AJ: Diagnostic role of Doppler echocardiography in constrictive pericarditis. *J Am Coll Cardiol* 23, 154–162 (1994)
90. Meyer TE, Sareli P, Marcus RH, Pocock W, Berk MR, McGregor M: Mechanism underlying Kussmaul's sign in chronic constrictive pericarditis. *Am J Cardiol* 64, 1069–1072 (1989)
91. Dalvi B: Kussmaul's sign: An artifact? (lett). *Lancet* 1, 1337 (1989)
92. Byrd BF III, Linden RW: Superior vena cava Doppler flow velocity patterns in pericardial disease. *Am J Cardiol* 65, 1464–1470 (1990)
93. Schrire V, Gotsman MS, Beck W: Unusual diastolic murmurs in constrictive pericarditis and constrictive endocarditis. *Am Heart J* 76, 4–12 (1968)
94. Fowler NO, Bove KE, Dunbar S, Meyer R: Fatigue, dyspnea, and abdominal swelling in a 13-year-old boy. *Am Heart J* 96, 533–542 (1978)
95. Pulvaneswary M, *et al.*: Constrictive pericarditis. Clinical, hemodynamic, and radiologic correlation. *Australas Radiol* 26, 53 (1982) Cited in *Heart Disease. A Textbook of Cardiovascular Medicine* (Ed. Braunwald E). W.B. Saunders, Philadelphia 4th ed. (1992) 1485
96. Surawicz B, Lasseter KC: Electrocardiogram in pericarditis. *Am J Cardiol* 26, 471–474 (1970)
97. Dalton JC, Pearson RJ Jr, White PD: Constrictive pericarditis: A review and long-term follow-up of 78 cases. *Ann Intern Med* 45, 445–458 (1952)
98. Levine HD: Myocardial fibrosis in constrictive pericarditis: Electrocardiographic and pathologic observations. *Circulation* 48, 1268–1281 (1973)

99. Chesler E, Mitha AS, Matisonn RE: The ECG of constrictive pericarditis. Pattern resembling right ventricular hypertrophy. *Am Heart J* 95, 420–424 (1976)
100. Chesler E, Mitha AS, Matisonn RE, Rogers MNA: Subpulmonic stenosis as a result of noncalcific constrictive pericarditis. *Chest* 69, 425–427 (1976)
101. Fukuda K, Nakamura Y, Ogawa S, Yokoyama H, Handa S: Constrictive pericarditis with electrocardiographic evidence of right ventricular hypertrophy. *Chest* 96, 691–693 (1989)
102. Engel PJ, Fowler NO, Tei C, Shah PM, Driedger H, Shabetai R, Harbin AD, Franch RH: M-mode echocardiography in constrictive pericarditis. *J Am Coll Cardiol* 6, 471–474 (1985)
103. Janos GG, Arjunan K, Meyer RA, Engel P, Kaplan S: Differentiation of constrictive pericarditis and restrictive cardiomyopathy using digitized echocardiography. *J Am Coll Cardiol* 1, 541–549 (1983)
104. D'Cruz IA, Dick A, Gross CM, Hand CR, Lalmalani GG: Abnormal left ventricular–left atrial posterior wall contour: A new two-dimensional echocardiographic sign in constrictive pericarditis. *Am Heart J* 118, 128–132 (1989)
105. Vilacosta I, Alberto J, Calvar SR, Prieto EI, Recio MG, Elbal LM: Transesophageal echocardiographic features of the atrial septum in constrictive pericarditis. *Am J Cardiol* 68, 271–273 (1991)
106. Chandraratna PAN: Echocardiography and Doppler ultrasound in the evaluation of pericardial disease. *Circulation* 84(suppl I) I, 303–310 (1991)
107. Schiavone WA, Calafiore PA, Currie PJ, Lytle BW: Doppler echocardiographic demonstration of pulmonary venous flow velocity in three patients with constrictive pericarditis before and after pericardiectomy. *Am J Cardiol* 63, 145–147 (1989)
108. Schiavone WA, Calafiore PA, Salcedo EE: Transesophageal Doppler echocardiographic demonstration of pulmonary venous flow velocity in restrictive cardiomyopathy and constrictive pericarditis. *Am J Cardiol* 63, 1286–1288 (1989)
109. Klein AL, Cohen GI, Pietrolungo JF, White RD, Bailey A, Pearce GL, Stewart WJ, Salcedo EE: Differentiation of constrictive pericarditis from restrictive cardiomyopathy by Doppler transesophageal echocardiographic measurements of respiratory variations in pulmonary venous flow. *J Am Coll Cardiol* 22, 1935–1943 (1993)
110. Von Bibra H, Schober K, Jenni R, Busch R, Sebening H, Blomer H: Diagnosis of constrictive pericarditis by pulsed Doppler echocardiography of the hepatic vein. *Am J Cardiol* 63, 483–488 (1989)
111. Hatle LK, Appleton CP, Popp RL: Differentiation of constrictive pericarditis and restrictive cardiomyopathy by Doppler echocardiography. *Circulation* 79, 357–370 (1989)
112. Mancuso L, D'Agostino A, Pitrolo F, Marchi S, Carmina MG, Celona G, Raspanti G, Figlia A: Constrictive pericarditis vs restrictive cardiomyopathy: The role of Doppler echocardiography in differential diagnosis. *Int J Cardiol* 31, 319–327 (1991)
113. Shabetai R, Fowler NO, Guntheroth WG: The hemodynamics of cardiac tamponade and constrictive pericarditis. *Am J Cardiol* 26, 480–489
114. Yu PNG, Lovejoy FW Jr, Joos HA, Nye RE Jr, Mahoney EB: Right auricular and ventricular pressure patterns in constrictive pericarditis. *Circulation* 7, 102–107 (1953)
115. Harvey RJ, Ferrer MI, Cathcart R, Richards DW, Cournand A: Mechanical and myocardial factors in chronic constrictive pericarditis. *Circulation* 8, 695–707 (1953)
116. Reddy PS: Hemodynamics of constrictive pericarditis. In *Pericardial Disease* (Eds. Reddy PS, Leon DF, Shaver JA). Raven Press, New York (1982) 275–297
117. Tyberg TI, Goodyer AVN, Hurst VW III, Alexander J, Langou RA: Left ventricular filling in differentiating restrictive amyloid cardiomyopathy and constrictive pericarditis. *Am J Cardiol* 47, 791–796 (1981)
118. Vallance PJT, Gray HH, Oldershaw PJ: Diagnostic features of localized pericardial constriction. *Int J Cardiol* 20, 416–419 (1988)
119. Nigri A, Mangieri E, Martuscelli E, Ruvolo G, Papalia U, Toscano M, Marino B, Reale A: Pulmonary trunk stenosis due to constriction by a pericardial band. *Am Heart J* 114, 448–450 (1987)
120. Nishimura RA, Kazmier FJ, Smith HC, Danielson GK: Right ventricular outflow obstruction caused by constrictive pericardial disease. *Am J Cardiol* 55, 1447–1448 (1985)
121. Wolozin MW, Ortola FV, Spodick DH, Seifter JL: Release of atrial natriuretic factor after pericardiectomy for chronic constructive pericarditis. *Am J Cardiol* 62, 1323–1325 (1988)
122. Svanegaard J, Thayssen P, Arendrup HK: Atrial natriuretic peptide and hemodynamic response to pericardiectomy for chronic constrictive pericarditis. *Am J Cardiol* 66, 117–120 (1990)
123. Anand IS, Ferrari R, Kalra GS, Wahi PL, Poole-Wilson PA, Harris PC: Pathogenesis of edema in constrictive pericarditis. Studies of body water and sodium, renal function, hemodynamics, and plasma hormones before and after pericardiectomy. *Circulation* 83, 1880–1887 (1991)
124. Spodick DH, Kumar S: Subacute constrictive pericarditis with tamponade. *Dis Chest* 54, 62–66 (1968)
125. Gerson MC, Colthar MA, Fowler NO: Differentiation of constrictive pericarditis and restrictive cardiomyopathy by radionuclide ventriculography. *Am Heart J* 118, 114–120 (1989)
126. Aroney CN, Ruddy TD, Dighero H, Fifer M, Boucher CA, Palacios IF: Differentiation of restrictive cardiomyopathy from pericardial constriction: Assessment of diastolic function by radionuclide angiography. *J Am Coll Cardiol* 13, 1007–1014 (1989)
127. Hoit BD: Imaging the pericardium. In *Diseases of the Pericardium* (Ed. Shabetai R). Cardiology Clinics Vol. 8, No. 4. W.B. Saunders, Philadelphia (1990) 587–600
128. Masui T, Fink S, Higgins CB: Constrictive pericarditis and restrictive cardiomyopathy: Evaluation with MR imaging. *Radiology* 182, 369–373 (1992)
129. Oren RM, Groer-McKay M, Stanford W, Weiss RM: Accurate preoperative diagnosis of pericardial constriction using cine computed tomography. *J Am Coll Cardiol* 22, 832–838 (1993)
130. Vaitkus PT, Kussmaul WG: Constrictive pericarditis versus restrictive cardiomyopathy: A reappraisal and update of diagnostic criteria. *Am Heart J* 122, 1431–1441 (1991)
131. Schoenfeld MH, Supple EW, Dec GWD Jr, Fallon JT, Palacios IF: Restrictive cardiomyopathy versus constrictive pericarditis: Role of endomyocardial biopsy in avoiding unnecessary thoracotomy. *Circulation* 75, 1012–1017 (1987)
132. Seifert FC, Miller DC, Oesterle SN, Oyer PE, Stinson EB, Shumway NE: Surgical treatment of constrictive pericarditis; analysis of outcome and diagnostic error. *Circulation* 72(suppl II), 264–273 (1985)
133. Bush CA, Stang JM, Wooley CF, Kilman JW: Occult constrictive pericardial disease: Diagnosis by rapid volume expansion and correction by pericardiectomy. *Circulation* 56, 924–930 (1977)