CHAPTER 3
Right ventricular imaging: cardiac magnetic resonance

3.1
Review: cardiac magnetic resonance imaging in postoperative congenital heart disease patients

Journal of Magnetic Resonance Imaging: Invited

A.E. van der Hulst
A.A.W. Roest
J.J.M. Westenberg
L.J.M. Kroft
A. de Roos
ABSTRACT

The survival of patients with congenital heart disease (CHD) has greatly improved over the last decades. Nevertheless, lifelong follow-up is required in postoperative CHD patients, and noninvasive imaging plays an important role during follow-up. Cardiac magnetic resonance (CMR) imaging enables comprehensive imaging of cardiac function and anatomy, and helps to detect patients who need re-intervention and to predict clinical outcome. Postoperative CHD patients who are frequently referred for CMR evaluation include those with coarctation of the aorta, tetralogy of Fallot, transposition of the great arteries and single ventricle patients after the Fontan procedure. This article reviews the current clinical role of CMR in these various subgroups of postoperative CHD patients. Furthermore, an overview of novel CMR applications and their clinical value in CHD patients is provided.

INTRODUCTION

Advances in diagnostic, surgical and peri-operative knowledge have greatly improved the survival of patients with congenital heart disease (CHD). As a result, the population with CHD has doubled over the last decades. In addition, the age distribution of the CHD patients is changing with an increasing number of patients aging into adulthood. However, surgical intervention may not lead to complete correction of CHD and re-interventions are frequently needed, especially in patients with complex CHD. Therefore, extensive life-long follow-up is required in CHD patients to monitor ventricular, valvar and electrophysiological function and to detect any cardiovascular or non-cardiovascular sequelae.

Postoperative CHD patients, MRI can provide valuable information on cardiac anatomy, ventricular function, valvar function and on the presence and location of myocardial scar tissue. Within the population of postoperative CHD patients, the diagnostic subgroups most frequently referred for CMR evaluation include those with coarctation of the aorta, tetralogy of Fallot (ToF), transposition of the great arteries and single ventricle patients after the Fontan procedure. This article provides an overview of the role of CMR during follow-up of postoperative CHD patients. The various CMR acquisition schemes and their applications in postoperative CHD patients are reviewed, with specific attention to the most frequently referred subgroups of CHD patients. Finally, we provide a concise overview of novel CMR applications and their clinical value in postoperative CHD patients.

TECHNICAL ASPECTS

Several technical aspects and acquisition schemes apply to CMR imaging. First, specific technical adaptations are needed with regard to cardiac and respiratory motion during image acquisition. In addition, a range of CMR-specific pulse sequences are used to image the various aspects of the heart that are of use in clinical practice. During CMR assessment, motion synchronization is needed to avoid respiratory and cardiac motion artifacts. To compensate for respiratory motion, various strategies (signal averaging, breath
Bright-blood gradient echo cine imaging

Bright blood GRE imaging yields T1 weighted images, with blood appearing white, hence the name “bright blood”. In current clinical practice, bright-blood GRE image acquisition is essentially obtained by the “balanced steady state free precession” (balanced SSFP) technique. Balanced SSFP is a fast gradient echo technique providing images with high spatial and temporal resolution. (13) Contrast in balanced SSFP images is generated by the differences in T1/T2 ratio in the imaged tissues. Balanced SSFP imaging yields a high signal from blood or fat (which have a short T1 time), hence appearing white in the reconstructed images, whereas other tissues, such as myocardium, will appear gray. Like BBSE, balanced SSFP allows imaging of cardiac anatomy. Moreover, balanced SSFP is specifically useful for the assessment of ventricular function(14,15). With whole-heart multi-slice balanced SSFP cine images, ventricular volumes (end-diastolic volume, end-systolic volume, stroke volume), ejection fraction and ventricular mass can be reliably assessed (Figure 2). (14,15) Since multiple contiguous parallel slices covering the entire ventricle are obtained, geometrical assumptions of ventricular shape are avoided. This is particularly important for the assessment of reliable ventricular volumes and ejection fraction in CHD patients, as these patients may have an abnormal ventricular shape.(14) The analysis of left ventricular (LV) volumes and function is preferably performed in balanced SSFP cine images acquired in the short axis, but may also be derived from axial images. In contrast, the RV has a more complex ventricular shape and coarse endocardial trabeculations, which complicates the identification of the endocardial border in the short axis during image analysis. Therefore, CMR assessment of volume and function of the RV may be more reliable when imaged in the axial plane (Figure 2). (16) Moreover, imaging in the axial plane provides additional valuable information regarding spatial relationships between the heart and its surrounding structures, as well as diagnostic information of the surrounding structures (e.g. pulmonary artery stenosis, conduit obstruction, pleura effusion). Finally, with bright-blood GRE cine acquisition, the presence of turbulent flow may be identified. Turbulent flow causes local signal loss during GRE acquisition, resulting in a black ‘flow void’ in the image.

Figure 1. Example of black blood spin echo image

Repaired tetralogy of Fallot. BBSE images in a 36-year old male. Axial images with view from below showing the spatial relationships between the heart, large vessels, and airways. Right aortic arch (a) and right descending aorta (b-f). Normal diameter of left pulmonary artery (b), but relatively small caliber right pulmonary artery (c), as complication after Waterston shunt in the past. Dilated right ventricular outflow tract (d) and dilated right ventricle with some right ventricular hypertrophy (e,f). Note that this is accompanied with slight ventral displacement of the left thoracic chest wall (c-e). The left ventricle was also enlarged (f). Patient had moderate-to-severe pulmonary regurgitation of 45%.
Accordingly, valvar regurgitation, stenosis, or turbulent flow over a ventricular septum defect may be identified with GRE cine imaging. However, the balanced SSFP technique is less sensitive for the identification of turbulent flow as compared to the conventional GRE techniques. Furthermore, to obtain quantitative flow measures, phase-encoded imaging is required.

**Phase-encoded flow imaging**

In contrast with BBSE and bright-blood GRE imaging, which rely on signal intensity for image reconstruction, flow imaging encodes the phase of moving spins in the structures of interest linearly to the velocity of their movement. Intravascular protons that flow along a magnetic gradient obtain phase-shifts that are proportional to their flow velocity. Therefore, after the application of a magnetic gradient along the flow direction, the flow velocity of the blood can be derived by analyzing the phase-shifts at a plane of interest.\(^{(17)}\)

Phase-encoded flow imaging is used to measure flow volumes, regurgitation fraction and peak flow velocities. Phase-encoded flow imaging can be performed in any arbitrary direction, perpendicular to an acquisition plane (i.e. through-plane encoding), or in a combination of directions (i.e., 2D in-plane encoding or 3D encoding).

Through-plane phase-encoded flow imaging enables the assessment of flow volumes across cardiac valves and vessels. For this purpose, the imaging plane is positioned perpendicular to the flow at the level of the valve or vessel of interest. Data acquisition with retrospective ECG-gating results in a time series of reconstructed phase-encoded images covering the full cardiac cycle. From these images, flow velocity curves are reconstructed from which flow volumes and regurgitation volumes can be derived (Figure 3). In CHD patients, through-plane flow imaging can furthermore be used to quantify shunt volumes, measuring the ratio between pulmonary flow volume (Qp) and aortic flow volume (Qs)\(^{(18)}\).

In the absence of a shunt, the aortic flow volume should equal the pulmonary flow volume, and the Qp/Qs ratio is equal to 1. In general, a Qp/Qs ratio over 1.5 represents a large left-to-right shunt that indicates the need for (re)intervention. Similarly, the relative flow volume to each lung can be measured to assess the hemodynamic significance of branch pulmonary artery stenosis.\(^{(19,20)}\)

**3D gadolinium-enhanced angiography (3D-MRA)**

3D-MRA provides three-dimensional images of the pulmonary and aortic vessels. 3D-MRA images are obtained by a T1-weighted sequence applied shortly after intravenous administration of a gadolinium-based contrast agent. The intravascular gadolinium-chelate shortens the T1 time of the surrounding tissue (blood) resulting in increased signal intensity, delineating bright blood against virtually no background tissue signal (Figure 4). 3D-MRA provides images with high resolution and is suitable for characterization of complex intravascular anatomy of the aorta and pulmonary vessels. In CHD patients, 3D-MRA is of great value to detect vascular anomalies, such as major aorta-to-pulmonary collateral arteries (MAPCAs).\(^{(21)}\) Furthermore, during long-term follow-up of postoperative CHD patients, 3D-MRA is an important imaging tool for the evaluation of (re)stenosis, aneurysms, or for the evaluation of surgical shunts.\(^{(22,23)}\)
Late gadolinium-enhanced imaging

Following 3D-MRA, late gadolinium-enhanced imaging may be performed. Images are acquired approximately ten minutes after intravenous gadolinium-chelate contrast agent administration. Late gadolinium-enhanced imaging enables the detection and quantification of myocardial fibrosis or scar tissue. The imaging sequence takes advantage of the T1 shortening effect of gadolinium and of the differences in 'wash-out' kinetics of the gadolinium-chelate between viable and non-viable myocardium. To obtain the late gadolinium-enhanced images, an inversion-recovery, T1 weighted GRE image is acquired. Myocardial fibrotic regions or scar tissue have a delayed wash-out of the contrast agent and appear bright; i.e. "late enhancement", whereas contrast agent in normal and/or viable tissue has been washed out (no enhancement). (24) Although limited data are available at present in CHD patients, late gadolinium-enhanced myocardial viability imaging may be of use in various postoperative CHD patients to quantify scar tissue. (25, 26)

CMR IN POSTOPERATIVE CHD PATIENTS

The subgroups of postoperative CHD patients most frequently referred for CMR evaluation include those with coarctation of the aorta, ToF patients, patients after correction of transposition of the great arteries and single ventricle patients after the Fontan procedure (6,7) The various subgroups require specific CMR imaging protocols based on postoperative anatomy and the specific clinical problems that may be encountered during follow-up.

Coarctation of the aorta

Coarctation of the aorta is characterized by stenosis of the aortic lumen. The stenotic region is typically located just distal to the left subclavian artery at the insertion of the ductus arteriosus (Figure 5). Coarctation severity may vary from a mild focal stenosis (presenting in young adults with hypertension) to a severe obstruction with an additional hypoplastic aortic arch (presenting in infants with severe systemic hypo-perfusion at the time of the physiologic closure of the ductus arteriosus). Coarctation of the aorta is associated with cardiac and vascular anomalies, of which a bicuspid aortic valve is the most common anomaly (75%). (27) Even in the absence of symptoms, relief of a significant hemodynamic coarctation (as indicated by a pressure gradient over the coarctation >20 mmHg, as assessed by catheterization) is indicated due to a high incidence of complications (including congestive heart failure, aortic rupture and infective endocarditis) if left untreated. (28) Relief of the obstruction may be performed surgically by resection of the stenotic region followed by end-to-end anastomosis or, less frequently, by the use of prosthetic material or a subclavian flap. (27, 29) Alternatively, in older patients, an endovascular approach may be used with transcatheter stenting or balloon angioplasty. (27, 30, 31) Common problems in postoperative coarctation patients include aneurysm formation at the surgical site, recoarctation with hypertension and subsequent LV dysfunction and collateral artery formation. (30, 31) During long term follow-up of postoperative coarctation patients, CMR imaging of the aortic arch should be routinely performed to detect aneurysm formation or recoarctation. (5, 32) Aneurysm formation at the region of coarctation correction is observed in up to 30% of postoperative patients, with the highest incidence after the use of prosthetic material. (33) To appreciate aortic aneurysms or recoarctation, anatomical BBSE images may be obtained by planning a stack of slices in the oblique sagittal plane through the aortic arch. However, when the coarctation...
tissue is thin or membranous, it may not be projected on BBSE images due to partial volume effects. Alternatively, bright-blood GRE cine imaging may yield a turbulent flow void at the site of obstruction. In case of re-coarctation, quantification of the hemodynamic severity is necessary to investigate the need for intervention. Although cardiac catheterization is currently the reference standard to quantify hemodynamic severity, it is an invasive procedure requiring ionizing radiation. Alternatively, for initial assessment of the hemodynamic significance of re-coarctation, CMR may be used (Figure 6). CMR has various approaches to non-invasively characterize coarctation severity. With 3D-MRA, the exact diameter, length, and location of the (re-)coarctation is visualized and can be measured. In addition, with 3D-MRA, the presence of collateral arteries is shown. In the presence of collaterals coarctation severity can be assessed by measuring collateral flow volume with the use of phase-contrast flow imaging. For this purpose, the aortic flow volume just proximal to the coarctation region is measured and compared with the flow volume in the descending aorta at the level of the diaphragm. In the presence of collaterals, the flow volume is increased at the distal aorta. The percent increase in volume provides a quantitative measurement of (re-)coarctation severity (34). In addition, CMR images in a 13-year old female with re-coarctation. BBSE images in axial orientation with view from below (a-c) and sagittal image with left lateral view showing the aortic arch with mildly hypoplastic descending aortic arch (a) and the aorta at the narrowest part of the coarctation (arrow in b, d) at the classic location distal from the origin of the left subclavian artery (SA, in d). Descending aorta distal from the coarctation (arrow, c). MRA showing the coarctation in original 2.8 mm slice image (e) and in thick 29 mm 3D maximum intensity projection image (f). Note the enlarged intercostal arteries and enlarged mammary artery that serve as collateral arteries as to bypass the coarctation (f).

MR images in a 13-year old female, same patient as in figure 5. 3D-MRA showing the locations for flow measurements with resulting flow volumes through the aorta. Flow was measured by velocity encoded imaging, immediately proximal to the coarctation, half-way below the coarctation, and at the level of the diaphragm. Note that the aortic flow increases from proximal to distal that indicates retrograde collateral flow towards the aorta. At the level of the diaphragm approximately 90% of flow through the aorta is due to collateral retrograde flow via chest-wall- and intercostal arteries. Output at the level of the aortic valve was 5.6 L/min.

CMR images in a 12-year old male with transposition of the great arteries after arterial switch. Axial black blood image with view from below (a) and MRA (e) showing the spatial relationships between the pulmonary trunk and ascending aorta. Lecompte maneuver locates the pulmonary trunk in front of the ascending aorta with the pulmonary arteries on either side of the aorta (a–c). Left pulmonary artery is partially obstructed due to the position of the main pulmonary artery in relation to the aorta. Axial GRE cine end-diastolic (b) and end-systolic (c) images showing the dynamic character of obstruction within the cardiac cycle. The neo-aortic root in this child was enlarged to 39 mm (d).
Tetralogy of Fallot
Tetralogy of Fallot (ToF) is the most common complex congenital heart disease with an incidence of 420 per million live births. In ToF patients, an anterior displacement of the RV outflow septum causes a ventricular septum defect, overriding of the aorta, pulmonic stenosis, and subsequent RV hypertrophy. Surgical repair of ToF involves closure of the ventricular septum defect and relief of the pulmonic stenosis by infundibular resection, valvulotomy and/or patch placement. The long term survival after surgical correction of ToF is excellent. Nevertheless, postoperative ToF patients may present with a range of functional and hemodynamic sequelae during follow-up. CMR plays an important role to assess these abnormalities. After surgical relief of the pulmonary stenosis, pulmonary regurgitation is common in ToF patients. As a result of the pulmonary regurgitation, longstanding chronic volume overload may cause RV dilatation and subsequent RV dysfunction (Figure 1 and Figure 2). Severe RV dilatation in postoperative ToF patients is associated with ventricular arrhythmia and sudden death. Therefore, in postoperative ToF patients with moderate or severe pulmonary regurgitation fraction ($\geq$25%) and severe right RV dilatation (RV end-diastolic volume $\geq$160-170 ml/m$^2$ or RV end-systolic volume $\geq$82-85 ml/m$^2$) or clinical symptoms, pulmonary valve replacement is recommended in order to improve clinical outcome.

Transposition of the great arteries
The population of postoperative patients with transposition of the great arteries consist of three subgroups of patients: those with D-transposition of the great arteries who underwent arterial switch operation, D-transposition patients after atrial switch operation (Mustard or Senning procedure), and patients with "congenitally corrected" transposition of the great arteries (L-transposition). The latter two subgroups have the morphological RV sustaining the systemic circulation, which largely determines the role of CMR during follow-up in these patients.

D-transposition of the great arteries after arterial switch
Patients with D-transposition of the great arteries have a discordant ventriculo-arterial connection. In these patients, the aorta arises from the morphological RV and the pulmonary artery arises from the morphological LV. The ‘D’ refers to the normal dextro-position of the embryonic bulboventricular loop (i.e. the morphological RV is on the right side). In patients with D-transposition, the atrial situs and the atrio-ventricular connections are normal. Common associated cardiac lesions include a ventricular septum defect and pulmonary outflow obstruction. In the absence of a sufficient shunt between the pulmonary and systemic circulation (such as a patent ductus arteriosus, a ventricular septum defect or an atrial septum defect), neonates with D-transposition present with severe cyanosis at the time of the physiological closure of the ductus
Manipulation, tension and flattening of pulmonary arteries may cause obstruction. Furthermore, the suture line of the pulmonary artery anastomosis may develop. Second, as a result of surgical complications, re-interventions are frequently performed during follow-up. The most common indications for re-intervention after arterial switch operation are RVOT obstruction and supra-valvar pulmonary stenosis, accounting for 40% of all reoperations. As a result, the compliance of the pulmonary trunk in patients with D-transposition may be reduced. Accordingly, even in the absence of significant pulmonary artery stenosis, increased peak flow velocities in the pulmonary trunk have been demonstrated in patients after arterial switch operation.

To appreciate possible pulmonary or RVOT stenosis and neo-aortic dilatation during CMR evaluation of postoperative D-transposition patients, specific attention should be paid to the LV and RV outflow tracts. Oblique sagittal and oblique coronal bright-blood GRE cines through the LV outflow tract and RVOT enable assessment of the regional diameters and geometry. Furthermore, multi-slice axial bright-blood GRE cines from the diaphragm up to the transverse aorta permit dynamic imaging of the supra-valvar aortic and pulmonary artery regions as well as assessment of ventricular volumes and function. In addition, with the use of 3D-MRA, more distal (branch) pulmonary artery stenosis can be identified. Finally, aortic and pulmonary valve competence and velocity can be quantified by phase-encoded flow imaging.

Finally, stenosis of the coronary arteries at the site of the surgical anastomosis may occur after arterial switch operation. Although these patients are often asymptomatic, they may be at risk for ischemia during follow-up. CMR enables various approaches for the detection of coronary artery disease, including late enhancement MRI, stress MRI (by supine bycicle ergometry or dobutamine infusion), contrast-enhanced myocardial perfusion MRI, coronary angiography MRI, and coronary flow MRI. Various authors have demonstrated the feasibility of these imaging techniques to detect coronary stenosis in young asymptomatic patients after the arterial switch operation. As a result, these imaging protocols are increasingly being incorporated into the CMR protocols of patients after arterial switch operation. However, at present, the clinical implications of the detection of coronary obstruction with CMR in asymptomatic patients after arterial switch operation remains under investigation.

D-transposition of the great arteries after atrial switch

Before the introduction of the arterial switch operation, which was first performed by Latane in 1975, D-transposition of the great arteries was surgically corrected at the atrial level. During atrial correction of D-transposition, a surgical baffle is constructed within the atria, redirecting the systemic venous blood to the LV and the pulmonary venous blood to the RV (Figure 8). The atrial baffle may be constructed with synthetic or pericardial tissue (Mustard procedure), or, alternatively, native atrial tissue is used (Senning procedure). In patients with D-transposition after atrial switch, the RV is supporting the systemic circulation. Consequently, the RV functions as a high pressure pump with increased oxygen demand, making it vulnerable for failure and/or ischemia. In a follow-up study including 91 patients with D-transposition after the Mustard procedure, over 50%...
Surgical management may be performed to improve outcome in patients with significant associated cardiac lesions. Most commonly, surgical repair for L-transposition of the great arteries includes closure of a ventricular septum defect, and/or tricuspid valve surgery or replacement. Alternatively, repair may be performed by means of the double switch procedure. The double switch procedure combines atrial rerouting (Senning or Mustard procedure) with the arterial switch. Hence, after the double switch operation, the morphological LV supports the systemic circulation. Currently, the double switch procedure is gaining ground in the surgical treatment of patients with L-transposition. During long term follow-up of L-transposition patients with a systemic RV, ventricular dysfunction and tricuspid regurgitation are common. In a multi-center study by Graham and colleagues on 132 L-transposition patients with various associated cardiac lesions, 67% presented with systemic RV failure at the age of 45.

CMR imaging plays an important prognostic role in the clinical follow-up of both patient subgroups with a systemic RV. Assessment of function and condition of the systemic RV can be studied in detail with various CMR techniques. CMR bright-blood GRE cines should be obtained to assess volumes and function of the systemic RV, as well as qualitative assessment of the presence of tricuspid regurgitation. In systemic RV patients with progressive RV failure and significant tricuspid regurgitation, tricuspid valve replacement may be performed. The importance of close clinical follow-up of RV ejection fraction is underlined by the results of a study of Van Son et al., who observed increased mortality in systemic RV patients after tricuspid replacement when pre-operative ejection fraction was below 44%. Image post-processing of the cine images requires special attention in systemic RV patients. During systemic RV analysis, hypertrophy of the RV myocardium may complicate identification of the endocardial border. When the hypertrophied RV trabeculations are included in the myocardial blood pool, RV volumes are may be significantly overestimated. A recent investigation showed that delineation of endocardial contours outside the trabeculations yields more reproducible and probably more accurate measurements.

Furthermore, stress MRI is of prognostic value in patients with a systemic RV. Winter et al. demonstrated that the inability of the systemic RV to increase ejection fraction and to reduce RV end-systolic volume upon exertion is significantly related to adverse outcome. In addition, late enhancement imaging enables further prognostic classification of patients with a systemic RV. Areas of late enhancement in the systemic RV have been associated with RV dysfunction, poor exercise tolerance, arrhythmia, and progressive clinical deterioration.

Finally, in D-transposition patients after the atrial switch operation and in L-transposition patients after the double switch procedure, another important goal of CMR imaging is the detailed assessment of baffle function and morphology. Obstruction of the intra-atrial baffle is frequently encountered during follow-up. Furthermore, baffle leaks may occur, causing left-to-right or right-to-left shunting at the atrial level. Baffle leaks may be quantified with phase-encoded flow imaging, by comparing aorta and pulmonary artery flow volumes (Qp/Qs). To delineate baffle obstruction, contiguous slices in the axial plane obtained by BBSE or bright-blood GRE cine imaging at the atrial level can be used. Fogel et al. demonstrated that a 3D image display of the obtained contiguous
Post processing of 3D time-resolved flow images for volumetric assessment of the tricuspid valve. Retrospective tracking of the tricuspid valve is performed by positioning a plane of interest on the tricuspid valve perpendicular to the flow in every cardiac phase in a four-chamber (a) and two-chamber (b) view (cine images: repetition time (TR) 3.9 ms, echo time (TE) 1.5 ms, flip angle 50°). Velocity-encoded images from three orthogonal directions (TR/TE 7.5 ms/4.3 ms, flip angle 10°) (c–e) reconstructed from the position of the valve determined with retrospective valve tracking based on the two-chamber (b) and four-chamber (a) views. Through-plane velocity-encoded images (f) are obtained by reformatting of the center valvular plane in each cardiac phase. The inner border of the tricuspid annulus is traced for flow analysis. A region within the free wall of the right ventricle is traced for background correction. Tricuspid flow velocity in time can be presented in a curve (g), from which flow volume can be calculated. Abbreviations: AP: anterior-posterior velocity-encoded, FH: feet-head velocity-encoded, RL: right-left velocity-encoded.

Abnormal vortical flow in a postoperative coarctation patient. Minimal residual narrowing (top arrow) of the mid aortic arch demonstrated with 3D-MRA (a) and BBSE (b) images. Streamlines are presented in mid (c) and late (d) systole. Disturbed flow is revealed (secondary flow features), with acceleration and signal drop out secondary to aliasing, as well as vortical-type flow (bottom arrow) downstream of the focal region of mild aortic narrowing.
slices is useful for this purpose, as complex spatial relationships are more easily clarified.(74) Furthermore, 3D-MRA is a valuable technique for anatomic imaging of baffle morphology in patients with D-transposition after atrial switch.(74)

Single ventricle patients after the Fontan procedure

The subgroup of CHD patients with a single ventricle is heterogeneous, with diverse complex cardiac anomalies. There may be a true ‘anatomic’ single ventricle, or, more commonly, there may be one ‘functional single ventricle’ (such as in hypoplast left heart syndrome or tricuspid atresia). The single ventricle can be of RV or LV morphology and a range of additional cardiac abnormalities may be present. The treatment of choice in patients with a single ventricle is the surgical construction of the Fontan circulation, performed in a staged process requiring at least two separate surgical interventions.(75) After completion of the Fontan circulation, the systemic and pulmonary circulations are separate and the single ventricle sustains the systemic circulation. Both caval veins are directly connected to the pulmonary arteries without interposition of a sub-pulmonary ventricle. Direct connection of the systemic venous return to the pulmonary arteries is constructed via an intra-atrial tunnel or an extra-cardiac conduit.(76) A fenestration may be created between the systemic venous return and the pulmonary venous pathways which allows for shunting towards the pulmonary venous circulation in case of increased pulmonary vascular resistance.

The Fontan procedure is a palliative procedure, although long term outcome is improving. In a recent report on 250 single ventricle patients who underwent the Fontan procedure, the 20-year survival after a successful Fontan procedure was 84%. (76) Failure of the Fontan circulation (defined as death, surgical ‘takedown’ of the Fontan, heart transplantation, or symptomatic heart failure in New York heart association (NYHA) functional class III or IV) was observed in 30% of patients after 20 years’ follow-up. (76) CMR evaluation can aid in the early recognition of single ventricle dysfunction. For this purpose, volumes, mass and ejection fraction can be studied by obtaining whole-heart bright blood GRE cine.(77) In addition to ventricular failure, other problems may complicate postoperative single ventricle with a Fontan circulation. Most commonly, obstruction of the pulmonary arteries or pulmonary veins occur, which can severely compromise the Fontan circulation. Flow obstruction to the pulmonary arteries may be due to anatomical narrowing or to increased pulmonary vascular resistance. Also, an enlarged right atrium or extra-cardiac conduit may compress the pulmonary veins.(76) Anatomic narrowing and obstruction in the pulmonary arterial and venous pathways can be visualized by static BBSE or bright-blood GRE cine acquisitions. Furthermore, obstruction can be revealed with the use of phase-encoded flow imaging by through-plane measurement of flow velocities. Of note, in Fontan patients, the maximum velocity-encoding during image acquisition should be anticipated upon the low flow velocities at the systemic venous side of the Fontan circulation. Moreover, as a result of the low blood flow velocities, the BBSE images should be comprehensively reviewed to identify possible thrombi. In addition to the BBSE images, bright blood GRE images and phase-encoded images, 3D-MRA can reveal possible obstruction of the pulmonary and venous pathways in Fontan patients. Furthermore, 3D-MRA is helpful in delineating possible collateral arteries. The detection of the presence of collateral arteries is of clinical importance in Fontan patients as collateral blood flow causes volume overload to the vulnerable single ventricle. Goo and co-workers recently evaluated time-resolved 3D-MRA for the assessment of flow dynamics in Fontan patients. In 15 young patients (median age 10 years), time-resolved 3D-MRA yielded data on preferential lung flow and collateral arteries with high spatial resolution.(23) Finally, imaging of bilateral lung perfusion is of clinical importance in Fontan patients.(79) Currently, perfusion scintigraphy is the method of choice to assess lung perfusion. However, Fratz and co-workers have demonstrated that in patients with a Fontan circulation, lung perfusion imaging with phase-encoded flow MRI is more accurate than perfusion scintigraphy.(79)

NOVEL CMR IMAGING TECHNIQUES IN CHD PATIENTS

In addition to the ‘conventional’ imaging sequences that are used during routine clinical follow-up, continuing research has provided several novel applications of CMR that have clinical value in postoperative CHD patients, including pulse wave velocity, 3D flow imaging, tissue velocity imaging and T1 mapping.

Aortic pulse wave velocity

The aortic pulse wave velocity is a surrogate marker for aortic wall compliance. It is defined as the velocity of the systolic wave front propagating through the aorta. Aortic pulse wave velocity, as assessed with Doppler echocardiography, is a strong predictor of cardiovascular mortality. In hypertensive patients, the aortic pulse-wave velocity, as assessed with CMR, has been shown to be associated with LV mass and lacunar brain infarcts. (82) With CMR, the aortic pulse wave velocity can be assessed at any site of the aorta even in the presence of a tortuous aortic vessel. To obtain the aortic pulse wave velocity with CMR, ‘conventional’ phase-encoded flow images at various regions of the aorta are acquired (usually at the ascending aorta, descending aorta and at the abdominal aorta just proximal to the iliac bifurcation). From the time-flow curves at each aortic region, the timing of the onset of the systolic flow wave can be obtained. (Figure 9) Subsequently, the time difference between the onset of flow between two aortic regions (transit time) is calculated. Finally, the pulse-wave velocity is obtained by dividing the distance measured between the two regions by the transit time. This CMR approach to assess the aortic pulse wave velocity was recently validated against intra-arterial pressure measurements during catheterization, and good agreement and high reproducibility were reported. (83) The application of CMR to assess the aortic pulse wave velocity has been performed in postoperative CHD patients. Grotenhuis and colleagues assessed the aortic pulse wave velocity of the aortic arch in 15 D-transposition patients after arterial switch operation. (58) A reduced elasticity, reflected by an increased pulse wave velocity, was observed in the patients as compared with 15 age-matched controls (patients: 5.1 ± 1.2 m/s, controls: 3.9 ± 0.7 m/s, p=0.004). (58) Another study reported the aortic pulse wave velocity of 16 ToF patients after pulmonary valve replacement. The pulse wave...
velocity at the aortic arch was increased in the ToF patients as compared with 16 healthy control subjects (5.5 ± 1.2 m/s vs. 4.6 ± 0.9 m/s, p<0.04). (47) Finally, Voges et al. assessed the aortic pulse wave velocity in 40 patients with a hypoplastic left heart syndrome and a Fontan circulation. In their study, the aortic pulse wave velocity was not significantly different in patients as compared with healthy controls. (84)

Current clinical research aims at further improvement of the assessment of the pulse wave velocity with CMR. Westenberg et al. recently validated the assessment of aortic pulse wave velocity by in-plane two-directional phase-encoded imaging of the aortic arch. (85) With this method, the entire aortic arch was depicted in three two-directional phase-encoded slices, which enabled the assessment of pulse-wave velocity among any two regions in the aorta. (85) The in-plane method was compared with the through-plane method and showed a better agreement with intra-arterial pressure measurement and an improved reproducibility. (85) Future follow-up studies using pulse wave velocity assessment with CMR are needed to reveal its prognostic value in CHD patients.

3D time-resolved flow imaging

Three-dimensional three-directional phase-encoded flow imaging (3D time-resolved flow imaging) enables quantification of valve flow as well as visualization of complex flow patterns. (86-90) Quantification of valve flow with conventional phase-encoded (two-dimensional) flow imaging is hampered by cardiac motion, since the acquisition plane is fixed throughout the cardiac cycle. Indeed, it has been shown that two-dimensional flow assessment of atrioventricular valve flow can lead to significant overestimation of flow volumes. (90) 3D time-resolved flow imaging solves the problem of valve motion by scanning a 3D volume with phase-encoding in three orthogonal directions. (87,90,91) This approach allows for ‘retrospective valve tracking’ during post-processing, a method that enables manual adjustment of the imaging plane in the 3D volume at any phase according to the angle and position of the valve of interest (Figure 10). (87,90) Furthermore, 3D time-resolved flow imaging allows simultaneous assessment of flow over all four valves, resulting in decreased scan time. (87) In postoperative ToF patients, 3D time-resolved flow has been validated against planimetry for the assessment of pulmonary and tricuspid valve flow. (89) As compared with two-dimensional flow MRI, 3D time-resolved flow imaging yielded better agreement with stroke volumes as assessed by SSFP planimetry. (89) Furthermore, the simultaneous assessment of flow over several valves enables evaluation of diastolic function of the RV in ToF patients with pulmonary regurgitation. (89) In the presence of pulmonary regurgitation, the flow pattern over the tricuspid valve does not reflect RV filling, since RV filling occurs also from the pulmonary regurgitation. Summation of the 3D time-resolved diastolic flow curves of the pulmonary valve and tricuspid valve allows the reconstruction of RV time-volume curves. (89) The RV time-volume curve represents RV filling and can be used to detect diastolic impairment in ToF patients. (89)

In addition to flow quantification, complex flow patterns can be visualized with 3D time resolved flow images. (86,88) This can be performed by particle tracing visualization in the 3D phase-encoded dataset. Particle tracing simulates the path taken in time by an imaginary particle (e.g., an erythrocyte in the vessel of interest). Particle tracing allows comprehensive visualization of complex flow pathways, for instance in patients with a Fontan circulation. (88) Furthermore, flow patterns can be characterized in static images with the use of so-called streamlines. Streamlines provide a snapshot of the instantaneous blood flow pattern in an image. In streamline images, laminar flow can be differentiated from ‘secondary flow features’ (e.g., vortices or helices) (Figure 11). Hope and colleagues performed 3D time-resolved systolic streamline imaging of the aorta in 26 coarctation patients and in eight healthy subjects. All healthy subjects showed laminar aortic flow patterns. However, vortical and helical flow patterns were observed in the descending aorta of 14 postoperative coarctation patients (54%). (86) Future studies are warranted to assess the clinical implications of flow visualization in postoperative CHD patients.

Tissue-velocity imaging

The echocardiographic assessment of peak systolic velocities with tissue Doppler imaging has shown to be of incremental value to conventional echocardiography to assess ventricular performance in various clinical conditions. (92-94) In addition, the assessment of the temporal occurrence of peak systolic velocities permits evaluation of mechanical dyssynchrony, which aids in the selection of heart failure patients that may benefit from cardiac resynchronization therapy. (95) With CMR, myocardial velocity (tissue-velocity) can be measured with phase-encoded imaging. (96-98) With tissue-velocity imaging, the maximum velocity is set at a lower threshold as compared to flow imaging (for example at 20 cm/s) which enables the assessment of myocardial motion velocity in time. Velocity can be encoded into any desired direction (usually along the longitudinal axis of the heart) and subsequently, regional time-velocity curves can be obtained. Tissue-velocity imaging with CMR has been validated against tissue Doppler imaging with echocardiography for the assessment of myocardial velocities and timings of myocardial velocities. (99-102) A recent study on tissue-velocity imaging of the RV with CMR in 33 ToF patients demonstrated significantly reduced peak systolic velocities at the RVFW and at the ROVT as compared with 19 healthy controls (RVFW: 8.2 cm/s (inter-quartile range 6.4-9.7) vs. 12.4 cm/s (10.8-13.8), p<0.01, RVOT: 4.7 cm/s (4.1-7.2) vs. 10.2 cm/s (8.7-11.2), p<0.01). (99) Furthermore, the mechanical activation pattern of the RV was evaluated and a significant time-delay within the healthy RV was observed, which was reduced in ToF patients. (99) Insight into RV mechanics may have clinical implications for future studies on pacing strategies in RV failure. In future trials, tissue-velocity MRI can provide detailed insight into myocardial performance, myocardial mechanics and possible dyssynchrony in postoperative CHD patients.

T1 mapping

The novel CMR application of clinical T1 mapping enables quantification of myocardial interstitial fibrosis. Interstitial fibrosis has been demonstrated within the myocardium of CHD patients (103) and may be an important substrate leading to heart failure. Currently, late enhancement imaging is the reference standard to image myocardial fibrosis or scar tissue. (5) However, with late enhancement imaging, diffuse myocardial fibrosis is not revealed, as the selected inversion time to
obtain late enhancement images is deliberately chosen to null normal myocardium. Hence, with late enhancement imaging, only macroscopic scarring can be visualized. By the use of contrast-enhanced T1 mapping (regional assessment of T1 time throughout the myocardium of interest), diffuse interstitial fibrosis may be quantified. Recently, various methods to obtain T1 maps in clinical practice have been proposed. In a heterogeneous group of adult CHD patients, Broberg et al. performed T1 mapping of the LV. The authors assessed the ratio of T1 in the blood pool and in the LV myocardium before and after gadolinium-chelate administration with a multi-phase inversion recovery GRE sequence (Look-Locker sequence). In the presence of myocardial fibrosis, the gadolinium has a delayed wash-out from the extracellular matrix of the myocardium, altering the T1 ratio of the surrounding blood and myocardium. With this technique, the authors observed significantly more interstitial fibrosis in CHD patients as compared to healthy controls. T1 mapping may be of great value in future research on the pathophysiology and treatment of heart failure in CHD patients.

CONCLUSION AND FUTURE PERSPECTIVE

Post-operative CHD patients constitute a heterogeneous group of patients that require life-long follow-up. In current clinical practice, noninvasive cardiac imaging with CMR plays an important role during follow-up of postoperative CHD patients, especially in those with complex CHD. CMR imaging yields 3D datasets that provide a comprehensive characterization of complex cardiac anatomy and reliable imaging of cardiac function. In addition, continuing clinical research on novel CMR techniques aims at providing new imaging parameters that may further improve clinical-decision making with regard to re-intervention and follow-up protocols in the growing CHD population.
REFERENCES

Right ventricular imaging: cardiac magnetic resonance

3.1 For the transposition of great arteries. Cardiol Young 2010;20:410-7.


