A rare type of ‘coronary arterial — left ventricular fistula’ via thebesian veins in a Fragile X syndrome carrier

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A 62 year-old woman, who was initially investigated for syncopal attacks, was referred to the cardiology department for further assessment of an incidentally found heart murmur. Clinical examination revealed a continuous murmur best heard in the pulmonary area. She was known to be a carrier for Fragile X syndrome. All the previous investigations performed to evaluate her syncopal episodes had been normal. A transthoracic echocardiogram showed multiple colour flow jets in the left ventricular (LV) cavity arising from lateral and septal walls.

Transesophageal imaging with colour Doppler revealed blood flow from the epicardial surface into the LV cavity through the lateral and septal walls of the myocardium in both systole and diastole. However, there was no evidence of any valvular or patent ductus arteriosus malformations. Coronary angiogram demonstrated a coronary arterial — LV fistula. Communication was seen between the left anterior descending artery and LV, as well as the obtuse marginal artery and LV via a leash of thin vessels, classically described as ‘thebesian veins’ [1], that drained from these arteries directly into the LV cavity (Figs. 1, 2).

Structural cardiac abnormalities such as mitral valve prolapse and dilatation of aorta have been re-
ported in patients suffering from Fragile X syndrome [2]. To the best of our knowledge, this is the first reported case of coronary arterial — LV fistula in a patient with a Fragile X syndrome carrier state.

Acknowledgements

The authors do not report any conflict of interest regarding this work.

References