

Role of Cardiac MRI in Assessment of Post Fallot Tetralogy Surgical Repair Cases

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Abstract

Background: Tetralogy of Fallot (TOF) is a congenital heart defect characterized by four anatomical abnormalities. Cardiac MRI has emerged as a valuable non-invasive imaging modality for comprehensive evaluation of cardiac structure and function. **This study aimed to** evaluate the role of magnetic resonance imaging in assessment of post Fallot tetralogy surgical repair. **Patients & Methods:** This observational study included 30 patients with repaired Tetralogy of Fallot and referred to diagnostic radiology department, at Benha University Hospitals. All patients were subjected to complete history taking, full clinical examination, 12 lead ECG, Echocardiography, and Cardiac MRI. For further evaluation of CMR parameters, the studied subjects were divided into two group according to RVEF: Group A were patients with RVEF <50% and Group B were patients with RVEF >50%. **Results:** The mean age was 13.13 years. Sixty percent of the studied subjects were males and 40% were females. According to CMR parameters, RVEF mean level was 51.54. RV cardiac output mean level was 6.06 L/minute, RV stroke volume mean was 99.94%, RVEDV was 164.18 ml and RVESD mean was 75.87 ml. Pulmonary regurgitation fraction mean was 41.44, RV mass was 91.03 and LVEF mean was 69.25%. **Conclusion:** Cardiac MRI plays a crucial role in the assessment of post-surgical repair cases of Tetralogy of Fallot. By providing detailed anatomical and functional information, it helps clinicians evaluate the success of the surgical repair, detect residual abnormalities, and guide further interventions.

Keywords: Tetralogy of Fallot; Surgical Repair; Cardiac MRI; Postoperative Assessment.

Introduction

Tetralogy of Fallot (TOF) is the most common form of congenital cyanotic heart disease, occurring in approximately 4 to 5 per 100 000 live births, and represents 7%-10% of all congenital heart defects. Although TOF is often thought of in terms of the tetrad of anomalies—pulmonary stenosis, ventricular septal defect (VSD), aorta overriding the ventricular septum, and right ventricular hypertrophy—it has been proposed that all of these features are the result of anterior malalignment of the infundibular septum with the muscular septum (1).

Today, most patients with TOF undergo complete repair in early infancy or early childhood with >90% survival. Children who have undergone palliative procedures often suffer from failure to thrive and erythrocytosis. Left untreated, patients with TOF have a 50%, 5–10-year survival, with mortality related to hypoxemia, endocarditis, brain abscesses, or cerebral vascular accident. Longevity beyond the fourth decade of life in unrepaired or palliated TOF is rare (2).

Surgical repair of Tetralogy of Fallot (rToF) has significantly improved long-term survival. However, this repair can result in pulmonary regurgitation (PR) and right ventricular (RV) volume overload, which has been associated with RV dilation, RV dysfunction, symptomatic heart failure, ventricular arrhythmia and sudden death (3).

Chronic pulmonary regurgitation (PR) is a common consequence of the surgical repair of Tetralogy of Fallot (rToF) and may result in right ventricular (RV) dilation and dysfunction, decreased exercise tolerance, ventricular arrhythmia, and sudden cardiac death. Pulmonary valve replacement (PVR) can improve functional class, decrease or normalize RV volume, and reduce the risk of arrhythmias (4).

However, PVR does not improve RV function once it is already impaired. Hence, the procedure should be performed before the development of overt RV failure. The threshold of RV dilatation linked to irreversible RV dysfunction can vary from patient to patient. Furthermore, the evidence on the impact of PVR on RV remodeling, QRS duration and arrhythmia risk is also conflicting (5).

On the other hand, the operative risk of PVR, and more importantly, the limited life expectancy of prosthetic valves (especially in younger patients), needs to be taken into consideration. At present, it is still not clear whether the benefits of PVR outweigh the complications associated with the operation and the limited life expectancy of the prosthetic valves currently used. In addition, there are no specific PVR criteria for the pediatric population (<18 years old) or for gender. (4).

The radiologist plays an important role, interpreting many different imaging modalities, thus requiring full understanding of TOF anatomical features, surgical approaches and potential postoperative complications (6).

Echocardiography still remains the main diagnostic tool for the evaluation of pre and postoperative CHD. Because of numerous technical limitations, transthoracic echocardiography can fail to assess adequately hemodynamic or anatomic information. Among noninvasive imaging modalities, computed tomography (CT) angiography allows to evaluate cardiovascular structures with an excellent spatial resolution in a very short examination time (7).

Magnetic resonance imaging (MRI) provides a better image quality of intracardiac anatomy, unlimited choice of imaging planes, accurate flow quantification and ventricular function evaluation without the use of ionizing radiation and can thus be repeated without concerns for radiation toxicity (7)

The purpose of this study was to evaluate the role of magnetic resonance imaging in assessment of post Fallot tetralogy surgical repair.

Patients and methods

This observational study included 30 patients with repaired Tetralogy of Fallot who were referred to the diagnostic

radiology department at Benha University Hospitals, during the period from January 2022 to December 2022.

The study was done after being approved by the research ethics committee, Faculty of Medicine, Benha University (MS.43.7.2022). Informed written consent was obtained from the patients or their parents before enrollment in the study.

Inclusion criteria were patients of both sexes, age range from 1-20 years old and diagnosed with Fallot tetralogy and underwent surgical repair for assessment by cardiac MRI after repair.

Exclusion criteria were patients with the following significant confounding congenital heart defects as: common atrioventricular canal, heterotaxy syndrome, double outlet RV, major aorto-pulmonary collateral arteries, RV-to-pulmonary artery conduits, regurgitation of the aortic and/or the mitral valve. Contraindications to MRI as: patients with cochlear implant, cardiac pacemaker and foreign bodies.

All studied cases were subjected to:

- 1. Detailed history taking, including** (Personal history, Present history: course of the disease and duration, past history of any medical condition or previous hospital admission, Family history of similar condition, and surgical data concerning the timing and type of surgery)

2. Full clinical examination:

A-General examination including

(General comment on patient conscious and mental state, Vital signs: pulse, blood pressure, capillary filling time, respiratory rate and temperature).

B-Systemic examination including (Cardiovascular System, Respiratory System, Central Nervous System (CNS) and Musculoskeletal System Assessment of Glasgow coma score, pupillary reaction, examination of motor system including power, tone and reflexes)

3. Routine laboratory investigations:

Complete blood count (Hb, WBCs, Platelets). Random blood sugar, Kidney function testes, Liver function tests.

4. Radiological investigations (Cardiac MRI, 12 lead ECG and Echocardiography).

Cardiac MRI: Cardiac MRI examinations were conducted using a 1.5 T scanner (AERA 1.5 T scanner, Siemens, Erlangen, Germany), following established imaging protocols. The MRI scans evaluated the anatomy of the heart, utilized cine steady-state free precession sequences to assess volume and function, and employed phase-contrast imaging to measure flow in the pulmonary valve, aortic valve, and both pulmonary arteries.

Image analysis of the cardiac MRI data involved several measurements and calculations. The left ventricle (LV) and

right ventricle (RV) volumes were determined by manually segmenting the endocardial border on short-axis cine images at end-diastole and end-systole. The ejection fraction (EF) was calculated based on the measured volumes. All volumes were indexed by body surface area (BSA), using the DuBois formula ($BSA (m^2) = 0.007184 \times Height (cm) 0.725 \times Weight (kg) 0.425$), and compared with normal values from Kawel-Boehm et al. (2015).

Blood flow measurements were obtained from phase-contrast images using a semiautomatic edge-detection algorithm with operator correction. The regurgitant fraction was calculated by dividing the retrograde flow by the forward flow. Pulmonary regurgitation severity was classified as mild (<20%), moderate (20-40%), or severe (>40%). The diagnosis of pulmonary artery/right ventricular outflow tract (RVOT) obstruction was made when at least two of the following MRI criteria were met (in addition to RV pressure ≥ 45 mmHg in echocardiography): (1) flow velocity across the RVOT or a branch pulmonary artery ≥ 3 m/s, (2) abnormal pulmonary artery size, and (3) blood flow maldistribution (RPA < 40%; LPA < 20%). RV dysfunction was defined as a right ventricular ejection fraction (RVEF) $\leq 47\%$, while LV dysfunction was indicated by RVEF $\leq 55\%$.

In asymptomatic patients, pulmonary valve regurgitation (PVR) was considered by assessing two or more MRI parameters: RV end-diastolic

volume indexed by BSA (RVEDVi) ≥ 150 mL/m², RV end-systolic volume indexed by BSA (RVESVi) ≥ 80 mL/m², RVEF $\leq 47\%$, PRF $\geq 40\%$, LVEF $\leq 55\%$, moderate or greater tricuspid regurgitation, and RVOT obstruction with RV systolic pressure ≥ 0.7 systemic.

12 lead ECG: Performing a 12-lead electrocardiogram (ECG) is an important part of the cardiac evaluation of patients with repaired Tetralogy of Fallot. The procedure involved placing a series of electrodes on the patient's chest, arms, and legs to record the electrical activity of the heart.

Echocardiography: Transthoracic echocardiography was performed by an experienced cardiologist, using a GE VIVID S70 ultrasound system. The examination included standard views such as parasternal long-axis, parasternal short-axis, apical four-chamber, and subcostal views.

The following parameters were assessed: Cardiac chamber sizes, wall thickness, and function, Presence and severity of any valvular stenosis or regurgitation, Presence and severity of any residual ventricular septal defect and assessment of the pulmonary artery pressure. Detection of any abnormalities such as thrombus, masses, or pericardial effusion.

Statistical analysis

The collected data was revised, coded, and tabulated using the Statistical Package for Social Science (IBM Corp.

Released 2017. IBM SPSS Statistics for Windows, Version 25.0. Armonk, NY: IBM Corp.). Quantitative data were assessed for normality by the Shapiro-Wilk test and direct data visualization methods. Numerical data were summarized as means and standard deviations or medians and ranges. Categorical data were summarized as numbers and percentages. All statistical tests were two-sided. P values less than 0.05 were considered significant.

Results

The current study was carried on 30 subjects with history of surgical repair of tetralogy of Fallot. Their age ranged from 6 to 20 years old. Mean age was 13.13 years. Sixty percent of the studied subjects were males and 40% were females. **Table 1**

According to CMR parameters, RVEF mean level was 51.54. RV cardiac output mean level was 6.06 L/minute, RV stroke volume mean was 99.94%, RVEDV was 164.18 ml and RVESD mean was 75.87 ml. Pulmonary regurgitation fraction mean was 41.44, RV mass was 91.03 and LVEF mean was 69.25%. For further evaluation of CMR parameters, the studied subjects were divided into two group according to RVEF. Group A subjects in whom RVEF was $< 50\%$ and group B with RVEF $> 50\%$. **Table 2**

Mean age in group A was 13.66 years and 12.77 years in group B. Males represent 75% of group A subjects and

fifty percent of group B. Females were 25% of group A and 50% of group B.

Table 3

According to CMR parameter differences between the two studied

groups, significant differences shown in RVEF, RV COP, RVEDV, RVESV, Pulmonary ejection fraction, RV mass and LVEF. **Table 4**

Table 1. Baseline criteria of the studied group.

Total subjects n=30			
		Age (yrs)	
	M±SD	13.13	± 4.08
	Range	6	- 20
Gender, n (%)		<i>n</i> (%)	
	Male	18	(60%)
	Female	12	(40%)

Table 2. Cardiac MRI parameters in the studied subjects

Total subjects n=30		
	<i>M±SD</i>	<i>Range</i>
CMR		
RV cardiac output (L/min)	51.54±11.99	32.82-71.5
RV stroke volume (mL)	6.06±0.85	4.32-8.08
RV stroke volume (mL)	99.94±33.55	40.69-163.55
RVEDV (mL)	164.18±50.4	50.19-249.11
RVESV (mL)	75.87±19.36	31.64-112.83
Pulmonary regurgitant fraction (%)	41.66±4.44	29.65-49.36
RV mass (g)	91.03±25.69	38.94-145.54
LVEF (%)	69.25±6.82	55.54-84.76

Table 3. Demographic data differences between the studied groups.

	Group A n=12	Group B n=18	T	p
Age (yrs)	13.66±2.93	12.77±4.74	0.577	0.568
Gender				
Males	9(75%)	9(50%)	1.875	0.260
Females	3(25%)	9(50%)		

Table 4. Comparison of CMR parameters in the studied groups

	Group A n=12	Group B n=18	T	p
RVEF (%)	38.07±3.09	60.62±4.86	2.663	< 0.001*
RV cardiac output (L/min)	5.29±0.41	6.57±0.67	2.409	< 0.001*
RV stroke volume (mL)	91.14±31.03	105.8±34.73	0.893	0.248
RVEDV (mL)	211.28±21.68	132.78±37.68	3.100	< 0.001*
RVESV (mL)	67.05±20.81	81.76±16.36	0.605	0.039*
Pulmonary regurgitant fraction (%)	45.77±2.05	36.92±3.32	1.101	< 0.001*
RV mass (g)	115.99±15.99	74.39±15.08	0.243	< 0.001*
LVEF (%)	63.33±4.44	73.19±5.05	0.022	< 0.001*

T: independent t test

Cases

Case 1: Severe TR in a 16-year-old male with TOF repaired in infancy. A dephasing jet (arrow) is seen in the right atrium during systole on cine SSFP image in the 4-chamber plane. TR fraction calculated from velocity-encoded phase contrast imaging was 43-46%. **Figure 1**

Case 2: Dilated aortic root in a 19-year-old male with TOF status post valve-sparing repair as an infant. The aortic root is measured from sinus-to-sinus and sinus-to commissure on the end-systolic phase of this cine SSFP image obtained perpendicular to the root, and the largest dimensions are reported. **Figure 2**

Case 3: A 23-year-old female patient, diagnosed as tetralogy of Fallot, underwent RVOT repair at age of 1-year-old and mitral valve replacement as well. The patient was referred for RV volumes, function and Qp/Qs

quantification. 4-chamber cine left 2-chamber cine and left 3-chamber cine images showing mild tricuspid regurgitation and moderate mitral regurgitation on top of mitral valve replacement. Three sequential images from VSD cine showing the systolic jet through the VSD. Coronal pulmonary cine images during systole showing stenotic jet and during diastole showing dephasing regurgitant jet. Sagittal pulmonary in-plane phase-contrast velocity encoding sequence (phase) showing aliasing across the pulmonary valve at Venc 2.5 m/s needing increasing the Venc for optimal flow assessment. Q CMR revealed: Dilated RV with fair systolic function, preserved LVEF, moderate PR, RF=36%, residual PS with maximum velocity=3.5 m/s, RPA branch stenosis 3 m/s and LPA branch stenosis 2.5 m/s, mild TR, moderate MR, residual VSD with Qp/Qs=1.5:1. **Figure 3**



Figure 1: A dephasing jet (arrow) is seen in the right atrium during systole on cine SSFP image in the 4-chamber plane.

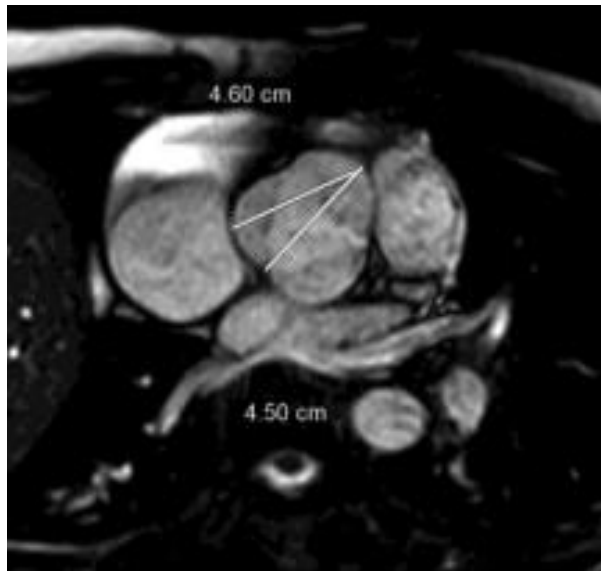


Figure 2: 19-year-old male with TOF status post valve-sparing repair as an infant.

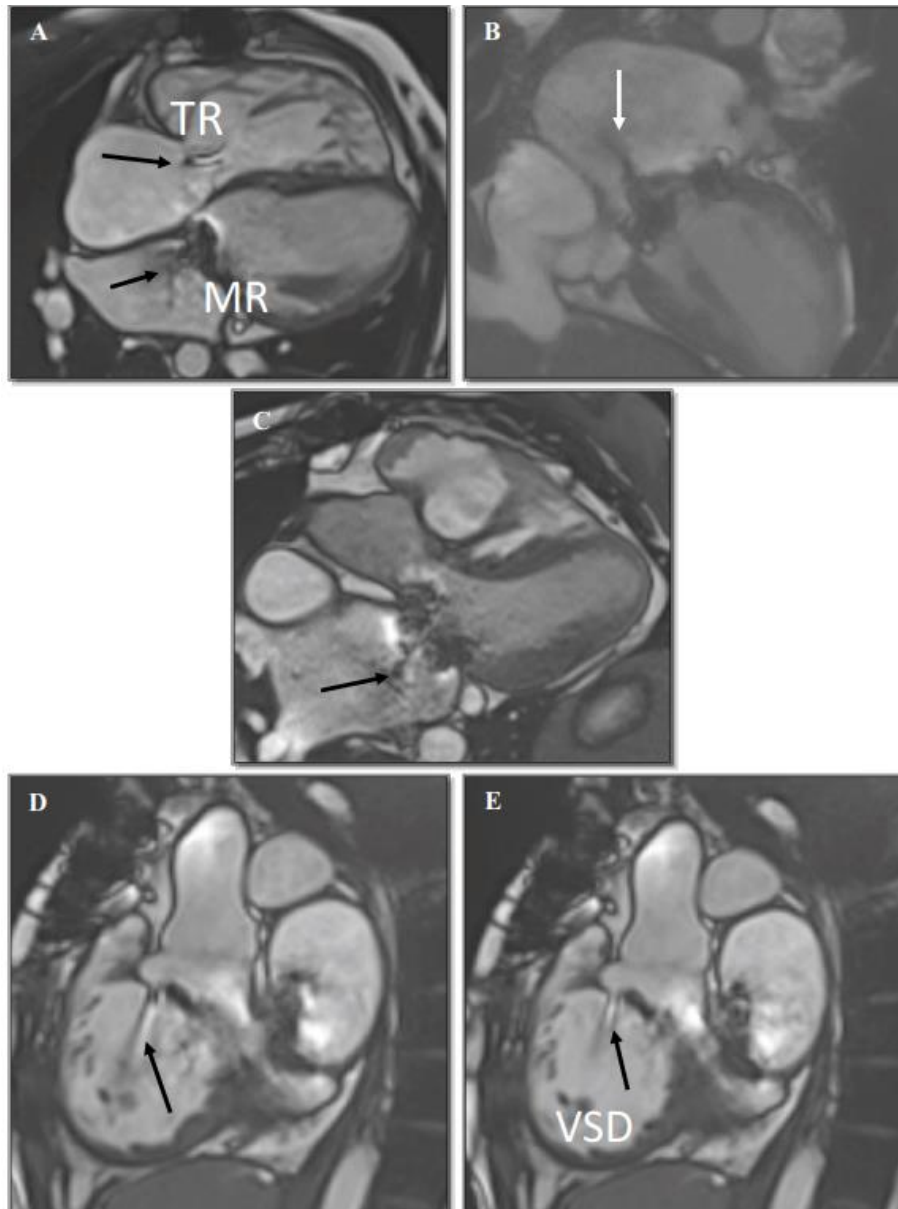


Figure 3: A, B, C 4-chamber cine, left 2-chamber cine and left 3-chamber cine images showing mild tricuspid regurgitation and moderate mitral regurgitation on top of mitral valve replacement. D, E

Discussion

In terms of demographic characteristics in the current study, the age ranged from 6 to 20 years old. The mean age was 13.13 years. Sixty percent of the studied subjects were males and 40% were females.

In agreement with the present study, as study was performed to evaluate the role

of cardiac MRI in functional and structural assessment of the right ventricle in TOF patients after surgical management. The study reported an age with a mean value of 21.20 (SD 13.59) years, also the study had a wide range of the age in their study as the age of patients ranged from 4 to 64 years. Regarding gender, 18 (60%) patients

were males while 12 (40%) patients were female with male to female ratio was 1.5:1(8).

In line with the current study, a study was carried out to identify independent factors associated with impaired clinical status in late survivors of tetralogy of Fallot (TOF) repair. The clinical and laboratory data of 100 consecutive patients with repaired TOF (median 21 years after repair) who completed a cardiac magnetic resonance imaging protocol were analyzed. The study reported a younger age at diagnosis and repair of this congenital heart defect, median age (IQR) (3.0 (0–31.8)). The majority of our patients were males, which is also consistent with previous reports (9).

In contrast, another study published aimed to evaluate the long-term outcomes of patients with repaired Tetralogy of Fallot using cardiovascular magnetic resonance imaging. The study included 93 adult patients with a mean age of 29.8 years, which is older than our patients (10).

According to cardiovascular magnetic resonance (CMR) parameters, RVEF mean level was 51.54. RV cardiac output mean level was 6.06 L/minute, RV stroke volume mean was 99.94%, RVEDV was 164.18 ml and RVESD mean was 75.87 ml. Pulmonary regurgitation fraction mean was 41.44, RV mass was 91.03 and LVEF mean was 69.25%.

This value is higher than what was reported by a study which found that the mean RVEF in repaired Tetralogy of Fallot patients was 44%. The RV cardiac output mean level was 6.06 L/minute, RV stroke volume mean was 99.94%,

RVEDV was 164.18 ml and RVESD mean was 75.87 ml (9).

For further evaluation of CMR parameters, the studied subjects were divided into two groups according to RVEF. Group A subjects in whom RVEF was <50% and group B with RVEF >50%. Mean age in group A was 13.66 years and 12.77 years in group B. Males represent 75% of group A subjects and fifty percent of group B. Females were 25% of group A and 50% of group B.

The studied subjects were divided into two groups according to RVEF. The differences in CMR parameters between the two groups showed significant differences in RVEF, RV COP, RVEDV, RVESV, Pulmonary ejection fraction, RV mass, and LVEF. This finding suggests that CMR parameters can be used to differentiate between patients with different degrees of RVEF impairment (11).

According to CMR parameter differences between the two studied groups, significant differences shown in RVEF, RV COP, RVEDV, RVESV, Pulmonary ejection fraction, RV mass and LVEF. TAPSE mean level in group A was 1.28 cm while 1.64 cm in group B. Statistically significant p value (<0.001*) was detected between the two groups.

In a retrospective study to present CMR findings of the asymptomatic patients with repaired TOF found eligible for PVR and collaborative pediatric cardiology and radiology experience with the largest CMR population. CMR examinations of 196 patients with repaired TOF performed between 2016 and 2018 were enrolled in this retrospective study. Only 165 were included in the study. CMR findings

were assessed according to the American College of Cardiology/American Heart Association guideline and recommendations of Geva. Among those 165 patients (median age 14 years [mean age 15.62 ± 7.42 years], M/F=114/61; 1.86/1), 73 patients were found eligible for PVR (59 patients for transcatheter while 14 patients for surgical). The mean QRS duration was 170.2 ± 16.89 ms. On CMR assessment, mean indexed right ventricular end-diastolic volume, end-systolic volume, right, and left ventricular ejection fraction were 187.64 ± 45.07 ml/m², $39.90 \pm 6.60\%$, and $47.83 \pm 6.12\%$, respectively (12).

A comparative study evaluated the diagnostic accuracy of cardiac MRI in the assessment of post-operative cardiac function in patients with tetralogy of Fallot. It was found that cardiac MRI had high sensitivity (96%) and specificity (90%) for the detection of right ventricular dysfunction, and that it provided superior diagnostic accuracy compared to echocardiography (13).

In addition to its superior diagnostic accuracy, cardiac MRI can also provide valuable information about cardiac anatomy and blood flow, which may be useful in the assessment of post-operative complications such as pulmonary regurgitation and right ventricular dilation. For example, in a study, cardiac MRI was used to evaluate the efficacy of pulmonary valve replacement in patients with repaired tetralogy of Fallot. The authors found that pulmonary valve replacement resulted in significant improvements in right ventricular volume, ejection fraction, and pulmonary regurgitation fraction (14).

Furthermore, a study evaluated the role of CMR in the assessment of pulmonary

regurgitation in post-operative TOF patients and found that pulmonary regurgitation fraction was significantly associated with adverse outcomes, such as arrhythmias, heart failure, and death.

A study found that CMR parameters, such as RV end-diastolic volume, RV mass index, and pulmonary regurgitant fraction, were strongly associated with adverse outcomes, including heart failure, arrhythmia, and death. The study concluded that CMR is a valuable tool for risk stratification and monitoring of patients with repaired Tetralogy of Fallot in the long-term (10).

In a study it was found that cardiac MRI is a valuable tool in assessing post-Fallot tetralogy surgical repair cases. The study reported that cardiac MRI provided accurate measurements of RV volumes and function, as well as assessment of pulmonary regurgitation, which is crucial for follow-up and decision-making regarding possible interventions (15).

Another study suggested that CMR parameters may not always accurately reflect clinical outcomes in patients with repaired Tetralogy of Fallot. The study reported that RV mass, RVEDV, and RVESV were not significantly associated with clinical outcomes, while other factors such as age at repair, pulmonary valve regurgitation, and RV function measured by echocardiography were more predictive of long-term outcomes (16).

Conclusion

Cardiac MRI plays a crucial role in the assessment of post-surgical repair cases of Tetralogy of Fallot. By providing detailed anatomical and functional information, it helps clinicians evaluate

the success of the surgical repair, detect residual abnormalities, and guide further interventions. Its non-invasive nature and excellent imaging capabilities make it a valuable tool in the management of TOF patients, ensuring optimal long-term outcomes.

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