

# Revisiting the Diagnostic Criteria of Constrictive Pericarditis

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## Background

- Constrictive pericarditis (CP) is a form of diastolic heart failure that arises from an inelastic pericardium inhibiting cardiac filling
- Men ages 16–65 years are at a higher risk for pericarditis than women with the highest risk difference among young adults compared with the overall population
- Risk factors for CP include prior cardiac surgery, radiation, trauma, drugs/toxins, metabolic disorders, rheumatologic diseases, and gastrointestinal diseases. However, most cases are considered idiopathic/viral
- Significant physical exam findings are signs of volume overload such as jugular venous pressure (JVP) elevation, symptoms of decreased cardiac output such as edema, pericardial knock, and Kussmaul's sign
- CP is a diagnosis of exclusion that requires extensive cardiac work-up as the definitive cure requires a pericardiectomy which holds a significant perioperative mortality rate ranging from 6 to 12%

## Purpose

The purpose of this case report is to increase clinician awareness of the clinical findings of constrictive pericarditis.

## Case Description

- 50-year-old man sent to the ED from pulmonology clinic due to bilateral pleural effusions seen on outpatient CT chest. Presenting with dyspnea on exertion and chronic cough productive of clear sputum
- Past medical history: only significant for pneumonia 4 months prior to symptoms onset
- Vitals: T 97.8, HR 90, BP 112/76, RR 20, O2 98% on room air, BMI 23.6
- Physical exam findings: Mild bilateral lower extremity edema, Crackles in mid and lower lung fields bilaterally, occasional extra heart sound prior to diastole on auscultation, JVP 9cm H2O, JVP elevation upon inspiration as observed on point-of-care ultrasound
- Underwent bilateral thoracenteses twice and the pleural studies pointed towards a transudative etiology and was negative for malignant cells
- Pertinent cardiac imaging included septal bounce and moderate pericardial effusion on TTE, equalization of pressures in the left and right ventricular end-diastolic pressures on right heart catheterization, and borderline thickened pericardium on Cardiac MRI
- HIV, hepatitis panel, and TB spot test negative. TSH normal.
- Extensive rheumatologic workup revealed only +ANA 1:40, weakly positive RNP IgG, and indeterminate anticardiolipin IgM
- Discharged on conservative management with colchicine to decrease inflammation as an initial treatment while further work-up is done in the multispecialty clinic prior to definitive CP treatment
- In the outpatient setting, he re-developed bilateral pleural effusions thrice within three months and, contrary to prior pleural studies, the latest appeared milky white and contained chylomicrons.
- Referred to interventional radiology for transjugular biopsy due to concern for cirrhosis as the etiology for the chylothorax given the patient's >20 year alcohol history
- Biopsy was negative for steatosis, active steatohepatitis, or cirrhosis. Instead, it showed focal centrilobular and mid-zonal sinusoidal congestion, which is consistent with hepatovascular outflow insufficiency related to the patient's clinical history of cardiomyopathy.
- At the most recent follow-up appointment, the next course of action was evaluation of the thoracic duct and then possible pericardial stripping.

## Discussion

This patient's imaging and physical exam findings were suggestive of CP, and the pneumonia he contracted previously may have been the inciting factor if it were viral. The development of a chylothorax is also interesting as there have been case reports that attribute CP as a rare etiology. Thus, the formation of a chylothorax may add further evidence of this pt having CP. However, despite these classic findings for CP, it is important that other causes are first ruled out due to the high mortality rate of a pericardiectomy. One etiology that must be ruled out is in fact acute pericarditis with constriction which is caused by inflammation that resolves once the inflammation is treated. As a result, before a pericardiectomy is recommended, a trial of anti-inflammatory medications for 2-3 months is an important consideration especially for those with constrictive features early in the course of their illness and are hemodynamically stable. On the other hand, delayed time until a pericardiectomy is performed has been attributed to longstanding disease with myocardial atrophy or fibrosis.

## Management & Complications

The only definitive treatment option for patients with chronic constrictive pericarditis with persistent and prominent symptoms is a pericardiectomy, also referred to as pericardial stripping. In one series of patients who underwent surgery between 1970 and 1985, the operative mortality was 12 percent. A lower mortality rate of between 4-8% was noted in patients who underwent pericardiectomy between 1977 and 2012. Furthermore, the 5-year survival rate post-op was 29%. However, long-term prognosis after a pericardiectomy depends on the etiology of CP and is best for idiopathic constriction but worst for radiation constriction. On immediate follow-up after surgery, most patients had improved symptomatically but some re-developed recurrent heart failure symptoms. Right-sided heart failure after pericardiectomy can be caused by incomplete pericardiectomy, recurrent constriction due to scar tissue, or extension of pericardial calcification into the myocardium. Additionally, progressive tricuspid annular dilatation/regurgitation is a possible complication from the surgery that can lead to poorer outcomes.

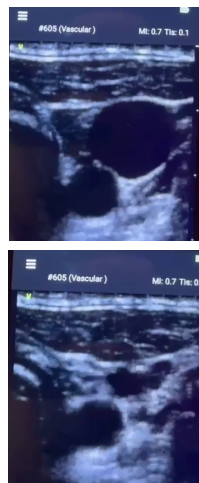


Figure 1: Increase in jugular venous pressure during inspiration (top) and decrease with expiration (bottom)

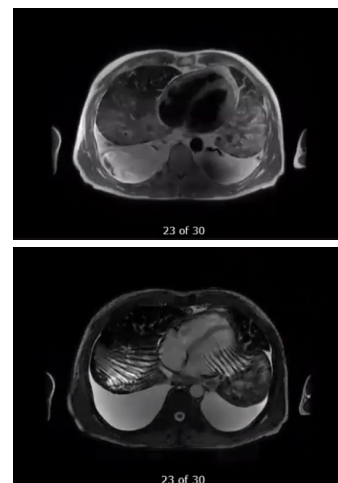


Figure 2: Single-shot fast spin-echo view (top) and Fast Imaging Employing Steady-state Acquisition view (bottom) on Cardiac MRI

## Conclusion

### Implications for Clinical Practice:

- Constrictive pericarditis has notable physical exam findings that should raise concern for CP if found. Extensive lab work and imaging should be performed in order to rule out other diagnoses before committing to a CP diagnosis.

### Implications for Future Research:

- It would be beneficial to patients if medications developed in the future would be able to prevent the pericardial lining from increasing and thus decrease the need for a pericardiectomy for mild cases. Severe cases would most likely continue to require a pericardiectomy.
- Furthermore, for patients that present with risk factors for CP, there may be some utility in researching screening methods to detect CP earlier prior to obtaining clinical symptoms.

## Contact Information

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