Operative approach for right coronary artery to coronary sinus fistula

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ABSTRACT

Coronary artery fistula is a rare congenital heart disease that is defined as an abnormal connection between a coronary artery and a cardiac vessel or cardiac chamber. Most coronary artery fistulas involve the right coronary artery draining into the right-sided heart structures. We present a patient with right coronary artery to coronary sinus fistula diagnosed by coronary angiography. The surgical approach of retrograde cardioplegia and transatrial resection allowed for protection of the myocardium and definitive visualization and closure of the abnormal fistula.

KEYWORDS Aneurysmal dilation; cardiac surgery; coronary artery fistula

Coronary artery fistula (CAF) is a rare congenital heart disease that is defined as an anomalous connection between a coronary artery and any other cardiac vessel or cardiac chamber. Most CAFs involve the right coronary artery (RCA) draining into the right heart structures. While presentation may initially be asymptomatic, untreated CAFs may result in long-term sequelae such as proximal aneurysmal dilation of involved vessels and symptoms of myocardial ischemia or congestive heart failure.¹ The definitive treatment of CAF requires either a catheter-based or surgical intervention, depending on patient-specific anatomy and other factors.

CASE REPORT

A 33-year-old woman with Von Willebrand disease presented with frequent chest pain exacerbated by activity and dyspnea for 1 year, and CAF was diagnosed by angiography. Her symptoms worsened after the diagnosis. The electrogram showed no abnormalities, and cardiac troponin levels remained normal. She had no abnormal findings on physical exam. Coronary angiography showed an RCA with a transverse diameter of 7 to 8 mm and an RCA CAF that drained into the right atrium (*Figure 1*). Preoperative color Doppler showed turbulent blood flow from shunt to right atrium (*Figure 2a*). She was given prophylactic desmopressin and Alphanate to prevent bleeding complications from Von

Willebrand disease. The surgical procedure was performed through a median sternotomy, and aneurysmal RCA was revealed after pericardial dissection. Aortic and bicaval

Figure 1. Coronary angiogram showing an aneurysmally dilated right coronary artery (RCA) with fistula creating anomalous blood flow to the right atrium (RA).

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Figure 2. Color Doppler echocardiography. **(a)** Preoperative image showing turbulent blood flow (*) in the right cardiac chambers due to right coronary artery to coronary sinus fistula. **(b)** Postoperative image confirming resolution of anomalous blood flow after surgical repair of the fistula.

Figure 3. Opening of the coronary sinus (*) in the exposed right atrium.

cannulation was performed and cardiopulmonary bypass was initiated, followed by antegrade cardioplegia. The right atrial wall was incised, and there was stenosis of the ostium of the coronary sinus. There was a fistula from the RCA to the coronary sinus with an opening diameter of 4 mm, about 1 cm distal to the coronary sinus ostia, and a diminutive posterior descending coronary artery (*Figure 3*). The cribriform flap at the ostium was removed and the ostium of the fistula was closed by primary surgical repair. Transesophageal echocardiogram with color Doppler imaging after closure showed no

shunt (*Figure 2b*). Her postoperative course was uncomplicated.

DISCUSSION

In our patient, coronary angiography revealed aneurysmal dilation of the RCA and a diminutive posterior descending coronary artery, and transesophageal echocardiogram revealed a shunt between the RCA and coronary sinus. The operation eliminated the shunt. We used the transcatheter approach as indicated for a single, large, symptomatic fistula presenting with aneurysmal formation.² Less-invasive transcatheter techniques using occlusion coils, detachable balloons, and covered stents have comparable outcomes in select patients. Transcatheter closure has optimal results in patients with small CAFs with nontortuous, nondilated vessels and a single drainage site.³ While the benefits of a less-invasive procedure were considered, the literature suggests that our patient with coronary artery aneurysm, symptomatic coronary steal, and a fistula to the right atrium required open intracardiac ligation. Definitive surgical treatment of the RCA to coronary sinus fistula resolved symptoms and hopefully prevented future complications, including further dilation or rupture of the RCA, heart failure, infective endocarditis, and thrombosis.¹

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