Bilateral Coronary Artery Fistula

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Abstract
Coronary artery anomalies include anomalies of origin, termination, structure or course. Coronary artery fistulae (CAF) are classified as abnormalities of termination and are considered a major congenital anomaly. A coronary artery fistula involves a sizable communication between a coronary artery, bypassing the myocardial capillary bed and entering either a chamber of the heart (coronary-cameral fistula) or a great vessel. Bilateral coronary artery fistula, are a rare variant. We describe a bilateral CAF with angina and significant coronary artery disease requiring percutaneous coronary intervention.

Introduction
Coronary artery fistula is a rare congenital or acquired anomalous shunt from a coronary artery to a cardiac chamber or great vessel. It is seen in 0.6-1.5% of patients who undergo coronary angiography and in 0.002% of the general population.¹ It is the most common congenital coronary anomaly leading to significant hemodynamic abnormalities. The persistence of intratrabecular gaps that are normally present in the intrauterine life results in coronary artery fistulas. Low-pressure structures are the most common sites of CAF drainage.² Though most CAFs are small and asymptomatic, they may cause many problems including continuous murmur, angina pectoris, acute myocardial infarction, sudden death, coronary steal, congestive heart failure, infective endocarditis, arrhythmia, coronary aneurysm formation and superior vena cava syndrome.

Case Report
A 49 year old male patient presented to our hospital with complaints of chest pain on and off for past 5 years which had worsened 7 hours prior to presentation. He was a smoker and had diabetes mellitus.

His physical examination was unremarkable with normal blood pressure. His ECG showed Q-waves in the inferior leads. 2D-echo showed normal size cardiac chambers with mild left ventricular hypertrophy and a left ventricular ejection fraction of 60%.

He was taken up for coronary angiogram subsequently and it revealed a critical stenotic lesion in mid RCA and D1 (Figures 1, 3). There was an AV fistula from the proximal LAD to the pulmonary artery and from conal branch of RCA to the pulmonary artery (Figures 1, 2, 3).

He was subsequently taken up for PTCA and an Everolimus-eluting stent 3.5 x 28 mm (Xience V, Abbott, USA) was deployed in mid RCA with no complications (Figures 4, 5). For the D1 lesion he was managed medically. As patient became asymptomatic after PCI of RCA lesion no intervention was performed for CAF. He has been on regular follow up and has remained asymptomatic post-procedure.

Discussion
Coronary artery fistula refers to a direct connection between a coronary artery and one of the cardiac chambers, large vessels, or other vascular structures. Clinical manifestations vary depending on the site and size of the shunt, pressure gradient between the

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Received: 02.06.2015; Accepted: 16.06.2015
origin and drainage site, and underlying valvular or coronary pathologies. Most patients are asymptomatic. However, myocardial ischemia, congestive heart failure, arrhythmia, sudden death, infective endocarditis and rupture may develop. The drainage sites can be listed in descending order of frequency as the right ventricle (41%), right atrium (26%), pulmonary artery (17%), left ventricle (3%), and superior vena cava (1%). Bilateral fistulas originating from both the coronary systems account for 5% of the total. These types of fistulas terminate more often into the pulmonary artery (56%) than the unilateral fistulas (17%). In a review in 2011, it was reported that the origin of the fistulas was from the LCA in 69% and from the RCA in 31% respectively. The majority, as expected, were unilateral fistulas (80%), followed by bilateral fistulas (18%) and finally multilateral fistulas (2%). Among the unilateral fistulas, the LAD was predominantly the origin (42%) of the fistulas, followed by the RCA (31%), the Cx (20%) and finally the left main trunk (LMT) (7%). The management of small and asymptomatic fistulae is controversial. Coronary fistulae require treatment in the presence of large fistulae, progressive left-to-right shunt, myocardial ischemia, left ventricular dysfunction, congestive heart failure and in order to prevent endocarditis/endarteritis. There are two interventional treatment modalities for CAF: surgical ligation and percutaneous transcatheter approach, both having similar success rates. As the patient had remained asymptomatic following PCI to RCA, no intervention was performed on the fistula and the patient has since been remaining asymptomatic on regular follow up.

References