A healthy man, aged 20 years, experienced sudden cardiac death caused by ventricular fibrillation while running a half marathon. Cardiopulmonary resuscitation was initiated within 2 minutes of latency and was continued for 9 minutes with return of spontaneous circulation after defibrillation. The ECG showed ST-elevations in leads II, III, and aVF (Figure 1).

A coronary artery disease could be excluded by coronary angiography. The left coronary artery had its orthotopic source on the left coronary cusp and arose anatomically regular with identification of a myocardial bridge in segment 7 (Figure 2 and Movies I and II in the online-only Data Supplement). The right coronary artery (RCA) arose atypically from the left coronary cusp. The proximal segment of the RCA was slightly narrowed with a length of 11 mm (Figure 3). The intubation of the RCA proved to be atypical. However, all of the coronary arteries showed thrombolysis in myocardial infarction 3 flow. No atherosclerotic lesions were detected (Figure 4 and Movies III through VII in the online-only Data Supplement).

Acute aortic dissection could be ruled out by transesophageal echocardiography. Furthermore, the left coronary artery was free-visible up to the bifurcation. A second small vessel was identified next to the left coronary artery in the left coronary cusp (Figure 5). Postresuscitation care phase and weaning proceeded rapidly with no neurologic dysfunctions.

Because of the treatment history of a common cold, the absence of coronary artery disease, and suggestive laboratory results (creatine kinase max, 35 pkat/L [normal value, <2.85 pkat/L], creatine kinase MB, 2.18 pkat/L [normal value, <0.42 pkat/L], high-sensitivity troponin T, 2068 ng/L [normal value, <14 ng/L], and d-dimer, 0.85 mg FEU/L [normal value, <0.5 FEU/L]), initially a myocarditis was considered as a relevant cause for the ventricular fibrillation. To test for myocarditis lesions, a cardiac magnetic resonance imaging (MRI) scan was performed. Surprisingly, a pattern of posterior wall infarction was shown by the MRI, yet there was no evidence of acute myocarditis or hemodynamic relevant muscle bridge of the left anterior descending artery with stress MRI. The perfusion sequences showed an inferior perfusion failure. Accordant to this, a late gadolinium enhancement was seen inferior in the late gadolinium enhancement sequences (Figure 6). The subsequent coronary MRI showed an interarterial course of the RCA between the pulmonary trunk and the ascending aorta. The proximal part of the RCA appeared to be (intussusceptive) intramural within the aortic root wall (Figure 7).

Surgical correction was performed with a reimplantation of the atypical originating RCA to the right side of the aortic root. The muscle bridge on the left anterior descending artery was not unroofed because of the missing perfusion failure in the left anterior descending artery territory and the clear target of fibrosis in the RCA territory. The operative correction proceeded without any difficulties.

The present case of a young athlete experiencing sudden cardiac death illustrates the importance of suspecting the presence of coronary anomalies, especially in younger patients. In this case, the RCA arose abnormally from the left coronary cusp coursing partially intramural in the aortic root and interarterial between the aorta and the pulmonary trunk. The intramural course leads to partially hypoplastic portions, which, especially during exertion, causes a further compression of these parts that lead to deterioration of the hemodynamic relevance of these arteries.1-3

Medical treatment, coronary angioplasty with stent deployment, and surgical repair (unroofing, debranching, or bypassing) are possible treatment options.1 In this case, the decision was made for surgical treatment. An off-label stent deployment would not have been feasible because of the difficult intubatable RCA, as well as the increased risk of restenosis and stent thrombosis with involvement of a large territory at risk for myocardial infarction.

The prevalence of coronary anomalies is reported with ≈5% of coronary anomalies.1 The subgroup of anomalous origination of a coronary artery from the opposite sinus is a serious anomaly with an incidence of ≈1% of all coronary anomalies. Coronary angiography and cardiac MRI should be performed in ambiguous situations. The relevance of myocardial bridges is controversially discussed. They were observed in >1% of coronary anomalies in an unselected population, which is considered to be a normal variant.2 The intramural-interarterial course of a coronary artery is an especially severe and rare disorder. The cause and the best medical treatment for this disorder remain unclear.
Disclosures

None.

References


Figure 1. Initial ECG with ST-elevations up to 0.2 mV in leads II, III, aVF, and subsequent ECG with normalized ST-elevations and T negativity in II, III, and aVF.

Figure 2. Myocardial bridge (right anterior oblique 30°). A myocardial bridge was identified above segment 7 of the left anterior descending artery.

Figure 3. Atypical origin of the right coronary artery (RCA) from the left coronary cusp (right anterior oblique 30°). Figure shows lateral compression indicating the intramural course labeled by double arrow, single arrow indicating normal diameter of the RCA, and pigtail catheter lying in the pulmonary trunk.

Figure 4. Atypical origin of the right coronary artery (RCA) from the left coronary cusp (right anterior oblique 30°). RCA is projected 3 mm superior and posterior to the left main stem. Figure shows inconspicuous left coronary artery and pigtail catheter lying in the pulmonary trunk.
Figure 5. Suspected origin of the right coronary artery (RCA) from the left coronary cusp (LCC): transesophageal echocardiography showing the atypical arising of the RCA from the LCC. The course of the RCA appears to be interarterial between the aortic root and the pulmonary trunk and partial intramural of the aorta. Single arrow indicates the RCA. LMS is labeled by a double arrow. Ao indicates aorta; AV, aortic valve; LMS, left main stem; PA, pulmonary artery; PV, pulmonary valve; and RVOT, right ventricular outflow tract. A, Midesophageal short-axis view in 30°; B, atypical midesophageal short-axis view; C, midesophageal long-axis view in 125°.

Figure 6. Late gadolinium enhancement (LGE) inferior. An LGE of 20% wall thickness was seen inferior in cardiac magnetic resonance imaging, representing a pattern of inferior wall infarction.

Figure 7. Magnetic resonance tomographic exposure of the arising right coronary artery from the left coronary cusp with interarterial course between aortic root and pulmonary trunk. LV indicates left ventricle; PA, pulmonary artery; RA, right atrium; and RV, right ventricle (RV).
Survived Sudden Cardiac Death in a Young Marathon Runner: Cardiac Ischemic Event Attributed to an Interarterial Course of the Right Coronary Artery

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Circulation. 2013;128:e104-e106
doi: 10.1161/CIRCULATIONAHA.113.001418

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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**Movie Legend:**

**Movie 1:** Muscle bridge LAD.
Coronary Angiography showing a muscle bridge in the middle LAD, best viewed within real player or windows media player.

**Movie 2:** Coronary Angiography LCA.
Inconspicuous left coronary artery. Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.

**Movie 3:** Coronary Angiography RCA.
Atypical origin of the RCA from the LCC (RAO 30[degree sign]). Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.

**Movie 4:** Coronary Angiography RCA.
Atypical origin of the RCA from the LCC. Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.

**Movie 5:** Coronary Angiography RCA.
Atypical origin of the RCA from the LCC. Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.

**Movie 6:** Coronary Angiography RCA.
Atypical origin of the RCA from the LCC. Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.
Movie 7: Coronary Angiography RCA.

Atypical origin of the RCA from the LCC. Pigtail catheter lying in the pulmonary trunc, best viewed within real player or windows media player.