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Higher left ventricular stroke volume is associated with aortic dilatation in repaired tetralogy of Fallot patients

Yuki Kawasaki[1,2], Daiji Takajo[2,3], Pooja Gupta[2] and Sanjeev Aggarwal[2]

[1]Department of Pediatric Cardiology, Osaka City General Hospital Pediatric Medical Center, Osaka, Japan; [2]Division of Cardiology, Carman and Ann Adams Department of Pediatrics, Children's Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA and [3]Heart Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

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Corresponding author:
Daiji Takajo; Email: daiji.takajo@cchmc.org

Abstract

Aortic root dilation has been reported commonly after repair of tetralogy of Fallot. However, the rate and risk factors of progression of the dilation are not fully understood. This is a single-centre, retrospective study to assess the rate and factors associated with progressive dilatation of the aortic root in repaired tetralogy of Fallot patients using cardiac MRI. The presence of the significant aortic dilation and the progression of dilation between initial and follow-up cardiac MRI were examined. The study cohort comprised 72 patients with repaired tetralogy of Fallot. The median age at the initial cardiac MRI scan was 19.6 (interquartile range: 14.6–31) years, and the median follow-up interval was 4.3 (2.9–5.7) years. Median dimension of ascending aorta at initial and follow-up cardiac MRI was 27.0 (22.3–31.0) mm and 29.2 (25.0–32.1) mm, respectively. Significant aortic dilation (the percentage predicted ascending aorta ≥150%) was observed in 11 (15.2%) patients at the initial cardiac MRI and 24 (33.3%) at the follow-up cardiac MRI. The significant aortic dilation at follow-up cardiac MRI was associated with increased indexed left ventricular stroke volume (odds ratio 1.062, $p = 0.023$). Thirteen patients demonstrated the significant progressive dilation of aorta between initial and follow-up cardiac MRI. The progressive dilation was associated with left ventricular ejection fraction at initial cardiac MRI (odds ratio 1.135, $p = 0.048$). In patients with repaired tetralogy of Fallot, aortic dilation is common and progresses over time. Cardiac MRI is a valuable tool for identifying individuals at risk for progressive aortic dilation.

Introduction

Tetralogy of Fallot is one of the most common cyanotic CHDs, occurring in approximately 1 in 3,500 births and accounting for 7–10% of all CHDs.[1] Aortic dilation is commonly reported in patients with repaired tetralogy of Fallot. While the precise mechanism of aortic dilation in this group of patients remains incompletely understood, it is believed to be multifactorial, primarily resulting from increased volume overload due to right-to-left shunting and inherent aortopathy.

Monitoring the progression of aortic dilation is crucial due to its potential complications, including aortic valve regurgitation, aortic aneurysm, and dissection. Egbe et al. reported that 312 out of 453 (69%) adult patients with repaired tetralogy of Fallot [mean age 37 ± 13 years, 216 males (49%)] demonstrated aortic aneurysm, with 12% of them presenting significant aortic aneurysm (aortic root or mid-ascending aorta dimension ≥50 mm) or aortic valve disease including aortic stenosis or regurgitation, and 15 requiring aortic surgery.[2] Although aortic dissection or rupture among patients with repaired tetralogy of Fallot has been reported in only five cases, it was noted that progressive aortic dilation appeared as the prerequisite for aortic dissection.[3] Aortic deformation in repaired tetralogy of Fallot patients should be monitored cautiously.

There are some reports that have evaluated the longitudinal changes in aortic root and ascending aorta dimensions using echocardiograms. However, aortic dimensions in adult patients with echocardiogram are not always reproducible due to image quality. Recently, cardiac MRI has emerged as the gold standard for assessing ventricular size, function, and aortic dimensions. However, there remains a paucity of data on the longitudinal changes in aortic dimensions assessed by cardiac MRI and the risk factors associated with progressive aortic dilation among children and young adults with repaired tetralogy of Fallot.

Given that cardiac MRI enables accurate measurement of aortic dimensions and right ventricle/left ventricle volumes, the primary objective of our study is to report the longitudinal changes in aortic dimensions, as measured by cardiac MRI, in patients with repaired tetralogy of Fallot. Additionally, we aim to identify demographic data and cardiac MRI parameters that serve as risk factors for progressive aortic dilation.

Materials and method

This was a single-centre, retrospective study that was approved by the Institutional Review Board. The study included patients with repaired tetralogy of Fallot who had undergone cardiac MRI at least twice between the years 2008 and 2019. Patients with poor image quality or incomplete data were excluded from the study. Additionally, patients with other CHDs or those with a pacemaker were also excluded from the analysis. In our institution, the patients with repaired tetralogy of Fallot typically undergo cardiac MRI every 2–5 years after adolescence, and the timing for cardiac MRI is decided by the primary cardiologist taking care of patient. The clinical and cardiac MRI data were collected including age, gender, height, weight, cardiac diagnosis, surgical history, dates of cardiac MRI, blood pressure measurements, medication information, and cardiac MRI parameters.

Cardiac MRI of eligible patients was evaluated by a single reader blinded to the clinical and demographic data. Cardiac MRI was performed using 1.5 T scanners (GE Medical Systems). A stack of short-axis cine images was obtained using breath-hold steady-state free precision method. Contrast-enhanced magnetic resonance-angiography data were also obtained with each MRI study. Maximum intensity projection images and cross-sectional reconstructions were obtained to measure various aortic dimensions.

Measurements of dimensions and cross-sectional area of the ascending aorta were conducted at three specific levels: the ascending aorta, sinotubular junction, and aortic sinus, following the methodology described by Kaiser et al.[4] Each measurement site was obtained using two double oblique planes. The first plane was aligned with the longitudinal axis of the vessel, while the second plane was set perpendicular to the first one. Subsequently, a cross-sectional view perpendicular to both longitudinal planes was generated. The diameters passing through the centre of the vessel in both planes were measured, averaged, and considered as the aortic dimension. All measurements were performed twice at each aortic site, and the averaged data were used for analysis.

The following data were also collected: left ventricular end-diastolic volume, left ventricular end-systolic volume, left ventricular stroke volume, left ventricular ejection fraction, left ventricular cardiac index, right ventricular end-diastolic volume, right ventricular end-systolic volume, right ventricular stroke volume, right ventricular ejection fraction, and right ventricular cardiac index. The endocardial borders were automatic drawn and manually adjusted on all left and right ventricular short-axis images by means of the previously validated software (Mass, MEDIS, The Netherlands). The ventricular volume and stroke volume were indexed for the body surface area (ml/m^2). The cine phase velocity pulse sequence was used to quantitate the aortic and pulmonary regurgitant fraction. We defined significant aortic regurgitation as aortic regurgitant fraction of more than 15%.[5]

The predicted dimensions of ascending aorta, sinotubular junction, and aortic sinus for the patients less than 21 years of age at the time of cardiac MRI were calculated by the following formula: ascending aorta = $-1.33 + 18.6$*body surface area$^{0.5}$, sinotubular junction = $-0.03 + 16.91$*body surface area$^{0.5}$*, and aortic sinus = $0.57 + 10.37$*body surface area$^{0.5}$ (mm) [4]. On the other hand, the predicted dimensions of ascending aorta, sinotubular junction, and aortic sinus for the patients more than or equal to 21 years of age at the time of cardiac MRI were calculated by the following formula: ascending aorta = 12.2*body surface area for male, ascending aorta = 12.0*body surface area for female; sinotubular junction = 13.4*body surface area for male, sinotubular junction = 13.6*body surface area for female; aortic sinus = 17.0*body surface area for male, aortic sinus = 16.8*body surface area for female.[6,7]

The percentage predicted for ascending aorta, sinotubular junction, and aortic sinus was calculated as a ratio of measured dimension to predicted dimension. Significant aortic dilation was defined as the percentage predicted ascending aorta (%ascending aorta) ≥150 (%). The progression of aortic dilation was defined as≥50% increase in %ascending aorta from initial cardiac MRI to follow-up cardiac MRI. The yearly rate of change from initial cardiac MRI to follow-up cardiac MRI was reported as annual growth (mm/year).

Statistical analysis was performed using SPSS version 28 (IBM SPSS Inc., Chicago, IL). Based on normality tests (Kolmogorov–Smirnov and Shapiro–Wilk), there were no normally distributed continuous variables. Therefore, continuous variables are reported as median and interquartile range while categorical variables are denoted by number and percentage. All pre-specified demographic and echocardiographic parameters between the groups were analysed using Mann–Whitney U test and Chi-square test as appropriate. Univariate logistic regression analysis was used to identify the significant parameters to predict %ascending aorta ≥150 and an ≥50% increase in %ascending aorta between initial and follow-up cardiac MRI. A $p < 0.05$ was considered statistically significant.

Results

The study cohort consisted of 72 patients. The demographics and clinical data of the entire cohort are summarised in Table 1. The median age at the time of the initial cardiac MRI scan was 19.6 years (interquartile range: 14.6–31 years), while the median age at the follow-up cardiac MRI scan was 23.7 years (19.2–34.9). The median interval between the initial and follow-up cardiac MRI scans was 4.3 years (2.9–5.7). The primary cardiac diagnosis was typical tetralogy of Fallot except 4 patients (5.6%) had pulmonary atresia. A systemic-to-pulmonary shunt was required in 26 patients (36.1%) during the neonatal period. Complete repair was performed at a median age of 1.1 years (0.5–3.2). The primary methods of complete repair included transannular patch in 60 patients (83.3%), valve-preserved repair in 7 patients (9.7%), and right ventricular to pulmonary artery conduit placement in 5 patients (6.9%). Pulmonary valve replacement was performed in 14 patients (19.4%) prior to the initial cardiac MRI scan, and during the interval between the two cardiac MRI, 30 patients (41.7%) underwent surgical pulmonary valve replacement. All the patients had tricuspid aortic valve with non or trace aortic regurgitation based on cardiac MRI. There were no patients with significant residual intracardiac shunts except for trivial shunts across ventricular septal defect that do not affect haemodynamics. All of the patients with pulmonary atresia had confluent branch pulmonary arteries.

Comparison between observed and predicted measurements of the aorta

Ascending aorta, sinotubular junction, and aortic sinus dimensions at initial cardiac MRI were 27.0 (22.3–31.0) mm, 26.0 (23.5–30.2) mm, and 30.0 (27.4–33.5) mm, respectively (Table S1). The range of ascending aorta dimension was 14.8–41.7 mm. In comparison, the median predicted values of ascending aorta, sinotubular junction, and aortic sinus dimensions

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Table 1. Demographics of the entire study cohort

Median (IQR) n (%)	$n = 72$
Age at initial CMR (years)	19.6 (14.6–31)
Age at follow-up CMR (years)	23.7 (19.2–34.9)
Interval between two CMR (years)	4.3 (2.9–5.7)
Gender (male)	34 (47)
Height (cm)	163 (155–170)
Weight (kg)	62.0 (46.0–77.3)
BSA (m^2)	1.7 (1.4–1.8)
Initial SP shunt	26 (36)
Primary Diagnosis TOF	
Pulmonary stenosis	68 (94)
Pulmonary atresia	4 (6)
Laterality of aortic arch	
Left aortic arch	48 (67)
Right aortic arch	24 (33)
Interval between SP shunt and complete repair (year)	1.2 (0.77–2.0)
Type of complete repair	
Valve-preserved repair	7 (10)
Transannular patch	60 (83)
RV-PA Conduit repair	5 (7)
Age at complete repair (years)	1.1 (0.5–3.2)
Timing of PVR	
No PVR	28 (39)
PVR before initial CMR	14 (19)
PVR between two CMR images	30 (42)
Hypertension (BP > 140/90 mmHg)	4 (6)
Medication	
beta blocker	5 (7)
ACE inhibitor	7 (10)
Ca^{++} channel blocker	1 (1)

ACE = angiotensin-converting enzyme; BSA = body surface area; CMR = cardiacovascular magnetic resonance; IQR = interquartile range; PVR = pulmonary valve replacement; RV-PA = right ventricle-to-pulmonary artery; SP = systemic-to-pulmonary; TOF = tetralogy of Fallot.

as calculated by the published equation were 21.3 (20.0–23.6) mm, 22.2 (19.9–24.3) mm, and 26.3 (23.6–29.7) mm, respectively. The measured dimension was significantly higher than predicted dimension at all levels of ascending aorta, sinotubular junction, and aortic sinus. The median percent predicted for ascending aorta, sinotubular junction, and aortic sinus dimensions were 120 (106–140) %, 117 (107–134) %, and 113 (104–121) %, respectively. Eleven patients (15.2%) had significant aortic dilation as defined as having ascending aorta dimensions ≥150% of the predicted values.

Ascending aorta, sinotubular junction, and aortic sinus dimensions at follow-up cardiac MRI were 29.2 (25.0–32.1) mm, 29.1 (25.4–31.7) mm, and 31.2 (28.4–35.2) mm, respectively. The range of ascending aorta dimension was 16–44.5 mm. The median predicted values of ascending aorta, sinotubular junction, and aortic sinus dimensions were 22.8 (19.9–24.1) mm, 25.1 (22.7–27.6) mm, and 27.7 (24.4–32.1) mm, respectively. The measured dimension was significantly higher than predicted dimension in all of ascending aorta, sinotubular junction, and aortic sinus. The median percent predicted for ascending aorta, sinotubular junction, and aortic sinus dimensions were 125 (114–145) %, 113 (99–129) %, and 115 (106–130) %, respectively. Twenty-four patients (33.3%) had %predicted ≥ 150 for ascending aorta (significant aortic dilation).

Comparison between initial and follow-up cardiac MRI

In the follow-up cardiac MRI scans, the percent predicted value for the ascending aorta (%ascending aorta) was found to be significantly higher compared to the initial cardiac MRI scans (125% vs. 120% $p = 0.004$) (Table S2). Left ventricular ejection fraction and indexed left ventricular stroke volume were significantly higher in follow-up cardiac MRI than those in initial cardiac MRI (left ventricular ejection fraction; 56.6% vs. 58.4%, $p < 0.001$, indexed left ventricular stroke volume; 39.8 vs. 42.1ml/m^2, $p < 0.001$), although they remained within normal range. On the other hand, indexed right ventricular stroke volume was significantly lower in follow-up cardiac MRI (62.9 vs. 45.0 ml/m^2, $p < 0.001$) in the context of 30 patients undergoing pulmonary valve replacement between initial and follow-up cardiac MRI. Right ventricular ejection fraction was similar between the two (40.5% vs. 41.6%, $p = 0.758$).

Comparison of demographics between patients with and without significant aortic dilation

The entire study cohort was divided into two groups based on the presence or absence of aortic dilation. The patients who exhibited a percent predicted ascending aorta (%ascending aorta) value of ≥150% at the follow-up cardiac MRI were categorised into the aortic dilation group, while those with a %ascending aorta value <150% at the follow-up cardiac MRI were categorised into the non-dilation group (Table S3). The aortic dilation group consisted of 24 patients, including 8 males (33.3%), with a median age of 22.2 years (15.9–27.5) at the time of the follow-up cardiac MRI. The non-dilation group consisted of 48 patients, including 26 males (54.2%), with a median age of 25.5 years (20.7–38.3) at the time of the follow-up cardiac MRI. The median age at follow-up cardiac MRI among non-dilation group is significantly older than those among aortic dilation group. However, there were no statistically significant differences in other demographic variables between the two groups.

Comparison of cardiac MRI measurements between patients with and without significant aortic dilation

As expected, %ascending aorta among aortic dilation group was found to be significantly higher in the aortic dilation group compared to the non-dilation group (172% vs. 117%, $p < 0.001$) (Table S4). Similarly, %sinotubular junction and %aortic sinus among aortic dilation group were also significantly higher than those among non-dilation group (%sinotubular junction; 147% vs. 99%, $p < 0.001$, %aortic sinus; 141% vs. 102%, $p < 0.001$). No statistically significant differences were observed in ventricular volume, ejection fraction, stroke volume, and cardiac index at initial MRI between the two groups. Indexed left ventricular stroke volume and left ventricular cardiac index at follow-up MRI were significantly higher in the aortic dilation group compared to the non-dilation group (46.8 vs. 40.1 ml/m^2,

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Table 2. Univariate logistic regression analysis to predict significant aortic dilation (%AA ≥150) at follow–up CMR

	Univariable analysis	
Predictors	Odds ratio (95% CI)	*P* value
Gender, male	0.423 (0.152–1.175)	0.099
Age at complete repair	0.814 (0.611–1.085)	0.814
History of PVR	0.385 (0.140–1.054)	0.063
Presence of right aortic arch	0.751 (0.260–2.168)	0.596
LVEDVI at initial CMR (ml/m^2)	1.030 (0.991–1.070)	0.138
RVEDVI at initial CMR (ml/m^2)	0.993 (0.982–1.00)	0.206
LVEF at initial CMR (%)	1.058 (0.975–1.148)	0.174
RVEF at initial CMR (%)	1.008 (0.955–1.064)	0.770
LVSVI at initial CMR (ml/m^2)	1.023 (0.963–1.087)	0.452
RVSVI at initial CMR (ml/m^2)	1.005 (0.981–1.030)	0.678
LVCI at initial CMR (ml/m^2)	2.100 (0.824–5.340)	0.120
RVCI at initial CMR (ml/m^2)	1.250 (0.892–1.760)	0.194
PRF at initial CMR (%)	0.984 (0.954–1.02)	0.333
LVEDVI at follow–up CMR (ml/m^2)	1.010 (0.983–1.03)	0.660
RVEDVI at follow–up CMR (ml/m^2)	1.000 (0.993–1.01)	0.521
LVEF at follow–up CMR (%)	1.050 (0.988–1.116)	0.115
RVEF at follow–up CMR (%)	1.019 (0.973–1.068)	0.423
LVSVI at follow–up CMR (ml/m^2)	1.062 (1.008–1.119)	**0.023***
RVSVI at follow–up CMR (ml/m^2)	1.001 (0.976–1.027)	0.937
LVCI at follow–up CMR (ml/m^2)	2.340 (1.180–4.640)	**0.014***
RVCI at follow–up CMR (ml/m^2)	1.010 (0.704–1.440)	0.969
PRF at follow–up CMR (%)	1.030 (0.998–1.050)	0.065

CI = confidence interval; CMR = cardiovascular magnetic resonance; LVCI = left ventricular cardiac index; LVEDVI = left ventricular end-diastolic volume index; LVEF = left ventricular ejection fraction; LVSVI = indexed left ventricular stroke volume; PRF = pulmonary regurgitant fraction; PVR = pulmonary valve replacement; RVCI = right ventricular cardiac index; RVEDVI = right ventricular end-diastolic volume index; RVEF = right ventricular ejection fraction; RVSVI = indexed right ventricular stroke volume; %AA = the percentage predicted ascending aorta.
*Indicates $p < 0.05$.

$p = 0.017$, 3.31 vs. 2.59 ml/min/m^2, $p = 0.00469$, respectively). Univariate regression analysis was performed to identify the significant parameters to predict %ascending aorta ≥150 (Table 2). Indexed left ventricular stroke volume and left ventricular cardiac index at follow-up cardiac MRI were significant parameters. The remaining parameters were not statistically significant.

Comparison between patients with and without progressive dilation

The entire cohort of 72 patients was divided into two groups based on the change in %ascending aorta between the initial cardiac MRI and follow-up cardiac MRI. Patients with a 50% or greater increase in %ascending aorta from the initial cardiac MRI to the follow-up cardiac MRI were classified as the progressive group, while the remaining patients were classified as the non-progressive group. Among the 72 patients, 13 exhibited progressive dilation. Demographic characteristics did not show any statistically significant differences between the progressive and non-progressive groups (Table S5). Table S6 presents a comparison of cardiac MRI parameters at the initial and follow-up cardiac MRI between the progressive and non-progressive groups. In the progressive dilation group, a median dilation rate of 3.68 mm/year (2.84–5.02 mm/year) was observed. Univariate regression analysis was performed to identify the significant parameters to predict the progressive dilation (Table S7). Left ventricular ejection fraction and left ventricular cardiac index were the significant parameters (odds ratio 1.135, $p = 0.048$, 4.180 $p = 0.022$, respectively). The remaining parameters were not statistically significant.

Discussion

Our study suggested that not a small number of patients (18%) demonstrated the progressive dilatation in aortic dimension within short follow-up period and 33% of patients reached our definition of significant aortic dilation (%ascending aorta ≥150%) at follow-up cardiac MRI. Our data have consistently shown that both progressive dilation (≥50% increase in %ascending aorta at follow-up), and significant aortic dilation (%ascending aorta≥150%) are associated with left ventricular haemodynamics.

There are some reports of aortic dilation in children with diagnosis tetralogy of Fallot.[8,9] Historically, volume overload, in the context of right ventricular outflow tract obstruction and right-to-left shunt through ventricular septal defect, has been considered a contributing factor to aortic dilation in children with unrepaired tetralogy of Fallot, and the aortic size would stabilise once the defect is repaired. In fact, a study has shown significant regression of indexed aortic diameters after early complete repair of tetralogy of Fallot.[10]

However, an increasing body of evidence, including our study, supports the idea that patients with repaired tetralogy of Fallot can develop progressive and significant aortic dilation. The mechanism of aortic dilation among patients with tetralogy of Fallot after complete repair is not yet clear. Histological changes such as fibrosis, cystic medial necrosis, and elastic fragmentation have been observed in the ascending aorta of patients with tetralogy of Fallot, even in infancy.[11] Additionally, a study conducted in the catheterisation laboratory suggested that the degree of dilation of the ascending aorta was significantly correlated with the decreased distensibility of the ascending aorta. Interestingly, this finding was not observed in the descending aorta.[12] Therefore, patients with tetralogy of Fallot may have histological changes limited to the ascending aorta, leading to stiffness. A stiff ascending aorta is associated with the loss of load-bearing characteristics and a reduced capacity for elastic recoil, resulting in gradual plastic deformation and outward expansion of the aorta.[13] Our study suggests that the ascending aorta in patients with tetralogy of Fallot has less tolerance for increased left ventricular stroke volume.

Only a few articles have analysed cardiac MRI indices of ventricular volume, function, and flow volume as risk factors for aortic dilatation.[9,14,15] Dennis et al. reported that 23% of patients had aortic root dilatation (>40 mm) as evaluated by cardiac MRI. Their study suggested that male gender is the only consistent significant factor contributing to aortic root dilation. However, the study also implied that higher left ventricular stroke volume was associated with aortic dilation because patients with aortic dilation had a larger left ventricular stroke volume compared to patients without dilation (90 ml vs. 79 ml, $p = 0.04$), although this association was not proven by regression analysis.[14] Our data support this implication, which demonstrated consistent association between left ventricular stroke volume and significant aortic

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dilation. On the contrary, our study did not show any gender difference in aortic dilation.

Longitudinal rate of change of aorta in repaired tetralogy of Fallot patients has reported in a few studies to best to our knowledge.[2,13,16] Siripornpitak et al. reported the findings of cardiac MRI assessments of aortic diameter in 94 patients with repaired tetralogy of Fallot (mean age 14.5 ± 4.4 years), observing slow aortic growth in 78-85% of patients during the median interval of 52 months (24–71 months). The median annual growth rate ranged from 0.37 mm (0.13–0.72) at annulus to 0.56 mm (0.22–0.91) at the ascending aorta.[17] Another study by Bonello et al. evaluated 110 patients [median age 30.9 years (22.9–39.4)] with repaired tetralogy of Fallot using cardiac MRI and found that 69% of the patients had aortic dilation, with varying degrees at different levels. Over a median follow-up period of 6.3 years, aortic diameters increased in 47% of the patients.[18] Egbe et al. reported the single-centre, retrospective review of adults with repaired tetralogy of Fallot, including 453 patients (mean age; 37 ± 13 years). A total of 52 (12%) patients had significant aortic aneurysm, defined by aortic root/sinus or Valsalva or mid-ascending aorta dimension ≥50 mm, at baseline. Of the 52 patients with significant aortic aneurysm, 15 (29%) patients underwent aortic surgical intervention. Among rest of 37 patients who did not require intervention, 4 (11%) patients showed progressive aortic dilation, defined by increase in aortic dimension ≥2 mm. For those who did not have significant aortic aneurysm at baseline (401 patients), progressive aortic dilation was seen in 36 (9%) patients at follow-up.[2]

Our longitudinal observation revealed that out of 72 repaired tetralogy of Fallot patients, 42 (58%) showed an increase in % ascending aorta at follow-up, with 13 (18%) developing a significant increase in %ascending aorta by ≥50% in a relatively short-term follow-up. Identifying risk factors to predict this progressive dilation group is particularly important in determining whether patients need close and frequent follow-up for aortic dilation. In our study, left ventricular ejection fraction and left ventricular cardiac index at the initial cardiac MRI were associated with this, although its clinical significance remains unknown. Further study is warranted to identify additional risk factors.

Currently, there is no verified consensus threshold for surgical intervention in tetralogy of Fallot patients with a dilated aorta. For patients with a bicuspid aortic valve, the 2020 ACC/AHA guidelines recommend a reduced threshold for surgery of 50 mm with risk factors, including a growth rate ≥5 mm/year for the maximal aortic diameter, or 55 mm otherwise.[19] Given the limited number of case reports of aortic dissection or ruptures in patients with tetralogy of Fallot,[20] the incidence of aortic dissection is believed to be lower compared to patients with other aortopathies such as Marfan syndrome.[21] Therefore, preventive surgery for aortic dilation according to the current ACC/AHA guidelines may result in over-treating patients with tetralogy of Fallot, although there is a lack of data to support this. The optimal timing and need for intervention on a dilated aortic root in these patients require further research.

This single-centre, retrospective study has a few limitations. First, due to the lack of a follow-up protocol in our institution, the intervals between the initial and follow-up cardiac MRI exams were inconsistent among patients. Additionally, our study primarily included young adults with limited follow-up. A longer follow-up period is warranted in future studies. Due to variations in patients' age and body size, we used predicted percentages for aortic dimensions to assess significant and progressive aortic dilation. We had to employ two different prediction models based on the patients' age, which may potentially result in over- or underestimation of the predicted aortic dimensions. Inter-observer and intra-observer variability in MRI measurements were not assessed at this time.

Progressive and significant aortic root dilation was observed in some young adults with repaired tetralogy of Fallot. Although preserving left ventricular systolic function is favourable in general, ejecting higher stroke volume against pathologic, stiff ascending aorta may cause significant dilation of ascending aorta. Identification of risk factors for progressive aortic dilation and follow-up with advanced imaging at appropriate timing are important considerations in patients with repaired tetralogy of Fallot.

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All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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