

Anomalous Origin of the Right Coronary Artery from the Left Main Coronary Artery in the Setting of Critical Bicuspid Aortic Valve Stenosis

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ABSTRACT: Anomalous origin of the right coronary artery (ARCA) is an extremely rare cardiac pathology. The coronary artery can potentially course between the aorta and the pulmonary artery, putting the patient at risk for sudden cardiac death. Even rarer is an ARCA that arises from the left main coronary artery (LMCA). To our knowledge, this is the first case where an ARCA arising entirely from the LMCA is associated with critical bicuspid aortic valve stenosis.

BACKGROUND

Anomalous origin of a coronary artery (ACA) is a rare entity with an incidence of 1% in those undergoing angiographic evaluation.¹ The most common benign variant is the anomalous origin of the left circumflex artery from a separate ostium or sinus.² Anomalous origin of a right coronary artery (ARCA) from the left main is an extremely rare phenomenon that has only been documented in case reports.^{3,4} In these variants, the most common origin was the left coronary cusp. Although typically benign, ACA can lead to ischemia, chest pain, and sudden cardiac death (SCD) when it courses between the aorta and pulmonary artery.⁵ Multiple different anatomical subtypes of ACA contribute to the underlying pathophysiology that leads to SCD and arrhythmias.⁶

Even rarer than ACA is a coexisting bicuspid aortic valve (BAV); there are only a few case reports documenting this combination.^{7,8} Patients with BAV are at risk for premature aortic stenosis and may present with chest pain, heart failure, syncope, or SCD. There have been few documented cases of ARCA arising entirely from the left main coronary artery (LMCA).^{3,4,9} Here, we present a case involving a young male who was diagnosed with critical bicuspid aortic stenosis and incidentally discovered to have a nondominant ARCA arising entirely from the LMCA.

CASE REPORT

A 47-year-old African American male with no pertinent medical history presented to our emergency room with angina. Vital signs upon arrival were as follows: blood pressure 112/78 mm Hg, heart rate 111 BPM, respiratory rate 22 breaths/min, and oxygen saturation 98% on noninvasive positive pressure ventilation. Electrocardiogram (ECG) was notable for left ventricular

hypertrophy with repolarization changes. First troponin I was 0.01 ng/mL, with subsequent troponin levels peaking at 0.07 ng/mL. There were no dynamic ST segment changes on serial ECGs. Additional imaging revealed pulmonary edema on chest x-ray and an elevated brain natriuretic peptide of 724 pg/mL. A 2-dimensional (2D) transthoracic echocardiography (TTE) revealed critical aortic stenosis with a valve area of 0.4 cm² and a mean gradient of 66 mm Hg; however, it was unclear if the patient had a bicuspid valve. He underwent a left heart catheterization that revealed a nondominant ARCA arising entirely from the LMCA (Figure 1). The rest of the epicardial anatomy was normal except for nonobstructive atherosclerotic coronary artery disease noted in the left circumflex. He was immediately referred to cardiac surgery for aortic valve replacement. His ARCA was examined intraoperatively, and since its anatomic course did not pose a risk for compression between the aorta and pulmonary artery, his coronary anatomy was left alone. The postoperative pathology revealed a BAV.

DISCUSSION

Bicuspid aortic stenosis with an ARCA is an extremely rare congenital cardiac pathology that has only been reported in case reports.^{7,8,10} This case is unique in that the ARCA arose entirely from the LMCA in the setting of critical aortic stenosis. The typical presentation of patients with ACA varies between chest pain, dizziness, and SCD. Although mostly benign, an ACA can lead to SCD when it courses between the aorta and pulmonary artery.⁵ Therefore, it is important to analyze the course of the coronary artery either during surgery or preoperatively with cardiac computed tomography.¹¹ The associated defect should be carefully elucidated because any surgical operation for valve management may need to be modified to include additional surgical techniques, such as coronary reimplantation, unroofing, or coronary artery bypass grafting with ligation of the ACA.⁶ It

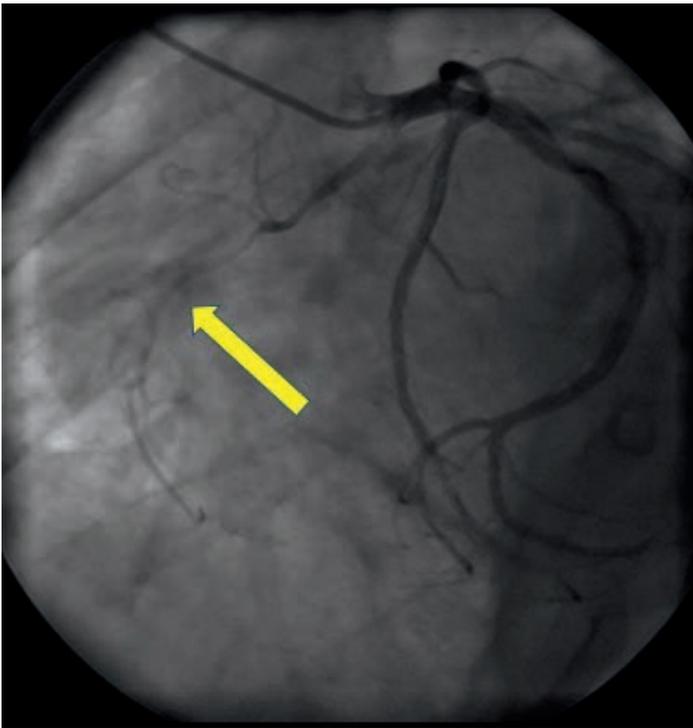


Figure 1 (Online Video 1)

Left anterior oblique view of a 6F Judkins left 4.0 catheter engaged into the left coronary cusp. An anomalous right coronary artery (yellow arrow) can be seen arising from the left main coronary artery.

is also important to identify an anomalous course of an artery to avoid inadvertent injury to the vessel during surgery.

A retrospective study from Naito et al. examining the morphology and vascular defects associated with BAV showed that there was a significantly higher incidence of coronary anomalies in bicuspid valves compared to tricuspid aortic valves, with incidence rates of 7% and 3%, respectively. This same study also showed that the more common ACA involved the ARCA. Looking at the secondary end point, patients with ACA and BAV who had received aortic valve replacement tended to have higher postoperative cardiac comorbidity, which required repeat surgery and/or coronary angiography.¹² Although the reason behind this was unclear, it may be due to coronary artery hypoplasia or to the intramural location of the anomalous artery, both of which may increase surgical risk during aortic valve replacement.¹³

Initial workup for BAV stenosis starts with a 2D TTE and Doppler waveforms.¹⁴ All patients require a left heart catheterization to evaluate the epicardial arteries for anomalies and atherosclerotic coronary artery disease. Although medical therapy can reduce symptoms of aortic stenosis, these patients require definitive

surgery to replace the aortic valve and, in some cases, repair or bypass the affected coronary artery.¹⁴ Age, anticoagulation preference, comorbidities, and underlying coronary artery anatomy all play a role in careful perioperative planning. The decision to implant a bioprosthetic versus mechanical aortic valve depends on shared decision making between the patient and physician(s) after weighing the risks and benefits of both options.¹⁵ Patients with underlying coronary artery defects that warrant cardiac surgery will require cautious outpatient monitoring with an experienced heart team. Further studies are needed to assess the long-term outcomes of these individuals.

CONCLUSION

Our patient was urgently referred to cardiac surgery. After shared decision making with careful assessment of risks and benefits, our patient opted for a tissue valve because he did not want long-term anticoagulation. He underwent successful replacement of his native valve. The course of his ARCA was examined intraoperatively and noted to be benign, so it was left alone. He did well postoperatively and was discharged with close follow-up.

Conflict of Interest Disclosure:

The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords:

bicuspid aortic valve, severe aortic valve stenosis, anomalous coronary artery, aortic valve replacement

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