

# The waffle procedure as treatment of a first episode of right heart failure: a case report

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## Background

Constrictive pericarditis is a rare condition which is characterized by pericardial layers thickening and it may not be relieved by conventional medical or surgical therapies. Some patients could profit from specific surgical techniques such as the waffle procedure which removes epicardial tissue strips of the visceral layer of the pericardium. It alleviates diastolic dysfunction, since it is known that the constrictive physiology observed in constrictive pericarditis depends mainly on the visceral layer.

## Case summary

We describe the case of a 59-year-old male complaining of predominantly right heart failure symptoms. Initially, a transthoracic echocardiogram showed pericardial effusion with several fibrous tracts and a constrictive pattern. The work-up was completed with thoracic computed tomography and right heart catheterization along with several microbiological, serological and immunological tests. The final diagnosis was idiopathic constrictive pericarditis. The clinical course was unfavourable, and the patient was referred for prompt surgical treatment using a technique called the waffle procedure. Subsequently, the patient has remained asymptomatic.

## Discussion

Constrictive pericarditis combines features of both constriction and tamponade. Some patients may present an unfavourable clinical course with conventional therapeutic approaches. They could benefit from the waffle procedure which allows an improvement in diastolic and, subsequently, in systolic function.

## Keywords

Waffle procedure • Constrictive pericarditis • Patched epicardiectomy • Case report • Tuberculous pericarditis

## Learning points

- Constrictive pericarditis results in mixed features of both constriction and tamponade and its symptoms are directly associated with right heart failure. As a consequence, diagnosis may be difficult and multiple imaging modalities are required in its diagnosis.
- Some patients could benefit from the waffle procedure which allows an improvement in diastolic and, subsequently, in systolic function.

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## Introduction

Constrictive pericarditis is a rare condition which is characterized by pericardial layers thickening with underlying constrictive physiology.<sup>1,2</sup> There are several causes of constrictive pericarditis: post-cardiac surgery, infectious or radiation therapy. Its diagnosis can be challenging requiring detailed analysis of a variety of investigations due to overlapping signs and symptoms of tamponade or right heart failure. On physical examination, different symptoms and signs such as lethargy, ascites, lower limb oedema, or hypotension along with jugular venous distension or pulsus paradoxus may be found. Some patients may benefit from surgical treatment which allows relief of the constriction.<sup>3,4</sup>

## Timeline

Day/month	Event
5 May 2016	Patient arrives in emergency department complaining of 1-month history of dyspnoea, lower-limb oedema, and ascites.
5 May 2016	Evaluated by a cardiological team, admitted to Cardiology ward. Diuretic treatment is started.
6 May 2016	A first transthoracic echocardiogram is performed—thickening of the pericardial layers, loculated pericardial effusion. Anti-inflammatory treatment is started.
9 May 2016	Computed tomography shows an anterior pericardial effusion.
11 May 2016	Patient undergoes right heart catheterization—elevation and near-equalization of end-diastolic pressures of both ventricles.
13 May 2016	Echocardiography performed—reports similar findings. Ongoing symptoms still present.
16 May 2016	Clinical deterioration. Cardiac magnetic resonance imaging is postponed.
17 May 2016	Surgical treatment (waffle procedure) is performed.
24 May 2016	Favourable post-operative course.
17 June 2016	All microbiological, serological and immunological tests are negative.
1 July 2016	Patient remains asymptomatic. No recurrence of pericardial effusion.

## Case presentation and diagnostic assessment

We describe the case of a 59-year-old Hispanic male with type 2 diabetes on treatment with metformin. He was admitted to our hospital with a 1-month history of heart failure symptoms. He complained of progressive dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, lower-limb oedema extended above the knee, and an increased

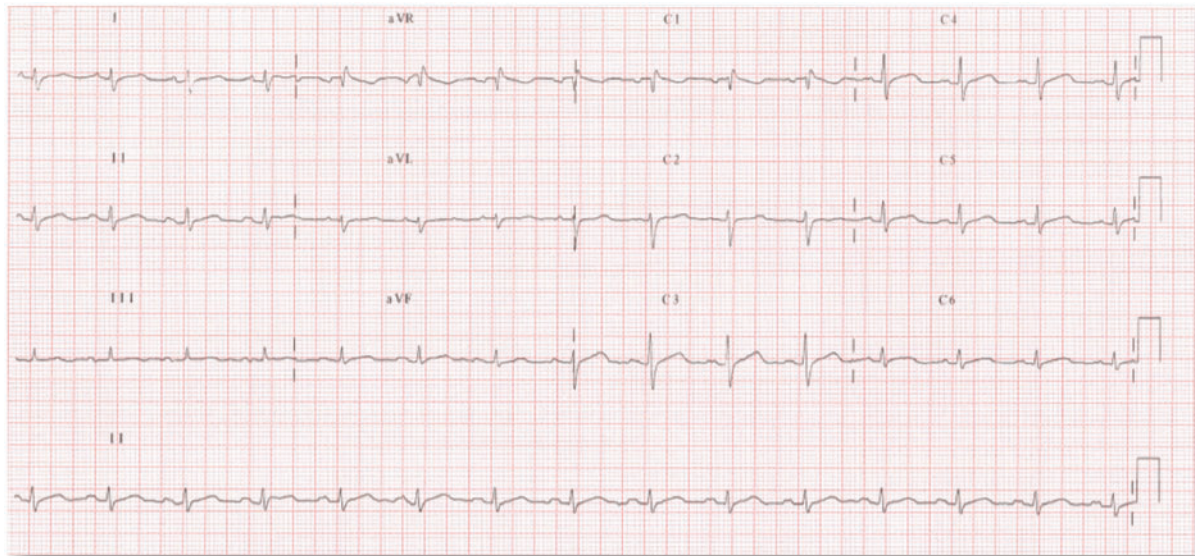
abdominal size. In addition, cough and chest pain worsening during inspiration were described. Blood pressure at admission was 106/68 mmHg and heart rate 90 b.p.m. Physical examination revealed ascites with visible bulging of the flanks and fluid wave, jugular venous distension (the filling level of the jugular vein was 5 cm vertical height above the manubriosternal joint), positive Kussmaul sign, and pulsus paradoxus (the systolic blood pressure fell to 92 mmHg in inspiration) as the most relevant symptoms. Upon cardiac auscultation: S1/S2 heard, no gallops or murmurs heard, no pericardial knock. Pulmonary auscultation: reduced vesicular breath sounds in the lung bases, no rales.

The blood test showed haemoglobin of 13 g/dL (13.5–17.5 g/dL), spontaneous INR of 1.3 (0.9–2), C-reactive protein of 4 mg/L (<0.29 mg/L), NT pro-BNP: 667 pg/mL, and GGT: 95 U/L. A posterio-anterior chest X-ray film showed cardiomegaly and bilateral pleural effusions. The 12-lead ECG is showed in *Figure 1*.

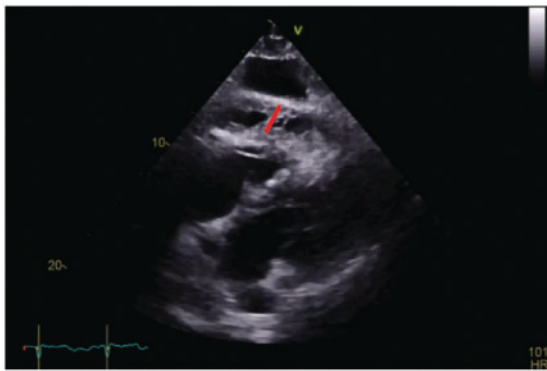
The episode was labelled as a first episode of heart failure, and the patient was admitted to work-up. A transthoracic echocardiogram displayed a preserved left ventricular ejection fraction and a mild right ventricular systolic dysfunction (right ventricular size normal, TAPSE 15 mm). Both atrial sizes were normal. The septum presented an abrupt posterior motion in early diastole with inspiration. It also appeared to 'flutter' as the left and right ventricles filled during diastole. Thickening of the pericardial layers and a loculated anterior pericardial effusion (19 mm) with several fibrous tracts were observed (*Figure 2*). Spectral PW Doppler imaging revealed a restrictive filling pattern in the mitral inflow—(E:A ratio 2.2). It also revealed significant respiratory variation: the mitral E wave velocity decreased more than 25% with inspiration. A contrast enhanced computed tomography (CT) of the thorax was performed to assess thickening and calcification of the pericardium in order to characterize the pericardial effusion (*Figure 3*). The scan also showed contrast in the suprahepatic veins as an indirect sign of systemic veins congestion and heart failure.

An intravenous diuretic treatment (furosemide and spironolactone) was started as well as anti-inflammatory therapy (Ibuprofen 1200 mg daily and colchicine 1 mg daily). In order to rule out infectious and oncological causes of pericardial effusion, tuberculin sensitivity test (Mantoux) and several infectious (*C. burnetti*, *Toxoplasma*, HIV, *Borrelia*, *Rickettsia*, *Brucella*, *Tripanosoma cruzi*, *M.* and *C. pneumoniae*) and oncological markers (alpha fetoprotein, beta-2-microglobulin, CA 19-9, and carcinoembryonic antigen) were tested and were all normal. In the 12-lead ECG there were sinus rhythm and low voltage QRS complexes. A coronary angiography revealed normal coronary arteries and the right heart catheterization showed the following results: (i) right atrial pressure: 21/12 mmHg; (ii) right ventricular pressure: 38/20 mmHg; (iii) left ventricular pressure: 110/24 mmHg; (iv) mean pulmonary artery pressure: 28 mmHg; (v) PWP: 17 mmHg; an (vi) cardiac index: 3.14 L/min/m<sup>2</sup>. They evidenced elevation and near-equalization of the end-diastolic pressures of both ventricles and high right atrial pressure with M morphology and Kussmaul sign. Evidence of dip and plateau was not found because of tachycardia and previous diuretic treatment.

Afterwards, the patient presented a marked clinical worsening due to a slight increase in pericardial effusion size and tamponade which prevented us from performing further tests, and the patient was directly referred to cardiac surgery.



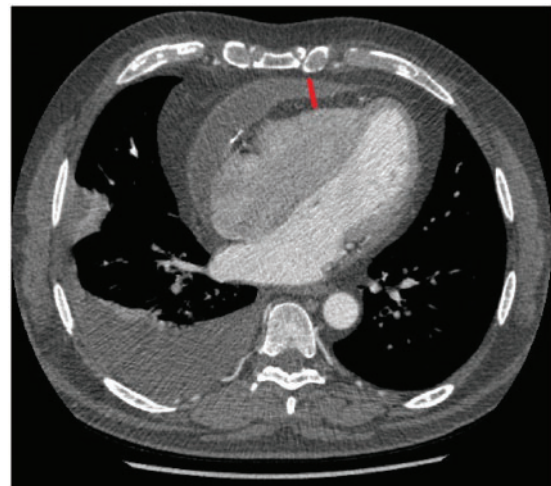
**Figure 1** 12-lead ECG that denotes sinus rhythm and low voltage.



**Figure 2** 2D transthoracic echocardiogram, subcostal view. There is pericardial effusion, fibrinous tracts and very thick pericardial layers.

## Interventions, follow-up, and outcomes

A subtotal phrenic-phrenic pericardiectomy was performed. Subsequently, an off-pump waffle procedure, with several transverse and longitudinal epicardial incisions, was performed ([Figure 4](#) and [Supplementary material online, Video S1](#)). Surgery showed a thickened pericardium with haematic fluid filling the pericardial sac and fibrin tracts. The right heart failure was resolved, and a control echocardiogram excluded pericardial effusion or constriction. The histology, including the polymerase chain reaction (PCR) testing for *Mycobacterium tuberculosis* on pericardial tissue, excluded



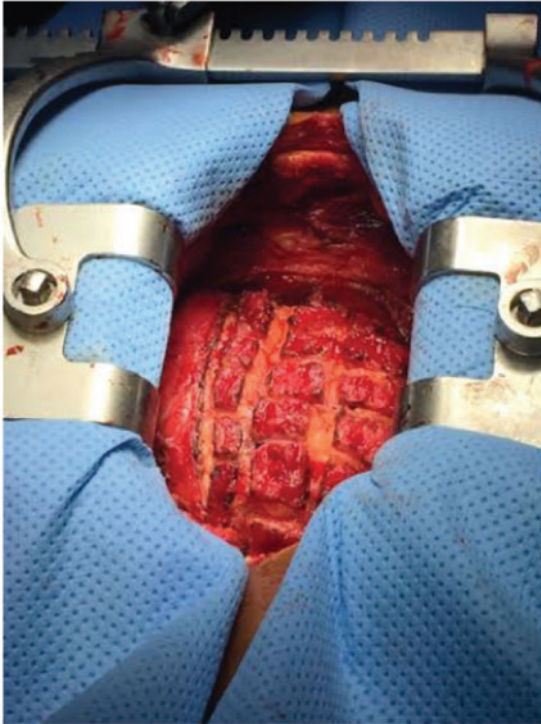
**Figure 3** Contrast enhanced in thoracic CT. Thickened pericardium (arrow), severe anterior pericardial effusion and right pleural effusion were observed.

tuberculosis. Other microbiological and serological tests were negative. The clinical diagnosis was idiopathic constrictive pericarditis. Subsequently, the patient has remained asymptomatic.

## Discussion

Constrictive pericarditis usually results in a combination of features of both constriction and tamponade, and its symptoms are directly





**Figure 4** Off-pump waffle subtotal pericardiectomy. Transverse and longitudinal incisions of visceral pericardium.

associated with right heart failure. In consequence, the diagnosis may be difficult and multiple imaging modalities are required to diagnose it. Magnetic resonance imaging, over recent years, has shown itself to be important not only to establish the severity of the disease but also to inform the clinical course,<sup>5</sup> but the clinical situation of the patient prevented us to perform it. In some cases, there can be concomitant pericardial effusion and elevation of the right atrial and pulmonary wedge pressures following drainage of the pericardial fluid which point to the constrictive process.<sup>4,6-8</sup> Unfortunately, despite considering performing a pericardiocentesis, both as a therapeutic method and also to define better the disease, the characteristics of the pericardial effusion with fibrous tracts together with the poor clinical status of the patient led us to choose surgery as the most effective and long-lasting treatment in this particular case.

Concerning the aetiology, the most common cause is idiopathic (up to 61%), followed by post-cardiac surgery (37%). Other causes are post-infectious tuberculous or purulent pericarditis (3–15%) and radiation therapy.<sup>2,4,9,10</sup> Imaging and analytical results of this case agree with an idiopathic constrictive pericarditis. Interestingly, all serological and microbiological tests, including tuberculosis with PCR testing for *M. tuberculosis* on pericardial tissue, were negative. The sensitivity of pericardial biopsy for diagnosis of tuberculous pericarditis ranges from 10 to 64%<sup>11,12</sup> and a normal pericardial biopsy does not exclude tuberculosis,<sup>13</sup> whereas PCR testing for *M. tuberculosis* is more sensitive (>80%) than other tests when it is performed on pericardial tissue.<sup>14</sup>

Regarding the surgery, pericardiectomy is an effective treatment. However, it is known that in constrictive pericarditis the constriction depends mainly on the visceral layer of the pericardium so a radical pericardiectomy alone could not be effective in patients with a marked epicardial constriction as described by Heimbecker et al.<sup>3,4,6,15</sup> The waffle procedure consists of removing longitudinal and transverse epicardial tissue strips of the visceral layer. This procedure must be started on the left ventricle and continued on the right ventricle, since the left ventricle would suffer from massive preload if a waffle incision were to be made first on the right ventricle.<sup>15</sup> Patched epicardiectomy allows an improvement in diastolic function by relieving the constriction and subsequently allows an improvement in systolic function.<sup>3,4,6</sup>

## Conclusions

The waffle procedure is an effective therapy in relieving pericardial constriction. Radical pericardiectomy alone may not be effective enough in some patients who require a more aggressive strategy to improve diastolic dysfunction.<sup>15</sup>

## Supplementary material

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

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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 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.

## References

- Hancock EW. Subacute effusive-constrictive pericarditis. *Circulation* 1971;**43**: 183.
- Cameron J, Oesterle SN, Baldwin JC, Hancock EW. The etiology spectrum of constrictive pericarditis. *Am Heart J* 1987;**113**:354.
- Adler Y, Charron P, Imazio M, Badano L, Barón-Esquivias G, Bogaert J, et al. 2015 ESC Guidelines for the diagnosis and management of pericardial diseases: the Task Force for the Diagnosis and Management of Pericardial Diseases of the European Society of Cardiology (ESC) Endorsed by: The European Association for Cardio-Thoracic Surgery (EACTS). *Eur Heart J* 2015; **36**:2921.
- Sagrístà-Sauleda J, Angel J, Sánchez A, Permanyer-Miralda G, Soler-Soler J. Effusive-constrictive pericarditis. *N Engl J Med* 2004;**350**:469.
- Cremer PC, Kumar A, Kontzias A, Tan CD, Rodriguez ER, Imazio M, Klein AL. Complicated pericarditis: understanding risk factors and pathophysiology to inform imaging and treatment. *J Am Coll Cardiol* 2016;**68**:2311–2328.
- Hancock EW. A clearer view of effusive-constrictive pericarditis. *N Engl J Med* 2004;**350**:435.

7. Shabetai R. *The Pericardium*. Norwell, MA: Kluwer Academic Publisher; 2003. p227.
8. Osterberg L, Vagelos R, Atwood JE. Case presentation and review: constrictive pericarditis. *West J Med* 1998;**169**:232–239.
9. Hugo-Hamman CT, Scher H, De Moor MM. Tuberculous pericarditis in children: a review of 44 cases. *Pediatr Infect Dis J* 1994;**13**:13.
10. Bansal R, Perez L, Razzouk A, Wang N, Bailey L. Pericardial constriction after cardiac transplantation. *J Heart Lung Transplant* 2010;**29**:371.
11. Komsouglu B, Goldeli O, Kulan K, Komsouglu SS. The diagnostic and prognostic value of adenosine deaminase in tuberculous pericarditis. *Eur Heart J* 1995;**16**:1126.
12. Schepers GW. Tuberculous pericarditis. *Am J Cardiol* 1962;**9**:248.
13. Cheitlin MD, Serfas LJ, Sbar SS, Glasser SP. Tuberculous pericarditis: is limited pericardial biopsy sufficient for diagnosis? Report of two cases. *Am Rev Respir Dis* 1968;**98**:287.
14. Cegielski JP, Devlin BH, Morris AJ, Kitinya JN, Pulipaka UP, Lema LE, Lwakatare J, Reller LB. Comparison of PCR, culture, and histopathology for diagnosis of tuberculous pericarditis. *J Clin Microbiol* 1997;**35**:3254.
15. Heimbecker RO, Smith D, Shimizu S, Kestle J. Surgical technique for the management of constrictive epicarditis complicating constrictive pericarditis (the Waffle procedure). *Ann Thorac Surg* 1983;**36**:605–606.