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CASE SERIES
Heart failure

Pericardial decompression syndrome with acute right ventricular failure: a case series

Dean M. Nelson *, Anthony P. Brennan, Andrew T. Burns, and Andrew I. MacIsaac

Department of Cardiology, St Vincent's Hospital Melbourne, 41 Victoria Parade, 3065, Fitzroy, Victoria, Australia

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Background Pericardial decompression syndrome (PDS) is an uncommon complication of pericardial drainage of large pericardial effusions and cardiac tamponade characterized by paradoxical haemodynamic instability following drainage. Pericardial decompression syndrome may occur immediately, or in the days following pericardial decompression, and presents with signs and symptoms suggestive of uni-/biventricular failure or acute pulmonary oedema.

Case summary This series describes two cases of this syndrome which demonstrates acute right ventricular failure as a mechanism of PDS and provides insights into the echocardiographic findings and clinical course of this poorly understood syndrome. Case 1 describes a patient who underwent pericardiocentesis, whilst Case 2 describes a patient who underwent surgical pericardiostomy. In both patients, acute right ventricular failure was observed following the release of tamponade and is favoured to be the cause of haemodynamic instability.

Discussion Pericardial decompression syndrome is a poorly understood, likely underreported complication of pericardial drainage for cardiac tamponade associated with high morbidity and mortality. Whilst a number of hypotheses exist as to the aetiology of PDS, this case series supports haemodynamic compromise being secondary to left ventricular compression following acute right ventricular dilatation.

Keywords Pericardial decompression syndrome • Tamponade • Case series

ESC Curriculum 2.2 Echocardiography • 6.4 Acute heart failure • 6.6 Pericardial disease • 6.7 Right heart dysfunction • 7.1 Haemodynamic instability

Learning points

- Understand pericardial decompression syndrome as an infrequent complication of pericardial drainage that may present with acute right ventricular failure.
- Observe echocardiographic progression from pericardial effusion to pericardial decompression syndrome and return to baseline function.

Introduction

Pericardial decompression syndrome (PDS) is a paradoxical deterioration of haemodynamic status or acute pulmonary oedema that uncommonly occurs following decompression of large pericardial effusions and cardiac tamponade.[1] Epidemiological data on the incidence of PDS are limited and likely underreported. The syndrome is rare and occasionally fatal. In this case series, we describe two patients with PDS that demonstrate the role of acute right ventricular (RV) failure in this syndrome.

PDS commonly manifests early post-drainage but can occur up to 48 h post-drainage. Symptoms are similar to acute heart failure, with

* Corresponding author. Tel: +613-9231-2211, Fax: +613-9231-3333, Email: dean.nelson3@svha.org.au
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acute pulmonary oedema, and can lead to cardiogenic shock. PDS should be considered a diagnosis of exclusion, and alternative diagnoses such as procedural trauma, pulmonary embolism, acute myocardial infarction, and sepsis should all be considered.

Timeline

Patient 1

Day	Salient features
0	Presentation to hospital with 7 days of worsening dyspnoea.
	Initial TTE revealing pericardial effusion with echocardiographic features of tamponade.
	Emergency pericardiocentesis with evidence of right ventricular failure occurring during drainage on concurrent echocardiography.
	Total 800 mL drained D0.
	Admission to CCU following procedure
1	Total 1300 mL drained.
	Normal LV size with moderate systolic dysfunction. Moderate dilated RV with severe systolic dysfunction and severe tricuspid regurgitation
8	Normalization of moderate LV and severe RV dysfunction
10	Resolution of symptoms and discharge to home
26	Repeat community TTE demonstrating normal biventricular systolic function, mild tricuspid regurgitation and no evidence of recurrence of significant effusion

Patient 2

Day	Salient features
0	Presentation to hospital with 6 days of worsening dyspnoea.
	Initial TTE revealing pericardial effusion with echocardiographic features of tamponade.
	Emergency surgical pericardiostomy with initial drainage of 3000 mL.
	Admission post-operatively to ICU for ventilatory support.
	Repeat D0 TTE showing moderately dilated RV with moderate-to-severe systolic dysfunction and mid-to-apical free wall hypokinesis
7	Repeat TTE demonstrating improvement in RV function to mild dysfunction with mild-to-moderate LV dysfunction
10	Removal of surgical drainage after ~4500 mL total drainage.
	Patient discharged against medical advice

Patient 1

A 20-year-old female attended the emergency department with 7 days of progressive dyspnoea. Her medical history included asthma and was on no regular medications. At presentation, the patient was tachypneic with a respiratory rate (RR) of 32 and hypoxic on minimal exertion (SpO2 90% on exertion from 100% at rest on room air) and exhibited elevated jugular venous pressure (JVP) without peripheral oedema. Blood pressure (BP) was 128/80 mmHg with a heart rate (HR) of 100 b.p.m. The initial investigation included a chest x-ray (CXR) which demonstrated an enlarged cardiac silhouette, bilateral pleural effusions, and an electrocardiogram (ECG) which showed sinus tachycardia with electrical alternans (*Figure 1*). Blood tests revealed iron deficiency anaemia (haemoglobin 8.6 g/dL, ferritin 22 ug/L) and an elevated white cell count (WCC) (14.9×10^9/L) and d-dimer (7.9 mg/L). C-reactive protein (CRP) and troponin were not elevated.

The patient underwent an urgent transthoracic echocardiogram (TTE), which showed a large circumferential pericardial effusion; the largest measurement was 5.5 cm in depth lateral to the left ventricle (LV). Echocardiogram features of tamponade were present with a 'swinging heart' and dilated inferior vena cava (IVC) unresponsive to inspiration with respiratory variation of mitral valve inflow (*Figure 2*).

Urgent pericardiocentesis was performed utilizing continuous echocardiographic imaging with fluoroscopic confirmation of drain position. Progressive enlargement of the RV was observed during pericardial aspiration as tamponade was relieved (*Figure 1* and see Supplementary material online, *Videos S1–S3*). Severe tricuspid regurgitation developed due to a large coaptation defect between the tricuspid valve leaflets as a result of annular dilation. Continuous wave Doppler through the tricuspid valve demonstrated RV systolic pressure of 23 mmHg, likely underestimating pulmonary pressure due to the severity of tricuspid regurgitation and rapid early equalization of atrial and ventricular pressures. In addition, 800 mL of blood-stained fluid was drained through an 8.3 Fr pigtail catheter.

After the initial improvement in clinical status during drainage (BP 140/80 mmHg, HR 100, RR 20), the patient became increasingly tachycardic (HR 130 b.p.m.), tachypneic (RR 28), and hypotensive (BP 85/60 mmHg) over the next 48 h, consistent with a clinical diagnosis of PDS. Despite the change in vital signs, there was no evidence of end-organ dysfunction. Arterial blood gas demonstrated mild respiratory alkalosis (pH 7.47, pCO2 32, lactate 0.7) whilst liver and renal function testing suggested no compromise. In the absence of end-organ dysfunction or rising lactate, the exhibited tachycardia was determined to be a physiological and sufficiently compensated response despite mild hypotension, and inotropic support was not instituted. The patient

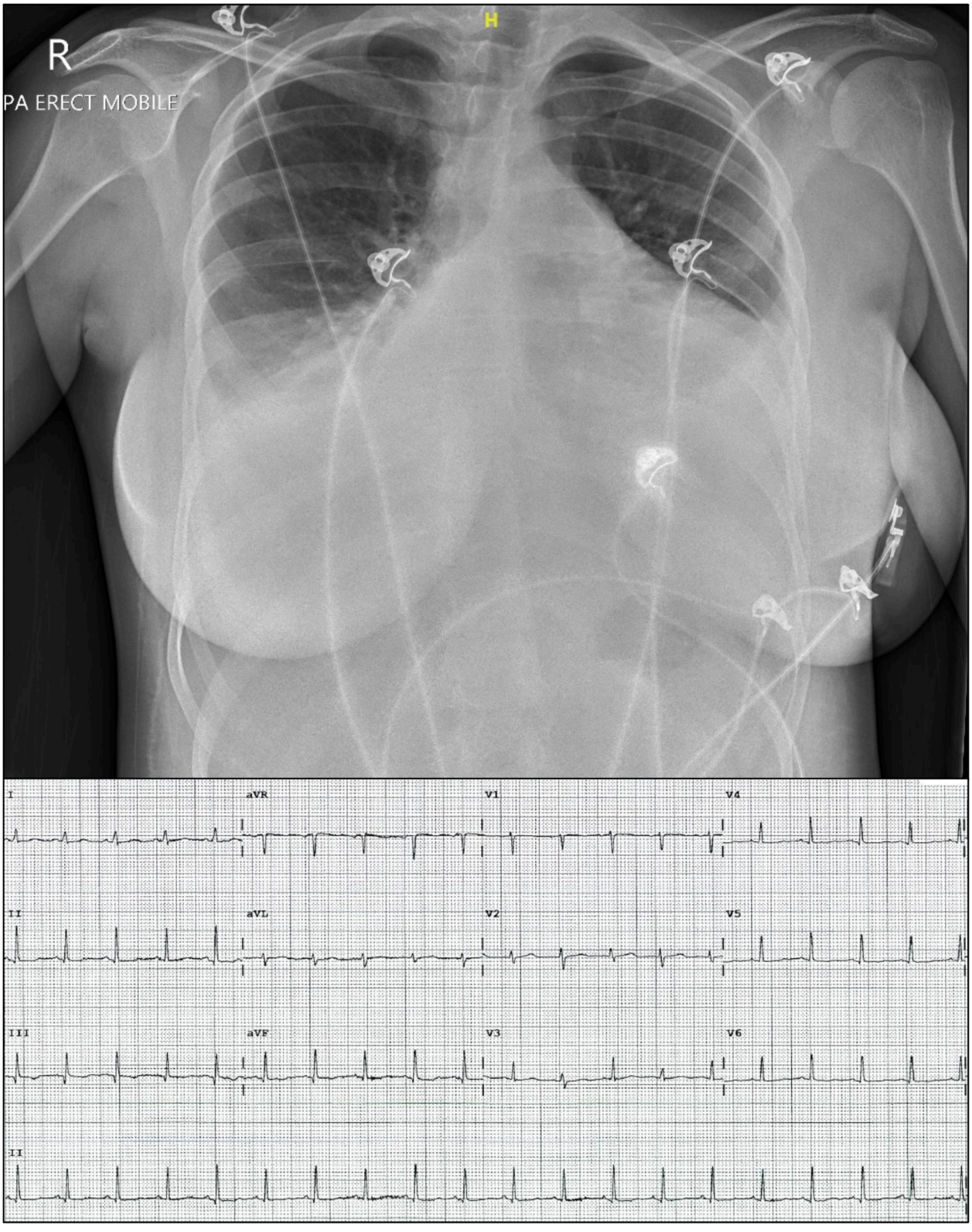

Figure 1 Posteroanterior chest x-ray and electrocardiogram of Patient 1. Posteroanterior chest x-ray demonstrating bilateral pleural effusions and enlarged cardiac silhouette. Electrocardiogram demonstrating small QRS complexes and electrical alternans.

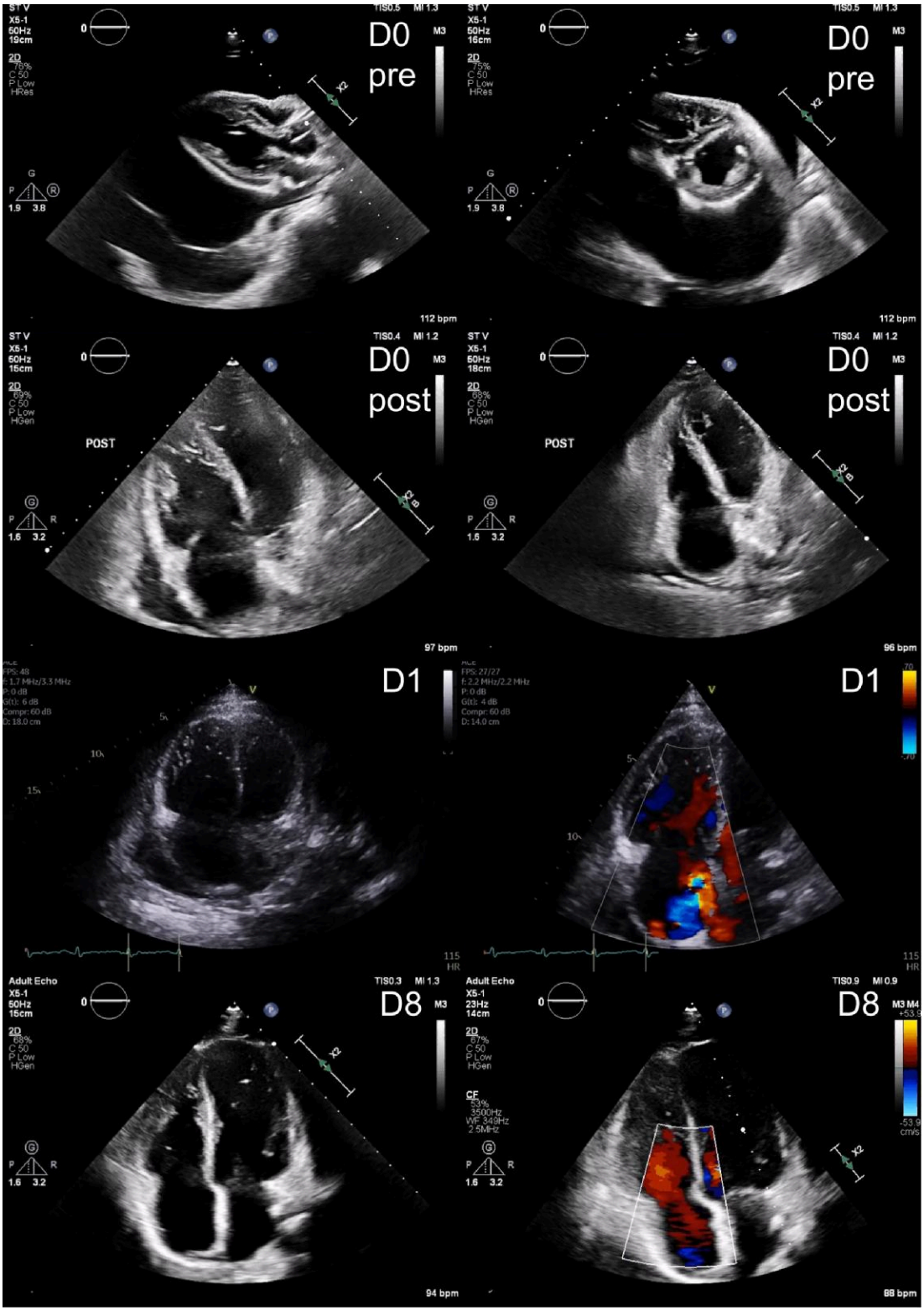

Figure 2 Echocardiographic progression from pericardial effusion with echocardiographic features of tamponade, to pericardial decompression syndrome, to resolution of Patient 1. (D0 pre-pericardiocentesis) Parasternal long- and short-axis views, demonstrating large pericardial effusion with right-sided chamber collapse. (D0 post-pericardiocentesis) Apical four-chamber view, demonstrating successive enlargement of the right ventricle and development of severe tricuspid regurgitation. (D1) Apical four-chamber view demonstrating severe right ventricle dilation and dysfunction, severe tricuspid regurgitation, and moderate left ventricle dysfunction. (D8) Apical four-chamber view showing normal right ventricle function, mild tricuspid regurgitation, and normalization of left ventricle function.

was closely monitored, and haemodynamic parameters improved without inotropic or mechanical support.

The following day 1300 mL had been drained. Fluid analysis showed elevated protein = 68 g/L (ref < 35 g/L), whilst microscopy, culture, and cytology were unremarkable—demonstrating only a moderate number of histiocytes, mesothelial cells, and lymphocytes. No malignant cells were identified. Serum rheumatological screening was also unremarkable. Repeat TTE revealed normal LV size with moderate systolic dysfunction. The RV was moderately dilated (RV basal diameter 44 mm) with severe systolic dysfunction and severe tricuspid regurgitation. A small residual pericardial effusion remained (*Figure 2* and Supplementary material online, *Video S4*). A CT pulmonary angiogram excluded pulmonary embolus as the cause for tachycardia and RV dilatation, and bilateral pleural effusions were again noted. Moreover, 600 mL of pleural fluid was aspirated for concern for contribution to respiratory distress.

Moderate LV and severe RV dysfunction initially persisted on serial echocardiograms with progressive improvement and normalization by Day 8 (*Figure 2* and see Supplementary material online, *Video S5*). The patient's symptoms resolved by Day 10, and she was discharged and recovered without complication. A repeat TTE on Day 26 demonstrated normal biventricular systolic function, mild tricuspid regurgitation, and no evidence of recurrence of significant effusion.

No cause for pericardial effusion was identified, and subsequently the patient's presentation is theorized to be of idiopathic/viral phenomenology. Without classical features for pericarditis and normalization of LV ejection fraction (EF) on serial echocardiography, anti-inflammatory agents and guideline-directed medical therapy were not instituted.

Patient 2

A 38-year-old male attended the emergency department with 6 days of worsening dyspnoea. Past history included schizophrenia (on paliperidone depot), obesity [body mass index (BMI) ~40], obstructive sleep apnoea, and polysubstance use (cannabis and methamphetamine). The patient presented to the hospital 1 year prior with dyspnoea and peripheral oedema but self-discharged before investigation.

At presentation, the patient was hypoxic (SpO2 94% on 1L via nasal prongs), tachypneic (RR 36), hypertensive (152/127 mmHg), and tachycardic (HR 121 b.p.m.). JVP was not visible given body habitus, whilst peripheral oedema was noted bilaterally to the ankles. A CXR demonstrated marked enlargement of the cardiac silhouette, and ECG revealed sinus tachycardia with low voltage QRS complexes and electrical alternans in lead II (*Figure 3*). Full blood analysis revealed elevated inflammatory markers (WCC 15.2×10^9/L and CRP of 53 mg/L), a mildly elevated B-natriuretic peptide of 110 ng/L, and antinuclear antigen titre of 1:160. TTE revealed a 6.5 cm circumferential pericardial effusion, with echocardiographic features of tamponade including dilated (3.0 cm) and non-collapsible IVC, with RV and RA compression (*Figure 4*).

Given the patient's body habitus and perceived potential for complications with pericardiocentesis, the patient underwent a surgical pericardiostomy after a heart-team discussion. In addition, 3000 mL of blood-stained fluid was drained, and the patient was admitted to the intensive care unit (ICU). Fluid analysis yielded protein = 69.0 g/L (ref < 35), lactate dehydrogenase (LDH) was not available for review, and cytological and microbiological analysis was unrevealing. Histological analysis revealed scarring and patches of mildly active chronic inflammation with areas of erosion and replacement with fibrin deposition. A TTE on Day 0 soon after drainage demonstrated a moderately dilated RV with moderate-to-severe systolic dysfunction and mid-to-apical free wall hypokinesis (*Figure 4*). There was moderate LV dysfunction, and whilst a diagnosis of stress cardiomyopathy was considered, the pattern of LV dysfunction was not consistent with this diagnosis.

At this time, the patient began to experience worsening pulmonary oedema with increasing oxygen and ventilation requirement, escalating from nasal prongs to high-flow nasal prongs to bilevel positive airway pressure (BiPAP) in the subsequent 24 h, consistent with PDS. The patient was weaned from all ventilatory support by 72 h post-procedure. In light of this and the unclear aetiology of pericardial effusion, the patient underwent contrast-enhanced CT brain chest, abdomen, and pelvis. No pulmonary embolus or features of occult malignancy were identified.

Repeat TTE on Day 7 showed mild RV dysfunction with mild-to-moderate LV dysfunction. No evidence of residual effusion was demonstrated (*Figure 4*). No apical views were possible due to the surgical site and drain tube. The patient was commenced on colchicine and ibuprofen in light of histological findings, as well as therapy for LV dysfunction given the persistent impairment of LV systolic function on echocardiogram. Drains were removed on Day 10 after drainage of ~4500 mL total, and the patient was subsequently discharged against medical advice. Further echocardiographic evaluation has not occurred to date.

Discussion

Pericardial decompression syndrome is a rare, life-threatening, and transient complication of pericardial drainage,[1] characterized by paradoxical haemodynamic instability and/or pulmonary oedema following an otherwise uncomplicated procedure. A comprehensive analysis of PDS cases published in the European Heart Journal in 2015 reported that in limited cases (totalling 35 patients), mortality from PDS was high (29%), and only significantly associated with surgical drainage.[2] In this comprehensive analysis, only three patients (9%) of those studies presented with RV failure as seen in our case series.

Further studies are required to better understand the pathophysiology of PDS; however, a number of hypotheses have been proposed:

The haemodynamic hypothesis describes the complex interplay between left and right ventricular systolic dysfunction. Pericardial drainage is followed by a net increase in pulmonary venous return (i.e. LV preload) whilst adaptive systemic vascular resistance (i.e. LV afterload) is still high. The rapid increase in preload is mismatched against high afterload and leads to decompensated LV failure, hypotension, and pulmonary oedema. This observation was first described by Vandyke *et al.* in 1983.[3] LV dysfunction increases pulmonary wedge/pulmonary arterial pressures, impeding RV outflow and precipitating RV failure.

The alternative mechanism of the haemodynamic hypothesis suggests RV failure precedes LV failure, through compression of the LV due to ventricular interdependence. As drainage occurs, the more compliant RV expands more rapidly and leads to acute compression of the LV. Pulmonary capillary wedge pressure is elevated and cardiac output is decreased leading to hypotension, pulmonary oedema, and respiratory failure.

Case studies demonstrating acute dilation of the RV, along with the development of severe tricuspid regurgitation during pericardiocentesis as seen in Patient 1 are extremely limited. Our case supports the haemodynamic hypothesis and the role the acute RV dysfunction plays in this condition; however, it is difficult to demonstrate whether RV dysfunction precedes LV dysfunction or is a consequence of this. We suggest that the RV free wall, being thinner and less muscular than the LV, is more at risk of dysfunction secondary to external forces (as seen in tamponade with RV chamber collapse) and may precede LV dysfunction, though this remains unproven. Our observations could be explained by a transient loss of RV free wall tensile strength secondary to raised pericardial pressure reducing the pressure gradient between the RV and the pericardium. It is unknown whether the duration of pericardial effusion existence prior to drainage is a factor in this syndrome.

The ischemic hypothesis postulates that increased pericardial pressure during tamponade impairs coronary perfusion.[4] Myocardial ischaemia and LV stunning may persist after pericardial drainage temporarily, resulting in diastolic dysfunction and subsequently PDS. Notably, our patients neither reported anginal symptoms nor had ECG changes to support this hypothesis.

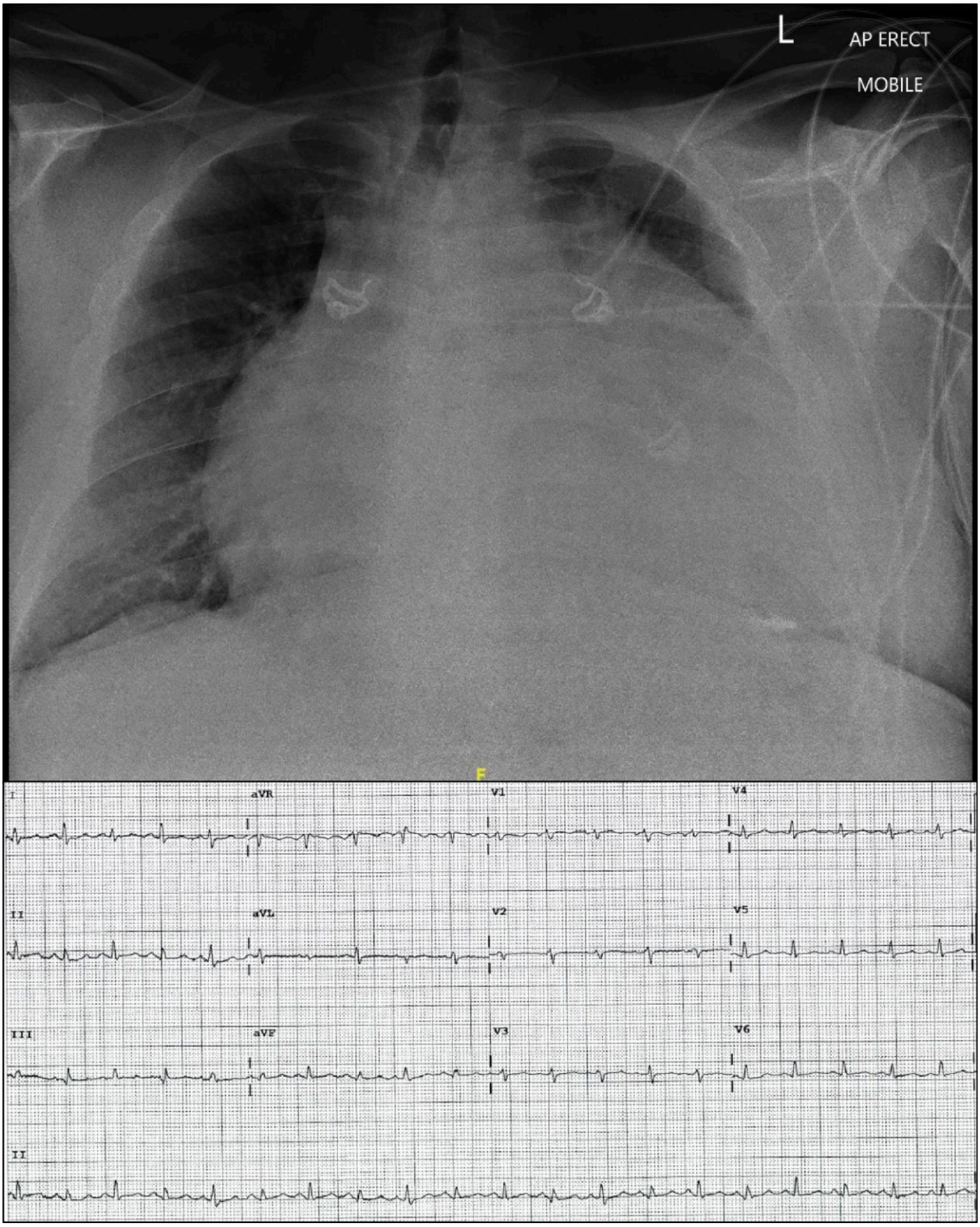

Figure 3 Posteroanterior chest x-ray and ECG of Patient 2. Posteroanterior chest x-ray demonstrating severe cardiomegaly. Electrocardiogram demonstrating small QRS complexes and electrical alternans.

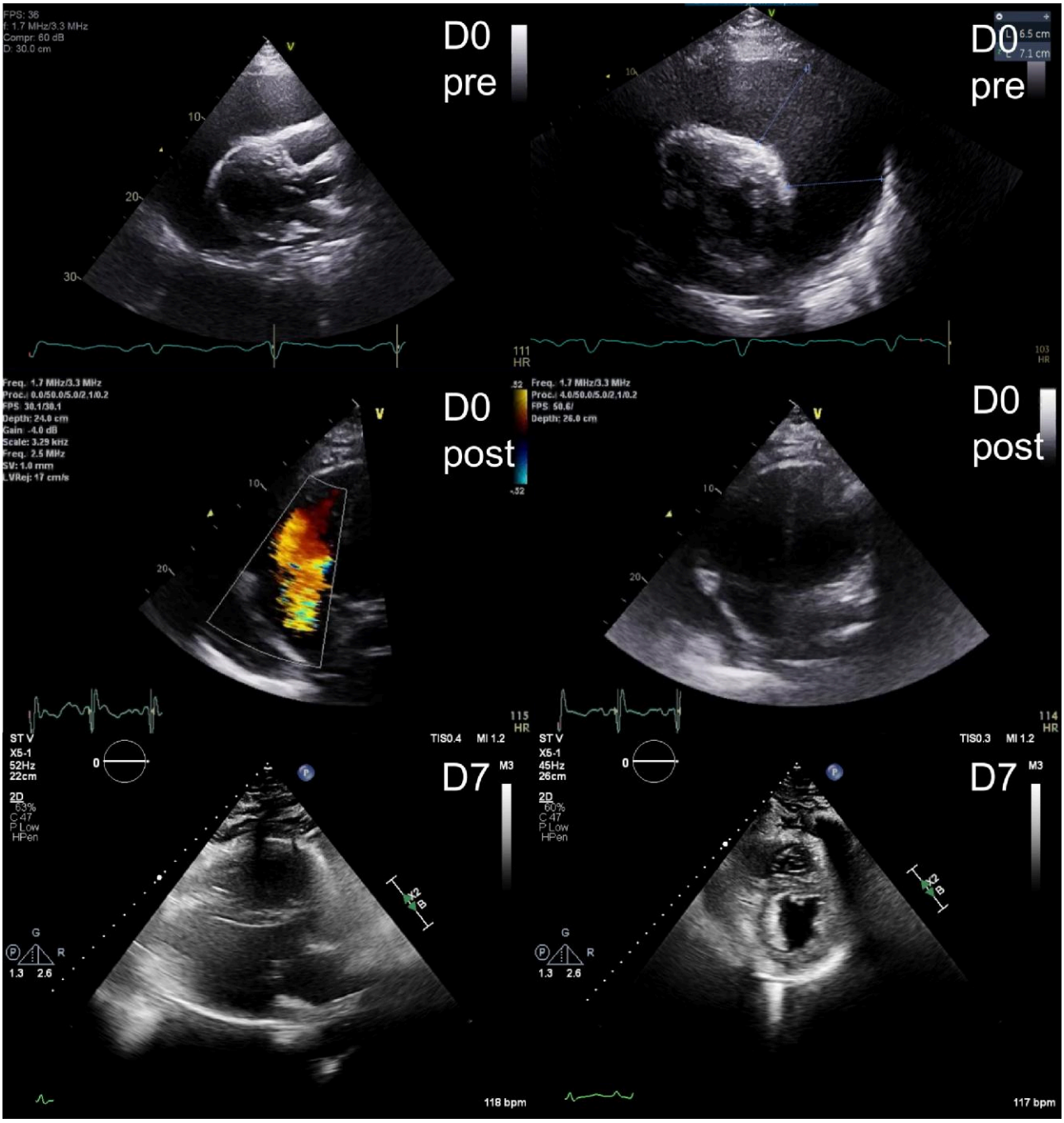

Figure 4 Echocardiographic progression from pericardial effusion with echocardiographic features of tamponade, to pericardial decompression syndrome, to resolution of Patient 2. (D0 pre-drainage) Parasternal long- and short-axis view, demonstrating massive pericardial effusion with right-sided chamber collapse. (D0 post-drainage) Apical four-chamber view, demonstrating severe right ventricle dilation and dysfunction, severe tricuspid regurgitation, and moderate left ventricle dysfunction. (D7) Parasternal long- and short-axis view, demonstrating mild right ventricle dysfunction, mild tricuspid regurgitation, and mild-to-moderate left ventricle dysfunction.

The autonomic/sympathetic overdrive hypothesis postulates acute withdrawal of sympathetic stimulus after decompression leads to an imbalance of the autonomic system.[5] Removal of sympathetic stimulus may lead to the unmasking of LV dysfunction that was previously compensated for by high endogenous catecholamine levels having a positive chronotropic and inotropic effect. This mechanism is thought to be similar to stress-induced cardiomyopathy and has elsewhere been suggested to potentially represent a clinical state in the same spectrum of disease as PDS, as opposed to being clinically distinct.[6] Notably, none of our patients had echocardiographic findings consistent with stress cardiomyopathy.

To date, there are no evidence-based guidelines for the prevention or treatment of PDS. The European Society of Cardiology 2015 Guidelines recommend drainage of fluid in < 1000 mL aliquots,[7] noting a comprehensive analysis conducted in the same year published in the European Heart Journal that reported PDS occurrence in drainages of < 500 mL.[2] An alternate approach suggests the removal of pericardial fluid until resolution of tamponade (evident by haemodynamic/echocardiographic measures), and then avoiding further rapid removal of additional fluid with a prolonged pericardial drainage protocol may help avoid PDS.[8] Some degree of chamber dilation is to be expected during pericardiocentesis; however, the development of progressive haemodynamic compromise with the development of ventricular dysfunction and severe tricuspid regurgitation may be indications to pause further drainage. Recommendations for management include monitoring patients with or at high risk for PDS in a coronary care unit (CCU) or ICU environment, institution of heart failure therapy, and consideration of inotropic support. In severe cases of PDS, external corporeal membrane oxygenation (ECMO) has been utilized.[9] Echocardiography during pericardiocentesis and at the onset of haemodynamic deterioration was valuable to guide management during each of the cases described in this series.

PDS is an uncommon complication of pericardial drainage; however, the haemodynamic effects have a significant impact on morbidity and mortality. Proceduralists should have a high index of suspicion to monitor for the development of PDS following both pericardiocentesis and surgical pericardiostomy. Further investigation into the aetiology and physiology of PDS is required in order to assist in the production of evidence-based guidelines to assist in the prediction, prevention, and treatment of PDS.

Lead author biography

Dr Dean Nelson is a physician trainee from St Vincent's Hospital Melbourne in Victoria, Australia. During his work in the Department of Cardiology, he has enjoyed the opportunity to explore interesting case studies in the area of interventional cardiology and echocardiography with the support and mentorship of his co-authors. Dean hopes to undertake advanced training in cardiology in the future and combine his interest in clinical cardiology with his passion for research.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Consent: The authors declare that consent has been given for publication of each of the detailed case studies in accordance with Committee on Publication Ethics (COPE) guidelines.

Conflict of interest: None declared.

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Data availability

The data underlying this article are available in the article and in its online supplementary material.

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