Recent Role of Imaging in the Diagnosis of Pericardial Disease

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Noninvasive cardiac imaging techniques have made a striking impact on the evaluation and management of pericardial disorders. Two-dimensional and Doppler echocardiography are the methods of choice in the evaluation of pericardial effusion and cardiac tamponade. Magnetic resonance imaging, computed tomography, and transesophageal echocardiography are valuable in the assessment of pericardial thickness in suspected cases of constrictive pericarditis. Filling dysfunction associated with constrictive pericarditis is well demonstrated by Doppler flow velocity recordings of intracardiac flow jets, and pulmonary and hepatic venous flow streams. Tissue Doppler echocardiography, by which tissue velocity of myocardial regions and mitral annulus are analyzed, offers additional information in the differentiation of constrictive pericarditis and restrictive cardiomyopathy. Magnetic resonance imaging and computed tomography are the techniques of choice in the recognition of unusual disorders such as pericardial cysts, tumors invading the pericardium, and congenital absence of pericardium.
Noninvasive imaging aids not only in the diagnosis of pericardial diseases, but also in the guidance of optimal therapy.

Introduction

Pericardial disorders result from a wide variety of cardiac and systemic pathology. The clinical impact of pericardial diseases varies from the innocuous trivial pericardial effusion, to life-threatening cardiac tamponade or progressive pericardial constriction. Prompt diagnosis of the pericardial disorder and definition of its severity are critical for optimal patient management. Until two decades ago, the diagnosis of pericardial disease relied on insensitive or invasive approaches, such as fluoroscopy and cardiac catheterization. In the recent decades, noninvasive cardiac imaging has emerged as the mainstay of the diagnosis of pericardial diseases. Among imaging techniques, echocardiography is the primary tool for this purpose; computed tomography (CT) and magnetic resonance imaging (MRI) are useful mainly in the assessment of pericardial thickness, or in the detection of unusual abnormalities such as absence of pericardium or pericardial cysts. This review focuses on how to integrate the various modalities of cardiac ultrasound, the clinical features of a patient, and other imaging techniques to detect pericardial diseases.

Pericardial Effusion

The pericardium is a unique, double-layered structure that surrounds the heart. The physiologic space between these layers, the pericardial space, usually contains about 10 to 50 cc of lubricating plasma ultrafiltrate. Accumulation of fluid above the normal amount constitutes a pericardial effusion. Pericardial effusion can be circumferential, enveloping the cardiac chambers, or localized, as often noted following cardiac surgery.

Echocardiographic findings

Pericardial effusion is easily identified by the presence of a space between the layers of the pericardium, not only by echocardiography, but also by CT and MRI [1–4]. On two-dimensional echocardiograms (2DE), an echo-free space seen surrounding the cardiac structures indicates the presence of a pericardial effusion (Fig. 1). If a space is visualized primarily during systole, it denotes normal pericardial fluid. An echo-free space less than 5 mm in width indicates a small pericardial effusion, 5 to 10 mm a moderate-sized effusion, and more than 10 mm a large effusion. Although a few approaches are available to quantify the volume of the effusion [5], such quantification is generally not necessary in clinical practice; evaluation of whether the effusion is small, moderate, or large is sufficient. Misinterpretation of epicardial fat as a pericardial effusion is a common pitfall in the diagnosis of pericardial effusions [6–8]. Epicardial adipose tissue is usually more prominent anteriorly, but can be circumferential, and thus can be easily misdiagnosed as a pericardial effusion. Fat, however, is usually recognized as a low intensity echogenic pericardial space, whereas a pericardial effusion is usually echolucent. Careful examination to identify such low-intensity echo signals with a granular appearance and their movement with the

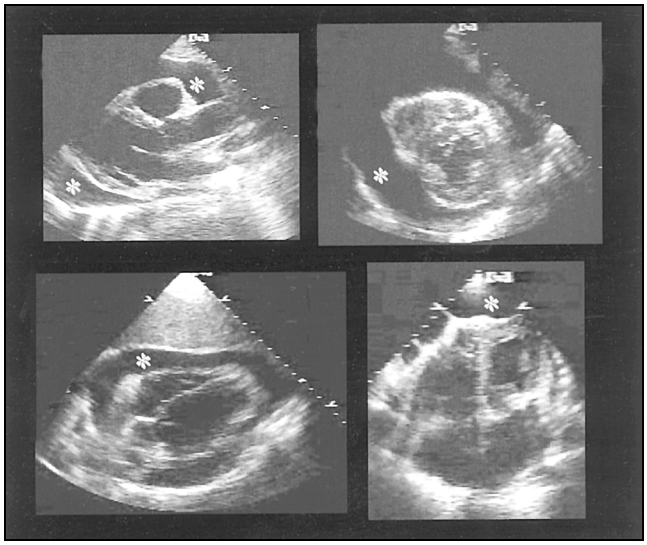

Figure 1. Two-dimensional echocardiographic images of pericardial effusion (*). **Top,** parasternal long axis (*left*) and short axis (*right*) views showing a circumferential effusion. **Bottom,** subcostal view (*left*) shows a large circumferential effusion. Apical four-chamber view shows a loculated effusion with septi in the region of the apex, causing an apical biventricular collapse.

myocardium would help in the differentiation of fat pads from a true effusion. An organized pericardial effusion can also be associated with ultrasound signals, but organized material does not slide with the myocardial movement like a fat pad. Although an organized effusion, a hematoma in the pericardium, or a purulent effusion may exhibit echogenic signals in the pericardial space, the precise nature of a pericardial effusion cannot generally be recognized by echocardiography.

The location of the pericardial effusion is also well recognized by ultrasound imaging. A small pericardial effusion is often posterior in echocardiographic studies performed with the patient in a recumbent posture. Pericardial fluid collection, however, can be localized to any region. Even though pericardial reflection is limited posterior to the atrium, a large pericardial effusion can extend behind the left atrial (LA) wall. A left pleural effusion may mimic a posterior pericardial effusion. Examination of the relation of the echo-free space to the descending aorta is useful in differentiating a pericardial effusion from a pleural effusion. Pericardial effusion adjoins the anterior and anterolateral aspect of the aorta, whereas a pleural effusion is seen along the posterolateral aspect of the aorta. Accurate localization of the pericardial fluid is important, particularly when deciding how to effectively drain an effusion. Pericardial effusion following cardiac surgery is often localized posteriorly. It may warrant echo-guidance during pericardiocentesis, or require a surgical approach if drainage of the effusion is mandated because of cardiac tamponade. If transthoracic imaging is technically suboptimal, transesophageal echocardiography (TEE) can clearly define the presence, location, and effects of pericardial effusions.

Cardiac Tamponade

The accumulation of fluid in the pericardium in an amount sufficient to cause significant impediment to the inflow of blood to the ventricles results in cardiac tamponade. The pericardium is able to adapt to increases in intrapericardial fluid volume up to a certain limit without any significant concomitant increases in the intrapericardial pressure. When the adaptive limit is reached, or if the fluid accumulates rapidly, the rise in intrapericardial pressure is exponential and causes external pressure on the cardiac chambers. The three principal features of tamponade are limitation of ventricular filling, elevation of cardiac filling pressures, and reduction of cardiac output. Although cardiac tamponade is usually associated with effusions of moderate or large size, rapid fluid accumulation in a short period (such as a hemopericardium due to trauma) can result in life-threatening tamponade. Prompt recognition of tamponade is important because of the need to perform life-saving pericardiocentesis. Clinical findings such as hypotension and pulsus paradoxus are limited in their sensitivity and specificity, and thus objective demonstration of a pericardial effusion and its hemodynamic effects is important.

Echocardiographic findings

Echocardiography is the best imaging modality for the diagnosis of cardiac tamponade. The first key to the diagnosis is, of course, the demonstration of a pericardial effusion. The hemodynamic effects of a compressive effusion are reflected by a number of 2DE and Doppler findings. In a large effusion with advanced compression, the chamber sizes can be small because of decreased filling volume. Otherwise, the size of the chambers is normal. The inferior and superior vena cavae, however, are dilated because of elevated filling pressures. High right atrial (RA) pressures associated with tamponade cause reduction or abolition of the normally seen collapse of the inferior vena cava during inspiration [9]. Other useful signs include right atrial collapse (RAC), right ventricular diastolic collapse (RVDC), both shown by 2DE imaging, [10–13], and exaggerated flow velocity variation with respiration identified by Doppler recordings (Figs. 2, 3) [14,15••]. Elevated intrapericardial pressure and the transmural pressure difference between intrapericardial and intracardiac pressure result in the characteristic deformation of chamber walls. RAC, invagination of the RA wall, is a common finding in tamponade. The RA wall, instead of maintaining a normal rounded contour during the cardiac cycle, exhibits a localized invagination toward the cavity during atrial relaxation, when the intracavitary RA pressure falls below the intrapericardial pressure. In cases of advanced tamponade, the duration of RAC is prolonged. If the pericardial effu-

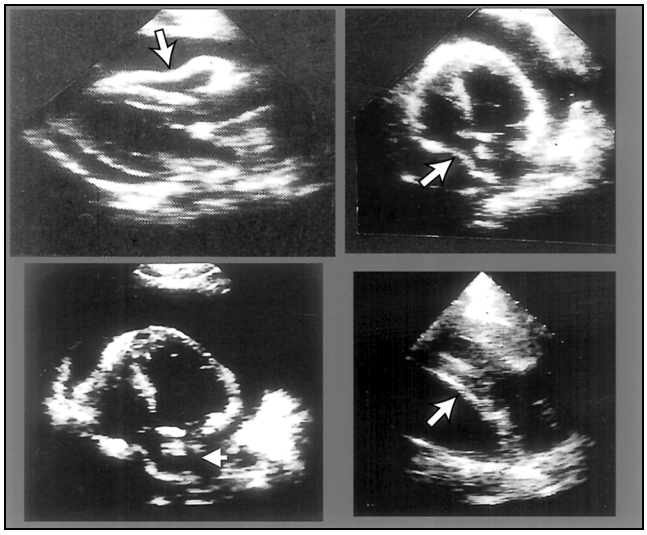

Figure 2. Two-dimensional echocardiographic images in cardiac tamponade. The *top left* figure demonstrates right ventricular collapse in the parasternal long axis view. The *top right* figure is an apical four chamber view showing right atrial collapse. The *bottom left* demonstrates left atrial collapse, and the *bottom right* picture shows left ventricular collapse.

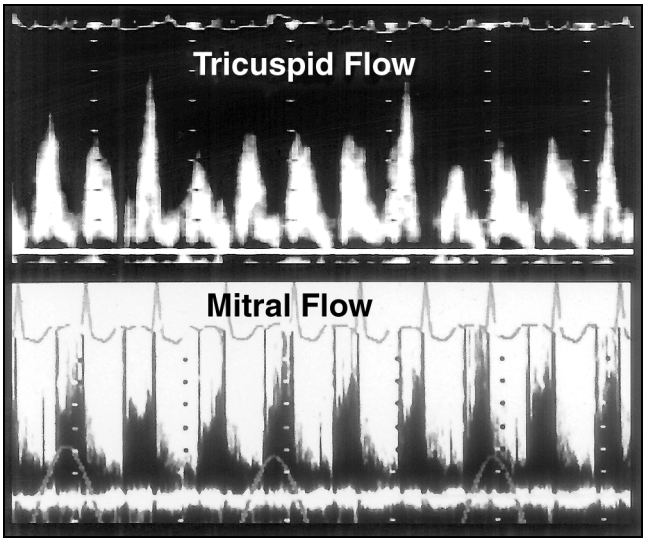

Figure 3. Doppler recordings of respiratory variation in tricuspid and mitral flow velocities in cardiac tamponade.

sion surrounds the LA, left atrial collapse (LAC) can occur based on similar mechanism.b a similar mechanism [13]. Both RAC and LAC are visualized in apical and subcostal four-chamber views on 2DE recordings (Fig. 2). Another echocardiographic sign of tamponade is RVDC. During early ventricular diastole, the RV cavity pressure falls below the intrapericardial pressure, resulting in a negative transmural pressure gradient. This causes invagination of the RV wall toward the cavity. RVDC is best recognized in parasternal long axis and short axis views. An extreme degree of tamponade can result in compression and collapse of the whole RV chamber. A similar mechanism can be operative in the manifestation of left ventricular diastolic collapse (LVDC), when a loculated effusion adjacent to the LV causes tamponade [16].

similar mechanism.b One or more of these signs (RAC, RVDC, and LAC) may be present in a given patient, and their predictive accuracy varies. RAC and LAC have been found to have about 90% sensitivity in tamponade caused by circumferential effusions. The sensitivity of RVDC is about 80% to 90%. These signs have an excellent negative predictive value in ruling out the diagnosis of cardiac tamponade, except in certain situations where the intracardiac pressures were unduly elevated prior to the development of tamponade.

The characteristic Doppler finding in tamponade is that of increased respiratory variation in flow velocities [14,15••]. The filling of cardiac chambers is dependent on respiratory changes in intrathoracic pressure, pulmonary capillary wedge pressure, and intrapericardial pressure (IPP). During inspiration, there is an increase in venous return resulting in more blood entering the RA, and consequently the RV and pulmonary artery. In cardiac tamponade the IPP is increased. When the RV fills in diastole during inspiration, it has less space to expand. Thus, the interventricular septum bulges in the LV cavity. Conversely, during expiration there is increased LV filling in diastole and the septum bulges in the RV cavity. This phenomenon is called *interventricular dependence*. The IPP also determines the LV diastolic pressure (LVDP). Therefore, the high IPP in tamponade results in an increase in the LVDP. During inspiration, the intrathoracic pressure decreases, but because of the tamponade, the LVDP does not decrease as much as it normally would. This results in a decreased gradient between the pulmonary capillary wedge pressure and the LVDP, *ie*, decreased LV filling pressure causing decreased mitral inflow during inspiration. In normal individuals, the right-sided flow velocities (tricuspid and pulmonic flow velocities) increase by approximately 30% during inspiration, and left-sided flow velocities (mitral and aortic) decrease less than 20%. In the setting of tamponade, there is greater respiratory variation in these flow velocities. This finding is best noted in tricuspid and mitral flow (Fig. 3) [14,15]. Similar respiratory variation can be noted in the pulmonary venous and hepatic vein flow velocities. Inspiratory increase in antegrade diastolic flow in the hepatic vein with an expiratory increase in reversed flow in the hepatic vein, and converse changes in pulmonary vein flow are registered by pulsed Doppler recordings.

Atypical presentations

In the setting of a loculated pericardial effusion or in the presence of coexisting diseases, the classic clinical as well as echocardiographic features of cardiac tamponade may not be present.

Regional tamponade

Because the RV and RA walls are often adherent to the chest wall following cardiac surgery, the most common

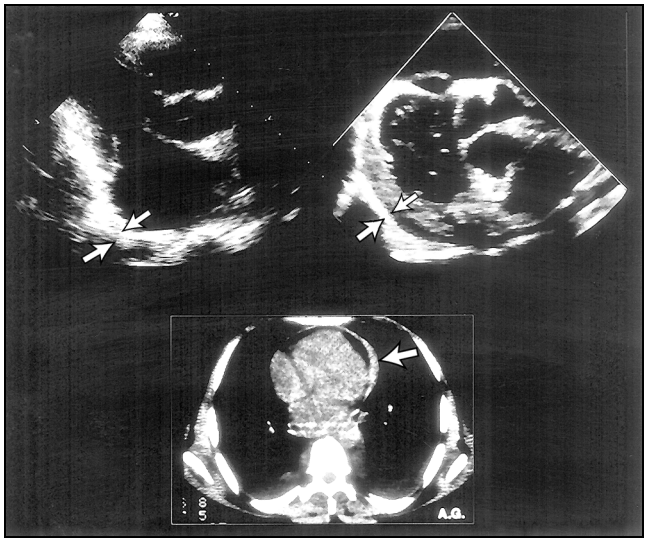

Figure 4. Images of thickened pericardium. **Top left,** transthoracic two dimensional echocardiography. **Top right,** transesophageal echocardiography. **Bottom,** computed tomography.

location of the effusion is posterior [16]. Consequently, the typical right-sided findings such as RAC and RVDC may not be seen. If the posterior pericardial effusion is under enough tension to cause tamponade, even the thick-walled LV posterior wall may exhibit diastolic collapse, or LVDC [17]. This finding, however, is subtle and therefore a careful examination of the LV in parasternal and apical views is essential to detect LVDC [18••]. Doppler examination to assess flow velocity variations is also of help.

Coexisting disorders

In the presence of pulmonary hypertension, RV hypertrophy and any condition that already has caused elevated right-sided filling pressures, RVDC and RAC could be absent despite cardiac tamponade physiology. Similarly pre-existing left-sided disease may mask the development of LVDC and LAC. Presence of an atrial septal defect and aortic regurgitation can mask flow velocity variations. The volume status of the patient also could influence the sensitivity of echocardiographic signs. If a patient is volume depleted either due to dehydration or excessive diuresis, there could be "low pressure tamponade" [19]. RAC and RVDC may still be noted in these patients. If they are absent, an echocardiographic study repeated after a volume challenge is useful in unmasking these and other signs.

Other scenarios to be kept in mind include severe chronic obstructive lung disease, in which pulsus paradoxus and respiratory flow velocity variation may be present, even in the absence of tamponade, as well as conditions such as pulmonary embolism and RV infarction, which may be associated with elevated RV pressures. Thus, a skillfully performed and interpreted echocardiogram, and consideration of the clinical context are essential to confirm or exclude cardiac tamponade.

Echocardiographic guidance for pericardiocentesis

The treatment of choice of cardiac tamponade is drainage of the pericardial fluid. Most centers prefer to employ 2DE guidance for percutaneous pericardiocentesis [20•]. This has lead to a low rate of pneumothorax, ventricular puncture, and death, all of which have been associated with the blind percutaneous techniques. Echocardiography also helps to monitor the results of the pericardiocentesis.

Constrictive Pericarditis

Constrictive pericarditis is a disease with two fundamental problems: a morphologic abnormality (thickened, nonpliable pericardium [Fig. 4]), and a physiologic abnormality (impaired diastolic filling). The classic picture of constrictive pericarditis has been that of a chronic disorder caused most often by tuberculosis. It is characterized by symptoms of dyspnea, ascites, and leg edema, physical findings of elevated jugular venous pressure and diastolic knock on auscultation, radiographic findings of calcified pericardium, and hemodynamic findings of elevated intracardiac diastolic pressures with equilibration, prominent Y descent in atrial pressure, and a dip and plateau shape in ventricular diastolic pressures. Pericardiectomy has been the therapeutic procedure of choice. The incidence of etiologic factors has changed during recent decades, with consequent shift in the clinical spectrum of constrictive pericarditis. Tuberculosis was considered to be the most common etiology in the past. Although it is still a common cause in a few countries where tuberculosis infection is prevalent, this etiology has become less common in western countries. An increasing frequency of constrictive pericarditis due to cardiac surgery and mediastinal radiation is noted today. Furthermore, it is being increasingly encountered in older patients rather than younger ones. The change in the profile and spectrum of etiologic mechanisms has also caused alterations in pathophysiology and clinical expression. The basic physiologic derangement is the inability of the ventricles to distend normally and to fill adequately because of the restraint posed by a thick, noncompliant pericardium. This is associated with an increase in intracardiac filling pressures, and a reduction in cardiac output. To cause a restraint in filling, the pericardium does not have to be calcified or markedly thickened. Even a noncalcified, mildly thickened pericardium can become noncompliant and cause impediment to filling. Consequently, symptoms and signs of constrictive pericarditis can develop not only over months and years, as in the past, but even over a matter of weeks or days. Postoperative constrictive pericarditis is an excellent example of the rapid evolution of constrictive physiology. Thus, constrictive pericarditis has evolved into a disorder that can manifest as a chronic, subacute, or acute syndrome.

Furthermore, some of the underlying etiologic processes can result in some degree of myocardial fibrosis and

pathology. This could alter the classic hemodynamic findings of constrictive pericarditis. Other confounding factors include coexisting diseases such as ischemic heart disease, valvular heart disease, LV dysfunction, and pulmonary hypertension, which by themselves result in abnormalities in ventricular diastolic performance and left or right-sided intracardiac pressures. These factors could modify the finding of equilibration of filling pressures. In other words, equilibration of filling pressures is not an absolute accompaniment of constrictive physiology in all cases.

The classic presentation of chronic constrictive pericarditis used to be characterized by dyspnea on exertion, orthopnea, leg edema, congestive hepatomegaly, ascites, elevated jugular venous pressure, inspiratory increase in jugular venous pressure (Kussmaul's sign), prominent Y descent in the jugular venous pulse, and diastolic knock. With the changing spectrum of constrictive pericarditis, and with patients presenting early in the course of the syndrome, a lesser degree of elevation in venous pressure may be encountered. Symptoms may involve those more of coexisting diseases. Pulsus paradoxus occurs in few cases, and is usually less severe than in tamponade.

The diagnosis of constrictive pericarditis requires demonstration that the pericardium is thickened, and that there is filling dysfunction. A variety of diagnostic methods are available to assess pericardial thickness (Fig. 4) [3,21–24]. Echocardiography and invasive hemodynamic recordings are employed to characterize filling dysfunction. Although classic abnormalities are seen in many patients, it is to be noted that many atypical scenarios may be encountered posing a diagnostic challenge.

Demonstration of thickened pericardium

In classic cases of chronic constrictive pericarditis, pericardial calcification can be noted in the chest radiograph or at fluoroscopy. Such a finding is easily recognized on CT scans and MRI [23,24]. Pericardial calcification can be localized and shaggy, or like an eggshell. Progression of the disease to a stage of pericardial calcification has become rare in western countries (less than one third of patients). Pericardial calcification indicates a more chronic disease and also implies higher perioperative mortality.

In most cases of constrictive pericarditis encountered these days, the pericardium, although thickened, is not calcified. There can be varying degrees of pericardial thickening, which can be circumferential or localized. Pericardial thickness of 4 mm or more on CT or MRI is considered to represent an abnormal, thickened pericardium. Transthoracic echocardiography may show a shell-like appearance around the heart. A markedly bright pericardial echo may be noted, despite turning down the ultrasound gain. However, transthoracic echocardiography is not always sensitive in the identification of thickened pericardium. Transesophageal echocardiography (TEE) is more sensitive and accurate for the detection of thickened pericardium. TEE determination of pericardial thickness has yielded excellent correlation with anatomic measurements and CT estimations [23]. Pericardial thickness of 3 mm or more on TEE is 95% sensitive and 86% specific for the detection of thickened pericardium. Although most patients have a markedly thickened pericardium, some may exhibit constrictive physiology, even without demonstrable pericardial thickening. Conversely, the presence of a thickened pericardium does not always mean that a patient has constrictive pericarditis. A thickened pericardium could still be compliant and not cause any filling impediment. The diagnosis of constrictive pericarditis requires demonstration of filling dysfunction as well.

Demonstration of filling dysfunction

Filling dysfunction is best identified by 2DE and Doppler echocardiography, and in some cases by invasive hemodynamic recordings. 2DE reveals features consistent with impeded filling, exaggerated ventricular interdependence, and elevated filling pressures. The ventricular chambers are usually normal in size, but the atria and the inferior and superior vena cava are dilated. The usual decrease in inferior vena caval size normally seen during inspiration is blunted or absent. The LV chamber may show rapid expansion during early diastole, with a plateau during the rest of diastole. The interventricular septum exhibits an abrupt bouncing motion toward the LV during inspiration, followed by a shift in the opposite direction during expiration, reflecting pressure gradient changes between the two ventricles during respiration. This septal abnormality may also be seen as diastolic notches in the septum on M-mode echocardiographic recordings [25]. The pulmonic valve may open prematurely because of elevated RV diastolic pressure. Antegrade diastolic flow into the pulmonary artery may be seen on Doppler recordings.

The mitral flow velocity pattern shows a higher E wave velocity, an increased E wave to A wave ratio, and a shortened E wave deceleration time (Fig. 5). This pattern is due to a rapid fall in the ventricular pressure during early diastole with unimpeded rapid filling, followed by an abrupt impediment to further ventricular filling because the thick pericardium does not allow the ventricle to expand anymore. A thick constraining pericardium also does not allow transmission of intrathoracic pressure changes during respiration to intracardiac chambers. This results in increased ventricular interdependence, and dissociation of intrathoracic and intracardiac pressures. This phenomenon is reflected in Doppler recordings of intracardiac and pulmonary venous, and systemic venous flow velocity recordings. In normal patients, left-sided flow velocities exhibit less than 20% reduction during inspiration, whereas right-sided flow velocities show less than 30% increase in flow velocities. This cyclical change is markedly exaggerated (more than 30%) in patients with constrictive pericarditis (Fig. 5). If tricuspid regurgitation is present, its velocity, duration, and time velocity integral exhibit an exaggerated change during inspiration com-

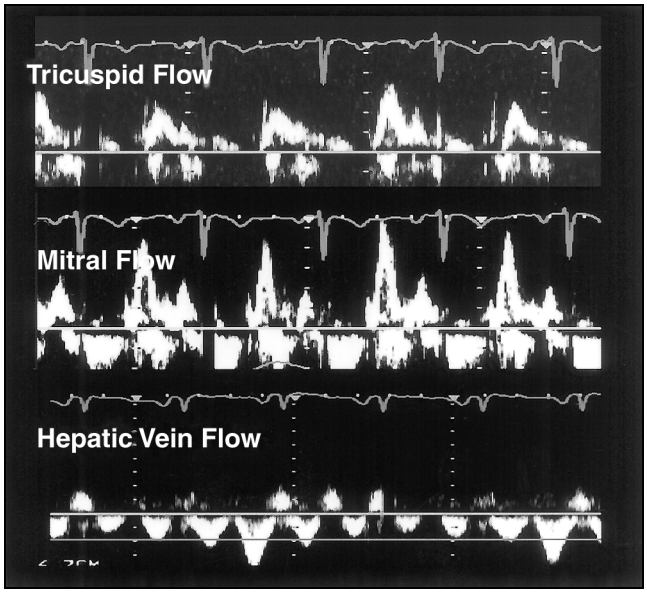

Figure 5. Doppler recordings of flow abnormalities in constrictive pericarditis. Tricuspid, mitral and hepatic vein flows show exaggerated respiratory variation. The early diastolic wave in the mitral flow is much taller than the atrial wave. Hepatic vein flow exhibits expiratory increase in late-diastolic reversal wave.

pared with normal subjects. If Doppler findings appear to be blunted or absent in patients suspected to have constrictive pericarditis, Doppler recordings repeated after maneuvers to decrease preload can be helpful in unmasking the characteristic findings.

Doppler assessment of the hepatic vein and superior vena cava in normal subjects demonstrates biphasic forward flow followed by flow reversal with atrial contraction and the peak systolic flow greater than peak diastolic flow (Fig. 5). In constrictive pericarditis, expiration results in a decrease in systolic or diastolic forward flow, or an increase in flow reversal.

It should be noted that many patients with constrictive pericarditis may continue to exhibit features of filling dysfunction even after pericardiectomy for a variable time period or even permanently. This generally implies either the development of epimyocardial fibrosis or incomplete pericardial resection. In patients with effusive-constrictive pericarditis, the symptoms and signs of cardiac tamponade are the presenting features. The continued presence of elevated cardiac pressures and features of constriction noted after pericardiocentesis establish the diagnosis of this condition [26•]. Hence, a follow-up echocardiogram after pericardial drainage is essential in all patients with cardiac tamponade.

Atypical presentations

The classic echocardiographic findings are altered in a number of scenarios. In occasional cases, particularly following cardiac surgery, there could be constrictive physiology even without demonstrable pericardial thickening. In many cases, this problem is temporary. The natural history of constrictive pericarditis in the absence of pericardial thickening has not been well studied. Pre-existing or coexisting disease processes can alter the typical echocardiographic features. Patients with disorders such as hypertension or coronary artery disease may have pre-existing diastolic dysfunction coupled with elevation of filling pressures. They may not exhibit the characteristic features of constrictive pericarditis. Likewise, findings associated with pre-existing right heart diseases or pulmonary hypertension may mask or mimic findings of constrictive pericarditis. It may not be easy to distinguish flow velocity changes associated with severe lung disease from those of constriction.

Differentiation from restrictive cardiomyopathy

In restrictive cardiomyopathies such as amyloidosis, cardiac imaging techniques reveal a pericardium of normal thickness in contrast to constrictive pericarditis. In contrast to constriction in which the myocardial wall thickness is generally normal, amyloidosis is associated with increased wall thickness. The combination of increased wall thickness and low voltages QRS complexes on the electrocardiogram is highly specific for infiltrative cardiomyopathy. The sparkling or speckled appearance on the 2DE described in amyloidosis is not sensitive, specific, or reliable, particularly in this era of harmonic 2DE. In cases of advanced amyloidosis with markedly increased myocardial thickness, the mitral flow velocity pattern may not be helpful in differentiating this condition from constrictive pericarditis, because both conditions could have increased E velocity with rapid deceleration. The respiratory variation in flow velocities seen in patients with constriction, however, are absent in restrictive cardiomyopathy, and thus is a useful distinguishing feature. Recordings of pulmonary venous and hepatic vein flow recordings, described earlier, also are of some help in the differential diagnosis. It has been suggested that tissue Doppler echocardiographic analysis of the mitral annulus, interventricular septum, and posterior LV wall can be useful in differentiating the two conditions. While mitral flow pattern could be similar in both amyloidosis and constrictive pericarditis, mitral annular velocities appear to be diminished in cases of restriction [27]. The precise role of tissue Doppler echocardiography is yet to be defined.

Tumors and Cysts

Pericardial tumors are a rare occurrence. They can be either primary pericardial tumors, or a result of metastatic extension of breast or lung carcinomas, leukemias, or lymphomas. The most common primary pericardial tumor is mesothelioma. Other pericardial tumors include malignant fibrosarcomas, teratomas, and angiosarcomas. Transthoracic and transesophageal echocardiography reveals them as echo-dense mass or masses involving the pericar-

dium, pericardial cavity, and adjacent regions. They are also invariably associated with hemorrhagic effusions. MRI and CT are helpful in localizing the extent of these tumors. Pericardial cysts are echo-free, localized, usually spherical or oval structures, contiguous with the pericardium. They are usually detected on chest radiographs incidentally as masses in the right costophrenic angle, left costophrenic angle, hilum, or superior mediastinum. 2DE helps distinguish between these cystic structures from solid tumors. MRI and CT scans also help confirm the diagnosis.

Absent Pericardium

The pericardium may be completely or partially absent in a patient. This syndrome is more commonly seen in men, and may be associated with other congenital anomalies such as atrial septal defect, bicuspid aortic valve, tetralogy of Fallot, patent ductus arteriosus, hiatal hernia, and bronchogenic cysts. It may also be seen in cases of therapeutic removal or stripping of the pericardium. This syndrome hardly manifests clinically, and is usually detected by echocardiography as an incidental finding. In the complete absence of pericardium, echocardiography reveals exaggerated cardiac motion and apparent RV enlargement. In case of partial absence, the part of the chamber that is uncovered may herniate through the defect, giving the false impression of a wall motion abnormality [28].

Conclusions

In the current era, noninvasive cardiac imaging techniques have become the primary diagnostic modes in the evaluation of pericardial disorders. Pericardial effusion and cardiac tamponade are readily and reliably recognized by 2-D and Doppler echocardiography. When constrictive pericarditis is suspected, MRI, CT, and TEE are valuable in the assessment of a morphologic abnormality (*ie*, the thickened pericardium), and Doppler examination of various flow jets yields valuable information on the presence or absence of filling dysfunction associated with this condition. Careful analysis of 2-D and Doppler studies, coupled when necessary with CT or MRI, can almost always differentiate constriction from restrictive cardiomyopathy. Tissue Doppler echocardiography is a promising technique to unmask alterations in myocardial and hemodynamic function caused by pericardial and myocardial diseases. Pericardial cysts and tumors, and absence of the pericardium, are well delineated by CT and MRI. The spectrum of pathogenesis and pathophysiology of pericardial disorders has changed during the recent decades, and it is not uncommon to encounter patients with atypical clinical presentations. Careful interpretation of noninvasive studies, and consideration of the clinical context are essential in the optimal evaluation and management of patients with pericardial pathology.

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