

Multimodality imaging and functional assessment in patients with systemic right ventricle and biventricular physiology: a retrospective single-center study

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Abstract

Systemic right ventricle (sRV) dysfunction is frequent in patients with congenitally corrected transposition of great arteries (cc-TGA) and those with dextro-transposition of great arteries (D-TGA) after Mustard/Senning operations. This condition should be identified promptly. We aimed to compare echocardiographic parameters with cardiac magnetic resonance (CMR)-derived parameters in patients with sRV and to evaluate their correlation with clinical variables and exercise capacity. Patients with cc-TGA and D-TGA after Mustard/Senning who underwent standard and advanced (speckle tracking and 3D) echocardiography and CMR (including feature-speckle tracking) were included. Clinical and imaging parameters were collected. Echocardiographicderived right ventricle end-diastolic area and end-systolic area correlated with 3D echocardiographic-derived right ventricle enddiastolic and end-systolic volume (r=0.6, p=0.006 and r=0.8, p=0.002). 3D ejection fraction (EF) correlated with fractional area change and tricuspid annular plane systolic excursion (TAPSE) (r=0.8, p=0.001 and r=0.7, p=0.03). sRV global longitudinal strain (GLS) correlated with systemic atrial strain (sAS) (r=-0.6, p=0.01). CMR-derived EF correlated with CMR-derived GLS both endocardial and myocardial (r=-0.7, p=0.007 and r=-0.6, p=0.005). sRV areas as assessed by echo correlated with CMRderived volumes (r=0.9, p=0.0001 for diastole and r=0.8, p=0.0001 for systole). Similarly, a correlation was found between sRV echo-derived GLS and CMR-derived GLS, both endocardial and myocardial (r=0.8, p=0.001 and r=0.7, p=0.01). The only imaging parameter that correlated with peak oxygen consumption was sAS (r=0.55, p=0.04). When comparing cc-TGA and D-TGA, the former showed better GLS-derived values as assessed by CMR (CMR-derived right ventricle endocardial longitudinal strain -23.2% versus -17.2%, p=0.002; CMR-derived right ventricle myocardial longitudinal strain -21.2% versus -16.7%; p=0.05), bigger systemic atrial area (20.2 cm²/m² versus 8.4 cm²/m², p=0.005) and higher TAPSE values (16.2 mm versus 12.2 mm, p=0.04). Echocardiography is valid to screen for sRV dilatation and function and to guide the timing for CMR. The investigation of atrial deformation imaging may help to better understand diastolic function. Patients with cc-TGA show better cardiac function compared to patients after atrial switch. Further investigations are needed to identify imaging parameters linked to exercise capacity.



The systemic right ventricle (sRV) condition, where the morphological right ventricle supports systemic circulation, accounts for approximately 10% to 12% of all congenital heart diseases (CHDs) [1,2]. This unique anatomical and functional adaptation occurs in both two-ventricle and single-ventricle arrangements, presenting a spectrum of clinical phenotypes and prognoses. Biventricular physiologies involving sRV are typically seen in patients with congenitally corrected transposition of the great arteries (cc-TGA) and those with dextro-transposition of the great arteries (D-TGA) who have undergone Mustard or Senning operations. While many of these patients survive into adulthood, they often face progressive sRV dysfunction, heart failure, exercise intolerance, arrhythmias, and premature death in early adulthood [3-7]. sRV failure is a significant concern for CHD practitioners, necessitating longitudinal follow-up to identify markers of cardiac dysfunction and implement appropriate treatments. A recent consensus paper has provided comprehensive guidelines and recommendations for the effective use of integrative imaging technologies in CHDs, with special insights on sRV [8]. Echocardiography remains the first-line diagnostic technique, yet its application is challenging due to the lack of specific cut-off values and quantitative parameters tailored to sRV. Additionally, obtaining optimal acoustic windows in these patients can be difficult [9]. Cardiac magnetic resonance (CMR) offers a more reliable quantitative assessment of sRV function but is limited by longer execution times, higher costs, and limited availability [10,11]. Currently, multimodality imaging with integration of different methodologies allows to overcome challenges of sRV, thus improving management and long-term prognoses of affected patients [12].

This study aims to: i) compare a comprehensive range of standard and advanced echocardiographic parameters with CMRderived parameters in patients with sRV and biventricular physiology (D-TGA post-Mustard/Senning operations and cc-TGA); ii) evaluate their correlation with clinical variables and exercise capacity.

Materials and Methods

Study population

This is a retrospective single-center study, carried out at the Department for Women's and Children's Health of Padua University Hospital between March and September 2023.

Patients were included if they met the following criteria: i) sRV and biventricular circulation in D-TGA with previous Mustard or Senning surgery or cc-TGA; ii) patients who had an echocardiogram and a CMR scan within one year from the last follow-up visit (September 2022-September 2023).

Patients excluded from the study were those implanted with pacemakers, cardioverter defibrillators, or any CMR-incompatible device.

Patients' records were reviewed from the medical platform of Padua University Hospital. Clinical data extracted from the medical record included age, body mass index, body surface area (BSA), New York Heart Association (NYHA) functional class, time since surgery, and ongoing medications. Moreover, data derived from cardiopulmonary exercise tests (CPET) done within 12 months from any imaging exam were collected, when available. The following parameters were considered: peak oxygen consumption (VO₂ peak), minute ventilation/carbon dioxide production slope, metabolic equivalents, and



peak respiratory exchange ratio. All data was collected keeping confidentiality and was anonymized for statistical analysis.

Cardiac resting imaging

All the echo studies were performed using the Vivid E95 ultrasound system (General Electric Healthcare, Horten, Norway). Standard and advanced transthoracic echocardiographic exams were performed by one experienced pediatric cardiologist (MA).

Chamber quantification measurements were assessed according to the current European Association of Echocardiography guidelines [13]. From the apical window, we measured the RV end-diastolic and end-systolic area (eRV-EDA, eRV-ESA), and ESA A. These values were then normalized for BSA.

Global sRV function was visually assessed from the apical four chamber view. Fractional area change (FAC) was calculated as the percentage of change between the eRV-EDA and eRV-ESA. Tricuspid annular plane systolic excursion (TAPSE) was determined in M-MODE. Tricuspid inflow PW Doppler velocities, lateral wall tissue Doppler imaging velocities, and the derived E/E' ratio were also assessed.

Speckle tracking global longitudinal strain (GLS) of sRV, including RV free wall and septum, was calculated in the apical four chamber view on the ultrasound device itself or on an offline workstation (Echo PAC version 112.99, Research Release, GE Healthcare), which allows semi-automated analysis.

The images selected for the speckle tracking analysis had a frame rate between 50 and 100 frames per second, with a $\leq 10\%$ variability in heart rate, as already detailed in previous articles on speckle tracking echocardiography and CHDs [14,15]. After having drawn manually 3 points (2 annular and 1 apical), the software tracked the myocardium semi-automatically throughout the heart cycle. Manual adjustments, if needed, were performed to optimize the region of interest. The automated algorithm allowed GLS to be calculated, dividing the right ventricles into 6 segments. GLS by speckle tracking was defined as the average peak negative value on the strain curve during the systole. Segments not well visualized were not included; in the case of ≥ 2 non-visualizable segments the patient was not included in the analysis.

From the apical four chamber view, the systemic atrium peak systolic strain (sAS) was also measured. The atrial wall was traced manually (3 points) and adjusted, if necessary, resulting in strain curves from a total of 3 atrial segments. Global peak atrial strain was defined as the average of the maximum positive values during RV systole of the 3 analyzed atrial segments.

When available, 3D images were elaborated to obtain 3D volumes (eRV-EDV, eRV-ESV) and ejection fraction (EF) (eRV-EF).

Tricuspid regurgitation was evaluated as none/trivial (0 = sin-gle narrow jet), mild (1 = multiple narrow jets), moderate (2 =wide jet reaching the mid part of the atrium), and severe (3 = wide jet reaching the roof of the atrium). The color scale was set at a Nyquist limit of 50-60 cm/s. Analysis of echocardiographic data was blind to CMR results, clinical data, and CPET findings.

Each CMR examination was carried out by one pediatric cardiologist experienced in pediatric CMR imaging (ER) and one pediatric radiologist (AC) with the same CMR scanner (Achieva 1.5 Tesla, Philips Healthcare; Best, the Netherlands). CMR measurements were performed by the same operator (ER), to empower data consistency and assessment was blind to Echo data.

A standard CMR protocol for evaluating patients with sRV includes real-time localization imaging in three planes without electrocardiogram (ECG)-gating and during free breathing; cine steady-



state free precession (SSFP) sequence with gated-breathing to report anatomy, size of ventricles and function; phase contrast sequences Qp:Qs; whole-heart isotropic 3D SSFP imaging for vascular evaluation without contrast material administration and visualization of proximal and mid-coronary arteries; late gadolinium enhancement (LGE) imaging along the long and the short axes [16].

In the following study, we evaluated SSFP end-inspiratory breath-hold ECG-gated cine-images in long-axis views and a stack of short-axis slices covering the ventricular cavities. Moreover, right ventricular outflow tract (RVOT) obstruction was assessed by a specific RVOT view, together with an analysis of the superior systemic venous pathways and the possible presence of baffle leaks.

All patients underwent Gadolinium 0.2 mmol/kg administration to assess the presence of LGE in the four-chamber and short-axis view.

Image analysis was performed by using Philips Intellispace Cardiovascular post-processing software. The sRV epi- and endocardium were manually segmented in cine short axis on end-diastolic and end- systolic images. The following data were considered: sRV volumes both absolute and indexed to BSA [CMRderived right ventricle end-diastolic volume (cRV-EDV), CMRderived right ventricle end-systolic volume (cRV-ESV)] and EF [CMR-derived right ventricle EF (cRV-EF)] [17,18].

Myocardial strain was assessed using feature tracking (FT) applied to SSFP cine images during post-processing with a dedicated software (Qstrain, Medis Suite Version 4.0.38.4, Leiden, Netherlands). GLS of the sRV [CMR-derived right ventricle endocardial longitudinal strain (cRV-GLSendo), CMR-derived right ventricle myocardial longitudinal strain (cRV-GLSmyo)] was evaluated on long-axis images [17,18]. Initially, the endocardial borders of the sRV were manually segmented in the end-diastolic

 Table 1. Demographic and clinical features of the study cohort.

 Values are expressed as mean + standard deviation, median (interquartile range), or n (%).

Variables	Study cohort (n=19)
Age at CMR	28 (IQR 17.25-33)
cc-TGA age (years)	21 (IQR 16.25-28.5)
D-TGA age (years)	32 (IQR 28.25-36.75)
Female (%)	11 (58)
cc-TGA, (%)	11 (57.9)
D-TGA (%)	8 (42.1)
s/p Mustard	3 (37)
s/p Senning	5 (63)
NYHA class \geq II, n (%)	7 (8%)
Ongoing treatment, n (%)	
β-blocker	4 (21)
ACE-I	4 (21)
Diuretics	2 (10.5)
Anticoagulants	2 (10.5)
Antiaggregant	1 (5.2)
Sacubitril/valsartan	2 (10.5)
Peak VO ₂ (mL/kg/min)	24.3 (SD 6.6)
VE/VCO ₂	32.6 (SD 3.9)
Peak RER	1.16 (SD 0.1)
METS	12.8 (SD 2.4)

SD, standard deviation; CMR, cardiac magnetic resonance; cc-TGA, congenitally corrected transposition of the great arteries; D-TGA, dextro transposition of great arteries; NYHA, New York Heart Association; ACE-I, angiotensin-converting enzyme inhibitors; peak VO₂, peak oxygen consumption; VE, minute ventilation; VCO₂, carbon dioxide production; peak RER, peak respiratory exchange ratio; METs, metabolic equivalent of task.

phase and then automatically expanded to all phases with an automatic border detection algorithm. The endocardial borders were checked for accuracy in all cardiac phases and manually adjusted if necessary. The software provided endocardial and myocardial peak systolic strain [19,20]. Finally, we compared Echo and CMR data with values measured by CPET.

Statistical analysis

Baseline characteristics are reported as percentages for categorical variables, as means and standard deviations for continuous variables when normality was verified, and as median and interquartile range (IQR) when normality was not verified by the Kolmogorov-Smirnov test. The Student's *t*-test for independent samples or the Mann-Whitney test when normality was not verified was used for the analysis of the variables. Differences between groups for continuous variables were analyzed based on distribution using the *t*-test, Kruskal-Wallis test, or Wilcoxon-Mann-Whitney test, as appropriate. The relationship between CMRderived data, echocardiographic parameters, and exercise test results was evaluated using the Pearson and Spearman correlation coefficient as appropriate. The statistical significance was set at p<0.05. Analysis was performed using SPSS statistic software version 22 (IBM SPSS Statistics Version 22, Chicago IL, USA).

Results

Patients

A total of 22 patients with sRV and biventricular physiology underwent echocardiography and CMR between September 2022 and September 2023. Both echocardiographic and CMR data were available for 19 patients. Median age at CMR was 28 years (IQR 17.25-33); 11 (58%) were female. 11 patients had cc-TGA, 4 of them had a previous physiologic repair, consisting of large ventricle septum defect closure (3 patients) and atrial septal defect closure (1 patient). Of the 8 patients with D-TGA, 5 had undergone Senning repair (63%). cc-TGA patients were significantly younger than patients with D-TGA (p 0.03; cc-TGA median age 21 years IQR 16.25-28.5; D-TGA median age 32 years IQR 28.25-36.75). Most patients were clinically asymptomatic. Demographic and clinical features of patients are summarized in Table 1.

Cardiac imaging

Echocardiographic and CMR data were available in 19 patients. 2D speckle tracking echocardiography was available in 15 patients, 3D echocardiography in 11 patients, and CMR FT was assessed in 18 performed scans. TR was assessed as mild in 4 patients (21%), moderate in 13 (68.4%) and severe in 2 (10.5%) patients. Echocardiographic and CMR variables are summarized in Table 2.

Echocardiographic results

A correlation between eRV-EDA and eRV-EDV and between eRV-ESA and eRV-ESV, both as absolute (rho=0.6, p=0.006; rho=0.8, p=0.002) and indexed values (rho=0.8 p=0.007; rho=0.9 p 0.0001) was found. In addition, a correlation between eRV-EF and FAC and TAPSE was identified (r=0.8, p=0.001 and r=0.7, p=0.03). Also, E/E' correlated inversely with sAS (r=-0.8, p=0.03). Lastly, the correlation between sAS and RV-GLS was also statistically significant (r=-0.6, p=0.01). No correlation was found between other echocardiographic data and NYHA class.



Cardiac magnetic resonance results

A correlation was found between cRV-EF and CMR-GLS, both endocardial and myocardial (r=-0.7, p=0.007, r=-0.6, p=0.005). No correlation was found between CMR-derived data and myocardial fibrosis or NYHA class.

Echo and cardiac magnetic resonance results

A statistically significant correlation was found between cRV-EDV and cRV-ESV and eRV-EDA and ESA (r=0.9, p=0.0001; r=0.8, p=0.0001 respectively). Also, cRV-GLSendo and cRV-GLSmyo correlated significantly with RV-GLS assessed by echocardiography (r=0.8, p=0.001; r=0.7, p=0.01 respectively) and cRV-GLSendo correlated with sAS (r=-0.6, p=0.04). These results are shown in Figure 1. No correlation was found between CMR-derived data and other echocardiographic parameters.

Table 2. Cardiac magnetic resonance and echo variables analyzed in the study. Values are mean + standard deviation or n (%).

Echocardiographic variables	Study cohort (n=19)
eRV-EDA (cm ²)	31.06 (SD 9.14)
eRV-EDAi (cm ² /m ²)	22.48 (SD 12.38)
eRV-ESA (cm ²)	20.13 (SD 8.61)
eRV-ESAi (cm ² /m ²)	14.24 (SD 8.34)
FAC (%)	37.41 (SD 7.62)
E/A	2 (SD 1.03)
TAPSE (mm)	13.93 (SD 5.24)
S'velocity (cm/s)	9.55 (SD 2.66)
E/E'	12.04 (SD 6.09)
E' velocity (m/s)	0.96 (SD 0.28)
eRV-EDV (mL)	127.18 (SD 40.67)
eRV-EDVi (mL/m ²)	81.32 (SD 32.51)
eRV-ESV (mL)	67.26 (SD 29.55)
eRV-ESVi (mL/m ²)	42.52 (SD 20.15)
eRV-EF (%)	47.44 (SD 8.88)
eRV-GLS (%)	-15.35 (SD 4.02)
$ESA_A (cm^2)$	21.50 (SD 12.13)
ESA_Ai (cm ² /m ²)	15.35 (SD 9.87)
sAS (%)	18.17 (SD 7.87)
Moderate-severe TR, n (%)	15 (78)
CMR variables	Study cohort (n=19)
cRV-EDV (mL)	170.25 (SD 73.56)
cRV-EDVi (mL/m ²)	104.61 (SD 22.79)
cRV-ESV (mL)	87.22 (SD 39.94)
cRV-ESVi (mL/m ²)	67.72 (SD 61.15)
cRV-EF (%)	50.11 (SD 8.44)
FAC (%)	34.16 (SD 10.45)
cRV-GLSendo	-20.62 (SD 5.35)
cRV-GLSmyo	-19.22 (SD 4.99)

SD, standard deviation; eRV, echo-derived right ventricle; cRV, CMR-derived right ventricle; EDA, end-diastolic area; ESA, end-systolic area; EDAi, indexed end-diastolic area; ESAi, indexed end-systolic area; FAC, fractional area change; TAPSE, tricuspid annular plane systolic excursion; EDV, end-diastolic volume; ESV, end-systolic volume; EDVi, indexed end-diastolic volume; ESVi, indexed end-systolic volume; ESVi, indexed end-systolic volume; ESV, end-diastolic volume; ESV, indexed end-systolic volume; ESV,

Cardiopulmonary exercise testing and cardiac imaging

CPET was available for 17 patients. No correlation was found between CMR imaging parameters and CPET functional data. This result did not change even when evaluating cc-TGA and TGA Mustard/Senning patients separately. The only echocardiographic parameter that correlated with peak VO₂ was sAS (r=0.55, p=0.04).

Corrected transposition of great arteries and transposition of great arteries-Mustard/Senning

When cc-TGA and D-TGA patients were compared, the former showed better GLS values as assessed by CMR (cRV-GLSendo -23.2% *versus* -17.2%, p=0.002; cRV-GLSmyo -21.2% *versus* -16.7%; p=0.05); and bigger indexed systemic atrial area (20.2 cm²/m² *versus* 8.4 cm²/m², p=0.005). TAPSE values were higher in cc-TGA group (16.2 mm *versus* 12.2 mm, p 0.04). Also, a trend was noticed towards worse values of sAS in D-TGA (13.8% *versus* 21.0%, SD 7.6 and 6.9 respectively). These results are shown in Figure 2. No differences were noticed in terms of tricuspid regurgitation and peak VO₂ between the two populations (D-TGA 22.2 mL/kg/min; ccTGA 25.8 mL/kg/min).

Discussion

To the best of our knowledge, only a few studies compared standard and advanced echocardiographic parameters with CMR parameters and CPET in patients with sRV and biventricular physiologies, sometimes even yielding conflicting results [21].

This study contributes to the limited body of research comparing standard and advanced echocardiographic parameters with CMR parameters and CPET in patients with sRV and biventricular physiologies. Our findings confirm that echocardiography is a well-established clinical tool for the clinical follow-up of these patients, to evaluate sRV dilatation and to assess systolic function. Also, further investigations of diastolic function in these patients, particularly by assessing atrial strain, may help in better understating the role of diastole in exercise capacity. Lastly, our findings suggest that some differences in terms of cardiac function exist in patients with sRV and different cardiac anatomies.

In this study, a significant correlation was found between 2D echo-derived areas and CMR-derived volumes, confirming that 2D echocardiography is a valid first tool to screen right ventricular dilatation, to assess patients longitudinally, and to guide the timing for advanced imaging such as CMR. Indeed, CMR remains a solid tool to assess sRV, but high costs, lower availability, and the need for dedicated skilled staff may limit its use [11].

3D echocardiography is an advanced imaging technique that could provide a wealth of information for the evaluation of sRV. Nevertheless, the semi-automated software for 3D analysis needs good-quality images, sometimes difficult to achieve when evaluating sRV in the TGA population [22]. Some studies reported a correlation between echo volumes and CMR volumes [23,24]. This was not identified in our study, but the small sample size may have limited the analysis. Indeed, considering that sRV areas by echocardiography correlated significantly with 3D volumes, we can speculate that with a bigger sample size, we might have reached the same result also in comparison with CMR data.





Figure 1. Scatterplot showing the correlation between CMR-and Echo derived parameters. eRV-ESA, echo-derived right ventricle endsystolic area; cRV-ESV, CMR-derived right ventricle end-systolic volume; eRV-EDA, echo-derived right ventricle end-diastolic area; cRV-EDV, CMR-derived right ventricle end-diastolic volume; RV-GLS, right ventricle global longitudinal strain; cRV-GLSendo, CMR-derived right ventricle endocardial longitudinal strain; cRV-GLSmyo, CMR-derived right ventricle myocardial longitudinal strain, sAS: systemic atrium peak systolic strain.





Figure 2. Distribution of GLS, ESA Ai and TAPSE in the two TGA subgroups. cRV-GLSmyo, CMR-derived right ventricle myocardial longitudinal strain; cRV-GLSendo, CMR-derived right ventricle endocardial longitudinal strain; ESA_Ai, indexed echo-derived systemic atrium end-systolic area; TAPSE, tricuspid annular plane systolic excursion.

Previous studies demonstrated a good correlation between strain measurements obtained by echocardiography and CMR, and data from our study supports this finding [10,25]. Also, in this study, a significant correlation was found between sAS and RV-GLS assessed by echo. Impaired atrial function, as it can be in patients after Mustard/Senning operations, can lead to inadequate ventricular filling [26,27]. This may affect firstly ventricular diastolic function and, later, systolic performance. This can potentially lead to decreased cardiac output and, ultimately, heart failure. Thus, monitoring both atrial and ventricular strain parameters longitudinally could provide valuable insights to early identify patients at risk for adverse outcomes and guide treatment strategies [28]. In addition, the systematic assessment of atrial strain may help to better understand diastolic function in these patients, which is still poorly understood [29-31].

The correlation between sAS and peak VO₂ may suggest a major role of diastolic function in determining exercise capacity in patients with sRV. The relationship between atrial deformation imaging and peak VO₂ has already been investigated in other CHDs [32,33]. By contrast, previous echocardiographic and CMR studies investigated the correlation between functional imaging parameters and CPET in patients with sRV, with no significant results [23,26]. However, atrial deformation imaging was not performed in these studies. Based on the results of the current study, we speculate that the presence of pulmonary vein baffle and tricuspid regurgitation can alter atrial compliance and elasticity, especially under exercise, resulting in an insufficient atrial output into sRV. Lastly, compared to subjects with D-TGA, patients with cc-

TGA showed better systolic function, as assessed by CMR-derived GLS values, higher values of TAPSE, systemic atrial area, and a trend toward better values of sAS. These results might be partially explained by the degree of TR, which leads to atrial enlargement and enhances TAPSE values [34,35]. However, in our study we did not find a difference when comparing the degree of TR among the two subgroups, thus other mechanisms such as altered ventriculo-atrial coupling and issues about atrial contraction may be involved. Ultimately, the absence of atrial surgery could explain the trend toward higher sAS values in the cc-TGA group.

Limitations

We acknowledge certain limitations of our study. This is a retrospective, single-institution cohort study; thus, advanced imaging was not available for all patients. Furthermore, some correlation coefficients are based on a small sample size, as measurements were not feasible/available in a considerable number of cases. However, this reflects practical applicability in everyday clinical practice, as echocardiographic assessment of sRV function is challenging and patients with sRV represent a rare population.

Conclusions

The study reaffirms the value of echocardiography as a primary tool for assessing sRV dimensions and function and guiding the timing for CMR use. The better systolic performance in cc-



TGA patients compared to those with D-TGA reflects the distinct characteristics and natural history of these conditions. Lastly, the use of 3D echocardiography and the assessment of diastolic function in this population remain areas that need further development.

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