Early-onset post-cardiotomy severe constrictive pericarditis: a case report

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Background
Constrictive pericarditis (CP) can be one of the most challenging conditions to diagnose within cardiovascular medicine. Iatrogenic causes of CP are increasingly recognized in higher income countries. This case provides insight into the need for clinical suspicion when diagnosing this relatively under recognized clinical entity as well as the need for multimodality imaging combined with invasive haemodynamic assessment.

Case summary
A 68-year-old man presented with decompensated heart failure 4 weeks after open-heart surgery. A diagnosis of early-onset post-cardiotomy CP was made using multimodality imaging and invasive haemodynamic assessment, which demonstrated the cardinal features of constrictive physiology. Surgical intervention with two pericardiectomy procedures was pursued given the aggressive and recalcitrant nature of his presentation. Our patient died shortly after his second surgery due to progressive multi-organ dysfunction.

Conclusion
Constrictive pericarditis is a challenging but important clinical entity to diagnose. Differentiating CP from restrictive cardiomyopathy is important as there are key differences in management and prognosis. Our case supports the clinical utility of multimodality imaging combined with invasive haemodynamic assessment in patients with suspected CP.

Keywords
Constrictive pericarditis • Pericardial disease • Iatrogenic pericarditis • Case report • Post-cardiotomy pericarditis

ESC Curriculum
2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 6.6 Pericardial disease

Learning points
• Recognize the increasing prevalence of iatrogenic pericarditis.
• Understand the role of multimodality imaging and invasive haemodynamic assessment in the diagnosis of CP.

Introduction
Constrictive pericarditis (CP) can be one of the most challenging clinical entities to diagnose within cardiovascular medicine potentially, which can lead to under recognition of this potentially fatal disease. High-income countries are increasingly recognizing the rising incidence of iatrogenic pericarditis. 1,2 Similarities in clinical presentation and diagnostic features of restrictive cardiomyopathy (RCM) and CP are often seen. Differentiating these distinct clinical entities is important as there are key differences in management options and prognosis. Multimodality imaging and invasive haemodynamic assessment are supported by consensus guidelines in the diagnostic pursuit of CP. 1 We present a case detailing the early-onset, post-cardiotomy CP demonstrating the cardinal diagnostic features of CP.

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Timeline

| Timepoint                      | Event Description                                                                 |
|-------------------------------|-----------------------------------------------------------------------------------|
| 4 weeks prior to initial      | Coronary artery bypass grafting (CABG) and aortic valve replacement (AVR) for     |
| presentation                  | severe aortic stenosis and concomitant stable coronary artery disease            |
| Initial presentation to       | Presented with features of acutely decompensated congestive cardiac failure      |
| hospital                      |                                                                                  |
| 1 week after initial          | Had been investigated with transthoracic echocardiogram, cardiac magnetic        |
| presentation                  | resonance imaging, and simultaneous left and right heart catheterization to       |
|                               | establish the unifying diagnosis of CP                                           |
| 2 weeks after initial         | Prolonged first pericardiectomy procedure complicated by tears to the right     |
| presentation                  | ventricular wall and pulmonary artery                                           |
|                               | (densely adherent pericardium difficult to decorticate leading to decision for a |
|                               | second, staged pericardiectomy procedure)                                       |
| 2 weeks + 3 days after        | Had second pericardiectomy procedure                                            |
| initial presentation          |                                                                                  |
| 3 weeks after initial         | Acute right ventricular dysfunction necessitating veno-arterial extracorporeal    |
| presentation                  | membrane oxygenation (VA-ECMO) support                                           |
| 4 weeks after initial         | Progressive clinical deterioration with multi-organ dysfunction leading to        |
| presentation                  | transition to comfort cares and patient passing away                             |

**Case presentation**

A 68-year-old man underwent CABG and bioprosthetic AVR without immediate peri-operative complications. One month post-operatively, he presented with dyspnoea on exertion, orthopnea, paroxysmal nocturnal dyspnoea, and extensive oedema. His physical examination was notable for pitting oedema extending from the feet up to the abdomen, a markedly elevated jugular venous pressure and a positive Kussmaul’s sign. Auscultation of the chest demonstrated reduced air entry at the bases extending up to the mid zones. The heart sounds were dull with no appreciable murmurs.

His past medical history included multivessel coronary artery disease and severe aortic valve regurgitation. He was treated with CABG to the left anterior descending artery (LAD) and posterior descending artery (PDA) as well as bioprosthetic AVR insertion. He also had paroxysmal atrial fibrillation, hypertension, and hypercholesterolaemia. He was a lifelong non-smoker. There was no prior history of impairment in left or right ventricular function including diastolic parameters. There was no history of pericarditis, myocarditis, autoimmune, or rheumatological conditions.

The patient’s clinical signs and symptoms were suggestive of acute decompensated biventricular failure. The differential diagnosis included acute left ventricular dysfunction although there was no clear precipitant for this identified from the patient’s clinical history. Other considerations included acute valvular pathology (although no murmur was identified on clinical exam), uncontrolled tachyarrhythmias (again there was no history of palpitations, pre-syncope, or syncope to support this), and pericardial/myocardial disease. Other diagnostic possibilities included superimposed pulmonary pathology such as pulmonary embolism or lower respiratory tract infection.

Chest roentgenogram demonstrated bilateral pleural effusions without evidence of pericardial calcification (Figure 1). A 12-lead electrocardiogram demonstrated sinus rhythm with no abnormalities (Figure 2). N-Terminal pro hormone B-type Natriuretic Peptide (NTproBNP) was elevated at 3034 ng/L (normal <126 ng/L). Transthoracic echocardiogram demonstrated normal sized cardiac chambers, normal left ventricular systolic function, and normal function of the aortic bioprosthesis. There was no regional wall motion abnormality noted. Note was made of a bright, thickened pericardium, and a small pericardial effusion. Respiratory sepal motion (septal bounce) consistent with ventricular interdependence was also noted. The inferior vena cava was noted to be distended without collapse on inspiration. Mitrall inflow velocities increased by >25% during expiration while tricuspid inflow velocities increased by more than 40% on inspiration (Figure 3).

Given the atypically rapid presentation, diagnostic certainty prior to referral for potential repeat sternotomy and pericardial decortication was deemed necessary. Invasive haemodynamic assessment was pursued to confirm the diagnosis of CP. Concurrent invasive coronary angiography was performed as an alternative to coronary CT to limit delays in clinical assessment due to the limited availability of coronary CT at our centre. Coronary angiography demonstrated severe native, multivessel coronary artery disease. The left internal mammary artery graft to the LAD was patent but the saphenous vein graft to the PDA was atretic. Ventriculography confirmed normal left ventricular systolic function with no appreciable gradient across the aortic valve on pigtail catheter pullback. Right heart catheterization with simultaneous left ventricular pressure measurements was performed. The following cardinal features of pericardial constrictions were demonstrated (Figure 4):  

1. Elevated mean right atrial pressure (17 mmHg) with rapid X and Y descents (the W or M sign) signifying rapid, early diastolic atrial emptying into an underfilled ventricle.
2. The square root sign in the right and left ventricular pressure waveforms indicating rapid early ventricular filling with a sudden plateau as the ventricles reach capacity within a stiff pericardium.
3. Equalization of left and right ventricular end-diastolic pressures reflective of a stiff, non-compliant pericardial sac.

![Figure 1](chest-roentgenograph-demonstrating-pleural-effusions.png)
Figure 2 12-Lead electrocardiogram demonstrating sinus rhythm without acute abnormalities. Note that the low peripheral lead voltages, which is one of the non-specific signs of constrictive pericarditis on electrocardiogram.

Figure 3 Transthoracic echocardiogram from the apical four-chamber view revealed respirophasic septal motion, seen at end-diastolic (A) inspiration and (B) expiration, and altered inflow velocities at the (C) mitral valve and (D) tricuspid valve.
Discordance in the left and right ventricular systolic pressures during the respiratory cycle indicating ventricular interdependence.

Cardiac magnetic resonance imaging (CMR) demonstrated normal cardiac chamber size and respirophasic septal motion consistent with ventricular interdependence. Crescentic thickening (up to 24 mm) of the pericardium around the left ventricular wall was demonstrated but Phase Sensitive Inversion Recover (PSIR) confirmed that the true pericardium measured $\sim 8$ mm with up to 16 mm of pericardial fluid identified around the left ventricle (Figure 5). Pericardial signal

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**Figure 4** Right and left heart catheterization pressure measurements demonstrated (A) elevated right atrial pressure with rapid x and y descents, (B) square root sign, (C) equalization of left ventricular and right ventricular end-diastolic pressures, (D) discordance in ventricular pressures during respiration.

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**Figure 5** On T2 cardiac magnetic resonance imaging, thickened, fibrous crescent-shaped pericardial mass adjacent to the anterolateral, lateral, and inferolateral walls, measuring up to 24 mm (A). Phase Sensitive Inversion Recover sequences with gadolinium contrast demonstrated an 8 mm thick pericardium with up to 16 mm of pericardial fluid around the left ventricle (B). Evident interventricular dependence on free breathing module, indicative of contractive physiology (Supplementary material online, Video).
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Discussion

Our case highlights the high index of clinical suspicion required in diagnosing CP as well as the challenges in involving this potentially life-threatening condition. In this case, the recalcitrant-to-treatment and unusually severe nature of the patient’s symptomatology led to consideration of CP with left and right heart catheterization and exclusion of other pathological mechanisms such as RCM. The high index of clinical suspicion required in diagnosing CP is essential to prevent delays in diagnosis and management. Multimodal cardiac imaging supported the diagnosis of CP and excluded other pathological mechanisms such as RCM.

Conclusions

Constrictive pericarditis is a rare but life-threatening condition. Iatrogenic causes are increasingly recognized. Diagnosis of CP and differentiation from RCM are challenging but necessary. Invasive haemodynamic assessment remains the gold standard for diagnosis.

Lead author biography

Dr Philopatir Mikhail is a Cardiology Advanced Trainee at Gosford Hospital in Australia with a special interest in coronary artery disease and coronary physiology. He hopes to pursue subspeciality training in Interventional and Structural Cardiology.

Supplementary material

Supplementary material is available at European Heart Journal—Case Reports online.

Slide sets:

A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent:

The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest:

None declared.

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