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Research Article

Echocardiographic Follow-Up of Right Ventricular Function After Tetralogy of Fallot Operation

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ABSTRACT

Aim: Although limited in assessing right ventricular (RV) function, echocardiography is widely used after correction for tetralogy of Fallot (TOF). The change in echocardiographic measurements of RV in asymptomatic patients after TOF repair over a long follow-up time is not explored yet. The variation in simple echocardiographic measurements during follow-up of our TOF patients are presented. The predictive value of those parameters in determining a future pulmonary valve replacement is sought.

Method: Asymptomatic patients surviving the first year after correction for simple TOF from February 2007 to December 2019 at Başkent University, Istanbul Hospital are enrolled. Patients are followed annually with echocardiography including: RV area, volume, length, RV outflow tract (RVOT) diameter and gradient, tricuspid annulus diameter, tricuspid lateral annular tissue velocities, tricuspid annular plane systolic excursion, TEI index RV ejection fraction (EF) measurements. The change in the consecutive echocardiographic measurements during follow-up is analysed. Patients are evaluated with a cardiac magnetic resonance (CMR) imaging when deemed necessary and compared with echocardiographic measurements.

Results: A total of 66 patients (54.5% males) are operated at age 14.4±9.3 months (78.8% with transannular patch). Twelve patients had pulmonary valve replacement (PVR) operation at an age 10.1±3.1 years. During follow-up of 7.2±4.3 years, tricuspid annulus diameter, RV area, RV inlet length, RV volume, RV volume index significantly increased (p=0.001 for all), whereas RV inlet length index, TEI and TEI z score decreased (p<0.0001 for all). When means are compared, tricuspid annulus (28.8mm vs 25.0mm; p=0.013), RV volume (72.2ml vs 52.2ml; p=0.042), RV inlet length index (77.9mm/m^2 vs 60.2mm/m^2; p=0.013), RVOT diameter (28.7 vs 23.0; p=0.007) are increased. RV EF is decreased (51.3% vs 60.5%; p=0.011) in those requiring PVR. Those with higher RV area index, RV volume index, tricuspid annulus diameter and tricuspid annular z score in their first echocardiography after the TOF repair, are more likely to have a PVR operation later on (p<0.05 for all). RV volume index ≥39ml/m^2 predicts a PVR within 7 years with 100% sensitivity and 74% specificity. Tricuspid annular z score less than -0.43 seems to eliminate the possibility of having a PVR within 7 years with a sensitivity of 44% and specificity of 100%.

Conclusion: Observing the sequential change in echocardiographic parameters like RV volume index, RV area index, tricuspid annulus z score is reliable in determining right ventricular function and can limit the costly CMR applications. Cut off values of RV volume index >39ml/m^2 and tricuspid annulus z score ≥ -0.43 after the initial TOF repair may determine patients with higher possibility of having early PVR and indicate a closer follow-up.

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Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease accounting for 7-10% of all congenital cardiac malformations. Total repair for TOF has been available for more than six decades with a favourable outcome in most patients [1]. Improved late survival of these patients provide an ever-increasing number of patients who require regular follow-up for complications after initial correction. Pulmonary regurgitation acquired after operation for TOF causes a chronic volume overload for the right ventricle and frequently leads to progressive loss of function [2-5]. Pulmonary valve replacement has shown potential to improve right ventricular hemodynamics, yet the best timing is controversial [2-4, 6, 7]. Therefore, longitudinal assessment of pulmonary insufficiency and right ventricle function is critical in the care of patients with repaired TOF.

Cardiac magnetic resonance (CMR) remains the gold standard imaging modality to determine the right ventricular size and function, but it is expensive, time consuming, needs special expertise, and it is not widely available [1]. Furthermore, CMR is not plausible in patients with an implanted device [8-10]. Although having limitations, echocardiography seems to be the best candidate for longitudinal follow-up of postoperative TOF patients, since it is cost-effective and readily available in almost every setting [11]. Up until now, various echocardiographic modalities have been used to assess right ventricular volumes and function and were compared with measurements obtained by CMR [8-10, 12-14]. Although the importance of serial follow-up of these parameters is predominantly emphasized, we are not aware of studies observing the change in echocardiographic measurements in operated TOF patients over time. In this study we aim to determine the temporal change in the right ventricular echocardiographic measurements in postoperative TOF patients during their follow-up, and to seek a parameter that offers a reliable evaluation of the right ventricular size and function.

Materials and Methods

This retrospective study is reviewed and approved by Medical and Health Sciences Research and Ethics Committee of our institution. We obtained a list of all patients who underwent repair of uncomplicated TOF and who survived the first postoperative period at Başkent University, Istanbul Research Hospital from February 2007 to December 2019. Patients with absent pulmonary valve, pulmonary atresia, atrioventricular septal defect, or those requiring a conduit at the initial repair were excluded. Those who became symptomatic because of right ventricular failure or arrhythmia were not assigned. TOF patients are evaluated annually in the outpatient clinic, which includes physical examination, echocardiography and EKG.

After the standard echocardiographic examination, the following measurements are additionally done for all TOF patients in the follow-up clinics, using Vivid S5, GE Vingmed Ultrasound (General Electric, Horten, Norway) echocardiography machine equipped with 3 MHz transducer (3S phased array probe; GE Vingmed Ultrasound).

I Apical Four Chamber Position

Tricuspid annular plane systolic excursion (TAPSE) is obtained from M-mode interrogation of lateral aspect of the tricuspid valve (Figure 1). Tricuspid annulus diameter, right ventricle inlet length (from midpoint of tricuspid valve to the right ventricle apex) are measured in diastole. Right ventricular endocardial surface is manually traced in end-diastole and in end-systole. End diastolic area of the RV is calculated by an automated formula (assuming a spherical shape). End diastolic and end-systolic volumes, and ejection fraction were calculated by using Simpson's formula (Figure 2).

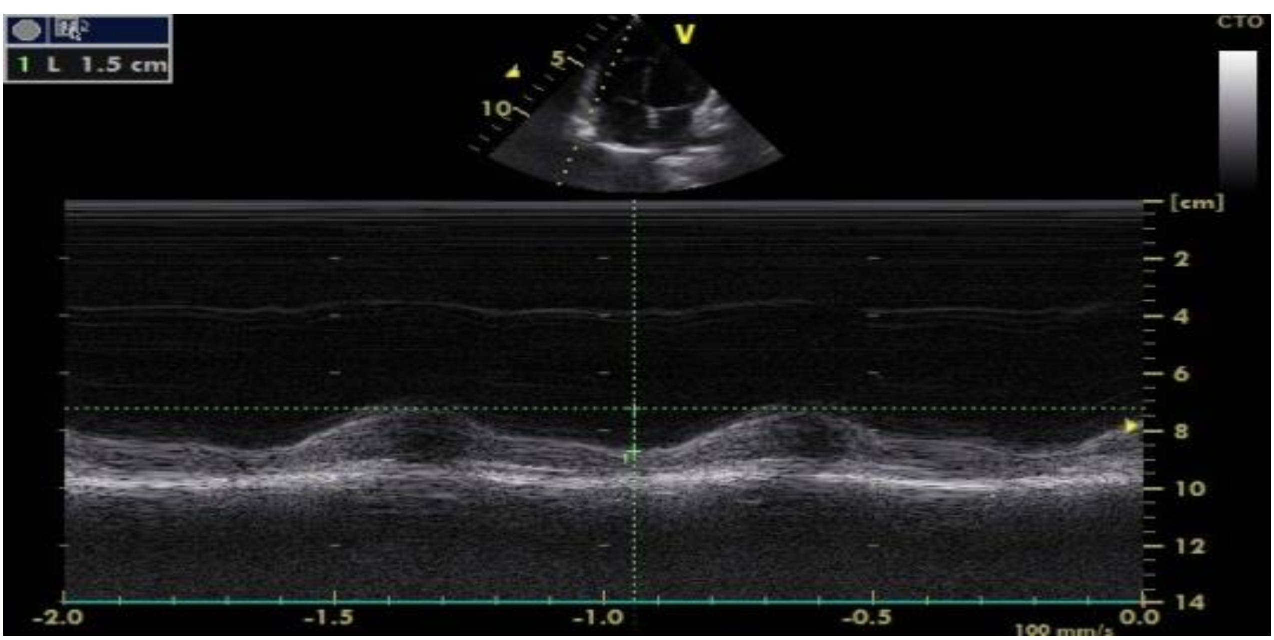

Figure 1: TAPSE (tricuspid annular plane systolic excursion).

A: Diastolic **B:** Systolic

Figure 2: Right ventricular ejection fraction (Simpson's method).

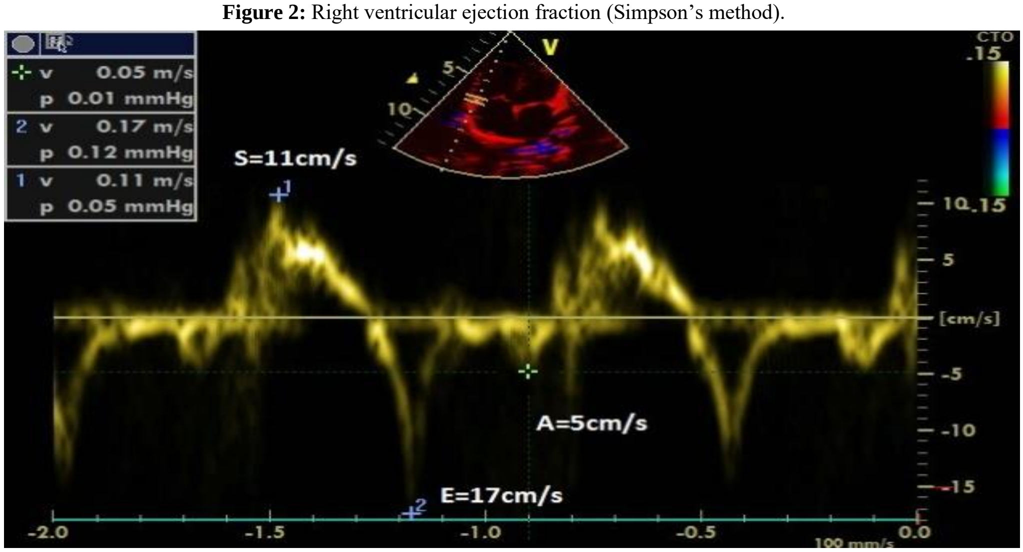

Figure 3: Tissue doppler: Tricuspid lateral annular tissue velocities: systolic (S), early diastolic (E), atrial systolic (A). S: systolic; E: early diastolic; A: atrial contraction

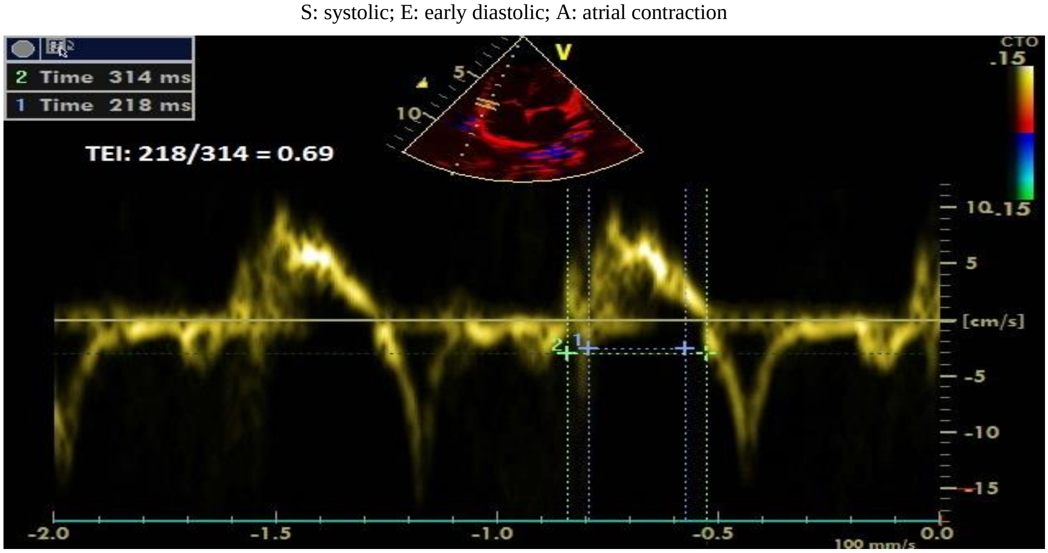

Figure 4: Tissue doppler: TEI index.

$$\text{TEI} = \frac{IVCT + IVRT}{ejection\ time} = \frac{2\ (diastolic\ time\ in\ milliseconds) - 1\ (systolic\ time\ in\ milliseconds)}{1\ (ejection\ time\ in\ milliseconds)}$$

IVCT: Isovolumic contraction time, IVRT: Isovolumic relaxation time

Tissue Doppler measurements are done at the lateral corner of the tricuspid valve annulus. Peak systolic (S), early diastolic (E), and atrial systolic (A) myocardial velocities are obtained at a standard speed of 100mm/second. TEI index is calculated by using the formula: [isovolumic contraction time (IVCT) + isovolumic relaxation time (IVRT)]/ejection time (Figures 3 & 4).

II Parasternal Short Axis Position

Right ventricular outflow tract diameter is measured. Peak velocity gradient across the right ventricular outflow tract is calculated from maximum velocity obtained from the continuous-wave Doppler signal using Bernoulli equation (Figure 5). Measurements of the right ventricular area, volume, inlet length and right ventricle outflow tract diameter are indexed to body surface area. Z scores are calculated for tricuspid annulus, TAPSE, and tricuspid lateral annulus S, E, A velocities using an automated calculator application developed by Evelina Children's Hospital, London, UK.

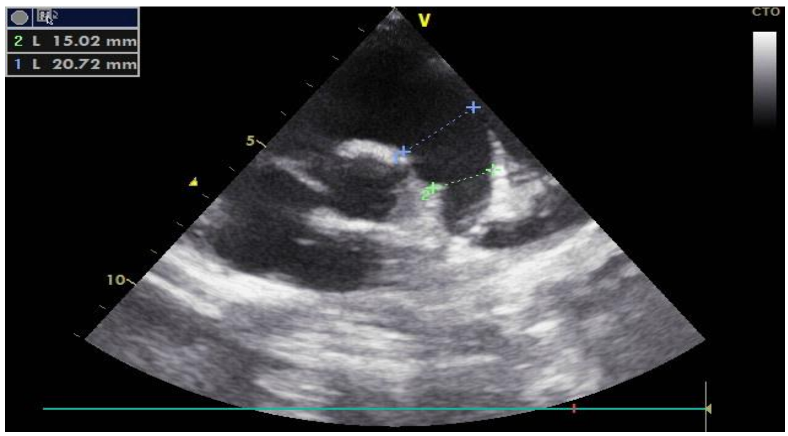

Figure 5: Parasternal short axis, right ventricular outflow tract.
1 L: Right ventricular outflow tract (measurement in mm); 2 L: Pulmonary annulus (measurement in mm)

III Statistics

All values are expressed as mean ± standard deviation; median and range of values are also provided for data with non-normal distribution. Friedman test was performed to analyze the change in the consecutive echocardiographic measurements during follow-up. The measurement(s) that changed significantly in time was/were explored with the Wilcoxon signed rank test. Mann-Whitney U-test was performed while comparing the mean values of certain echocardiographic measurements. A P value of <0.05 was considered significant.

Results

Sixty-six patients (54,5% male, 45,5% female), whose complete data sets were available, are included in the study. The patients were operated for TOF at a mean age of 14.4±9.3 months (median 12.7 months). Transannular patch was used in 78.8% (52 patients). The patients were followed for a mean of 7.2±4.3 years (median 7 years; minimum 0.1, maximum 12.9 years). After total repair for TOF, the following measurements significantly changed during follow-up:

Tricuspid annulus diameter increased (p=0.001), but no significant change was observed in the z score of the tricuspid annulus (p>0.05). Similarly, right ventricle area increased over time (p=0.001), but the right vertical area index did not change significantly (p>0.05). Right ventricle inlet length, volume and volume index increased significantly, whereas right ventricle inlet length index decreased during follow-up (p<0.0001 for each). TEI and TEI z scores decreased significantly (p<0.0001 for both measurements). Tricuspid valve lateral annulus myocardial velocities did not change significantly over time (Figure 6).

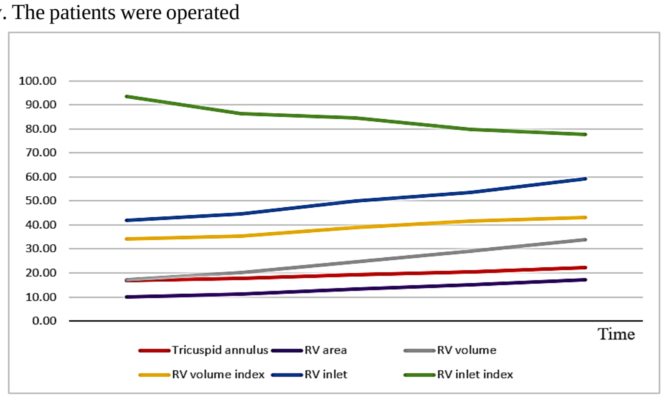

Figure 6: Change in echocardiographic measurements during follow-up after Fallot repair.
Tricuspid annulus (mm), RV area (cm^2), RV volume (ml), RV volume index (ml/m^2), RV inlet (mm), RV inlet index (mm/m^2); RV: right ventricle

Pulmonary valve replacement was done on 12 patients (6 male) at a mean age of 10.1±3.1 years (median 8.8 years; minimum 7.1, maximum 16.7 years). None of the patients were symptomatic at the time of operation, and pulmonary valve replacement was indicated because at least two of the following were present: end diastolic right ventricle volume more than 150ml/m^2, end systolic right ventricle volume more than 80ml/m^2, right ventricle ejection fraction less than 47%, pulmonary regurgitation fraction more than or equal to 25%. The mean follow-up time for those having pulmonary valve replacement was 8.5±3.0 years (significantly longer than those without replacement; p=0.001). More patients with a PVR pulmonary valve replacement operation, had a transannular patch placed during the initial repair (83.3% vs 77.8%), but this was statistically insignificant (p>0.05).

The echocardiographic measurements before the pulmonary valve replacement operation, were compared with those of unoperated patients at their last visit. The tricuspid annulus (mean 28.8mm vs mean 25.0mm; p=0.013), right ventricle volume (mean 72.2ml vs mean 52.2ml; p=0.042), right ventricle inlet length index (mean 77.9mm/m^2 vs 60.2mm/m^2; p=0.013), right ventricular outflow tract diameter (mean 28.7 vs 23.0; p=0.007) were significantly increased in patients before vale replacement. Whereas the right ventricle ejection fraction was significantly decreased (mean 51.3 vs 60.5; p0.011) (Table 1).

Table 1: Echocardiographic measurements compared between patients with and without pulmonary valve replacement (PVR) operation.

	No PVR operation (echocardiographic examination during last visit)	**PVR operation** (last echocardiographic examination before surgery)	**p value**
Tricuspid annulus (mm)	**25.0±4.1**	**28.8±5.7**	**0.013**
Tricuspid annulus z score	0.20±0.60	-0.04±1.10	>0.05
RV area (cm^2)	21.8±8.1	27.0±6.3	>0.05
RV area index (cm^2/m^2)	24.7±15.5	20.4±6.0	>0.05
RV volume (ml)	**52.2±28.2**	**72.2±27.2**	**0.042**
RV volume index (ml/m^2)	50.0±14.9	53.6±18.8	>0.05
RV inlet length (mm)	68.3±17.9	79.0±8.9	>0.05
RV inlet index (mm/m^2)	**60.2±15.7**	**77.9±20.7**	**0.013**
RVOT diameter (mm)	**23.0±5.3**	**28.7±5.2**	**0.007**
RVOT diameter index (mm/m^2)	23.0±7.9	21.5±8.0	>0.05
RVEF (%)	**60.5±7.9**	**51.3±17.1**	**0.011**
QRS	126±47	132±18	>0.05

PVR: Pulmonary Valve Replacement; RV: Right Ventricle; RVOT: Right Ventricular Outflow Tract; RVEF: Right Ventricular Ejection Fraction (p<0.05 is statistically significant).

Table 2: Significant correlations between echocardiographic and cardiac magnetic resonance imaging measurements.

Echocardiography	**MRI**	**p value**	**r value**
RV area	RV diastolic volume	p=0.023	r=0.581
RV diastolic volume	RV diastolic volume	p=0.041	r=0.532
RV inlet	RV systolic volume index	p=0.006	r=0.671
RV EF	RV EF RV end systolic volume index	p=0.023 p=0.043	r=0.623 r= -0.568

MRI: Magnetic Resonance Imaging; EF: Ejection Fraction; RV: Right Ventricle.

All patients who had a pulmonary valve replacement were evaluated with a CMR before the operation. On the other hand, six patients had a CMR but were not considered a candidate for pulmonary valve replacement, based on the right ventricular measurements on CMR. Right ventricular measurements obtained by echocardiography were compared with those from CMR in these 18 patients. The significant correlations are depicted in (Table 2).

Right ventricle area, and right ventricle end diastolic volume measured by echocardiography were correlated with right ventricle end diastolic volume measured by CMR (p = 0.023, r = 0.581; and p = 0.041, r = 0.532 respectively). Right ventricle inlet length was correlated with right ventricle systolic volume index in CMR (p=0.006, r=0.671). Right ventricle ejection fraction measured by echocardiography using Simpson formula was strongly correlated with right ventricle ejection fraction (p = 0.023, r = 0.623) whereas it was inversely correlated with right ventricle end systolic volume index at CMR (p = 0.043, r = -0.568). None of the tissue Doppler velocities or TAPSE measured by echocardiography were correlated with any of the CMR right ventricle measurements.

The patients eventually having pulmonary valve replacement had significantly larger tricuspid valve annulus diameter (21.6±6.0mm vs 17.7± 4.1mm; p=0.011), higher tricuspid annular z score (0.2±0.5 vs ±0.6, p=0.008), right ventricle area index (25.8±4.6cm^2/m^2 vs 21.6±4.2cm^2/m^2, p=0.006) and right ventricle volume index (49.5±12.2ml/m^2 vs 36.2±12.6ml/m^2, p=0.003) on their first echocardiogram after the initial TOF repair. However, the age and the body surface area at the time of initial echocardiography were similar to those not having valve replacement.

Having observed this relation, we tried to determine a cut-off value that would predict a pulmonary valve replacement after a complete repair for TOF. Right ventricle volume index ≥39ml/m^2 predicts a pulmonary valve replacement within 7 years with 100% sensitivity and 74% specificity. The other cut off values and the corresponding sensitivities and specificities are given in (Table 3).

Table 3: Cut-off values of certain echocardiographic measurements, their sensitivity and specificity to predict a pulmonary valve replacement.

Echocardiographic Measurement	Cut off Value	Sensitivity	Specificity	Positive Predictive Value	Negative Predictive Value
RV volume index (ml/m^2)	>39	100%	74%	42%	100%
RV area index (cm^2/m^2)	>23.5	80%	74%	36%	95%
Tricuspid annulus (mm)	>19.5	70%	80%	39%	93%
Tricuspid annulus z score	≥-0.43	100%	44%	40%	100%

RV: Right Ventricle

After pulmonary valve replacement operation, no significant changes were observed in tricuspid annulus, RV volume, area, or inlet measurements during a mean follow up of 22.5±24.1 months (median 21.2 months). However, TAPSE and TAPSE z score increased significantly (p=0.043, p=0.027 respectively), as well as the RVOT gradient (p=0.006).

Discussion

In this study, 18% of the patients with total repair of TOF needed a pulmonary valve replacement operation within a mean of 7.2 years. During follow-up, tricuspid annulus diameter, right ventricle area, inlet length, volume, volume index significantly increased, whereas right ventricle inlet length index, TEI and TEI z score decreased. While observing an increase in right ventricle length, the decrease in right ventricle length index may be explained by the change in the geometry of the right ventricle, assuming a more spherical shape during follow-up.

Expectedly, the four chamber echocardiographic measurements of the right ventricle were greater in those requiring pulmonary valve replacement. Those with higher right ventricle area index, right ventricle volume index, tricuspid annulus diameter and tricuspid annular z score even in their first echocardiography after the TOF repair, were more likely to have a pulmonary valve replacement operation later on.

Pulmonary regurgitation is a common finding in patients after correction of TOF. Right ventricle dysfunction and arrhythmia have been attributed to chronic pulmonary regurgitation and progressive right ventricle dilatation after repair [10]. Pulmonary valve replacement is the standard treatment for pulmonary regurgitation, and the timing mainly depends on symptoms. Marked right ventricle enlargement and dysfunction can be present before the onset of symptoms, which explains the discouraging outcomes in some of the symptomatic patients. Hence, early intervention in asymptomatic patients is advocated [4, 7]. Assuming that the less severe the ventricular dilatation prior to valve replacement, the more likely the ventricle will remodel, many investigators tried to determine the right ventricle measurements beyond which return to normal size is unlikely [5, 15-17]. Thus, there is an evolving strategy of earlier pulmonary valve replacement.

Accurate assessment of right ventricle volumes and function is very important in the follow-up of patients with TOF. Right ventricle measurements obtained by CMR are considered as reference in guiding the management decisions [8]. Although CMR is available in most centers managing patients with congenital heart disease, its use is expensive and time consuming. Acquisition and interpretation of the images requires special expertise [9, 10]. Furthermore, it is contraindicated in patients with implanted devices. All these features limit its frequent use in the clinical setting. While standard echocardiography is the most widely available and cost-effective imaging modality, its use is hampered by its less accurate right ventricle measurements based on geometry assumptions that do not apply to the right ventricle [4]. Many novel techniques like real time 3D echocardiography and strain echocardiography have been used with the hope that they will help refine the decision making process in these patients [8, 12-14]. However, these methods have been predominantly utilized as research tools. Their clinical application, although promising, has not gained widespread attention in everyday practice. Therefore, a simple echocardiographic measurement that reliably and accurately predicts right ventricle volume and function would be ideal to monitor in outpatient postoperative TOF patients.

We suggest that two dimensional echocardiographic measurements of right ventricle in four chamber view can be used reliably in the follow-up of patients after TOF repair. The temporal change in these measurements will signal a right ventricular dilatation and dysfunction and a call for CMR, thereby limiting the costly CMR applications. Especially tricuspid annulus z score and RV volume index measurements seem to be good predictors of a future pulmonary valve replacement operation. Even in the first echocardiographic examination after the TOF repair, a right ventricle volume index of more than 39 ml/m^2 is 100% sensitive and 74% specific in predicting a PVR in 7 years. Similarly, tricuspid annular z score less than -0.43 may eliminate the possibility of having a PVR within 7 years with a sensitivity of 44%. Those patients having greater measurements than the cut-off values right after the repair may be candidates for closer follow-up and more frequent CMR studies.

We found that four chamber measurements of right ventricle volume and area are correlated well with right ventricle diastolic volumes in CMR.

Also, right ventricle inlet length is correlated with right ventricle systolic volume index in CMR, and right ventricle ejection fraction measured by Simpson's formula is correlated with right ventricle ejection fraction measured in CMR. Longitudinal shortening is the main component of right ventricular systolic function, therefore these simple measurements in four chamber view reflect ventricular function fairly well. These findings are in accordance with other studies reporting a correlation between four chamber measurements and CMR measurements [8, 12].

On the other hand, as with other investigators, we did not observe any relation between tissue Doppler velocities, TEI or TAPSE and the CMR findings of the right ventricle [3, 9, 12, 18, 19]. TEI is a reliable echocardiographic parameter showing ventricular function. Although in our study group right ventricular TEI progressively increased over time suggesting a developing ventricular dysfunction, this may be misleading. With high preload and low pulmonary pressures, such as seen in TOF, the isovolumic relaxation time becomes very short. Since right ventricular end diastolic volume is increased, ejection time is also increased. As a result, pseudonormalization of TEI is often seen in TOF patients with pulmonary regurgitation. Therefore, the change in TEI may actually reflect the change in right ventricular loading rather than intrinsic myocardial function [12].

There is no correlation between right ventricle measurements in CMR and myocardial velocities of lateral tricuspid annulus or TAPSE in echocardiography either. This may be attributed to a local dysfunction in the myocardial contraction pattern after TOF operation, which does not translate to global ventricular activity. Measurements integrating longitudinal and circumferential shortening of the right ventricle (i.e. fractional area/volume change) may analyse its global systolic function better. Tricuspid annular plane systolic excursion of the lateral annulus depends on ventricular load rather than the right ventricular ejection fraction. Furthermore, decrease in tricuspid annular motion has been observed in patients even after operations that do not affect the right ventricular volume and when right ventricular ejection fraction is normal [12].

Frigola A *et al.* noted that virtually all of their patients more than 35 years of age and free of pulmonary valve replacement had a pulmonary valve *z* scores <0.5, suggesting that ideal PA diameter z score in postoperative TOF is -1.3-0.5 [20]. Although most of our patients had a transannular patch, it was unexpected to see any correlation between right ventricular outflow tract diameter or pulmonary annulus on echocardiography and right ventricular measurements on CMR. This may be due to the small number of patients studied. However, we did find a correlation between tricuspid annulus and CMR measurements. We are tempted to add that the tricuspid annulus z score should be no more than -0.43 after the repair, so that it voids a pulmonary valve replacement operation in 7 years.

Although right ventricle size decreases and functional class improves after pulmonary valve replacement, right ventricular normalization does not occur very often and modest amount of patients achieve increased systolic function [2, 5, 7, 13, 14, 16, 21-26]. We failed to observe any significant change in the right ventricle measurements after pulmonary valve replacement, only TAPSE and right ventricular outflow tract gradient increased significantly. This may be due to short term follow-up after valve replacement, since 33% of our patients were followed less than a year. Besides, CMR was not repeated after the pulmonary valve replacement to confirm any change in right ventricle volumes.

Twelve of our patents had relatively early pulmonary valve replacement operations. This may be because majority of our patients had a TOF repair with a transannular patch. Performing pulmonary valve replacement early in the follow-up in younger patients could be associated with a better right ventricular modeling, but it probably increases the number of surgeries or percutaneous interventions in a lifetime [3, 4]. Whether there is definitive improvement in right ventricle function after pulmonary valve replacement, even with less significant right ventricular dilation, remains unclear [5]. Therefore, determining clear indications for secondary pulmonary valve replacement requires longitudinal data. Unfortunately, most of the available data are from cross sectional studies which mostly focus on patients who are already being considered for valve replacement [20]. Frequent, serial volumetric follow-up of the right ventricle over a prolonged time is best achieved with echocardiography. Controlled, multicenter study with an extended follow-up time span is necessary to collect data on asymptomatic patients.

Limitations

This study has a number of limitations; retrospective design of the study may have caused bias in case selection. The postoperative follow-up period after pulmonary valve replacement is short, therefore the findings may not be applicable in the long term. Our study group is relatively small which may have decreased the power of statistical analysis.

Conclusion

The two dimensional right ventricular assessment in the four chamber view, being highly feasible, easily obtainable, and having good correlation with CMR measurements, is a good monitoring tool for right ventricular function during follow-up of asymptomatic postoperative TOF patients. Especially observing the sequential change in echocardiographic parameters like right ventricle volume index, right ventricle area index, tricuspid annulus z score may be valuable in determining right ventricular function. We suggest that cut off values of right ventricle volume index >39ml/m^2 and tricuspid annulus z score ≥ -0.43 after the initial TOF repair may determine patients with higher possibility of having early pulmonary valve replacement and indicate a closer follow-up, whereas the others may be spared with less frequent CMR's.

Conflicts of Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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