9 Pericardium and Pericardial Diseases

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9.1 Introduction

The normal pericardium is a thin, linear structure that echocardiographically is outlined as a bright, highly echogenic line. Within the two layers of the pericardium, the inner serosal layer and the outer fibrous pericardium, about 10–50 ml of pericardial fluid accumulates physiologically; it is generally visualized only in systole as an echo-free space surrounding the heart.

Transthoracic echocardiography is a milestone and the first-line imaging modality for diagnosing pericardial diseases such as pericardial effusion (PE) and cardiac tamponade, constrictive pericarditis, pericardial cysts and tumors, and partial and complete absence of the pericardium. The limitations of this imaging are very few, mainly represented by a poor echocardiographic window and the lack of details of the pericardium's anatomy, which need to be investigated with a further examination (CT or MRI). Although transthoracic echocardiography is almost always the ideal technique to detect and grade pericardial diseases, several factors in the period after cardiac surgery may contribute to the need for transesophageal echocardiography.

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9.2 Pericardial Effusion

The causes of PE range from inflammatory effusions (secondary to bacterial or viral infections, myocardial infarction, trauma, or neoplasm) to hemorrhagic effusions, mostly common in the period after cardiac surgery. The clinical signs and symptoms are related to the cause of the PE and to the hemodynamic significance of the effusion itself. If the PE is secondary to pericarditis, a prodrome of fever, malaise, and myalgia could precede major symptoms such as retrosternal or left precordial chest pain and shortness of breath. A pericardial friction rub could be present, although it could be transient, and the heart rate is usually rapid and regular. If the PE develops slowly, it can be remarkably asymptomatic, whereas rapidly accumulating effusions manifest themselves clinically with tamponade (see the next section). Echocardiography should be the first modality to determine the size and the hemodynamic significance of PE, as it is an even more accurate imaging technique than CT in quantitative assessment of nonloculated PE, always keeping in mind the few limitations that this examination has. The hemodynamic consequences of PE will be examined in the next section. In this section we will examine the classification and the characteristics of PE.

The size of the effusion has been graded by the European Society of Cardiology as small, moderate, large, and very large (Table 9.1). PE may be seen not only in systole (as it is common for the physiological pericardial fluid), but they

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Table 9.1 Classification of pericardial effusion by the European Society of Cardiology

Classification of pericardial effusion	Dimension in diastole (mm)	Location
Small	<10	Posterior atrioventricular groove
Moderate	10–20	
Large	>20	Usually extends behind the left atrium, may determine a compression of the heart
Very large	>20	Extends behind the left atrium and determines a compression of the heart

may become visible throughout the entire cardiac cycle. As the amount of pericardial fluid increases, fluid moves from the posterobasilar left ventricle to the apex and anterior wall and then laterally and posteriorly to the left atrium (Fig. 9.1). It is important to emphasize that the mere presence of an effusion, even when large, does not indicate hemodynamic significance. Since the rapidity of pericardial fluid accumulation and the compliance of the pericardium influence the pressure elevation for any given fluid volume, effusion volume alone does not determine hemodynamic significance. Therefore, the presence of an effusion must be related to other echocardiographic parameters of cardiac filling and transvalvular flow and must be correlated with the clinical features (see the next section).

Although PE generally appears as an echo-free space encircling the heart, sometimes echogenic materials such as fibrinous strands and a shaggy exudative coating are found. These findings are important as well, because the initial echocardiographic characteristics of PE determine the pericardial complication. More precisely, echogenic PE, including an exudative frond-like coating and fibrinous strands in PE, are the major risk factors for pericardial complications such as constrictive pericarditis and PE recurrence. Diffuse echogenic PE results in the highest incidence of events, followed by PE with an exudative coating, and fibrinous strands. Echocardiography could, then, be used not only for diagnostic purposes, but also for a prognostic evaluation.

PE are a possible complication of cardiac surgery, with a reported incidence that ranges between 50 and 64 % of cases depending on the study definitions and designs. They compromise cardiac function in 0.8–7 % of cases and have a peak incidence on the tenth postoperative day. Echocardiographic monitoring of surgical patients should be, therefore, done up to 20 days to 1 month after surgical intervention, because late PE are an important cause of morbidity. Persisting PE is more frequent after coronary artery bypass graft surgery than after valve replacement surgery. In contrast, the incidence of late cardiac tamponade is higher in patients undergoing valve replacement surgery. The use of transthoracic echocardiography is important not only for the follow-up of cardiac surgical patients, but it has also been validated for the classification of postoperative PE for predicting late postoperative cardiac tamponade. Indeed, the incidence of late cardiac tamponade is significantly increased in patients with a loculated effusion larger than 15 mm or a circumferential effusion larger than 10 mm on transthoracic echocardiography on postoperative day 20.

The risk of PE has to be considered not only in the adult population, but also after congenital cardiac surgery. Serial echocardiographic monitoring up to 28 days postoperatively is indicated in selected high-risk patients such as those with symptoms of postpericardiotomy syndrome and those given warfarin. Interestingly, PE that eventually becomes moderate to large tends to occur later and occurs more commonly after Fontan-type procedures.

Although transthoracic echocardiography is almost always the ideal technique to detect and grade PE, several factors in the postoperative

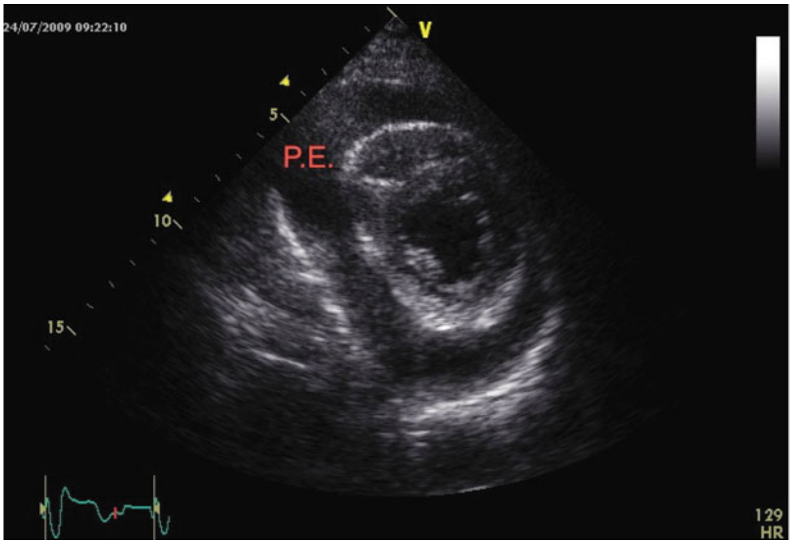

Fig. 9.1 Transesophageal echocardiography transgastric short-axis view of the left ventricle showing a circumferential large pericardial effusion (*P.E.*)

patient after cardiac surgery may contribute to the need for transesophageal echocardiography: the surgical site may preclude the use of the optimal transthoracic window, chest tubes may prevent proper positioning of the patient, and some loculated effusions or intrapericardial clots may not be amenable to transthoracic imaging.

As a final consideration, the differential diagnosis of echo-free spaces should include PE, pleural effusions, and pericardial fat. As a rule, in the different views pericardial fluid reflects at the posterior atrioventricular groove, whereas PE continues under the left atrium, posterior to the descending aorta. Pericardial fat can be identified as the hypoechoic space anterior to the epicardial fat; it is more prominent anteriorly, but may appear circumferentially, thus mimicking effusion. Pericardial fat is slightly echogenic and tends to move with the heart; in contrast, the effusion is generally echolucent and motionless.

Echocardiography is very important not only for diagnostic purposes, but also as a guide for percutaneous needle pericardiocentesis: it has an excellent profile in terms of simplicity, safety, and efficacy. Echocardiography identifies the shortest route by which the pericardium can be entered intercostally, allows clear localization of the needle, and shows the immediate benefit of the removal of the excess of pericardial fluid. Finally, the approach to the PE should be selected according to the distribution of the PE on echocardiography. If the effusion is equally large in the apical position and in front of the right ventricle from the subxiphoid view, both an apical and a subxiphoid approach can be attempted, according to the operator's preference. However, if the effusion is significantly asymmetrically distributed, it should be approached from the side where the accumulation of fluid is largest. The reported incidence of major complication ranges from 1.3 to 1.6 %.

9.3 Cardiac Tamponade

When the accumulation of pericardial fluid causes an increase in the pericardial pressure exceeding the intracardiac pressure, the positive transmural pressure gradient compresses cardiac chambers at different points in the cardiac cycle, compromising cardiac filling. Depending on the type and the severity of tamponade, a variety of physical findings may be present: chest pain radiating to the neck and jaw, orthopnea, cough, and dysphagia. The jugular venous pressure is elevated and there is commonly an exaggeration of the normal variation in the pulse pressure during the inspiratory phase of respiration greater than 10 mmHg (pulsus paradoxus) as well as an elevation in the venous jugular pressure during inspiration (Kussmaul sign). On chest radiography, large effusions are depicted as globular cardiomegaly with sharp margins ("water bottle" silhouette). Electrocardiography may demonstrate diminished QRS and T-wave voltages, PR-segment depression, ST-T changes, bundle-branch block, and electrical alternans. Patients after cardiac surgery present a much more specific diagnostic challenge, since effusions may be localized, underlying cardiac disease is present, and positive pressure ventilation is used, all factors likely to alter the classic clinical findings.

Echocardiography is a powerful tool to quickly identify the hemodynamic significance of the PE and the cardiac tamponade. The basic elements that allow this identification are as follows:

- The low pressure of the right chambers makes them the first structures susceptible to the increased transmural pressure. Right atrial wall inversion during ventricular systole is usually an early sign of cardiac tamponade, followed by diastolic compression of the right ventricular outflow tract (evaluated by both M-mode and 2D echocardiography) (Fig. 9.2). The longer is the duration of the right atrial invagination relative to the length of the cardiac cycle, the greater is the likelihood of significant hemodynamic compromise: a duration of right atrial collapse exceeding one third of the cardiac cycle increases specificity without sacrificing sensitivity. Right chamber collapse may be delayed in the setting of pulmonary hypertension; in such cases, left atrial collapse may precede right atrial collapse.
- "Swinging heart": when a large PE accumulates, the heart will be swinging in the pericardial fluid beat-to-beat.
- Ventricular interdependence: during inspiration the interventricular septum bulges into the left ventricle owing to increased systemic venous return to the right ventricle and limited expansion of the free wall of the right ventricle.
- Respiratory variation in tricuspid and pulmonary flow detected with Doppler echocardiography. Tricuspid flow increases and mitral flow decreases during inspiration: the respiratory variations in mitral inflow of more than 35% and tricuspid inflow of more than 40% correlate well with cardiac tamponade. These respiratory variations cannot be seen in the invasively ventilated patient.
- Plethora of the inferior vena cava: there is a lack of change in vena cava caliper (less than 50% reduction in diameter) during inspiration.
- Prominence of diastolic reversals in hepatic veins by pulsed Doppler echocardiography: with expiration, systemic venous return decreases with reversal of diastolic flow in the hepatic veins.

9.4 Constrictive Pericarditis

Chronic inflammation of the pericardium results in thickening fibrosis and fusion of both layers of the pericardium. Constrictive pericarditis represents the end stage of this chronic inflammatory process leading to a limit in diastolic filling and resulting in diastolic failure, with relatively preserved global systolic function. The most common symptoms are related to either fluid overload (peripheral edema, elevated central venous pressure, hepatomegaly, pleural effusion, ascites, and anasarca) or decreased cardiac output (dyspnea, fatigue, palpitations, weakness, and exercise intolerance). An important reason to use echocardiography early in the diagnostic process is to rule out more common causes of right-sided heart failure, including left or right ventricular systolic dysfunction, severe pulmonary hypertension, or unrecognized left-sided valvular diseases. Once these diseases have been excluded, echocardiography is very useful in recognizing the pericardial thickening and the elements that characterize constrictive pericarditis. Visualization of the pericardium thickening (Fig. 9.3) could be done in a more accurate way with transoesophageal echocardiography, with a significant cutoff value of 3 mm.

The echocardiographic characteristics of constrictive pericarditis are secondary to the impaired diastolic cardiac filling and elevated ventricular filling pressures:

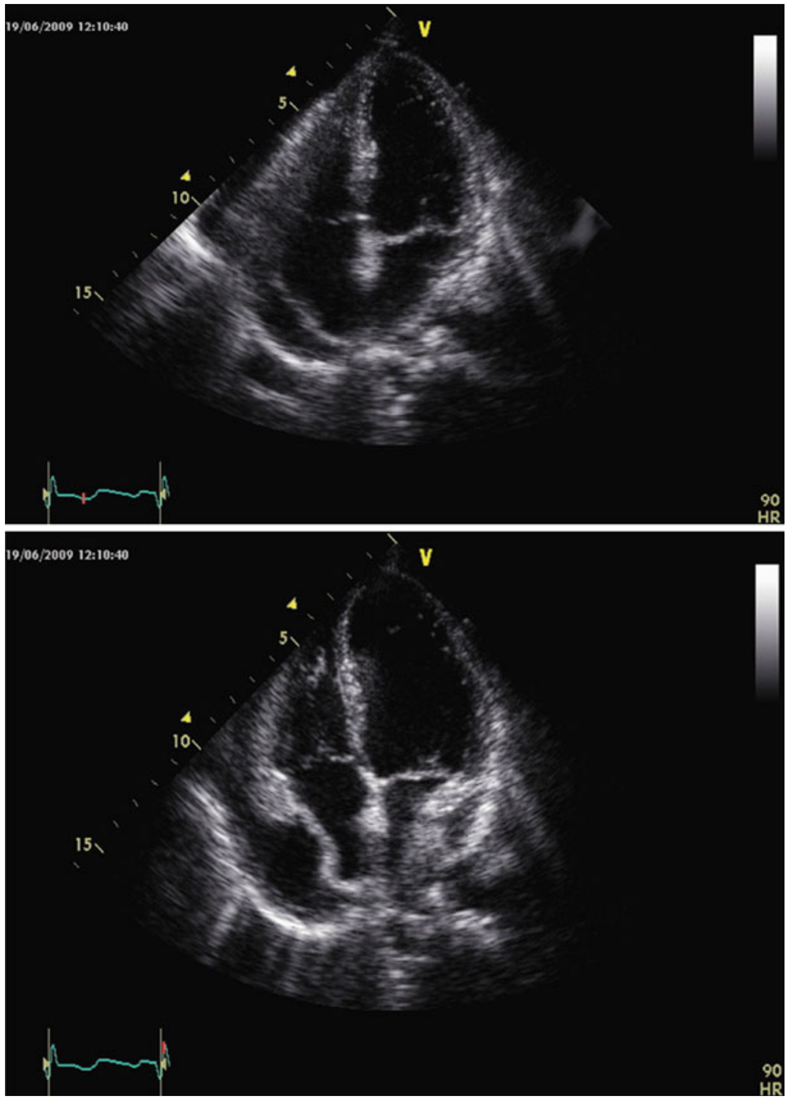

Fig. 9.2 Transthoracic echocardiography apical four-chamber view showing a large pericardial effusion with right atrial wall inversion during ventricular systole (*bottom*)

- Mitral inflow assessed by Doppler echocardiography demonstrates a rapid increase in ventricular diastolic pressure that creates the dip-and-plateau pattern with an increased early diastolic filling (E-wave) velocity with a rapid deceleration time and a small or absent A wave.
- Marked respiratory variation in left and right ventricular inflow velocities are seen with Doppler echocardiography. There is an increase in early diastolic mitral inflow of more than 25% during expiration. After complete pericardiectomy, mitral inflow patterns return to normal, and little respiratory variation is seen.
- Tissue Doppler imaging of the mitral annulus shows a prominent early diastolic velocity (Ea). A lateral or septal early diastolic mitral annular velocity of more than 8 cm/s on pulsed tissue Doppler imaging is in general the accepted cutoff value to diagnose constrictive pericarditis. Mitral annular velocities are particularly useful when pronounced respiratory variations in peak early mitral inflow velocities are not seen.
- There is increased ventricular interdependence with a classic respiratory shift in the position of the interventricular septum

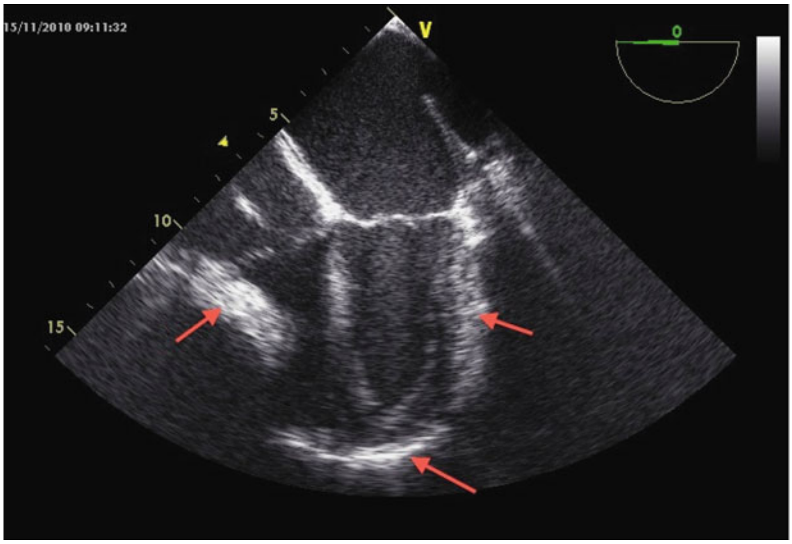

Fig. 9.3 Transesophageal echocardiography mid-esophageal four-chamber view showing pericardial thickening and calcification in constrictive pericarditis

towards the left ventricle during inspiration ("septal bounce").

- The flow propagation velocity into the left ventricle detected by color Doppler M-mode echocardiography is greater than 45 cm/s.
- Marked diastolic flow reversal that increases in expiration is evident in the hepatic veins.
- There is rapid flattening of the posterior wall of the left ventricle in early diastole with normal or exaggerated longitudinal deformation of the left ventricle.

The above criteria are important not only to diagnose constrictive pericarditis, but they also help to distinguish it from restrictive cardiomyopathy (see Chap. 13). Doppler echocardiographic techniques, in particular, have been shown to be useful in differentiating between these two diseases: a marked respiratory variation in mitral inflow and pulmonary venous flow is present in patients with constrictive pericarditis but is absent in those with restrictive cardiomyopathy. Recently, the newer echocardiographic modalities of tissue Doppler imaging and color M-mode flow propagation have been validated as ancillary tools to distinguish between these diseases, although they are equivalent and complementary to Doppler respiratory variation in distinguishing between constrictive pericarditis and restrictive cardiomyopathy. The additive role of the new methods needs to be established in difficult cases of constrictive pericarditis where respiratory variation may be absent or decreased. Of the two newer modalities, tissue Doppler imaging of the mitral annulus tends to have greater specificity and sensitivity than color M-mode flow propagation and is generally easier to use. As the velocity of propagation of early ventricular inflow from color M-mode echocardiography and the early mitral annular velocity from tissue Doppler imaging are markers of myocardial relaxation, their values are generally normal or supranormal in pure constrictive pericarditis, in which myocardial relaxation is normal or raised. By contrast, these values are decreased in restrictive cardiomyopathy, in which myocardial relaxation is impaired.

Patients with chronic obstructive pulmonary diseases or severe right ventricular dysfunction and large respiratory variations in intrathoracic pressure may also show inspiratory decreases in early mitral inflow velocities, such as in constrictive pericarditis. To distinguish these two conditions, the intensivist should consider that in chronic obstructive pulmonary disease there is a greater decrease in intrathoracic pressure in inspiration, which generates greater negative pressure changes in the thoracic cavity, and augments blood flow to the right atrium from the superior vena cava during inspiration.

9.5 Effusive-Constrictive Pericarditis

The features of cardiac tamponade and constrictive pericarditis may combine and cause effusive-constrictive pericarditis. These rare and particular cases should be diagnosed not only with the important help of echocardiography, but also taking into consideration hemodynamic variations before and after the removal of the excess pericardial fluid. The clinical diagnosis of this condition is based on the demonstration that in a patient with PE and tamponade a clinical and hemodynamic picture consistent with pericardial constriction persists after the removal of enough pericardial fluid to lower the intrapericardial pressure to the normal level.

9.6 Pericardial Masses

Primary pericardial tumors are rare; secondary tumors are far most common. Pericardial masses are often detected incidentally during routine echocardiography; although echocardiography should be considered important in the identification of these masses, CT or cardiac MRI should always be performed as they are the imaging modalities of choice when further evaluating these tumors.

9.7 Pericardial Cysts

Pericardial cysts are rare, benign congenital or inflammatory malformations; they can also be acquired after cardiothoracic surgery. They are usually found incidentally during routine X-ray or echocardiographic examinations and appear as echo-free fluid-filled loculated masses located mostly at the right costophrenic angle, and less frequently in the left costophrenic angle or in the posterior or anterior superior mediastinum. Color flow and pulsed Doppler interrogation at a low-velocity setting can be used to ensure there is no phasic flow within the structure and can be used to differentiate pericardial cysts from coronary aneurysm, left ventricular aneurysm, prominent left atrial appendage, aortic aneurysm, or solid tumors. Transesophageal echocardiography can be useful if transthoracic echocardiography is inadequate in delineating the diagnosis and can help to identify a pericardial cyst in atypical locations and distinguish it from other posteriorly located lesions.

9.8 Congenital Absence of the Pericardium

Congenital absence of the pericardium is a rare anomaly whose reported prevalence is 0.002–0.004 %. This defect can be partial or complete, mostly located on the left side of the heart and more frequently asymptomatic and detected incidentally. The foramen-type defects are the most dangerous subgroup of partial defects, because they can be fatal when they allow herniation of part of the heart. The absence of the pericardium results in an exaggerated cardiac motion, particularly of the posterior wall of the left ventricle. Traditional echocardiography shows prominence of the right-sided cardiac chambers and abnormal septal motion. If the right ventricle shifts to the left, its cavity may falsely appear enlarged. Finally, there is typically a displacement of the apical imaging window into the axilla and the atria appear compressed. An echocardiogram should be useful as well in the identification of the associated heart defects, such as atrial septal defects and bicuspid aortic valve.

Further Reading

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