

Case Report

Heart Murmurs Detected During Newborn Examination: A Case Report Highlighting the Need for Caution

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ABSTRACT

In this case report, we examine a 2-day-old infant referred for evaluation due to the detection of a heart murmur. Coronary artery fistula, an anomalous connection between coronary arteries and a cardiac chamber or vascular structure bypassing the typical capillary network, is the focus of our discussion. The newborn displayed no symptoms, and all other physical assessments appeared normal. Transthoracic echocardiography uncovered an enlargement of the coronary arteries, notably the proximal right coronary artery. Furthermore, a color Doppler scan indicated turbulent flow at the terminal portion of the ventricular septum, revealing a fistula linking the right coronary artery to the right ventricle. Throughout a 1-year follow-up period, the fistula spontaneously closed, resulting in the resolution of the heart murmur. Remarkably, the diameters of the coronary arteries reverted to the standard range. It is noteworthy that while surgical or interventional methods are sometimes used to treat these patients, spontaneous closure may occur in asymptomatic cases with close monitoring over time. (*Iranian Heart Journal 2025; 26(1): 107-112*)

KEYWORDS: Neonate, Congenital coronary fistula, Murmur

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Coronary artery fistula (CAF) involves an abnormal connection between 1 or more coronary arteries and either a cardiac chamber, known as a coronary-cameral fistula, or another vessel, referred to as a coronary arteriovenous malformation.¹ CAF is a rare occurrence, with a reported prevalence of 0.002% in the general population and accounting for 0.3% of congenital heart disease (CHD) cases.^{2,3} CAF most commonly originates from the right coronary artery (RCA) (55.0%), the left coronary artery (35.0%), or, in some instances, both coronary arteries (5.0%).^{4,5} The selection of an appropriate treatment

strategy for patients with CAF relies on various factors, such as fistula size, the presence or absence of symptoms, fistula anatomy, patient age, and any coexisting cardiovascular anomalies.^{5, 6} Treatment modalities encompass surgical ligation, percutaneous transcatheter closure, and conservative management.

This case report highlights a rare example of asymptomatic CAF occurring during the neonatal period.

CASE REPORT

A 2-day-old male newborn was referred to our clinic for evaluation following the

identification of a heart murmur before being released from the hospital. The neonate was born at full term via a normal delivery, and the mother had no health complications. Weighing 3200 g at birth, the baby exhibited strong Apgar scores of 9 and 10 in the first and fifth minutes postpartum, respectively. At our clinic, the newborn appeared healthy with no indications of illness. The physical examination revealed a blood pressure of 70/40 mm Hg, with no significant increase in pulse pressure. There were no signs of tachypnea, tachycardia, or hepatomegaly. Auscultation identified a grade II/VI continuous heart murmur with a diastolic focus at the left sternal border. Oxygen saturation levels were 98.0% in both the right hand and legs. The ECG yielded normal results, and the chest X-ray demonstrated a standard heart-to-chest ratio. Transthoracic echocardiography (TTE) disclosed a dilated coronary artery, with the proximal RCA diameter measuring 2.21 mm (Z score > 2.5). Furthermore, the left main coronary artery (LMCA), the left anterior descending artery (LAD), and the left circumflex artery (LCX) exhibited diameters of 2.57 mm (Z score > 2.5), 2.17 mm (Z score > 2.5), and 1.16 mm (Z score < 2), respectively. An anomalous flow pattern was detected at the terminal portion of the ventricular septum, leading to the discovery of a fistula between the RCA and the right ventricle. This post-tricuspid left-to-right shunt displayed a pulmonary-to-systemic blood flow ratio (Q_p/Q_s) of 1.2/1. TTE also revealed normal cardiac function and ventricular dimensions. There was no indication of reverse flow in the descending aorta, and no additional cardiac anomalies were detected (Figs. 1 & 2).

Following discharge, the newborn was closely monitored on an outpatient basis. The parents received thorough instruction on essential signs of heart failure to watch for in their child.

During the 1-month follow-up visit, the patient exhibited appropriate weight gain and displayed no clinical symptoms. Upon auscultation, a heart murmur could still be detected at the left sternal border. No significant changes were noted in the echocardiographic results when compared with the previous assessment. Coronary computed tomography angiography was conducted to evaluate the coronary arteries, revealing a dominant RCA and LAD. Notably, the RCA extended toward the apical inferior region of the right ventricle after branching from the distal LAD and exhibited a fistula connecting to the inferior right ventricular apex (Fig. 3). Throughout the follow-up period, as the newborn remained asymptomatic, the decision was taken to prioritize his growth and schedule any further interventions on an elective basis.

At the 1-year follow-up examination, no heart murmurs were identified during the physical assessment, and echocardiography results indicated that the fistula had closed. Additionally, there was a reduction in the diameters of the coronary arteries. The diameter of the RCA decreased from 2.21 mm to 1.71 mm, while the diameters of the LMCA, LAD, and LCX decreased from 2.57 mm, 2.17 mm, and 1.16 mm to 2.01 mm, 1.41 mm, and 1.10 mm, respectively. Importantly, all Z-score changes were below 2 (Fig. 4).

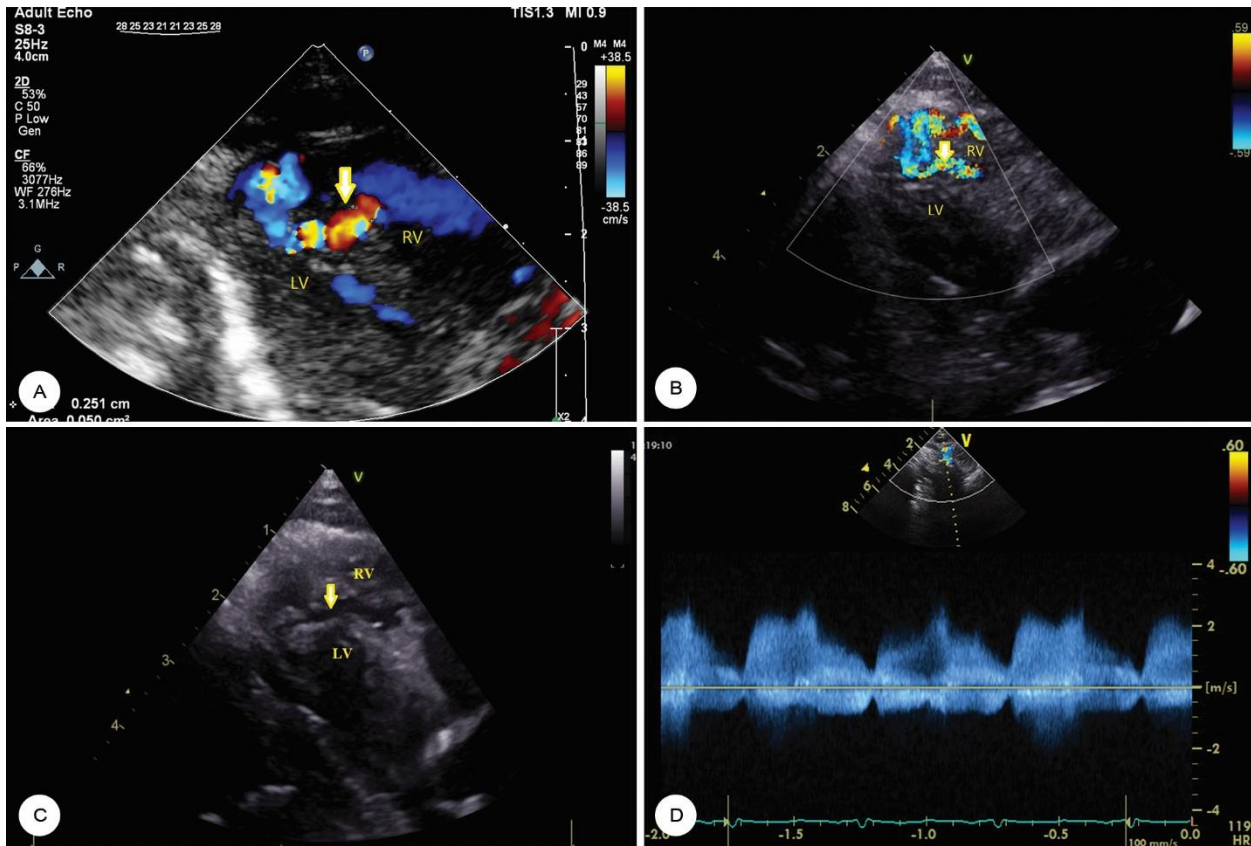


Figure 1: The images show a right coronary artery-RV fistula. A and B) The images showcase modified long-axis views (tilted anteriorly). Color Doppler indicates a turbulent, fast flow (the arrows). C) The image illustrates the course of the fistula over the RV septum (the arrows). D) Continuous wave Doppler shows a continuous spectrum of the fistula, predominantly in diastole, consistent with a coronary artery fistula.

RV: right ventricle, LV: left ventricle



Figure 2: A) The 4-chamber view shows a normal chamber size. B and C) The short-axis views indicate dilatation of the LMCA, the LAD, and the RCA.

LMCA: left main coronary artery, LAD: left anterior descending artery, RCA: right coronary artery

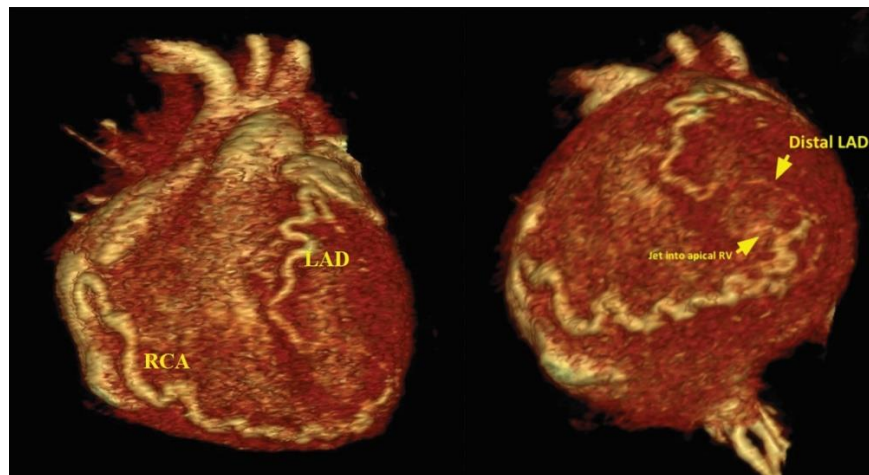


Figure 3: A and B) Volume-rendered computed tomography images display a coronary fistula originating from a dilated RCA and draining into the RV.

LAD: left anterior descending artery, RCA: right coronary artery

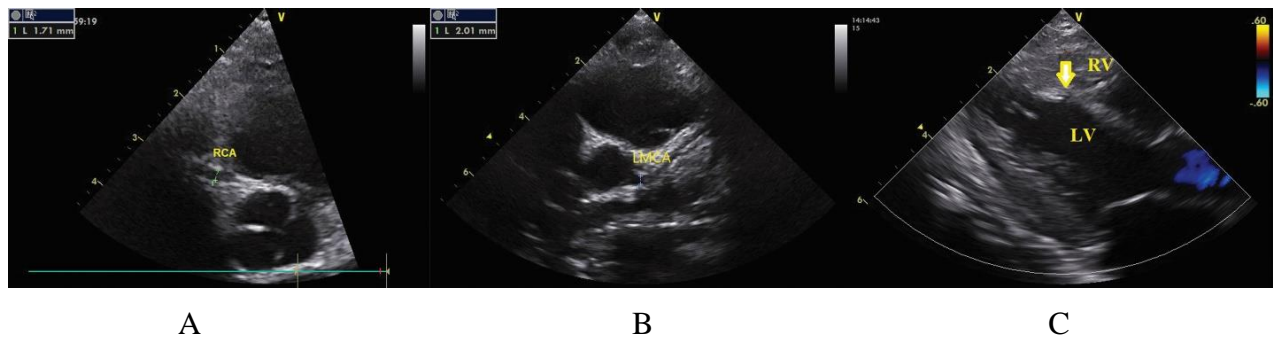


Figure 4: A and B) The short-axis views indicate the regression of the RCA, the LMCA, and the LAD to normal size after the closure of the fistula. C) The modified long-axis view (tilted anteriorly) represents the closure drainage of the fistula to the RV.

RCA: right coronary artery, LMCA: left main coronary artery, LAD: left anterior descending artery, RV: right ventricle

DISCUSSION

Heart murmurs that are incidentally detected are frequently observed in neonates during routine check-ups. While some heart murmurs during the neonatal period may be physiologic, CAF as the underlying cause of neonatal heart murmurs is exceedingly rare. The murmur linked to a CAF exhibits unique qualities, typically being most prominent during the latter portion of diastole. Thus, the term “innocent murmur” should be applied judiciously, particularly regarding diastolic murmurs. Distinguishing this murmur from those associated with other types of cardiac shunts, such as patent

ductus arteriosus or ventricular septal defect, relies upon identifying the anatomical location and the specific cardiac cycle phase in which they transpire. Diagnosing a CAF is achievable via TTE, but coronary computed tomography angiography serves as a dependable tool for assessing both coronary artery dimensions and fistula anatomy.⁷

There are 2 primary approaches for the treatment and management of CAF. Patients presenting with a substantial fistula, overt symptoms, or complications should undergo fistula closure (either through surgical

ligation or percutaneous transcatheter techniques).^{8,9}

Conversely, patients with a smaller fistula or smaller body size can be closely monitored. Throughout the follow-up period, it is crucial to evaluate clinical signs, physical symptoms, and echocardiographic results. One potential advantage of monitoring patients over time is the possibility of observing spontaneous fistula closure in some cases.

In cases where clinical symptoms become apparent and the condition has progressed, such as cases involving cardiac ischemia, arrhythmias, unexplained heart failure, or coronary artery enlargement, performing the procedure at a later age may be associated with fewer complications.^{10,11}

Our patient presented without clinical symptoms, and echocardiography did not reveal impaired cardiac function or progressive coronary dilation. Nevertheless, a significant concern pertained to the involvement of the LAD branch, which could potentially result in myocardial ischemia. Considering the patient's young age and low body weight, it was deemed appropriate to opt for a close monitoring strategy.

Although some experts advocate for closing small, asymptomatic CAFs during childhood before the patient reaches adulthood, our approach leans toward observing these patients and reassessing their condition over time through follow-up evaluations.¹²

Studies indicate that patients under 2 years of age have a higher probability of successful occlusion, particularly in cases involving smaller fistulas connected with right-sided structures, such as the right ventricle, and a QP: QS ratio below 1.5.⁹

CONCLUSIONS

The findings suggest that a substantial proportion of patients with CAF can be closely monitored, with the final decision on

definitive treatment determined during follow-up evaluations, particularly in neonates with a small fistula, minimal shunt volume, and adequate growth. Nonetheless, the treatment approach for pediatric patients should be individualized based on each patient's unique circumstances.

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Consent for Publication: The authors confirm that written consent for submitting and publishing this case report, including images and associated texts, has been obtained from the patient's parents.

REFERENCES

1. Qureshi SA. Coronary arterial fistulas. *Orphanet J Rare Dis.* 2006; 1:51.
2. Abdelmohsen G, Abd El Rahman MY, Dohain A, Latif SA, Attia W. Left circumflex coronary artery to coronary sinus fistula diagnosed in infancy. *J Cardiol Cases.* 2016; 15:97-99.
3. Song G, Ren W, Tang L, Hou Y, Zhou K. Coronary artery fistula from the left circumflex to coronary sinus in infant: Case report with literature review. *Int J Cardiol.* 2015; 188:37-39.
4. Song G, Zhang J, Ren W, Li Y, Zhou K. Pediatric coronary artery fistula: echocardiographic case reports and literature

- review of the treatment strategy. Springerplus. 2016; 5:1583.
5. Yun G, Nam TH, Chun EJ. Coronary Artery Fistulas: Pathophysiology, Imaging Findings, and Management. *Radiographics*. 2018; 38:688-703.
 6. Peighambari M, Pakbaz M, Alizadehasl A, Hosseini S, Pouraliakbar H. A Case of Coronary Cameral Fistula: When and How to Intervene? *J Tehran Heart Cent*. 2020; 15(4):189-194.
 7. Gupta-Malhotra M. Coronary artery fistulas. Available from <https://emedicine.medscape.com/article/895749-overview> (17 May 2023).
 8. Jenab Y, Khaledi AK, Ranjbarnejad H, Zoroufian A, Shahzadi M. Coronary Artery Fistula with Double Outlet Right Ventricle: a Case Report. *J Tehran Heart Cent*. 2007; 2(2):115-116.
 9. Yilmazer MM, Demir F, Yolbaş I, Bilici M. Spontaneous closure of a symptomatic coronary artery fistula just within a few days of newborn period. *Congenit Heart Dis*. 2014; 9:E27-30.
 10. Ismail AQ, Gandhi A, Desai T, Stumper O. A neonatal case of congenital coronary artery fistula. *BMJ Case Rep*. 2012; 2012:bcr0920114773.
 11. Kaldararova M, Tittel P, Zahorec M. Giant Coronary Artery Fistula: Prenatal Diagnosis, Newborn Manifestation. *Rev Esp Cardiol (Engl Ed)*. 2016; 69:1100.
 12. Lo MH, Lin IC, Hsieh KS, Huang CF, Chien SJ, Kuo HC, et al. Mid- to long-term follow-up of pediatric patients with coronary artery fistula. *J Formos Med Assoc*. 2016;115:571-576.