Coronary arteriovenous fistula with coexisting atrial septal defect

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DESCRIPTION
A 36-year-old Hispanic woman was referred for an asymptomatic cardiac murmur and auscultation revealed wide and fixed second heart sound. Transthoracic echocardiogram showed severely dilated right atrium and ventricle. Transesophageal echocardiogram (TEE) confirmed left-to-right shunt through secundum atrial septal defect (ASD) (1.5 cm×2.0 cm×2.5 cm), with Qp:Qs 2:1 on Doppler studies (figure 1). In view of large ASD size and inadequate superior rim of tissue, the patient was planned for surgical repair. Routine coronary angiogram showed a significant coronary arteriovenous fistula (CAVF) from the sinoatrial branch of the right coronary artery to the common pulmonary artery (PA; figure 2). The patient underwent autologous pericardial patch repair of ASD with CAVF ligation, with no post-operative complications.

CAVF is a congenital coronary anomaly noted in 0.002% of the general population and 0.08–0.30% of angiographic series, hypothesised to be due to persistent sinusoidal connections between the coronary arteries and primitive heart tubules.1–3 In 20–45% cases, other associated congenital cardiac anomalies are evident.4 Drainage is primarily into the right heart system (90%) with 15–17% PA drainage.2–4 Complications include angina (coronary steal), heart failure, myocardial infarction and death.1–4 Concurrent ASD is reported to have a significantly higher Qp:Qs ratio.5 Despite coronary angiogram being the gold standard of diagnosis, alternate modalities such as TEE, CT angiogram and MRI are increasingly being advocated.1–4

Figure 1 Three-dimensional view on transesophageal echocardiogram demonstrating atrial septal defect.
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REFERENCES