Anomalous origin of the left coronary artery from the pulmonary artery - retrospective study of 23 years of experience.

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Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) causes severe myocardial ischemia, global left ventricular dysfunction, and varying degrees of mitral regurgitation. Surgical strategy to construct a two-coronary system for a patient with ALCAPA has evolved with time but many questions remain unanswered.

Methods: We designed a retrospective, longitudinal, descriptive study that included patients with ALCAPA. We describe perioperative details such as clinical manifestations, variations in the surgical technique and the postoperative morbidity and mortality.

Results: Eighteen children underwent surgical reconstruction of a two-coronary system because of ALCAPA between 1991 and July 2014. Two patients were lost to follow-up, one early, one in recent years. Heart failure was the principal cause of hospitalization in 14/18 of our patients. Left ventricular (LV) dysfunction was present in 14/18 and 12/14 had moderate or severe mitral regurgitation (MR). Surgery was performed with direct coronary reimplantation in 14/17 patients and intrapulmonary tunnel (Takeuchi repair) in 3/17. The most common immediate postoperative complications were: low cardiac output (10/13), pleural effusion (3/13). There was one early postoperative death (30 days) due to heart failure and ventricular arrhythmia. Mean follow-up (16 patients) was 12 years and 4 months (5 months–23 years). There was 1 late death at the age of 21 caused by VF. LV function and MR significantly improved during follow-up in all surviving patients. Global LV function by echocardiography was 66% (55-81%, Teichholz). Six patients had minor regional hypokinesis which was related to the presence of myocardial scar confirmed by perfusion scintigraphy and/or magnetic resonance imaging. Moderate MR was present in one patient, severe in none.

Conclusions: Long-term prognosis after surgical repair of ALCAPA is unclear. Standard echocardiography may underestimate LV scars and perfusion deficits. Lifelong surveillance of these patients, including magnetic resonance imaging, is recommended.