

Sudden Cardiac Death Revealed by an Anomalous Origin of the Right Coronary Artery From the Left Sinus of Valsalva



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Aberrant origin of the coronary artery from the opposite sinus of Valsalva is a rare congenital coronary anomaly associated with increased risk of myocardial ischemia and sudden death in young patients. We report a case of resuscitated sudden cardiac death in a patient with an anomalous origin of the right coronary artery, arising from the left sinus of Valsalva and coursing between the ascending aorta and the pulmonary artery. Successfully coronary arterial bypass grafting using the left radial artery was performed. Despite the risk of fatal issue, surgical management of patient with this coronary anomaly still remains controversial.

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Congenital anomalies of the coronary artery occur in 0.3% to 1.3% of the population.¹ Most anomalies never cause symptoms of cardiac dysfunction, and they are often revealed incidentally during cardiac catheterization or computed tomography evaluation for other cardiac diseases. However, some of these coronary anomalies are associated with disruption of myocardial perfusion, and they can lead to fatal complications. Anomalous origin of the right coronary artery (ARCA) from the left sinus of Valsalva is one of the highest-risk lesions, but it is relatively rare, with an incidence ranging from 0.019% to 0.17%² and an estimated angiographic prevalence of 0.92%.³ Although ARCA is potentially life threatening, surgical correction remains controversial except in symptomatic patient. We report a resuscitated sudden death patient with ARCA who underwent coronary artery bypass grafting (CABG) using a radial artery (RA), and we discuss the advantages and the limitations of this treatment.

A 37-year-old man was admitted to the hospital after he was resuscitated from a cardiorespiratory arrest that occurred while he was vomiting. Except for tobacco smoking and alcohol consumption, the patient did not have any significant medical history. An emergency

coronary angiography showed ARCA arising from the left sinus of Valsalva (Figure 1A) without evidence of atherosclerosis disease.

This anomaly was confirmed with coronary computed tomography (CT; Figures 1B-1D) and cardiac magnetic resonance imaging, which revealed the initial compression of the right coronary artery (RCA) between the aorta and the pulmonary artery with an acute angulation between the ostium and the RCA. After a discussion with the heart team, we decided to perform a surgical correction through a median sternotomy approach with cardiopulmonary bypass and cardiac arrest. Inspection of the right coronary origin confirmed that the RCA originated from the left coronary sinus with an intramural course between the ascending aorta and the pulmonary artery (Figure 2). Surgical examination of the RCA showed in its first segment several initial branches (infundibular branch or the sinus node artery) and a fibrously thickened wall. We decided to perform a CABG using the left RA anastomosing to the RCA just distal to the anomalous segment when it emerged from behind the pulmonary artery. Ultrasonic Doppler imaging showed a good flow pattern. The origin of the right coronary was clipped with the goal of avoiding competitive flow.

After an uneventful recovery with no complication, the patient was discharged from the hospital on post-operative day 7. At 6-year follow-up, coronary CT confirmed the patency of the left RA (Figure 3). The patient was well and asymptomatic with no ischemic events.

Comment

The majority of coronary anomalies never cause symptoms but some occasionally result to arrhythmias, angina, syncope myocardial infarction or sudden death with an incidence of 33%.^{4,5} There is currently no clinical factor that can predict sudden death, although young patients appear to be at lower risk.⁶ The mechanism of sudden death remains unclear. Several pathophysiologic theories have been suggested to explain the compromised coronary blood flow responsible of myocardial ischemia. In our patient, the ARCA originated from the left sinus of Valsalva and coursed between the aortic root and the pulmonary artery; therefore, the hypothetical mechanism was related to be the compression between these vessels.

The treatment of patients with ARCA is controversial, but surgery is generally recommended in cases of symptoms or evidence of myocardial ischemia being related to the coronary anomaly.⁶ Several surgical options have been described,⁷ including CABG, unroofing procedure, reimplantation of the coronary ostia to the correct sinus, main pulmonary artery translocation, and neo-ostium formation. Within this variety of reported techniques, CABG and unroofing are the most commonly used procedures. The selection of the best surgical strategy is still a source of debate, and it might depend on the

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Figure 1. (A) On initial coronary angiography (left anterior oblique projection, 20 degrees) the right coronary artery originates aberrantly from the left coronary sinus. (B, C) Coronary computed tomography demonstrated an intramural course of the ARCA (white and black arrows) with a compression between the ascending aorta and the pulmonary artery. (D) Volume-rendered image display of the RCA (black arrow) originating from the left coronary sinus with an interarterial course. (Ao, ascending aorta; ARCA, anomalous right coronary artery; LM, left main; MPA, main pulmonary artery; RCA, right coronary artery.)

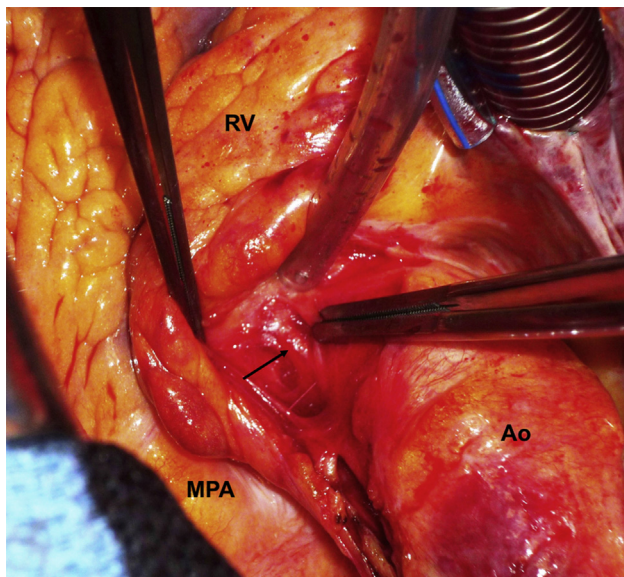
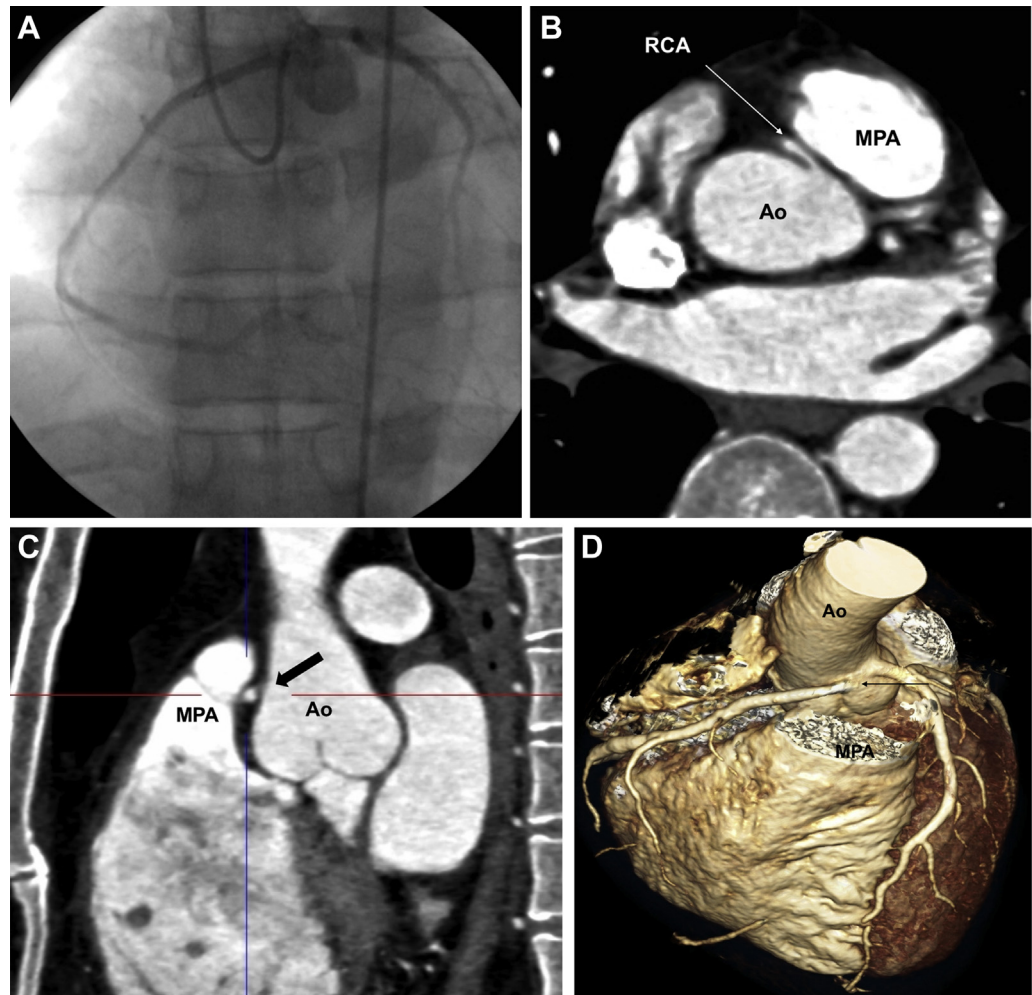


Figure 2. Operative findings. Black arrow indicates the anomalous right coronary artery from the left coronary sinus. (Ao, ascending aorta; MPA, main pulmonary artery; RV, right ventricle.)

patient's age, the specific ARCA anatomic details, the pathophysiologic mechanisms, and the operative findings.

CABG remains the standard procedure for restoring normal distal coronary flow with good long-term patency results. However, the ideal bypass graft selection for the ARCA remains a source of debate. In our patient, we chose to perform CABG using the left RA. Herein, we report using the left RA in CABG for ARCA arising from the left sinus of Valsalva. Our revascularization strategy was based on (1) the presence of a fibrously thickened wall and several initial branches in the first segment of ARCA; (2) the specific ARCA anatomic details, such as acute angulation and location between the great arteries; (3) the advantages of sparing the left internal mammary artery in the event that the patient develops coronary disease later and the lower risk of size mismatch with the use of the RA compared with the use of internal mammary artery; and (4) the excellent long-term outcomes with RA patency rates similar to those for the left internal thoracic artery and much better than for saphenous vein graft, as confirmed recently.⁸ According to the CT obtained 6 years postoperatively, the RA seems to be the

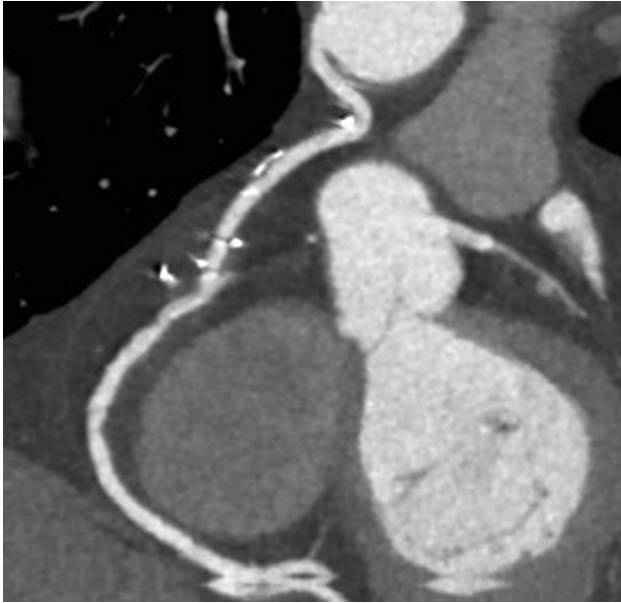


Figure 3. Three-dimensional volume rendering of a computed tomographic image obtained 6 years postoperatively confirmed a patent left radial artery bypass.

appropriate choice in our patient. Undoubtedly, a longer follow-up period should be required to determine the long-term patency rate in case of bypass for ARCA using an RA.

ARCA from the left sinus of Valsalva is a potentially life-threatening condition that can lead to sudden death. Therefore, we recommended surgical correction in symptomatic patients. CABG using the left radial artery

represents a safe, durable, and valuable alternative to the left internal mammary artery or vein graft in appropriate patients. Additional multicentric studies are necessary to design the optimal surgical management in symptomatic and asymptomatic patients with ARCA.

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