A Multimodality Imaging and Multidisciplinary Approach to Manage Anomalous Right Coronary Artery from the Pulmonary Artery in Pregnancy

Offdan D. Narvaez-Guerra, MD, Nouran Sorour, MD, Gerard P. Aurigemma, MD, FASE, Matthew W. Parker, MD, FASE, David J. Kanter, MD, and Lara C. Kovell, MD, *Worcester, Massachusetts*

INTRODUCTION

Anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital coronary anomaly that is characterized by an abnormal connection of the right coronary artery (RCA) to the pulmonary artery (PA), as opposed to its typical origin from the aorta. This anatomical anomaly disrupts normal coronary circulation, leading to a spectrum of clinical syndromes from silent myocardial ischemia to infarction and left ventricular dysfunction and can be fatal if left untreated. We present a case of ARCAPA previously diagnosed in a young woman, which later represented a delivery planning challenge during a subsequent pregnancy. Identification of this condition, risk stratification, and multidisciplinary team planning during pregnancy are key elements in preventing complications.

CASE PRESENTATION

A 37-year-old patient, gravida 3 para 2, with a history significant for recurrent urinary tract infections, nephrolithiasis, and gastroesophageal reflux disease, presented to the cardiology clinic to reestablish care. Fifteen years prior, the patient emigrated to the United States, and a positive tuberculin test led to tuberculosis screening, which revealed an enlarged cardiac silhouette on chest x-ray. Transthoracic echocardiography (TTE) demonstrated preserved biventricular systolic function without regional wall motion abnormalities, dilated left main (LM) coronary artery originating from the left sinus of Valsalva, and an RCA originating from the PA (Figures 1-4, Videos 1-5). Color-flow Doppler demonstrated diastolic flow through prominent septal collateral arteries as seen in Video 1. A subsequent cardiac computed tomography (CCT) showed marked dilation of the LM and RCA with evidence of multiple collaterals between the left anterior descending (LAD) artery and RCA. The origin of the RCA was confirmed to be from the proximal PA (ARCAPA; Figure 5).

Management decisions over the subsequent 15 years involved a multidisciplinary approach (Figure 6). The patient had 2 uneventful pregnancies at the ages of 25 and 27 years old with spontaneous vaginal deliveries at term. Over the years, the patient developed occasional, transient chest pain that was evaluated with treadmill stress echocardiography, in which the patient exercised for over 10 minutes on the Bruce protocol with no echocardiographic or electrocardiographic evidence of ischemia. When chest pain and shortness of breath recurred in 2019, the patient was offered surgical intervention, specifically RCA unroofing, but initially declined and subsequently had intermittent limited interactions with health care. Three years before the current presentation, the patient was evaluated for dyspnea, worsening exercise tolerance, and fatigue for which an exercise treadmill stress test was conducted, which was nondiagnostic due to failure to achieve the target heart rate, although asymptomatic and without ischemia at the workload achieved (Figure 6).

While remaining largely asymptomatic until the third pregnancy in 2023, fatigue and mild exertional dyspnea surfaced during the prenatal period. In light of the history of ARCAPA and for preeclampsia prevention, preventive therapy was initiated with aspirin. Upon reestablishing care with cardiology, the patient was carefully monitored throughout their pregnancy. A repeat TTE showed preserved biventricular function, with better visualization of intramyocardial collaterals (Figure 7). At 27 weeks of gestation, the patient underwent a repeat treadmill stress echocardiogram, exercising for 7 minutes and 3 seconds on the Bruce protocol, reaching a heart rate of 155 bpm (85% of the age-predicted maximum heart rate). There was no evidence of ischemia observed in the electrocardiogram or echocardiographic assessments, and the resting blood pressure was 115/66 mm Hg with a normal response to stress.

A 7-day cardiac event monitor did not reveal any tachyarrhythmias, and lab testing revealed a brain natriuretic peptide of 20 pg/mL. A fetal echocardiogram was normal. Ultimately, after a thorough multidisciplinary discussion between maternal fetal medicine, cardiology, and anesthesia, the patient proceeded with a spontaneous vaginal delivery, supported by continuous cardiac telemetry monitoring and early epidural anesthesia. The consideration of anticoagulation was eventually dismissed, as there is a dearth of comprehensive guidance endorsing its application in individuals with ARCAPA and the patient had no prior thromboembolic event. Additionally, no evidence suggests that ARCAPA patients exhibit a higher level of hypercoagulability compared to the general pregnant population. The patient successfully delivered a healthy child at 38 weeks and 5 days of gestation after spontaneous rupture of

From the Division of Cardiovascular Medicine, Department of Medicine (O.D.N.-G., N.S., G.P.A., M.W.P., L.C.K.), and Division of Maternal-Fetal Medicine, Department of Obstetrics and Gynecology, University of Massachusetts Chan School of Medicine, Worcester, Massachusetts (D.J.K.).

Keywords: Cardio-obstetrics, Anomalous coronary, Congenital heart disease, Pregnancy

Correspondence: Lara C. Kovell, MD, UMass Chan Medical School, 55 Lake Avenue North, Worcester, MA 01655. (E-mail: *lara.kovell@umassmemorial.org*).

2468-6441

https://doi.org/10.1016/j.case.2023.12.013

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal short-axis view with color-flow Doppler, demonstrates flow through septal collaterals, as well as intermittent visualization of the anomalous RCA.

Video 2: Two-dimensional TTE, right ventricular inflow view, demonstrates dilation of anomalous RCA.

Video 3: Two-dimensional TTE demonstrating marked LM coronary dilation.

Video 4: Two-dimensional TTE, parasternal short-axis view with color-flow Doppler, demonstrates retrograde flow from RCA into PA.

Video 5: Two-dimensional TTE, parasternal short-axis view, demonstrates both marked dilation of the LM coronary artery during late diastole and marked dilation of an anomalous RCA originating from the PA during systole.

View the video content online at www.cvcasejournal.com.

membranes and was discharged home without complications on postpartum day 3.

DISCUSSION

ARCAPA is a rare congenital coronary abnormality with an incidence in those undergoing coronary angiography of approximately 0.002%, with a slight male predominance.[1] Anomalies of coronary artery origin may be the result of malrotation of the coronary buds and/or malrotation of the spiral septum during division of the truncus arteriosus.[2] Even though it frequently presents as an isolated lesion, 40% of patients with ARCAPA have other congenital cardiac defects, including aortopulmonary window (most common), atrial septal defects, ventricular septal defects, bicuspid aortic valve, and tetralogy of Fallot.[3] While anomalous origin of the left coronary artery from the PA (ALCAPA) tends to exhibit a more severe clinical presentation characterized by malignant arrhythmias, systolic dysfunction, myocardial infarction, and/or sudden cardiac death, ARCAPA is frequently diagnosed incidentally likely because of the lower right ventricular oxygen demands and the relatively lower contribution of the RCA to left ventricular blood supply as compared to the left coronary system.[4] However, patients with a dominant RCA system do not tolerate the ARCAPA circulation when compared to those with a left dominant circulation.[5] More than two-thirds of patients with ARCAPA are asymptomatic, and the most common clinical finding is a murmur.[1] When overtly symptomatic, dyspnea and chest pain are the most common symptoms.[1]

ARCAPA results in low arterial intracoronary oxygen levels that trigger coronary steal syndrome and eventually myocardial ischemia. During the immediate neonatal period, high PA pressure causes antegrade flow of deoxygenated blood in the RCA; this will be reversed as PA pressure decreases and flow through the RCA follows a PA-to-RCA direction.[6] Subsequently, collateralization between the RCA and the left system, usually through the LAD artery, takes place, confirming a left-to-right shunt leading to RCA dilation, with the LM being the main supplier of oxygenated blood to the myocardium.[1] These adaptations are the pathophysiologic basis of imaging findings in ARCAPA. Transthoracic echocardiography is an ideal, noninvasive tool that safely identifies various hallmark findings. Right coronary artery dilation, coronary collaterals, and retrograde RCA flow from the PA are seen in 32%, 28%, and 25% of cases, respectively.[3] It is important to note that off-axis echocardiography can help identify the anomalous origin of the RCA from the anterior aspect of the PA trunk, especially if there is blood flow reversal in the proximal RCA. Anterior tilting of the ultrasound probe from the parasternal short axis at the level of the great vessels can help bring into view different segments of the PA including the one containing the anomalous RCA. Collaterals between the RCA and the LAD are predominantly located in the anterior RV wall as well as the interventricular septum.[7] These connections are not as pronounced as in cases of ALCAPA where color-flow Doppler echocardiography of the myocardium can reveal extensive collaterization, conforming the so-called heart on fire sign.[8] In our patient, the use of CCT allowed for identification of the anterior RV and septal collaterals. Cardiac computed tomography is fundamental in evaluating coronary anatomy and high-risk features of anomalous coronaries including intramural course, slit-like orifice, or acute-angle takeoff, all of which were not present in this patient.[4] A comprehensive review and graphical depiction of such features

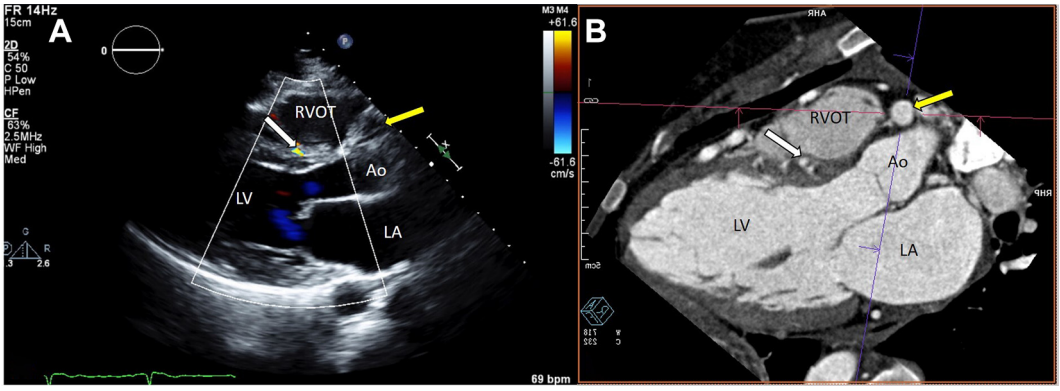

Figure 1 Two-dimensional TTE, parasternal long-axis view, color-flow Doppler **(A)**, shows flow during diastole through septal collaterals (*white arrow*), as well as the anomalous RCA (*yellow arrow*), which is dilated and can be visualized between the RVOT and proximal ascending aorta. Multiplanar reconstruction of CCT images at the same echocardiographic level **(B)** allows for better visualization of the aforementioned structures. *Ao*, Ascending aorta; *LA*, left atrium; *LV*, left ventricle; *RVOT*, right ventricular outflow tract.

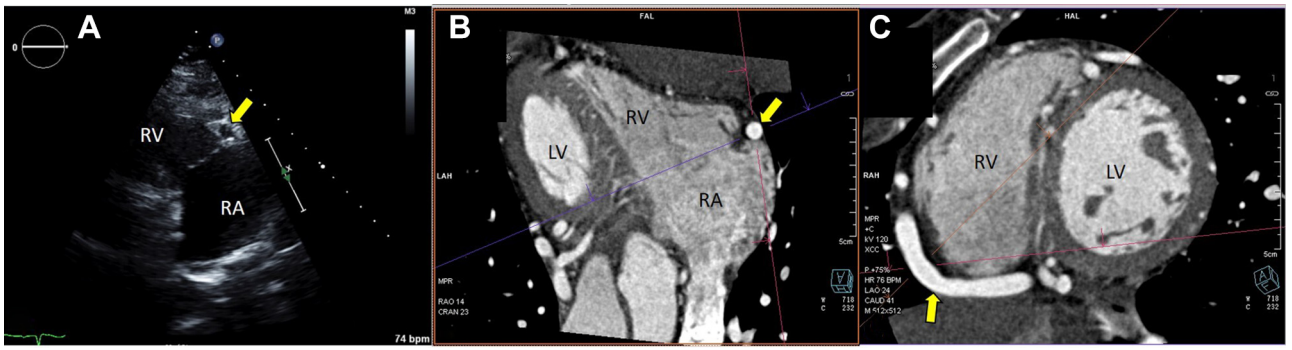

Figure 2 Two-dimensional TTE, parasternal long-axis view of the right ventricular inflow **(A)**, demonstrates a dilated anomalous RCA (*yellow arrow*) located in the right atrioventricular groove, which is better visualized in the corresponding multiplanar reconstruction view from CCT **(B)**. The dilated RCA can be seen making an angle between the inferolateral and inferior right ventricular wall **(C)**. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

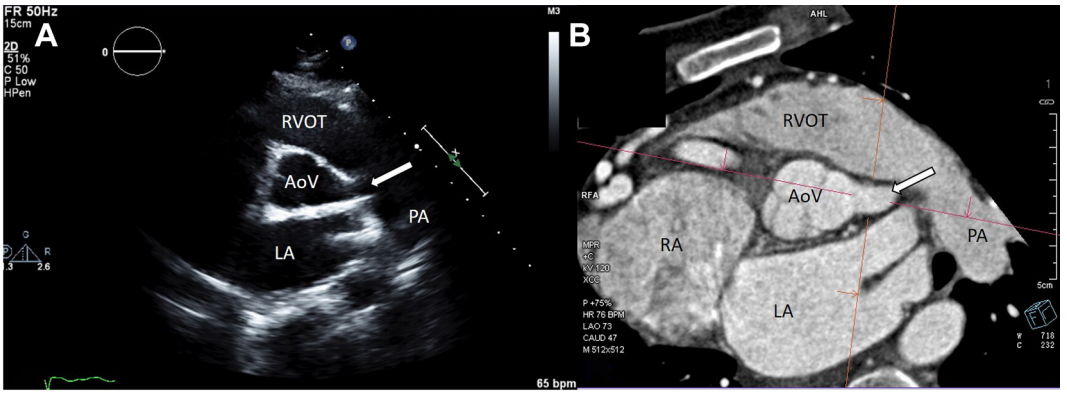

Figure 3 Two-dimensional TTE, parasternal short-axis view at the level of the great vessels during diastole **(A)** and corresponding reconstructed view from CCT at the same level **(B)**, demonstrates a markedly dilated LM coronary artery (*white arrow*) originating from the left coronary cusp. *AoV*, Aortic valve; *LA*, left atrium; *RA*, right atrium; *RVOT*, right ventricular outflow tract.

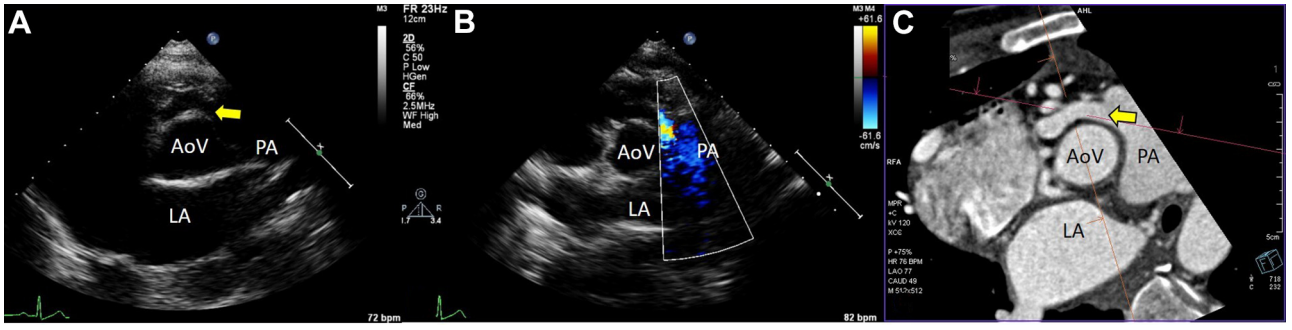

Figure 4 Two-dimensional TTE, parasternal short-axis view **(A)** demonstrates anomalous origin of RCA from the proximal PA (*yellow arrow*) located anteriorly in relationship to the aortic valve. Color-flow Doppler during diastole **(B)** shows flow acceleration at the origin of the anomalous RCA, indicative of retrograde flow from RCA into PA. Cardiac computed tomography multiplanar reconstruction image at the same echocardiographic level **(C)** details the anomalous origin from RCA in relationship to surrounding structures. *AoV*, Aortic valve; *LA*, left atrium.

have been compiled by Gräni *et al.*[9] The detailed origin of the RCA should be carefully examined with CCT, as ARCAPA can be mistaken for a coronary fistula when its origin is not clearly identified.[3]

Although our patient did not have any modified World Health Organization (mWHO) class IV risk factors for which pregnancy is contraindicated (pulmonary hypertension from any cause, severe ventricular dysfunction, severe mitral stenosis, or aortic stenosis), the only generic mWHO criteria met was mWHO class III as "other complex heart disease."[10] Similarly, other scores that predict cardiovascular morbidity and mortality among pregnant women with heart disease,

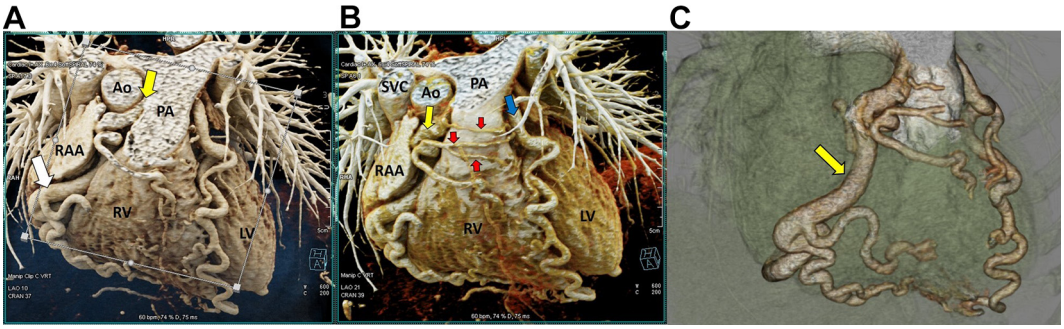

Figure 5 Cardiac computed tomography panels utilizing the cinematic volume rendering technique demonstrating three-dimensional coronary and pulmonary anatomy. **(A)** Highlights of the origin of the RCA from the PA, indicated by the *yellow arrow*. Additionally, it illustrates the marked tortuosity of the RCA, emphasized by the *white arrow*. **(B)** Collaterals (*red arrows*) between the proximal anomalous RCA and the LAD (*blue arrow*), which is enhanced by use of color coding. **(C)** Transillumination of myocardial tissue allows for visualization of a dilated and tortuous RCA, which can be seen crossing across the right atrioventricular groove (*yellow arrow*). *Ao*, Ascending aorta; *RAA*, right atrial appendage; *SVC*, superior vena cava.

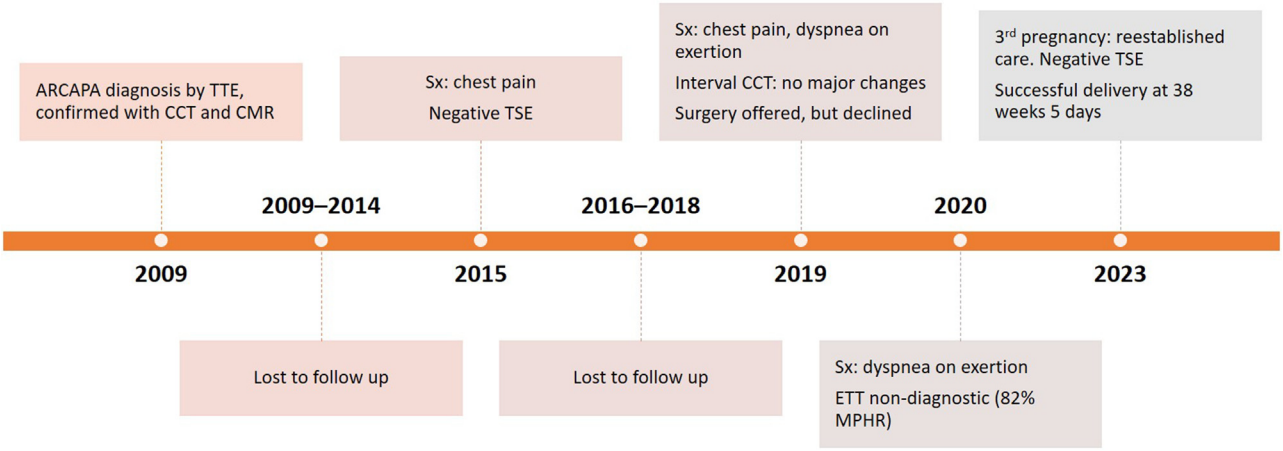

Figure 6 Clinical timeline. *ETT*, Exercise treadmill stress test; *Sx*, symptom; *TSE*, treadmill stress echocardiogram.

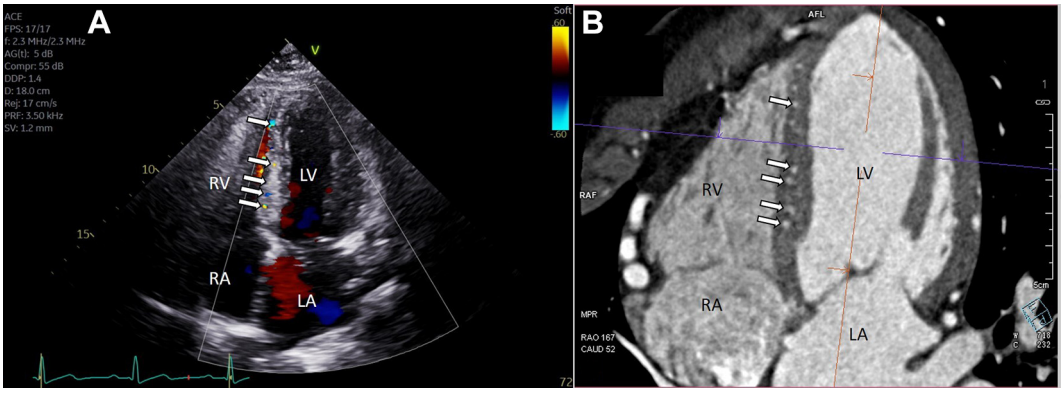

Figure 7 Two-dimensional TTE, apical 4-chamber view, color-flow Doppler **(A)**, demonstrates diastolic flow through myocardial septal collaterals (*white arrows*), basal to apical. Multiplanar reconstruction CCT images of the same echocardiographic level **(B)** detail the location of these collaterals. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

including the CARPREG-II and ZAHARA scores, do not specifically consider congenital coronary anomalies in their point allocation system.[10-12] However, the CARPREG-II score does consider coronary artery disease, defined as angiographically proven coronary obstruction or past myocardial infarction, which may have been relevant for this patient given the underlying coronary anatomy and risk for

ischemia.[10] Although the patient underwent an uneventful vaginal delivery, cesarean delivery would have been considered if ischemic symptoms recurred or if the stress test revealed evidence of ischemia.

Our patient became more symptomatic with dyspnea on exertion during the second trimester. This can be explained in part by 2 phenomena. Pregnancy-induced increase in pulmonary blood flow and a small increase in PA pressure may limit left-to-right shunting through the anomalous RCA, with a possible subsequent increase in intravascular RCA pressure and interstitial myocardial pressure.[5,12,13] It is also possible that normal pregnancy-induced upregulation of vascular endothelial growth factor and coronary angiogenesis is not completely normal in an anomalous coronary artery with retrograde flow.[5,12]

Current American Heart Association/American College of Cardiology guidelines on the management of congenital heart disease provide a class I recommendation for treatment with surgery in symptomatic adults with ARCAPA with symptoms attributed to the anomalous coronary.[14] Nonetheless, it is unknown whether a surgical intervention in the anomalous origin of coronary arteries improves survival.[14] While initially declining surgical intervention, our patient is now undergoing reevaluation in the postpartum period. Available surgical techniques include reimplantation of the RCA directly into the aorta, which is performed in approximately 60% of patients,[1] or ligation of the RCA at the level of the PA with coronary artery bypass grafting.[14]

CONCLUSION

This patient illustrates the complementary roles of echocardiography and CCT in detailing the anatomical and hemodynamic features of ARCAPA, a rare congenital anomaly that can present with a wide range of clinical manifestations. Adequate risk stratification, multidisciplinary team approach, and tailored management are crucial in delivery planning for pregnant patients with congenital anomalies of the coronaries.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a noninterventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

ACKNOWLEDGMENTS

We thank Christopher Stuart and Matthew Geary for their time and effort in obtaining the computed tomography images for the case and our wonderful sonographers for their excellent images.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.013.

REFERENCES

1. Guenther TM, Sherazee EA, Wisneski AD, Gustafson JD, Wozniak CJ, Raff GW. Anomalous origin of the right coronary artery from the pulmonary artery: a systematic review. Ann Thorac Surg 2020;110:1063-71.
2. Vergara-Uzcategui CE, das Neves B, Salinas P, Fernández-Ortiz A, Núñez-Gil IJ. Anomalous origin of coronary arteries from pulmonary artery in adults: a case series. Eur Heart J Case Rep 2020;4:1-5.
3. Ajam A, Rahnamoun Z, Sahebjam M, Sattartabar B, Razminia Y, Ahmadi Tafti SH, et al. Cardiac imaging findings in anomalous origin of the coronary arteries from the pulmonary artery; narrative review of the literature. Echo Res Pract 2022;9:12.
4. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller G-P, et al. 2020 ESC guidelines for the management of adult congenital heart disease: the task force for the management of adult congenital heart disease of the European Society of cardiology (ESC). Endorsed by: Association for European Paediatric and congenital cardiology (AEPC), International Society for adult congenital heart disease (ISACHD). Eur Heart J 2020;42:563-645.
5. Sanghavi M, Rutherford JD. Cardiovascular physiology of pregnancy. Circulation 2014;130:1003-8.
6. Williams IA, Gersony WM, Hellenbrand WE. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. Am heart J 2006;152:1004. e9-17.
7. Wu LP, Zhang YQ, Chen LJ, Liu YQ. Diagnosis of anomalous origin of the right coronary artery from the pulmonary artery by echocardiography. J Med Ultrason (2001) 2019;46:335-41.
8. Kerut EK, Kogos PG, Anderson JH, Turner M, Ascuitto R, Ross-Ascuitto N, et al. Adult presentation of ALCAPA: echo and CT diagnosis. Echocardiography 2018;35:1045-8.
9. Gräni C, Buechel RR, Kaufmann PA, Kwong RY. Multimodality imaging in individuals with anomalous coronary arteries. JACC Cardiovasc Imaging 2017;10:471-81.
10. Elkayam U, Goland S, Pieper PG, Silversides CK. High-risk cardiac disease in pregnancy. J Am Coll Cardiol 2016;68:396-410.
11. Silversides CK, Grewal J, Mason J, Sermer M, Kiess M, Rychel V, et al. Pregnancy outcomes in women with heart disease: the CARPREG II study. J Am Coll Cardiol 2018;71:2419-30.
12. Mehta LS, Warnes CA, Bradley E, Burton T, Economy K, Mehran R, et al. Cardiovascular considerations in caring for pregnant patients: a scientific statement from the American Heart Association. Circulation 2020;141:e884-903.
13. Sharma R, Kumar A, Aneja GK. Serial changes in pulmonary hemodynamics during pregnancy: a non-invasive study using Doppler echocardiography. Cardiol Res 2016;7:25-31.
14. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. Circulation 2019;139:e698-800.