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SHORT REPORT

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Congenital atresia of the left main coronary artery: successful surgical treatment (myocardial revascularisation and mitral valve repair) in a 1-year-old boy

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Congenital atresia of the left main coronary artery (LMCA) is an extremely rare coronary anomaly [2,3]. The variety of clinical symptoms of this condition depend on the degree of the collateral circulation between the right and left coronary arteries [1]. We report on a 1-year-old boy with congenital atresia of the LMCA and mitral insufficiency (MI), who successfully underwent two separate surgical procedures at the same time.

The patient first demonstrated a heart murmur at 7 months of age and was diagnosed as having significant MI. At 10 months of age, he had poor weight gain and was admitted to our hospital for catheter and angiographic examinations. A grade 3/6 holosystolic murmur and mid diastolic rumble were heard in the mitral area. The X-ray examination revealed significant cardiomegaly (cardio-thoracic ratio 62%). The ECG showed high QRS voltage in the left precordial leads and negative P waves in the right precordial leads with no abnormal ST-T and Q waves. An echocardiogram showed a dilated left atrium, left ventricle and significant MI with anterior leaflet prolapse. Coronary arteries were seen to be normal except that there was mild dilatation (2.5 mm) of the right coronary artery (RCA). On aortic root angiography, the RCA was dilated and filled the left coronary artery (LCA) via collaterals at a delayed rate. On selective right coronary angiography, the RCA with a normal route was dilated with a retrograde filling of the proximal connected left anterior descending coronary artery (LAD) and left circumflex arteries with a normal route via collaterals (Fig. 1). The left main coronary trunk was not seen. The LCA ostium could not be selectively cannulated. On the basis of these results, the patient was diagnosed as having congenital atresia of the LMCA with significant MI. When he was 12 months of age, the boy underwent surgery because of continued poor weight gain and mild tachypnoea due to significant MI. Mitral valve repair and revascularisation of the

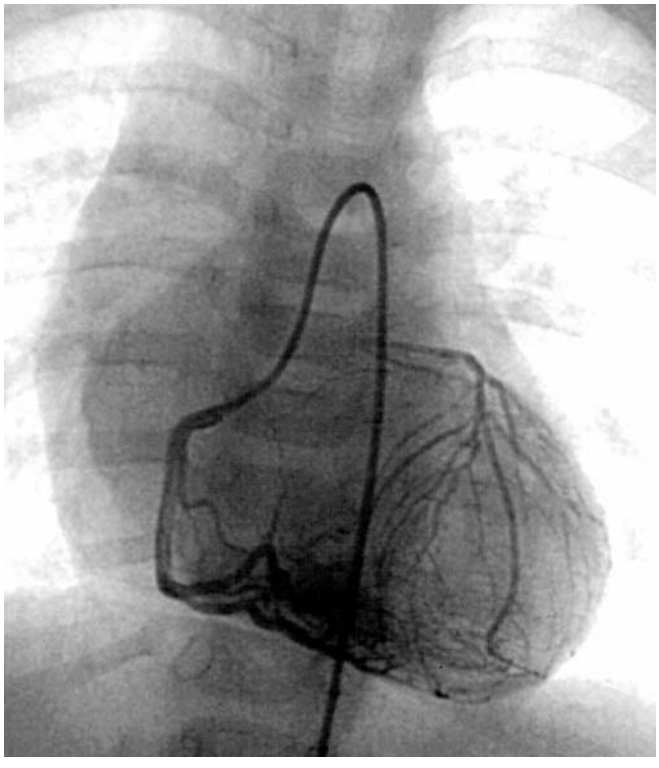

Fig. 1 Preoperative angiogram of the RCA showing a dilated RCA with retrograde filling of the left coronary system by collaterals and atresia of the left main coronary artery

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LCA with a bypass grafting from the left internal mammary artery (IMA) to segment 6 of the LAD was also performed at the same time. A follow up invasive study at 30 months after surgery showed the patency of the left IMA-LAD graft that filled all of the left coronaries, and the feeding collaterals had mostly disappeared.

Our patient presented congenital atresia of the LMCA and MI. We considered MI to be secondary to papillary muscle dysfunction. As it is difficult to diagnose this rare disease noninvasively, diagnostic confirmation was made by invasive examinations. One of the first presenting symptoms is heart murmur due to MI. Therefore, this rare condition should be considered whenever significant MI is first diagnosed, regardless of the severity of cardiac decompensation. Also, surgery should be considered as soon as diagnostic confirmation is obtained. Long-term prognosis and the quality of life of the patient with this rare condition are unclear, especially in cases with MI, even if revasculalisation and mitral valve repair are successful. Such patients must be closely monitored.

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