Left coronary sinus to pulmonary artery fistula, complicated by pulmonary valve endocarditis

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A 63-year-old woman presented to hospital with a 4-day history of constitutional symptoms and a continuous murmur on examination. There was no significant past medical history. Inflammatory markers were markedly elevated with a C-reactive protein of 190 mg/L and a WCC of $22.5 \times 10^9$ per litre. Transthoracic echocardiography was non-diagnostic. A two-dimensional transoesophageal echocardiogram demonstrated a communication between the aortic root and pulmonary artery, with a probable vegetation on the pulmonary valve and mild pulmonary regurgitation (Panel A; AoV, aortic valve; TV, tricuspid valve; PV, pulmonary valve). A three-dimensional full-volume acquisition (Panel B) demonstrated a well-defined fistula between the left coronary sinus and main pulmonary artery, 2–3 cm above the pulmonary valve. The fistula appeared separate to the left coronary ostium. A large vegetation was clearly evident on the pulmonary valve.

Clinically, the patient was completely asymptomatic after empirical antibiotic therapy. The organism was later identified as one of the HACEK group, Aggregatibacter actinomycetemcomitans.

CT and invasive coronary angiography confirmed that the fistula was separate to the left coronary artery ostium and ruled out communication with the coronary circulation (Panels C and D; LCA, left coronary artery; RCA, right coronary artery; PA, pulmonary artery).

It is very rare for aorto-pulmonary fistulae to occur in the absence of Sinus of Valsava aneurysms. It was agreed at a Joint Cardiology/Cardiothoracic meeting that this was a rare congenital abnormality and the presence of pulmonary hypertension at right heart catheterization necessitated intervention. She has been referred for surgical ligation.

Supplementary material
Supplementary material is available at European Heart Journal online.