Cardiology in the Young

cambridge.org/cty

A rare disease detected in a school-age child aortopulmonary window with anomalous right coronary artery from the pulmonary artery

Perver Arslan[1], Sezen Atik Ugan[2] and Alper Guzeltas[3]

[1]Pediatric Cardiology, İstanbul Mehmet Akif Ersoy Göğüs Kalp Ve Damar Cerrahisi Eğitim Ve Araştırma Hastanesi, Istanbul, Turkey; [2]Pediatric Cardiology, Cerrahpaşa Medical Faculity, Istanbul, Turkey and [3]Saglik Bilimleri Universitesi, Istanbul, Turkey

Brief Report

Cite this article: Arslan P, Atik Ugan S, and Guzeltas A (2023) A rare disease detected in a school-age child aortopulmonary window with anomalous right coronary artery from the pulmonary artery. *Cardiology in the Young* **33**: 2427–2429. doi: 10.1017/S1047951123001713

Received: 3 March 2023
Revised: 31 May 2023
Accepted: 1 June 2023
First published online: 11 July 2023

Keywords:
Aortopulmonary window; Anomalous right coronary artery from the pulmonary artery; Coronary anomalies

Corresponding author: P. Arslan;
Email: perwerars93@gmail.com

Abstract

Aortopulmonary window is a condition characterized by a communication between the pulmonary artery and the ascending aorta. The coexistence of aortopulmonary window and an anomalous right coronary artery originating from the pulmonary artery is rarely observed together, as mentioned in previous studies. In this report, we aim to describe our diagnostic and treatment experiences with a 6-year-old patient diagnosed with aortopulmonary window associated with an abnormal origin of the right coronary artery from the pulmonary artery.

Case report

A 6-year-old boy presented to our clinic due to a heart murmur detected during routine health screening. A systolic thrill was noted at the left sternal edge, accompanied by a grade 4/6 continuous ejection systolic murmur. The electrocardiogram revealed left ventricular dilatation. Chest radiography showed a dilated main pulmonary artery, resulting in a cardiothoracic ratio of 65%. Echocardiographic examination indicated enlargement of the left heart chambers. The color flow started just above the pulmonary valve and extended to the left pulmonary artery (Fig. 1). Although the outflow of the left coronary artery was observed, the outflow of the right coronary artery could not be clearly evaluated. As a result, it was planned to perform computerized tomography and angiography procedures for further differential diagnosis.

Computerized tomography imaging revealed a 7 mm diameter aortopulmonary window at the ascending aorta and proximal region of the pulmonary artery. Furthermore, it was determined that the right coronary artery originated from the proximal part of the pulmonary artery.

During cardiac catheterization, when injecting into the ascending aorta, it was observed that the pulmonary artery filled simultaneously, providing support for the diagnosis of an aortopulmonary window (Fig. 2a). Additionally, retrograde filling of the right coronary artery was observed during selective left coronary arteriography (Fig. 2b).

Based on these findings, a surgical operation was planned for the patient. During the operation, a 7 mm diameter defect between the aorta and the main pulmonary artery was observed and primarily repaired. It was also noted that the right coronary artery was located adjacent to the pulmonary artery. The right coronary artery was prepared as a button and reimplanted into the ascending aorta. The patient was extubated on the first day of intensive care follow-up and discharged on the fifth day of hospitalization without any complications. Subsequent follow-up at our pediatric cardiology clinic revealed that the patient remained asymptomatic.

Discussion

The coexistence of aortopulmonary window and an abnormal origin of the right coronary artery from the pulmonary artery is rare. There are limited studies in the literature demonstrating the presence of both conditions together[1,2]. Typically, this condition is detected in the neonatal period. However, in our case, it is noteworthy that the patient was 6 years old and remained asymptomatic.

Agarwala et al[3] reported a case of a 5-month-old female infant with aortopulmonary window and an abnormal origin of the right coronary artery from the pulmonary artery. The patient was born prematurely at 34 weeks with a birth weight of 2.15 kg. Continuous heart murmur was detected, and after undergoing surgical operation, the patient was discharged without complications. Similarly, Alakhfash et al[4] described a premature neonate who was diagnosed with aortopulmonary window and underwent a successful operation at the age of 4 months.

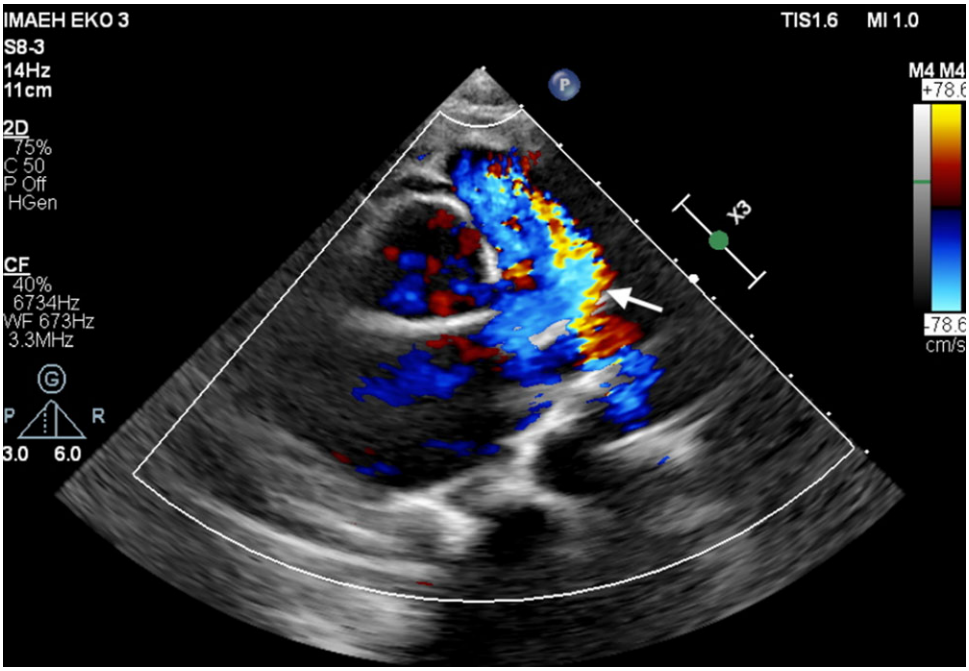

Figure 1. The color Doppler echocardiography image shows the flow of blood from the aortic root to the pulmonary artery and the right coronary artery flow jet directed into the pulmonary artery.

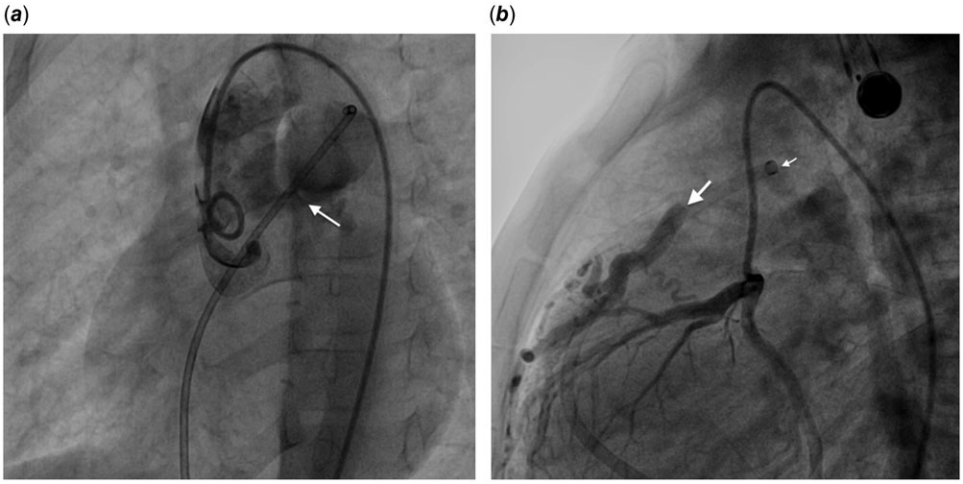

Figure 2. (***a***) The injection into the aortic root and the passage of contrast material into the pulmonary artery through the aortopulmonary defect. (***b***) The right coronary artery showed retrograde filling during selective injection of the left coronary artery.

In both of these cases, the patients were in the neonatal age group, and they were treated with early diagnosis. In contrast, our case was 6 years old at the time of diagnosis and had not yet developed any clinical symptoms.

Atalay et al[5] reported a case of a 45-day-old male infant who presented with congestive cardiac failure, murmur, and tachycardia. The patient underwent a successful one-step surgery. In this case, the detected diameter of the aortopulmonary window was around 7 mm, similar to our patient. However, it is important to note that the patient in this case had additional pathologies with left-right shunt, such as atrial septal defect and ventricular septal defect.

Wadhawa et al[6] described a case of a 19-year-old patient with tetralogy of Fallot and an anomalous origin of the right coronary artery from the aortopulmonary window. The patient presented with progressive dyspnea on exertion since childhood.

It is worth noting that in our case, the patient did not have any additional conditions other than the coexistence of the aortopulmonary window and the abnormal origin of the right coronary artery from the pulmonary artery. Furthermore, our patient was asymptomatic at the time of diagnosis.

In conclusion, when evaluating pediatric patients with only a murmur and no other clinical findings, the possibility of an aortopulmonary window should be considered. Although rare, the association between aortopulmonary window and coronary artery anomalies should be investigated in diagnosed cases. In situations where the coronary artery anatomy cannot be clearly

https://doi.org/10.1017/S1047951123001713 Published online by Cambridge University Press

evaluated or when there is suspicion, differential and diagnostic methods such as catheterization and computed tomography should be employed.

Acknowledgements. We would like to thank the paediatric cardiac surgeons (Dr Sertaç Haydin), who was involved in the care of this patient.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interests. None.

Ethical standard. The patient's parents provided informed consent for the publication of this case report.

References

1. Gabbieri D, Guadalupi M, Stefanelli G. Aortopulmonary window and anomalous origin of the right coronary artery from the pulmonary artery: long-term result after Takeuchi-type repair. Heart Lung Circ 2008; 17: 339–342. DOI: 10.1016/j.hlc.2007.05.002.
2. Barnes ME, Mitchell ME, Tweddell JS. Aortopulmonary Window. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2011; 14: 67–74. DOI: 10.1053/j.pcsu.2011.01.017.
3. Agarwala BN, Varga P, Hijazi ZM, Ziemer G. Rare case of aortopulmonary window with Anomalous origin of right coronary artery. Ann Thorac Surg 2015; 99: 1803–1805. DOI: 10.1016/j.athoracsur.2014.06.116.
4. Alakhfash AA, Tagelden M, Almesned A, Alqwaiee A. Aortopulmonary window with anomalous right coronary artery from the pulmonary artery. Case report and literature review. Cardiol Young 2020; 30: 47–49. DOI: 10.1017/S1047951119002543.
5. Atalay A, Soran Turkcan B, Kulahcioglu E, Eris D, Ece İ., Aydın H. Aortopulmonary window with anomalous origin of the right coronary artery from the pulmonary trunk. Cardiol Young 2021; 31: 838–841. DOI: 10.1017/S1047951120004783.
6. Wadhawa V, Mishra A, Kothari J. Anomalous right coronary from aortopulmonary window in tetralogy of Fallot. Asian Cardiovasc Thorac Ann 2015; 23: 1140–1140. DOI: 10.1177/0218492314537505.