Characteristics of Cardiovascular Magnetic Resonance Imaging and Outcomes in Adults With Repaired Truncus Arteriosus

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The cardiovascular magnetic resonance imaging (CMR) features of adults with repaired truncus arteriosus (rTA) are largely undefined. We sought to explore CMR characteristics in rTA and to identify associations between imaging findings and cardiovascular outcomes. Adults with rTA and CMR were identified and anatomic subtypes (1-4) were assigned (Collett and Edwards classification). CMR characteristics, clinical data at last follow-up and adverse cardiovascular outcome were recorded. Twenty-seven adults (19% male) were studied (median age at cardiovascular magnetic resonance 26 years [interquartile range 18 to 40]) over 5.2-year duration [interquartile range 2.5 to 7.5]. With the exception of mildly increased RV mass (30 ± 12 g/m^2), cardiac chamber measurements were within the normal range. In CMR measurements, only pulmonary artery peak velocity differed in subtypes (highest in subtype 3, 318 ± 26 cm/s, p = 0.029). Number of cardiovascular interventions in adulthood was moderately correlated with left ventricular end-diastolic volume (r = 0.463, p = 0.015), left ventricular ejection fraction (r = 0.425, p = 0.027) and neoaortic root size (r = 0.398, p = 0.039). Cardiovascular events (nonmutually exclusive) in 5 of 27 patients (19%) included death (n = 1), heart failure (n = 1), ventricular tachycardia (n = 1), and atrial tachycardia (n = 3). Increased cardiovascular risk was associated with decreased right ventricular ejection fraction (odds ratio 1.153, confidence interval 1.003 to 1.326, p = 0.046) and smaller ascending aorta diameter (odds ratio 1.758, confidence interval 1.037 to 2.976, p = 0.036). In conclusion, decreased right ventricular ejection fraction and smaller ascending aorta on cardiovascular magnetic resonance were associated with adverse events in rTA. **(Am J Cardiol 2019;124:1636−1642)**

Truncus arteriosus (TA) is a rare form of complex cyanotic congenital heart disease (CHD) (accounting for ∼2% to 4% of CHD lesions) where the pulmonary arteries (PAs), aorta and coronary arteries arise from a single great vessel.[1,2] Complete surgical repair typically occurs in early infancy and involves PA reconstruction, insertion of a valved conduit or homograft between the right ventricular outflow tract (RVOT) and PAs, ventricular septal defect closure and, in some instances, repair or replacement of the native truncal valve. Cardiovascular magnetic resonance imaging (CMR) is an essential tool for surveillance of adults with CHD. It is considered the reference standard for quantification of ventricular volumes and mass[3] and can comprehensively assess vascular anatomy and flows, conduit integrity, and valve dimensions. The CMR characteristics of adults with repaired truncus arteriosus (rTA) have not been systematically studied and associations between CMR measurements and adverse outcomes have not been reported. We aimed to define CMR findings in adults with rTA and to explore the associations between CMR features and adverse clinical outcomes.

Methods

Adults with rTA in infancy were identified from an institutional database and were included if they had ongoing follow-up at our center along with a contemporary CMR study which included multiplanar cine imaging and magnetic resonance angiography (MRA). Patients with incomplete or insufficient CMR data for study analyses were excluded (short axis and 4-chamber cine views as well as MRA sequences were minimum requirements for inclusion). The study was approved by the institutional research ethics review board and the need to obtain informed consent was waived given its retrospective nature.

All CMR studies were performed on a 1.5 Tesla imager (Siemens Avanto Fit or GE Signa Excite). Our clinical CMR protocol for rTA includes multiplanar steady state free-precession (SSFP) cine imaging (axial and short axis stacks, sagittal oblique RVOT and cross-sectional views across the neoaortic root for native valve anatomy), static and time resolved MRA in the coronal plane, and non-breath-held phase contrast flow analysis (PAs ± aorta). Of note, late gadolinium enhancement imaging is not included

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Funding: Dr. Wald has received funding from the Canadian Institutes of Health Research (MOP-119353).

See page 1641 for disclosure information.

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0002-9149/
https://doi.org/10.1016/j.amjcard.2019.08.007

in our routine clinical protocol but is instead reserved for select indications (such as fibrosis/scar assessment in patients with ventricular dysfunction, ventricular tachyarrhythmias, etc) and therefore occurs only rarely during rTA CMR. All measurements were completed by an experienced observer (LRV) who was blinded to clinical outcomes and measurements were compared with published normal values.[4] The intra- and interobserver ICCs for CMR measurements were good to excellent (range 0.88 to 0.99) based on our established and previously published lab protocol.[5] Ventricular end-diastolic and end-systolic volumes, ejection fraction (EF), and mass for RV and left ventricle (LV) were completed on short axis cine SSFP images using commercially available software (QMASS, Medis, Leiden, Netherlands). As previously described, RV and LV endocardial and epicardial borders were manually defined.[5] Atrial dimensions were measured on the 4-chamber cine SSFP view. Cross-sectional measurements for neoaortic root dimensions were made using cross-sectional cine SSFP views across the neoaortic root; measurements were not completed in patients with mechanical valves due to susceptibility artifact. Flow analysis within the PAs ± aorta (when available) was completed using commercial software (QFLOW, Medis, Leiden, Netherlands). High resolution static MRA allowed for double oblique measurements of the aorta and PAs at sequential locations across the length of the vessel using commercially available postprocessing software (TeraRecon, Foster City, California). Vessels with indwelling stents were excluded from analysis due to susceptibility artifact. Measurements were indexed to body surface area.[6]

Demographic characteristics, details of cardiac anatomy, interventional procedures (surgical and catheter) and clinical outcomes were recorded following review of electronic patient records. Data from available electrocardiography, echocardiography, and exercise tests within 6 months of CMR completion were recorded. Anatomic subtypes of TA were assigned following review of pediatric imaging data and surgical reports according to the classification described by Collett and Edwards (Figure 1).[7] The primary endpoint was defined as occurrence of any of the following major adverse cardiac events at the time of last follow-up: death, cardiac transplantation, heart failure (requiring hospital admission for escalation of therapy) and/or hemodynamically significant arrhythmia (sustained atrial or ventricular tachyarrhythmia [AT or VT] >30 seconds). Clinical outcomes were independently ascertained by 2 experienced observers (LRV and RW) and discrepancies were adjudicated as necessary.

Continuous variables were assessed for normality of distribution using the Shapiro-Wilk test and presented as mean ± standard deviation or median with interquartile range, as appropriate. Categorical variables were presented as frequency (percentage). A one-way ANOVA with post-hoc analysis using the Bonferroni correction was used for comparisons of continuous variables within different subtypes of TA. Correlations between continuous variables were assessed using either the Pearson or Spearman correlation coefficient, as appropriate. Univariable logistic regression analysis was performed to evaluate association between CMR parameters and clinical events. A 2-tailed p value <0.05 was considered clinically significant. Statistical analysis was performed using STATA v14.1 (StataCorp, College Station, Texas).

Results

In total, 27 adults with rTA were identified for inclusion (CMR studies completed between 2004 and 2016). Clinical characteristics, CMR measurements and adverse outcomes are shown (Table 1). The median number of cardiac interventions (surgical or catheter) following the initial repair in infancy to the date of last follow-up were 4 (interquartile range 3 to 6) and was not significantly different in the TA subtypes (p = 0.528); the majority of interventions were completed in pediatric life. In adult life, 8 cardiac surgeries were completed in 6 patients (in total, 8 RVOT-PA conduit replacement procedures which included 2 neoaortic valve surgeries [valve repair in 1 patient and valve replacement in 1 patient] and 8 percutaneous catheter interventions in 7 patients (in total, 3 percutaneous pulmonary valve implantations, 4 RVOT-PA conduit interventions for dilation +/− stent implantation, and 1 pulmonary artery stent implantation). Multiple or combined surgical and/or catheter interventions were performed in 6 subjects. There were no statistically significant differences in electrocardiography, echocardiographic or exercise study findings in rTA anatomic subtypes. Adverse cardiovascular events (nonmutually exclusive) were identified in 5 of 27 patients (19%) and included death (n = 1), VT (n = 1, tachycardia of indeterminate ventricular origin), heart failure (n = 1 with pulmonary congestion), and AT (n = 3, all with atrial flutter requiring escalation of medical and/or interventional therapy). There was no association between adverse outcome and rTA subtype. The one death in this cohort occurred as a result of multiorgan dysfunction in a patient with overwhelming sepsis. Additional cardiovascular outcomes of interest, not included in the primary outcome but worthy of mention, included endocarditis (n = 3 on the prosthetic pulmonary valve and n = 1 on a native truncal valve), nonsustained VT (n = 3), nonsustained AT (n = 1), and decline in New York Heart Association Functional Class (n = 1).

The only CMR value which was outside of the normal range was RV mass, which was mildly increased (30 g/m^2 ± 12 [upper limit of normal 28 g/m^2]. Although CMR measurements were within normal range for biventricular volumes, biventricular EF, biatrial measurements and LV mass, it is noteworthy that the LV end-diastolic volume was at the upper limit of normal (98 ± 27 ml/m^2), systolic function was at the lower limit of normal for the RV (RVEF 52% ± 10%) and was borderline low for the LV (LVEF 55% ± 6%). Although the mean neoaortic root size was dilated (22 ± 4 mm/m^2), there was no evidence of dilation in the remainder of the thoracic aorta. The only CMR parameter to differ between the anatomic subtypes was PA systolic peak velocity which was increased in subtype 3 (318 ± 26 cm/s) as compared with subtype 1 (204 ± 81 cm/s) and subtype 2 (164 ± 18 cm/s) (p = 0.029). Correlations of moderate intensity were observed between the total number of cardiovascular interventions in adult life and the following CMR parameters: LV end-diastolic volume

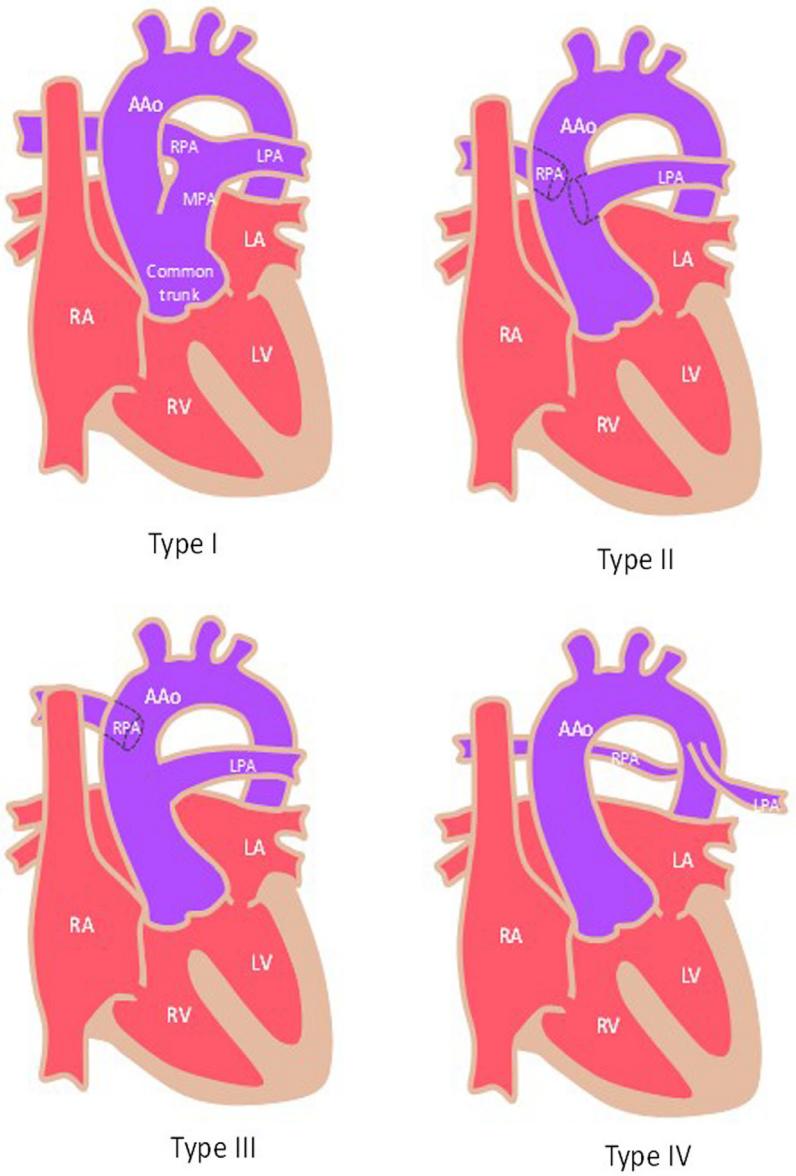

Figure 1. Illustration demonstrating the Collet and Edwards classification of truncus arteriosus subtypes (1-4).

($r = 0.463$, $p = 0.015$), LVEF ($r = -0.425$, $p = 0.027$), and neoaortic root diameter ($r = 0.398$, $p = 0.039$). Characteristic ventricular and vascular abnormalities seen on CMR are demonstrated (Figure 2).

Univariable associations between CMR measurements and the composite primary outcome were analyzed. Increased risk of adverse cardiovascular outcomes was associated with decreased RVEF (odds ratio 1.153, confidence interval 1.003 to 1.323, $p = 0.046$) and decreased ascending aorta diameter (odds ratio 1.758, confidence interval 1.037 to 2.976, $p = 0.036$). Given the relatively low number of adverse outcomes, independent associations using multivariable modeling could not be explored.

Discussion

Unlike the more commonly encountered forms of conotruncal defects, including tetralogy of Fallot or transposition of the great arteries, where substantial literature exists describing imaging characteristics and the prognostic relevance of these, there is a relative paucity of published data pertaining to adults with rTA. In the spectrum of CHD lesions repaired in infancy, TA uniquely predisposes a patient to obligate reintervention over the course of a lifetime, in both the right heart (for RVOT and/or PA rehabilitation) and the left heart (due to neoaortic valve dysfunction or occasionally arch obstruction). Our study provides the first description of CMR features in a cohort of adults with rTA along with identification of CMR measurements which are associated with reintervention in adult life and later development of major adverse cardiovascular events.

Clinical findings and imaging characteristics, of a cohort similar in size to ours, although exclusively pediatric, were described.[8] Direct comparison of the CMR findings revealed that children and adults had identical LV volumes (at the

Table 1
Demographic and imaging features of the study population

Patient number	Age at time of CMR study (years)	Follow-up duration (years)	DiGeorge syndrome (+, present; -, absent)	TA subtype*	LVEDVi (mll/m^2)	LVEF (%)	RVEDVi (ml/m^2)	RVEF (%)	RV mass indexed (g/m^2)	Area of RA, LA (cm^2)	Aortic root (mm)	Ascending aorta (mm)	MPA, RPA, LPA (mm)	Re-intervention (surgery or catheterization) following initial repair	Adverse Cardiovascular outcome
1	18.4	1.1	+	I	136	53	144	58	22	12, 12	41	31	25,15,15	2	0
2	18.6	5.2	+	I	82	53	75	53	28	8,9	41	36	16,8,14	7	0
3	19.0	3.6	-	II	82	60	122	58	33	22,22	41	32	15,12,14	3	0
4	19.4	6.1	-	I	94	58	81	57	46	21,15	39	31	18,16,26	4	0
5	19.4	4.6	-	I	117	62	105	65	33	17,15	45	36	18,12,14	5	0
6	19.5	2.5	-	I	73	61	67	63	29	19,17	41	35	24,10,14	4	0
7	19.9	5.5	-	II	130	52	110	51	23	21,17	39	36	11,7,7	7	0
8	19.9	9.6	-	II	97	51	89	54	35	14,15	38	35	21,14,12	4	0
9	20.0	9.5	-	I	82	56	112	52	30	35,24	35	35	20,13,17	4	0
10	20.4	7.6	+	I	147	54	113	40	59	30,16	40	35	20,13,11	3	0
11	20.8	6.7	-	II	66	56	78	45	35	17,11	34	24	12,10,15	4	0
12	20.8	13.3	-	I	82	52	104	54	18	20,16	35	27	16,10,12	6	Death
13	20.8	2.0	+	II	133	52	84	56	25	19,16	40	25	23,16,15	4	0
14	20.9	3.4	-	III	58	61	106	48	25	22,8	36	30	22,15,17	7	0
15	21.1	2.6	-	I	120	60	143	40	23	22,21	41	28	18,16,11	4	0
16	21.7	3.5	-	I	108	52	67	71	20	26,26	34	29	32,8,9	9	0
17	22.0	5.0	-	III	101	61	111	44	33	21,7	40	26	21,13,15	5	Sustained atrial arrhythmia
18	25.0	5.7	-	II	116	55	94	36	25	20,29	34	34	16,12,16	1	Heart failure
19	25.4	5.6	+	I	84	63	73	72	22	13,8	33	36	10,8,8	4	0
20	25.5	12.3	-	II	71	52	89	53	32	29,11	48	28	19,10,18	8	Sustained atrial arrhythmia
21	27.7	5.0	-	I	81	58	88	58	22	20,12	41	36	22,10,12	2	0
22	28.3	1.2	-	II	71	45	61	56	21	25,17	35	32	16,10,12	2	0
23	30.0	10.6	-	I	158	45	89	52		26,25	43	36	16,10,13	13	0
24	30.2	2.0	-	I	61	47	101	48	27	19,15	35	35	14,9,8	5	0
25	30.2	1.9	-	II	99	59	95	49	19	23,18	31	36	24,19,19	3	0
26	34.0	8.7	-	I	92	42	201	26	71	39,17	45	35	24,11,23	4	Sustained ventricular tachycardia
27	39.8	1.4	-	I	94	60	89	45	20	22,20	43	35	11,8,11	3	0

LA = left atrium; LPA = left pulmonary artery; LVEDVi = indexed left ventricular end diastolic volume; LVEF = left ventricular ejection fraction; MPA = main pulmonary artery; RA = right atrium; RPA = right pulmonary artery; RVEDVi = indexed right ventricular end diastolic volume; RVEF = right ventricular ejection fraction; TA = truncus arteriosus.

* Collett and Edwards classification of truncus arteriosus.

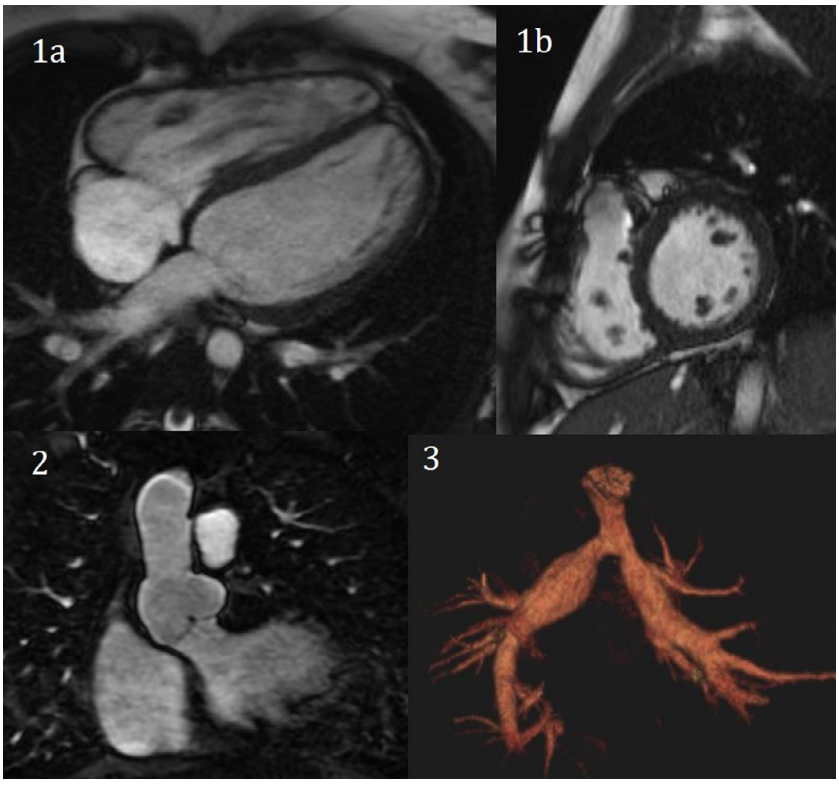

Figure 2. Characteristic ventricular and vascular abnormalities in the adult late after repair truncus arteriosus repair. Enlargement of the left ventricle with left ventricular systolic dysfunction in a patient with symptomatic heart failure (steady state free precession 4 chamber view, *panel 1A*, and short axis view, *panel 1B*); dilation of the neoaortic root in a patient with aortic insufficiency (steady state free precession coronal left ventricular outflow tract view, *panel 2*); and narrowing of the proximal pulmonary arteries (magnetic resonance angiography volume rendered image, *panel 3*).

upper limit of normal) and similar RV volumes (within the normal range); although RVEF and LVEF were well within the normal range for the pediatric patients, EF of the ventricles was at the lower limit of normal versus borderline decreased for the adults, an observation in keeping with other conotruncal lesions reflecting the expected decline in systolic function with age.[9–11] Although RV mass was mildly increased in our cohort (30 g/m^2 ± 12), likely an adaptive response to conduit dysfunction and/or PA stenoses, there was a strikingly high value for RV mass in the pediatric cohort (71 g/m^2 ± 19).[8] Whether this discrepancy reflects underlying differences in ventricular morphology, survival bias or variation in CMR technique is uncertain. It is worth noting that even in conotruncal lesions where RV hypertrophy is a diagnostic feature, such as tetralogy of Fallot, the RV mass was found to be only mildly elevated in multiple large CMR series (ranging between 34 and 36 g/m^2),[10,12] not dissimilar to measurements seen in our cohort. Absent data on aortic dimensions, PA measures, and flow velocities in the pediatric cohort precluded comparison of vascular characteristics in children versus adults.

Despite successful repair in childhood, adults with rTA can anticipate cardiovascular re-intervention over the course of a lifetime, most commonly due to RVOT-PA dysfunction, although intervention to address PA stenoses or neoaortic valve disease may also be required. In our cohort, the number of cardiovascular interventions in adulthood was moderately correlated with various parameters in the left heart, including increased LV size, decreased LVEF and enlarged neo-aortic diameter. Taken together, these findings underscore the importance of left heart disease, perhaps as a surrogate of severity of underlying TA and point to the possibility of adverse RV-LV interactions, as seen in other conotruncal lesions[13] as well as other anatomies.[14]

Two CMR variables measured at baseline were associated with adverse cardiovascular events at last clinical follow-up: impaired RV systolic function and small ascending aorta diameter. The association between decreased RVEF and cardiovascular outcome in rTA is in keeping with other forms of conotruncal disease[11,15,16] and may arise as a result of the need for multiple reinterventions throughout life and/or adaptation to conduit dysfunction. The pathophysiologic link between smaller ascending aorta dimension and cardiovascular outcomes, while less apparent, is hypothesis generating. Whether ascending aorta dimension is a surrogate for disease severity/complexity of surgery at the time of initial repair (likely reflecting the site of PA dislocation), is reflective of abnormal compliance within the aorta or is associated with limitations in cardiac output remains speculative and is beyond the scope of our present study. Of note, there were no patients with arch interruption or arch obstruction requiring surgical repair.

The utility of CMR for evaluation of the adult with CHD, at baseline assessment, and for longer term surveillance, has been firmly and irrefutably established, as this imaging modality can provide comprehensive measurements of anatomy, systolic function and vascular flows, safely and reproducibly.[17,18] Paradoxically, while CMR is

typically the advanced imaging modality of choice for patients with the most complex forms congenital heart disease, given the rarity of such lesions, the clinical context associated with these measurements is often scarce or altogether absent. The present study is a step forward in our understanding of the range of measurements in adults with rTA and the potential prognostic relevance of select variables. Our observations would benefit from further validation in larger populations with longer follow-up.

There are several strengths and limitations to our retrospective which are worthy of mention. Our study is strengthened by the fact that CMR measurements were all completed by a single experienced observer which eliminates variability which could be imposed by multiple observers. Although cine SSFP imaging for assessment of ventricular volumes, function and mass as well as atrial dimensions could be found for all patients studied, data pertaining to valve anatomy and function (stenosis or insufficiency) were inconsistently available; additionally, assessment of replacement fibrosis (using late gadolinium enhancement imaging) or diffuse fibrosis (by parametric mapping) was largely absent although potentially of clinical relevance. Because we reviewed a select population of patients with CMR studies, our clinical findings may not be reflective of the full spectrum of adults with rTA. The size of our study population is modest, reflecting a relatively rare form of CHD, where subtypes of extreme complexity are rarer still. Although we report on a population with predominance of Collet and Edwards subtypes 1 and 2, which is similar to previous reports, we were unable to adequately characterize subtypes which were under-represented (only 2 patients in this cohort had subtype 3) or absent altogether (no patients with subtype 4). Of note, the elevated PA velocities in subtype 3 are not unexpected and likely reflect a combination of native anatomy and sequelae from surgical repair. Finally, the small number of adverse events precluded exploration of the independent associations between CMR measurements and outcomes. Study of a larger population with longer follow-up, along with serial CMR studies, would likely yield more robust data regarding the prognostic relevance of CMR measurements in this high-risk population.

In conclusion, in a cohort of adults with rTA and contemporary CMR studies, RV mass was mildly increased although biventricular volumes, biventricular systolic function, LV mass, and atrial dimensions were within the normal range. Two CMR variables were associated with major adverse cardiovascular events at the time of last follow-up: decreased RVEF and smaller ascending aorta.

Acknowledgment

The authors gratefully acknowledge Elyar Abbasi Bavil for his work on the figures included in this manuscript.

Disclosures

The authors have no conflicts of interest to disclose.

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