Constrictive Pericarditis

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KEYWORDS

• Constrictive pericarditis • Heart failure • Cardiac MR • Echocradiography

KEY POINTS

- Constrictive pericarditis is a disorder of cardiac filling caused by a diseased, inelastic pericardium that restricts cardiac chamber expansion.
- Patients present with heart failure symptoms and signs, including dyspnea on exertion, increased venous pressure, and edema.
- Key pathophysiologic features include dissociation of intrathoracic and intracardiac pressures and enhanced ventricular interaction.
- Key echocardiographic features include respiration-related changes in the position of the ventricular septum and Doppler velocities; the myocardium exhibits unique diastolic properties and systolic strain.
- Invasive hemodynamic assessment is the gold standard diagnostic test and requires careful simultaneous recordings of right and left ventricular pressures, ideally with high-fidelity manometer-tipped catheters.

INTRODUCTION

Constrictive pericarditis masquerades as diastolic heart failure, but is distinct in terms of its cause and treatment. A diseased, inelastic pericardium restricts cardiac filling and leads to unique hemodynamic derangements. Recognizing this disorder requires an index of suspicion, a careful history and physical examination, meticulous cardiac imaging, and often invasive hemodynamic assessment. Accurate identification of constrictive pericarditis markedly changes the therapeutic plan for the affected patient. A minority may find relief through antiinflammatory therapies. Others require and benefit from surgical pericardiectomy.

ETIOLOGIES

Tuberculosis was historically the most common cause for constrictive pericarditis in North America; a report from 1962 cited tuberculosis as the cause of 48% of cases of constrictive pericarditis.[1] Worldwide, this likely remains the case, particularly in areas where human immunodeficiency virus and AIDS are most prevalent. A South African institution reported 121 cases of constrictive pericarditis over 22 years (1990–2012) and of these, tuberculosis was confirmed as the cause in 29.8% of cases and suspected in an additional 61.2% of cases.[2]

In North America and Europe, tuberculosis is now a relatively rare cause of constrictive pericarditis, with reports ranging from less than 1% to 5.6% of cases.[3–6]

- The 3 dominant etiologies reported now in North America and Europe are idiopathic, prior cardiac surgery, and radiation therapy.

For example, the Mayo Clinic reported 135 cases over 10 years (1985–1995) and classified 80% as being idiopathic or owing to prior cardiac surgery, acute pericarditis, or radiation therapy.[3] Also noted was an increase in frequency of cases

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owing to prior cardiac surgery or radiation therapy compared with an historic cohort. The remaining cases were due to rheumatologic disease, infection, malignancy, trauma, asbestosis, drug-induced causes, and complicated pacemaker lead replacement. Other presumably rare causes of constrictive pericarditis include immunoglobulin G4-related disease and Whipple's disease.[7,8]

EPIDEMIOLOGY

Constrictive pericarditis seems to be a relatively rare disorder. Neither the prevalence nor overall incidence of constrictive pericarditis is known. A specific etiologic type, constrictive pericarditis occurring after an episode of acute pericarditis, has been studied and has a low incidence of 1.8%.[9] Constrictive pericarditis is less common after idiopathic or viral pericarditis than after pericarditis owing to connective tissue disease, pericardial injury syndrome, neoplasm, or bacterial infection.

PATHOLOGY

Pathologic changes in constriction most commonly affect the parietal pericardium, but may also affect the visceral pericardium and even the underlying epicardium. In chronic constrictive pericarditis, which develops over months to years, the pericardium is typically fibrotic and calcified. Thickening of the pericardium is often found, but is not required for the disorder to occur. In fact, the thickness of surgically removed constrictive pericardium has been reported to be normal in 18% of cases.[10] In subacute constrictive pericarditis, which develops over days to weeks, inflammation seems to be the dominant pathologic abnormality. These cases of constrictive pericarditis are more likely to be transient and medically treatable.

PATHOPHYSIOLOGY

The diseased pericardium loses its reserve volume and begins to restrict cardiac chamber expansion. Two fundamental pathophysiologic principles are recognized when this occurs:

1. Dissociation of intrapleural and intracardiac pressures, and
2. Enhanced ventricular interaction.[11]

The principle of dissociation of intrapleural and intracardiac pressures is due to the insulating effects of the diseased pericardium on the cardiac chambers, such that intracardiac pressures no longer change to the same degree as intrapleural pressures during the respiratory cycle.

- In inspiration, intrapleural pressure decreases, but left atrial pressure decreases more modestly, if at all. This reduces the pressure gradient from pulmonary veins (which are intrapleural) to the left atrium, which in turn reduces left heart filling. The interventricular septum shifts toward the left ventricle and right heart filling is favored, which illustrates the principle of enhanced ventricular interaction (**Fig. 1**).
- During expiration, these phenomena are reversed: intrapleural pressure increases, the pressure gradient between the pulmonary veins and the left atrium is restored, left heart filling increases, the interventricular septum shifts to the right, and right heart filling decreases. The principles of dissociation of intrapleural and intracardiac pressures and

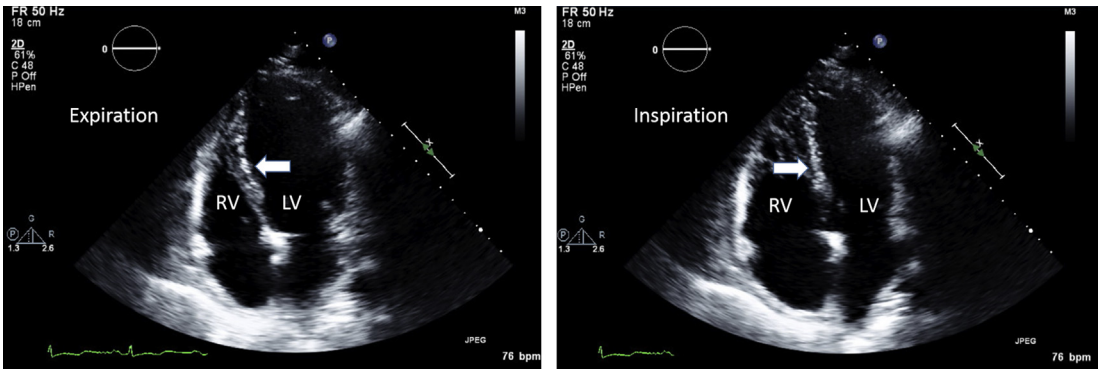

Fig. 1. Enhanced ventricular interaction in constrictive pericarditis. Note the respiration-related shift in the position of the interventricular septum (and relative sizes of the right and left ventricles) in these apical 4-chamber images. The septum shifts (*arrows*) toward the right during expiration (*left panel*) and toward the left during inspiration (*right panel*). LV, left ventricle; RV, right ventricle.