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Case Report

Cardiovascular Magnetic Resonance Evaluation of Patients with Transposition of the Great Arteries Following Atrial Switch Surgical Correction

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We present a method of imaging patients with surgically corrected transposition of the great arteries using cardiac magnetic resonance imaging and we describe the findings from a typical case. Cardiac magnetic resonance is superior to other imaging methods for patients with complex congenital heart diseases, both for establishing the initial diagnosis and for subsequent follow-up after surgical therapy.

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The most common anomaly of ventriculoarterial relations in congenital heart disease is transposition of the great arteries (TGA),[1] characterised by misplacement of the aorta and the pulmonary artery in relation to the ventricular septum. Thus, in patients with TGA each great vessel arises entirely from the morphologically wrong ventricle; the aorta from the right ventricle (RV) and the pulmonary artery from the left ventricle (LV). This results in systemic (poorly oxygenated) venous blood being recirculated to the systemic circulation and in pulmonary (highly oxygenated) venous blood being directed back to the pulmonary circulation.

If left uncorrected, TGA is a lethal malformation. Medical and surgical interventions within the last four decades have changed the prognosis of the disease. The definitive treatment is surgical and there are two main surgical approaches that have succeeded in allowing patients with TGA to reach adulthood.

- The atrial switch procedure (Senning or Mustard) aims at the physiological correction of the malformation, by redirecting the blood flow at the atrial level. Accordingly, surgical baffles direct the pulmonary venous blood to the tricuspid valve, right ventricle and aorta, and systemic venous blood to the mitral valve, left ventricle and pulmonary artery.[2,3] In this way the RV becomes the systemic ventricle and the LV becomes the pulmonary ventricle.
- The arterial switch procedure (Jatene) aims at the anatomical correction of the malformation. The aorta and the pulmonary artery are dissected and reconnected to the appropriate ventricle, restoring the proper anatomical and physiological sequence of the cardiac structures.[4]

Patients with surgically corrected TGA require close follow-up. Particularly for those who have undergone atrial switch operations, evaluation of the ventricular and baffle structure and function may be challenging as the patients grow into adulthood. Cardiovascular magnetic resonance imaging (CMR) overcomes many of the limitations of conventional imaging modalities (e.g. echocardiography and angiography) and has become the preferred method for serial follow-up of these patients. In this report we present the current rationale and approach for the imaging of such a patient with surgically corrected TGA.

Case presentation

An 18-year old man with D-TGA corrected with a Senning procedure in infancy underwent CMR evaluation of cardiac anatomy and function.

The study was performed with a 1.5 Tesla Philips Intera CV MR scanner (Philips Medical Systems, Best, The Netherlands) equipped with advanced cardiac software and a 5-element synergy coil. After acquiring localising images, 2- and 4-chamber views of the heart were obtained using a steady-state free precession (SSFP) breath-hold sequence. These images were used to plan the acquisition of a series of contiguous short axis cine images from the base (valves) to the apex of the heart, in order to quantify left and right ventricular function. Cine SSFP images in contiguous coronal and oblique planes were also used for evaluation of the systemic venous baffles and transverse SSFP images were obtained for assessment of the pulmonary venous system. Black-blood fast spin echo images in coronal and transverse planes were also obtained to assess the structure of the heart and great vessels. Phase-contrast (PC) sequences were used to quantify flow in cross-sectional areas of the aorta and pulmonary artery. Magnetic resonance angiography (MRA) using Gd-DTPA at a dose of 0.2 mmol/kg was performed to visualise the intracardiac communications and extracardiac vasculature. Finally, delayed inversion-recovery images were obtained along the 2-chamber, 4-chamber and short-axis orientations to evaluate the possibility of surgery-related scars in the ventricular myocardium. Image analysis was performed off-line on a workstation with dedicated cardiac analysis software (EasyVision 5, Philips Medical Systems, Best, The Netherlands).

Morphologically the RV was dilated, heavily trabeculated and hypertrophied and the LV was smaller in size and wall thickness (Figure 1). The RV mass (69 g, 35 g/m^2) was increased, while the LV mass (80 g, 40 g/m^2) was decreased compared to reference values,

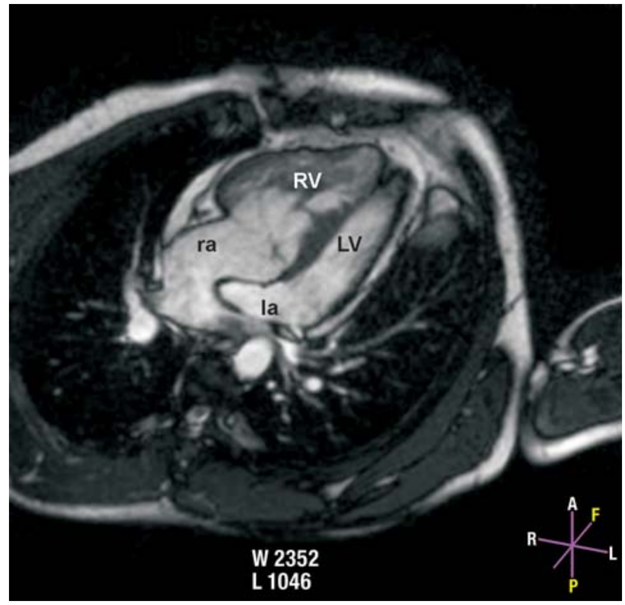

Figure 1. Four chamber bright blood image demonstrating the pulmonary vein to right atrium (ra) and systemic venous to left atrium (la) baffles. The heavily trabeculated right ventricle (RV) is dilated and hypertrophic, while the left ventricle (LV) is smaller in size.

as would be expected given the fact that the RV serves as the systemic ventricle in this patient. Both ventricles had preserved systolic function, with an RV ejection fraction of 55% and an LV ejection fraction of 60%. The interventricular septum was flattened and bowed towards the LV during systole. The stroke volumes in the pulmonary and systemic circulations were equal, suggesting that there was no left-to-right or right-to-left shunt. By combining volumetric and PC data, it was estimated that there was moderate tricuspid regurgitation (calculated tricuspid regurgitant fraction of 25%). The semilunar valves were competent without significant regurgitation.

Evaluation of the baffles demonstrated normally functioning surgical pathways. Black-blood and cine imaging of the systemic venous baffle revealed a patent pathway directing the blood of the superior and inferior venae cavae (SVC and IVC) to the mitral valve and left ventricle (Figures 1-3). In the cine images there were no signal voids, suggesting the absence of turbulent blood flow and therefore implying that there was no significant baffle stenosis or leak. Similarly, transverse anatomical and functional images of the pulmonary venous chamber revealed no anatomical or functional stenoses. The MRA corroborated these findings for both systemic and pulmonary venous baffles (Figure 4). Delayed imaging showed no hyperenhancement, sug-

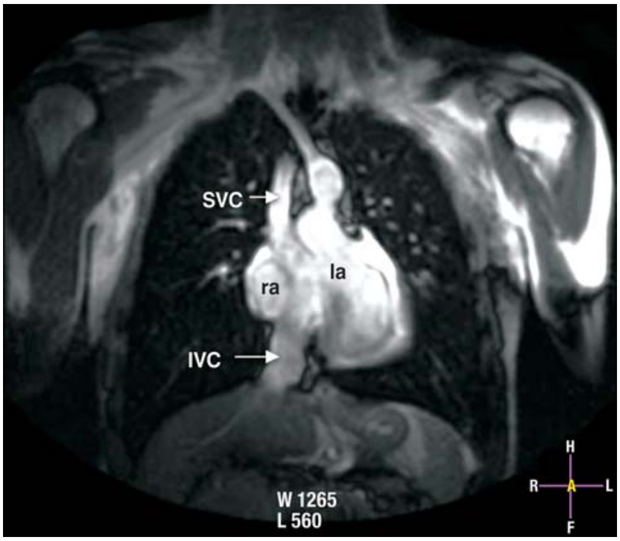

Figure 2. Coronal bright blood image demonstrating the superior and inferior vena cava (SVC and IVC, respectively) surgically constructed communication to the left atrium (la). ra: right atrium.

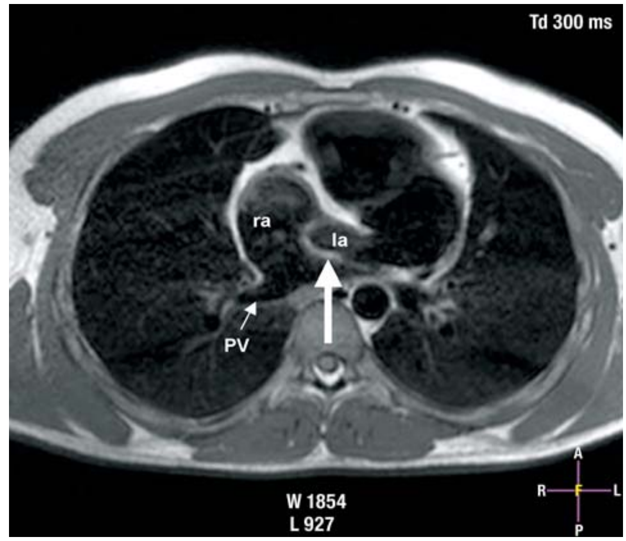

Figure 3. Black blood transverse image demonstrating the pulmonary vein (pv) to right atrium (ra) communication and the systemic venous to left atrium (la) baffle (arrow).

gesting the absence of scar or fibrosis in either of the ventricles.

Discussion

It has been almost four decades since the introduction of the Senning and Mustard procedures for the surgical correction of the transposition of the great arteries.[2,3] A significant number of patients who have undergone these procedures have reached adulthood and now present for follow-up. Their postoperative evaluation requires a thorough evaluation of the right ventricular function and possible tricuspid valve regurgitation, left ventricular outflow tract obstruction, arrhythmias, baffle leaks and systemic and pulmonary venous pathway obstruction.[5]

Among the anatomical complications of the Senning (and Mustard) procedure, systemic or pulmonary venous pathway obstruction is the most common. This complication may cause significant morbidity and it is the most difficult to assess accurately, as the patients become older, with a conventional non-invasive modality. In several studies it has been shown that CMR provides excellent visualisation of both extracardiac venous structures as well as intracardiac baffles, and can detect obstruction with good sensitivity and excellent specificity.[7-10] Compared to transthoracic echocardiography, CMR offers superb spatial resolution, is inherently a 3-D tomographic modality that allows imaging and reconstruction of the venous pathways in any orientation, and is not limited by body mass or poor acoustic penetration.[11] Although cardiac catheterisation and angiography can provide highly accurate anatomic and functional information, the invasive nature of the procedure and the exposure to ionising radiation are significant disadvantages in children and young adults who require frequent evaluation.

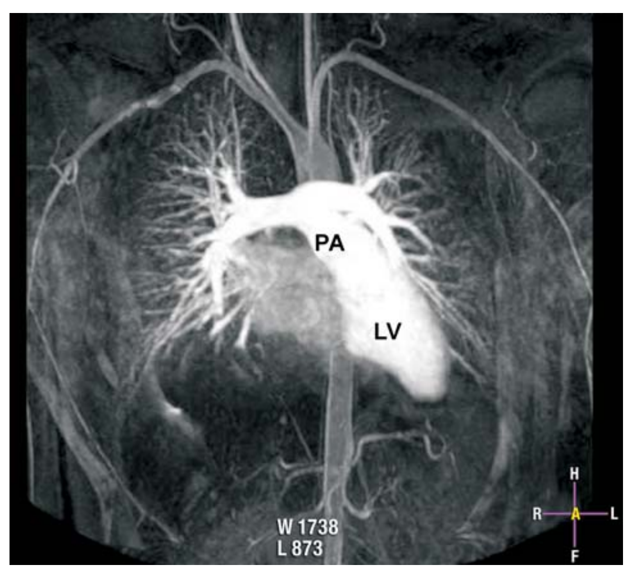

Figure 4. Maximal intensity projection from contrast-enhanced pulmonary angiography. The pulmonary artery (PA) originates from the left ventricle (LV), and there are no peripheral pulmonary artery stenoses.

Both gradient and spin echo sequences (white blood and black blood images, respectively) can be used to obtain information about the systemic and pulmonary venous patency and are complementary to each other. Spin echo imaging gives great anatomic detail even in the presence of foreign material in the systemic venous system, such as stents; cine MRI imaging, on the other hand, can give information about dynamic obstruction and can help visualise residual baffle leaks.

RV dilatation and failure are other common problems that occur later in life, as the RV fails to overcome the systemic arterial pressure. The tricuspid valve also frequently becomes incompetent, a finding that may sometimes be independent of RV dysfunction. Both RV failure and tricuspid insufficiency are important determinants of late morbidity and mortality, and their early detection and serial follow-up are essential in deciding the need for intervention to prevent further deterioration. CMR compares favourably and is superior to echocardiography, radionuclide angiography and conventional angiography for the assessment of the stroke volume and ejection fraction of the RV.[12-15] Minor early changes in ventricular size and function are more readily detectable, given the reproducibility of CMR, and may lead to an early appropriate medical intervention.

Along with the evaluation of the systolic function, CMR can also provide information regarding the diastolic function of the systemic ventricle in patients with atrial inversion. PC mapping of diastolic tricuspid flow can be reproducibly quantified and provides an assessment of RV diastolic function comparable to that obtained by echocardiography.[16] The same technique can also quantify blood flow through valves and vessels and give valuable information concerning outflow tract stenosis and regurgitatant fractions that can be followed over time and aid in management decisions. By quantifying the pulmonary and systemic blood flow, PC CMR can accurately assess residual left to right shunts, thus obviating the need for catheterisation.[17]

Contrast-enhanced CMR can be useful both for angiographic imaging of the extracardiac vasculature and for the assessment of scarring and fibrosis of the often seriously hypertrophied systemic right ventricle. Data from single-photon emission computed tomography suggest that myocardial perfusion defects could be a sensitive predictor of systemic ventricular impairment.[18] CMR may be of value in this context.

Our patient's diagnostic assessment was planned and performed taking in consideration all the above mentioned capabilities of CMR. The images that we obtained answered all the clinical questions regarding the patient's anatomy and function and will serve as the baseline data for subsequent evaluations. CMR is an ideal diagnostic modality for the evaluation of patients who have undergone an atrial switch procedure for TGA, be it a Mustard or a Senning. CMR is noninvasive, can provide detailed anatomic and functional assessment, can be repeated readily and reliably, and can be helpful in timely management decision making in patients with surgically corrected TGA.

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