

[CASE REPORT]

Cardiac Arrest in a 33-year-old Marathon Runner with Anomalous Right Coronary Artery Originating from the Pulmonary Artery

Koichi Ohashi¹, Ryo Itagaki², Takuto Mukaida¹, Kazuki Miyazaki³, Keiko Ohashi³, Masaaki Kawada⁴ and Daisuke Abe¹

Abstract:

A 33-year-old marathon runner presented with anomalous right coronary artery originating from the pulmonary artery after being admitted for cardiac arrest. Surgical re-implantation of the right coronary artery to the aortic root to re-establish right coronary ostial circulation was successful. The patient resumed exercise and required no further medical therapy.

Key words: ARCAPA, cardiac arrest, surgical transposition, marathon

(Intern Med 61: 673-677, 2022)

(DOI: 10.2169/internalmedicine.7612-21)

Introduction

Coronary artery anomalies occur in 1.3% of the overall population. They are usually asymptomatic and discovered incidentally during coronary arteriography or at an autopsy. Some anomalies present with symptoms or potentially serious sequelae that require surgical treatment. They are often the cause of sudden death in young athletes. Clinicians should suspect the presence of a coronary artery anomalies in young patients presenting with exertional syncope, myocardial infarction, exercise-induced arrhythmias, or cardiac arrest. Although congenital coronary anomalies are relatively rare, some entities, such as anomalous dominant right coronary artery originating from the pulmonary artery (ARCAPA), may lead to life-threatening presentations, including myocardial infarction, arrhythmia, or sudden cardiac death (1, 2).

We herein report a case of ARCAPA diagnosed in a 33-year-old man who experienced cardiac arrest while running a marathon.

Case Report

A 33-year-old man collapsed at the 20-km line of a marathon (42.195 km) due to cardiac arrest after 100 minutes of running. An advanced cardiac life support team immediately started advanced cardiopulmonary resuscitation. The initial electrocardiogram (ECG) showed ventricular fibrillation. He achieved return of spontaneous circulation after the first shock of an automated external defibrillator (AED) and was transferred, comatose, to our Emergency Department.

The patient had no heart murmur, and an ECG showed a normal sinus rhythm without ST-T changes (Fig. 1), no structural cardiovascular disease, and a normal ventricular function. Blood tests showed metabolic acidosis, renal insufficiency, and elevated levels of transaminases and lactate dehydrogenase. Electrolytes, creatinine kinase, and troponin levels were normal. The patient had no medical history of heart disease.

Emergency coronary angiography was performed, revealing an ARCAPA. There were extensive collaterals from the left anterior descending artery to the ARCAPA without any stenotic lesion (Fig. 2). A 64-slice coronary computed to-

¹Department of Cardiology, Tokyo Metropolitan Bokutoh Hospital, Japan, ²Department of Cardiovascular surgery, Tokyo Metropolitan Bokutoh Hospital, Japan, ³Tertiary Emergency Medical Center (Trauma and Critical Care), Tokyo Metropolitan Bokutoh Hospital, Japan and ⁴Division of Pediatric and Congenital Cardiovascular Surgery, Jichi Children's Medical Center Tochigi, Japan

Received: March 23, 2021; Accepted: July 13, 2021; Advance Publication by J-STAGE: August 31, 2021

Correspondence to Dr. Koichi Ohashi, wang.hao.yin@gmail.com

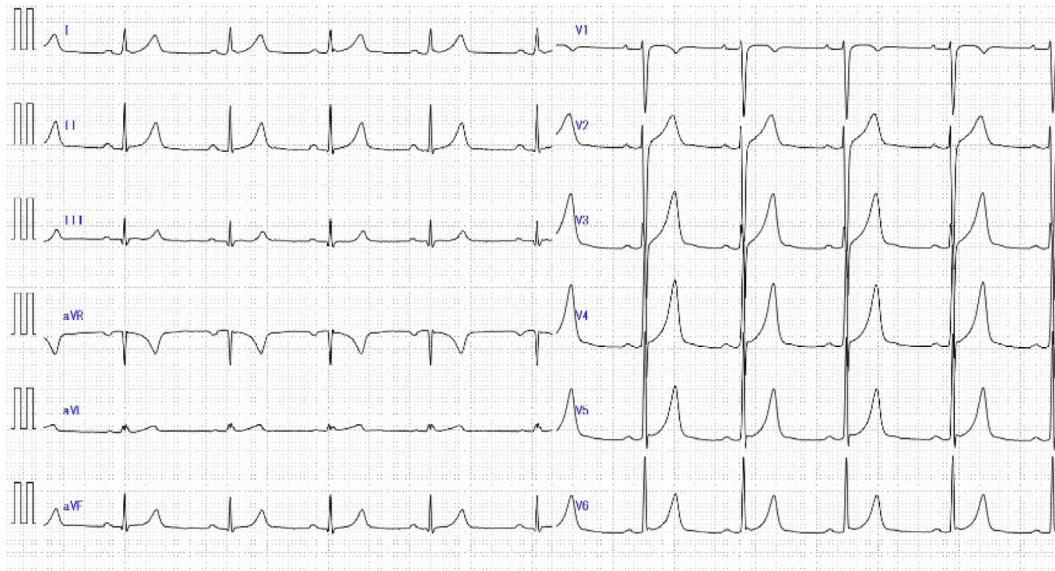


Figure 1. An electrocardiogram (ECG) showing a normal sinus rhythm without ST-T changes.

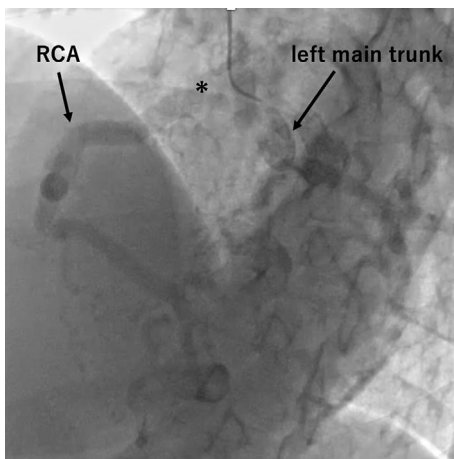


Figure 2. An aortogram of the left coronary artery in the left anterior oblique cranial view showing a delayed filling of the right coronary artery (RCA) and pulmonary artery (*) through collaterals in the late phase.

mography angiogram was performed, confirming ARCAPA (Fig. 3).

Gadolinium-enhanced cardiovascular magnetic resonance imaging revealed no significant ischemic changes. A right heart catheterization examination was also performed, which showed a normal pressure, oxygen saturation step-up from the right ventricle to pulmonary artery (PA) and increased pulmonary blood flow/systemic blood flow ratio 1.21, indicating a left to right shunt from the left coronary artery (LCA) to PA via the right coronary artery (RCA). An echocardiogram performed on day 12 showed a normal left ventricular ejection fraction without regional wall motion abnormalities.

The patient was referred for exercise testing using standard Bruce protocol on day 15. The test was stopped at minute 7 of the protocol because the patient was experiencing fatigue, had an increasing heart rate of 161 beats/minute

(87% of the maximal predicted heart rate), a maximal blood pressure of 223/68 mmHg, and a workload of 16.9 metabolic equivalents without clinical symptoms. An ECG showed a normal sinus rhythm and ST segment horizontal depression at several leads from V3 to V6. The depression of the ST segments returned to the normal position at minute 3 of the recovery phase.

On day 24, to prevent further cardiac events, the patient underwent surgical repair (Fig. 4) by direct re-implantation of the anomalous right coronary artery along with a cuff of the pulmonary artery into the aorta. Intraoperative transesophageal echocardiography revealed the ARCAPA, which was implanted into the right Valsalva sinus (Fig. 5).

The postoperative course was uneventful. An electrophysiological study (EPS) on day 40 indicated no fatal arrhythmia, and the patient was discharged without requiring an implantable cardioverter defibrillator (ICD). One year after surgery, the patient was healthy, requiring no medical therapy. An echocardiogram showed a normal biventricular function with forward flow from the aorta to the implanted RCA (Fig. 5b). He resumed running after a negative treadmill exercise tolerance test.

Discussion

Sudden cardiac arrest is the leading cause of death in athletes during exercise. In Japan, the number of cardiac arrests, especially during marathon races, has been gradually increasing. Cardiac arrest can occur not only in the elderly but in individuals across a wide age range. Previous studies reported that heart disease is the most common cause of cardiac arrest during marathon races (3). Kuroyanagi et al. reported that, in Japan, 81% of sudden death in sports was caused by heart-related diseases (4). Since 2004, the use of an AED by citizens had been permitted and has led to a rapid improvement in the survival rate during marathon

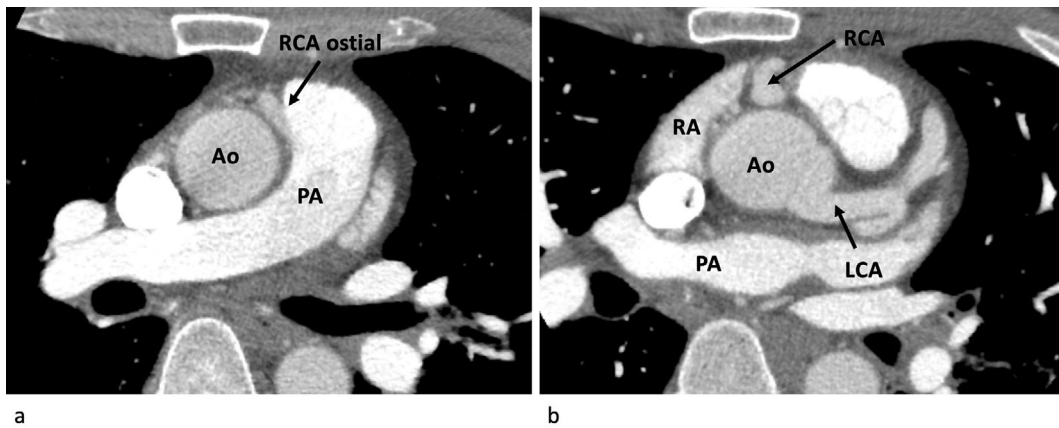


Figure 3. A 64-slice coronary computed tomography angiogram, confirming ARCAPA. Ao: aortic artery, RCA: right coronary artery, PA: pulmonary artery, LCA: left coronary artery, RA: right atrium

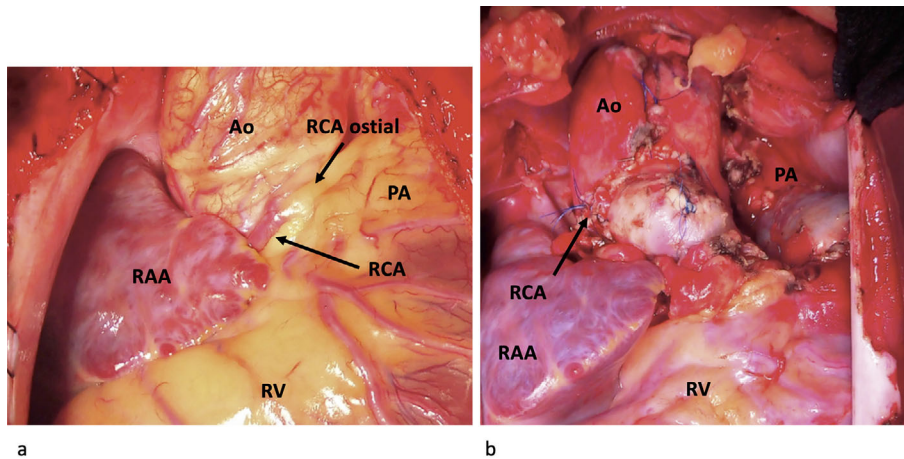


Figure 4. Surgical repair by direct implantation of the anomalous right coronary artery along with a cuff of the pulmonary artery into the aorta. Isolated RCA ostial, which was thick enough to implant directly without making a flap, from the main PA wall. Implanted directly into the aorta wall. Ao: aortic artery, RCA: right coronary artery, PA: pulmonary artery, RV: right ventricle, RAA: right atrial appendage

ances. Several cases of AED use for cardiac arrest during marathon have been reported. As shown in our case, the installation of an AED can prevent sudden cardiac death during marathons and other sporting events.

After Bregmann et al. reported in 1979, ours is only the second reported case of ARCAPA with cardiac arrest during exercise. It is occasionally diagnosed during childhood due to cardiac murmur, chest pain, or congestive heart failure and may present with subclinical myocardial ischemia and pose a risk of sudden cardiac death. However, the diagnosis of ARCAPA is often missed due to its asymptomatic nature (5). The timing and severity of symptoms depend on the type of anomaly, direction of blood flow in the anomalous vessels, and extent of collateralization. Its small perfusion area generally means it is asymptomatic, but an increase in oxygen demand may lead to exhaustion of the coronary physiologic reserve, which triggers extensive car-

diac ischemia and causes ventricular arrhythmia.

The most commonly recorded echocardiography findings for ARCAPA are the presence of inter-coronary collateral blood flow within the ventricular septum and a dilated LCA (5). Imaging using a 64-slice coronary computed tomography angiogram and coronary angiography are useful for detecting ARCAPA. Emergency coronary angiography may identify acute culprit lesions of acute coronary syndrome in patients with out-of-hospital cardiac arrest, regardless of any ST segment changes (6). We therefore performed emergency coronary angiography as soon as the patient's vital signs stabilized and found that the left main coronary artery arose from the appropriate sinus without aneurysm or stenosis. After a slight delay, the posterior descending artery and RCA were filled up in a retrograde fashion through collaterals from the left coronary artery. Cardiac catheterization showed normal pressures and little oxygen saturation step-up

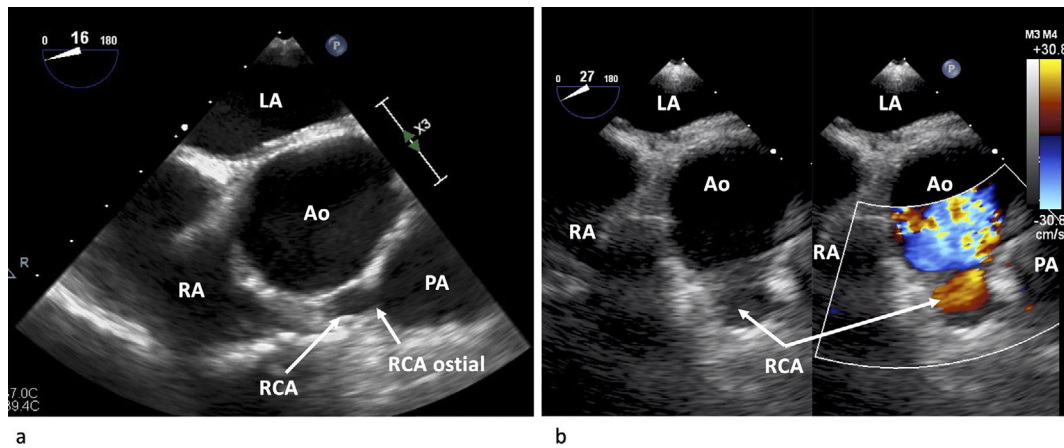


Figure 5. Intraoperative transesophageal echocardiography revealed the ARCAPA, which was implanted into the right Valsalva sinus. (a) Transesophageal echocardiography (before) (b) Transesophageal echocardiography (after). Ao: aortic artery, RCA: right coronary artery, PA: pulmonary artery, LA: left atrium, RA: right atrium

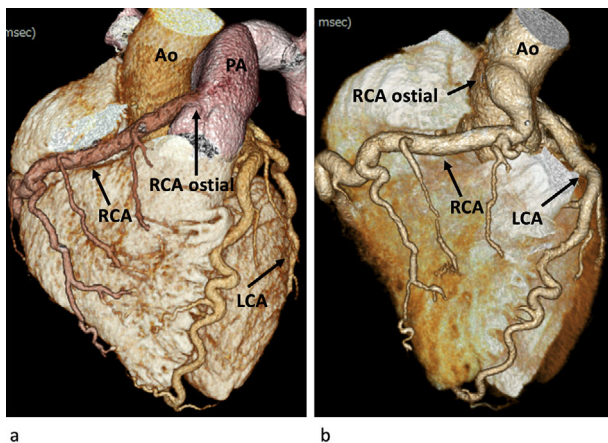


Figure 6. Direct implantation of the anomalous right coronary artery along with a cuff of the pulmonary artery into the aorta. (a) Before surgery. (b) After surgery. Ao: aortic artery, RCA: right coronary artery, PA: pulmonary artery, LCA: left coronary artery

in the pulmonary artery.

Due to the risk of myocardial ischemia or sudden death, surgical correction with the re-establishment of antegrade coronary circulation is recommended, even in asymptomatic patients with evidence of myocardial ischemia. According to a previous review, some cases did not undergo surgical operations because of no evidence of myocardial ischemia. In our case, we performed an ECG during exercise and detected evidence of cardiac ischemia. We also considered the involvement of a steal phenomenon due to the relative differences in diastolic pressure between the pulmonary and systemic arterial beds produced by adequate collateralization (7). We therefore decided to perform surgical re-implantation of the artery, despite not detecting significant ischemic changes in the study at rest. After the surgery, EPS results indicated that an ICD would not be necessary.

Even in asymptomatic patients, surgical correction should

be considered to prevent the risk of myocardial ischemia or sudden death. Especially in cases of ostial stenosis at the origin of the right coronary artery in the pulmonary artery, a relatively low degree of coronary runoff into the low-pressure pulmonary artery can be present. Our patient had no evidence of coronary ischemia at rest, probably because of excessive collateralization of the right coronary artery from the left coronary artery. However, we detected evidence of cardiac ischemia upon exercise. Patients with anomalous origin of the coronary artery, including ARCAPA, should be examined carefully using several imaging modalities to detect myocardial ischemic changes.

The most promising surgical strategy is direct implantation of the anomalous RCA along with a cuff of the pulmonary artery into the aorta (Fig. 6a, 6b, 7). This is technically feasible, since the proximal portion of RCA is long enough without significant branches, and the inclusion of a cuff of pulmonary artery permits a large anastomosis. This type of operation does not necessarily require cardiopulmonary bypass. The long-term patency of the direct arterial anastomosis is probably higher than that of a venous graft especially in a relatively young cohort (7). When anatomical issues, such as calcification of the aorta or a too-short RCA, preclude re-implantation of the RCA into the aorta, ligation of the abnormal RCA and bypass grafting should be considered as alternative therapeutic options.

Prevention of sudden death in athletes and sports enthusiasts is one of the most important topics for cardiologists. Several autopsy studies have demonstrated that patients with anomalous origin of the coronary artery have an associated increased risk for sudden cardiac death (SCD) (8). In these studies, the risk for SCD appeared highest in young individuals, particularly in patients with anomalous left coronary artery, during or following a period of strenuous exertion. However, anomalous RCA, including ARCAPA, is also not rare in patients who develop SCD during exercise. The absence of symptoms before sudden death in a significant por-

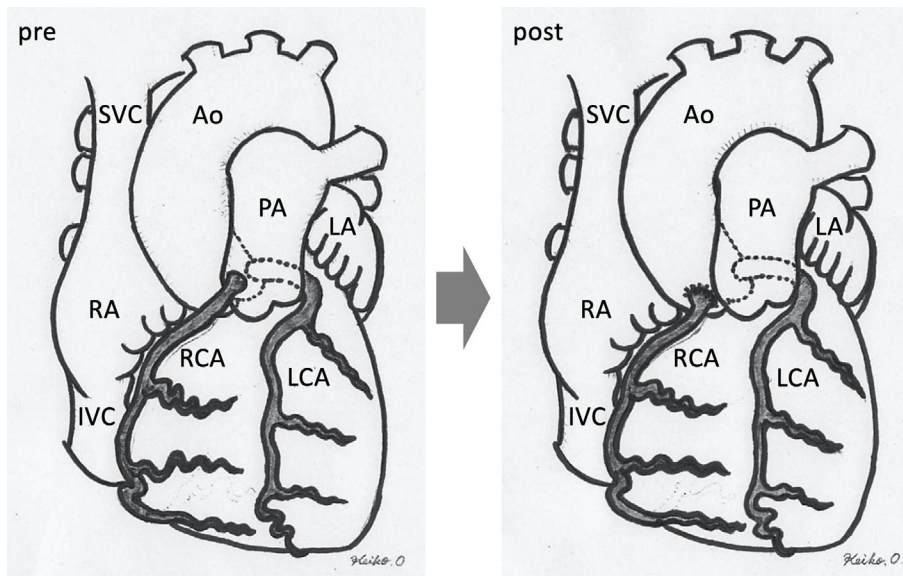


Figure 7. Ao: aortic artery, RCA: right coronary artery, PA: pulmonary artery, LCA: left coronary artery, RA: right atrium, LA: left atrium, SVC: superior vena cava, IVC: inferior vena cava

tion of patients with anomalous coronary artery has been a limitation in detecting these patients. Because of the rarity of these cases, current guidelines do not support universal preparticipation cardiac testing to screen for anomalous origin of the coronary artery in asymptomatic athletes. This remains an evolving issue, with studies and clarification needed to consider screening options. At present, the American Heart Association and American Academy of Pediatrics offer various tools for individualized risk assessments (9, 10).

We treated a 33-year-old man presenting with ARCAPA after cardiac arrest during a marathon. We successfully performed direct coronary transposition of coronary ostial circulation. Surgical transposition of coronary arteries should be considered to prevent further ischemic events and cardiac death due to ARCAPA.

The authors state that they have no Conflict of Interest (COI).

References

1. Algolini P. Coronary artery anomalies; an entity in search of an identity. *Circulation* **115**: 1296-1305, 2007.
2. Maron BJ, Thompson PD, Puffer JC, et al. Cardiovascular preparticipation screening of competitive athletes: a statement for health professionals from the Sudden Death Committee (clinical cardiology) and Congenital Cardiac Defects Committee (cardiovascular disease in the young), American Heart Association. *Circulation* **94**: 850-856, 1996.
3. Joggerst S, Monge J, Angelini P, et al. Sudden cardiac arrest at the finish line: in coronary ectopia, the cause of ischemia is from intramural course, not ostial location. *Tex Heart Inst J* **41**: 212-216, 2014.
4. Kuroyanagi K, Matsuo Y, Kojimahara M, et al. Sudden death during exercise. *J Jpn Soc Clin Sports Med* **10**: 479-489, 2002.
5. Ismee AW, Welton MG, William EH. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. *Am Heart J* **152**: 1004, 2006.
6. Tateishi K, Abe D, Iwama T, et al. Clinical value of ST-segment change after return of spontaneous cardiac arrest and emergent coronary angiography in patients with out-of-hospital cardiac arrest: diagnostic and therapeutic importance of vasospastic angina. *Eur Heart J Acute Cardiovasc Care* **7**: 405-413, 2018.
7. Haider W, Robina M, Sajid S, et al. Anomalous right coronary artery arising from the pulmonary artery. *Ann Thorac Surg* **93**: e75, 2012.
8. Michael KC, Richard RL, Ron B, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of Valsalva. *J Am Coll Cardiol* **62**: 1592-1608, 2017.
9. Maron BJ, Levine BD, Washington RL, Baggish AL, Kovacs RJ, Maron MS. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 2: Preparticipation Screening for Cardiovascular Disease in Competitive Athletes: a scientific statement from the American Heart Association and American College of Cardiology. *J Am Coll Cardiol* **66**: 2356-2361, 2015.
10. Kühn A, Kasnar-Samprec J, Schreiber C, et al. Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA). *Int J Cardiol* **139**: e27-e28, 2010.

The Internal Medicine is an Open Access journal distributed under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view the details of this license, please visit (<https://creativecommons.org/licenses/by-nc-nd/4.0/>).