Effusive-Constrictive Pericarditis

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KEYWORDS

• Effusive-constrictive pericarditis • Pericardiocentesis • Echocardiography

KEY POINTS

- In effusive-constrictive pericarditis (ECP), a hemodynamically significant pericardial effusion coexists with decreased pericardial compliance.
- The hallmark of ECP is the persistence of elevated right atrial pressure postpericardiocentesis.
- The reported prevalence of ECP has varied significantly, but it seems higher in tuberculous pericarditis and lower in idiopathic cases.
- A diagnosis of ECP is traditionally based on invasive hemodynamics. The presence of echocardiographic features of constrictive pericarditis postpericardiocentesisis, however, can detect ECP.
- Data on the prognosis and optimal treatment of ECP are still limited. Anti-inflammatory therapy should be the first line of treatment. Pericardiectomy should be reserved for refractory cases.

INTRODUCTION

The coexistence of pericardial effusive and constrictive features was first brought to attention more than 50 years ago[1,2] and was also described by Paul Wood in the subacute phase of tuberculous pericarditis.[3] Better characterization of this clinical entity did not occur until the series reported by Hancock[4] and Sagrista-Sauleda and colleagues.[5] Almost 15 years later, the understanding and appreciation of effusive-constrictive pericarditis (ECP) is still incomplete, with data limited to case reports,[6–9] small case series,[4] and only a few prospective studies.[5,10–12] This review summarizes the available data on ECP and describes the authors' own clinical experience.

PATHOPHYSIOLOGY

In ECP, a hemodynamically significant pericardial effusion coexists with decreased pericardial compliance. Consequently, as the cardiac compression is relieved by pericardial fluid drainage and intrapericardial pressure falls, the features of constrictive pericarditis (CP) become predominant. This is manifested by the persistence of elevated right atrial pressure after pericardiocentesis, the hemodynamic hallmark of ECP.[13]

Although cardiac tamponade and CP are both pericardial disorders leading to abnormal cardiac filling, their pathophysiologies are markedly different. In tamponade, early diastolic filling is significantly impaired due to elevated intrapericardial pressures. Systemic venous pressure rises and adrenergic tone increases to overcome the decrease in cardiac output. If tamponade continues to progress and compensatory mechanisms are overwhelmed, shock ensues unless pericardiocentesis is performed. In contrast, in CP early diastolic filling prevails — the increase in filling pressures secondary to the inelastic pericardium promotes increased ventricular filling in early diastole. As the pericardial reserve is exhausted in mid to late diastole, however, ventricular filling markedly decreases. These differences lead to their classic hemodynamic features[14,15]: blunting of the *y* descent in tamponade

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and deep, rapid ventricular filling waves in CP (square root sign or dip-and-plateau pattern).

Although ECP is frequently thought of as the conversion from tamponade to constrictive physiology after pericardiocentesis, ECP cases represent a spectrum of patients with pericardial effusions, elevated intrapericardial pressures, and decreased pericardial compliance. Therefore, patients with ECP have hemodynamics that fall on a continuum between pure effusive tamponade on the one hand and chronic CP on the other. This accounts for the clinical variability and for lack of full understanding of the underlying pathophysiology, because ECP is actually part of a clinical spectrum. This spectrum is evident in Sagrista-Sauleda and colleagues' series[5] where, despite all patients having clinical tamponade and documented decreased right atrial transmural pressure, right atrial contours pre-pericardiocentesis were not typical of tamponade. Hence, despite the elevated intrapericardial pressures and low right atrial transmural pressure (as seen in tamponade), ECP has different underlying hemodynamics. This was better exemplified by Hancock,[4] who showed that right atrial contour in ECP differed from both tamponade and CP, with the relationship between *x* and *y* descents in ECP intermediate between the 2 entities. Lastly, in the authors' experience, overt tamponade is not a mandatory component of ECP. A similar observation was reported by Ntsekhe and colleagues[16] in a series of 68 patients with tuberculous pericardial effusions; tamponade confirmed by cardiac catheterization was present in only 56% of patients. There was no difference in the prevalence of tamponade between ECP and non-ECP groups in their cohort. Patients with ECP, however, had higher right atrial pressures, suggesting that the hemodynamic effects of the effusion and reduced pericardial compliance are additive.

Bloody effusions are common in patients with ECP[4,5] and it has been postulated that the presence of blood in the pericardial space might promote more exuberant inflammation and decreased pericardial compliance. Patients with tuberculous ECP were found to have higher levels of interleukin 10 and interferon gamma in the pericardial fluid as well as serum interleukin 10 and tissue growth factor β compared with patients with non-ECP tuberculous pericarditis.[16] These cytokines may foster pericardial inflammation/fibrosis and be related to higher prevalence of ECP in tuberculous pericarditis. In a series of patients with nontuberculous pericardial effusions, the authors observed that, although no differences in pericardial total white blood cell count was seen, the percentage of neutrophils was higher in ECP patients. This also suggests more florid inflammation in these patients.[17] Although the hemodynamic derangements in ECP are believed secondary to constriction from the visceral pericardium,[18] findings at time of pericardiectomy are of thickening of both visceral and parietal pericardia, with associated pericardial fluid and adhesions.[4,5,13] Pericardial edema with active inflammation might also be seen, whereas pericardial calcifications are not typical.

EPIDEMIOLOGY

The reported prevalence of ECP among patients with a pericardial effusion has ranged widely, from 1% to 2%[5,19] to more than 50%.[10,11] This variation can be attributed to differences in methodology (diagnosis of ECP based on catheterization vs echocardiography) and the cohorts studied. It is clear, however, that the prevalence with ECP is closely related to the associated etiologies.[13] ECP seems much more prevalent in tuberculous pericarditis than in idiopathic pericarditis. In a series of 68 patients with tuberculous pericardial effusions, 53% were diagnosed with ECP.[13] Similar results have been reported in other groups of patients with tuberculous pericarditis.[11] In contrast, in a single-center European series, ECP was diagnosed in 1.3% of patients undergoing pericardiocentesis, corresponding to 7.9% of tamponade cases.[5] The most common etiologies were idiopathic, neoplastic, and postradiation, in order of frequency. These etiologies are in agreement with observations from a North American center,[18] in which idiopathic cases corresponded to the vast majority of ECP patients identified, followed by radiation-heart disease. This epidemiologic profile would also be akin to the one currently observed for CP in Europe and the United States, where idiopathic and radiation are among leading causes. Compared with CP in developed countries, however, the lack of postoperative/postprocedural cases in these 2 ECP series differentiates the populations and may be related to lower volumes of cardiac surgery and percutaneous interventions during the study periods (4 decades and 2 decades ago, respectively). In a series of 205 consecutive patients undergoing pericardiocentesis at the authors' institution,[17] post–cardiac surgery and percutaneous procedure-related effusions corresponded to more than 50% of cases. The overall prevalence of ECP (diagnosed by echocardiography) in the authors' cohort was 16%. Other causes reported in the literature include ECP secondary to purulent pericarditis, trauma, neoplastic involvement, and end-stage renal disease.[13] Nevertheless, it is