Multiple coronary fistula closure in patient with pulmonary atresia and intact ventricular septum

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Introduction

Multiple coronary fistula are often diagnosed in patients with pulmonary atresia and intact ventricular septum (PA/IVS). Re-opening of the RVOT can cause significant impairment of coronary circulation what leads to myocardial infarction and consecutive heart failure in the early stage of life (RV-dependent coronary perfusion). In those patients univentricular palliation is chosen due to significant coronary fistula blood flow. We report on a patient with PA/IVS in whom closure of multiple coronary fistulas enabled to achieve biventricular circulation.

Methods

A 2.5-year-old boy with pulmonary atresia and intact ventricular septum and multiple coronary fistula and suprasystemic RV pressure was treated initially with Formula Stent implantation into the intra atrial septum and a BT-Shunt implantation after 2 unsuccessful attempts of ductus arteriosus stenting. The size of the right ventricle and tricuspid valve annulus were in the normal range so biventricular circulation seemed to be possible.

Results

At the age of 2 months the first three coronary fistulas (RV-RCA) were closed (one with ADO AS5/2 occluder and Coil 4/2, the other with ADO AS4/2 occluder and the third one with ADO AS3/2). At the age of 4 months the atretic pulmonary valve was perforated with the Baylis radiofrequency system and dilated with 4 mm balloons. After a month the next catheterization was scheduled and this time the fourth RV-RCA fistula was closed with a Covinien Microplug 5 and two RV-LCA fistulas were closed with an Amplatz plug 6-6 and an Amplatz Plug AS5-2 respectively. The RVOT was dilated with a 6 mm balloon. The last RV-LCA fistula was closed 8 months later with an Amplatzer Plug II 8/7 and the RVOT was dilated with an 8 mm balloon. One year later in echocardiography the RVSP was estimated at 40 mmHg and good ventricular function after occlusion of all coronary fistulas was observed.

Conclusion

Percutaneous closure of multiple coronary fistula is feasible and effective and may enable to avoid necessity to perform univentricular palliation in patients with PA/IVS with multiple coronary fistula. Of course this is only feasible if RV-dependent coronary circulation is absent.