Late coronary complications in patients with Bland-White-Garland syndrome (ALCAPA)

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Objectives: Anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) is rare and accounts for 0.5% of all congenital heart disease. Age and mode of presentation vary. Surgical treatment aims to establish a normal coronary blood supply to the left coronary artery (LCA) and includes reimplantation of the LCA into the aortic root, ligation of the origin of the LCA, bypass grafting and the Takeuchi-procedure.

Methods: Registry Search of the German Competence Network for Congenital Heart Defects with the aim to assess morbidity in this patient cohort.

Results: 107 patients with ALCAPA were identified within the registry. In 9/107 patients (8.4%) coronary artery problems occurred after a median follow-up of 4.3 years from initial repair, performed at a median age of 6.8 years (LCA reimplantation n=3, Tacheuchi-Operation n=2, bypass grafting n=1, other n =3). Coronary problems reported were myocardial infarction (n=2), coronary artery occlusion (n=1), stenosis (n=5) and aneurysmatic ectasia of the LCA (n=1). Two patients underwent percutaneous transluminal coronary angioplasty (PTCA) with stent insertion, one underwent surgical augmentation of the stenosed coronary artery and two patients received a coronary artery bypass graft. One patient with initial PTCA and stent placement received a bypass graft following in-stent stenosis. Four patients had no specific revascularization procedures. 8/9 patients were still alive at 24 years (median; 3.6-47.6 years) of follow-up. 5/8 survivors had a normal left ventricular function. In 3/8 patients contractility was impaired. Associated risk factors for coronary artery disease were not reported in all but one patient (arterial hypertension and obesity). Four patients had additional valvar procedures (mitral valve replacement n=2, aortic valve replacement n=1, Contegra graft for supravalvar pulmonary stenosis after reconstruction of the pulmonary root). One patient required ICD implantation for episodes of sustained ventricular tachycardia.

Conclusions: Although rare after surgical treatment of ALCAPA, late coronary artery problems occur and account for significant morbidity and need for subsequent intervention or surgery. Valve disease (mainly left sided) also contribute to morbidity. Thorough life-long follow-up for and awareness of these complications are warranted to improve the long-term outcome for patients with ALCAPA.