cambridge.org/cty

Original Article

Cite this article: Jones AL, White BR, Ghosh RM, Mondal A, Ampah S, Ho DY, Whitehead K, Harris MA, Biko DM, Partington S, Fuller S, Cohen MS, and Fogel MA (2024) Cardiac magnetic resonance predictors for successful primary biventricular repair of unbalanced complete common atrioventricular canal. Cardiology in the Young 34: 387–394. doi: 10.1017/S1047951123001786

Received: 24 March 2023 Revised: 22 May 2023 Accepted: 7 June 2023 First published online: 18 July 2023

Keywords:

CHD; cardiac MRI; echocardiography; atrioventricular canal

Corresponding author: A. L. Jones; Email: Jonesa21@chop.edu

Cardiac magnetic resonance predictors for successful primary biventricular repair of unbalanced complete common atrioventricular canal

Andrea L. Jones¹, Brian R. White¹, Reena M. Ghosh¹, Antara Mondal², Steve Ampah², Deborah Y. Ho³, Kevin Whitehead¹, Matthew A. Harris¹, David M. Biko⁴, Sara Partington¹, Stephanie Fuller⁵, Meryl S. Cohen¹ and Mark A. Fogel¹

¹Division of Pediatric Cardiology, Department of Pediatrics, The Children's Hospital of Philadelphia, Philadelphia, PA, USA; ²Department of Biomedical & Health Informatics, The Children's Hospital of Philadelphia, Philadelphia, PA, USA; ³Division of Pediatric Cardiology, Department of Pediatrics, Stanford University School of Medicine, Palo Alto, CA, USA; ⁴Department of Radiology, The Children's Hospital of Philadelphia, Philadelphia, PA, USA and ⁵Division of Cardiothoracic Surgery, The Children's Hospital of Philadelphia and the University of Pennsylvania, Philadelphia, PA, USA

Abstract

Background: Patients with unbalanced common atrioventricular canal can be difficult to manage. Surgical planning often depends on pre-operative echocardiographic measurements. We aimed to determine the added utility of cardiac MRI in predicting successful biventricular repair in common atrioventricular canal. Methods: We conducted a retrospective cohort study of children with common atrioventricular canal who underwent MRI prior to repair. Associations between MRI and echocardiographic measures and surgical outcome were tested using logistic regression, and models were compared using area under the receiver operator characteristic curve. Results: We included 28 patients (median age at MRI: 5.2 months). The optimal MRI model included the novel end-diastolic volume index (using the ratio of left ventricular end-diastolic volume to total end-diastolic volume) and the left ventricle-right ventricle angle in diastole (area under the curve 0.83, p = 0.041). End-diastolic volume index \leq 0.18 and left ventricle-right ventricle angle in diastole ≤ 72° yield a sensitivity of 83% and specificity of 81% for successful biventricular repair. The optimal multimodality model included the end-diastolic volume index and the echocardiographic atrioventricular valve index with an area under the curve of 0.87 (p = 0.026). Conclusions: Cardiac MRI can successfully predict successful biventricular repair in patients with unbalanced common atrioventricular canal utilising the end-diastolic volume index alone or in combination with the MRI left ventricle-right ventricle angle in diastole or the echocardiographic atrioventricular valve index. A prospective cardiac MRI study is warranted to better define the multimodality characteristic predictive of successful biventricular surgery.

In patients with unbalanced complete common atrioventricular canal defect, surgical planning can be difficult. ^{1,2} Echocardiography has traditionally been used to determine the probability of success of a biventricular repair using indices such as the atrioventricular valve index, ^{3,4} the left ventricle–right ventricle in diastole, ^{5,6} and the left ventricle inflow index. ⁷ However, two-dimensional echocardiography is inherently limited in its ability to describe three-dimensional structures, and there is an opportunity to improve the prediction of successful biventricular canal repair.

Cardiac MRI offers advantages over echocardiography in its ability to accurately calculate three-dimensional anatomic and functional measurements (i.e., it is the gold standard of ventricular volumes and performance) and quantify flow. Previous cardiac MRI studies predicted successful biventricular repair in a small, heterogeneous cohort of patients with left ventricular hypoplasia, as well as successful univentricular to biventricular conversion in patients with common atrioventricular canal. 9,10 The utility of cardiac MRI in primary common atrioventricular canal repair has not been studied.

The objective of this investigation is to determine the utility of cardiac MRI in predicting biventricular repair in unbalanced common atrioventricular canal defect. We hypothesise that novel cardiac MRI metrics will predict successful biventricular repair.

© The Author(s), 2023. Published by Cambridge University Press.



388 A. L. Jones et al.

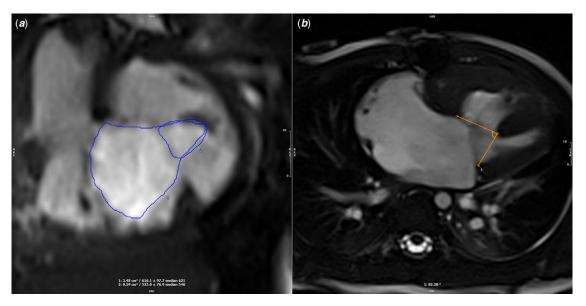


Figure 1. Example CMR measurements. a. Left and total AV valve area measured from a short-axis view. b. LV-RV angle in diastole measured from four-chamber view. AV = atrioventricular; CMR = cardiac MRI; LV = left ventricle; RV = right ventricle.

Materials and method

Subjects

We conducted a single-centre, retrospective cohort study of all patients undergoing cardiac MRI at our institution from July 2010 to February 2021. Complete common atrioventricular canal subjects were enrolled consecutively if they underwent cardiac MRI and at least one echocardiogram with adequate images for analysis prior to definitive repair (biventricular repair or superior cavopulmonary anastomosis), decision to list for transplant, or death. Cardiac MRI referral was at the discretion of the clinical care team. Subjects with conotruncal defects were excluded if they underwent univentricular palliation primarily due to the conotruncal defect, based on chart review (six patients). Subjects were also excluded if they died due to non-cardiac (e.g., malignancy) or unknown causes (two patients). This study was approved by our Institutional Review Board and was exempt from the need to obtain informed consent.

Cardiac magnetic resonance

Cardiac MRIs were obtained on Siemens Avanto or Avanto FIT systems (Erlangen, Germany). The cardiac MRI protocol is detailed in supplemental methods. Cardiac MRI variables were re-measured offline by a single observer (RMG) using CVI42 (Circle Cardiovascular, Calgary, ON, Canada) (Fig. 1). Measurements included (a) left ventricle and right ventricle end-diastolic and end-systolic volumes, (b) atrioventricular valve areas (short-axis images, Fig. 1a), (c) left ventricle and right ventricle inflow diameters (four-chamber view), (d) through-plane and (when available) in-plane PCMR left ventricle and right ventricle inflow volumes, (e) left ventricle-right ventricle angle in systole and diastole (four-chamber view, Fig. 1b), (f) ventricular septal defect size in systole and diastole (four-chamber view), (g) pulmonary and systemic blood flow, and (h) common atrioventricular canal inflow. We then calculated (a) left ventricle and right ventricle ejection fraction, (b) ratios of ventricular volumes, (c) ratios of atrioventricular valve areas, (d) ratios of inflow volumes, (e) ratios of inflow volumes to ventricular

volumes, (f) ratio of pulmonary to systemic blood flow, and (g) common atrioventricular canal regurgitant volume. Absolute measurements, such as ventricular volume, were indexed to body surface area for all subjects.

Due to the inclusion of both right- and left-dominant canals in the cohort, several variables were standardised to their absolute deviation from the expected values for a balanced canal: $M_{standardized} = |M_{measured} - M_{expected}|.$

The ratio of the left atrioventricular valve area to total common atrioventricular valve area (the MRI equivalent of the modified atrioventricular valve index) and the ratio of the left ventricle inflow volume to the total common inflow were standardised to an expected value of 0.5.³ For example, patients with a modified atrioventricular valve index of 0.3 and 0.7 would both have a standardised atrioventricular valve index of 0.2, indicating a similar level of deviation from balance. The ratio of the left ventricle end-diastolic volume to total end-diastolic volume was standardised to an expected value of 0.483 based on normal values published by Altmayer et al.¹¹ The standardised left ventricle end-diastolic volume ratio is hereafter referred to as the end-diastolic volume index.

Echocardiography

The echocardiogram obtained closest in time to the cardiac MRI and prior to definitive repair with views adequate for measuring all variables was used. Echocardiograms were unsedated. Echocardiographic variables were re-measured offline by a single observer (ALJ) using Syngo (Siemens Healthineers, Erlangen, Germany). Measurements included (a) left and right atrioventricular valve areas (subcostal view), (b) left ventricle and right ventricle inflow diameter (apical four-chamber view), (c) left ventricle and right ventricle colour inflow diameter (apical four-chamber view), and (d) left ventricle–right ventricle angle in systole and diastole (apical four-chamber view). From these measurements, the modified atrioventricular valve index and left ventricle inflow index were calculated. A standardised atrioventricular valve index was calculated as the difference from an expected value of 0.5.3

Statistical analysis

The primary outcome was successful biventricular canal repair, defined as survival with a biventricular circulation at last follow-up. Patients who underwent univentricular palliation with a superior cavopulmonary anastomosis, were listed for transplant, remained palliated with a pulmonary artery band, or died were classified as not having a successful biventricular repair. The Wilcoxon rank-sum test and Pearson's chi-squared test or Fisher's exact test were used to compare continuous and categorical clinical characteristics between groups, as appropriate. *P*-values < 0.05 were considered statistically significant. All calculations were performed in R Statistical Software, version 4.1.2 (Vienna, Austria).

Intra-rater, inter-rater, and inter-modality reliability were calculated using intra-class correlation coefficients across 20 subjects in a blinded fashion. Echocardiographic measurements were compared across three observers (ALJ, BRW, and DYH). Intra-rater comparisons were performed across measurements repeated at least 2 weeks apart. Inter-modality reliability was calculated between echocardiographic measurements and analogous cardiac MRI measurements.

We first used logistic regression to develop univariable and multivariable models using cardiac MRI measurements. We explored if the predictive power of the model could be improved by allowing non-linear relationships using restricted cubic splines. The optimal univariable model was selected with a high receiver operator characteristic area and statistical significance. For the multivariable models, predictors were limited to 2 due to sample size. Given the number of cardiac MRI variables, only predictors with *P*-values < 0.2 in the univariable models were considered for the cardiac MRI multivariable model. The optimal multivariable model was selected using the area under the receiver operator characteristic curve, good model calibration accuracy, and overall statistical significance. The final multivariable model was validated using the Bootstrap method with re-estimation of 500 random samples.

Univariable and multivariable prediction models were developed for the echocardiographic parameters similarly. The optimal cardiac MRI model was compared to the optimal echocardiographic model using the area under the receiver operator characteristic curve. The area under the receiver operator characteristic curve and likelihood ratio test were used to determine the added value of the best single cardiac MRI predictor to the best single echocardiographic predictor.

Results

Cohort description

Twenty-eight patients were included (Table 1). Eleven (39%) had successful biventricular repairs. Eight (47%) had primary single-ventricle palliations, one (5%) initially underwent biventricular repair and did not survive to hospital discharge, two (12%) remained palliated with a pulmonary artery band, one (5%) was transplanted, and five (29%) died prior to repair. There were no significant differences between groups.

Cardiac magnetic resonance imaging model

The best single predictor of successful biventricular repair (Table 2) was the end-diastolic volume index with an area under the receiver operator characteristic curve of 0.82 (p < 0.01, Fig. 2a). The standardised atrioventricular index was also statistically

significant; however, the area under the receiver operator characteristic curve was lower (area under the curve 0.66, p = 0.01). The optimal cut-point for the end-diastolic volume index for successful biventricular repair was \leq 0.18, which corresponds to a left ventricle end-diastolic volume to total end-diastolic volume ratio of 0.30–0.66, yielding a sensitivity of 91% and a specificity of 65%. In a sensitivity analysis using restricted cubic splines, our data lacked sufficient evidence that the predictive ability of developed models could be improved by assuming non-linear relationships of predictors with the outcome (Supplemental Table 1).

Three multivariable models were created (Table 3), with the optimal multivariable model combining the end-diastolic volume index and the left ventricle–right ventricle angle in diastole, resulting in an area under the receiver operator characteristic curve of 0.83 (p = 0.041) (Fig. 2b). This improvement in area under the receiver operator characteristic curve compared to the univariable model was not statistically significant (likelihood ratio test, p = 0.19). Cut-points of end-diastolic volume index \leq 0.18 and left ventricle–right ventricle angle in diastole \leq 72° yield a sensitivity of 83% and specificity of 81%.

Echocardiographic model

The best single predictor of successful biventricular repair (Table 2) was the standardised atrioventricular valve index with an area under the receiver operator characteristic curve of 0.83 (p < 0.01) (Fig. 2c). An optimal cut-point of standardised atrioventricular valve index ≤ 0.11 (equivalent to a modified atrioventricular valve index of 0.39–0.61) had a sensitivity of 73% and a specificity of 76%. The optimal multivariable model (Table 4) combined the standardised atrioventricular valve index and the left ventricle–right ventricle angle in diastole with an area under the receiver operator characteristic curve of 0.85 (p = 0.042). Optimal cut-points of standardised atrioventricular valve index ≤ 0.11 and left ventricle–right ventricle angle in diastole $\leq 100^\circ$ had a sensitivity of 73% and a specificity of 82%. The multivariable model was not statistically significantly improved compared to the univariable model (likelihood ratio test, p = 0.44).

Multimodality model

The optimal multimodality model (Table 5) combining cardiac MRI end-diastolic volume index and echocardiographic standardised atrioventricular valve index yielded a high area under the receiver operator characteristic curve of 0.87 (p = 0.026, Fig. 2d). Optimal cut-points of cardiac magnetic resonance imaging end-diastolic volume index \leq 0.18 and echocardiographic standardised atrioventricular valve index \leq 0.11 had a sensitivity of 73% and specificity of 76%. The improvement in area under the receiver operator characteristic curve from 0.83 to 0.87 with the addition of the end-diastolic volume index to the echocardiographic atrioventricular valve index was not statistically significant (likelihood ratio test, p = 0.22).

Reliability

Cardiac MRI intra-rater reliability was excellent (≥0.97, Table 6). Left ventricle inflow index could not be calculated by cardiac MRI because only four of the 28 patients had in-plane velocity mapping of ventricular inflows. By echocardiography, the modified atrioventricular valve index and left ventricle–right ventricle angle in diastole showed moderate to good intra- and inter-rater

390 A. L. Jones et al.

Table 1. Cohort clinical characteristics grouped by status of biventricular outcome.

	Total	Biventricular outcome	Non-biventricular outcome	
Variable	(N = 28)	(N = 11)	(N = 17)	р
Age at CMR (months)	5.2 (3.9-6.8)	5.2 (4.9–7.5)	4.7 (2.2-6.7)	0.25
Time between CMR and echocardiogram (days)	29 (7–56)	30 (20–56)	27 (4–60)	0.96
Female	15 (54%)	8 (73%)	7 (41%)	0.14
Larger right ventricle	17 (61%)	4 (36%)	13 (76%)	0.05
Genetic syndrome	13 (46%)	6 (55%)	7 (41%)	0.49
Heterotaxy syndrome	7 (25%)	2 (18%)	5 (29%)	0.67
Conotruncal anomaly	18 (64%)	7 (64%)	11 (65%)	> 0.99
Type of conotruncal anomaly				0.25
Tetralogy of Fallot	8 (29%)	4 (36%)	4 (24%)	
Double-outlet right ventricle	5 (18%)	1 (9%)	4 (24%)	
Right ventricle to aorta and pulmonary atresia	2 (7%)	0 (0%)	2 (12%)	
Transposition of the great arteries	2 (7%)	2 (18%)	0 (0%)	
Truncus arteriosus	1 (4%)	0 (0%)	1 (6%)	
Palliative procedure	17 (61%)	7 (64%)	10 (59%)	0.80
Type of palliative procedure				0.63
Pulmonary artery band	9 (32%)	4 (36%)	5 (29%)	
Aortopulmonary shunt or stent	6 (21%)	3 (18%)	3 (27%)	
Stage I	2 (7%)	0 (0%)	2 (12%)	
Common atrioventricular valve regurgitation (%)	9 (2, 17)	6 (0, 12)	9.5 (4, 17.5)	0.49
More than one CMR	4 (14%)	2 (18%)	2 (12%)	> 0.99
Age at repair (months)	6.8 (6.1–11.0)	6.6 (6.2–13.2)	7.4 (5.7–10.4)	0.97
Follow-up time after repair (months)	20.6 (2.0, 66.5)	26.2 (2.0, 67.0)	17.1 (2.1, 61.1)	0.79

 $\mathsf{CMR} = \mathsf{cardiac} \; \mathsf{MRI}.$

Summary statistics shown as median (IQR) or n (%).

reliability (\geq 0.72). The left ventricle inflow index showed poor intra- and inter-rater reliability and thus was not used in further analyses. Inter-modality reliability for the modified atrioventricular valve index was good but was poor for the left ventricle-right ventricle angle.

Discussion

Surgical decision-making in patients with borderline unbalanced common atrioventricular canal remains challenging. Failure of a biventricular circulation results in re-interventions to address residual lesions and increased mortality often due to left heart failure and pulmonary vascular disease.² Creating a univentricular circulation when a biventricular repair could have been performed exposes patients to the long-term risks of a Fontan circulation, including hepatic fibrosis, heart failure, poor exercise performance, and lymphatic congestion.¹² Our retrospective study demonstrated that cardiac MRI can predict biventricular repair. The best cardiac MRI predictors measured different characteristics of common atrioventricular canal anatomy than commonly used echocardiographic indices. Further, cardiac MRI intra-rater reliability was higher than echocardiography, emphasising cardiac MRI

reliability. Thus, we believe that cardiac magnetic resonance imaging can play a complementary role in echocardiography.

Historically, the surgical approach for patients with common atrioventricular canal has been primarily determined using echocardiography. In 2008, Grosse-Wortmann et al reported on 20 patients with borderline left ventricular hypoplasia. Using a left ventricle end-diastolic volume cut-off of at least 20 mL/m², cardiac MRI correctly predicted biventricular repair in all 16 patients who survived biventricular repair. However, only five patients had common atrioventricular canal defects, and patients with hypoplastic right ventricles were not considered. Thus, the generalisability of these results to common atrioventricular canal is unclear. Nathan et al and Banka et al have reported on cardiac MRI metrics in patients with common atrioventricular canal undergoing univentricular to biventricular conversion. 9,10 They reported left ventricle end-diastolic volume cut-offs of > 20 mL/m² and 22 mL/m² as predictive of successful conversion. It is unknown if the criteria for successful biventricular conversion also apply to primary repair.

Our study examines cardiac MRI predictors of primary biventricular repair in patients with common atrioventricular canal and includes patients with both left- and right-dominant

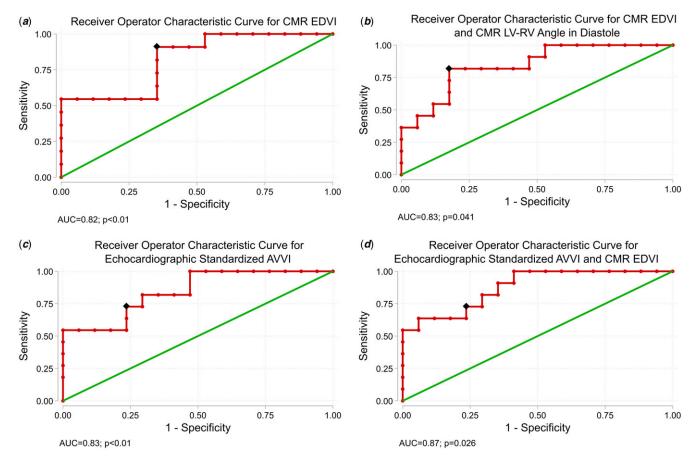


Figure 2. ROC curves for models predicting bi-ventricular repair. \boldsymbol{a} . ROC curve for CMR EDVI. The optimal cut-point (black diamond) of the EDVI is ≤ 0.18 : sensitivity of 92% and specificity of 65%. \boldsymbol{b} . ROC curve for CMR EDVI and CMR LV-RV angle in diastole. The optimal cut-point is an EDVI of ≤ 0.18 and LV-RV angle in diastole of $\leq 72^\circ$: sensitivity of 83% and specificity of 81%. \boldsymbol{c} . ROC curve for echocardiographic standardised AVVI. The optimal cut-point of the standardised AVVI is ≤ 0.11 : sensitivity of 73% and specificity of 76%. \boldsymbol{d} . ROC curve for the multimodality model of echocardiographic standardised AVVI and CMR EDVI. The optimal cut-point is an EDVI of ≤ 0.18 and a standardised AVVI of ≤ 0.11 : sensitivity of 73% and specificity of 76%. AUC = area under the curve; AVVI = atrioventricular valve index; CMR = cardiac MRI; EDVI = end-diastolic volume index; LV = left ventricle; ROC = receiver operator characteristic; RV = right ventricle.

ventricles. Like the above studies, we found that ventricular volume measurements had the highest predictive value. In our cohort, the minimum left ventricle end-diastolic volume for successful biventricular repair was 18 ml/m². We found that an end-diastolic volume index ≤ 0.18 (left ventricle end-diastolic volume to total end-diastolic volume ratio of 0.30–0.66) and a left ventricle–right ventricle angle in diastole $\leq 72^{\circ}$ resulted in a sensitivity of 83% and a specificity of 81% of predicting successful biventricular repair (Fig. 2).

Our findings regarding the direction of association of the left ventricle–right ventricle angle in diastole differ from previously published echocardiographic literature. Cohen et al reported that larger left ventricle–right ventricle angles were associated with biventricular repair in common atrioventricular canal.⁵ A smaller angle may be the result of a larger ventricular septal defect, and larger ventricular septal defects have been reported as risk factors in patients with common atrioventricular canal for unsuccessful biventricular repair.^{3,6} In our cohort, patients with a larger left ventricle–right ventricle angle had lower odds of successful biventricular repair. One possible explanation is that echocardiography tends to foreshorten the four-chamber view, whereas cardiac MRI, due to its 3D nature, can obtain a true four-chamber view. Another explanation may be that our cohort included 64%

with conotruncal anomalies. As such, a larger ventricular septal defect may have allowed more flexibility in the overall repair.

We aimed to determine the added value of cardiac MRI to commonly used echocardiographic parameters such as the modified atrioventricular valve index (left atrioventricular valve area/total atrioventricular valve area).^{3,4} In our cohort, a modified atrioventricular valve index by echocardiography of 0.39-0.61 predicted biventricular repair, similar to previous studies. The addition of cardiac MRI end-diastolic volume index in a multimodality model improved the area under the receiver operator characteristic curve from 0.83 to 0.87; however, the change was not statistically significant, most likely due to our small sample size. We postulate that the addition of cardiac MRI parameters to echocardiographic parameters may improve accuracy in a larger cohort. We did find higher reliability of cardiac MRI measurements than echocardiographic measurements, giving added value to these metrics. We hope that the present study will inspire a prospective study of the use of preoperative cardiac MRI in patients with common atrioventricular canal, which would additionally allow us to capture a more representative cohort of common atrioventricular canal patients.

There are the usual inherent limitations to a retrospective study such as this. In addition, our small sample size limited our

Table 2. Summary statistics and univariable logistic regression for the association between CMR and echocardiographic parameters and successful biventricular repair (median (IQR)).

	Total	Biventricular outcome	Non-biventricular outcome		Univariable logis	ic regression	
Variable	N = 28	N = 11	N = 17	OR	95% CI	Р	AUC
CMR variables							
Ventricular volume							
EDVI	0.18 (0.09–0.24)	0.06 (0.05-0.18)	0.22 (0.14–0.28)	0.21*	0.06, 0.70*	< 0.01	0.82
Ventricular inflow							
Standardised LV inflow volume: total inflow volume	0.11 (0.09-0.15)	0.10 (0.03-0.16)	0.11 (0.10-0.14)	0.48*	0.12, 1.95*	0.28	0.60
Atrioventricular valve area							
Standardised AVVI	0.13 (0.10-0.16)	0.13 (0.04–0.15)	0.13 (0.12–0.19)	0.14*	0.02, 0.90*	0.01	0.66
VSD size							
VSD size (diastole, cm/m²)	4.47 (3.52–6.00)	5.16 (4.19–6.00)	4.25 (3.13–6.01)	1.13	0.76, 1.71	0.53	0.63
VSD size (systole, cm/m²)	2.32 (1.98–3.56)	3.00 (2.00–3.90)	2.27 (2.02–3.52)	1.22	0.59, 2.54	0.59	0.54
LV–RV angle (diastole, degrees)	82 (71–95)	75 (69–83)	88 (81–96)	0.95	0.89, 1.00	0.06	0.71
LV–RV angle (systole, degrees)	99 (85–112)	93 (83–102)	103 (88–116)	0.97	0.92, 1.02	0.20	0.65
Other							
LV EF (%)	60 (56–69)	60 (56–70)	63 (56–68)	1.01	0.95, 1.09	0.72	0.52
RV EF (%)	63 (58–72)	68 (62–72)	62 (57–72)	1.05	0.97, 1.15	0.28	0.60
Total atrioventricular valve regurgitation (%)	9 (2–17)	6 (0–12)	10 (5–17)	0.95	0.82, 1.07	0.44	0.61
Qp:Qs	1.90 (1.16–2.69)	1.74 (1.26–2.57)	2.00 (1.07–2.65)	0.86	0.47, 1.35	0.53	0.50
Echocardiographic variables							
Atrioventricular valve area							
Standardised AVVI	0.12 (0.06-0.21)	0.05 (0.02–0.11)	0.16 (0.11–0.23)	0.14*	0.03, 0.61*	< 0.01	0.83
VSD size							
LV–RV angle (diastole, degrees)	76 (68–89)	72 (69–83)	78 (64–91)	0.99	0.94, 1.04	0.67	0.55
LV–RV angle (systole, degrees)	93 (80–109)	89 (84–104)	98 (78–109)	1.00	0.95, 1.04	0.93	0.54

AUC = area under the curve; AVVI = atrioventricular valve index; CMR = cardiac MRI; EDVI = standardised LV EDV to total EDV ratio; LV = left ventricle; Qp = pulmonary blood flow; Qs = systemic blood flow; RV = right ventricle; VSD = ventricular septal defect.
*OR and CI for change in metric by 0.1.

Table 3. Multivariable models using CMR parameters to predict successful biventricular repair.

Variable 1	Variable 2	AUC	Р
EDVI	Standardised AVVI	0.77	0.07
EDVI	LV–RV angle (diastole, degrees)	0.83	0.04
Standardised AVVI	LV–RV angle (diastole, degrees)	0.81	0.10

AUC = area under the curve; AVVI = atrioventricular valve index; CMR = cardiac MRI; EDVI = standardised LV EDV to total EDV ratio; LV = left ventricle; RV = right ventricle.

Table 4. Multivariable models using echocardiographic parameters to predict successful biventricular repair.

Variable 1	Variable 2	AUC	Р
Standardised AVVI	LV–RV angle (diastole, degrees)	0.85	0.04
Standardised AVVI	LV–RV angle (systole, degrees)	0.83	0.04

AUC = area under the curve; AVVI = atrioventricular valve index; LV = left ventricle; RV = right ventricle

Table 5. Multivariable models using CMR and echocardiographic parameters to predict successful biventricular repair.

Echocardiographic variable	CMR variable	AUC	Р
Standardised AVVI	EDVI	0.87	0.03
Standardised AVVI	Standardised modified AVVI	0.85	0.05
Standardised AVVI	LV–RV angle (diastole, degrees)	0.88	0.04

AUC = area under the curve; AVVI = atrioventricular valve index; CMR = cardiac MRI; EDVI = standardised LV EDV to total EDV ratio; LV = left ventricle; RV = right ventricle.

statistical power. We were also restricted to two variables in the multivariable models, which limited our ability to investigate more complex interactions between various measurements. Additionally, we did not have adequate sample size for both a derivation and validation cohort. We addressed this limitation using Bootstrap cross-validation analysis.

All patients in our study were referred for clinically indicated cardiac MRI, which is not current standard of care. Thus, our cohort of common atrioventricular canal patients were more likely to have complex cardiac defects such as conotruncal anomalies that led to the decision to undergo cardiac MRI and may have affected surgical outcome. Overall, the presence of additional cardiac anomalies was not itself associated with surgical outcome, and our echocardiography results align with prior published literature. Thus, we can have confidence that our results should generalise to common atrioventricular canal patients as a whole.

Finally, we were limited in our analysis by the images available. Based on the predictive value of the left atrioventricular inflow index measured by echocardiography, we hypothesised that a similar calculation using the ventricular inflow by cardiac MRI would also have predictive value.⁷ However, very few patients had in-plane velocity mapping of the ventricular inflows; thus, we were

Table 6. Intra-class correlation coefficients.

Measurement	Intra-rater reliability	Inter-rater reliability	Inter-modality reliability
Echocardiography			
Modified AVVI	0.74	0.78	0.87
LVII	0.53	0.40	N/A
LV–RV angle (diastole)	0.89	0.72	0.50
CMR			
Left atrioventricular valve area	0.99		
Total atrioventricular valve area	0.99		
LV–RV angle (systole)	0.99		
LV–RV angle (diastole)	0.98		
LV 2D inflow	0.99		
RV 2D inflow	0.99		
VSD size (systole)	0.97		
VSD size (diastole)	0.99		

 $AVVI = a trioventricular \ valve \ index; \ CMR = cardiac \ MRI; \ LV = left \ ventricle; \ LVII = left \ ventricular \ inflow \ index; \ RV = right \ ventricle; \ VSD = ventricular \ septal \ defect.$

unable to calculate a cardiac MRI inflow index. Further investigation is needed with a prospective study to validate our findings and explore other possible cardiac MRI predictors of surgical outcome in common atrioventricular canal.

Conclusions

Cardiac MRI can successfully predict biventricular repair in patients with unbalanced common atrioventricular canal utilising the end-diastolic volume index alone or in combination with the left ventricle-right ventricle angle in diastole. Cut-points of enddiastolic volume index ≤ 0.18 (left ventricle end-diastolic volume to total end-diastolic volume ratio of 0.30-0.66) and left ventricleright ventricle angle in diastole ≤ 72° yield a sensitivity of 83% and specificity of 81% to predict successful biventricular repair. The end-diastolic volume index in combination with the echocardiographic modified atrioventricular valve index can make this prediction with a maximum area under the receiver operator characteristic curve of 0.87. As cardiac MRI predictors measure ventricular volumes and echocardiographic predictors measure atrioventricular valve geometry, along with higher cardiac MRI reliability, cardiac MRI is complementary to echocardiography to predict successful biventricular repair. At our institution, cardiac MRI is becoming a more commonly used tool in the evaluation of unbalanced common atrioventricular canal, in particular in the setting of borderline left ventricular volume and conotruncal abnormalities. We hope to undertake a prospective cardiac MRI investigation to better define the physiology and anatomy of

394 A. L. Jones et al.

unbalanced common atrioventricular canal to determine which patients will have successful biventricular surgery.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123001786.

Acknowledgements. None.

Financial support. This work was supported by a grant from Matthew's Hearts of Hope and by the Children's Hospital of Philadelphia Cardiac Center. Dr Andrea Jones was supported by T32-HL007915. Dr Brian White was supported by K08-NS117897.

Competing interests. None.

Ethical standards. This study protocol was approved by the Institutional Review Board at our institution and deemed to be exempt from the need to obtain informed consent (11/07/2019).

References

- Shaddy RE, Lippincott Williams & Wilkins. Moss & Adams' heart disease in infants, children, and adolescents including the fetus and young adult. 10th edn. Philadelphia: Wolters Kluwer, 2022.
- Hickey EJ, Caldarone CA, Blackstone EH, et al. Critical left ventricular outflow tract obstruction: the disproportionate impact of biventricular repair in borderline cases. J Thorac Cardiovasc Surg2007; 134: 1429–1436. DOI: 10.1016/j.jtcvs.2007.07.052.
- Cohen MS, Jacobs ML, Weinberg PM, Rychik J. Morphometric analysis of unbalanced common atrioventricular canal using two-dimensional echocardiography. J Am Coll Cardiol 1996; 28: 1017–1023. DOI: 10. 1016/s0735-1097(96)00262-8.
- 4. Jegatheeswaran A, Pizarro C, Caldarone CA, et al. Echocardiographic definition and surgical decision-making in unbalanced atrioventricular

- septal defect: a congenital heart surgeons' society multiinstitutional study. Circulation 2010; 14 122: S209–15. DOI: 10.1161/CIRCULATIONAHA. 109.925636.
- Cohen MS, Jegatheeswaran A, Baffa JM, et al. Echocardiographic features defining right dominant unbalanced atrioventricular septal defect: a multiinstitutional congenital heart surgeons' society study. Circ Cardiovasc Imaging 2013; 6: 508–513. DOI: 10.1161/CIRCIMAGING.112.000189.
- Arunamata A, Balasubramanian S, Mainwaring R, Maeda K, Selamet Tierney ES. Right-dominant unbalanced atrioventricular septal defect: echocardiography in surgical decision making. J Am Soc Echocardiogr 2017; 30: 216–226. DOI: 10.1016/j.echo.2016.10.017.
- Szwast AL, Marino BS, Rychik J, Gaynor JW, Spray TL, Cohen MS. Usefulness of left ventricular inflow index to predict successful biventricular repair in right-dominant unbalanced atrioventricular canal. Am J Cardiol 2011; 107: 103–109. DOI: 10.1016/j.amjcard.2010.08.052.
- 8. Grosse-Wortmann L, Yun TJ, Al-Radi O, et al. Borderline hypoplasia of the left ventricle in neonates: insights for decision-making from functional assessment with magnetic resonance imaging. J Thorac Cardiovasc Surg 2008; 136: 1429–1436. DOI: 10.1016/j.jtcvs.2008.04.027.
- Nathan M, Liu H, Pigula FA, et al. Biventricular conversion after singleventricle palliation in unbalanced atrioventricular canal defects. Ann Thorac Surg 2013; 95: 2086–2095. DOI: 10.1016/j.athoracsur.2013.01.075.
- Banka P, Schaetzle B, Komarlu R, Emani S, Geva T, Powell AJ. Cardiovascular magnetic resonance parameters associated with early transplant-free survival in children with small left hearts following conversion from a univentricular to biventricular circulation. J Cardiovasc Magn Reson 2014; 7 16: 73. DOI: 10.1186/s12968-014-0073-1.
- Altmayer SP, Patel AR, Addetia K, Gomberg-Maitland M, Forfia PR, Han Y. Cardiac MRI right ventricle / left ventricle (RV/LV) volume ratio improves detection of RV enlargement. J Magn Reson Imaging 2016; 43: 1379–1385. DOI: 10.1002/jmri.25110.
- Metcalf MK, Rychik J. Outcomes in hypoplastic left heart syndrome. Pediatr Clin North Am 2020; 67: 945–962. DOI: 10.1016/j.pcl.2020.06.008.