Pericardial Diseases

131

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Abstract

The pericardium is a thin fibrous membrane that separates the heart from the remaining mediastinal structures. Disease of the pericardium presenting as acute pericarditis, pericardial effusion, and cardiac tamponade can be life-threatening in some cases. The subsequent development of chronic or recurrent pericarditis is a feared complication, although less frequent in the pediatric population. Post-pericardiotomy syndrome is a medical syndrome referring to an inflammatory phenomenon that occurs a few weeks after surgical incision of the pericardium. Structural abnormalities including congenitally absent pericardium and pericardial cysts are usually asymptomatic.

The etiology of pericardial disease is often difficult to determine and remains idiopathic in a large number of cases. The accurate diagnosis of pericardial diseases is a well-recognized clinical challenge that often requires the integration of imaging and invasive hemodynamic measurements. Recent advances in multimodality noninvasive cardiac imaging with cardiac computed tomography (CT) and magnetic resonance imaging (MRI) have aid in determining the etiology of pericardial pathology.

This chapter described those different pathologies of the pericardium, the clinical manifestations, diagnosis, and treatment.

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Introduction

Pericardial diseases are defined as abnormalities of the visceral or parietal pericardium that may or may not have an impact on cardiac function. Pericardial diseases are divided into diseases affecting the pericardial layers, including acute, chronic, or recurring pericarditis and constrictive pericarditis, and those affecting the pericardial sac, including pericardial effusion and tamponade. In children, diseases of the pericardium consist mostly of infectious and inflammatory processes. Rarely, neoplastic lesions as well as congenital structural defects may be the cause. Pericardial diseases may present with chest pain, pericardial friction rub, fever, and acute hemodynamic compromise or can be asymptomatic, depending on their etiology.

Anatomy and Physiology

The pericardium consists of a sac surrounding the heart. It is made of a thin inner visceral layer made up of mesothelial cells, called the *serous (visceral) pericardium or epicardium*, and a thick outer layer made up of collagen and elastic fibers, called the *fibrous (parietal) pericardium*. Those two layers are separated by a virtual space containing a small amount of physiological fluid which serves as lubricant [1]. The pericardial space normally contains <30 ml of fluid in the adult and considerably less in infants and children. The pericardial fluid is produced by the visceral pericardium and is an ultrafiltrate of plasma. The pericardial fluid normally drains through the right lymphatic duct via the right pleural space and through the thoracic duct via the parietal pericardium [2].

The pericardium envelops the heart and great vessels and reflects around the great vessels forming the pericardial recesses and sinuses. The pericardium is anchored to the diaphragm by the pericardiophrenic ligament and to the sternum by the sternopericardial ligament, providing support for the heart within the thoracic cage. It is thought that the presence of the parietal pericardium helps maintain a functionally optimal cardiac shape.

Although an intact pericardium is not critical to the cardiovascular function, it assumes some minor functions including:

- Limitation of intrathoracic cardiac motion and acute cardiac dilatation
- Preservation of diastolic and systolic interactions between the right and left ventricles
- Lubricant effect that minimizes friction between cardiac chambers and surrounding structures
- Lymphatic and immunological functions, helping prevent spread of infection from contiguous structures, especially the lungs

The normal pericardium limits cardiac distension, thereby coupling the ventricles and enhancing their interactions [3, 4]. Pressure or volume overload of one ventricle influences the compliance and filling of the contralateral ventricle via interventricular septal diastolic interactions called interventricular coupling. By influencing the effects of diastolic pressure and dimensions between the ventricles, the pericardium helps to balance the right and left ventricular output.

The normal intrapericardial pressure is subatmospheric and several millimeters of mercury (mmHg) less than the right atrial (RA) pressure. As the right ventricle (RV) fills, this passively increases pericardial pressure, which restricts further ventricular filling due to limited extensibility of the pericardium near end diastole. The intrapericardial pressure varies with the pleural pressure: the inspiratory decrease in pleural pressure slightly reduces the pericardial, right atrial (RA), right ventricular (RV), pulmonary capillary wedge (PCW), and systemic arterial pressures. Under physiologic conditions, respiratory variations influence cardiac filling and hemodynamics, but their effects on the right and left heart are different, secondary to the differences in the anatomic relationship of

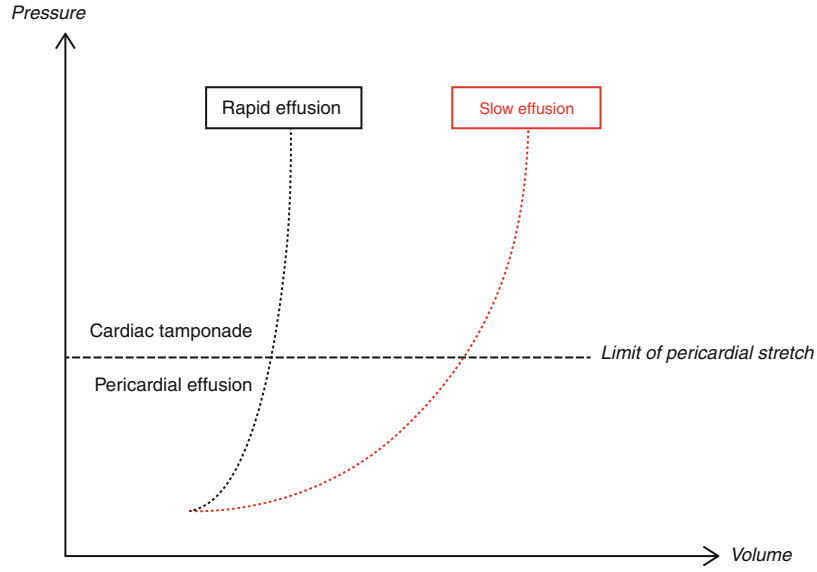


Fig. 131.1 Pericardial pressure-volume curve showing effect on pressure of rapid and slow increase of pericardial volume over time, as seen in acute and chronic pericardial effusion

the venous return to the intrapleural space [5]. The systemic venous system is extrapleural as opposed to the pulmonary venous return which is intrapleural. As a consequence, a decrease in intrathoracic pressure during inspiration has a different effect on the systemic and pulmonary venous return. During inspiration, the systemic venous return is increased by about 50 %, which increases right heart filling and output. Since the pulmonary venous return is intrathoracic, the pleural pressure changes are evenly distributed to the left heart chambers and pulmonary veins with minimal change in left heart filling and output throughout the respiratory cycle [6].

Abnormal pericardial fluid production is usually secondary to injury (postoperative pericardial effusion) or inflammation (acute pericarditis, postpericardiotomy syndrome). Transudative fluid results from obstruction of fluid drainage, while exudative fluid is secondary to inflammatory, infectious, malignant, or autoimmune processes. The normal pericardium has a small capacitance volume (about 150 ml) limited by the relative noncompliance of the parietal pericardial layer. When reserve capacitance has been reached, further increases in intrapericardial volume result in a steep increment of intrapericardial pressure. The hemodynamic repercussion of pericardial fluid accumulation is highly dependent upon the rate of accumulation in the pericardial sac. Rapid accumulation of pericardial fluid causes a sudden increase in intrapericardial pressure and hemodynamic compromise. Slow accumulation of pericardial fluid can be asymptomatic even when large fluid volumes are present (Fig. 131.1) [7].

Due to the pericardial sac's small reserved volume, an increase in pericardial volume that occurs in pathological states can exert a significant hemodynamic hindrance on ventricular compliance. Cardiac tamponade is a consequence of markedly diminished diastolic filling that occurs when the atrial and ventricular distending pressures are insufficient to overcome the increased intrapericardial pressure. During tamponade, inspiration increases inflow to the right ventricle, causing an abrupt expansion of the right ventricle during diastole at the expense of the left ventricle (Fig. 131.2) [8]. Conversely during expiration, left ventricular expansion causes right ventricular and atrial diastolic collapse. This reciprocating behavior of the ventricles during respiration is responsible for a *paradoxical pulse*, defined as an exaggeration (>10 mmHg) of the normal inspiratory decrease in systolic blood pressure [9].

Inflammation of the pericardium can manifest as a fibrinous reaction with an exudative effusion leading to thickening, fibrosis, and obliteration of the space between the two pericardial layers.

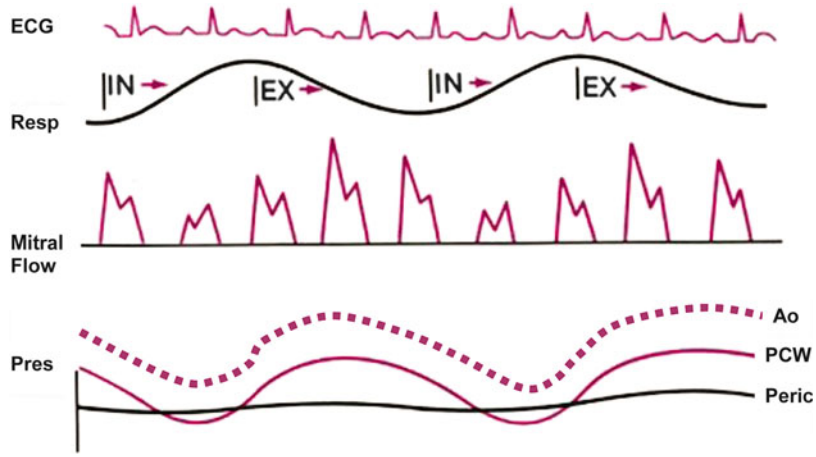


Fig. 131.2 Mechanism of pulsus paradoxus with exaggeration of inspiratory decrease in blood pressure, as seen in pericardial effusion and tamponade. *Abbreviations: ECG* electrocardiogram, *Resp* respiration, *IN* inspiration, *EX* expiration, *Pres* pressure, *Ao* aortic pressure, *PCW* pulmonary capillary wedge pressure, *Peric* intrapericardial pressure

As a result, adhesions can occur between the pericardium and myocardium leading to decreased pericardial compliance and constrictive pericarditis. This results in diminished ventricular distensibility with inability to maintain adequate preload and biventricular diastolic dysfunction. As opposed to a pericardial effusion, early ventricular filling is not altered in constrictive pericarditis. However, as the ventricles fill, they meet the resistance of the stiff pericardium, and filling pressure rises rapidly to an elevated plateau. This late diastolic phenomenon is due to a change in the volume-elasticity curve, a small increase in volume resulting in a considerable increase in end-diastolic pressure. In this situation, atrial filling pressures are elevated, reflecting both ventricular noncompliance and atrial constraint from the thick pericardium. Because of the isolated encasement of the pericardium and not the systemic veins or lungs, there is dissociation between intrathoracic and intracardiac pressures with marked respiratory variation in right and left heart filling pattern [10, 11].

Through analysis of the atrial waveforms, it is possible to understand the dynamic effect of intrapericardial and intrathoracic pressures and respiratory variations during the cardiac cycle [12]. The atrial pressure waveforms are constituted by two major positive deflections, the A and V waves, and two negative waves, the x and y descents (Fig. 131.3):

- The *A wave* (atrial contraction) follows the P wave of the electrocardiogram and is generated by *A*trial contraction.
- The *C wave* (atrioventricular valve closure) corresponds to ventricular *C*ontraction causing the atrioventricular valve to bulge toward the atrium.
- The *x descent* (atrial diastole) corresponds to atrial relaxation and rapid atrial filling and is influenced by pericardial compliance.
- The *V wave* (ventricular contraction) reflects venous return when the atrioventricular valve is closed resulting in atrial filling and increased atrial pressure during ventricular systole and is influenced by atrioventricular valve competence.
- The diastolic *y descent* (atrial emptying) represents opening of the atrioventricular valve and rapid filling of the ventricle and is influenced by the resistance to atrial emptying (atrioventricular valve opening, ventricular and pericardial compliance).

Acute Pericarditis

Etiology and Epidemiology

Acute pericarditis is the most common pericardial disease. Most cases are idiopathic (75–80 %) and presumed to be due to viral infection, particularly Enterovirus and Coxsackievirus B (Table 131.1) [13], although identifying the cause may be challenging. It may occur

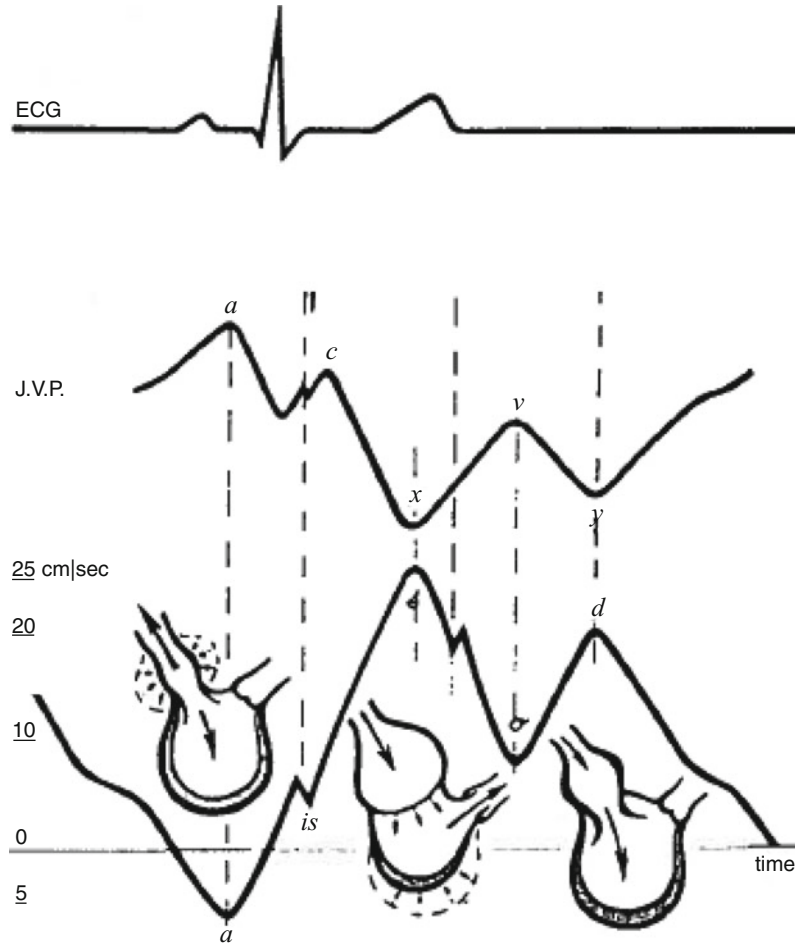


Fig. 131.3 Mechanics of jugular venous pressure and atrial wave form: - The "A" wave corresponds to atrial contraction. - The "C" wave corresponds to ventricular contraction and atrioventricular valve closure. - The "x" descent corresponds to atrial relaxation and rapid atrial filling. - The "V" wave corresponds to passive atrial filling during ventricular contraction. - The "y" descent corresponds to the rapid emptying of the atrium into the ventricle following the opening of the atrioventricular valve. *Abbreviations: ECG* electrocardiogram, *JVP* jugular venous pressure (Adapted from Kalmanson [12])

secondary to collagen vascular or rheumatic diseases, drug therapy, and cardiac surgery, as a manifestation of rheumatic fever or in association with chronic renal failure and dialysis. Effusive-constrictive pericarditis is usually secondary to infections creating a thick exudate, such as pyogenic bacteria or tuberculosis. Purulent pericarditis is a medical and surgical emergency and requires prompt antibiotic treatment and pericardial drainage to prevent adhesions and constriction. It is a life-threatening illness with mortality between 25 % and 75 % and may be primary or secondary to dissemination from another site of infection (pneumonia, septic arthritis, meningitis, osteomyelitis). Tuberculous pericarditis occurs as a result of direct spread of mycobacterium tuberculosis from mediastinal lymph nodes or secondary to hematogenous dissemination and may occur in the absence of pulmonary infiltrates. It is more common in underdeveloped countries and is fatal in 90 % of cases without antibiotic therapy. The incidence is increasing particularly in Africa, as a result of the human immunodeficiency virus (HIV) epidemic [14].

Chronic pericarditis refers to a pericarditis lasting more than 3 months and is generally secondary to inflammatory diseases or congestive heart disease. *Recurrent pericarditis* refers to either an intermittent or an incessant form of pericarditis in which recurrence occurs as soon as the therapy is discontinued and may be seen in rheumatic disease of childhood (lupus erythematosus, juvenile rheumatoid arthritis) and in post-pericardiotomy syndrome.

Table 131.1 Infectious causes of pericarditis

Causes of pericarditis	
Idiopathic	
Viral	Coxsackievirus A and B
	Adenovirus
	Cytomegalovirus
	Epstein-Barr virus
	Varicella zoster
	Mumps virus
	Influenza virus
	Hepatitis virus
	Human immunodeficiency virus
	Variola and vaccinia viruses
Pyogenic	Streptococcus pneumoniae
	Streptococcus pyogenes
	Staphylococci aureus
	Haemophilus influenzae
	Neisseria meningitidis
	Neisseria gonorrhoeae
	Pseudomonas aeruginosa
	Francisella tularensis
	Bartonella henselae
	Cardiobacterium hominis
	Salmonella spp
	Actinomyces spp
	Nocardia spp
	Coxiella burnetii
	Legionella spp
Tuberculous	Mycobacterium tuberculosis
Fungal	Histoplasmosis
	Coccidioidomycosis
	Candidosis
	Aspergillosis
	Blastomycosis
	Echinococcosis
	Amebiasis
Parasites	Entamoeba hystolitica
	Echinococcus spp
Miscellaneous	Mycoplasma
	Chlamydia
	Rickettsiae
	Spirochetes

Signs and Symptoms

The predominant symptom is chest pain, usually retrosternal or precordial, and is present in as many as 80 % of children. The pain usually is described as sharp or stabbing, is made worse with inspiration, coughing, or movement and patients are more comfortable in the upright position. Pain may be of sudden or gradual onset and may radiate to the back, neck, or left shoulder. Associated signs and symptoms include low-grade intermittent fever, dyspnea, tachypnea, cough, and dysphagia. Acute abdominal pain is not uncommon in children. The most common and important physical finding is a pericardial friction rub, best heard at the lower left sternal border or apex when the patient is sitting forward, and may be transient. Tachycardia, out of proportion to the degree of fever, may indicate pericarditis and/or myocarditis. With bacterial pericarditis, the patient is febrile and appears toxic. In the setting of viral or autoimmune pericarditis, fever and evidence of toxicity are generally milder.

Diagnostic Workup

- Laboratory

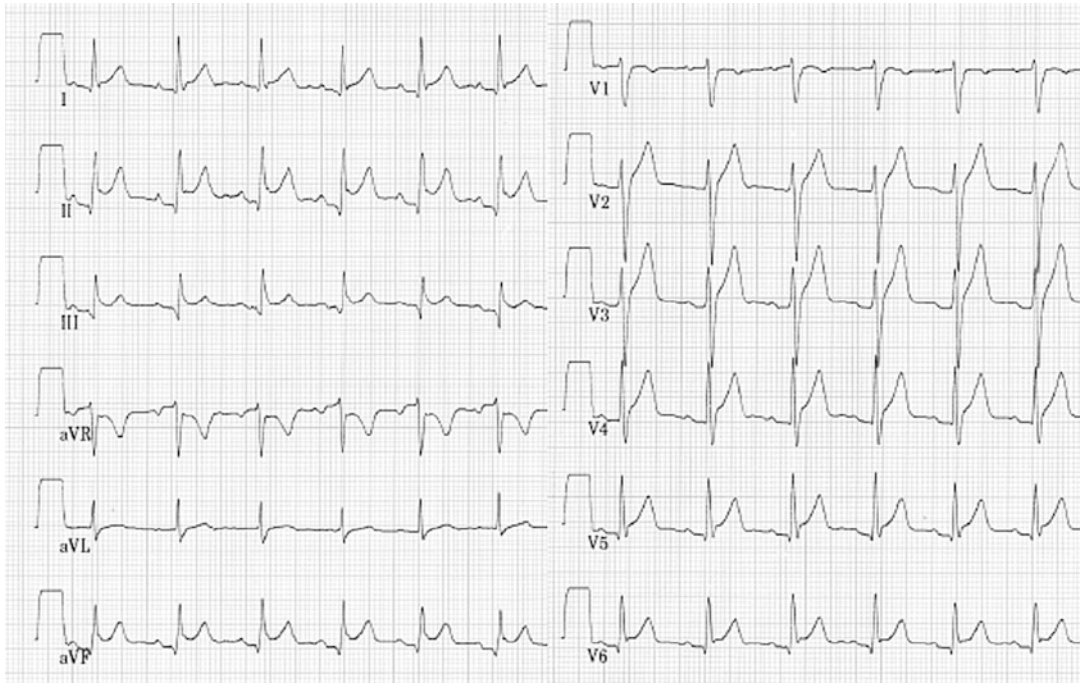
Inflammatory markers are usually elevated (C-reactive protein, ESR). Cardiac troponin I rise has been described as detectable in acute pericarditis in about 30 % of patients, reflecting injury of the underlying myocardium, and is often associated with ST-segment elevation and the presence of a pericardial effusion [15].

- Chest X-Ray

Without pericardial effusion, the chest X-ray is unremarkable except in the case of calcifications, and absence of cardiomegaly does not preclude the diagnosis of pericarditis.

- Electrocardiogram

The ECG can be diagnostic in acute pericarditis. It evolves in four stages. The first stage is characterized by ST-segment elevation and concave upward ST segments, noted in all leads except V_1 (Picture 131.1). In the second stage, normal ST segments with T- wave flattening are noted. Third stage is characterized by T wave inversion without Q wave formation with normalization of the ECG in the fourth stage (Table 131.2). Another important ECG finding is PR-segment depression.



Picture 131.1 Acute pericarditis stage 1: electrocardiogram with diffuse ST-segment elevation and PR depression except in aVR, where there is ST-segment depression and PR elevation

Table 131.2 Electrocardiographic changes in acute pericarditis

Time	ST segment	T wave	PR segment
Stage 1: hours	Diffuse elevation	Upright	Leads aVR, V1: elevation
			All others lead: depression
Stage 2	Resolution	Flattening	Resolution
Stage 3: days	Resolution	Inversion	Resolution
Stage 4: weeks	Resolution	Normalization	Resolution

- Echocardiography

The echocardiogram is often normal, unless acute pericarditis is associated with a pericardial effusion. While the finding of a pericardial effusion supports the diagnosis of acute pericarditis, its absence does not exclude it. In pericarditis, the pericardium may have a normal appearance.

- Cardiac computed tomography (CT) scan and magnetic resonance imaging (MRI)

The normal thickness of the pericardium as measured by CT scanning is less than 2 and 4 mm by MRI. The limitation of CT scan is the difficulty in differentiating fluid from thickened pericardium.

Medical Management

If an infectious cause of pericarditis is identified, appropriate antimicrobial therapy should be started. Prior to identification of the etiologic agent, broad-spectrum antibiotic coverage should be directed toward the treatment of Staphylococcus aureus and Haemophilus influenzae, and penicillinase-resistant penicillin in combination with a third-generation cephalosporin is a good association. Nevertheless, antibiotic therapy alone is not sufficient to adequately treat bacterial pericarditis as antibiotic penetration is not sufficient and will require pericardial fluid drainage.

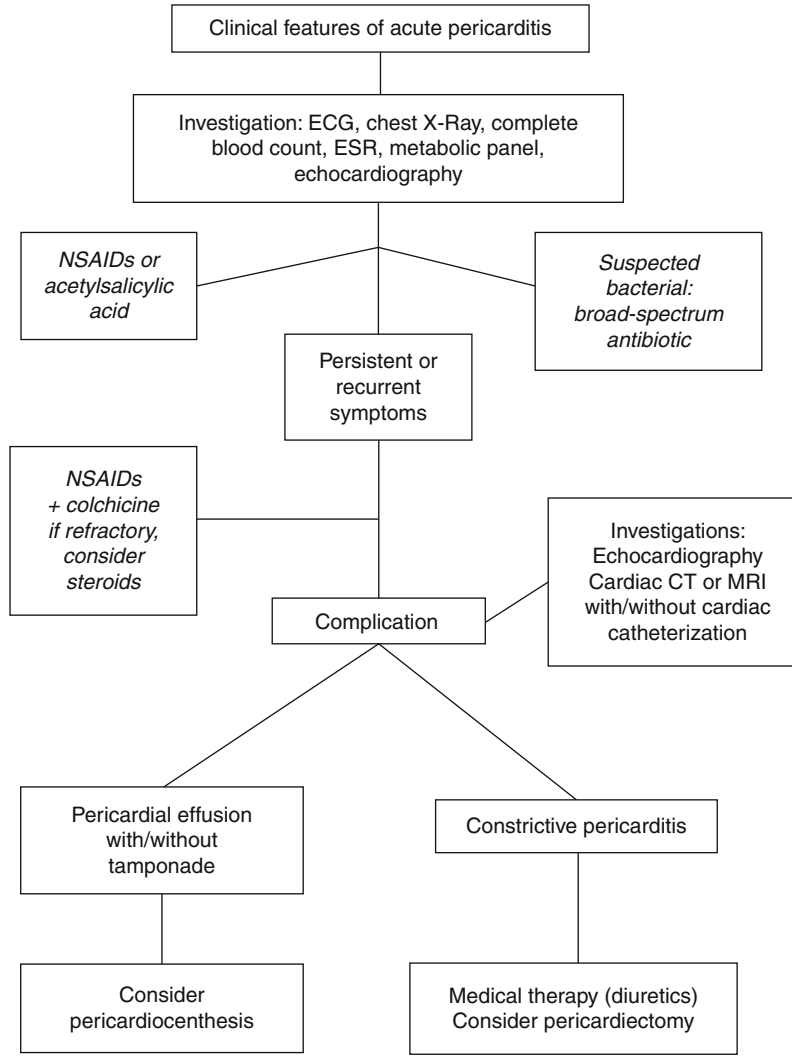


Fig. 131.4 Proposed algorithm for management of acute pericarditis. *Abbreviations: ECG* electrocardiogram, *ESR* sedimentation rate, *NSAIDs* nonsteroidal anti-inflammatory drugs, *CT* computed tomography, *MRI* magnetic resonance imaging

Treatment of viral pericarditis is predominantly symptomatic. Nonsteroidal anti-inflammatory drugs or acetylsalicylic acid is the mainstay of initial treatment and are usually used to decrease the inflammatory reaction. Acetylsalicylic acid (80–100 mg/kg/day) during 7–10 days, tapered down over 3–4 weeks, is usually the first-line treatment. Steroids therapy is rarely indicated and should be considered only when nonsteroidal agents have failed (Fig. 131.4). In adults, colchicine has been used as an initial therapy along with nonsteroidal drugs or alone and has been found to be effective in the immediate management and in the prevention of recurrences [16]. In tuberculous pericarditis, because of the high prevalence of drug-resistant organisms, initial therapy should consist of isoniazid, pyrazinamide, rifampin, and streptomycin for at least 9 months. The development of constrictive pericarditis in as many as 35 % of patients results from diffuse inflammation with subsequent pericardial thickening. It is uncertain whether adjunctive corticosteroids for 1–2 months are effective in reducing mortality or pericardial constriction in tuberculous pericarditis.

Surgical Management

As many as 50 % of patients will require pericardial fluid drainage, accomplished by pericardiocentesis or by placement of a pericardial catheter. In some patients with bacterial pericarditis, the pericardial fluid is so viscous that open drainage via a pericardial window or pericardiectomy is required. Intrapericardial administration of streptokinase has been reported to facilitate successful drainage of pericardial fluid [17].

Pericardial Effusion and Tamponade

Etiology and Epidemiology

Pericardial effusions can be associated with pericarditis or be secondary to cardiac surgery (Table 131.1) [18]. Common causes of pericardial effusions in children include prior cardiac surgery, bacterial pericarditis, malignancy, and connective tissue disorders. In a significant number of children, however, despite extensive investigation, it is not possible to identify a clear etiology. A viral cause is often considered, though rarely confirmed [19]. In developing countries, tuberculosis is responsible for approximately 70 % of cases of large pericardial effusions and most cases of constrictive pericarditis. However, in industrialized countries, tuberculosis accounts for only 4 % of cases of pericardial effusion and an even smaller proportion of constrictive pericarditis [20].

Postoperative pericardial effusions can occur in isolation or be secondary to postpericardiotomy syndrome. *Post-pericardiotomy syndrome* occurs following cardiac surgery in which the pericardium has been opened and is similar to that occurring after myocardial infarction (Dressler syndrome). The incidence approaches 30 %, although children younger than 2 years old appear to be less commonly affected. An immunologic mechanism has been postulated, with the demonstration of specific antisarcolemmal and antifibrillatory antibodies supporting this theory [21]. A viral pathogenesis has also been postulated due to seasonal variation in the incidence with a significant rise in antibody titer to several viral agents [22]. In the postoperative period, even small amount of loculated pericardial fluid, particularly when localized along the free wall of the right atrium or ventricle, can have significant hemodynamic repercussion [23]. In post-cardiac transplant patients, delayed-onset pericardial effusion (after 30 days post-transplantation) may represent acute or chronic rejection, and any effusion should be evaluated with careful attention to the immune status of the transplant recipient and promptly treated for rejection.

In acute rheumatic fever, cardiovascular involvement is characterized by pancarditis with inflammation of the endocardium, myocardium, and pericardium. Pericardial involvement is seen in 5–10 % of patients with acute rheumatic fever, almost always in association with valvulitis or myocardial dysfunction. During the acute phase of Kawasaki disease, one third of patients are found to have pericardial effusion. In children with connective tissue disorders, pericarditis occurs in approximately 10 % of patients with juvenile rheumatoid arthritis and in up to 25–50 % of those with systemic lupus erythematosus.

Hemopericardium should be suspected in any patient who complains of severe chest pain following traumatic injury. *Chylopericardium* is a pericardial effusion consisting of chyle and may be primary (idiopathic) or secondary to injury of the thoracic duct and associated with chylothorax (post-surgery).

Signs and Symptoms

Chest pain or discomfort relieved by sitting up and leaning forward and intensified by lying supine is typical. Respiratory symptoms of cough and dyspnea can dominate the clinical picture. The physical exam reveals a pericardial friction rub, heard most frequently during expiration with the patient upright and leaning forward [24]. The friction rub may not be heard in patients with large effusions. Tachypnea, tachycardia, widened pulse pressure, and

hepatojugular reflux are signs of impending hemodynamic compromise. The classic Beck triad of pericardial tamponade includes hypotension, muffled heart sounds, and jugular venous distension [25]. Pulsus paradoxus, defined as an exaggerated decrease in systolic blood pressure (>10 mmHg) with inspiration, is a sign of falling cardiac output. Late findings are cyanosis and decreased level of consciousness.

Post-pericardiotomy syndrome typically occurs after cardiac surgery and is usually mild. The patient can suffer from fatigue and low-grade fever. Anterior precordial chest pain that increases on deep inspiration is common. The typical clinical finding is that of a pericardial friction rub. When a pericardial effusion is associated, the friction rub can disappear, the heart sounds are attenuated and tamponade is a possibility.

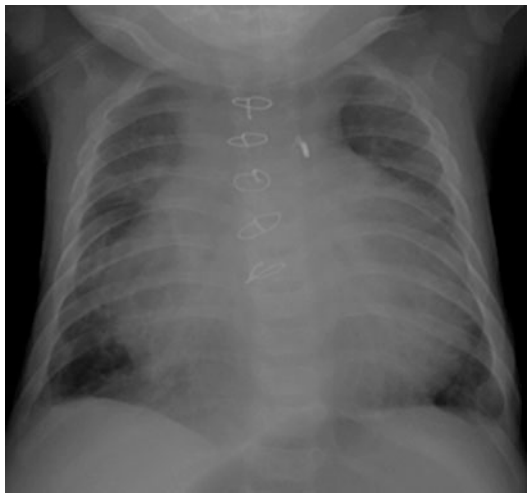
Diagnostic Workup

- Laboratory

Blood work should be directed toward identifying the etiology (Tables 131.2 and 131.3). Diagnostic studies can be performed on the pericardial fluid including cell count and differential, protein, lactate dehydrogenase, glucose, gram stain, bacterial and fungal cultures, viral PCR, mycobacterial acid-fast stain, and tumor cytology. When connective tissue disease is suspected, rheumatoid factor, antinuclear antibodies, and complement levels can be added. Specific bacterial antigens may be identified with immunologic techniques. In tuberculous pericarditis, the pericardial fluid will be serosanguineous with a predominance of lymphocytes, and pericardial fluid adenosine deaminase activity, a T-lymphocyte product, has been shown to be useful in the diagnosis of tuberculous pericarditis [26]. Acid-fast bacilli are present on auramine-rhodamine fluorescent stained smears in 15–40 % of patients, and incubation of Mycobacterium tuberculosis may require 6 weeks using standard cultures. In chylopericardium, pericardial fluid triglyceride levels are elevated. In patients with post-pericardiotomy syndrome, elevated white blood cell count with a left shift and elevated erythrocyte sedimentation rate (ESR) are usual. Analysis of pericardial fluid should include cell count.

Table 131.3 Causes of pericardial effusion

Causes of pericardial effusion	
Idiopathic	
Viral	Coxsackievirus A and B
	Hepatitis
	HIV
Pyogenic	Pneumococci
	Streptococci
	Staphylococci
	Neisseria species
	Legionella species
	Haemophilus influenzae
Tuberculous	Mycobacterium tuberculosis
Fungal	Histoplasmosis
	Coccidioidomycosis
	Candidosis
Other infections	Syphilitic
	Protozoal
	Parasitic
Acute rheumatic fever	
Uremia	
Hypothyroidism	
Neoplasia	Metastases
	Leukemia
	Lymphoma
Post-cardiac surgery	Post-pericardiotomy syndrome
Acute myocardial infarction	Dressler syndrome
Collagen vascular diseases	Rheumatoid juvenile arthritis
	Systemic lupus erythematosus
Kawasaki disease	
Hemopericardium	Trauma
Chylopericardium	Primary
	Secondary: post-cardiac surgery
Drug-induced	Hydralazine
	Isoniazid
	Procainamide
Postradiation therapy	
Post-cardiac transplant	
Others	Hypercholesterolemia
	Sarcoidosis
	Whipple disease



Picture 131.2 Chest X-ray in pericardial effusion: water bottle-shaped heart

- Chest X-Ray

An increased cardiac silhouette that is globular (water bottle-shaped heart) can be seen with excessive pericardial fluid accumulation (Picture 131.2) [27]. Another finding is visualization of the pericardial fat stripe within the cardiac silhouette. The lung fields are usually oligemic and a pleural effusion is often associated.

- Electrocardiogram

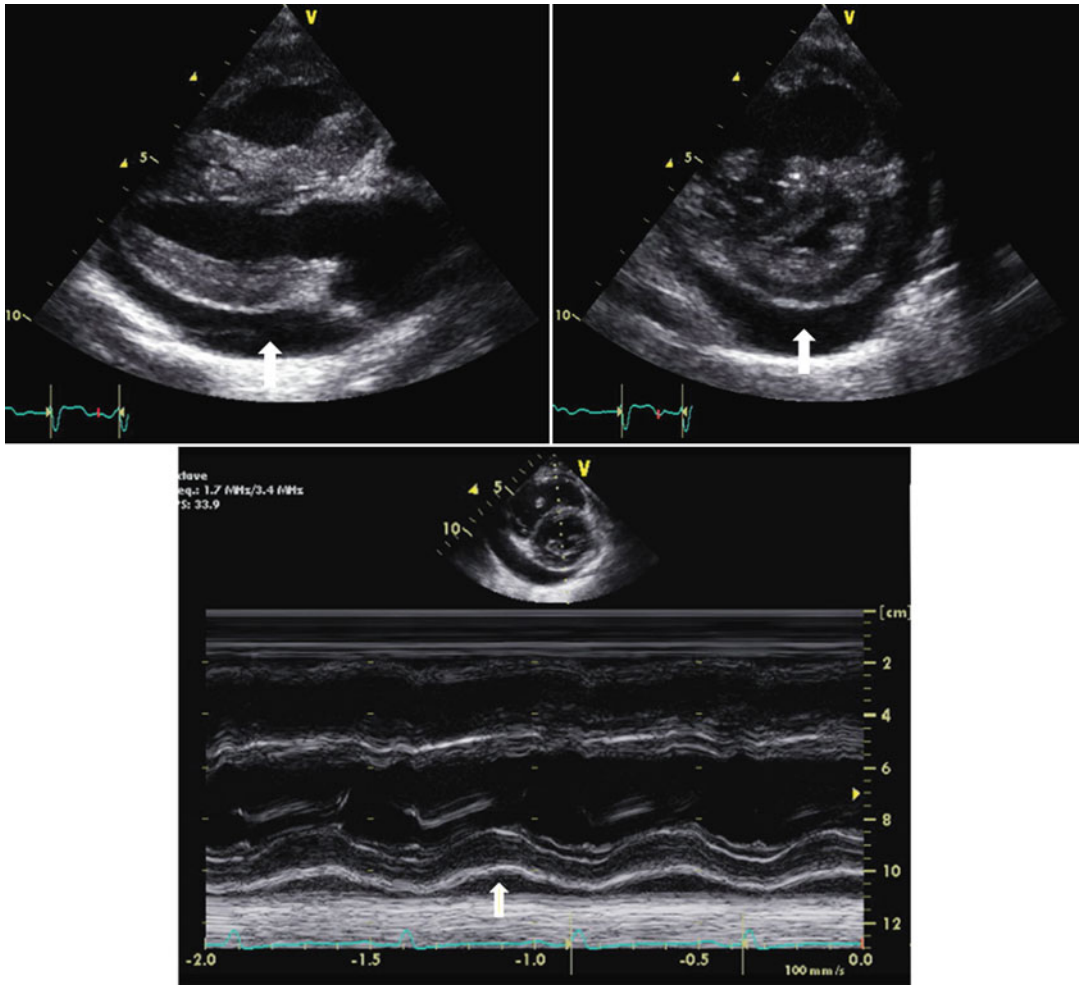
Low voltage QRS with diffuse nonspecific ST-segment changes are present with large effusions. Electrical alternans is pathognomonic of cardiac tamponade and is characterized by alternating P wave, QRS complex, and T wave voltages attributable to swinging motion of the heart [28, 29]. In patient with post-pericardiotomy syndrome, nonspecific abnormalities of the T waves (flattening in leads I and lateral chest) and decrease in voltage are common findings, especially with large pericardial effusions.

- Echocardiography

Pericardial effusion appears as an "echo-free" space between the visceral and parietal pericardium on M-mode echocardiography (Picture 131.3) [30]. Effusions tend to accumulate posterior and inferior to the left ventricle initially. However, on echocardiographic imaging, fluid visualized above the right atrium in the four chamber view is the most sensitive indication as this is the first place where a pericardial effusion is seen. Moderate effusions (10–20 mm in size) extend toward the apex of the heart, and large effusions (more than 20 mm) circumscribe the heart (Picture 131.3, Videos 131.1 and 131.2).

The rapidity of fluid accumulation and the compliance of the pericardium influence the hemodynamic significance for a given fluid volume. As pericardial fluid accumulates, intrapericardial pressure increases until it exceeds normal filling pressure of the heart, leading to tamponade. The first sign of hemodynamic compromise is *expiratory right ventricular free wall collapse early in diastole*, reflecting the brief period when intrapericardial pressure is greater than the right ventricular transmural distending pressure [31, 32]. The right ventricle is the first to collapse due to its lower intracardiac pressure compared to the left ventricle. Although right ventricular collapse is generally a specific indicator of tamponade, the sensitivity can be reduced in conditions with increased right ventricular pressure [33]. *Expiratory right atrial collapse* occurs in *late diastole* (Picture 131.4). The sensitivity of expiratory diastolic right atrial collapse for diagnosing tamponade is low (55 %), but the specificity is high (90 %). Extension of collapse greater than 1/3 of the cardiac cycle increases the sensitivity of this finding to more than 90 % [34, 35]. Absence of expiratory right atrial collapse virtually excludes tamponade. Another sensitive marker of tamponade by echocardiogram is absence of inspiratory collapse (plethora) of the inferior vena cava, defined by less than 5 mm decrease in diameter during inspiration [36]. The sensitivity of inferior vena cava plethora is high (97 %), but the specificity is low (66 %). Diastolic collapse of the left atrium and rarely the left ventricle occurs during inspiration, related to the increased right heart inflow and abrupt expansion of the right ventricle.

Doppler echocardiography shows large swinging amplitudes of the mitral and tricuspid inflow, the aortic and pulmonary outflow, and the hepatic veins. Normally, there is no more than 10 % variation in the amplitude of inflow and outflow signals with respiration, but this exceeds 30 % in tamponade (Picture 131.5). The classic

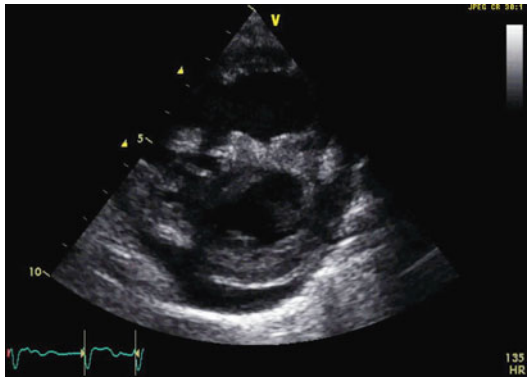


Picture 131.3 Parasternal long and short axis 2D-echocardiography: large pericardial effusion (*arrow*) circumscribing the heart. M-mode echocardiography: echo-free space (*arrow*) between the visceral and parietal pericardium, posterior to the left ventricle

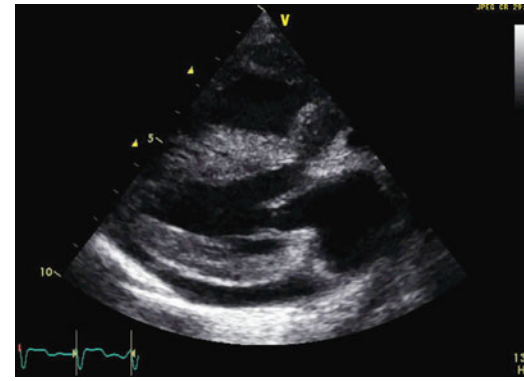
Doppler patterns of cardiac tamponade (Fig. 131.4) [37, 38] are as follows:

- Mitral inflow: During inspiration, E wave velocity decreases by more than 30 % compared with expiration.
- Pulmonary veins: During inspiration, D wave velocity decreases.
- Tricuspid inflow: During inspiration, E wave velocity increases more than 50 % compared with expiration.
- Hepatic veins: During inspiration, S is greater than D and during expiration, there is a very limited or absent D wave with prominent reversal.
- Cardiac CT and MRI

CT can potentially determine composition of fluid and may detect as little as 50 mL of fluid. MRI can detect as little as 30 ml of pericardial fluid and may be able to distinguish hemorrhagic and nonhemorrhagic effusions. Both MRI and CT scan may be superior to echocardiography in detecting loculated pericardial effusions and pericardial thickening.



Video 131.1 Implantation of Artificial Chordae. The mitral valve is exposed through a left atriotomy, and anterior and posterior mitral leaflets are tested for mobility and coaptation using 2 nerve hooks. The prolapse and increased mobility of the anterior mitral leaflet when compared to the posterior leaflet is noted here. Since the size of the mitral annulus corresponds to the surface area of the anterior mitral leaflet, this equates to the inter-trigonal distance. The anterior mitral leaflet is unfurled, and the corresponding sizer, that matches the surface area of the anterior leaflet and inter-trigonal distance, is chosen as the correct size for the annuloplasty ring. Pledgeted 4-0 poly-tetra-fluoro-ethylene (PTFE, Gortex, USA) sutures are inserted through the body of the anterior and posterior papillary muscles below, and through the free-margin of the anterior mitral leaflet at the appropriate distance. Any clefts/indentations in the posterior mitral leaflet are closed using interrupted 5-0 polypropylene sutures. One of the needles of the biodegradable annuloplasty ring is inserted into the posterior mitral annulus along an intra-annular plane, starting from the level of the posterior commissure. The ring is gently pulled through and advanced along the entire length of the posterior annulus, until it exits at the level of the anterior mitral commissure. The length of the artificial chordae are measured such that the coaptation height of both the leaflets are equal, and the anterior leaflet prolapse is corrected. Saline injection testing of the valve, and intra-operative trans-esophageal echocardiography confirm adequacy of mitral valve repair, and obliteration of mitral regurgitation.

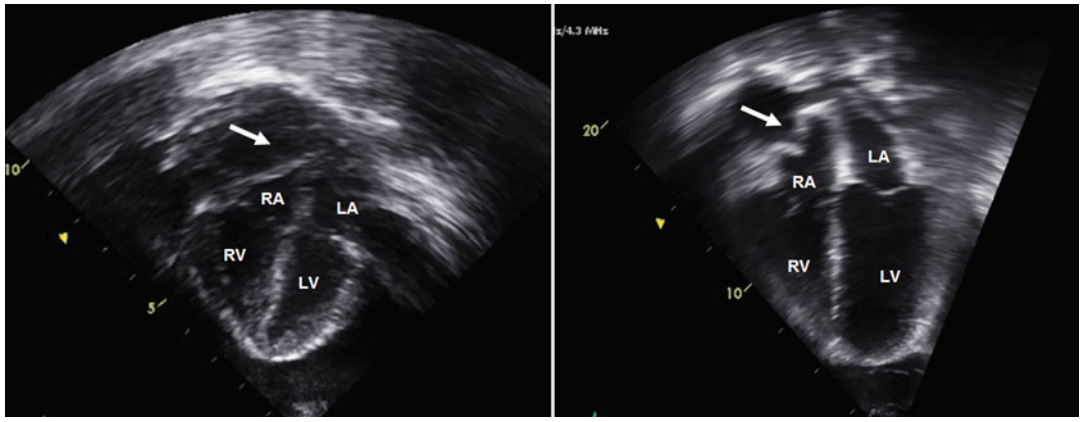


Video 131.2 Fundoplasty. Inspection of the mitral valve using 2 nerve hooks demonstrates a typical stenotic "fish mouth" rheumatic valve, characterized by thickened, fibrotic leaflets with markedly limitation in mobility, and commissural fusion. Anterior and posterior commissurotomies are performed using a No. 11 blade, leaving a 2 mm margin between the incision and the annulus. Fused sub-valvular tissue is also divided and released. Re-inspection of the valves demonstrates retraction of the P2-P3 scallops of the posterior mitral leaflet. A 3-4 cm incision is made on the posterior mitral annulus, extending from P3 to P2, leaving a 2 cm rim away from the free margin of the posterior leaflet. Full thickness polypropylene sutures are taken through the 3 O'clock and 9 O'clock positions of the incision. Similarly, multiple interrupted sutures are taken through the 10 O' clock and 2 O' clock, 8 O'clock and 4 O'clock positions, etc. When these sutures are tied down, the orientation of this incision changes from a horizontal incision into a vertical incision, thereby improving the coaptation height of the P2 and P3 segments. The mitral annulus is then supported by conventional ring annuloplasty. Saline injection testing and intra-operative trans-esophageal echocardiography confirm adequacy of repair, with good leaflet coaptation and no mitral regurgitation.

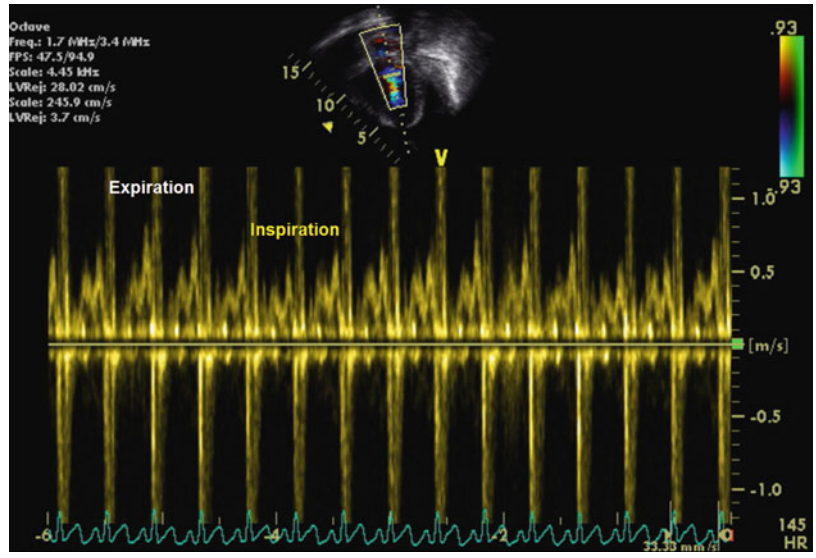
Medical Management

In the case of a small pericardial effusion in the postoperative period, an increase in the diuretic regimen can be attempted. Nonsteroidal anti-inflammatory drugs or acetylsalicylic acid is usually used to decrease the inflammatory reaction. Aspirin (80–100 mg/kg/day) during 7–10 days, tapered down over 3–4 weeks, is usually the first-line treatment. Steroids are reserved for severe and recurrent cases, as cases of corticoid-dependent pericardial effusion has been described. *Post-pericardiotomy syndrome* is usually self-limited, but relapses can occur. Medical treatment includes bed rest and anti-inflammatory drugs:

- Acetylsalicylic acid (aspirin 80–100 mg/kg/day or 800 mg every 6 h in adults) is thought to reduce inflammation and fever and is administered for 10 days and then gradually tapered down over 3 weeks. High-dose acetylsalicylic acid should be associated with gastroduodenal prophylaxis.
- Nonsteroidal anti-inflammatory drugs (NSAIDs), like ibuprofen (10 mg/kg/day), is an alternative to acetylsalicylic acid therapy.



Picture 131.4 Apical four-chamber and subcostal 2D-echocardiography views in pericardial effusion right atrial collapse (*arrow*)



Picture 131.5 Doppler echocardiography in pericardial effusion: respiratory variation in mitral inflow with increased E wave velocity during expiration

- Colchicine has been shown to be safe and effective in the treatment and prevention of recurrent pericarditis after failure of conventional treatment, especially in idiopathic cases [16]. The dose for adults is 1–2 mg for the first day followed by a maintenance dose of 0.5–1 mg daily for 3 months. No data are available in children.
- Corticosteroids (prednisone 1–2 mg/kg/day) are preferably avoided but can be used in severe or recurrent cases during 2–4 weeks followed by gradual tapering off.
- High doses intravenous immunoglobulins (IVIG) have been described in the treatment of recurrent post-pericardiotomy syndrome [39].

In pericarditis associated with acute rheumatic fever, treatment with anti-inflammatory agents usually effects a rapid reduction in the amount of pericardial effusion.

Percutaneous Management

- Echocardiography-guided pericardiocentesis

The approach is to assess the size, distribution, and ideal needle entry site and trajectory to safely evacuate the pericardial fluid.

The echocardiographic transducer is placed approximately 3–5 cm from the parasternal border, and the area of maximal pericardial fluid accumulation is identified. The needle trajectory is established by the angle of the transducer [40–42]. The precordium is entered in the angle formed between the xiphoid process and the left costal cartilages using an 18-mm gauge needle directed at an approximate 15° posterior angle toward the shoulder. The needle is advanced with the tip bent downward, while continuous suction is performed with a syringe until fluid is obtained. Adequate drainage of the pericardial fluid is assessed by echocardiography. Echocardiography-guided pericardiocentesis is simple, safe, and effective for primary treatment of clinically significant pericardial effusion, even in the postoperative period [43]. Complications include transient arrhythmia and cardiac injury with possible hemopericardium.

- Percutaneous pericardial drainage

Frequently, pericardiocentesis is accompanied by insertion of a drainage catheter to reduce the rate of recurrence that may complicate simple needle drainage. The precordium is entered from the standard subxiphoid approach using an 18-mm gauge needle until fluid is obtained. To assess the position of the needle in the pericardial sac, saline solution can be injected and monitored via echocardiography [44]. A 0.035″ guide wire is advanced into the pericardial space, under echoguidance. The needle is subsequently removed and the tract is dilated with a 7F or 8F dilator. A 7F or 8F pigtail catheter is then inserted over the guide wire, positioned in the posterior pericardial space at the level of the left atrioventricular groove.

- Percutaneous balloon pericardiotomy

The initial part of the procedure is similar to percutaneous pericardial drainage but is performed in the catheterization laboratory under fluoroscopic guidance. The parietal pericardium is dilated using a 10F dilator. Further dilation is performed using either a single balloon (20 mm wide, 3 cm long) or trefoil (triple) balloons. The balloon is filled with a mixture of contrast and saline and is manually inflated to a maximum pressure of 3.5 atm until the balloon "waist" disappears. Three inflations of 15 s each are recommended. At the end of the procedure, a pigtail catheter is exchanged over the wire and left in place to allow complete drainage of the effusion [45]. Complications include fever, pneumothorax, pleural effusion, and severe chest pain. The success rate of this procedure is high, and yet many would disagree that this technique is successful or durable [46].

- Pericardial sclerosing therapy

When significant pericardial effusion recurs, a more definitive intervention is needed. Pericardiocentesis with instillation of sclerosing agents has been shown to be successful for malignant pericardial effusions, with a low recurrence rate. Most commonly used are tetracyclin or bleomycin, instilled through a pigtail catheter [47, 48]. Common side effects include transient pyrexia, severe retrosternal chest pain, and transient atrial arrhythmia. Few data are available in children [49].

Surgical Management

- Subxiphoid pericardial drainage

Subxiphoid pericardiotomy is often preferred to percutaneous pericardiocentesis in critically ill patients or when echo-guided pericardiocentesis has failed. It is performed via a midline incision from the xiphisternal junction to 6–8 cm below the tip of the xiphoid. The xiphisternal junction is split and the xiphoid process removed to expose the diaphragm. The sternum is lifted so the pericardium can be reached. The pericardium is incised allowing the fluid to drain freely and a pericardial drain is left in place [44]. Minor complications include wound dehiscence and transient pneumothorax.

- Pleuropericardial window

Limited pericardiectomy is performed via a left thoracotomy. No attempt is made to excise all pericardial tissue, the main objective being to provide drainage of the pericardial sac into the left pleural space. This procedure can also be performed under direct thoracoscopic vision with excellent visualization of the pericardium and pleura.

Constrictive Pericarditis

Etiology and Epidemiology

Constrictive pericarditis is rare in children in developed countries, but as mentioned above, the incidence in undeveloped countries is much higher due to higher rates of tuberculosis. Clinical presentation depends on etiology and the rate of onset and severity of disease (Table 131.4) [50, 51].

Table 131.4 Causes of constrictive pericarditis

Causes of constrictive pericarditis	
Idiopathic	
Post-acute pericarditis	
Tuberculosis	
Infectious	Virus
	Bacteria: staphylococci, streptococci
	Fungi: histoplasmosis
Rheumatoid disease	
Sarcoidosis	
Mediastinal radiation	
Hemopericardium	
Post-cardiac surgery	
Uremia	
Neoplasia	
Metabolic disorders	
Genetic disorders	

Signs and Symptoms

The history reveals symptoms of congestive heart failure, such as dyspnea, orthopnea, paroxysmal nocturnal dyspnea, diaphoresis, easy fatigability, and tachycardia. Precordial pain is unusual in chronic constrictive pericarditis, as opposed to acute pericarditis. The hallmarks of physical diagnosis include absence of a drop in jugular venous pulsations during inspiration (Kussmaul sign) and elevated jugular pressure with prominent y descent (Friedreich's sign). Unlike other forms of pericardial disease, such as acute pericarditis, a friction rub is usually not audible. A protodiastolic knock, usually heard along the left sternal border, corresponds to the abrupt cessation of ventricular filling during diastole. As the systemic venous pressure becomes elevated, signs of right-sided heart failure develop, such as neck vein distention, hepatomegaly, ascites, hepatojugular reflux, and peripheral edema [52, 53]. Signs of diminished cardiac output include diminished pulse pressure, pulsus paradoxus, and a prominent third heart sound.

Diagnostic Workup

- Laboratory

Brain natriuretic peptide (BNP) is usually normal or just above normal in patients with constrictive pericarditis as opposed to elevated (>600 pg/ml) in patients with restrictive cardiomyopathy, helping differentiate between these two conditions [54]. No data on BNP levels in this setting are available in children.

- Chest X-Ray

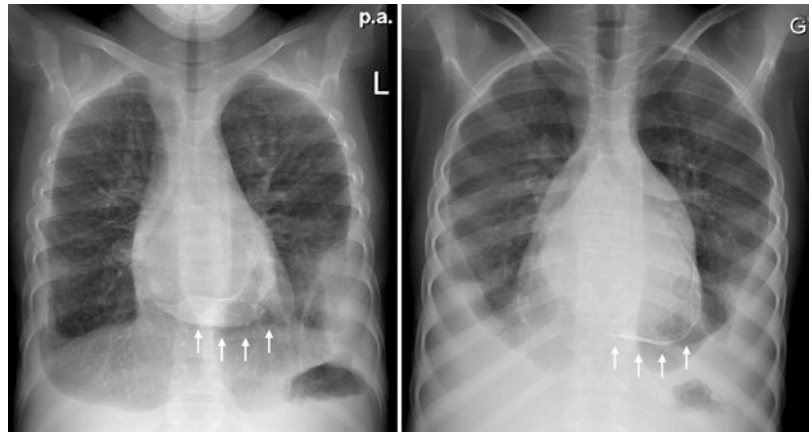
The chest X-ray is usually unremarkable. Pericardial calcifications are present in 40–50 % of patients, giving an eggshell appearance of the cardiac silhouette [55] (Pictures 131.6 and 131.7). The right superior mediastinum may be enlarged owing to dilation of the superior vena cava. Pleural effusions may be present, reflecting chronic elevation of right heart filling pressures. Pulmonary infiltrates are uncommon.

- Electrocardiogram

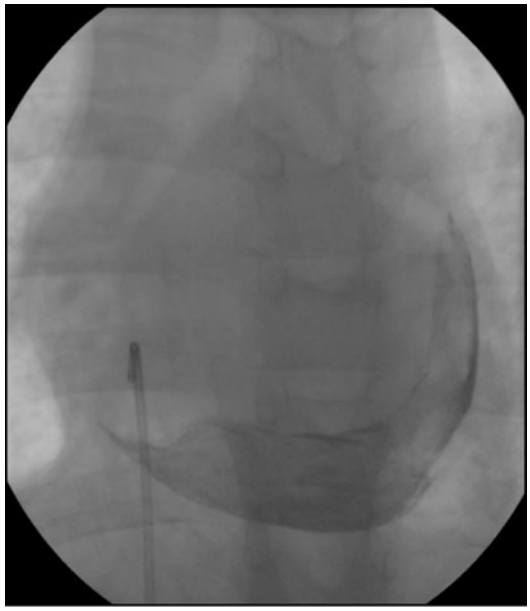
ECG is nonspecific but usually demonstrates diffuse low voltage and nonspecific ST-T wave changes. Atrial dysrhythmias are common.

- Cardiac catheterization

The hallmark finding in constrictive pericarditis is elevation and near equalization of end-diastolic pressures in the right atrium, right ventricle, pulmonary capillary wedge (left atrium), and left ventricle. The jugular venous or right atrial pulse waveform is characterized by a prominent A wave, reflecting rapid early diastolic filling of the ventricle, a sharp x descent, due to accelerated atrial relaxation, and a sharp y descent reflecting the early resistance free right



Picture 131.6 Chest X-ray in constrictive pericarditis: eggshell appearance of the cardiac silhouette with calcification of the pericardium (*arrows*)



Picture 131.7 Cardiac catheterization in constrictive pericarditis: calcification of the cardiac silhouette

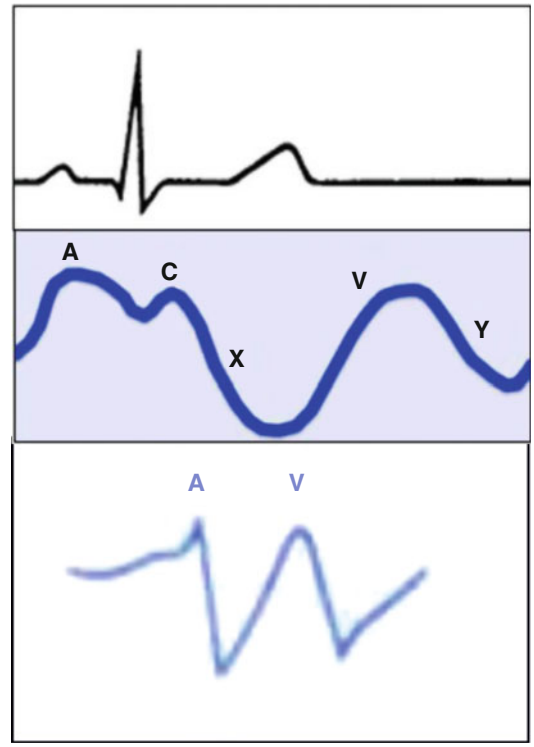


Fig. 131.5 Jugular venous pressure and atrial wave form: normal (*upper tracing* below the ECG) and in constrictive pericarditis (*bottom tracing*) with rapid x and y descents described as an "M-" or "W-" shaped pattern

ventricular filling (Fig. 131.5). The mean jugular venous and right atrial pressures are elevated.

The right ventricular waveform is distinctive, with a "dip and plateau" or "square-root sign" pattern (Fig. 131.6), reflecting the rapid relaxation, followed by a sharp increase in filling pressure as the expanding ventricle meets the constraints of the pericardium [56]. The left ventricular pressure tracing is usually similar. Other hemodynamic findings include a right ventricular diastolic pressure exceeding one third of the right ventricular systolic pressure and a pulmonary artery pressure of less than 50 mmHg [57].

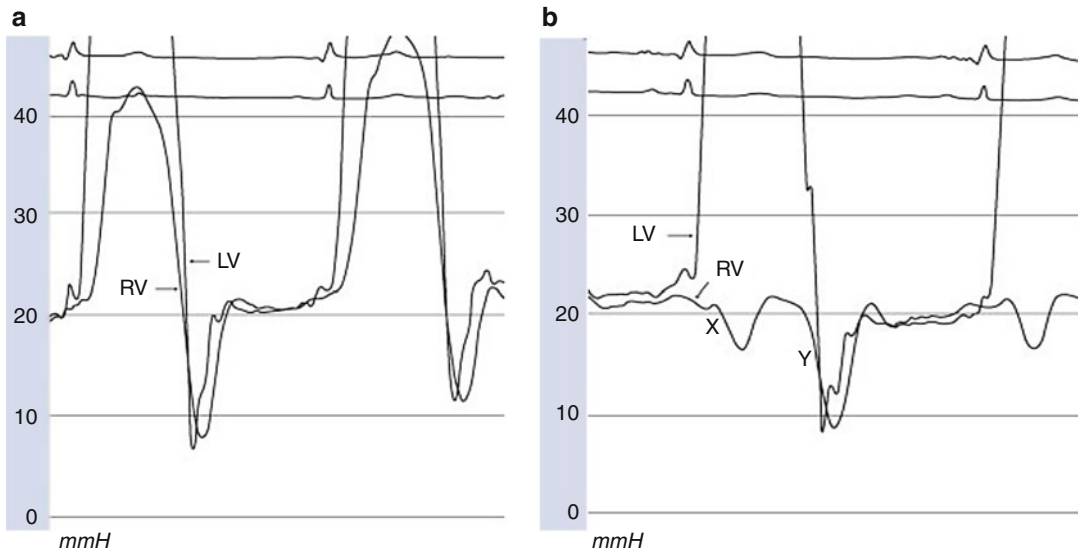


Fig. 131.6 Cardiac catheterization pressure tracings in constrictive pericarditis: (**a**) Simultaneous right and left ventricular pressure showing equalization of diastolic pressures and characteristic "dip and plateau" or "square-root" pattern of the right ventricular pressure curve. (**b**) Simultaneous left ventricular and right atrial pressure showing equalization of diastolic pressures and characteristic waves of the right atrial pressures with sharp x and y descents. *Abbreviations*: *RV* right ventricle, *LV* left ventricle, *RA* right atrium

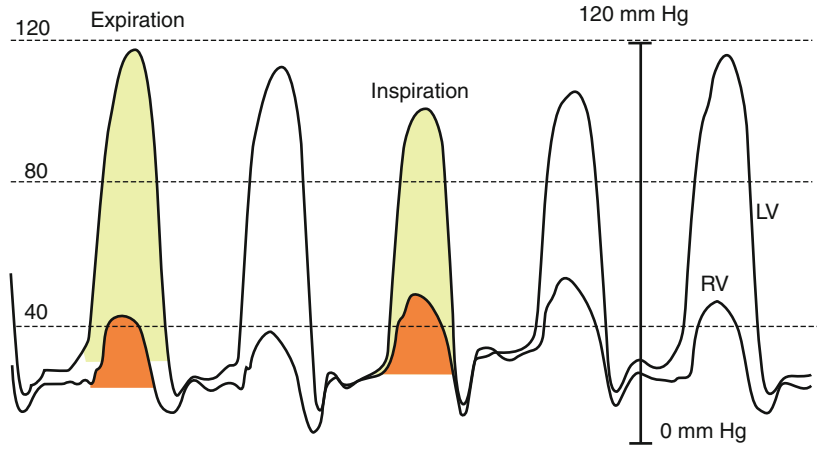


Fig. 131.7 Pressure tracing in constrictive pericarditis: during inspiration, there is an increase in the area under the RV pressure curve (*orange-shaded area*) compared with expiration, the area of the LV pressure curve (*yellow-shaded area*) decreases during inspiration as compared with expiration (Adapted from Talreja et al. [89]). *Abbreviations*: *RV* right ventricle, *LV* left ventricle

Another hallmark of constriction is increased ventricular interdependence. Because pericardial constraint limits total cardiac volume, there is a reciprocal relation between left and right heart filling due to enhanced septal interaction. There is opposite directional changes in ventricular systolic pressure and reciprocal changes with respiration, with inspiration inducing an increase in right ventricular but a decrease in left ventricular pressure, a phenomenon called ventricular discordance (Fig. 131.7). The opposite changes occur during expiration, with increased left heart filling and reduced right heart filling. This may be the most reliable hemodynamic indicator of

constriction [58]. When hemodynamic studies are equivocal, a bolus fluid can be administered and reveals striking elevation of the filling pressures in the case of constrictive pericarditis [59, 60].

- Echocardiography

Echocardiography remains the best tool in the initial assessment of constrictive pericarditis [61]. A thickened pericardium with some degree of pericardial effusion may be observed by 2D-echocardiography [62]. Transthoracic echocardiography is insensitive, as mildly increased pericardial thickening can be missed and false positives can be obtained if the gain is set too high. Pericardial calcifications with localized tethering of atrial or ventricular cavities may be noted, while separation of the entire pericardium by a small fixed space is known as the "halo sign." The systemic veins are usually dilated, with the inferior vena cava showing absent collapse with inspiration (plethora). Septal "bounce" is typical, defined as abrupt posterior movement of the interventricular septum in early diastole during inspiration, and is caused by underfilling of the left ventricle and redistribution of blood from the left to the right ventricle. This "bounce" represents the first and best clue for the presence of constriction [63].

The right and left ventricular size is decreased, and both atria are mildly enlarged, related to the compliance abnormality of the ventricles. The ventricles have an elongated appearance giving the heart a tubular shape. The biventricular systolic function is usually normal. Interventricular septal motion may be paradoxical or flat as a sign of ventricular interdependence. A characteristic septal notch has been described in early diastole (Picture 131.6), corresponding to the septal bounce seen by 2D-echocardiography [64, 65]. Extensive areas of adhesions seen posteriorly by M-mode provide evidence for generalized pericardial thickening and constriction.

The hallmark of Doppler examination is reciprocal respiratory variation of right and left heart flows caused by interventricular dependence. The classical Doppler pattern consists of the following (Fig. 131.8) [66–68]:

– Mitral inflow: During inspiration, E wave to A wave ratio (E > A) is lower, while during expiration, there is larger E wave to A wave (E > A) ratio. E wave is typically increased more than 25 % with expiration and the IVRT increased more than 25 % with inspiration (Picture 131.8).
– Pulmonary veins: During inspiration, S and D waves are near equal in size. During expiration, larger S and D waves are noted.
– Tricuspid inflow: It shows the same pattern with reciprocal changes compared to the mitral inflow. During expiration, smaller E wave to A wave (E > A) ratio is noted, while during inspiration, there is larger E wave to A wave (E > A) ratio. E wave is typically increased more than 40 % with inspiration.
– Hepatic veins: During inspiration, S wave is greater than D wave, with a small A wave reversal. During expiration, S wave is greater than D wave, with small or absent D wave and larger A wave reversal.

Also described in constrictive pericarditis is an inspiratory increase in the tricuspid regurgitant jet velocity and duration of the signal [58]. As opposed to restrictive cardiomyopathy, respiratory variation in the filling phase is more pronounced in constrictive pericarditis. Tissue Doppler echocardiography shows a normal or high early mitral annular velocity (Em wave) in constrictive pericarditis (Picture 131.9), as opposed to restrictive cardiomyopathy where it is reduced [69]. The usually positive linear relation between mitral Doppler E and tissue Doppler Em (E/Em) is useful to assess left atrial pressure and is found to be reversed in constrictive pericarditis [70].

- Cardiac MRI and CT

Both CT and MRI can detect a thickened pericardium (≥4 mm), but this is an insensitive finding. An advantage of CT is the ability to detect calcification (Picture 131.10), indicative of constrictive pericarditis [71, 72]. However, CT may have difficulty differentiating pericardial fluid from thickened pericardium. The absence of pericardial thickening does not rule out hemodynamically significant constrictive pericarditis.

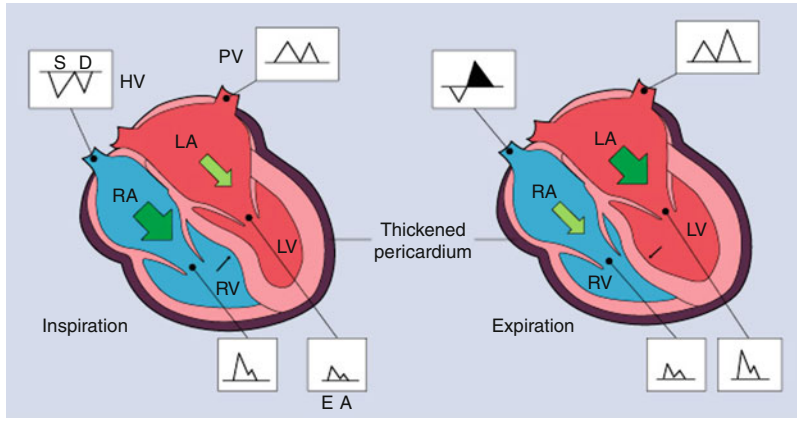
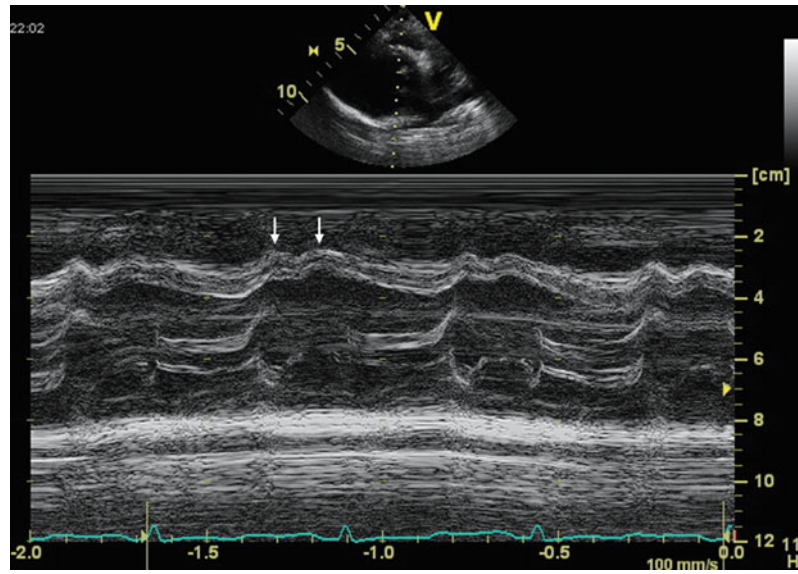


Fig. 131.8 Ventricular interdependence and respiratory variation in transvalvular and central venous flow velocities in constrictive pericarditis. With inspiration, the driving pressure gradient from the pulmonary bed to the left cardiac chambers decreases, resulting in a decrease in mitral inflow and diastolic pulmonary venous velocities. Decreased left ventricular filling results in ventricular septal shift to the left, allowing increased flow to the right-sided cardiac chambers, resulting in increased tricuspid inflow and diastolic hepatic venous velocities. The opposite changes occur during expiration (Adapted from Oh [90]). *Abbreviations*: *A* late diastolic (atrial reversal) Doppler wave, *D* diastolic Doppler wave, *E* early diastolic Doppler wave, *HV* hepatic vein, *LA* left atrium, *LV* left ventricle, *PV* pulmonary vein, *RA* right atrium, *RV* right ventricle, *S* systolic Doppler wave



Picture 131.8 M-mode echocardiography in constrictive pericarditis: notched interventricular septum (*arrow*)

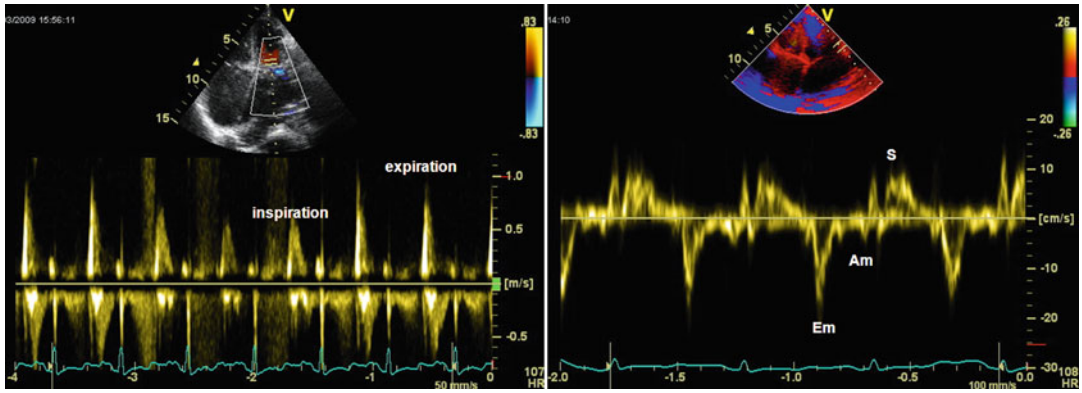
Medical Management

The treatment is essentially symptomatic, with diuretics to reduce right heart failure and pulmonary edema. The only curative treatment is pericardiectomy.

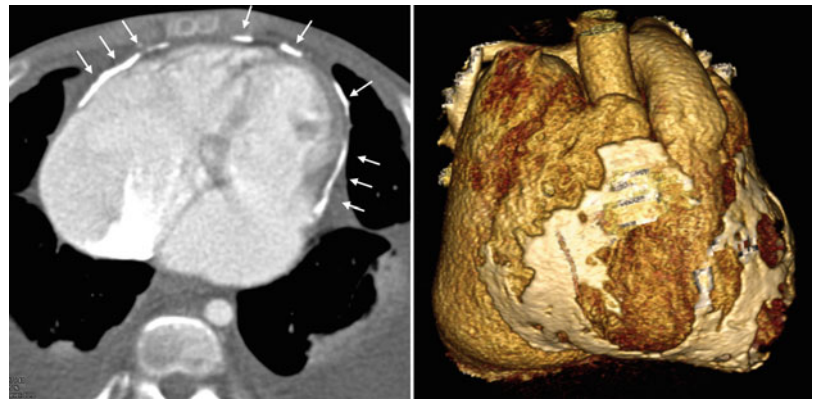
Surgical Management

- Pericardiectomy

Pericardiectomy is the treatment of choice for symptomatic patients with typical constrictive hemodynamics. Limited pericardiectomy is usually



Picture 131.9 Doppler echocardiography in constrictive pericarditis: respiratory variation in mitral inflow with increased E wave velocity with expiration, secondary to ventricular interdependence. Tissue Doppler imaging (TDI): normal or increased early diastolic (Em) wave velocity



Picture 131.10 Cardiac computed tomography (CT): pericardial calcifications (*arrows*). 3D reconstruction: calcified pericardium

performed via a left thoracotomy, but does not allow access to the right atrium and vena cava. Total pericardiectomy is defined as wide excision of the pericardium from around both ventricles, the great vessels, the vena cava, and the right atrium. It is important to include pericardium both anterior and posterior to the phrenic nerves, which must be properly identified and spared. It is usually performed via a median sternotomy or bilateral trans-sternal anterior thoracotomy. A median sternotomy with cardiopulmonary bypass standby is usually the preferred approach as it offers better exposure to the right side of the heart [73].

Poor results with persistent elevation of ventricular filling pressures have been attributed to inadequate decortication and remodeling of the ventricles after pericardiectomy. Complications include excessive bleeding and low cardiac output syndrome, thought to be secondary to fibrosis and atrophy of the myocardial fibers. Reoperation for recurrent constrictive pericarditis after partial pericardiectomy is common [74]. Improvement of symptoms and normalization of the intracardiac pressures occurs more quickly after extensive pericardiectomy [75].

Congenital Anomalies of the Pericardium

Etiology and Epidemiology

Congenital anomalies of the pericardium are uncommon but should be considered in children. *Pericardial cyst* is rare and usually congenital but may also be acquired after cardiothoracic

surgery. Cysts are typically located at the right cardiophrenic angle (50–70 %) or at the left cardiophrenic angle (28–38 %). *Congenital absence of the pericardium* can be complete or partial and can be isolated or associated in one third of the cases with other congenital cardiac anomalies (patent ductus arteriosus, atrial septal defect, mitral stenosis, tetralogy of Fallot) or congenital pulmonary anomalies (sequestration, bronchogenic cyst, diaphragmatic hernia). Eighty percent of defects occur on the left side and are related to premature atrophy of the left duct of Curvier during embryologic development.

Signs and Symptoms

Although most *pericardial cysts* are asymptomatic, patients may present with atypical chest pain, dyspnea, or persistent cough [76]. Complete *congenital absence of the pericardium* is often an incidental finding with chest imaging demonstrating deviation of the heart into the left chest. In those with symptoms, paroxysmal stabbing chest pain, largely nonexertional and mimicking coronary artery disease, dyspnea, and syncope can be associated [77]. Life-threatening complications include herniation of a cardiac chamber through the defect, most commonly the left atrial appendage, torsion of the great arteries, or constriction of a coronary artery at the rim of the defect. Displacement of the left ventricular impulse on clinical exam is the most common feature.

Diagnostic Workup

- Chest X-Ray

  A *pericardial cyst* is typically suspected after an abnormal chest X-ray consisting of a round, discrete mass in the right cardiophrenic angle, which is the most common location of these cysts [78]. In *congenital absence of the pericardium*, the chest X-ray reveals levoposition of the heart with loss of the right heart border hidden by the spine [77]. Prominence of the main pulmonary artery and interposition of a tongue of lung tissue between the pulmonary artery and the aorta (opacification of the aortopulmonary window) or between the inferior border of the heart and the left hemidiaphragm are other findings.
- Electrocardiogram

  In *congenital absence of the pericardium*, right bundle branch block is common. Right axis deviation with leftward displacement of the transition zone in the precordial leads can be seen.
- Echocardiography

  Pericardial cysts are difficult to detect with transthoracic echocardiography. They present as an echo-free space which is more localized and spherical than a pericardial effusion [79].

  Complete absence of the pericardium leads to enlargement of the right ventricle, excessive motion of the posterior left ventricular wall, paradoxical motion of the interventricular septum, and a shift of the heart to the left resulting in more of the right ventricle being seen on the left parasternal long axis view. All of these findings mimic right ventricular volume overload and thus this diagnosis should be excluded [80]. Partial absence of the pericardium sometimes results in herniation of a chamber through the defect, with the false appearance of wall motion abnormality. The biventricular function is usually normal. True wall motion abnormality is seen if a coronary artery is compressed.
- Cardiac CT and MRI

  CT and MRI are the preferred methods to confirm a suspected diagnosis of *pericardial cyst* [76, 81]. On CT scan, pericardial cysts are thin-walled, sharply defined, oval homogeneous masses. Their attenuation is slightly higher than water density, 30–40 HU, and the cyst fails to enhance with intravenous contrast [82]. The most reliable finding in *congenital absence of the pericardium* is interposition of lung tissue between the main pulmonary artery and the aorta. The heart can be completely displaced in the left hemithorax and its apex elevated. The main pulmonary artery and the left atrial appendage can be seen extending far beyond the mediastinal margins.

Surgical Management

Surgical procedures employed for patients with *absence of the pericardium* include left atrial appendectomy, division of adhesions, pericardiectomy, or pericardioplasty. The latter is usually reserved for symptomatic patients, as the symptoms are thought to be secondary to excessive cardiac motion. It is controversial as to whether asymptomatic patients with moderate-sized pericardial defects should undergo prophylactic operation to reduce the risk of death from cardiac structure herniation or incarceration [83]. Surgical reconstruction of the pericardium (pericardioplasty) can be performed with PTFE (polytetroflouroethylene) material or xenograft pericardium. The lateral and anterior surfaces of the newly reconstructed pericardium are then sutured to the lateral and medial aspect of the diaphragmatic surface to avoid excessive cardiac motion. Careful attention must be paid to the left phrenic nerve [77].

For patients with *pericardial cyst*, surgical excision is recommended only in symptomatic patients, while asymptomatic patients can be managed conservatively [84]. Minimally invasive thoracoscopic resection of the cyst is a good alternative, as it minimizes postoperative pain and has a better cosmetic outcome [85].

Postoperative Management

Among the pericardial disease processes, a common postoperative strategy may be employed. In large part, postoperative care is supportive with additional treatment directed at the underlying etiology of the disease. Management of the patient in ICU consists of fluid balance monitoring, sedation and analgesia, respiratory management, inotropic and vasodilator therapy, and recognition of anticipated complications.

In many patients, the duration of the pericardial disease process will impact their postoperative course.

- Monitoring

Continuous cardiorespiratory monitoring remains standard for these patients. Attention should be paid to alterations in heart rate, blood pressure, and respiratory rate. Most patients should be maintained in the physiologic range after returning to the intensive care unit.

- Fluid Management

The fluid status of a patient with pericardial disease returning to the ICU environment should be monitored carefully. Many postoperative patients may have undergone aggressive diuresis prior to surgery, and a few may be diuretic dependent. Maintenance intravenous fluid therapy should be initiated on patients unless contraindicated by concurrent illness. Most will not need the aggressive fluid management of other postoperative cardiac patients. The exceptions are those patients with restrictive physiology in whom fluid balance will become important as their disease process progresses.

- Sedation and Analgesia

Most patients should be extubated in the operating room or upon return to the intensive care unit (ICU). Sedation should not be a significant issue in the postoperative period. Pain control can be achieved with continuous narcotic infusion or boluses. Pain from thoracotomy should not be underestimated, especially in the older patients, as atelectasis secondary to shallow breathing can be a serious complication. The transition to oral pain management should occur when the patient is tolerating an oral diet. In the older child or adolescent, intermittent oral or intravenous benzodiazepines may be used for anxiolysis. Additionally, intravenous ketorolac or nonsteroidal anti-inflammatory drugs (NSAIDs) may be useful in patients with an inflammatory component to their pericardial disease.

- Respiratory Management

Most patients are extubated in the operating room or immediately upon return to the intensive care unit. Adequate pain control helps to avoid one of the most common complications after thoracotomy or sternotomy: atelectasis.

- Inotropic and Vasodilator Therapy

A low cardiac output state may be treated with volume resuscitation, inotropic support, and afterload reduction. Additionally, vasoactive medications may be used depending upon the patient's clinical state.

- Anticipated Complications

In-hospital mortality after pericardiectomy for constrictive pericarditis is not negligible, around 15 %. Complications after surgery include low cardiac output syndrome and hemorrhage. Patients should be monitored for persistent effusion or restrictive physiology after surgical intervention.

Long-Term Outcome

Pericardiectomy improves symptomatology in the majority of patients during late follow-up. A subgroup of patients do not experience an amelioration in clinical symptoms, probably because myocardial function does not completely recover [86]. This is particularly true for patients with long-standing constriction, especially in the setting of tuberculosis. Right ventricular dysfunction has been associated with myocardial involvement and absence of clinical improvement after pericardiectomy [87]. Recurrence is the most troublesome complication of pericarditis, occurs in 15–50 % of patients and is probably an autoimmune process. The overall prognosis in idiopathic recurrent pericarditis is excellent and complications are uncommon. Even after numerous recurrences of pericarditis, constrictive pericarditis as a complication is extremely rare. The risk of evolution to constrictive pericarditis in idiopathic acute pericarditis is estimated to be around 1 % [88]. The risk of progression to constriction is higher in tuberculous, neoplastic, or purulent pericarditis.

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