The risk of arrhythmias after surgical repair of anomalous left coronary artery arising from the pulmonary artery (ALCAPA).

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Background: Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare defect (0.1% of new cases of congenital heart disease), resulting in congestive heart failure secondary to myocardial ischemia and infarction in early infancy. Surgical strategy to construct a two-coronary system for a patient with ALCAPA has evolved with time. Despite apparent normalization of cardiac function in the first years in survivors with a patent dual coronary system, the scar persists. This may be a substrate for ventricular arrhythmias. This study evaluated the risk of arrhythmias after surgical repair of ALCAPA.

Methods: We designed a retrospective, longitudinal, descriptive study that included 18 patients with ALCAPA operated from January 1991 to July 2014. One patient was lost to follow up early and one in recent years. Surgery was performed with direct coronary reimplantation in 14/17 patients and intrapulmonary tunnel (Takeuchi repair) in 3/17.

Results: Mean follow-up was 12 years and 4 months (5 months - 23 years). There were 2 postoperative deaths (overall mortality 11.1%); one early (30 days), due to ventricular arrhythmia and heart failure and the second due to ventricular fibrillation 20 years after surgical repair. Electrocardiographic abnormalities were noted in 10/16 patients; 8 patients had PVCs or ventricular tachycardia; six patients had repolarization abnormalities in inferior and lateral leads. Three of them were on antiarrhythmics, none was considered for ICD implantation or catheter ablation. Mean preoperative left ventricular ejection fraction (LVEF) was 32% (Teichholz). At last follow-up visit, the LVEF improved in all patients to a mean of 66% (ranging from 55 to 81%), with six patients having regional hypokinesis. Fibroelastosis persists in all but one patient. The presence of scar was confirmed by myocardial perfusion scintigraphy and magnetic resonance imaging. These findings correlated with electrical instability of the myocardium.

Conclusions: Treatment of ALCAPA aims to stop process of myocardial ischemia and to restore the normal anatomy of the coronary arteries but deaths may occur due to severe myocardial injury and malignant ventricular arrhythmias. Further research is needed, aiming at early identification of patients at risk of late arrhythmic deaths.