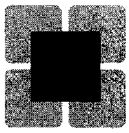

Pericardial disease: current diagnosis and management methods

WILLIAM A. SCHIAVONE, DO AND THOMAS W. RICE, MD

■ Diseases of the pericardium can be indolent or have a sudden onset. They may be primary, but more often are secondary to a systemic disease or previous therapy. Pericardial diseases imitate more common cardiac diseases and therefore can be difficult to diagnose. This paper's four case presentations illustrate use of recently developed methods that facilitate diagnosis. Magnetic resonance imaging is useful for the diagnosis of congenital absence of the pericardium. Echocardiography and computed tomography are useful for the diagnosis of pericardial cysts. Diagnosis of cardiac tamponade is aided by echocardiography, and Doppler echocardiography can help diagnose constrictive pericarditis. Effective management of pericardial diseases requires an understanding of the pathophysiology and natural history of each disease entity, knowledge of the individual patient, and realistic application of therapy.

□INDEX TERMS: CARDIAC TAMPONADE; MEDIASTINAL CYST; PERICARDITIS; PERICARDITIS, CONSTRICTIVE; PERICARDIUM □CLEVE CLIN J MED 1989; 56:639–645

DISEASES of the pericardium can present subtly, with minimal symptoms that become noticeable only after overcoming the compensatory mechanisms, or they can present suddenly, with disabling symptoms that can progress rapidly to death. Although some are primarily pericardial, most diseases of the pericardium are local expressions of systemic disease or distant malignancy.

Since diseases of the pericardium are relatively uncommon and may imitate more common ischemic, valvular, or myocardial diseases, they may be misdiagnosed. Acute pericarditis can mimic pulmonary embolism and can mimic or accompany myocardial infarction. Cardiac tamponade can resemble acute asthma, or can be silent and so gradual in onset that it is unrecognized until it is very severe. Constrictive pericarditis is usually so gradual in onset that antecedent cardiac disease is forgotten and the presenting signs and symptoms can be misdiagnosed as primary hepatic or pulmonary disease. Congenital diseases of the pericardium, partial absence of the pericardium, and pericardial cyst are most often silent and clinically unimportant. In this age of preventive medicine, however, they can be misdiagnosed on a screening chest radiograph as an intracardiac defect or shunt, or as a lung or mediastinal neoplasm, and lead to further evaluation and costly, potentially harmful diagnostic procedures.

In recent years several methods have been developed to facilitate prompt and accurate diagnosis and treatment of diseases of the pericardium. These methods must be used only if history and physical examination point to the pericardium as the likely source of signs and symptoms. One must maintain a level of suspicion that

From the Departments of Cardiology (W.A.S.) and Cardiothoracic Surgery (T.W.R.), The Cleveland Clinic Foundation. Submitted Sep 1988; accepted Feb 1989.

Address reprint requests to W.A.S., Cardiology Specialists, Inc., 3963 Loomis Parkway, Ravenna, Ohio 44266.

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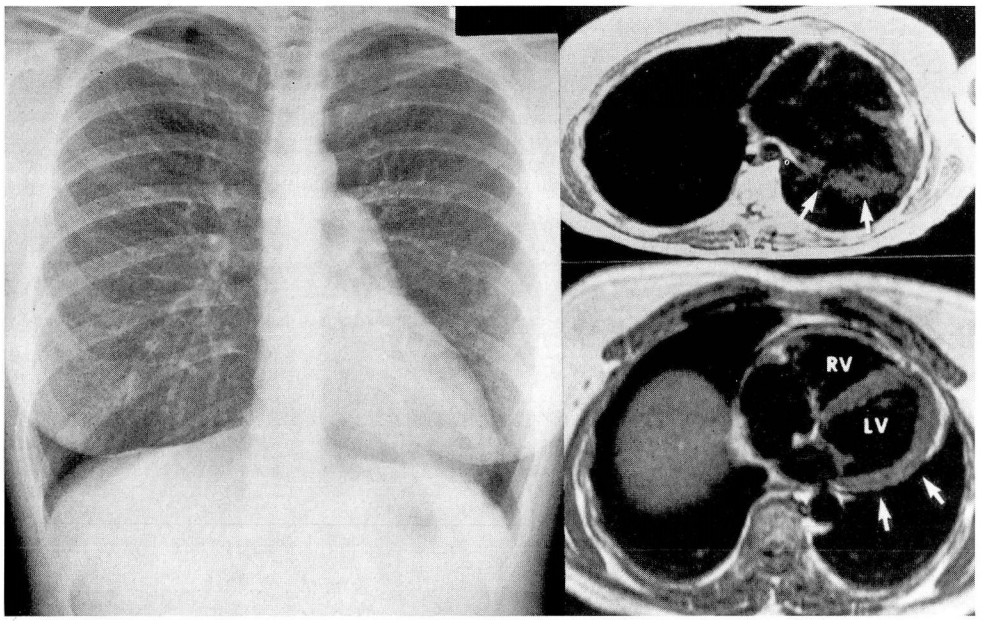

FIGURE 1A. Posteroanterior chest radiograph shows enlargement of the pulmonary artery segment and the heart shifted into the left hemithorax. FIGURE 1B. Transverse gated magnetic resonance image shows rotation of the heart into the left hemithorax and blurring of the posterior left ventricular border (arrows). FIGURE 1C. Transverse gated magnetic resonance image shows normal biventricular size and axis. The left posterior cardiac border is distinct (arrows). *RV*=right ventricle; *LV*=left ventricle. (Reproduced from Schiavone and O'Donnell[1] with permission.

permits consideration of diseases of the pericardium when other diagnostic avenues are not successful.

The following cases describe patients who presented with pericardial diseases that mimicked other diseases. These cases illustrate the application of current methods of diagnosing and managing pericardial diseases and permit discussion of the differential diagnosis and the treatment options.

CASE REPORTS

Case 1

A 35-year-old woman was evaluated by her internist for excessive fatigue. She was found to have mild hypothyroidism and was treated with levothyroxine. A grade I/VI systolic murmur was heard in the pulmonic area and a routine chest radiograph (*Figure 1A*) showed prominence of the pulmonary trunk with normal pulmonary vasculature. She was referred to a cardiologist to rule out valvular pulmonic stenosis or atrial septal defect. Echocardiography showed abnormal ventricular septal motion suggesting right ventricular volume overload. By Doppler echocardiography, however, there was no evidence of left to right shunting, tricuspid regurgitation, pulmonic regurgitation, or pulmonic stenosis. Right-sided heart catheterization showed normal pressures, no gradient across the pulmonic valve, and normal oxygen saturation without a step-up from venae cavae to the pulmonary arteries. Magnetic resonance imaging (MRI) (*Figure 1B*) showed the heart to be totally contained in the left hemithorax and the left and posterior cardiopulmonary interface was less distinct than normal (*Figure 1C*), due to complete absence of the left portion of the parietal pericardium.[1] Since the patient was asymptomatic, the defect was large (complete left pericardium), and the possibility of herniation, incarceration, and infarction was small, no treatment was necessary and close follow-up was undertaken.

Most patients with pericardial defects are asymptomatic and the defects are diagnosed at autopsy. They may complain of palpitations and chest pain. The use of MRI to demonstrate absence of the left portion of the parietal pericardium is less invasive than the previous method, which required creation of a left pneumothorax for radiographic demonstration of air outlining the right side of the heart and its pericardium. In this asymptomatic patient, the absence of a containing pericardium explains the shift of the heart into the left hemithorax and the apparent enlargement of the pulmonary trunk. This alteration of the pulmonary outflow tract produced the murmur heard in the pulmonic area.

More than 330 cases of congenital absence of the pericardium have been reported,[2] but only five deaths have been reported.[3,4] These defects were all single, left-sided, and partial. In these and other situations where herniation is possible, surgical intervention is indicated. Treatment has consisted of primary closure, pericardiectomy, left atrial appendectomy, and patch-closure with an autograft (fascia lata), xenograft (bovine pericardium), or synthetic materials (Dacron or Gortex).

Case 2

A 50-year-old woman was referred to a pulmonary physician for evaluation of a right cardiophrenic angle mass seen on chest radiograph (*Figure 2A*). She was asymptomatic. The physical examination was normal. Computed tomography (CT) further characterized the mass as homogeneous and of water density (*Figure 2B*). An echocardiogram, using the subcostal imaging plane (*Figure 2C*), demonstrated the mass at the right cardiophrenic angle to be a pericardial cyst since it was fluid-filled and did not communicate with a cardiac chamber. No treatment was necessary since the patient was asymptomatic and at follow-up the cyst had not enlarged.

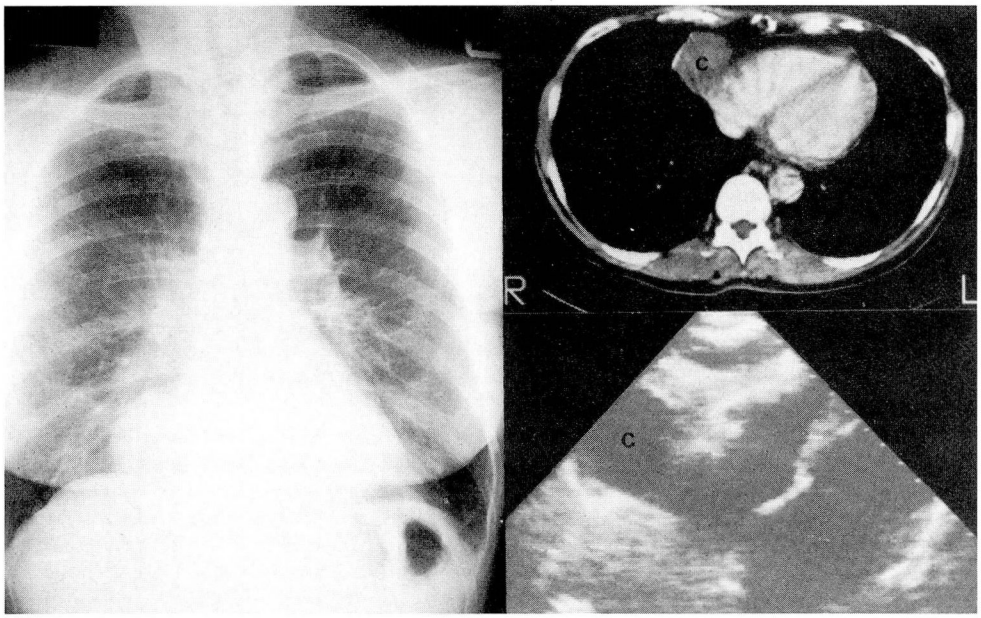

FIGURE 2A. Posteroanterior chest radiograph shows a right cardiophrenic angle mass. FIGURE 2B. Transverse CT scan shows the pericardial cyst (c) anterior to the heart in the right hemithorax. FIGURE 2C. Subcostal two-dimensional echocardiographic image shows the pericardial cyst (c) to be sonolucent and not to communicate with a cardiac chamber.

Pericardial cysts are asymptomatic in 70% of patients. In the other 30%, chest pain, dyspnea, cough, or palpitations may occur.[5] Rarely, these cysts may reach a size sufficient to displace the heart and the patient may present with heart failure.[6,7] Spontaneous regression or rupture is uncommon.[8] Ninety percent of pericardial cysts are found at the cardiophrenic angle. The balance are remote from this area and CT or echocardiography can be helpful in locating them.

The diagnosis can be confirmed and patients with symptomatic cysts who are poor surgical candidates can be treated with needle aspiration guided by CT or echocardiography. In our experience, mediastinal cysts drained in this manner tend to recur. The majority of pericardial cysts only require close follow-up; however, symptomatic cysts, cysts that increase in size, and those that evade diagnosis should be managed surgically.

Case 3

A 38-year-old man with an ascending aortic aneurysm and severe aortic insufficiency underwent replacement of the aortic root, aortic valve, and proximal ascending aorta with a St. Jude-valved conduit, and the coronary arteries were reimplanted. He had an uneventful postoperative recovery and received warfarin for anticoagulation. Prior to his aortic surgery, he had had several episodes of idiopathic acute pericarditis, which had required at least 7.5 mg of prednisone daily for relief. One month after aortic surgery he presented to the emergency room with dyspnea, cyanosis, and a 20-lb (9-kg) weight gain. His blood pressure was 110/90 mmHg with 20 mmHg of pulsus paradoxus and a central venous pressure of 25 mmHg. The pulse was 120 beats per minute and regular. He was taking 7.5 mg of prednisone and 5 mg of warfarin daily. The prothrombin time was 20 sec (control 12–13 sec). An emergency echocardiogram showed a large circumferential pericardial effusion with M-mode (*Figure 3A*) and two-dimensional echocardiographic signs of cardiac tamponade (*Figure 3B*). Pericardiocentesis was performed using a current method of echocardiographic guidance,[9] with needle insertion at the left sixth intercostal space in the anterior axillary line. Hepatic engorgement, clearly demonstrated echocardiographically, made the subxiphoid approach undesirably hazardous in a patient with a prolonged prothrombin time. Echocardiography also defined the nature of the pericardial fluid as circumferential, nonloculated, and without thrombus, tumor masses, or adhesions; therefore it was considered likely to be drainable with a needle and an indwelling catheter. The procedure was monitored and recorded echocardiographically using a sterile sleeve surrounding a

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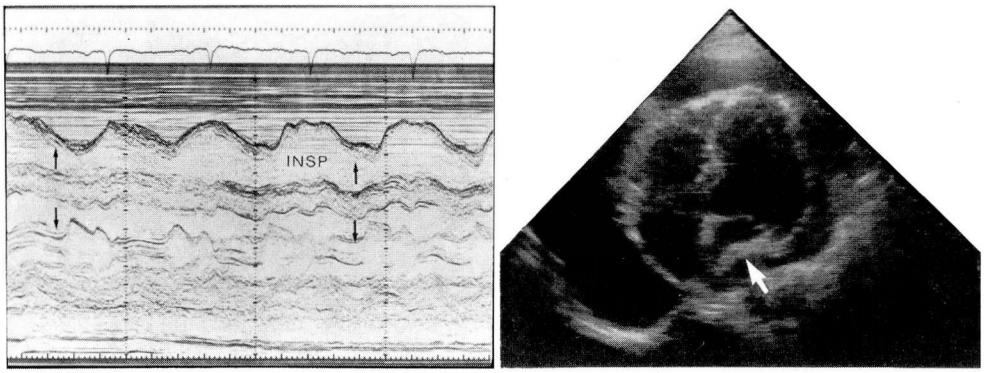

FIGURE 3A. M-mode echocardiographic signs of cardiac tamponade are displayed. Arrows are located in the right ventricle and the left ventricular outflow tract. Each arrow is the same size. With inspiration (*INSP*) right ventricular filling is increased at the expense of left ventricular filling. Notice that when the right ventricle is less well filled the left ventricular outflow tract is larger. This is the M-mode echocardiographic sign of pulsus paradoxus. FIGURE 3B. Apical four-chamber two-dimensional echocardiogram demonstrates left atrial collapse (arrow), which can be an early sign of cardiac tamponade.

2.5-MHz transducer from the left parasternal long axis image plane (*Figure 4*). The pericardiocentesis needle was shown to enter the pericardial space, injection of a saline solution provided contrast and verified its location in the pericardial space, and the guide wire was shown entering the pericardial space. Over the guide wire a multi-holed pericardial drainage catheter was placed in the pericardial space to drain the fluid totally.

In this case the fluid was serosanguinous, transudative, and stained negative for bacteria, mycobacteria, and fungi. The catheter continued to drain 1000 mL/day of serous fluid following complete fluid removal on the day of pericardiocentesis. With no evidence of infection, and with the history of recurrent pericarditis, postpericardiotomy syndrome was diagnosed. The prednisone dose was increased to 60 mg/day in a single morning dose, but no effect was noted. Only when the prednisone dose was divided into 20 mg t.i.d. did the pericardial fluid production cease. Five days postpericardiocentesis the drainage tube was removed. After discharge, the prednisone dose was gradually decreased, repeat echocardiography showed that the fluid did not reaccumulate, and the patient returned to work in good health one month after the pericardiocentesis.

Echocardiography has become the standard imaging technique used to confirm the clinical suspicion of cardiac tamponade. The most sensitive but least specific echocardiographic sign of cardiac tamponade is atrial collapse seen by two-dimensional echocardiography. The most specific signs of cardiac tamponade are the two-dimensional echocardiographic and M-mode echocardiographic demonstration of right ventricular diastolic collapse and left ventricular outflow tract narrowing that accompanies the augmentation in right ventricular filling during inspiration.

Surgical management of effusive pericarditis is indicated when medical management fails, such as when anti-inflammatory drugs do not control acute pericarditis or dialysis fails in uremic pericarditis, when cardiac tamponade occurs, or when there is metastatic involvement of the pericardium in a patient with a good life expectancy. Surgical management requires an adequate pericardiostomy (pericardial window) that will allow prolonged drainage into an adjacent body cavity. The window should be large enough to allow unobstructed drainage and to prevent early closure. There are three surgical approaches: thoracotomy, median sternotomy, and subxiphoid.

A left thoracotomy, either anterior or posterolateral (a muscle-sparing posterolateral incision is favored because it provides the greatest exposure and thus the largest window with minimal trauma), is chosen when the pleural space is the most favorable cavity into which to drain. This is generally used in patients with ascites, previous upper abdominal surgery, or in patients with uremic pericarditis where the peritoneal cavity may be used for dialysis. Left thoracotomy also allows an effective pericardiectomy over the left and right ventricles when there is constrictive pericarditis that predominantly involves these chambers.

Median sternotomy is chosen if there is a severe degree of constrictive pericarditis requiring a complete anterior pericardiectomy. This approach is also indicated when concomitant cardiac surgery is required.

The subxiphoid approach may be used with local anesthesia, but general anesthesia is preferred. The xiphoid is not excised and the anterior mediastinum is not approached, but rather a large window (4–6 cm diameter) is constructed through the pericardial portion of the diaphragm. The subxiphoid approach allows only limited inspection of the pericardium and the cardiac surface. The introduction of a mediastinoscope or sterile fiberop-

tic bronchoscope through the window will allow more complete inspection of the pericardium and facilitate distant pericardial biopsy.[10,11] Constrictive pericarditis is not well treated or examined by this approach.

Pericardiocentesis may be used preoperatively to prepare the patient for intended surgery. The improvement in hemodynamics will be a predictor of successful surgical results and can allow anesthesia and surgery to proceed with less risk. This procedure is best used for patients with free pericardial fluid that is not loculated or clotted (which may sometimes look like homogeneous, free fluid) in whom there is no chance of lacerating coronary bypass grafts and in whom there is enough fluid to permit safe needle insertion without cardiac laceration (at least 1.5 cm circumferentially when done electively).

Pericardiocentesis also allows sampling of fluid for chemical, microbiological, and cytological analysis in preparation for therapy. This latter therapy can be as diverse as antibiotic therapy for bacterial infection, antituberculous therapy for mycobacterial infection, surgical thoracic duct ligation for a traumatic pericardial-thoracic duct fistula with chylous pericardial effusion, or surgical pericardiostomy or chemical pericardiodesis (pericardial sclerosis) for metastatic malignant pericardial effusion.

Intrapericardial instillation of tetracycline following pericardiocentesis and catheter placement is a current method of palliative treatment for malignant cardiac tamponade.[12] In patients whose tumors are not likely to respond to chemotherapy or radiation therapy, it effectively prevents reaccumulation or reduces the rate of reaccumulation of pericardial fluid in approximately 80% of patients treated.[13] If the patient is premedicated with a narcotic analgesic and lidocaine is mixed with the sclerosing agent, this painful procedure can be performed with less discomfort. When a single intrapericardial instillation of a sclerosing agent is successful, it usually requires a shorter hospital recovery time than formal surgical pericardiostomy. If repeated instillations are required, the length of the hospital stay approaches that for surgical pericardiostomy. For this reason pericardial sclerosis is used for patients with terminal disease and the number of attempts at sclerosis is limited to one. Patients who have an expected survival of greater than two months are better served by a surgical pericardiostomy.

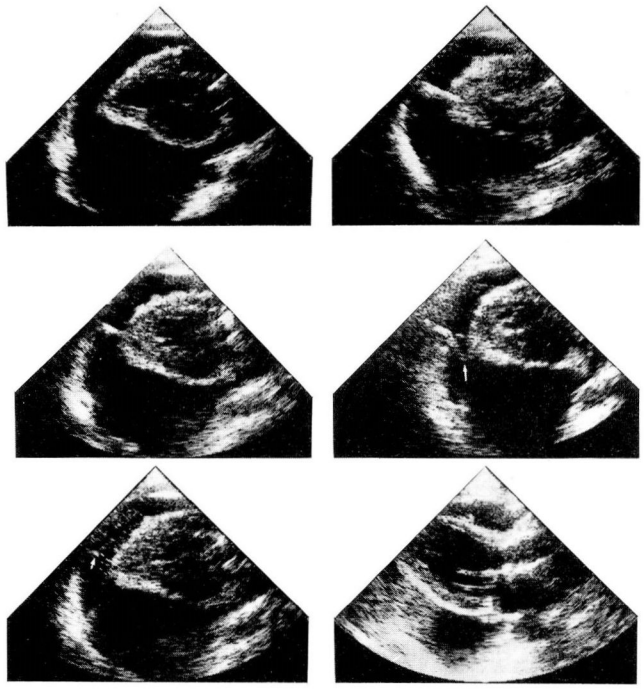

FIGURE 4. Parasternal long-axis two-dimensional echocardiographic images show a large pericardial effusion with the pericardiocentesis needle inserted (center left), injection of contrast material to verify location (arrow, lower left), guide wire advanced into the pericardial space (upper right), and drainage catheter placement (arrow, center right) to drain the fluid nearly totally.

Case 4

A 44-year-old man with abdominal fullness and elevated alkaline phosphatase levels was referred to a gastroenterologist by an internist. Because of increased jugular venous pressure, a cardiologist was consulted. The patient's exercise tolerance was decreased and he had increased abdominal girth and mild peripheral edema. He had had a nonspecific viral illness nine months prior to his visit to the cardiologist. Although his viral illness had been transiently disabling, he did not mention this illness until directly questioned since he made no association between it and his present illness. His blood pressure was 120/90 mmHg and pulse was 100 beats per minute and regular. The lungs were clear to auscultation, but the heart examination showed an early diastolic high-pitched filling sound. The electrocardiogram showed nonspecific ST-segment and T-wave changes. The chest radiograph showed a normal cardiac silhouette with mild blunting of the right costophrenic angle. An M-mode echocardiogram (*Figure 5*) showed a thickened pericardium, abnormal ventricular septal motion, and a brisk halt to diastolic filling of the ventricles. Cardiac catheterization showed equal di-

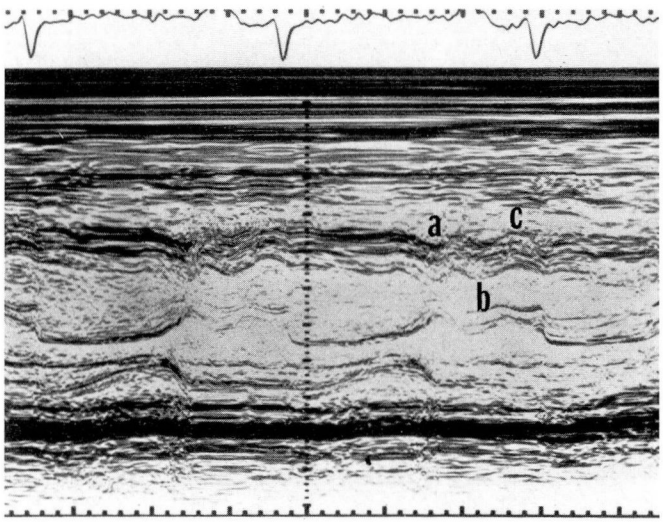

FIGURE 5. M-mode echocardiogram demonstrates the echocardiographic features of constrictive pericarditis. The posterior pericardium is thickened. There is abrupt halt to diastolic filling with a flat posterior left ventricular wall and abnormal septal motion. The early systolic notch (*a*) coincides with the deep Y descent in the jugular venous tracing. The second posterior displacement in the ventricular septum (*b*) occurs between *a* and *c*. Point *c* shows a brisk anterior motion of the ventricular septum during atrial mechanical systole that occurs prior to the electrocardiographic QRS segment.

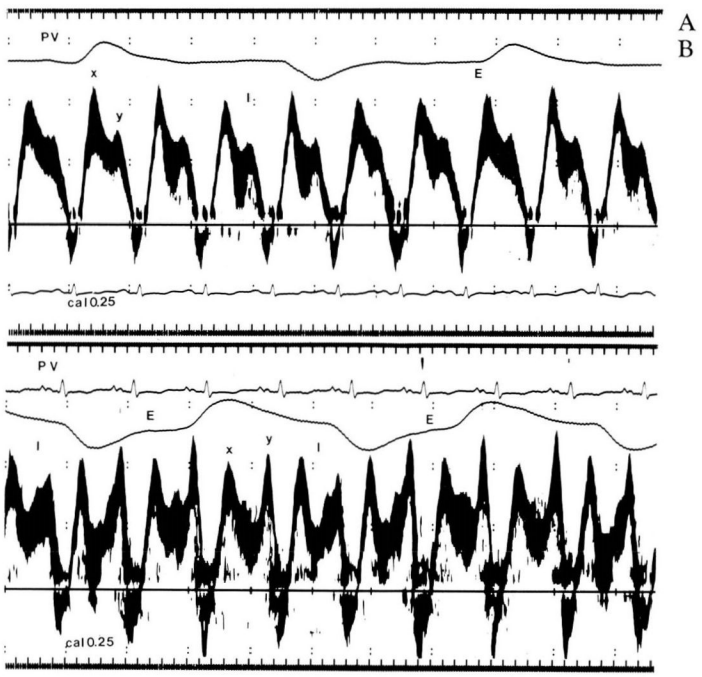

FIGURE 6A. Pulsed-wave Doppler recording of pulmonary venous flow velocity in patient 4 with constrictive pericarditis, obtained using a transesophageal transducer. It demonstrates systolic (*x*) flow velocity greater than diastolic (*y*) flow velocity. During inspiration (*I*), pulmonary venous flow is attenuated and during expiration (*E*), both *x* and *y* are augmented. FIGURE 6B. Pulmonary venous pulsed-wave Doppler recording, obtained in the same manner in patient 4 following pericardiectomy. Diastolic flow velocity (*y*) is greatly increased.

astolic pressures of 24 mmHg on the right and left sides of the heart. There were deep X and Y waves in the right atrium and the square root sign was present in the right ventricle. Constrictive pericarditis was diagnosed and pericardiectomy was successfully performed. He returned to active life one month following surgery.

A study of pulmonary venous return to the left atrium using a new application of transesophageal echocardiography was conducted in this patient prior to and following complete pericardiectomy. Color flow Doppler transesophageal echocardiography allowed precise pulsed-wave Doppler recording of pulmonary venous flow. Respiration was monitored using nasal capnography (measurement of nasal CO_2) simultaneously with Doppler recording of pulmonary venous flow.

Like the jugular venous pulse tracing, Doppler-echocardiographically recorded pulmonary venous flow is characterized by systolic (X) and diastolic (Y) components that represent pulmonary venous return to the left atrium. Inscription above the baseline represents antegrade flow toward the left atrium. In patient 4, with constrictive pericarditis, during inspiration there is an overall decrease in pulmonary venous flow velocity, and systolic flow velocity (X) is higher than diastolic flow velocity (Y) (*Figure* 6A). In this same patient following pericardiectomy (*Figure* 6B), pulmonary venous diastolic flow velocity (Y) is greater than systolic flow velocity (X), especially during expiration. These patterns of pulmonary venous flow velocity and their changes with respiration seen in constrictive pericarditis are clearly different from those found in the normal heart in which X and Y are relatively equal and respiratory variation is negligible.

The theory explaining these alterations in pulmonary venous flow in constrictive pericarditis is that the heart is encased in a rigid shell that insulates it from the intrathoracic pressure changes in respiration. With inspiration the intrathoracic pressure decreases but the intracardiac pressure remains high. This inspiratory decrease in intrathoracic pressure (and thus pulmonary venous pressure) causes pulmonary venous blood to well up in situ rather than to return to the higher-pressure

left atrium. The opposite occurs during expiration, as the expiratory increase in intrathoracic pressure forces pulmonary venous blood toward the left atrium, whose pressure has not changed or perhaps has decreased slightly due to decreased inspiratory left atrial filling. After the rigid constricting pericardium has been removed, the heart is no longer insulated from the intrathoracic pressure changes of respiration, and the pattern of pulmonary venous flow resembles that of the undiseased heart; however, the diastolic flow velocity (Y) is higher. Since constrictive pericarditis is a disease of diastolic filling, it is diastolic (Y) pulmonary venous flow that is most attenuated, and this same flow velocity that is most augmented following pericardiectomy.[14]

SUMMARY

Pericardial diseases, which are seldom considered but may masquerade as other diseases, must be included in the differential diagnosis of dyspnea and weight gain along with chest radiographic findings of an enlarging cardiac silhouette (pericardial effusion or cardiac tamponade) or a small cardiac silhouette (constrictive pericarditis). Heightened awareness of pericardial diseases is necessary, especially in view of longer survival of patients with cancer, renal failure, connective tissue diseases, and trauma. This longer survival is the result of earlier and more successful application of chemotherapy, radiation therapy, dialysis, and cardiac surgery.[15]

Diseases of the pericardium can be elucidated more effectively with such current diagnostic methods as CT, MRI, Doppler echocardiography, and transesophageal echocardiography and pericardioscopy. Once diagnosed, management of pericardial diseases, repair of congenitally absent pericardium, echocardiographically directed pericardiocentesis, chemical pericardiodesis, pericardiostomy, and complete pericardiectomy can be appropriately applied to afford greater comfort for the patient, or cure.

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