

# Prognostic implications of pulmonary hypertension due to constrictive pericarditis

## Abstract

Constrictive pericarditis (CP) is a pathological process leading to scarring, fibrosis and thickening of the pericardial layers with subsequent compression of the cardiac chambers, resulting in diastolic dysfunction. Pericardiectomy remains the only acceptable and curative surgical procedure in most CP patients. Pulmonary hypertension (PH) is defined as a mean pulmonary arterial systolic pressure greater than 25mmHG at rest and 30mmHg during exercise. The prevalence of PH in CP has previously been reported to be rare; as a result the main aim of this paper is to extensively review the prevalence of PH in CP patients.

**Keywords:** constrictive pericarditis, mean pulmonary arterial systolic pressure, pericardiectomy

Volume 9 Issue 5 - 2017

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Received: June 11, 2017 | Published: September 8, 2017

**Abbreviations:** CP, constrictive pericarditis; LVEF, left ventricular ejection fraction; mPASP, mean pulmonary arterial systolic pressure; PH, pulmonary hypertension; RV, right ventricle; TR, tricuspid valve

## Introduction

Constrictive pericarditis (CP) is commonly a symmetrical pathological process from scarring, fibrosis and thickening involving the parietal and visceral pericardial layers; due to its compressive phenomenon on the cardiac chambers, CP results in diastolic dysfunction.<sup>1-9</sup> Although CP is regarded as a chronic process, some patients may develop mild pericardial constriction which may resolve spontaneously within few weeks or months following the initial insult. Constrictive pericarditis is thought to be a rare cause of pulmonary hypertension (PH); as a result the main objective of this paper is to review PH due to CP.

## Overview of constrictive pericarditis

**Etiology of CP:** Tuberculous pericarditis remains an important cause of CP in the developing world, however in developed countries prior pericarditis, idiopathic, prior cardiac surgery and radiotherapy in particular prior mediastinal radiation exposure are the common causes of CP. Other causes should also be evaluated which include systemic inflammatory or connective tissue disease, uremia particularly in patients with chronic renal failure or end-stage renal diseases, malignancy etc.

**Clinical characteristics of CP:** During the early stages of CP, patients may remain completely asymptomatic and clinical signs may be subtle. However, in patients with established or advanced CP, the typical presentation is similarly to that of right heart failure. Where patients may present with lower extremity edema, elevated jugular venous pressure which is classically characterized by a prominent y-descent and Kussmaul sign.

**The role of echocardiography in CP:** Echocardiography is the most easily accessible, cheap, non-invasive and readily available in almost every center; and is useful for both diagnostic and could also assist to exclude possible differential diagnoses. Echocardiography has evolved extensively in the recent past, with the introduction of tissue

Doppler and speckle tracking imaging modalities which are useful in guiding treatment and also have important prognostic implications in CP patients.

**Differential diagnosis of CP:** although restrictive cardiomyopathy has been widely reported as the main mimic for CP, other possible diagnosis should be entertained during the initial evaluation of patients suspicious of constriction, and these include dilated cardiomyopathies, pericardial effusion and cardiac tumours.

**Treatment modalities for CP:** Although the primary cause of constriction should be evaluated and managed accordingly, surgery in the form of a complete pericardiectomy remains a definitive and curative management strategy in patient with chronic CP.

## Pulmonary hypertension related to constrictive pericarditis

**Demographics characteristics and risk factors of pulmonary hypertension in constriction:** Data on the prevalence and risk factors of PH in CP are still rather limited. The factors presumed to be associated with the occurrence or severity of PH in patients with CP include: old age, prior cardiac surgery, pre-existing PH, severity and chronicity of PH at the time of surgery, primary etiology of CP particularly prior mediastinal radiotherapy, collagen vascular or connective tissue diseases, chronic respiratory diseases and pre-existing myocardial diseases.<sup>2,3,10</sup> Talreja et al.<sup>11,12</sup> reported comparison of etiologies between groups with PH and showed that prior cardiovascular surgery (18% vs 38%) and irradiation (13% vs 19%) were more common in the PH group with mean pulmonary arterial systolic pressure (mPASP) of greater than 50mmHg, while idiopathic (likely viral) etiology was more common in the PH group with mPASP of less than 50 mmHg. Although left ventricular (LV) systolic dysfunction and the presence of valvular lesion are presumed to be among the leading causes or risk factors for PH in CP; Talreja et al.<sup>11,12</sup> reported no difference in LV ejection fraction (56% vs 58%) or frequency of significant mitral incompetence (2% vs 0%) or TR (5% vs 4%), in patients with mPASP >50mmHg versus <50mmHg. However, more data is warranted to address these controversies.

**Mechanisms of pulmonary hypertension in constrictive pericarditis:** In the true sense mPASP and right ventricular systolic pressures are estimated to be normally between 35 and 45mmHg;

however mPASP may be as low as possible reflecting reduced RV stroke or pressure and sometimes mildly elevated if the left atrial pressure has caused a secondary increase in pulmonary vascular resistance. In rare instances localized constriction can cause external valvular constriction or pinching of the epicardial coronaries. Up until recently, there has been case reports in patients with severe PH in the setting of CP.<sup>10</sup> Severe PH has been reported as a rare feature of CP and may indicate co-existing cardiac or pulmonary disease, particularly in patient with prior cardiac surgery or radiotherapy.

Talreja et al.<sup>13</sup> reported a case of severe PH due to CP in a 58years old female, who had a reversible constriction associated with chronic pleural effusion and hypoxemia.<sup>10</sup> The magnitude of mPASP improved significantly before and after pericardiectomy following the initiation of medical treatment with sildenafil and nitric oxide. This approach remains controversial as there is a paucity of evidence regarding the optimal medical management of PH secondary to CP. Treatment of PH in CP patients using pulmonary vasodilators may be hazardous given their potential to increase LV filling pressures further, thus worsening pulmonary edema. Brunner et al.<sup>10,14</sup> reported a case where there was deterioration in a patient's condition after the initiation of sildenafil.

**Impact of pericardiectomy on pulmonary hypertension in constriction:** Pericardiectomy remains the only curative treatment for CP, irrespective of the chronicity or severity of constriction. Reports have highlighted the impact of pericardiectomy on PH in patients with surgery or autopsy confirmed CP.<sup>3</sup> In the same reports pericardiectomy significantly improves the level of PH in those who survived surgery.<sup>3</sup> However, long-term follow-up data on patients undergoing surgery with pre-operative severe PH are still limited, as a result this opens a window of opportunities to further study these patients in long-term.

**Prognosis in constriction patient with pulmonary hypertension:** Long-term survival after pericardiectomy for CP is related to the underlying etiology, left ventricular systolic function, renal function, serum sodium and pulmonary pressures. High pulmonary pressures may reflect the severity of constriction, concomitant myocardial dysfunction or pulmonary pathology. Talreja et al.<sup>11</sup> reported data of 224 patients with surgically or autopsy confirmed CP, where 20% of these patients had PH. Pericardiectomy significantly improved the level of mPASP and survival in those who survived the surgical procedure. Improvement on survival for those with PH was almost similar to their counterparts with normal preoperative mPASP. Furthermore, Talreja et al.<sup>11</sup> demonstrated a significantly higher perioperative or 30 day mortality particularly in the group with higher mPASP, mostly the group with mPASP of more than 50 mmHg. This group consisted mainly of patients with prior cardiac surgery or mediastinal radiotherapy.

### Future directions

Each patient with PH presumed to be due to CP should be properly evaluated and managed individually as the presence of PH in CP may declare poor prognostic in some patients. Data are still limited on mechanisms and outcome; as a result a window of opportunities is opened for future research to address many unanswered questions related to the pathological mechanisms and long-term implications of PH in CP irrespective of the primary etiology of CP.

### Conclusion

Although the prevalence PH in CP is presumed to be rare, PH due to CP could be a reversible entity with use of vasodilatation and pericardiectomy. Pericardiectomy seems to improve the level mPASP

and survival in those who survive surgery, similarly to those with normal baseline mPASP; however larger studies with longer follow-up are still warranted.

### Acknowledgments

None

### Conflicts of interest

There were no financial interest or conflict of interest.

### Funding

None.

### Disclosures

None related to this study.

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