

# Anomalous Right Coronary Artery Arising from the Pulmonary Artery with Associated Enlargement of the Left Coronary Artery: A Case Report

Yueqiu Su, MD,<sup>1</sup> Zhongze Cao, MD,<sup>2</sup> Yunfei Ling, MD, PhD,<sup>3</sup> Yongjun Qian, MD, PhD<sup>3</sup>

<sup>1</sup>Department of Anesthesiology, West China Hospital, Sichuan University, Chengdu, Sichuan, China;

<sup>2</sup>West China School of Medicine, Sichuan University, Chengdu, Sichuan, China<sup>a</sup>, Chengdu, Sichuan, China;

<sup>3</sup>Department of Cardiovascular Surgery, West China Hospital, Sichuan University, Chengdu, Sichuan, China

## ABSTRACT

An anomalous right coronary artery arising from the pulmonary artery (ARCAPA) is among the least common form of congenital coronary anomalies, accounted for the incidence of only 0.002% in the general population. Most ARCAPA patients have no symptoms but may develop myocardial ischemia. Surgical correction of the anomaly is recommended to prevent subsequent fatal outcomes.

Here, we reported a case of a 2-year-old female child initially hospitalized for diarrhea, but later diagnosed with an ARCAPA through echocardiogram and computed tomography. Surgical reimplantation of the right coronary artery from the pulmonary artery to the ascending aorta was performed. The patient recovered well from the surgery with no postoperative complications. In the follow-up assessments, normal coronary function and myocardial effusion were demonstrated.

## INTRODUCTION

Anomalous right coronary artery arising from the pulmonary artery (ARCAPA) is rare and accounted for the incidence of only 0.002% in the general population [Williams 2006], which represents the least common form of congenital anomalies. To date, only approximately 200 cases have been reported in literature since the first case series of ARCAPA was described by Brooks in 1885 [Wu 2019].

In patients with ARCAPA, the symptoms and patterns of disease progression vary widely depending on the development of functional myocardial collateral branches and variations in pulmonary arterial pressure [Guzeltas 2017]. In the majority, ARCAPA patients will have no significant or specific symptoms. However, the anomaly is considered to be lethal, given that sudden death from myocardial infarction or heart failure may ensue even in completely asymptomatic patients [Guzeltas 2017]. Therefore, surgical correction is strongly

recommended for all patients diagnosed with ARCAPA to normalize cardiac perfusion.

## CASE REPORT

A 2-year-old female child was initially admitted to the local hospital for diarrhea, but unfortunately initial medical records were unavailable. The echocardiogram at the time revealed findings suggestive of a septal ventricular defect. However, a year later, a repeat echocardiogram showed the likelihood of an ARCAPA. There was no other significant past medical history identified. The patient was therefore transferred to West China Hospital for further management.

Upon admission, inspection of the chest showed that the apical pulse was located at 0.5 cm interior of the midclavicular line and the fifth intercostal space. There was no abnormal uplift or pit at the precordial area. On palpation, the apical pulse appeared normal with no pericardial friction or tremor. The percussion examination suggested a normal heart border. There was no murmur on auscultation, and heart rhythms were normal. A transthoracic echocardiogram was performed, which revealed a normal systolic function of the left ventricle with no valvular malfunction or septal defect, as well as normal right ventricular function. The ARCAPA was suggested in addition to the aneurysm-like enlargement of the left coronary artery.

To validate the diagnosis further, a multislice computed tomography angiography was performed. The origin of the anomalous right coronary artery was located at the root of the pulmonary artery (Figure 1A, 1B), 6 mm above the valvular leaflets on the right position with an inner diameter of 3.1 mm. The artery then ran past the front of the aorta and followed the outline of the right atrium toward the right atrioventricular groove, eventually divided into four major branches. (Figure 2) The left coronary artery had originated from the left coronary aortic sinus and appeared tortuous and dilated. (Figure 3) The inner diameter measured 2.9 mm in the main branch and 3.1 mm in the anterior descending branch. (Figure 1)

The patient then underwent surgical correction of the anomaly. In surgery, the anomalous right coronary artery was re-implanted to the right coronary sinus and a temporary pacemaker was implanted. The aneurysm-like enlargement of the left coronary artery did not require any surgical intervention given its mild abnormality.

Received November 20, 2020; received in revised form January 27, 2021; accepted January 28, 2021

Yongjun Qian MD PhD, West China Hospital Sichuan University 37th Guoxue Lane Chengdu 610041 Sichuan China (e-mail: [qianyongjun@scu.edu.cn](mailto:qianyongjun@scu.edu.cn))

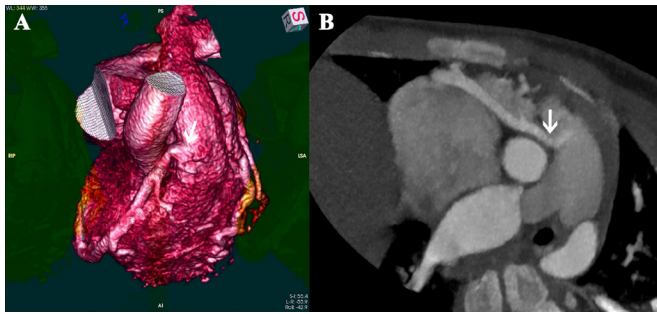


Figure 1. A) Three-dimensional reconstruction showing the right coronary artery emerging from the main pulmonary artery. B) Short axis view showing the right coronary artery emerging from the main pulmonary artery.

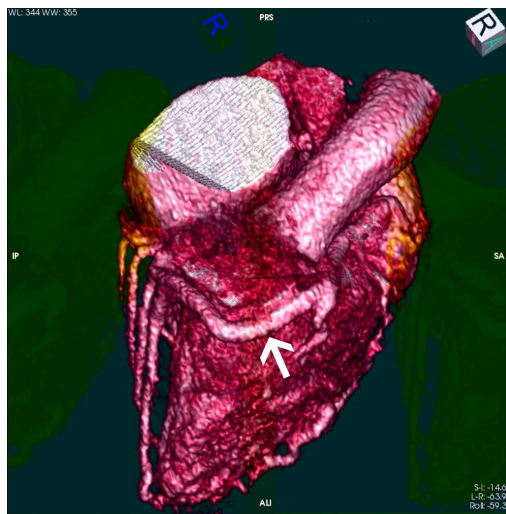


Figure 2. Three-dimensional reconstruction showing the course of the right coronary artery.

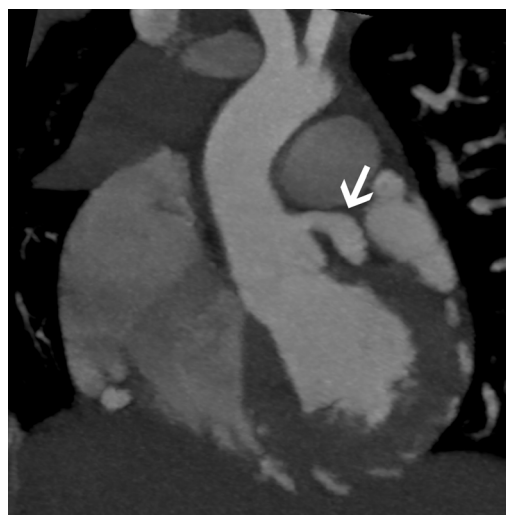


Figure 3. Long axis view showing the aneurysm-like enlargement of the left coronary artery.

The patient recovered from the surgery uneventfully. Post-operative echocardiogram revealed a normal laminar flow of the implanted right coronary artery into the right coronary sinus accompanied by a normal myocardial function. Upon hospital discharge, the patient was prescribed a two-week course of oral furosemide 10 mg per day and oral potassium chloride solution 6 ml per day, and a three-month course of oral aspirin 50 mg per day. The patient remained well at the last follow up.

## DISCUSSION

ARCAPA is an extremely rare type of congenital coronary anomaly with a very small pool of patients. During a normal fetal developmental period, blood flows from the pulmonary artery into the anomalous coronary artery as a result of high pulmonary vascular resistance [Williams 2006]. After birth, collaterals develop and blood flow reverses, from the anomalous coronary artery to the pulmonary artery due to decreased pulmonary vascular resistance [Afolabi-Brown 2014]. Diversity in patients' clinical signs and symptoms is attributed to variations in coronary blood flow and myocardial perfusions. Most patients with ARCAPA remain asymptomatic and may be diagnosed incidentally because of a heart murmur. At times, coronary steal may occur from the left to the right coronaries through the development of intermediate collaterals, leading to symptoms of myocardial ischemia, including chest pain, dyspnea, fatigue, congestive heart failure, myocardial infarction, and even sudden death [Saavedra 2018]. In our case, the patient was diagnosed to have an ARCAPA at a relatively young age with no cardiac symptoms. She presented initially with diarrhea and thus, the diagnosis of ARCAPA was likely to be incidental.

Not uncommonly, an ARCAPA is accompanied by other cardiac defects, which have a protective role against the steal from the left to the right coronary, such as the stenosis at the origin of the anomalous right coronary artery and dominant left coronary circulation. Left-to-right shunts, including the patent ductus arteriosus, aortopulmonary window and ventricular septal defect, can increase pulmonary vascular resistance and promote blood flow from the pulmonary artery into the anomalous right coronary artery [Ramani 2017]. In this case, the dominant left coronary perfusion might have exerted a protective function. The aneurysm-like enlargement of the proximal left coronary artery is likely a parallel anomaly.

Several investigative modalities can be used to diagnose ARCAPA. Coronary angiography is considered the gold standard for confirming an ARCAPA, given that it has a superior spatial and temporal resolution. However, it is an invasive procedure which therefore has limitations in clinical utility [Wu 2019]. On the other hand, computed tomography can delineate detailed anatomical information regarding the origin, course, and collaterals of the anomalous coronary artery, but it exposes patients to ionizing radiation and requires contrast agents [Restrepo-Cordoba 2016]. Echocardiography is noninvasive, convenient, and safe but provides insufficient visualization of the size and course of the entire coronary artery, due to suboptimal image quality [Robinson 2014]. In our case, the initial diagnosis was suggested by the

transthoracic echocardiography, which was further validated by the computed tomography angiography.

Surgery is recommended upon the diagnosis of ARCAPA, even in completely asymptomatic patients to prevent sudden death from myocardial ischemia [Al-Dairy 2017]. Surgical approach includes either reimplantation of the anomalous right coronary artery or ligation of the right coronary artery at its origin from the pulmonary artery with or without concomitant grafting to itself [Al-Dairy 2017]. The former technique involves reimplanting the right coronary artery from the pulmonary artery to the ascending aorta to re-establish dual systems of coronary artery perfusion. The practice of the latter technique is debatable given the uncertainty of the long-term graft patency [Hauser 2005]. For our patient, we performed reimplantation of the anomalous vessel to the right wall of the ascending aorta because the origin of the anomalous right coronary artery was in close vicinity of the aorta. Results during follow up suggested this surgical approach can lead to satisfactory results for ARCAPA originating closely from the aorta.

In conclusion, ARCAPA is a rare, congenital coronary anomaly usually with no specific symptoms but may cause sudden, unexpected death due to myocardial ischemia. Several investigative modalities can be applied for diagnosing ARCAPA, and the treatment involves surgical correction, which is recommended for all patients with the disorder.

## REFERENCES

- Afolabi-Brown O, Witzke C, Moldovan R, Pressman G. 2014. A different kind of Christmas tree: anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA). *Echocardiography*. 31(2):E52-4.
- Al-Dairy A, Rezaei Y, Pouraliakbar H, Mahdavi M, Bayati P, Gholampour-Dehaki M. 2017. Surgical Repair for Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery. *Korean Circ J*. Jan; 47(1):144-147.
- Guzeltas A, Ozturk E, Tanidir IC, Kasar T, Haydin S. 2017. Evaluation of Anomalous Coronary Arteries from the Pulmonary Artery. *Braz J Cardiovasc Surg*. Jan-Feb 32(1):29-37.
- Hauser M. 2005. Congenital anomalies of the coronary arteries. *Heart*. Sep;91(9):1240-5.
- Ramani J, Ananthanarayanan C, Pujara J, Thakkar B, Jain I, Mishra A. 2017. Intramural Anomalous Right Coronary Artery From the Main Pulmonary Artery. *World J Pediatr Congenit Heart Surg*. Nov; 8(6):745-749.
- Restrepo-Cordoba MA, Arellano-Serrano C, Mingo-Santos S. 2016. Right Coronary Artery With Anomalous Origin: The Role of Imaging Techniques. *JACC Cardiovasc Interv*. Jul 25; 9(14):e137-9.
- Robinson JA, Maclellan-Tobert SG, Horner JM, Cetta F. 2014. Pitfalls in diagnosis: suspected anomalous origin of the right coronary artery from the pulmonary artery. *Tex Heart Inst J*. Feb; 41(1):51-4.
- Saavedra MJ, Mozzi J, Nápoli N, Villa A, Barretta J, Marantz P. 2018. Anomalous origin of the right coronary artery from the pulmonary artery in an infant with a heart murmur: Case report. *Arch Argent Pediatr*. Dec 1; 116(6):e789-e792.
- Williams IA, Gersony WM, Hellenbrand WE. 2006. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. *Am Heart J*. 152(5):1004.e9-17.
- Wu LP, Zhang YQ, Chen LJ, Liu YQ. 2019. Diagnosis of anomalous origin of the right coronary artery from the pulmonary artery by echocardiography. *J Med Ultrason* (2001). 46(3):335-341.