Pericardial Disease

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Pericardial Disease

The heart is protected by tissue layers that make up the pericardial sac. The parietal pericardium is a thick fibrous structure that can be congenitally absent without clinical significance. The visceral pericardium is adherent to the epicardial layer of the heart. The potential space between the two layers contains a small amount of serous pericardial fluid to cushion and protect the heart, allowing it to move freely within the pericardial sac. It is important to recognize that the parietal pericardium extends up to and includes the ascending root of the aorta (Fig. 23.1). This becomes clinically relevant with aortic dissection. Blood tracking outside the aorta will collect in the pericardial space with resultant hemodynamically significant effusion and clinical tamponade (Chap. 28).

Pericarditis

Pericarditis is inflammation of the pericardial sac surrounding the heart. Acute pericarditis is the most common disorder involving the pericardium and is often secondary to a viral process [1]. Other common causes are systemic autoimmune diseases and neoplastic disease with pericardial involvement (Table 23.1) [2]. The clinical diagnosis is made when two of the four criteria listed in Table 23.2 are present. Commonly, a patient will present with inability to lay flat due to sharp and stabbing chest pain. The pain is usually sudden in onset and often described as centrally located, worse with deep inspiration, and pleuritic in nature. Pain frequently improves when sitting forward. Patients may describe radiation to the trapezius ridge, the area from the neck to the shoulder region. Recurrence of inflammation will have very similar symptoms to initial presentation. Many also complain of associated fatigue, shortness of breath, orthopnea, or PND.

The classification of pericarditis is related to the duration and recurrence of symptoms (Table 23.3).

Physical Exam

Commonly, the physical exam is unremarkable. Occasionally, a pericardial rub may be heard with the patient leaning forward, during expiration at the left lower sternal edge. It may have one, two, or three components. Usually, a single sound during ventricular systole can be heard intermittently. It has been described sounding like the separation of Velcro™ and can be mistaken for a systolic murmur. The presence of a pericardial effusion often negates the presence of the rub as

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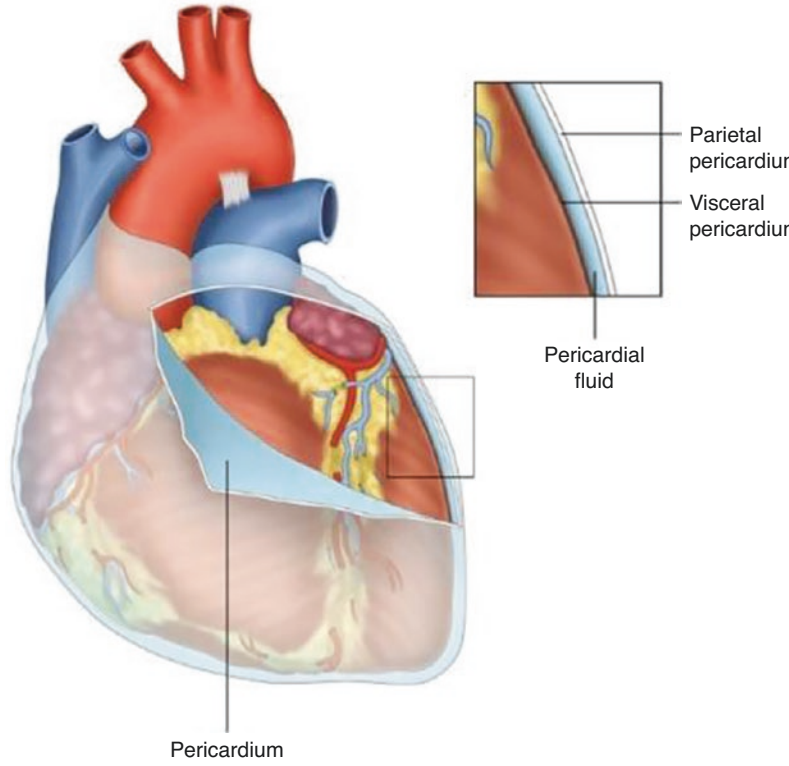

Fig. 23.1 Anatomy of pericardium. (Adapted from Poorsattar, S.P., Maus, T.M. (2022). Pericardium. In: Maus, T.M., Tainter, C.R. (eds) Essential Echocardiography. Springer, Cham. https://doi.org/10.1007/978-3-030-84349-6_14)

Table 23.1 Causes of pericarditis

Etiologies	Examples
Infectious	Viral: Idiopathic. COVID-19-common Bacterial: TB—more common in underdeveloped countries Fungal-rare
Uremic pericarditis	ESRD patients
Post-myocardial infarction (Dressler's syndrome)	Less commonly seen due to early MI intervention
Autoimmune disease	RA, SLE serositis—common
Malignancies	Metastatic cancer-lung, melanoma, and breast—common

Table 23.2 Diagnostic criteria by the American College of Cardiology requires two of four criteria

Chest pain that is positional and pleuritic
Physical exam finding of friction rub
EKG changes characteristic of pericarditis
New or worsening pericardial effusion

Table 23.3 Classification of pericarditis

Acute	Criteria present in Table 23.2, CRP and sedimentation rate elevation, CT or MR imaging showing pericardial inflammation
Incessant	Symptoms lasting >4–6 weeks and less than 3 months
Recurrent	Symptom recurrence after 4–6 weeks or longer of a symptom-free interval
Chronic	Symptoms greater than 3 months

Adapted from Yehuda Adler, Philippe Charron, Massimo Imazio, Luigi Badano, Gonzalo Barón-Esquivias, Jan Bogaert, Antonio Brucato, Pascal Gueret, Karin Klingel, Christos Lionis, Bernhard Maisch, Bongani Mayosi, Alain Pavie, Arsen D Ristić, Manel Sabaté Tenas, Petar Seferovic, Karl Swedberg, Witold Tomkowski, ESC Scientific Document Group, 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC) Endorsed by: The European Association for Cardio-Thoracic Surgery (EACTS), European Heart Journal, Volume 36, Issue 42, 7 November 2015, Pages 2921–2964, https://doi.org/10.1093/eurheartj/ehv318

the fluid lubricates the inflamed pericardial layers. A current or recent fever may be associated if infectious process is present.

Diagnostic Testing

EKG may show diffuse concave or saddle-shaped ST elevation in multiple vascular distributions. If these changes were all true STEMI, hemodynamic collapse and cardiogenic shock would be clinically present. Diffuse PR segment depression with isolated PR segment elevation in aVR is usually present simultaneous with the ST segment changes (Fig. 23.2).

Transthoracic Echocardiogram: Since pericarditis is a clinical diagnosis, there are no specific echocardiographic findings in acute pericarditis. The imaging modality may be a helpful diagnostic tool to assess for pericardial effusion and tamponade physiology. Assessment for a focal wall motion abnormality should be undertaken potentially indicating post-infarct pericarditis or myocarditis.

Labs: Inflammatory markers may be elevated including white blood cell count, C-reactive protein, and sedimentation rate. High sensitivity troponin is used to assess for myocardial damage and associated myocarditis or ischemic etiology. If there is a classic presentation of positional chest pain with other clinical signs of pericarditis, an elevation of troponins suggests myopericarditis (Table 23.6).

Cardiac MRI (cMRI) may be useful to assess for pericardial inflammation and to evaluate associated myocarditis (Fig. 23.3).

Management of Acute Pericarditis

Uncomplicated pericarditis should be managed in the ambulatory setting (Table 23.4). High-risk features including an effusion with hemodynamic compromise necessitating admission for inpatient management (Fig. 23.4). Acute inpatient management includes Ketorolac 15–30 mg IV for 1–2 doses for initial pain management and immediate initiation of colchicine.

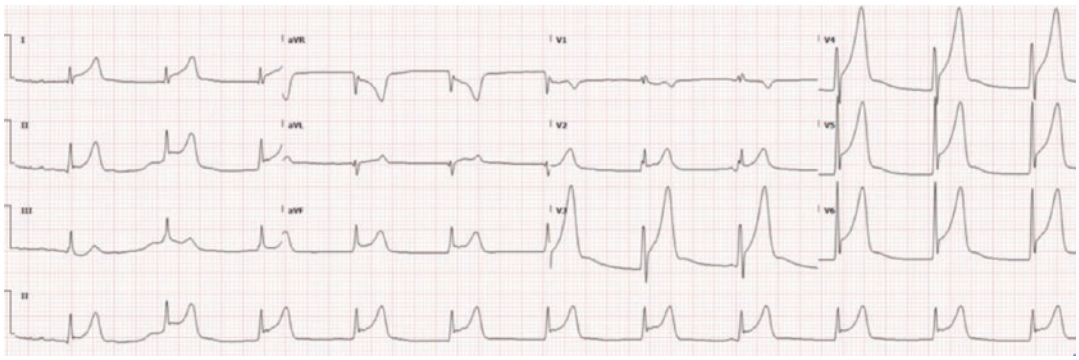

Fig. 23.2 EKG suggestive of acute pericarditis

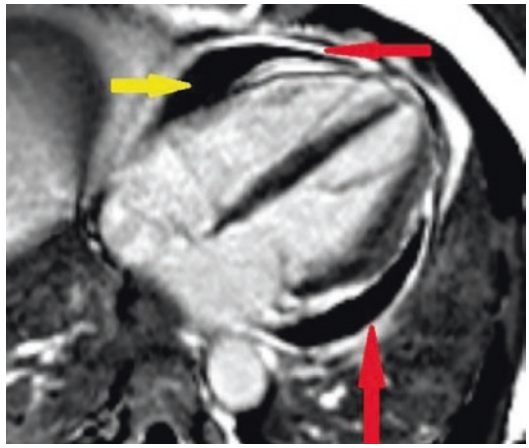

Fig. 23.3 MRI of pericarditis and effusion. Yellow arrows indicate pericardial effusion. Red arrows are gadolinium enhancement of inflamed pericardium

Table 23.4 Treatment options for acute pericarditis

Drug	Usual dosing	Duration of therapy	Tapering
High-dose aspirin	750–1000 mg every 8 h	1–2 weeks	Decrease by 250–500 mg every 1–2 weeks
Ibuprofen	600 mg every 8 h	1–2 weeks	Decrease by 200–400 every 1–2 weeks
Colchicine	0.5 mg daily (<70 kg) or 0.5 mg twice daily (≥70 kg)	3 months	Continue for duration

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Corticosteroids should be avoided as initial treatment since corticosteroids could suppress the patient's immune response to a virus and therefore maintain the trigger for inflammation [3]. This may lead to increased risk of chronic pericarditis and constrictive pericardial physiology. If recurrent pericarditis does occur, colchicine for 6 months along with NSAIDs or high-dose ASA should be continued until the relief of symptoms occurs [4]. Cardiac MRI is now recommended in cases of recurrent pericarditis. Corticosteroids are considered to treat recurrence if the serum CRP is low and the patient is without cardiac MRI abnormalities. Recently, a new agent, rilonacept, was approved to treat recurrent pericarditis, especially if abnormal MRI findings of late gadolinium enhancement are seen within the pericardium. This injectable agent inhibits IL-1 alpha and IL-1 beta and thus alters the autoinflammatory pathway associated with recurrent pericarditis.

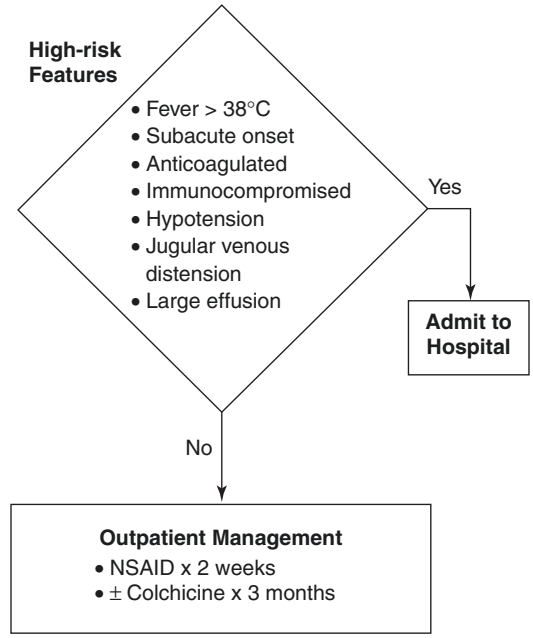

Fig. 23.4 Pathway for treatment of acute pericarditis. (Adapted from Lilly, L., 2013. Treatment of Acute and Recurrent Idiopathic Pericarditis. *Circulation*, 127(16), pp.1723–1726)

Clinical Pearls

- Pericarditis is a clinical diagnosis. A good history of present illness is all that is needed.
- Inquire about recent cold symptoms or fever as recent viral illness is a very common cause of pericarditis.
- Always obtain an EKG to rule out ischemia as an etiology.
- If hemodynamically unstable, echocardiography and clinical bedside assessment to evaluate for tamponade.

- Avoid corticosteroids as they can often lead to recurrent pericarditis.
- Exercise restriction for 3 months or until resolution of symptoms and normalization of CRP, ECG, MRI, and echocardiogram.

Myocarditis

Myocarditis is inflammation of the heart muscle tissue. Clinically, myocarditis and pericarditis can present at the same time. Similar to pericarditis, viral infection and replication in myocarditis can cause myocardial injury and cell death. The injury occurs through direct invasion, production of cardiotoxic substances, or chronic inflammation [5]. Clinical pericarditis with significant troponin elevation and LV dysfunction suggests myocarditis (see Chap. 20). This diagnosis should be suspected in patients who present with or without new cardiac symptoms with a rise in cardiac biomarkers, abnormal LV function, or change in EKG [6].

Constrictive Pericarditis

Constrictive pericarditis is typically a chronic condition that results when granulation tissue forms scar/fibrosis that encases the heart. This causes a loss of elasticity of the pericardial sac. Scarring can become calcified and lead to a compressive syndrome where the ventricles are unable to adequately fill [7]. The disease is often progressive in nature, and nonspecific early signs of cor pulmonale often delay the diagnosis. Traditional treatment often is not successful, and this diagnosis must be considered if there are risk factors for constriction (Table 23.5).

Table 23.5 Common causes of constrictive pericardial disease

Acute or relapsing viral or idiopathic pericarditis
Any type of cardiac surgery
Trauma with organized blood within pericardial space
Mediastinal irradiation
Neoplastic disease
Rheumatoid arthritis
Systemic lupus erythematosus
ESRD/chronic dialysis patients

Adapted from Hoit, B., 2022. *UpToDate*. [online] Uptodate.com

Physiological Characteristics

Constriction causes hemodynamic compromise due to the fixed obstruction to rapid early diastolic filling. An effusion does not need to be present for constrictive pericardial disease. If it is present, it is referred to as constrictive-effusive disease. Pericardiocentesis can alleviate the additional obstruction if an effusion is present, but the underlying constriction will still cause clinical cor pulmonale.

Normally, LV filling is independent of respiratory variation. In constrictive pericarditis, elevated pericardial pressures impede diastolic filling of the RV and eventually the LV. As respiration occurs, there is increased blood return to the fixed RV. The pressures equilibrate due to constriction or an effusion, and the septum pushes into the LV. The abnormal septal motion will impede LV filling and results in lower stroke volume and cardiac output. When this occurs, the ventricles become interdependent. This interdependence causes variation of LV filling associated with respiration. Echocardiography can image this interdependence, and the abnormal septal motion with inspiration is easily seen. Constriction is a clinical diagnosis supported by cardiac imaging. Occasionally, cardiac catheterization is necessary to confirm equalization of diastolic pressures.

Constriction and restrictive cardiomyopathy can be a clinical dilemma. The main difference is the presence of pericardial abnormalities with ventricular interdependence on imaging in constrictive disease. Restriction has abnormalities of the myocardium with evidence of pulmonary hypertension due to chronic HFpEF [8].

Physical Exam

The examination is very similar to pericardial tamponade. The chronicity of the constriction results in more extensive signs of cor pulmonale as seen in Table 23.6.

Table 23.6 Clinical examination findings of constrictive pericarditis

Elevated JVP with a deep, steep Y descent
Kussmaul sign—lack of an inspiratory decline in JVP
Peripheral edema
Ascites and hepatomegaly
Pleural effusion
Pericardial knock—slightly earlier than a third heart sound
Pulsus paradoxus—uncommon without effusion

Adapted from Hoit, B., 2022. *UpToDate*. [online] Uptodate.com

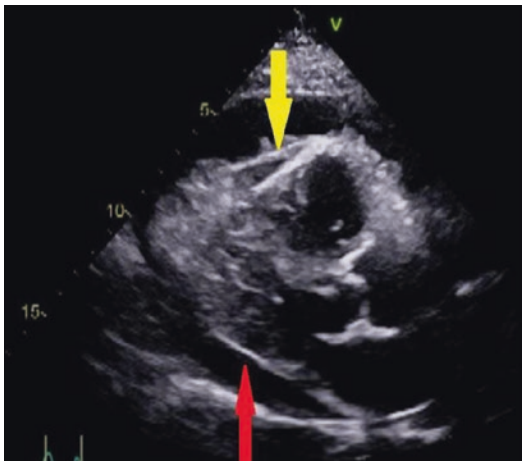

Fig. 23.5 Constrictive effusive pericarditis. The yellow arrows show the thickened parietal pericardium with fibrinous exudate attached suggestive of a chronic disease. The red arrow shows the pericardial effusion

Noninvasive Testing

Echocardiography is the initial test of choice. Signs of chronic effusion with thickening of the visceral and parietal pericardium are often noted. Abnormal septal bounce (ventricular interdependence) is seen with ventricular contraction and varies with respiration (Fig. 23.5).

Cardiac CT is a very good test to evaluate for pericardial thickening as is MRI. CT imaging is more readily available and has excellent diagnostic accuracy. An effusion does not have to be present for constriction physiology (Fig. 23.6).

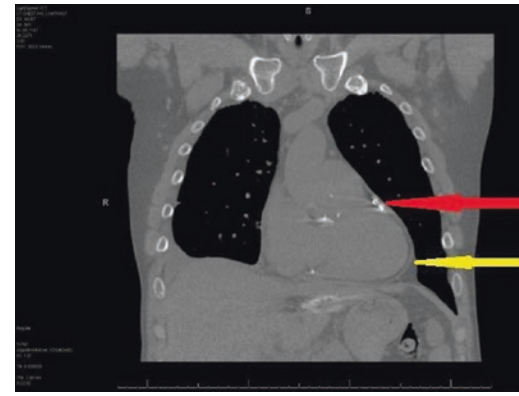

Fig. 23.6 CT of pericardium. Red arrow shows calcification of the pericardium. Yellow arrow shows marked thickening of the pericardium. Both finding suggest constrictive pericardial disease. Note the absence of a significant effusion

Treatment

Constrictive pericardial disease is difficult to diagnose and treat. Diuretic therapy to manage symptoms of cor pulmonale is the cornerstone of treatment. Often, this becomes inadequate and signs of intravascular volume depletion and end-organ hypoperfusion become problematic. Surgical pericardial stripping can be performed but only after the failure of conservative management. This is a high-risk procedure that has high surgical mortality but can significantly improve this disease. During this procedure, parietal pericardium is removed allowing passive filling of the ventricles. If the disease has progressed significantly, the visceral pericardium may be involved, and the patient may have minimal improvement postoperatively. Appropriate patient selection is essential prior to this procedure.

Clinical Pearls

- Consider constriction in patients with risk factors and treatment-resistant cor pulmonale
- Diuretics are used aggressively before high-risk surgery is considered

Pericardial Effusion

The normal pericardial sac contains 10–50 ml of serous fluid. This fluid is designed to lubricate the heart during contraction. Certain disease states result in an increased production of this physiologic fluid. A pericardial effusion may be present and asymptomatic. The effusion is often an incidental finding on other imaging studies and only significant if its size causes hemodynamic instability. A pericardiocentesis may be performed to rapidly treat hemodynamic instability or may be performed for diagnostic evaluation to determine the etiology (Table 23.7).

Physical Examination

Heart sounds may be faint due to increased distance between the chest and the heart. Clinically, the measurement of pulsus paradoxus is the bedside examination finding of ventricular interdependence. Pulses paradoxus is a clinical clue to tamponade and consists of a greater than 10 mmHg inspiratory decline in systolic arterial pressure. This evaluation is performed by increasing the pressure of sphygmomanometer cuff until there are no Korotkoff sounds. Air is slowly released from the cuff until sounds are intermittently heard. The cuff is deflated until the sounds are present continuously. The difference in numbers from beginning of intermittent and continuous is calculated. You should also palpate the radial pulse, and if you note the pulse weakens or disappears during inspiration, this is also a positive sign [5]. Normal is less than 10 mmHg. Hemodynamic significance is seen at 30 mmHg.

Table 23.7 Common causes of pericardial effusion

Common causes of cardiac tamponade	Uncommon causes of cardiac tamponade
Pericarditis	Collagen vascular disease (SLE, RA, scleroderma)
Tuberculosis	Radiation induced
Iatrogenic/procedure	Post myocardial infarction
Trauma	Uremia
Neoplasm/malignancy	Aortic dissection
	Bacterial infection
	Pneumopericardium

Adapted from Yehuda Adler, Philippe Charron, Massimo Imazio, Luigi Badano, Gonzalo Barón-Esquivias, Jan Bogaert, Antonio Brucato, Pascal Gueret, Karin Klingel, Christos Lionis, Bernhard Maisch, Bongani Mayosi, Alain Pavie, Arsen D Ristić, Manel Sabaté Tenas, Petar Seferovic, Karl Swedberg, Witold Tomkowski, ESC Scientific Document Group, 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: The Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC)

Diagnosis

The gold standard for diagnosis of a pericardial effusion is with echocardiography (Fig. 23.7). It is rapidly available and simple test to evaluate hemodynamic stability. Stable pericardial effusion can be appreciated on a CT scan or MRI. Hemodynamic abnormalities can be seen on these imaging modalities, but echocardiography is more readily available. CT can sometimes overestimate the size of effusion, and the size should be confirmed with echocardiography.

EKG findings are nonspecific. When a pericardial effusion is present and large, you may see electrical alternans on the EKG. This is caused by the heart "floating" in the fluid and moving with respiration and contraction toward and away from the EKG leads.

The cardiac silhouette may be enlarged on chest X-ray due to the presence of fluid.

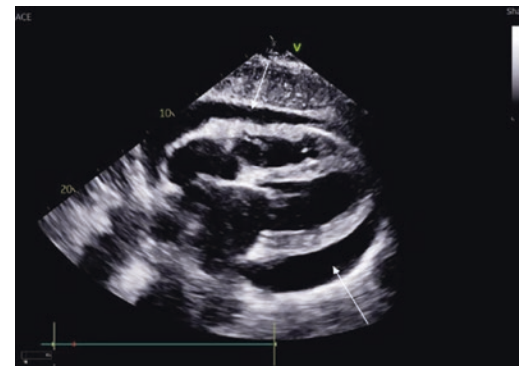

Fig. 23.7 Pericardial effusion on echocardiography. White arrows point to large circumferential pericardial effusion

Treatment

Asymptomatic effusions are usually observed without treatment. If the etiology is unclear and enough fluid is present, pericardiocentesis may be used as a diagnostic test. Treatment of an underlying disease is often indicated. Rarely, empiric anti-inflammatory agents are given if the sed rate and CRP are elevated (Table 23.4).

Cardiac Tamponade

Cardiac tamponade is a life-threatening condition that occurs when the accumulation of fluid in the pericardial space compresses the ventricle resulting in obstruction to ventricular filling. The quantity of fluid is of less concern but more so how rapidly it accumulates. The amount of fluid needed to cause tamponade could be as small as 200 ml if it develops rapidly. When pericardial effusions are chronic or develop more slowly, the pericardium has time to adapt, and there is less chance of hemodynamic compromise.

Clinical Signs

Three principal clinical features of tamponade are hypotension, soft or absent heart sounds, and jugular venous distention with a prominent descent during systole. These three clinical features are known as Beck's triad. Since the main issue is reduced preload to the ventricles from external compression, the heart rate increases to maintain a cardiac output. The SVR increases to maintain blood pressure. The patient in tamponade presents cool, diaphoretic, tachycardic, and hypotensive. *Never* give beta blockers to reduce the heart rate as this will result in cardiovascular collapse.

Diagnosis

Echocardiography will show collapse of the RA and RV walls during diastole. The collapse confirms impaired filling of the ventricle. Blood flow across the mitral valve is also assessed to determine if there is variation with respiration. These signs together confirm tamponade.

Treatment

Pericardiocentesis is indicated if there is hemodynamic compromise with tamponade physiology. If immediate removal of fluid is not possible, aggressive fluid resuscitation should be administered to optimize preload. Occasionally, recurrent significant effusions may require surgical removal of a section of pericardium (pericardial window) to avoid recurrent tamponade, often seen with malignant effusions and is usually palliative in nature [9].

Clinical Pearls

- CT scan can sometimes overestimate the size of effusion. Always confirm size and hemodynamic significance with echocardiography
- Tamponade is an emergency and requires immediate action
- Never give beta blockers to a patient in tamponade

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