Constrictive Pericarditis

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Constrictive pericarditis (CP) is characterized by scarring and loss of elasticity of the pericardium, resulting in external impedance of cardiac filling. In the developed world, CP is most frequently encountered as a consequence of previous cardiac surgery, thoracic irradiation, viral or idiopathic causes. Tuberculosis still remains a common cause of CP in the developing world, immigrants from underdeveloped nations, and immunosuppressed patients. Clinical signs and symptoms of right heart failure coupled with risk factors for pericardial disease should raise suspicion for CP. Echocardiographic evaluation and often cardiac catheterization are essential components of accurate diagnosis of CP. Enhanced interventricular dependence, with respiratory variation in the ventricular pressures, and ventricular discordance are the pathophysiologic hallmarks of CP. Imaging findings such as increased pericardial thickness or pericardial calcification on computed tomography can be supportive, but are not necessary for the diagnosis of CP. Pericardiectomy remains the most effective therapy for symptomatic CP. (Circ J 2008; 72: 1555–1562)

Key Words: Constrictive pericarditis; Diastole; Heart failure; Pericardiectomy

The pericardium is a fibroserous membrane composed of visceral and parietal pericardium. The pericardium envelopes the cardiac chambers and under physiological conditions exerts several important functions, including limitation of intrathoracic cardiac motion and acute dilatation, suction filling, lubricant effects that minimize friction between the cardiac chambers and surrounding structures, and balancing right and left ventricular (LV) output through diastolic and systolic interactions. Constrictive pericarditis (CP) occurs when a thickened, fibrotic, and frequently calcified shell-like pericardium, from whatever cause, impedes normal diastolic filling. It is usually a long-term consequence of either acute or chronic forms of pericarditis. Over the past two decades, the evolution of noninvasive imaging techniques has facilitated early clinical recognition of CP and its differentiation from restrictive cardiomyopathy (RCM).

History of CP

Avenzoar (1113–1162) from medieval Spain described serofibrous pericarditis, and Lancisi (1654–1720) described the clinical significance of pericardial adhesions. In 1669, Richard Lower from London gave a description of chronic CP in a 30-year-old woman. Chevers in 1842 discussed compression of heart chambers by pericardial adhesions. Kussmaul in 1873 described pulsatia paradoxus, and Pick (1867–1926) reported pseudocirrhosis of the liver secondary to adhesive pericarditis. Brauer reported resection of ribs and costal cartilages for surgical management of CP, and Weill in 1895, and later Delorme in 1898 introduced the concept of cardiac decortication. The modern description of the disease was given by White in 1935. In his historic paper, White described the major features of the disease and reported the results of the first 12 patients who underwent operation at the Massachusetts General Hospital.

CP is caused by fibrosis and thickening of the pericardium, processes that predominantly inhibit the diastolic filling of the heart. Recognition of CP is clinically important, but often difficult. Radiographic evidence may provide a clue, but is not diagnostic and may be absent. Because pericardiectomy can result in complete relief of symptoms in many patients, accurate diagnosis is important. Before the advent of 2-dimensional (D) and Doppler echocardiography, differentiation of CP from other causes of right-sided heart failure (ie, pulmonary hypertension, pulmonary embolism, right ventricular (RV) infarction, LV systolic dysfunction etc) was cumbersome. Now those conditions often can be easily identified or excluded by echocardiography, so the main challenge today remains that of determining whether restrictive filling is caused by CP, RCM or both. In patients with disease features suggesting both pericardial and endocardial involvement, determination of the relative contribution of each can be extremely difficult. The important distinction of CP from RCM allows a select group of patients with predominantly constrictive physiology to potentially benefit from surgery. In contrast to the past when tuberculosis was the predominant etiology of CP, in the current era radiation-induced restrictive physiology is an important condition that can manifest as involvement of both the pericardium and myocardium.

Etiology of CP

Though CP can occur from a number of causes, significant changes in its etiologic spectrum have occurred in recent years. Before the 1960s, tuberculosis was a common cause worldwide but its incidence has since declined and is now rare in Western countries. However, tuberculosis CP is still an important cause in developing countries. Radiation-induced CP was recognized in the 1960s which prompted introduction of techniques to reduce the radiation dose to cardiac structures in patients requiring radiotherapy. Despite...
these protective measures, post-radiotherapy injury continues
to be an important cause of CP. In the 1970s, reports
highlighted the occurrence of CP after open-heart surgery.
At present, idiopathic CP leads the list of causes in the
Western world, followed by cardiac surgery and mediastinal
irradiation.

Pathophysiology of CP

The pathophysiologic manifestations of CP are mostly
because of loss of pericardial compliance, which causes ex-
ternal impedance of cardiac filling. Normally, the inspira-	ory decrease in the intrathoracic pressure is transmitted to
all cardiac chambers and to the pulmonary veins. The pres-
sure gradient between the pulmonary veins and the left-
sided structures thus remains unchanged. In patients with
CP, however, the encasing pericardium effectively isolates
the cardiac chambers from changes in intrathoracic pressure.
The pressure gradient between the pulmonary veins and the
left chambers thus decreases with inspiration, resulting in an
inspiratory reduction in the velocity of diastolic flow in the
pulmonary veins and hence a reduction in left-sided filling.
Also, in CP the ventricles are pathologically coupled and
function within a rigid, non-compliant pericardium. The
pericardial restraint limits the total cardiac volume and,
consequently, an increase in filling on one side of the heart
impedes contralateral filling through septal interaction, thus
making the two ventricles interdependent. The total volume
of blood entering the heart changes little during the respira-
tory cycle.

Two factors, the rigid pericardium not allowing transmis-
sion of changes in intrathoracic pressure with respiration to
the cardiac chambers and the exaggerated interventricular
dependence, result in dynamic respiratory changes. The
inspiratory reduction in LV filling is associated with a
simultaneous increase in RV diastolic filling and a septal
shift to the left. An opposite physiologic effect on the fill-
ing gradients and septal shift is seen in expiration. Expira-
tion results in increased intrathoracic pressure, which results
in increased pulmonary venous pressure. This increase,
however, is not transmitted to the cardiac chambers. The
gradient between the pulmonary veins and the left cham-
bers increases with expiration with an attendant increase in
diastolic flow, which is associated with a simultaneous de-
crease in right-sided filling. These changes exist in contrast
to cardiac tamponade, in which the diastolic pressures are
elevated, but changes in intrathoracic pressure with respira-
tion are transmitted to the cardiac chambers.

The fibrotic encasement of the heart in CP limits diastolic
filling of all cardiac chambers, resulting in elevation of the
diastolic pressures. The right atrial (RA), RV, pulmonary
capillary wedge and LV filling pressures are elevated and
equalized, reflecting the common constraining effects of the
fibrotic pericardium. In cardiac tamponade, a tense effusion
imposes a pandiastolic resistance to ventricular filling,
whereas in CP early diastolic filling is resistance-free. Ven-
tricular filling is very rapid in early diastole because of
elevated atrial pressures. Up to 75% of filling may occur in
the first 25% of diastole in CP. This rapid filling abruptly
decreases by mid-diastole because of the limitation of the
non-compliant pericardium. This hemodynamic pattern is
reflected in the jugular venous pulse (JVP) and RA pres-
ture waveforms as elevated mean filling pressure with prominent a wave, a sharp y descent, reflecting early dias-
tolic free rapid RV filling, as well as a preserved x descent
because of accelerated atrial relaxation. This pattern with
prominent x and y descents may in some patients result in an
“M-shaped” pattern. The RV waveform is similarly distinc-
tive, with a dip and plateau, or “square-root” pattern reflect-
ing the rapid ventricular relaxation and a sharp increase in
filling pressure as the expanding ventricle meets the con-
straints of the pericardium. Analysis of LV mechanics in CP
has shown that LV deformation and early diastolic recoil
are constrained in the circumferential direction, reflecting a

Table 1  Hemodynamic and Echocardiographic Features of Constrictive Pericarditis and Restrictive Cardiomyopathy

<table>
<thead>
<tr>
<th>Feature</th>
<th>Constrictive pericarditis</th>
<th>Restrictive cardiomyopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paradoxical pulse</td>
<td>Present in 1/3 of cases</td>
<td>Absent</td>
</tr>
<tr>
<td>Pericardial knock</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Prominent y descent in JVP</td>
<td>Present</td>
<td>Variable</td>
</tr>
<tr>
<td>Right- and left-sided filling pressures</td>
<td>Equalized within 5 mmHg</td>
<td>Left-sided pressures at least 3–5 mmHg more than right</td>
</tr>
<tr>
<td>Filling pressures &gt;25 mmHg</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>RVSP &gt;50 mmHg</td>
<td>No</td>
<td>Common</td>
</tr>
<tr>
<td>“Square-root” sign</td>
<td>Present</td>
<td>Variable</td>
</tr>
<tr>
<td>RV/EDP/RVSP ≥0.3</td>
<td>Right and left ventricular peak systolic pressure variations are out-of-phase</td>
<td>Right and left ventricular peak systolic pressure variations are in-phase</td>
</tr>
<tr>
<td>Discordant respiratory variation of ventricular peak systolic pressures</td>
<td>Usually increased</td>
<td>Normal</td>
</tr>
<tr>
<td>Pericardial thickness</td>
<td>Mild enlargement, usually of the left atrium</td>
<td>Bi-atrial enlargement, usually severe</td>
</tr>
<tr>
<td>Atrial size</td>
<td>Normal</td>
<td>Usually increased</td>
</tr>
<tr>
<td>Ventricular wall thickness</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Septal bounce</td>
<td>Usually absent or mild</td>
<td>Present</td>
</tr>
<tr>
<td>Mitral or tricuspid regurgitation</td>
<td>Inspiratory E less than expiratory</td>
<td>Inspiratory E greater than expiratory</td>
</tr>
<tr>
<td>Respiratory variation in left-right pressures or flow</td>
<td>Inspiratory E greater than expiratory</td>
<td>Mild respiratory variation in E velocity (≤15%)</td>
</tr>
</tbody>
</table>
| Mitral inflow                        | Decreased diastolic forward flow with expiration; marked diastolic flow reversal, which increases with expiration compared to inspiration | Systolic forward flow less than diastolic forward flow;
diastolic flow reversal in the hepatic vein is more prominent with inspiration |
| Tricuspid inflow                     | Inspiratory E less than expiratory                             | No respiratory variation of E velocity; increased E/A ratio ≥2.0; DT <160 ms |
| Pulsed wave Doppler of hepatic vein  | Increased E/A ratio ≥2.0; DT <160 ms                           | Increased E/A ratio ≥2.0; DT <160 ms                    |

JVP, jugular venous pulse; RVSP, right ventricular systolic pressure; RV/EDP, right ventricular end-diastolic pressure; E, early rapid filling wave; A, filling wave because of atrial systole; DT, deceleration time.

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The clinical, hemodynamic and echocardiographic findings should all be assessed before arriving at the diagnosis of CP in a patient with right-sided heart failure. Symptoms in CP are typically insidious in onset and may develop weeks to decades after an episode of pericarditis or chest trauma. Patients typically present with signs of systemic venous congestion with ascites, hepatomegaly and pedal edema. Elevated JVP with a rapid and deep y descent, with preserved x descent, is characteristic of CP. The inspiratory increase in venous return may not be accommodated in the right atrium because of high RA pressure, thus resulting in increased JVP with inspiration. This finding, termed Kussmaul’s sign, is not specific for CP and may be observed in any condition with elevated right-sided pressures, such as RCM, tricuspid stenosis etc.

The presence of pericardial calcification on a plain radiograph strongly suggests CP in patients with heart failure. However, calcific CP is less common in the United States because the incidence of tuberculosis has decreased. In a retrospective cohort study of a consecutive series of 135 patients with CP confirmed surgically, pericardial calcification was seen in only 36 patients (27%). The cause of constrictive pericardial disease was indeterminate in 67% of patients with pericardial calcification and in 21% of patients without pericardial calcification (p<0.001). In that study, patients with pericardial calcification had symptoms of longer duration and were more likely to have pericardial knock, larger atrial size, and atrial arrhythmia and significantly more perioperative deaths. However, the incidence of late survival was similar in both groups.

Pericardial thickness is generally increased in CP. However, it is important to note that normal pericardial thickness does not exclude CP, as an inelastic fibrous visceral epicardium can physiologically constrict without a demonstrable increase in thickness. In a recent study of 143 patients, baseline pericardial thickness was not increased in 18% of patients with surgically proven CP, although the histopathologic appearance was focally abnormal in all cases. When the clinical, echocardiographic, or invasive hemodynamic features indicate CP in patients with right-sided heart failure, pericardiectomy should not be denied on the basis of normal thickness as demonstrated by non-invasive imaging. The etiologies reported to be more commonly associated with normal pericardial thickness with constrictive physiology are previous cardiac surgery and previous thoracic irradiation; whereas infective pathology, especially tuberculosis, is generally associated with increased pericardial thickness.

Echocardiographic Assessment of CP

**M-Mode Echocardiography**

M-mode echocardiography in the parasternal long-axis view can identify a thickened pericardium.

**M-Mode Signs Reflecting Rapid Early Diastolic Filling**

1. The LV posterior wall demonstrates rapid early relaxation with posterior movement during early diastole, followed by abrupt cessation of such movement during mid- and late diastole. This flat motion of the LV posterior wall during mid- and late diastole corresponds to the abrupt transition of rapid ventricular filling in patients with CP. Also, the net diastolic LV posterior wall endocardial movement posteriorly is less than 1 mm in CP compared with normal controls in whom the posterior wall endocardial posterior movement ranges from 1.5 to 4 mm. There is no gradual downward motion in mid-diastole or with atrial systole. This sign, however, is not pathognomonic of CP.

2. A steep E–F slope in M-mode tracing of the mitral valve is seen in CP, suggesting rapid early diastolic filling.

3. Another sign of rapid early ventricular filling is a sharp downward motion of the posterior aortic root in early diastole on M-mode tracing of the aorta.

**M-Mode Signs Reflecting Ventricular Interdependence**

1. Abrupt anterior or posterior motion of the septum in early diastole is seen in most patients with CP and reflects the rapid change in transseptal pressure gradient during early diastole caused by unusually vigorous early ventricular filling. The direction of the motion of the septum in early diastole depends on a number of factors, including uneven distribution of fibrosis or calcification in the pericardial sac, timing of mitral and tricuspid opening, relative compliance of the right and left ventricles, and phase of respiration. The
The interventricular septum shows an abrupt bouncing motion towards the left ventricle during inspiration, followed by a shift in the opposite direction during expiration, reflecting an exaggerated interventricular dependence. Candell-Riera et al studied the interventricular septum in chronic CP. A brisk, early diastolic anterior movement followed by a rebound towards the LV posterior wall occurs (Fig. 1). The beginning of this anomalous movement coincides with the pericardial knock in the phonocardiogram and its peak coincides with the simultaneously recorded deep trough in the jugular pulse tracing (2). There is also an exaggerated anterior motion of the septum with atrial filling, which occurs because of the restriction of posterior free wall motion by the pericardium and results in septal displacement towards the RV. This anterior motion of the septum occurs at the end of the electrocardiographic P wave.

M-Mode Sign Reflecting Increased Right-Sided Diastolic Pressure Premature opening of the pulmonary valve (opening independent of atrial or ventricular systole) can be observed in patients with CP. Premature pulmonary valve opening is caused by the restriction of diastolic filling of the RV with subsequent mid-diastolic pressure rise, which exceeds the pulmonary artery diastolic pressure. However, it is a non-specific finding and can be recorded in other conditions such as sinus of Valsalva rupture into the RA, RCM etc.

2-D Echocardiography

The initial role of 2-D echocardiography is to exclude other causes of right heart failure, such as unsuspected mitral valve disease, pulmonary hypertension, LV systolic dysfunction etc. Increased systemic venous pressure is reflected by a dilated inferior vena cava without or with minimal respiratory variation. Spontaneous echocardiographic contrast is frequently seen in the inferior vena cava and the hepatic veins. Pericardial thickness is usually increased in CP. Pericardial thickness and calcification can be assessed on 2-D echo, though other investigative modalities, such as computed tomography (CT), are better suited for assessing pericardial thickness. However, pericardial thickness is also a function of gain settings and has been shown to correlate poorly with the degree of thickening found at surgery. Pericardial effusion, if present, can also be easily identified. Some dilatation of the atria, especially the left atrium (LA), can be noted in CP, but severe bi-atrial enlargement, together with thickened ventricular walls with unusual texture, generally go in favor of RCM. In patients with CP, septal bounce resulting from exaggerated ventricular interaction can be detected. Displacement of the interatrial septum towards the LA during inspiration is another reported sign of CP. The angle between the posterior LV wall and posterior LA is decreased in CP, because the thickened, constricting pericardium affects the posterior LV more than the posterior LA. Thus, the posterior LA wall expands at a more acute angle to the LV wall.

Pulsed Wave Doppler

The Doppler pattern of mitral inflow demonstrates an early increased diastolic filling velocity followed by a rapid deceleration, leading to a short filling period. Mitral E wave deceleration time is usually, but not always, less than 160 ms. As first reported by Hatle et al., dynamic changes with respiration occur in patients with CP, but not in patients with RCM. Two factors responsible for these changes are the dissociation of respiratory changes in intrathoracic pressure with intracardiac pressure and enhanced interventricular dependence. In CP, early diastolic mitral flow is reduced with the onset of inspiration and isovolumic relaxation time is prolonged. With expiration, mitral flow returns to normal and little variation is noted with respiration. After pericardiectomy, the flow patterns return to normal and respiratory variation is lessened. Typically, patients with CP demonstrate an increase in mitral inflow early diastolic velocity greater than or equal to 25% during expiration compared with inspiration. There is a reciprocal relationship with tricuspid flow. These changes can be seen with the first beat of inspiration, when a decrease in the E velocity on the transmitral flow velocity tracing is noted and reverse occurs with expiration. After pericardiectomy, the flow patterns return to normal and little variation is noted with respiration. Pulsed Doppler assessment of respiratory variation has recently been reported to be useful for evaluating the outcome of pericardiectomy. Patients who had minimal respiratory variation after pericardiectomy were asymptomatic compared with those who continued to show respiratory variation.

Fig 2. Diffuse pericardial thickening on axial computed tomography (A) and characteristic respiratory variations in transmitral pulsed wave Doppler (B) and hepatic venous pulsed wave Doppler (C) tracings.
A subset of patients with CP (≥20%), do not exhibit typical respiratory changes, because of a mixed restrictive–restrictive physiology or a markedly increased LA pressure. Typical respiratory changes in the latter situation may not be observed, because if LA pressure is markedly increased, the mitral valve opening occurs at a steep portion of the LV pressure curve, at which time the respiratory change has little effect on the transmural pressure gradient. In these patients maneuvers that decrease preload (head-up tilt or sitting) can unmask the characteristic respiratory variation in mitral E velocity. Atrial fibrillation makes the interpretation of respiratory variation in Doppler velocities difficult, but even in the presence of atrial fibrillation the respiratory variation can still be appreciated, regardless of the cardiac cycle length. Usually this requires longer recording periods of Doppler tracings. Aortic flow in patients with CP is also decreased markedly with inspiration.

This respiratory variation in transmitral flow is not seen in RCM; however, these phasic changes in mitral flow pattern with respiration can also be observed in other conditions such as chronic obstructive pulmonary disease (COPD), pulmonary embolism and RV infarction. However, their character may be different in these other conditions. In patients with COPD, large respiratory variations in intrathoracic pressure may result in inspiratory decrease in the E velocity at mitral inflow; however, the changes are more gradual, occurring later in the respiratory cycle.

Superior vena caval velocities can be helpful in distinguishing between CP and COPD. Patients with COPD display a marked increase in inspiratory superior vena caval systolic forward flow velocity, which is not seen in CP and occurs because, in COPD, there is a greater decrease in intrathoracic pressure in inspiration, which generates greater negative pressure changes in the thoracic cavity. This enhances flow to the RA from the superior vena cava.

The pulsed Doppler recording of hepatic venous flow mirrors the RA pressure tracing. Pulsed wave Doppler recordings from the hepatic vein in CP show marked diastolic flow reversal, which increases with expiration compared with inspiration although it is not unusual to see significant diastolic flow reversals during both inspiration and expiration in patients with advanced constriction or with mixed restrictive–restrictive physiology. In contrast, diastolic flow reversal in the hepatic vein flow is more prominent with inspiration in RCM.

Evaluation of pulmonary venous flow by Doppler shows marked respiratory change in CP. The pulmonary venous systolic wave and early diastolic wave velocities, especially the early diastolic wave velocity, are increased during expiration and decreased during inspiration. The changes in pulmonary venous flow velocities have been reported to be more pronounced than changes in mitral inflow velocities. The combination of a systolic wave velocity and early diastolic wave velocity ratio of more than 0.65 in inspiration and a respiratory variation of early diastolic velocity of more than 40% correctly classified 86% of patients with CP. Similar respiratory variation can also be observed in patients with CP and atrial fibrillation, regardless of the irregular cycle lengths. In contrast, patients with RCM show blunting of the systolic wave velocity and decreased systolic wave velocity and early diastolic wave velocity ratio throughout the respiratory cycle with a large atrial reversal wave without any significant respiratory variation.

**Tissue Doppler Imaging (TDI)**

Both CP and RCM cause diastolic filling abnormalities with relatively preserved global systolic function. Diastolic dysfunction in CP is the result of a fibrosed, non-compliant pericardium, whereas in RCM it is caused by a stiff and non-compliant ventricular myocardium. Both disease processes limit diastolic filling and result in diastolic heart failure. TDI is a relatively new echocardiographic method of quantifying regional and global myocardial function and has been shown to be helpful in the clinical diagnosis of CP. The quantitative assessment of longitudinal mitral annular motion by this method provides an accurate estimate of global LV function. Because the mechno-elastic properties of the myocardium are preserved in CP, the longitudinal mitral annular velocities are normal. It has
been reported that an early diastolic mitral annular velocity (Ea) >8 cm/s at the lateral or septal mitral annular corner and an Ea/E <0.11 can distinguish between patients with CP and those with RCM. Mitral annular velocities are particularly useful in CP when a pronounced respiratory variation in peak mitral E velocities is not seen. In patients with RCM, who have intrinsic myocardial relaxation abnormalities, early diastolic mitral annular velocities are reduced. The overall sensitivity and specificity for diagnosing CP using tissue Doppler incrementally with M-mode, 2-D, and transmitral flow Doppler is 88.8% and 94.8%, respectively.

The overall sensitivity and specificity for diagnosing CP ties, early diastolic mitral annular velocities are reduced. Mitral annular velocities help with the diagnosis and differentiation of CP in most cases, except in the presence of extensive annular calcification, LV systolic dysfunction, or segmental non-uniformity in myocardial velocities. Sengupta et al. have reported that in RCM, annular velocities may not always be representative of a disease process that is heterogeneous and this may limit the differentiation of RCM caused by endomyocardial fibrosis from CP by mitral annular velocities assessment. In two patients with endomyocardial fibrosis in their series, the mitral annular velocities were normal at the lateral mitral annulus, and the mitral annular velocity at the septal corner was also normal in 1 of them. Regional heterogeneity of the disease process, which has been reported earlier in patients with endomyocardial fibrosis, has been suggested as responsible for the normal velocities. Thus, hypokinetic areas accompanied by hyperkinesis in neighboring normal areas could result in relatively unaltered overall annular motion. Recently, high velocity (>7 cm/s) early diastolic biphasic motion with or without multiple recoil waves (polyphasic diastolic septal flattening) of the interventricular septum by pulsed wave Doppler tissue imaging was shown to have high sensitivity (82.5%) and specificity (92.7%) in differentiating CP patients from controls.

2-D Speckle Tracking
The gray-scale image is seen to consist of a speckled pattern. The served pattern is not an actual image of the scatterers in the tissue itself, but the interference pattern generated by the reflected ultrasound. The speckle pattern remains reasonably stable, and the speckles follow myocardial motion. The ability of 2-D speckle tracking to detect longitudinal and circumferential myocardial deformation has made it a useful modality for differentiating CP from RCM. Echocardiographic assessment by 2-D speckle tracking highlights the two distinct patterns of abnormal diastolic restoration mechanics seen in CP and RCM. In a study of 26 patients with CP, 19 with RCM, and 21 age-matched controls, Sengupta et al. characterized and compared their LV longitudinal, circumferential and radial mechanics. Patients with RCM had abnormal longitudinal mechanics (reduced longitudinal strain, particularly at the LV base) with relative sparing of LV rotation. In contrast, CP patients had markedly abnormal circumferential deformation, torsion and untwisting velocity, with relative sparing of the longitudinal mechanics. Furthermore, a comparison of the LV mechanics with pericardial thickness, as measured by CT, in patients with CP has shown a significant correlation between decreased circumferential strain and degree of pericardial thickening at the apex.

Transesophageal Echocardiography (TEE)
TEE can be a useful adjunct to the assessment of patients with CP. It is superior to transthoracic echocardiography in measuring the pericardial thickness and has an excellent correlation with CT for this purpose. The transgastric view by TEE has been reported to give high-quality images of the pericardium, which might be useful in diagnosing CP. Further, when mitral inflow velocities by transthoracic echocardiography are technically inadequate, measurement of pulmonary venous Doppler velocities using TEE can be helpful in demonstrating the pronounced respiratory variation.

Effusive-CP
Effusive-CP is a form of subacute CP characterized by pericardial effusion together with constriction of the visceral pericardium. The effusion may be concentric, regional, or loculated. Also, over time the effusion may organize and the pericardial layers may become thickened. The result is a spectrum of hemodynamic patterns from pure tamponade through an effusive-constrictive physiology to pure constriction. Effusive-CP is characterized by mixed clinical and hemodynamic findings. The effusive-constrictive nature of the physiology can only be revealed when pericardiocentesis fails to completely reduce the diastolic pressures. Generally, the hemodynamic findings before pericardiocentesis resemble tamponade, and after pericardial fluid drainage the characteristics of constriction become apparent. Before pericardiocentesis, the diastolic pressures are elevated and equalized with a prominent paradoxical pulse, but with preserved y descent in contrast to classical tamponade. These findings, in conjunction with Kussmaul’s sign, suggest the existence of a constrictive component. Effusive-CP may be transient and may not always progress to frank CP, reflecting the dynamic healing process within the pericardium. It is commonly seen in post-pericardiectomy patients, but can also be seen with post-infectious pericarditis or uremia.

Transient CP
In some patients with acute CP, the symptoms and constrictive physiologic features resolve with medical therapy alone, a phenomenon that has been labeled transient CP and was first described in 1987. Although most patients with acute pericarditis proceed directly to complete resolution, in some cases recovery may be preceded by transient constriction; others may develop subacute effusive-CP, sometimes followed by classical CP, or their condition may directly progress to subacute or chronic constriction. In a recent study of 212 patients who had echocardiographic findings of CP, 36 of these patients on follow-up echocardiograms showed resolution of the constrictive hemodynamics without pericardiectomy. The causes for the CP were diverse, the most common being prior cardiovascular surgery (25%). In a subset of 22 patients who were followed serially during the course of their illness, resolution of the constrictive physiologic features occurred at an average of 8.3 weeks after diagnosis. The awareness of this entity may obviate the need for surgery in some patients.

In summary, there are numerous echocardiographic signs of CP, but no single finding is pathognomonic of the disease. A detailed echocardiographic study including M-mode, 2-D, Doppler and TDI will help to confirm or rule out the diagnosis of CP. However, equivocal echocardiographic findings may be present in up to 25% of patients with possible CP, especially those with mixed pericardial and myocardial disease, necessitating further studies such as
cardiac catheterization and CT.

Cardiac Catheterization

Right- and left-heart catheterization in patients suspected of having CP provide documentation of the hemodynamics of constrictive physiology and assist in differentiating between CP and RCM. Right- and left-heart pressures should be recorded simultaneously at equisensitive gains, with meticulous attention to calibration. The equilibration of LV and RV pressures with impaired diastolic filling pattern is a hallmark of constrictive physiology. RA, RV diastolic, pulmonary capillary wedge and pre-a wave LV diastolic pressure are elevated and equal, or nearly so (within less than 5 mmHg), at approximately 20 mmHg. The RA pressure tracing shows a preserved x descent, a prominent y descent, and almost equal a and v wave heights, with the resultant M- or W-shaped configuration. Both the LV and RV pressures reveal an early, marked diastolic dip followed by a plateau, (“dip and plateau” or “square-root” sign). Pulmonary artery systolic and RV systolic pressures are modestly elevated, usually less than 45 mmHg. Pulmonary hypertension is not a feature of CP and is indicative of coexisting cardiac or pulmonary disease. Hypovolemia, as may occur with aggressive diuretic therapy, may mask the typical hemodynamic features of CP and may require rapid volume challenge of normal saline of 1,000 ml over 6–8 min to unmask the hemodynamic features. Although the stroke volume is almost always reduced because of the decreased diastolic filling, compensatory tachycardia maintains normal or near-normal resting cardiac output. Simultaneous systolic pressure tracings of the LV and RV pressures demonstrate ventricular pressure discordance, whereby while during inspiration the RV systolic pressure increases, the LV systolic pressure falls. During expiration the reverse is seen, with RV systolic pressure decreasing and the LV systolic pressure increasing. This phenomenon, caused by exaggerated interventricular dependence, has been shown to have a sensitivity of 100% and specificity of 95% for diagnosing CP. In contrast, in patients with congestive heart failure both the LV and RV systolic pressures decrease during peak inspiration. Recently the ratio of the RV to LV systolic area during inspiration and expiration has been shown to be a reliable catheterization criteria for the diagnosis of CP. During inspiration, the area of the RV pressure curve increases, while the area of the LV pressure curve decreases, as compared with expiration. This finding has a sensitivity of 97% and a predictive accuracy of 100% for the identification of patients with surgically proven CP.

CT and Magnetic Resonance Imaging (MRI)

CT and MRI provide excellent visualization of the pericardium and can lend support to the diagnosis of CP. The CT and MRI hallmarks of CP include pericardial thickening greater than 4 mm, and narrow, tubular deformation of the RV with a straightened or sigmoid-shaped interventricular septum (Fig 2). Dilatation of the inferior vena cava, hepatomegaly and ascites are also frequently seen. CT has the added ability of detecting pericardial calcifications. When accompanied by clinical signs of heart failure, these findings are highly suggestive, but not diagnostic, of CP

Pericardectomy

The results of several large series of patients undergoing pericardectomy for CP have reported improvement in both hemodynamics and clinical symptoms in the majority of patients. A low output syndrome persists in a considerable number of patients after pericardectomy, which may result from long-standing pericardial restriction causing atrophic changes in the myocardium or from the epicardial fibrosis penetrating deeply with resultant myocardial involvement. In patients requiring pericardectomy, early surgery allows easier and more complete decortication with low surgical mortality and preservation of myocardial function. Tricuspid regurgitation frequently accompanies CP and when severe is associated with increased mortality. Concomitant pericardectomy and tricuspid valve repair in patients with moderate to severe tricuspid regurgitation has not been shown to improve late mortality in CP.

Conclusion

CP is a heterogeneous disease characterized primarily by fibrosis and often thickening of the pericardium, leading to abnormal diastolic function. New advances in echocardiographic techniques, including TDI and 2-D speckle tracking, have made for easier differentiation of CP from RCM. Cardiac catheterization, CT and MRI remain useful aids in the diagnosis of CP. Accurate diagnosis of CP is essential in order to identify patients who may benefit from pericardectomy.

References

15. Voelkel AG, Pietro DA, Folland ED, Fisher ML, Parisi AF. Echocar-