

Congenital absence of the right coronary artery with acute myocardial infarction: report of two cases and review of the literature

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Abstract

Congenital absence of the right coronary artery (RCA) is a rare coronary anomaly. Few cases of this condition have been reported. Congenital absence of the RCA is considered as a benign anomaly. However, in certain cases, these patients may develop life-threatening clinical complications that include acute myocardial infarction, stroke, or sudden death. We report two patients who were diagnosed with congenital absence of the RCA and presented with acute myocardial infarction. We discuss our experience in diagnosis and treatment of this disease. Congenital absence of the RCA with acute myocardial infarction is an uncommon clinical emergency. Therefore, early detection, correct diagnosis, and appropriate treatment are important.

Keywords

Coronary anomaly, right coronary artery, acute myocardial infarction, coronary angiography, percutaneous intervention, chest pain

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Background

Congenital absence of the right coronary artery (RCA) is an uncommon coronary anomaly. To date, there have been fewer than 40 cases of congenital absence of the RCA. Patients who are diagnosed with congenital absence of the RCA with acute

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myocardial infarction are even more infrequent. Individuals with congenital absence of the RCA are usually asymptomatic and this disorder is considered benign. However, a small proportion of these patients develop life-threatening clinical complications, including acute myocardial infarction, stroke, or sudden death. Therefore, making an early and accurate diagnosis of congenital absence of the RCA and treating it effectively are important.

We report here two patients who were diagnosed with congenital absence of the RCA and presented with acute myocardial infarction. We performed a PubMed search for reported cases of a single coronary artery (SCA) in the last decade. Case reports that met the diagnostic criteria for congenital absence of the RCA were summarized to assess their clinical characteristics. This study aimed to share our experience of diagnosis and treatment of this disease. We also discuss previously reported cases, especially those reported in the last decade. We assessed diagnostic and clinical characteristics of congenital absence of the RCA and its association with atherosclerosis and other congenital heart diseases.

Case presentation

Case 1

A 54-year-old woman who developed chest pain and dyspnea, without any cardiovascular risk factors and a family history of coronary heart disease, was admitted to our hospital with the diagnosis of acute myocardial infarction. A physical examination showed a body temperature of 36.5°C, pulse rate of 84 beats/minute, respiratory rate of 20 breaths/minute, and blood pressure of 149/104 mmHg (1 mmHg = 0.133 kPa). Coarse breathing sounds and moist rales were heard in both lungs. A cardiac examination was unremarkable. An electrocardiogram (ECG) showed sinus rhythm and ST-segment elevation in leads II, III, and aVF (Figure 1). Laboratory tests showed a creatine kinase-MB (CK-MB) level of 70.00 ng/mL (reference range, 0–4.3 ng/mL), myoglobin level of 759.00 ng/mL (reference range, 0–107 ng/mL), and cardiac troponin I (cTNI) level of 7.43 ng/mL (reference range, 0–0.05 ng/mL). The patient was diagnosed with acute inferior ST-elevation myocardial infarction (STEMI). After obtaining

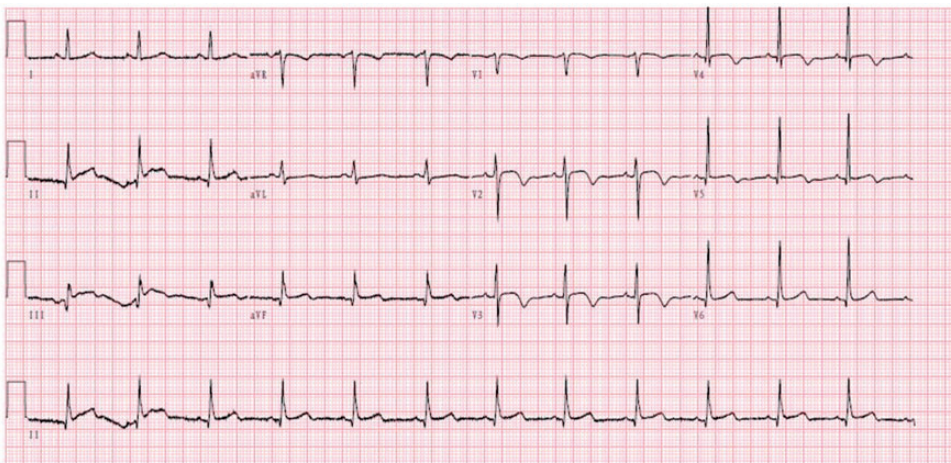


Figure 1. Electrocardiogram shows sinus rhythm and ST-segment elevation in leads II, III, and aVF.

informed consent, the patient underwent diagnostic coronary angiography (CAG). CAG showed an SCA arising from the left sinus of Valsalva, without an RCA from the right sinus of Valsalva. Additionally, there was severe stenosis, with up to 95% in the mid-distal left anterior descending (LAD) artery, and a normal left circumflex artery. The proximal LAD gave rise to the right ventricular branch and the distal LAD artery

continued to be the posterior descending artery (Figure 2).

A 3.0 × 29-mm drug-eluting stent was deployed in the mid-distal LAD artery. Percutaneous coronary intervention (PCI) was successful, without any perioperative complications. The patient was managed with antiplatelets, anticoagulants, coronary artery dilators, lipid-lowering agents, and symptomatically. One week later, the patient was discharged from the hospital.

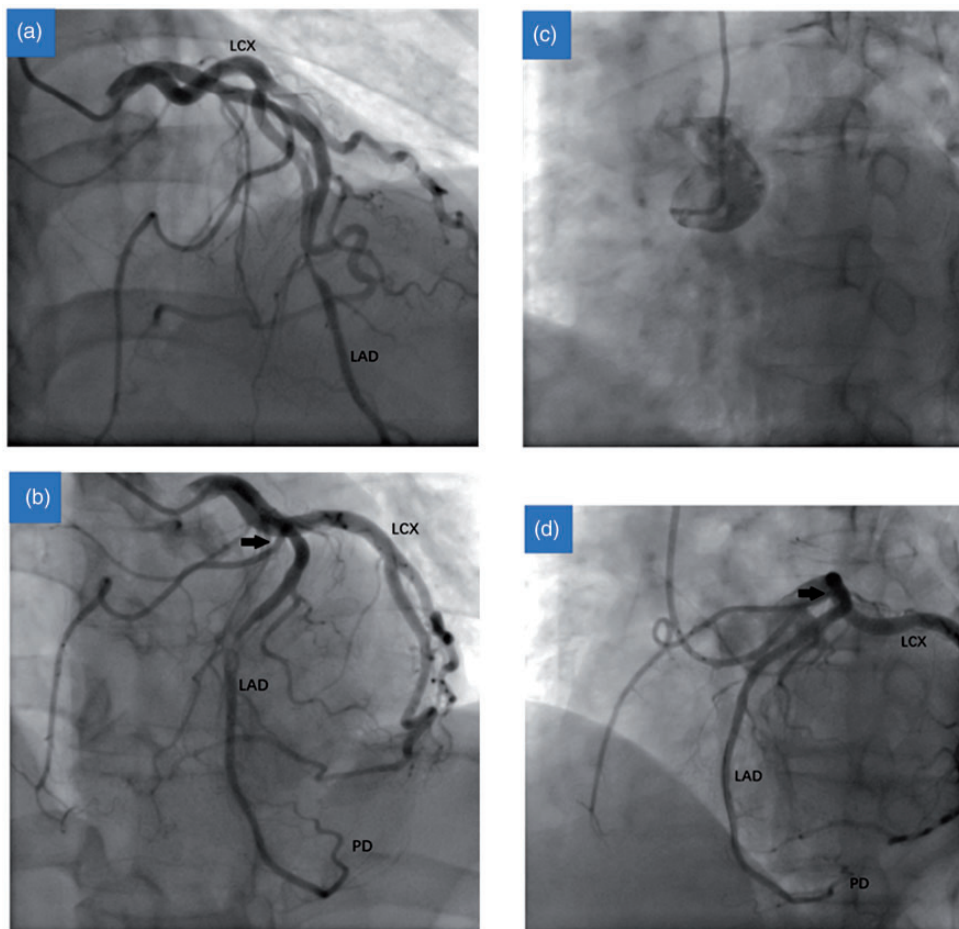


Figure 2. Severe stenosis, with up to 95% in the mid-distal LAD. (a) Right anterior oblique cranial view, (b) left anterior oblique cranial view, and (c) absent RCA in the right coronary sinus of Valsalva. (d) The proximal LAD artery gives rise to the right ventricular branch and the distal LAD continues to the PD branch. The arrow points to the right branch originating from the LAD artery. LAD, left anterior descending; RCA, right coronary artery; PD, posterior descending.

Case 2

A 67-year-old man who had a history of hypertension and diabetes for 6 years, without a family history of coronary heart disease, presented with complaints of chest pain and shortness of breath for 2 hours. A physical examination showed a body temperature of 36.0°C, pulse rate of 72 beats/minute, respiratory rate of 19 breaths/minute, blood pressure of 162/98 mmHg, and moist rales was heard in both lungs. An ECG showed sinus rhythm and ST elevation in leads V1 to V4 (Figure 3). Laboratory tests showed a CK-MB level of 25.70 ng/mL and a cTNI level of 7.16 ng/mL. The diagnosis of acute extensive anterior STEMI was made. CAG showed total occlusion of the proximal LAD artery with thrombus burden, no major stenosis of the left circumflex artery, and an invisible RCA. The distal LAD artery gave rise to the right ventricular branch and posterior descending branch (Figure 4).

After obtaining consent from the family, we proceeded to primary PCI, thrombus aspiration was performed, and a 4.0 × 32-mm drug-eluting stent was

implanted in the proximal LAD artery. The procedure was successful. The patient's vital signs were stable during the operation, without any complications. This patient was also managed with antiplatelets, anticoagulants, coronary artery dilators, lipid-lowering agents, and symptomatically. Six days later, the patient was discharged from the hospital.

Discussion

Embryonic development of the coronary arteries can be mainly divided into two stages. In the first stage, endothelial cells derived from venous sinuses form immature primary coronary vascular networks in the subepicardial and myocardial layers. In the second stage, epicardial cells undergo epithelial—mesenchymal transition to form smooth muscle cells and fibroblasts, which are remodeled to form a mature cardiac coronary artery system.^{1,2} Abnormal embryonic development leads to a coronary artery anomaly. An SCA means only one coronary artery that arises from the aortic trunk from a single coronary ostium and supplies blood to the entire heart.³ An SCA is a relatively rare coronary artery

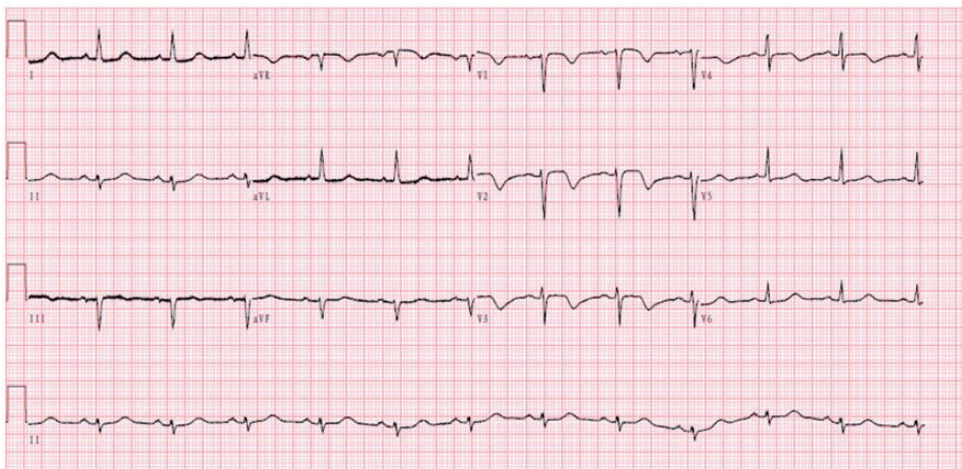


Figure 3. Electrocardiogram showed sinus rhythm and ST elevation in leads V1 to V4.

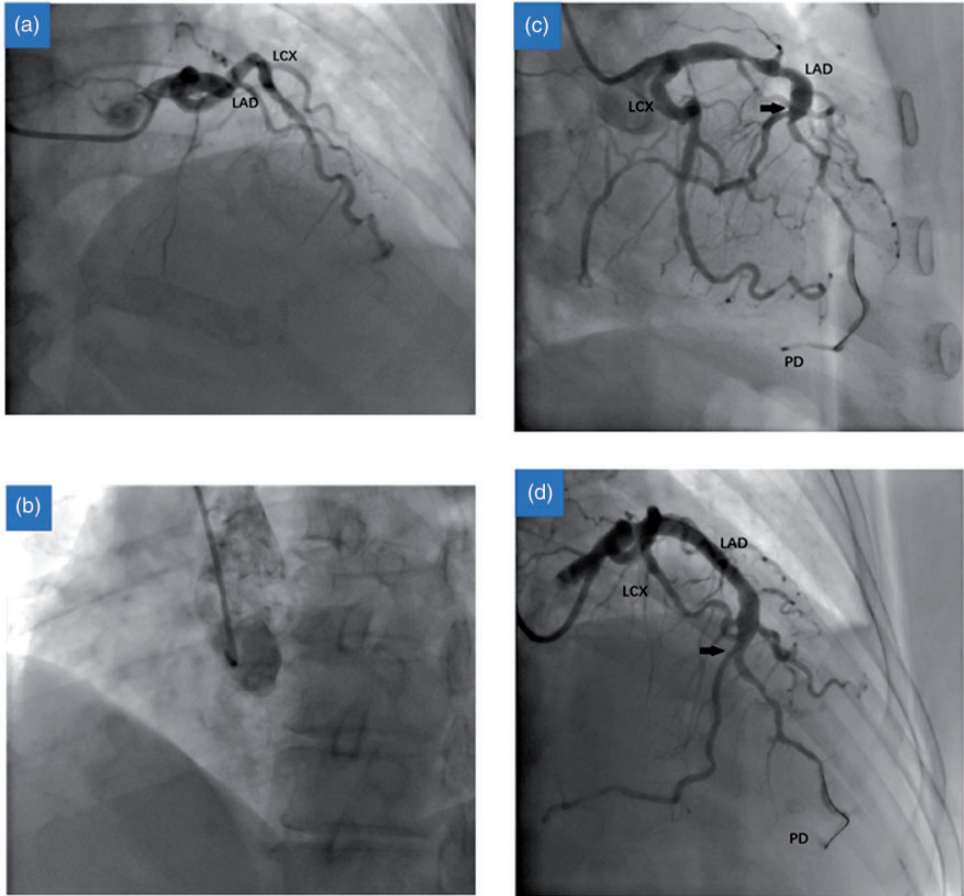


Figure 4. (a) Total occlusion of the proximal LAD artery with thrombus burden. (b) The RCA is not visualized in the aortic root. (c) The distal LAD artery gives rise to the right ventricular branch and PD branch. The arrow points to the right branch originating from the LAD artery. LAD, left anterior descending; RCA, right coronary artery, PD, posterior descending.

anomaly with a prevalence of approximately 0.014% to 0.066% of the general population.^{4,5} Congenital absence of the RCA is a subtype of SCA that was first described in 1948 by White and Edwards.⁶ The classification of congenital absence of the RCA based on coronary angiography was initially proposed by Lipton et al. in 1979⁴ and this was later modified by Yamanaka et al.⁷ The underlying mechanism of this condition may be related to a fetal developmental defect or congenital coronary occlusion. We performed a PubMed search for reported

cases of an SCA from January 2010 to December 2020. Twenty-six cases of congenital absence of the RCA have been reported in 24 articles in the last decade (Table 1).^{8–31} The mean age of patients at the time of diagnosis of congenital absence of the RCA was 53 years. Of the 26 reported cases, 14 patients were women, and 5 of them presented with acute myocardial infarction.

Based on the anatomical course of the vessel and the course of the transverse trunk, congenital absence of the RCA is

Table 1. Clinical characteristics of cases with congenital absence of the RCA that have been reported in the last decade.

Author	Year	Number of cases	Sex	Age (years)	Atherosclerosis	AMI	Associated CHD	Diagnostic tool	Lipton classification
Forste, et al. ²³	2020	1	F	45	N	N	N	CTA	L-I pattern
Liu, et al. ³⁰	2020	1	F	53	Y	Y	N	CTA + CAG	L-II pattern
Zolowska, et al. ²⁹	2018	1	M	45	Y	N	N	CAG	L-II pattern
Yan, et al. ²²	2018	1	M	63	Y	N	N	CTA + CAG	L-I pattern
Lai, et al. ²¹	2018	1	M	43	N	N	N	CTA	L-I pattern
Elbadawi, et al. ²⁰	2018	1	F	56	N	N	N	CAG	L-I pattern
Witkowska, et al. ¹⁹	2017	1	M	40	Y	Y	N	CAG + CTA	L-I pattern
Hansen, et al. ¹⁸	2017	1	M	70	N	N	N	CAG	L-I pattern
Gholoobi and Poorzand ²⁸	2017	1	M	43	Y	N	Y	CAG	L-II pattern
Jung, et al. ²⁷	2016	1	F	73	N	N	N	CAG + CTA	L-II pattern
Kim, et al. ¹⁷	2015	1	F	13	N	Y	N	CAG + CTA	L-I pattern
Gupta, et al. ¹⁶	2015	1	M	62	N	N	N	CAG + CTA	L-I pattern
Mishra, et al. ¹⁵	2014	1	M	55	N	Y	N	CAG + CTA	L-I pattern
Gul, et al. ²⁶	2013	1	M	73	Y	N	N	CAG + CTA	L-I pattern
Toyono, et al. ¹⁴	2013	1	F	2	N	N	Y	CAG	L-II pattern
Phasalkar, et al. ¹³	2013	1	F	3	N	N	Y	CAG + CTA	L-I pattern
Devidutta, et al. ¹²	2013	1	F	52	N	N	N	CAG + CTA	L-I pattern
Nasir, et al. ¹¹	2012	1	M	51	Y	Y	N	CAG + CTA	L-I pattern
Morimoto, et al. ¹⁰	2012	1	F	89	N	N	N	MDCT	L-I pattern
Zhu, et al. ²⁵	2011	3	F	77	Y	N	N	CAG + MDCT	L-I pattern
			F	72	Y	N	N	CAG + MDCT	L-I pattern
			M	77	Y	N	N	CAG	L-II pattern
Rudan, et al. ⁹	2010	1	F	57	Y	N	N	CAG	L-I pattern
Lim, et al. ²⁴	2010	1	F	84	Y	N	N	CAG	L-II pattern
Ishii, et al. ⁸	2010	1	M	6	N	N	Y	CAG	L-I pattern
Choi, et al. ³¹	2010	1	F	68	N	N	N	CAG	L-II pattern
Chen, et al. (present cases)	2020	2	F	54	N	Y	N	CAG	L-I pattern
			M	67	Y	Y	N	CAG	L-II pattern

RCA, right coronary artery; AMI, acute myocardial infarction; CHD, congenital heart disease; F, female; M, male; N, no; Y, yes; CTA, computed tomography angiography; CAG, coronary angiography; MDCT, multidetector computed tomography.

classified as an L-I pattern or L-II pattern. The L-I pattern refers to extension of the left circumflex artery or the LAD artery that dominates the area that should be supplied by the RCA. The L-II pattern represents a branch divided from the proximity of the left coronary artery that dominates the area that should be supplied by the RCA. Based on the above-mentioned classification, case 1 had the L-I pattern and case 2 had the L-II pattern. Of the 26 previously reported cases, 16 had an L-I pattern.⁸⁻²³ Therefore, the L-I pattern may be more likely to occur than the L-II pattern.

Individuals with congenital absence of the RCA present with non-specific clinical features and ECG manifestations. Most of these patients present with symptoms of myocardial ischemia, which includes angina pectoris, chest tightness, palpitations, and atypical chest pain. These symptoms are caused by arterial stenosis and obstruction. There is obstruction of blood flow due to coronary atherosclerosis. Among the 26 previously reported cases of congenital absence of the RCA, severe atherosclerotic lesions were present in 12 cases.^{9,11,22,24-30} Patients with this condition sometimes present with acute myocardial infarction. Certain mechanisms could be considered, such as the coronary steal phenomenon due to abnormal vessel or microvascular damage, and slow controlled ischemia caused by a long travel distance of coronary flow due to an abnormal coronary artery.¹⁹ Of the 26 cases of congenital absence of the RCA, 5 presented with acute myocardial infarction.^{11,15,17,19,30} Patients with a diagnosis of congenital absence of the RCA and acute myocardial infarction should undergo more aggressive management because these patients have a high risk for cardiovascular adverse events. Our two cases had congenital absence of the RCA and presented with acute myocardial infarction. Considering the risk of sudden

cardiac death, both patients received revascularization with PCI. The patients were discharged 1 week after surgery and both of them were asymptomatic without recurrence of any adverse events. Two months after discharge, the patients were followed up without obvious chest pain symptoms.

CAG is still the gold standard for diagnosing congenital absence of the RCA. CAG is extensively used in clinical diagnosis, with several advantages. However, there are some limitations of CAG. Coronary artery malformation often creates great confusion for the operator during angiography, which requires repeated non-standard angiographic projections. This technique not only causes consumption of a large amount of contrast medium, a long operation time, and a high risk of complications, but also leads to misdiagnosis and missed diagnosis.³⁰ As a non-invasive examination, computed tomography angiography (CTA) and multidetector computed tomography (MDCT) are widely performed for diagnosis, treatment, and follow-up of coronary artery disease, coronary artery malformation, and other diseases.^{5,32} CTA and MDCT can also detect anatomical variations of cardiovascular structure and are used to diagnose congenital heart diseases. Another advantage of CTA and MDCT is that the left and right coronary artery can be evaluated simultaneously and they provide excellent definition of ostial morphology and the course of the anomalous coronary, including the intraseptal course.³³ A combination of these imaging modalities yields a more robust assessment of the coronary arteries and their neighboring structures.¹⁰ These techniques also better reflect the anatomical course of the vessels and the course of the transverse trunk than other modalities. Therefore, MDCT and CTA might also be helpful in diagnosing congenital absence of the RCA.

Currently, there are no proper recommendations on treatment of congenital

absence of the RCA. Treatment options include medical therapy, interventional therapy (coronary artery revascularization, pacemaker implantation), and cardiac surgical procedures. Medical therapy includes treatment with antiplatelets, anticoagulants, coronary artery dilators, and lipid-lowering agents.³⁴ When the patient does not have coronary atherosclerosis or the degree of coronary stenosis is not severe, conservative management can be performed with drug therapy and regular follow-up. When a patient is diagnosed with congenital absence of the RCA and acute myocardial infarction, coronary artery revascularization should be performed.

Conclusion

Congenital absence of the RCA is an uncommon type of coronary anomaly that is considered as a benign disorder. However, in certain cases, patients with this condition may develop life-threatening clinical complications, such as acute myocardial infarction, stroke, or sudden death. This anomaly is more commonly found in women. Additionally, patients with the L-I pattern are more likely to develop acute myocardial infarction and are associated with congenital heart disease. Furthermore, the L-I pattern is more common than the L-II pattern. Although coronary angiography is the gold standard for diagnosing the absence of the RCA, MDCT and CTA might also be helpful. Coronary artery stenting is important for patients who are diagnosed with acute myocardial infarction, along with congenital absence of the RCA.

Author contributions

ZBC: Conceptualization and writing the original draft of the manuscript.

JHY, BKA: Writing, review, and editing of the manuscript.

XRH, JZ, YZ: Writing, review, and editing of the manuscript.

JS, YGW: Supervision and administration of the project.

All authors read and approved the manuscript.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

Ethical approval was obtained from the Ethics Committee from The First Hospital of Jilin University, China. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

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