Closure of coronary artery fistula in childhood: treatment indications and long-term follow-up

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Introduction: Coronary artery fistula (CAF) is a rare congenital anomalous connection between the coronary arteries (CA) and a cardiac chamber or great vessel bypassing the myocardial capillary network. Treatment options of symptomatic CAF consist of transcatheter or surgical closure.

Methods: Retrospective analysis of all patients with CAF in our outpatient clinics database diagnosed between 1986 - 2014 concerning significance, treatment approaches and follow-up after closure, if applicable.

Results: In a cohort of more than 25000 patients, 194 (<0.01%) were diagnosed to have CAF and were enrolled in the analysis. Median age at first diagnosis was 6 months (0d–18y). In 79 patients (40.7%) CAF were isolated, whereas in 115 patients (59.3%) CAF were found in association with congenital heart disease (CHD). Origin of CAF was from the LCA in 77.2% with a left to right shunt in 86.5%. Treatment indication was given in 10 patients (5.2%): volume overload of the right sided cardiac chambers (n=6, 60%), heart insufficiency (n=2, 20%), aneurysmatic dilation of the CA with risk of ischemia (n=2, 20%). Six patients (60%) were treated by catheter interventional approach (3 Coils, 2 Amplatzer Vascular Plugs, 1 Amplatzer Duct Occluder). Follow-up (median 7 years (2-12 years) revealed one major complication with dislocation of the occluder and thrombosis of CA one week after implantation. Persistent dilated CAs were seen in 3 patients (control angiographies in 2 and echocardiography in 1 patient) and normal CA dimensions in 2 patients (echocardiography). In these five patients, no thrombosis of CA occurred and CAF remained closed. Surgical closure of CAF was performed in 4 patients (median follow-up 2 years (0-7y), during correction of CHD (n=2), and, in the era before transcatheter closure of CAF, in patients with isolated CAF (before year 2000, n=2).

Conclusions: In a pre-selected cohort of pediatric cardiology patients CAF is a very rare finding, although our numbers are probably underestimated. Intervention in childhood is rarely needed, nevertheless it is known, that small fistulas may become relevant in adulthood. Myocardial ischemia is rarely found in childhood CAF. Transcatheter closure techniques are considered the treatment of choice, especially in isolated CAF, and promote good long-term results.