JACC: CASE REPORTS VOL. 29, 2024

CASE REPORT

CLINICAL CASE

ARCAPA in Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collateral Arteries

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ABSTRACT

We report a case of anomalous origin of the right coronary artery from the pulmonary artery in a patient with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries. The diagnosis was made during a proposed hybrid approach to stent the native right ventricular outflow tract, and an alternative surgical strategy was created. (J Am Coll Cardiol Case Rep 2024;29:102318) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A female infant with an antenatal diagnosis of pulmonary atresia, ventricular septal defect (VSD), and major aortopulmonary collateral arteries (MAPCAs) was born in good condition with a birth weight of 3.65 kg and oxygen saturations that were stable in the mid to high 80s in room air. On examination, she had a normal first heart sound, a single second heart sound, and a 3/6 continuous murmur heard throughout the precordium and radiating to her back. Following discussion at the joint cardiac conference, she was referred from a nonsurgical cardiac center at day 18 of life for a semiurgent diagnostic cardiac catheterization under general anesthesia. The agreed plan was to attempt transcatheter right ventricular outflow tract (RVOT) perforation or hybrid subxiphoid periventricular needle perforation with the aim to provide antegrade blood flow to the hypoplastic native branch pulmonary arteries and encourage native pulmonary arterial growth.

LEARNING OBJECTIVES

- To appreciate the importance of the use of multimodality imaging to fully assess the unique anatomical variations that can be present in patients with pulmonary atresia, VSD, and MAPCAs, including MAPCA delineation, native pulmonary artery architecture, great artery relationship, and coronary artery variations.
- To understand the significance of delineation of coronary arteries in these patients prior to consideration of RVOT stenting.

PAST MEDICAL HISTORY

The patient had a normal microarray and no extracardiac morbidities.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received December 1, 2023; revised manuscript received February 7, 2024, accepted March 6, 2024.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2024.102318

ABBREVIATIONS AND ACRONYMS

ARCAPA = anomalous origin of the right coronary artery from the pulmonary artery

CT = computed tomography

MAPCAs = major aortopulmonary collateral arteries

MPA = main pulmonary artery

RCA = right coronary artery

RVOT = right ventricular outflow tract

VSD = ventricular septal defect

INVESTIGATIONS

Initial echocardiogram confirmed the antenatal diagnosis (Videos 1 and 2) and demonstrated multiple collaterals, some of which appeared quite proximal from descending aorta. Cardiac computed tomography (CT) had been performed in her local cardiac center prior to the cardiac catheterization. The CT illustrated diminutive branch pulmonary arteries of 1 to 2 mm in diameter, 2 large MAPCAs from the right and left subclavian arteries, respectively, and 3 additional MAPCAs that had not been previously seen on echocardiogram. The CT was nongated, and the report did not comment regarding coronary artery position.

She was consented for a transcatheter procedure, with potential for a subxiphoid/hybrid approach if the pulmonary outflow was truly atretic or if direct hybrid puncture was not achievable, sternotomy and central shunt insertion.

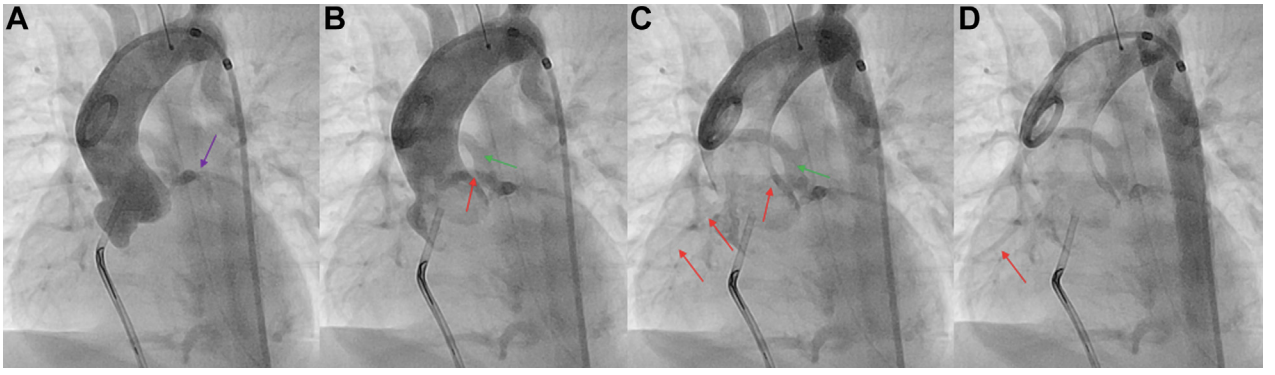
Cardiac catheterization and angiography showed there was no connection through the RVOT connection to the main pulmonary artery (MPA). Aortic angiography was suspicious for coronary anomalies (**Figure 1**, Video 3), and a decision to proceed to a subxiphoid approach was made to attempt direct perforation of the atretic pulmonary plate. On inspection, the surgical team noted that the anterior wall of the right ventricle appeared somewhat hypokinetic, which further increased the suspicion of a potential coronary anomaly. A median sternotomy was performed with direct inspection of the pulmonary artery that illustrated that the right coronary artery (RCA) originated from the anterior aspect of the diminutive main pulmonary artery, which confirmed the diagnosis of anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA). The presumed mechanism for the RV hypokinesia was increased coronary steal with full ventilation and 100% oxygen post intubation, which likely dropped the pulmonary vascular resistance and increased the coronary steal from the ARCAPA. Further review of the angiography and preoperative imaging (**Figures 2 and 3**) was clearer in the context of the findings on direct inspection and enabled a discussion regarding an alternative palliative approach.

MANAGEMENT

The chosen approach was to directly surgically anastomose the proximal main pulmonary artery, which fed into the anomalous coronary artery, onto the aortic root to facilitate normal coronary artery perfusion. The disconnected distal MPA and pulmonary artery confluence was connected to a central shunt to the ascending aorta.

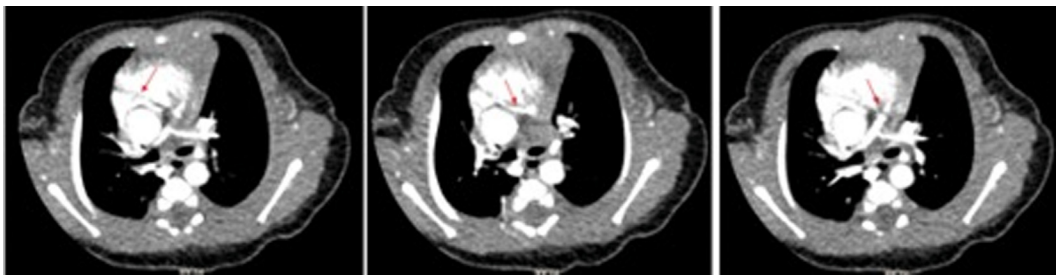
Once cardiopulmonary bypass and aortic cross clamp was established, the ascending aorta was

FIGURE 1 Serial Angiographic Images From Aortogram in the Ascending Aorta With 4-F Pigtail Catheter



(A) Contrast fills the ascending aorta, and the left coronary artery is delineated by the purple arrow. (B) Contrast passes from the aorta via the major aortopulmonary collateral arteries to the main pulmonary artery and branches, denoted by the green arrow. Early filling of the proximal anomalous origin of the right coronary artery from the pulmonary artery is highlighted by the red arrow. (C) The anomalous origin of the right coronary artery from the pulmonary artery then fills with contrast highlighted by red arrows. (D) Distal right coronary artery filling.

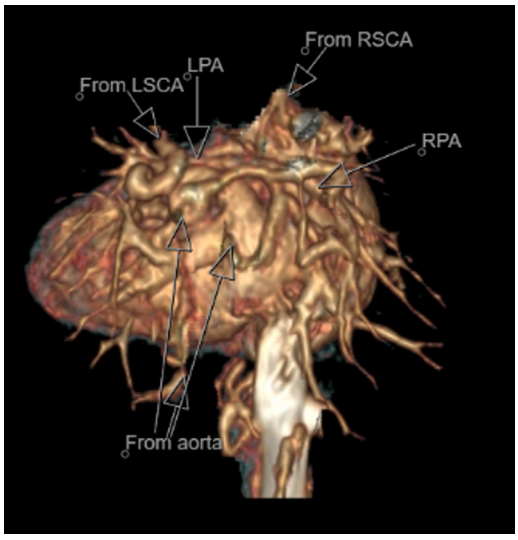
FIGURE 2 Cardiac CT Axial Images Demonstrating Anomalous Origin RCA From the Proximal MPA



Anomalous origin of the right coronary artery from the pulmonary artery indicated by red arrows. CT = computed tomography; MPA = main pulmonary artery; RCA = right coronary artery.

opened anteriorly and incised along the left side of the right coronary sinus and the incision was extended along the adjacent side of main pulmonary artery root for a V-shaped anastomosis. Autologous pericardial patch was used to augment aortic closure to relieve tension at the pulmonary root. A 4-mm punch was made on the ascending aorta for the 4-mm Propaten graft (Gore Medical) to be anastomosed to the pulmonary artery confluence. The patient came off bypass successfully with a bypass time of 89 minutes and a cross clamp time of 59 minutes. When weaning off bypass, there was still some dysfunction of the ventricle and moderate inotropic support, so the chest was left open so as not to compromise the heart further as the ventricular function improved over the next few days.

FIGURE 3 CT 3D Reconstruction Illustrating MAPCAs Supply to Confluent Branch Pulmonary Arteries



This computed tomography (CT) 3-dimensional (3D) reconstruction shows how the major aortopulmonary collateral arteries (MAPCAs) supply blood to the confluent branch pulmonary arteries. LPA = left pulmonary artery; LSCA = left subclavian artery; RPA = right pulmonary artery; RSCA = right subclavian artery.

The patient made good postoperative progress in the intensive care unit. She had delayed sternal closure until day 6 postoperatively. The patient was then extubated and transferred to the ward at day 9 post procedure (Video 4), and she made consistent progress weaning from high flow oxygenation at day 10. She was subsequently transferred back to her local center at day 11 postoperatively to continue to re-establish breast feeds and wean from nasogastric feeding.

FOLLOW-UP

Following discharge from hospital, the patient was able to establish breastfeeding and gain weight. Follow-up echocardiograms have demonstrated normal biventricular systolic function. Although her pulmonary artery branches remain small, there is good flow supplied via the central shunt, reflected by her oxygen saturation levels in the low 90s. Outpatient electrocardiograms have illustrated normal sinus rhythm with no evidence of ischemic changes.

DISCUSSION

ARCAPA is an extremely rare congenital cardiac lesion; the true incidence of which remains unknown.[1] There are no reported cases in the literature of ARCAPA in the setting of pulmonary atresia, VSD,

and MAPCAs with confluent branch pulmonary arteries. Although this was previously reported in the setting of aortopulmonary window and tetralogy of Fallot.[2-5] A similar case was noted in a previous ARCAPA case series published in 2006,[5] in a patient with tetralogy, pulmonary atresia with right pulmonary artery off the descending aorta, and left pulmonary artery off the ascending aorta. This patient was diagnosed with ARCAPA intraoperatively following RCA ligation and died on the first postoperative day. We feel that this comparable case only further serves to highlight the importance of delineation of coronary artery anatomy in patients with pulmonary atresia, VSD, and MAPCAs prior to consideration of intervention or surgery.

In our patient with pulmonary atresia, VSD, and MAPCAs, the ARCAPA diagnosis was made during the cardiac catheterization hybrid procedure. If she had proceeded to have RVOT stenting, this may have resulted in significant RCA diastolic steal and reduced RCA perfusion pressure following reduction in the pulmonary arterial diastolic pressures, or occlusion of the origin of the RCA by the stent, with subsequent myocardial ischemia and dysfunction.

Analysis of CT and echocardiographic imaging in these patients can be particularly difficult,[6,7] given the diminutive nature of the main pulmonary artery and the native branch pulmonary arteries and their proximity to the aortic root. MAPCA flow on echocardiogram can also make flow analysis within the native branch pulmonary vasculature challenging given the potential for antegrade and retrograde flow depending on MAPCA size and location.

CONCLUSIONS

This case highlights the importance of correct delineation of coronary arteries in these patients prior to consideration of RVOT stenting via transcatheter or hybrid/subxiphoid approach.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS anomalous, major aortopulmonary collateral arteries, pulmonary atresia, right coronary, ventricular septal defect

APPENDIX For supplemental videos, please see the online version of this paper.