Congenital Ostial Atresia of the Left Anterior Descending Artery

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Figure 1. (A) Right coronary angiogram showing normal right coronary artery (RCA) with some small collateral vessels from the RCA to the left anterior descending artery (LAD). (B) Left coronary angiogram showing ostial atresia of the LAD and the collaterals from the left circumflex coronary arteries into the LAD. (C) Right coronary angiogram showing collaterals from the right ventricular branch and the distal RCA to the distal LAD. (D) Left coronary angiogram showing ostial atresia of the LAD at the bifurcation of the first diagonal branch and the first septal branch.
Left main coronary artery (LMCA) atresia is one of the least frequently observed congenital coronary anomalies. Herein we report the case of an 18-year-old man with congenital ostial atresia of the left anterior descending coronary artery (LAD).

An 18-year-old man had cardiopulmonary arrest (CPA) in the high school gymnasium after playing basketball with a high fever of 39°C due to a cold. Spontaneous circulation was restored on bystander cardiopulmonary resuscitation performed by his teacher using an automated external defibrillator (AED). On later AED analysis the CPA was found to be due to ventricular fibrillation (VF). At hospital, electrocardiography (ECG) showed elevation in the ST segment in leads V3–V6, I, II, aVL and aVF, but left ventricle (LV) wall motion disorder was absent. He received therapeutic hypothermia for 24h. On the second hospital day, he abruptly developed cardiogenic shock, and ECG showed bradycardia (45 beats/min). Echocardiogram indicated hypokinesis of the whole LV wall with an ejection fraction of 40%. Circulatory collapse with diffuse LV dysfunction might have been due to catecholamine-induced cardiomyopathy or diffuse coronary spasm secondary to VF/CPA, therefore, percutaneous cardiopulmonary support (PCPS) and intra-aortic balloon pumping (IABP) were induced and maintained. At the same time, coronary angiography showed mid-LAD atresia with collaterals (Figure 1). This was confirmed on coronary computed tomography angiogram (CTA), which also showed origin of the LMCA from the aorta, and retrograde filling of the LAD by the collaterals from the right coronary artery (RCA) and the circumflex artery (Figure 2A). There were no calcium deposits, arteriosclerosis, or continuity between the LMCA and the proximal tip of the LAD (Figure 2B, C). The proximal tip of the LAD seemed to approach the main pulmonary artery (Figure 2D, E).

He was thereafter weaned from PCPS on the fifth hospital day and from IABP on the sixth hospital day. On resting 12-lead ECG, early repolarization (J waves) was noted in leads V4–V6, II and aVF. Adenosine triphosphate-loading myocardial single-photon emission computed tomography (ATP-loading SPECT) was consistent with myocardial ischemia of the LV anteroseptal and apex walls. It was clear that the main pathology of VF was myocardial ischemia, but he and his family did not agree to open-chest coronary artery bypass grafting (CABG); therefore, given that the early repolarization ECG findings indicated VF, he underwent implantable cardioverter defibrillator (ICD) implantation. After discharge from the intensive care unit, he had no cardiac events and was discharged from hospital on the 57th hospital day. After 6 months, neurological outcome on Glasgow outcome scale was good with mild left peroneal nerve palsy. He had been

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**Figure 2.** (A–C) Coronary computed tomography angiogram (CTA) showing the left anterior descending artery (LAD) occluded at the bifurcation of the first diagonal branch and the first septal branch (A, red arrowhead), with (B, C) no evidence of ostial stamp of the LAD distal tip (red arrowhead). (D, E) The proximal tip of the LAD seemed to approach the main pulmonary artery while leaving a funicular trace (red arrow).
asymptomatic until the episode of VF, and at 2 years after the current event he had not received any appropriate ICD shocks and had no cardiac events. Genetic analysis for early repolarization syndrome was negative.

Embryologically, when the aorta and pulmonary trunks completely separate, coronary buds emerge from the semilunar sinuses and arise from the wall of the aorta. Rudimentary structures of the right, circumflex, and LAD, coronary arteries are isolated in situ in vascular networks. Congenital atresia of the LMCA is a very rare disease, and only a small number of reports have been published.3 In the present case, the absence of aortitis, Kawasaki disease history, calcium deposits, arteriosclerosis, and of continuity between the LMCA and the proximal tip of the LAD suggested a congenital phenotype. Furthermore, on CTA the LAD was occluded, with no evidence of ostial stamp of the LAD distal tip, and the proximal tip of the LAD seemed to approach the main pulmonary artery while leaving a trabecular trace. The survival rate of anomalous origin of the LAD from the pulmonary artery (ALADPA) is higher than in anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, in which the LMCA originates from the pulmonary tract, because the extent of ischemia is smaller.4 We consider that the present case is likely to be a subtype of ALADPA, although there was no definite evidence. Although ATP-loading SPECT showed poor perfusion in the anteroseptal and apex walls of the LV, the patient had no symptoms of angina before or after the CPA event. Intense exercise during a condition of high fever might have exacerbated the myocardial ischemia. CABG seems to be the treatment of choice in adult congenital atresia of the LMCA.5 At the time of writing the patient was 21 years old, and he and his family had agreed to grafting to the distal LAD using mammary artery bypass, which has superior long-term patency and great physiological adaptability to various flow patterns,6 and which had been scheduled for the near future.

Disclosures
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References