A 56-year-old male patient presented with dyspnea on mild exertion (New York Heart Association-Class III), which had remained stable over previous few years. His medical history was otherwise unremarkable. On admission, physical examination revealed a holosystolic III/VI murmur over the apex and an audible third heart sound on auscultation. The patient also had bilateral fine crackles and pedal edema. Echocardiography showed biatrial enlargement, severe mitral regurgitation with an eccentric jet, and severe pulmonary artery hypertension with an estimated left ventricular ejection fraction (LVEF) of 50% (Video 1*). Additionally, a continuous flow (arrows pointing to the fistulous dilatation) was noted in the coronary sinus indicating a pattern compatible with an arteriovenous (AV) fistula in the short-axis view of the left ventricle (Figure A, Video 2 and 3*). The mitral valve annulus seemed to be intact despite the tortuous course of AV malformation. Three-dimensional, volume rendered reconstruction of CT angiographic images were obtained, and showed the course of the left circumflex artery connecting to the coronary sinus over the course of aneurysmal dilatation of the left circumflex artery and tortuous clusters of AV malformation (Figure B and C). Coronary AV malformation was further confirmed by conventional coronary angiography (Figure D, Video 4*). No underlying coronary artery disease was found in coronary angiographic studies. The patient underwent surgical ligation of the fistula and Alfieri edge-to-edge mitral valve repair, following which there was no residual fistula and only trace mitral regurgitation.

**Figures**—(A) An echocardiographic image from short-axis view of left ventricle. Arrows point to the fistulous dilation of circumflex artery. (B) Three-dimensional, volume-rendered reconstruction of CT angiographic image showing the course of the left circumflex artery to coronary sinus fistula. (C) CT angiographic image showing the course of the coronary fistula. (D) Conventional coronary angiography showing the course of the coronary arteriovenous fistula. (AML: Anterior mitral leaflet; AVM: Arteriovenous malformation; AVM of LCx: Arteriovenous malformation of left circumflex artery; CS: Coronary sinus; LAD: Left anterior descending artery; LMC: Left main coronary artery; LCx: Left circumflex artery; PML: Posterior mitral leaflet, RA: Right atrium; RV: Right ventricle; SVC: Superior vena cava) *Supplementary video files associated with this presentation can be found in the online version of the journal.