## Pericardial Diseases

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#### ANATOMY AND PHYSIOLOGY

- The pericardium is comprised of two principal layers, the visceral pericardium and the parietal pericardium.
- The visceral pericardium, or epicardium, is a single serous layer covering the surface of the heart and proximal great vessels.



#### The parietal pericardium

- The innermost layer is a serous layer, continuous with the serous visceral pericardium.
- The space between the visceral serous and parietal serous layers is the pericardial space, and contains a small amount of serous fluid for lubrication (<20 to 30 cc in adults, less in children).</p>
- ► The middle layer of the parietal pericardium is fibrous
- ▶ the outer layer is epicardial collagenous connective tissue.
- The pericardium receives arterial blood supply from the descending aorta and internal mammary artery, and is innervated from the phrenic and vagus nerves.







- The pericardium provides mechanical protection to the heart from the spread of neoplastic, infectious, and inflammatory diseases from adjacent structures.
- The presence of a small amount of fluid in the pericardial space allows free movement of the heart throughout the cardiac cycle.
- The pericardium limits acute distension of the heart, and therefore limits enddiastolic volume. It permits diastolic coupling of the two ventricles, whereby filling pressure abnormalities of one ventricle affect the other.
- Slow progressive accumulation of fluid within the pericardium is tolerated by stretching and growth of the parietal pericardium, however, rapid accumulation of even a small amount of fluid is poorly tolerated.

#### Pressure volume curve



#### **ACUTE PERICARDITIS**



#### **Symptoms**

- precordial or substernal chest pain. The pain is described as squeezing, sharp or dull, and characteristically is worse in the supine position.
- The patient will prefer to sit upright leaning forward, and may refuse to lie down to be examined.
- ▶ The pain worsens with inspiration, coughing, and movement.
- > Younger children may present with atypical symptoms.
- Respiratory distress is uncommon unless tamponade or pulmonary disease is present.
- Rarely, abdominal pain can result from hepatic distension in patients with quickly accumulating effusions.
- Fever may be present.



#### **Physical Examination**

- The pathognomonic physical examination finding with acute pericarditis is a friction rub.
- This is a high-frequency, scratching or sandpaper-like sound caused by friction between the inflamed pericardial surfaces. The rub can be heard throughout the cardiac cycle; however, it may be intermittent.
- It classically has been described as having three components: (1) systolic, (2) early diastolic, and (3) late diastolic.
- These components correspond to ventricular contraction, ventricular relaxation, and atrial contraction.
- Absence of a friction rub does not exclude pericarditis, particularly with large effusions or tamponade when the heart sounds may be muffled



#### **Cardiac Tamponade**

- ► Tamponade is characterized by **Beck's triad**:
- ▶ (1) distant heart sounds,
- ▶ (2) hypotension, and
- ▶ (3) elevated central venous pressure with jugular venous distension.
- Patients will have tachycardia, tachypnea, and a narrow pulse pressure with pulsus paradoxus.

#### Pulsus paradoxus

- decrease in systolic blood pressure of greater than 10 mm Hg during inspiration. Normally during inspiration, systolic blood pressure decreases by 4 to 6 mm Hg due to decreased intrathoracic pressure and increased capacity of the pulmonary venous bed. With cardiac tamponade, the left ventricular diastolic volume is restricted by increased pericardial pressure, decreased pulmonary venous return, and shifting of the interventricular septum.
- To determine the clinical presence of pulsus paradoxus, the patient should be supine. A blood pressure cuff is inflated until the radial pulse is not palpable. With slow release of pressure, one should listen for the initial Korotkoff sounds. With inspiration, the Korotkoff sounds disappear, particularly in the presence of pulsus paradoxus. Cuff pressure should be slowly released until the Korotkoff sounds are heard throughout the respiratory cycle.



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ORIGINAL ARTICLE

Detection of Pulsus Paradoxus by Pulse Oximetry in Pediatric Patients After Cardiac Surgery

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#### Kussmaul sign

During normal inspiration, intrathoracic pressure decreases with an increase in venous return to the right atrium. In patients with cardiac tamponade, however, expansion of the right atrium is limited by pericardial pressure. Thus, during inspiration there may be a paradoxical increase in central venous pressure.

#### Chest Radiography

- The absence of cardiomegaly by chest radiography does not exclude pericarditis or pericardial effusion.
- Tamponade can be present with a normal-appearing cardiac silhouette. With a progressively increasing effusion, the cardiac silhouette may assume a triangular or "waterbottle" shape, with normal pulmonary vascular markings.

Patients with chronic pericarditis may have calcification of the pericardium





#### Electrocardiography

- Acute pericarditis is the most frequent pathologic cause of ST segment elevation in children.
- Low QRS voltages occur in all leads in the presence of large pericardial effusions or chronic pericarditis.
- Electrical alternans, a cyclical variation of the QRS amplitude, may occur secondary to the pendular motion of the heart within a large pericardial effusion.

# stages of electrocardiographic changes

- Stage 1 consists of ST segment elevation in the lateral/inferior leads (I, II, aVF, aVL, V4–V6)
- Reciprocal ST segment depression may be present in leads aVR and V1. PR interval depression can occur if atrial tissue is inflamed.
- **stage 2**, the ST segment normalizes, and T-wave amplitude diminishes.
- stage 3, the ST segment remains normal, but the T waves become inverted in the lateral/inferior leads (aVF, aVL, V4–V6).
- Stage 4 is characterized by the relative normalization of the ECG, although some T-wave changes may persist.



#### Echocardiography

- Echocardiography is the primary imaging modality for the diagnosis of pericardial effusion.
- Fibrinous strands, thrombi, adhesions, or metastases may be noted in the effusions.
- With the patient in the supine position, a small effusion most commonly is seen posteriorly, and may be detectable only in systole.
- ► A tiny effusion in systole that is not evident in diastole is normal.
- An effusion that is present during both systole and diastole is abnormal.
- As the volume of the effusion increases, fluid may be detected both anterior and posterior to the hear.
- With large effusions, the heart may swing to-and-fro within the pericardial space

#### Echo ...

- The earliest sign of hemodynamic impairment in cardiac tamponade is collapse of the right ventricular free wall in early to mid-diastole
- ▶ The right atrial free wall may appear indented later in diastole.
- Inferior vena cava dilation without normal inspiratory variation and abnormal ventricular septal motion also may occur. Thrombus in the pericardial space suggests the presence of a hemopericardium







- During inspiration in tamponade, the intrathoracic pressure declines to a greater degree than the intrapericardial pressure. Thus, the gradient between the pulmonary capillary wedge pressure and left ventricular diastolic pressures decreases with inspiration resulting in an exaggerated decrease in the mitral inflow velocity (E velocity) and velocity-time integral by at least 30%, with a relatively increased atrial component during inspiration (A velocity).
- Conversely, there is an exaggerated increase in tricuspid inflow velocity (tricuspid E velocity) and the velocity-time integral by at least 70% during inspiration.
- The aortic and pulmonary outflow changes mirror those of their respective atrioventricular valves







#### **Cardiac** Catheterization

- With large accumulation of pericardial fluid, diastolic pressures rise in all four chambers, and ultimately equalize.
- Right ventricular and pulmonary artery pressures may be elevated. Pulsus paradoxus can be seen on a femoral artery pressure tracing. In patients with constrictive physiology, due to the equalization of the left ventricular and right ventricular end-diastolic pressures, the characteristic "square root" sign may be present on the left ventricular pressure tracina



500 ms

## **Other Imaging Modalities**

- MRI or CT can be useful if constrictive pericarditis is suspected. Active pericardial inflammation (a treatable cause of constriction) and hemodynamic evidence of constriction may be apparent using MRI
- MRI is helpful in characterizing pericardial masses and congenital anomalies of the pericardium, such as absence of the pericardium and pericardial cysts.
- FDG-PET/CT has been reported to help identify hypermetabolic tissue associated with malignancy or inflammation and even assess response to antiinflammatory medication, though this modality is not utilized commonly in clinical practice.



## Indications for pericardiocentesis

- Iow cardiac output
- ► Hypotension
- Pulsus paradoxus >10 mm Hg
- suspected bacterial pericarditis, pericardial effusions
- in immunocompromised hosts, or for diagnostic purposes when the etiology of the effusion is unclear.

#### Pericardiocentesis



▶ 30-degree head-up position and adequately sedated.

Subxiphoid

- Continuous monitoring of heart rate, blood pressure, and pulse oximetry should be performed. In an emergent situation, the needle is introduced subxiphoid, and is advanced toward the left shoulder. In nonemergent situations, echocardiographic guidance allows accuracy in entering the pericardial space and reduces complications.
- One can perform an agitated saline injection to confirm the location of the needle in the pericardial space.
- Potential complications of pericardiocentesis can include hemopericardium, pneumothorax, arrhythmias, myocardial puncture, coronary artery, aorta, or internal mammary artery injury.

#### Pericardiotomy

Skin entry -Abdominal wall (One cm below Xyphoid process)

**Parietal peritoneum** 

Diaphrgmatic peritoneum

Anterior aspect of the diaphragm ?

Para-cardiac fat

Sternopericardial /Pericardio-diaphragmatic ligaments

**Parietal pericardium** 







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Image used from Daniel A. Jones & Ajay K. Jain, Journal of thoracic Oncology, 2011

#### Management of Tamponade

- after diagnosing cardiac tamponade, one should administer IV fluid to increase diastolic filling pressure temporarily in order to stabilize the patient.
- Medications that decrease systemic arterial blood pressure such as vasodilators and diuretics should be avoided.

## Pericardial fluid should be analyzed for

- cell content,
- Glucose concentration,
- protein concentration,
- Gram stain, acid-fast bacilli stain,
- cultures (bacterial, viral, and fungal), and microscopic analysis.
- Specific bacterial- or viral-specific antigens may be assessed using polymerase chain reaction (PCR) studies and latex agglutination studies.
- ▶ High triglyceride levels are diagnostic of chylopericardium.
- Adenosine deaminase activity levels can be measured to assist in the diagnosis of tuberculous pericarditis. Increased levels of adenosine deaminase (>40 U/L) accurately diagnose tuberculous pleural effusions



- In viral pericarditis, the fluid usually is serous or serosanguinous
- Iymphocyte predominance, although neutrophils may be common in the early stages of the illness.
- Viral cultures can be obtained from the pericardial fluid, nasopharynx, or stool.
- PCR studies often are useful in determining a specific viral cause

## Viral Pericarditis

	TABLE 61.1 Pericarditis: Potential Viral Causes			
Enterovirus (primarily Coxsackie B)			Measles	
Adenovirus			Cytomegalovirus (CMV)	
Influenza virus (A and B)			Respiratory syncytial virus (RSV)	
Rubella			Herpes simplex	
Mumps			Hepatitis B	
Epstein–Barr virus (EBV)			HIV	



- Coxsackievirus is the most common viral cause of pericarditis in children.
- Patients often present 10 to 14 days after an upper respiratory or gastrointestinal infection with precordial chest pain, fever, and a friction rub.



- Specific treatment for viral pericarditis is symptomatic, including bed rest and oral nonsteroidal anti-inflammatory drugs (NSAIDs).
- If NSAIDs are unsuccessful, steroids may be considered once bacterial causes have been excluded.
- The use of colchicine with aspirin as first-line combination therapy decreases the likelihood of recurrence in adults.
- Colchicine has not been well studied in the pediatric population, but has good anecdotal success and is used in many centers.
- Clinical improvement occurs in days to weeks, with complete resolution usually within 6 weeks.
- Relapse occurs in a small subset of patients, typically improving with reinstitution of NSAIDs or steroids. Constrictive pericarditis rarely occurs as a late complication of viral pericarditis

#### **Bacterial Pericarditis**

most frequently in patients under 2 years of age.

Patients present with symptoms of fever, chest pain, dyspnea, friction rub, and muffled heart sounds. They may be very ill and hypotensive on presentation, and display tachycardia out of proportion to the fever. Bacterial pericarditis can result from hematogenous dissemination or direct contact.

The lung is the most common origin of dissemination, particularly when the agent is Staphylococcus aureus, Haemophilus influenzae, and Streptococcus pneumoniae. Septic arthritis, osteomyelitis, meningitis, or soft tissue infection may be sources for hematogenous dissemination.

#### TABLE 61.2 Pericarditis: Potential Bacterial Causes

Staphylococcus aureus	Chlamydia psittaci	
Haemophilus influenzae	Nocardia asteroides	
Streptococcus pneumoniae	Brucella	
Other Streptococci species	Yersinia	
Neisseria meningitidis	Salmonella	
Neisseria gonorrhoeae	Actinomyces	
Campylobacter fetus	Mycobacterium tuberculosis	
Pseudomonas aeruginosa	Escherichia coli	
Mycoplasma pneumoniae	Listeria monocytogenes	
Mycoplasma hominis	Pasteurella multocida	
Legionella	Klebsiella	
Francisella tularensis	Anaerobes	



- Marked predominance of neutrophils, and cultures typically are positive for the causative organism.
- Latex agglutination studies of the pericardial fluid, serum, or urine may be helpful if antibiotics have been given before obtaining a sample of pericardial fluid.
- Staphylococcus aureus is the most common bacterium isolated, accounting for half of the cases of bacterial pericarditis.
- Also, it is the most common cause of postoperative bacterial pericarditis (i.e., within 3 months of a cardiac procedure).

### Survival

- Survival for patients with bacterial pericarditis is >90%.
- Risk factors for poor outcome include young age at diagnosis, septicemia, tamponade, delayed diagnosis, inadequate drainage, concurrent myocarditis, and a staphylococcal etiology

#### **Tuberculous Pericarditis**

#### occurs frequently in developing nations.

- The typical onset is insidious with symptoms of low-grade fever, night sweats, weight loss, malaise, dyspnea, and chest pain.
- The presentation may be complicated by subacute pericardial tamponade.
- Tuberculous pericarditis often occurs due to direct extension of miliary tuberculosis or lymphatic spread into the pericardium. Hematogenous spread may occur without evidence of pulmonary infiltrates.
- Most patients have a positive Mantoux skin test



- Aspirated pericardial fluid is serosanguineous or hemorrhagic with lymphocyte predominance.
- Acid-fast bacilli may be seen on auramine- rhodamine fluorescent-stained smears. Pericardial biopsy is useful to provide histologic confirmation. Pericardial fluid adenosine deaminase levels are diagnostic for tuberculous pericarditis if >40 U/L.
- Mycobacterium cultures may take up to 6 weeks to grow, and thus treatment should be initiated well before confirmation of diagnosis.



Multidrug therapy is essential due to the risk of drug-resistant tuberculosis. A treatment regime of rifampicin, isoniazid, pyrazinamide, and ethambutol for at least 2 months, followed by isoniazid and rifampicin for another 4 months is highly effective.

#### **HIV and Other Infections**

#### ▶ nearly 25% of children with HIV infection.

- Children with HIV rarely develop tamponade. Pericardial fluid cultures typically are negative.
- Immunocompromised patients, including patients with HIV, are at risk for more rare causes of pericarditis, including parasitic and fungal infections



TABLE 61.3	Pericarditis:	Other In	fectious	Causes
				Caaboo

Fungal	Candida, Aspergillus, Blastomyces, Coccidioides, Histoplasma, Cryptococcus
Parasitic	Entamoeba histolytica, Echinococcus
Protozoal	Toxoplasma gondii
Rickettsial	Typhus, Q fever
Spirochetal	Syphilis, leptospirosis

#### **Renal Failure**

- Pericarditis occurs in 10% of patients with chronic renal failure. It is more common in patients with concurrent systemic lupus erythematosus (SLE).
- It can also be drug induced in those receiving hydralazine. Most pericardial effusions will resolve with efficient dialysis.
- The pericardial fluid typically is serous. Care should be taken with anticoagulation in these patients, as a pericardial hemorrhage can occur with resultant tamponade.
- NSAIDs may relieve chest pain, but do not affect the size of the effusion (and can affect renal function).
- Pericardiocentesis should be performed for bacterial pericarditis and in those with hemodynamic compromise. If the effusion fails to resolve with dialysis, or if constriction develops, pericardiectomy may be necessary

#### Kawasaki Disease

During the acute phase of Kawasaki disease, one-third of patients have a pericardial effusion, which typically resolves within 2 weeks. Effusions related to this inflammatory process rarely progress to tamponade

#### **Drug-Induced Pericarditis**

- Antinuclear antibodies often are elevated in these patients. Effusions due to a lupus-like reaction rarely progress to tamponade.
- Treatment includes discontinuing the offending drugs and initiation of NSAIDs. Patients with hypersensitivity reactions to penicillin and cromolyn sodium may develop pericardial effusions.

TABLE 61.4 Drug-Induced Pericarditis			
Cromolyn sodium	Isoniazid		
Cyclophosphamide	Methysergide		
Cyclosporine	Penicillin		
Dactinomycin	Phenytoin		
Doxorubicin	Procainamide		
Hydralazine			

### Hypothyroidism

- Patients with hypothyroidism and a pericardial effusion usually are asymptomatic.
- Pericardial effusion occurs in most patients with myxedema, but it is rare in patients with mild hypothyroidism.
- Cardiac tamponade is exceedingly uncommon due to the slow accumulation of fluid.
- In hypothyroidism with pericardial effusion, patients present with paradoxical bradycardia, rather than tachycardia. If pericardiocentesis is required, the fluid contains elevated protein and mucopolysaccharides.
- Most effusions resolve gradually after initiation of thyroid hormone replacement therapy.



#### Chylopericardium

- Chylous pericardial effusion may occur in patients following congenital heart surgery, particularly after trauma to the thoracic duct or in patients with elevated central venous pressures.
- Concurrent chylous pleural effusion may be present. Chylopericardium can occur in patients with mediastinal masses that obstruct lymphatic drainage, and in association with cystic hygromas, radiation therapy, or pancreatitis.
- The pericardial fluid is milky-colored, with elevated triglyceride and protein levels. Patients should receive a low-fat or medium-chain triglyceride diet, initiated after a period of parenteral nutrition without oral intake, to allow resolution of the chylous effusion.
- Persistent chylous effusions can be treated by thoracic duct ligation. Some patients may require palliation with a pericardial window, pericardiectomy, or placement of a pericardioperitoneal or pleuroperitoneal shunt.
- Successful use of intravenous octreotide has been reported in some patients with chronic chylous pleural effusions.

#### Trauma

- Blunt and penetrating cardiac trauma may result in hemorrhagic pericardial effusion. In cases of trauma, Beck triad (distant heart sounds, hypotension, and jugular venous distension) rarely occurs, and echocardiography is diagnostic.
- All symptomatic patients require emergent pericardiocentesis. latrogenic hemopericardium also can occur during placement of central venous lines and invasive cardiac procedures.
- A pneumopericardium may be present in patients after an esophageal rupture



#### **Neoplastic Disease**

- Primary tumors of the pericardium are rare. Metastatic disease is more common.
- Primary neoplasms include lymphoma, malignant teratoma, mesothelioma, and angiosarcoma.
- Metastatic tumors include Hodgkin disease, non-Hodgkin lymphoma, leukemia, malignant melanoma, Wilms tumor, neuroblastoma, Kaposi sarcoma, and bone/soft tissue sarcomas.
- Nonmalignant congenital intrapericardial lesions may occur, including pericardial cysts, extralobar pulmonary sequestration, cystic lymphangioma, bronchogenic cysts, and pericardial teratomas.

#### Neoplastic ..

- Pericardial effusions often occur in patients with malignancies, independent of the primary tumor, due to infectious causes or metastatic invasion of pericardial lymphatics. Pericarditis can occur as a complication of certain chemotherapy agents
- Patients who have received mediastinal irradiation are at increased risk of pericardial involvement after chemotherapy.
- Nearly 5% of patients receiving mediastinal irradiation will develop pericarditis within2 months to 2 years after treatment.

#### Postpericardiotomy Syndrome

- Postpericardiotomy syndrome (PPS) usually occurs at least 1 week after intracardiac or pericardial surgery. There is pericardial and pleural inflammation so these patients frequently have pleuritic chest pain. It has been estimated to occur in up to 30% of patients following surgery.
- PPS typically occurs as a single episode, but can recur for weeks to years. Children younger than 2 years rarely are affected.
- PPS may produce irritability, malaise, decreased appetite, and arthralgias. Physical examination will reveal a friction rub, tachycardia, and signs of fluid retention. Cardiac tamponade is rare.



- PPS can occur after blunt cardiac trauma, pacemaker lead placement, and in Dressler syndrome (postmyocardial infarction syndrome).80 While PPS may occur after any cardiac operation,
- it is most frequent after repair of atrial septal defects, ventricular septal defects, and tetralogy of Fallot.
- ▶ It occurs in nearly50% of pediatric patients after cardiac transplantation.

#### PPP..

- Serum laboratory evaluation may reveal nonspecific inflammatory markers, including elevated erythrocyte sedimentation rate or C-reactive protein, and an elevated white blood cell count.
- effusion, which typically reaches its maximal size by the 10th postoperative day
- Aspirin is the primary anti-inflammatory medication recommended, in doses as high as 30 to 75 mg/kg/day in four divided doses for 4 to 6 weeks. As symptoms resolve, aspirin is tapered.
- In patients refractory to NSAIDs, or in those with large effusions, prednisone (2 mg/kg/day, maximum dose 60 mg daily, for 1 week with a 4-week taper) can be effective.

#### Autoimmune and Connective Tissue Diseases

- Pericarditis and pericardial effusions occur in many autoimmune and connective tissue diseases: SLE, juvenile rheumatoid arthritis, dermatomyositis, periarteritis nodosa, mixed connective tissue diseases, Wegener granulomatosis, Takayasu arteritis, and spondyloarthropathies. It has been estimated that 25% of pediatric patients with SLE develop pericarditis This usually is treated with oral NSAIDs, but could require pericardiocentesis if hemodynamic compromise develops.
- Pericarditis occurs in 10% of patients with juvenile rheumatoid arthritis at the time of diagnosis. Oral NSAIDs are effective treatment, but short courses of steroids hasten resolution of symptoms.
- Patients with acute rheumatic fever rarely will develop pericarditis.
- Pericardial effusions in patients with acute rheumatic fever respond well to NSAIDs.

#### **Recurrent and Chronic Pericarditis**

- This occurs most frequently in patients with PPP, juvenile rheumatoid arthritis, or SLE.
- Treatment strategies include reinstitution of NSAIDs, colchicine, or oral steroids.
- Patients have been treated successfully with immune modulators including azathioprine, cyclophosphamide, and intravenous immune globulin.
- Pericardiectomy should be reserved for the patient with multiple recurrences with or without chest pain

#### Chronic pericarditis

- Chronic pericarditis is defined as pericardial inflammation lasting greater than 3 months. Treatment for symptomatic patients includes NSAIDs, steroids, and percutaneous or surgical drainage
- Intravenous immunoglobulin has been reported to be effective in some patients with chronic pericarditis.

#### Thanks for your attention

