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## Assessing outcomes after paediatric cardiac surgery: which test is best?

## Albert de Roos

Imaging techniques are routinely used in the evaluation and follow-up of paediatric patients with a variety of congenital heart diseases. The selection of the 'best test' in these patients will depend on a number of factors, including the medical relevance of the outcome parameters for prognosis and guiding treatment options, the accuracy and reproducibility for measuring key parameters, the perceived pathophysiological implications of those measurements, the experience and preference of clinicians as to how to use the information, the convenience for the patient to undergo a specific test, the non-invasiveness and lack of complications of the test, the costs and the availability.

Cardiac ultrasound may fulfil most of these requirements and has traditionally been the most widely applied and accepted non-invasive imaging tool in diagnosis and follow-up of patients with congenital heart disease. Is there a need for other non-invasive imaging tests and do diagnostic tools, like MRI, offer better, just the same or perhaps slightly different parameters than those we get from ultrasound? Is there also a role for cardiovascular multidetector computed tomography (MDCT) for evaluating congenital heart disease?

MRI provides a large number of measurements of cardiac function and structure that are similar to those of cardiac ultrasound. The estimation of ventricular ejection fraction, assessment of cardiac dimensions, analysis of flow profiles across valves, estimation of diastolic functional parameters and filling pressures rely on similar approaches for MRI and ultrasound. The main advantage of ultrasound over MRI is the real-time capability, easy access and wide acceptance in routine practice. The main advantage of MRI over ultrasound is its three-dimensional representation, providing anatomical and functional data with very high image quality, thereby allowing direct measurements of key parameters that may be more difficult to obtain by ultrasound. The high dimensional accuracy of MRI measurements may impact the relevance of these outcomes for clinical decision-making directly. However, broad acceptance and application of MRI measures of cardiovascular function is still evolving, because some measures are not widely used yet (eg, ejection fraction corrected for pulmonary regurgitation in patients with Fallot's tetralogy) and others are emerging as new ways to assess cardiovascular disease (eg, assessment of vascular stiffness in the aorta and pulmonary artery).

MDCT shares with MRI the threeof the dimensional representation anatomy and large field of view. When using retrospective gating techniques, MDCT also allows evaluation of ventricular function, although at the expense of higher radiation exposure than when just performing anatomical imaging during breath-holding or using prospective cardiac gating.<sup>1</sup> The main advantages of MDCT for evaluating congenital heart disease are the exquisite high spatial resolution allowing visualisation of small structures, the large field of view for evaluating extracardiac vasculature, the visualisation of the bronchial tree and lung parenchyma. From a practical point of view, MDCT is feasible in small children with no or short-term sedation, is easier and faster in ventilated and very sick patients who need life support and close supervision. However, MDCT lacks most of the functional options, like flow dynamics, available with MRI and ultrasound technology. The main limitation of MDCT is the need for radiation exposure, which is highly undesirable in small children and for repeated follow-up studies.

Cardiac MRI has been available for over 25 years with some major improvements in technical capabilities in more recent years. The technique has now been introduced and accepted for evaluation of paediatric congenital heart disease in many centres, particularly in the follow-up of repaired congenital defects. What are the major developments that have led to more widespread acceptance of this technology? Although MRI techniques may be technically demanding, the uniqueness and accuracy of the diagnostic information, the lack of radiation exposure, the wide field of view and the versatility of the MRI sequences have greatly contributed to the acceptance of the technology for application in both acquired and congenital cardiovascular disease.

As a case in point, in this issue of the journal, Puranik et al<sup>2</sup> illustrate the versatility of MRI techniques to assess functional outcomes after the Ross procedure (see page 304). State-of-the-art MRI techniques were used to assess a number of outcome parameters in one comprehensive imaging protocol, including autograft and homograft stenosis and regurgitation, vascular dimensions, function of both ventricles as well as a direct measure of myocardial scar tissue. The study showed that minor dysfunction of the autograft and homograft is very common after the Ross procedure, that myocardial scar is found in one-third of the patients, but that ventricular function and exercise capacity are still preserved despite these functional and structural alterations.

This study underlines the use of MRI to monitor subclinical alterations in cardiovascular function for both the right and left ventricle and its associated grafts in one comprehensive imaging protocol. Previous MRI studies have shown that mild degrees of homograft stenosis may lead to right ventricular hypertrophy and diastolic dysfunction and that autograft dilatation may lead to impaired left ventricular function in patients who have undergone the Ross procedure.<sup>3 4</sup> The long-term implications of these observations have to be awaited in this particular patient population.

In general, the concept of comprehensive MRI evaluation of biventricular function and ventriculoarterial coupling has great potential in both congenital and acquired cardiovascular disease. MRI is well suited to assess the dimensions, distensibility and stiffness (as estimated by pulse wave velocity) of the large vessels in conjunction with evaluation of ventricular mass, systolic as well as diastolic function. MRI assessment of the aortic pulse wave velocity in defined segments of the aorta may reflect intrinsic aortic wall disease. MRI assessment of

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aortic stiffness is an emerging new MRI application for assessing vascular function, which is independent of knowledge of the central arterial pressure or geometrical assumptions about the estimated path length. Increased aortic stiffness may affect left ventricular function and may have prognostic significance. Ventriculoarterial interaction has been demonstrated by MRI techniques-for example, in patients with non-stenotic bicuspid aortic valve disease, in whom MRI-based estimates of aortic pulse wave velocity have shown reduced aortic elasticity and dilatation leading to concomitant aortic regurgitation and left ventricular hypertrophy.<sup>5</sup> Interestingly, a recent MRI study showed abnormal aortic stiffness in patients with hypertrophic cardiomyopathy, particularly in the presence of myocardial scar tissue.<sup>6</sup>

Furthermore, many MRI studies have shown the added value of evaluating right heart function in conjunction with flow across the pulmonary artery in patients with a variety of repaired congenital heart defects. As a case in point, many MRI studies have shown that right and left ventricular dysfunction and myocardial scar predict adverse outcomes in patients after repair for tetralogy of Fallot.7 8 Assessment of the function of the right ventricle may be challenging by ultrasound, particularly in patients with distorted anatomy and concomitant pulmonary regurgitation, as frequently occurs in patients with tetralogy of Fallot. MRI has been shown to be a reliable tool for assessing pulmonary regurgitation, right ventricular systolic and diastolic function, ventricular mass and ventricular interaction after Fallot repair.9 10 These outcome measures are now widely used to monitor cardiac function after Fallot repair over time and to select the best timing for reintervention.<sup>11</sup> MRI has a unique potential allowing correction of the right ventricular ejection fraction for pulmonary regurgitation, and shunts to effectively demonstrate right ventricular output and clinical outcome.<sup>12</sup>

MRI protocols are currently performed mostly under resting conditions. Stress MRI may be performed using pharmacological or physical exercise to further improve the diagnostic capabilities of cardiac MRI.<sup>13 14</sup> It is conceivable that ventricular response to stress, flow alterations under stress, myocardial rest-stress perfusion imaging, and response of aortic pulse wave velocity to stress may provide further insight in the pathophysiology of congenital heart disease. MRI techniques are providing many options for evaluating traditional and new emerging outcomes in congenital heart disease. Further studies are needed to show whether these options will translate into better risk stratification and, ultimately, better patient care.

Finally, MRI-guided cardiopulmonary interventions may increasingly become adopted to guide device and stent placement, to monitor stent position and to assess the haemodynamic effect on flow and vascular resistance.<sup>15</sup> <sup>16</sup> MRI techniques may ultimately be used for diagnosis and treatment of congenital heart disease, thereby improving physiological evaluation of outcomes and avoiding the radiation risks associated with x-ray technology. When these options are all realised MRI may qualify as the 'best test' for the management and treatment of patients with congenital heart disease.

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