Pericardiectomy for Chronic Constrictive Pericarditis: Where are we after 100 Years?

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Abstract

This study attempts to address several controversies that surround an important disease entity, chronic constrictive pericarditis with reference to an optimal surgical approach, the extent of decortication which can be achieved by individual surgical approaches, its optimal surgical timings, the requirement for cardiopulmonary bypass, and the management of postoperative low cardiac output syndrome following pericardiectomy in the present era.

Introduction

Perspective on pericardiectomy for constrictive pericarditis: Indications, surgical timings, surgical approach and postoperative low cardiac output syndrome

Described first, 300 years ago as concretio cordis, Chronic Constrictive Pericarditis (CCP) commands substantial clinical interest because the disease continues to elude the clinicians mimicking restrictive cardiomyopathy, endomyocardial fibrosis and chronic liver disease [1]. Pericardial constriction does have a country-specific pathology [2-6]. Unlike other diseases linked to underdevelopment or inflammation such as rheumatic heart disease, end myocardial fibrosis and aorto-aortitis, CCP has not shown a declining trend despite socio-economic development [2-6].

The aetiology of CCP also has changed during the past few decades leading to diagnostic uncertainties. The major specific causes to be ruled out are tuberculous pericarditis, neoplastic pericarditis and pericarditis associated with a systemic disease including an autoimmune disease [2-7]. Tuberculosis continues to be the leading cause of CCP in developing countries with a reported incidence of 38% to 83% [2-7]. Due to the emergence of drug-resistant strains of tuberculosis in association with AIDS, the prevalence has increased to ≥90% [7]. Tubercular pericarditis may present with dense fibrosis without direct evidence of tuberculosis, similar to other etiologies of CCP. The advent of antitubercular chemotherapy brought down the mortality from 90% to 40% [4,5,7]. In patients with tubercular pericarditis, our policy is to institute anti-tubercular chemotherapy for a minimum period of 12 months [6]. In developed countries, other causes of CCP continues to dominate such as mediastinal radiation and previous open heart surgery [5,8]. Emerging additional causes include iatrogenic origins such as percutaneous coronary interventions, pacemaker insertion and catheter ablation [3]. The reported prevalence of idiopathic CCP has varied from 24% to 61% in Indian studies [6,9].

This condition has posed a diagnostic dilemma since it was first recognized. The problem of misdiagnosis with other disease entities has also not been adequately addressed. No single approach can be used to diagnose all cases of constrictive pericarditis. The diagnostic approach taken should be individualized for each patient. In some patients, the diagnosis may be made on the basis of the history, physical examination and CXR. In other patients, echocardiography, cardiac catheterization and visualization of the pericardium may all be required. The most important diagnostic tool is the clinical suspicion of constrictive pericarditis in a patient with signs and symptoms of right sided heart failure that are disproportionate to pulmonary or left-sided heart disease.

Clinically, it is necessary to differentiate constrictive pericarditis from other causes of right sided heart failure, such as pulmonary embolism, pulmonary hypertension, right ventricular infarction, mitral stenosis and left ventricular systolic dysfunction. Kussmaul’s sign may be positive but it lacks specificity, as it is also seen in patients with restrictive cardiomyopathy, endomyocardial fibrosis, right ventricular failure and tricuspid stenosis [5,10-12]. In constrictive pericarditis ascites appears first followed by pedal oedema. This sequence is one of the cardinal features in CCP. Published literature does not satisfactorily explain the cause of this ‘ascites precox’. Disproportionately high right atrial pressures, hypoalbuminemia secondary to protein losing enteropathy, cardiac cirrhosis,
increased capillary permeability, impedance to lymph flow and disproportionately high atrial natriuretic peptide have been variously implicated as the causative factors for ascites precox.

Although chest radiography as a single non-invasive imaging modality is not helpful in the diagnosis of CCP, certain findings suggest the existence of CCP [13]. In a typical patient with CCP, the cardiac silhouette is not enlarged; however the cardiac silhouette can be enlarged because of a co-existing pericardial effusion or extracardiac masses. Eggshell calcifications or cocoon calcifications can be enlarged because of a co-existing pericardial effusion or postsurgical and post radiation can occur in around 18% of patients with normal pericardial thickness, where-in microscopic examination revealed focal fibrosis, focal calcification or inflammation [25].

Figure 1: A) Intraoperative views of the steps of left modified anterolateral thoracotomy (UKC’s modification) for radical pericardiectomy. The left pleural cavity is entered through fourth intercostal space. B) Left lung is retracted posteriorly with a wet sponge for adequate exposure. Left phrenic pedicle is identified. C, D) Using cautery, a new cleavage plane is created between posterior surface of the sternum and anterior surface of the pericardium. E) The cleavage plane is extended beyond sternum to identify the right phrenic pedicle. F) Using cautery, a new dissection plane is developed between the diaphragmatic pericardium and diaphragm.

Other findings which may be suggestive of CCP include i) abnormal ventricular septal motion, ii) dilatation and absence of collapse of the inferior vena cava and hepatic veins, iii) preserved or increased or medial mitral annulus early diastolic e' -velocity and iv) increased hepatic flow reversal with expiration reflecting the ventricular interaction and the dissociation of the intra-cardiac and intra-thoracic pressures. However, echocardiography was of limited value in the evaluation of pericardial thickening anterior to the right ventricle (RV) and near the right atrio-ventricular groove [16-19].

Using speckle tracking echocardiography, several investigators have demonstrated that left ventricular mechanics in constrictive pericarditis is limited in circumferential rather than the longitudinal direction and vice-versa in restrictive cardiomyopathy [20-22].

Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are superior imaging modalities for detection of pericardial masses, loculated pericardial effusions, pericardial calcification and asymmetric pericardial thickening which may be helpful in determining the optimal surgical approach for pericardial resection [23,24]. Additionally, MRI defines both morphological (left atrial, superior and inferior vena caval dilatation, ventricular elongation, myocardial atrophy, fibrosis) and functional changes (constriction, septal bounce) [24]. However, constrictive pericarditis mostly post-surgical and post radiation can occur in around 18% of patients with normal pericardial thickness, where-in microscopic examination revealed focal fibrosis, focal calcification or inflammation [25].

Despite the difference in pathophysiologic mechanisms of restriction and constriction, considerable overlap is seen in the parameters of these entities. Increased atrial pressures, equalization of end-diastolic pressures, and dip-and-plateau or square root sign of the ventricular diastolic pressure have traditionally been considered hemodynamic features typical of CCP. However, identical hemodynamic pressures traces can be obtained in patients with restrictive cardiomyopathy.

Although cardiac catheterization may not confirm the diagnosis of constrictive pericarditis, Vatikus and Kussmaul demonstrated overall predictive accuracy of three major hemodynamic criteria in the diagnosis of constrictive pericarditis. A difference between right ventricular end-diastolic pressure and left ventricular end-diastolic pressure of 5 mmHg or less, a right ventricular systolic pressure of 50 mmHg or less and a ratio of right ventricular end-diastolic pressure to right ventricular systolic pressure of >1.3 are 85%, 70% and 76% sensitive, respectively for correctly diagnosing constrictive pericarditis. Presence of all three criteria correctly establishes the diagnosis of constrictive pericarditis in more than 90% of patients [26]. Thus, patients today have symptoms and signs of right heart failure disproportionate to left ventricular dysfunction or valvular heart disease. The challenge is to determine whether abnormalities are caused by pericardial restraint, myocardial restriction, or both [26].

Pericardiectomy is the only accepted treatment for CCP. Its origin dates back to 1898, when DeLorme first suggested it. However, the German group Rehn and Sauer Bruch in 1913 performed successful pericardial resection for CCP through a left anterolateral thoracotomy approach [27]. Other surgical approaches for pericardiectomy include Churchill’s approach, left anterolateral thoracotomy, median sternotomy (Holman and Willett’s approach), a U-incision with the base of “U” at the left sternal border (Harrington’s approach) and
bilateral anterolateral thoracotomy [27-29].

Despite the experience spanning over 100 years, there is no fool-proof formula in the published literature to decide the optimal approach for a given patient. The literature is rife with descriptions of pericardiectomy by either left anterolateral thoracotomy or median sternotomy. Despite the effectiveness of surgery, there are disparate opinions regarding the role of corticosteroids in treating tuberculous pericarditis, timing of operation, surgical approach, and extent of decortication and requirement of Cardiopulmonary Bypass (CPB) [27-32]. The efficacy of pericardiocentesis in preventing CCP in pericardial effusion (serous/or hemorrhagic) has been inadequately investigated [32]. The terms “radical”, “total”, “extensive”, “complete”, “subtotal”, “adequate”, “near-total” and partial pericardiectomy have been variably used to describe the procedure, often without precise definition of the limits of pericardial resection [27-32].

Published reports attest to the unpredictable and variable pattern of CCP and lend support to radical decortication. In 2005, for the sake of uniformity, total pericardiectomy was defined as wide excision of the pericardium with the phrenic nerves defining the posterior extent, the great vessels including the intrapericardial portion of superior vena cava and superior vena cava right atrial junction defining the superior extent, and the diaphragmatic surface, including the inferior vena cava right atrial junction defining the inferior extent of the pericardial resection [6]. Constricting layers of the epicardium were removed whenever possible and the atria and venae cavae were decorticated in all cases in this study group. Pericardiectomy was considered partial if both ventricles could not be decorticated completely because of dense my pericardial adhesions or calcification [6]. Radical pericardiectomy was defined as removal of the entire pericardium over the anterolateral, diaphragmatic surfaces of left ventricle, portion of pericardium posterior to the phrenic nerve and the left ventricle and the anterior and diaphragmatic surfaces of RV until the atroventricular groove leaving behind intact left and right phrenic pedicles [6].

Secondly, the importance of unrecognized constricting epicardial (visceral pericardial) peel was described by Harrington in 1944 and successful pericardiectomy requires decortication of the ventricular epicardium and relief of all constricting layers [28]. In a study, the normalization of pressure volume loop was used as an indicator of operative success of pericardiectomy [33].

In 2005, we compared two surgical approaches for the treatment of CCP i.e., median sternotomy and conventional left anterolateral thoracotomy in 395 patients. The surgical approach was primarily based on surgeon’s preference and remained uniform [6]. However, the median sternotomy approach was preferred in the following conditions: (1) annular CCP, (2) extracardiac intrapericardial mass, (3) presence of a gradient between the superior or inferior vena cavae and Right Atrium (RA) of 2 mmHg or greater, (4) calcific pericardial patch compressing the RA and right ventricular outflow tract, (5) previous open heart surgery, (6) circumferential ‘cococon’ calcification of the pericardium, and (7) recurrent CCP after partial pericardiectomy [6]. We demonstrated that the maximum benefit occurs after total pericardiectomy which is best achieved through a median sternotomy and is very difficult through a conventional left anterolateral thoracotomy [6].

While Cardiopulmonary Bypass (CPB) may not be necessary for effusive or inflammatory pericarditis, it does all depend on how the patient tolerates cardiac manipulation; likely the most important reason to use CPB in order to facilitate a complete pericardiectomy because we know that this is more favourable in terms of long-term functional outcomes compared to a partial pericardiectomy. Studies in which CPB was associated with lower survival and higher risk including the Stanford series, demonstrate that this is a reflection of a more advanced disease process when CPB is needed [30]. So, we should not be reluctant to utilize CPB if needed to facilitate a complete resection with the thought process that this will lower survival. If it enables a more complete resection, this will undoubtedly impact patient’s outcome more favourably compared to the use of CPB lowering survival. Although routine use of CPB to achieve total pericardiectomy was an issue of debate, it requires to be employed in special circumstances, namely (1) inadvertent damage to a cardiac chamber, (2) cardiac operation, or previous partial pericardiectomy, (3) presence of calcific pericardial “cococon” encompassing all cardiac chambers, (4) pericardiectomy following mediastinal irradiation and (5) coexistent cardiac lesion [6,13,14,30,34,35].

However, a left anterolateral thoracotomy was the preferred approach in cases of purulent pericarditis and effusive constrictive pericarditis is because of the presence of concomitant pyothorax and the concerns of sternal infection. We could achieve total pericardiectomy in these patients because of loculations and flimsy...
adhesions which could be easily peeled-off [6,34,35].

**Criteria for decision-making on the timing of operation and selection of surgical approach for patients undergoing pericardiectomy**

Several investigators including ourselves advocate early pericardiectomy before the occurrence of severe constriction and myocardial atrophy [3-7,9,13-15,30,31,34,35]. In our previous study, we compared the outcomes after total vs. partial pericardiectomy. Our study demonstrated that total pericardiectomy was associated with lower operative mortality and Low Cardiac Output Syndrome (LCOS), abbreviated hospitalization, and better long-term survival than partial pericardiectomy. Ascites, low ejection fraction (0.40 or less), renal dysfunction, hyperbilirubinemia, high preoperative RA pressure (>24 mmHg), atrial fibrillation, pericardial calcification, tricuspid regurgitation, mitral regurgitation, partial pericardiectomy, thoracotomy approach and postoperative LCOS negatively affected survival. In this study, the risk of death was 4.5 times higher (95% CI: 2.05 to 9.75) in patients undergoing partial pericardiectomy compared to total pericardiectomy [6].

Despite total pericardiectomy, the operative mortality rate was 7.6% in our series and 6% to 19% in several large series published after 1985 [3,6,14,15,29,31,34,35]. Unlike others, there was no correlation with age, tuberculous etiology and advanced NYHA symptoms on late survival, presumably because of young patient population and timely institution of chemotherapy and surgery [6,15,34,35].

Although the median sternotomy approach allowed a more radical clearance of pericardium overlying the RA and venae cava including the cavaatrial junctions, these areas usually are of little hemodynamic significance in the majority of patients. Furthermore, it is impossible to excise the portion of the pericardium posterior to the phrenic nerve using this approach [6,15,34,35].

**Criteria for decision-making and selection of surgical approach for patients undergoing radical pericardiectomy via left anterolateral thoracotomy without cardiopulmonary bypass (UKC’s modification)**

As enunciated above, the median sternotomy approach was the preferred option of the author (UKC) in the selected heterogeneous group of patients undergoing pericardiectomy [6,15]. In an effort to decrease the hospital mortality rates and postoperative LCOS, the author proceeded to perform several technical modifications of the conventional left anterolateral thoracotomy approach to achieve further radical excision of the pericardium posterior to the phrenic nerve and diaphragmatic pericardium without utilizing CPB [34,35]. Thus, there were seven forces driving our decision-making towards improvement of the results after pericardiectomy via modified anterolateral thoracotomy.

- The desire to obtain improved operative exposure of the RV and RA by developing a new dissection plane between the posterior surface of the sternum and anterior surface of the pericardium.
- The desire to dissect the pericardium posterior to the phrenic nerve overlaying the left atrium and posterolateral surface of the left ventricle.
- The desire to develop a new cleavage plane between the diaphragmatic pericardium and diaphragm.
- The desire to minimize cardiac manipulation at the time of dissection by dividing the anterior and posterior pericardial flap in two halves respectively.

- The desire to minimize postoperative auto transfusion by inserting a peritoneal dialysis catheter before surgical incision and placing it on gravity drainage intraoperatively.
- The desire to maintain oxygenation and hemodynamic stability during pericardiectomy via left anterolateral thoracotomy by placing an intercostal chest drain on the opposite side in case of right-sided significant pleural effusion.
- The desire to keep both groins prepared at the time of pericardiectomy via modified left anterolateral thoracotomy in case of inadvertent injury to the cardiac chambers and/or great vessels and urgent institution of CPB.

The step-by-step technical details of the conventional median sternotomy (n=55) and the authors modification of the left anterolateral thoracotomy (n=67) to achieve radical pericardiectomy without utilizing cardiopulmonary bypass have been alluded to in our previous publications. The following specific maneuvers facilitated performance of radical pericardiectomy via modified left anterolateral thoracotomy (Figures 1A-F and 2A-F):

1. Development of a new cleavage plane between the sternum and the anterior surface of the pericardium using cautery and a right angled deep blade sternal retractor.
2. Extension of the dissection plane beyond the mid sternum to the right phrenic pedicle.
3. Development of a new cleavage plane between the diaphragmatic pericardium and diaphragm.
4. Dissection of the pericardium posterior to the left phrenic nerve and division of the posterior pericardium in two halves.
5. Dissection of the pericardium anterior to the phrenic nerve, division of the anterior pericardium in two halves and detachment of the anterior pericardium 1 cm away from the right atrio-ventricular groove.

Using these modifications, radical pericardiectomy was associated with a further reduction of operative mortality as compared to total pericardiectomy of our initial publication (2.9% vs. 7.6%) and patients undergoing total pericardiectomy of our second publication (2.9% vs. 7.2%) [6,15,34,35]. By employing these modifications, we have been able to reduce the incidence of postoperative Low Cardiac Output Syndrome (LCOS) from 69% (total pericardiectomy) to 26.8% (radical pericardiectomy) [6,15,34,35].

Despite improved accuracy of diagnosis with echocardiography, and computed tomography often obviating the need for cardiac catheterization, aggressive preoperative stabilization, improvements in cardiac anaesthesia, perioperative intensive care, hemodynamic monitoring and advances in surgical techniques during the past 100 years, pericardiectomy for CCP continues to be associated with mortality ranging from 6% to 19% [3,6,14,15,29,31,34,35]. In 2005, we reported worse outcomes of pericardiectomy in patients with preoperative high RA pressure >24 mmHg Hyperbilirubinemia, renal dysfunction, atrial fibrillation and pericardial calcification [6].

Careful analysis of the published literature substantiates significant incidence of LCOS following decortication of the pericardium. The culprit patho-physiological mechanisms responsible for LCOS
immediately following pericardiectomy although not specifically addressed or analysed is multifactorial in nature. Immobilisation atrophy, myopericardial involvement by the same pathologic process, imperfect or incomplete decortication, remodelling of the ventricle, abnormal diastolic filling characteristics, worsening tricuspid regurgitation and postoperative mitral regurgitation secondary to papillary muscle elongation have been variously implicated as the causative factors for LCOS [36-39]. Several investigators including ourselves have observed that regardless of the operative approach or extent of pericardial resection, a subset of patients with chronic CCP will develop LCOS.

The hemodynamic hallmark of CCP is impairment of ventricular diastolic compliance. On completion of a successful pericardiectomy, there are major fluid shifts from extravascular to intravascular compartments. We had previously demonstrated that this auto transfusion results in failure of Frank-Starling mechanism causing acute cardiac dilatation and this almost mimics acute LV dysfunction from volume overload [6,15]. We often see some worsening of valve function due to acute stretching of the annuli resulting in functional regurgitation. Studies have shown, that massive ascites was a significant negative factor for survival [6,15].

Additionally, due to repeated mechanical compression during the process of pericardial mobilization, there is myocardial oedema, which subsides over time [6,15,34,35]. It is indeed impossible to pinpoint a specific causative factor for LCOS following pericardiectomy. Although high right atrial pressure and atrial fibrillation are associated with poor outcomes, we do not advocate an aggressive surgical approach to treat tricuspid regurgitation or atrial fibrillation at the time of pericardiectomy. In these patients, the use of CPB allows one to control these fluid shifts and ultra filtrate some of this fluid off. So, this may be a concept that is not appreciated the use of CPB to avoid cardiac distension. If the cardiac output cannot be sustained by the currently available medical treatment, the next strategy may be to assist the failing heart by mechanical circulatory assistance.

Although the use of Intra-Aortic Balloon Counterpulsation (IABC) is universal in adults with acute left ventricular dysfunction after myocardial infarction or cardiac surgery, its use in patients undergoing pericardiectomy for chronic CCP remains sporadic [40]. Intra-Aortic Balloon Counterpulsation (IABC) facilitates recovery of left ventricular function by decreasing left ventricular end-diastolic and left atrial pressure, thus helping the systemic ventricle and indirectly helping the pulmonary ventricle by the phenomenon of ventricular interdependence [40,41]. The advantages of IABC over left atrial-aortic assist devices is the ease of application [40,41]. Other assist device like axial flow pumps and veno-arterial extracorporeal membrane oxygenation as a salvage procedure [42,43]. Use of IABC for supporting the failing myocardium also remains limited in small children, along with sparse availability and inability to track rapid heart rates and narrow pulse pressures of children in shock. Additionally, complications like limb ischaemia, mesenteric ischaemia and renal failure are greater in children because of inappropriate balloon length [41].

The insertable balloon lengths of the commercially available intra-aortic balloon catheters are 16.5 cm, 22.1 cm and 25.8 cm for 25 CC, 34 CC and 40 CC balloons respectively. The pediatric patients with preserved RV and pulmonary function requiring mechanical circulatory assistance fulfilling the above-mentioned mandated insertable balloon lengths may be candidates for IABC. The timing and indications of balloon deployment is a matter of judgment. In patients who suddenly deteriorate after total pericardiectomy and are unresponsive to medical therapy, the decision to initiate IABC is relatively straightforward. The other clinical scenario would be in cases of progressive deterioration of ventricular function and unresponsive to adequate inotropic support [40].

The decision to address this disease entity is based on the belief that surgery for chronic CCP is safe and reproducible, it is therefore teachable. I recognize that the techniques and concept of others may be different from ours and may give results that are as good. Yet, I am confident that the concepts and techniques reflected in this manuscript, if carefully followed will deliver excellent results for the surgical treatment of CCP. No single laboratory tests or diagnostic findings should be considered pathognomic of CCP. A combination of clinical and investigative results should be thoughtfully analysed to diagnose this disease entity. The most important diagnostic tool is clinical suspicion of CCP in patients with signs and symptoms of right-sided heart failure that are disproportionate to pulmonary or left-sided heart disease. The extent of contribution by the researchers in dealing with this disease is reflected in the large bibliography in the Medline search. Through the usual scientific channels, all of us have pooled our knowledge in such a way that we have built upon the shoulders of each other in preparing this manuscript.

References


