Echocardiography in the diagnosis and management of pericardial disease

Mauro Pepi and Manuela Muratori

This review covers the role of echocardiography in the diagnosis and management of the main pericardial disorders. The sensitivity of echocardiography in detecting pericardial fluid is very high and this technique allows the detection of effusion as well as the definition of the size of effusion (small, moderate and severe). The evaluation of the pericardial sac should be carefully performed through all the echocardiographic windows, differentiating diffuse from loculated (regional) effusions. Several echocardiographic and Doppler signs allow an accurate diagnosis of cardiac tamponade. The role of echocardiography is extremely important in atypical clinical presentation such as in patients in the postoperative period after cardiac surgery. Moreover, drainage of the effusion is mandatory in the presence of cardiac tamponade and in this regard echo-guided pericardiocentesis is the gold-standard method. Finally this review covers the echocardiographic diagnosis of pericardial cysts and masses and constrictive pericarditis. *J Cardiovasc Med* **7:533–544 © 2006 Italian Federation of Cardiology.**

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Introduction

Echocardiography has proved to be highly valuable in evaluating pericardial disorders. Pericardial involvement in disease processes is manifested in a multitude of ways ranging from trivial pericardial effusion to cardiac tamponade, from pericardial thickening to constrictive pericarditis. This review covers the role of echocardiography in the detection of pericardial effusion and signs of cardiac tamponade as well as the contribution of ultrasound techniques in the diagnosis and management of main pericardial disorders.

Pericardial effusion

Echocardiography aids in the detection, localization and quantification of pericardial effusion. The appearance of the normal pericardium in M-mode or two-dimensional echocardiography is that of a bright, dense layer of echoes inseparable from the epicardial echo (generally the pericardial echo is slightly greater than 2 mm). The sensitivity of echocardiography in detecting pericardial fluid is very high and even 20 ml of fluid inside the pericardium may be visualized. Since even healthy individuals occasionally may have up to 50 ml of pericardial fluid, echocardiographic visualization of a very small amount of pericardial fluid (in an otherwise healthy individual) should not be cause for concern. The size of the effusion may be graded as: small (echo-free space in diastole < 10 mm, corresponding approximately to 300 ml); moderate (10–20 mm, corresponding to 500 ml); large (> 20 mm, > 700 ml). Alternatively, Horowitz *et al.* [1] classified pericardial effusion as: type A, no effusion; type B, systolic separation of epicardium and pericardium (3–16 ml); type C1, systolic and diastolic separation of epicardium and pericardium (> 16 ml); type C2, systolic and diastolic separation of pericardium with attenuated pericardial motion; type D, pronounced separation of epicardium and pericardium with large echo-free space; type E, pericardial thickening (> 4 mm). Both classifications may be useful, however, even in diffused and circumferential effusion dimensions of the echo-free space may be different in the examined views and therefore it is more correct and easy to measure and annotate the dimension of the effusion and to report where it has been evaluated (i.e. 12 mm in the left ventricular lateral wall in the apical four-chamber view; 10 mm along the right atrium in the subcostal view) (Fig. 1). This methodology not only facilitates the definition of effusion size, but it allows follow-up studies, detecting changes in the amount of pericardial fluid after therapy. Most pericardial effusions are circumferential, and therefore moderate and large effusions in particular result in echo-free spaces along the entire heart profile in the different views. On the contrary small or loculated effusion may be observed only in the posterior regions or (particularly iatrogenic and post-surgical effusion) along one or more heart regions (more frequently along the right side of the heart). With regard to technical rules it is important to visualize the pericardium from as many planes as possible, including off-axis views, and to utilize the gray scale and gain settings correctly. In fact, a small pericardial effusion may be more precisely defined by setting the gain low so that only the posterior pericardium is

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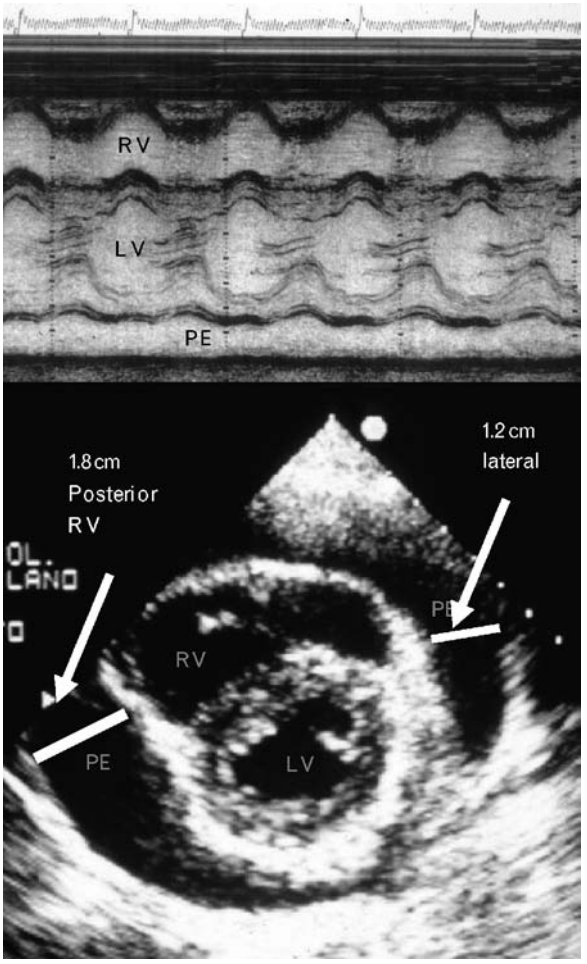
Fig. 1



M-mode (top) and parasternal short axis view (bottom) showing patients with large diffuse pericardial effusions (PE). Arrows indicate measurements of the dimension of the PE. RV, right ventricle; LV, left ventricle.

visualized. On the contrary, fibrin strands, masses and clots within the pericardial cavity may be missed if the gain is too low. Therefore ultrasound gain and gray scale should be optimized throughout the examination.

In general it is not possible to assess the character of fluid (serous effusion, hemopericardium and chylopericardium as all appear as similar clear spaces), however hematomas and neoplastic diseases may be suspected in the presence of solid masses of echoes inside the pericardium; fibrin strands are frequently seen in chronic serous or serous–hematic effusions. Echogenic masses attached to the visceral pericardium may suggest the presence of metastases.

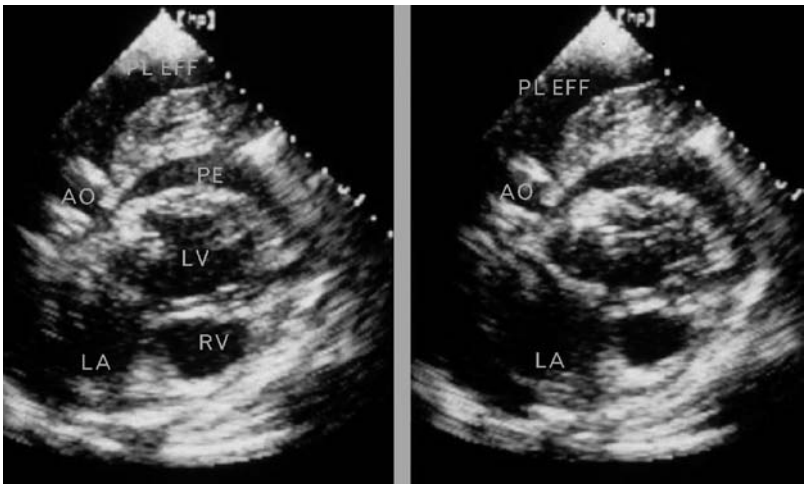
Several conditions and pitfalls in echocardiographic examination may mimic the presence of pericardial fluid. Epicardial fat in particular may be sometimes mistaken for pericardial effusion. Left pleural effusions may be differentiated from pericardial effusion by utilizing all echo-windows including the posterior approach. This view facilitates the recognition of pleural space, pericardial layers and posterior pericardial effusion. Figure 2 shows an example of the usefulness of the posterior approach in a patient with a combined pericardial and pleural effusion. 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. Other diagnostic pitfalls are: cysts, foramen of Morgagni hernia, epicardial fat inferior left pulmonary vein, left ventricular pseudoaneurysm. Other techniques may be utilized under difficult conditions and particularly when masses along the pericardium are suspected, in the presence of suboptimal imaging, or when pericardial thickening is suspected. Transesophageal echocardiography, computed tomography and magnetic resonance have been demonstrated to be very useful in several pericardial diseases including neoplastic diseases, cysts, constrictive pericarditis, hernias and differentiation between fat and pericardial fluid.

Cardiac tamponade and echo-guided pericardiocentesis

Definition and physiology

Cardiac tamponade is defined as significant compression of the heart by accumulating pericardial contents (effusion fluids, clots, pus and gas, singly or in combination). When liquid is injected into the pericardial sac of an experimental animal, intrapericardial and right and left atrial pressures begin to rise equally; this may occur also in the presence of small amount of fluid (20–40 ml). As more fluid is added cardiac stroke volume falls while cardiac output falls less because of compensating tachycardia. In fact output of both ventricles depends on adequate diastolic filling; normally intrapericardial pressure is zero or slightly negative and the transmural pressure gradient across the myocardium during diastole is positive (i.e. the intraventricular pressures are greater than the intrapericardial pressures), thus facilitating ventricular filling. The accumulation of fluids into the pericardial cavity causes an increase of both the intrapericardial and intracardial pressures but the difference between them narrows, reducing the distending force for ventricular filling. Even in cases with diffuse circumferential effusion the hemodynamic effects of tamponade are due primarily to right heart compression; in fact the pressures of these thinner chambers equilibrate with rising pericardial pressure before the left atrial and ventricular pressures. With overt tamponade, rising of the pericardial pressure progressively reduces and ultimately makes phasically negative the transmural pressure of first the right and then the left heart chambers. Filling of the heart is therefore maintained by a parallel increase in systemic and pulmonary venous

Fig. 2



Posterior view (from a back transducer location). From the posterior view the association of pericardial effusion (PE) with a left pleural effusion (PL EFF) is easily detected. AO, descending thoracic aorta; LA, left atrium; LV, left ventricle; RV, right ventricle.

pressures; as compensatory mechanisms (primarily tachycardia) are defeated, cardiac filling decreases, pericardial pressure equilibrates with left ventricular diastolic pressure and cardiac output decreases critically. Inspiration increases the filling gradient across the right heart but not the left heart; the augmented right ventricular filling occurs at the expense of reduced filling and stroke output of the left ventricle; these mechanisms cause a typical clinical sign of tamponade, pulsus paradoxus (systolic drop in arterial pressure of 10 mmHg or more during normal breathing) [2–4].

Clinical signs

Although the clinical conditions may be markedly varied in patients with cardiac tamponade (due mainly to different etiologies of the syndrome), three signs (Bech's triade) hypotension, jugular venous distension (elevated systemic venous pressure) and pulsus paradoxus are typically associated with cardiac tamponade. Tachycardia, dyspnea, cough and dysphagia, chest discomfort, shock and unconsciousness may also be present. However, rapid bleeding or fluid accumulation into the pericardial sac may present with more overt signs and symptoms than medical patients who have more gradual onset of the syndrome and preserved blood pressure.

Echocardiographic Doppler features of cardiac tamponade

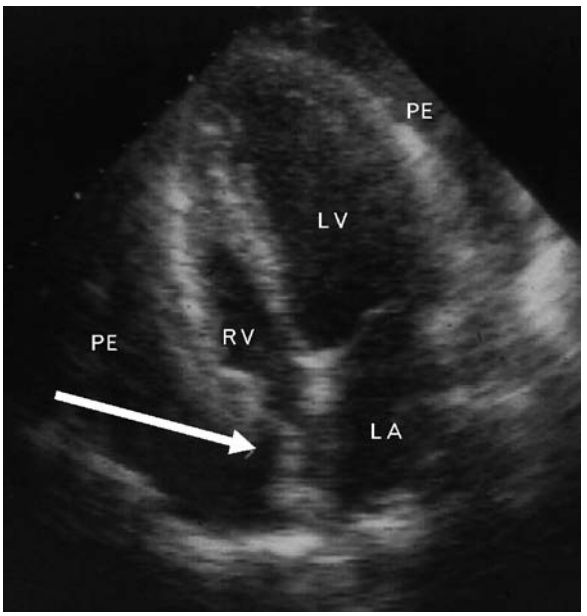
Pericardial effusion is easily demonstrated by echocardiography; when tamponade is suspected the evaluation of the pericardial sac should be carefully performed through all the echocardiographic windows, in an attempt to quantify pericardial fluid and differentiating diffuse circumferential effusions from loculated regional ones. As previously reported, echocardiography allows discrimination between small, moderate and large effusions and may also detect the association with pleural effusions [5–8]. Excessive cardiac motion up to the so-called 'swinging heart' is frequently seen in severe pericardial effusion with chronically accumulated effusion and a minimum of adhesions. This movement has been observed in malignancies, chronic tuberculous pericarditis and also benign viral pericarditis. Table 1 shows a list of the echocardiographic and Doppler signs that have been described in cardiac tamponade.

An exaggerated inspiratory expansion of the right ventricle (RV) and simultaneous compression of the left ventricle is a nonspecific sign of increased direct interdependence and has therefore a low specificity. Diastolic collapse of the right atrium (RA) and RV free walls are accepted signs of cardiac tamponade; they can be evaluated by both M-mode and two-dimensional echocardiography [9–11]. Right ventricular collapse is a transient invagination of the RV free wall which occurs in early diastole, whereas RA collapse is a transient invagination of this wall, which occurs in late diastole and early systole (Fig. 3). The timings of these two collapses are related to

Table 1 Echo-Doppler signs of cardiac tamponade

Exaggerated inspiratory variation of the two ventricles (inspiratory expansion of the right ventricle and simultaneous compression of the left ventricle; reciprocal changes in the expiratory phase)
Right atrial collapse
Right ventricular collapse
Left atrial collapse
Left ventricular collapse
Inferior vena cava plethora
Abnormal increased respiratory variation in transvalvular blood flow velocities (mitral and aortic flow reduction in the inspiratory phase)

Fig. 3



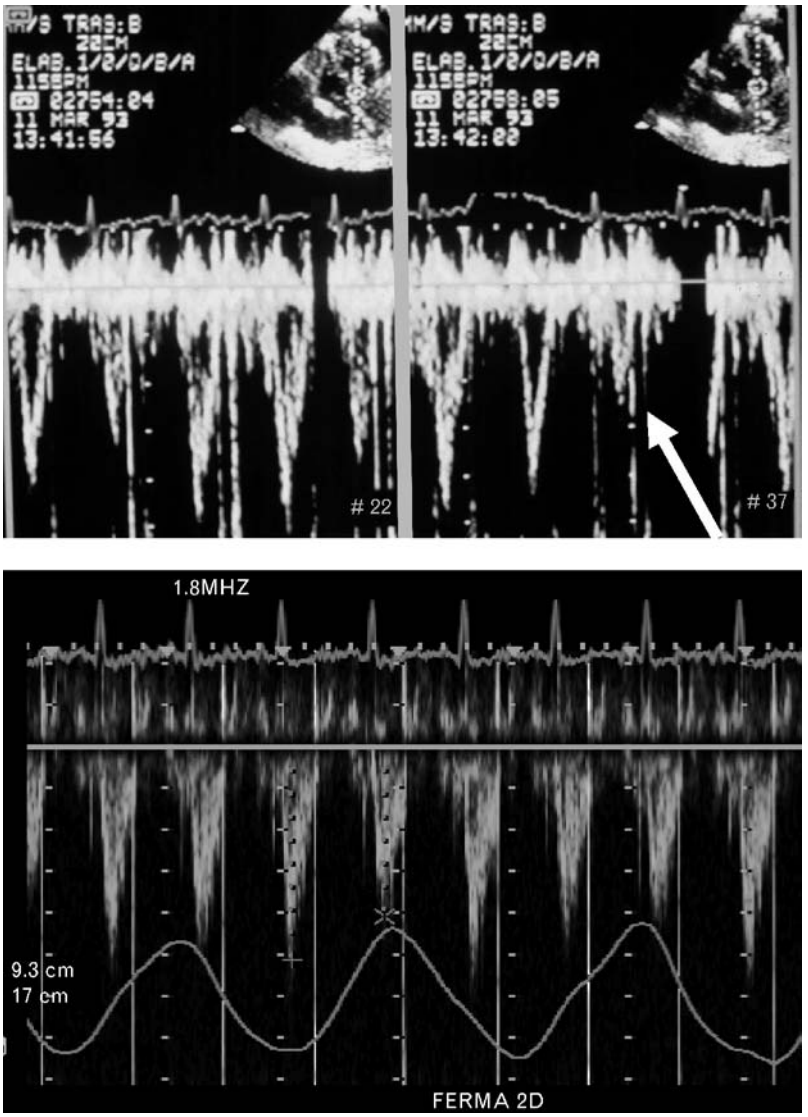
Apical four-chamber echocardiogram of a patient with pericardial effusion (PE) causing cardiac tamponade. Arrow indicates the presence of right atrial collapse. LA, left atrium; LV, left ventricle.

the lowest intracavitary pressures occurring in the two chambers in early (RV) or late (RA) diastole, respectively. These two signs of tamponade may be too sensitive on the one hand and lack specificity on the other. Duration of the collapse should therefore be taken into account. In fact, duration of collapses is directly related to severity of tamponade and improves the specificity and predictive value of these diagnostic signs [12]. With increasing severity, RA collapse tends to begin earlier and RV collapse to extend later in diastole. Even though these two signs are too sensitive, the presence of both RA and RV collapses always indicates that the effusion is hemodynamically significant. Leimgruber *et al.* [13] in an experimental animal study showed that the earliest appearance of RV diastolic collapse is associated with a 21% reduction in cardiac output at a time when mean aortic pressure is unchanged. Lopez-Sendon *et al.* [14] in an open-chest dog model showed the sequence and characteristics of RA collapse: long before hemodynamic alterations of fully established cardiac tamponade are present, RA compression becomes apparent as a quick inward motion of a small portion of the posterior RA wall, and as the intrapericardial pressure increases, a wider portion of the RA wall presents an abnormal motion. With a further increase in intrapericardial pressure, RA inversion throughout the entire cardiac cycle becomes progressively apparent, and finally the complete distortion of the RA shape and dimensions indicates an extreme situation of cardiac tamponade. These old experimental studies underline the concept that echocardiography may detect early tamponade, and that we may always consider tamponade as a continuum of events: in the early phase of cardiac compression even minor elevations of intrapericardial pressure produce some effects on ventricular filling and echocardiographic signs could be present even in the absence of overt clinical tamponade [15–19]. There are few exceptions to the use of right-sided collapse for diagnosing tamponade: right ventricular hypertrophy and high RV intracavitary pressures may prevent the occurrence of these collapses. Left atrial and left ventricular collapse are rarely seen in patients with cardiac tamponade. This is mainly due to local factors; first, the left ventricle is much thicker and stiffer than the other chambers and for that reason it resists collapse. The left atrium is posteriorly positioned and it is tightly clasped by the pericardium; rarely, in cases with very large effusions, fluid does get behind the left atrium causing wall collapse. Facilitating factors of left atrial and ventricular wall collapse are therefore posteriorly loculated effusions and conditions in which pressures in these chambers are relatively low. Left ventricular collapse has been described in postoperative effusion [20].

Inferior vena cava plethora with reduced inspiratory collapse is another sign of cardiac tamponade [21]; several studies have shown that dilatation and particularly blunted inspiratory variation of the inferior vena cava correlate with increased right atrial pressure. Therefore, this is a sensitive but unspecific sign of the syndrome, which may be seen also in constrictive pericarditis, RV infarction, pulmonary hypertension and tricuspid regurgitation. Through this method (inspiratory variation during regular respiration) RA pressure can be estimated and classified as normal (6 mmHg in the presence of an inspiratory reduction of the inferior vena cava diameter $> 45\%$), moderately elevated (9 mmHg; between 35 and 45%) and markedly elevated (16 mmHg; $< 35\%$) [22].

Cardiac tamponade is associated with an abnormally increased respiratory variation in transvalvular blood flow velocities. Normally inspiration causes a minimal increase in systemic venous, tricuspid and pulmonary valvular blood flow and a corresponding decrease in pulmonary venous, mitral and aortic flow velocities. Thus in a normal subject inspiratory variations in these measurements are less than 20%. With cardiac tamponade, ventricular interdependence is exaggerated and inspiration produces a significant decrease in left-sided filling. Accordingly mitral and aortic valvular flows are reduced by more than 40% (and a corresponding increase is observed in right-sided flow velocities) [23,24]. From a technical point of view it is important to control the positioning of the sample volume during the examination trying to exclude differences in location during respiration, and to utilize the electrocardiographic cable as a

Fig. 4



Aortic flow (top) during respiratory phases in a patient with severe cardiac tamponade. Inspiration produces a marked decrease in the aortic flow (arrow). In a second patient (bottom) with mild hemodynamic consequences of pericardial effusion respiratory changes are approximately 15% (inspiratory versus expiratory percentage changes in the maximal aortic flow velocities).

respiratory monitoring (several ultrasound companies have this option) (Fig. 4).

Pericardial effusion and cardiac tamponade after cardiac surgery

Pericardial effusion is not a rare complication of cardiac surgery [25–29]. Although it is generally reversible and not life threatening, it may sometimes evolve towards cardiac tamponade. The role of echocardiography is extremely important in the diagnosis of cardiac tamponade in the postoperative period since the clinical and imaging presentations could be atypical and therefore a specific knowledge of this pathology is very useful [30]. Virtually all pericardial effusions are generally found by the fifth postoperative day; they peak on the tenth day and resolve within 1 month. In a large prospective study [31] pericardial effusion was detected in 64% and was more often associated with coronary artery bypass grafting (75%) than with valve replacement (52%) or other types of surgery (50%). It was small in 68%, moderate in 30% and large in 2%. Loculated effusions (57.8%) were more frequent than diffuse ones (42.2%). The size and site of effusions were related to the type of surgery; in particular small pericardial effusions were slightly more frequent after valve replacement, than after coronary artery bypass grafting. After valve replacement, diffuse

fluid accumulations were more frequent (55%) than loculated ones, whereas after coronary artery bypass grafting loculated effusions were more common (63.5%). In particular anterior loculated effusions were more common after coronary artery bypass grafting, and postero-lateral ones after valve replacement. Interestingly 6% of the patients had isolated effusions along the right atrial wall; this type of effusion can compress the heart and can be difficult to diagnose in patients with postoperative low output failure. Fifteen out of the series of 780 cases had cardiac tamponade; this event was significantly more common after valve replacement than after coronary artery bypass grafting or other types of surgery. All these epidemiological data are extremely important since the recognition of cardiac tamponade in the early or late postoperative periods is not easy.

Loculated effusions may be difficult to explore and off-axis views should be attempted in cases in which the differential diagnosis with pleural effusion is difficult or in the presence of atypical presentation. In particular, localized pericardial effusion or clot at the level of the right atrial wall, right ventricle, left ventricle and left atrium may occur after cardiac surgery and present with unique clinical and echo-Doppler features. Modified apical, subcostal and posterior views are extremely useful in the evaluation of all these isolated effusions; however, in several cases, particularly when transthoracic echocardiography does not provide complete imaging of the pericardial sac and unstable hemodynamics with suspected cardiac tamponade coexist, then transesophageal echocardiography is mandatory [32–35]. Russo *et al.* [30] demonstrated that nine out of 10 patients with cardiac tamponade (10 out of 510 consecutive patients evaluated after cardiac surgery) had either atypical clinical, hemodynamic and/or echocardiographic findings. The authors found that frequently patients had vague and nonspecific initial symptoms, including general malaise, lethargy, anorexia, palpitations whereas dyspnea, chest pain, confusion, diaphoresis, oliguria generally occur later. Selective chamber compression was the more frequent cause of atypical clinical and echo presentation. Left and right ventricular, and left and right atrial compressions have been demonstrated after cardiac surgery. Pericardial clots, particularly at the level of the right chambers may produce low cardiac output soon after open-heart surgery with hemodynamic signs that are very similar to constrictive pericarditis; Beppu *et al.* [33] showed the indispensability of transesophageal echocardiography in these cases. Several reports confirmed that after cardiac surgery and tamponade caused by loculated regional RA compression pulsus paradoxus may be absent, whereas regional compression of the left ventricle and tamponade may occur despite normal RA and peripheral venous pressure. Atypical clinical presentation is very frequent after cardiac surgery and the three clinical signs of tamponade (in particular the pulsus paradoxus) may be absent in several settings: left ventricular dysfunction, regional RA tamponade, positive-pressure breathing, atrial septal defect, pulmonary arterial obstruction, severe aortic regurgitation. In terms of hemodynamics effects Fowler *et al.* [36] demonstrated that right-sided cardiac compression has more important effects than left-sided compression; however, left-sided tamponade still makes a significant contribution to the total hemodynamic picture of cardiac tamponade. Therefore echocardiographic research of regional pericardial effusion should be very accurate. Postoperative cardiac tamponade might be suspected in the presence of loculated effusions along the right atrium and/or along its junction with the superior vena cava, left atrial or bilateral atrial compression, right or left ventricular effusions. Figure 5 shows two examples of loculated pericardial effusions (clots) causing cardiac tamponade. We investigated in this setting the type and frequency of Doppler and echocardiographic findings of cardiac tamponade [31]. We invariably (15/15 cases in our series) recorded an inspiratory decrease ($> 25\%$) in the velocity of flow through the aortic and mitral valves; five patients had right atrial collapse, right ventricular collapse and inferior vena cava plethora; eight had two of these signs, and two had one. In four patients moderate pericardial effusions were associated with one or two echocardiographic signs of cardiac tamponade, but not with clinical evidence of hemodynamic embarrassment or with Doppler signs of tamponade.

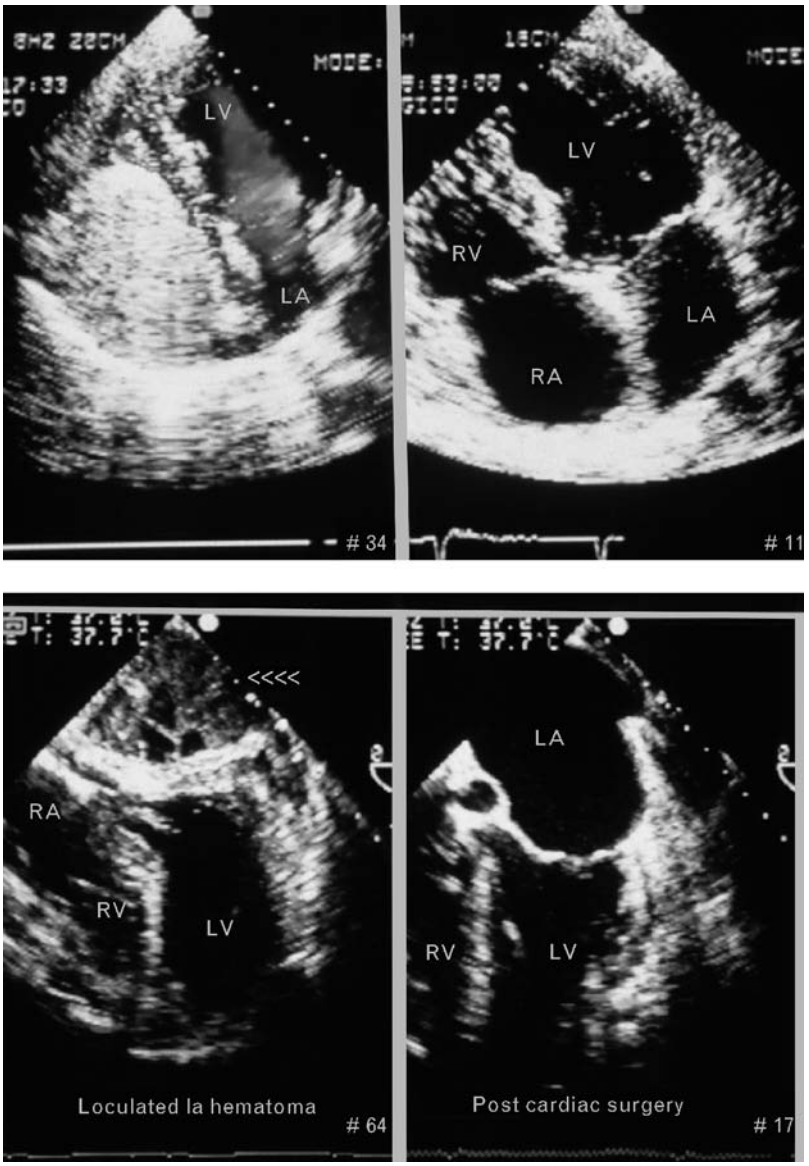
Echo-guided pericardiocentesis

Early recognition and treatment of patients with suspected tamponade represents an important clinical goal and, once diagnosed, decompression by a safe, simple and rapid method is required.

Drainage of an acute effusion producing cardiac tamponade may be performed by two alternative techniques: subxiphoid pericardiotomy and percutaneous pericardiocentesis. Subxiphoid pericardiotomy [37] is performed using either local or general anesthesia via a midline longitudinal incision from the xiphosternal junction to 6 to 8 cm below the tip of the xiphoid. The xiphosternal point is then split and the xiphoid process removed to expose the subxiphoid diaphragm. The anterior diaphragm is depressed and the sternum can be lifted to allow dissection and removal of fatty tissue until the pericardium is well reached; then a finger is introduced to palpate the tension of the pericardial sac, and the pericardium is grasped with forceps and incised allowing the fluid to escape. Samples of the pericardial effusion can be collected in sterile tubes and sent for bacteriologic analysis, and finally a pericardial drain may be left in place for 24 h or more.

Percutaneous pericardiocentesis has been described in detail by several authors [38–42], who proposed improvement of the technique starting from the blind

Fig. 5



Examples of loculated pericardial hematomas. Top left panel: apical four-chamber view showing an echo-dense voluminous mass (pericardial clot) at the level of the right atrium. Due to this compression the right atrial cavity is virtual. Top right panel: after pericardial drainage the pericardial clot has been completely removed and the size of the right atrium is normal. Bottom left panel: transesophageal four-chamber view in a patient with hemodynamic instability immediately after a coronary artery by-pass grafting. A large pericardial hematoma compressing the left atrium was clearly demonstrated. Bottom right panel: normalization of the left atrial cavity was clearly documented after surgical drainage of the pericardium. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

procedure, passing through echo-guided and contrast echo-guided pericardiocentesis. Callahan *et al.* [40] emphasized visualization of the pericardial needle during his study on echo-guided pericardiocentesis with the assistance of contrast echocardiography, thus reducing the likelihood of heart puncture. An improvement of ultrasonically guided pericardiocentesis has been proposed by our group [41], with the specific purpose of facilitating posterior drainage. The Tuohy needle, utilized in this study, thanks to its curved tip, greatly facilitated guidance of the wire and catheter to the posterior pericardial space so that the standard Seldinger technique was successful not only in massive and diffused effusions, but also in loculated posterior ones. We also compared the combined use of two-dimensional echo monitoring with the described technique, with surgical subxiphoid pericardiotomy in the treatment of acute cardiac tamponade due to pericardial effusion occurring after cardiac surgery [42]. Forty-two patients were included in the study: during the first period one of

the two methods was chosen by the clinical staff, whereas in the second period percutaneous pericardiocentesis was the treatment of choice. Complete drainage of pericardial fluid by percutaneous pericardiocentesis was obtained in 26 out of 29 patients (86%). No major complication occurred with the use of the two techniques. In three cases, in the second study period, the minimal amount of fluid or pericardial hematoma indicated surgical approach as the first therapeutic choice; percutaneous pericardiocentesis was unsuccessful in three cases without complications and surgical drainage was therefore performed. This study suggested that the more invasive technique (subxiphoid surgical pericardiotomy) should be selected in cases in which percutaneous pericardiocentesis is unsuccessful or when the echocardiographic examination discourages a percutaneous approach.

Tsang *et al.* [43] have reported the Mayo clinic experience in cardiac tamponade caused by cardiac perforation as a complication of interventional catheterization. Echocardiography guided pericardiocentesis was safe and effective for rescuing patients from tamponade and reversing hemodynamic instability complicating invasive cardiac-based procedures. They proposed in this clinical setting as well as in cardiac tamponade due to other etiologies a specific technique based on two-dimensional characteristics of effusions. Two-dimensional echocardiography allowed the examiner to locate the largest collection of pericardial fluid in closest proximity to the transducer, thereby identifying the ideal entry site. The trajectory of the pericardiocentesis needle was defined by transducer angulation. After local infiltration with 1% lidocaine, a polytef-sheathed needle (16 to 18 gauge) was used for the initial pericardial entry. On reaching the pericardial fluid, the steel core of the needle was immediately withdrawn, leaving only the polytef sheath in the pericardial space. In the presence of hemorrhagic effusion, injection of a small volume of agitated saline as a contrast was used for echocardiographic confirmation of the sheath position. The sheath was withdrawn and a standard dilator and introducer (6–7F) advanced over the guide wire. A pigtail angiographic catheter (65 cm; 6–7F) was then introduced into the pericardial space. Rescue echo-guided pericardiocentesis with the described technique was the only treatment required for the majority of these patients. Surgical exploration or intervention is indicated when complete control of the bleeding is in question or when hemodynamic stability is not rapidly restored with rescue pericardiocentesis alone.

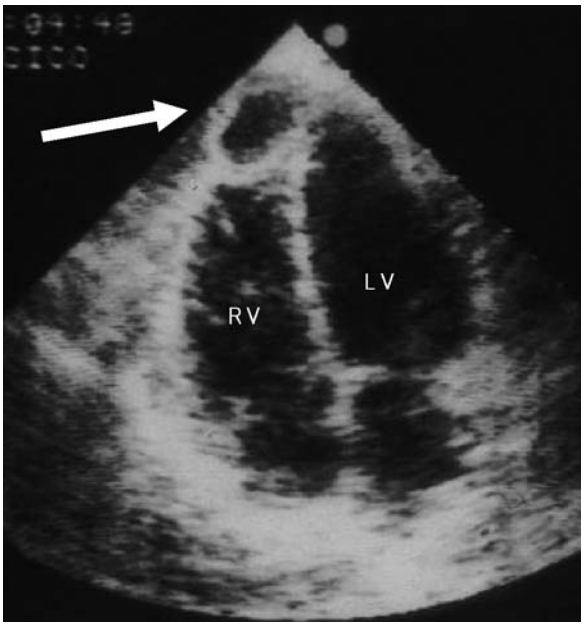
Many investigators have suggested two-dimensional echocardiography to guide not only the placement of the pericardiocentesis needle, but also biopsy forceps, balloon catheters (pericardioplasty) and other new percutaneous techniques [44,45]. Instillation in the pericardial sac of different agents, with sclerosing, anti-inflammatory or cytostatic activity has been also proposed under echocardiographic assistance [46].

Pericardial cysts and masses

Pericardial cysts can be congenital or inflammatory. Congenital cysts are uncommon, mostly unilocular and located at the right cardiodiaphragmatic angle, with a diameter from 1 to 5 cm, and clinically silent [47]. However, sometimes they are associated with chest pain, dyspnea, cough and/or significant arrhythmias, due to the compression of the heart. Inflammatory cysts can be caused by rheumatic pericarditis, bacterial infection, particularly tuberculosis, trauma and cardiac surgery. Most cysts are discovered accidentally on chest X-ray, or at echocardiography [particularly at transesophageal echocardiography (TEE)]. Usually computerized tomography (CT) and magnetic resonance imaging (MRI) are diagnostic. Pericardial cysts are seen as an echolucent unilocular cavity which can cause extrinsic compression of the atrium. Sometimes they may have a grossly heterogeneous echodense appearance on TEE, due to the collection of mucilaginous material or embryonic residua (bronchogenic cyst). Echinococcal cysts, usually originated from ruptured cysts in the liver and lungs, are the typical hydatid, multiseptate cysts (Fig. 6).

The treatment for congenital and inflammatory cysts is percutaneous aspiration and ethanol sclerosis. Video thoracotomy or surgical resection may be necessary. Only

Fig. 6



Pericardial cyst: at the apical level of the right ventricle an oval-shaped cyst (arrow) due to pericardial echinococcosis is clearly shown. LV, left ventricle; RV, right ventricle.

surgical excision of echinococcal cysts is not recommended. Pericardial cysts and particularly hydatid cysts require differential diagnosis, especially if atypically located and if imaging suggests solid tumors [48].

Pericardial masses can be primary benign tumors (as teratoma, especially found in children) or malignancies, mainly discovered in the third and fourth decades. The most important pericardial malignancies are mesotheliomas and sarcomas (particularly angiosarcoma). Secondary malignancies are metastatic (lymphoma, Hodgkin's disease, melanomas and multiple myeloma) or from neighboring structures (tumors of the lung, esophagus or chest wall).

Constrictive pericarditis

Constrictive pericarditis is a rare but severely disabling consequence of the chronic inflammation of the pericardium, which causes chronic cardiac compression. This results in restricted diastolic filling of the heart, which manifests as venous engorgement and diminished cardiac output. In the past tuberculosis was the most commonly known cause; at present tubercular etiology has become rare in developed countries, while new etiologies have appeared. Two of them, previous cardiac surgery and previous radiotherapy (for Hodgkin's disease or breast cancer) are now responsible for up to one-third of cases [49].

A distinction between elastic (subacute) and rigid shell (chronic) constriction has been proposed, other additional clinical profiles include the recognition of effusive–constrictive pericarditis, occult constriction, localized constriction and reversible constriction. Each of these variant forms of constrictive pericarditis may present with some features that differ from the classic chronic pericarditis [50]. When constrictive pericarditis is suspected, echocardiography and Doppler combined with CT or MRI are the diagnostic techniques used in most cases.

Pathophysiology

The pathophysiologic hallmarks of constrictive pericarditis occur due to a loss of pericardial compliance, which limits diastolic filling of all cardiac chambers, resulting in elevation of end-diastolic pressures. Early diastolic filling is unrestrained and up to 75% of filling may occur in the first 25% of diastole, but subsequently this rapid filling abruptly declines by mid-diastole. End-diastolic ventricular pressures and mean atrial pressures increase to nearly equal levels. As a consequence the end-diastolic volume, stroke volume and cardiac output are reduced. Moreover the encasing pericardium isolates chambers from changes in intrathoracic pressures resulting in an inspiratory reduction in the diastolic flow in the pulmonary veins and left filling and a simultaneous increase in right ventricular filling with a leftward movement of the septum (exaggerated ventricular interdependence) [51]. The opposite physiologic effects on septal shift and filling gradients of the ventricles are seen with expiration. As the intrathoracic pressure increases with expiration, flow into the left ventricle increases and the septum shifts rightward, resulting in an expiratory reduction of flow in the venae cavae, increased hepatic venous diastolic flow reversal, and decreased transtricuspid flow velocity.

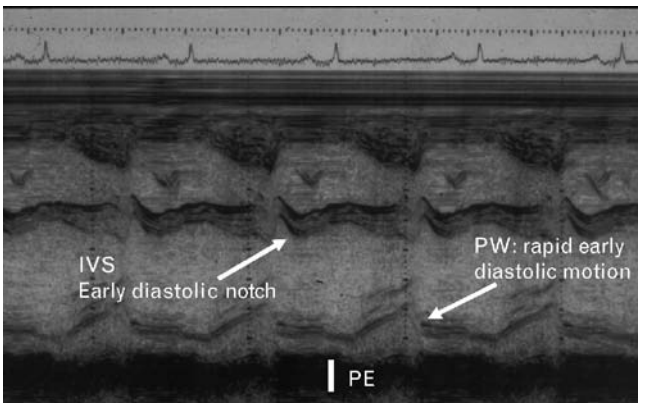
Clinical signs

Signs and symptoms of constrictive pericarditis are myriad and non-specific. Symptoms generally develop slowly over a period of years. The most common complaints described are dyspnea on exertion (78%), abdominal swelling and secondary digestive disturbance such as anorexia, abdominal fullness or pain (68%), edema (54%) and fatigue (25%). Chest pain is present in 24% of patients as a result of active inflammation [52]. Physical findings are sinus tachycardia, because cardiac output becomes dependent on heart rate. Heart sounds appear distant, but a pericardial knock may be present and signifies the end of rapid ventricular diastolic filling. Pulsus paradoxus is often found in subacute constrictive pericarditis with pericardial effusion. Hepatosplenomegaly occurs quite early and neck veins are constantly engorged in 86% of patients.

Echocardiography and Doppler features

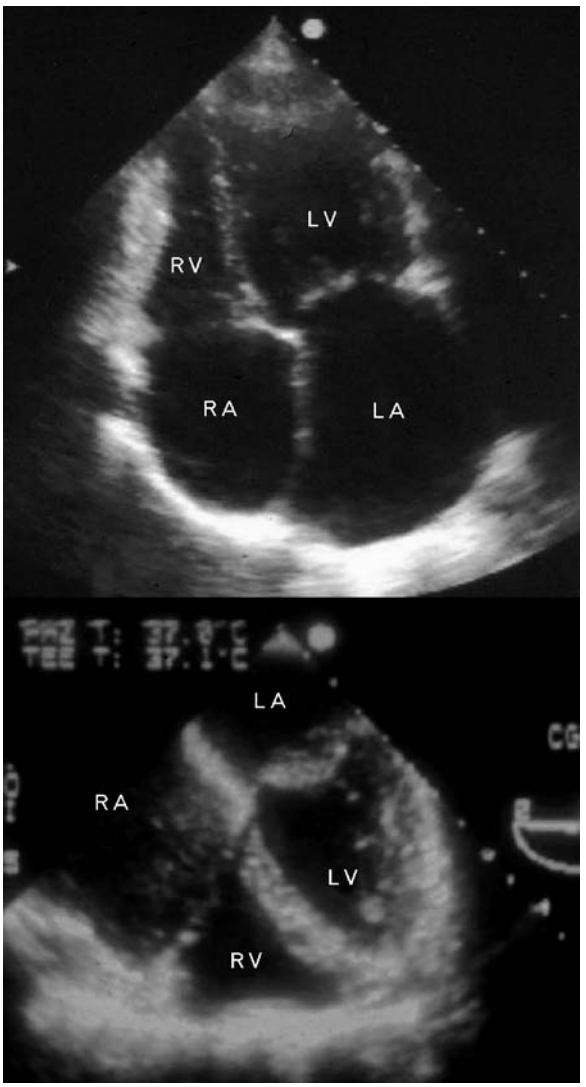
Although non-specific some M-mode and two-dimensional findings suggest constrictive pericarditis. M-mode may show an abnormal septal motion with an atrial diastolic notch and early diastolic notch, rapid early diastolic motion of the posterior left ventricular wall, and rapid early closure of mitral valve, reflecting a sudden decline in diastolic filling (Fig. 7). Abnormal diastolic septal bounce is also frequently seen, as a consequence of

Fig. 7



M-mode tracing in constrictive pericarditis. The posterior pericardium is markedly hyperechogenic and calcified; its thickness is approximately 1 cm; arrows indicate typical altered motion of the interventricular septum (IVS) and posterior left ventricle (LV) wall. PE, pericardial effusion; PW, posterior wall.

Fig. 8



Examples of constrictive pericarditis. The transthoracic apical four-chamber (top) and the transesophageal four-chamber view (bottom) show marked thickening of the pericardium that is markedly calcified. The two ventricles are normal whereas the two atria are enlarged. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

sudden changes in the trans-septal gradient during diastole. Atrial enlargement, inferior vena caval and hepatic venous dilatation are also seen at two-dimensional echocardiography, whereas transesophageal echocardiography is superior to transthoracic for detection of pericardial thickening (Fig. 8). Thickened pericardium is most easily visualized anteriorly over the right ventricular free wall, and, when a cut-off of 3 mm was used for pericardial thickness, the diagnostic sensitivity for constrictive pericarditis was 95% and specificity 86% [53].

Doppler signs are similar to those noted in patients with cardiac tamponade. Therefore with inspiration, the driving pressure gradient from the pulmonary capillaries to the left cardiac chambers decreases, resulting in a marked decreasing in diastolic pulmonary venous flow velocity and in early mitral flow velocity. The opposite change occurs with the expiration with a significant increase in early mitral flow velocity (E wave $> 25\%$) and in systolic, and more markedly diastolic pulmonary venous flow velocities increase. At the right side, because of the relatively fixed cardiac volume, with inspiration there is an increase in right ventricular filling, with a ventricular septal shift to the left. The opposite change occurs with the expiration and ventricular septum shift to the right, tricuspid inflow velocity significantly decreases and diastolic hepatic venous flow reversals significantly increase ($> 25\%$) [54]. The interpretation of Doppler flow velocities and their respiratory variation are complicated in the presence of arrhythmia or atrial fibrillation or in patients with comorbid clinical features, mimicking similar respiratory variation (chronic obstructive lung disease, mechanical ventilation, right ventricular infarction or pulmonary embolism). The restrictive mitral Doppler pattern poses a more difficult problem in a patient with clinical feature of constriction because it does not exclude constriction.

Differential diagnosis versus restrictive cardiomyopathy

As the clinical presentations of constrictive pericarditis may resemble those of restrictive cardiomyopathy, identification of echo-Doppler features of these two conditions is very important. The fundamental difference between these conditions is the absence of respiratory variation in mitral inflow and pulmonary venous Doppler velocities in restrictive cardiomyopathy [55,56]. Recently new technologies, in particular tissue Doppler, which allows direct measurement of the myocardial velocity relaxation can be useful in differentiation of constrictive pericarditis versus restrictive cardiomyopathy. Two recent studies validated the diagnostic role of E' measurement at the septal annulus by tissue Doppler echocardiography. In fact E' was significantly higher in patients with constrictive pericarditis than in those with primary restrictive cardiomyopathy or cardial amyloidosis with an E' cut-off value > 8 cm/s resulting in 95% sensitivity and 96% specificity for the diagnosis of constrictive pericarditis [57,58].

Constrictive pericarditis management

Pericardiectomy is the only treatment for permanent constriction. The indications are based upon clinical symptoms, echocardiography and CT/MRI findings. The mortality rate of pericardiectomy was 6–12% in the current series, with a complete normalization of cardiac hemodynamics in 60% of cases. Major complications include acute perioperative cardiac insufficiency and ventricular wall rupture. Cardiac mortality and morbidity is mainly caused by the presence of myocardial atrophy or myocardial fibrosis that was not recognized prior to surgery [49,59].

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