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ORIGINAL ARTICLE

Cardiovascular magnetic resonance assessment of right ventricular function and pulmonary regurgitation in patients with repaired tetralogy of fallot: a single-center experience from Pakistan

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ABSTRACT

Background: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease requiring lifelong surveillance post-repair. Cardiovascular magnetic resonance (CMR) is the gold standard for assessing right ventricular function and pulmonary regurgitation, but limited data exists from South Asian populations. This study evaluates CMR findings in repaired TOF patients and assesses its role in pulmonary valve replacement (PVR) decision-making in a Pakistani cohort.

Methods: This cross-sectional study included 80 consecutive patients with surgically repaired TOF who underwent CMR evaluation ≥2 years post-repair at Hayatabad Medical Complex, Peshawar (January 2020-December 2024). CMR was performed using 1.5 Tesla scanners with comprehensive protocols. Primary outcomes included right ventricular volumes, ejection fraction, and pulmonary regurgitation severity.

Results: The cohort comprised 80 patients (mean age 14.5 ± 3.2 years, 56.25% male) with median 8.9 years since repair. Right ventricular dysfunction was prevalent with mean RVEF of 39 ± 11% and dilatation in 87.5% (mean RVEDVi - right ventricular end-diastolic volume index) 128.6 ± 32.4 mL/m²). Left ventricular function remained preserved (Left ventricular ejection fraction LVEF 53 ± 6%). Severe pulmonary regurgitation (>40%) occurred in 56.25% with mean regurgitation fraction of 36 ± 13%. Strong correlations existed between pulmonary regurgitation severity and right ventricular parameters ($p < 0.001$). PVR (Pulmonary valve replacement) was recommended in 81.3% of patients with 92.3% guideline adherence. Residual anatomical defects significantly worsened outcomes. Time since surgery correlated with progressive right ventricular deterioration ($p < 0.001$).

Conclusions: High prevalence of right ventricular dysfunction and severe pulmonary regurgitation exists in Pakistani repaired TOF patients. CMR demonstrated strong correlations with clinical outcomes and proved essential for treatment planning. These findings emphasize CMR's critical role in guiding timely interventions and optimizing long-term management in resource-limited settings.

Keywords: Cardiovascular Diseases, Congenital Heart Disease, Magnetic Resonance Imaging, Pulmonary Regurgitation, Pulmonary Valve Replacement, Right Ventricular Dysfunction, Tetralogy of Fallot.

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Introduction

TOF represents the most prevalent form of cyanotic congenital heart disease, characterized by a complex interplay of anatomical abnormalities including an overriding aorta, pulmonary stenosis, ventricular septal defect, and right ventricular hypertrophy (1). This complex combination of lesions typically manifests in approximately 3 out of every 10,000 live births, accounting for a significant proportion (7–10%) of all congenital cardiac malformations (2). While surgical correction typically occurs within the first year of life, often between three and six months of age, advancements in surgical techniques and post-operative care have significantly improved long-term survival rates, leading to a growing population of adult congenital heart disease patients (3). However, these patients frequently experience residual cardiac pathologies, with pulmonary regurgitation and right ventricular dysfunction being prominent concerns that necessitate ongoing surveillance and potential re-intervention (4). CMR has emerged as the gold standard for comprehensive assessment of right ventricular volumes, function, and pulmonary regurgitation fraction in these patients, offering superior reproducibility and accuracy compared to other imaging modalities (5). This study aims to evaluate the utility of CMR in characterizing the long-term sequelae of repaired TOF within the Pakistani population, specifically focusing on the prevalence and severity of right ventricular dysfunction and pulmonary regurgitation (6). Given the significant burden of congenital heart disease in Pakistan and the resource limitations prevalent in the region, understanding the precise role of advanced imaging techniques like CMR in guiding clinical management and predicting long-term outcomes in this specific demographic is critical. This research provides invaluable insights into the adaptation of cardiac structures post-repair, particularly highlighting changes in right ventricular reserve and its association with future heart failure hospitalizations (7, 8). Furthermore, this study seeks to identify potential correlations between CMR-derived parameters and clinical outcomes, thereby optimizing risk stratification and guiding timely interventions for this vulnerable patient cohort. This is particularly relevant in a developing country like Pakistan, where the epidemiological transition and an alarming increment of diseases ,including cardiovascular conditions necessitate a detailed understanding of long-term cardiac health in congenital heart disease survivors (9).

Methods

This descriptive cross-sectional study was conducted in the Department of Cardiology, Hayatabad Medical Complex Peshawar, Pakistan from January 2020 to December 2024. The primary objective was to evaluate cardiovascular magnetic resonance (CMR) findings in patients with repaired Tetralogy of Fallot and assess the role of CMR in pulmonary valve replacement decision-making. Ethical approval was obtained from the Institutional Review Board of Hayatabad Medical Complex via letter no. 2847. This study included patients with surgically repaired TOF who had undergone complete repair ≥2 years prior to CMR, were of appropriate age for cooperative imaging, and

had a clinical indication for CMR. All participants provided informed consent. Exclusion criteria comprised CMR contraindications (e.g., metallic implants, severe claustrophobia), severe renal dysfunction, inability to perform breath-holds, and incomplete/palliated TOF repairs. A total of 80 consecutive patients with repaired TOF who underwent CMR evaluation at our institution were included in this study. Patient demographics including age, gender, weight, and body surface area were recorded. The time interval between surgical repair and CMR evaluation was documented for all patients.

After detailed clinical evaluation, including detailed symptom assessment (dyspnea, exercise intolerance, palpitations, syncope), physical examination with cardiovascular focus, New York Heart Association (NYHA) functional class determination, 12-lead electrocardiography (ECG) with QRS duration measurement, and baseline transthoracic echocardiography, CMR imaging was performed (1.5 Tesla MRI-Philips Ingenia) using standardized protocols. Patients were prepared with peripheral intravenous access for contrast administration, optimal ECG lead placement for cardiac gating, and breath-hold instruction.

The imaging protocol included localizing images, cine images, phase-contrast flow quantification and late gadolinium enhancement (LGS) were evaluated using protocols previously described (10).

Images were reviewed by Pediatric cardiologist and the following analysis were done. Ventricular volumes, ejection fractions (EF), and mass were indexed to body surface area (BSA). Flow analysis quantified pulmonary regurgitation (PR) fraction/volume, while valve dysfunction was graded as mild (<25%), moderate (25-40%), or severe (>40%). Residual anatomical abnormalities (septal defects, right ventricular outflow tract [RVOT] aneurysms, branch pulmonary artery stenosis) were systematically documented.Therapeutic recommendations (e.g., pulmonary valve replacement (PVR) was based on integrated assessment of PR severity, ventricular remodeling (right ventricular end-diastolic volume index (RVEDVi), right ventricular ejection fraction (RVEF), residual lesions, and functional status per current guidelines. This standardized protocol ensured comprehensive evaluation of post-TOF repair pathophysiology to guide management.

Data were analyzed using SPSS (v22.0). Continuous variables were presented as mean ± standard deviation while categorical variables were expressed as frequencies and percentages. Descriptive analysis included patient demographics, surgical characteristics, residual anatomical abnormalities, ventricular function parameters, and valve regurgitation patterns. Comparative analysis assessed correlations between right and left ventricular parameters, associations between time since surgery and ventricular function, and the influence of residual lesions on dysfunction. Specific analyses evaluated pulmonary regurgitation severity versus right ventricular (RV) dimensions, RV ejection fraction's role in intervention decisions, residual lesions' impact on biventricular function, and age-related functional changes. Paired sample t-test was performed to find the mean differences between different parameters which was applied on categorical variables and $p<0.05$ was considered significant.

Results

A total of 80 patients with repaired tetralogy of Fallot were included in this analysis. The cohort demonstrated a mean age of 14.5 ± 3.2

years with a male predominance (n=45, 56.25%). The mean body surface area was 1.2 ± 0.3 m². The median time elapsed since initial surgical repair was 8.9 years (interquartile range: 6.5-11.2 years; mean: 10.2 ± 4.5 years).

Significant right ventricular dysfunction was prevalent throughout the cohort. The mean right ventricular ejection fraction (RVEF) was 39 ± 11%, with the majority of patients demonstrating systolic dysfunction (RVEF <45%). Right ventricular dilatation was observed in 87.5% of patients (n=70), with a mean RVEDVi of 128.6 ± 32.4 mL/m² and RVESVi of 76.3 ± 22.1 mL/m².

In contrast to right ventricular impairment, left ventricular systolic function remained preserved across the entire cohort. All patients maintained a left ventricular ejection fraction (LVEF) ≥50%, with a mean LVEF of 53 ± 6%.

The mean pulmonary regurgitation fraction was 36 ± 13%. According to established severity criteria, 45 patients (56.25%) demonstrated severe pulmonary regurgitation (>40%), 25 patients (31.25%) had Moderate regurgitation (25-40%), and 10 patients (12.5%) exhibited mild regurgitation (<25%). Strong correlations were identified between pulmonary regurgitation severity and right ventricular remodeling parameters. A significant positive correlation was observed between pulmonary regurgitant fraction and RVEDVi ($p < 0.001$), indicating that increased regurgitation severity was associated with progressive right ventricular dilatation. Conversely, a strong inverse correlation existed between pulmonary regurgitant fraction and RVEF ($p < 0.001$), demonstrating that higher regurgitation burden corresponded to diminished right ventricular systolic function. Stratified analysis by regurgitation severity revealed progressive deterioration in right ventricular parameters, as detailed in Table 1.

Table 1: Right Ventricular Parameters by Pulmonary Regurgitation Severity

PR Severity	N(%)	RVEDVi (mL/m²)	RVEF	PR Fraction	P-value
Severe (>40%)	45 (56.25)	142.3 ± 28.1	0.35 ± 0.09	>0.40	<0.001
Moderate (25-40%)	25 (31.25)	118.2 ± 25.6	0.43 ± 0.08	0.25-0.40	<0.001
Mild (<25%)	10 (12.5)	96.4 ± 18.3	0.51 ± 0.07	<0.25	<0.001

Abbreviations: RVEF; right ventricular ejection fraction, RVEDVi; right ventricular end-diastolic volume index, PR; pulmonary regurgitation

PVR was recommended for 65 patients (81.3%), while 15 patients (18.7%) were managed with conservative medical follow-up. The distribution of primary indications for PVR and adherence to guidelines are summarized in Table 2.

Table 2: Indications for Pulmonary Valve Replacement and Guideline Adherence

Parameter	Value
Primary Indications for PVR	
RVEDVi >150 mL/m²	28.1% of PVR cases
RVESVi >80 mL/m²	32.5% of PVR cases
PR fraction >40%	56.3% of PVR candidates
Symptom progression (NYHA II-III)	68.8% of PVR patients
Guideline Adherence	
AHA/ESC guideline compliance	92.3% (60/65 PVR cases)
Patients meeting RVEDVi >150 mL/m²	35 patients (43.75%)
PVR recommended in RVEDVi >150 mL/m²	32/35 patients (91.4%)

Significant correlations were observed between time elapsed since surgery and right ventricular parameters. RVEDVi demonstrated a positive correlation with time ($p < 0.001$), while RVEF showed a negative correlation ($p = 0.01$), indicating progressive right ventricular deterioration over time. Patients with >10 years post-repair demonstrated 28% higher RVEDVi compared to those <10 years post-repair ($p = 0.007$). Male patients exhibited significantly larger RVEDVi compared to females (135.4 vs. 119.8 mL/m², $p = 0.02$), while no significant difference in RVEF was observed between sexes ($p = 0.31$).

Residual anatomical defects were identified in 21 patients (26.25%) and significantly impacted right ventricular parameters. Patients with residual ventricular septal defects demonstrated the most severe impairment, while those with right ventricular outflow tract aneurysms and branch pulmonary artery stenosis were associated with universally high PVR rates. The detailed impact of each residual lesion type on ventricular function and treatment decisions is presented in Table 3.

Table 3: Impact of Residual Anatomical Lesions on RV Function

Residual Lesion	N (%)	RVEDVi (mL/m²)	RVEF	PVR Rate (%)
Residual VSD	6 (7.5)	158.3 ± 19.8	0.32 ± 0.07	100
RVOT aneurysm	7 (8.8)	146.2 ± 15.4	0.36 ± 0.08	100
PV stenosis	15 (18.8)	134.7 ± 28.2	0.37 ± 0.09	86.7
BranchPA stenosis	3 (3.8)	142.1 ± 24.6	0.34 ± 0.08	100
No residual defects	49 (61.3)	122.4 ± 30.1	0.41 ± 0.10	76.3

Patients recommended for PVR (n=65) were compared with those managed conservatively (n=15). While no significant differences existed in age or sex distribution, the PVR group had significantly longer time since initial repair. As expected, the PVR group demonstrated significantly worse right ventricular parameters and higher pulmonary regurgitant burden, while left ventricular function remained comparable between groups (Table 4).

Table 4: Baseline Demographics and CMR Parameters by Treatment Group

Parameter	Overall (n=80)	PVR Group (n=65)	Medical Follow-up (n=15)	p-value
Age (years)	14.5 ± 3.2	14.7 ± 3.1	14.1 ± 3.5	0.52
Male sex, n (%)	45 (56.25)	38 (58.5)	7 (46.7)	0.38
Time since surgery (years)	10.2 ± 4.5	10.8 ± 4.3	8.1 ± 4.9	0.03
RVEDVi (mL/m²)	128.6 ± 32.4	135.8 ± 30.1	106.7 ± 21.2	<0.001
RVESVi (mL/m²)	76.3 ± 22.1	80.4 ± 20.8	62.1 ± 18.5	0.002
RVEF	0.39 ± 0.11	0.36 ± 0.09	0.47 ± 0.06	0.002
PR fraction	0.36 ± 0.13	0.40 ± 0.10	0.25 ± 0.08	<0.001
LVEF	0.53 ± 0.06	0.52 ± 0.06	0.55 ± 0.05	0.09

Discussion

This study provides important insights into the long-term cardiovascular sequelae of repaired Tetralogy of Fallot in a Pakistani population, demonstrating the critical role of CMR in guiding clinical management. Our findings reveal a high prevalence of right ventricular dysfunction and severe pulmonary regurgitation, consistent with international cohorts but with some notable regional variations. The prevalence of severe pulmonary regurgitation (56.25%) in our study aligns with previous studies reporting rates of 50-70% in post-TOF

repair patients (11, 12). However, our observed mean RVEF of 39 ± 11% indicates more pronounced systolic dysfunction compared to Western cohorts, where RVEF typically ranges from 45-50% (13). This difference may reflect variations in surgical techniques, timing of intervention, or genetic factors specific to South Asian populations. The strong correlation between pulmonary regurgitation severity and right ventricular parameters ($p < 0.001$) reinforces established pathophysiological mechanisms where chronic volume overload leads to progressive ventricular remodeling (14, 15). Our finding that 87.5% of patients demonstrated right ventricular dilatation (mean RVEDVi 128.6 ± 32.4 mL/m²) is concerning, as RVEDVi >150 mL/m² is associated with reduced exercise capacity and increased arrhythmia risk (16). The temporal correlation between time since surgery and progressive dysfunction supports the concept of ongoing hemodynamic burden despite successful repair. Male patients exhibited significantly larger RVEDVi, consistent with previous observations suggesting sex-related differences in ventricular remodeling patterns (17). The high PVR recommendation rate (81.3%) with excellent guideline adherence (92.3%) demonstrates appropriate clinical integration of CMR findings. Our data support current guidelines emphasizing CMR-guided intervention timing, particularly the RVEDVi threshold of 150 mL/m² (18, 19). The universal PVR recommendation in patients with residual anatomical defects underscores the compounding effects of multiple hemodynamic burdens. The preserved left ventricular function across all patients contrasts with some studies reporting biventricular dysfunction, possibly reflecting differences in patient selection or surgical approaches (20). However, the observed biventricular correlation suggests ongoing monitoring remains essential.

Study Limitations

This single-center study may not represent the broader Pakistani TOF population. The cross-sectional design limits assessment of temporal changes, and lack of exercise testing or clinical outcomes reduces prognostic insights.

Future Recommendation

Future longitudinal studies with larger cohorts are needed to establish region-specific management protocols.

Conclusion

This study demonstrates that CMR provides comprehensive assessment of repaired Tetralogy of Fallot patients, revealing high prevalence of right ventricular dysfunction (mean RVEF 39 ± 11%) and severe pulmonary regurgitation (56.25%) in our Pakistani cohort. Strong correlations between pulmonary regurgitation severity and right ventricular parameters ($p < 0.001$) validate CMR as the gold standard for therapeutic decision-making. The high pulmonary valve replacement recommendation rate (81.3%) with excellent guideline adherence (92.3%) underscores CMR's clinical utility. These findings support standardized CMR protocols for long-term surveillance of repaired TOF patients and contribute valuable data on congenital heart disease outcomes in South Asian populations.

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Data acquisition, analysis and interpretation	HZ, HDW
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All the authors agree to take responsibility for every facet of the work, making sure that any concerns about its integrity or veracity are thoroughly examined and addressed.	