**Clinician Update**

Management of Effusive and Constrictive Pericardial Heart Disease

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Case study: A 69-year-old white male was referred for transesophageal echocardiography (TEE) two days after urgent coronary artery bypass grafting. In the first 24 postoperative hours, pleural and pericardial drainage was nearly 3 L, during which time the blood pressure was supported with inotropic agents. The next day, chest tube drainage was minimal, a transthoracic echocardiogram was unremarkable, and the patient was extubated. Shortly thereafter, the patient became hypotensive and dyspneic. TEE revealed a large loculated effusion with linear adhesions and thrombus that compressed both atria, as well as a large pleural effusion. There was marked respiratory variation in Doppler velocities across the tricuspid and mitral valves. At thoracotomy, 1 L of blood was removed from the pericardial space, bleeding at the site of aortic cannulation was sutured, and the patient made an uneventful recovery.

Treatment of effusive and constrictive pericardial disease is often simple and gratifying, but frustration and unforeseen challenges await the unwary clinician. The symptoms and signs of pericardial disease, at times unmistakable (as in the patient described above), may be overshadowed by extracardiac manifestations of a systemic disorder; at other times, they are insidious and conceal their true nature. Pericardial constriction mimics hepatic cirrhosis and myocardial failure and may be virtually indistinguishable from restrictive cardiomyopathy. Another important problem is the lack of placebo-controlled trials from which appropriate therapy may be selected and of guidelines that assist in important clinical decisions; as a result, the practitioner must rely heavily on clinical judgment. Finally, therapeutic options in most cases are limited to antiinflammatory agents, drainage of pericardial fluid, and pericardiectomy. However, despite general agreement on how these measures should be applied in patients with either very mild or severe disease, there is little consensus on the management of the large number of cases encountered with clinical manifestations between these two extremes.

**Effusive Pericardial Disease**

Patients presenting with a pericardial effusion for the first time are usually hospitalized to determine the cause of the effusion and to observe for the development of cardiac tamponade. Disease-specific and adjunctive therapy is given to those in whom pericarditis represents one manifestation of systemic illness. For example, bacterial pericarditis is treated with appropriate systemic antibiotics, surgical exploration, and drainage. Pericardial effusion that persists or recurs during antituberculous therapy occasionally responds to the addition of corticosteroids. Dialysis-associated effusive pericarditis usually improves with intensification of dialysis and regional heparinization or by switching to peritoneal dialysis. Uremic- and dialytic-associated effusions also may benefit from pericardial instillation of corticosteroids, but when they become intractable, pericardiectomy is occasionally necessary. In contrast, myxedematous pericardial effusions are rapidly responsive to thyroid hormone replacement.

In the absence of tamponade or suspected purulent pericarditis, there are few indications for pericardial drainage (Figure 1). Large effusions (when associated with pericarditis) that are unresponsive to nonsteroidal antiinflammatory drugs, corticosteroids, or colchicine, and unexplained effusions, especially when tuberculosis is suspect or when present for >3 months, warrant pericardiocenteses. Data suggest that pericardial drainage cures nearly one half of patients with idiopathic chronic pericardial effusion; in addition, the significant incidence of unforeseen cardiac tamponade is eliminated. Occasionally, a suspicion of malignancy or systemic disease demands pericardial drainage and biopsy. However, routine drainage of large effusions (20-mm echo-free space in diastole) has a very low diagnostic yield (7%) and no therapeutic benefit.
Management of Moderate-Large Pericardial Effusions

**Figure 1.** Algorithm for the management of moderate to large pericardial effusions.

*Antiflammatory treatment if there are signs of pericarditis.

**Right heart catheterization may be required.**

Echocardiography rapidly confirms the presence and hemodynamic impact of an effusion. However, the echocardiographic "signs of tamponade" (right atrial and ventricular collapse, vena caval plethora, respiratory variation in cardiac chamber dimension, and transvalvular and venous flow velocities) that have been introduced to make this assessment have created confusion as to the definition of tamponade. Chamber collapse indicates a transient negative transmural pressure (ie, elevated pericardial, relative to intracardiac pressures) and this typically occurs before hemodynamic embarrassment is evident clinically. Indeed, right cardiac chamber collapse is common in patients with moderate and large pericardial effusion and is correlated weakly with clinical features of tamponade. In a large prospective series in which the reference standard was clinical tamponade, the finding of any chamber collapse had a high (92%) negative predictive value (58% positive predictive value), whereas the positive predictive value (58% positive predictive value) was reduced.6 The positive and negative predictive values for abnormal right-sided venous flows (systolic predominance and expiratory diastolic reversals) but the latter could not be evaluated in more than one third of patients. During cardiac tamponade, tricuspid and pulmonic valve flow velocities increase markedly with inspiration and mitral, aortic, and pulmonary vein flow velocities decrease compared with normal controls and with patients with asymptomatic effusions. However, associated conditions can both create (eg, pleural effusions, chronic obstructive pulmonary disease, left ventricular [LV] dysfunction) and obscure (eg, right ventricular [RV] hypertension) the echocardiographic signs of tamponade. The decision to drain an effusion must therefore take into account not only the echocardiographic findings but also the clinical presentation and the risk-benefit ratio of the procedure. As illustrated in the case report, postoperative pericardial effusions present with atypical features (eg, posterior loculation, basal hematoma, LV diastolic collapse) and require a high index of suspicion. Transesophageal echocardiographic or CT imaging may be required for the correct diagnosis.7

Mild, or low-pressure, tamponade (ie, central venous pressure <10 mm Hg, absent pulsus paradoxus, normal arterial blood pressure) does not require pericardiocentesis, particularly when idiopathic, viral in origin, or responsive to specific therapy (eg, thyroid hormone). At the other extreme, hyperacute tamponade (usually traumatic or iatrogenic) calls for immediate pericardiocentesis; it is noteworthy that cardiac tamponade resulting from "new" percutaneous coronary interventions and temporary pacemaker wires is not a rare occurrence (≈0.2%).8 However, compression due to effusive pericardial disease that falls between these two extremes will require pericardiocentesis, open surgical drainage, or pericardiectomy. Intradural saline solution should be given to patients with cardiac tamponade awaiting pericardial drainage in an effort to expand the intravascular volume. Dobutamine or nitroprusside is used to increase cardiac output after the blood volume has been expanded, but only as a temporizing measure. Vagal reflexes complicating tamponade or pericardiocentesis are treated with atropine. Positive pressure breathing should be avoided.

Unless the situation is immediately life threatening, experienced staff should perform pericardiocentesis in a facility equipped for radiographic, echocardiographic, and hemodynamic monitoring to optimize the success and safety of the procedure. Monitoring the cardiac rhythm and systemic blood pressure is a minimum requirement; invasive hemodynamics and measurement of pericardial pressures are useful for the diagnosis, particularly in questionable cases. Monitoring the local ECG from the needle tip is not recommended by all authors. If such monitoring is employed, however, it is essential that that the apparatus employ equipotential grounding. The use of 2D echo guidance has increased the safety of the procedure.9

Removal of small amounts of pericardial fluid produces considerable symptomatic and hemodynamic improvement because of the steep relation between pericardial pressure and volume. Removal of all of the pericardial fluid normalizes
pericardial, atrial, ventricular diastolic and arterial pressures, and cardiac output, unless there is concomitant cardiac disease or coexisting constriction (ie, effusive-constrictive pericarditis). Drainage of the pericardial fluid with a thin-walled multi-hole catheter (5F to 8F) minimizes trauma, allows measurement of pericardial pressure and instillation of drugs into the pericardium, and helps prevent (but does not guarantee) reaccumulation of pericardial fluid. The catheter may remain in the pericardial space for several days and sclerosing agents, steroids, urokinase, and specific chemotherapeutic agents may be given through the catheter.10

Although pericardiocentesis is usually well tolerated, pulmonary edema, circulatory collapse, and acute RV and LV dysfunction have been reported after drainage.11,12 Patients should be monitored (vital signs, serial echocardiograms) for recurrent tamponade, particularly those with hemorrhagic effusions, which may occur despite the presence of an intrapericardial catheter. Dilute heparin or fibrinolitics may be instilled in the catheter to prevent clotting or fibrin deposition. Patients generally should be observed for 24 hours in an intensive care unit. Major complications of pericardiocentesis include laceration of a coronary vessel, perforation of the myocardium (the thin-walled coronary veins and right heart chambers are particularly prone to brisk bleeding) or lung, hypotension (often reflex in origin), and arrhythmia (both atrial and ventricular).

Although pericardiocentesis may provide effective relief, percutaneous balloon pericardiomyotomy, subxiphoid pericardiomyotomy, or the surgical creation of a pleuropericardial or a peritoneal-pericardial window may be required. Pericardial drainage may also be accomplished by surgical means, either through a subxiphoid incision, with video-assisted thoracoscopy, via thoracotomy, or percutaneously, with a balloon catheter.

The advantages of needle pericardiocentesis include the ability to perform careful hemodynamic measurements, and relatively simple logistic and personnel requirements. Pericardiocentesis is ill advised when there is <1 cm of effusion, loculation, or evidence of fibrin and adhesion. Open surgical procedures offer several advantages, including complete drainage, access to pericardial tissue for histopathological and microbiological diagnoses, the ability to evacuate loculated effusions, and the absence of traumatic injury attributable to the blind placement of a needle into the pericardial space. The choice between needle pericardiocentesis and surgical drainage depends on institutional resources and physician experience, the pathogenesis of the effusion, the need for diagnostic tissue samples, and the prognosis of the patient. Needle pericardiocentesis is often the best option when the pathogenesis is known and/or the diagnosis of tamponade is in question, and surgical drainage is optimal when the presence of tamponade is certain but the pathogenesis is unclear. It should be recognized that surgical approaches (subxiphoid pericardiomyotomy or thoracoscopic drainage) performed with the use of local anesthesia are associated with little attendant morbidity. Irrespective of the method of retrieval, pericardial fluid should be sent for hematocrit and cell count; glucose; smears, culture and cytology.

Recurrent effusions (occurring in up to 40% of cases) may be treated by either repeat pericardiocentesis, intrapericardial instillation of agents with sclerosing or cytostatic activity (eg, tetracycline, bleomycin, thiopeta), surgical creation of a pericardial window, or pericardiectomy. Sclerotherapy produces good results in terms of recurrence prevention (~70% to 90% at 30 days) but is painful and may not be tolerated. A pleuropericardial window provides a large area for fluid reabsorption and is often performed to treat malignant effusions. In critically ill patients, a pericardial window may be created percutaneously with a balloon catheter. Subtotal pericardiectomy is preferred when the patient is expected to survive >1 year.

Constrictive Pericarditis
Constrictive pericarditis results from a thickened, scarred, and often calcified pericardium that limits diastolic ventricular filling. Idiopathic pericarditis and pericardial involvement from cardiac trauma (including surgery), mediastinal irradiation, tuberculosis and other infectious diseases, neoplasms, and renal failure are common antecedents, although acute pericarditis from most causes may bring about constrictive pericarditis.

Chronic constrictive pericarditis is less frequently encountered than in the past, whereas subacute constrictive pericarditis is increasingly common. Postoperative constrictive pericarditis is an important but relatively uncommon cause of constriction with a reported incidence of 0.2%.13 In asymptomatic patients, exercise testing and if available, maximal O2 consumption, should be quantified, jugular venous pressure carefully estimated, and liver function tests measured. The presence of increasing jugular venous pressure, the need for diuretic therapy, evidence of hepatic insufficiency, or reduced exercise tolerance indicates the need for surgery.

Pericardial thickening, calcification, and abnormal ventricular filling produce characteristic changes (eg, flattening of the LV posterior wall endocardium, abnormal septal motion, premature opening of the pulmonic valve, dilated atria) on the echocardiogram. Although these findings lack the specificity to be clinically useful, a normal study virtually rules out the diagnosis.14 Moreover, transvalvular and venous flow velocities are diagnostically effective and play an important role in the infrequent but clinically crucial need for differentiation between restrictive cardiomyopathy and constrictive pericarditis. It is important to examine the respiratory variation of flow profiles, in that constrictive pericarditis waveforms often display marked respiratory variation (patients with very high left atrial pressures may require a reduction in preload to observe the variation), whereas restrictive cardiomyopathy waveforms are generally unaltered (Figure 2). In addition to conventional Doppler, tissue imaging, color M-mode, and myocardial velocity gradients of the posterior LV wall during diastole have been proposed to make the distinction between restrictive cardiomyopathy and constrictive pericarditis. The additive benefit of these newer methods remains to be determined, but their importance increases when respiratory changes are equivocal.16,17 In addition, patients with mixed constriction and restriction, marked obesity, chronic obstructive pulmonary disease, and other conditions that may increase respiratory variation of transvalvular flow velocities are generally not represented in the small series that have been published. Thus, although Doppler echocardiographic studies are promising, the ability to discriminate restrictive cardiomyopathy from constrictive pericarditis with certainty...
Pericardiectomy is the definitive treatment for constrictive pericarditis but is unwarranted either in very early constriction (occult and functional class I) or in severe, advanced disease (functional Class IV), when the risk of surgery is excessive (operative mortality 30% to 40% versus 6% to 19%) and the benefits are diminished.\textsuperscript{18} \textsuperscript{19} Constriction may be transitory with a course lasting weeks to a few months in patients recovering from acute effusive pericarditis. In these patients, the procedure should be delayed until it is clear that the constructive process is not transitory. Symptomatic relief and normalization of cardiac pressures may take several months after pericardiectomy but occurs sooner when the operation is performed before the disease is too chronic and when the pericardiectomy is almost complete. Complete or extensive pericardial resection is desirable, although recent data suggest that in some instances, subtotal pericardectomy may be preferred.\textsuperscript{20} Data from the Mayo Clinic suggest that despite reduced perioperative mortality, the late survival of contemporary patients after pericardiectomy is inferior to that of an age- and sex-matched group of historical controls. The long-term outcome was predicted by three variables in a recent stepwise logistic regression analysis; specifically, the prognosis was worse with increasing age, New York Heart Class IV, and the benefits are diminished.\textsuperscript{18} \textsuperscript{19} Constriction may be transitory with a course lasting weeks to a few months in patients recovering from acute effusive pericarditis. In these patients, the procedure should be delayed until it is clear that the constructive process is not transitory. Symptomatic relief and normalization of cardiac pressures may take several months after pericardiectomy but occurs sooner when the operation is performed before the disease is too chronic and when the pericardiectomy is almost complete. Complete or extensive pericardial resection is desirable, although recent data suggest that in some instances, subtotal pericardectomy may be preferred.\textsuperscript{20} Data from the Mayo Clinic suggest that despite reduced perioperative mortality, the late survival of contemporary patients after pericardiectomy is inferior to that of an age- and sex-matched group of historical controls. The long-term outcome was predicted by three variables in a recent stepwise logistic regression analysis; specifically, the prognosis was worse with increasing age, New York Heart Association class, and a postirradiation pathogenesis.\textsuperscript{19} Pericardiectomy is commonly performed via a median sternotomy, although some surgeons prefer access through a thoracotomy. Despite a decline, the risk of mortality remains \( \approx 6\% \) to 19%. Heavy calcification and involvement of the visceral pericardium increase the risk. LV systolic dysfunction may occur after decorticating a severely constricted heart. Although the LV dysfunction may require treatment for several months, it usually resolves completely. In highly selected patients, orthotopic transplantation may be considered.

Medical therapy of constrictive pericarditis has a small but important role. In some patients, constrictive pericarditis resolves either spontaneously or in response to various combinations of nonsteroidal antiinflammatory agents, steroids, and antibiotics;\textsuperscript{21} in the remaining patients, medical therapy is adjunctive. Specific antibiotic (eg, antituberculous) therapy should be initiated before surgery and continued afterward. Preoperative diuretics should be used sparingly with the goal of reducing, not eliminating, elevated jugular pressure, edema, and ascites. Postoperatively, diuretics may be useful. Diuretics and digoxin (in the presence of atrial fibrillation) are useful in patients who are not candidates for pericardiectomy because of their high surgical risk.

Prevention of pericardial constriction consists of appropriate therapy of acute pericarditis and adequate pericardial drainage. Although instillation of fibrinolytics (eg, urokinase 400 000 U per instillation to 1 600 000 U; streptokinase 400 000 U per instillation to 1 000 000 IU) is promising, corticosteroid instillation is often ineffective.\textsuperscript{22} Evaluation of patients with effusive and constrictive pericardial disease affords clinicians the opportunity to integrate bedside observations and noninvasive testing to expeditiously arrive at a management plan.

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\textbf{References}

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