Anomalous right coronary artery from the pulmonary artery: An institutional report

Joseph A. Policarpio, a Marvin J. Derrick, MD, b and Sanjay S. Mehta, MD, c Champaign, Ill

The anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is rare, with a reported incidence of 0.002%, accounting for 0.12% of all congenital heart defects. 1-3 This institutional report of 2 cases at our institution is the first to our knowledge to address medical versus surgical management of ARCAPA (Video 1 provides a brief introduction and significance of this report).

CLINICAL SUMMARY

The first patient is a 59-year-old man who presented with chest pain, mild elevation of troponin, altered mental status, and hypoxia due to opioid abuse. Comorbidities included asthma, epilepsy, and a body mass index of 31. Coronary angiogram revealed a large tortuous left anterior descending artery and left circumflex artery that collateralized to a large right coronary artery (RCA), which drained into the pulmonary artery (PA). Computed tomography angiography confirmed ARCAPA. Catheter oximetry showed no significant step-up in oxygen saturation from the right ventricle to the PA. Cardiotothoracic surgical consultation was done and after discussing the risks and benefits, including the risk of sudden cardiac death, the patient elected to defer surgery. The patient's condition was managed medically for 4 years, with recurrent presentations to the emergency department leading to several hospitalizations for complaints of shortness of breath, chest pain, acute kidney injury, dehydration, and opiate abuse. This patient has not yet experienced a major cardiac event.

The second patient is a 51-year-old woman who presented with dyspnea, lower-extremity edema, and increasing abdominal girth. Comorbidities included hypertension, diabetes mellitus, and a body mass index of 41. A diagnosis of heart failure with reduced ejection fraction was made (ejection fraction 25%-30%) and a right- and left-heart catheterization revealed elevated right- and left-sided filling pressures, pulmonary hypertension, and reduced cardiac output. The left anterior descending artery and left circumflex were severely dilated with retrograde flow into the RCA, which drained into the PA with no step-up in oxygenation (Figure 1). Intraoperatively, the anomalous RCA was visible from the PA anteriorly (Figure 3). After ligation, an immediate drop of PA pressure was observed from 55/33 mm Hg to 37/24. The patient recovered well and was discharged on postoperative day 4. Follow-up was uncomplicated but challenging due to patient
noncompliance over an 8-month period. An echocardiogram at 5.5 months postoperatively showed ejection fraction of 45% to 50% and calculated right ventricular systolic pressure of 34 mm Hg and pulmonary artery systolic pressure of 33 mm Hg. The patient reported no cardiovascular symptoms since the procedure.

This report is exempt from institutional review board approval as it avoids identifiable patient information. Patient consent for the publication of the study was received verbally and documented in the patient chart.

DISCUSSION

While the surgical correction of ARCAPA is described in the literature, this report is the first to our knowledge to address medical versus surgical management in two patients of similar age and comorbidity, presenting late in the disease course. The first patient decided on medical management and has presented several times to the hospital with chest pain but has not yet experienced a major cardiac event. Although this patient declined surgery, which is the recommended treatment, an alternative approach might include closure of the anomalous RCA with a vascular plug or coil. However, we favored medical management over endovascular occlusion for a few reasons. We believe that leaving the anomalous RCA open would allow for a

FIGURE 1. Preoperative coronary angiogram of patient 2. The catheter is in the ascending aorta with the tip positioned in the proximal LCA. Notice the subtle radio-opacity in the PA due to retrograde flow from the LAD and LCx into the RCA. PA, Pulmonary artery; LCA, left coronary artery; LAD, left anterior descending artery; LCx, left circumflex artery; RCA, right coronary artery.

FIGURE 2. Preoperative computed tomography angiogram of patient 2. The RCA arises from the PA. RCA, Right coronary artery; PA, pulmonary artery; AA, ascending aorta.

FIGURE 3. Intraoperative anterior view of patient 2 before ligation and anastomosis. AA, Ascending aorta; PA, pulmonary artery; RA, right atrium; RCA, right coronary artery; RV, right ventricle.
shunt reversal if a plaque rupture and occlusion of the left system were to occur in the future. The anomalous RCA would then perfuse the myocardium, albeit with deoxygenated flow. This might allow the patient to receive medical care and thus help prevent sudden cardiac mortality. While minimally invasive, this procedure would still expose the patient to risks such as bleeding, infection, or embolization of the percutaneous device. Although percutaneous treatment has been well-established for coronary artery aneurysm and fistula, there are no data to support percutaneous closure of ARCAPA at this time.

The second patient who underwent surgical repair of ARCAPA with saphenous vein graft and ligation of the proximal anomalous artery performed without cardiopulmonary bypass recovered well and remained asymptomatic.

CONCLUSIONS
Surgical treatment remains the standard of care for management of ARCAPA, and those reluctant to undergo surgery should be followed closely with medical management and regular follow-up.

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