MRI Is the Preferred Method for Evaluating Right Ventricular Size and Function in Patients with Congenital Heart Disease

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In contrast to adult patients with acquired heart disease, abnormalities of the right ventricle (RV) are ubiquitous in children and adults with congenital heart disease (CHD). The RV is exposed to volume overload in shunt lesions (e.g., atrial septal defect, anomalous pulmonary venous connections) as well as congenital or acquired tricuspid and/or pulmonary valve regurgitation. RV pressure overload characterizes numerous congenital anomalies, including pulmonary valve stenosis or atresia, large ventricular septal defect, single ventricle, tetralogy of Fallot (TOF), truncus arteriosus, and transposition of the great arteries (TGA), to name a few. Importantly, many surgical and transcatheter treatments of CHD result in persistent or acquired volume and/or pressure overload of the RV. In some CHD patients, the RV functions as the systemic ventricle (e.g., palliated hypoplastic left heart syndrome, physiologically corrected TGA, and D-loop TGA following atrial switch procedure). Furthermore, exposure to cyanosis and to surgical procedures in the RV often lead to myocardial abnormalities, including scar tissue and diffuse fibrosis.

Given the frequent involvement of the RV in CHD, it is not surprising that assessment of RV size and function is key for guiding clinical decisions in these patients.[1] Among the diagnostic imaging tools available to clinicians for RV imaging, cardiac magnetic resonance (CMR) has emerged as the reference standard. In the following sections I will review the evidence supporting this contention, highlight how CMR data is used to guide clinical decisions, and discuss the strengths and weaknesses of CMR in comparison with other modalities, including echocardiography, computed tomography (CT), conventional x-ray angiography, and nuclear scintigraphy.

Versatility of CMR

CMR is ideally suited for assessment of the RV because it allows comprehensive assessment of cardiovascular morphology and physiology without most of the limitations that hinder alternative imaging modalities. Specifically, without restrictions related to acoustic windows, body size, scar tissue and other postoperative changes, exposure to harmful ionizing radiation, or the morbidity associated with invasive diagnostic catheterization,

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CMR provides high-resolution time-resolved 3-dimensional (3D) visualization of the right heart (Fig. 1). It allows depiction and quantification of blood flow, measurements of valve regurgitation (Figs. 2 and 3), and assessment of tissue characteristics (e.g., scar tissue) (Fig. 4).[2] No other imaging modality currently provides such comprehensive information in the clinical arena. The limitations of CMR — higher cost in comparison with echocardiography (but not in comparison with other modalities),[3] lack of portability, limited availability, artifacts from implants containing stainless steel (though no longer used in most modern implants),[4] and relative contraindication in patients with pacemaker or defibrillator[5] — are well documented. It should be noted that the risk of nephrogenic systemic fibrosis that has been linked to gadolinium-based contrast has largely been eliminated or greatly reduced by avoiding its use in patients with reduced glomerular filtration rate.[6] Importantly, when it comes to evaluation of the RV by CMR, use of a contrast agent is not required. Hence, on balance, the clinical benefits of the data obtained by CMR greatly outweigh it limitations as detailed in the following sections.

CMR is the Gold Standard for Noninvasive Measurements of RV Size and Function

Accuracy and Reproducibility

For any diagnostic test to be clinically useful, it must be accurate and reproducible. Accuracy can be determined by comparing measurements obtained by the technique or modality in question with those obtained by a reference standard. Accuracy determines how close to the "truth" a measurement is. Reproducibility addresses measurement variability, which can relate to the individual(s) performing the measurement (intra- and interobserver variability) as well variability related to repeated measurements (test-retest or interstudy variability). Reproducibility is especially crucial for tests that are being used for clinical surveillance over time, as is the case in serial follow-up of the RV in patients with CHD.

CMR has been shown to be both accurate and reproducible with regard to quantitative RV assessment. The combination of a time-resolved 3D dataset, clear distinction between the blood pool and the myocardium, and high spatial and temporal resolutions allow for accurate measurements of the RV regardless of its morphology or orientation within the thorax, and without geometrical assumptions. The accuracy of ventricular volume measurements by CMR was determined in the late 1980s and early 1990s using in-vitro phantoms, animal models, and in human subjects.[7–9] Experiments aimed specifically at the RV showed similarly excellent results.[10] For example, Koch et al. compared the accuracy of *in-vivo* RV volume assessment by CMR with *ex-vivo* measurements in 8 pig hearts.[10] Compared with volume measurements in the explanted hearts, observers 1 and 2 underestimated RV volume by a mean of 0.70 mL and 0.2 mL (1.6% and 0.45%), respectively. In another study, Beygui et al. compared the accuracy CMR measurements of RV mass with *ex-vivo* measurements in minipigs.[11] The correlation coefficient between *in-vivo* and *ex-vivo* measurements was 0.98 and the mean bias was 2.5 g.

The reproducibility of RV measurements is a notable strength of CMR over other modalities. Over the last decade, several groups have reported on inter- and intraobserver as

well as interstudy reproducibility of CMR measurements of RV volumes, ejection fraction (EF), and mass (Table 1).[12–15] Mooij et al. demonstrated low intra- and interobserver coefficients of variation in 60 children, most with abnormalities affecting the right heart.[12] The interobserver coefficient of variation for RV volumes and mass ranged from 6.4% to 11.3%; for LV volumes and mass variations ranged from 3.6% to 10.5%. Studies by Hudsmith et al.[16] and Grothues et al.[15] reported similar interobserver coefficients of variations for RV measurements. Clarke et al.[17] compared the observer variability of RV volume measurements between images obtained in the short-axis plane versus the axial plane in 50 patients with CHD. The intra- and interobserver reliability of RV end-diastolic volume, end-systolic volume, and stroke volume measurements was excellent for both contouring methods. In most measurements observer reliability was not influenced by the imaging plane except for RV end-systolic volume, which slightly favored the axial plane (p = 0.047). Blalock et al. demonstrated good interstudy reproducibility of RV measurements in 30 patients with repaired TOF, demonstrating the utility of CMR for serial evaluations of the RV in patients with CHD.[18]

Use of CMR as a Reference Standard for Other Modalities

CMR has been considered by many investigators as the gold standard for RV assessment since the late 1990s.[19] Over the past 15 years numerous publications have documented the use of CMR as reference standard for comparison of echocardiographic (2D, 3D, tissue Doppler, strain),[20–26] computed tomography,[27, 28] and radionuclear scintigraphy[29] measurements. In general, the level of agreement between echocardiographic variables and CMR depends on the subjects included (with influence from factors such as diagnosis or age) and the parameters evaluated. The overall picture that emerges from the literature highlights several consistent observations: 1) compared with CMR, the reliability of 2D echocardiographic measurements of RV size and function is modest with large limits of agreement; 2) RV volumes by 3D echocardiography correlate better with CMR measurements than 2D measurements, though systematic underestimation is common;[30, 31] 3) unlike promising results in adult patients with acquired cardiopulmonary diseases,[32] echocardiographic indices of longitudinal shortening (e.g., tricuspid annular plane excursion, TAPSE) in CHD are not as robust;[33, 34] and 4) RV myocardial velocities (by tissue Doppler) and deformation (by speckle tracking) are topics of intense interest but results are too preliminary to draw firm conclusions.[35] These and numerous other reports confirm that CMR is the reference standard for noninvasive assessment of the RV in patients with CHD.

Role of CMR in Guiding Clinical Decisions

The ultimate goal of any diagnostic test is to guide clinical management. In the context of managing patients with CHD that involves the RV, assessment of chamber size, global and regional function, pressure, scar tissue, thrombus formation, AV valve and semilunar valve regurgitation, and shunt quantification are all essential pieces of the diagnostic puzzle used to inform clinical decisions. Although some of these data can be determined by different diagnostic modalities, CMR has an advantage because it is capable of accurately and reproducibly providing most diagnostic information noninvasively and without exposure to harmful ionizing radiation.[36]

CMR has been shown to be useful in informing clinical decisions in several types of CHD that affect the RV. Repaired TOF is a good example in which CMR data is paramount to clinical management, as stated in the ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: "MRI is now seen as the reference standard for assessment of RV volume and systolic function."[37] Indeed, RV size and function, pulmonary regurgitation fraction, tricuspid regurgitation, differential pulmonary artery blood flow and anatomy, right ventricular outflow tract aneurysm, and residual shunts and sites of obstruction impact management decisions.[38–40] For example, criteria for pulmonary valve replacement rely on CMR-measured parameters such as RV volumes and ejection fraction (Table 2).[2] Several investigators have proposed threshold criteria for CMR-measured RV end-diastolic volume index as an important criterion for pulmonary valve replacement. Others have emphasized the importance of RV end-systolic volume index as an important criterion because it integrates both RV size and function.[41] Similarly, the importance of RV dysfunction measured by ejection fraction as a criterion for pulmonary valve replacement has been shown by several groups and accepted by the ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease.[37] More recently, data from an international multicenter cohort of patients with repaired TOF showed that lower left and right ventricular ejection fractions and higher RV mass-to-volume ratio measured by CMR are strong independent predictors of major adverse clinical outcomes, namely death and sustained ventricular tachycardia.[42] These observations highlight the utility of CMR in assessing prognosis and guiding clinical decisions in patients with repaired TOF, which comprises a substantial proportion of adolescents and adult patients with moderate or severe CHD.[37, 38, 43]

CMR is also valuable in other CHD that affects the RV. Examples in patients with unrepaired CHD include superior and inferior sinus venosus defects,[44] partially or totally anomalous pulmonary venous connection,[45] atypical atrial communications such as coronary sinus defect, Ebstein anomaly and other forms of dysplastic tricuspid valve,[46] anomalies of the RV myocardium such as arrhythmogenic RV cardiomyopathy,[47] RV outflow tract obstruction in patients with poor echocardiographic windows, absent pulmonary valve syndrome, and pulmonary hypertension.[48] In patients who underwent transcatheter and/or surgical management of lesions affecting the right heart, CMR is frequently being used to inform clinical management. Examples include assessment of pulmonary regurgitation and RV size and function following balloon dilation of pulmonary valve stenosis,[49] tricuspid valvuloplasty, residual shunts after management of septal defects,[36] and residual or recurrent RV outflow tract obstruction or pulmonary regurgitation.[41]

Role of Multimodality Imaging

Although CMR is the preferred modality for RV assessment, multiple diagnostic tools are used in clinical practice. The choice of which and when to obtain an echocardiogram, CT, nuclear scintigraphy, diagnostic catheterization, or a combination of these diagnostic procedures is dictated by the clinical question and by a host of patient-, modality-, provider-, and institution-related considerations.[3] The patient's clinical circumstance and the specific information sought constitute the first step in the decision-making process. Once those are

determined, patient-, modality- provider-, and institution-related considerations are weighted. Examples of patient-related factors include age, body size, ability to cooperate with the test, and presence of implantable metallic devices or pacemaker/defibrillator. Examples of modality-related considerations include accuracy, reproducibility, patient acceptance, and procedural risk versus benefit. Examples of provider-related factors include level of comfort and trust with specific modalities and their interpretation. Examples of institution-related considerations include access to different modalities, quality of hardware and software, level of expertise, and charges.

In clinical pediatric/congenital practice echocardiography is the first line of investigation. With regard to RV assessment, echocardiography is capable of providing the necessary diagnostic information to inform clinical decisions in many scenarios. Examples include the presence or absence of RV volume overload in a young child with a secundum atrial septal defect, abnormalities of the tricuspid valve with mild regurgitation, pulmonary valve regurgitation in a patient followed after balloon dilation of pulmonary valve stenosis, and infants and young children after repair of tetralogy of Fallot with uncomplicated clinical course and reassuring echocardiographic findings. Common to these circumstances is that precise determination of RV size and function and flow measurements (e.g., pulmonary regurgitation, differential pulmonary artery flow) are not essential for clinical decision making. In contrast, when accurate assessment of the RV is essential for clinical management (e.g., adolescent or adult patient with repaired TOF), CMR is the best tool currently available in the clinical arena. Due the to increased risk of cancer associated with ionizing radiation,[50] CT, nuclear scintigraphy, and diagnostic catheterization are used for RV assessment in this patient population only when the diagnostic information cannot be obtained by echocardiography or CMR.

Summary

A large body of evidence published during the past 15 years clearly indicates that CMR is presently the best diagnostic modality for assessment of RV size and function in patients with CHD. Moreover, a growing literature informs clinicians on how to use CMR data to guide patient management. Echocardiography, which is more widely available, provides useful diagnostic information in many clinical circumstances that affect the right heart. However, when precise quantitative data is required to make important clinical decisions (e.g., when to recommend pulmonary valve replacement), CMR remains the diagnostic modality of choice. As new echocardiographic, CMR, and other imaging techniques continue to evolve, it would be interesting to revisit this controversy in the future.

Acknowledgments

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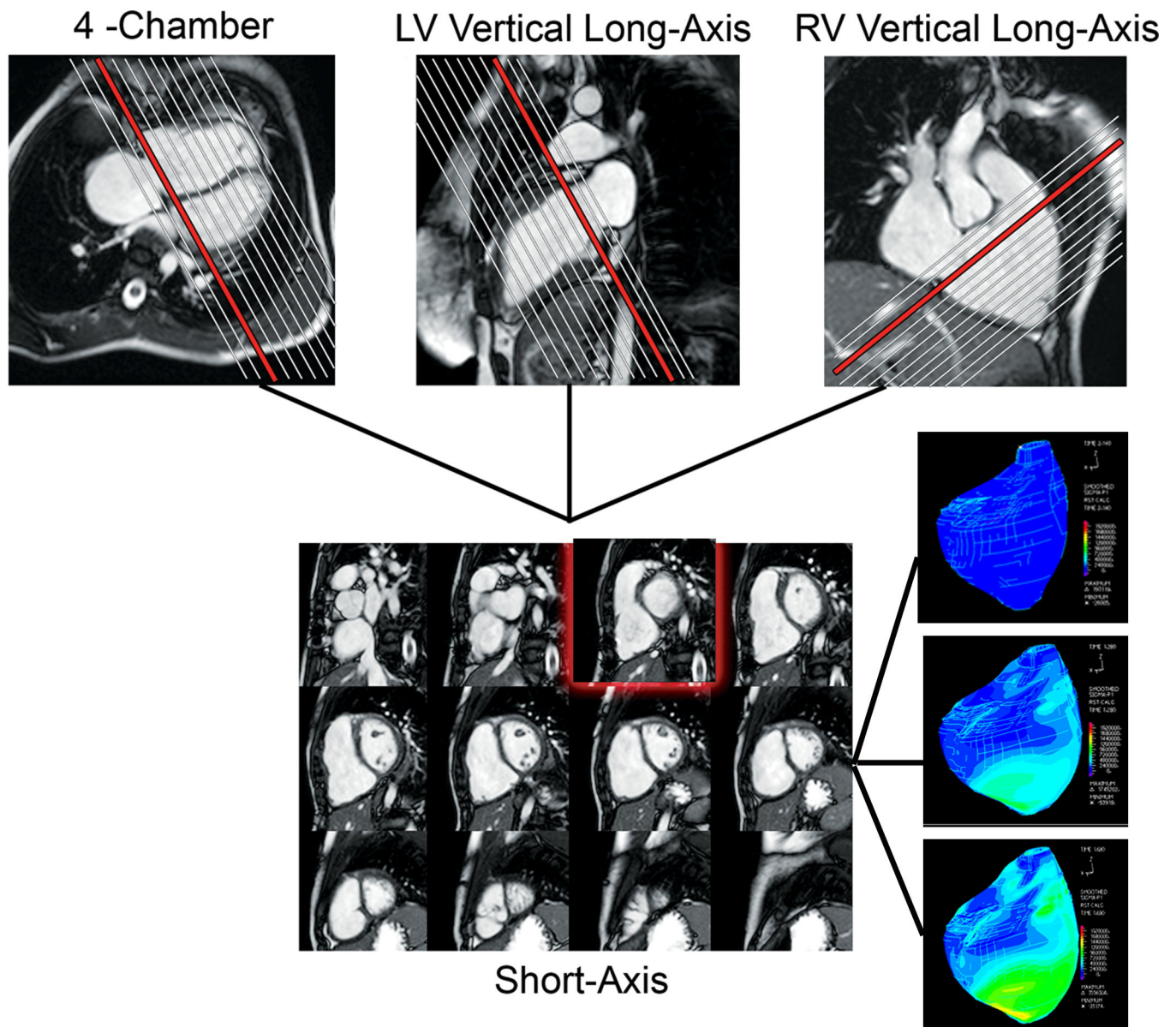
Figure 1.
CMR assessment of biventricular volumes and mass in a patient with repaired TOF. Cross-referencing between ventricular long- and short-axis imaging planes aids determining inclusion of basal slices in the ventricular volume analysis. **Right lower panel:** 3D strain maps of the RV at end-diastole (top), mid-systole (middle), and late systole (bottom).
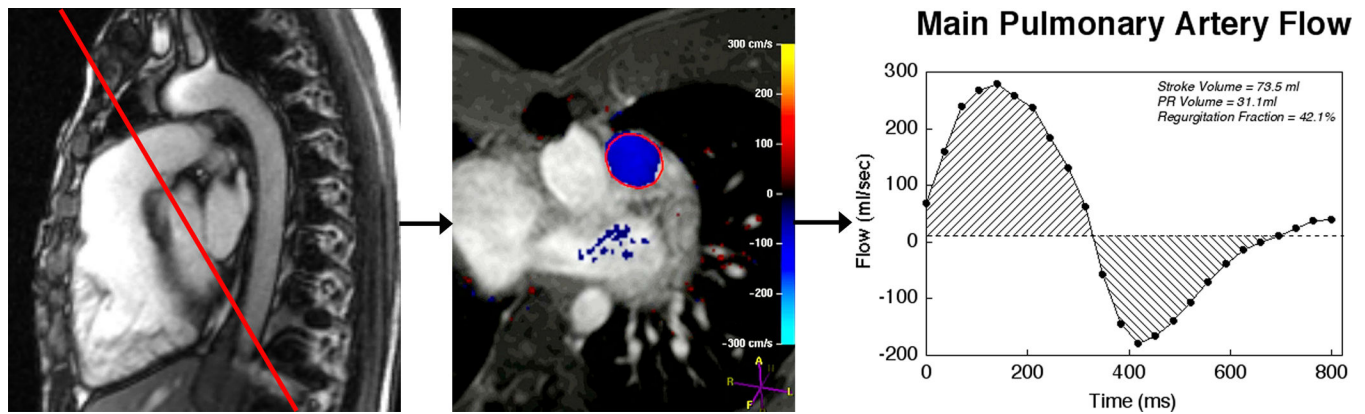
Figure 2.
Evaluation of pulmonary regurgitation by ECG-gated cine phase contrast MR. **Left panel:** The imaging plane is placed perpendicular to the long-axis of the main pulmonary artery (MPA); **Middle panel:** Color-coded flow map with the region of interest contour shown at peak systole; **Right panel:** MPA flow rate versus time. Flow above the baseline represents antegrade flow and flow below the baseline represents retrograde (regurgitation) flow.
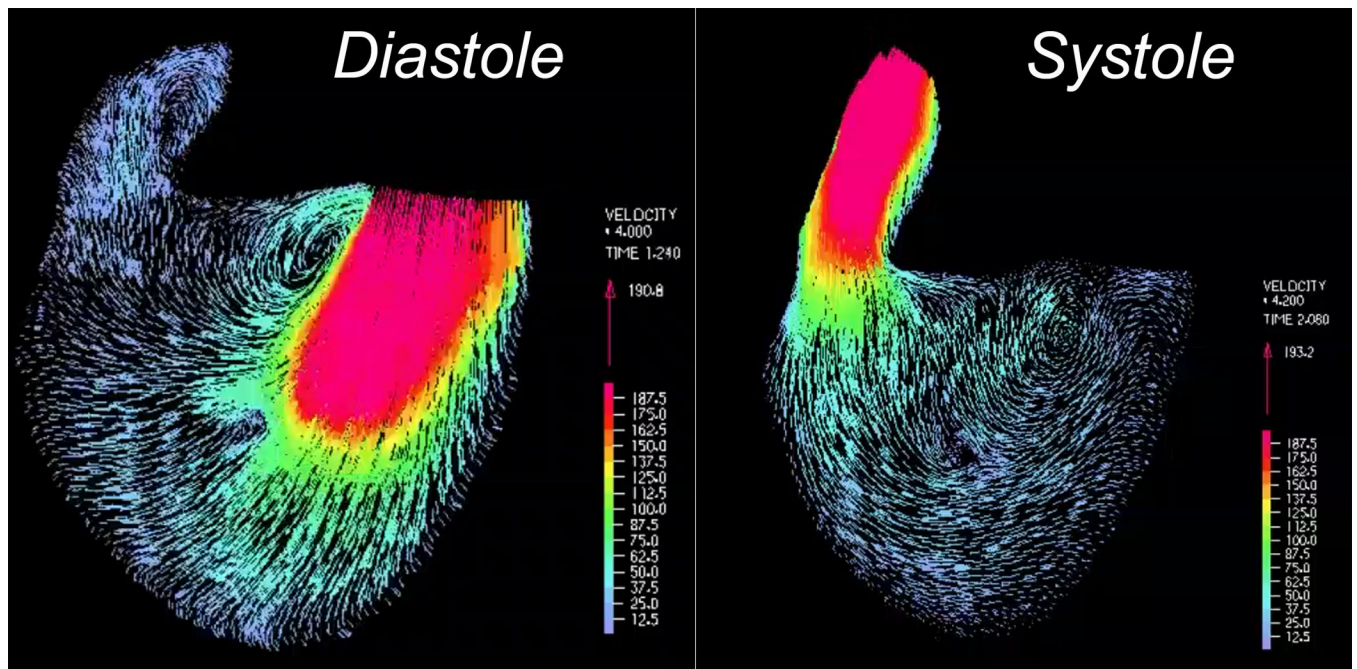
Figure 3.
Four-dimensional depiction of right ventricular blood flow based on cine phase contrast MR. **Left panel:** Early-diastolic frame showing blood flow through the tricuspid valve; **Right panel:** Mid-systolic frame showing blood flow through the right ventricular outflow tract.
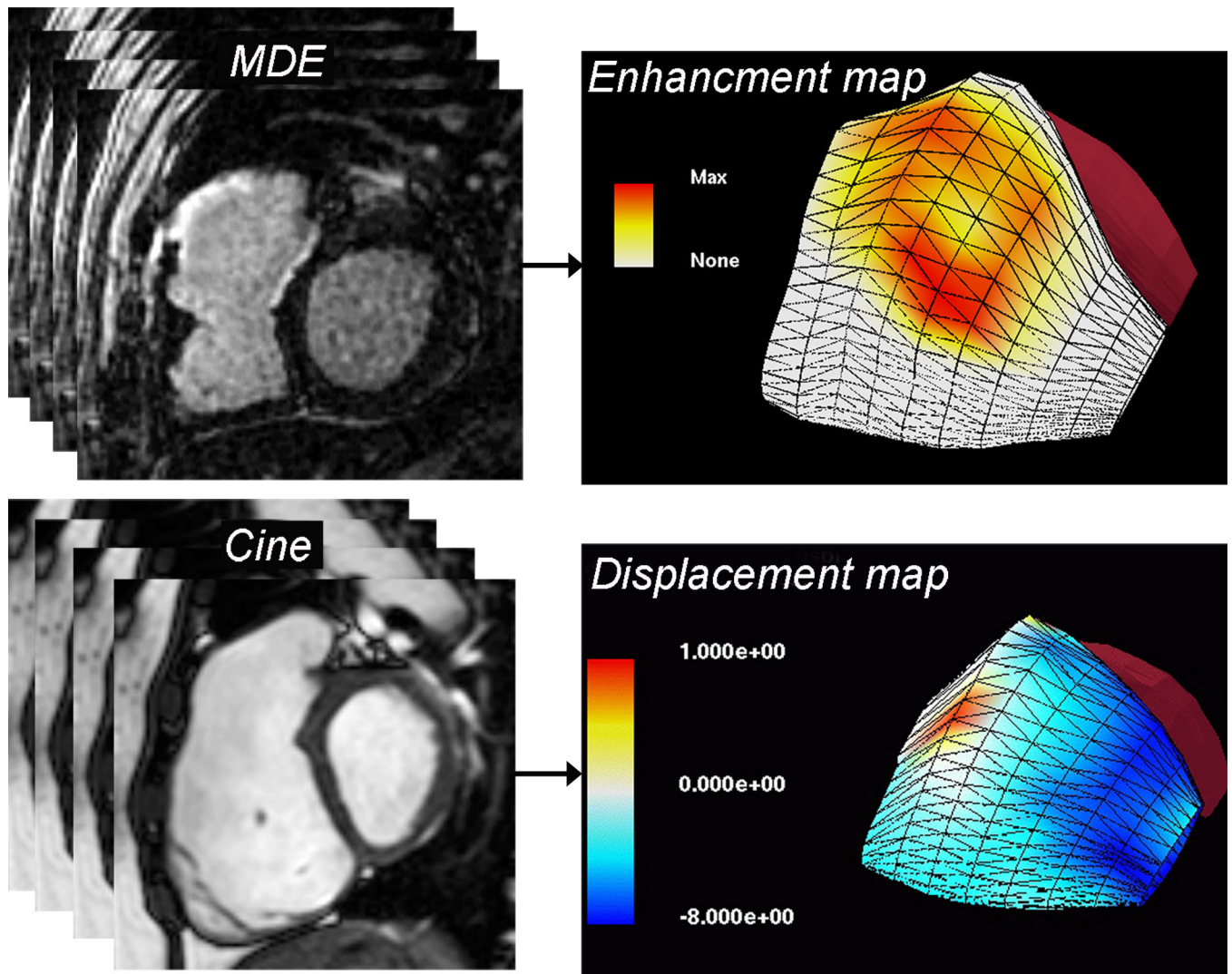
Figure 4.
Three-dimensional surface maps of RV scar tissue and motion.[2] The models were reconstructed from multi-slice 2-dimensional short- and long-axis images. **Top panel:** Scar tissue map based on late gadolinium enhancement (LGE) imaging showing extensive late hyperenhancement of the RVOT (yellow and orange). **Bottom panel:** Displacement map based on multi-slice cine SSFP showing dyskinesis of the RVOT (red).

Table 1

Reproducibility[*] of ventricular size and function measured by CMR (from Mooij et al.[12])

	Mooij et al.[12]	Grothues et al.[15]	Hudsmith et al.[16]	Karamitsos et al. (post-training)[13]	Karamitsos et al. (expert)[13]	Moon et al.[9]
No. of patients	60	60	12	10	10	20
Diagnosis	Normal/ASD/TOF	Normal/CHF/LVH	Normal	Normal	Normal	Normal/CHF
CMR technique	SSFP	FLASH	SSFP	SSFP	SSFP	SSFP
Right ventricle						
EDV	6.4%	6.2%	9.6%			
ESV	13.0%	14.1%				
EF	8.0%	8.3%	10.7%			
Mass	11.3%	8.7%				
Left ventricle						
EDV	3.6%		2.7%	4.6%	2.6%	2.6%
ESV	10.5%			7.4%	6.9%	10.5%
EF	5.8%		3.3%	3.7%	2.9%	6%
Mass	5.3%		5.2%	6.7%	5.8%	6%

[*] Reproducibility in this table is expressed as coefficient of variability (expressed as percentage)

Abbreviations: ASD, atrial septal defect; CHF, congestive heart failure; EDV, end-diastolic volume; EF, ejection fraction; ESV, end-systolic volume; FLASH, fast low-angle shot; LVH, left ventricular hypertrophy; SSFP, steady-state free precession; TOF, tetralogy of Fallot

Table 2

Role of CMR in informing the decision for pulmonary valve replacement in patients with repaired tetralogy of Fallot. Criteria based on CMR are marked with (*)

Indications for pulmonary valve replacement in patients with repaired TOF or similar physiology with moderate or severe pulmonary regurgitation (regurgitation fraction 25%)

I. Asymptomatic patient with 2 or more of the following criteria:

    a. *RV end-diastolic volume index >150 ml/m^2 or Z-score >4. In patients whose body surface area falls outside published normal data: RV/LV end-diastolic volume ratio >2

    b. *RV end-systolic volume index >80 ml/m^2

    c. *RV ejection fraction <47%

    d. *LV ejection fraction <55%

    e. *Large RVOT aneurysm

    f. QRS duration >140 ms

    g. Sustained tachyarrhythmia related to right heart volume load

    h. Other hemodynamically significant abnormalities:

        i. RVOT obstruction with RV systolic pressure 2/3 systemic

        ii. Severe branch pulmonary artery stenosis (<30% flow to affected lung) not amenable to transcatheter therapy

        iii. Moderate tricuspid regurgitation

        iv. Left-to-right shunt from residual atrial or ventricular septal defects with pulmonary-to-systemic flow ratio 1.5

        v. Severe aortic regurgitation

        vi. Severe aortic dilatation (diameter >5 cm or progressive dilatation >0.5 cm/year)[37]

II. Symptoms and signs attributable to severe RV volume load documented by CMR or alternative imaging modality, fulfilling 1 of the quantitative criteria detailed above. Examples of symptoms and signs include:

    a. Exercise intolerance not explained by extra-cardiac causes (e.g., lung disease, musculoskeletal anomalies, genetic anomalies, obesity), with documentation by exercise testing with metabolic cart (70% predicted peak VO$_2$ for age and gender not explained by chronotropic incompetence)

    b. Signs and symptoms of heart failure (e.g., dyspnea with mild effort or at rest not explained by extra-cardiac causes, peripheral edema)

    c. Syncope attributable to arrhythmia

III. Special considerations

    a. Due to higher risk of adverse clinical outcomes in patients who underwent TOF repair at age 3 years,[39] PVR may be considered if fulfill 1 of the quantitative criteria in section I

    b. Women with severe PR and RV dilatation and/or dysfunction may be at risk for pregnancy-related complications.[40] Although no evidence is available to support benefit from pre-pregnancy PVR, the procedure may be considered if fulfilling 1 of the quantitative criteria in section I

Adapted from Geva T.[2]