Watching the right ventricle in treated congenital heart disease

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The success of congenital heart surgery over the past four decades has been such that the surgical correction of a myriad of congenital heart defects is now routine. Nevertheless, as time goes by it has become increasingly hard to find a surgically treated lesion where there is a negligible risk of reoperation, arrhythmia, or other complication.

Anticipating the “natural” history of postoperative congenital heart disease has become a significant challenge for the cardiologist and cardiac surgeon. This task is complicated by the slow onset of clinically detectable problems, the relatively small number of patients involved, and heterogeneous and constantly changing treatment strategies. The last of these are a reflection of the innovative nature of the specialty as clinicians devise new strategies to circumvent problems encountered or anticipated during long-term follow-up.

Experience with the atrial switch operation for transposition of the great arteries and repair of tetralogy of Fallot has demonstrated that the right ventricle is especially vulnerable to chronic pressure or volume load.^{1,2} Recent technological advances, including tissue Doppler imaging and cardiac magnetic resonance imaging have generated new insight into the evolution and mechanisms of right ventricular (RV) pathology.^{3–5} These sensitive, non-invasive imaging techniques have the potential to detect abnormalities of ventricular structure and function early in the course of the “natural” history of the postoperative state. Therapeutic intervention can be expedited and the original operative strategy revised, with the potential to avoid problems in future patients. The downside is that these “abnormalities” may have no long-term consequence. In the worst situation, needless anxiety may be generated, unnecessary treatment provided and operative strategies changed, with adverse consequences.

In this issue of *Heart*, Grotenhuis and coworkers present a well-designed study using cardiac magnetic resonance imaging to examine the relationship of RV mass and diastolic function with pulmonary artery flow 10–20 years after the arterial switch operation (ASO) for transposition of the great arteries (see article on page 1604).⁶ Those patients studied did not have significant pulmonary artery stenosis, although their main pulmonary artery diameter was slightly less than

that of matched controls. Pulmonary flow velocities were increased in a way suggestive of increased afterload. Although RV size and systolic function were normal, RV mass was increased by an average of 50%, and diastolic function indices were reduced compared with controls. RV mass loosely correlated with pulmonary flow velocities so it is unlikely that the increase in mass was solely a marker of another unrelated issue (eg, neonatal cardiac surgery or transposition itself).

The authors hypothesise that scar tissue limits the distensibility of the proximal pulmonary arteries. They consider that this, together with minor degrees of stenosis and the abnormal anatomical relation of the right ventricle pulmonary trunk and branch pulmonary arteries, increases afterload, causing ventricular hypertrophy with associated diastolic relaxation abnormalities. They speculate that this type of suprapulmonary stenosis is more significant than a similar degree of pulmonary valve stenosis, and that delayed relaxation might be an earlier marker of diastolic dysfunction, presenting a future prognostic risk.

At this point in follow-up, major concerns after the ASO relate to coronary insufficiency,⁷ and the progression of neo-aortic regurgitation.⁸ Although pulmonary stenosis is the commonest cause of reintervention, it has not been identified otherwise as a risk factor for long-term morbidity or mortality. Usually it involves the supra valvar area (the main or branch pulmonary arteries, or both), sometimes the valve and, less commonly, the subvalvar area. Reoperation or catheter-based treatment rates are around 5–15%,^{9,10} with variance related to surgical technique,^{10,11} and probably also to diagnostic and treatment criteria.

In general, supra valvar stenosis has a greater impact on the ventricle than valvar stenosis, because it presents an increased load throughout systole, whereas the load associated with valvar stenosis is limited to early systole. Early reflection of the arterial waveform further adds to the ventricular workload. This is certainly the case on the left side of the heart, where coarctation of the aorta has a significantly greater impact on left ventricular structure and function than a similar degree of aortic valve stenosis.¹² After the ASO, energy loss related to the acute angle of the main pulmonary artery in relation to the branch pulmonary arteries may also be important.¹³

Abbreviations: ASO, arterial switch operation; RV, right ventricular

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Should the results of the study of Grotenhuis *et al* lower the threshold for intervention in those with relatively minor degrees of pulmonary artery stenosis? Should our surgical colleagues further refine their techniques to achieve a more anatomical pulmonary artery configuration? As the authors state, RV diastolic dysfunction is associated with a poor outcome, but data to date are for elderly populations with dilated cardiomyopathy, where the association of RV diastolic dysfunction with left ventricular dysfunction is a prognostic risk factor.¹⁴ We do need to look to the future—most patients who have benefited from an ASO will, we hope, survive to old age. Nevertheless, the findings of this study should stimulate further investigation rather than immediate reaction. Do these abnormalities of RV structure and function increase with time? Are they associated with a decline in indices of general wellbeing? RV dysfunction is often accompanied by increased cardiac hormones and impaired exercise capacity.^{15–16} It is reassuring that exercise capacity is normal after an ASO—at least to this point of follow-up.¹⁷ A correlation between RV mass and exercise capacity or natriuretic peptide levels would raise this issue from an observation to an abnormality with potential consequences, and might assist longitudinal assessment.

We need to remain vigilant and continue to watch the right ventricle. The “natural” history of the ASO is in its youth with the vast majority of patients aged <20 years. As with other postoperative conditions, our understanding will surely evolve as this surgical cohort passes from adolescence to adulthood.

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