

# Endocarditis and coronary artery fistula: a case report<sup>†</sup>

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## Introduction

Coronary artery fistulae are rare abnormal congenital communications between a coronary artery and a cardiac chamber or great vessel. The majority of adult patients are asymptomatic, and it is most commonly discovered incidentally on coronary angiography.

## Case Presentation

We present the case of a 50-year-old woman, with a known fistula connecting the right coronary artery (RCA) and right atrium (RA), presenting with aortic valve endocarditis and pulmonary emboli. We detail the presentation and echocardiographic findings of aortic valve endocarditis with extension of the vegetation into the RA via the giant RCA fistula. We describe the clinical course including initial therapy, embolization of the right atrial vegetation to the lungs, and ultimately successful surgical correction after prolonged antibiotic therapy.

## Discussion

Patients with coronary artery fistulae are susceptible to potentially serious complications including myocardial ischaemia, shunting and in this case infective endocarditis. We review the literature and discuss timings for corrective intervention.

## Keywords

Coronary artery fistula • Endocarditis • Echocardiography • Cardiac computerized tomography • Case report

## Learning points

- Coronary artery fistulae are rare abnormal congenital communications between a coronary artery and a cardiac chamber or great vessel. Most fistulae drain from the right coronary artery into the right ventricle but up to 24% connect to the right atrium or central veins.
- Long term complications arise from myocardial ischaemia, heart failure due to shunting, or bacterial endocarditis and as such patients should be routinely followed up. Infective endocarditis complicating coronary artery fistulae predominantly affects those with dilated vessels and a unilateral fistula draining into the right heart. Due to the potential severity of complications some authors recommend closure (surgical or transcatheter) in all cases but the majority still advocate intervention only in the presence of symptoms or complications.

## Introduction

Coronary artery fistulae are an abnormal congenital communication between a coronary artery and a cardiac chamber or great vessel. The majority of adult patients are asymptomatic, and it is most commonly discovered incidentally on coronary angiography.<sup>1</sup> It is a rare finding: during a 10 year study of invasive coronary angiography a fistula was uncovered in only 0.2% of patients.<sup>2</sup> The most common presenting symptoms are dyspnoea and chest pain whilst clinical examination may reveal a continuous murmur.<sup>3</sup> Most fistulae drain from the right coronary artery (RCA) into the right ventricle but up to 24% connect to the right atrium (RA) or central veins.<sup>4</sup> Long term complications arise from myocardial ischaemia, heart failure due to shunting, or bacterial endocarditis. Due to the potential severity of complications some authors recommend closure (surgical or transcatheter) in all cases<sup>5</sup> but the majority still advocate intervention only in the presence of symptoms or these complications.<sup>1</sup> Infective endocarditis complicating coronary artery fistulae predominantly affects those with dilated vessels and a unilateral fistula draining into the right heart.<sup>6</sup>

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<sup>†</sup> ESC Clinical Case Gallery - Best Cases of the Year (2017): Second Place.

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## Timeline

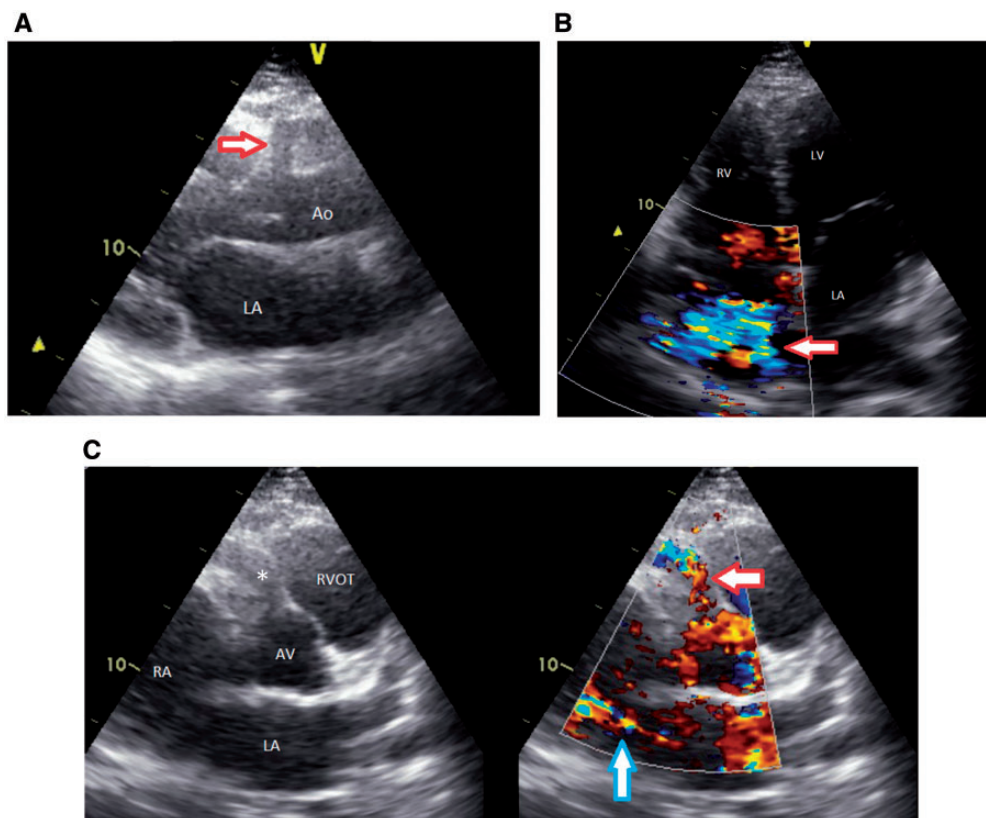
| Date | Events   |
|------|--|
| 2013 | <p>Incidental finding of a murmur</p> <p>Open access echocardiogram identifies a dilated vessel from the right coronary cusp with abnormal colour flow into the right atrium (RA)</p> <p>Computerized tomography confirms giant right coronary artery (RCA) with fistula to the RA</p>   |
| 2015 | <p>Presented with dyspnoea, low grade fevers, weight loss, and lower back discomfort</p> <p>Echocardiography and blood cultures confirm aortic valve endocarditis with extension of the vegetation into the RA via the giant RCA fistula</p> <p>Deterioration with presumed septic emboli to the lungs</p> <p>Successfully underwent aortic valve replacement and RCA fistula ligation after a prolonged course of intravenous antibiotics</p> |

## Case Presentation

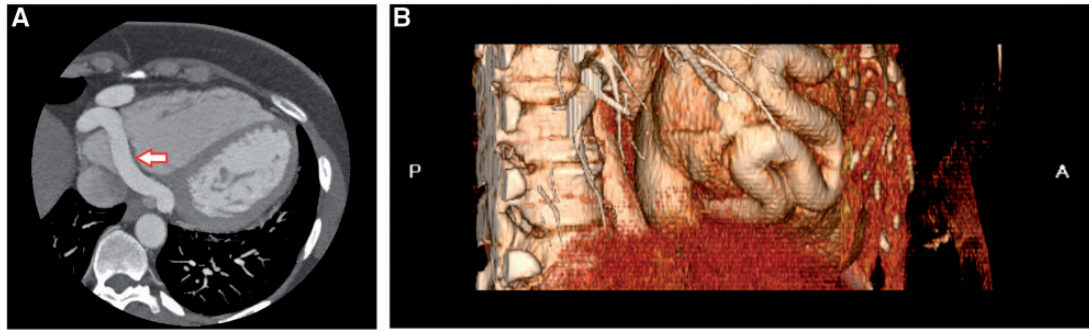
A 50-year-old woman initially presented in 2013 through the open access echocardiography service with the incidental finding of a systolic murmur. Her echocardiogram (*Figure 1*) revealed a dilated vessel arising from the right coronary cusp with consequential turbulent colour flow in the RA. She denied any symptoms, and there was no significant past medical history.

She went on to have a cardiac computerized tomography (CT) scan (*Figure 2*), which confirmed a giant RCA with a connecting fistula into the RA. A myocardial perfusion scan was negative for ischaemia. It was felt there was no indication for intervention, at that time, given the patient was asymptomatic with normal biventricular function and no inducible ischaemia. She remained under routine follow-up due to the potential for complications.

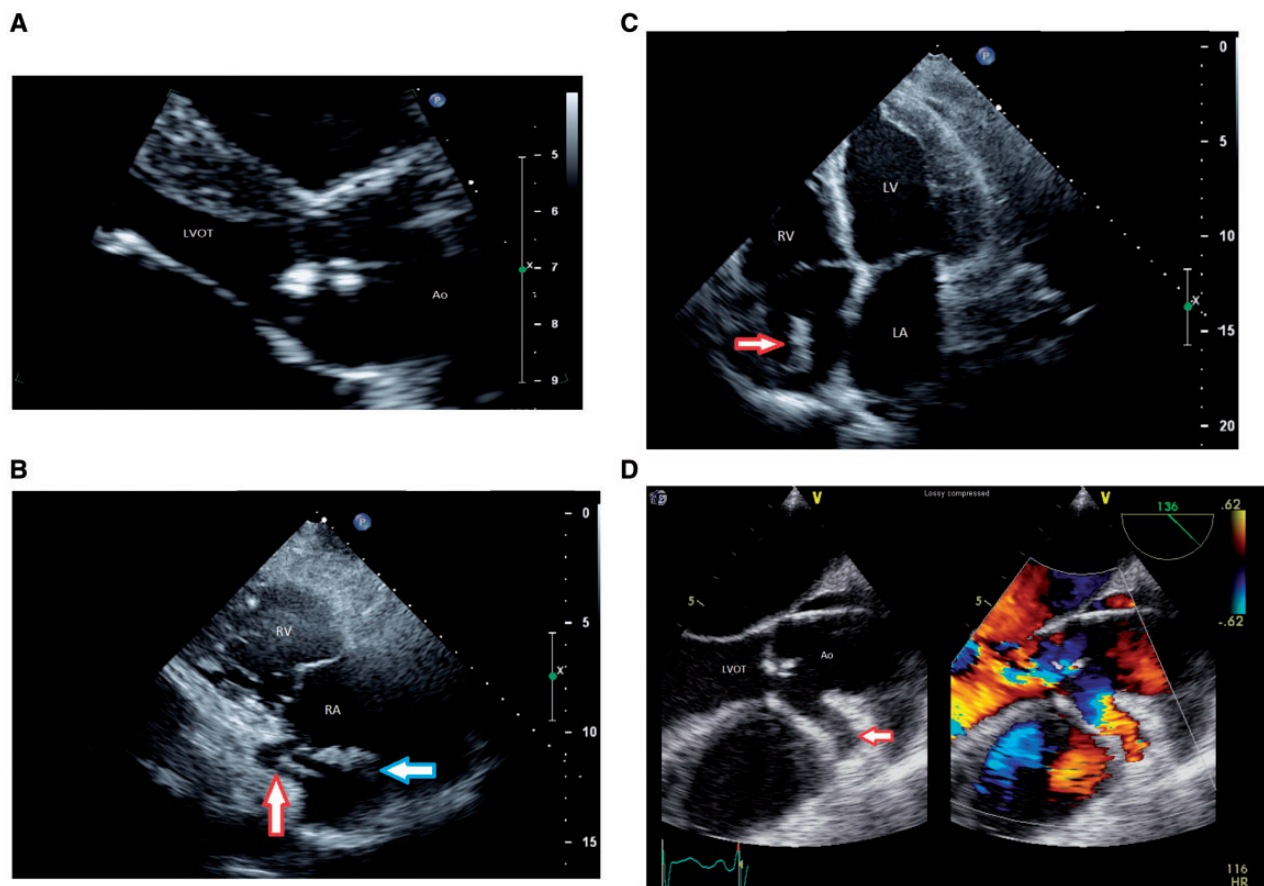
The patient represented 2 years later—between follow-up appointments—with a few months history of dyspnoea, low grade fevers, weight loss, and lower back discomfort. She was pyrexial and tachycardic. Initial laboratory data revealed a microcytic anaemia and raised C-reactive protein (CRP): haemoglobin 64 g/L (normal range 115–165), mean corpuscular volume 78.9fl (83–101), and CRP 142 mg/L (0–5). The admission chest X-ray was clear and the electrocardiogram showed sinus tachycardia only. In view of the indolent



**Figure 1** Original echocardiography pictures from 2013 detailing the dilated vessel arising from the right coronary cusp with consequential turbulent colour flow in the right atrium. (A) Parasternal long axis two-dimensional view of the dilated vessel (red arrow). (B) Apical four chamber illustrating abnormal colour flow in the right atrium (red arrow). (C) Parasternal short axis demonstrating the dilated vessel (star) with colour flow in the vessel (red arrow) and into the right atrium (blue arrow). Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle.



**Figure 2** (A) Computerised tomography demonstrating the giant right coronary artery (red arrow). (B) Three-dimensional reconstruction displaying the size and tortuosity of the vessel.



**Figure 3** Echocardiography demonstrating aortic valve endocarditis, aortic incompetence and vegetation protruding into the right atrium. (A) Parasternal long axis view of the aortic valve masses. (B) Angulated parasternal view of the right ventricular inflow illustrating the right coronary fistula (red arrow) and vegetation into the right atrium (blue arrow). (C) Apical four chamber view with tip of the vegetation seen in the right atrium (red arrow). (D) Transoesophageal echocardiogram highlighting the abnormal aortic valve, aortic regurgitation, and giant right coronary artery (red arrow). Ao, aorta; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; RA, right atrium; RV, right ventricle.

history the differential diagnoses included malignancy, autoimmune conditions, and infective processes such as discitis or bacterial endocarditis. Computerised tomography chest/abdomen/pelvis did not

reveal any signs of malignancy or spinal lesions but highlighted an occlusive pulmonary filling defect in the left lower lobe consistent with a pulmonary embolus. The patient went on to have transthoracic

echocardiography (Figure 3) demonstrating new dilatation of the left ventricle with mildly impaired systolic function. There were new echobright masses on the right and non-coronary cusps of the aortic valve with associated moderate aortic incompetence. There was a large and highly mobile structure in the RA originating from the distal communication of the RCA fistula. She was diagnosed with aortic valve endocarditis with extension of the vegetation down the giant RCA into the RA via the fistula. Subsequent blood cultures grew a gram positive cocci and the patient was treated according to local antimicrobial policy. The patient went on to have a transoesophageal echocardiogram (Figure 3) to further detail the aortic valve and regurgitation.

The patient initially improved with medical therapy but unfortunately then became more breathless with a rise in her oxygen requirements. Computerised tomography pulmonary angiography revealed bilateral lobar pulmonary emboli with associated infarction and consolidation. Repeat echocardiography showed progressive left ventricular dilatation with worsening of the aortic regurgitation. The right atrial vegetation was no longer visible suggesting embolization to the lungs.

The patient completed a prolonged course of intravenous antibiotics with clinical and radiological recovery from the septic emboli. She successfully underwent aortic valve replacement and ligation of the RCA fistula. She remains well and symptom free at 2 years after discharge.

## Discussion

This case demonstrates the rare, often incidental initial finding of coronary artery fistulae. When first diagnosed the patient was completely asymptomatic and clinically well without evidence of heart failure, significant shunting, or ischaemia. The decision was made, after consultation with the regional congenital heart disease team, not to proceed to surgical correction. This approach is supported by most authors.<sup>1</sup> A recent small case series by Gräni *et al.*<sup>7</sup> suggested hybrid imaging with coronary computed tomography angiography and positron emission tomography is superior to perfusion imaging alone at detecting impaired myocardial blood flow in patients with complex coronary artery anomalies. The significance of such changes has yet to be assessed prognostically but could potentially aid risk stratification in such patients. Interestingly, there is no clear correlation between shunt size and either symptoms or likelihood of developing infective endocarditis in patients with coronary artery fistulae.<sup>6</sup>

Unfortunately, she presented 2 years later with the recognized complication of endocarditis. Although the history was indolent and non-specific, echocardiography and blood cultures proved diagnostic. The patient's clinical course was complicated by septic emboli to the lungs and progressive aortic regurgitation. A review of 25 such cases by Said in 2016<sup>6</sup> found septic emboli were reported in 24% of cases

and the author recommended prophylactic antibiotics for all patients with coronary artery fistulae.

The timing of surgical intervention in endocarditis can be difficult and in this case urgent intervention was felt to be high risk. The European Society for Cardiology guidance is for urgent surgery in the presence of locally uncontrolled infection (abscess, false aneurysm, fistula, or enlarging vegetation) or persistent vegetations >10 mm after  $\geq 1$  embolic episode despite appropriate antibiotic therapy.<sup>8</sup> At presentation—after detailed discussions between the Cardiology and Cardiothoracic surgical teams—the consensus was that in the absence of these features and given the indolent presentation, it was appropriate to treat with antibiotics and observe. Once the right atrial vegetation had embolized to the lungs both teams felt prolonged antibiotics and supportive care were still the best option: the aortic valve vegetations were <10 mm, there had been no embolization to the systemic circulation, and any surgery would be better undertaken once the infection had cleared. The patient responded well to a prolonged course of antibiotics and was able to undergo successful aortic valve replacement and ligation of the RCA fistula when aseptic. The patient ultimately had a good outcome and remained well and symptom free at follow-up.

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

**Conflict of interest:** none declared.

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