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Essentials of Pericardial Diseases

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The pericardial diseases result in cardiovascular perturbations ranging from asymptomatic electrocardiographic findings (in pericarditis of end-stage renal disease) to circulatory shock (in acute hemorrhagic pericardial tamponade). Despite of clinical features that resemble right-, or left-side failure, the clinical management of pericardial pathology may differ significantly from those of ventricular systolic or diastolic dysfunction. Therefore, prompt diagnosis and appropriate medical or surgical therapy is imperative.

Anatomy

The pericardium consists of three layers: (a) a fibrous, that blends with the adventitia of the great vessels, (b) a parietal, lining the inner surface of the fibrous layer composed of a mesh of collagen and elastic fibers that provide for the flexibility of the pericardium, and (c) a visceral, that attaches to the surface of the heart and epicardial fat and forms the epicardium. The visceral layer is comprised of mesothelial cells that produce the pericardial fluid. This layer reflects back on itself to line the outer fibrous layer and becomes the parietal pericardium. The pericardium functions as a barrier that protects the heart from spread of infection and malignancy, reduces the friction between the heart and the adjacent tissues, and controls the hydrostatic forces of the heart, preventing acute chamber dilatation and maintaining diastolic coupling between the ventricles [1].

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Pericarditis

Pericarditis, the inflammation of the pericardium, develops as an acute illness or complicating rheumatic, renal and malignant disease or treatment with certain drugs. The causes of pericarditis are numerous (see table 1)

Table 1. Causes of pericarditis

Infectious *viral* *tuberculosis* *bacterial* *fungal* *toxoplasmosis* *amebiasis*	**Autoimmune** *acute rheumatic fever* *systemic lupus erythymatosus* *rheumatoid arthritis* **Medications** *hydralazin* *procainamide* *phenyntoin* *penicillin*
Uremia	**Idiopathic**
Neoplasms	**Sarcoidosis**
Radiation	**Acute myocardial infarction**
Trauma	**Postcardiotomy** for cardiac surgery
Amyloidosis	**Dissecting aortic aneurysm**

The microscopic findings include infiltration with leukocytes (lymphocytes, polymorphonuclear leukocytes, macrophages), alterations in vascularity, and fibrin depositon. The amount of pericardial fluid increases, and depending on the rate and the amount of accumulation, may result in cardiac tamponade. Following the acute inflammatory phase, fibrous adhesions may be present between the pericardium and the epicardium and any other contiguous tissues

surrounding the heart, resulting in varying degree of constrictive pericarditis.

The usually febrile patient complains of mild or severe retrosternal chest pain, variable in intensity and quality, indistinguishable from acute abdomen or myocardial ischemia. The typical pericardial pain is exacerbated by inspiration and relieved by leaning forward. The physical findings are comprised of a pericardial rub, distant heart sounds, increased jugular venous pressure, edema, ascites, and Kussmaul's sign (increased right atrial pressure during inspiration that results in a paradoxical jugular venous distension). These clinical signs are often identical to those of severe right ventricular dysfunction and/or tricuspid insufficiency, whereby hepatic congestion, abdominal ascites and lower extremity edema are prominent. The electrocardiogram, during the acute phase, will typically demonstrate diffuse ST segment elevation, a current injury from the superficial layers of the myocardium. The ST changes are not accompanied by reciprocal depression, as seen in myocardial ischemia, while the voltage pattern is low. In the chronic state, the chest radiographs may show calcification of the pericardium principally on lateral views with or without cardiomegaly. Pleural effusions can also be present [2].

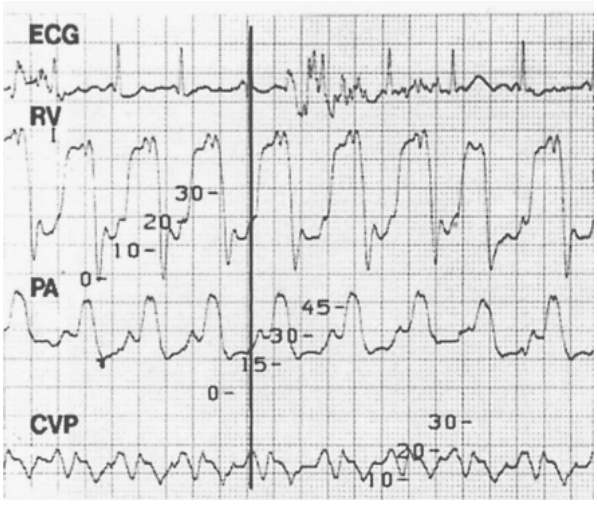
Relapsing or recurrent pericarditis causes episodes of chest pain without a friction rub, sometimes with pericardial and even pleural effusions. The episodes are accompanied by fever, and may complicate recovery from acute myocardial infarction (Dressler's syndrome) or cardiac surgery (the postcardiotomy syndrome). Although the condition is benign and does not lead to constriction, it may be quite debilitating because of recurrences. Mild cases respond to aspirin or other non-steroidal anti-inflammatory drugs, while steroids are reserved for severe cases. Colchicine is the treatment of choice and can also provide effective prophylaxis. Pericardiotomy fails to prevent relapses of pain [3].

Contstrictive Pericarditis

The majority of the cases of non-idiopathic constrictive pericarditis are caused by previous cardiac surgery, or radiation exposure. The thickened, fibrotic, heavily calcified pericardial sac has decreased compliance and restraints ventricular filling. Constrictive pericarditis can be surgically cured with pericardial stripping, with acceptable surgical mortality and symptomatic benefits to most patients. The physical findings in advanced disease reflect the signs of cardiac congestion.

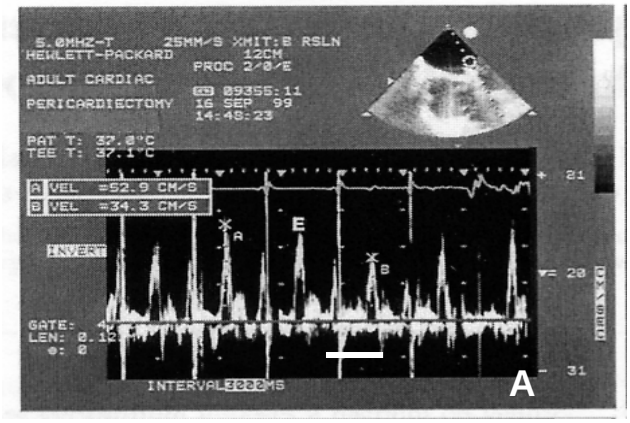
Isolated constrictive pericarditis is characterized by an abnormality in compliance. Although the myocardium is not diseased, the ventricular filling is impaired because of external interference to diastolic filling from the rigid pericardium. The stroke volume is either fixed or progressively decreases and any increase in physiologic demands can only be met via increases in heart rate. Following isovolumetric relaxation, chamber filling is initially rapid and uncompromised. However, ventricular as well as atrial volumes are reined by the noncompliant pericardium, so that once ventricular diastolic filling reaches the limitations imposed by the non-elastic pericardial sac, the pressure and volume inside the cardiac cavity rises, the filling ceases and congestion occurs. Characteristic hemodynamic tracings include the "W" or "M" central venous pressure (CVP) waveform and the "square root" right ventricular (RV) sign (Figure 1). In constrictive pericarditis, the thickened pericardium shields the heart, but not the extrapericardially, and only partially intrathoracically located systemic (superior [SVC] and inferior [IVC] vena cavae) veins, or the

Figure 1: Hemodynamic tracings in constrictive pericarditis



entirely intrathoracic pulmonary veins (PV) from changes in intrathoracic pressure during respiration. Furthermore, since (i) the filling pressure gradient for the RV is the difference between the systemic veins and the RV diastolic pressure, while the filling pressure gradient for the left ventricle (LV) is between the PVs and the LV diastolic pressure, and (ii) the intracardiac blood volume is relatively fixed, constrictive pericarditis results in an exaggerated RV and LV interdependence. That means that there is a reciprocal relation between the left and right heart filling: during spontaneous inspiration the right heart fills, the septum shifts to the left and the filling of the left heart is restricted. The opposite occur during spontaneous exhalation: the pressure gradient between the PVs and the left atrium increases, and the left heart fills at the expense of the right heart [4].

Figure 2: Trans-mitral flow in constrictive pericarditis



Echocardiographic assessment

The echocardiographic assessment of the thickened pericardium by the transthoracic (TTE) approach is often limited and is diagnostic in less than one third of cases of constrictive pericarditis. A comprehensive transesophageal echocardiographic (TEE) approach has the advantage of better image resolution and enhanced definition of the pericardial interface with fat, fluid and surrounding tissue. Pericardial thickening appears as increased echogenecity at the pericardial reflections on 2D echo and multiple parallel reflections posterior to the LV on M-mode. Pericardial thickening of greater than 3 mm is considered abnormal. Doppler examination of flow dynamics is often diagnostic of constrictive pericarditis and is an important adjunct in differentiating pericardial processes from myocardial pathology associated with diastolic dysfunction. Both RV and LV diastolic inflow velocities show high early (E) filling velocities consistent with rapid early filling, rapid equalization of pressures between atria and ventricles, and decreased filling of the ventricles in late diastole, as shown by the extremely low or virtually absent late (A) filling velocities (Figure 2). The RV and LV filling in constrictive pericarditis is also reflected by the hepatic and pulmonary vein flow respectively: the diastolic (D) velocities are prominent and higher than the systolic (S) velocities (Figure 3). The hallmark of the Doppler echocardiographic examination is the reciprocal respiratory variation of right and left heart filling caused by ventricular interdependence. Patients with constrictive pericarditis exhibit a characteristic respiratory variation of the LV filling-associated pulsed-wave Doppler velocities: the velocities decrease with inspiration and increase with expiration. The magnitude of the change is about 25% [5]. Increased respiratory variation in mitral inflow E velocities in conjunction with pulmonary vein inflow velocity variation is virtually pathognomonic for constrictive pericarditis [6]. This is an important finding that helps to distinguish constrictive pericarditis from restrictive cardiomyopathy where there is not respiratory variation in velocities and there is a lack of pericardial thickening on 2D echo.

Figure 3: Pulmonary vein flow in constrictive pericarditis

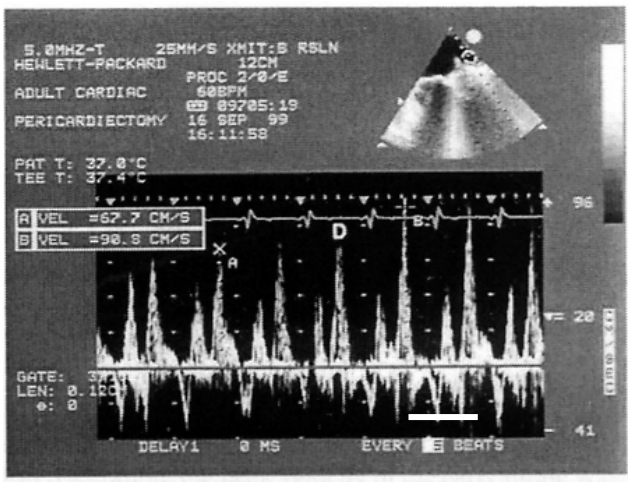
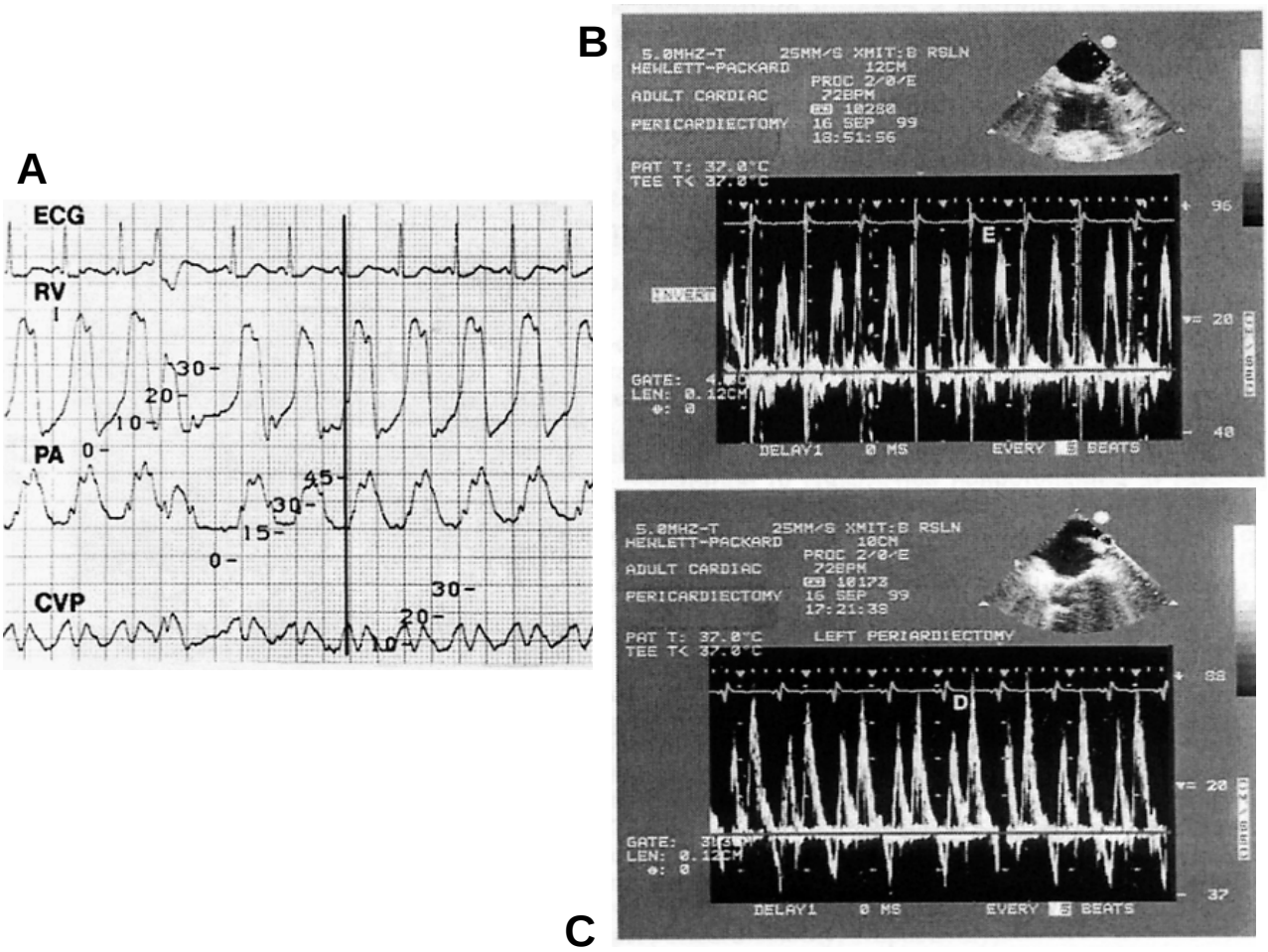


Figure 4: Hemodynamic tracings and Doppler echocardiography immediately after pericardial stripping. A: The "square root" sign has disappeared after pericardial stripping and the "M" waveform of the CVP is less prominent (compare with figure 2). B (trans-mitral flow velocities) and C (pulmonary venous velocities) reveal decreased respiratory variation after pericardial stripping (compare with figures 3 and 4, respectively). Abbreviations as in previous figures. (Adapted with permission from Ref. 8)



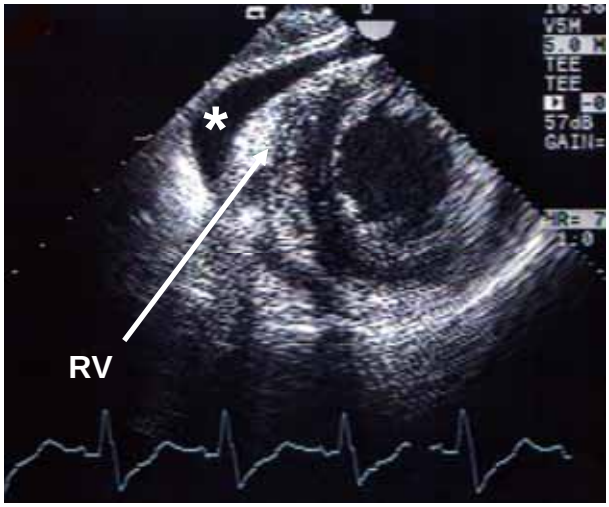
Of note, there is a subset of patients with constrictive pericarditis who do not exhibit respiratory variation, namely, those with atrial fibrillation or severely elevated LA pressures. In the anesthetized patient, positive pressure inspiration increases intrathoracic pressure, thus decreasing blood return to the RV, while the LV filling is enhanced because of augmented pressure gradient between the PVs and the intrapericardial left atrium [7]. During positive pressure exhalation, the intrathoracic pressure returns to atmospheric pressure, the gradient between PVs and left atrium decreases, the LV filling is decreased, while the RV filling is increased.(Figure 2, 3) Although not all patients will revert to a normal pattern of LV diastolic filling immediately after pericardiectomy, a more normalized respiratory variation in trans-mitral and pulmonary venous blood flow should accompany disappearance of the square root sign and decrease of right heart filling pressures (Figure 4) [8].

Cardiac Tamponade

In cardiac tamponade, the heart and the peri-cardial contents (effusion fluids, blood clots, pus, gas, singly or in combination) compete continuously for the relatively fixed intraperi-cardial volume (figure 5). Life threatening cardiac tamponade is a result of progressive, ultimately critical, reduction in cardiac output

that can occur from as little as 150 ml of blood flooding the pericardial space after a cardiac wound, to much more than 1 liter of fluid in slowly accumulating pericardial effusions [9].

Figure 5. Cardiac tamponade



The pathophysiologic mechanism behind cardiac tamponade is the reduction in cardiac inflow due to compression of all cardiac chambers, progresssively reducing their diastolic compliance and ultimately equalizing the mean diastolic pressures in all of them. The earlier targets of significant compression are the thinner right atrium and ventricle. The compressive effects of pericardial collection are seen most clearly when the respective cardiac chamber pressure is at its lowest: during atrial relaxation (i.e., ventricular systole) for the right atrium and during diastole for the RV. The transmural pressure (intracardiac pressure minus pericardial pressure) is the determinant of the filling of any chamber, and it fluctuates with respiration. During spontaneous inspiration, negative intrapleural pressure is generated facilitating venous return to the right heart whereas venous return to the left heart is diminished: (i) lung expansion increases its reservoir for blood and (ii) the increase in RV filling, causes the inter-ventricular septum to "bulge" leftwards reducing LV dimension and altering its compliance and filling (ventricular interaction). During spontaneous exhalation, the reverse process occurs. In cardiac tamponade, the total intrapericardial volume (intracardiac blood volume and pericardial contents) is fixed. The tamponading pericardial fluids compress the heart throughout the cardiac cycle, in diastole as well as in systole. Although the atria fill continuously, blood enters the heart when blood is leaving it during right and left ventricular ejection: systole ejects blood, thus reducing the intracardiac volume, and, transiently, reducing the intra-pericardial pressure, while the floor of the atria is pulled towards the apex increasing the atrial volume. Therefore, net blood flow inside the heart chambers occurs during systole, while during diastole there is a "redistribution" of the intracardiac volume between the atria and the ventricles. The respiratory variation is pronounced in cardiac tamponade and there is a reciprocal relation between the right and the left heart filling: during spontaneous inspiration, the right heart fills at the expense of the left, while during spontaneous exhalation the left heart fills at the expense of the right. This cyclical respiratory difference in left and right ventricular stroke volume is expressed clinically as pulsus paradoxus. Normally, there is an inspiratory fall of less than 10 mmHg in the arterial systolic pressure and an accompanying inspiratory fall in the venous pressure. However, a paradoxical pulse differs from the normal situation in two aspects: (i) the inspiratory fall of the arterial pressure exceeds 10 mmHg and (ii) the inspiratory venous pressure remains steady or increases (Kussmal's sign). Pulsus paradoxus is not unique to cardiac tamponade. The phasic respiratory changes in ventricular dimensions with exaggerated decrease in systemic systolic arterial pressure during inspiration can be also present in constrictive pericarditis, respiratory distress, airway obstruction, COPD, and pulmo-

Figure 6. Regional cardiac tamponade

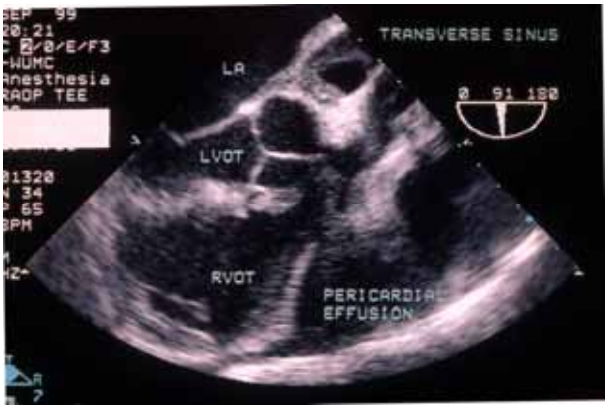
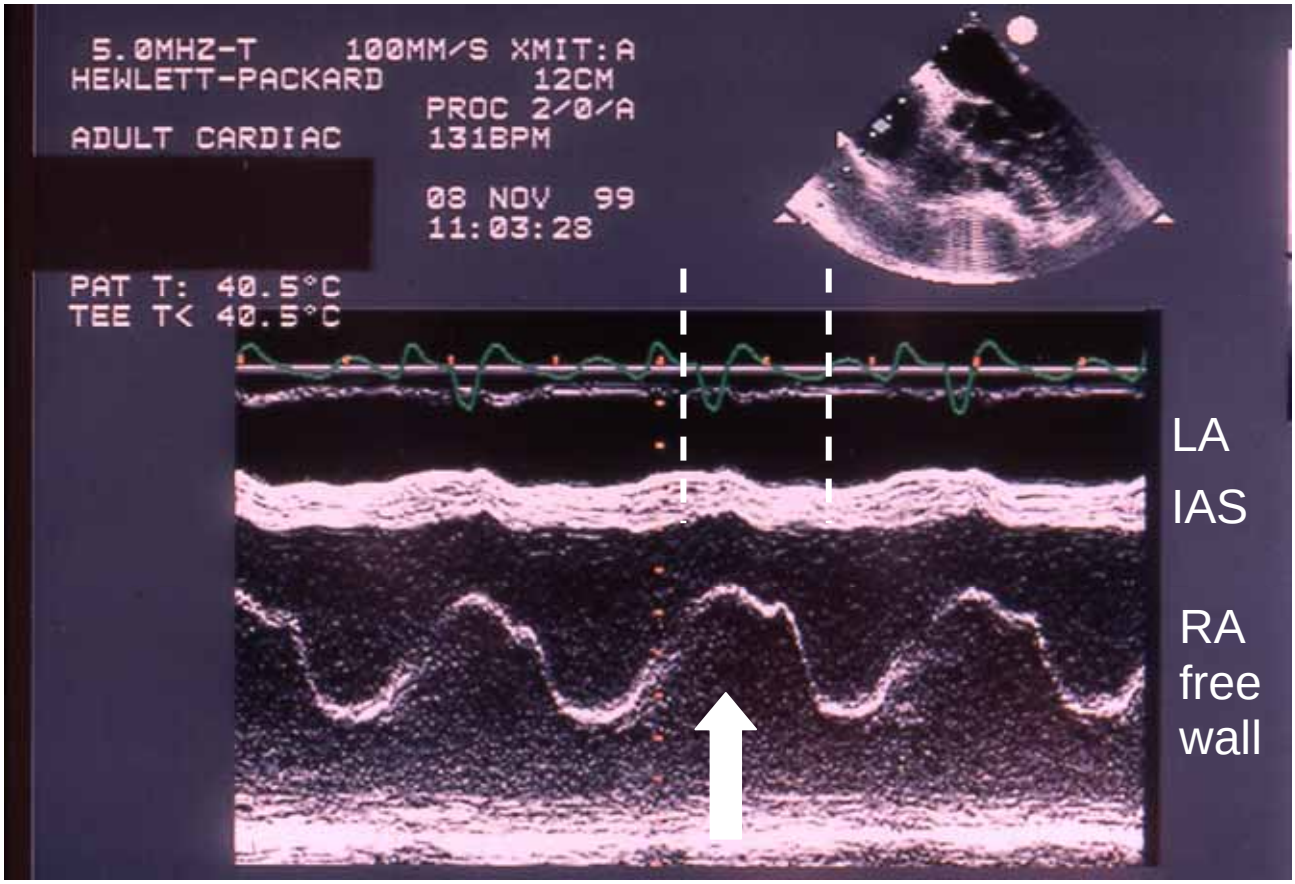


Figure 7: Right atria free wall inversion in cardiac tamponade



nary embolism. Although a prominent feature of cardiac tamponade, pulsus paradoxus may not be present at all in patients with chest wall trauma, neuromuscular disease, and pneumothorax, for they cannot produce sufficient negative intrapleural pressure during inspiration to produce the typical changes in chamber dimension and fall in left ventricular stroke volume. Similarly, those who are receiving positive pressure mechanical ventilation (early postoperative period) and patients with severe aortic regurgitation do not exhibit pulsus paradoxus. In general, coronary blood flow is reduced in cardiac tamponade, but this reduction, in the absence of coronary artery disease, is not sufficient to add an ischemic injury to the myocardium because both preload and afterload are proportionally decreased. In contrast, patients with coronary artery disease may be at increased risk for myocardial ischemia and/or infarction [10].

Clinicians should be able to recognize and treat cardiac tamponade rapidly. The syndrome of acute cardiac compression was first described in 1935 as Beck's triad: hypotension, increased jugular venous pressure, and muffled heart sounds. The causes are numerous (malignancy or any cause of acute or chronic pericarditis), but in the daily practice most cases occur after cardiac surgery.

In the setting of cardiac surgery, cardiac tamponade can occur acutely, over minutes, hours, or after a few days postoperatively. The reported incidence of acute cardiac tamponade is 0.5-5.8%. The typical patient has significant chest tube output (>200 ml/hr) in the immediate postoperative period associated with or without hemodynamic signs of inadequate cardiac output. Alternatively, the chest tubes may become obstructed by blood clots impeding mediastinal drainage. Delayed tamponade has been arbitrarily defined as cardiac tamponade occurring greater than 5-7 days after pericardiotomy. The incidence is 0.3 to2.6% and is often misdiagnosed because of a low index of suspicion and because the clinical signs and symptoms can be comparable to those of congestive heart failure, pulmonary embolism,

and generalized fatigue (i.e., failure to thrive postoperatively) [11].

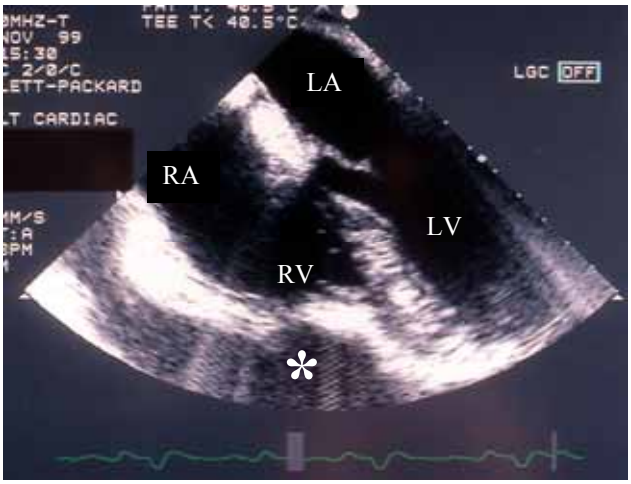
Regional cardiac tamponade can occur when one or more cardiac chamber becomes compressed. Postcardiotomy, diastolic regional collapse of the right atrium or the RV is the most common echocardiographic finding in "early" cardiac tamponade.(figure 6) Postoperative right atrial hematoma often become localized to the anterior and lateral walls, whereas left atrial clots are more commonly found behind the left atrium where they become encysted in the oblique sinus. The clinical presentation may be mistaken for congestive heart failure, acute LV or RV infarction, septic shock, or pulmonary embolism. Therefore, the diagnosis of postoperative cardiac tamponade should be considered whenever hemodynamic deterioration is encountered, particularly when reduction in cardiac output and/or blood pressure are not readily responsive to conventional management. Further, in delayed tamponade, the complaints tend to be vague and less hemodynamic data is available (i.e., PAC) to allow prompt diagnosis of tamponade.

The diagnosis of cardiac tamponade following cardiac surgery is often difficult and requires a high degree of clinical suspicion, proficient knowledge of pulmonary artery catheter (PAC) derived measures, physical examination, and diagnostic tools such as echocardiography and chest roentnography. The classical teaching of equalization of diastolic blood pressures in cardiac tamponade (CVP = PAD = PAOP) is infrequently observed postoperatively because the pericardium is left open: often only the CVP is elevated, while the PAD and the PAOP can be normal, elevated or, in some cases, reduced. The work up of cardiac tamponade can be further facilitated by several characteristic clinical and laboratory. Electrocardiography may reveal nonspecific ST-T wave abnormalities, low voltage QRS complex, signs of myocardial ischemia and pericarditis, and electrical alternans. The latter is seen in large effusions and is characterized by inter-beat shifts in electrical axis. This beat-to-beat alteration may by due to increased distance between the heart and the chest wall resulting in a "pendulum" like motion of the heart. This finding, although sensitive for cardiac tamponade, is not very specific (very few patients with tamponade present with electrical alternans). On chest x-ray the cardiac silhouette appears "widened" with or without features such as obscuring of the pulmonary vessels at the hilium and a globular or "water bottle" configuration of the heart [12].

Echocardiographic assessment

TEE usually overcomes many of the imaging constraints of TTE echocardiography: the surgical site may preclude use of the optimal transthoracic window, chest tubes can affect imaging and proper positioning of the patient, compression of the chest wall with the probe may worsen incisional pain, while some loculated effusions may be difficult to image [13]. Pericardial effusion is graded as small if the visceral and parietal pericardial layers are separated by less than 0.5 cm, moderate by 0.5 – 2 cm, and large when greater than 2 cm [14]. When the effusion is significant, ventricular function can be normal or depressed, particularly if the pericardial pressure overwhelms ventricular diastolic pressures adding to resistance to myocardial and endocardial blood flow. Of note, absence of pericardial contents around the LV should not necessary exclude the absence of pericardial collection around other heart structures, such as, the RV or atria (figure 6). The right atrium is a thin and flexible structure, and brief wall inversion can occur in the absence of tamponade physiology. That is why the specificity of right atrial systolic

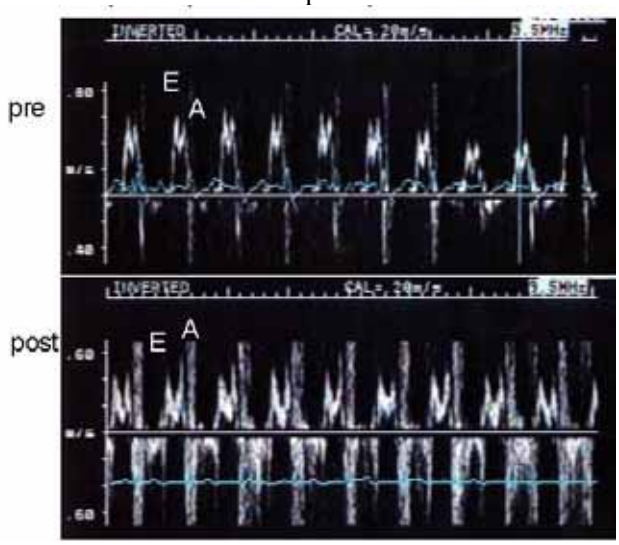
Figure 8: Right ventricular collapse in cardiac tamponade



collapse for tamponade increases if the systolic compression continues for 1/3 of the cardiac cycle (figure 7) [15,16]. RV collapse may also occur when the pericardial pressure exceeds RV pressure. This occurs during diastole, when the RV pressure is at its lowest. Diastolic RV collapse is more specific than right atrial collapse for confirming tamponade and can be identified by an abnormal posterior motion of the anterior RV wall during diastole [17,18]. This is readily apparent on M-mode and 2-D echocardiography (figure 8). It is important to note that RV collapse may not occur in the presence of RV hypertrophy with significantly elevated RV end diastolic pressure, or when a regional process such as blood clot is compressing the heart at an area distant from the RV. Pulsed-wave Doppler may reveal respiratory variation of the RV and LV inflow (hepatic vein and trans-tricuspid, and PV and transmitral flows respectively) [19]. Of note, the ventricular filling pattern is normal with impaired relaxation (figure 9).

Experimental and clinical studies have shown that tamponade is not an all-or-none phenomenon but a continuum from slight elevations of pericardial pressure to those associated with hemodynamic collapse. There is no echocardiographic sign that is 100 % sensitive and specific. The diagnosis is mainly a clinical one. Echocardiography however with a combination of M-mode, two-dimensional and Doppler techniques is the tool of choice for confirming a clinical suspicion.

Figure 9: Doppler echocardiography pre and post evacuation of pericardial effusion



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