

## MINI-FOCUS ISSUE: HEART FAILURE

### IMAGING VIGNETTE: CLINICAL VIGNETTE

# Idiopathic Constrictive Pericarditis and Eggshell Calcification of the Heart



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#### ABSTRACT

Extensive pericardial calcification is rare in patients with chronic constrictive pericarditis (CCP). We report the case of a young man who had CCP with “eggshell” calcification of the pericardium and the classic features of CCP on echocardiography and cardiac catheterization. The patient had an uneventful recovery following surgical pericardiectomy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1542-4) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 30-year-old man presented with gradually increasing pedal edema, abdominal distention, and dyspnea on exertion of 5 months’ duration. He had distended jugular veins with a positive Kussmaul sign and prominent x and y venous descent. He also had tender hepatomegaly, ascites, and grade II pedal edema. Cardiac auscultation revealed a diastolic pericardial knock.

Echocardiography revealed the following: calcified pericardium that was approximately 11 mm thick; septal bounce; medial and lateral mitral e’ velocity of 12 and 7 cm/s, respectively; more than 25% variation in mitral inflow velocity with respiratory movements (**Figure 1A**); a dilated inferior vena cava; and expiratory diastolic flow reversal in hepatic veins (**Figure 1B**). Fluoroscopy revealed dense circumferential pericardial calcification (**Figure 1C**, **Video 1**), and computed tomography demonstrated classic “eggshell” calcification encircling the heart (**Figures 1D and 1E**). Constrictive physiology was further confirmed on cardiac catheterization, which revealed elevated and equalization of all pressures including mean right atrial pressure, pulmonary capillary wedge pressure, right ventricular pressure, and left ventricular end-diastolic pressure (i.e., 30 mm Hg). Biventricular pressure tracings showed a typical “dip-and-plateau” configuration, as well as ventricular discordance (**Figure 1F**, white arrow) suggestive of CCP. The initial tracing (**Figure 1F**) also showed ventricular concordance suggestive of underlying myocardial involvement (restrictive physiology) associated with extensive pericardial calcification.

Surgical pericardiectomy was performed through a median thoracotomy. The calcified, firmly adherent pericardium was resected from the anterior and left lateral aspect of the heart. The patient had uneventful recovery. Histopathologic examination of resected pericardial tissue revealed extensive fibrosis, hyalinization, and calcification, without any granulomatous or giant cell inflammation.

CCP manifests as right-sided heart failure. The dyspnea on exertion results from raised filling pressures, whereas easy fatigability is caused by decreased cardiac output. The most common type of CCP is idiopathic (1), as in the index case, followed by CCP caused by infections, post-cardiac surgery status, radiation therapy.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors’ institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

Manuscript received April 24, 2020; revised manuscript received June 1, 2020, accepted June 10, 2020.

Although mild calcification is common in pericarditis, extensive calcification as seen in the index case is extremely rare in CCP (2).

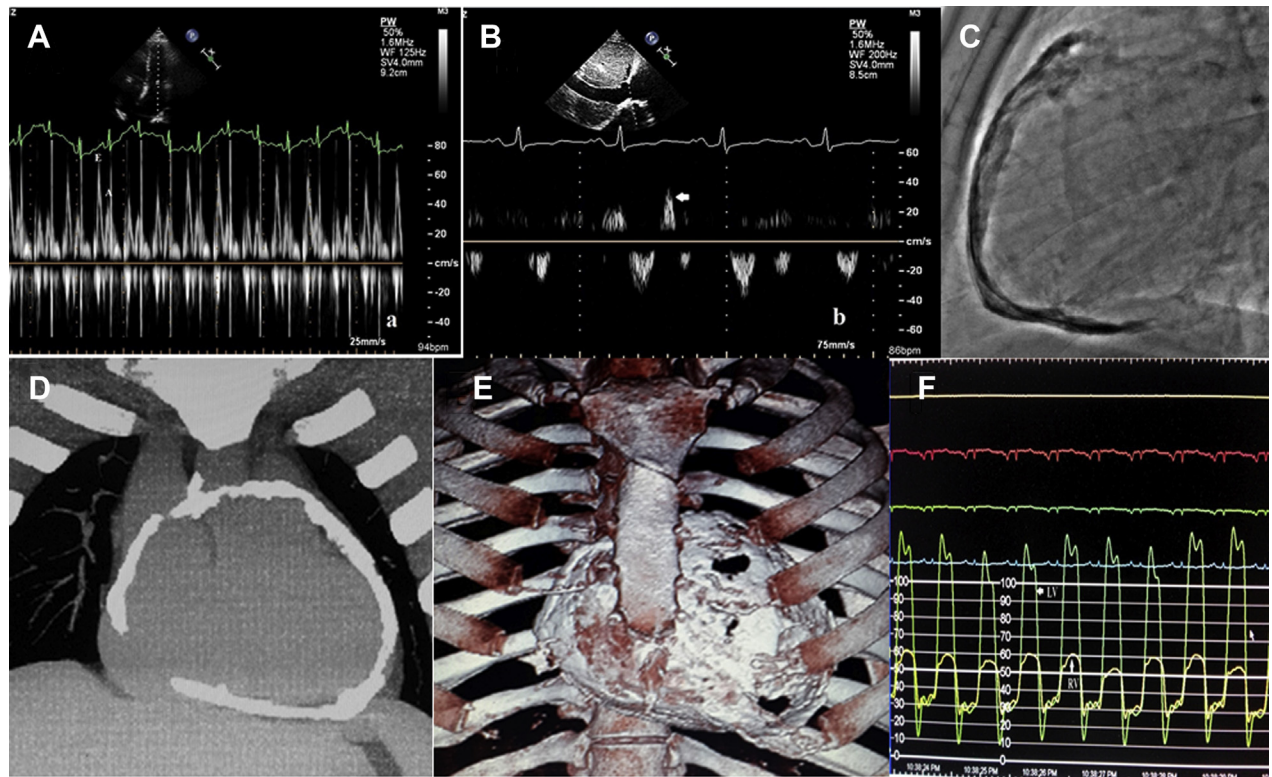
The diagnosis of CCP is made on the basis of echocardiographic findings of increased ventricular interdependence in the form of septal bounce and respiratory variation in atrioventricular valve inflow velocities, annulus reversus with medial e' velocity more than lateral, and late diastolic expiratory flow reversal in hepatic veins (3). Pericardial calcification and thickening can be clearly seen on computed tomography, and cardiac catheterization confirms the diagnosis.

Typically, all chambers have raised and equalization of diastolic pressures, as in the index case (3). Ventricular discordance is a specific hemodynamic finding in CCP (3). Because of cessation of negative intrathoracic pressure transmission to the heart in CCP, there are decreases in left ventricle filling and systolic pressure during inspiration, whereas right ventricular filling and systolic pressure are relatively maintained, thus resulting in exaggerated interdependence and ventricular discordance (3). Surgical pericardiectomy is associated with increased morbidity and mortality in such cases because of incomplete resection of the adherent calcified pericardium and poor hemodynamic recovery (2). Nonetheless, our patient had an uneventful post-operative recovery and remained asymptomatic for the next 5 years of follow-up.

**ABBREVIATIONS  
AND ACRONYMS**

**CCP** = chronic constrictive pericarditis

**FIGURE 1** Idiopathic Constrictive Pericarditis and Eggshell Calcification of the Heart



**(A)** Transmittal pulse-wave Doppler showing respiratory variation in inflow velocities. **(B)** Echocardiography showing a distended inferior vena cava and expiratory diastolic flow reversal of the hepatic veins. **(C)** Fluoroscopy image in the lateral view showing thickened calcified pericardium around the heart. **(D)** Computed tomography reconstructed image in the coronal plane showing thick, calcified pericardium around the heart. **(E)** Volume rendered reconstructed computed tomography image showing "eggshell" calcification of the heart. **(F)** Biventricular pressure tracing showing raised and equal end-diastolic ventricular pressure, a "dip-and-plateau" configuration, and ventricular discordance suggestive of chronic constrictive pericarditis.

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**KEY WORDS** calcification, constrictive pericarditis, pericardiectomy, right-sided heart failure

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 **APPENDIX** For a supplemental video, please see the online version of this paper.