

Cardiac MRI in surgically repaired tetralogy of Fallot: Our initial experience

 Sercin Ozkok,¹  Oyku Tosun,²  Ilker Kemal Yucel,³  Ahmet Celebi³

¹Department of Radiology, Istanbul Medeniyet University, Goztepe Training and Research Hospital, Istanbul, Turkiye

²Department of Pediatric Cardiology, Istanbul Medeniyet University, Goztepe Training and Research Hospital, Istanbul, Turkiye

³Department of Pediatric Cardiology, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkiye

ABSTRACT

OBJECTIVE: Pulmonary regurgitation (PR) required pulmonary valve replacement (PVR) is usually seen after surgically repaired tetralogy of Fallot (TOF). Assessment by cardiac magnetic resonance imaging (CMR) plays a crucial role in the decision of PVR. Herein, we presented our 3-year interdisciplinary CMR experience in the assessment of repaired TOF.

METHODS: CMR examinations of 196 patients with repaired TOF performed between 2016 and 2018 were enrolled in this retrospective study. Only 165 were included in the study. CMR findings were assessed according to the American College of Cardiology/American Heart Association guideline and recommendations of Geva.

RESULTS: Among those 165 patients (median age 14 years [mean age 15.62±7.42 years], M/F=114/61; 1.86/1), 73 patients were found eligible for PVR (59 patients for transcatheter while 14 patients for surgical). The mean QRS duration was 170.2±16.89 ms. On CMR assessment, mean indexed right ventricular end-diastolic volume, end-systolic volume, right, and left ventricular ejection fraction were 187.64±45.07 ml/m², 39.90±6.60%, and 47.83±6.12%, respectively. The PR fraction was as 50.10±2.54% and 2.25±1.92. Balloon dilatation and/or stenting of branch pulmonary arteries in 12 patients and ventricular septal defect closure in four patients were performed at the same session of percutaneous PVR. At the time of the surgical PVR, repair of partial anomalous pulmonary venous return in one patient, ventricular septal defect in two patients, and subaortic membrane in one patient were performed. An implantable cardioverter-defibrillator was also performed in one patient.

CONCLUSION: Our CMR experience has the largest patient population in our country and may contribute to the national data pool. We believe that our collaborative experience between radiologists, cardiologists, and cardiovascular surgeons may also enhance the use of CMR in determining the appropriate technique or timing for PVR.

Keywords: Cardiac magnetic resonance imaging; pulmonary valve replacement; tetralogy of Fallot.

Cite this article as: Ozkok S, Tosun O, Yucel IK, Celebi A. Cardiac MRI in surgically repaired tetralogy of Fallot: Our initial experience. *North Clin Istanbul* 2022;9(6):622–631.

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with the incidence of approximately 0.5/1000 live births and 7–10% of all congenital heart diseases [1]. The classical tetrad of the disease consists of a right ventricular outflow tract (RVOT) obstruction, a ventricular septal defect, an overriding aorta, and hypertrophy of the right ventricle (RV) [1]. The severity of the disease and the timing of

the surgery are depended on the degree of RVOT obstruction or hypoplasia of the pulmonary valve [1]. The initial surgical repair is consists of the ventricular septal defect closure and relieving the RVOT which varies from infundibulectomy with/without commissurotomy, or transannular patch repair in the presence of severe stenosis, or a RV-to-main pulmonary artery conduit if pulmonary atresia was present [1–3].

Received: January 27, 2021

Revised: February 23, 2021

Accepted: October 18, 2021

Online: November 17, 2022

Correspondence: Sercin OZKOK, MD. Istanbul Medeniyet Universitesi, Goztepe Egitim ve Arastirma Hastanesi, Radyoloji Klinigi, Istanbul, Turkiye.

Tel: +90 216 606 52 00 e-mail: sercinbas2005@gmail.com

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Although surgical advances in one- or two-stage repair facilitate survival of the patients into adulthood with good quality of life [1–3], long-term complications and residual lesions can be seen [4, 5]. In most cases, pulmonary regurgitation (PR) required a pulmonary valve replacement (PVR) may be seen and be resulted in a progressive RV dilatation and dysfunction [6, 7]. Right heart failure with severe PR and/or pulmonary stenosis is unequivocally indicated for PVR in the American College of Cardiology/American Heart Association (ACC/AHA), European Society of Cardiology (ESC), and Canadian Cardiovascular Society (CCS) guidelines and recommendation of Geva [8–11]. Nevertheless, the appropriate timing for PVR is still controversial in asymptomatic patients [12–14]. In our clinic, there has been an increased request of cardiac magnetic resonance imaging (CMR) to assess the timing and technique of PVR. We strongly believe that interdisciplinary cooperation and increased examination quality will enhance the diagnostic value of CMR and the frequency of its use in the clinic. Herein, we aimed to present CMR findings of the asymptomatic patients with repaired TOF found eligible for PVR and collaborative pediatric cardiology and radiology experience with the largest CMR population in the Turkish literature.

MATERIALS AND METHODS

Ethical approval of this retrospective study was obtained from the Ethical Committee (no: 2018/0484; date: December 12, 2018). This study was conducted in accordance with principles for human experimentation as defined in the Declaration of Helsinki. Informed consent for CMR examination was obtained from all patients and parents under 18 years old before the examination.

Study Group

We retrospectively reviewed 196 patients with repaired TOF referred to our department between 2016 and 2019. Patients' demographic data such as gender, age at surgery, and age at CMR examination were obtained from the hospital information system. Transthoracic two-dimensional echocardiographic evaluations of the patients were performed with the Philips Epiq 7c Echocardiography Systems (Philips Healthcare; Holland) and S5-1 probe by an experienced pediatric cardiologist before the CMR examination.

Highlight key points

- Moderate-to-severe PR is required a PVR in late after surgical repair of TOF.
- PVR in asymptomatic patients should be assessed by CMR following the listed criteria.
- Our experience may provide important clues for the widespread use of CMR imaging and multidisciplinary cooperation in the follow-up of repaired TOF in our country.

Asymptomatic patients with repaired TOF are referred to our clinic to perform CMR examinations. Timing and the appropriate technique for PVR in patients with TOF repair were assessed by CMR findings collaboratively. All the CMR examinations were evaluated according to the criteria listed in the ACC/AHA guideline [9] and recommendations of Geva [11]. Asymptomatic patients (New York Heart Association Class I or II) were defined as those who have no symptom during ordinary physical activity or mild symptoms such as mild shortness of breath and/or angina.

The patients who have syncope caused by arrhythmia or symptoms of heart failure, implantable cardioverter-defibrillator, history of PVR, contraindications for magnetic resonance imaging (MRI), and repetitive control CMR were excluded from the study.

CMR Imaging

All CMR examinations were performed using a 1.5 Tesla scanner (Signa HDx; GE Medical Systems, Milwaukee, WI, USA) with a 32-channel phased-array abdominal coil with electrocardiographic gating followed our institutional standardized CMR protocol with imaging parameters below. No sedation was used during imaging. All the patients were trained about how to breath-hold before the examination. The images were acquired during one or two breath-hold of 8–12 s duration depends on the heart rate during end-expiratory breath-hold. The duration of the examination changes between 30 and 45 s. The weight and height of the patients were recorded before the examination.

Functional Evaluation

Sagittal, coronal, and axial localizers through thorax revealed by steady-state free precession sequence first. Then, cine steady-state free precession sequence images of two-chamber and four-chamber views were obtained to plan the short-axis plane covering the entire ventricles.

The slices were positioned perpendicular to the interventricular septum. Cine images of the RVOT were revealed by positioning parallel to the plane of the RVOT. Each set of images was acquired with retrospective gating, and 20 reconstructed cardiac phases.

Magnetic Resonance Angiography and Blood Flow Assessment of Vascular Structures

A 0.2 mmol/kg of gadolinium-based contrast media was performed for contrast-enhanced magnetic resonance angiography. Phase-contrast imaging of the main pulmonary artery, branch pulmonary arteries, and ascending aorta was performed perpendicular to the vascular structures. Phase-contrast images of the main pulmonary artery revealed just below the bifurcation, left pulmonary artery, and right pulmonary artery on the mid-segment level. The optimal velocity encoding value of the pulmonary artery was calculated by the Bernoulli equation reported gradients in the echocardiography report [12]. Each set of images was acquired with 20 reconstructed cardiac phases. Late gadolinium enhancement sequences also revealed after 10 min of injection for the assessment of myocardial contrast enhancement.

Image Analysis and Post-Processing Workflow

All CMR examinations were reviewed by a 10-year experienced radiologist (trained in congenital CMR and computed tomography, with an experience of more than 700 CMR examinations) and a pediatric cardiologist. All the images were reviewed using a commercially available software program (5.6i report card, GE Medical Systems, Milwaukee, WI, USA) on a workstation. The endocardial layer of ventricles was contoured manually on short-axis cine images by including the papillary muscles and the trabeculations through all slices on end-diastolic and end-systolic phases. Body surface area (with Mosteller's formula), biventricular end-diastolic volume index (EDVI), end-systolic volume index (ESVI), stroke volume index (SVI), and ejection fraction (EF) were calculated automatically by the workstation. The presence of late gadolinium enhancement of both ventricles was also assessed.

The size of the branch pulmonary arteries was measured at the narrowest area perpendicular to the long axis on a multiplanar reconstruction image of magnetic resonance angiography. Z-score of branch pulmo-

nary arteries was also recorded [13]. The presence of stenosis or aneurysmatic dilatation of the main pulmonary artery was assessed on three-dimensional volume rendered images.

For the flow analysis, the contour of the vascular structures was traced manually. Forward flow volume, regurgitant flow volume, and net flow volumes were calculated by a software program. PR fraction (regurgitant flow volume/forward flow volume \times 100 in%) and blood flow distribution of the right-to-left pulmonary artery [net right pulmonary artery flow volume/(right pulmonary artery + left pulmonary artery flow volume) \times 100 in%] were also calculated. The presence of end-diastolic antegrade flow was also recorded from flow diagrams. The systemic-to-pulmonary flow ratio is calculated to assess the degree of the left-to-right shunt. It is calculated as dividing the net flow volume of the pulmonary artery to ascending aorta.

Statistical Analysis

The analysis of the obtained data was performed with SPSS version 20.0 software (IBM Corporation, Armonk, NY, USA). The mean and standard deviation values were used for the presentation of the descriptive values and presented as mean \pm standard deviation in quantitative variables. Median values were also used for the body surface area and age of the patients.

RESULTS

We retrospectively reviewed 196 patients with repaired TOF referred to our clinic between 2016 and 2018. Repetitive control MRI of nine patients and 22 patients had already PVR excluded from the study (Fig. 1). CMR examinations of the 165 patients (M/F=104/61; 1.70/1) with repaired TOF were involved in the study. The median age of patients was 14 years (mean age 15.62 ± 7.42 years, range 5–50 years) and the body surface area was 1 m^2 (mean body surface area $1.47 \pm 0.32 \text{ m}^2$, range 0.7–2.16 m^2). The median age of initial repair was 24 months (mean age 35.93 ± 35.69 months, 5 months–172 months) using the transannular patch repair approach. A palliative modified Blalock-Taussig shunt procedure has been performed in four patients. Before CMR examination, the various transcatheter procedures have already been performed in 16 patients (11 patients had branch pulmonary artery stenting and/or balloon dilatation of branch pulmonary artery,

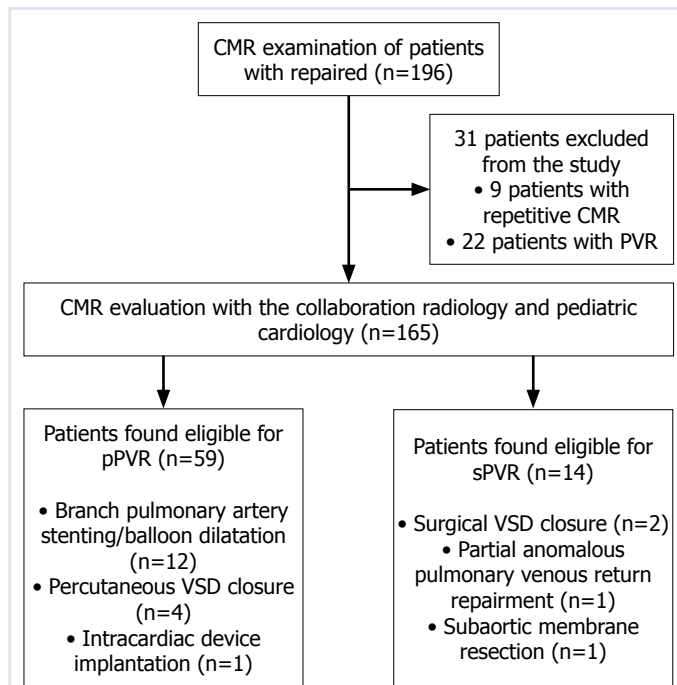


FIGURE 1. Determination of the study cohort.

CMR: Cardiac magnetic resonance imaging; TOF: Tetralogy of Fallot; VSD: Ventricular septal defect; PVR: Pulmonary valve replacement; pPVR: Percutaneous pulmonary valve replacement; sPVR: Surgical pulmonary valve replacement.

four patients had balloon dilatation of main pulmonary artery, and one patient had balloon dilatation of aortic coarctation). The mean RV and LV diameter were 3.65 ± 0.98 cm and 4.19 ± 0.62 cm, respectively, on two-dimensional echocardiography and the mean pulmonary valve gradient was 26.43 ± 9.89 mm/Hg on Doppler echocardiography. The mean TAPSE value was 17.08 ± 3.16 mm and the mean QRS duration was 151.47 ± 21.60 ms. All the patients had a functional status that can walk 2–3 floor stairs up (New York Heart Association Class I or II). The demographic data of the patients' are shown in Table 1.

Calculated mean EDVI, ESVI, SVI, and EF were 158.49 ± 44.48 ml/m², 93.11 ± 32.39 ml/m², 65.88 ± 11.09 ml/m², and $42.06 \pm 7.26\%$ for RV and 89.30 ± 18.95 ml/m², 46.29 ± 12.68 ml/m², 43.39 ± 9.77 ml/m², and $48.40 \pm 6.23\%$ for LV, respectively. PR fraction was found $43.69 \pm 4.92\%$ in pulmonary artery flow analysis. Preferential blood flow was 1.67 ± 0.55 by calculating the ratio of net right pulmonary artery-to-left pulmonary artery flow (Fig. 2). In the morphologic evaluation of short-axis and four-chamber cine mages, a residual septal defect was detected in 15 patients (atrial septal defect in six patients and ventricular septal de-

TABLE 1. Demographic and echocardiographic data of the patients with repaired tetralogy of Fallot

	Patients (n=165)
Gender (male/female)	104/61
Median age at CMR examination (years)	14
Median BSA (m ²)	1
Median age at TOF repair (months)	24
Mean RV diameter in echocardiography (cm)	3.65 ± 0.98
Mean LV diameter in echocardiography (cm)	4.19 ± 0.62
Mean TAPSE (mm)	17.08 ± 3.16
Mean gradient of pulmonary artery (mmHg)	26.43 ± 9.89
Mean QRS duration (ms)	151.47 ± 21.60

CMR: Cardiac magnetic resonance imaging; BSA: Body surface area; TOF: Tetralogy of Fallot; RV: Right ventricle; LV: Left ventricle; TAPSE: Tricuspid annular plane systolic excursion.

TABLE 2. Cardiac magnetic resonance imaging of the patients with repaired tetralogy of Fallot

Cardiac magnetic resonance imaging parameters	Patients (n=165)
Left ventricle	
Mean EDV index (ml/m ²)	89.30 ± 18.95
Mean ESV index (ml/m ²)	46.29 ± 12.68
Mean SV index (ml/m ²)	43.39 ± 9.77
Mean EF (%)	48.40 ± 6.23
Right ventricle	
Mean EDV index (ml/m ²)	158.49 ± 44.48
Mean ESV index (ml/m ²)	93.11 ± 32.39
Mean SV index (ml/m ²)	65.88 ± 11.09
Mean EF (%)	42.06 ± 7.26
Mean regurgitation fraction of MPA (%)	43.69 ± 4.92
Preferential blood flow (RPA/LPA)	1.67 ± 0.55
Mean diameter of RPA (mm)	18.42 ± 4.80
Mean Z-score of RPA	1.57 ± 1.64
Mean diameter of LPA (mm)	18.5 ± 7.25
Mean Z-score of LPA	2.7 ± 2.14
Mean diameter of AAO (mm)	28.12 ± 4.87
Mean regurgitation fraction of AAO (%)	12.39 ± 6.32

EDV: End-diastolic volume; ESV: End-systolic volume; SV: Stroke volume; EF: Ejection fraction; MPA: Main pulmonary artery; RPA: Right pulmonary artery; LPA: Left pulmonary artery; AAO: Ascending aorta.

fect in nine patients), moderate tricuspid regurgitation was determined in 90 patients. During the assessment of three-dimensional volume rendered images of contrast-enhanced magnetic resonance angiography, a sac-

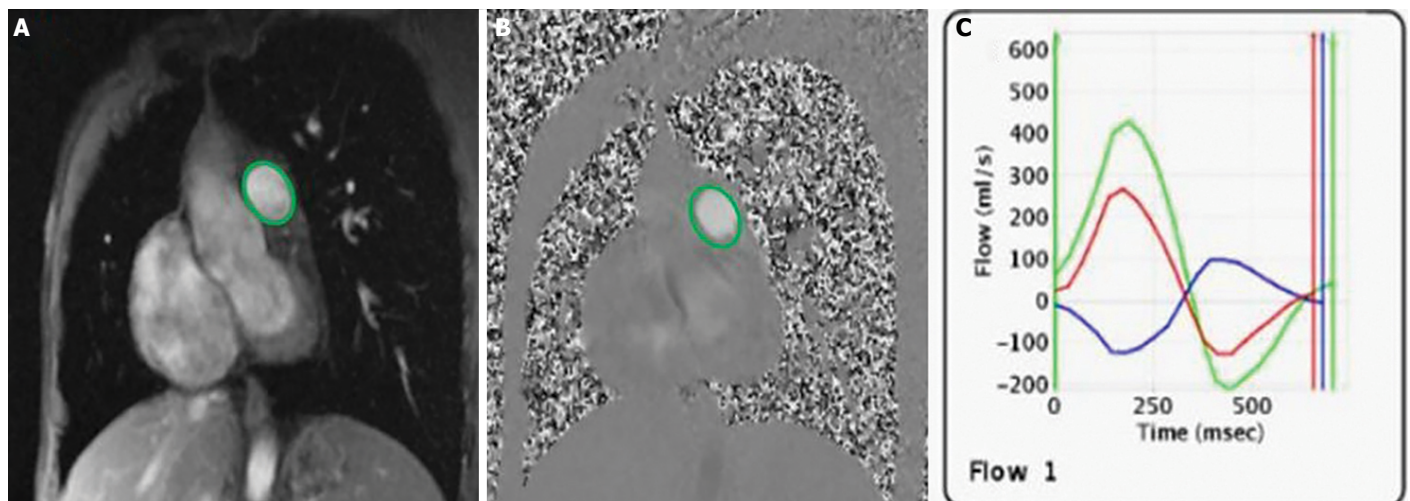


FIGURE 2. Phase-contrast imaging with magnitude **(A)** phase-velocity, **(B)** image of the main pulmonary artery in cross-section. Flow analysis **(C)** of the main pulmonary artery (green line) shows systolic forward flow, diastolic backward flow, and late diastolic forward flow at end-diastole. The left pulmonary artery flow (red line) and right pulmonary artery flow (blue line) also demonstrated.

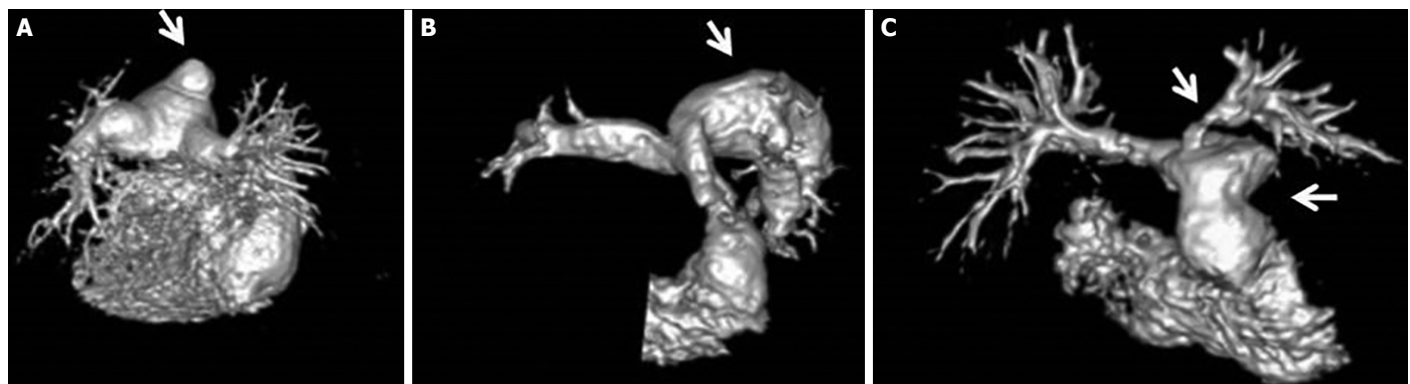


FIGURE 3. Saccular aneurysmatic dilatation at the bifurcation of the main pulmonary artery (MPA) and valvular stenosis of the main pulmonary artery from posterior aspect **(A)**, dilated left pulmonary artery and extension of the aneurysm **(B)**, valvular stenosis with post-stenotic dilatation of MPA and narrowing of the proximal left pulmonary artery **(C)** demonstrated in 3D reconstructed images of contrast-enhanced magnetic resonance angiography.

cular aneurysmatic dilatation of the main pulmonary artery was revealed in three patients, RVOT dilatation extended to the left pulmonary artery was revealed in 12 patients (Fig. 3). The mean diameter of the left pulmonary artery and right pulmonary artery was found 18.5 ± 7.25 mm and 18.42 ± 4.80 mm, respectively. The mean diameter of the ascending aorta and regurgitation fraction was 28.12 ± 4.87 mm and $12.39 \pm 6.32\%$, respectively. A right-sided aortic arch was revealed in 30 patients. Myocardial delayed enhancement of RV free wall extended to the right atrial wall, interventricular septum, RV insertion point, and RVOT was found in only one patient (Fig. 4). CMR parameters of the patients are demonstrated in Table 2.

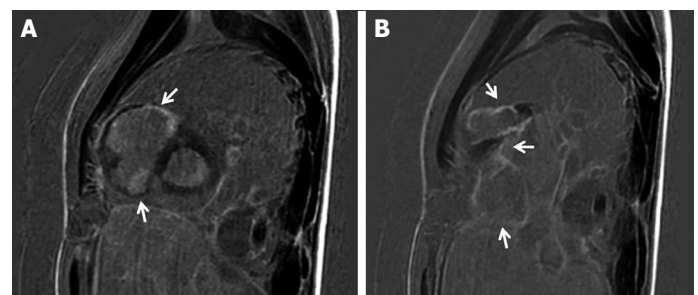


FIGURE 4. Late gadolinium enhancement imaging in the ventricular short-axis view showing contrast enhancement in the free wall of the right ventricle extends to the right atrial wall and interventricular septum, atrioventricular hinge point **(A)**, and right ventricular outflow tract **(B)**.

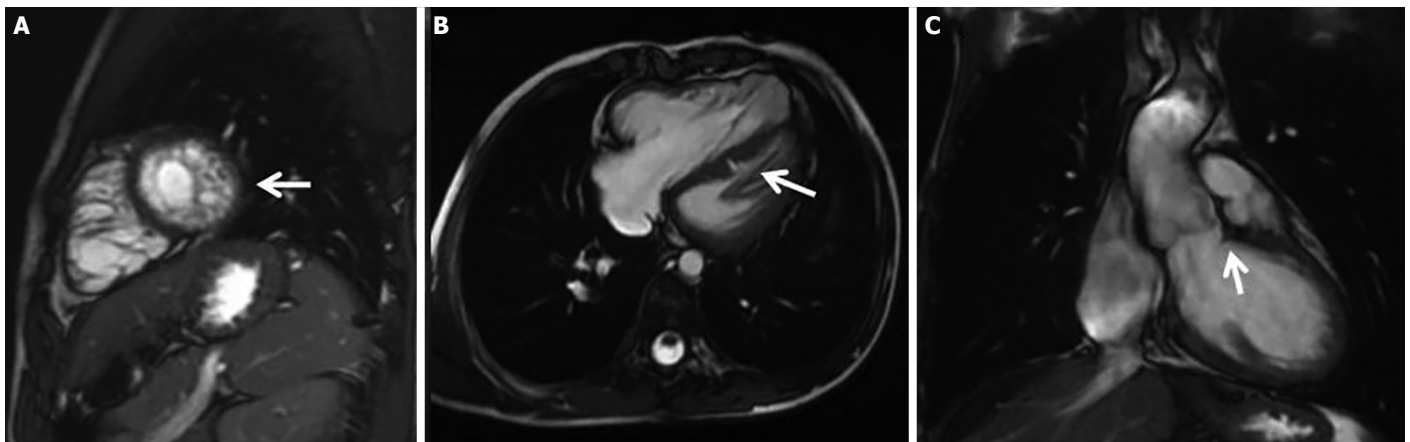


FIGURE 5. Steady-state free precession cine SA views showing hypertrabeculation (the ratio of non-compacted/compacted layer higher than 3.1 indicative for the left ventricular non-compaction cardiomyopathy **(A)**, myocardial crypt in interventricular septum on a four-chamber view **(B)**, and thin transverse subaortic membrane on the left ventricular outflow tract view **(C)**.

According to the ACC/AHA guideline [9] and recommendations of Geva [11], 73 patients (mean age: 18.88 ± 5 years and body surface area: 1.71 ± 0.26 m²) were found eligible for PVR. The mean RV diameter, LV diameter, and pulmonary valve gradient pressure were 4.54 ± 1.19 cm, 4.38 ± 0.22 cm, and 27 ± 40 mm/Hg, respectively, on echocardiography. The mean QRS duration and TAPSE values of the patients referred to PVR were 170.2 ± 16.89 ms and 19.33 ± 1.03 mm, respectively. On CMR assessment, mean RV-EDVI, RV-ESVI, RV-EF, RV-EF, and LV-EF were 187.64 ± 45.07 ml/m², 113.69 ± 33.29 ml/m², $39.90 \pm 6.60\%$, and $47.83 \pm 6.12\%$, respectively. In pulmonary artery flow analysis, the PR fraction and preferential blood flow ratio were calculated as $50.10 \pm 2.54\%$ and 2.25 ± 1.92 , respectively. The mean diameter of the left and the right pulmonary artery was 16.71 ± 4.19 mm and 20 ± 5.71 mm, respectively. The mean diameter of the ascending aorta and regurgitation fraction was 30.66 ± 5.03 mm and $14.53 \pm 6.46\%$, respectively.

According to the ACC/AHA guideline [9] and recommendations of Geva [11], 73 patients were referred to PVR (59 for percutaneous and 14 for surgical PVR) (Fig. 1). During the PVR, various transcatheter or surgical procedures were also performed. Balloon dilatation and/or stenting of branch pulmonary arteries were performed in 12 patients and ventricular septal defect closure was performed in four patients at the same session of percutaneous PVR (Fig. 1).

Fourteen patients were found eligible for surgical PVR, hence, the anatomy of the RVOT is unsuitable for the percutaneous approach and additional lesions indi-

cated surgery. At the time of the surgical PVR, surgical repair of partial anomalous pulmonary venous return was performed in one patient, ventricular septal defect was performed in two patients, and subaortic membrane was performed in one patient (Fig. 1). An implantable cardioverter-defibrillator was also performed in one patient. The mean systemic-to-pulmonary flow ratio was 1.67 ± 0.68 in six patients with ventricular septal defect and was 1.5 in a patient with partial anomalous pulmonary venous return calculated by CMR (Fig. 1).

Fourteen patients were found to have additional congenital cardiac anomalies (four patients had persistence left superior vena cava, two patients had subaortic membrane, one patient had left ventricular non-compaction cardiomyopathy, right ventricular non-compaction cardiomyopathy biventricular non-compaction cardiomyopathy, interrupted inferior vena cava-azygos continuation, mitral valve prolapse, bicuspid aortic valve, myocardial crypt, and partial anomalous pulmonary venous return, respectively) (Fig. 5).

DISCUSSION

The right heart failure with severe PR and/or pulmonary stenosis is a widely accepted criterion for PVR [8–11]. However, the appropriate timing for PVR in asymptomatic patients is still controversial [14–16]. In asymptomatic patients, it is crucial to perform PVR before irreversible RV remodeling is seen [5, 14, 17]. Therefore, the role of the CMR becomes a decision-making choice with the guidance of management and planning of the PVR according to listed criteria [8–11, 18–20].

Several options such as surgical, percutaneous, or hybrid approaches exist for PVR [21]. However, the 2020 ESC Guidelines for the management of adult congenital heart disease recommended transcatheter and surgical PVR under the same indications [10].

Surgical PVR with the repairment of RVOT aneurysm and proximal branch pulmonary artery stenoses is accepted as a gold standard technique for PR [22]. However, requiring sternotomy, cardiopulmonary bypass, and increased risk of bleeding limit the technique [23].

On the other hand, percutaneous PVR is an alternative less invasive technique in selected patients with tubular-shaped, non-aneurysmatic RVOT, RV-PA conduits, and bioprosthetic valves. Non-compressed coronary artery, diameter between 14 and 25 mm in native RVOT [24], and smaller than 16 mm in conduits [25] are the main indications for percutaneous PVR.

Herein, we collaboratively assessed the CMR findings of 165 asymptomatic patients with repaired TOF according to the ACC/AHA guideline [9], of whom 73 patients were found eligible for percutaneous PVR and 14 patients for surgical PVR.

Size and Function of the RV

Evaluation of RV size and function is the most common indication for referrals of CMR in asymptomatic patients, because of the principal advantage of CMR over transthoracic echocardiography with the gold standard, accurate quantification of biventricular size and function [26].

The ACC/AHA, ESC, and CCS guidelines [8–10] require the listed degree of PR or pulmonary stenosis and one additional listed criterion, whereas the Geva suggests a more aggressive approach as two additional listed criteria [11]. In many studies, it is stated that the improvement of cardiac parameters is irreversible if indexed right ventricular end-diastolic and end-systolic volume higher than 160–170 ml/m² and 80–85 ml/m² at the time of the procedure [15, 27–29]. In our study group, mean RV-EDVI, RV-ESVI, and RV-EF that referred to PVR were 187.64±45.07 ml/m², 113.69±33.29 ml/m², and 39.90±6.60%, respectively. Hence, change in ventricular volume and function with serial examination is crucial for the management of the timing of the PVR during lifelong follow-up.

Functional and Anatomical Evaluation of Pulmonary Arteries

Moderate-to-severe PR as required the surgical technique of patch augmentation of RVOT is usually seen in postoperatively [6, 14]. And also, stenosis and/or aneurysm in any segment from RVOT-to-branch pulmonary arteries required surgical or transcatheter procedure can be seen in late after the initial surgery [30, 31]. CMR is the gold standard imaging tool in the assessment of PR fraction and preferential blood with the separate assessment of the left and right pulmonary artery flow to each lung by phase-contrast sequences [22, 32]. Percutaneous PVR is not recommended in severe branch pulmonary artery stenosis causing impaired blood flow more than 30% toward to the affected lung by Geva [11]. And also, blood flow asymmetry between both lungs (at least 35: 65%) is indicated for pulmonary artery stenting or balloon dilatation [33]. Hence, preferential pulmonary blood flow with percentage to each lung has significant importance. The blood flow ratio of the right-to-left pulmonary was mentioned to be between 61/39 and 43/57 (approximately between 1.56 and 0.75) in usual in an MRI study [23]. In our study, blood flow ratio of the right-to-left pulmonary artery was calculated as 2.25±1.92 in patients referred to PVR. That means the patients had impaired blood flow toward the left pulmonary artery.

When we assessed the three-dimensional volume rendered images, saccular aneurysmatic dilatation of the main pulmonary artery was observed in three patients and RVOT dilatation extended to the left pulmonary artery in 12 patients. The mean diameter of the left and right pulmonary arteries was measured as 16.71±4.19 mm and 20±5.71 mm in patients that we referred to PVR. During the percutaneous PVR, balloon dilatation and/or stenting of branch pulmonary arteries were performed to relieve the pulmonary artery flow in 12 patients by a percutaneous approach according to pulmonary artery flow analysis and anatomic evaluation on phase-contrast images and magnetic resonance angiography. Assessment of the stenosis and the blood flow ratio of the right-to-left pulmonary are important to evaluate the necessity of any surgical or percutaneous procedure during the long-term follow-up.

Diastolic Dysfunction and Restrictive Physiology

Diastolic dysfunction of the RV is suggested to be associated with multiple anatomical and functional abnormalities such as ventricular volume overload, valvular in-

sufficiency, systolic dysfunction, pulmonary stenosis, or increased pulmonary arterial resistance [34]. Moreover, the end-diastolic forward flow of the pulmonary artery was reported to be correlated with the severity of PR, reduced exercise capacity, and arrhythmia [35]. During the assessment of pulmonary artery flow analysis, the end-diastolic forward flow was found in 79 patients. All the patients that we referred to PVR in our study had end-diastolic forward flow. We suggest that not only the PR fraction but also the presence of the reverse flow in the late diastolic phase should be assessed on CMR examination to evaluate diastolic dysfunction, follow-up functional status before, and post-procedural period.

Residual Intracardiac Shunts

Residual septal defects can be visualized by the signal void by cine images. In CMR evaluation, residual septal defect was revealed in 15 patients (six patients had atrial and nine patients had ventricular septal defect) by morphologic evaluation on cine images. The systemic-to-pulmonary flow ratio can also be calculated to assess the degree of the left-to-right shunt by phase-contrast CMR [36]. The mean systemic-to-pulmonary flow ratio was also calculated as 1.67 ± 0.68 for six patients with ventricular septal defect. Closure of residual ventricular septal defect was performed during the PVR (four patients percutaneously and two patients surgically). The presence of a residual septal defect with the degree of systemic-to-pulmonary flow ratio calculated by CMR should be mentioned on the radiology report to guide the clinicians for the additional procedures and the technique of the procedure.

Tricuspid Regurgitation

Almost 10% of patients after surgical repair of TOF have reported having at least moderate tricuspid insufficiency because of the progressive right ventricular dilatation and concomitant tricuspid valve annulus enlargement and may be associated with disease progression [6, 18]. Although threshold for classifying the degree of tricuspid regurgitation on cardiac MRI is not well described, thresholds determined in echocardiography can still help to diagnose severity. In mild tricuspid regurgitation, the jet extends up to 2 cm into the right atrium. In moderate regurgitation, it extends 3–5 cm [37]. In our study, we evaluated the presence of atrioventricular valve insufficiency in four-chamber cine images according to signal void through regurgitation flow, and 90 patients were found to have moderate tricuspid regurgitation.

Aortic Regurgitation and Size of the Ascending Aorta

Dilatation of the aorta complicated with progressive aortic regurgitation is more commonly seen in repaired TOF and a surgical approach is recommended if the aortic root size is greater than 5.5 cm [38, 39]. CMR is the appropriate imaging modality for quantifying aortic regurgitation and evaluating aortic root dimension and it's recommended to follow up on the size and the regurgitation fraction of the aorta after initial repair [3]. In our study, the mean diameter and regurgitation fraction of the aorta were found as 30.66 ± 5.03 mm and $14.53 \pm 6.46\%$ in patients referred to PVR. Although the mean diameter of the aorta is <5.5 cm and the aortic regurgitation fraction was not severe, we recommended following anatomical and physiological changes by CMR during follow-up.

Myocardial Contrast Enhancement

MRI has the ability to assess either focal or interstitial fibrosis and is found to be associated with the risk of RV dysfunction, ventricular arrhythmias, and sudden cardiac [18, 40]. Late gadolinium enhancement on RV free wall extends to the right atrial wall and outflow tract, interventricular septum, and right ventricular insertion point with ventricular septal defect patch area were observed in only one patient. The presence of myocardial delayed enhancement should be included in the protocol in case of the clinical indication.

The Other Indications

Recommendation listed by Geva [11], as indicators for PVR suggest a cutoff of longer than 160 ms. The mean QRS duration of the patients was calculated as 170.2 ± 16.89 ms that we referred to PVR. Two patients were found to have bigeminy extrasystole, 10 patients were found to have right bundle branch block on Holter echocardiography, and one patient had an implantable cardioverter-defibrillator during surgical PVR.

Besides these factors, age at PVR is one of the most important predictor factors for post-operative death and sustained ventricular tachycardia [18]. The mean age of the patients that we referred to PVR was 18.88 ± 5 years. The retrospective study 452 patients in the study by Valente and Geva reported the threshold of age at PVR as 25 years [41].

CMR examination for symptomatic patients should be indicated routinely every 3 years with the combination of echocardiography according to guidelines [8–10].

To the best of our knowledge, this is the first study with the largest CMR examination of repaired TOF in our country. However, our study is limited by its retrospective design and lack of CMR findings after PVR. Further prospective CMR studies should be planned to assessing the outcomes of PVR.

Conclusion

Moderate-to-severe pulmonary PR is usually seen after repairment of TOF and requires PVR. In asymptomatic patients, requirement of PVR should be assessed by CMR following the listed criteria. A detailed functional and morphological assessment by CMR following the initial surgery plays an important role in the decision of PVR. Moreover, a collaborative assessment will promote a timely replacement of pulmonary valves and prevent unnecessary procedures. We believe that our experience with the largest patient population in our country may contribute to the national data pool and provides important clues for the widespread use of CMR and multidisciplinary cooperation in the follow-up of repaired TOF.

Acknowledgements: We would like to thank Prof. Dr. Adnan Kabaalioglu, Dr. Ali Yikilmaz for his support in setting up the cardiac MRI unit and cardiac MRI technologists (Serhat Once, Firat Avanes).

Ethics Committee Approval: The Istanbul Medeniyet University Clinical Research Ethics Committee granted approval for this study (date: 12.12.2018, number: 2018/0484).

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

Authorship Contributions: Concept – SO; Design – SO; Supervision – AC, SO; Fundings – SO; Materials – SO; Data collection and/or processing – SO, OT; Analysis and/or interpretation – IKY, SO; Literature review – SO- OT; Writing – SO- OT; Critical review – AC, IKY.

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