Successful transapical Melody valve implant in a mitral bioprosthesis in a 3-year old child with dextrocardia and Fontan palliation

Kerst G., Akintürk H., Moysich A., Müller M., Schmidt D., Recla S., Schranz D.
Pediatric Heart Centre, University Children’s Hospital, Giessen, Germany

Background:
Fontan failure with consecutive bronchitis fibroplastica or exudative enteropathy might be caused by pre- or postcapillary pulmonary vascular obstruction or by a mixed form. We describe the first-in-child transapical melody valve-in-valve (ViV) implantation in complex congenital heart disease with failing Fontan circulation.

Case report:
A 3.5-year old, 15kg weighing girl was referred for evaluation of heart transplantation. She presented with bronchitis fibroplastica (BF) based on secondary pre- (23mmHg mean pulmonary artery pressure, PAP) and post capillary (PCWP, 14mmHg) hypertension within the Fontan circulation. Based on a complex anatomy consisting of situs inversus with dextrocardia, asplenia and imbalanced atro-ventricular (AV) septal defects and transposition of the great arteries bidirectional Glenn shunt followed by a fenestrated 16mm extra-cardiac Fontan completion with the need of re-fenestration was performed. Additionally, due to a severe AV regurgitation an artificial tricuspid vale atresia together with implantation of 27mm mitral valve bioprosthesis became necessary. In a follow-up of six months the artificial valve showed a severe regurgitation, again. Considering the clinical situation, the echocardiography and magnetic resonance data a customized 22mm Melody valve was implanted through a minimally invasive right intercostal thoracotomy and an 21F sheath. The intervention was performed without complications, despite the BF she recovered well, and the transthoracic echo at her discharge home revealed a minimal residual regurgitation without any trans-valvular pressure gradient.

Conclusion:
The transapical implantation of a Melody valve allowed successful sealing following a ViV, but the anatomical dimension of the patient with body surface area of about 0.65qm was borderline.