Anomalous Right Coronary Artery off the Pulmonary Artery Strikes When You Least Expect It!

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INTRODUCTION

Anomalous right coronary artery off the pulmonary artery (ARCAPA) is an exceedingly rare anomaly that can be found in isolation or in association with other congenital heart diseases.[1] Here we describe ARCAPA in an infant with an inferior sinus venosus defect and partial anomalous pulmonary venous return.

CASE PRESENTATION

The patient was an 8-month-old male child who was initially referred at 1 month of age following diagnosis of a moderate inferior sinus venosus defect in the neonatal intensive care unit. At his 6-month cardiology clinic follow-up, he had persistently poor oral intake, failure to thrive without clear alternative etiology, and mild intermittent tachypnea. He was started on an oral diuretic regimen and admitted to the hospital for nasogastric tube supplemental nutrition. Despite feed fortification and gavage, he continued to exhibit suboptimal growth. Gastroenterologic workup was nonrevealing.

The patient was born at 37 weeks to a G6P1 mother with normal prenatal laboratory findings via induced vaginal delivery for intrauterine growth restriction. The pregnancy was also complicated by gestational hypertension. He had a 3-week stay in the neonatal intensive care unit following birth for hypoglycemia, hypothermia, and poor feeding. He was also diagnosed with profound bilateral sensorineural hearing loss necessitating hearing aids, a right inguinal hernia that was repaired at 1 month of age, hypospadias, right-sided undescended testicle, and 10q23.33q24.1 duplication of unclear significance.

Transthoracic echocardiography demonstrated a moderate inferior sinus venosus defect with anomalous drainage of the right lower pulmonary veins. The right upper pulmonary veins also appeared to drain into the superior vena cava–right atrium junction (Figure 1A). There was normal biventricular function and right ventricular enlargement. ARCAPA was not considered at the time, as coronary origins appeared deceptively normal by two-dimensional imaging (Figure 1B, Video 1). Follow-up images obtained at 8 months of age were technically challenging in visualizing the right coronary artery origin and instead suggested possible right coronary artery–to–pulmonary artery fistula (Figure 2A). The left main coronary artery also appeared prominent, with an additional branch suggestive of dual left anterior descending coronary arteries (Figure 2B, Video 2). Electrocardiography demonstrated normal sinus rhythm with right atrial enlargement, right-axis deviation, and incomplete right bundle branch block with nonspecific T-wave abnormality.

For anatomic delineation of pulmonary venous anatomy and no prior suspicion of anomalous coronary artery origins, the patient underwent cardiac magnetic resonance imaging showing the inferior sinus venosus defect, partial anomalous pulmonary venous return of the right upper pulmonary veins into the superior vena cava–right atrium junction, and the right lower pulmonary veins draining into the right atrium at the inferior sinus venosus defect (Figure 3, Video 3). The right ventricle was noted to be severely enlarged with normal systolic function. Estimated Q_p/Q_s was 3:1. The coronary artery origins could not be evaluated on cardiac magnetic resonance imaging because of patient size and the spatial resolution of cardiac magnetic resonance imaging (~1-mm voxel size).

The patient underwent elective surgical repair of the inferior sinus venosus defect and right-sided anomalous pulmonary venous drainage. Preoperative transesophageal echocardiography showed all the anticipated findings, but closer interrogation of the coronary arteries revealed flow reversal in the right coronary artery with dilation of the left main coronary artery, concerning for ARCAPA (Figure 4, Video 4). On visual surgical inspection, the anomalous right coronary artery was visualized originating from the right pulmonary sinus. The pulmonary arteries were snared before the administration of cardioplegia to prevent run-off into the pulmonary circulation. The right coronary artery button was excised and sutured to the left side of the mid ascending aorta via button transfer onto a prebypass marked location. The excised part of the pulmonary artery was reconstructed with autologous pericardium. The inferior sinus venosus defect was closed by patch, and the anomalous right upper pulmonary veins were patch-baffled to the left atrium using the same patch. The superior vena cava was enlarged laterally using autologous pericardium. Postoperative transesophageal echocardiography demonstrated good antegrade flow across the right coronary artery (Figure 5B), no residual atrial septal defects, no obstruction of the pulmonary veins, and good biventricular function.

DISCUSSION

ARCAPA, though largely asymptomatic in nature, has been implicated in heart failure, myocardial ischemia, and sudden cardiac death.[2] Myocardial ischemia occurs as a result of flow reversal across the coronary artery into the pulmonary artery, as pulmonary vascular resistance and pulmonary artery pressure decreases after birth.[3] Unlike anomalous left coronary artery from pulmonary artery, ARCAPA appears to convey lower risk for heart failure. In our case, the patient was experiencing mild intermittent tachypnea and failure

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Keywords: Congenital heart disease, Anomalous coronary arteries, ARCAPA, Pediatric echocardiography, Cardiac MR

Conflicts of interest: The authors report no conflicts of interest relative to this document.

2468-6441

https://doi.org/10.1016/j.case.2020.11.007

VIDEO HIGHLIGHTS

Video 1: Deceptive right coronary artery origin by two-dimensional echocardiography.
Video 2: Color Doppler echocardiography demonstrating anomalous right coronary artery off the pulmonary artery.
Video 3: Cardiac magnetic resonance imaging showing anomalous pulmonary venous drainage and sinus venosus defect.
Video 4: Preoperative transesophageal echocardiography showing anomalous right coronary artery off the pulmonary artery
Video 5: Postoperative transthoracic echocardiography showing complete repair.

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to thrive, which are not frequently seen even with large atrial-level shunt lesions in near-term infants. There was normal biventricular function preoperatively, likely due to collateralization from the left main coronary artery and higher pulmonary artery pressures as a result of the large atrial shunt.

ARCAPA is an important yet ever unanticipated diagnosis, with potential clinical consequence at any point from infancy to late adulthood. Approximately one third of ARCAPA cases are associated with other congenital heart disease, most commonly aortopulmonary window.[2,4] Other associated defects included atrial and ventricular septal defects, bicuspid aortic valve, coarctation of the aorta, and patent ductus arteriosus. To date, this is the first case describing ARCAPA associated with partial anomalous pulmonary venous return.

Coronary imaging by echocardiography can be challenging. Although two-dimensional imaging suggested normal coronary origins, confirmation with color Doppler imaging is always necessary.[5] In retrospect, Figure 2A likely demonstrates ARCAPA with flow reversal of the right coronary artery into the pulmonary artery, and

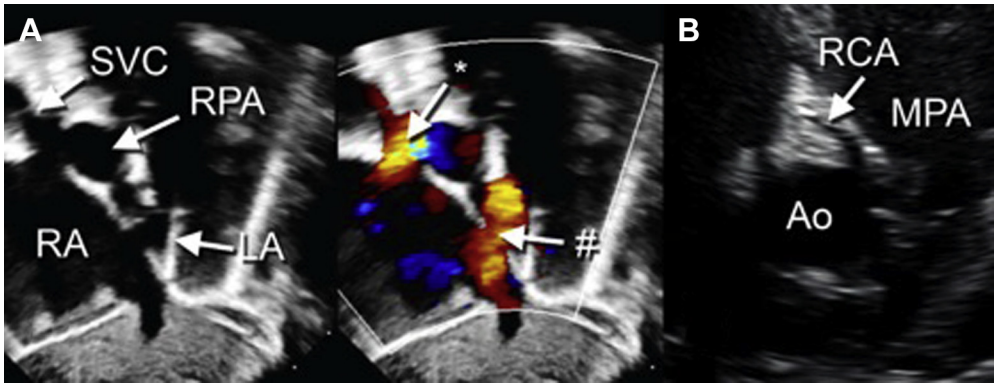

Figure 1 Preoperative transthoracic echocardiography at 8 months of age. **(A)** Subcostal sagittal view demonstrates inferior sinus venosus defect *(pound sign)* and partial anomalous pulmonary venous return of the right upper pulmonary veins to the superior vena cava (SVC)–right atrium (RA) junction *(asterisk)*. **(B)** Deceptive two-dimensional imaging from the parasternal short-axis view suggested normal origin of the right coronary artery (RCA) into the aorta (Ao). *LA*, Left atrium; *MPA*, main pulmonary artery; *RPA*, right pulmonary artery.

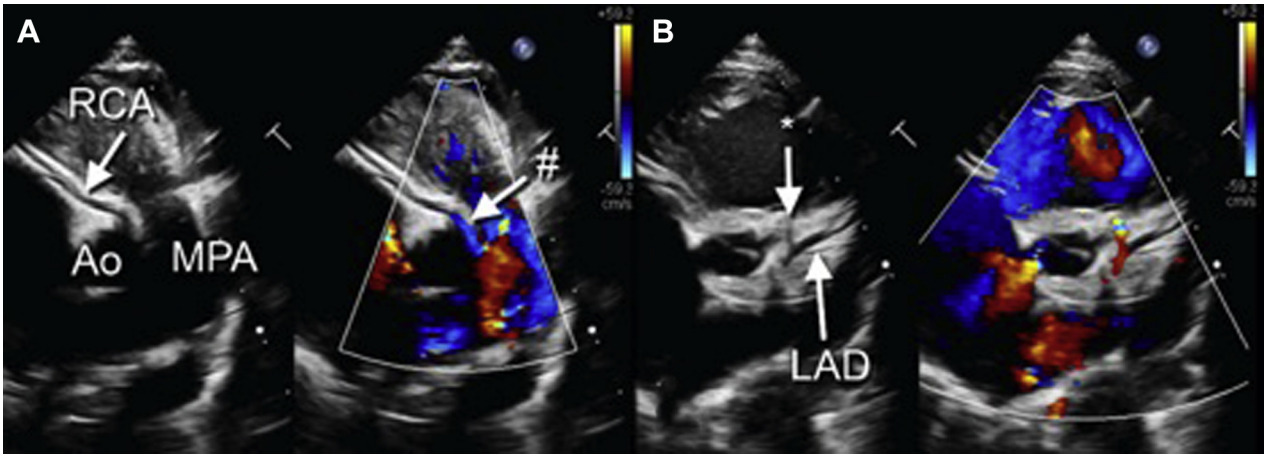

Figure 2 Preoperative transthoracic echocardiography with color Doppler imaging. **(A)** Flow reversal was noted across the right coronary artery; this was initially thought to be a possible right coronary artery (RCA)–to–main pulmonary artery (MPA) fistula. **(B)** Prominent left coronary artery with an additional branch initially suggestive of dual left anterior descending coronary artery (LAD) supply. *Ao*, Aorta.

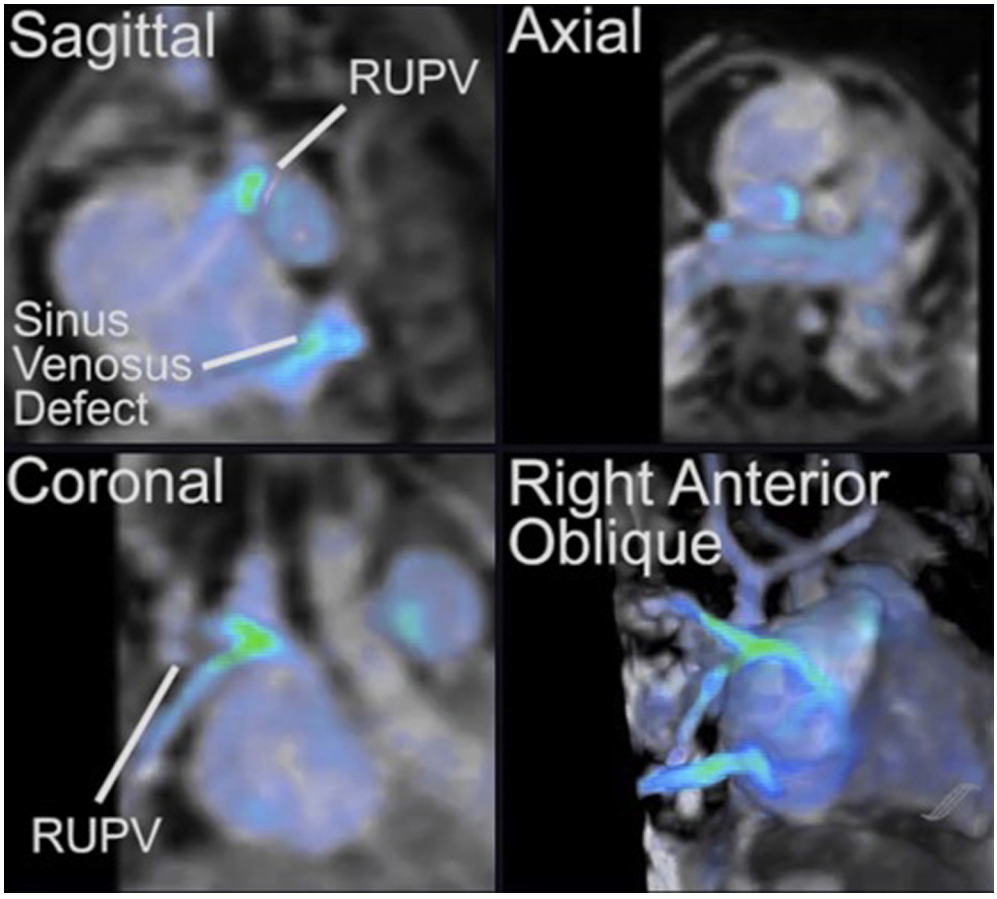

Figure 3 Cardiac magnetic resonance imaging with four-dimensional flow sequence from the sagittal view, coronal view, axial view, and right anterior oblique view as labeled, demonstrating pulmonary venous and atrial anatomy, inferior sinus venosus defect, and partial anomalous pulmonary venous return of the right upper pulmonary veins (RUPVs) to the superior vena cava–right atrium junction.

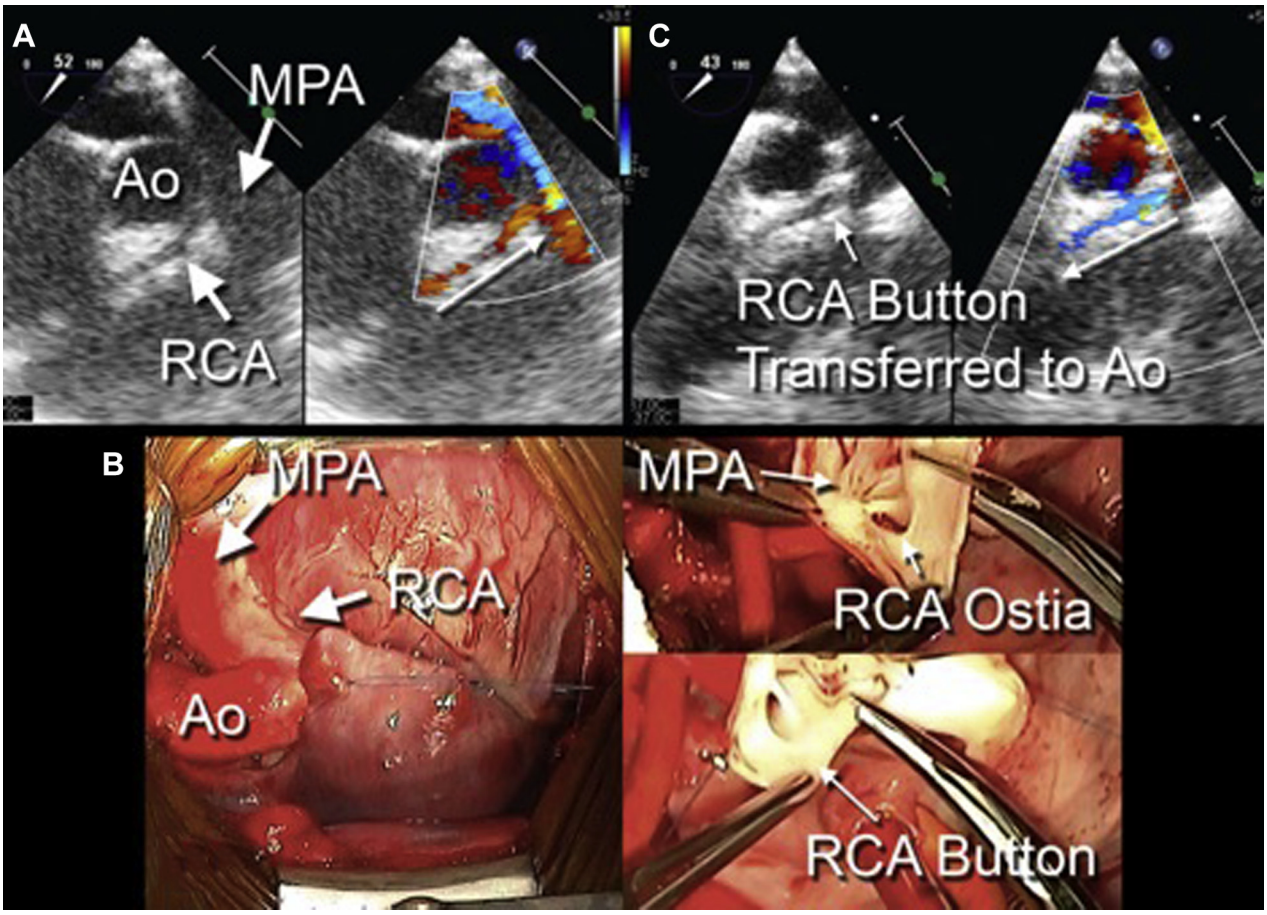

Figure 4 Preoperative transthoracic echocardiogram shows flow reversal in the right coronary artery (RCA), concerning for anomalous right coronary artery off the pulmonary artery **(A)**. Intraoperative findings are displayed **(B)**, along with postoperative transthoracic echocardiography showing normal flow directionality visualized in the RCA after reimplantation **(C)**. *Ao*, Aorta; *MPA*, main pulmonary artery.

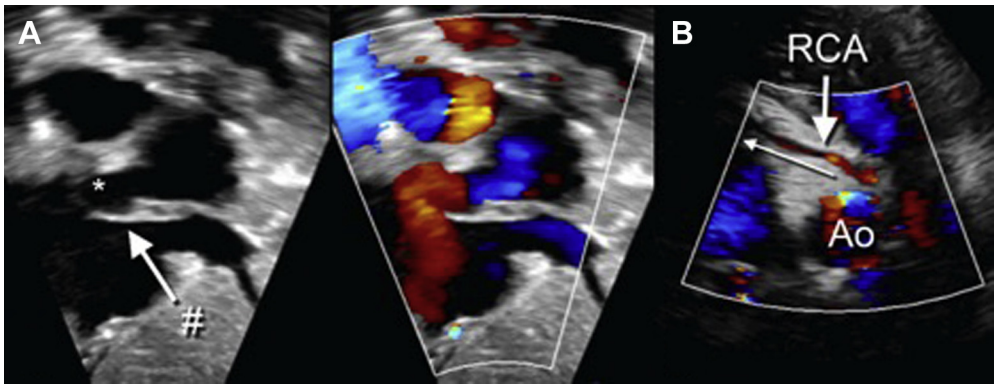

Figure 5 Postoperative transthoracic echocardiography before patient discharge. **(A)** Subcostal sagittal view demonstrates the patch *(pound sign)* that baffles the right upper pulmonary veins *(asterisk)* back into the left atrium. **(B)** Reimplanted right coronary artery (RCA) with antegrade flow from the aorta (Ao) into myocardium.

Figure 2B likely demonstrates collateralization from the left main coronary artery. In this case, the right coronary artery–to–pulmonary artery fistula was considered. Right coronary os atresia is another important consideration in the differential diagnosis of ARCAPA and should be evaluated for when a dilated left coronary artery is noted.[6]

In this case, transthoracic echocardiography was initially deceptive in definitive delineation of the right coronary artery origin. In retrospect, the imaging should have prompted preoperative coronary angiography. Fortunately, the flow reversal within the right coronary artery as noted on transesophageal echocardiography with scale optimization prompted visual confirmation of the diagnosis. If the run-off into the pulmonary arteries had not been addressed by snaring the pulmonary arteries distal to the ARCAPA site, cardioplegia instilled into the aortic root could have had run-off into the pulmonary circulation with suboptimal penetration into the myocardium, leading to myocardial ischemia and a poor postoperative outcome. Although long-term outcomes following surgical reimplantation of the right coronary artery are not well known, the general risk/benefit ratio appears to favor surgical correction when the diagnosis is known.

Ultimately, the patient did well and deescalated well following surgical repair with good antegrade flow across the reimplanted right coronary artery and no other residual cardiac disease found on transthoracic imaging (Video 5). He was discharged home on the fifth postoperative day with routine outpatient cardiology follow-up.

CONCLUSION

ARCAPA is an uncommon but important diagnosis to consider when flow reversal is seen within the right main coronary artery along with relative dilation of the left coronary artery system. Coronary artery origins should always be confirmed with color Doppler imaging. Detailed interrogation of coronary artery anatomy should be pursued through alternative imaging modalities when diagnosis by echocardiography is not completely clear.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.11.007.

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