The development of right ventricular (RV) dysfunction and failure is a significant clinical problem in patients with congenital heart disease or pulmonary arterial hypertension. The RV remodels when exposed to abnormal loading conditions and when the intrinsic myocardial dysfunction is present. After congenital heart surgery this remodelling process is further influenced by the effect of right ventriculotomy particularly involving the right ventricular outflow tract (RVOT).

The RV is a tripartite structure with an inflow part including the tricuspid valve apparatus; a trabecular part that includes pronounced trabeculations that function as an absorptive sponge, filling during diastole and releasing blood in systole; and an outflow tract that consists of a muscular infundibulum, separating the tricuspid from the pulmonary valve. How these different regions of the RV remodell in different disease conditions is poorly studied; however, how remodelling progresses in each region likely has significant influence on how the RV adjusts to different disease conditions and could provide insight into the mechanisms of RV failure. Part of this lack of knowledge is probably related to complex three-dimensional RV geometry and the difficulty of completely imaging the RV shape by two-dimensional imaging.

The paper by Leonardi et al. published in this issue is an important manuscript, as it describes and quantifies changes in the RV shape in patients after tetralogy of Fallot (TOF) repair. The authors extracted a detailed RV geometry and shape from cardiac magnetic resonance imaging (cMRI) volumetric data sets and performed an unbiased shape analysis using a principal component analysis (PCA) approach. They generated a typical RV shape, known as a template, from 49 patient-specific models. Thirty-eight individual RV shapes were then compared with the template using PCA, which quantified the major shape differences between the template and the individual RVs. As such this might seem a purely descriptive and mathematical project, but yields highly innovative information in that the authors found a correlation between the shape differences and the degree of pulmonary regurgitation (PR) as quantified by cMRI. Not surprisingly, the RVs were more dilated in the case of more severe PR. The RV dilatation primarily involved the RVOT and the RV apex, which were more significantly dilated in severe PR. The aneurysmal remodelling of the RVOT is probably related to the presence of the surgical outflow patch. Wald et al. described the presence of significant fibrosis in the region of the surgical patch extending into the RV anterior wall. The fibrotic RVOT region often is dyskinetic, which reduces the RV ejection fraction.

Dilatation of the RV apex seems to be a consistent finding also reported by two other studies using different methods. Sheehan et al. described a larger cross-sectional area and a more circular shape in the RV apical region. Additionally, they found pronounced basal bulging with tilting of the tricuspid annulus. Zhong et al. quantified RV wall curvedness and found decreased curvedness of the RV apex compared with normal RVs. They suggest that this might be the first region that dilates in TOF patients. All three studies suggest that in TOF patients important remodelling occurs in the RV apical segments, with the RV apex becoming rounder and larger. Leary et al. recently described increased apical rounding and basal bulging as the two most important characteristics of RV remodelling in patients with pulmonary arterial hypertension. Thus, these characteristics seem to be consistent traits of RV remodelling, even in different disease states.

A further question is how regional differences in RV remodelling relate to changes in the regional and global RV function. Leonardi et al. did not address this question, which is an important limitation of the paper. Theoretically increased eccentricity and decreased curvedness of the RV apex should result in increased regional radii of curvature and wall thinning. Both factors result in increased regional wall stress, which could impact regional myocardial function significantly.

The regional RV function has so far been poorly studied and is difficult to quantify. Our group used speckle tracking echocardiography and recently showed that in postoperative TOF patients, apical longitudinal deformation in the RV free wall is more significantly reduced compared with the RV basal segments. We hypothesized this could be one of the earlier changes in the RV dysfunction. Zhong et al. used cMRI-based area-strain calculations, which integrate deformation in different directions (longitudinal, radial and circumferential). In TOF patients area strain was reduced in all RV segments including the RV apex but was more significantly decreased at the basal RV level. A confounding
factor, however, is the inclusion of the basal RV with the RVOT. Van der Hulst et al. used three-dimensional echocardiography in post-operative TOF patients. They could show that the RV apex was the most significantly remodelled part of the RV; however, the local ejection fraction was preserved. A significant reduction in the regional RV apical function was also described in patients with pulmonary arterial hypertension and after atrial septal defect closure. Together, these data show that more research with pulmonary arterial hypertension and after atrial septal defect closure is required to better understand how the RV adjusts to maintain cardiac output in different disease conditions. The relationship between regional RV remodelling and the RV regional myocardial function could be essential for identifying the early RV dysfunction and failure, and its quantification should help in therapeutic decision-making and prognostication.

References

Onion-like masses in the left ventricle

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A 50-year-old male presented at the emergency department with a hypoglycaemia due to liver failure. One year earlier he was diagnosed with a severe dilated cardiomyopathy related to alcohol and cocaine abuse. Because of otherwise unexplained hypoxia, a CT-scan was performed, which confirmed the diagnosis of pulmonary embolism. By serendipity, a large mass was observed in the left ventricle (Panel 1A). Transthoracal echocardiography confirmed mobile masses in the apex and centre of the left ventricle (Panel 1B). No flow was detectable in these masses, consistent with thrombi. Despite the start of anticoagulant treatment, the patient’s condition deteriorated and he died due to liver and heart failure. Post-mortem examination (Panel 1C and D) showed an enlarged heart with multiple biventricular thrombi and one large trombus (6 x 1 x 1.5 cm) in the left ventricle. The large thrombus showed an onion-like layering, suggesting a slow formation, resulting from the decreased flow related to cardiomyopathy.

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