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# Echocardiography vs cardiac magnetic resonance imaging assessment of the systemic right ventricle for patients with d-transposition of the great arteries status post atrial switch

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## Abstract

**Objective:** Patients with Dextro-transposition of the great arteries status post atrial switch (dTGA s/p atrial switch) are "at-risk" for systemic right ventricular (RV) dysfunction. Due to complex RV geometry, echocardiography (Echo) does not allow accurate determination of ejection fraction (EF), but cardiac magnetic resonance imaging (CMR) allows quantitative right ventricular assessment. Measures of ventricular deformation may be precursors to global ventricular dysfunction. The primary aim of this study was to characterize imaging and clinical findings for adult patients with dTGA s/p atrial switch.

**Design:** This was a retrospective cohort study of patients with dTGA s/p atrial switch operation (February 1966 to August 1988) with CMR performed at Children's Hospital of Wisconsin (from September 2005 to May 2015). Eligible patients had clinic visit, Echo, and exercise stress test within 1 year of CMR.

**Results:** This study enrolled twenty-seven patients (16 males, 11 females) with dTGA s/p atrial switch (18 with Mustard operation and 9 with Senning operation; median age 30 years; 74% New York Heart Association class 1 and 26% class 2). Seventy-four percentage had normal RV systolic function (RV EF >45% by CMR). No correlation was observed between Echo strain data and clinical status (EF, exercise endurance, $VO_2$ max, or New York Heart Association class). Cardiac magnetic resonance imaging RV global circumferential strain GCS and RV EF had moderate negative correlation ($r = -0.65$, $P < .001$). Global circumferential strain was significantly different for those with RV EF above and below 45%, while global peak longitudinal strain (GLS) was not. Patients had reduced CMR myocardial strain values compared with healthy controls.

**Conclusions:** Reduced RV CMR GCS (for those with RV EF <45%) suggests that CMR evaluation may enhance early detection of detrimental changes in the systemic RV myocardium.

KEYWORDS

cardiac magnetic resonance imaging (CMR), d-transposition of the great arteries (dTGA), myocardial strain

  

## 1 | INTRODUCTION

Dextro-transposition of the great arteries (dTGA) occurs when there are concordant atrio-ventricular and discordant ventriculo-arterial relationships leading to systemic cyanosis. Repair by an atrial switch operation (either a Mustard or Senning procedure) was crucial for survival in the cardiovascular surgical era before the refinement of the arterial switch procedure in the 1980s.[1] The atrial switch operation redirected systemic venous and pulmonary venous blood flow, via baffled pathways through the atria (using native atrial tissue in the Senning or pericardial patch material in the Mustard operation). Thus, the atrial switch operation allowed deoxygenated systemic venous blood to flow to the left ventricle (LV) and main pulmonary artery (MPA), while oxygenated pulmonary venous blood was redirected into right ventricle (RV) and aorta (Ao). In this manner, the RV became the systemic ventricle for a patient after atrial switch operation.

Many young adults with dTGA s/p atrial switch are alive today. Survival rates are estimated to be 76% by 20 years of age with mean age at death being 27 years.[2,3] Although no registry exists, it is estimated that at least 3500 patients with dTGA s/p atrial switch are alive today.[4] Long-term problems are encountered after atrial switch. These include anatomic issues (eg, baffle obstruction and leakage, LV outflow tract obstruction, declining systolic function for the systemic hypertrophic RV) which may lead to clinical problems (eg, decreased exercise tolerance, dysrhythmia, and congestive heart failure).[5-7] Mechanisms of morbidity and mortality for patients with dTGA patients s/p atrial switch are poorly characterized, although myocardial fibrosis may be related to the systemic RV dysfunction.[8]

Cardiac magnetic resonance imaging (CMR) is the gold standard for quantitative assessment of the RV in many congenital heart diseases (eg, Tetralogy of Fallot) and in some centers has been used for those with dTGA s/p atrial switch.[9,10] Echocardiography (Echo), though, is extensively and routinely used for clinical assessment, because it is readily accessible. Echocardiography assessment of RV systolic function is typically subjective, because the complex RV geometry does not allow accurate quantitative determination of ejection fraction (EF).[9] Simpson's formula, which can be applied to the LV, cannot be accurately applied to the RV to calculate RV EF.

Velocity vector imaging (VVI) is a novel way to quantitatively assess RV function based on 2D speckle tracking Echo data. Analysis by VVI allows for ventricular (myocardial) strain measurement by endocardial border tracking that is independent of the angle of the ultrasound probe. Increasingly, strain data are moving from a research interest to clinical practice.[11-13] Of the VVI measures, global peak longitudinal strain (GLS) may be the best overall assessment of myocardial health with a few small studies showing decreased RV GLS associated with increased NT-BNP and worsening clinical status for patients with a systemic RV.[13-16] Recently, regional myocardial strain analyses by CMR have become feasible. Little is known, though, about how myocardial strain is correlated with the clinical status of patients with dTGA s/p atrial switch.[14,15]

The primary aim of this imaging research was to characterize CMR and Echo findings for patients with dTGA s/p atrial switch who have a systemic RV. Secondarily, correlations were sought between these imaging data and clinical status. We hypothesized that RV GLS, in patients with dTGA s/p atrial switch procedure, predicts RV EF.

## 2 | METHODS

### 2.1 | Study population

This was a retrospective cohort study with approval from the Children's Hospital of Wisconsin Institutional Review Board (IRB). Clinical data were reviewed for patients with dTGA s/p atrial switch (performed between February 1966 and August 1988), having CMR performed at Children's Hospital of Wisconsin (September 2005 to May 2015). A primary diagnosis of dTGA s/p atrial switch procedure was confirmed in the medical records. Patients were considered eligible for this research, if they had a clinic visit, Echo, and Bruce protocol exercise stress test within 1 year of their CMR scan. Other clinical data points extracted from the medical record included: secondary diagnoses, age at atrial switch, operations/procedures before atrial switch, operations/procedures after the atrial switch, New York Heart Association (NYHA) class, and medications.

### 2.2 | Cardiac magnetic resonance imaging

Clinically indicated CMR scans were performed on 1.5T or 3T Siemens magnets (Avanto, Symphony, and Skyra, Siemens Medical Solutions USA, Malvern PA), according to standard clinical imaging protocol to allow anatomic definition of the atrial baffles, determination of ventricular volumes and systolic function, and quantification of vascular blood flow. The short-axis ventricular stack was imaged with the following parameters: (FOV 192 × 156 or better, slice thickness 7-9 mm, repetition time 45-75 msec, echo train length (TE) 1-2 msec, and NEX 1) using either steady state-free precession (SSFP) or gradient echo (FLASH) cine imaging, as clinical practice varied in some cases. Blinded examination of the CMR data were undertaken by two independent cardiologists with CMR expertise (BG and MS) using Circle Cardiovascular CVI 42 software (Circle Cardiovascular Imaging, Calgary Alberta, Canada) for planimetry of the ventricular short-axis stack according to standard methods.[17] Assessment of RV mass indices was performed in two ways for the short-axis stack of images—first with attribution of the interventricular septal (IVS) mass to the LV mass calculation and second with the IVS given to the RV mass calculation. RV mass/volume ratios, calculated by the two methods, were compared (Figure 1). Finally, regional myocardial mechanics was assessed by a third reader (KH) using Diogenes CMR FT software Version 4.6 (TOMTEC Imaging Systems, Munich, Germany).

### 2.3 | Echocardiography

Echocardiography (Echo) was performed using a Sequoia ultrasound system (Siemens Medical Solutions USA, Malvern, PA) using

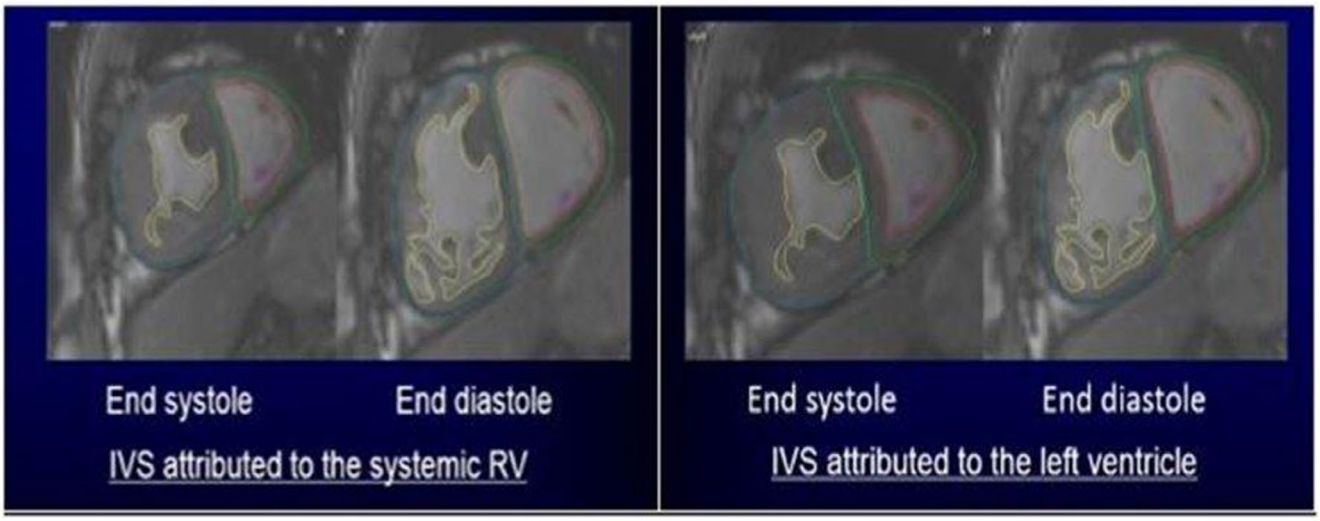


**FIGURE 1** Planimetry of short-axis plane with interventricular septum attributed to the right ventricle (RV mass index = 119 g/m$^2$) vs. to the left ventricle (LV mass index = 100 g/m$^2$) (Siemens 1.5T Avanto scanner)

a standard EKG-triggered digital capture of 2-dimensional, color flow, and Doppler flow per institutional protocol. Echocardiography speckle tracking VVI analyses were performed by a reader (CM with overread by cardiologists SC and DS, who were blinded to CMR feature tracking data); these analyses were made using TOMTEC Cardiac Performance Analysis software (TOMTEC Corporation, Chicago IL). Single beat acoustic capture digital cine images (with frame rates 70-120 per cardiac cycle), best visualizing the entire RV, were used for analyses, with manual planimetry of the subendocardial border by methods described previously.[18] Longitudinal strain and strain rate were determined by tracing the endocardial border of the RV and septal wall from an apical four chamber view. The RV myocardium was partitioned into six segments including: basal RV free wall, mid RV free wall, and apical RV free wall with corresponding basal, mid, and apical RV septal regions. Circumferential RV strain and strain rate were determined for the RV, using a short-axis window focused on the RV at the region of the LV papillary muscles.

Echocardiography anatomic images were also reviewed by physician readers (SC and DS blinded to CMR data) to determine the presence of pulmonary stenosis (peak gradient greater than 25 mm Hg), left ventricular outflow tract obstruction (greater than 25 mm Hg), and tricuspid regurgitation (moderate or greater). Echocardiography measures of ventricular diastolic function were also assessed (SC and DS) by Doppler and tissue Doppler imaging techniques, as described by others.[19] The myocardial performance index (MPI) was calculated as previously described, as the sum of the isovolumic relaxation time (IVRT) and the isovolumic contraction time (IVCT) divided by the RV ejection time.[20-22]

## 2.4 | Statistical analyses

Medians and ranges were calculated for continuous demographic variables, while frequencies and percentages for categorical variables. For the agreement between two readers, scatterplots and Bland–Altman plots were generated, and the concordance correlation coefficients (CCC)[23] were calculated for each pair of variables. For the correlation between two variables, scatterplots and Pearson's correlation coefficients were calculated. Mann-Whitney test was used to compare the two groups of subjects (ie, those with CMR-derived RV EF < 45% and RV EF > 45%). *P* value < .05 was considered as statistically significant.

**TABLE 1** Demographics

| | dTGA *N* = 27 |
|---|---|
| Gender (male) (*n*, %) | 16 (59%) |
| Age at atrial switch (years) | 0.9 (0.01-10.50) |
| Age at CMR scan (years) | 30 (19-50) |
| Race (*n*, %) Caucasian | 26 (96%) |
| Other | 1 (4%) |
| Weight (kg) | 78.1 (54.0-134.9) |
| Height (cm) | 171.5 (155.0-200.0) |
| BSA (m$^2$) | 1.92 (1.54-2.54) |

*Note:* For continuous variables, medians with ranges are displayed.

## 3 | RESULTS

Twenty-seven patients (16 male) with dTGA s/p atrial switch (9 Senning and 18 Mustard procedures) were enrolled. Demographic and anatomic data are shown in Tables 1 and 2. Few interventions were required for other congenital heart issues (ie, ventricular septal defect repair (four patients) and coarctation repair (one patient). Median age at atrial switch was 0.9 years (range 0.01-10.50 years). Atrial baffle reconstruction occurred for a minority (4/27 patients). The median age at CMR scan was 30 years (range 19-50 years). The patients were generally healthy (74% NYHA class 1% and 26% class 2). Medication use varied greatly among the patients (Table 3) with five patients not taking any

chronic medications. During Bruce protocol treadmill exercise stress test, 89% had an induced arrhythmia, although most were minor (ie, isolated premature atrial contractions, isolated premature ventricular contractions, with rare short runs of ectopy). None required anti-arrhythmic medication long-term, and only one patient had a pacemaker/AICD (placed after CMR). The median endurance on Bruce protocol treadmill testing was 9.0 minutes (range 3.7-12.6 minutes) with median $VO_2$ max was 2.1 L/min (range 1.0-3.7 L/min).

For the whole cohort, using the original clinical data, most patients (74%) had normal RV systolic function with RV EF > 45% by CMR. The systemic RV was hypertrophied when either method (ie, IVS attributed to RV or IVS attributed to LV) of assessing the RV mass index was employed. With IVS attributed to RV, the RV mass index was 94.0 g/m$^2$ (range, 57.0-200.3 g/m$^2$); when IVS was attributed to LV, RV mass index was 78.1 g/m$^2$ (range 49.2-168.0 g/m$^2$). The RV appeared moderately dilated (>120 ml/m$^2$) for two subjects (123 and 136 ml/m$^2$) and severely dilated for one (177 ml/m$^2$), but otherwise was normal or only mildly enlarged. The pulmonic (morphologic left) ventricle typically had normal to hyperdynamic systolic function (median LV EF = 68%, range 27%-86%) with only one patient having reduced LV systolic function.

Cardiac magnetic resonance imaging data were analyzed by two independent reviewers. Despite one outlier (subject 22), reasonable correlation existed between these two readers for most CMR variables. Concordance correlation coefficient[23] was 0.75, 0.82, and 0.82 for RV EDV index, RV ESV index, and RV EF, respectively. Concordance correlation coefficient was 0.79, 0.91, and 0.87 for LV EDV index, LV ESV index, and LV EF, respectively. Not surprisingly, there was slightly lower agreement for RV mass index determination, where CCC was 0.60 if the interventricular septum was attributed to the RV and 0.79 if attributed to the LV (Figure 2).

**TABLE 2** Cardiac anatomic and operative data

| | *N* (%) |
|---|---|
| Secondary diagnosis | 5 (18.5) |
| VSD | 4 (14.8) |
| Coarctation | 1 (3.7) |
| Senning procedure | 9 (33.3) |
| Mustard procedure | 18 (66.7) |
| Operation before atrial switch | 11 (40.7) |
| Septectomy | 8 (29.6) |
| PA band | 2 (7.4) |
| Coarctation repair | 1 (3.7) |
| Atrial septal defect | 27 (100) |
| No (ASD at birth) | 3 (11.1) |
| Septostomy | 16 (59.3) |
| Septectomy | 8 (29.6) |
| Operation after atrial switch | 4 (14.8) |
| Reconstructed systemic venous baffle | 1 (3.7) |
| Reconstructed pulmonary venous baffle | 2 (7.4) |
| Reconstructed both baffles | 1 (3.7) |

Cardiac magnetic resonance imaging determined regional myocardial mechanics are highlighted in Table 4 and show reduced strain.

Echocardiography-derived VVI data are in Table 5 with moderate agreement between readers. Concordance correlation coefficient for GLS was 0.862 (CI 0.764-0.960) and for GCS was 0.710 (CI 0.489-0.931). Significant Echo diastolic data for the cohort are given in Table 6.

## 3.1 | Correlations between CMR data and clinical status

The only significant correlation found between CMR data and clinical status was a weak correlation between RV mass index (when IVS was given to the LV) and exercise endurance ($r$ = 0.39, $P$ = .044). No other correlations were found (for this relatively healthy cohort of dTGA s/p atrial switch patients) for any CMR parameter including CMR strain assessment and clinical status (as measured by endurance on treadmill, $VO_2$ max, or NYHA class).

### 3.1.1 | Correlations between Echo data and clinical status

No correlation was observed between Echo strain data (GLS or GCS) and clinical status, as measured by exercise endurance, $VO_2$ max, or NYHA class.

As shown in Table 6, statistically different values were seen for some Echo tissue Doppler data when those dTGA s/p atrial switch patients with reduced RV systolic function (RV EF < 45% based on clinical CMR report) were compared with those good RV systolic function (RV EF > 45%). For those with lower RV EF (<45%), the tricuspid valve S' was lower (medians 0.06 vs 0.09, $P$ = .026) and A' was lower (medians 0.05 vs 0.07, $P$ = .018), while E/E' was higher 13.0 vs 8.1, $P$ = .049). Despite these correlations with RV EF, no correlation was seen for Echo measures of diastolic function and clinical status. As highlighted in Table 7, complete Echo data were examined

**TABLE 3** Clinical outcomes

| NYHA (*n*, %) | Class 1: 20 (74%)<br>Class 2: 7 (26%) |
|---|---|
| Stress test endurance (min), $n$ = 27 | 9.0 (3.7-12.6) |
| Stress test VO2 max (L/min), $n$ = 27 | 2.1 (1.0-3.7) |
| Stress test arrhythmia (yes, %) | 24 (89%) |
| Medications (*n*, %) | 22 (81%) |
| Afterload reduction | 19 (70%) |
| Digoxin | 12 (44%) |
| Diuretic | 6 (22%) |
| Anticoagulation | 8 (30%) |
| Pacemaker/AICD (*n*, %) | 1 (3.7%) |

*Note:* For continuous variables, medians with ranges are displayed.

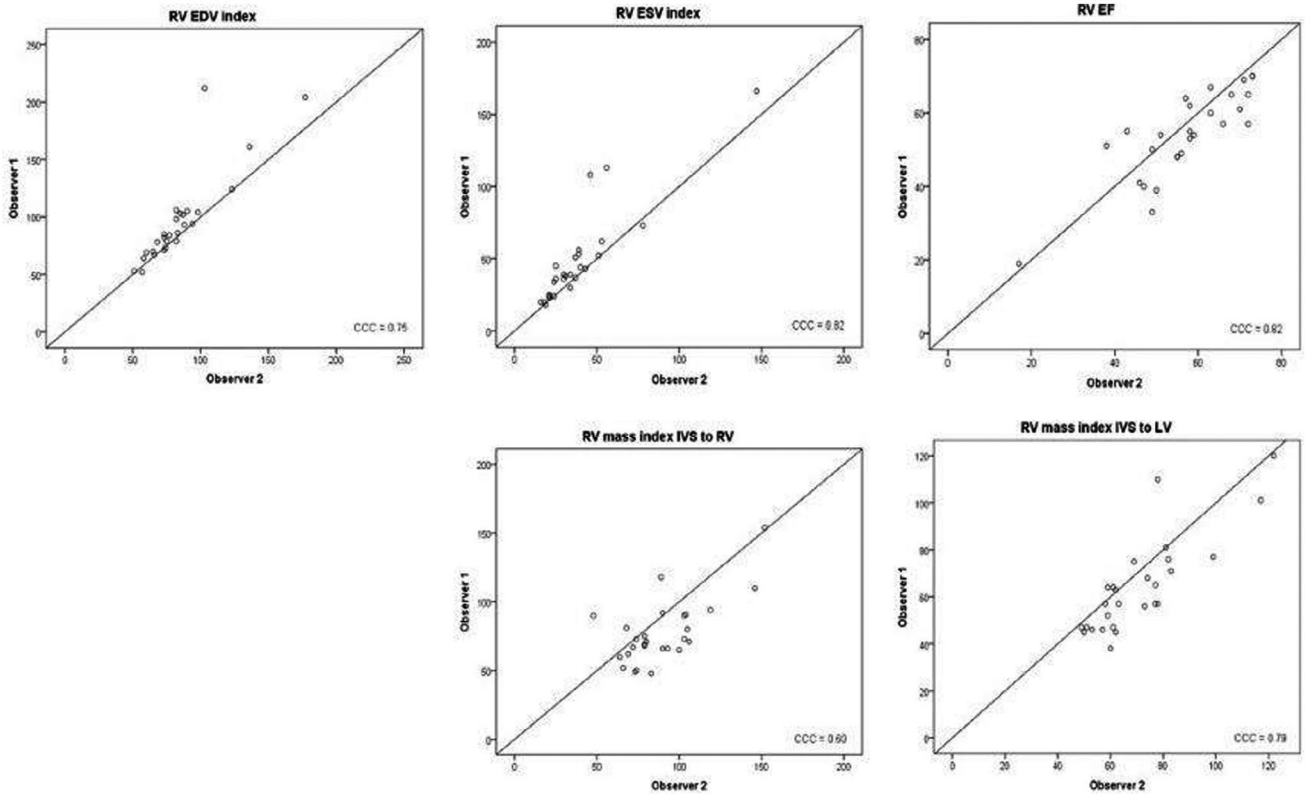


**FIGURE 2** dTGA s/p atrial switch: Agreement (using concordance correlation coefficients) is shown between independent, blinded cardiologists for CMR determined RV quantitative data

**TABLE 4** RV myocardial mechanics via CMR speckle tracking

| | | *N* | Median (range) | *P* value |
|---|---|---|---|---|
| CMR RV GLS | CMR RV EF < 45% | 7 | −9.8 (−14.7 to −2.9) | .42 |
| | CMR RV EF ≥ 45% | 20 | −10.2 (−13.5 to −6.8) | |
| | Total | 27 | −10.0 (−14.7 to −2.9) | |
| CMR RV GCS | CMR RV EF < 45% | 7 | −9.3 (−13.1 to −3.5) | .031 |
| | CMR RV EF ≥ 45% | 20 | −13.9 (−19.9 to −4.4) | |
| | TOTAL | 27 | −11.5 (−19.9 to −3.5) | |

*Note:* For continuous variables, medians with ranges are displayed.

for correlations with clinical data (exercise endurance, $VO_2$ max, or NYHA class), and no significant correlations were seen.

## 3.2 | Correlations between myocardial mechanics and CMR volumetric/functional data

### 3.2.1 | CMR-derived myocardial mechanics

Moderate negative correlation was seen for CMR RV global longitudinal strain (GLS) and RV EF ($r$ = −0.50, $P$ = .0084), but it seems to be driven by a *single outlier*. With this patient removed, there was no correlation. Cardiac magnetic resonance imaging RV global circumferential strain (GCS) and RV EF had moderate negative correlation despite the outlier ($r$ = −0.65, $P$ < .001). Cardiac magnetic resonance imaging GCS was significantly different for those with RV EF above and below 45%, while GLS was not able to discriminate between RV EF above and below 45% (Table 4, Figure 3). While RV GLS data were driven by a single outlier, RV GCS data showed modest correlation with RV EDV index and RV ESV index ($r$ = 0.56, $P$ = .0025 and $r$ = 0.55, $P$ = .0028, respectively). Finally, whether the interventricular septum was attributed to the RV mass calculation or to the LV mass, RV GCS showed some correlation with RV mass index ($r$ = 0.52, $P$ = .0054).

### 3.2.2 | Echo-derived myocardial mechanics compared with CMR volumetric data

As Echo-derived GLS becomes more negative, the CMR-derived RV EF improves ($r$ = −0.57, $P$ = .002). As Echo-derived RV GCS becomes

**TABLE 5** RV myocardial mechanics via echo VVI

| | | *N* | Median (range) | *P* value |
|---|---|---|---|---|
| Echo RV GLS | CMR RV EF < 45% | 7 | −10.5 (−15.5 to −7.5) | 0.21 |
| | CMR RV EF ≥ 45% | 20 | −13.5 (-23.4 to −6.8) | |
| | Total | 27 | −12.8 (−23.4 to −6.8) | |
| Echo RV GCS | CMR RV EF < 45% | 5 | −9.0 (−13.1 to −4.3) | 0.38 |
| | CMR RV EF ≥ 45% | 15 | −8.8 (−17.2 to −5.2) | |
| | TOTAL | 20 | −8.9 (−17.2 to −4.3) | |

*Note:* Seven had Echo data suboptimal for assessment of GCS due to poor visualization of the anterior RV wall. For continuous variables, medians with ranges are displayed.

**TABLE 6** Significant differences for echo measures of RV Diastolic Function When CMR Denoted RV EF < 45% vs. RV EF > 45%

| | All subjects | | CMR RV EF < 45% | | CMR RV EF ≥ 45% | | *P* value |
|---|---|---|---|---|---|---|---|
| | n | (median, range) | n | (median, range) | n | (median, range) | |
| TV E | 21 | 0.86 (0.61 - 1.25) | 5 | 1.02 (0.67 – 1.25) | 16 | 0.84 (0.61 – 1.16) | 0.083 |
| TV S' | 21 | 0.08 (0.03 – 0.14) | 6 | 0.06 (0.05 – 0.08) | 15 | 0.09 (0.03 – 0.14) | 0.026 |
| TV A' | 19 | 0.06 (0.04 – 0.10 | 5 | 0.05 (0.04 – 0.06) | 14 | 0.07 (0.04 – 0.1.) | 0.018 |
| TV E/E' | 18 | 10.47 (4.05 – 22.75) | 5 | 13.00 (10.56 – 22.23) | 13 | 8.10 (4.05 – 22.75) | 0.049 |

*Note:* For continuous variables, medians with ranges are displayed.

**TABLE 7** Echo variables compared with patient clinical status

| |
|---|
| Echo-derived GLS |
| Echo time to peak GLS |
| Echo-derived GCS |
| Echo time to peak GCS |
| Tricuspid valve (TV) E wave |
| TV A wave |
| TV E/A ratio |
| Tissue Doppler TV S' wave |
| Tissue Doppler TV A' wave |
| TV E/E' ratio |
| TV E'/A' ratio |
| Time from isovolumic contraction to end of isovolumic relaxation |
| RV ejection time |
| Myocardial performance index (MPI) |

*Note:* Clinical status was assessed by exercise endurance, $VO_2$ max, and NYHA class.

more negative, the CMR RV EF improves ($r$ = −0.66, $P$ = .0016), as expected (Figure 4).

### 3.2.3 | Correlations between CMR and Echo strain data

VVI assessment of RV GCS by Echo had a modest correlation with that determined by CMR ($r$ = 0.53, $P$ = .017). RV GLS as measured by Echo vs. CMR did not correlate well ($r$ = 0.23, $P$ = .25) (Figure 5).

## 4 | DISCUSSION

The dTGA s/p atrial switch population may suffer from complications including atrial dysrhythmias (arising from scarring around Mustard/ Senning baffle suture lines and compounded by baffle obstruction and reconstruction) and myocardial failure of the systemic morphologic RV.[24] The present cross-sectional study of a relatively healthy cohort of young adults with dTGA s/p atrial switch (majority NYHA class 1), who were able to complete a CMR scan and treadmill test, had few of these problems. Our cohort is similar to other published series, where NYHA Class 1 and 2 predominated (96% of 137 subjects reviewed by Dos et al), in having relatively little need for reintervention within the first few decades post atrial switch.[25] Literature review shows patients with dTGA s/p atrial switch, classified as NYHA class 3 or greater, though, have an increased risk for functional deterioration leading to fatal arrhythmias and heart failure.[26] Individuals with NYHA three or four symptoms were not in the present cohort.

Most of the present cohort had *few to no symptoms*. These patients, therefore, likely exist earlier on the Frank-Starling curve and are able to maintain cardiac output despite having a systemic RV. The patients, though, have compromised myocardium, as reflected by their reduced myocardial strain values. Although reduced myocardial strain was present, it was not sufficiently severe to lead to significant alterations in *global* ventricular EF (with few having RV EF < 45%); thus, it is not surprising that Echo data and most CMR data did not correlate with clinical status in this healthy, asymptomatic cohort. Only a weak correlation was seen between systemic RV CMR-derived mass index (when IVS was given to the LV) and exercise

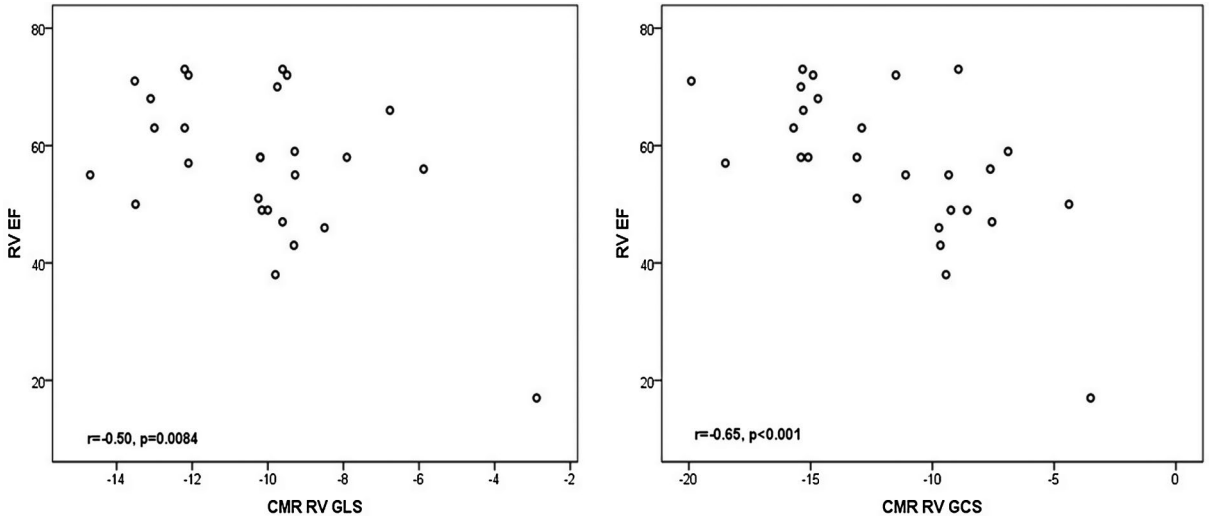


**FIGURE 3** CMR-derived RV Strain and CMR RV EF

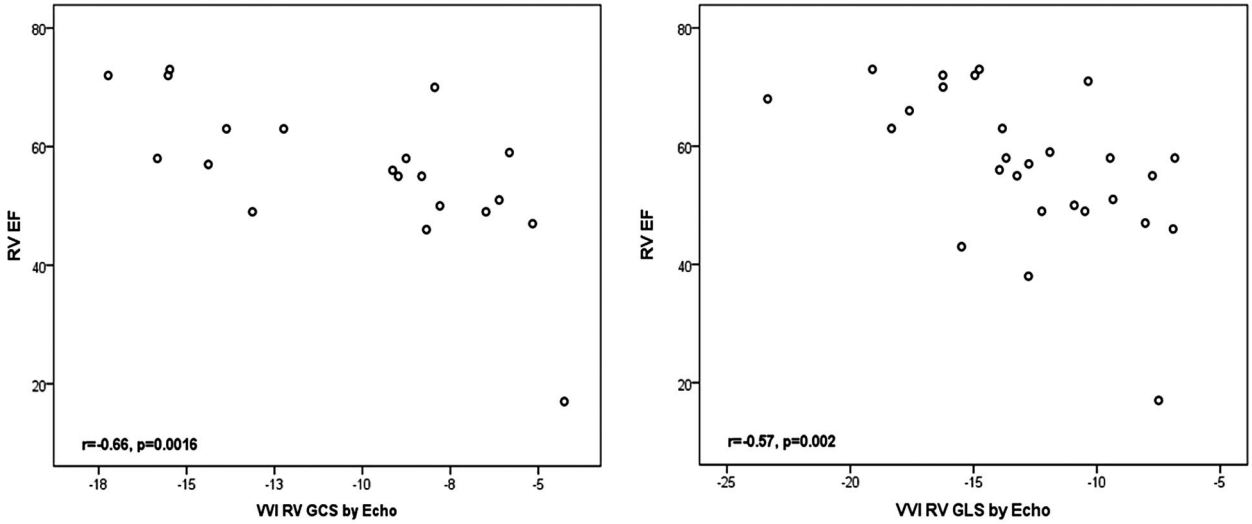


**FIGURE 4** Echo-derived RV Strain and CMR RV EF

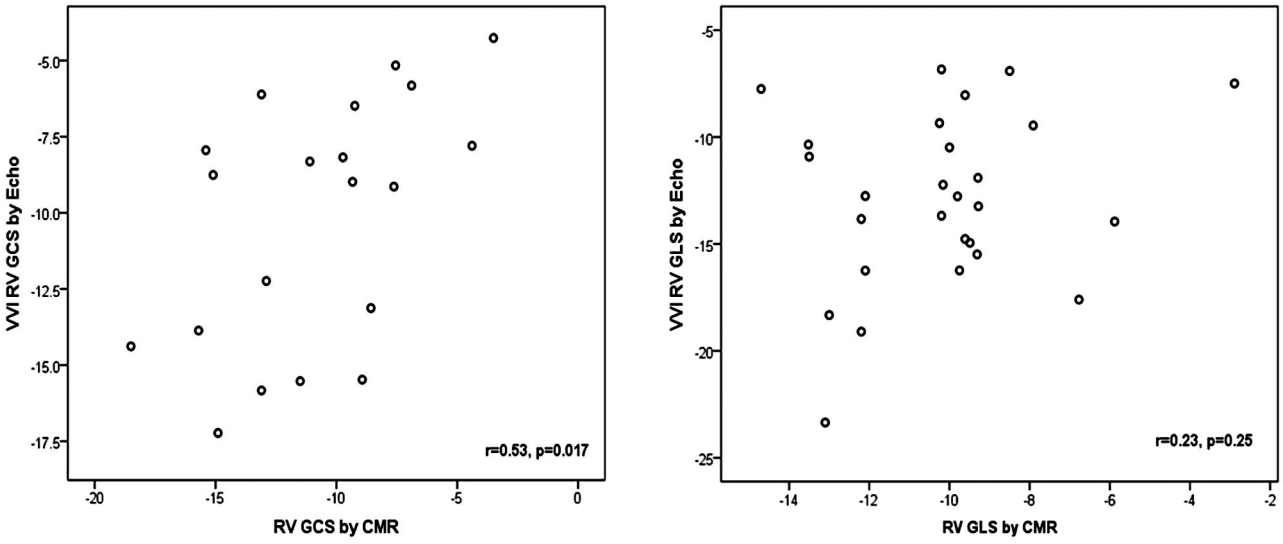


**FIGURE 5** Correlation between CMR- and Echo-derived RV Myocardial Strain

endurance ($r$ = 0.39, $P$ = .044). A similar correlation was expected for exercise endurance and RV mass index when IVS was given to the systemic RV, but this was not seen. While ventricular hypertrophy is induced by *extensive* strength training and/or endurance training,[27] review of clinical data suggests that the patients' lifestyles did not factor into their observed myocardial hypertrophy. Right ventricular hypertrophy, seen in this cohort, was related to the RV's chronic exposure to the increased afterload of systemic vascular resistance. Right ventricular hypertrophy, therefore, is a physiologic adaptational process that allows for building up systemic pressure, as described by Grothoff et al.[28]

With blinded review of CMR data and great care to trace the muscular trabeculations (giving them to the mass calculation and excluding them from ventricular volume), reasonable interobserver agreement was seen for calculated CMR short-axis cine data for the systemic RV (CCC of 0.75, 0.82, and 0.82 for RV EDV index, RV ESV index, and RV EF), similar to that described by Jimenez-Juan et al, using smooth contours (ignoring trabeculation) with short-axis imaging of adults with dTGA s/p atrial switch population.[29] Winter et al, using smooth ventricular contours, like Jimenez-Juan et al., obtained larger RV EDV and ESV with lower RV EF.[30] Manual tracing of the trabeculations, with attribution to the mass, had lower reproducibility for Winter et al., than was seen in our study.[30] Consistency in data analyses is, therefore, important for any CMR Lab, especially with regard to the process for handling trabeculations during planimetry. Consistent processes for data analyses may permit more valuable serial data interpretation, as repeated CMR scans are performed for aging patients with dTGA s/p atrial switch. Our study shows that CMR provides complete evaluation of the hypertrophied systemic RV, which can be fully visualized by this imaging modality. By extrapolation, periodic CMR scanning (to visualize the whole heart) may aid in the evaluation of other patients who live with a systemic RV (eg, patients with Fontan palliation for hypoplastic left heart syndrome and patients with pulmonary hypertension (PH)).

In the present cohort, most had good global RV systolic function (RV EF > 45%), despite the presence of reduced CMR-derived myocardial strain values. RV strain for our cohort was compromised when compared with published CMR strain values from healthy volunteers (age 48 ± 13 years), where normal RV GLS was −26.0% (−23.8% to −28.2%), LV GLS −22.3% (−20.5% to −24.0%), and LV GCS -25.0 (−24.0% to −26.1%).[31] Cardiac magnetic resonance imaging-derived RV strain data for our cohort, were similar to CMR strain values (RV GLS −9.9% ± 0.5% and RV GCS −11.2% ± 0.7%) from a young adult cohort of 20 dTGA s/p atrial patients (28.7 ± 1.8 years with CMR-derived RV EF = 45% ± 2.5%), described by Thattaliyath et al.[32]

In our study, CMR-derived GCS had modest correlation with increasing RV size. Thus, we speculate that with chronic pressure load on the systemic RV, progressive deterioration in CMR-derived RV strain precedes changes in ventricular volumes and EF, which may worsen clinical status over time. Thus, the present study gives an intriguing "signal" from CMR-derived myocardial strain that warrants further study in a larger, *longitudinal*, multi-center trial of a more heterogenous population of patients with dTGA s/p atrial switch—including sicker patients (NYHA Class 3 and 4).

By employing CMR (for not only traditional ventricular volumetric and global systolic assessment, but also for an assessment of myocardial mechanics), our research showed that RV GCS could discriminate the failing RV with EF < 45%, but GLS could not. Lipczynska et al, who studied a population of 40 patients with dTGA s/p atrial switch (mean age, 25.6 ± 5 years; 25 men) similar in age to the present cohort, though, demonstrated a linear correlation between RV GLS and CMR systemic RV EF values. As the longitudinal strain values became more negative, myocardial strain was better and CMR EF, not surprisingly, improved ($r$ = 0.4; $P$ = .01). Recently, Lipczynska proposed that RV GLS is able to discriminate between RV EF above and below 45%.[19] We did not see this in our small healthy cohort. While normal RV function seems to be highly dependent on longitudinal shortening,[33] it has been suggested that *transverse* wall motion of the RV (measured by GCS) may be a better marker of systolic function when the RV faces high resistance, as exists for example in the population of patients with severe PH. As PH advances and the RV becomes more hypertrophied, *circumferential* deformation becomes relatively more important, resembling LV contractility patterns.[34] Likewise, this RV circumferential deformation seems important in our dTGA s/p atrial switch cohort, where RV hypertrophy exists as the RV pumps against the chronic pressure load of systemic vascular resistance.

Turning attention now to Echo assessment of the systemic RV, it is clear from the missing data (Table 6) that challenges existed for Echo due to poor visualization of the RV. Unlike CMR strain, Echo measures of myocardial mechanics (strain) could not distinguish between the failing RV with EF less than 45% and the thriving RV with EF above 45%. Although in the present study, no correlations existed between Echo strain data (GLS or GCS) and clinical status, the Echo strain data (Table 5) (like the CMR RV strain data) are lower than published normal Echo-derived RV GLS data from healthy adult controls with no structural heart disease (RV GLS = −24.7 ± 2.6 for men and −26.7 ± 3.1 for females).[35] These Echo strain data are consistent with the presence of myocardial disease for the chronically pressure overloaded systemic RV of dTGA s/p atrial switch patients. Our study's Echo-derived RV GLS data are also reduced when compared with normative data for the *systemic LV*, where Echo-derived LV GLS has been reported as: −21.3 ± 1.9 for ages 30-39 years and 20.7 ± 2.2 for 40-49 year olds.[36] This implies that the RV may be less well adapted than the LV to perform as the systemic pump.

Our Echo-derived RV GLS data are similar to published values from a prospective study of 49 (majority NYHA class 1) adult patients (32 ± 4 years old) with dTGA s/p atrial switch, where cycle ergometry was employed to classify the patients as "healthy or compromised," using a median peak oxygen uptake ($VO_2$) cutoff of >64.5.5% as "healthy" ($n$ = 23) and $VO_2$ ≤ 64.5% ($n$ = 24) as "compromised." For the compromised group, Ladouceur et al noted that Echo-derived GLS was significantly *reduced* compared to the healthy dTGA s/p atrial switch group (−10.9% ± 2.9% vs −13.1% ± 2.3%, $P$ < .05).[37] This was primarily due to significant decrease of IVS longitudinal strain. As in our study, Ladoucer et al found that no other echocardiographic systolic parameters were significantly different between these groups.[37]

### 4.1 | Limitations

There were some limitations to this retrospective, single center study of a small cohort of patients with dTGA s/p atrial switch. All data were collected from a single time point without longitudinal follow-up. Selection bias may have existed, as only dTGA s/p atrial switch patients who were able to participate in CMR scanning (ie, those without pacemaker) and Bruce protocol exercise treadmill testing were included; thus, the study included a disproportionately healthy group.

Despite full visualization of the RV by CMR, an era effect may have altered image quality, as advances in CMR pulse sequences allowed for improved image quality for scans performed during 2010-2015, compared with CMR scans performed in the earlier years (2005-2009) of the Children's Hospital of Wisconsin CMR Program. In addition, including patients scanned using 1.5T and 3T magnets, while expanding our patient population, likely added variability to the data quality due to differences in image resolution. Employing experienced CMR physicians/personnel for data analyses is key to achieving good interobserver agreement. The goal is to have uniformity in ventricular planimetry, especially regarding the Lab's handling of trabeculations (ie, with detailed contours vs. smooth contouring which ignores trabeculations). Even then, partial volume averaging may occur from variability in image resolution and slice thickness between patients.

This study chose to segment the ventricles by using short-axis imaging planes, as this is the standard CMR practice at the Herma Heart Institute; others have suggested that the axial plane through the hypertrophied, systemic RV may have less variability,[29] as has also been advocated by Fratz et al. for the dilated RV present in other congenital heart diseases, like tetralogy of Fallot.[38] Image resolution and slice thickness may lead to partial volume effects which cause suboptimal endocardial-blood pool border detection (particularly at the apical and basal portions of the ventricles) which further degrades accuracy of volumetric calculations. Thus, in the future, to mitigate this effect, imaging based on a further standardized protocol (with adherence to a uniform slice thickness and image resolution) may aid serial study of this population.

Finally, the sample size was small with poor quality Echo (ie, inability to see the anterior RV) limiting the VVI strain data collection. For circumferential strain, there were five subjects' data were excluded, as the images were suboptimal for analyses. A blinded Echo review did take place with two reviewers for Echo VVI strain data, but only a single reviewer evaluated Echo tissue Doppler data.

## 5 | CONCLUSIONS

This cross-sectional study shows the utility of CMR scanning for complete visualization of the systemic RV. Variability is inherent in planimetry of heavily trabeculated ventricles; thus, any given CMR Lab should establish and adhere to a consistent protocol by which data analyses occur—either with careful tracing of trabeculae or with smooth contouring, as both have reasonable interobserver agreement. While regional ventricular deformation (ie, myocardial strain) assessment is currently in vogue, its clinical relevance may require longitudinal study with an emphasis on consistency in acquisition and analyses. Cardiac magnetic resonance imaging strain measurements may be more complete than Echo strain measurements, as the latter may be encumbered by poor visualization of the RV. A prospective, multi-center, longitudinal imaging study of patients with dTGA s/p atrial switch may increase recruitment of a more diverse clinical cohort (NYHA 1-4), enhancing detection of early markers of clinically important progressive myocardial disease related to chronic pressure overload. While the population living with dTGA s/p atrial switch is small, longitudinal study of this population with a systemic RV is relevant today, because information can be applied to the care of other populations, who live with a systemic RV—especially, those patients with hypoplastic left heart syndrome status post Fontan and those patients with severe PH.

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## CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest with the contents of this article.

## AUTHOR CONTRIBUTION

*Study design, supervision of data collection, CMR analyses, interpretation of data analyses, draft article, approval of submission, and creation of revised manuscript*: MS

*Statistics, critical revision of article, and approval of article*: KY

*Data collection and approval of article*: CM

*CMR data analyses, critical revision of article, and approval of article*: BG

*Echocardiography data analyses, critical revision of article, and approval of article*: DS

*Initial data management, statistics and interpretation, manuscript review, and approval*: JL

*Initial data management, statistics and interpretation, manuscript review, and approval*: AK

*Study project coordination, assistance with IRB submission, and manuscript review for clarity with approval of submission*: MK

Cardiac magnetic resonance imaging *strain analyses and critical revision of article, and approval of article*: KH

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