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EDITORIAL COMMENT

The Multidisciplinary Heart Team Approach to Management of Coronary Artery Fistula With the Assistance of 3D Image Reconstruction*

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In this issue of *JACC: Case Reports*, Brantner et al. (1) from University Hospital of Basel in Switzerland described a wonderful case of a large, complex coronary venous fistula with additional feeders from the descending aorta. Three-dimensional (3D) printing allowed precise anatomic visualization of the origin, course and drainage site of the fistula. This case wonderfully illustrates the value of imaging in complex coronary artery fistulas.

Coronary artery fistula, also known as a coronary artery venous malformation, is an abnormal communication between a coronary artery and another cardiovascular structure, which may include a cardiac chamber (otherwise referred to as a cameral fistula), coronary sinus, superior vena cava, or pulmonary artery occurring in absence of the myocardial capillary bed. The incidence of coronary artery fistula is 0.1% to 0.2% (2) in all patients undergoing coronary angiography, and most commonly occurs without other associated congenital anomalies. Fistulous communications may be congenital or acquired and can be identified at any age or stage of life. The unique location and physiology of coronary arteriovenous fistula lends to the use of burgeoning 3D imaging technologies in conjunction with a highly integrated heart team for management.

The origin of the fistula may drain from a main coronary artery and is usually a dilated and tortuous artery terminating in one of the cardiac chambers or a vessel. The more proximal the origin from the main coronary artery, the more dilated it is. If the fistula drains to the right atrium with a proximally arising feeding artery, it tends to be considerably dilated but less tortuous. If there is a more distal origin, as when the fistulas originate from the left coronary artery and drain to the left ventricle, they may be very tortuous, presenting a challenge for catheter closure. However, in the less frequently encountered right coronary artery to coronary sinus, the fistula vessel may be large and very tortuous. There may be multiple feeding arteries to a single coronary arterial fistula drainage point or there may be multiple drainage sites.

Fistulas originate from the right coronary artery in 52% of cases, the left anterior descending is the next most frequently involved in approximately 30% of cases, and the LCX in about 18% of cases. Over 90% of the fistulas regardless of origin egress to the right side of the heart, and the remainder drain to the left side of the heart. In the right heart, drainage occurs most frequently to the right ventricle (in about 40%), followed by the right atrium, coronary sinus, and pulmonary trunk. In adults, occasionally fistulas may be encountered that originate from both coronary arteries and drain into the pulmonary trunk.

When the fistula drains to the right side of the heart, there is right-sided volume loading as well as to the pulmonary vascular bed, the left atrium, and the left ventricle. However, when the fistula drains into the left heart, although there is left-sided volume

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loading, there is no increase in the pulmonary blood flow, thus resulting in different echocardiographic appearances. The size of the shunt is determined by the size of the fistula and by the pressure difference between the coronary artery and the chamber into which the fistula drains. Occasionally, congestive heart failure occurs, and rarely, in adults, myocardial ischemia may be seen.

Coronary arterial fistulas are usually asymptomatic in the first 2 decades of life, particularly when they are small, and may even close spontaneously. Complications include "steal" from the adjacent myocardium causing myocardial ischemia, thrombosis and embolism, heart failure, atrial fibrillation, rupture, endocarditis/endarteritis, and arrhythmias. Thrombosis within the fistula is rare, but may cause acute myocardial infarction and atrial and ventricular arrhythmias. The largest shunts occur when a proximal coronary artery connects to the right side of the heart rather than the left heart chambers.

The majority of the patients are asymptomatic. In the older patients, symptoms may include dyspnea on exertion or arrhythmias. Patients with large left-to-right shunts may develop congestive heart failure. If angina is documented, it may be due to coronary artery steal; pulmonary hypertension secondary to long-term high-volume R>L shunts can also be seen.

Specific management strategies, which can include surgical repair or catheter embolization, are somewhat controversial. In the largest recorded series of 46 patients treated with surgery, pre-operative symptoms included angina and HF (3). Importantly, post-operative myocardial infarction occurred in 11% because of low flow in the dilated coronary artery proximal to fistula closure. An interventional approach is best evaluated by a multidisciplinary team including cardiothoracic surgery, interventional cardiology, structural cardiology, and advanced imaging.

Most patients are referred because of a continuous murmur, loudest over the precordium, which may be thought to be due to patent arterial ductus. However, the murmur is heard over the mid-chest, or even lower, rather than below the left clavicle and typically peaks in mid to late diastole rather than systole. If the fistula connects to the left ventricle, only an early diastolic murmur may be heard. Some patients with large shunts may present with signs of congestive cardiac failure and angina, usually at the extremes of life.

Two-dimensional and color Doppler echocardiography are helpful in demonstrating dilation of the affected coronary artery and on color flow mapping may show the site of drainage, but it is difficult to define the detailed anatomy of the fistula with echo. Cardiac magnetic resonance imaging may be helpful in confirming the diagnosis, as the proximal coronary arteries or even the whole length of the fistula vessel may be seen. Fractional flow reserve cardiac computed tomography angiography and 3D volumetric rendering of cardiac computed tomography angiography are valuable in procedural planning.

Procedural options can be optimized by careful identification of the number of fistulous connections, nature of the feeding vessel or vessels, sites of drainage, and quantification of myocardium at risk. The goal of treatment is the occlusion of the fistula, while maintaining normal myocardial perfusion. The indications for treatment of CAVF include the presence of a large or increasing left-to-right shunt, left ventricular volume overload, myocardial ischemia, left ventricular dysfunction, or congestive cardiac failure, and for prevention of endocarditis/endarteritis.

The treatment options for CAVF include surgery or catheter closure. Surgery involves internal closure of the fistula within the receiving chamber or vessel whenever feasible, but when the fistula is associated with a large aneurysm of the feeding artery, it may need to be ligated from within the aneurysm. Surgery is associated with a low morbidity and mortality rate; infarction may occur in <5% of cases and there is a risk of recurrence. The reason for the recurrence includes the fact that there may be multiple fistulas present that are difficult to visualize intraoperatively, advocating for a hybrid approach.

Catheter closure of the fistulas is an effective and safe alternative to surgery. The aim of catheter closure is to occlude the fistula artery as distally and as close to its termination point as possible, so as to avoid any possibility of occluding branches to the normal myocardium. Whichever technique of catheter closure is used, the occlusion should be at a precise point. In some patients, it may be easier to enter the fistula from the right side of the heart. These may be suitable for occlusion with Amplatzer occluder devices (Abbott, Abbott Park, Illinois), such as vascular plug, duct occluder, or atrial or ventricular septal occluder.

After occluding the main fistulous vessel, repeat selective coronary angiography in both coronary arteries is essential, as a second branch feeding the fistula or multiple feeding vessels may be visualized. With catheter-based closure techniques, complete occlusion of the fistula may be achieved in >95% of the patients. In the remaining patients, either further procedures may be required to close the fistulas or

they may be managed conservatively if the residual fistulas are small. All complications are rare and include premature deflation of a detachable balloon, inadvertent coil embolization, transient T-wave changes, transient bundle branch block, and myocardial infarction.

3D imaging and printing now allow for manipulation of the aneurysm both in a virtual environment as well as an in vivo simulation to determine the most efficient method to address the fistulae. This potentially reduces fluoroscopy time as well as radiation dose and decreases the likelihood of device failure or migration should device placement be considered. The management of the fistula clinically is more important at the egress than the origin. There are a variety of approaches that may be appropriate and should all be considered in addressing fistula closure. Strategies including coil embolization, vascular plugs, and septal occluder devices can all be electronically modeled and implanted in a virtual environment as well as trial therapy in a 3D printed environment. The collaborative environment for management of late-presenting symptomatic coronary fistulae includes the multidisciplinary heart team approach. The presence of coronary artery fistula requires review by a knowledgeable heart team that may include congenital and noncongenital cardiologists and surgeons to determine the role of percutaneous and/or surgical closure.

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