A case of coronary cameral fistula

Gareth J. Padfield*

Department of Cardiology, Edinburgh Royal Infirmary, Edinburgh, UK

Received 10 February 2009; accepted after revision 10 April 2009; online publish-ahead-of-print 4 May 2009

An 85-year-old woman underwent transthoracic echocardiography for the investigation of breathlessness and atypical chest discomfort. Clinical examination was unremarkable. A standard 12 lead ECG demonstrated anterior T wave inversion, but was otherwise normal. Transthoracic echocardiography demonstrated a normally functioning left ventricle with hypertrophy and trabeculation of the apical and lateral segments. Imaging with colour flow Doppler demonstrated blood flow from the epicardial surface into the left ventricular cavity through the hypertrophied segment of myocardium during diastole. A diagnosis of multiple, diffuse coronary-left ventricular fistulae predominantly of a large diagonal branch of the left anterior descending artery was made at coronary angiography. The patient responded well to oral beta-blockade, reporting an improvement in symptoms 2 months later in the outpatient clinic. The echocardiographic appearances of coronary fistulae may cause diagnostic confusion, particularly in the presence of myocardial hypertrophy and trabeculation.

KEYWORDS
Coronary fistula; Congenital abnormality

Discussion

Coronary fistulae with the cardiac chambers (Cameral fistulae) are rare congenital vascular anomalies reported to be found in approximately 0.08–0.3% of unselected patients undergoing diagnostic coronary angiography.1,2 Such fistulae have been described as arterio-luminal, where there is direct and focal communication with the cardiac chamber concerned, or arterio-sinusoidal, as in this case where arterial blood communicates with the cardiac chambers via a sinusoidal network. An arterio-capillary variant has also been described.3 The vast majority (~90%) of cameral fistulae communicate with the right-sided chambers of the heart and in the remainder of cases will drain to the left side of the heart or to both.4 Fistulae usually arise predominantly from one of the two major coronary arteries, however, in a small proportion of cases (~5%) communications may arise from both coronary arteries.5

Clinical presentation will generally depend on the haemodynamic significance of the anomaly and most commonly coronary artery fistulae are asymptomatic and are found incidentally.2 Anginal symptoms may be the presenting feature, particularly in patients with multiple fistulae, and

* Corresponding author: Centre for Cardiovascular Science, The University of Edinburgh, Chancellor’s Building, Edinburgh EH16 4SU, UK.
Tel: +44 131 242 9475; fax: +44 131 242 6379.
E-mail address: gareth.padfield@ed.ac.uk

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in those patients with a single fistula, exertional dyspnoea is more likely to predominate.6 Inducible ischaemia has been well demonstrated in these patients and is thought to occur as a result of left to left shunting causing a coronary steal phenomenon and diastolic overload.4 Coronary cameral fistulae may also cause myocardial infarction,7 congestive heart failure, arrhythmias, and aneurysmal formation and rupture of affected vessels may also occur.8 Turbulent blood flow may predispose to the development of infectious endocarditis although this is not well substantiated. Such complications are more common in older patients, in part due to the progressive enlargement of coronary cameral fistulae over time secondary to abnormal haemodynamics. Cameral fistulae are recognized to be associated with regional hypertrophy as in this case although it is unclear whether hypertrophy in this context is a cause or effect of the abnormal coronary anatomy.9 The appearance of blood flow within the myocardium may cause diagnostic uncertainty. The trabeculated appearance of the LV on transthoracic echocardiography in this case raised the suspicion of left ventricular non-compaction; however, inspection of the colour Doppler imaging clearly revealed a sustained flow of blood from the epicardial surface to the LV cavity throughout diastole more suggestive of a coronary fistula. In this case, the diagnosis was established by conventional coronary angiography, though the coronary anatomy may alternatively be delineated using computed tomography coronary angiography.10

The best way to manage cameral fistulae is uncertain largely due to the rarity of the condition. Patients in whom focal fistulae with large shunts exist may benefit from closure of the shunt, and if this is to be performed it is probably best done as early as possible.8 Multiple fistulae of the sinusoidal type, such as in this case, are unlikely to be amenable to surgical correction and have, as in this case, been treated successfully with beta-blockade.11,12

Supplementary material
Supplementary material is available at European Journal of Echocardiography online.

Acknowledgements
I would like to acknowledge Catherine Quinn of the Echocardiography department of Edinburgh Royal Infirmary for performing the echocardiographic study.

References

