Has biventricular repair become an option in patients with pulmonary atresia, intact ventricular septum and coronary to right ventricular fistulae?

Schneider M. (1), Hraska V. (2), Zartner P.(1)
Cardiology (1), Cardio-thoracic Surgery (2), German Paediatric Heart Centre, Stankt Augustin, Germany

Purpose: Patients with pulmonary atresia and intact ventricular septum (PAIVS) are usually planed for uni-ventricular palliation as, in the presence of coronary to right ventricular (RV) fistulae (CAF), decompression of the right ventricle caused severe problems for the coronary perfusion and myocardial function. In patients with PAIVS and a well developed RV a bi-ventricular approach is possible if the CAF can be closed.

Methods: In a male newborn with PAIVS, reasonably sized RV, hypoplastic pulmonary arteries, multiple aorto-pulmonary collateral arteries (MAPCA) and multiple CAF a coronary stent was placed in the arterial duct to secure pulmonary perfusion. At the age of 3 months and with a bodyweight of 4 kg several CAF from the right and left coronary artery were occluded using retrograde and antegrade approaches. All, but one small, CAF could be closed by coils and vascular plugs. In a second step the duct was surgically ligated and the RV connected to the small pulmonary artery by patch insertion. Due to poor biventricular performance two big aorto pulmonary collaterals were closed in the cathlab the other day. The RV contractility improved. Patient was discharged 4 months ago and is awaiting bi-ventricular repair with homograft implantation in RV to PA position.

Results: Despite high pressures in the RV the global myocardial function improved during follow-up and the antegrade pulmonary perfusion secures oxygen saturations above 80% with only two aorto-pulmonary collaterals and one CAF left.

Conclusions: Highly selective placement of coils and vascular plugs can effectively reduce coronary arterial run-off in patients with PAIVS after opening of a RV to PA connection. The RV can regain its contractility and biventricular repair seems to become a realistic option in this patient. The further course of this patient and the evaluation of comparable other patients have to prove the reliability of this approach. Selective and supra selective coronary angiographies are necessary for the decision whether closure of CAF would be possible, thus, making patients with diagnosis of PAIVS and CAF candidates for biventricular repair.