Pericardium and Pericardial Disease

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10.1 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, accumulate physiologically about 10–50 ml of pericardial fluid, 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 like pericardial effusion 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 in case need to be investigated with further examination. CT scan and MRI provide a larger field of view, allowing the detection of loculated pericardial effusion and pericardial thickening and masses, as well as associated chest abnormalities. Although transthoracic echocardiography is almost always the ideal technique to detect and graduate pericardial diseases, several factors in the postoperative period after cardiac surgery may contribute to the need of transesophageal echocardiography.

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10.2 Pericardial Effusion

The etiology of pericardial effusions (PE) ranges from inflammatory effusions (secondary to bacterial or viral infections, myocardial infarction, trauma, or neoplasm) to hemorrhagic effusions, mostly common in the postoperative cardiac surgery setting. Different criteria are used to classify pericardial effusion: the onset (acute, subacute, chronic), the distribution (circumferential or loculated), the hemodynamic impact (none, cardiac tamponade, effusive-constrictive), the composition (exudates, transudate, blood), and especially the size. The clinical signs and symptoms are related to the etiology 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 like retrosternal or left precordial chest pain and shortness of breath. A pericardial friction rub could be present, though it could be transient, and heart rate is usually rapid and regular. If the PE develops slowly, it can be remarkably asymptomatic, while rapidly accumulating effusions manifest clinically with tamponade (see next paragraph).

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Echocardiography should be the first modality to determine the size and the hemodynamic significance of PE, as it is even a more accurate imaging technique than computed tomography in quantitative assessing of nonloculated PE, always keeping in mind the few limitations that this exam holds. The hemodynamic consequences of PE will be examined in the next paragraph. In this paragraph we will examine the classification and the characteristics of PE.

The size of the effusion has been graded by the European Society of Cardiology in small, moderate, large, and very large (Table 10.1). Pericardial effusions may be seen not only in systole (as it is common for the physiological pericardial fluid), but they become visible throughout the entire cardiac cycle. As the amount of pericardial fluid increases, fluid distributes from the posterobasilar left ventricle to the apex and anterior wall and then laterally and posteriorly to the left atrium (Fig. 10.1). 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 next paragraph).

Although PE generally appears as an echo-free space encircling the heart, sometimes echo-

Table 10.1 Classification of pericardial effusion by the European Society of Cardiology

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

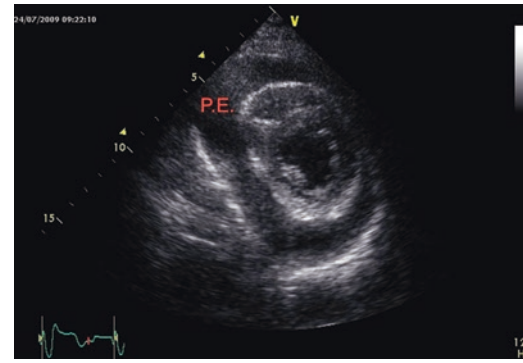

Fig. 10.1 A TEE TG short-axis left ventricle view showing a circumferential large pericardial effusion

genic materials such as fibrinous strands and shaggy exudative coating are found. These findings are important as well, because initial echocardiographic characteristics of PE determine the pericardial complication. More precisely, echogenic PE, including exudative frond-like coating and fibrinous strands in PE, are the major risk factor for pericardial complication such as constrictive pericarditis and PE recurrence. Diffuse echogenic PE results in the highest incidence of events, followed by PE with 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 study definitions and designs. They compromise cardiac function in 0.8–7% of cases and have a peak incidence on the 10th postoperative day. Echocardiographic monitoring of the surgical patients should be done up to 20 days–1 month after surgical intervention, because late pericardial effusions are an important cause of morbidity. Persisting PE is more frequent after undergoing CABG surgery than after undergoing valve replacement surgery, on the contrary, the incidence of late cardiac tamponade is higher in the patients undergoing valve replacement surgery. The use of TTE is important not only for the follow-up of the cardiac surgical patients, but it has been validated for the classification of postoperative pericardial effusion for predicting late postoperative cardiac tamponade. Indeed, the

incidence of late cardiac tamponade is significantly increased in patients with a TTE loculated effusion >15 mm or circumferential effusion >10 mm on postoperative day 20.

The risk of pericardial effusion 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 post-pericardiotomy syndrome and those given warfarin. Interestingly, PE that eventually becomes moderate to large in amount tends to present later and occurs more commonly after Fontan-type procedures.

Although transthoracic echocardiography is almost always the ideal technique to detect and graduate PE, several factors in the postoperative patient after cardiac surgery may contribute to the need of 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 pericardial effusions, pleural effusions, and pericardial fat. As a rule, in the different views, pericardial fluid reflects at the posterior atrioventricular groove, whereas pleural effusion 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; on the contrary the effusion is generally echolucent and motionless.

Echocardiography is very important not only for the diagnostic purposes but also as a guide of percutaneous needle pericardiocentesis: it has an excellent profile in simplicity, safety, and efficacy. Echocardiography identifies the shortest route where the pericardium can be entered intercostally, allows a clear localization of the needle, and shows the immediate benefit of the removal of the exceeding pericardial fluid. Finally, approach to pericardial effusion should be selected according to the distribution of pericardial effusion in echocardiography. If the effusion is equally large in the apical position and in front of the right ventricle from the subxiphoid view, both apical and 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%.

10.3 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; an exaggeration of the normal variation in the pulse pressure during the inspiratory phase of respiration greater than 10 mmHg is commonly found (pulsus paradoxus) as well as an elevation in the venous jugular pressure during inspiration (Kussmaul sign). In 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 provide a much more specific diagnostic challenge, since effusions may be localized, underlying cardiac pathology is present, and positive pressure ventilation is used, all factors likely to alter the classical clinical findings.

Echocardiography is a powerful tool to quickly identify the hemodynamic significance of the pericardial effusion and the cardiac

tamponade. The basic elements that allow this identification are the following:

- The low pressures of the right chambers make them the first structures susceptible to the increased transmural pressure. Right atrial wall inversion during ventricular systole (the indentation is seen near the peak of the R wave) is usually an early sign of cardiac tamponade, followed by diastolic compression of the RVOT (evaluated by both M-mode and 2D echocardiography) (Fig. 10.2). 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: 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 the right atrial one. False-positive right ventricle diastolic collapse may be present in hypovolemic states (because of the very low RV diastolic pressure) and in patients with large pleural effusions.
- "Swinging heart": when large pericardial effusion 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 due to increased systemic venous return to the right ventricle and limited expansion of the right ventricle free wall.
- Respiratory variation in tricuspid and pulmonary flow detected with Doppler echocardiography. Tricuspid flow increases and mitral flow decreases during inspiration: the percent respiratory variations in mitral inflow E-wave of >35% and tricuspid inflow E-wave of >40% correlate well with tamponade physiology. These respiratory variations could not be seen in the invasively ventilated patient.
- Plethora of the inferior vena cava: IVC diameter >21 mm and a lack of change in vena cava caliper (<50% reduction in diameter) during inspiration.
- Prominence of diastolic reversals in hepatic veins by pulsed Doppler: with expiration systemic venous return decreases with reversal diastolic flow in the hepatic veins.

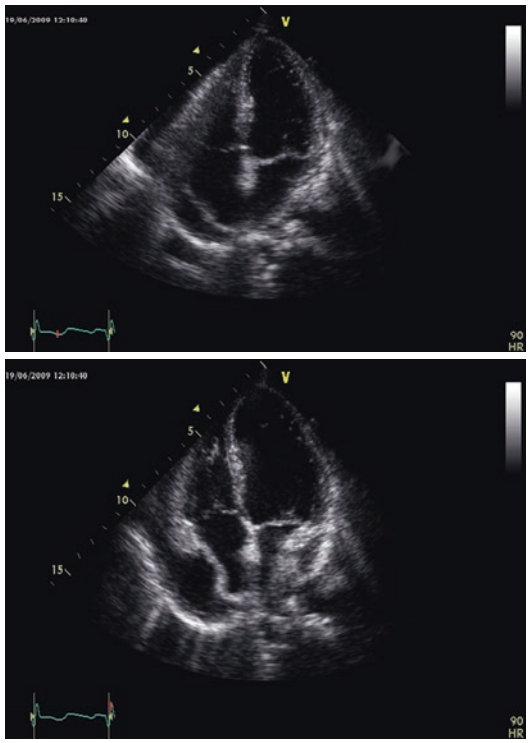

Fig. 10.2 TTE apical four-chamber view showing a large pericardial effusion with right atrial wall inversion during ventricular systole (on the bottom)

10.4 Constrictive Pericarditis

The most common reported causes of constrictive pericarditis in developed countries are idiopathic or viral (42–49%), post-cardiac surgery (11–37%), postradiation therapy (9–31%), connective tissue disorder (3–7%), post-infectious causes (3–6%), and miscellaneous causes (malignancy, trauma, drug-induced, asbestosis, sarcoidosis, uremic pericarditis in 10%). 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). The impaired ventricular filling determines particular patterns at the direct measurements of pressures: M- or W-shaped atrial pressure waveforms and "dip-and-plateau" right ventricular pressure waveforms. End-diastolic pressure equalization (typically within 5 mmHg) occurs between these cardiac chambers in constrictive pericarditis because of the fixed and limited space within the thickened and stiff pericardium. Pulmonary artery systolic pressures are usually normal in pericardial constriction; higher pulmonary pressures suggest a restrictive cardiomyopathy.

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 are excluded, echocardiography is very useful in recognizing the pericardial thickening and the elements that characterize constrictive pericarditis. Visualization of the pericardium thickening (Fig. 10.3) could be done in a more accurate way with transesophageal echocardiography, with a significant cutoff value of 3 mm.

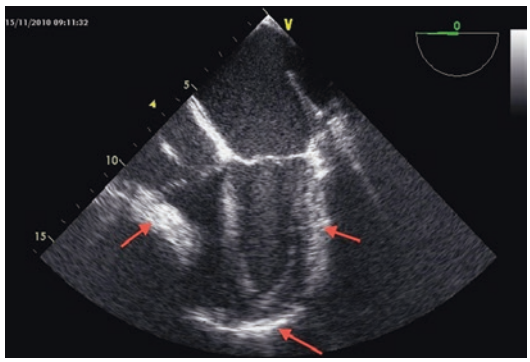

Fig. 10.3 TEE ME four-chamber view showing pericardial thickening and calcification in constrictive pericarditis

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

- 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 is seen with Doppler echocardiography. There is an increase in early diastolic mitral inflow of >25% during expiration and an increase of the tricuspid inflow velocity by at least 40% with inspiration. After complete pericardiectomy mitral inflow patterns return to normal, and little respiratory variation is seen.
- Tissue Doppler (TDI) of the mitral annulus shows a prominent early diastolic velocity. A lateral or septal early diastolic mitral annular velocity of >8 cm/s on pulsed tissue Doppler 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. The tethering of the lateral annulus secondary to the constrictive process causes the "annulus reversus" phenomenon: the medial mitral e' velocity becomes equal to or greater than the lateral e' velocity on average, in contrast to what is seen in other forms of heart failure and in the absence of cardiac disease.
- Increased ventricular interdependence with a classic respiratory shift in the position of the interventricular septum toward the left ventricle during inspiration ("septal bounce"). Beyond the respirophasic motion, a septal "bounce," also referred to as a "shudder" or "diastolic checking," may be present. The mechanism of this "bounce" is a combination of differential timing of filling and pericardial constraint, leading to rapid cessation of filling.

- Flow propagation velocity into the left ventricle detected by color-Doppler M-mode is greater than 45 cm/s.
- Marked diastolic flow reversal that increases in expiration is evident in the hepatic veins.
- The evaluation with the two-dimensional speckle tracking echocardiography shows a rapid flattening of the posterior wall of the left ventricle in early diastole with normal or exaggerated longitudinal deformation of the left ventricle. In general, circumferential strain, torsion, and early diastolic twisting are decreased in constrictive pericarditis. In contrast, global longitudinal strain is usually preserved. Patients with restrictive cardiomyopathy can demonstrate the converse pattern: global longitudinal strain is decreased, while circumferential strain and early diastolic untwisting are preserved.

The above criteria are important not only to diagnose constrictive pericarditis, but they help distinguishing it from restrictive cardiomyopathy. 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 echocardiography and color M-mode flow propagation have been validated as ancillary tools to distinguish between these diseases, though 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 echocardiography 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 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 disease or severe right ventricle disfunction and large respiratory variations in intrathoracic pressure may also show inspiratory decreases in early mitral inflow velocities, like 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. This augments blood flow to the right atrium from the superior vena cava during inspiration.

10.5 Effusive-Constrictive Pericarditis

The features of cardiac tamponade and constrictive pericarditis may combine originating the effusive-constrictive pericarditis. These rare and particular cases should be diagnosed not only with the important help of the echocardiographic imaging but also taking into consideration hemodynamic variations before and after the removal of the exceeding pericardial fluid. The clinical diagnosis of this condition is based, indeed, on the demonstration that in a patient with pericardial effusion 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 normal.

10.6 Pericardial Masses

Primary pericardial tumors are rare; secondary tumors are far most common. Primary tumors can be benign, including lipoma, hemangioma, and teratoma or malignant, including

mesothelioma, sarcoma, and lymphoma. Metastatic tumors are usually of breast, lung, and bone marrow origin.

Pericardial masses are often detected incidentally during routine echocardiography; though echocardiography should be considered important in the identification of these masses, CT scan or CMR should always be performed as they are the imaging modalities of choice when further evaluating these tumors. Currently, 2D echocardiography techniques are most often used to assess tumor characteristics such as shape, size, tumor attachment, and location as related to adjacent structures. Transesophageal echocardiography (TEE) can be a complementary diagnostic test that is especially helpful in characterizing masses in the posterior structures of the heart, particularly in the left atrium, left atrial appendage, right heart, and descending thoracic aorta. Further use of 3D echocardiography can be helpful by allowing for measurement of the entire volume of a mass, which may be underestimated by 2D echocardiography.

10.7 Pericardial Cysts

Pericardial cysts are rare, benign congenital, or inflammatory malformation; they can also be acquired after cardiothoracic surgery. They are usually found incidentally during routine x-ray or echo examinations and appear as echo-free fluid-filled loculated masses located mostly at the right costophrenic angle, less frequently in the left costophrenic angle, or in the posterior or anterior superior mediastinum. Color-flow and pulsed-Doppler interrogation at low-velocity setting can be used to ensure there is no phasic flow within the structure and 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 identifying a pericardial cyst in atypical locations and distinguishing it from other posteriorly located lesions.

10.8 Congenital Absence of the Pericardium

Congenital absence of the pericardium is a rare anomaly whose reported prevalence is of 0.002–0.004%. This defect could 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 determines 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. Echocardiogram should be useful as well in the identification of the associated heart defects such as atrial septal defects and bicuspid aortic valve.

Suggested Reading

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