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Chapter

Constrictive Pericarditis: Surgical Management

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Abstract

Constrictive pericarditis represents an uncommon sequela of multiple pathologic processes. It involves the pericardium, a tri-layered sac that encases the heart within the mediastinum. Inflammation of the pericardium can lead to formation of fibrous adhesions between the outer wall of this sac and the surface of the heart. Due to the stiff, inflexible structure of the pericardium, its adherence to the heart negatively impacts normal diastolic filling and hemodynamics. Over time, this can lead to reduced cardiac output and severe heart failure. This condition is typically refractory to medical treatment. The definitive treatment of constrictive pericarditis involves surgical decortication and removal of the pericardium to alleviate the constriction and restore normal diastolic filling capacity. This procedure has evolved since its inception and is now the gold standard in curing constrictive pericarditis. However, despite its necessity in the treatment of constrictive pericarditis, this procedure carries considerable risk of intra- and post-operative complications and poor outcomes. The poor prognosis is often related to the patient’s pre-surgical status, which must be considered when identifying candidates for surgery. When successful, though, pericardiectomy can produce immediate and progressive improvements in hemodynamic parameters.

Keywords: surgery, constrictive pericarditis, pericardiectomy, surgical treatment of constrictive pericarditis, diagnosis of constrictive pericarditis

1. Introduction

Constrictive pericarditis is an uncommon, diagnostically challenging disease in which the layers of the pericardium become fused and impede normal cardiac function. Historically, the majority of cases were idiopathic. While this is still a leading cause of the disease today, the drastic increase in number of individuals undergoing cardiac surgery, interventional, and electrophysiologic procedures has led to iatrogenic causes becoming a more common source of constriction in the United States and Europe [1, 2]. Elsewhere, tuberculosis infection represents the leading cause of constrictive pericarditis. Studies in India, for example have attributed up to 93% of constrictive pericarditis cases to tuberculosis compared to 4% in one US study [1, 3, 4]. As case numbers continue to rise, the importance of appropriate diagnosis and treatment methodologies will also rise.
Located within the mediastinum, the pericardium is comprised of 3 layers that encircle the heart. It serves important purposes including protecting the heart from friction-related damage, regulating diastolic filling, and preventing overexpansion. The outermost layer of the pericardium, called the fibrous pericardium, is comprised of dense irregular connective tissue and, due to its lack of elastic properties, helps limit ventricular volume capacity. The fibrous pericardium is anchored superiorly to the great vessels at the base of the heart and is continuous with the tunica adventitia. It attaches to the diaphragm inferiorly. The inner portion of the pericardium is formed by a serous bilayer, collectively referred to as the serous pericardium. The outermost layer of the serous pericardium is called the parietal pericardium and is fused with the fibrous pericardium to form a single outer envelope. The parietal layer reflects around the roots of the great vessels where the fibrous pericardium emerges from the adventitia and covers the outermost surface of the heart as the visceral pericardium. The visceral pericardium is also referred to as the epicardium and provides an external covering for the coronary vessels and myocardial cells. The visceral pericardium also contains mesothelial cells that are responsible for manufacturing and secreting pericardial fluid into the pericardial cavity. This fluid helps lubricate the layers of the pericardium as they come into contact with one another during the cardiac cycle.

Constrictive pericarditis occurs following inflammation or injury to the pericardium. As the pericardium heals, fibrous adhesions can form, anchoring the layers of serous pericardium to one another. This results in anchoring of the fibrous pericardium to the surface of the heart, progressively reducing diastolic filling capacity and leading to symptoms of heart failure. This can be further complicated by formation of calcifications that may extend deep into the myocardium, making cardiac function and treatment more difficult.

Currently, the most effective treatment for constrictive pericarditis is total pericardectomy [5–9]. This procedure has evolved over the years from the previously favored partial or “phrenic-to-phrenic” procedure which, while less technically challenging, did not resolve all constrictive foci. This resulted in continued constriction of the posterior surfaces of the heart and less-favorable patient outcomes in many cases. With the shift toward complete pericardial resection, survival rates following surgical treatment for constrictive pericarditis have improved. However, the underlying etiology of constriction and patient condition at the time of surgery do play crucial roles in predicting a particular individual’s prognosis. While resolution of idiopathic and tuberculosis-related constriction has produced 5-years survival rates around 80%, rates in cases stemming from previous thoracic surgery and prior radiation treatment are much lower [5]. Similarly, patients with advanced disease or poor hemodynamic parameters at the time of treatment experience a perioperative mortality rate of up to 60% [10].

In this chapter, we will review the relevant mediastinal anatomy, discuss the pathophysiology and clinical presentation of constrictive pericarditis, as well as the common diagnostic findings. We will cover, in depth, the surgical treatment of constrictive pericarditis, including the varying approaches and prognostic factors.

2. Mediastinal anatomy

The pericardium forms a 3-layered envelope that surrounds the heart. It consists of a dense, inelastic outer layer called the fibrous pericardium and a serous bilayer. The bilayer consists of a visceral pericardium that lies adherent to the heart, also known as
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the epicardium, as well as a parietal layer that is fused with the fibrous pericardium. The visceral pericardium, comprised of mesothelial cells, secretes pericardial fluid that helps reduce friction as the heart pumps within the envelope.

The pericardium defines the mediastinum within the thoracic cavity, separating the heart and great vessels from the pleural spaces. The pericardium arises from the tunica adventitia of the great vessels superiorly and is anchored to the central tendon of the diaphragm inferiorly. Its lateral borders lie adjacent to the pleura. Anteriorly, the pericardium attaches to the posterior surface of the sternum via weak sterno-pericardial ligaments. The pericardium extends circumferentially to cover the dorsal surface of the heart. Of surgical consideration, the left phrenic nerve lies superficial to the pericardium of the left ventricle and the right phrenic nerve lies to the right of the pericardium.

The reflections of the serous pericardium contribute to the overall anatomy of the mediastinum by creating two sinuses. The oblique sinus spans the distance between the right and left pulmonary veins on the dorsal surface of the heart. This space is encapsulated posteriorly by the serous pericardium and anteriorly by the left atrium. The oblique sinus allows for posterior expansion of the left atrium in to accommodate additional blood volume. The transverse sinus is a result of the visceral serous pericardium reflecting off the posterior pulmonary trunk and aorta to adhere to the atri. This creates an open channel behind the pulmonary trunk and aorta that emerges anterior to the superior vena cava.

The pericardium performs physiologic functions as well. The dense connective tissue that comprises the fibrous pericardium prevents overexpansion of the heart and limits diastolic filling. The fluid produced by the visceral mesothelial cells provides a barrier around the heart to reduce friction during contraction, and the pericardial attachments help to reduce motion of the heart within the thoracic cavity.

The pericardium primarily receives its blood supply from branches of the internal thoracic arteries called the pericardiophrenic arteries. Additional blood supply is delivered from branches of the musculophrenic, bronchial, esophageal, and superior phrenic arteries. The visceral pericardium also receives a portion of its blood supply from the coronary arteries. Venous drainage occurs via the pericardiophrenic veins, which drain into the brachiocephalic trunk and the azygos system. The visceral pericardium drains lymph into the tracheal and bronchial lymphatic chain, while the parietal pericardium empties into mediastinal lymph nodes. Ventral pericardial lymphatics travel over the cranial portion of the phrenic nerves. On the posterior and lateral surfaces, the lymphatics join with lymphatic vessels of the mediastinal pleura.

The parietal pericardium receives somatic sensory innervation from the phrenic nerve arising from C3–C5. The visceral pericardium lacks sensory innervation. Autonomic innervation arises from the sympathetic trunk and vagus nerve.

3. Etiology of constrictive pericarditis

In the western world, acute pericarditis most often lacks a definitive diagnostic origin [5, 11–15]. Of those cases determined to be of viral origin, infection by Epstein Barr Virus (EBV), Cytomegalovirus (CMV), influenza, HIV, Adenoviruses, Echovirus, Parvovirus B19, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), and Coxsackieviruses A and B have all been implicated [16–26]. As mentioned previously, tuberculosis is the leading cause of constrictive pericarditis in many areas of the world, though it is not the only bacterial source of this disease [3, 4, 27]. In rare cases,
pericarditis may result from bacterial infection by Meningococcus, Pneumococcus, Coxiella burnetii, Staphylococcus, and Streptococcus species [28]. Acute pericarditis may also, in extremely rare cases, be the result of fungal infection by Coccidiodes, Candida, Histoplasma, or Blastomyces or parasites such as Echinococcus and Toxoplasma [29]. The inflammatory reaction induced by these microbial species can lead to formation of fibrous adhesion between the parietal and visceral pericardia, resulting in constriction.

Another leading cause of constrictive pericarditis in western countries is previous cardiac surgery. Rates of constrictive pericarditis have actually increased as cardiac surgery becomes more commonplace and is now seen over a wider variety of age groups compared to previous years [2]. Surgeries in which the pericardium is damaged or manipulated in some way predispose the patient to the development of fibrous adhesions and, later, constriction. Therefore, any patient with a prior history of corrected congenital heart disease, valvular surgeries, coronary artery bypass grafting, or other procedures involving opening of the mediastinum should be evaluated for the development of pericardial thickening or calcification. One twenty-year study found that patients diagnosed with constrictive pericarditis were more likely to have been treated surgically for valvular disease and atrial fibrillation and demonstrated a significantly increased 7-year mortality rate compared to controls [30].

Patients who have undergone mediastinal irradiation for treatment of primary or metastatic cancer are also at increased risk of developing constrictive pericarditis. Some evidence suggests that patients who develop constrictive pericarditis following radiation treatment are also at increased risk of mortality following pericardiectomy compared with patients receiving the same procedure for other causative states [10]. In fact, mediastinal irradiation is considered an independent prognostic factor for mortality following surgical correction of constrictive pericarditis, with 5-year survival at a dismal 11.0% and an increased 10-year mortality rate [13]. One retrospective study reported an overall intraoperative mortality of 10.1% for patients with a history of mediastinal irradiation over a seventeen-year period, though it should be noted that the majority of those patients underwent concomitant procedures that may have attributed to their outcome [31]. While there have been numerous reports indicating poor outcomes for patients undergoing pericardiectomy following mediastinal radiation treatment, it cannot be overlooked that the increased mortality rates among these patients may be related to their original need for radiation therapy, rather than an interaction between pericardiectomy and irradiated tissue.

In developing countries, tuberculosis infection is most often the causative agent of constrictive pericarditis. This is especially pronounced in HIV-positive patients who may lack the robust immune system needed to protect the pericardium from inflammation. Current estimates suggest that tuberculosis is second only to purulent disease as the cause of constrictive pericarditis in areas such as sub-Saharan Africa and Asia [27]. Exact data is difficult to acquire, however, due to the challenges of diagnosing this disease. Definitive diagnosis of tuberculous pericarditis is based on the presence of tubercle bacilli in samples of a patient’s pericardial fluid or histologic section of their pericardium. It is possible to identify “probable” cases in patients with known tuberculosis infection and otherwise idiopathic pericarditis [4].

Other, less common, causes of constrictive pericarditis include connective tissue diseases such as rheumatoid arthritis, systemic lupus erythematosus, Behçet’s disease, scleroderma, and Sjögren syndrome [7, 32–34]. Amyloidosis and sarcoidosis can result in fibrous adhesion between pericardial layers, as can metabolic pathologies such as uremia [35–40]. Inflammation secondary to acute coronary syndromes have also been
shown to precipitate constrictive pericarditis, a condition known as Dressler’s syndrome [41, 42]. Additionally, purulent pericarditis may lead to constriction in cases of incomplete drainage [41, 43].

Lastly, certain pharmaceuticals have been implicated in the pathogenesis of constrictive pericarditis. Procainamide, hydralazine, and isoniazid have all been reported as causative agents of drug-induced systemic lupus erythematosus (SLE) [33, 44]. SLE, as previously discussed, can lead to pericarditis through the induction of serositis [33, 34]. The resultant inflammation can produce constrictive adhesions in such instances. Cardiotoxic drugs, such as Ipilimumab and Nivolumab, have also been attributed to development of constrictive pericarditis [45–48]. Patients taking these drugs should be monitored for changes in hemodynamic parameters and evidence of constriction.

4. Pathophysiology of constrictive pericarditis

Constrictive pericarditis is the result of a chronic inflammatory process that causes fibrous adhesions and calcifications to form between the epicardium and parietal pericardium. The ongoing inflammation leads to calcium deposition and remodeling, resulting in thickening and scarring of the tissue. This scarring can reach deep into the adjacent myocardium, further reducing cardiac function. This inflammation may be attributed to episode(s) of acute pericarditis, chronic pericarditis, or the other mechanisms described above.

Inflammation of the pericardium can result from a number of processes and occurs in both acute and chronic forms. Acute pericarditis is one of the most common disorders involving the pericardium and occurs in approximately 0.1–0.2% of hospitalized patients and 5% of patients admitted to the emergency department for nonischemic chest pain [49, 50]. Clinical presentation of acute pericarditis includes sharp, pleuritic chest pain that is alleviated with the patient leans forward, thereby decreasing contact of the pericardium with nearby structures. On auscultation, a pericardial friction rub can be heard at the left sternal border. Electrocardiogram changes commonly associated with acute pericarditis include depression of the PR segment and widespread ST elevation early in the disease process. It should be noted, however, that these ECG findings may change over the course of the disease.

Under normal physiologic conditions, the pericardium is not especially compliant but is capable of accommodating small changes in preload experienced by the heart. It can also expand over time in cases of prolonged, slowly accumulating pericardial effusion. However, the pericardium is not typically distensible or capable of elastic recoil. This becomes relevant when discussing the pathologic changes observed in constrictive pericarditis. In the normal cardiac cycle, diastolic volume increases by approximately 70 mL over systolic volume. This expansion in volume causes the lateral walls of the ventricles to expand outward, which is accommodated by the pericardial cavity. This helps to regulate filling volume while also allowing for appropriate preload.

Another part of normal physiology is the interplay between respiration and cardiac filling. This is represented by the relationship between intrathoracic and intracardiac pressures. During inspiration, there is an increase in right-sided venous return, which causes expansion of the right ventricle and pushes the interventricular septum into the left ventricle to accommodate the added volume. This produces a transient reduction in left ventricular size and transmural filling pressure, leading to
a drop in left ventricular end diastolic volume. Reduced end diastolic volume results in reduced stroke volume. These changes tend to be minimal and produce only small changes to hemodynamic parameters.

When adhesions form between the layers of the pericardium, the pericardial cavity is lost, and the outer fibrous pericardium must move in synchrony with the expanding ventricles during diastole. As mentioned, though, the fibrous pericardium lacks elasticity. This limits the ability of the ventricle to expand outwardly and accommodate its normal diastolic volume. As the constriction becomes more severe, ventricular filling can become severely impeded and even lead to transient displacement of the interventricular septum into the left ventricle, called septal bounce. Decreasing the preload capacity of the heart leads to reduced cardiac output and venous congestion. Over time, the restricted ventricular filling leads to dissociation of intracardiac and intrathoracic pressures with respiration and equalization of intracardiac diastolic filling pressures, which increase until the patient develops right heart dysfunction [51]. Reductions in caval flow velocity during expiration, decreased mitral flow velocity, reduced heart rate, and increased hepatic venous diastolic flow also result [52]. Reduced end diastolic volume, stroke volume, and cardiac output result. These changes mimic, and can eventually lead to, heart failure.

5. Patient presentation and symptom progression

Patients with constrictive pericarditis often present with symptoms that mimic heart failure with preserved ejection fraction. As adhesions form between the parietal pericardium and epicardium, myocardial function is progressively hindered. This results in reduced cardiac output as well as pulmonary and systemic venous congestion. These abnormal physiologies can lead to symptoms of progressive exertional dyspnea, fatigue, tachypnea, peripheral edema, and gastrointestinal upset [1, 7, 53]. The patient may also experience exertion-independent tachycardia. Some patients may report chest pain or present with atrial arrhythmias, or symptoms of cardiac tamponade [1, 7, 53].

It is important to obtain a thorough patient history in a patient presenting with signs of heart failure. Those who report previous episodes of acute pericarditis, tuberculosis, mediastinal radiation treatment, prior cardiothoracic surgery, or previous chest trauma should produce a high index of suspicion for constrictive pericarditis.

5.1 Physical exam

Constrictive pericarditis can be identified on physical exam of the patient through palpation of the precordium. A ‘diastolic apex beat’ or diastolic precordial impulse represents a positive finding, as this beat should normally be felt during systole [51]. The abrupt termination of early diastolic filling, which is characteristic in constrictive pericarditis, is responsible for this switch. Note that a positive finding should be confirmed by palpating the impulse at multiple areas along the sternum and epigastric region and comparing the beat against the carotid pulse [51, 54].

While precordial palpation can reveal characteristic signs of constrictive pericarditis, the most common, though nonspecific, finding is elevated jugular venous pressure [51, 54]. Distension can frequently be observed by reclining the patient to an elevation of thirty degrees and having them look to their left. The clinician can then evaluate the jugular vein for distension and abnormal pulsation. It should be noted that elevated jugular venous pressure may not be observed in patients with mild to
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moderate constriction [54]. However, a high index of suspicion should be aroused in
patients presenting with unexplained jugular vein distension and a history of known
predisposition to constrictive pericarditis.

Other features that may be observed include pericardial knock or friction rub,
Kussmaul sign, or pulsus paradoxus [2]. As mentioned previously, peripheral edema
is also a frequent finding in patients with constrictive pericarditis, which may be
accompanied by ascites and hepatomegaly [51]. Pleural effusion is also often found
during the physical exam [2].

6. Diagnostic criteria and the differential

Early diagnosis of constrictive pericarditis is of vital importance to the success
of treatment as well as the long-term prognosis of the patient. Early treatment via
pericardiectomy is associated with lower intra- and post-operative risk and reduced
incidence of complications [51]. Surgical intervention prior to the onset of NYHA
Class III or IV symptoms—that is, heart failure symptoms with minimal exertion or
at rest, respectively, is associated with significantly reduced risk of morbidity and
mortality in the 30-days following pericardiectomy [7, 51, 55].

6.1 Imaging

The diagnosis of constrictive pericarditis is based on guidelines set by the American
College of Cardiology and the European Society of Cardiology, both of which recom-
mand evaluation by 2-D echocardiogram [7, 56]. This test allows for visualization of
calcifications or increased thickness of the pericardium—both diagnostic indicators of
constrictive pericarditis. Echocardiogram also detects two other characteristic changes
associated with constriction: ventricular interdependence and loss of intrathoracic
pressure variation with breathing [7, 10, 56–58]. The abnormal rigidity of the peri-
cardium in constrictive pericarditis prevents independent activity of the ventricles,
which can be seen as decreased diastolic filling time and septal bounce [7, 10, 56–58].
Additionally, echocardiogram may detect the presence of dilation of the inferior vena
cava due to decreased preload [7, 10, 56–58].

M-mode ultrasound may also be used to exclude constrictive pericarditis from
a list of differential diagnoses. In constrictive pericarditis, when a patient inhales,
M-mode ultrasound should show posterior movement of the interventricular septum
during the early diastolic phase [59–61]. Another inspiratory feature indicative of
pericardial constriction is the absence of increased systemic venous return, again
visible on ultrasound [61, 62]. A third feature that would suggest a patient may be
experiencing constrictive pericarditis is the premature opening of the pulmonic valve
due to increased ventricular diastolic pressure [61, 62]. Absence of any of these three
features should help the clinician to rule out constrictive pericarditis [61].

Doppler echocardiography is also useful in the diagnosis of constrictive pericar-
ditis. Key indicators of this state include abnormal filling of the ventricles in early
diastole and changes in flow velocity across the tricuspid valve during the respiratory
cycle [63–65]. More specifically, during inspiration the clinician should expect to see
an increase in diastolic flow velocity followed by a decrease during expiration [56].
Additionally, the pulmonary veins and mitral valve should experience a drastic reduc-
tion in flow velocity during inspiration accompanied by a shift of the interventricular
septum toward the left ventricle [56].
Computed tomography (CT) and cardiovascular magnetic resonance (CMR) imaging are commonly used as adjuncts to echocardiography when making a definitive diagnosis of constrictive pericarditis. These imaging modalities are particularly useful, though, in differentiating constrictive pericarditis from restrictive cardiomyopathy, a challenging distinction and common misdiagnosis. Cardiac MRI and CT provide higher resolution and a broader field of view than more traditional imaging modalities [66]. Cardiac MRI, in particular, is able to provide high-resolution and contrast of the heart and related cardiac structures, including the pericardium. Use of cardiac MRI allows for accurate measurement of pericardial thickness, which can be used as diagnostic criteria in constrictive pericarditis. Pericardium that exceeds 4 mm in thickness produces a signal intensity that is equal to that of the myocardium and characteristic calcification of the tissue can be visualized as well [67]. Other abnormalities associated with constrictive pericarditis that may be detected with cardiac MRI include “tubing” of the right ventricle, enlargement of the atria, abnormal motion of the interventricular septum, and enlargement of the inferior vena cava due to decreased preload [67].

Chest radiographs may also be useful in the diagnosis of constrictive pericarditis. Patients who exhibit calcifications on radiograph in the presence of a consistent clinical picture should produce a high index of suspicion. More specifically, lateral and anterior oblique images of patients with constrictive pericarditis may show concentric, linear rings of calcification surrounding the heart [7]. It should be noted that evidence of pericardial calcifications on chest radiograph is not in itself diagnostic of constrictive pericarditis as calcification can occur for a number of reasons, however the pattern in non-constrictive disease is often more diffuse or patchy [7, 68, 69]. Some patients with constrictive pericarditis may not show evidence of calcification at all and therefore radiograph should not be used to eliminate this diagnosis from the differential in the setting of other, more characteristic findings.

6.2 Electrocardiogram

In contrast to acute pericarditis, constrictive pericarditis does not demonstrate pathognomonic changes on electrocardiogram (ECG). A wide variety of ECG changes, ranging from a normal QRS complex to low voltage, and generalized T-wave flattening, or inversion may be exhibited [70]. Other commonly seen ECG changes are those of right ventricular hypertrophy and right axis deviation [70]. Some patients may display non-specific ST-segment changes, but the most common abnormality observed is low voltage but, as stated, this is not diagnostic of constriction. Also, in patients with long-standing or advanced constriction, the chronic elevation of left atrial pressures may manifest as atrial fibrillation [6, 71]. While ECG findings may aid in the diagnosis of constrictive pericarditis, any changes should be evaluated in conjunction with echocardiography and CT or CMR imaging.

6.3 Cardiac catheterization

While more noninvasive diagnostic techniques are generally favored in the diagnosis of constrictive pericarditis, right heart catheterization remains the gold standard and should be performed in patients being considered for surgical treatment. Cardiac catheterization is particularly useful when other imaging modalities produce inconclusive results, or the diagnosis is particularly challenging. Cardiac catheterization allows for monitoring of the hemodynamic changes characteristic of constrictive pericarditis. In particular, the abnormal ventricular filling associated with different
phases of the respiratory cycle and eventual equalization and interdependence of right and left diastolic pressures can be measured with cardiac catheterization, then used in the making of a definitive diagnosis [72].

Pressure changes during cardiac catheterization can also be of substantial use in confirming a diagnosis of constrictive pericarditis. Indications include a notable drop in pulmonary capillary wedge pressure compared to left ventricular diastolic pressure during inspiration and sharp decreases in x and y descents of atrial and venous pressure tracings [7, 73]. Diastolic pressure changes in the right and left ventricles may produce a “square root” sign with an absent wave, which reflects the rapid ventricular filling during early diastole, followed by diastolic plateau caused by compression [7]. Findings may also include increased right atrial pressure and increased right ventricular end-diastolic pressure [7].

6.4 Differential diagnoses

Diagnosis of constrictive pericarditis can be particularly challenging as the clinical presentation of this disease closely resembles heart failure with preserved ejection fraction, which can have a number of underlying etiologies. Therefore, it is important for clinicians to maintain a high index of suspicion in patients who report predisposing factors, such as prior cardiothoracic surgery, previous mediastinal radiation or malignancy, thoracic trauma, tuberculosis, or a history of connective tissue disorders (see Section 3). A thorough physical exam and utilization of the imaging modalities described above can help to rule in or rule out a diagnosis of constrictive pericarditis. Still, definitive diagnosis can be difficult.

On the list of differentials in a patient presenting with signs of constrictive pericarditis should be restrictive cardiomyopathy. While constrictive pericarditis results from pericardial thickening and formation of adhesions between pericardial layers that results in reduced ventricular compliance, restrictive cardiomyopathy is due to progressive myocardial stiffness which likewise produces a decrease in ventricular compliance. Restrictive cardiomyopathy is often considered to be the most diagnosis similar to constrictive pericarditis and may therefore be difficult identify. Proper differentiation between these two conditions is crucial, as the treatment methodologies vary drastically. Constrictive pericarditis, in many cases, can undergo definitive treatment with pericardiectomy. Restrictive cardiomyopathy, on the other hand, has no curative therapeutic options and often requires cardiac transplantation.

As mentioned previously, constrictive pericarditis is frequently associated with a history of one or more predisposing factors. Prior treatment for mediastinal malignancy, mediastinal radiation, or with cardiothoracic surgery should increase suspicion of underlying constriction [5, 7, 11, 12, 15, 74]. Likewise, constrictive pericarditis, unlike restrictive cardiomyopathy, is associated with a history of tuberculosis, viral infections, trauma, and connective tissue disease [17, 18, 20, 22–26, 75]. Restrictive cardiomyopathy, in contrast, is often related to sarcoidosis, amyloidosis, or inherited mutations in one of several genes related to the sarcomere subunit [6].

In both constrictive pericarditis and restrictive cardiomyopathy, patients can have increased right and left sided filling pressures and preserved ejection fractions [6, 7]. Both conditions can also present as diastolic heart failure in their later stages [6, 7]. Fortunately, appropriate diagnostic testing and a thorough patient history can help elucidate the underlying cause of a patient’s symptoms and ensure proper treatment. Though not an exhaustive list, some useful diagnostic differences between constrictive pericarditis and restrictive cardiomyopathy include:
1. Whereas constrictive pericarditis generally lacks notable ECG changes, restrictive cardiomyopathy can present with changes in depolarization, pathologic Q waves, impaired conduction, repolarization abnormalities, or ventricular hypertrophy [76]. Though it may be possible to observe abnormal repolarization or low voltage in constrictive pericarditis though or nonspecific ST or T wave changes, again, this is not common [60]. Also of note, in their later stages both conditions may predispose a patient to atrial fibrillation [6, 71].

2. The difference in the underlying pathophysiology of both diseases produces varying results in B-type natriuretic peptide (BNP) values and imaging studies. BNP levels tend to be higher in patients experiencing restrictive cardiomyopathy compared to constrictive pericarditis, likely owing to lack of ventricular wall compliance in the former [77–79].

3. Pericardial calcification is sometimes seen in constrictive pericarditis but has not been commonly associated with restrictive cardiomyopathy [7, 68, 69]. Rarely, calcification of the ventricle may be seen in restrictive physiology and may contribute to the pathogenesis of this disease [80].

4. Perhaps of no surprise, pericardial changes are not commonly observed in the setting of restrictive cardiomyopathy [81]. Increased pericardial thickness (>4 mm) may, however, be indicative of constrictive pericarditis when seen on imaging [60]. It should be noted that pericardial thickening is not always observed in patients with constrictive pericarditis and the absence of thickening should not be used to definitively eliminate constriction from the differential [60, 81].

5. As mentioned previously, constrictive pericarditis frequently presents with significant respiration-dependent changes in ventricular filling on Doppler studies [82]. Any such filling changes in restrictive cardiomyopathy are usually minimal [82]. When measuring pulmonary venous flow using transesophageal echocardiography, it has been reported that peak systolic flow variations and flow velocities during the respiratory cycle were also greater in patients with confirmed constrictive pericarditis, compared with restrictive cardiomyopathy [65].

6. The fibrous myocardium that is characteristic of restrictive cardiomyopathy limits movement of the muscle. This is translated into markedly reduced septal bounce during diastole. As discussed previously, constrictive pericarditis produces notably increased septal bounce as movement of the outer walls of the ventricle become more impeded [7, 10, 56–58].

7. Measures of tissue strain using CMR tend to be significantly higher in patients with restrictive cardiomyopathy as compared to restrictive pericarditis, likely owing to the decrease in myocardial compliance associated with fibrous infiltrates [83].

7. Surgical treatment of constrictive pericarditis

In 1898, the French physician, Dr. Delorme, first proposed surgical intervention in the treatment of pericarditis [84]. However, another fifteen years would pass before
Dr. Ludwig Rehn would perform the first successful pericardiectomy as a treatment for constrictive pericarditis [85]. In the United States, the first successful surgical treatment for constrictive pericarditis was performed by Dr. Edward Delos Churchill at Massachusetts General Hospital in 1928 [86]. The surgery has evolved since then and continues to be a mainstay of treatment for pericardial disease. Today, pericardiectomy is the gold standard of treatment for constrictive pericarditis and considered the only curative, rather than palliative, option. For most patients, medical therapy is only effective in the treatment of acute pericarditis, where corticosteroids and anti-inflammatory medications have shown to produce acceptable outcomes.

7.1 Indications for surgery

When determining whether a patient is a good candidate for pericardiectomy in the treatment of constrictive pericarditis, numerous factors must be taken into consideration. This procedure, while curative, is not without notable risks to the patient. Research into prognostic indicators is ongoing, though current recommendations focus on preoperative state and the patient’s medical history.

Pericardiectomy produces the most positive outcomes in the treatment of constrictive pericarditis when performed early in the disease course [87, 88]. Clinical judgment is used to determine which patients are best suited to undergo pericardiectomy. Patients who fall into NYHA heart failure Classes I and II may remain clinically stable for years and be placed at unnecessary risk through surgery [41]. However, a delicate balance must be struck, as patients with advanced pericardial disease in NYHA Class IV who have significant left ventricular dysfunction or advanced fibrosis and calcification tend to have high mortality rates [41]. Therefore, outcomes depend largely on individual patient factors and a thorough risk-benefit analysis should be employed.

7.2 Midline sternotomy approach

The surgeon must not only determine whether the patient is a good candidate for pericardiectomy, but also which surgical technique is most appropriate given the patient’s specific condition.

Approaching the pericardium through a median sternotomy is the most common technique used in the decortication procedure. This access provides the broadest view of the heart and its related structures, as well as the lungs and, crucially, the phrenic nerves. The initial opening of the pericardium after a midline sternotomy may be easiest at the lower portion of the right ventricle, over the epicardial fat pad by the diaphragm. This provides the safest avenue of identifying the appropriate dissection planes, as the likelihood of damaging underlying structures is low. Dissection continues as the surgeon identifies the dissection plane that will separate the epicardium from its parietal pericardial adhesions, taking special care to avoid damaging the coronary vessels. It is also important to consider which portion of the heart will be decorticated first. Traditionally, the left ventricle is freed first as, this helps to prevent pulmonary edema that may otherwise occur if the right ventricle were freed first. This particular approach can be quite challenging, though, and some surgeons elect to begin with the right ventricle and relieve the anterior plane first.

Avoiding damage to the underlying cardiac structures is of paramount importance when performing a pericardiectomy. As such, the surgeon must be cognizant of the dissection plane at all times. This can be particularly challenging in areas of especially thickened adhesions. In such instances, it may be necessary to dissect around a focal
adhesion and perform a waffle procedure to minimize the risk of inadvertent damage to underlying myocardium. A waffle procedure involves making multiple longitudinal and transverse incisions in the thickened area of pericardium to create a more distensible surface.

As the surgical treatment for constrictive pericarditis has evolved and outcomes have been analyzed, the preferred techniques for removal of the pericardium have also changed. Historically, a partial, or anterior, pericardiectomy was performed. Following decortication of the mid-anterior portion of the pericardium, dissection proceeds laterally toward the phrenic nerves, carefully separating the planes of tissue and ending approximately 1 cm anterior to the nerves. Immediate hemodynamic improvement is observed upon removal of the diseased pericardium from the ventricles. The surgeon then turns their attention to the atria and all stiff pericardial tissue is resected. At completion, only the anterior section of the pericardium is removed, leaving the posterior surfaces adhered. While this approach is thought to be considerably less challenging and, therefore, safer than the alternative total pericardiectomy, it leaves intact any posterior adhesions and does not provide full resolution of the constriction. It also leaves an opportunity for further adhesions to form on the posterior surface of the heart and lead to progressively increased constriction and worsened hemodynamics. As will be discussed in more depth in the “Outcomes” section, patients who undergo partial pericardiectomy tend to experience sub-optimal outcomes and increased risk of complications [5, 8, 9].

Today, the more accepted approach is the total, or radical, pericardiectomy. Most modern studies report improved outcomes with total pericardiectomy. Improved hemodynamics, as measured by right ventricular pressures and reduced instances of tricuspid regurgitation, have been noted with complete pericardial removal as compared with the partial removal procedure [9]. Lower long term mortality rates have also been reported in total versus partial pericardiectomy [9]. Despite the more favorable outcomes of total pericardiectomy, some patients may be more suited to the partial approach. This includes those with advanced pericardial disease, poor cardiac function, or those at risk of acute heart failure following surgery [41].

This procedure begins at the right atrium, where the appropriate dissection plane is identified, and the right atrium is freed from its pericardial adhesions. This dissection continues to the level of the pulmonary veins and inferior vena cava. It is at this point that the right phrenic nerve is delicately removed as a fat pedicle, and the pericardium can be resected from around the entirety of the inferior vena cava. The surgeon then turns their attention to the left side of the heart, dissecting over the left atrium to the diaphragmatic surface of the heart, again taking special care around the coronary arteries and particularly dense adhesions. The left phrenic nerve is detached and protected as a fat pedicle. The dissection continues, detaching the pericardium from the diaphragm, pulmonary ligaments, posterior mediastinum, and major blood vessels until it can be extracted in its entirety.

Once all visible pericardial adhesions have been relieved, thoracic drains are inserted, the patient is monitored for hemodynamic stability, and echocardiography confirms appropriate cardiac blood flow.

7.3 Anterolateral thoracotomy

An alternative approach that is favored in some instances is the anterolateral thoracotomy. It provides for sufficient visualization of the lateral and diaphragmatic
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surfaces of the left ventricle without the need for excessive manipulation of the heart required of the midsternal approach. This approach is particularly beneficial for patients whose adhesions are primarily focused on the left side of the heart. It is less useful when the right side of the heart is involved as the field of view is very limited in that area.

The process of an anterolateral approach to pericardiectomy involves opening of the chest wall through the fourth or fifth intercostal space. If an expanded view is needed, the incision can be extended to the right side of the chest.

Once the thoracic cavity has been accessed, the left lung is displaced posteriorly, revealing the left side of the heart and left phrenic nerve. The pericardium is dissected anteriorly and posteriorly to the left phrenic nerve to a depth sufficient to identify the desired plane. Once the plane between the epicardium and parietal pericardium is localized, the pericardium is dissected away, beginning at the left ventricle, and proceeding over to the right ventricle. Finally, the adhesions overlying the pulmonary artery and aorta are removed, freeing the heart.

As previously stated, this approach does not allow for easy access to the right side of the heart. If pericardial adhesions extend to this area, it may be necessary to extend the thoracotomy to the right side of the chest. Then, a similar approach to that used on the left side can be taken to resolve any constrictions.

7.4 Cardiopulmonary bypass

Cardiopulmonary bypass in the surgical treatment of constrictive is not commonly utilized unless additional procedures are to be performed concomitantly and require it. More often, patients undergo the pericardiectomy with the femoral vessels prepared in case emergency bypass, but not as a standard part of the procedure. Having cardiopulmonary bypass at the ready can be useful in cases of extreme blood loss, large calcifications, or accidental damage to the heart during surgery [10].

Some research indicates that the use of cardiopulmonary bypass during a pericardiectomy procedure is an independent predictor of post-procedure complications [89]. It should be noted, though that because the use of cardiopulmonary bypass has traditionally been reserved for more hemodynamically unstable or higher-risk patients, it may not actually be a causative factor in negative outcomes but rather a marker for those already predisposed to such results [8]. Therefore, it seems to largely depend on surgeon preference whether a patient should undergo bypass during pericardial surgery.

8. Post-surgical prognoses

Post-pericardiectomy outcomes have been the subject of much study in recent years. Common avenues of research investigate the relationship between the etiology of constrictive pericarditis and surgical outcomes. As mentioned previously, the most common underlying causes of constrictive pericarditis include tuberculosis infection, previous cardiac surgery, mediastinal radiation, and idiopathic means [2–5, 10–15, 27, 30]. It appears that, despite the common resultant pathophysiology, unique causative etiologies are associated with variable long-term prognoses. One study reported that patients presenting with constrictive disease arising from tuberculosis infection and idiopathic sources tend to experience
longer event-free survival than those having previously undergone cardiac surgery [90]. A second center found that the 5-year survival rate of patients treated for constrictive pericarditis arising from idiopathic causes stood at 79.8%, while those treated post-cardiac surgery or following mediastinal radiation demonstrated rates of 55.9% and 11.0%, respectively [13]. Previous mediastinal radiation, in particular, seems to be implicated with relative frequency in poor post-pericardiectomy outcomes [9].

Underlying etiology is not the only prognostic factor of post-surgical outcomes for constrictive pericarditis. Preexisting illness also seems to be significant contributor to patient prognosis. A retrospective study of patients at the Asian Medical Center found that diabetes mellitus represented an independent risk factor for post-procedure mortality, as did high early diastolic mitral inflow [91]. They also report that the patients who died following pericardiectomy had higher levels of aspartate aminotransferase, smaller left ventricular end-systolic dimension index, and higher early diastolic mitral inflow velocity prior to surgery compared with the patients who survived [91]. Other pre-surgery hemodynamic and structural parameters including reduced left ventricular ejection fraction, right ventricular dilation, central venous pressure, myocardial atrophy or fibrosis, and tricuspid regurgitation also appear to contribute to poor outcomes [41]. Perhaps unsurprisingly, advanced heart failure symptoms (NYHA III-IV) and arrhythmias are also associated with poor outcomes [41]. Likewise, advanced age and patients with end-stage renal disease, coronary artery disease, chronic obstructive pulmonary disease, sepsis, and other severe comorbidities also appear to experience poorer outcomes than other pericardiectomy patients [41, 92, 93].

9. Conclusions

While constrictive pericarditis represents a relatively rare disease process, it provides several diagnostic and treatment challenges. Constriction of the heart within the pericardium negatively impacts ventricular filling, leading to poor hemodynamics which, over time, can result in heart failure. Early diagnosis and management are key to improving patient prognoses and minimizing complications. Diagnosis of this condition requires a high degree of suspicion from the treating physician and a thorough exam. Imaging modalities, including computed tomography and cardiac magnetic resonance imaging, help to differentiate constrictive pericarditis from other conditions that may present with similar exam findings. Constrictive pericarditis responds poorly to medical management and typically requires surgical decortication of the fibrous adhesions holding the pericardium to the heart. The evolution of this procedure from the partial removal of the pericardium to the radical pericardiectomy has led to improved patient outcomes.

Conflict of interest

The authors declare no conflict of interest.
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