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Right ventricular function late after total repair of tetralogy of Fallot

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Abstract Over the past decades, life expectancy in patients with congenital heart disease has increased dramatically. However, serious complications may develop late after total repair in infancy. These complications are usually the result of longstanding pulmonary regurgitation, which leads to dilatation of the right ventricle and an increased risk for severe arrhythmias. Therefore lifelong follow-up in these patients is required. Cardiac magnetic resonance imaging is the current imaging tool of choice because it offers superior imaging quality and enables accurate quantification of functional parameters such as flow volumes and systolic and diastolic performance.

Keywords Magnetic resonance · Heart · Review · Tetralogy of Fallot

Introduction

Presently, the vast majority of congenital heart diseases can be surgically repaired. However, even after corrective surgery, the right ventricle (RV) may remain subject to an abnormal pressure and/or volume overload, due to longstanding residual pulmonary stenosis and/or regurgitation (PR) [1, 2]. Hence, RV function has been shown to be a major determinant of clinical outcome in congenital heart disease (CHD) patients. With the growing number of long-term survivors amongst CHD patients, the need for accurate follow-up becomes increasingly important in clinical management. Careful monitoring of functional parameters such as cardiac function and vascular flow will help to allow early detection of the most important postoperative complications and aid in the timing of re-interventions [3]. Timely detection of late complications requires adequate assessment of right ventricular size and function as well as quantification of intracardiac blood flow in these patient groups. Widely used techniques, such as echocardiography and radionuclide studies, have limitations when applied for this purpose, especially in patients with abnormal right ventricular morphology.

Transthoracic echocardiography is still the most commonly used technique in the non-invasive assessment of CHD, especially in young children whose small thoracic diameter provides an optimal acoustic window [4]. After surgical intervention, however, the use of ultrasound is often restricted because of scar tissue and thoracic deformations [5].

The current gold standard in RV function assessment is cardiac MRI. The role of MRI for assessing pre- and post-

surgical aspects of CHD is well established [6]. Unlike echocardiography, MRI provides unlimited access to the thoracic cavity and different techniques are available for detailed visualization and accurate measurement of the complex post-surgical morphology and functional status. Biventricular volumes and blood flow can be accurately measured during a single examination, allowing complete evaluation of both right and left ventricular systolic and diastolic function as well as intracardiac and vascular flow.

With an incidence of 0.3–0.5 per 1000 live births, tetralogy of Fallot (TOF) is the most common cause of cyanotic CHD (Fig. 1) [7]. Traditionally, corrective surgery was preceded by a palliative shunt connecting the systemic arterial circulation to the pulmonary arterial system. It was thought that these shunts would promote growth of the pulmonary arteries and total correction could be delayed until the patient was larger. Nowadays, patients undergo total repair in infancy, which may decrease the risk of late sudden death, but may increase the severity of residual PR in some patients, due to the higher number of trans-annular patch procedures needed in young infants. In this review, we will focus on the MRI assessment of RV function in post-repair Fallot patients.

Right ventricular function

Systolic function

Ventricular function is traditionally divided into systole and diastole, describing ventricular emptying and filling, respectively. In tetralogy of Fallot, usually both systolic and diastolic right ventricular function is impaired, due to severe dilatation and hypertrophy. Assessment of emptying and filling patterns can be performed by echocardiography, or cardiac MRI.

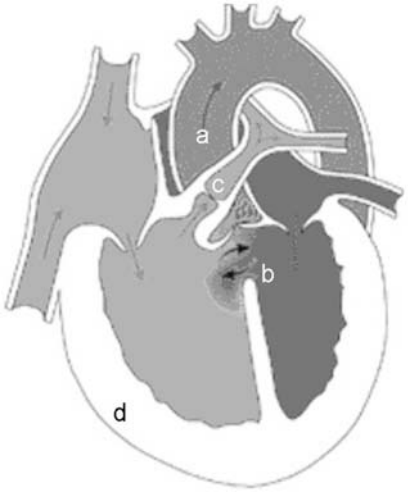

Fig. 1 Schematic view of the Fallot heart. **a** Overriding of the ventricular septum by the aorta, **b** ventricular septal defect, **c** pulmonary stenosis, **d** right ventricular hypertrophy

Nowadays, gradient-echo MR is the most accurate imaging modality for measuring ventricular volumes [8, 9]. This technique has been validated extensively for measuring right ventricular function [9–11]. The practical applicability of this method has also been demonstrated both in adults and in children. Helbing et al. [12] studied RV volumetrics using MRI in 20 children with CHD and 22 healthy children. In this study, a success rate of 97% was achieved for measuring RV function, while nearly half of the children were less than 10 years of age. The systolic function is usually expressed as the ejection fraction, calculated according to the Simpson's method (stroke volume/end-diastolic volume). In severe PR, the ejection fraction is usually diminished due to extreme dilation of the right ventricle. Since the increase in RV end-diastolic volume in these patients is more than the increase in stroke volume, these ventricles are probably acting on the down slope of the Frank Starling curve, stressing the need for timely PVR.

Since the stroke volume is largely determined by the volume of regurgitant flow, the ejection fraction is not useful in load-dependent conditions, e.g. before and after PVR. Therefore, RV ejection fraction should be corrected for pulmonary and tricuspid regurgitation as well as residual intracardiac shunts (EF_{cor}) by using the net pulmonary flow, instead of the stroke volume based on end-diastolic and end-systolic volumes. In a recent study by our group in 26 adult Fallot patients undergoing PVR for severe regurgitation, we found a dramatic increase in RV-EF_{cor} from 25.2±8.0% to 43.3±13.7% (P<0.001) using this method. Furthermore, the improvement of systolic function after PVR was associated with improved validity class, underscoring the practical implication of this approach. Global left ventricular function is a major prognostic factor in patients with heart failure. It is known that severe impairment of RV function may hamper left ventricular function. In a recent study, Dornier et al. [13] describe a new method for assessment of both global and regional left ventricular function in a single imaging session by using tagged MRI. Since the effects of PVR on left ventricular function are largely unknown, this method could be a valuable tool in assessment of pre- and post-PVR left ventricular function in TOF patients.

Diastolic function

Diastolic filling of the right ventricle can be divided into three different phases. After closure of the pulmonary valve, but before opening of the tricuspid valve, the ventricle relaxes, while the volume remains constant. This so-called isovolumetric relaxation is followed by the first phase, the early filling of the ventricle. The relaxed right ventricle is filled until equilibrium with atrial pressure is reached. The second phase, the diastasis, is recognized by

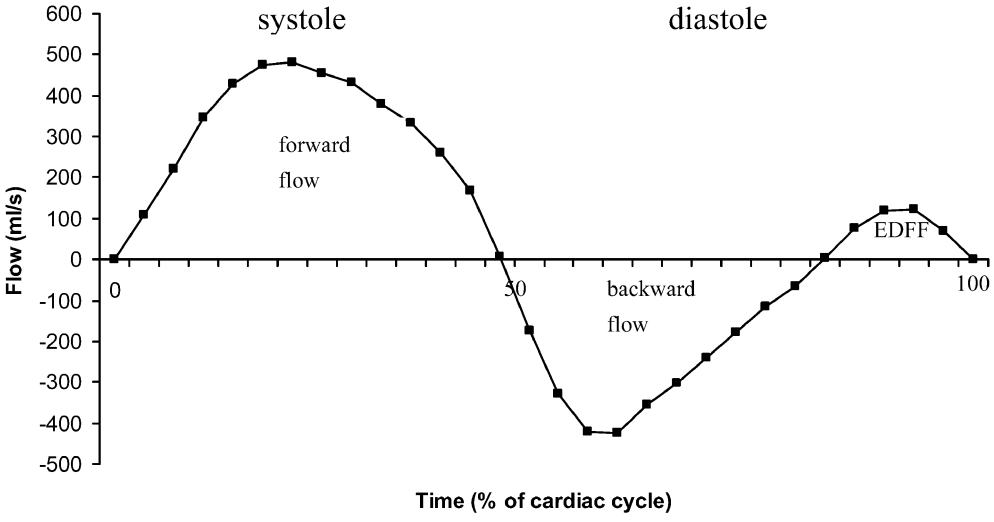

Fig. 2 Pulmonary flow pattern of Fallot patient

very slow diastolic filling. During the final phase, filling of the RV is the result of atrial contraction. Global diastolic function of the right ventricle can be derived from time–volume curves obtained with phase contrast MR imaging. Velocity encoding of both pulmonary (Fig. 2) and tricuspid (Fig. 3) flow provides accurate information on the diastolic filling pattern of the right ventricle, even in the presence of severe pulmonary regurgitation. Decreased diastolic function is frequently observed in CHD [14–16]. Diastolic dysfunction is characterized by an abnormal filling pattern which is marked by delayed relaxation and impaired diastolic filling with or without increased stiffness [17]. When diastolic filling is severely hampered, the ventricle may become restrictive. The restrictive right ventricle acts as a passive conduit between the right atrium and pulmonary artery, hence directly forwarding the blood flow during the atrial contraction to the main pulmonary artery [15–18]. The clinical implications of restriction are subject to debate; Helbing et al. [14] reported a decreased exercise performance in children with restrictive RVs after repair of TOF compared to similar children without signs of restriction. Gatzoulis et al. [15], however, found an improvement of exercise performance in combination with less cardiomegaly in repaired TOF patients with restriction, as compared to repaired TOF patients without restriction.

In recent years, steady-state free precession (SSFP) sequences such as sensitivity encoding (SENSE) have dramatically improved image quality. This is due to the higher signal-to-noise ratio, contrast-to-noise ratio and temporal resolution compared with conventional gradient-echo sequences. SSFP sequences have therefore become the sequence of choice for evaluation of cardiac (systolic) function. A recent study by Bernd et al. [19] showed that further improvement of temporal resolution can be

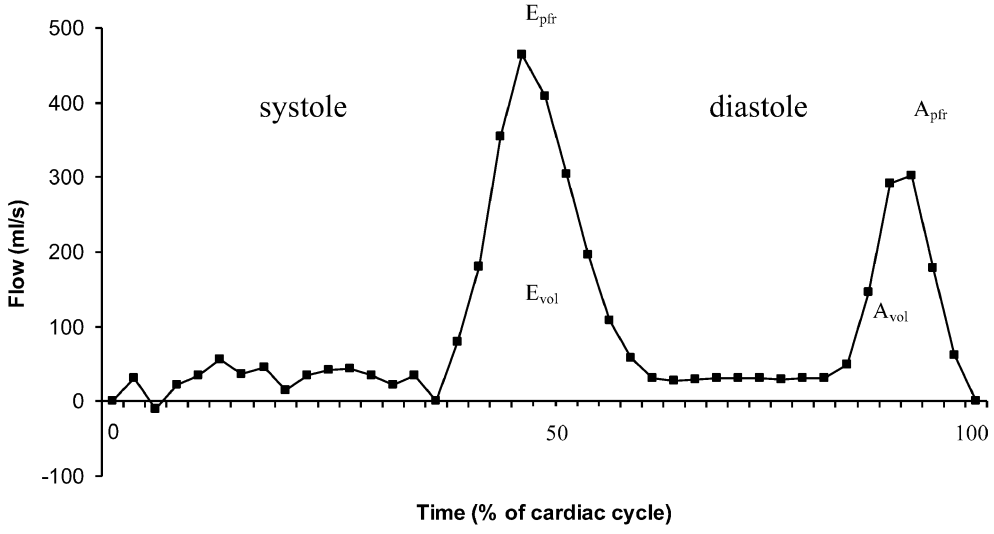

Fig. 3 Diastolic function assessment by tricuspid flow mapping. E_{pfr} Early filling, peak flow rate; E_{vol} early filling volume; A_{pfr} atrial contraction, peak flow rate; A_{vol} atrial contraction, volume

achieved by using a parallel imaging technique in comparison with standard segmented single-slice SSFP techniques.

Right ventricular outflow tract

The current interest in aetiological factors in the development of RV failure has focussed on the RV outflow tract (RVOT). Pulmonary regurgitation relates to RVOT reconstruction and, in particular, the usage of a patch, either transannular or confined to the right ventricle [20, 21]. Therefore, surgical management has moved towards preservation of pulmonary valve function if possible and limitation of the extent of patching when a patch is needed. Adequate relief of the RVOT without inducing significant PR, however, remains a surgical challenge. Since the emergence of cardiovascular MRI as the gold standard for evaluation of cardiac morphology and function, accurate quantification of valvular flow and ventricular volumes has been an important step towards optimal timing of pulmonary valve replacement. Recent studies have stressed the clinical importance of aneurysm formation and akinesia of the RVOT [22] (Fig. 4). Davlouros et al. found the combination of chronic PR and the presence of RVOT aneurysmal or akinetic regions to be the main predisposing factors for RV failure in adult patients late after repair of TOF. Furthermore, reconstruction of the RVOT during initial repair with a patch, whether transannular or confined to the right ventricle, was associated with higher PR fractions compared to patients who did not receive a patch, which is in accordance with previous studies [23]. Finally, in two other studies, the correlation between RVOT aneurysms and sustained ventricular tachycardia has been reported [24, 25].

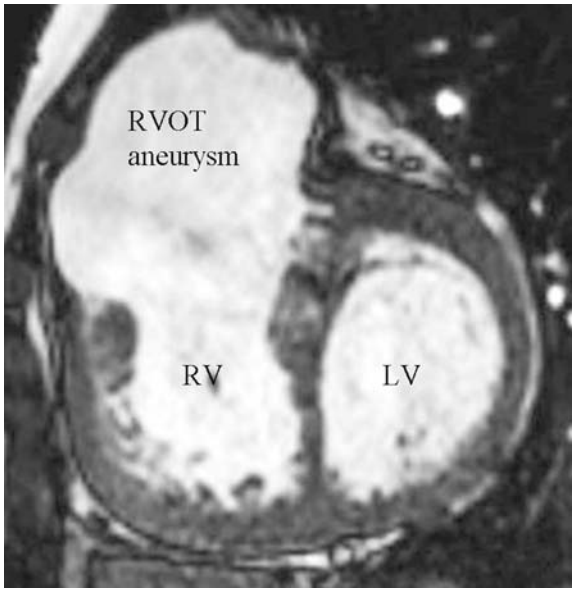

Fig. 4 Right ventricular outflow tract aneurysm late after total repair of tetralogy of Fallot. *RV* right ventricle, *LV* left ventricle, *RVOT* right ventricular outflow tract

Pulmonary valve replacement

Repeat operations late after total repair of TOF are not uncommon. The deleterious effects of longstanding PR and consequently RV dilatation (Fig. 5a, b) and arrhythmias, necessitate additional reconstructions in some, but not all, patients. The promising results of studies on RV function improvement in Fallot patients after pulmonary valve replacement advocate a less restrictive strategy concerning pulmonary valve replacement in patients with moderate to severe PR and RV dilatation late after total repair for tetralogy of Fallot [26, 27]. Pulmonary valve replacement may be combined, if necessary, with RVOT reconstruction, closure of residual VSD, tricuspid valve repair and reconstruction of peripheral pulmonary arteries. A regurgitant pulmonary valve may be replaced by a pulmonary homograft. Pulmonary homografts last longer than aortic homografts in pulmonary position. Nevertheless, homografts may suffer from calcific degeneration and usually need later replacement [28]. Some surgeons use mechanical valves in pulmonary position, but since the patients are usually young, and females may later wish to become pregnant, life-long anticoagulation should be avoided if possible. Before PVR, a full hemodynamic assessment is essential. In our institution, all patients who are eligible for PVR undergo a cardiac MR examination to assess the presence of valve insufficiencies and intracardiac shunting and to determine systolic and diastolic function. MR angiography is performed to detect possible branch pulmonary artery stenoses. Our experience with homograft pulmonary valve replacement late after correction of TOF now consists of 60 patients. Patients with pulmonary atresia or a conduit inserted at initial repair, patients with absent pulmonary valve syndrome or those presenting with a significant residual VSD or predominantly residual pulmonary stenosis are not included in this series. The mean interval between Fallot repair and later pulmonary homograft insertion is 19.3 years (range 2.7–40.3 years). Hospital mortality is 1.7% while one patient died 18 months after uncomplicated surgery, probably because of ventricular rhythm disturbances. Symptomatic improvement after PVR was striking: 6 months post-PVR validity class had improved from a mean NYHA class 2.3 to a mean class 1.4. Normalization of validity class was not observed in patients with preoperative validity class III. Comparison of pre- and postoperative MRI scans showed significant reduction of right ventricular volumes 6 months after insertion of the pulmonary homograft [27]. Furthermore, the RV function improvement was sustained at 19 months follow-up in the majority of patients. However, recurrence of pulmonary regurgitation after PVR appeared to reduce

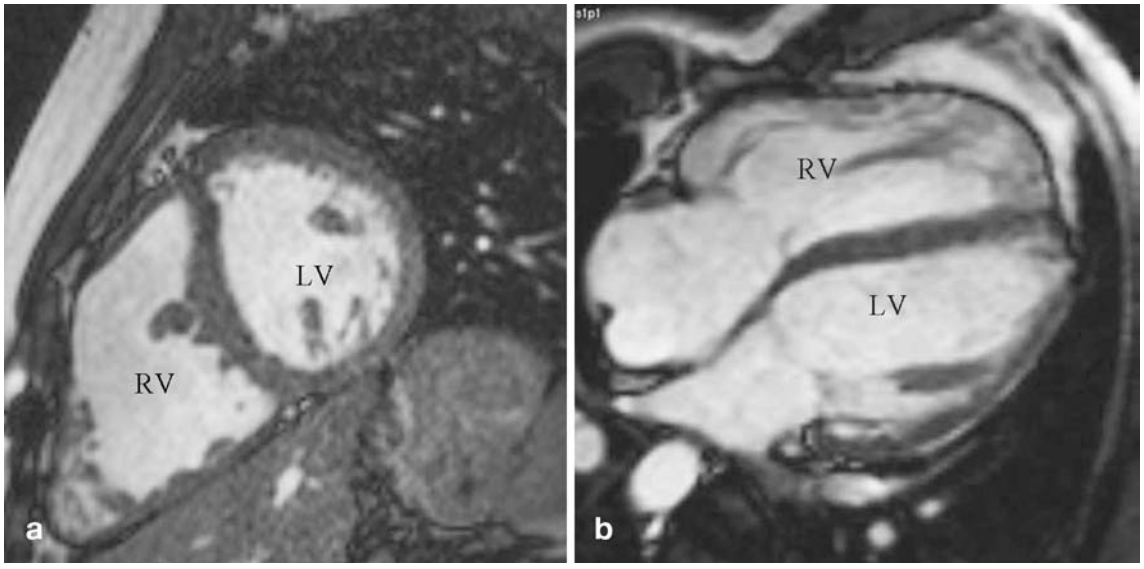

Fig. 5 **a** Dilated right ventricle due to chronic overload as a result of longstanding pulmonary regurgitation, short-axis view. *RV* Right ventricle, *LV* left ventricle. **b** Dilated right ventricle, four-chamber view. *RV* right ventricle, LV left ventricle

recovery of right ventricular systolic function [29]. Diastolic function, however, did not show early improvement 6 months after PVR. At 19 months, there was a significant improvement of diastolic function. Possibly the rapid volume unloading after PVR increases systolic performance, whereas improvement in diastolic function requires long-term remodeling [30]. Optimal timing of pulmonary valve replacement is still a subject of debate, as the amelioration of right ventricular function has to be weighed against the risk of later reoperation for homograft failure. A scoring system is under development to define the optimal moment of pulmonary valve replacement. Several parameters should be included such as functional validity class, right ventricular dimensions and ejection fraction, degree of pulmonary regurgitation, degree of tricuspid regurgitation, degree and nature of arrhythmia's, QRS duration, residual ventricular septal defect, and right ventricular outflow tract obstruction.

Other MRI applications

The recent implementation of fast MR imaging sequences along with the ability of MR imaging to acquire images in any given orientation and with high soft-tissue contrast makes this technique attractive for guiding interventional procedures [31, 32]. Razavi et al. [31] showed that cardiac catheterisation guided by MRI is safe and practical in a clinical setting, allows better soft tissue visualisation, provides more pertinent physiological information, and results in lower radiation exposure than do fluoroscopically guided procedures. MRI guidance could become the method of choice for diagnostic cardiac catheterisation in patients with congenital heart disease, and an important tool in interventional cardiac catheterisation and radiofrequency ablation. MRI guided cardiopulmonary interventions, however, are challenging due to motion artefacts caused by the beating heart and respiration. Moreover, the tortuous anatomy of the right cardiac chambers and pulmonary arteries makes monitoring of the passage of guide wires and endovascular catheters and the deployment of stents with MR imaging more difficult. Kuehne et al. [32] showed that interactive real-time MR imaging has the potential to guide stent placement in the pulmonary valve or main pulmonary artery and measure blood flow volume in the stent lumen immediately after the intervention. Several other authors have reported the use of MR imaging-compatible catheters and guide wires [33, 34]. However, none of the investigated instruments possessed the properties required for complex cardiovascular interventions, such as (1) fast and reliable detection of the tip; (2) curvature of the shaft; and (3) material properties such as tip flexibility, torque and tracking ability, shaft strength, and flexibility. More work is needed to develop new MR imaging-compatible catheters and guide wires that are appropriate for use in cardiovascular interventions. In the future, research methods such as use of resonance circuits as fiducial markers should be investigated to provide tip and shaft detection without incorporating the risk of heating effects inherent with active catheter tracking methods [35].

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