Detection of myocardial ischaemia caused by coronary artery-left ventricular fistulae using myocardial contrast echocardiography

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A coronary artery-left ventricular fistula (CAF) is an extremely rare anatomical abnormality in which blood drains directly from a coronary artery into the left ventricle. CAF may cause myocardial ischaemia and angina. Myocardial contrast echocardiography (MCE) is a non-invasive technique which assesses myocardial perfusion. We describe a patient with CAF in whom transmural myocardial ischaemia was demonstrated using MCE.

KEYWORDS
Myocardial ischaemia; Coronary artery-left ventricular fistula; Myocardial contrast echocardiography

Case report
A 73-year-old man was admitted with history of cardiac sounding chest pain at rest. Apart from mild chronic obstructive pulmonary disease (COPD) which was well controlled with inhalers, there was no other significant past medical history. Systemic examination was unremarkable. Serial ECGs and 12-h post-admission troponin were normal. A 12-lead exercise treadmill ECG was stopped at 3 min and 28 s due to chest tightness and breathlessness. There were no significant ECG changes. Transthoracic echocardiography was normal. Selective coronary angiography demonstrated a normal left main stem, normal right coronary artery, and a non-flow limiting stenosis in the proximal circumflex artery. In the left anterior descending artery, multiple coronary artery-left ventricular fistulae (CAF) were observed to arise from a large proximal diagonal vessel and drain directly into the left ventricle (see arrow in Figure 1 and Supplementary data, Video 1). We performed both real-time imaging to assess wall motion, and low power myocardial contrast echocardiography (MCE) with triggered imaging following microbubble destruction to assess myocardial perfusion, with dobutamine as the stressor agent. The patient was on no anti-anginal medications and in the light of patient’s history of chronic obstructive pulmonary disease, dipyridamole was not used. End-systolic images were obtained with a low mechanical index (MI) of 0.1 with high-intensity pulses to facilitate microbubble destruction followed by acquisition of 10 end-systolic frames. Resting images showed both normal wall motion and myocardial perfusion (Figure 2 and Supplementary data, Video 2). As peak stress wall motion remained normal, however, following microbubble destruction, there was a transmural perfusion defect in the apical anterior and apical anterolateral walls, with normal perfusion in the remaining walls (Figure 3 and Supplementary data, Video 3). Owing to the size and multiplicity of the fistulae, medical management in the form of anti-anginal agents was opted. The patient has not had a recurrence of his angina 1 year after his initial presentation.

Discussion
A coronary artery fistula is a communication between one of the coronary arteries and a cardiac chamber or vein. Coronary artery fistulae have an incidence of 0.2% in patients undergoing diagnostic cardiac catheterization.1 The right and left coronary arteries are involved in 55 and 35% of cases, respectively, with involvement of both vessels in 5% of cases.2 Coronary artery fistulae may manifest as a single large communication in an individual or as multiple tiny communications.2 They represent a shunt between the coronary system and the cardiac chamber into which they drain. The most common drainage sites are the right ventricle (40%) and right atrium (23%).1 Coronary artery-left

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ventricular fistulae are exceedingly rare with the incidence being reported as 1.2% of all coronary artery fistulae. In the presence of a small-shunt myocardial blood flow is not compromised. However, large shunts may present with pulmonary oedema, pulmonary hypertension, infective endocarditis, rupture, or thrombosis of the fistula, an associated arterial aneurysm or myocardial ischemia distal to the fistula (‘myocardial steal phenomenon’.) Transcatheter and surgical closure techniques have been described for the treatment of this anomaly. However, interventional procedures are reserved for large, clinically significant coronary artery fistulae. The presence of multiple small fistulae and the control of symptoms with medical management in our patient precluded the use of these options.

Transthoracic echocardiography has been shown to be able to visualize large coronary artery fistulae directly. The vessels from which these fistulae arose were dilated and continuously turbulent on colour Doppler echocardiography. However, the majority of patients were children with large and proximally located congenital coronary artery fistulae. The authors concluded that the findings could not be extrapolated to an adult population.

There are several possible reasons why the CAF were not visualized directly on transthoracic echocardiography. First, the multiple CAF in this case were distally positioned in the coronary circulation and were of smaller calibre. These factors made them much more difficult to detect, particularly during ventricular systole. However, it is possible that they may have been visualized with contrast if a higher MI had been used. This could have allowed visualization of both myocardial tissue and contrast agent simultaneously. The reason for a lack of wall motion defect is due to the lower sensitivity of wall motion in comparison with perfusion in the detection of ischaemia. Contrast-enhanced transoesophageal echocardiography has also been used to diagnose CAF in adults and is thought to be more sensitive.

Cardiac magnetic resonance imaging has been shown recently to demonstrate global subendocardial ischaemia due to multiple coronary-ventricular fistulae arising from all three major coronary arteries. Our group used low-power MCE, which demonstrated a transmural myocardial perfusion defect in the territory of the LAD wall despite normal wall motion. To our knowledge, this is the first report in which MCE has demonstrated myocardial ischaemia in the context of CAF.

**Supplementary data**

Supplementary data are available at European Journal of Echocardiography online.

**References**

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