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Arrhythmias in repaired pediatric and adolescent Fallot tetralogy, correlation with cardiac MRI parameters

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Abstract

Background: Tetralogy of Fallot (TOF) is a common congenital cyanotic heart disease in which arrhythmias could develop even after successful operative repair. Pulmonary regurgitation and right ventricular dilatation develop in many cases. The relation between arrhythmias and right ventricular dilatation is not established. Our aim is to assess the relation in between the degree of right ventricular volume affection and the severity of the pulmonary regurgitation, associated arrhythmias and the need for pulmonary valve repair in Egyptian pediatric and adolescent cases after successful TOF repair.

Results: A cross sectional descriptive study was conducted on 32 cases after successful surgical repair. Transthoracic Doppler echocardiography, 24 h Holter monitoring and cardiac MRI for assessment of pulmonary regurgitation fraction (PRF), ventricular volumes and function were measured. Cases were classified according to right ventricular end diastolic volume index (RVEDVI) into 2 groups with cut off value 150 ml/m². Mean age of the studied cases was (12.96 ± 3.384) years, mean age at time of surgical repair was (34.23 ± 22.1) months, and mean duration postoperatively was (121.72 ± 41.028) months. Eighteen cases (56%) had RVEDVI ≥ 150 ml/m², PRF was significantly higher in cases with increased RVEDVI (*p* value 0.007), with positive significant correlation between RVEDVI and PRF (*p* value = 0.0001, *r* = 0.61). Arrhythmias were detected in 18 cases (56%), the most common of which was infrequent supraventricular ectopy. No significant difference in incidence of arrhythmias between the 2 groups (*p* value = 1) with also no significant correlation between arrhythmias and increased RVEDVI (*p* value = 0.76, *r* = 0.05). No difference between cases with and without arrhythmias regarding RVEDVI (*p* value = 0.56) or PRF (*p* value = 0.5).

Conclusion: Holter detected arrhythmias after successful surgical repair of TOF were significantly associated with increased postoperative duration but not with PRF or RVEDVI.

Keywords: TOF, Arrhythmias, Cardiac MRI, RVEDVI, PR fraction

Background

The surgical techniques used to relieve right ventricular outflow tract (RVOT) obstruction in surgical repair of tetralogy of Fallot (TOF) were found to result in pulmonary regurgitation (PR), with subsequent increase in

right ventricular (RV) volume [1]. Chronic hemodynamically relevant PR resulting in important right ventricular dilation and ventricular dysfunction can result in late adverse clinical outcomes, including exercise intolerance, arrhythmias, heart failure and/or death accelerated in the third decade of life [2]. Ventricular and supraventricular arrhythmias were reported long time ago to occur in cases of TOF even after successful surgical repair, and could lead to sudden death [3].

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Cardiac magnetic resonance (CMR) imaging is considered an accurate and reproducible technique for assessing RV size and severity of PR [4]. The impact of abnormal RV mechanics on the left ventricle (LV) has been appreciated, with the assessment of LV function assuming higher priority in decision-making regarding possible pulmonary valve replacement (PVR) [5]. It was suggested that with aging, the increase for PVR will increase to overcome the problem of progressive RV dilatation with subsequent arrhythmias [5].

In the presence of PR, the presence of clinical symptoms clearly corresponds to class I evidence of indication for pulmonary valve (PV) implantation in both the European [6] and North-American guidelines [7].

In asymptomatic patients the indications for PV implantation criteria, and particularly the choice of the best timing, are less clearly defined [8]. General agreement exists on the indication in asymptomatic patients in the presence of any of the following criteria, as judged by echocardiography and/or magnetic resonance imaging: a. PV regurgitation > 20%. b. Indexed end-diastolic right ventricular volume > 120–150 ml/m². Indexed end-systolic right ventricular volume > 80–90 ml/m² [9].

We aimed to assess the degree of RV volume affection in relation to severity of PR, associated arrhythmias and the need for PVR in Egyptian pediatric and adolescent cases after successful TOF repair.

Methods

A cross sectional descriptive study was conducted in the postoperative cardiac surgery clinic, at our University. It was done during the period from August 2016 to December 2018 after the approval of the research ethic committee of the Radiology and Pediatric departments at our institute. Surgical repair was done in the period from 2000 to 2017. Thirty-two cases of successfully repaired TOF were included in this study. Those with pre-operative arrhythmias, a residual ventricular septal defect (VSD) or significant pulmonary stenosis (PS) (PG ≥ 40 mmHg) and those with permanent pacemaker were excluded.

Clinical examination (including weight and height and calculation of body surface area) was done using Mosteller formula $BSA(m^2) = \sqrt{Wt(Kg) \times Ht(cm) / 3600}$ [10].

A written informed consent to participate in this study was given by all patients or the parents or legal guardians for the patients less than 16 years old or unconscious patients at the time of the exam.

Symptoms suggestive of arrhythmias as chest pain, palpitation and syncopal attacks, and those suggestive of exercise intolerance were asked about. Routine laboratory investigations as CBC, serum sodium and potassium were requested.

Transthoracic color doppler echocardiography

Using GE Vivid S5 machine was carried out for all children. We retrieved the measurements of left ventricular end diastolic diameter (LVEDD), left ventricular end systolic diameter (LVESD), fractional shortening (FS), M-mode was used to assess the previous measurements. Assessment of degree of PR using color Doppler echocardiography was done (Grade 1 if less than 20%, Grade 2 if 20–40% and Grade 3 if more than 40%).

Subcostal view and other standard views as parasternal, suprasternal and apical views were used in chamber morphologies and function assessment with the color flow Doppler was used to provide additional hemodynamic information.

Holter monitoring

24 h Holter monitoring was performed for all cases. Significant premature ventricular contractions (PVCs) were defined as presence of PVCs greater than 10% of the total QRS complexes [11]. Clinically important arrhythmias as sick sinus syndrome, 2nd or 3rd degree atrioventricular block, atrial tachyarrhythmias, premature ventricular beats more than couplet, and non-sustained or sustained ventricular tachycardia were assessed using standard methods [12].

Cardiac functional MRI

Magnetic resonance imaging studies were performed using 1.5 Tesla Philips systems [Gyrosan Intera (1.5 T) whole-body MRI scanner] by using a phased-array cardiac coil.

Children above 4 years tolerated the examination without sedation. They were given instructions to avoid any movement during the procedure. Infants and younger children were sedated by oral chloral hydrate (75 mg/kg body weight), they were wrapped in a blanket and placed on a thin, firm mattress.

C1 coil was used for infants while older children used a synergy body coil. All patients were examined in supine position, head first. ECG leads and PEAR (phase encoding artifact reduction) sensors were applied.

The protocol included ECG-gated steady-state free precession (SSFP) imaging sequences with retrospective gating (TE 1.5–2.0 ms, TR 2.8–4.0 ms, flip angle 45°, views per segment 10–20, 30 reconstructed images per cardiac cycle) where a stack of 12 contiguous short-axis slabs perpendicular to the long axis of ventricles (slice thickness 6–8 mm, interslice distance 0–2 mm) was obtained assuring full coverage of ventricles. The axial planes were used for the evaluation of

the anatomy while ventricular volumes were assessed in both axial and short axis planes.

Images were interpreted using dedicated software (ViewForum, Philips Medical Systems, Best, The Netherlands) by two radiologists (9- and 10-years' experience) being blind to the results of the echocardiography as well as to each other results. Ventricular volumes were calculated using manual contour tracing of short-axis slices from the apex to the base (excluding papillary muscles and trabeculations) to obtain end-diastolic and end-systolic volumes. The stroke volume (SV) was calculated by subtracting the end systolic volume (ESV) from the end diastolic volume (EDV). The ejection fraction (EF) was calculated by dividing the SV by the EDV.

Assessment of pulmonary regurgitation fraction (defined as the retrograde pulmonary flow/ ante grade pulmonary flow $\times 100\%$) was performed using through plane phase contrast sequences [13].

Right ventricular end diastolic volume was indexed to body surface area (RVEDI). Value ≥ 150 ml/m² was used as cut off value as it was reported that pulmonary valve replacement should be done even in asymptomatic patients with RVEDI ≥ 150 ml/m² to prevent irreversible RV dysfunction [13] (Figs. 1, 2, 3). According to this value cases were classified into 2 groups, group1: cases with RVEDI less than 150 ml/m² and group 2: cases with RVEDI more than or equal to 150 ml/m² and comparison between the 2 groups was performed.

Statistical analysis

Statistical calculations were done using computer program IBM SPSS (Statistical Package for the Social Science; IBM Corp, Armonk, NY, USA) release 20 for Microsoft Windows. Data were statistically described in terms of mean \pm standard, median and range, or frequencies (number of cases) and percentages when appropriate. Comparison of numerical variables between the study groups was done using Student-*t* test for

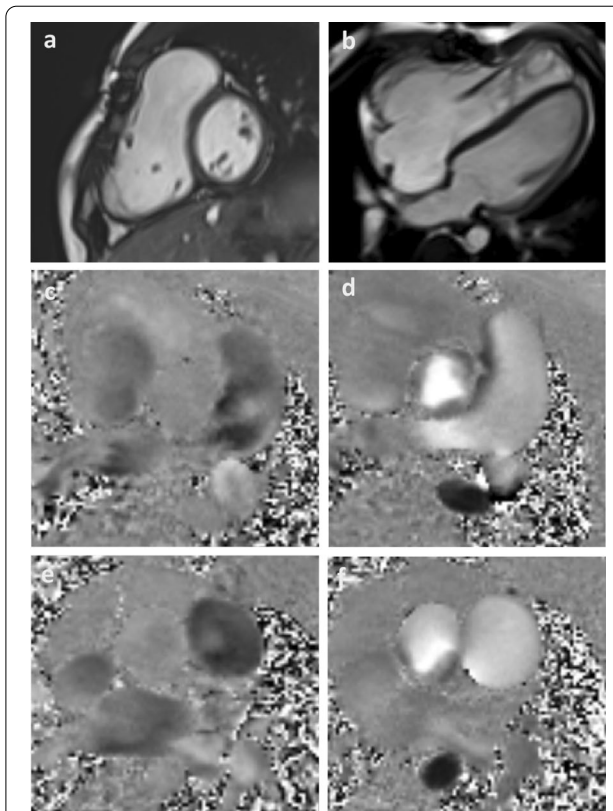


Fig. 1 Male child 16 year old came for follow after 10 years post TOF repair, Holter showed frequent PVCs, CMR showed marked RV dilatation (RVEDI = 170 ml/m², mildly impaired RV systolic function EF = 44%, and free PR (50%), **a** short axis SSFP view showing marked RV dilatation, **b** axial SSFP view showing marked RV dilatation, **c** and **d** in-plane phase contrast images showing backward (**c**) and forward (**d**) blood flow through the pulmonary artery, **e** and **f** through-plane phase contrast images showing backward (**e**) and forward (**f**) blood flow through the pulmonary artery

independent samples in comparing 2 groups of normally distributed data/large samples. For comparing categorical data, Chi-square test was performed. Exact test was

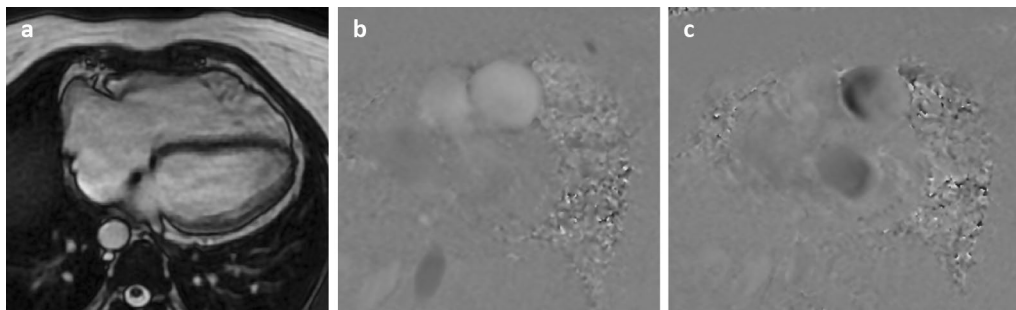


Fig. 2 Male child 12 years old came for follow after 8 years post Fallot repair, Holter showed infrequent PVCs, CMR showed mild RV dilatation (RVEDI = 132 ml/m², fair RV systolic function EF = 49%, and mild PR (22%), **a** axial SSFP view showing RV dilatation, **b** and **c** through-plane phase contrast images showing forward and mild backward blood flow through the pulmonary artery

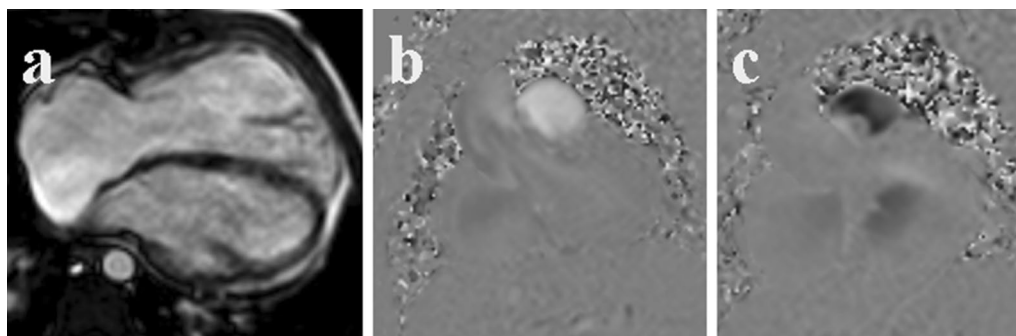


Fig. 3 Female child 9 years old came for follow after 7 years post Fallot repair, Holter showed no evidence of arrhythmia, CMR showed moderate RV dilatation (RVEDVI = 149 ml/m², fair RV systolic function EF = 46%, and moderate PR (35%), **a** axial SSFP view showing RV dilatation, **b** and **c** through-plane phase contrast images showing forward and backward blood flow through the pulmonary artery

used instead when the expected frequency is less than 5. Correlation between various variables was done using Pearson moment correlation equation for linear relation

Table 1 Comparison between cases with and without increased RVEDVI

Parameter	Group 1 RVEDVI < 150 (N°=14) mean ± SD	Group 2 RVEDVI ≥ 150 (N°=18) mean ± SD	p Value
Age (year)	13.29 ± 3.7	14.44 ± 5.6	0.5
Age at surgery (mo)	40 ± 26.9	29.47 ± 16.5	0.22
Duration (mo)	119.4 ± 49.7	123.5 ± 34.2	0.7
Weight (kg)	44.16 ± 18.1	36.19 ± 11.3	0.16
Weight centile	34.14 ± 26.2	27.47 ± 20.7	0.45
Height (cm)	145.14 ± 27.1	143.17 ± 17.5	0.81
Height centile	36.50 ± 31.61	28.35 ± 22.1	0.42
Na	138.86 ± 8.8	137.00 ± 5.2	0.64
K	4.11 ± 0.5	4.40 ± 0.57	0.32
Hb	12.40 ± 1.2	11.70 ± 1.37	0.18
WBCs	8.45 ± 3.2	8.67 ± 9.69	0.93
Platelets	306.4 ± 117.6	298.33 ± 69.1	0.89
<i>Echocardiographic findings</i>			
LVEDD	36.25 ± 6.7	30.01 ± 16.2	0.1
FS	39.98 ± 9.1	36.25 ± 7.5	0.28
<i>MRI findings</i>			
RVEF (%)	47.5 ± 6.6	56.86 ± 6.4	0.0001*
RVESVI (ml/m ²)	77.6 ± 29.2	82.7 ± 40.9	0.6
RVEDVI (ml/m ²)	112.21 ± 26.4	195.50 ± 54.9	0.0001*
LVEDVI (ml/m ²)	90.3 ± 41.58	102.6 ± 27.2	0.3
RVEDV/LVEDV	1.8664 ± .37	1.9112 ± .36	0.7
LVEF	61.6 ± 4.8	55.8 ± 4.7	0.002*
PR fraction	36.22 ± 7.4	47.72 ± 14.2	0.007*

FS fractional shortening, LVEDD left ventricular end diastolic diameter, RVEF right ventricular ejection fraction, RVESVI right ventricular end diastolic volume index, LVEDV left ventricular end diastolic volume, LVEF left ventricular ejection fraction, PR fraction pulmonary regurgitation fraction

*p Value is significant if less than 0.05

of normally distributed variables and Spearman rank correlation equation for non-normal variables/nonlinear monotonic relation. *p* values less than 0.05 was considered statistically significant.

Results

The mean age of the studied cases at the time of the study was (12.96 ± 3.384) years [155.6 ± 40.6 months], ranging from 4 to 21 years, with mean age at time of surgical repair was (34.23 ± 22.1) months, median 24 months, range 12–84 months and mean duration of follow-up postoperatively was (121.72 ± 41.028) months, range 30–192 months (2.5–16 years). Ten females (31.25%) and 22 males (68.75%) were included in this study. Although weight and height centiles were less in those with increased RVEDVI, the difference did not reach a statistical significance (*p* value 0.45 and 0.42 respectively) (Table 1).

Color Doppler echocardiography revealed grade 3 PR in 23 cases, grade 2 in 8 cases and grade 1 in one case.

A positive significant correlation between PRF and RVEDVI was detected (*p* value: 0.0001, *r*: 0.61) (Fig. 4).

No significant correlation between PRF and RVEF (*p* value: 0.07, *r*: −0.33). Negative significant correlation between RVEDVI and LVEDVI (*p* value: 0.001, *r*: −0.59), but not with FS (*p* value: 0.4, *r*: −0.15). LVEF was significantly lower in cases with increased RVEDVI than those without increased RVEDVI (*p* value: 0.002). Positive significant correlation between RVEF and LVEF (*p* value: 0.0001, *r*: 0.56).

Twenty-four hours Holter monitoring detected arrhythmias in 18 cases (56%) while it was normal in 14 cases. Holter detected arrhythmias are demonstrated in Table 2.

No significant correlation between arrhythmia and increased RVEDVI (*p* value: 0.76, *r*: 0.05).

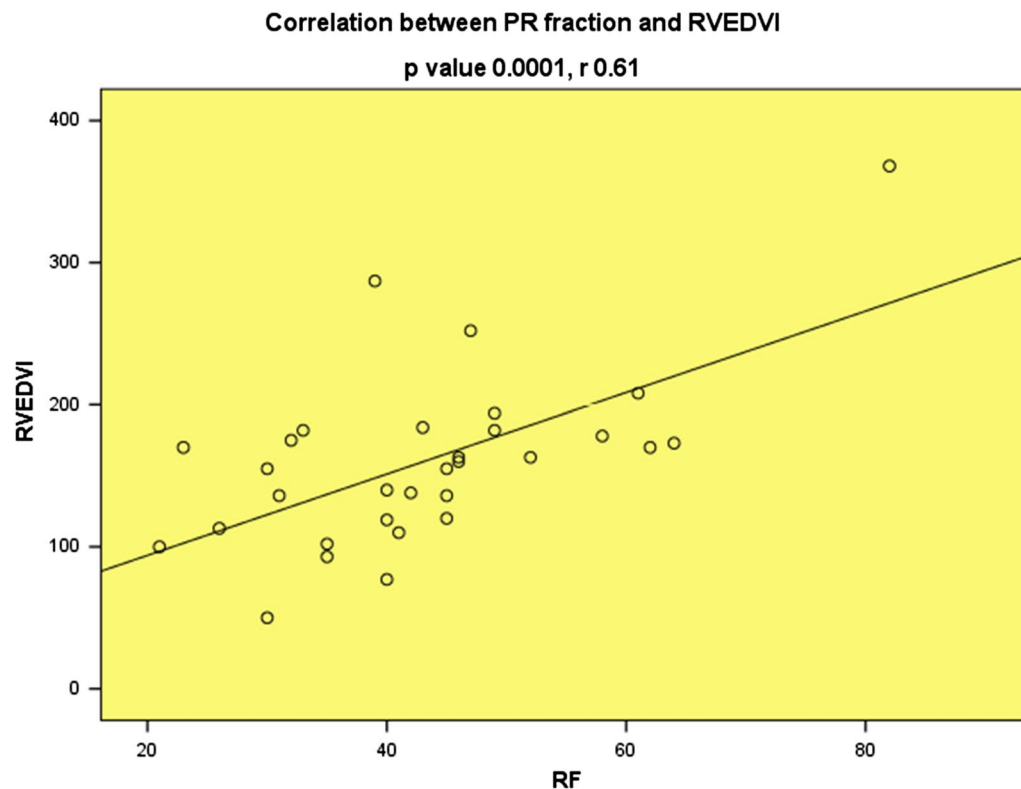


Fig. 4 Correlation between RF and RVEDVI

Table 2 24 h-Holter findings in the studied cases

Holter finding	Group1 RVEDVI < 150 (No 14) No (%)	Group2 RVEDVI ≥ 150 (N 18) No (%)	p Value
Normal	5(35.7)	9(50)	0.4
Sick sinus syndrome	1(7.1)	0(0)	0.4
Heart block	1(7.1)	1(5.5)	1
Junctional Tachycardia	2(14.2)	2(11.1)	1
Frequent PVCs	1(7.1)	1(5.5)	1
Infrequent PVCs	1(7.1)	3(16.6)	0.6
Infrequent SVE	3(21.4)	2(11.1)	0.6
Total cases of arrhythmias	9(64.2)	9(50)	

PVCs premature ventricular complexes, SVE supraventricular ectopy

Out of the studied cases, 2 cases had significant ventricular arrhythmia in the form of frequent PVCs. Arrhythmias were detected in 50% of cases with significantly dilated RV. Cases were classified into 2 groups according to the presence or the absence of arrhythmias and comparison between the 2 groups was done (Table 3).

Discussion

Tetralogy of Fallot is a common congenital cyanotic heart disease in which arrhythmias could develop even after successful operative repair [2]. PR and RV dilatation develop in many cases leading to arrhythmias, heart failure and could lead to sudden death [3]. In this study the role of cardiac MRI in the assessment of the degree of right ventricular volume affection in relation to severity of pulmonary regurgitation and associated arrhythmias was assessed helping to decide the need and timing for pulmonary valve repair in cases after successful TOF repair.

In the current study, 18 cases had dilated RV ($\text{RVEDVI} \geq 150 \text{ ml/m}^2$). This is in accordance with the results of Shin et al. [14] who reported progressive RV dilatation in this group of patients with variable rates and without detecting any predictive factor for rapid progression of RV dilatation. This is contradictory to the study performed by Rutz et al. [15] who reported no progression of RV dilatation in patients after surgical repair of TOF with moderately dilated RVs and significant PR during a 3-year follow-up. This difference could be attributed to the shorter duration of follow-up of their studied population in comparison to ours (2.5–16 years).

Table 3 Comparison between cases with and without arrhythmias in clinical, Echocardiographic and MRI findings

	Cases with arrhythmias (18 cases) mean \pm SD (range)	Cases without arrhythmias (14 cases) mean \pm SD (range)	p value
Age (year)	14.82 \pm 2.6	10.87 \pm 2.95	0.001*
Mo	177.88 \pm 31.24	130.40 \pm 35.385	
Age at surgery (mo)	35 \pm 21	32.47 \pm 23.04	0.7
Duration post operatively (mo)	142.71 \pm 33.11	97.93 \pm 36.52	0.001*
Weight centile	37 \pm 25.87	22.57 \pm 17.18	0.08
Height centile	35.53 \pm 28.05	27.79 \pm 25.22	0.4
Na	135.29 \pm 7.5	140.13 \pm 5.92	0.18
K	4.13 \pm 0.5	4.38 \pm 0.6	0.39
Hb	11.58 \pm 1.40	12.57 \pm 0.94	0.06
WBCs	6.57 \pm 1.89	11.59 \pm 11.56	0.11
Platelets	279.20 \pm 83.022	335.1 \pm 92.1	0.12
QRS duration (msec)	117.6 \pm 22.2	117.6 \pm 24	0.95
<i>Echocardiographic findings</i>			
LVEDD (mm)	32.62 \pm 16.03	32.76 \pm 9.53	0.9
FS %	38 \pm 9.39	37.52 \pm 7.13	0.88
<i>MRI findings</i>			
RVEF (%)	49.88 \pm 6.86	54 \pm 8.85	0.16
RVEDVI (ml/m ²)	153.18 \pm 48.94	165.73 \pm 73.31	0.56
RVESVI (ml/m ²)	85.1 \pm 39.1	71.6 \pm 21.2	0.4
LVEDVI (ml/m ²)	91.98 \pm 56.7	79 \pm 36.3	0.47
LVEF (%)	56.82 \pm 4.75	60.5 \pm 5.91	0.06
RVEDVI/LVEDVI	1.9 \pm 0.3	1.85 \pm 0.3	0.57
PR fraction (%)	41.44 \pm 13.05	44.6 \pm 13.34	0.5

FS fractional shortening, LVEDD left ventricular end diastolic diameter, RVEF right ventricular ejection fraction, RVEDVI right ventricular end diastolic volume index, LVEDVI left ventricular end diastolic volume, LVEF left ventricular ejection fraction, PR fraction pulmonary regurgitation fraction

*p Value is significant if less than 0.05

A positive significant correlation between PRF and RVEDVI was detected. Wald et al. [16] suggested that chronic significant PR has late adverse clinical outcomes (at their third decade of life) and were causes for considerable concern. To our knowledge, no published data about the effects of PR in the pediatric and adolescent age group. We found no significant correlation between RF and RVEF. This is coinciding with the results reported by Kordybach et al. [17].

Śpiwak et al. [18] reported that RVEDV, RVEF, and LVEF remained stable over a mean follow-up of approximately 2 years in the majority of adult patients after TOF repair with significant PR and a wide range of RVEDV. This is concordant with our findings with the difference that mean duration of follow-up in our study was 10 years.

We found that LVEF was significantly lower in cases of increased RVEDVI and significantly correlated positively with RVEF, this is coinciding with that reported by Koestenberger et al. [19] who reported in an adult and pediatric study that LVEF was decreased in patients with significantly reduced RVEF. This finding could be

explained by the theory of ventricular-ventricular interaction [20].

Significant ventricular arrhythmia was detected in 2 cases in the form of frequent PVCs. This is contradictory to the study performed by Agha et al. [21] who reported absence of ventricular arrhythmia. The difference could be attributed to the fact that they performed only 12 leads ECG, while in the current study, 24 h Holter monitoring was done, which has higher yield of arrhythmias than 12 leads ECG. In a Japanese multicenter adult study [12], there were 54 patients (10.5%) who had clinically important arrhythmias at follow-up, duration of follow-up ranged from 1 month to 30 years with an average of 11.7 \pm 7.2 years.

We encountered 2 cases of first-degree heart block in the studied population. Although it is generally considered a benign process, there is emerging evidence that prolonged PR interval may be associated with adverse outcomes. Data from Kwok et al. [22] suggests a possible association between prolonged PR interval and significant increases in atrial fibrillation, heart failure and mortality.

We found no significant difference in occurrence of arrhythmias in cases with increased RVEDVI than those without significant RVEDVI increase, this could be explained by non-development of fibrosis (which may require longer duration according to literature) in spite of RV dilatation. Irreversible myocardial injury in cases with severe prolonged volume overload was suggested to be associated with fibrosis and increased interstitial collagen in the left ventricle in cases of aortic regurgitation [23]. Same mechanism could be suggested to apply in RV in cases of longstanding PR and its effect on RV.

In the current study, cases with arrhythmias had significantly longer duration postoperatively and this could favor suggesting the theory of fibrosis that could be time dependent. We observed that cases with arrhythmias to be significantly older in age than those without arrhythmias. This is coinciding with that reported in an earlier young adult study by Arya et al. [24] Also mean QRS duration did not differ significantly between the 2 groups either in the current study or in the study of Arya et al. [24].

Some surgeons prefer to preserve pulmonary valve even at the expenses of residual mild PS (RVOT gradient of 25 mmHg or higher) to preserve RV volume subsequently from progressive dilatation [25].

An adult study done by O'Meagher et al. [26] demonstrated that "Normalization" of RVEDVI ($\leq 108 \text{ mL/m}^2$) was achieved in only 7 out of 18 patients, 14 ± 3 months after PVR. They found that normalization of RV volume is unlikely to be achieved above a pre-PVR RVEDVI of 165 mL/m^2 or more. In particular, an enlarged RVOT prior to PVR predicts suboptimal structural and functional outcomes. Accordingly, 18 (56%) of the studied cases were candidate for PVR. Although, it was reported that young age at pulmonary valve placement is associated with a higher rate of valve failure and early re-operation [13]. The possibility of irreversible RV changes and its impact should be taken into consideration.

The limitations of the current study were small sample size, where correlation with a larger sample size might be need. Also, T1 mapping and late gadolinium imaging were not done, as all of our cases had RV dilatation at variable degrees (mean \pm SD of 112.21 ± 26.4 at group 1 and 195.50 ± 54.9 at group 2), where RV wall thinning hinder the application of T1 mapping or the visual recognition of delayed right ventricular enhancement.

Conclusion

Holter detected arrhythmias after successful surgical repair of TOF were significantly associated with increased postoperative duration but not with PRF or RVEDVI.

Abbreviations

MR: Magnetic resonance; TOF: Tetralogy of Fallot; PR: Pulmonary regurge; PRF: Pulmonary regurge fraction; RVEDVI: Right ventricular end diastolic volume index; RVOT: Right ventricular outflow tract; RV: Right ventricle; CMR: Cardiovascular magnetic resonance; LV: Left ventricle; PVR: Pulmonary valve replacement; PV: Pulmonary valve; VSD: Ventricular septal defect; PS: Pulmonary stenosis; LVEDD: Left ventricular end diastolic diameter; LVESD: Left ventricular end systolic diameter; FS: Fractional shortening; PVC: Premature ventricular contractions; SV: Stroke volume; ESV: End systolic volume; EDV: End diastolic volume; EF: Ejection fraction; RVEF: Right ventricular ejection fraction; LVEF: Left ventricular ejection fraction.

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Authors' contributions

HAA, AR, AAM, GH, FAZM, SE, MAR, MO, SS contributed to this work. HAA, SS, GH, FAZM and SE designed research. HAA, AAM, SS and AR performed research. GH, FAZM and MAR analyzed data. MAR, SS, AR, MO and AAM wrote the paper. All authors have read and approved the manuscript.

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Availability of data and material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

This study was approved by the research ethic committee of the Radiology and Pediatric departments of the Faculty of medicine at Cairo University. All patients included in this study gave a written informed consent to participate in this research. If the patient was less than 16 years old or unconscious at the time of the study, written informed consent for their participation was given by their parent or legal guardian.

Consent for publication

All patients included in this research gave written informed consent to publish the data contained within this study. If the patients were less than 16-year-old, deceased, or unconscious when consent for publication was requested, written informed consent for the publication of this data was given by their parents or legal guardians.

Competing interests

The authors declare that they have no competing interests.

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