

Chapter 2

Hemodynamics and Heart Failure

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Introduction

Every year in the United States, there are approximately 550,000 newly diagnosed heart failure patients. Five million patients suffer from chronic heart failure. Acute heart failure exacerbation is the leading cause for hospitalization in Medicare patients over the age of 65. A fundamental understanding in the definition, etiology, pathophysiology and hemodynamics has led to advances in treatments.

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Definition

The complete definition of heart failure is not confined solely to the heart, but involves a complex interplay between the heart and other organs. Definitions in heart failure have mainly focused on impaired pump function and clinical manifestations of venous congestion. Katz [1] states “*heart failure is a clinical syndrome in which heart disease reduces cardiac output, increases venous pressures, and is accompanied by molecular and other abnormalities that cause progressive deterioration of the failing heart*”.

Etiology

The end result of heart failure is caused by a multitude of disease abnormalities. Damage to the heart varies in its clinical presentation, systemic effects on the body, in treatment and prognosis. Ischemic heart disease, pulmonary hypertension, systemic hypertension, primary heart muscle abnormalities and valvular abnormalities are a few of the causes of heart failure (Table 2.1) [2].

Clinical Presentation

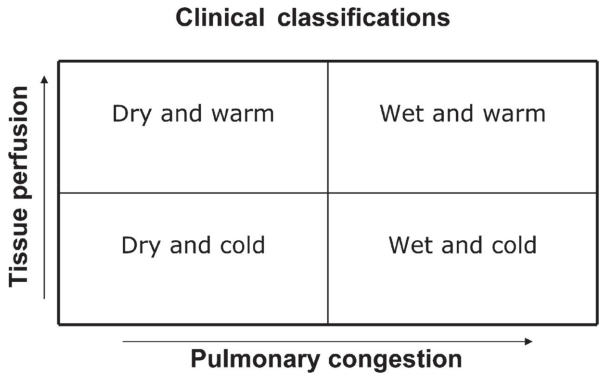
Heart failure can be classified as acute or chronic, compensated or decompensated, and combinations of these variables. History and physical examination skills are paramount to diagnosis in patients presenting with heart failure and their correlation

Table 2.1 Common etiologies of heart failure

Coronary heart disease	Acute coronary syndromes
Hypertension	Often associated with left ventricular hypertrophy and preserved ejection fraction
Cardiomyopathies	Familial/genetic or non-familial/non-genetic (including acquired, e.g. myocarditis)
	Hypertrophic (HCM), dilated (DCM), restrictive (RCM), arrhythmogenic right ventricular (ARVC), unclassified
Drugs	β -Blockers, calcium antagonists, antiarrhythmics, cytotoxic agents
Toxins	Alcohol, medication, cocaine, trace elements (mercury, cobalt, arsenic)
Endocrine	Diabetes mellitus, hypo/hyperthyroidism, Cushing syndrome, adrenal insufficiency, excessive growth hormone, pheochromocytoma
Nutritional	Deficiency of thiamine, selenium, carnitine. Obesity, cachexia
Infiltrative	Sarcoidosis, amyloidosis, haemochromatosis, connective tissue disease
Others	Chagas' disease, HIV infection, peripartum cardiomyopathy, end-stage renal failure

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Fig. 2.1 Correlation of hemodynamic profile and invasive hemodynamic findings (Reprinted from Dickstein et al. [2], © 2008, by permission of Oxford University Press)



to invasive hemodynamic alterations. Since the results from the Evaluation Study of Congestive Heart Failure and Pulmonary Artery Catheterization Effectiveness (ESCAPE) trial, the routine use of the invasive pulmonary artery catheter has largely fallen out of favor. Although the trial demonstrated no changes in primary endpoint with the routine utilization of right heart catheterization, there were correlations in the accuracy of jugular venous pressure (JVP) and right atrial pressure. An elevation of left ventricular filling pressures was associated with the findings of orthopnea and increased JVP. Discharge assessment of fluid status via elevated JVP and orthopnea (“wet”) or decreased cardiac output with reduced perfusion (“cold”) correlated with a 50 % increase in risk of death or re-hospitalization at 6 months [3, 4]. Invasive pulmonary artery catheter hemodynamic assessment is utilized to aid in the understanding and diagnosis of pathophysiology in patients that do not respond to typical initial treatments (Fig. 2.1) [2]. Recognition of these hemodynamic characteristics can lead to alteration in therapeutic decision making.

Acute Decompensated Heart Failure

A decrease in EF due to ischemia or infarction results in primary pump failure. Acutely depressed LV function results in depressed cardiac output and venous congestion.

As the heart suffers a decrease in pump capacity there is an increase in the pressures of the venous system. The increase in venous pressure is a result of the inability of the heart to adequately accept the blood returning to the heart. Right and left atrial pressures increase. These hemodynamic alterations from a decrease in cardiac output occur within a few seconds. With the inability of a low cardiac output to provide adequate systemic perfusion, there is a response in the compensatory mechanism that results in an increase in sympathetic nervous tone.

Sympathetic stimulation results from a complex system of neurohormonal feedback. A decrease in pump function results in lower systemic arterial pressure that in turn activates the baroreceptor reflex mechanism. Ischemic heart responses,

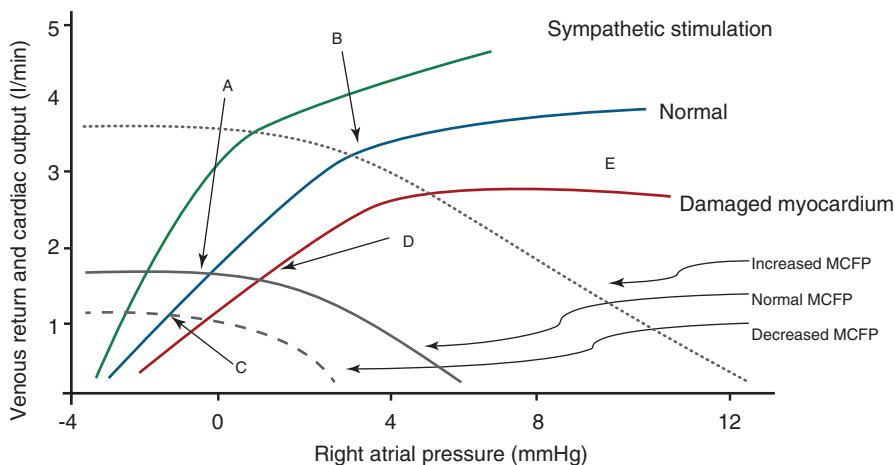


Fig. 2.2 Cardiac output and right atrial pressure relationship with varying myocardial functional states (Reprinted from Guyton [5], © 1955, with permission from The American Physiological Society)

intracardiac reflexes, and other components of this feedback system contribute to sympathetic nervous system activation. Parasympathetic inhibition and sympathetic stimulation occurs within a few seconds to compensate for the acute fall in cardiac output. As sympathetic stimulation occurs, the target and main effects are to the peripheral vasculature and the heart. An increase in cardiac function occurs with sympathetic stimulation to increase the recruitment of cardiac reserves within the normal and the remaining partially functional damaged myocardium. If there is diffuse damage to the ventricular myocardium during an ischemic insult, there is strengthening in the remaining functional myocytes via sympathetic stimulation. If there is no function of a portion of the ventricle, sympathetic stimulation results in the stimulation of the remaining normal myocardium. The normal myocardium attempts to compensate for the shortcomings of the damaged myocardium.

In addition to increasing myocardial muscle function, sympathetic stimulation leads to changes in the peripheral vasculature. This increase in tone in the peripheral vessels, leads to an increase in venous return. The mean systemic filling pressures are elevated increasing the flow from the venous system to the heart. The increased flow leads to increased filling of the damaged heart that in turn leads to increase in priming the heart to aid in pump function. Less than a minute is needed for the sympathetic nervous system to be completely activated (Fig. 2.2) [5, 6].

Chronic Heart Failure

The ischemic insult is followed by no, partial, or full recovery over weeks to months. In addition to ventricular myocardial recovery, fluid retention via renal mechanisms also occurs to compensate for this new cardiac pump status and alters the normal

Table 2.2 Normal hemodynamic parameters

Normal hemodynamic parameters		Pressure (mmHg)
Right atrium	<i>a</i> -wave / <i>v</i> -wave / mean	1–7 / 1–7 / 0–5
Right ventricle	Systolic / end diastolic	17–32 / 1–7
Pulmonary artery	Systolic / end diastolic	17–32 / 1–7
Left atrium	<i>a</i> -wave / <i>v</i> -wave / mean	4–12 / 4–15 / 4–15
Left ventricle	Systolic / end diastolic	90–140 / 5–12
Aorta	Systolic / end diastolic / mean	90–140 / 60–90 / 70–105

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physiologic hemodynamics found in invasive assessments (Table 2.2) [7]. Renal function is extremely sensitive to alterations in perfusion. A low cardiac output state can lead to a decline in renal function manifested to the point of anuria. The decrease in urine output can persist until there is normalization in systemic blood pressure and cardiac output.

Blood volume is altered via renal mechanisms to affect cardiac function. Initially, moderate retention of fluid results in an increase in blood volume that is beneficial to the diminished pumping function of the heart. The increase in fluid retention increases venous return, thereby increasing blood flow to the heart.

As the damaged heart receives the increased venous return, there is a gain of cardiac function. If cardiac output becomes too low, the kidneys respond with the inability to excrete adequate amounts of sodium and water. Excess fluid retention is no longer beneficial to myocardial function, and only serves to increase cardiac workload in the damaged heart and manifests as edema.

Extravasation of fluid from the pulmonary vasculature leading to hypoxia from pulmonary edema, and systemic edema develops in various organs and contributes to their dysfunction. Myocardial functional recovery can range from full to none. After partial recovery there is fluid retention that occurs to establish a new hemodynamic state. The increase in blood volume results in an increase in venous return that provides assistance in the pumping function of the heart. The elevated venous pressure persists as the cardiac output improves. As this new steady state is established and this resting cardiac output improves, the sympathetic tone progressively abates over several weeks following an acute ischemic insult. Altered renal function that results in fluid retention persists in this new hemodynamic state. As the pumping function of the heart compensates, the sympathetic tone begins to gradually decrease transitioning from an acute phase to a chronic heart failure state.

Compensated and De-compensated Chronic Heart Failure

As partial recovery of the ventricular myocardial function occurs, the resting pump output from the heart normalizes with the help of an increase in atrial pressure. This increased filling pressure helps in recruitment of myocardium and improved output

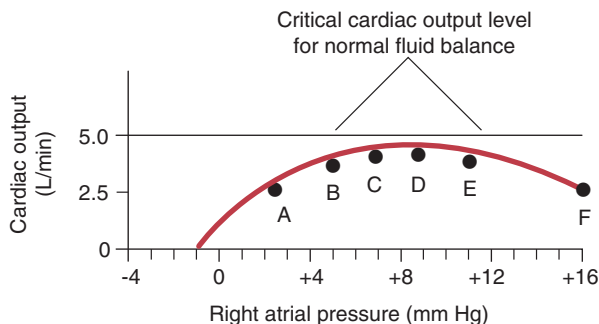


Fig. 2.3 The rise and fall of cardiac output as right atrial pressure increases. Further increase in RAP leads to a decline in cardiac output (Reprinted from Hall [8], © 2016, with permission from Elsevier)

in the resting state. As a patient begins to exercise, the already maximized heart lacks reserve and symptoms of heart failure return. The lack of cardiac reserve is a common occurrence in heart failure patients as they achieve a resting compensated state and attempt to demand more cardiac output with exercise or systemically stressed state.

In severe cardiac failure, de-compensation occurs as a consequence of the inability of the heart to provide additional cardiac output when there are increased systemic demands. Neither sympathetic stimulation nor fluid retention can increase cardiac output to normal. Fluid retention results as the heart is unable to provide sufficient blood flow to the kidneys to excrete sodium and water.

Correlation of cardiac output on the y axis and the atrial filling pressures on the x axis is represented in Fig. 2.3 [8]. As a poorly functional ventricle responds to gradually increasing filling pressures, the cardiac output rises. After a certain point of maximal myocardial stretch, cardiac output falls and higher filling pressures no longer provide additional aid in cardiac function. A progressive increase in fluid retention increases filling pressures beyond the ideal ventricular size and dilatation with overstretching ensues. As progressive increases in fluid retention occur, the mean systemic filling pressures are translated to the heart, which then leads to the gradual rise and fall of cardiac output.

If the cardiac output never reaches a point of providing sufficient perfusion, specifically to the kidney, then cardiac failure is imminent, leading to systemic edema and pulmonary edema, hypoxia, pump failure and eventually death.

Pathophysiology

The Frank-Starling mechanism or Starling's Law of the heart dictates that with increasing volume in the heart there is an increase in myocardial performance that includes an increased stroke volume (Fig. 2.4) [9]. This is a manifestation of the

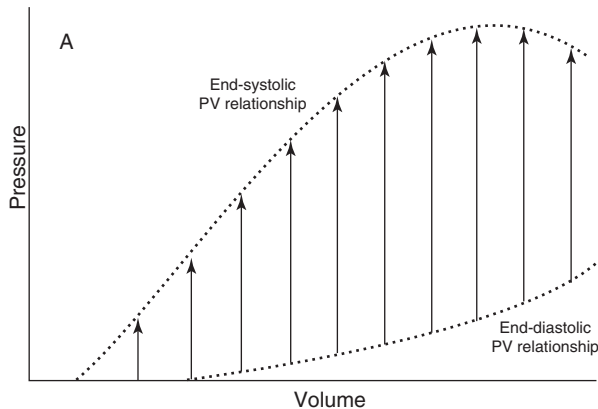


Fig. 2.4 Pressure-volume relationship within the left ventricle. As end diastolic volume increases, resulting end diastolic and systolic pressures increase (Reprinted with permission from Katz [9], © 2011, with permission from Wolters Kluwer Health)

sarcomere length-tension relationship. As the ventricle fills with blood there is distention of cardiac myocytes and less sarcomere overlap. As the myocytes are “stretched”, the heart is able to increase the volume of blood it ejects. After the optimal point of overlap, the ventricle can be overstretched leading to a decrease in the amount of volume ejected from the heart. The ascending portion of the Starling curve illustrates how the increase in preload leads to the increase in cardiac output. Ventricular over filling can be detrimental. End diastolic pressures rise, and the overly “stretched” myocardium transitions to the descending portion of the Starling curve and a decrease in stroke volume and systolic pressure.

Valvular Heart Failure

Normal valvular function provides a mechanism for unidirectional flow without resistance. The limitation of blood flow during diastole or systole is caused by a stenosis in the atrio-ventricular or ventricular-arterial valves respectively.

Valvular Stenosis

Aortic stenosis and pulmonic stenosis result in a decrease in cardiac output due to increased resistance to emptying of the ventricle. This increase in resistance to cardiac output results in a measurable pressure gradient across the valve which can be measured via a dual lumen pigtail catheter, separate aortic root and left ventricular catheters, or arterial sheath and left ventricular catheter. In aortic stenosis with

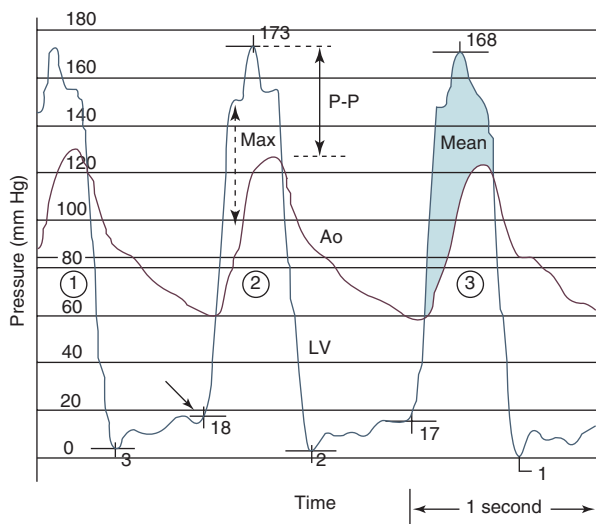


Fig. 2.5 Transaortic pressure gradient. The gradient between the left ventricle (LV) and the aorta (Ao) in aortic stenosis can be described by three invasive measures. The mean gradient (beat #3) represents the area under the left ventricular–aortic pressure curve. The peak-to-peak (P-P) gradient (beat #2) is the difference between the maximum aortic pressure and the maximum left ventricular systolic pressure. The maximum (*Max*) gradient (beat #2) is the maximum difference that can be measured between the left ventricle and aorta during systole (Reprinted with permission from Shavelle [10], © 2014, with permission from Elsevier)

preserved left ventricular function, as the severity in aortic valvular stenosis increases, there is an increase in the LV chamber pressure generation. Aortic and left ventricular pressure tracings are used to measure peak to peak, maximum, and mean pressure gradients (Fig. 2.5) [10]. With the advent of increased diagnostic accuracy of echocardiography, the utilization of direct hemodynamic measurement is most strongly indicated when there is a discrepancy between clinical and echocardiographic findings.

After analysis of hemodynamic tracings, the Gorlin formula is utilized in the calculation of aortic valve area. Special circumstances with decreased systolic function, “low-output, low gradient aortic stenosis”, are a subset of patients that pose diagnostic dilemmas.

Differentiation of *pseudo* aortic stenosis from true aortic stenosis in the setting of decreased cardiac output is crucial in effective management of the patient. Three particular scenarios are made apparent during dobutamine infusion with simultaneous aortic and left ventricular hemodynamic tracings obtained.

Illustrated in Fig. 2.6 is the potential findings during dobutamine challenge in patients with “low-output, low gradient” aortic stenosis [10, 11].

The far left clinical scenario both cardiac output and aortic valve mean gradient increase as a result of dobutamine infusion, thus true aortic stenosis. The middle scenario finds an increase in cardiac output with no dramatic increase in aortic valve

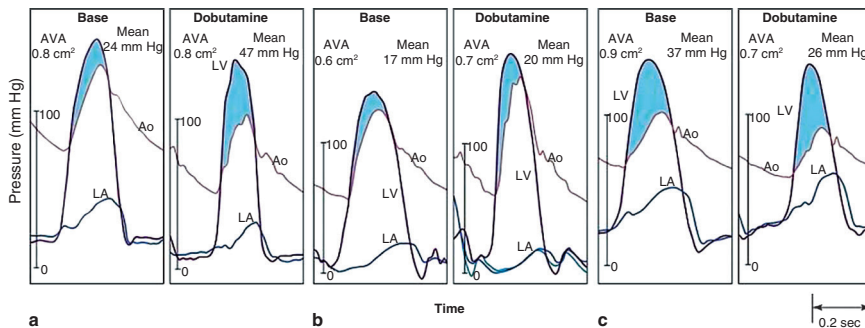


Fig. 2.6 Differentiation of three invasive measurement scenarios as a result of dobutamine infusion in “low-output, low gradient” aortic stenosis (Reprinted with permission from Nishimura et al. [11], © 2002, with permission from Wolters Kluwer Health. And reprinted with permission from Shaville [10], © 2014, with permission from Elsevier)

pressure gradient, a finding of mild aortic stenosis. In the right most clinical scenario, there was no change in the aortic valve pressure gradient as a result of dobutamine infusion, truly severe aortic stenosis.

In addition to valvular stenosis that limits cardiac output, is valvular stenosis that limits cardiac filling. Mitral and tricuspid valvular stenosis affects the ability to provide adequate chamber preload. Hemodynamic findings result in an elevation in PCWP but inaccurately reflect LVEDP. PCWP in mitral stenosis is reflection of left atrial pressure but not left ventricular end diastolic pressure. Mitral stenosis results in a pressure gradient between the left atrium and the left ventricle. The classic finding on hemodynamic tracings is the elevation of pulmonary pressures, prominent “a” and “v” waves on PCWP (Figs. 2.7a, b) [12]. Simultaneous tracings within the left ventricle reveal an evident pressure gradient between PCWP and LVEDP.

One must take into account the temporal delay as the pressure of the fluid column is transmitted to the right heart catheter from the left atrium through the pulmonary vascular bed. It is therefore ideal to obtain a direct measurement of left atrial pressure via septal puncture with simultaneous left ventricular hemodynamic tracings (Fig. 2.7c) [13].

Valvular Regurgitation

Aortic valvular regurgitation results in an increase in LV diastolic pressures. Acute aortic regurgitation results in the acute volume overload of the left ventricular chamber, this in turn leads to pulmonary edema, premature closure of the mitral valve and can also result in systemic shock (Fig. 2.8) [12].

Chronic aortic regurgitation results in a wide pulse pressure, high systolic and low diastolic pressures. In chronic, compensated aortic valve regurgitation, the left ventricle and systemic hemodynamics have had time to adjust, thus resulting in a

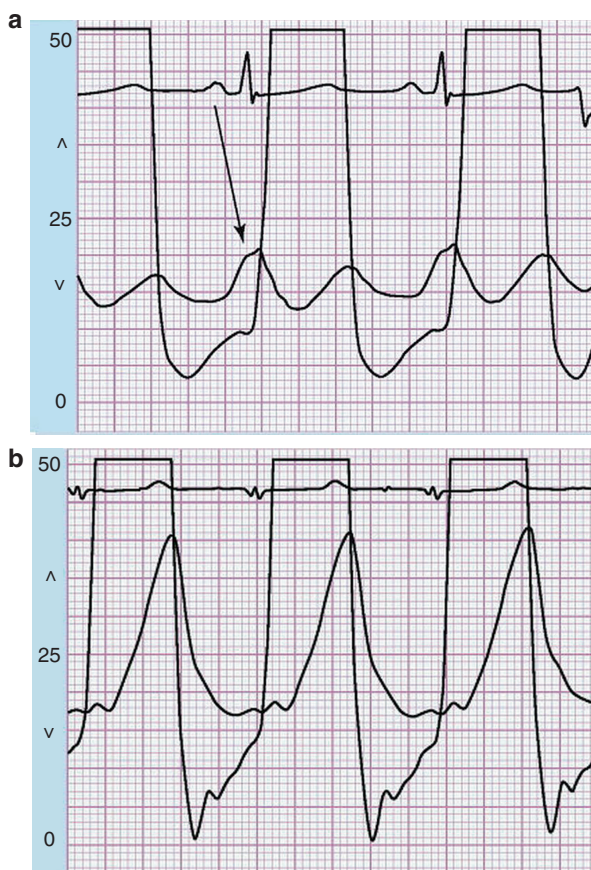


Fig. 2.7 (a) The “a” wave on a left atrial (shown here) or pulmonary capillary wedge pressure is accentuated in mitral stenosis (Reprinted with permission from Ragosta [12], © 2010, with permission from Elsevier) (b) The “v” wave may also be markedly increased in patients with mitral stenosis. This patient with mitral stenosis has no mitral regurgitation and normal systolic function (Reprinted with permission from Ragosta [12], © 2010, with permission from Elsevier) (c) Measurement of the transmitral gradient by cardiac catheterization is frequently made with a simultaneous pulmonary artery wedge pressure (PAWP) and left ventricular (LV) pressure. However, as a result of the delay in transmission of the change in pressure contour and a phase shift, the gradient using a pulmonary artery wedge pressure will frequently overestimate the true transmitral gradient. *Left*, Simultaneous left ventricular and pulmonary artery wedge pressure in a patient with mitral stenosis. The measured mean gradient is 15 mmHg. *Right*, In the same patient, the transmitral gradient is measured with a left ventricular and direct left atrial (LA) pressure. The true mean transmitral gradient is only 6 mmHg (Reprinted with permission Nishimura and Carabello [13], © 2012, with permission from Wolters Kluwer Health)

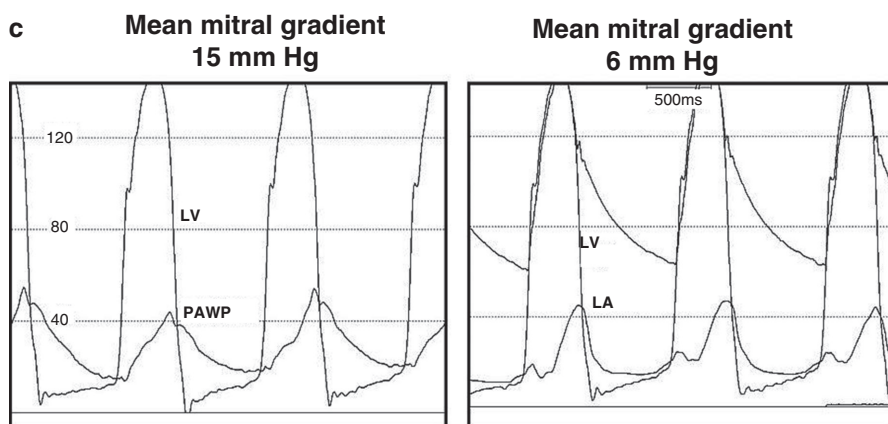
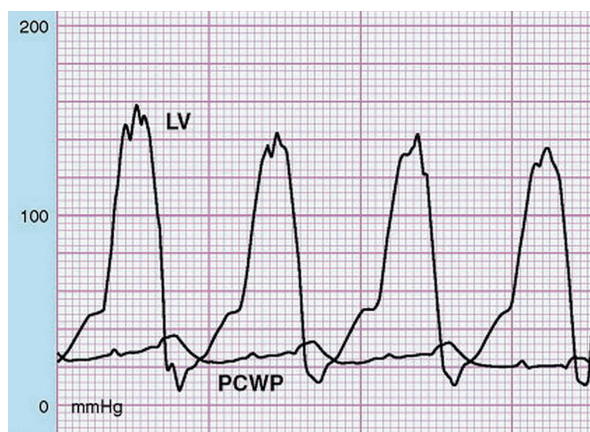


Fig. 2.7 (continued)

Fig. 2.8 Acute severe aortic valvular regurgitation resulting in LVEDP > PCWP
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normal early LVEDP. As the cardiac cycle approaches the end of the diastolic filling period, the LVEDP approaches the level of diastolic pressure. The rise of LVEDP to meet diastolic pressure is known as diastasis (Fig. 2.9) [10].

Similarly, chronicity of mitral valvular regurgitation determines the findings on hemodynamic tracings. Acute mitral valvular regurgitation results in pulmonary edema and prominent “v” waves on PA and PCWP hemodynamic tracing illustrated with simultaneous Aortic hemodynamic tracings (Fig. 2.10) [14].

Chronic valvular regurgitation may not manifest with the classic finding of prominent “v” waves on PCWP and pulmonary artery tracings as in acute mitral regurgitation. The presence of prominent “v” waves is determined by the compliance of the left atrium in addition to its size and pressure. Given this, chronic mitral regurgitation and may manifest as normal physiologic “v” waves on hemodynamic tracings.

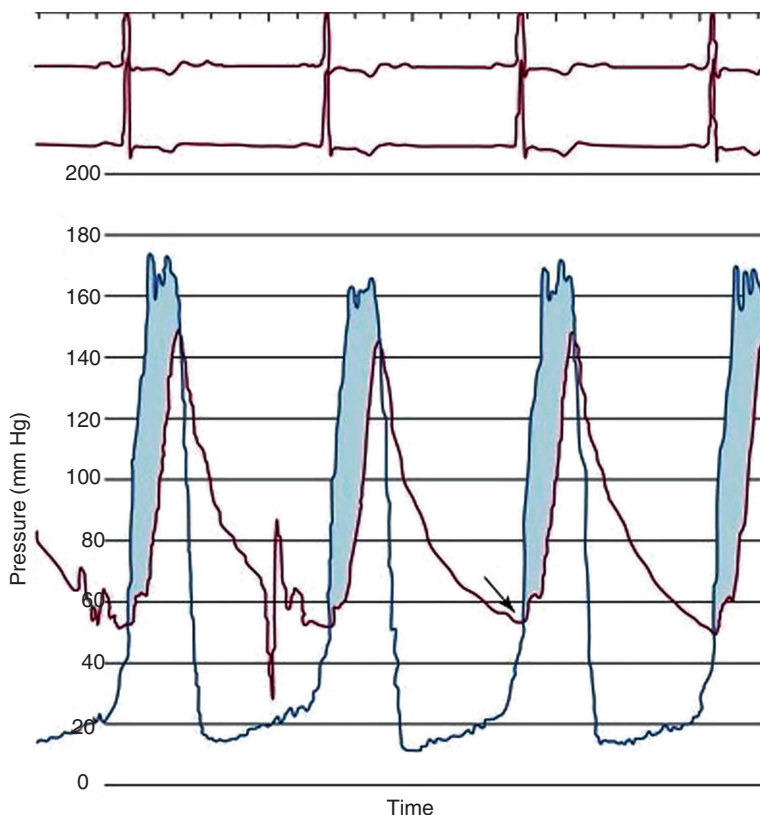


Fig. 2.9 Hemodynamics in severe aortic regurgitation. Simultaneous aortic and left ventricular pressures in a patient with mild aortic stenosis and severe aortic regurgitation. Note that the pulse pressure is wide (approximately 100 mmHg) and the aortic diastolic pressure (*arrow*) is low (Reprinted with permission from Shavelle [10], © 2014, with permission from Elsevier)

Unilateral Heart Failure

There are unique clinical presentations of unilateral heart failure that alter the hemodynamics related to the affected ventricle, specifically left sided heart failure in absence of right sided heart failure and vice versa.

Left sided heart failure in isolation, with normally functioning right sided ventricle leads to an increase in mean pulmonary artery pressures. In the setting of inadequate left ventricular pump function, the blood accumulates in the pulmonary vasculature and the backup in volume shifts from the systemic arterial circulation to the pulmonary vasculature. Pulmonary vasculature capillary pressures rise as a consequence of increased volume. Fluid accumulation in the pulmonary bed gradually rises to a point where the capillary network can no longer tolerate the additional fluid and begins to leak out into the interstitium. With the rise in pulmonary

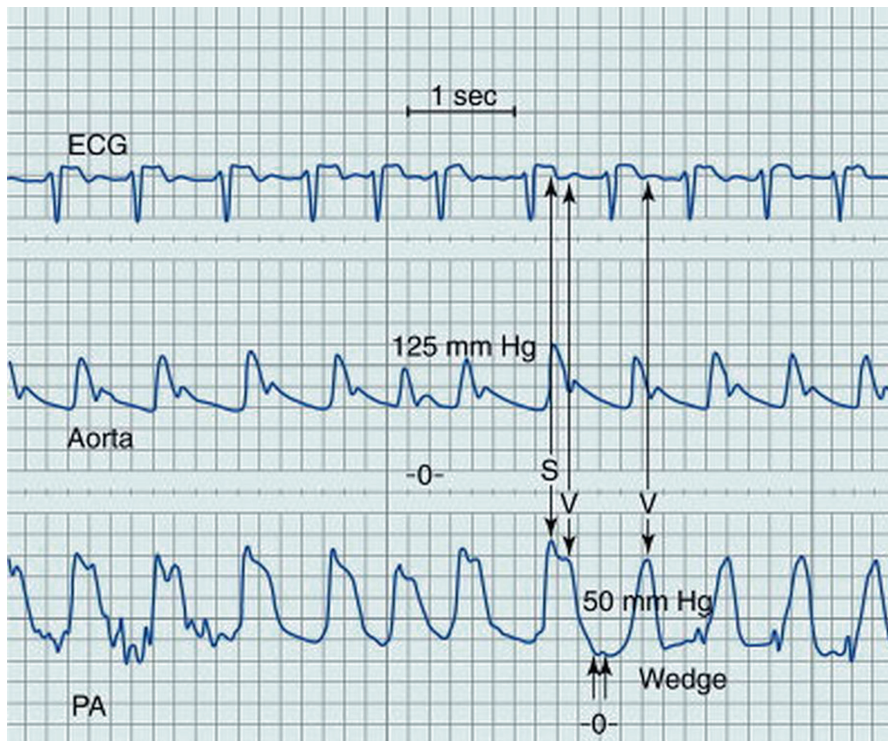


Fig. 2.10 Acute severe mitral regurgitation. Electrocardiogram (ECG), aortic (Aorta), pulmonary artery (PA) pressure (*left*), and pulmonary capillary wedge (wedge) pressure (*right*) tracings in a patient with acute severe mitral regurgitation. A prominent v wave is present in both the pulmonary artery and wedge pressure tracings. The pulmonary artery pressure is bifid because of the presence of both the pulmonary artery systolic wave (S) and the v wave. The large v wave can cause the wedge tracing to be confused with a pulmonary artery tracing (Reprinted from Sharkey [14], © 1987, with permission from Elsevier)

pressures beyond the ability of the capillary bed to hold in the fluid via osmotic mechanisms, pulmonary edema is manifested clinically as rales and dyspnea. The clinical presentation of severe pulmonary edema in the setting of acute myocardial infarction and cardiogenic shock can be dramatic resulting in profound hypoxia in less than an hour.

Isolated right ventricular failure results in a gradual increase in systemic venous pressures and a loss of preload to the left ventricular side. Right and left ventricular interdependence is a key concept in understanding the pathophysiologic alterations in hemodynamics of isolated right ventricular dysfunction. In acute right ventricular infarction, the systolic function of the pump is compromised. As right sided ventricular dysfunction occurs, right sided chamber dilatation along with a fall in systolic pressures occurs. The diastolic filling pressure of the ventricle rises and demonstrates a characteristic prominent “y” descent manifested as a dip and plateau on hemodynamic tracings (Fig. 2.11) [15].

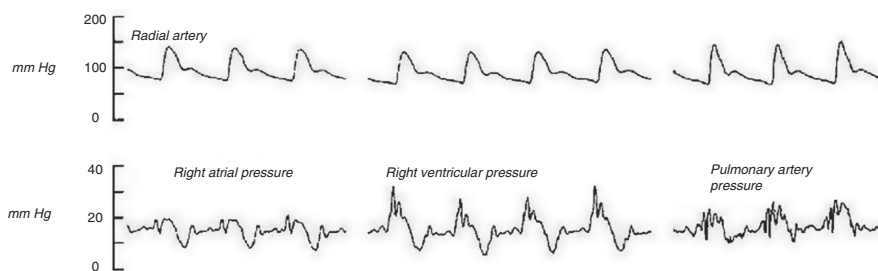


Fig. 2.11 Right ventricular infarction hemodynamic tracings (Reprinted from Lorell et al. [15], © 1979, with permission from Elsevier)

The depressed ventricular function leads to an increase in right atrial filling pressures and a backup into venous system. Acute right ventricular chamber dilatation encroaches upon the left ventricular chamber filling. As the shared space within the pericardium is fixed, acute RV dilation results in a decrease in LV chamber filling. The enhancement of the right and left interdependence is manifested dramatically as hypotension, clear lung fields, prominent JVD all mimicking the syndrome of pericardial constriction. Experimental models of isolated RV infarct have demonstrated the inability to induce hypotension in animals where the pericardium is surgically absent. Without acute reperfusion and hemodynamic support RV infarction can be life threatening.

Constrictive Pericarditis and Restrictive Cardiomyopathy

Restrictive cardiomyopathy is a rare form of cardiomyopathy in which the ventricle becomes abnormally rigid and lacks the flexibility to adequately expand as it fills with blood. This leads to venous congestion and heart failure.

It has distinct morphologic features allowing it to be separated from other cardiomyopathies. For example with restriction the left ventricle is usually normal in size and function unlike dilated or hypertrophic cardiomyopathies. A syndrome that poses a diagnostic dilemma with restrictive cardiomyopathy is constrictive pericarditis. They both exhibit normal ventricular size and function as well as some similar hemodynamic abnormalities.

It is extremely important for the clinician to be able to distinguish between constrictive pericarditis and restrictive cardiomyopathy since they require markedly different treatment. In some cases, invasive cardiac catheterization may be necessary to help differentiate the two. In addition the hemodynamic features of restrictive cardiomyopathy and constrictive pericarditis can be similar to tamponade and right ventricle infarction. This section will discuss the hemodynamics of these entities and how the hemodynamics can be used and differentiate between them.

Constrictive Pericarditis

Constrictive pericarditis is the end-stage of an inflammatory process affecting the pericardium, resulting in scarring and subsequent loss of the normal elasticity of the pericardial sac. In the developed world idiopathic or viral illness is the most common cause followed by post-surgical and radiation therapy (Table 2.3) [16–19].

Pathophysiology

The pericardium is a fluid filled fibro-elastic sac that surrounds the heart. It functions to keep the heart contained in the chest cavity and stretches to accommodate physiologic changes in cardiac volume [20, 21]

In constrictive pericarditis, the total cardiac volume is fixed as a result of the non-compliant shell around the heart. The abnormal pericardium prohibits outward ventricular expansion, which is necessary to accommodate venous return. As a result, an adaptive process occurs in which the inter-ventricular septum bulges inward either into the left or right ventricle in order to accommodate myocardial blood flow into the adjacent ventricle. This maladaptive process will allow filling in one ventricle while compromising blood flow into the other. This is known as ventricular interdependence, since the amount of blood flow into one ventricle is dependent on the amount of blood flow into the other.

Ventricular interdependence is the result of the rigid pericardium, which prevents the normal reduction in intra-thoracic pressure during inspiration from being transmitted to the heart chambers. As a result during inspiration pulmonary venous pressure will decrease while left ventricular pressure remains constant. This ultimately leads to a reduction in the gradient driving filling into the LV. Reduction in LV volume allows RV filling to occur. As the RV fills, the inter-ventricular septum will

Table 2.3 Causes of constrictive pericarditis

Idiopathic
Irradiation
Postsurgical
Infectious
Neoplastic
Autoimmune(connective tissue) disorders
Uremia
Post trauma
Sarcoid
Methylsergide therapy
Implantable defibrillator patches

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shift toward the left further reducing LV filling. This septal shift occurs because outward expansion is not possible due to the constraining effects of the rigid pericardium.

During expiration the opposite sequence will occur. Pulmonary venous pressures will rise resulting in an increased trans-pulmonary gradient into the LV. As LV fillings increases the inter-ventricular septum will bulge to the right resulting in decrease flow into the RV (Fig. 2.12) [19].

With constrictive pericarditis, early diastolic filling is preserved since it is a myocardial process and not dependent upon a functioning pericardium. In fact, early filling occurs even more rapidly than normal due to elevated atrial pressures and increased suctioning from the ventricle. However; in early to mid-diastole, ventricular filling will cease abruptly due to the non-compliant pericardium. Consequently, almost all filling occurs in early diastole and systemic venous congestion results causing peripheral edema, hepatic congestion, and ascites.

Typical constrictive physiology will be evident on right atrial and left ventricle pressure recording at the time of cardiac catheterization. During early diastole the ventricular pressure initially decreases rapidly causing a steep y descent on the right atrial pressure waveform. In mid to late diastole flow abruptly ceases which is evident as a plateau after the initial downward dip (Fig. 2.13) [19, 22].

In patients with constriction right and left heart pressure recording should be performed simultaneously. Typical measurements will reveal elevated and equal RA, RV diastolic, LV diastolic, and pulmonary wedge pressures. The fillings pressures are typically elevated at approximately 20 mmHg with no more than a 3–5 mmHg difference. The RA pressure will show a preserved x descent and prominent y descent. Both

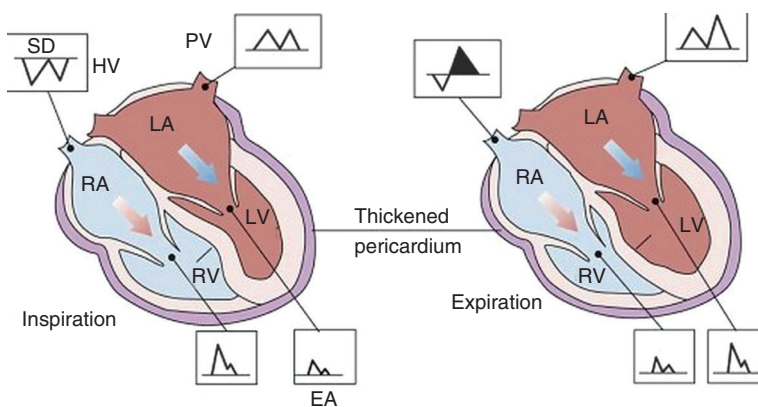


Fig. 2.12 Schematic representation of transvalvular and central venous flow velocities in constrictive pericarditis. During inspiration the decrease in LV filling results in a leftward septal shift that allows augmented flow into the right ventricle. The opposite occurs during expiration. *D* diastole, *EA* mitral inflow, *HV* hepatic vein, *LA* left atrium, *LV* left ventricle, *PV* pulmonary venous flow, *RA* right atrium, *RV* right ventricle, *S* systole (Reprinted from LeWinter and Hopkins [19], © 2015, with permission from Elsevier)

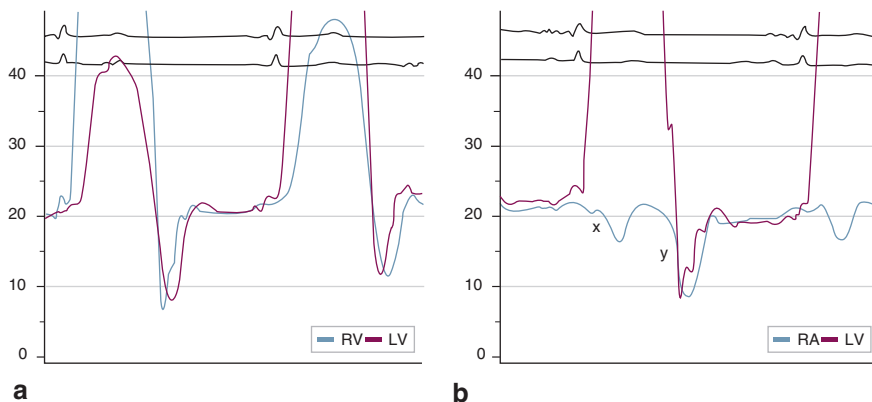


Fig. 2.13 Pressure recording in a patient with constrictive pericarditis. **(a)** Simultaneous LV and RV pressure tracing with the “dip-and-plateau waveform” or “square root sign” seen on right or left ventricular pressure waveform tracings. The black arrow represents the rapid descent, and the white arrow represents the plateau portion. **(b)** Right atrial pressure with prominent Y descent (Reprinted from Vaitkus et al. [22], © 1996, with permission from Wolters Kluwer Health, Inc. and Reprinted from LeWinter and Hopkins [19], © 2015, with permission from Elsevier)

RV and LV pressure tracings will show a early diastole dip followed by a plateau in mid to late diastole. Pulmonary hypertension is not usually a feature of constriction. The PA pressures are typically between 35-40 mmHg. If hypovolemia is present rapid volume challenge of 1000 cc may help to reveal typical hemodynamic features, which can be masked when fillings pressures are low [21, 23].

Clinical Presentation

Constrictive pericarditis can present with a myriad of symptoms, thus making a diagnosis solely on the basis of the clinical history is difficult. The usual presentation consists of predominantly right heart failure. Dyspnea is the most common presenting symptom and occurs in virtually all patients. Fatigue and orthopnea are common. Lower extremity edema and abdominal swelling are also common. Constrictive can mimic and be mistaken for other causes of right heart failure as well as end-stage liver disease. However it is important to note that in primary liver disease jugular venous pressures are not elevated.

The vast majority of patients with constriction have elevated jugular venous pressure. Elevated JVP has been reported in as many as 93 % of patients with surgically confirmed constrictive pericarditis. A pericardial knock, which corresponds with the sudden cessation of ventricular filling occurs in approximately half the cases. Kussmaul’s sign, which is the lack of an inspiratory decline in JVP, can be present in patients with constrictive pericarditis, however it is nonspecific [17].

Table 2.4 Causes of restrictive cardiomyopathy

Hemochromatosis
Amyloidosis (most common cause in the United States)
Sarcoidosis
Scleroderma
Carcinoid heart disease
Glycogen storage disease of the heart
Radiation
Metastatic malignancy
Anthracycline toxicity

Restrictive Cardiomyopathy

Restrictive cardiomyopathy may be caused by various local and systemic disorders that may be categorized into 4 groups. They includes idiopathic, infiltrative, treatment-induced, and malignancy (Table 2.4) [24].

Clinical Presentation

Affected patients have signs and symptoms of pulmonary and systemic congestion resulting in dyspnea, peripheral edema, palpitations and fatigue. In advanced cases hepatosplenomegaly, ascites, and anasarca can occur from marked elevation in venous pressures.

Jugular venous pressure is generally elevated. Kussmaul’s sign, which is the lack of an inspiratory decline in JVP, can be present. Also a prominent y descent may appear; however, it may not be obvious in patients with mild disease. The cardiovascular examination is often indistinguishable from that of constrictive pericarditis [25, 26].

Constrictive Pericarditis Versus Restrictive Cardiomyopathy

The history, physical examination, and radiographic findings may suggest a particular diagnosis. For example constrictive pericarditis may be suggested in patients with prior radiation or cardiothoracic surgery. Restrictive cardiomyopathy is more likely in a patient with a predisposing systemic disease such as amyloidosis. However, in many instances their clinical presentation and course overlap, making the ability to distinguish between the two syndromes difficult.

In some cases, invasive cardiac catheterization may be necessary to help differentiate (Table 2.5) [19].

Table 2.5 Hemodynamic features of constrictive vs restrictive cardiomyopathy

	Constriction	Restriction
Prominent Y descent	Present	Variable
Filling pressures >25 mmHg	Rare	Common
Pulmonary systolic pressure >60 mmHg	No	Common
Square root sign	Present	Variable
Respiratory variation in LV and RV pressure	Discordant	Concordant

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In both conditions, RV and LV diastolic pressure will be elevated. However in restrictive cardiomyopathy, the difference between diastolic pressures of the LV and RV normally exceed 3–5 mmHg. In constriction those pressures usually are within 3–5 mmHg of each other. Pulmonary hypertension is rare in constriction but common in restriction. The absolute level of diastolic pressure in restriction commonly exceeds 25 mmHg whereas in constriction pressure rarely exceeds 20 mmHg. [27].

One of the most sensitive ways to differentiate rarely constrictive pericarditis from restriction is through the respiratory effects observed between the RV and LV systolic pressures. With constriction there is discordance between RV and LV systolic pressure with respiration due to ventricular interdependence. However, with restriction ventricular interdependence does not exist because the pericardium is normal. Thus in restriction RV and LV systolic pressure move concordantly with each other during the respiratory cycle (Fig. 2.14) [27].

Constriction Versus RV Infarction and Tamponade

Compressive hemodynamic effects resulting in diastolic dysfunction may be seen in other conditions besides constrictive pericarditis. In fact, any condition that causes elevated intra-pericardial pressure, i.e. from pericardial tamponade to abrupt chamber dilatation with RV infarction, will lead to increased intra-pericardial pressure and constrictive physiology. All three conditions including RV infarction may show elevation and equalization of diastolic filling pressures in the RA, RV, and PCW, as well as the LV [14, 15].

However, only constriction will exhibit dissociation between intra-thoracic and intra-cardiac pressures during the respiratory cycle (Fig. 2.15) [28]. Normally with inspiration the negative pressure created within the intra-thoracic cavity results in a simultaneous decrease in pulmonary and ventricular pressures. In constriction the rigid pericardium acts to isolate the heart leaving it unaffected from these physiologic changes. As a result only patients with constriction will exhibit dissociation between intra-thoracic and intra-cardiac pressures, which distinguishes it from RV infarction and tamponade where the LV and wedge pressures will rise and fall concordantly.

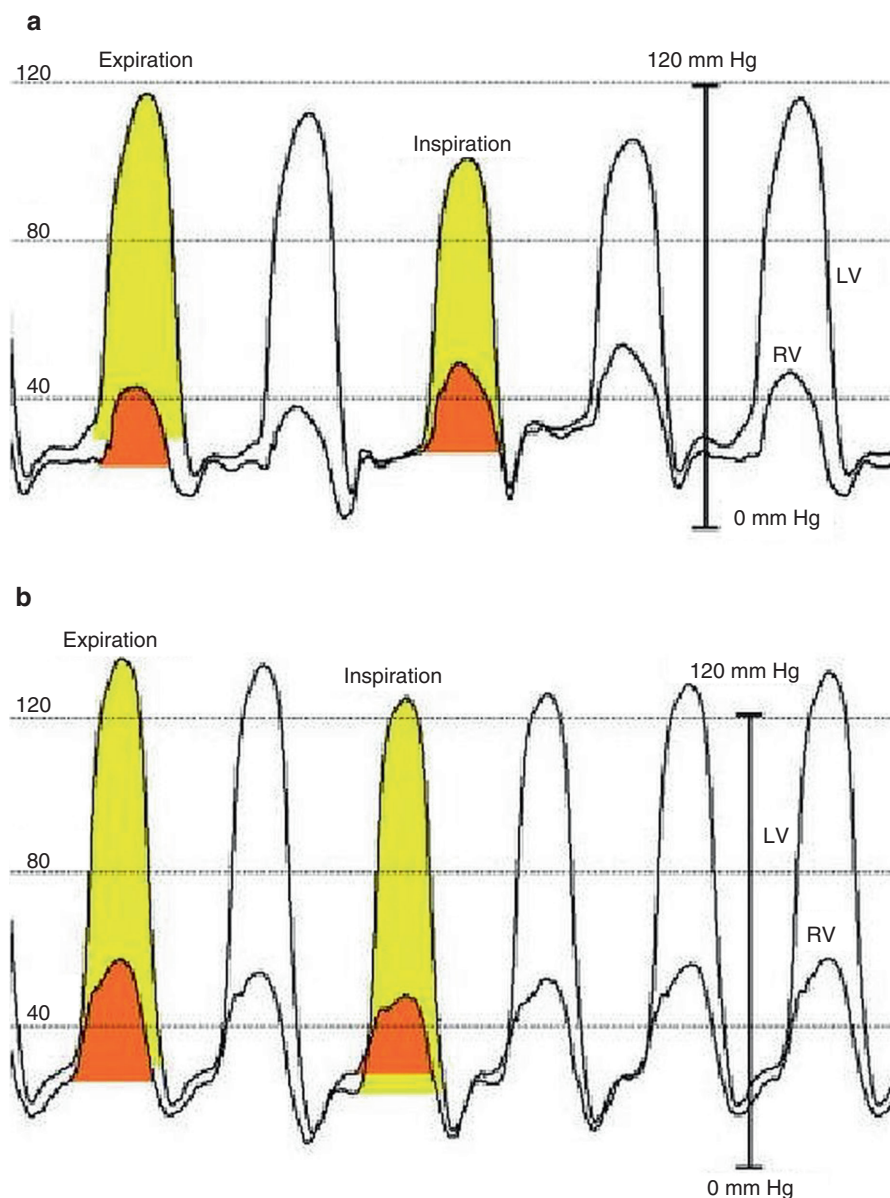


Fig. 2.14 LV and RV Pressure Recording From 2 Patients. During Expiration and Inspiration (a) With constrictive pericarditis LV and RV systolic pressure are discordant with respiration. During inspiration there is an increase in the RV pressure and a decrease in LV pressure. The opposite occurs during expiration (b) With restriction LV and RV systolic pressures are concordant during respiration. During inspiration there is a decrease in the RV pressure and a decrease in LV pressure. The opposite occurs during expiration (Reprinted from Talreja et al. [27], © 2008, with permission from Elsevier)

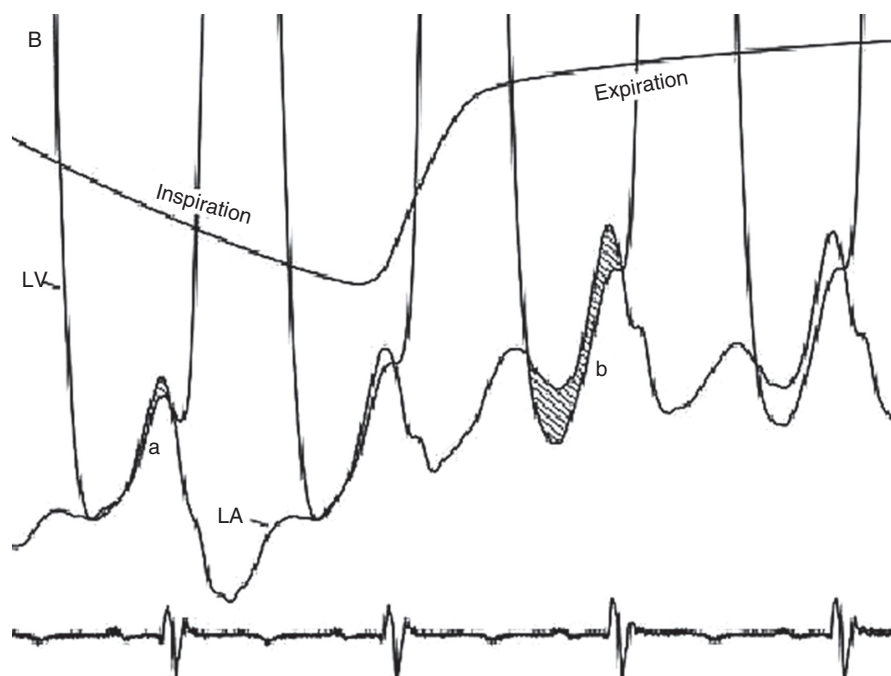


Fig. 2.15 Simultaneous pressure recording of LV and PCWP. There is significant respiratory change suggesting dissociation of intra-thoracic and intra-cardiac pressures (Reprinted from Doshi et al. [28], © 2015, with permission from Elsevier)

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