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Severe myocardial ischaemia after neonatal arterial switch operation

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A 24-year-old male patient presented for a routine yearly check-up. As a neonate, he underwent an arterial switch operation for d-transposition of the great arteries. He complained of shortness of breath on exertion and typical chest pain since 6 weeks. Transthoracic echocardiography was unremarkable. Treadmill examination showed no signs of ischaemia but was stopped due to severe dyspnoea. For further assessment, the patient was referred to cardiac magnetic resonance (CMR) imaging. Cine images showed normal left and right ventricular volumes and systolic function. Adenosine myocardial first-pass stress perfusion imaging revealed a severe perfusion deficit in the septal, anterior, and lateral left ventricular wall (*Panel A*). No myocardial scarring was present on late gadolinium enhancement images (*Panel B*). Three-dimensional whole heart imaging indicated a severe ostial stenosis of the reinserted left main coronary artery (LM) (*Panels C and D*). Subsequently, an invasive coronary angiography was performed and confirmed a severe ostial stenosis of the LM with a rather small circumflex artery originating from the same ostium (*Panel E*) and no abnormalities of the right coronary artery. The patient underwent surgery with a patch-extension of the insertion-site of the LM into the aorta and an elongation of the left pulmonary artery to reduce external coronary compression. Two months after the operation the patient was asymptomatic and follow-up adenosine myocardial first-pass stress perfusion was unremarkable (*Panel F*). This case illustrates the potential advantage of CMR for the non-invasive evaluation of young patients with congenital heart disease. It offers a unique comprehensive anatomical and functional assessment without ionizing radiation providing therapy planning and monitoring.

