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CASE REPORT

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Anomalous origin of the right coronary artery with severe stenosis of the left coronary artery orifice: a case study

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Abstract

Background Anomalous aortic origin of the coronary artery (AAOCA) represents a form of congenital heart disease. Several primary types of anomalous origin of the coronary artery (AOCA) exist. Patients with certain AOCA types may be at risk of sudden cardiac death (SCD) if timely intervention is delayed.

Case description This study reports a case of an 11-year-old female pediatric patient who experienced multiple episodes of syncope before being evaluated at our hospital. She was diagnosed with an anomalous origin of the right coronary artery from the left coronary sinus, along with severe stenosis at the left coronary artery orifice, following transthoracic echocardiography (TTE) and other detailed examinations. The patient subsequently responded well to surgical unroofing for AAOCA combined with single internal mammary artery-coronary artery bypass grafting.

Conclusion As advancements in ultrasonic medicine progress, TTE has proven to be a valuable non-invasive method for diagnosing AAOCA. In this paper, the findings from echocardiography, computed tomography angiography (CTA), and ascending aortography of the patient were analyzed retrospectively to explore the diagnostic value of echocardiography for this condition and to enhance its diagnostic efficacy.

Keywords Cardiogenic syncope, Congenital, Coronary artery disease, Myocardial infarction, Transthoracic echocardiography

Background

Coronary artery anomalies encompass a range of irregularities, including anomalous origin and course of the coronary arteries, atypical internal coronary anatomy, anomalous termination, and variant vascular anastomosis. Anomalous origin of the coronary artery (AOCA) is categorized based on the origin location and subsequent course, primarily classified into anomalous origin of the left coronary artery, anomalous origin of the right coronary artery, and origin from the pulmonary artery. When the left main coronary artery arises from the right aortic sinus, it is termed an anomalous origin of the left coronary artery from the right sinus; conversely the right coronary artery originating from the left sinus is referred to

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as an anomalous origin of the right coronary artery from the left sinus.

Individuals with AOCA, particularly those with intramural segments in either the right or left coronary artery, are more susceptible to myocardial ischemia, a condition that can lead to sudden cardiac death (SCD) in rare cases. The anomalous origin of the right coronary artery is observed 3 to 8 times more frequently than that of the left coronary artery and generally carries a reduced risk of cardiac arrest. However, not all cases of right coronary artery anomalies are benign; approximately 10% are linked to with ischemia and a potential for cardiac arrest. Chest pain is the primary symptom associated with myocarditis—a non-ischemic inflammation—and isolated coronary artery anomalies. Both myocarditis and coronary artery anomalies are potential risk factors for sudden death in children and adolescents presenting with chest pain [1].

Case description

A female pediatric patient aged 11-years and 7 months was admitted to our hospital following two episodes of syncope within a span of 5 days. Previously, she had been evaluated at another hospital for multiple syncope episodes, where left ventricular anterior myocardial infarction and an anomalous origin of the right coronary artery were suspected. Due to the critical nature of her condition, transfer to a higher-level facility was recommended.

Upon referral, she was diagnosed with cardiogenic shock, acute anterior myocardial infarction, an anomalous origin of the right coronary artery, and fulminant myocarditis. A family member reported that the patient had experienced a previous syncope episode in 2022, which had not been further evaluated. The patient had no significant medical history.

An emergency transthoracic echocardiography (TTE) conducted on the day of admission revealed that the right coronary artery originated from the left coronary sinus, featuring an intramural aortic segment measuring approximately 0.8 cm in length and 0.14 cm in diameter, as well as an extramural aortic segment with a diameter of about 0.31 cm. The left main coronary artery opening was displayed unclearly (Fig. 1A), and the anterior descending branch measured approximately 0.15 cm (Fig. 1B). The left ventricle exhibited an increased internal diameter with a slightly rounded apex.

Notably, the myocardium in the medial and inferior segments of the interventricular septum, as well as the medial and inferior segments of the anterior wall and apical segments of the lateral and inferior walls of the left ventricle, was thinned to approximately 0.4 cm, exhibiting reduced mobility. Myocardial thickness in the remaining segments was slightly reduced. Additionally, a small echo-free area suggestive of fluid accumulation was detected in the pericardial cavity.

Color Doppler flow imaging (CDFI) identified intramural blood flow in the right coronary artery, indicating accelerated blood flow (Fig. 2). Blood flow in the left coronary artery remained unclear, with small, low-velocity blood flow signals observed in the anterior descending branch. The left ventricular ejection fraction (LVEF) was assessed at 38%.

Based on these findings, the right coronary artery was observed to anomalously originate from the left coronary sinus, containing intramural aortic segments, while the left coronary artery exhibited dysplasia, indicating proximal stenosis. Ultrasonography identified an extensive anterior myocardial infarction in the left ventricle, along with a minor pericardial effusion.

Computed tomography angiography (CTA) of the great vessels of the heart confirmed that the right coronary artery originated from the left coronary sinus of the aorta (Fig. 3A), while the left main coronary artery was not clearly visualized. The anterior descending branch (Fig. 3B) and the circumflex branch of the left coronary artery were visible. However, there was not clearly retrograde filling of the left anterior descending branch and circumflex coronary arteries. Additionally, the pulmonary trunk appeared widened, measuring approximately 2.5×2.1 cm in diameter. These findings were further supported by 3D reconstruction (Fig. 4).

CTA results reinforced the presence of an anomalous origin of the right coronary artery; however, the left main coronary artery was not accurately visualized, necessitating clinical correlation for proper assessment. The widening of the pulmonary trunk suggested pulmonary arterial hypertension. The final diagnosis, based on the imaging examinations, included anomalous origin of the right coronary artery, stenosis progressing to near-atresia of the left coronary artery, and acute anterior myocardial infarction.

An ascending aortography was performed the following day. Intraoperative findings confirmed that the right coronary artery originated from the left coronary sinus and appeared thickened. There was a slight delay in the development of both the anterior descending and circumflex branches of the left coronary artery, with filamentary blood flow observed in the left main coronary artery (Fig. 5). This provided an accurate diagnosis.

Based on the patient's diagnosis, surgical treatment was necessary to address two key issues: (1) an anomalous origin of the right coronary artery from the left coronary sinus with intramural deformation, and (2) severe proximal stenosis of the left coronary artery, approaching atresia. The planned procedure included coronary angioplasty - right coronary artery unroofing and left coronary

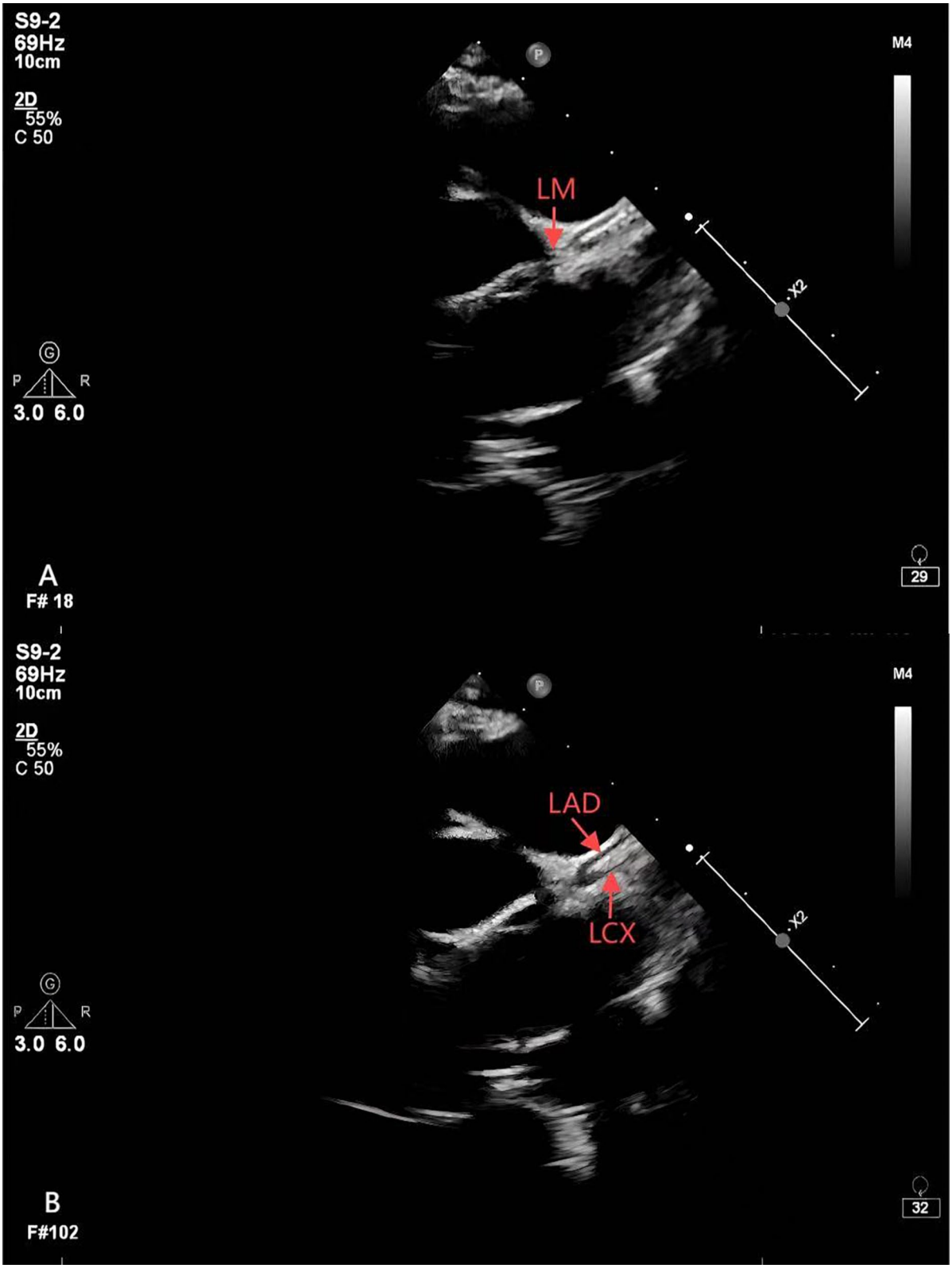

Fig. 1 Modified view of the parasternal short axis in echocardiography: (**A**) The left main coronary artery exhibits stenosis and is not clearly displayed at its opening. (**B**) The left anterior descending branch and the left circumflex branch are distinctly visualized
Abbreviations: LM: left main coronary artery; LAD: left anterior descending branch; LCX: left circumflex branch

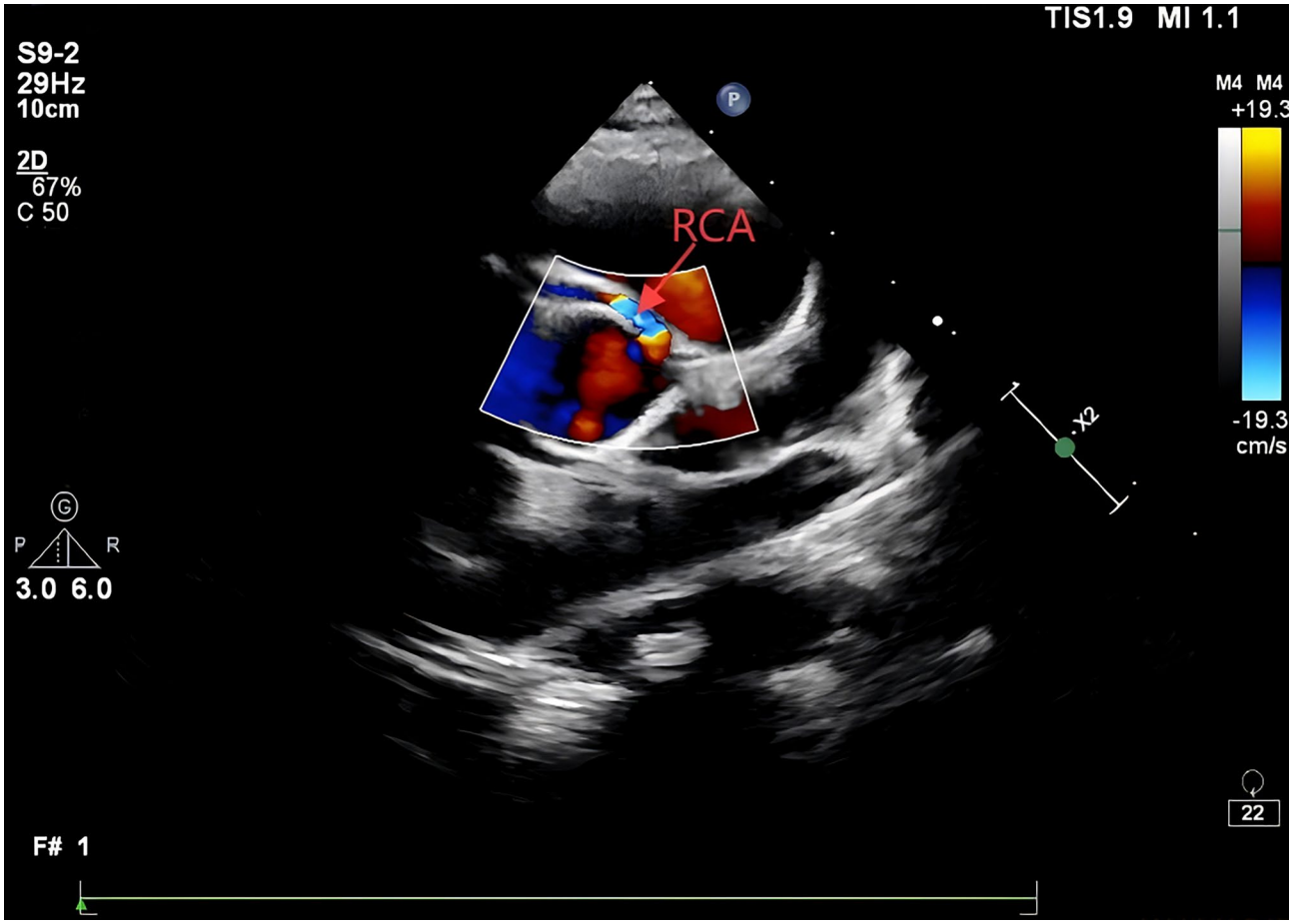

Fig. 2 The white box indicates the sampling location for color Doppler imaging. Color Doppler images show the right coronary artery originating from the left coronary sinus of the aorta, along with accelerated blood flow within the intramural segment
Abbreviation: RCA: right coronary artery

artery trunk enlargement - performed under general anesthesia with extracorporeal circulation.

Intraoperatively, the right coronary artery was found to originate from the left coronary sinus, adjacent to the lower edge of the petrous column, and travel within the aortic wall. The left main trunk exhibited severe stenosis, with near atresia and luminal narrowing throughout its course.

The aortic wall was incised along the right coronary artery until its exit from the ascending aorta. A pulmonary artery patch was used to enlarge the left main stem, the aortic incision was structured, and the pulmonary artery defect was repaired with a bovine pericardial patch. (Fig. 6) However, difficulties in weaning the patient from cardiopulmonary bypass were encountered, and left ventricular contractility remained poor.

To improve myocardial perfusion, the left internal mammary artery (LIMA) was harvested, and a LIMA-LAD anastomosis was performed, leading to hemodynamic stabilization. The final procedure consisted of coronary angioplasty combined with a single internal mammary artery-coronary artery bypass graft (CABG). The initial difficulty in discontinuing bypass was likely due to persistent ischemia caused by severe left main stenosis and near atresia, which was not fully corrected by left main enlargement alone.

Follow-up assessments were conducted at two and four months after coronary artery bypass grafting. Echocardiographic evaluations revealed a gradual improvement in left ventricular systolic function, with ejection fractions of 54% and 57%, respectively. Myocardial motion in the mid-to-lower segments of the interventricular septum, left ventricular anterior wall, and inferior wall also showed gradual improvement. These segments, primarily supplied by the left coronary artery, confirmed that the myocardial infarction was primarily caused by the near occlusion of the left main coronary artery.

Discussion

Anomalous origin of the coronary artery arises from embryonic dysplasia that leads to an atypical coronary artery location. On the 25th day of embryonic

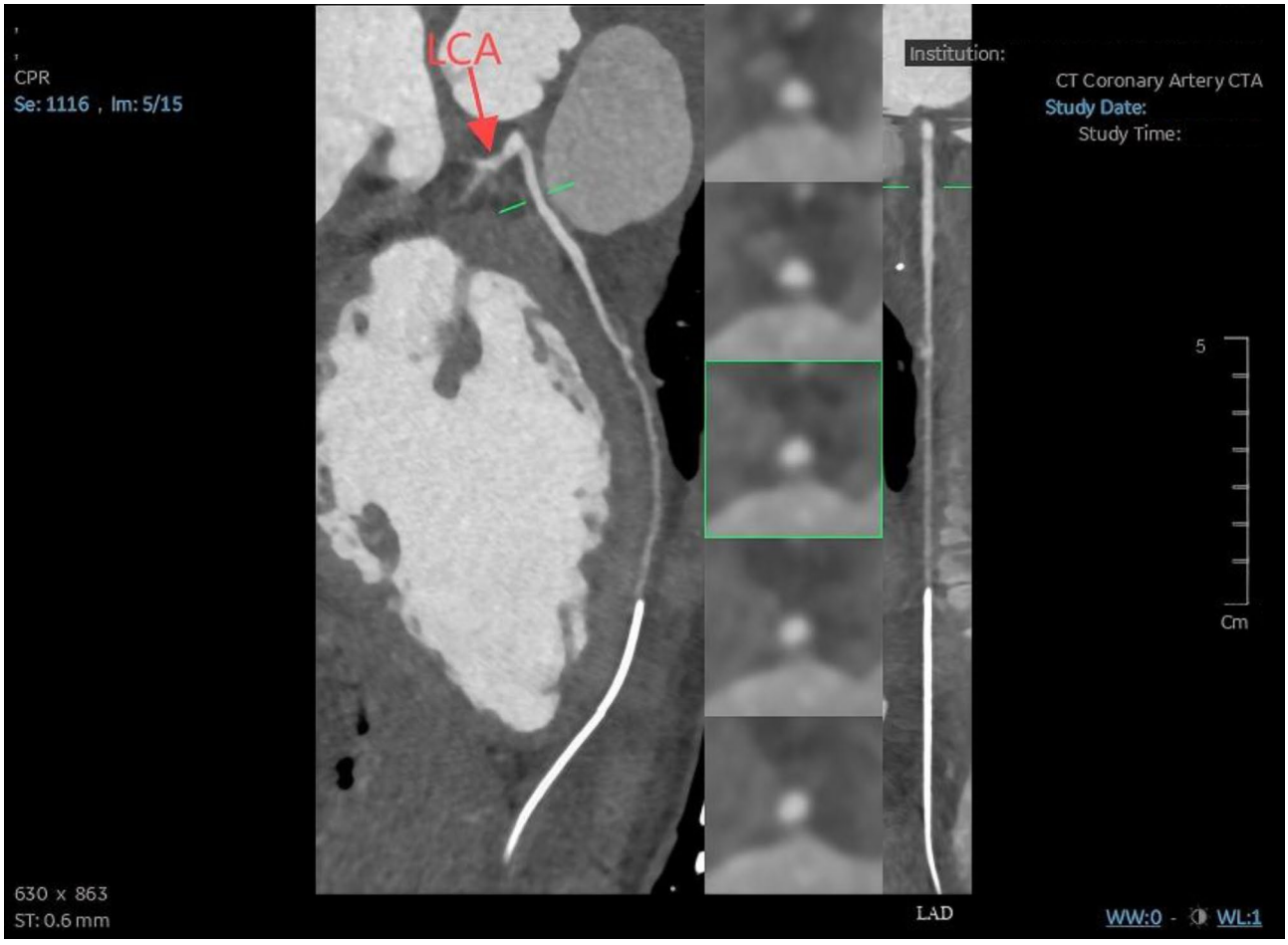

Fig. 3 CTA results: (**A**) The right coronary artery is observed originating from the left coronary sinus of the aorta. (**B**) The left main coronary artery is not clearly visualized, while the left anterior descending branch is distinctly displayed

development, discontinuous and angioid structures, initially devoid of blood flow, appear between the epicardium and myocardial cells. As embryonic development progresses, these angioid structures gradually fuse and penetrate the aorta, producing blood flow under pressure, which facilitates vascular maturation. Disruptions in this process can result in congenital dysplasia of the coronary arteries, where malrotation of the spiral septum or malposition of the coronary artery buds may lead to the anomalous origin of the coronary artery.

Advancements in echocardiography have enabled increased detection of anomalous origins of the right coronary artery, while the anomalous origin of the left coronary artery from the right coronary sinus remains rare [2]. Few patients with anomalous origin of the right coronary artery develop clinical manifestations, often remaining asymptomatic in infancy or early childhood, while angina may occur during adolescence [3]. This anomaly is closely related to SCD in children and adolescents, thus warranting clinical attention [4]. Intramural segments of coronary arteries in the aorta can create conditions conducive to myocardial ischemia or SCD. The pathophysiological mechanisms responsible for myocardial ischemia mainly involve several aspects: compression of coronary artery segments situated between the aorta and the pulmonary artery, especially during increased cardiac output; fissured openings or valvular ridges at the coronary artery opening that lead to stenosis; sharp angles at which intramural segments begin; and dysplastic characteristics of variant coronary arteries, which are prone to spasm due to endothelial injuries [5].

Conventionally, patients with ischemia are treated surgically, focusing on mitigating factors contributing to coronary artery occlusion [6]. Procedures include unroofing of intramural segments, correction, transposition, or reimplantation of the coronary artery into the appropriate sinus, with coronary artery bypass grafting as a less common approach [7]. Current perspective suggests that all anomalous components of the vessels must be treated to maintain the coronary arteries as closely aligned to their normal anatomical structure as possible. That is, all problems related to the opening and course of coronary

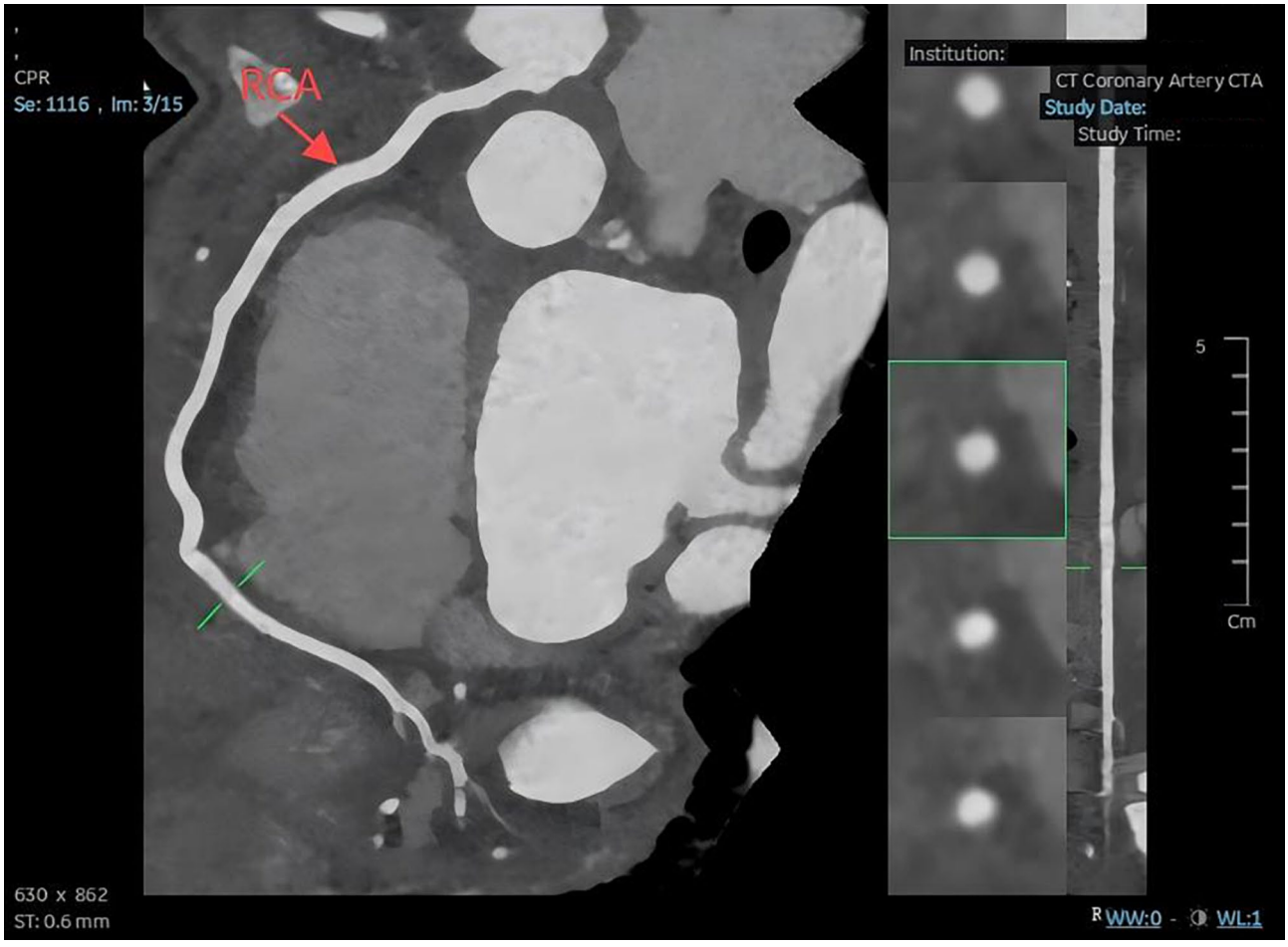

Fig. 4 CT 3D reconstruction: The long white arrow on the left indicates the right coronary artery, while the short white arrow on the right points to the left coronary artery

arteries must be resolved while ensuring the normal connection between the coronary artery and the sinus [8].

This rare case involves double coronary artery anomalies, wherein the patient presented with an anomalous origin of the right coronary artery alongside severe stenosis of the anatomical structure of the left coronary artery. The anomalous origin of the right coronary artery is a potentially fatal congenital condition. Although the incidence of sudden cardiac death due to the isolated anomalous origin of the right coronary artery from the left coronary sinus is low, it can lead to cardiac fibrosis and ventricular arrhythmias secondary to recurrent subclinical ischemic episodes.

When diagnosing this condition, it is crucial to differentiate between severe stenosis of the left coronary artery opening and a single coronary artery with left coronary artery atresia. In patients with left coronary artery atresia, the left coronary artery opening is not visible on 2D echocardiography, and the main coronary artery appears as a blind end. Additionally, left ventricular dilation may also be present, along with possible collateral circulation to the left ventricular myocardium. Color Doppler ultrasound can reveal bidirectional blood flow signals in the left coronary artery. Furthermore, aortic root radiography or selective right coronary arteriography can demonstrate retrograde perfusion of the right coronary artery to the branches of the left coronary artery, along with the development of the left main coronary artery and its blind-end appearance.

Echocardiography is generally employed to detect congenital coronary artery anomalies; however, diagnosing intramural segments of coronary arteries via 2D imaging can be somewhat challenging due to artifacts. Specifically, the anomalous coronary artery, upon leaving the aortic wall, may appear to originate from the correct coronary artery sinus. In such cases, color Doppler imaging plays a critical role in distinguishing between these anomalies by displaying blood flow signals and the course of the intramural segments [9, 10]. In this instance, the color Doppler ultrasound demonstrated intramural blood flow signals of the right coronary artery with accelerated

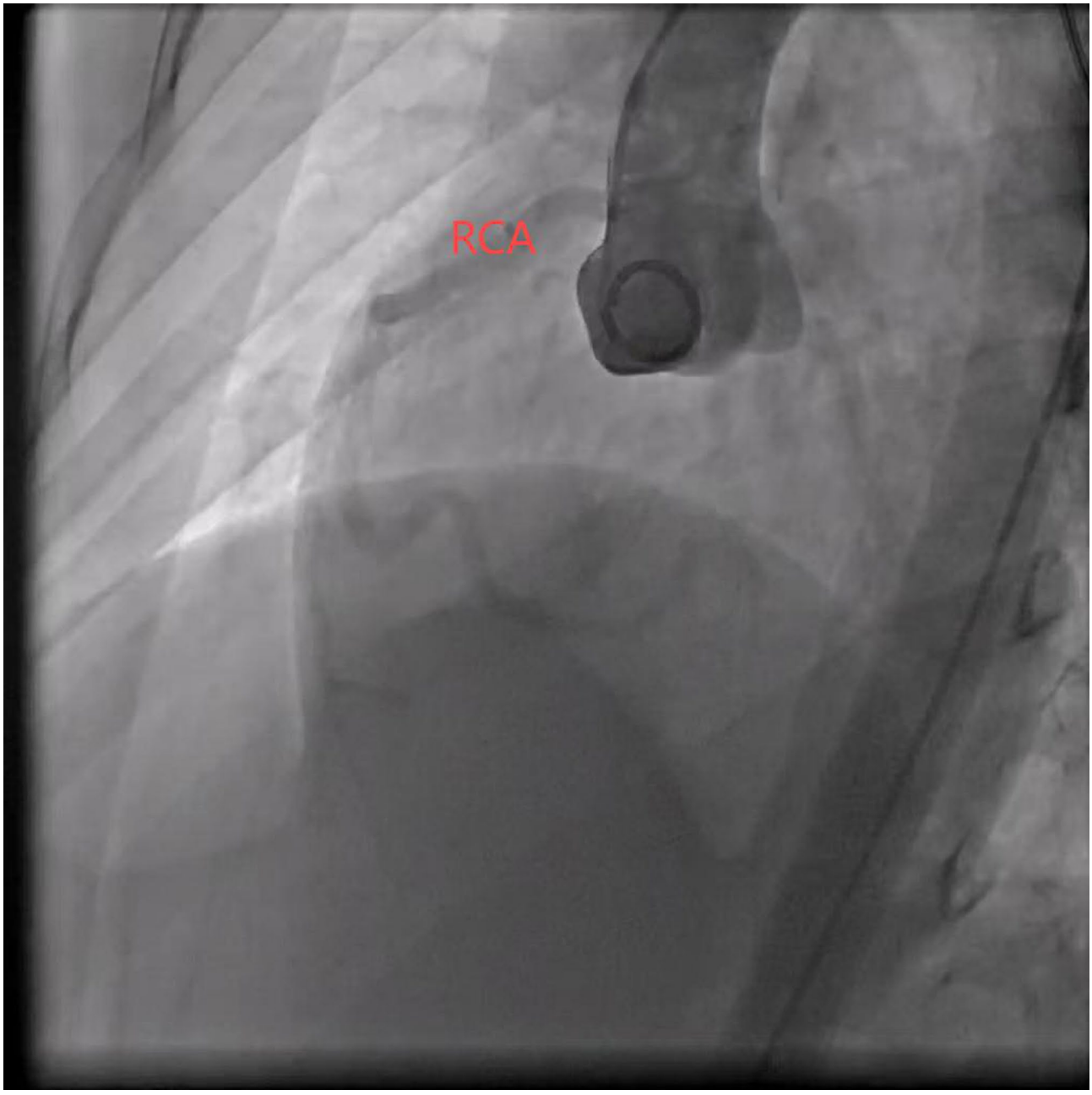

Fig. 5 Ascending aortography image: Both the right coronary artery and the left main coronary artery develop simultaneously. The right coronary artery appears thick, while the left main coronary artery exhibits filamentary blood flow. Additionally, there is a slight delay in the development of the anterior descending branch and circumflex branch of the left coronary artery

blood flow, indicating the presence of intramural segments within the aorta.

TTE remains the most valuable non-invasive imaging modality for accurately detecting coronary artery anomalies in children without the need for additional imaging techniques. Echocardiography can often identify anomalous coronary artery origins early on, providing both direct and indirect signs that facilitate diagnosis. Direct signs include the absence of the coronary artery opening in the corresponding aortic sinus. Indirect signs primarily indicate structural anomalies, such as increased left ventricular diameter, a slightly rounded apex, and myocardial motion abnormalities suggesting ischemic areas [11].

Echocardiography is a safe, non-invasive, and convenient method that effectively displays the origin and course of the coronary artery, ventricular structure, myocardial motion, and other relevant information. This makes it crucial for early diagnosis, surgical decision-making, prognosis assessment, and follow-up observations. Moreover, electrocardiograms can also provide

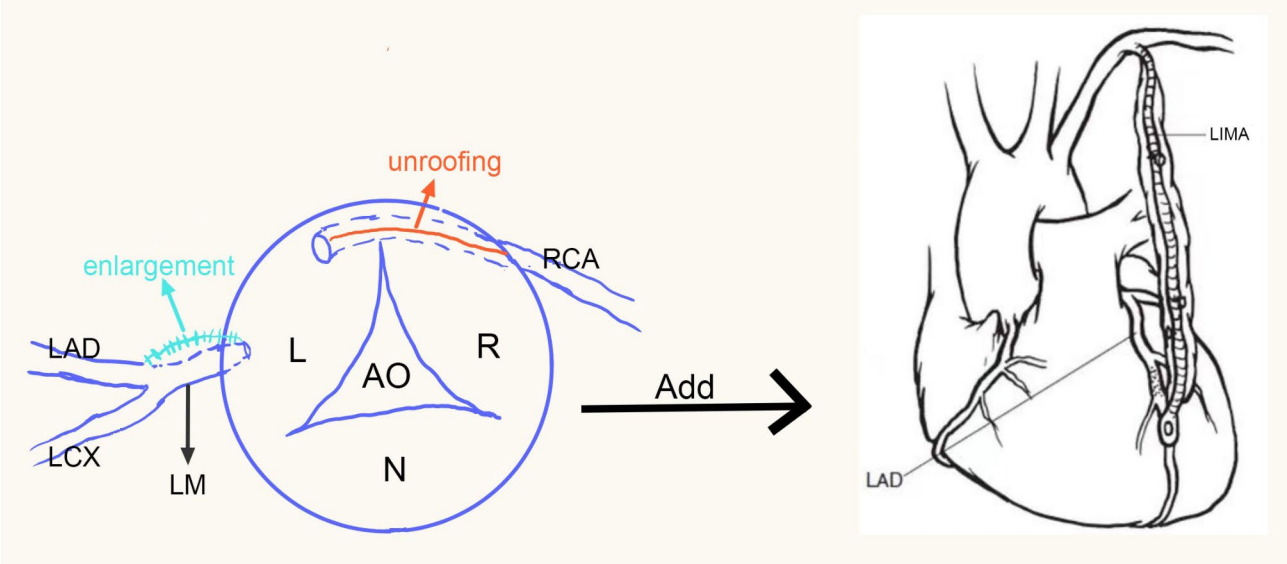

Fig. 6 Surgical management of anomalous right coronary artery and severe left main stenosis
Abbreviations: LM: left main coronary artery; LAD: left anterior descending branch; LCX: left circumflex branch; RCA: right coronary artery; LIMA: left internal mammary artery

valuable insights regarding AAOCA. In cases where suspicious Q waves, ST segment abnormalities, T wave abnormalities, or voltage abnormalities are present on the electrocardiograms of children and adolescents, it is essential for physicians to perform echocardiography and thoroughly examine the origin and course of the coronary artery whenever possible.

When coronary artery images obtained through echocardiography are of poor quality due to artifacts, diagnosis can be effectively made using CTA or aortography. These methods can directly visualize the opening location, opening pattern, intramural course, septal course, interarterial course of vessels, and other crucial details. As the gold standard for evaluating coronary artery stenosis, coronary arteriography is also essential for assessing the anomalous origin of coronary arteries. Selective coronary angiography or CTA is necessary for developing surgical plans for patients who were diagnosed.

This study has several limitations. First, due to the urgent clinical condition of the patient, bedside echocardiography was performed, which resulted in suboptimal imaging quality. Second, postoperative coronary CT angiography (CTA) follow-up was not performed, as the patient's family declined the examination due to concerns related to the patient's gender and the satisfactory postoperative recovery. Instead, follow-up was limited to echocardiography, which may have affected the comprehensive assessment of postoperative coronary artery status. These factors could have influenced the accuracy and completeness of the study findings.

Conclusion

In this study, AAOCA was identified as a consequence of fulminant myocarditis and myocardial infarction. The patient underwent timely surgery to mitigate the risk of SCD. Patients with AAOCA often experience clinical symptoms during physical activities, such as recruit training and student military exercises, leading to a higher incidence of cases in the autumn. However, clinicians should remain vigilant during all seasons when symptoms or abnormal electrocardiograms suggest the possibility of this condition, warranting further examinations to confirm or exclude the diagnosis.

Ultrasonography should be prioritized, and sonographers must be proficient in interpreting echocardiographic results to minimize the risk of misdiagnosis or missed diagnosis. When echocardiographic evaluation is inconclusive due to poor image quality, CTA or selective coronary angiography, though essential diagnostic imaging methods, should be considered secondary options, as echocardiography is typically the first choice due to its simplicity and ease of use. A comprehensive array of imaging modalities is essential to facilitate early diagnosis and to devise a more tailored, individualized treatment plan for patients.

Abbreviations

AAOCA	Anomalous aortic origin of the coronary artery
TTE	Trans thoracic echocardiography
CTA	Computed tomography angiography
CDFI	Color doppler flow imaging
LVEF	Left ventricular ejection fractions
LM	Left main coronary artery
LAD	Anterior descending branch
LCX	Left circumflex artery

RCA Right coronary artery

Author contributions
Rong-Hui Zheng: Formal Analysis, Writing – original draft. An-Biao Wang: Conceptualization, Project administration. Chun Zhao: Data curation, Writing – review & editing. Jie Zhang: Data curation, Project administration. Bo Han: Conceptualization, Writing – review & editing. Mei Zhu: Conceptualization, Data curation, Funding acquisition. All authors read and approved the final draft.

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Data availability
The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
This study was conducted with approval from the Ethics Committee of Shandong Provincial Hospital Affiliated to Shandong First Medical University (Approval Number: 2024 – 455). This study was conducted in accordance with the declaration of Helsinki. Written informed consent was obtained from the participant's legal guardians.

Consent for publication
The patient guardians signed a document of informed consent.

Competing interests
The authors declare no competing interests.

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