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The Conjoint Role of Echocardiography and Cardiac Magnetic Resonance Imaging in Follow up of Patients Post Tetralogy of Fallot Repair

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Abstract

Background: Tetralogy of Fallot is the most common form of cyanotic CHD. Surgical repair of TOF may be followed by various residual findings. CMR (Cardiac magnetic resonance) is the gold standard for evaluation of right ventricle (RV) volumes and quantification of degree of pulmonary regurgitation, meanwhile echocardiography represents the main line of follow up of these patients. Methods: This was a cross sectional observational study including 50 patients after TOF repair, presented to Ain Shams University Hospital for follow up, over 24 months. Transthoracic echocardiography (TTE) examination was done for RV linear diameters, RV function by fractional area change (FAC), tricuspid annulus plane systolic excursion (TAPSE), RV longitudinal strain, pulmonary regurgitation (PR) by diastolic flow reversal grading, Deceleration time (DT), PR jet width / pulmonary valve (PV) annulus ratio, and PR index (Time duration of PR/total diastole time) and full cardiac magnetic resonance for ten patients; measuring RV volumes, RV ejection fraction (EF) and PR fraction. Results: The study included 50 patients post TOF surgical repair, 26 (52%) males and 24 (48%) females, with a mean age of 11.88 years. Mean RV FAC by TTE was 51%, TAPSE mean value was 15, and mean GLS of RV was -19. The residual peak PG across the RVOT mean value was 35mmHg. Thirty-two of our patients (64%) had severe PR by diastolic flow reversal, fourteen patients (28%) had moderate to severe PR, and four patients (8%) had moderate PR. In the ten patients who had CMR, the mean RVEF was 50%, and the PR fraction mean value was 54%. There was a strong correlation between the RV diameters measured by TTE and RV volumes measured by CMR, accordingly a regression analysis equation to calculate RV volumes from a given RV diameter measured by TTE can be done. Conclusion: The follow up post TOF repair should be directed towards early and accurate assessment of post repair sequel and defining the intervention threshold. Multimodality imaging provides more accurate and practical protocol. The echocardiographic assessment can be used as a triage to decide who will benefit from expensive and not readily available CMR.

Keywords

Fallot Tetralogy, Right Ventricle, Pulmonary Regurgitation, Cardiac Magnetic Resonance Imaging

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1. Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease. Total repair for TOF has been available for 50 years, with a favorable outcome in most patients [1]. Survivors of TOF repair constitute a large and growing population of patients. Although postsurgical outcome is generally favorable, as these patients move into

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adulthood, late morbidity is becoming more prevalent and the notion that TOF has been "definitively repaired" is increasingly being challenged [2].

Surgical repair of TOF may be followed by various conditions and residual findings, early postoperatively or late during follow-up. Most of these conditions affect the right ventricular outflow tract and the pulmonary arteries and thus, indirectly, the right ventricle (RV) leading to RV dilatation and dysfunction, atrial and ventricular arrhythmias, congestive heart failure, and sudden death [3]. Pulmonary regurgitation (PR), the most common sequel of trans-annular or RV outflow tract patch repair, is present in nearly all TOF patients. Chronic PR is generally well tolerated; however, the evidence is beginning to point to PR as an important contributing cause of long term morbidities, including atrial and ventricular arrhythmia, RV dilatation, and, possibly, sudden death [4].

Reliable serial follow up techniques after repair can help determine when to intervene to preserve ventricular function and improve long-term outcome. No single modality is able to delineate all aspects of the intracardiac and extracardiac anatomy and evaluate hemodynamic consequences of TOF repair [5]. To date, CMR (cardiac magnetic resonance) is the gold standard for evaluation of RV volumes and quantification of degree of pulmonary and tricuspid regurgitation (TR). Meanwhile transthoracic echocardiography (TTE) still represents the main line of diagnosis and follow up of patients with congenital heart disease as it is readily available at an affordable cost [6].

2. Methods

This study included 50 patients who underwent total surgical repair for TOF, presented to the Congenital and structural heart disease Unit - Cardiology Department at Ain Shams University Hospitals during the years 2016-2017, and had more than mild residual PR by TTE during routine follow up after repair

Exclusion criteria: patients with any congenital heart disease other than isolated TOF, patients with prosthetic valves, critically ill patients e.g. end stage decompensated heart failure, patients with claustrophobia (in adult patients), patients with non-MRI compatible Permanent Pace Maker, patients with severe renal impairment (creatinine clearance below 30 ml/min), and any uncooperative patient who would need general anesthesia and had a contraindication or was high risk for it.

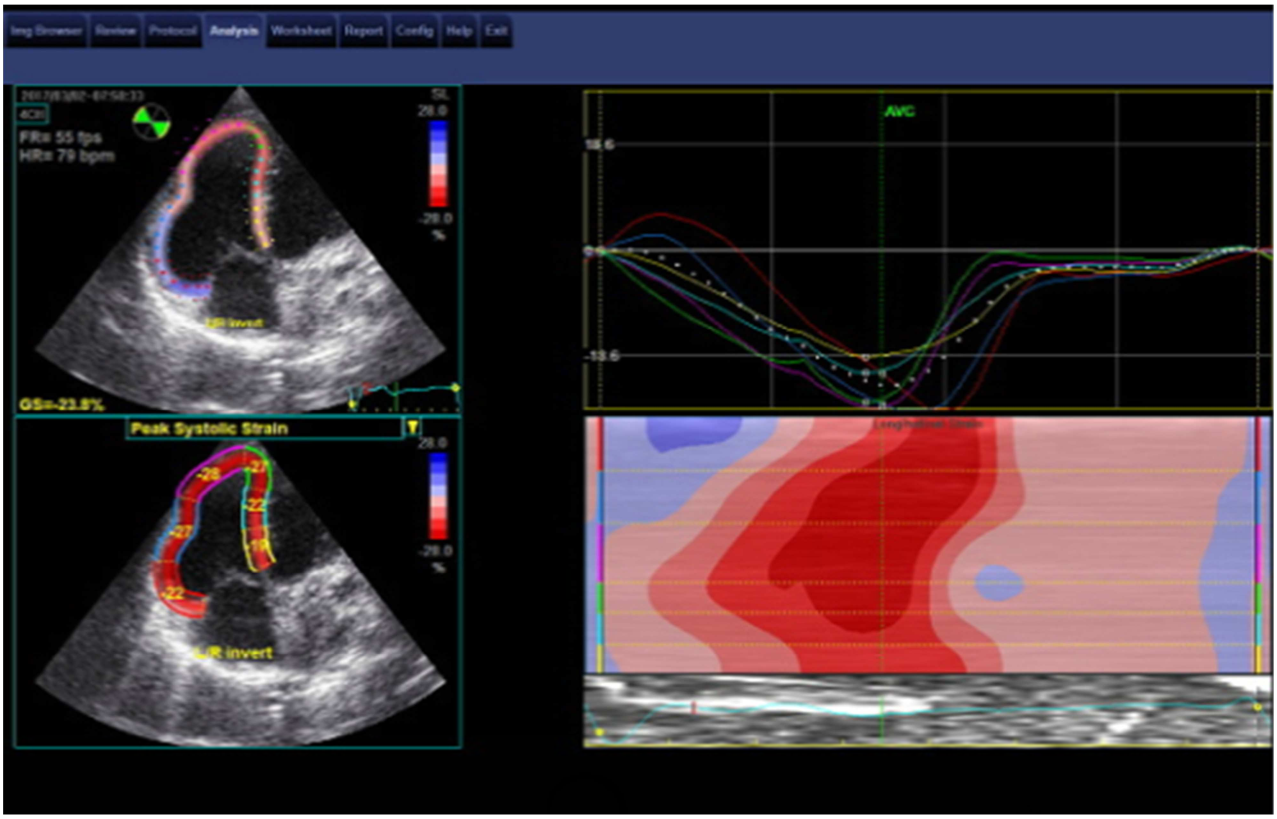

Figure 1. RV Longitudinal strain analysis.

RV right ventricle

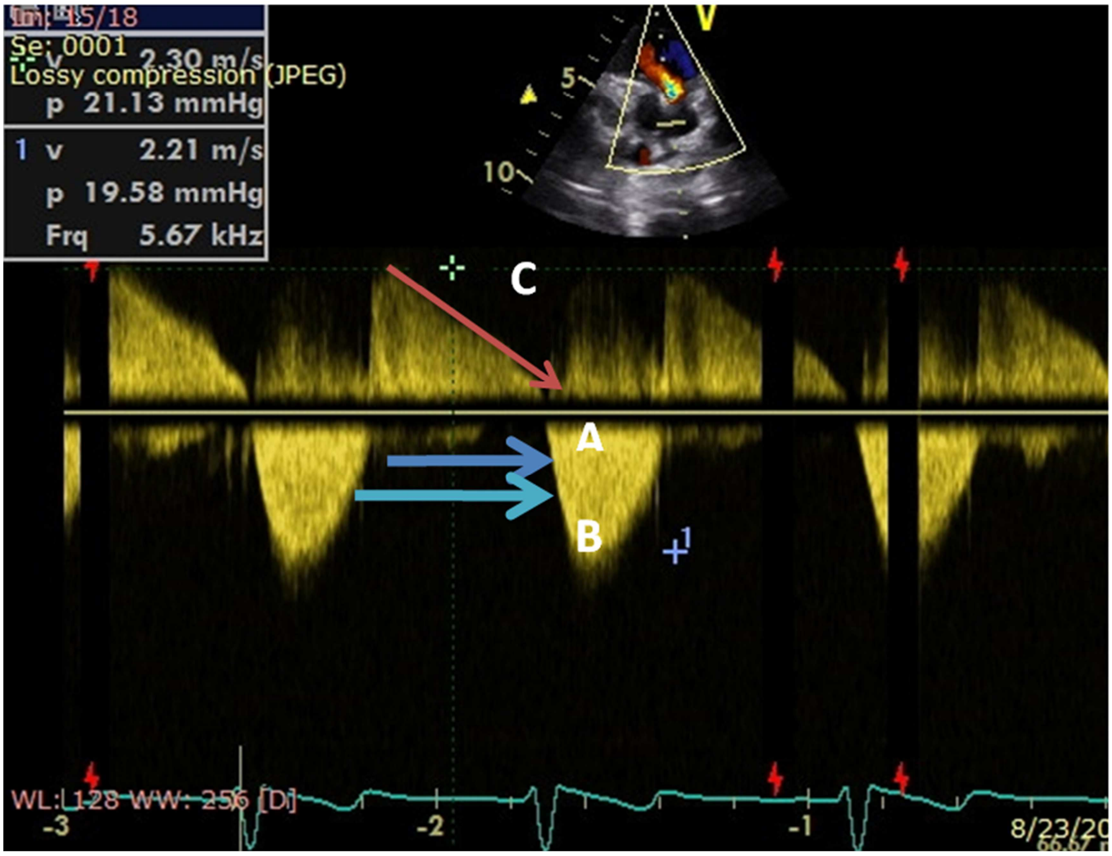

Figure 2. The figure shows CW Doppler of the PV flow, [A] represents the measurement of the duration of the PR signal, [B] represents the total duration of diastole, and [C] represents the measurement of the deceleration time of the PR spectral Doppler signal.

CW continuous wave, PV pulmonary valve, PR pulmonary regurgitation

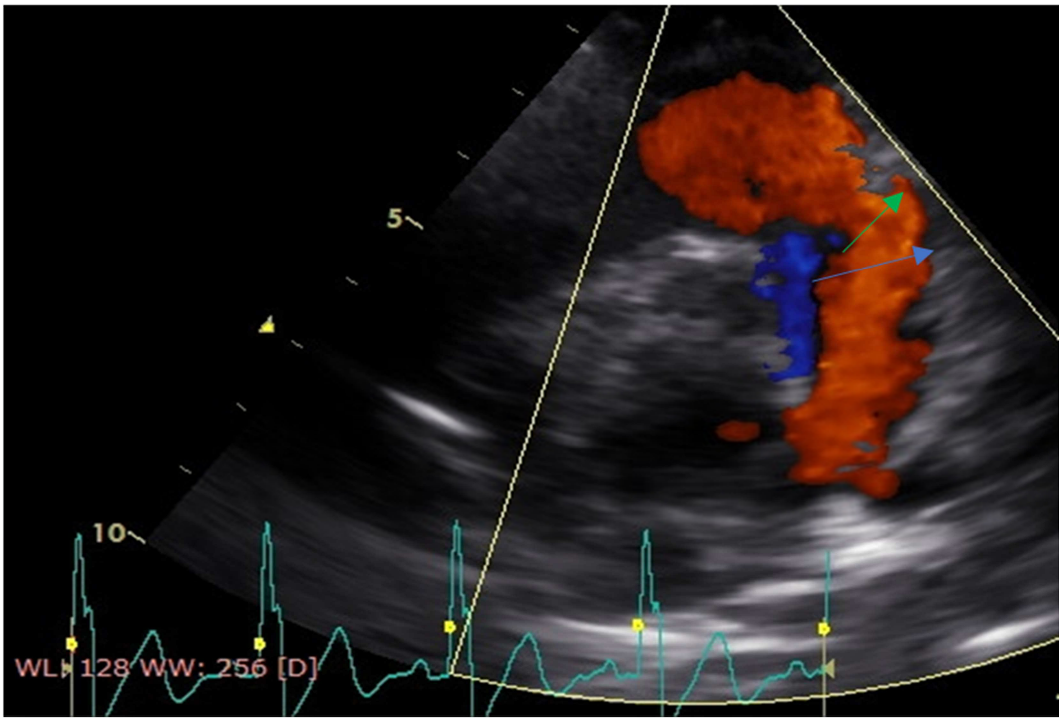

Figure 3. The figure shows parasternal short axis view at the level of PA with color Doppler flow of the PA showing diastolic flow reversal of the PR with the green arrow represents the measurement of the PR jet width and the blue arrow represents the measurement of the PV annulus.

PA pulmonary artery, PR pulmonary regurgitation, PV pulmonary valve

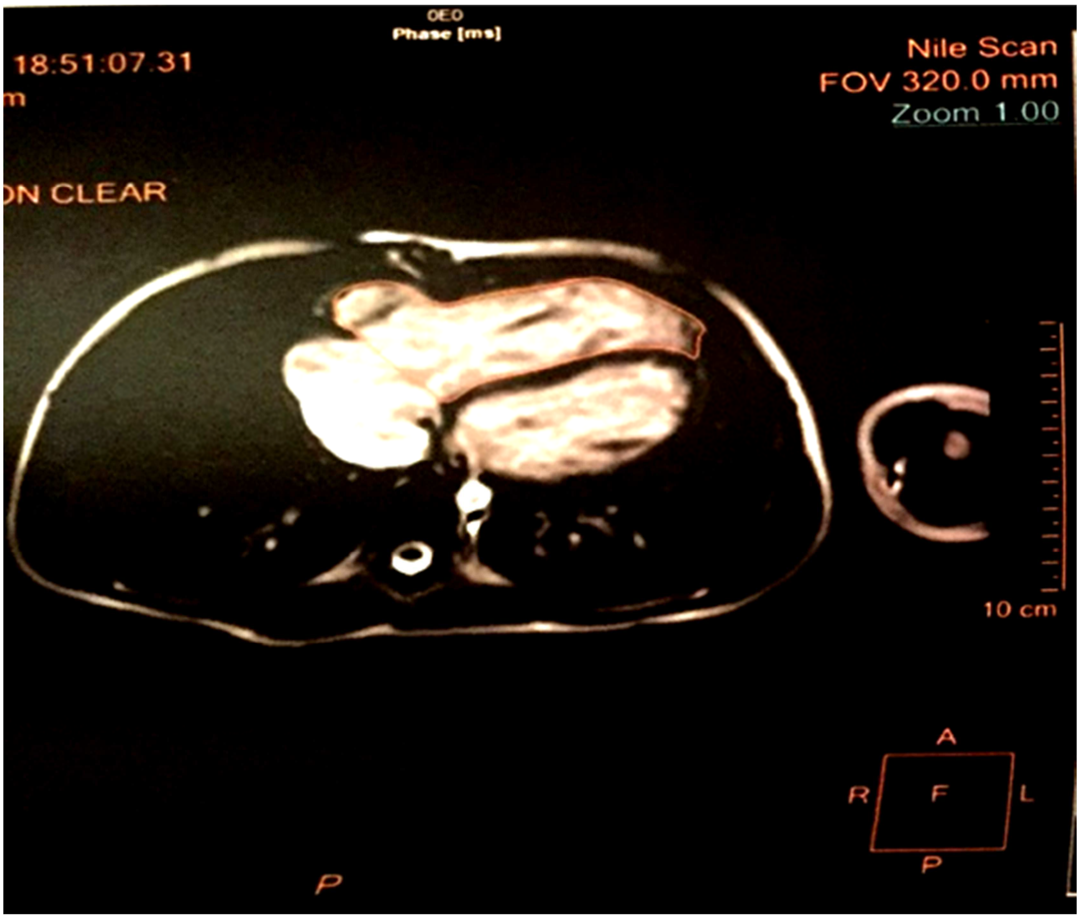

Figure 4. Showing modified axial sequence for tracing the RV myocardium at different levels for calculation of the RV volume and function.

RV right ventricle

Figure 5. Shows a modified RAO view showing a dilated RVOT.

RAO right anterior oblique, RVOT right ventricular outflow tract

This was a cross sectional observational study included 50 patients with total TOF repair, all patients were subjected to thorough history taking with particular stress on age at time of surgery, NYHA class, and revising the previous procedures' reports and documents for the surgical details and type of repair, thorough clinical examination, twelve-lead surface electrocardiogram, and standard TTE including RV size assessment by measurement of RV linear diameters [longitudinal, transverse mid RV, and basal diameters], right ventricle (RV) function assessment by fractional area change (FAC), tricuspid annular plane systolic excursion (TAPSE) and RV global longitudinal strain (GLS) measurement (figure 1) [7, 8], and pulmonary regurgitation (PR) assessment by diastolic flow reversal grading, Deceleration time (DT) (figure 2), PR jet width / pulmonary valve (PV) annulus ratio (figure 3), and PR index (Time duration of PR/total diastole time) and full CMR examination was conducted to ten patients who were sought to be in need for intervention post TOF repair based on the TTE assessment; including RV volumes measurement [end diastolic volume (EDV), and end systolic volume (ESV)], RV ejection fraction (EF) measurement (figure 4) [9, 10], RVOT diameter measurement (figure 5) [11], and PR fraction (PRF) measurement [12]. Data from echocardiography and CMR for the ten patients who were offered CMR were compared to each other.

The study was done after approval of ethical board of our university and an informed written consent was taken from each participant in the study.

3. Results

The present study included 50 patients with TOF post total surgical correction, 44 (88%) pediatric patients below the age of 18 years and 6 (12%) adult patients, 26 (52%) males and 24 (48%) females. The youngest patient was 4-year-old and the oldest was 40-year-old with a mean age of 11.88 years (table 1).

Table 1. Demographic characteristics of the study group.

		No. = 50
Age (years)	Mean±SD	11.88 ± 7.44
	Median (IQR)	10.0 (6 – 16)
	Range	4 – 40
Gender	Male	26 (52%)
	Female	24 (48%)
Weight (kg)	Mean±SD	41.00 ± 18.25
	Range	17 – 90
Height (cm)	Mean±SD	122.12 ± 38.14
	Range	65 – 175
BSA m^2	Mean±SD	1.180 ± 0.419
	Range	0.494 – 2.056

BSA: body surface area

All of them had full TTE assessment and ten of them had CMR. RV longitudinal, transverse mid RV, and basal diameters mean values were 61mm, 31mm, and 31mm, respectively. RV FAC by TTE mean value was 51%, TAPSE mean value was 15 (table 2), and GLS of RV mean value was -19. The RVOT dimeters in the parasternal long axis, parasternal short axis and subcostal RV inflow-outflow views mean value were 25mm, 22mm, and 19mm, respectively. The residual peak PG across the RVOT mean value was 35mmHg (table 2). Thirty-two patients (64%) had severe PR by diastolic flow reversal, 14 patients (28%) had moderate to severe PR, and 4 patients (8%) had moderate PR. PR to PV annulus ratio mean value was 0.7, deceleration time of PR Doppler signal mean value was 346m sec., PR index mean value was 0.9. In the ten patients who had CMR, the EDV, and ESV mean values were 227ml, and 115ml, respectively, the RV EF by CMR mean value was 50%, and the PR fraction mean value was 54% (table 3). There was non-significant difference between the RVOT diameters measured by TTE in the parasternal short axis and subcostal inflow-outflow views and the RVOT diameters measured by CMR. There was a strong correlation between the RV diameters measured by TTE and RV volumes measured by CMR (table 4), according to which a regression analysis equation to calculate RV volumes from a given RV diameter measured by TTE can be done (table 5)

Regression analysis equations:

RV EDV = 68.054 - (4.444 x RV longitudinal diameter)

RV EDV = 72.715 + (4.010 x RV transverse diameter)

RV EDV = 21.985 + (5.822 x RV basal diameter)

RV ESV = 89.646 - (3.076 x RV longitudinal diameter)

RV ESV = 9.048 - (3.211 x RV transverse diameter)

RV ESV = 29.744 - (4.098 x RV basal diameter)

Table 2. Descriptive analysis of RV size, functions and RVOT Diameters data.

Echocardiography data		No. = 50
RV size		
Longitudinal diameter		
Mean±SD		60.90 ± 15.36mm
Range		40-96mm
Long. Diam. i		
Mean±SD		55.1 ± 13.6mm/m^2
Range		29.2 – 88.6mm/m^2
Transverse diameter		
Mean±SD		31.74 ± 11.16mm
Range		19 – 70mm
Transv. Diam. i		
Mean±SD		28.4 ± 76mm/m^2
Range		11.8 – 45.7mm/m^2
Basal diameter		
Mean±SD		31.70 ± 10.41
Range		17 – 70mm
Basal diam. i		
Mean±SD		28.3 ± 71mm/m^2
Range		11.8 – 46.2mm/m^2
RV function		
FAC		
Mean±SD		51.64 ± 11.80
Range		25 – 78%
TAPSE		
Mean±SD		15.22 ± 4.26
Range		5 – 27
RVOT		
Parasternal Long axis	Mean±SD	25.12 ± 9.10
	Range	10 – 40mm
Parasternal Short axis	Mean±SD	21.72 ± 9.17
	Range	9 – 45mm
Inflow-outflow	Mean±SD	18.86 ± 7.42
	Range	5 – 35mm
PG	Mean±SD	34.98 ± 21.48
	Range	5 – 85mmHg

RV: right ventricle, RVOT: right ventricular outflow tract, FAC: fractional area change, TAPSE: tricuspid annular plane systolic excursion, Long. Diam.i: Longitudinal RV diameter indexed to body surface area, Transv. Diam.i: Transverse diameter indexed to body surface area, Basal diam. I: Basal diameter indexed to body surface area, PG: pressure gradient across the RVOT

Table 3. Descriptive analysis of RV volumes, functions and PR fraction by CMR.

RV	No. = 10
EDV	
Mean±SD	227.50 ± 114.33ml
Range	83 – 459ml
EDV i	
Mean±SD	150.5 ± 42.0ml/m^2
Range	90.5 – 220.7ml/m^2
ESV	
Mean±SD	114.90 ± 75.38ml
Range	26 – 290ml
ESV i	
Mean±SD	75.4 ± 35.3ml/m^2
Range	29.5 – 139.4ml/m^2
SV	
Mean±SD	107.40 ± 48.5ml
Range	50 – 19ml
EF	
Mean±SD	50.80 ± 10.21%
Range	36 – 68%
PR fraction	54.10 ± 14.65

EDV: end diastolic volume, ESV: end systolic volume, EVD i: End diastolic volume indexed to body surface area, ESV i: End systolic volume indexed to body surface area, SV: stroke volume, EF: ejection fraction, PR; pulmonary regurgitation

Table 4. RV diameters by TTE and RV volumes by CMR.

RV	RV EDV		RV ESV	
	r	p-value	r	p-value
RV longitudinal diameter	0.688*	0.028	0.722*	0.018
RV transverse diameter	0.635*	0.049	0.771**	0.009
RV Basal diameter	0.645*	0.044	0.689*	0.028

RV: right ventricle, EDV: end diastolic volume, ESD: end systolic volume

Table 5. Regression analysis of the correlations between RV diameters by TTE and RV volumes by CMR.

RV EDV	Unstandardized Coefficients		Standardized Coefficients	t	Sig.
	B	Std. Error	Beta		
RV longitudinal diameter	4.444	1.657	0.688	2.683	0.028
RV transverse diameter	4.010	1.726	0.635	2.324	0.049
RV Basal diameter	5.822	2.439	0.645	2.388	0.044

RV ESV	Unstandardized Coefficients		Standardized Coefficients	t	Sig.
	B	Std. Error	Beta		
RV longitudinal diameter	3.076	1.041	0.722	2.955	0.018
RV transverse diameter	3.211	0.938	0.771	3.425	0.009
RV Basal diameter	4.098	1.526	0.689	2.686	0.028

RV: right ventricle, EDV: end diastolic volume, ESD: end systolic volume

4. Discussion

In the current study, 50 patients with repaired TOF were included, with a mean age of 11.88±7.44 years, all our patients had transannular patch, Chaowalit et al. included 45 patients with mean age 27.4 ± 11.2 years (21 of the included patients had trans-annular patch), It was noticed that although the mean age of Chaowalit et al. [13] study group is older than ours, the mean values for RVOT diameters in this study are slightly higher than the mean values of RVOT diameters of Chaowalit et al., which were 17.9±4mm/m^2 for the patients with non-trans-annular patch repair and 17.8± 4mm/m^2 for trans-annular patch repair patients

Chaowalit and his colleagues concluded that there was a strong correlation between EDDi and ESDi measured by TTE and EDVi by CMR with P value <0.0001, which provided an indicator of significant RV dilatation in patients with repaired TOF with a predictive accuracy of positive and negative results 85.0% and 52.4% for RV ESDi ≥19.4 mm/m^2, and 87.5% and 64.7% for RV EDDi ≥24.5 mm/m^2 respectively. That was similar to the strong correlation found between the three RV diameters measured by TTE and the RV ESV and EDV by CMR in the ten patients who had CMR with P value <0.05. According to this correlation a regression analysis derived equation for calculating RV EDV and ESV from a given longitudinal, transverse or basal RV diameter can be provided.

By correlating the results of the four parameters used in the current study for assessment of PR by TTE to the standard method of PR assessment; PRF acquired by CMR, in the ten patients had CMR. None of the four parameters had significant correlation to the PRF, specially PR index and diastolic flow reversal which their statistical correlation to the PRF didn't result in a correlation factor at all, that was partly not consistent with what Agha et al [14] found when they included 50 patients, twenty-five of them were asymptomatic children after TOF repair (9.2 ± 4 years) and were compared to 25 age matched healthy children. They found that PR index and no flow time measured by TTE had a strong correlation to PRF measured by CMR with a conclusion of cutoff values for significant PR, PR index of <0.8 has sensitivity of 86.36% and specificity of 100% (AUC=0.924) and no flow time of >64 m sec has sensitivity of 81% and specificity of 100% (AUC=0.894) in identifying significant pulmonary regurgitation

The explanation for this result was that the 10 patients who were referred to have a CMR in our study, all had severe PR with the same value for diastolic flow reversal which is 4/4 and the same PR index which is 1, while the study population of Agha et al. had different degrees of PR 3 patients (12%) had mild PR with PRF <20%, 17 patients (68%) had moderate PR with PRF 20-40% and only 5 patients (20%) had severe PR with PRF >40% and all of them had CMR, which gave a spectrum for creating significant correlations

Del Toro et al. [15] carried out a study which included 57 patients with repaired TOF with a mean age of 13.0±3.6 years. Traditional echocardiographic function parameters and RV GLS were compared to CMR-derived RV EF, then subjects were divided into two groups based on CMR RV EF (group I: RV EF ≥45%; and group II: RV EF <45%). The mean RV GLS was significantly abnormal in group II (−15.3±3.8%) compared to group I (−20.9±3.3%; P<.001).

Yet in the current study, there was no correlation between RV EF by CMR and RV GLS. The suggested explanation is that patients' selection for CMR was based on the degree of PR and RVOT dilatation rather than impaired RV function

5. Conclusion

The follow up of post TOF patients should be directed towards early detection, accurate assessment of the post repair sequel and defining the threshold for intervention. Multimodality imaging approach of follow up for such patients provides more accurate and practical protocol for follow up. CMR will remain the gold standard for assessment of RV function and PR severity and its role in the follow up of patients post TOF repair will remain un-exchangeable, however the echocardiographic assessment of RV dimensions can be used as an indicator for triage of these patients and deciding who will benefit from this expensive and not readily available technique

Statistical Analysis

Results were analyzed by statistical package for social sciences (SPSS) Data were expressed as mean ± SD and percentage.

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