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Case Report

# Congenital left main coronary artery atresia presenting as syncope and generalized seizure during exercise in a 13-year-old boy

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ABSTRACT

We report the case of a 13-year-old boy who, while running in a school gymnasium, experienced sudden syncope and seizure. CPR was started immediately, and an automated external defibrillator (AED) was attached, but shock was not induced. He was referred to our hospital for loss of consciousness and intermittent general tonic-clonic seizure. A 12-lead electrocardiogram showed normal sinus rhythm and no ST-T wave abnormalities. Echocardiography showed normal structural heart and normal cardiac function. On the second day of hospitalization, AED electrocardiogram showed complete atrioventricular (AV) block at syncope and seizure. After the patient recovered from this neurological state, we performed the treadmill exercise test, and it did not show ST-T wave abnormalities or AV block, and he did not complain of chest pain. Coronary angiography showed atresia of the left main trunk and the collateral vessel from the right coronary artery connected to the left coronary artery. He was diagnosed with congenital left main coronary artery atresia. We began administration of calcium antagonist and aspirin to prevent a coronary artery spasm and then performed a coronary artery bypass graft (CABG) to prevent sudden cardiac death. After CABG, he has had no syncope episodes at rest or during light exercise.
<**Learning objective:** In pediatric patients, syncope during strenuous exercise should mandate exclusion of cardiac events, especially coronary artery anomalies. Coronary artery anomalies that could cause sudden cardiac death sometimes show no abnormalities at rest or even during exercise stress on 12-lead electrocardiogram. It is very important to suspect cardiogenic syncope during strenuous exercise.>


## Introduction

Congenital left main coronary artery atresia (LMCAA) is an extremely rare coronary artery anomaly in which the proximal left main trunk ends blindly and blood flows from the right coronary artery to the left coronary artery through collateral vessels [1–3]. Depending on the development of collateral vessels from the right coronary artery to the left coronary artery and on metabolic demands of the heart, clinical symptoms can vary widely from infants to adults. Here, we report the case of a 13-year-old boy with LMCAA who experienced syncope and seizure during strenuous exercise.

## Case report

A 13-year-old boy was transferred to our hospital emergency room. He had suddenly fallen while running in a school gymnasium in mid-August and experienced generalized tonic-clonic seizure. At school, an AED was attached immediately, but AED analysis indicated no need for shock, and shock was not delivered. However, since his pulse was weak, his teachers started CPR. When ambulance crews arrived, his pulse had returned to normal and he was showing spontaneous breathing, but he continued to experience loss of consciousness. The patient's height was 156 cm and weight was 42 kg. Upon arrival at our hospital, his consciousness level was 300 on the Japan Coma Scale, and electrocardiogram monitoring showed normal sinus rhythm without ST-T wave abnormalities. Oxygen saturation was maintained at 92% on 10 L/min of oxygen. Heart rate was 120 bpm and blood pressure was 116/94 mmHg. He had a decerebrate rigid posture with shivering, with all limbs moving intermittently. His

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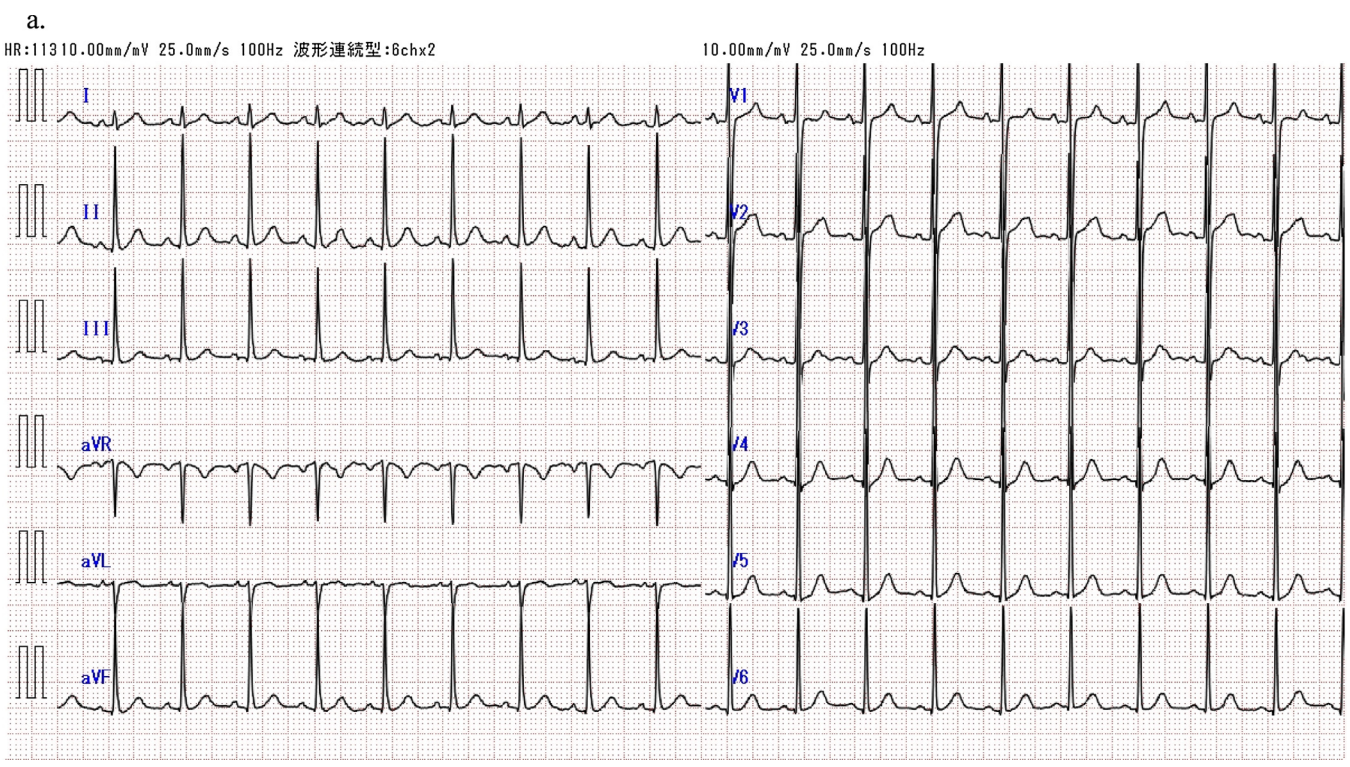


**Fig. 1.** Electrocardiogram findings. (A) 12-lead electrocardiogram on admission; (B) electrocardiogram of automated electrical defibrillator. Electrocardiogram showing complete atrioventricular block with a QRS rate of 30 bpm and a P rate of 60 bpm. Solid arrows show QRS waves; dotted arrows show P waves. (C) Electrocardiogram taken 6 min later showing sinus rhythm and escape junctional heart rate of 100 bpm.

body temperature was 37.9 °C. Chest X-ray showed a thoracic ratio of 54% and slight pulmonary congestion. Twelve-lead electrocardiogram showed normal sinus rhythm with no abnormal findings, including no abnormal Q wave, ST elevation, QT prolongation, or Brugada-type changes (Fig. 1a). Although echocardiography showed normal cardiac function and wall motion, it was difficult to locate the left main coronary artery. Right coronary artery relatively dilated. Conversely, anterior descending and circumflex branch were relatively hypoplastic. (Fig. 2a and b). Brain computed tomography did not show hemorrhagic lesions, and brain magnetic resonance imaging did not show findings suggestive of encephalitis, encephalopathy, or cerebral infarction. Because the patient continued to experience intermittent generalized tonic-clonic seizure, an anticonvulsant drug was administered in the intensive care unit, and he was intubated and received mechanical ventilation under sedation. Upon admission to the intensive care

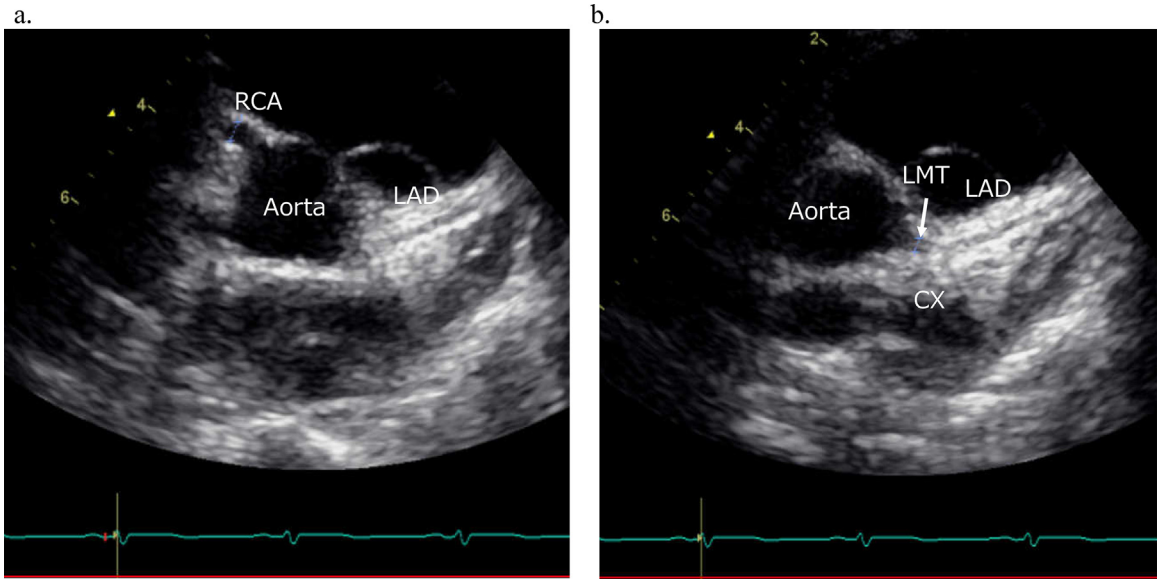


**Fig. 2.** Transthoracic echocardiogram: (A) short axis view showing the right coronary artery 3.5 mm; (B) short axis view showing the left main trunk 2.5 mm. But it was difficult to demonstrate the orifice of the left main trunk. Anterior descending 2.1 mm; circumflex branch 2.1 mm. Arrow shows the left main trunk. Abbreviations: RCA, right coronary artery; LAD, left anterior descending artery; CX, circumflex artery.

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