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Case Reports

Repeated Syncope During Exercise as a Result of Anomalous Origin of Left Coronary Artery With Intramural Aortic Course in a Teenage Boy

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Anomalous origin of the left coronary artery from the opposite sinus of Valsalva with an intramural aortic course (L-ACAOS-IM) can cause syncope, sometimes as a prodrome of lethal events, including sudden cardiac death, in young athletes. The detailed mechanism of syncope in patients with L-ACAOS-IM is still unclear. This case report describes a 17-year-old boy who presented to the hospital because of syncope following chest pain with increasing frequency during exercise, such as playing soccer and running. In a treadmill exercise test, a decrease in blood pressure was seen (from 99/56 mm Hg to 68/38 mm Hg); chest pain and faintness accompanied by ST-segment elevation in lead aVR and ST-segment depression at other leads on electrocardiography were noted. These findings and symptoms disappeared spontaneously within a few minutes while clinicians prepared for emergency medications. Coronary computed tomography angiography (CCTA) showed that the origin of the left coronary artery (LCA) was the opposite sinus of Valsalva, and the course of the LCA was through the aortic wall toward the left coronary sinus. He was diagnosed with L-ACAOS-IM. After surgical treatment by unroofing the intramural part of the LCA and reconstructing a neo-ostium, he no longer experienced syncope during exercise. This case suggests that low cardiac output caused by myocardial ischemia, not life-threatening arrythmia, is a main mechanism of syncope in patients with L-ACAOS-IM. Consideration should be given to performing CCTA before an exercise stress test for young patients with syncope and chest pain to avoid the risk of severe myocardial ischemia. ***(Tex Heart Inst J. 2022;49(6):e217677)***

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Anomalous origin of the left coronary artery from the opposite sinus of Valsalva with an intramural aortic course (L-ACAOS-IM) is an extremely rare congenital coronary artery anomaly.[1,2] This anomaly can cause syncope and sometimes lethal events, including ventricular arrhythmias, myocardial infarction, and sudden cardiac death (SCD) in young competitive athletes.[3] Syncope is a very important sign leading to the diagnosis of L-ACAOS-IM. However, it is still unclear whether the main mechanism of low cardiac output syndrome (LCOS) resulting in syncope in patients with anomalous aortic origin of a coronary artery is life-threatening arrhythmia originating from existing ischemic scars or myocardial ischemia itself. This case report describes L-ACAOS-IM in a young male patient who experienced chest pain and presyncope accompanied by ischemic changes without ventricular arrythmia on an electrocardiogram induced by an exercise stress test.

Case Report

A 17-year-old boy presented to the hospital as a result of syncope. He had been experiencing unexplained repeated syncope during exercise, such as playing soccer and run-

ning, which was sometimes accompanied by chest pain, for 5 years, and the frequency of syncope was increasing. He had undergone various medical tests at several hospitals, but the cause of syncope had not been determined.

A 12-lead electrocardiogram (ECG) showed incomplete right bundle branch block with normal sinus rhythm (Fig. 1A). Echocardiography showed that left ventricular wall motion was normal and there was no obvious structural abnormality. He then underwent a treadmill exercise test to reproduce the situation in which syncope occurred. On the treadmill exercise test, he showed ST-segment elevation at lead aVR and ST-segment depression at other leads accompanied by a decrease in blood pressure (99/56 mm Hg to 68/38 mm Hg), chest pain, and faintness (Fig. 1B). These changes in the ECG as well as his symptoms disappeared spontaneously within a few minutes when he rested while physicians prepared for emergency medications, including catecholamines. There was no life-threatening arrhythmia such as ventricular tachycardia or ventricular fibrillation on the ECG during the treadmill exercise test. He underwent coronary computed tomography angiography (CCTA) because involvement of myocardial ischemia was suspected as the cause of syncope. The CCTA showed that the origin of the left coronary artery (LCA) was the right sinus of Valsalva, that the takeoff angle to the ascending aorta of the LCA was acute (24°) (Fig. 2A), and that the ostium of the LCA was on its lateral side. In addition, the orifice of the LCA was a slit-like shape (Fig. 2B). The results of right coronary angiography were normal (Fig. 3A), whereas the region just proximal of the LCA showed severe stenosis on left coronary angiography (Fig. 3B). Based on these findings, he was diagnosed with L-ACAOS-IM. The patient underwent an unroofing operation for resolution of myocardial ischemia caused by L-ACAOS-IM. At the time of the aortic incision, it was confirmed that both the right coronary artery and the LCA were arising from the right sinus of Valsalva. Subsequently, the intramural part of the LCA was unroofed to create a neo-orifice in the left sinus of Valsalva. In addition, because the aortic commissure between the right and left cusps was involved in the unroofing procedure, it had to be incised and then reconstructed with sutures. CCTA performed after the operation showed that the origin of the LCA was corrected to the left sinus of Valsalva (Fig. 4A) and the orifice of the LCA had become a normal shape (Fig. 4B). After the unroofing operation, the ST-segment change on a 12-lead ECG and clinical symptoms such as chest pain and syncope during a treadmill exercise test disappeared. Five years after the operation, syncope had not recurred, even with high-intensity exercise.

Discussion

In a report by Romme et al,[4] vasovagal syncope accounted for 67.7% of the causative diseases of syncope in patients younger than 40 years, whereas the proportion of patients with cardiac syncope was only 1.3%. Anomalous origin of coronary artery from the opposite sinus of Valsalva (ACAOS) is a very rare cause of syncope in young people, but it is a structural disease that is sometimes fatal and should be diagnosed and treated. Frescura et al[5] reported that SCD was the first manifestation in 50% of patients with ACAOS, and Taylor et al[6,7] reported that age younger than 30 years was a risk factor associated with an increased incidence of SCD. Therefore, the possibility of ACAOS in young patients with syncope should be considered to prevent SCD. CCTA is a noninvasive and useful diagnostic tool for detecting ACAOS, and Cheezum et al[8] reported that proximal vessel narrowing, acute takeoff, and separation from another artery of ACAOS on CCTA were important characteristics of the accompanying intramural course.

On the other hand, as mentioned above, it is not still clear whether the cause of a decrease in cerebral blood flow resulting in syncope in patients with ACAOS is LCOS associated with myocardial ischemia itself or life-threatening arrhythmia originating from an ischemic scar. In this case, the exercise stress test induced chest pain accompanied by ST-segment elevation at lead aVR and ST-segment depression at other leads on the ECG, followed by decreased blood pressure and presyncope. These clinical and examination findings were strong evidence that structural narrowing of the left main coronary trunk of the patient caused widespread myocardial ischemia and led to syncope repeatedly resulting from low cardiac output while playing competitive sports. It should also be noted that in patients who show severe myocardial ischemia during an exercise stress test, as in this patient, a nitric acid agent cannot be used as emergent treatment because of decreased blood pressure and its inefficacy for structural narrowing of the coronary artery with an intramural course. The expert consensus guidelines of ACAOS recommended that patients with unrepaired L-ACAOS-IM should be restricted from participation in all competitive sports because of the substantial high risk for SCD, even if they do not show clinical symptoms including chest pain and syncope.[9] Therefore, it is extremely dangerous to perform an exercise stress test in patients with L-ACAOS-IM who have already shown clinical symptoms. It seems reasonable to consider performing noninvasive examinations, such as CCTA, for structural evaluation before performing an exercise stress test in young patients with syncope accompanied by chest pain. In addition,

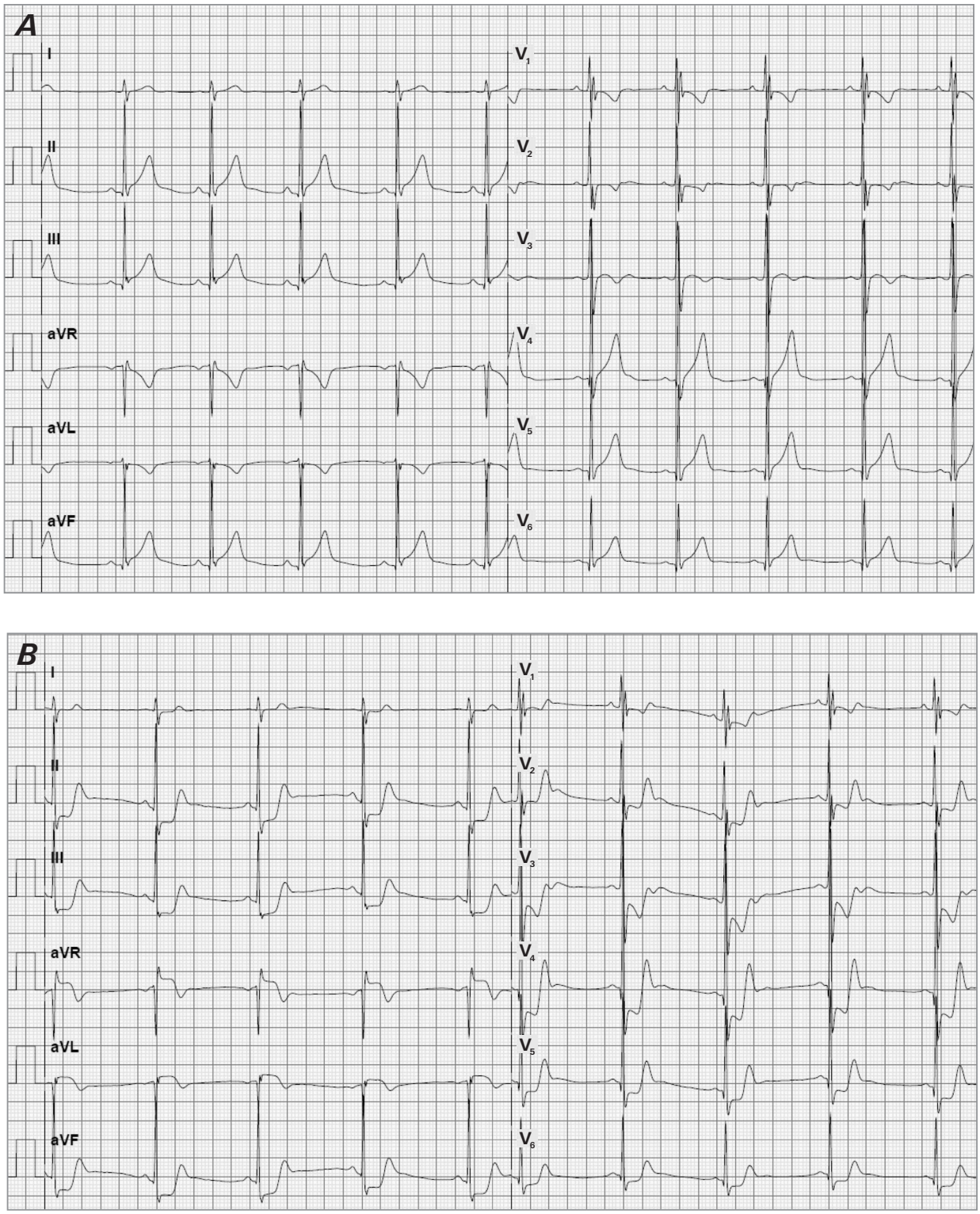

Fig. 1 *Findings of an ECG before and after a treadmill exercise stress test.* ***A****) Before exercise, there was no specific finding on the ECG other than incomplete right bundle branch block.* ***B****) On the exercise stress test, the ECG showed ST-segment elevation at lead aVR and ST-segment depression at other leads accompanied by a decrease in blood pressure (99/56 mm Hg to 68/38 mm Hg), chest pain, and faintness.*

ECG, electrocardiogram.

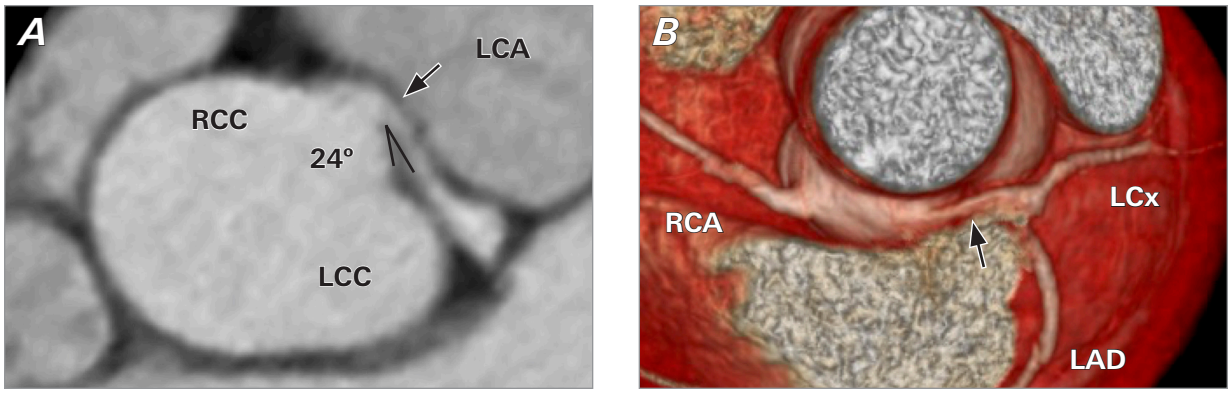

Fig. 2 *Findings of coronary computed tomography angiography in the patient before the operation.* ***A****) The origin of the LCA was the right sinus of Valsalva (arrow) and the takeoff angle was acute (24°), and* ***B****) it showed a slit-like orifice, as indicated by the arrow.*

LAD, left anterior descending artery; LCA, left coronary artery; LCC, left coronary cusp; LCx, left circumflex artery; RCA, right coronary artery; RCC, right coronary cusp.

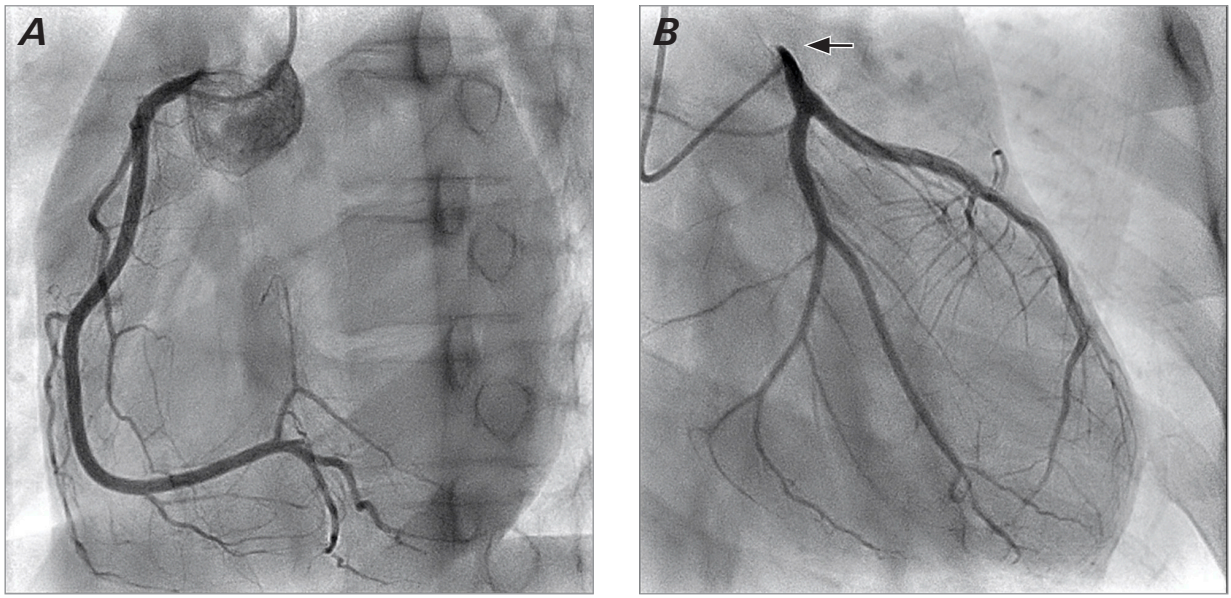

Fig. 3 *Findings of CAG in the patient.* ***A****) No abnormality was found on right CAG (left anterior oblique 50° view).* ***B****) On the other hand, the region just proximal of the left coronary artery showed severe stenosis, as indicated by the arrow (right anterior oblique 30° and caudal 30° view) on left CAG.*

CAG, coronary angiography.

it may be possible to evaluate myocardial ischemia in patients with anomalous aortic origin of a coronary artery by computed tomography–derived fractional flow reserve as an additional test for CCTA.[10] On the other hand, Angelini et al[11,12] reported the importance of intravascular ultrasound (IVUS) as a tool for evaluation of L-ACAOS-IM because IVUS can show the objective nature and the severity of intramural obstruction. Although IVUS was not performed in the present case, it should be considered as a presurgical evaluation tool for L-ACAOS-IM.

In conclusion, the present case suggested that the mechanism of syncope in patients with L-ACAOS-IM was LCOS caused by global left ventricular myocardial ischemia. CCTA must be performed before exercise stress tests in young patients with syncope during exercise accompanied by chest pain to avoid SCD during the examination.

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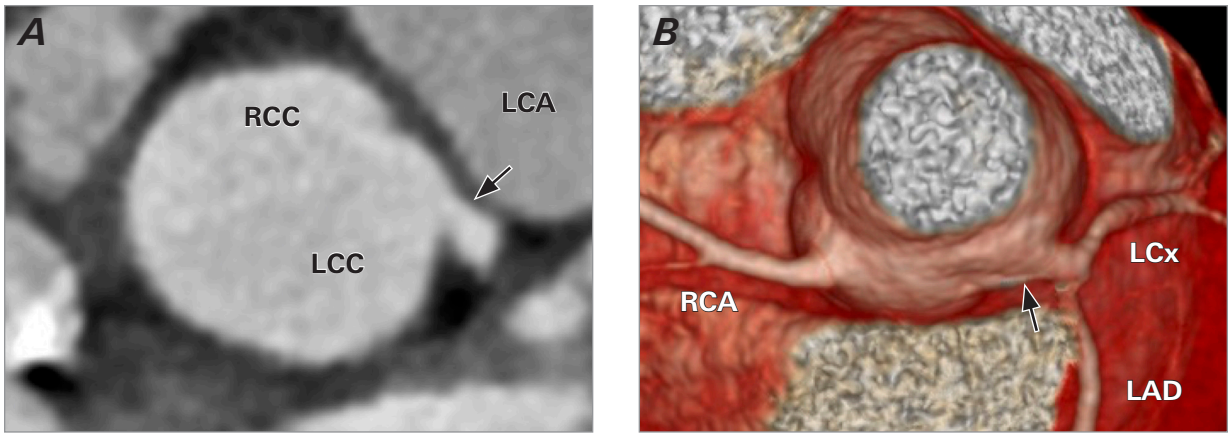

Fig. 4 *Findings of coronary computed tomography angiography in the patient after the unroofing operation.* ***A****) The origin of the LCA was corrected to the left sinus of Valsalva (arrow), and* ***B****) the orifice of the LCA had become a normal shape (arrow).*

LAD, left anterior descending artery; LCA, left coronary artery; LCC, left coronary cusp; LCx, left circumflex artery; RCA, right coronary artery; RCC, right coronary cusp.

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