REVIEW

Review of the role of cardiovascular magnetic resonance in congenital heart disease, with a focus on right ventricle assessment

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Summary Adult patients with congenital heart disease (ACHD) represent a growing population due to progress in management. Surgical procedures generally fall short of restoring entirely normal anatomical and functional relations. Further procedures can be needed and lifelong follow-up is required. The right ventricle (RV) plays an important role in congenital heart disease and cardiac magnetic resonance (CMR) imaging has become the imaging method of choice for its assessment. CMR can provide relatively accurate measurements of RV volume and function, and arterial flow, with additional anatomical information provided by three-dimensional contrast angiography and late gadolinium imaging of fibrosis. Here we focus our review on three categories of ACHD in which evaluation of the RV is important: repaired tetralogy of Fallot, the systemic RV and Ebstein anomaly. We demonstrate how CMR contributes to decision-making regarding the types and timings of interventions. A dedicated CMR service should be regarded as a necessary facility of a centre specializing in the care of ACHD patients.
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KEYWORDS
Magnetic resonance imaging; Congenital heart disease; Tetralogy of Fallot; Ebstein anomaly; Systemic right ventricle

Abbreviations: ACHD, adult congenital heart disease; CCTGA, congenitally corrected transposition of the great arteries; rTOF, repaired tetralogy of Fallot; RV, right ventricle/ventricular; RVOT, right ventricular outflow tract; TGA, transposition of the great arteries; TOF, tetralogy of Fallot.

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Imagerie par résonance magnétique ; Cardiopathies congénitales ; Tétralogie de Fallot ; Anomalie d’Ebstein ; Ventricule droit systémique

Résumé Les adultes atteints de cardiopathies congénitales représentent une population de patients qui augmente grâce aux progrès dans leur prise en charge. Les interventions chirurgicales ne peuvent aboutir à un cœur complètement normal dans son anatomie et sa fonction. D’autres interventions peuvent s’avérer nécessaires et un suivi à vie est requis. Le ventricule droit (VD) a un rôle important dans les cardiopathies congénitales et l’imagerie par résonance magnétique (IRM) est la méthode de choix pour son évaluation. Elle permet d’estimer de façon relativement rigoureuse le volume et la fonction du VD, ainsi que la mesure des flux artériels. Elle permet également d’apporter des informations complémentaires anatomiques par l’angiographie 3D et de caractérisation tissulaire par la mise en évidence de fibrose. Nous concentrerons cette revue sur trois catégories de pathologie pour lesquelles l’évaluation du VD est essentielle : la tétralogie de Fallot réparée, le VD en position systémique et l’anomalie d’Ebstein. Nous montrons comment l’IRM intervient dans la prise en charge décisionnelle pour le type et le timing d’intervention. Un service d’imagerie dédié est souhaitable dans les centres spécialisés dans la prise en charge des patients atteints de cardiopathie congénitale.
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Background

Congenital heart disease occurs in about six to eight of 1000 live births [1,2]. Progresses in paediatric cardiology and cardiac surgery have revolutionized patients management, allowing most affected children to survive into adulthood. However, surgical procedures generally fall short of restoring entirely normal anatomical and functional relations. Most patients need lifelong follow-up and many require further intervention. The need to understand and manage appropriately the growing population with adult congenital heart disease (ACHD) or grown-up congenital heart disease has led to an expanding cardiological subspecialty in which imaging plays a key role.

In a number of conditions, the right ventricle (RV) is prone to dilatation and arrhythmia and it has received increasing attention in recent decades [3]. Two-dimensional echocardiography is the first-line cardiovascular imaging modality in ACHD, but RV volumetric and functional assessments remain challenging due to the asymmetric shape, trabeculated structure and the particular combination of long- and short-axis functions of the RV [4]. Cardiovascular magnetic resonance (CMR) imaging allows a segmental analysis and has gained importance in the measurement and visualization of RV volume and function [5,6]. RV function and volume evaluation is of importance in most repaired or unrepaird ACHDs, such as atrial septal defect, pulmonary atresia with intact ventricular septum, double outlet RV, tetralogy of Fallot (TOF), systemic RV and Ebstein anomaly. Repaired double outlet RV with subaortic ventricular septal defect and subpulmonary stenosis can be integrated into the section covering analysis of TOF. Finally, CMR contributes to decision-making regarding the types and timings of interventions.

Here we will focus on three categories of ACHD in which RV evaluation by CMR is crucial in clinical decision-making: repaired TOF (rTOF), the systemic RV and Ebstein anomaly.

Right ventricular volume and pulmonary regurgitation

The surgical repair consists of ventricular septal defect closure with enlargement of the right ventricular outflow tract (RVOT) by patch insertion. The abnormalities encountered are summarized in Table 1. The most frequent residual

<table>
<thead>
<tr>
<th>Table 1 Structural abnormalities encountered after tetralogy of Fallot repair.</th>
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</thead>
<tbody>
<tr>
<td><strong>Secondary to surgical repair</strong></td>
</tr>
<tr>
<td>Pulmonary regurgitation</td>
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<tr>
<td>RVOT scar</td>
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<tr>
<td>Ventricular septal defect patch</td>
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<tr>
<td><strong>Residual or recurrent lesion</strong></td>
</tr>
<tr>
<td>RVOT obstruction</td>
</tr>
<tr>
<td>Branch pulmonary artery stenosis</td>
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<tr>
<td>Ventricular septal defect</td>
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<tr>
<td>Atrial septal defect</td>
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<tr>
<td><strong>Acquired lesion</strong></td>
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<tr>
<td>Tricuspid regurgitation</td>
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<tr>
<td>RVOT aneurysm</td>
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<tr>
<td>Aortic root dilatation</td>
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<tr>
<td>Aortic regurgitation</td>
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<tr>
<td>Left ventricle dysfunction</td>
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<tr>
<td><strong>Associated anomalies</strong></td>
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<tr>
<td>Systemic-to-pulmonary artery collateral</td>
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<tr>
<td>Aortic branch pattern</td>
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RVOT: right ventricular outflow tract.
lesion is free pulmonary regurgitation leading to RV dilatation, which is well tolerated during infancy and childhood. The RV compensatory mechanisms tend to fail during the third decade and after, predisposing to arrhythmias, exercise intolerance, heart failure and death [11,12]. Severe chronic pulmonary regurgitation has been reported to be the main cause of RV dilatation and arrhythmias. The treatment is pulmonary valve replacement, but the optimal timing is still under debate [13,14]. Criteria for pulmonary valve replacement in rTOF patients include moderate or severe pulmonary regurgitation (regurgitation fraction ≥ 25%), as summarized in Table 2.

CMR has become the imaging method of choice in patients with rTOF [15–17]. It provides quantitative assessment of left ventricular and RV volume, mass, stroke volume and ejection fraction (Fig. 1-1). Several aspects of the RV
Table 2 Summary of indications for pulmonary valve replacement in repaired tetralogy of Fallot patients [37].

<table>
<thead>
<tr>
<th>Asymptomatic patients with two or more of the following criteria</th>
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<tbody>
<tr>
<td>RV end-diastolic volume index &gt; 150 mL/m² or Z-score &gt; 4 (in patients whose body surface area falls outside published normal data, RV/LV end-diastolic volume ratio &gt; 2)</td>
</tr>
<tr>
<td>RV end-systolic volume index &gt; 80 mL/m²</td>
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<tr>
<td>RV ejection fraction &lt; 47%</td>
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<tr>
<td>LV ejection fraction &lt; 55%</td>
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<tr>
<td>Large RVOT aneurysm</td>
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<tr>
<td>QRS duration &gt; 140 ms</td>
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<tr>
<td>Sustained tachyarrhythmia related to right heart volume load</td>
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<tr>
<td>Other haemodynamically significant abnormalities</td>
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<tr>
<td>RVOT obstruction with RV systolic pressure ≥ ½ systemic</td>
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<tr>
<td>Severe branch pulmonary artery stenosis (&lt; 30% flow to affected lung) not amenable to transcatheter therapy</td>
</tr>
<tr>
<td>At least moderate tricuspid regurgitation</td>
</tr>
<tr>
<td>Left-to-right shunt from residual atrial or ventricular septal defects with pulmonary-to-systemic flow ratio ≥ 1.5</td>
</tr>
<tr>
<td>Severe aortic regurgitation</td>
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<tr>
<td>Severe aortic dilatation (diameter ≥ 5 cm)</td>
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</table>

Symptomatic patients

- Symptoms and signs attributable to severe RV volume load documented by CMR or alternative imaging modality, fulfilling at least one of the quantitative criteria detailed above. Examples of symptoms and signs include:
- Exercise intolerance not explained by extracardiac causes (e.g. lung disease, musculoskeletal anomalies, genetic anomalies, obesity), with documentation by exercise testing with metabolic cart (≤ 70% predicted peak VO₂ for age and sex not explained by chronotropic incompetence)
- Signs and symptoms of heart failure (e.g. dyspnoea with mild effort or at rest not explained by extracardiac causes, peripheral oedema)
- Syncope attributable to arrhythmia

Special considerations

- Due to higher risk of adverse clinical outcomes in patients who underwent TOF repair at age ≥ 3 years [15], PVR may be considered if the patient fulfils at least one of the quantitative criteria in section I.
- Women with severe PR and RV dilatation and/or dysfunction may be at risk of pregnancy-related complications [38]. Although no evidence is available to support benefit from prepregnancy PVR, the procedure may be considered if the patient fulfils at least one of the quantitative criteria in section I.

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CMR: cardiac magnetic resonance imaging; LV: left ventricle; PR: pulmonary regurgitation; PVR: pulmonary valve replacement; RV: right ventricle; RVOT: right ventricular outflow tract; TOF: tetralogy of Fallot.

present challenges for reproducible volumetric measurement by CMR. Different software packages are available, but there is not yet a consensus as to which is preferable. The free wall and apical regions of the RV are normally extensively trabeculated, even more with hypertrophy. A decision has to be made about whether to include or exclude as cavity volume the blood signal among the trabeculations. Smooth tracing outside the trabeculations and inside the compact layer is probably the most reproducible, particularly at end-diastole, but is not necessarily the most accurate, especially in the presence of RV hypertrophy. The long-axis displacement of the base of the RV is usually greater than that of the left ventricle and it is important to include this basal region when locating the first short-axis slice at end-diastole. After r TOF, any thin-walled akinetic region of the RVOT should be included in the RV volume, up to the level of the pulmonary valve annulus. Whichever software or tracing method is used, it is important to maintain consistent methods, with a view to longitudinal comparison between studies [5]. CMR has been found to give good reproducibility in rTOF, with intraclass correlations of 0.966 for RV end-diastolic volume, 0.932 for RV end-systolic volume, 0.817 for RV ejection fraction and 0.831 for RV mass [17].

CMR phase contrast flow velocity mapping can quantify the pulmonary regurgitant fraction (Fig. 1-2); it can also quantify relative flow volumes through the right and left pulmonary artery branches, which can be further visualized by CMR cine imaging and contrast angiography. In the presence of unilateral pulmonary artery stenosis, a preferential flow (more than 2:1) to the unaffected side may be considered significant [18].
Right ventricular outflow tract

CMR may contribute to the stratification of risk (Fig. 1-2). The presence of RVOT aneurysm or akinesia [18], and the presence of fibrosis in regions outside those of direct surgical intervention was demonstrated by CMR late gadolinium enhancement [19] and was found to correlate with adverse clinical events. CMR angiography can be useful for providing three-dimensional visualization of a complex RVOT. Finally, CMR can detect other residual lesions, such as atrial or ventricular septal defect, quantifying the associated shunt by measurement of the pulmonary-to-ascending aortic flow ratio. CMR can also provide information about the severity of tricuspid regurgitation. Abnormal aortic branch pattern, which is frequently associated with conotruncal abnormalities and aortic dilatation and regurgitation, can also be studied.

Ebstein anomaly

Ebstein anomaly is an uncommon congenital malformation involving failure in the delamination of the tricuspid valve from the septal and inferior walls of the RV (Fig. 2). The inferior parts of the septal and mural leaflets of the tricuspid valve can therefore appear displaced towards the apex, with atrialization of the base of the RV. The functional part of the RV then consists mainly of the apical and outlet portions.

In severe cases, the tricuspid valve plane appears rotated, orientated in an almost horizontal plane beneath a dilated RVOT. Moderate or severe tricuspid regurgitation is common. The anomaly can also include a spectrum and abnormalities of the RVOT, such as pulmonary valve or pulmonary branch stenosis.

The functional RV tends to show progressive dilatation and dysfunction as tricuspid regurgitation increases [20]. CMR can provide useful information for the surgeon. The mural leaflet can be more fully visualized than by echocardiography [21] and the severity of regurgitation assessed [5,6]. Stacks of transaxial and short-axis cines and four-chamber and oblique coronal (or RV vertical long-axis) cines show the displacement of the inferior insertion of the tricuspid valve. The severity of tricuspid regurgitation can be assessed using through-plane CMR velocity mapping, the velocity encoding range typically set at 250 cm/s, to depict the cross section of the regurgitant stream through a plane transecting the jet on the atrial side of the defect. A regurgitant jet cross-section of 6 × 6 mm or more, reflecting the regurgitant defect, can be regarded as severe. An atrial
Figure 3. 3-1: cardiac magnetic resonance features of the systemic right ventricle (RV). Transposition of the great arteries repaired by Mustard operation in a 34-year-old patient. A. The pulmonary venous pathway (PVP) drains via a relatively right subsided atrial pathway (A and D) connected to the right-sided RV, which is hypertrophied and connected to the aorta (Ao). B. Both outflow tracks lie parallel to one another. C. The superior vena cava (SVC) and inferior vena cava (IVC) each drain via channels to the left of the baffle to the left-sided left ventricle (LV), which is connected to the pulmonary artery (PA). D. Chamber view of the PVP draining to the right-sided atrium connected to a hypertrophied systemic RV (B); 3-2: cardiac magnetic resonance features of the systemic right ventricle (RV). Atrioventricular and ventriculoarterial discordance (A and B) in an unoperated 45-year-old patient. A. The morphologically RV is hypertrophied and located to the patient’s left. The septal insertion of the tricuspid valve (Tri) is apically displaced relative to that of the mitral valve (Mi). B. The two outflow tracts lie parallel to one another (B). C. In the mid short-axis image, the left ventricle (LV) cavity has the smoother, less trabeculated septal aspect. Ao: aorta; LA: left atrium; PA: pulmonary artery; RA: right atrium.
septal defect can be present in about 50% of adult Ebstein patients, possibly due to the gaping of a patent foramen ovale, and should be sought by a CMR atrial short-axis cine stack.

As in other congenital heart disease, decline in exercise capacity may be taken as one of the relative indications for surgery, but outcomes may be better if surgery is performed before the heart dilates and the clinical status deteriorates [22].

The cardiothoracic ratio measured on a chest X-ray reflects RV dilatation and has been taken as a guide to the need for surgery, but CMR may now provide more comprehensive assessments of RV and tricuspid valve function in Ebstein anomaly [19,20].

However, it may not be easy to delineate the functional from the atrialized RV from short-axis volumetric measurements alone. Yalonetsky et al. found better reproducibility with transaxial imaging of the RV, in terms of both intra- and interobserver quantification in patients with the Ebstein anomaly [23] and correlation between RV atrialized volume and exercise capacity [24].

Systemic right ventricle

Systemic RV in adulthood falls mainly into two categories (Fig. 3-1 and 3-2): transposition of the great arteries (TGA) unrepaired or repaired by atrial switch (Fig. 3-1); and congenitally corrected transposition of the great arteries (CCTGA), meaning discordant atrioventricular and ventriculoarterial connections (Fig. 3-2).

The systemic RV and its tricuspid valve are exposed to systemic arterial load. In CCTGA, there can also be an Ebstein-like anomaly of the tricuspid valve. The RV adapts by hypertrophy, but does not seem ideally adapted to deliver systemic pressure lifelong. Systemic RV failure and tricuspid regurgitation tend to develop in the third decade of life [25–28]. Prieto found that 30% of CCTGA patients had significant RV failure after 20 years of follow-up, while more than 40% developed severe tricuspid regurgitation; 26% died or required cardiac transplantation at an average age of 25 years [29]. Subclinical RV dysfunction precedes the development of symptomatic heart failure [27,30–33] and worsens outcomes after tricuspid valve replacement [26]. CMR is the gold standard for systemic RV evaluation [6]. Which ventricle is morphologically “right” and which is “left” can be determined by the presence of multiple coarse trabeculations, including the moderator band, on the RV side of the septum. Each atrioventricular valve and ventriculoarterial connection should be visualized by appropriately aligned views. The choice of method for RV cavity delineation for volume calculation is challenging, but delineation immediately outside the trabeculations may give more reproducible measurements [3]. CMR has demonstrated good correlation between RV function and clinical status [31,34]. Furthermore, the presence of fibrosis as detected by late gadolinium enhancement is associated with adverse outcomes [31,35].

For patients with TGA and atrial switch, CMR can assess the atrial pathways as well. Cines and velocity maps can be aligned with respect to systemic and pulmonary venous atrial pathways [36]. Comprehensive coverage can, however, be achieved using a stack of contiguous transaxial or coronal cines or a three-dimensional SSFP sequence. Baffle-leaks may not be easy to identify by CMR, the suture line being long and tortuous. The measurement of pulmonary relative to aortic flow (Qp:Qs) may be useful, and any incompetence of the tricuspid valve into the systemic RV needs to be evaluated.

Limitations

The availability of CMR may still be limited, in part due to the cost of purchasing the equipment and the time needed for acquisition and analysis of images.

Claustrophobia is occasionally problematic. Anxiolytics can be considered in certain cases. Patient compliance is important for breath-hold acquisitions, which can present problems in patients with cognitive or behavioural disorders.

CMR image quality and measurement accuracy may be compromised by arrhythmias, although more rapid acquisitions may help to minimize the problem.

Patients with a pacemaker or implantable cardioverter-defibrillator should not approach the magnet because of the risk of device dysfunction and possible heating or voltage effects when wires are in the magnet during imaging. Further studies are required to evaluate the risk with new devices. Metallic objects such as wires, clips, stents, occlusion devices and recent prosthetic valves within the chest are safe in a 1.5T magnet, causing only local image artefact.

Safety information relating to the implant should, however, be consulted at the following website: http://www.mrisafety.com/.

Conclusions

CMR is the imaging method of choice for RV assessment in ACHD patients with rTOF, Ebstein anomaly and systemic RV. It can provide relatively accurate measurements of RV volume and function, and arterial flow, with additional anatomical information provided by three-dimensional contrast angiography and late gadolinium imaging of fibrosis. CMR visualization and measurements contribute to decision-making regarding intervention. A dedicated CMR service should be regarded as a necessary facility of a centre specializing in the care of ACHD patients.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


