

# 16.8 Pericardial disease

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**ESSENTIALS** The most common clinical presentations of pericardial disease are pericarditis, effusion, tamponade, and constriction. Acute pericarditis The most common proven causes are viral infection or as a complication of myocardial infarction, but a wide range of other conditions including autoimmune rheumatic disorders and tuberculosis need to be considered. No firm cause is established in many cases, which are regarded as 'idiopathic' (presumed viral). The main clinical features are chest pain, the presence of a pericardial rub, and widespread ST segment elevation on the ECG. Idiopathic disease is self-limiting: treatment is with analgesics and/or nonsteroidal anti-inflammatory agents and colchicine. Pericardial effusion Acute rapid collection is usually caused by traumatic injury, iatrogenic ventricular puncture, or aortic dissection. Presentation is with pericardial tamponade. Chronic fluid accumulation is most commonly caused by viral infection, uraemia, autoimmune rheumatic disease, myocardial infarction, myxoedema, or malignancy. Patients may remain asymptomatic despite the presence of a large volume of fluid in the pericardium due to corresponding increase in the capacity of the pericardial cavity. Examination may reveal distant heart sounds and increase in the area of cardiac dullness to percussion. The chest radiograph typically shows a large globular heart and clear lung fields. Echocardiography is the investigation of choice for confirming the presence of effusion and for assessing its volume. Pericardial tamponade Pericardial tamponade is a condition of haemodynamic instability caused by chamber compression, because increased intrapericardial pressure is greater than the filling pressure of the right and left ventricles. Presentation is typically with shortness of breath or circulatory collapse. The key physical findings are tachycardia, pulsus paradoxus (an exaggeration of the normal fall in systolic blood pressure on inspiration) of greater than 10 mm Hg, and elevation of the venous pressure. Echocardiography is the most important investigation, providing clear evidence of fluid collection around the heart and presence of diastolic right ventricular or right atrial collapse. Immediate management is by pericardial aspiration. Pericardial constriction A stiff pericardium loses its stretching ability to accommodate normal changes in intracardiac pressures. Most patients present with leg or abdominal swelling and dyspnoea. The key physical findings are elevated venous pressure (with a characteristic 'M' or 'W' waveform), a pericardial knock, hepatomegaly, ascites, and oedema. Investigation and diagnosis—Doppler echocardiography is the best

noninvasive investigation. Cardiac catheterization demonstrates a difference of less than 5 mm Hg between end-diastolic pressures in the two ventricles, persisting with respiration and fluid loading; a peak right ventricular pressure of less than 50 mm Hg; and a ratio of end-diastolic to peak right ventricular pressure of more than 0.33. Management—fluid retention in early pericardial constriction can be managed by diuretics, with pericardiectomy recommended for patients who are resistant. Anatomy and physiology The pericardium consists of two layers, a visceral layer lined by mesothelial cells and a parietal or fibrous layer also lined by mesothelial cells, but with attached fat and fibrous tissue. The mesothelial layer secretes about 50 ml of clear pericardial fluid that allows both surfaces to slide together during the cardiac cycle. The innermost layer of the visceral pericardium is adherent to the outer myocardial layer, the epicardium. The fibrous layer is usually 1 mm in thickness, and the visceral layer is a transparent membrane on the surface of the heart. The fibrous pericardium attaches the heart to the diaphragm below and the great vessels above. Intrapericardial pressure normally ranges between  $-2$  and

- 2 mm Hg, thus it is less than that of the right heart. It falls with the intrapleural pressure during inspiration, resulting in a fall in right-sided cardiac pressures. This causes a modest increase in right heart filling velocities with inspiration. These effects are often exaggerated in patients with clinically significant pericardial disease. 16.8 Pericardial disease Michael Henein

section 16 Cardiovascular disorders 3502 The most common clinical presentations of pericardial disease are pericarditis, effusion, tamponade, and constriction. Pericarditis Causes Infection The most common causes of acute viral pericarditis are coxsackie B, flu, mumps, hepatitis B, rubella, echovirus 8, and HIV. The typical presentation is with 'flu-like' upper respiratory tract infection along with chest pain that is related to breathing. Sending blood tests for viral titres is not usually conclusive in routine clinical practice. The condition is usually self-limiting. Bacterial infection (other than tuberculous) is a very rare cause of pericarditis, usually caused by staphylococci, pneumococci, or streptococci spreading directly from the lungs or pleura, particularly in patients with impaired immunity. Tuberculous infection is an important cause of bacterial pericardial disease, particularly in developing countries. It may take the form of acute pericarditis, pericardial effusion, or constriction. The primary response is an acute pericarditis due to allergic reaction. Chronic pericardial effusion and constriction both reflect granulomatous disease complicated by fibrosis and calcification. Both parietal and visceral layers of the pericardium may be involved, including the epicardial layer of the myocardium. In sub-Saharan Africa, most patients (>80%) with tuberculous pericarditis will be HIV positive. This needs to be established before treatment: antituberculous chemotherapy is the first line of management for all, but if there is pericardial effusion or constriction steroids are used for the first few weeks to limit the development of adhesions and hence the need for pericardiectomy in those who are HIV-negative. Actinomycosis, coccidioidomycosis, histoplasmosis, and hydatid disease can rarely cause pericarditis in endemic areas. Myocardial infarction This may be complicated by acute pericarditis in 15% of cases, particularly in patients with transmural infarction, when electrocardiography (ECG) demonstrates ST and T-wave changes that are more generalized than the segmental distribution of the infarct. A friction rub may be heard and a small effusion may be seen on transthoracic echocardiographic examination. A delayed response 3 to 4 weeks after an acute infarct may present as Dressler's syndrome, with fever and pericardial rub. Although this condition is self-limiting it may respond to nonsteroidal anti-inflammatory medications (NSAIDs) and (if

needed) steroids. Autoimmune diseases Pericardial involvement can be a serious manifestation of rheumatoid disease, systemic lupus erythematosus, systemic sclerosis, and Churg–Strauss syndrome. Presentation can be with pericardial pain, effusion, or even constriction. A small pericardial effusion is seen in most cases of rheumatic fever, but this hardly ever develops into a significant problem. If adhesions develop they may later mature in the form of constriction, a pathology which can be confirmed at the time of valve surgery. Other medical conditions Inadequately treated chronic renal failure may be complicated by pericarditis and pericardial effusion. Pericardial tamponade may develop if the effusion remains untreated. Hypothyroidism may be complicated by pericarditis, usually accompanied by a small fluid collection that is unlikely to require drainage. Irradiation Irradiation can cause pericarditis soon after treatment, with typical ECG presentation, fluid collection, or even constriction. Late presentation can be seen years after irradiation, when the pericardium is thickened and fibrosed. Clinical features and management There are three main features of the clinical syndrome of acute pericarditis—chest pain, pericardial rub, and ECG changes. The chest pain occurs at rest and varies with posture and respiration. It is typically sudden in onset (although often preceded by the nonspecific symptoms of a viral illness), retrosternal, continuous, sharp, or ‘raw’ in character, worse on inspiration, radiating to the trapezius ridge, and relieved by sitting up. It needs to be distinguished from ischaemic cardiac pain (particularly in the context of recent myocardial infarction), oesophageal pain, and musculoskeletal pain. On examination the main feature is the presence of a pericardial rub. This scratching or creaking sound, variably described as being like ‘walking on fresh snow’ or the ‘creaking of new leather’, is usually loudest at the left sternal border, but may be heard anywhere in the chest. It often changes with posture, may be louder with inspiration, and can be fleeting, recurring in hours. In isolated pericarditis, other elements of the cardiovascular examination are usually normal unless pericarditis is associated with the presence of significant pericardial fluid to cause tamponade, or with pericardial constriction. The typical ECG change is generalized ST elevation, usually concave upwards, by 1 mm or more. The extent of ST change (unlike that of myocardial infarction) does not usually conform to a single coronary artery territory. Similar ECG features may be seen in individuals with early repolarization and in cases without a typical presentation it is important to repeat the ECG in the convalescent phase to ensure that the changes are not fixed to avoid over diagnosis of the condition. Depression of the PR segment is a subtle but characteristic feature. Nonspecific T-wave changes may follow after the acute episode has resolved. ST changes usually resolve, but T-wave changes may persist for years afterwards in some cases. A minor troponin rise reflecting myocardial involvement is not uncommon. Inflammatory markers—C-reactive protein and erythrocyte sedimentation rate (ESR)—are also commonly raised. The chest radiograph is not usually helpful in diagnosis: it may show cardiac enlargement, but may be completely normal. Echocardiography is the best technique to show any fluid collection around the heart and to assess its physiological significance. Similar findings can be shown by cardiac MRI, if available. Although the underlying cause of pericarditis should always be sought, the final diagnosis is often ‘idiopathic’ or ‘presumed viral’. Idiopathic pericarditis is self-limiting and needs only analgesics. Small effusions due to other causes rarely need drainage, but with symptoms the patient may benefit from NSAIDs. Colchicine is traditionally used as second line therapy in patients with recurrent or

16.8 Pericardial disease 3503 persistent pericarditis. However recent evidence suggests that the use of colchicine as a first-line agent (at a dose of 0.5 mg twice daily for 3 months for patients weighing >70 kg or 0.5 mg once daily for patients weighing ≤70 kg) may also reduce the rate of

symptom persistence and recurrence. Pericardial effusion The diagnosis of pericardial effusion is only made when the volume of the fluid in the pericardial space is more than the physiological amount of 50 ml. Two-dimensional echocardiography can detect 100 ml fluid in the pericardial space. Pericardial effusion can be secondary to cardiac or noncardiac causes. Acute rapid collection is usually caused by traumatic injury, iatrogenic ventricular puncture, or aortic dissection. The common causes of chronic fluid accumulation are viral infection, uraemia, autoimmune rheumatic disease, myocardial infarction, myxoedema, and malignancy. Conditions associated with generalized salt and water retention such as congestive cardiac failure, renal failure, and hepatic cirrhosis may also be complicated by pericardial effusion. A small, rapidly accumulated effusion may result in raised pericardial pressure and development of symptoms (see 'Pericardial tamponade'), whereas with a slowly accumulating effusion patients may remain asymptomatic despite the presence of a large volume of fluid because of the corresponding increase in the capacity of the pericardial cavity. Symptoms in uncomplicated pericardial effusion are nonspecific—reduced exercise tolerance or dull aching chest pain—but patients may develop symptoms of mediastinal syndrome: cough caused by bronchial compression, dyspnoea due to lung compression, or hoarseness of voice caused by recurrent laryngeal nerve compression. On examination distant heart sounds and increase in the area of cardiac dullness to percussion may be the only physical signs until tamponade develops. The chest radiograph does not always confirm the presence of pericardial effusion if it is less than 250 ml, but when the effusion is large there is an increased cardiothoracic ratio and the cardiac shadow is globally enlarged (Fig. 16.8.1). The ECG may show low-voltage QRS complexes, and electrical alternans may be present if the effusion is large, with the heart swinging to and fro within it (Fig. 16.8.2). Echocardiography is the investigation of choice for confirming the presence of pericardial effusion and for assessing its volume (Fig. 16.8.3). An echo-free space in the pericardium, on both M-mode and two-dimensional images, should be distinguished from anterior pericardial fat pad. Quantitation of pericardial effusion is quite reliable from two-dimensional echocardiographic images: a 1-cm global collection around the heart suggests an approximate amount of 200 ml. With localized effusion, a comparative assessment of the effusion size with that of the left ventricle gives a rough estimation of the collection volume. The haemodynamic effects of pericardial effusion depend on the pressure-volume relation of the pericardium, the speed of fluid collection and the volume of the effusion. Changes in ventricular compliance may also influence the haemodynamic effects of pericardial effusion in patients with ventricular disease.

**Pericardial tamponade** Pericardial tamponade is a condition of haemodynamic instability caused by chamber compression because increased intrapericardial pressure is greater than the filling pressure of the right and left ventricles.

**Pathophysiology** Provided the pericardium can stretch slowly, more than 2000 ml of fluid can be accumulated without a significant increase in pressure, but rapid accumulation of as little as 200 ml increases pericardial pressure. Inability of the pericardium to distend acutely causes its pressure to rise above right atrial pressure, followed by

Fig. 16.8.1 Posteroanterior chest radiograph of a patient with a large pericardial effusion. The heart shadow is greatly enlarged and globular in shape. The parts of the lung fields that can be seen are normal.

Fig. 16.8.2 ECG from a patient with a massive malignant pericardial effusion. All QRS complexes are sinus beats with a constant PR interval, but the QRS axis alternates, hence the term 'electrical alternans'.

section 16 Cardiovascular disorders 3504 right ventricular pressure, and eventually results in right ventricular collapse. Intrapericardial and intrapleural pressures normally fall equally during inspiration, but with tamponade intrapericardial pressure does not fall as much, resulting in a

reduced pressure gradient between intrathoracic pressure/pulmonary veins and left atrium/left ventricle. This results in reduced left-sided filling velocities during inspiration and hence stroke volume. On the right side of the heart the normal increase in right ventricular dimensions during inspiration enhances right-sided filling and ejection. Progressive increase in pericardial pressure and right ventricular pressure may affect the left heart further, adding to the compromise of its filling during inspiration and exacerbating reduction in stroke volume. The combined effect of these two mechanisms eventually compromises cardiac output. Pericardial pressure greater than 10 mm Hg results in right ventricular collapse and raised diastolic pressures of both ventricles as well as increased capillary wedge pressure. This leads to inspiratory fall of aortic pressure and hence hypotension with pulsus paradoxus (see 'Clinical features'). Left ventricular and left atrial collapse are much less commonly seen with tamponade. Causes The most common cause of tamponade is malignant effusion or acute fluid collection after cardiac surgery. Intrapericardial clot formation after cardiac surgery or as a complication of an interventional procedure (e.g. transseptal puncture) may result in signs of tamponade due to the rapid increase in intrapericardial pressure, even in the absence of a significantly large fluid volume. Left ventricular invagination caused by localized collection around the free wall has been reported after open heart surgery. Significant localized posterior effusion is usually caused by anterior adhesions between the right ventricle, the right atrium, and pericardium. Clinical features Patients with cardiac tamponade present with shortness of breath or circulatory collapse. The key physical findings to make the diagnosis are tachycardia, pulsus paradoxus of more than 10 mm Hg, and elevation of the venous pressure. Tachycardia (>100 beat/min) is almost invariable, but clearly not specific for tamponade. Pulsus paradoxus describes an exaggeration of the normal fall in systolic blood pressure (up to 10–12 mm Hg) on inspiration. With the patient breathing normally, the best way to detect this sign is to stop deflation of the blood pressure cuff as soon as the first Korotkoff sound is heard, in which case in the presence of pulsus paradoxus the sound will disappear on every inhalation and reappear on every exhalation. After noting the systolic pressure reading, the cuff is then gradually deflated until the Korotkoff sound is heard throughout the respiratory cycle, at which point the pressure is again noted—the difference between the two readings is the measurement of the amount of paradox. A pulsus paradoxus greater than 10 mm Hg is found in 98% of patients with tamponade, greater than 20 mm Hg in 78%, greater than 30 mm Hg in 49%, greater than 40 mm Hg in 38%, and total (pulse not palpable on inspiration) in 23%. The most common reason for pulsus paradoxus to be absent in tamponade is compromised stroke volume. However, although pulsus paradoxus is a sensitive sign of tamponade, it must be noted that it is not specific: it can be seen not infrequently in severe asthma, also (uncommonly) in constrictive pericarditis, right ventricular infarction, and pulmonary embolism. The venous pressure is always high in cardiac tamponade: if it is not, then the diagnosis is wrong. Usually it is very high, which can make it difficult to see the top. The venous pressure normally falls on inspiration because right heart pressures drop as intrathoracic pressure decreases. Kussmaul's sign is an increase in venous pressure during inspiration, which can be observed (infrequently if at all in most series) in tamponade because of the inability of the right atrium and ventricle to accommodate greater influx of blood. Abnormalities of the venous wave form are not helpful in making the diagnosis of tamponade. In addition to tachycardia, pulsus paradoxus, and elevated venous pressure, patients with tamponade will usually have tachypnoea and cool peripheries, and they may have a pericardial rub. Differential diagnosis The main requirement is for the doctor to consider the diagnosis, even if only briefly, when confronted with any patient in unexplained circulatory shock. Tamponade must be distinguished from the common causes of such a presentation, namely hypovolaemia,

overwhelming sepsis, severe ventricular disease (e.g. acute myocardial infarction), and pulmonary embolism. If the patient is shocked with a high venous pressure, then particular consideration needs to be given to pulmonary embolism, right ventricular infarction, and (less commonly) pericardial constriction. Investigations The chest radiograph typically shows a large globular heart, which unlike congestive heart failure, is not associated with pulmonary venous congestion (Fig. 16.8.1): if pulmonary oedema is present, it suggests additional myocardial disease. The ECG shows tachycardia, often with low-voltage QRS complexes, and may reveal electrical alternans (Fig. 16.8.2). Echocardiography is the most important investigation. It provides clear evidence for fluid collection around the heart, which is usually large with tamponade (Fig. 16.8.3), and is likely to show evidence for diastolic right ventricular or right atrial collapse. Right ventricular collapse is a sensitive (92%) and highly specific (100%) PE LV LA Fig. 16.8.3 Parasternal long axis echocardiographic view from a patient with a large pericardial effusion (PE), located mostly posteriorly to the left ventricle (LV). LA, left atrium.

16.8 Pericardial disease 3505 diagnostic sign for tamponade, reflecting transient negative transmural early diastolic pressure as pericardial pressure exceeds right ventricular pressure. Right atrial collapse is less sensitive (82%) but equally specific (100%). In the absence of a haemodynamically significant pericardial effusion, right ventricular diastolic collapse may be caused by bilateral large pleural effusions (Fig. 16.8.4). Right ventricular collapse may be delayed by myocardial hypertrophy, pulmonary hypertension, or free wall adhesions, commonly associated with malignant effusions. Swinging of the heart inside the pericardial fluid may be seen. Doppler recordings of right and left cardiac filling and ejection show inspiratory dominance in the right with reciprocal changes in the left (Fig. 16.8.5). Finally, echocardiography can exclude the presence of large pleural effusion as a potential cause of the clinical and physiological disturbances: it is essential for the echocardiographer to identify the high-intensity echo of the fibrous pericardium posterior to the left ventricle on the left parasternal image—a pericardial effusion is inside this layer and a pleural effusion outside it. Management Pericardial tamponade is a medical emergency, particularly when there is clear evidence for arterial paradox, or if the effusion is collecting rapidly. Pericardial aspiration should be performed in an area where resuscitation facilities are available. Echocardiography is used to determine where to insert the needle and to estimate the depth and direction of advancement. The subcostal route is the most popular because it avoids possible injury to the coronary artery (left anterior descending). After administration of a local anaesthetic a larger needle or polythene cannula is introduced into the effusion, followed by a pigtail catheter inserted over a guide wire. An injection of agitated saline through the drain can help to confirm that it is in the pericardial space. A maximum of 500 ml of fluid is removed initially to relieve haemodynamic instability: rapid withdrawal of a larger volume can provoke cardiovascular collapse. Continuous drainage is then commenced and the rest of the effusion drained over the next few hours. Many pericardial effusions are heavily bloodstained, particularly those that are malignant. The aspirated fluid can be distinguished from blood (usually to the great relief of the doctor performing the procedure) by its colour (dark because very desaturated) and failure to clot (because defibrinated). Fluid should be sent for culture and cytological analysis. Biochemical analysis (glucose and protein) can sometimes be useful but is diagnostically less reliable than for pleural effusions. Surgical creation of a pericardial window (usually with video-assisted thoracoscopy) is recommended for recurrent or rapidly accumulating effusions and permits therapeutic pressure relief, fluid drainage, and pericardial biopsy (for culture and histological examination). Constrictive pericarditis Pericardial constriction is a pathological

condition characterized by pericardial thickening and fibrosis that results in adhesion of its two layers. Chronic constrictive pericarditis frequently proves to be 'idiopathic'; a (presumed) viral aetiology is frequently invoked when no other cause is found. Tuberculosis is currently an uncommon cause, particularly in developed countries. Other causes include radiation, autoimmune rheumatic disease, chronic renal failure, neoplastic disease, and previous cardiac surgery. Pathophysiology The stiff pericardium loses its stretching ability to accommodate normal changes in intracardiac pressures. This is demonstrated by equalization of a raised end-diastolic pressures in the right and left ventricles, with the dip-plateau pattern a cardinal sign for diagnosing pericardial constriction (see Fig. 16.3.4.1). Pericardial constriction also leads to characteristic abnormalities in the venous pressure waveform, with prominent 'x' and 'y' descents. The 'x' descent, which occurs after atrial contraction ('a' wave), is caused by two processes: (1) right atrial relaxation (followed by a positive 'c' wave that is not visible on inspection), and (2) the atrioventricular tricuspid valve ring moving downwards during LA Pleural Fig. 16.8.4 Parasternal long axis view showing pleural effusion posterior to the left ventricle. Expiration Inspiration I E Fig. 16.8.5 Transmitral pulsed Doppler velocities from a patient with large pericardial effusion and tamponade demonstrating significant fall in left ventricular filling velocities with inspiration (arrow, I) compared to expiration (arrow, E).

section 16 Cardiovascular disorders 3506 systole, increasing the volume of the right atrium. A fibrosed and unstretchable pericardium, being adherent to the epicardial layer of the myocardium, can limit its normal movement during the cardiac cycle along the ventricular transverse axis, particularly in systole. It cannot, however, affect shortening and lengthening of the longitudinal myocardial fibres that are located in the subendocardium, hence the downward displacement of the tricuspid ring and valve in systole is preserved, allowing a column of blood to enter the atrium rapidly, thereby producing a characteristic exaggerated 'x' descent (Fig. 16.8.6). Following the 'x' descent the 'v' wave represents right atrial filling, with the 'y' descent beginning the moment that the tricuspid valve opens at the beginning of diastole and allows blood to enter the ventricle. In pericardial constriction the 'y' descent is prominent because, from a high venous pressure, diastolic filling is not impaired at the beginning of diastole, only when the relaxing ventricle meets the rigid pericardium. Similar features can be seen in the left heart physiology. It should be noted that simply the presence of a thickened pericardium on imaging techniques (echo or MRI) is not a sufficient diagnostic criterion for constrictive physiology. Furthermore, in rare cases of rapidly increasing ventricular volumes, as in dilated cardiomyopathy, the pericardium may be completely normal and yet demonstrate an external constricting effect, thus adding to the deterioration of the clinical condition. Clinical features Most patients with constrictive pericarditis present with leg or abdominal swelling and dyspnoea. Rarely the patient can present with jaundice, or with features of nephrotic syndrome or protein-losing enteropathy. The key physical findings are elevated venous pressure, a pericardial 'knock', hepatomegaly, ascites, and oedema. As with pericardial tamponade, the venous pressure is always high; if it is not, then the diagnosis is almost certainly wrong. However, unlike with pericardial tamponade, the form of the venous waveform is characteristic, with exaggerated 'x' followed by 'y' descent (for reasons explained earlier) that create two conspicuous dips per cardiac cycle, making the waveform appear to follow an 'M' or 'W' pattern with each arterial pulse (Fig. 16.8.6). Kussmaul's sign (an increase in venous pressure during inspiration) is seen in 50% of cases. Sometimes the changes in venous pressure are transmitted to the liver, which then pulses twice with each cardiac cycle. A pericardial knock is a loud, high-frequency sound, typically best heard between the left lower sternal border and the

apex. It is caused by sudden cessation of ventricular filling as it meets the constriction and is reported in about 50% of cases in most series. Other common findings are atrial fibrillation, a mild degree of pulsus paradoxus ( $\leq 20$  mm Hg), and systolic retraction of the apical impulse. A pericardial rub can be heard in some cases.

**Differential diagnosis** The main differential diagnosis of constrictive pericarditis is restrictive myocardial disease (see Chapters 16.7.2 and 16.7.3). Distinction between these can be difficult. In restriction, ventricular filling becomes limited to early diastole, with high acceleration and deceleration frequently associated with right-sided third heart sound. Respiratory variation of ventricular filling and ejection velocities may be present in constriction but is absent in restrictive right ventricular disease. Imaging can help to determine if the main abnormality is likely to be pericardial or myocardial. Pericardial constriction also needs to be distinguished from other causes of raised venous pressure, including obstruction of the superior vena cava, tricuspid stenosis, or tricuspid regurgitation.

**Investigations** The chest radiograph is usually normal but may show pericardial calcification, either as multiple plaques or as a rim covering the diaphragmatic and anterior surfaces of the heart. The ECG is not diagnostic, but can show low-voltage QRS complexes and nonspecific T-wave changes. On CT or MRI, the pericardium may appear thickened, but this is an insensitive marker for constriction. Doppler echocardiography is the best noninvasive investigation to demonstrate the systolic descent in the jugular venous pressure and systolic filling of the right atrium from the superior and inferior vena cavae during ventricular systole. Ventricular filling is nonspecific, depending on additional myocardial disease. In constrictive pericarditis there is less intracardiac than extracardiac respiratory variation, particularly on the right side, when compared to that seen with pericardial tamponade. A raised right atrial pressure during inspiration (Kussmaul's sign) and dilated inferior vena cava are nonspecific signs of constriction. Spontaneous contrast in the inferior vena cava, resulting from the limited venous return, may also be an additional finding in favour of constrictive pericarditis. Cardiac catheterization demonstrates the following diagnostic features for constriction:

- a difference of less than 5 mm Hg between end-diastolic pressures in the two ventricles, persisting with respiration and fluid loading;

Fig. 16.8.6 Systemic venous pulse from a patient with constrictive pericarditis before (top) and after (bottom) pericardiectomy. Note the 'M' or 'W' pattern, with deep 'x' descent that disappears after pericardiectomy.

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- a peak right ventricular pressure of less than 50 mm Hg;
- a ratio of end-diastolic to peak right ventricular pressure of more than 0.33.

**Management** Fluid retention in early pericardial constriction can be managed by diuretics, with pericardiectomy recommended for patients who are resistant. After surgical removal of the pericardium, which is often very difficult, the venous pressure drops and the 'x' descent disappears from the jugular venous pressure (Fig. 16.8.6). This is not always instantaneous and may take up to a few days or even weeks to settle.

**Pericardial complications after open**

**heart surgery** Apart from the commonly seen pericardial collection, other significant complications may occur that have a major impact on clinical management.

**Pericardial clot** A collection of clots in the pericardial space, with or without pericardial effusion, is often associated with delayed postoperative clinical recovery. It may have an important physiological effect on overall cardiac function, irrespective of the amount present. The clinical presentation is typically with cooling of the peripheries, hypotension, and fall in urine output over minutes to hours, and the condition should be suspected particularly in any patient who has bled rather heavily at operation, especially if the blood flow from the chest drains suddenly falls. There are no specific abnormalities on the chest radiograph or ECG; transthoracic echocardiography rarely gives good images immediately

postoperatively; transoesophageal echocardiography may show clotting alongside the heart. Reopening the chest to remove the clot surgically is the best management, with early detection and removal securing complete recovery. 'Tight' pericardium In the absence of postoperative pericardial effusion, intrapericardial pressure may be raised to the extent that it affects right-sided physiology so that superior vena caval flow occurs only during inspiration. This condition mimics left ventricular disease and may lead to inappropriate administration of inotropic agents. The jugular venous pressure is raised, and right-sided filling and ejection is predominantly inspiratory. On two-dimensional echocardiographic images there is no evidence for right atrial or ventricular collapse. Although these signs usually resolve with time, delayed sternal closure has proved beneficial when the clinical manifestations are severe. The condition tends to settle within days or weeks after surgery, with complete normalization of venous pressure. Restrictive pericarditis This is a rare clinical presentation that has been documented after open heart surgery, presenting with resistant fluid retention and raised venous pressure. Two-dimensional echocardiographic images may not show any specific abnormality, although MRI may demonstrate a thickened pericardium. The underlying pathology seems to be chronic combined pericardial and epicardial inflammation that results in massive fibrosis and adhesions between the two layers, with myocardial involvement. Patients with this condition are usually resistant to medical therapy, demonstrating signs of restrictive physiology on both sides of the heart with a dominant early diastolic filling component and short deceleration time. Cases resistant to medical therapy may respond to surgical decortication of the pericardium. Pericardial tumours The commonest tumours of the pericardium are secondaries from elsewhere, most frequently carcinoma of the breast and lung, malignant melanoma, lymphoma, and leukaemia. They invade the pericardium either directly, or via lymphatics, or by haematogenous dissemination. Primary tumours are rare but include malignant mesothelioma and sarcomas. Whereas carcinomas metastasize in the pericardium in the form of localized masses, lymphomas, and leukaemia present in the form of uniform pericardial infiltration and thickening, which may cause tumour incarceration of the heart and hence the clinical syndrome of 'constrictive physiology'. A mild degree of pericardial thickening can easily be missed on echocardiography, but the pattern of ventricular wall motion is characteristic. MRI or CT may be better at showing pericardial thickening. Recurrent pericardial effusion of unknown aetiology should always suggest malignancy until otherwise proved, as should pericardial effusion in the presence of an intracardiac mass. Congenital pericardial disease Congenital anomalies of the pericardium are rare. Pleuropericardial defect, either complete or partial (80% of cases), is the most common form. The left side is most commonly involved in the partial form, allowing the left atrial appendage or part of the left ventricle (if the defect is large) to herniate through the defects. The chest radiograph is characteristic, demonstrating a shift of the heart to the left and prominent main pulmonary artery. Defects in the diaphragmatic portion of the pericardium are extremely rare. In most instances pericardial defects are asymptomatic, but about one-third of cases are associated with congenital abnormalities of the heart and lungs. Pericardial cysts are very rare, difficult to diagnose, and if present do not cause any clinical problem. Their presence can only be confirmed when found surgically and removed. Pericardial constriction due to fibrosis of unknown cause may contribute to the clinical picture of Mulibrey (muscle, liver, brain, eye) nanism (OMIM 253250), which is an autosomal recessive condition characterized by growth failure, a triangular face (often with a hydrocephalus), hypotonia, a peculiar voice, large liver, and yellowish dots and pigment dispersion in the optic fundi. FURTHER READING Athappan G, Sorajja P (2017). Invasive hemodynamics of pericardial disease. *Interv Cardiol Clin*, 6, 309-17. Callahan JA, et al. (1985). Two-dimensional echocardiographically guided pericardiocentesis: experience in 117

consecutive patients. *Am J Cardiol*, 55, 476–9.

section 16 Cardiovascular disorders 3508 Cameron J, et al. (1987). The etiologic spectrum of constrictive peri- carditis. *Am Heart J*, 113, 354–60. Chang SA (2017). Tuberculous and infectious pericarditis. *Cardiol Clin*, 35, 615–22. Guberman BA, et al. (1981). Cardiac tamponade in medical patients. *Circulation*, 64, 633–40. Henein MY, et al. (1999). Restrictive pericarditis. *Heart*, 82, 389–92. Henein MY (2012). *Clinical echocardiography*, 2nd edition. Springer, London. Imazio M, et al. (2013). A randomized trial of colchicine for acute peri- carditis. *N Engl J Med*, 369, 1522–8. Imazio M, Gribaudo E, Gaita F (2017). Recurrent pericarditis. *Prog Cardiovasc Dis*, 59, 360–8. McGee SR (2001). *Evidence-based physical diagnosis*. W. B. Saunders, Philadelphia, PA. Miranda WR, Oh JK (2017). Effusive-constrictive pericarditis. *Cardiol Clin*, 35, 551–8. Murashita T, et al. (2017). Experience with pericardiectomy for constrictive pericarditis over eight decades. *Ann Thorac Surg*, 104, 742–50. Reddy PS, et al. (1978). Cardiac tamponade: hemodynamic obser- vations in man. *Circulation*, 58, 265–72. Sagrista-Sauleda J, et al. (2004). Effusive-constrictive pericarditis. *N Engl J Med*, 350, 469–75. Shabetai R, et al. (1965). Pulsus paradoxus. *J Clin Invest*, 44, 1882–98. Shabetai R, Oh JK (2017). Pericardial effusion and compressive disorders of the heart: influence of new technology on unrav- eling its pathophysiology and hemodynamics. *Cardiol Clin*, 35, 467–79. Welch TD, Oh JK (2017). Constrictive pericarditis. *Cardiol Clin*, 35, 539–49. Xu B, Kwon DH, Klein AL (2017). Imaging of the pericardium: a multimodality cardiovascular imaging update. *Cardiol Clin*, 35, 491–503.

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