

Lessons from a case with stiff left atrial syndrome complicated by iatrogenic inter-atrial shunts: a case report

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Background

In patients with stiff left atrial (LA) syndrome, reservoir function is significantly impaired due to extensive LA fibrosis; consequently, the increase in LA pressure during haemodynamic stress is prominent, easily leading to pulmonary venous hypertension and subsequent pulmonary congestion, and eventually results in intractable heart failure.

Case summary

A 79-year-old female with mitral stenosis and atrial fibrillation underwent valve replacement, Cox-Maze IV procedure, LA plication, and appendage ligation 4 years prior to presentation. Thereafter, she underwent a total of two catheter ablation procedures for recurrent atrial tachycardia. Transthoracic echocardiography revealed two continuous colour jets across the interatrial septum, with a peak pressure gradient of 23 mmHg, which was consistent with the residual puncture hole at the catheter ablation procedures. Although transoesophageal echocardiography showed no evidence of prosthetic valve dysfunction, the pulmonary venous flow signal showed a significantly blunted systolic forward flow, extremely small retrograde reversal flow during atrial contraction, and prominent diastolic flow velocities, all of which indicated significantly impaired LA function. Cardiac catheter examination revealed a characteristic pulmonary capillary wedge pressure waveform, which consisted of a steep ascending limb of v wave with a large peak, consistent with stiff LA syndrome.

Discussion

Treatment of patients with stiff LA syndrome is quite challenging and restricted to the use of diuretics only, which has limited efficacy and eventually results in intractable heart failure. In this case, owing to the inter-atrial pressure-relieving gateway, the patient was only mildly symptomatic despite the existence of a non-compliant LA.

Keywords

Stiff left atrial syndrome • Left atrial compliance • Left atrial fibrosis • Inter-atrial shunt • Case report

ESC curriculum

2.1 Imaging modalities • 2.2 Echocardiography • 6.1 Symptoms and signs of heart failure • 6.3 Heart failure with preserved ejection fraction • 7.1 Haemodynamic instability

Learning points

- In patients with stiff left atrial (LA) syndrome, reservoir function is significantly impaired due to extensive LA fibrosis, easily leading to pulmonary venous hypertension and subsequent pulmonary congestion, and eventually results in intractable heart failure.
- In patients with inter-atrial communication, the shunt functions as a pressure-relieving gateway, thereby preventing elevation in LA pressure.
- These unique haemodynamics could be extrapolated to an analogous pathophysiology, owing to which the patient remained disproportionately oligosymptomatic despite a stiff LA.

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Introduction

The principal role of the left atrium (LA) is to modulate left ventricular (LV) diastolic filling and cardiac performance through reservoir, conduit, and booster pump functions, without an elevation in LA pressure.¹ However, in patients with stiff LA, reservoir function is significantly impaired due to extensive LA fibrosis; consequently, the increase in LA pressure during haemodynamic stress is prominent, easily leading to pulmonary venous hypertension and subsequent pulmonary congestion, and eventually results in intractable heart failure.² Although stiff LA syndrome was first described as a complication after mitral valve replacement for long-standing mitral valve diseases,³ it has recently been recognized as a potential sequela of extensive catheter ablation for atrial arrhythmias.^{4,5}

Here, we report a unique case of only mildly symptomatic patient with stiff LA syndrome like haemodynamics after mitral valve surgery and repeated catheter ablation, presumably because iatrogenic inter-atrial shunts function as a pressure-relieving gateway.

Summary figure

Time	Events
75 yrs	She underwent MVR for severe mitral stenosis, Cox-Maze IV procedure for longstanding atrial fibrillation, LA plication due to severely enlarged atrium, and prophylactic LA appendage ligation. Atrial tachycardia recurred soon after the surgery; she underwent a catheter ablation procedure.
76 yrs	She underwent a catheter ablation procedure again.
Initial presentation: 77 yrs	She visited our institution for regular check-up after open-heart surgery. She felt mild shortness of breath when carrying heavy objects or walking long distance.
Admission: 79 yrs	Transthoracic echocardiography revealed pseudo-normal transmitral flow configuration. Pulmonary venous flow signal showed a significantly blunted systolic forward flow, extremely small retrograde flow reversal during atrial contraction, and prominent diastolic flow velocities, all of which indicated significantly devastated LA reservoir function with elevated LA pressure. Cardiac catheter examination revealed characteristic pulmonary capillary wedge pressure waveform, which consisted of a steep ascending limb of v wave with a large peak, consistent with a stiff LA and back pressure of the prominent v wave to the pulmonary circulation which is evident in pulmonary artery pressure waveform.

Continued

Continued

Time	Events
	Cardio-pulmonary exercise testing revealed significantly impaired exercise capacity with peak VO ₂ of only 11.7 mL/min/kg (48% of expected value). Integration of the non-invasive and invasive findings led to a final diagnosis of 'stiff LA syndrome'.
After discharge: 80 yrs	One year after discharge, she has been an outpatient without causing hospitalization for heart failure.

Case presentation

A 79-year-old female with a past medical history of decompensated left-sided heart failure which resulted from severe mitral stenosis and atrial fibrillation visited our institution for regular check-up after open-heart surgery. In 2018, she underwent mitral valve replacement for severe mitral stenosis, Cox-Maze IV procedure for longstanding atrial fibrillation, extensive LA plication, and prophylactic LA appendage ligation. Because atrial tachycardia recurred after the surgery, catheter ablation was performed on the pulmonary veins and mitral isthmus in 2018. Moreover, in 2019, repeated catheter ablation was performed for recurrent atrial tachycardia which originated from pulmonary veins. Thereafter, she felt mild shortness of breath when carrying heavy objects or walking long distance (NYHA functional class II).

She was in a dry-and-warm heart failure profile without leg oedema, jugular venous distension, or cold extremities. Cardiac auscultation revealed a grade 2/6 systolic ejection murmur in the third left intercostal space without extra heart sounds. Electrocardiography showed a normal sinus rhythm with non-specific ST depression on precordial leads. Chest radiography revealed cardiomegaly and bilateral pulmonary arterial dilatation. Interestingly, however, laboratory findings showed a disproportionately low brain natriuretic peptide concentration of only 25.4 pg/mL. Pulmonary venous stenosis was excluded by computed tomography. Although LV systolic function was preserved (see [Supplementary material online, Movie 1](#)), transthoracic echocardiography revealed E wave dominant transmitral flow configuration ([Figure 1A](#)). Moreover, two continuous colour jet flows across the inter-atrial septum were incidentally observed on subxiphoid view (see [Supplementary material online, Movie 2](#)), which was consistent with residual puncture holes at catheter ablation procedures. Of note, peak pressure gradient of the left-to-right flow was measured at 23 mmHg ([Figure 1B](#)), suggesting the existence of severely elevated LA pressure. On the other hand, the LA was not so dilated, with a LA diameter of 38.5 mm and a LA volume of only 36 mL/m². With regard to right ventricular function, TAPSE was measured at 13.6 mm. There was no evidence of prosthetic valve dysfunction with the mean gradient across the prosthetic valve of 6.1 mmHg, and the mitral valve area was calculated as 1.7 cm² (see [Supplementary material online, Movie 3](#)). By Bernoulli's equation, systolic pulmonary arterial pressure was estimated to be 43 mmHg. To further delineate haemodynamic abnormalities, she underwent transoesophageal echocardiography. Although no abnormalities in the prosthetic valve function were found (see [Supplementary material online, Movie 4](#)), transmitral Doppler signal showed prominent E wave of 176 cm/sec followed by rudimentary A wave of only 58 cm/sec. Of note, pulmonary venous flow signal

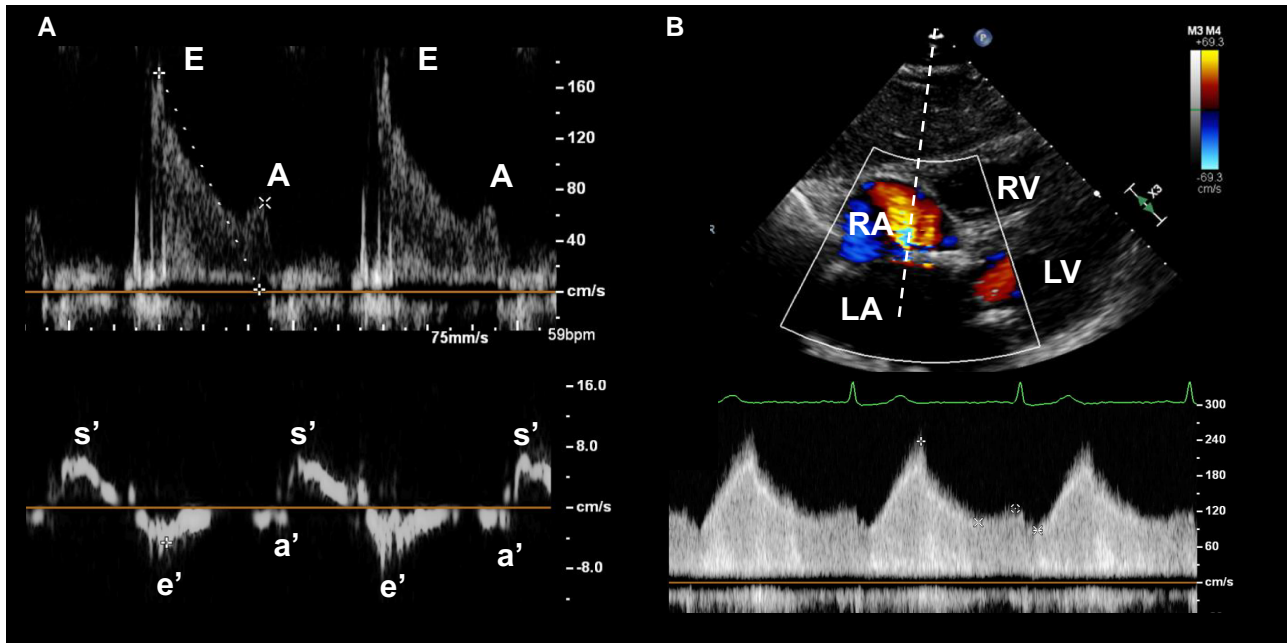


Figure 1 Transthoracic echocardiogram of a 79-year-old woman. Pulsed wave Doppler signal showing significantly enlarged E wave and blunted A wave, resulting in high E/A ratio of 2.40 and the E/e' ratio of 27.4 (A). Subxiphoid view revealed continuous colour flow from the LA to the right atrium (RA). A continuous wave Doppler echocardiography revealed a peak pressure gradient of 23 mmHg, suggesting significantly elevated LA pressure (B). LV, left ventricle; RV, right ventricle.

showed a significantly blunted systolic forward flow, extremely small retrograde flow reversal during atrial contraction, and prominent diastolic flow velocities, all of which indicated significantly devastated LA reservoir function with elevated LA pressure (Figure 2A). Three-dimensional echocardiography clearly shows two inter-atrial shunt flow, and a peak pressure gradient was measured at 36 mmHg (Figure 2B and see Supplementary material online, Movie 5). Speckle-tracking strain analysis revealed LA reservoir strain was critically impaired as 12.1% (Figure 3, normal value: 36–45%).^{6–8} Cardiac catheter examination revealed characteristic pulmonary capillary wedge pressure waveform, which consisted of a steep ascending limb of v wave with a large peak of 17 mmHg (8 mmHg above mean pressure), consistent with a stiff LA (Figure 4). The pulmonary-to-systemic blood flow ratio was calculated as 1.16 by Fick's equation.

Although she was only mildly symptomatic in her daily life, cardiopulmonary exercise testing revealed significantly impaired exercise capacity with peak VO_2 of only 11.7 mL/min/kg (48% of the expected value). Although LV end-diastolic pressure or LV diastolic capacity could not be assessed during invasive haemodynamic study, integration of the non-invasive and invasive findings led to a final diagnosis of stiff LA syndrome.

One year after discharge, she remains oligosymptomatic without experiencing heart failure hospitalization.

Discussion

Stiff LA syndrome was first reported by Pilote *et al.* in 1988.³ They described a patient with pulmonary oedema and intractable heart failure 7 years after mitral valve replacement, along with pulmonary hypertension and a characteristic large v wave on pulmonary capillary wedge pressure waveform despite no prosthetic valve dysfunction. Recent years, however, stiff LA syndrome has been recognized as a sequela

after aggressive catheter ablation due to extensive LA scarring, thus, gained significant prominence.⁴ In this case, several important factors supposed to superimposed and ultimately resulted in non-compliant LA. First of all, chronic pressure overload and overstretch imposed on the LA myocardium due to long-standing severe mitral stenosis and chronic atrial fibrillation presumably resulted in considerable LA fibrosis. In addition to the preoperative fibrotic derangement, extensive Cox-Maze IV procedure appeared to further precipitate iatrogenic LA fibrosis.⁵ Moreover, surgical appendage ligation would further reduce LA compliance; because LA appendage is proposed to be a compliant portion with great reservoir capacity, protecting from elevation in LA internal pressure.⁹ In addition to these surgical procedures, post-operative repeated catheter ablation supposed to contribute to further LA fibrosis.^{3–5} Therefore, one can speculate that these superimposed LA damage and resultant extensive fibrosis would ultimately result in a significantly non-compliant LA in this case.

Moreover, 'small LA' is supposed to further contribute to stiff LA pathophysiology. At the surgery, she underwent aggressive LA plication, which significantly reduced the LA volume from 103 to only 36 mL/m² postoperatively. From a physiological point of view, a small LA is undoubtedly disadvantageous for a stiff LA, because a given increase in volume will produce a much larger increase in the internal pressure for a stiff as well as small chamber based on the LA pressure–volume relationship.

At present, treatment of patients with the stiff LA syndrome is quite challenging and, indeed, limited to diuretics, while its effect is limited and generally results in intractable heart failure. In a recently conducted experimental study, however, inter-atrial 'shunt creation' with the use of dedicated shunt device successfully attenuated the exercise-induced elevation in LA pressure and it showed favourable trends in several haemodynamic and functional end points for patients with heart failure with preserved ejection fraction.¹⁰ This innovative therapy may open

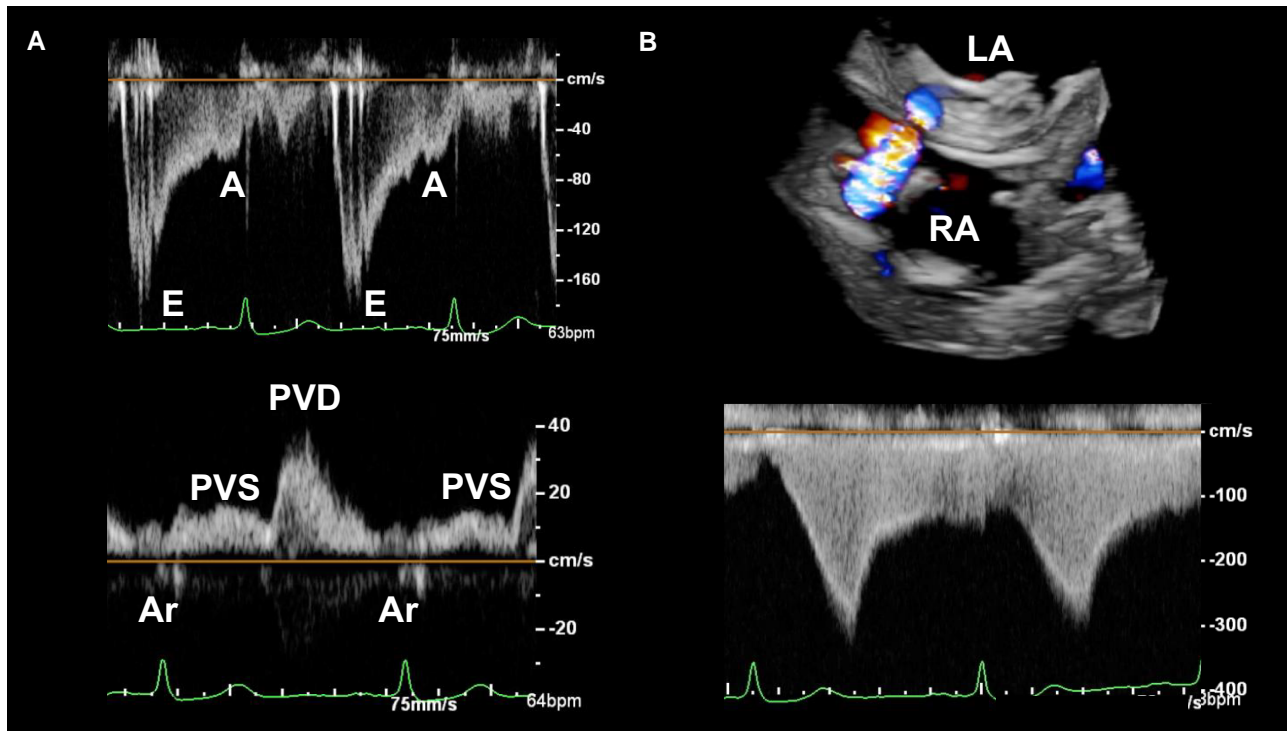


Figure 2 Transesophageal echocardiogram of a 79-year-old woman. Transmitral Doppler signal shows a prominent E wave of 176 cm/sec followed by rudimentary A wave of only 58 cm/sec. Pulmonary venous flow showed a significantly blunted systolic forward (PVS) flow, extremely small retrograde flow reversal (Ar) during atrial contraction, and a prominent diastolic wave (PVD) velocity (A). Three-dimensional transoesophageal echocardiography shows that a peak pressure gradient across the atrial septum was measured at 36 mmHg at end-systole (B). RA, right atrium.

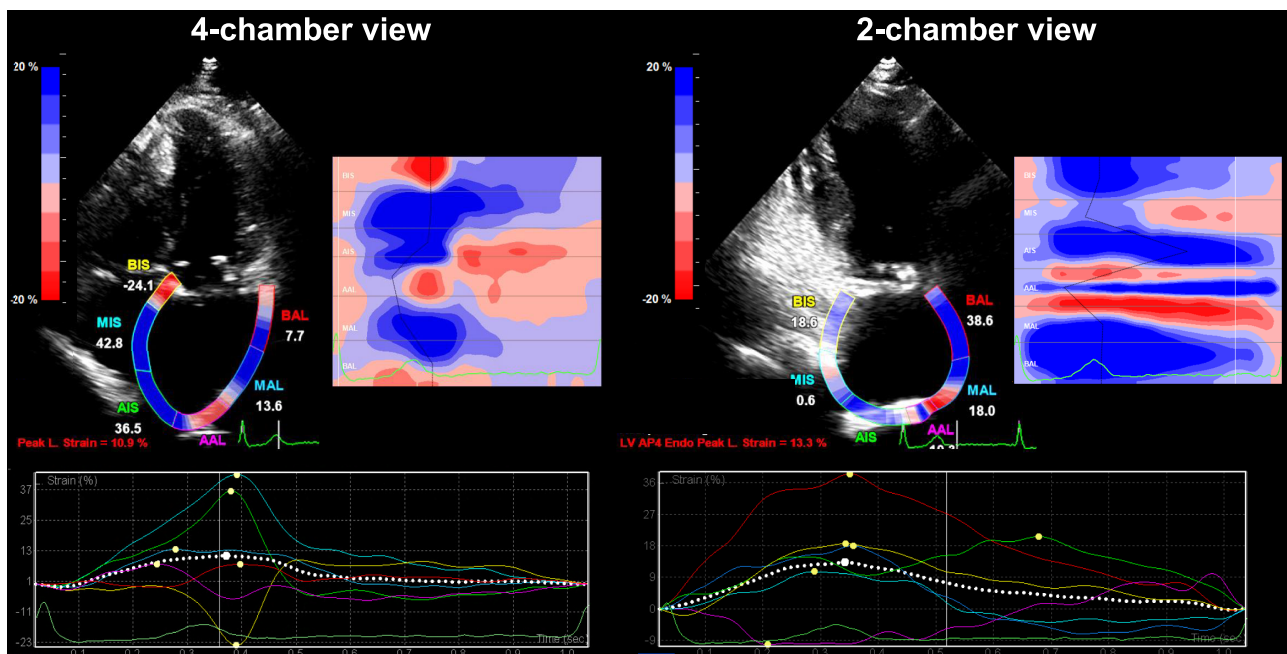
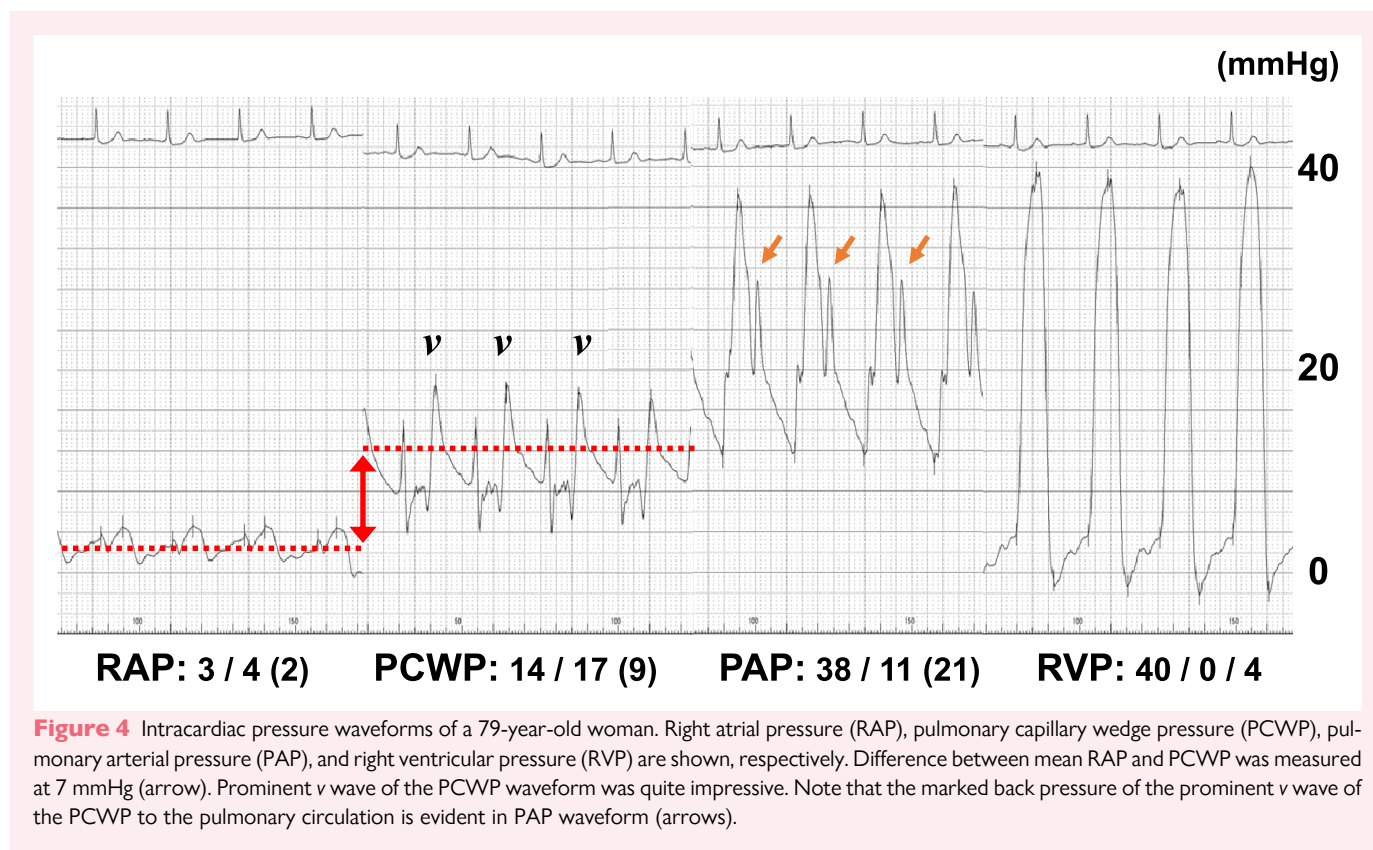


Figure 3 Result of left atrial speckle-tracking strain analysis of a 79-year-old woman. Left atrial (LA) reservoir strain was obtained from both four- and two-chamber views. Averaged value of LA reservoir strain revealed to be critically impaired as 12.1%.

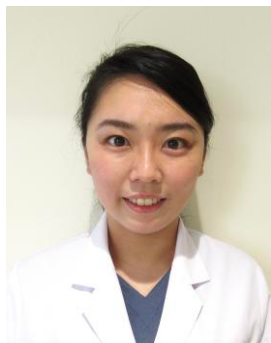


the perspectives to therapeutic breakthrough for patients with intractable stiff LA syndrome.

Conclusion

We experienced a unique case of stiff LA syndrome-like haemodynamics caused by a combination of surgical and catheter procedures. Owing to the inter-atrial pressure-relieving gateway, the patient was only mildly symptomatic despite the existence of a non-compliant LA. This case report may offer unique insights into emerging managements for stiff LA syndrome.

Lead author biography



After graduating from Kobe University in 2018, the author traversed the path of General Internal Medicine and General Cardiology at Ako City Hospital and Prefectural Tamba Medical Centre after 2 years of residency.

Supplementary material

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

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

Conflict of interest: The authors declare no conflicts of interest.

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Data availability

Unidentified data are available upon request.

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