

# Severe pulmonary regurgitation mimicking constrictive pericarditis: a case report—the sac or the content? That is the question

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## Background

Constrictive pericarditis (CP) is a pathological condition of the pericardium, resulting from fibrosis, scarring, and calcification of the pericardium. Other conditions have been reported to mimic ‘constrictive physiology’ despite the presence of an intact pericardium. However, there has been no report of pulmonary regurgitation (PR) mimicking the haemodynamic characteristics of CP.

## Case summary

A 51-year-old woman was admitted to our institute because of severe right-sided heart failure. Transthoracic echocardiography revealed severe PR concomitant with significant dilatation of the right-sided heart. Septal bounce and the respiratory reciprocation of the transmitral and transtricuspid inflow velocities were also observed, indicating exacerbated ventricular interdependence. Cardiac catheter examination demonstrated elevated right atrial pressure with a prominent y descent, dip, and plateau waveform in the right ventricular pressure, and equalization of the diastolic pressure of all cardiac chambers, which are quite consistent with CP. On surgical inspection, however, there was no pericardial thickening or adhesion, indicating no obvious signs of CP.

## Discussion

Pericardial constriction results from the relative relationship between intrapericardial volume and pericardial reserve. When the intrapericardial volume exceeds the physiological limit, the cardiac chambers compete with each other in a fixed pericardial space. In this case, prominent dilation of the right-sided chambers caused by severe PR resulted in overstretching of the pericardium above the pericardial reserve, which led to a characteristic haemodynamic picture that resembled CP. Thus, it is important to recognize the diagnostic pitfall in the preoperative evaluation of a ‘CP mimic physiology’.

## Keywords

Constrictive pericarditis • Right-sided heart failure • Pulmonary regurgitation • Constrictive physiology • Case report

## Learning points

- Constrictive pericarditis (CP) is a pathological condition of the pericardium that results from fibrosis, scarring, and occasionally calcification of the pericardium.
- Severe pulmonary regurgitation with an enlarged right heart can mimic the haemodynamic findings of CP because of interaction between intrapericardial volume and pericardial capacity.
- Repeated haemodynamic evaluation after volume reduction therapy may provide important clues for accurate diagnosis and therapeutic strategies in patients with ‘relative pericardial constriction’.

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## Introduction

Constrictive pericarditis (CP) is one of the pathological conditions of the pericardium resulting from fibrosis, scarring, and occasionally calcification of the pericardium.<sup>1</sup> Other conditions, including severe tricuspid regurgitation (TR),<sup>2,3</sup> right ventricular (RV) infarction,<sup>4</sup> and acute pulmonary thromboembolism<sup>5</sup> have been reported to mimic 'constrictive physiology' even with intact pericardium. However, no case of pulmonary regurgitation (PR) mimicking haemodynamic characteristics of CP has been reported to date.

Here, we report an extremely rare case of a patient with enlarged right-sided heart owing to severe PR who demonstrated CP-mimicking haemodynamics.

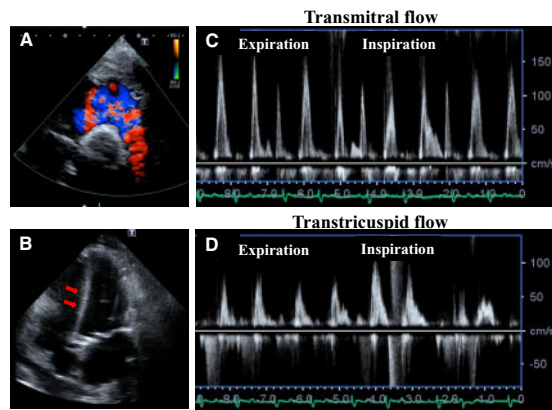
## Timeline

Time	Events
Before admission	<p>The patient underwent surgical pulmonary commissurotomy for pulmonary stenosis and closure of a secundum atrial septal defect at the age of 7 years.</p> <p>The patient received haemodialysis for end-stage renal failure from chronic glomerulonephritis at the age of 20 years.</p> <p>The patient underwent living-donor kidney transplantation twice owing to a rejection reaction at the age of 21 and 39 years, respectively.</p> <p>The patient underwent surgical aortic valve replacement for aortic regurgitation at the age of 27 years. Transthoracic echocardiography revealed severe pulmonary regurgitation (PR), and oral diuretics were prescribed to treat right-sided heart failure at the age of 49 years.</p> <p>The heart failure became refractory to medical therapy from the age of 50 years.</p>
After admission	<p>The patient was admitted to our institution because of a leg blister owing to stasis dermatitis resulting from severe chronic right-sided heart failure at the age of 51 years.</p> <p>Transthoracic echocardiography and right heart catheterization after admission demonstrated typical constrictive physiology.</p> <p>Cardiac magnetic resonance imaging showed significant right ventricular dilatation owing to severe PR.</p> <p>Computed tomography showed slightly thickened pericardium with scattered pericardial calcification.</p> <p>The patient underwent continuous haemodiafiltration to remove the excessive circulatory volume, and the cardiac catheter examination was re-analysed.</p> <p>Because the pericardium was intact on surgical inspection, only pulmonary valve replacement was performed. The right-sided heart failure improved after surgery.</p>
End result	<p>The patient died of renal failure on the 60th post-operative day triggered by urinary tract infection.</p>

## Case presentation

This case report concerns a 51-year-old woman who eventually developed refractory right-sided heart failure. Her past medical history consisted of surgical pulmonary commissurotomy for pulmonary stenosis and closure of atrial septal defect at the age of 7 years and aortic valve replacement using a mechanical valve due to aortic regurgitation at 27 years of age. She also had a medical history of haemodialysis for 17 years because of end-stage renal failure, and then underwent living kidney transplantation. Since the age of 49 years, she suffered from right-sided heart failure owing to severe PR; thus, oral diuretics were prescribed to treat symptoms of heart failure. However, systemic oedema and weight gain gradually worsened despite use of a large amount of diuretics, and eventually her heart failure became refractory to the medical treatment. Finally, she was admitted to our institution because of leg blister developed because of stasis dermatitis resulting from severe right-sided heart failure.

On admission, her blood pressure was 116/64 mmHg, heart rate 64 beats/min, and oxygen saturation 96% (room air). Physical examination revealed jugular venous distention, abdominal distension, and bilateral stasis dermatitis with blister. On auscultation, a harsh holodiastolic murmur was observed at the left upper sternal border. Electrocardiogram showed a normal sinus rhythm with Wenckebach atrioventricular block concomitant with complete right bundle branch block. Chest radiography showed cardiomegaly and bilateral pleural effusion. Transthoracic echocardiography revealed normal left ventricular contractile function; however, severe PR and resultant RV dilatation were observed ([Figure 1A](#), [Supplementary material online, Video S1](#)). Furthermore, a leftward shift of the ventricular septum during inspiration (i.e. septal bounce) was clearly confirmed ([Figure 1B](#), [Supplementary material online, Video S2](#)). On pulsed-wave Doppler echocardiogram, mitral *E* velocity increased and decreased on expiration and inspiration (respiratory variation of 25%), respectively, while the tricuspid *E* velocity decreased and increased on expiration and inspiration (respiratory variation of 43%), respectively ([Figure 1C and D](#)), indicating an exacerbated ventricular interdependence. Cardiac magnetic resonance imaging revealed RV end-diastolic volume index of 146 mL/m<sup>2</sup>, right ventricular ejection fraction of 47%, and regurgitant fraction of PR of 44% ([Figure 2](#), [Supplementary material online, Video S3](#)). The patient underwent cardiac catheter examination for further haemodynamic evaluation, which revealed the elevation and equalization of the diastolic pressure in all cardiac chambers ([Table 1](#)). In addition, the right atrial (RA) pressure waveform, consisting of significantly elevated mean RA pressure with a prominent *y* descent was consistent with the constrictive physiology ([Figure 3A](#)). Right ventricular pressure tracing revealed a typical dip and plateau waveform ([Figure 3B](#)). Although respiratory reciprocation of both ventricular pressures was not confirmed because of the mechanical valve at the aortic position; nevertheless, these haemodynamic findings were consistent with typical CP. Computed tomography showed mildly thickened pericardium with scattered pericardial calcification and severe abdominal visceral calcification due to secondary hyperparathyroidism resulting from chronic renal failure ([Figure 4](#)). Because the severe right-sided heart failure was intractable despite continuous administration of large amount of loop diuretics, she underwent continuous haemodiafiltration (CHDF) to



**Figure 1** (A) Parasternal short-axis view shows a significant regurgitant colour flow signal that occupies the entire right ventricular outflow, indicating severe pulmonary regurgitation. (B) The apical four-chamber view shows leftward shift of the ventricular septum during inspiration (red arrows). (C) Pulsed-wave Doppler E wave velocity of transmitral flow decreases during inspiration, while that of transtricuspid flow increases during inspiration (D).

remove excessive circulatory volume. After CHDF, although cardiac catheter examination showed a prominent  $\gamma$  descent in RA pressure and a dip and plateau waveform in RV pressure (Figure 3C and D), the equalization of diastolic pressure was absent (Table 1).

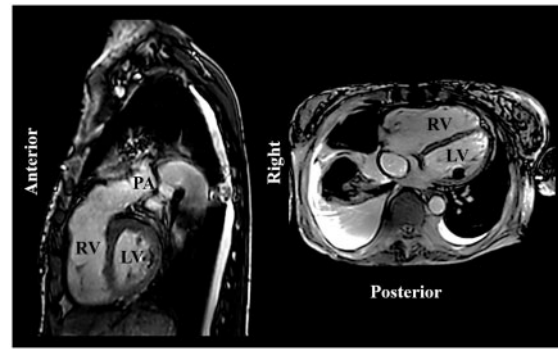
After a thorough discussion within the heart team, we decided to perform pericardiectomy simultaneous with pulmonary valve replacement. On surgical inspection, however, there was no pericardial thickening or adhesion, indicating no obvious signs of CP. Eventually, she underwent only pulmonary valve replacement. The patient did well just after the surgery, without the need for intravenous diuretics. However, she died owing to renal failure on the 60th post-operative day triggered by urinary tract infection.

## Discussion

The report presented the rare case of a patient with severe PR who demonstrated CP-mimicking haemodynamics.

The physiological role of the pericardium is to prevent excessive dilation of the cardiac chambers in response to an abrupt increase in the circulatory volume, to contribute to the haemodynamic connection between the cardiac chambers, and to generate adequate diastolic chamber stiffness.<sup>6</sup> However, in cases with CP, all cardiac chambers are confined within a constricting pericardium; thus venous return is significantly impaired, and the fluctuations in intrathoracic pressure do not transmit into intracardiac pressure.<sup>1</sup> Therefore, the primary haemodynamic consequences are impairment of the ventricular filling and exacerbation of ventricular interdependence.

Pericardial restraint can be resulted from the relative relationship between intrapericardial volume and pericardial reserve. The normal pericardium stretches over time without increase in intrapericardial pressure in response to the chronic volume overload. However, when intrapericardial volume exceeds the physiological limit, the



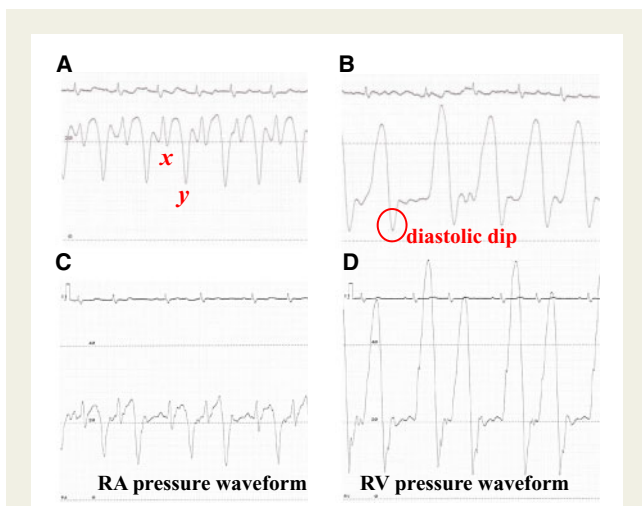
**Figure 2** (A) Sagittal view of the cardiac magnetic resonance imaging shows regression of the pulmonary valve and dilatation of the right ventricle. (B) Axial view shows dilated right-sided heart and bilateral pleural effusion. LV, left ventricle; PA, pulmonary artery; RV, right ventricle.

**Table 1** Pressure data measured by right heart catheterization before and after continuous haemodiafiltration

Pressure (mmHg)	Before CHDF	After CHDF
PCW: <i>a</i> wave/ <i>v</i> wave (mean)	32/17 (22)	26/20 (22)
PA: systolic/diastolic (mean)	58/20 (34)	53/15 (28)
RV: systolic/diastolic/end-diastolic	56/5/18	57/6/29
RA: <i>a</i> wave/ <i>v</i> wave (mean)	23/25 (21)	26/12 (20)
Ao: systolic/diastolic (mean)	161/75 (143)	137/39 (72)

Ao, aorta; CHDF, continuous haemodiafiltration; PA, pulmonary artery; PCW, pulmonary capillary wedge; RA, right atrium; RV, right ventricle.

cardiac chambers compete with each other in a fixed pericardial space. Thus, the diastolic pressures of all cardiac chambers will elevate and equalize in the end; moreover, ventricular interdependence will be exaggerated as observed in cases with CP. Thus, there may be two phenotypes that lead to constrictive physiology. First phenotype is the pericardial constriction of cardiac chambers by thickened, adhered, and rigid pericardium, that is true CP. Another phenotype is the 'relative pericardial constriction' resulting from enlargement of the cardiac chambers above the physiological limit of the pericardial reserve, that is the 'CP mimic physiology'.<sup>2-5</sup> In this case, prominent dilation of the right-sided chambers caused by severe PR resulted in overstretching of the pericardium beyond the pericardial reserve, which ultimately led to significant elevation of the intrapericardial pressure. Thus, based on the relative relationship between the enlarged cardiac chambers and overstretched non-compliant pericardium, a characteristic haemodynamic picture resembling CP manifested itself. Although the pericardial reserve may depend on its distensibility, mildly thickened pericardium with scattered calcification owing to prolonged haemodialysis and two cardiac surgeries might have reduced the pericardial reserve in this case. The continuous wave Doppler configuration derived from the PR signal, in which the



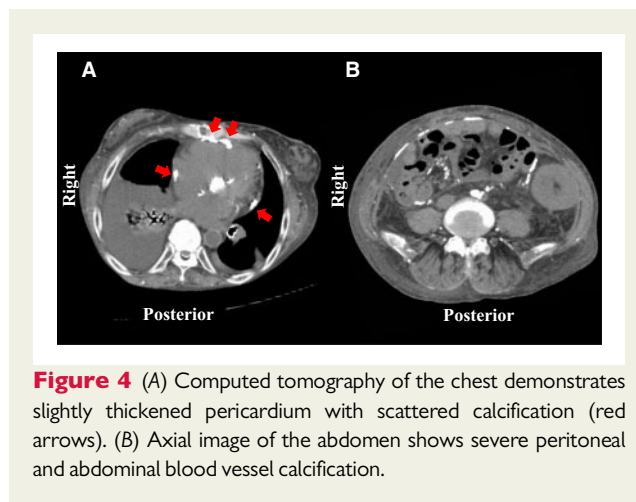
**Figure 3** Pressure waveforms of the right atrium and right ventricle before and after continuous haemodiafiltration are shown. (A) The right atrial pressure waveform shows a typical 'W shape' consisting of preserved x and deep y descent, consistent with constrictive pericarditis. (B) The right ventricular pressure tracing exhibits a typical 'dip and plateau' waveform. After continuous haemodiafiltration, the morphology of both right atrial (C) and right ventricular (D) pressure waveforms are similar, which is consistent with constrictive pericarditis. CHDF, continuous haemodiafiltration.

regurgitant flow velocity abruptly declines after early diastolic peak followed by a low-velocity plateau (i.e. reciprocal to RV dip and plateau pressure waveform), could enhance the detection of CP.<sup>7</sup> From a theoretical perspective, however, analysis of the PR waveform may not be effective for discriminating the 'relative pericardial constriction' from 'true CP' in this situation.

Another unique haemodynamic characteristic of this case was the fluctuated constrictive physiology by the volume reduction therapy. Kato et al.<sup>2</sup> previously reported an elderly patient who showed CP-mimicking haemodynamics owing to significant dilation of the right-sided chambers caused by severe TR. In this case, the CP-mimicking haemodynamics was completely resolved by medical therapy, including diuretics and a vasodilator. In the present case, although prominent y descent in RA pressure tracing and dip and plateau waveform in RV pressure tracing persisted after volume reduction therapy by CHDF, the equalization of diastolic pressure was absent, which may have provided evidence of 'relative constriction'. Therefore, repeated haemodynamic evaluation after volume reduction therapy may provide the important clues for accurate diagnosis and therapeutic strategy for patients with relative pericardial constriction.

## Conclusion

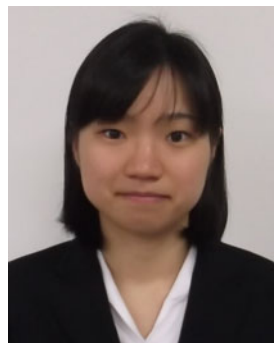
To the best of our knowledge, this is the first reported case of 'CP mimic haemodynamics' resulting from severe PR caused by the restraining effect of the enlarged right-sided heart within the intact pericardium. We experienced a highly suggestive case in considering



**Figure 4** (A) Computed tomography of the chest demonstrates slightly thickened pericardium with scattered calcification (red arrows). (B) Axial image of the abdomen shows severe peritoneal and abdominal blood vessel calcification.

the physiological and pathological relationships between the heart (i.e. content) and the pericardium (i.e. sac).

## Lead author biography



After graduating from university in 2011, Makiko Suto went into the path of cardiology after 2 years of residency. After working as a general cardiologist for 3 years, she devoted herself to studying echocardiography for 3 years at Kobe University Graduate School. Now, she is a physician specialized in echocardiography especially for adult congenital heart disease.

## Supplementary material

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

## Acknowledgements

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text were obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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