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ORIGINAL ARTICLE
Adult congenital heart disease

Aortic arch shape after arch repair predicts exercise capacity: a multicentre analysis

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Aims

Coarctation of the aorta is associated with long-term morbidity including decreased exercise capacity, despite successful repair. In the absence of discrete recoarctation, the haemodynamic mechanism remains unknown. This multicentre study evaluated the relationship between aorta shape, flow, and exercise capacity in patients after arch repair, specifically through the lens of aortic size mismatch and descending aortic (DAo) flow and their association with exercise.

Methods and results

Cardiac magnetic resonance, cardiopulmonary exercise test, and echocardiogram data within 1 year were analysed from 58 patients (age 28 ± 10 years, 48% male) across four centres with history of isolated arch repair. Aortic arch measurements were correlated with % predicted $VO2_{max}$ with subgroup analyses of those with residual arch obstruction, bicuspid aortic valve, and hypertension. Ascending aorta (AAo) to DAo diameter ratio (D_{AAo}/D_{DAo}) was negatively correlated with % predicted $VO2_{max}$. %DAo flow positively correlated with $VO2_{max}$. Sub-analyses demonstrated that the negative correlation of D_{AAo}/D_{DAo} with $VO2_{max}$ was maintained only in patients without arch obstruction and with a bicuspid aortic valve. Smaller aortic arch measurements were associated with both hypertension and exercise-induced hypertension.

Conclusion

Aorta size mismatch, due to AAo dilation or small DAo, and associated decreased %DAo flow, correlated significantly with decreased exercise capacity after aortic arch repair. These correlations were stronger in patients without arch obstruction and with a bicuspid aortic valve. Aorta size mismatch and %DAo flow capture multiple mechanisms of altered haemodynamics beyond blood pressure gradient or discrete obstruction and can inform the definition of a successful repair.

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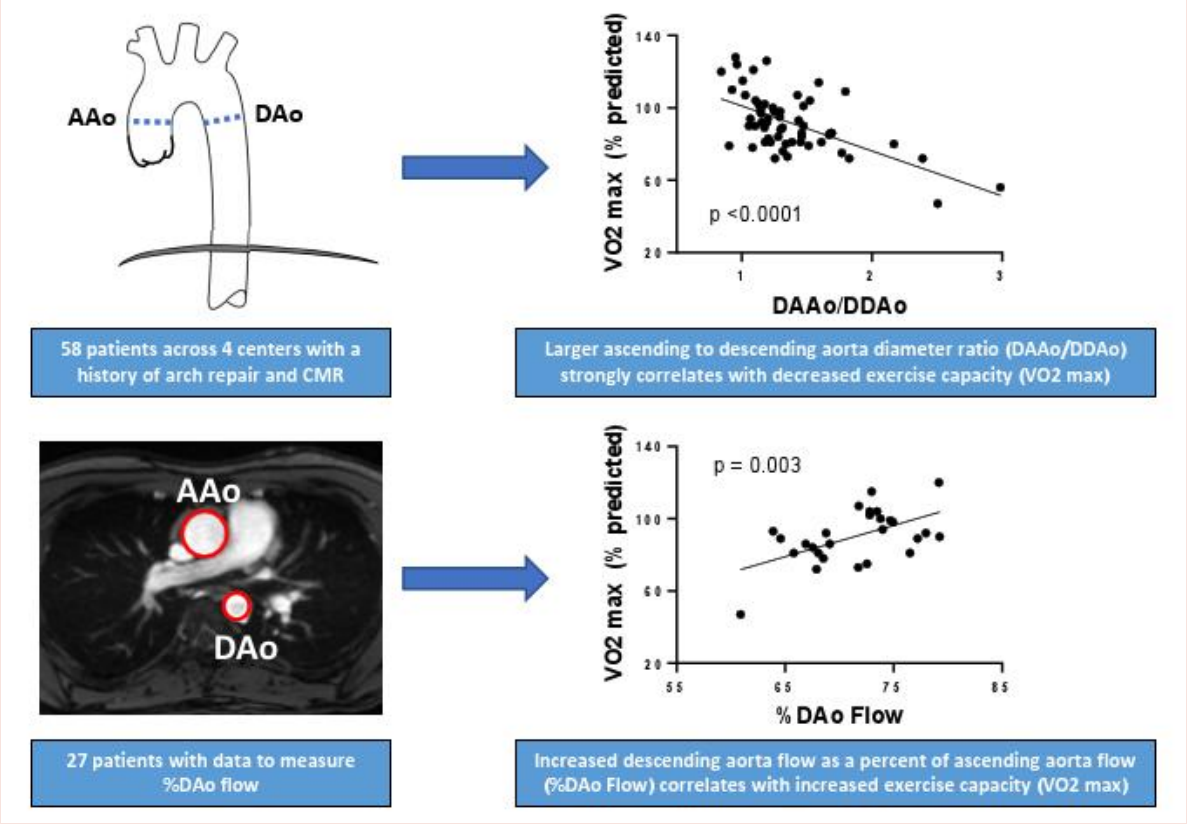
Graphical Abstract



Fifty-eight patients across four centres with a history of isolated arch repair were analysed. Ascending aorta (AAo) and descending aorta (DAo) diameter was measured from cardiac magnetic resonance, and the ratio (D_{AAo}/D_{DAo}) was calculated. Peak exercise capacity (% predicted VO2 max) ranged from severely decreased to normal. D_{AAo}/D_{DAo} negatively correlated with exercise capacity (% predicted VO2 max). Twenty-seven patients had sufficient data to calculate %DAo flow (DAo flow/AAo flow). %DAo flow positively correlated with % predicted VO2 max.

Keywords CMR imaging • Coarctation • Exercise • Congenital heart disease

Introduction

Coarctation of the aorta (CoA) is defined by a discrete stenosis of the aortic isthmus, and may be associated with aortic arch hypoplasia, left-sided obstructive lesions, and frequently, a bicuspid aortic valve (BAV).[1] Current treatment options typically result in excellent early and mid-term survival with overall re-intervention rates reported to be 16–31% depending on technique, age and size at repair, and surgical era.[2,3] However, there remains significant long-term morbidity[4] including hypertension,[3,5] decreased exercise capacity,[6–8] and reduced long-term survival.[3] Current definitions of successful arch repair are based on relief of pressure gradient,[9,10] however persistent flow abnormalities in the absence of a residual pressure gradient are apparent, which contribute to long-term outcomes.[8,11] In the absence of discrete re-coarctation, indications for re-intervention remain unclear.

Exercise intolerance in congenital heart disease is associated with increased risk of hospitalization and death.[12] The haemodynamic mechanism of exercise intolerance after CoA repair is not completely understood. Our previous single-centre study exploring the connection between exercise capacity and post-operative arch shape demonstrated strong associations between arch geometry, flow characteristics, and exercise capacity in a population with 'successfully' repaired CoA. Specifically, increased aorta size mismatch, defined by the ascending-to-descending aorta diameter ratio (D_{AAo}/D_{DAo}), and decreased descending aorta (DAo) flow correlated significantly with decreased exercise capacity in the absence of discrete recoarctation. An underlying mechanism of flow maldistribution with exercise was suggested by further *in vitro* studies.[7] Subsequent 4D flow analysis identified important changes in secondary flow characteristics suggesting that aorta size mismatch contributed to increased turbulence and inefficient flow.[8]

Exercise capacity is an important clinical indicator of patients at increased risk of hospitalization and death in the adult congenital heart disease population, including repaired CoA.[12] Given the wide range of outcomes in patients with repaired CoA, even in the absence of a residual pressure gradient, it is important to understand optimal arch shape to more clearly define goals of intervention. This multicentre study seeks to evaluate the relationship between aorta shape and exercise capacity in repaired CoA patients, specifically through the lens of aortic size mismatch and DAo flow and their association with exercise.

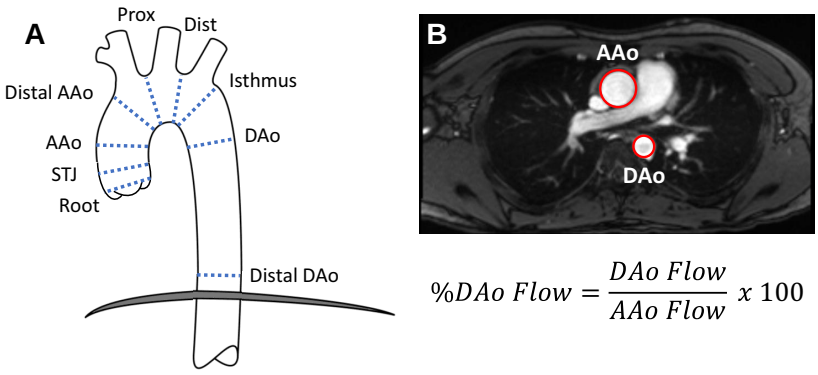


Figure 1 (*A*) Location of measurements of the aorta at the root, sinotubular junction (STJ), ascending aorta (AAo), distal AAo measured proximal to the origin of the innominate artery, proximal transverse arch (prox), distal transverse arch (dist), isthmus, proximal descending aorta (DAo), and distal DAo at the level of the diaphragm. (*B*) Axial plane from phase contrast cardiac magnetic resonance imaging at the level of the right pulmonary artery demonstrating location of flow measurements at the AAo and DAo. %DAo flow was defined by DAo flow/AAo flow.

Methods

Subjects

This retrospective study was approved for secondary data use with waiver of consent by the Institutional Review Board of each participating institution, including Children's National Hospital (CNH), Boston Children's Hospital (BCH), Advocate Children's Hospital (ACH), and University of Chicago Medical Center (UCMC). Patients were identified based on review of local imaging databases with review of the chart to identify inclusion/exclusion criteria. Patients with a history of aortic arch repair with cardiac magnetic resonance (CMR) study and cardiopulmonary exercise testing (CPET) that measured peak oxygen consumption ($VO2_{max}$) within one year of each other were included. Subjects were excluded from the study group with submaximal effort [respiratory exchange ratio (RER) < 1.09], more than mild valve dysfunction, more than mild outflow tract obstruction, or need for cardiac bypass outside of the arch repair. Thus, patients with a repaired ventricular or atrial septal defect were excluded. Bicuspid aortic valve and residual arch obstruction were not excluded.

Cardiopulmonary exercise testing

Data from CPET done closest to the CMR study were recorded, including equipment (treadmill or cycle ergometer), protocol (ramp or Bruce), $VO2_{max}$, ventilatory anaerobic threshold (VAT), RER, as well as baseline and peak heart rates and blood pressures. Exercise capacity was defined as decreased if $VO2_{max}$ was <90% predicted by age and sex.[6] Arch obstruction was defined by a baseline blood pressure gradient between arm and leg systolic cuff pressure ≥ 20 mmHg. Exercise-induced hypertension was defined by peak systolic arm blood pressure ≥ 200 mmHg.

Echocardiogram data

Clinically reported data from the echocardiogram closest to CPET were recorded, including left ventricular ejection fraction, aortic valve morphology, residual arch obstruction (peak velocity ≥ 2.2 m/s over the arch), valvar insufficiency and/or stenosis, and intracardiac shunt. As there were no patients with significant left ventricular outflow tract obstruction, DAo velocity was not corrected for proximal obstruction. Given imaging limitations, echocardiograms were not used to assess aorta diameters.

Cardiac magnetic resonance data

Ejection fraction by CMR was obtained from the clinical report. Any reported valvar stenosis or insufficiency, or intracardiac shunt was also noted. Aortic diameter and flow measurements were performed by a single investigator at the core lab (JGM) using offline analysis software (Arterys, San Francisco, CA, USA). Two-dimensional measurements of the aortic arch were done on the best quality 3D dataset available (ungated magnetic resonance angiogram or 3D steady-state free precession). Measurements were made at the aortic root, sinotubular junction (STJ), ascending aorta (AAo) at the level of the right pulmonary artery, distal AAo measured proximal to the origin of the innominate artery, proximal transverse arch, distal transverse arch, isthmus, proximal DAo, and distal DAo using standard double oblique technique[13] (*Figure 1A*). Diameters were normalized to body surface area. D_{AAo}/D_{DAo} was calculated as the ratio of AAo (at the level of the right pulmonary artery) to proximal DAo diameter.[7,8] Ascending aorta dilation was defined by CMR *z*-score[13] ≥ 2.0 for patients ≤ 20 years of age and a cut-off of 34 mm for males, 33 mm for females for patients > 20 years of age.[14] Where available, flow was measured by 2D phase contrast in the mid-AAo and proximal DAo at the level of the right pulmonary artery (*Figure 1B*). %DAo flow was defined by DAo flow/AAo flow.

Statistical analysis

All statistical analyses were performed using Prism 8 (Graphpad, San Diego, CA, USA). An unpaired two-tailed *t*-test or Fisher's exact test was used to evaluate differences between groups with normal and decreased exercise capacity, with and without hypertension treatment, and with and without exercise-induced hypertension. All correlations were performed using Pearson's correlation coefficient (*r*) including each aorta measurement with % predicted $VO2_{max}$, aorta size mismatch with VAT/predicted VO2 max, and %DAo flow with $VO2_{max}$.

Results

Fifty-eight patients (age 28 ± 10 years, 48% male) from four centres (*Figure 2*) were included in the study, including 57 CoA and one interrupted arch repair. Multiple types of primary repair were represented in this cohort, including end-to-end anastomosis ($n = 30$), subclavian flap repair ($n = 11$), long segment patch ($n = 4$), stent angioplasty ($n = 3$), balloon angioplasty ($n = 4$), interposition graft ($n = 4$), and two were unknown. Fifteen patients required secondary repair (12 interventional, 3 operative) and four patients required a tertiary repair (3 interventional, 1 operative). The overall mean % predicted VO2 max was normal at 92% with 30 (54%) having normal exercise capacity and 28 (46%) having decreased exercise capacity. Notably, 29% of the cohort had an excellent exercise capacity with a predicted VO2 max of 100% or greater. Overall absolute VO2 max ranged from 13.8 to 57.8 mL/kg/min with a mean of 32.8 mL/kg/min. A histogram of the distribution of exercise capacity is shown in *Figure 3*. Comparison between the two groups of normal and

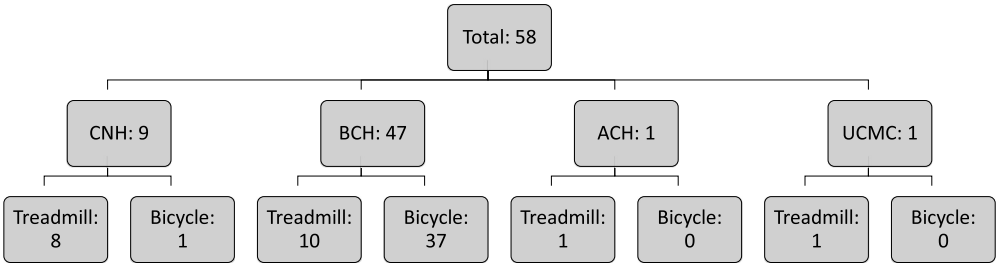


Figure 2 Distribution of patients from each of the four centres: Children's National Hospital (CNH), Boston Children's Hospital (BCH), Advocate Children's Hospital (ACH), and University of Chicago Medical Center (UCMC), and exercise equipment used.

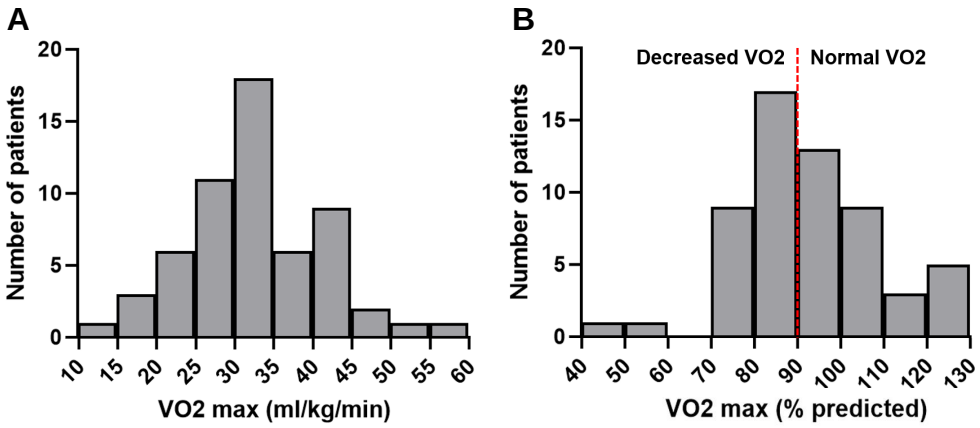


Figure 3 (*A*) Histogram of absolute peak oxygen consumption (VO2 max) among the cohort. (*B*) Histogram of % predicted VO2 max. The dashed line at 90% predicted VO2 max separates the groups of decreased capacity ($n = 28$) and normal exercise capacity ($n = 30$).

Table 1 Comparison of subjects with normal exercise capacity to those with decreased exercise capacity

	Normal VO2 (≥90% pred.) ($n = 30$)	Decreased VO2 (<90% pred.) ($n = 28$)	*P* value
Mean VO2 max % predicted (median)	**104 ± 11% (102)**	**79 ± 9% (81)**	**<0.0001**
Age at CMR, years	28 ± 12	27 ± 8	0.96
Male sex	37%	61%	0.11
Body mass index, kg/m^2	24 ± 3.1	25 ± 4.5	0.96
Diagnosis of hypertension	37%	64%	0.07
Hypertension medication (beta-blocker)	**23%** (13%)	**57%** (21%)	**0.02** (0.50)
Age at primary repair (range)	5 ± 7.4 (0–30)	7 ± 8.3 (0–35)	0.33
% requiring secondary repair	27%	25%	0.99
% requiring tertiary repair	3%	7%	0.34
Tricuspid aortic valve	37%	25%	0.40
LVEF (CMR)	65% ± 6%	61% ± 8%	0.08
LVEF (Echo)	65% ± 5%	64% ± 6%	0.63
Rest BP gradient ≥ 20 mmHg	14%	17%	1.0
Exercise BP gradient ≥ 20 mmHg	84%	90%	0.65
Arch obstruction (≥2.2 m/s by echo)	40%	36%	0.79

Bold values indicate statistical significance ($P < 0.05$). BP, blood pressure; CMR, cardiac magnetic resonance; Echo, echocardiogram; LVEF, left ventricular ejection fraction; VO2, peak oxygen consumption.

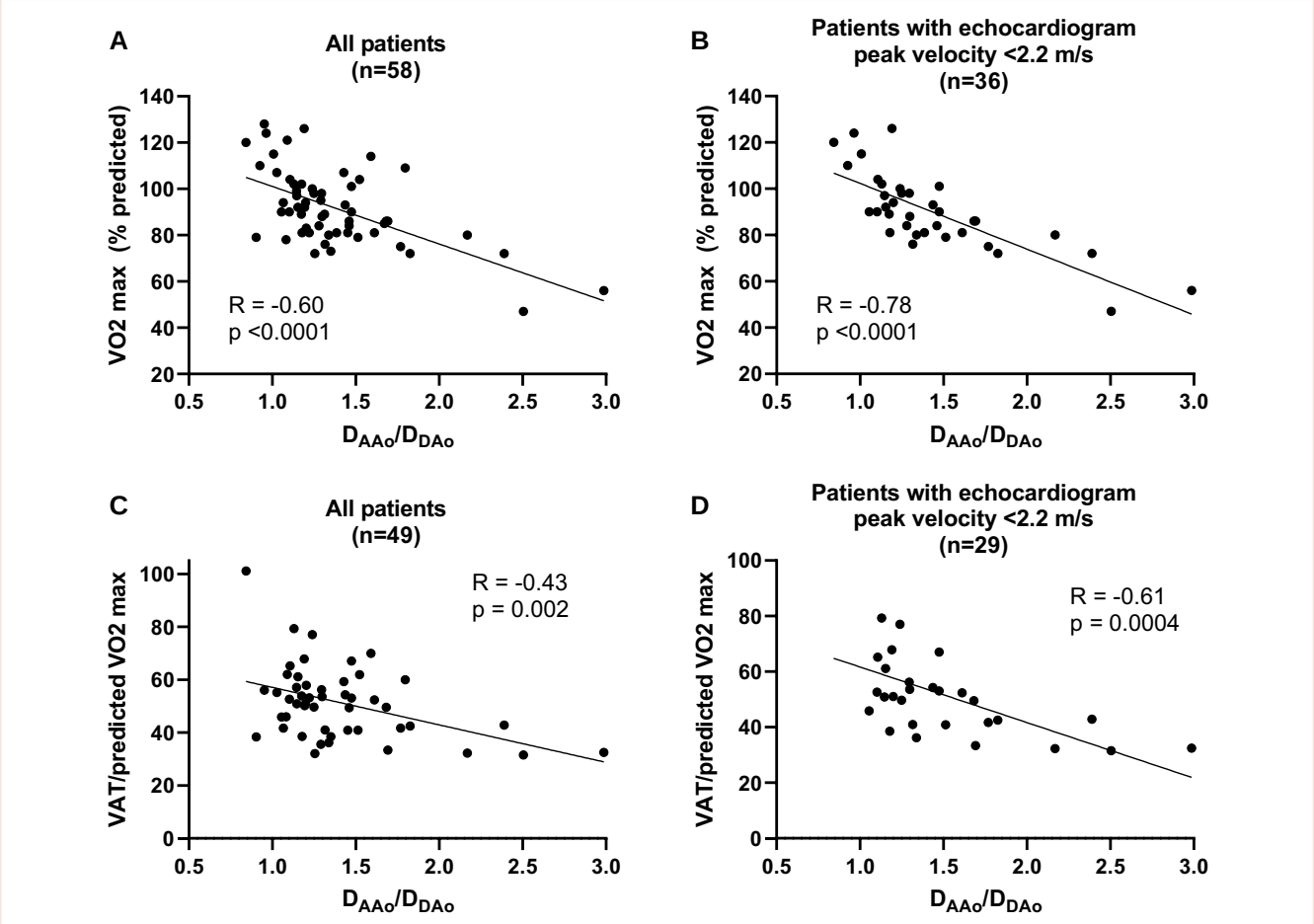


Figure 4 (*A*) Aorta size mismatch (D_{AAo}/D_{DAo}) negatively correlated with per cent predicted peak oxygen consumption (VO2 max). (*B*) A sub-analysis was performed on patients without evidence of arch obstruction by echocardiogram, defined by peak velocity < 2.2 m/s. The negative correlation of D_{AAo}/D_{DAo} and VO2 max was stronger in this group without arch obstruction. (*C*) D_{AAo}/D_{DAo} also negatively correlated with the ventilatory anaerobic threshold (VAT) as a percentage of predicted VO2 max. (*D*) Similar to peak exercise capacity, sub-analysis of patients without arch obstruction by echocardiogram demonstrated a stronger negative correlation between D_{AAo}/D_{DAo} and predicted VAT.

decreased exercise capacity demonstrated no difference in the prevalence of arch obstruction by cuff gradient or echocardiogram (*Table 1*). Additionally, there was a higher prevalence of hypertension in patients with decreased exercise capacity, though no difference in the use of beta-blockers between the two groups. There were no differences between males ($n = 28$) and females ($n = 30$) in age (average 26 vs. 29 years, $P = 0.19$), BMI (24 vs. 25, $P = 0.07$), indexed AAo diameter (15.7 vs. 15.8 mm/m^2, $P = 0.91$), indexed DAo diameter (11.9 vs. 11.9 mm/m^2, $P = 0.70$), or D_{AAo}/D_{DAo} (1.36 vs. 1.39 mm/m^2, $P = 0.16$). $VO2_{max}$ of the male group was an average of 89% vs. 93% in females that was significantly different ($P = 0.03$), though likely not clinically meaningful. Notably, of the previously reported 15 patients, 8 were included in the current study, with the remainder excluded due to the more restrictive exclusion criteria of a need for cardiac bypass outside of arch repair.[7,8]

Correlation of exercise with arch anatomy and flow

Among the entire cohort, a larger D_{AAo}/D_{DAo} correlated with lower exercise capacity (% predicted VO2 max) in patients s/p arch repair. Additionally, DAo size was positively correlated with $VO2_{max}$ (*Figure 4A*). *Table 2* summarizes correlations of CMR aorta measurements with VO2 max. In a sub-analysis of patients without arch obstruction by echocardiogram ($n = 36$), measurements of the STJ, AAo, distal AAo, DAo, and D_{AAo}/D_{DAo} all correlated significantly with exercise capacity (*Table 2*, *Figure 4B*). D_{AAo}/D_{DAo} also negatively correlated with the VAT as a percentage of predicted $VO2_{max}$ for all patients (*Figure 4C*) and in patients without arch obstruction (*Figure 4D*). For patients with residual arch obstruction by echocardiogram, there was no significant correlation between D_{AAo}/D_{DAo} and $VO2_{max}$ or predicted VAT. An analysis was performed excluding the previously published patients (remaining $n = 50$) demonstrating a significant correlation of D_{AAo}/D_{DAo} and VO2 max ($R = 0.51$, $P < 0.001$). There were 27 patients with sufficient data to measure %DAo flow that had a positive correlation with exercise capacity, i.e. more flow to the DAo at rest is associated with better exercise capacity (*Figure 5*). %DAo also correlated with D_{AAo}/D_{DAo} ($R = 0.61$, $P < 0.001$). There were insufficient data to reliably perform a sub-analysis based on arch obstruction for %DAo flow. Re-analysis of the correlation of %DAo flow and $VO2_{max}$ excluding previously published patients was no longer significant ($R = 0.24$, $P = 0.32$),

Table 2 Correlation of indexed aorta measurements by cardiac magnetic resonance with per cent predicted VO2 max including sub-analysis based on arch obstruction defined by echocardiogram

	All ($n = 58$)		V < 2.2 m/s ($n = 36$)		V ≥ 2.2 m/s ($n = 22$)	
Diameter/BSA	***r***	***P* value**	***r***	***P* value**	***r***	***P* value**
Root	−0.19	0.29	−0.33	0.14	−0.05	0.89
STJ	−0.11	0.40	**−0.42**	**0.01**	0.33	0.14
AAo	**−0.26**	**0.05**	**−0.61**	**<0.0001**	0.19	0.40
Distal AAo	−0.04	0.76	**−0.37**	**0.026**	0.30	0.17
Proximal arch	−0.06	0.70	−0.07	0.74	−0.29	0.30
Distal arch	0.08	0.56	−0.01	0.95	0.10	0.67
Isthmus	0.03	0.83	0.09	0.61	−0.17	0.45
DAo	**0.42**	**0.0009**	**0.46**	**0.0047**	0.25	0.26
Distal DAo	0.25	0.06	0.23	0.18	0.05	0.83
D_{AAo}/D_{DAo}	**−0.60**	**<0.0001**	**−0.78**	**<0.0001**	−0.01	0.98

Bold values indicate statistical significance ($P < 0.05$). AAo, ascending aorta; BSA, body surface area (m^2); DAo, descending aorta; D_{AAo}/D_{DAo}, aorta size mismatch; *r*, Pearson's correlation coefficient; STJ, sinotubular junction; V, velocity be echocardiogram.

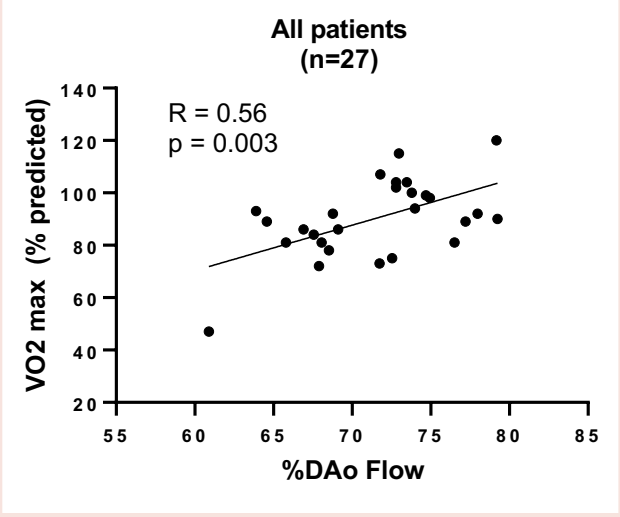


Figure 5 Per cent descending aorta (%DAo) flow positively correlated with per cent predicted peak oxygen consumption (VO2 max), i.e. more flow to the descending aorta at rest is associated with better exercise capacity.

though only 20 patients remained making this an even smaller subset of the total study cohort.

Effect of aortic valve morphology

Of the 58 total patients, 19 (33%) had a tricuspid aortic valve, 37 (64%) had a BAV, and 2 (3%) were defined as unicuspid. Patients with a unicuspid or BAV had a larger normalized AAo diameter compared to those with a normal tricuspid aortic valve (16.3 mm/m^2 vs. 14.4 mm/m^2, $P = 0.042$). There were 12 patients total with AAo dilation, 11 with a BAV and 1 with a tricuspid aortic valve. There was no difference in DAo diameter between the two groups (11.7 mm/m^2 vs. 12.0 mm/m^2, $P = 0.73$). Patients with a unicuspid or BAV maintained a significant negative correlation between $VO2_{max}$ and aorta size mismatch ($P < 0.0001$, *Figure 6*). Aorta size mismatch in patients with a tricuspid aortic valve was not significantly correlated with exercise capacity, however dilation of the ascending aorta was rare in the tricuspid aortic valve subgroup. Removing the 12 patients with AAo dilation, the correlation of D_{AAo}/D_{DAo} and $VO2_{max}$ remained significant ($R = 0.52$, $P < 0.001$). Those with AAo dilation had a lower $VO2_{max}$ compared to patients without AAo dilation (82% vs. 94%, $P = 0.018$). Despite differences in AAo size between patients with a BAV and tricuspid aortic valve, $VO2_{max}$ was not significantly different when classifying by aortic valve morphology alone (91% vs. 93% predicted, $P = 0.59$).

Hypertension and aortic arch geometry

Patients treated for hypertension had smaller measurements of the distal AAo, distal transverse arch, DAo, and distal DAo by CMR. Patients with exercise-induced hypertension had smaller measurements of the aortic root, STJ, AAo, distal AAo, and proximal transverse arch by CMR (*Table 3*).

Discussion

In this multicentre study in patients following aortic arch repair, we identified important aortic measurements that correlated with exercise capacity. Most significantly, a greater degree of aorta size mismatch—due to AAo dilation or a small DAo—correlated strongly with lower peak exercise capacity. The correlation with submaximal exercise capacity was maintained, corroborating the relationship with peak exercise.

Aorta narrowing, abnormal curvature, and vascular stiffness are known to contribute to increased left ventricular afterload and lead to diastolic dysfunction, effects that may be underestimated by imaging and blood pressure gradients.[11,15–19] Echocardiographic evidence of arch obstruction was present equally in both groups in this cohort: normal and decreased exercise capacity. Among patients without arch obstruction, a large AAo, small DAo, and thus larger D_{AAo}/D_{DAo}, correlated with decreased exercise capacity. This relationship was lost when assessing only patients with arch obstruction. This suggests that the mechanism of exercise intolerance in this population is multifactorial, and that in patients without significant arch obstruction, aorta diameter remains an important contributor. Further, the relationship between aorta size mismatch and exercise capacity remains significant even when removing patients with AAo dilation. Aorta size mismatch

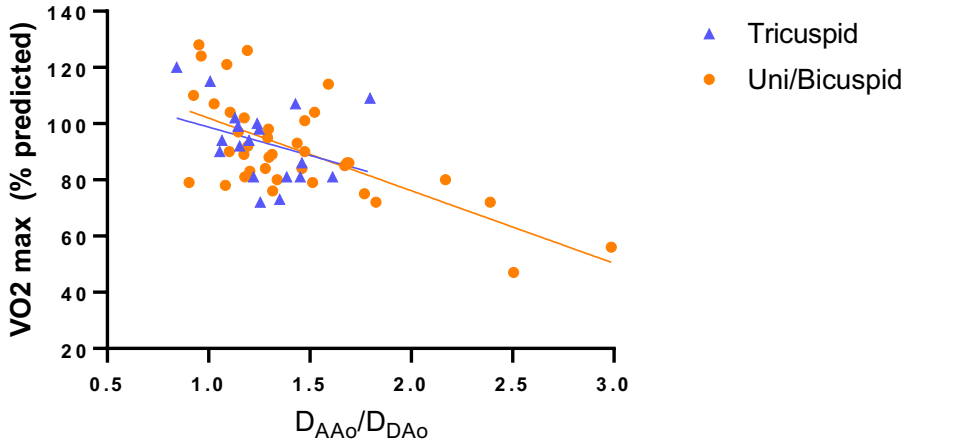


Figure 6 The negative correlation of aorta size mismatch (D_{AAo}/D_{DAo}) with exercise capacity (VO2 max) is maintained in patients with a unicuspid or bicuspid aortic valve ($r = -0.66$, $P < 0.0001$). The correlation is not significant when assessing only patients with a tricuspid aortic valve ($r = -0.33$, $P = 0.16$).

Table 3 Comparison of indexed aorta measurements by cardiac magnetic resonance between patients with and without hypertension treatment and with and without exercise-induced hypertension

	HTN treatment			Exercise-induced HTN		
Diameter/BSA (mean mm/m²)	**Yes**	**No**	***P* value**	**Yes**	**No**	***P* value**
Root	16.0	17.2	0.28	**15.1**	**17.4**	**0.028**
STJ	13.9	15.3	0.09	**13.5**	**15.4**	**0.024**
AAo	15.1	16.2	0.24	**14.4**	**16.4**	**0.033**
Distal AAo	**12.4**	**13.9**	**0.03**	**12.3**	**13.9**	**0.03**
Proximal arch	10.7	11.8	0.09	**10.5**	**12.0**	**0.031**
Distal arch	**8.7**	**10.6**	**<0.001**	9.2	10.3	0.08
Isthmus	9.2	10.2	0.17	9.7	9.7	0.96
DAo	**10.8**	**12.6**	**0.016**	11.4	12.3	0.29
Distal DAo	**9.4**	**10.7**	**0.004**	9.7	10.5	0.10

Bold values indicate statistical significance ($P < 0.05$). AAo, ascending aorta; BSA, body surface area (m^2); DAo, descending aorta; HTN, hypertension; STJ, sinotubular junction.

captures important haemodynamic effects of size discrepancy, even in the absence of significant AAo dilation.

Consistent with previous studies,[20,21] analyses of patients by valve morphology demonstrated the expected finding of a larger AAo in those with a BAV compared to a normal tricuspid aortic valve, and no difference in DAo size. As there is no follow-up data as part of this study, the degree of the progression of AAo dilation after arch repair is not known. We find that only those with a BAV maintained the negative correlation of D_{AAo}/D_{DAo} with exercise capacity. There were differences in the range of VO2 among the two groups, though it is likely that the degree of AAo dilation secondary to the vasculopathy of the BAV is an important haemodynamic mechanism not present in those with a normal aortic valve. Indeed, we found that patients with AAo dilation had lower exercise capacity compared to those without AAo dilation, even when there was no significant difference in exercise capacity by valve morphology. This suggests that the vasculopathy of a BAV is variable among patients leading to variability in the impact on exercise haemodynamics. Recent work found that patients with CoA and a BAV had a stiffer AAo compared to those with a tricuspid aortic valve as measured by distensibility.[22] This suggests that the aortopathy of the AAo with a BAV reduces the Windkessel effect, which describes the ability for the aorta to store energy in systole that is then released in diastole.[23] Therefore, it may be decreased distensibility and subsequent energy loss driving the relationship of aorta size mismatch and exercise capacity in patients with a BAV.

We additionally found that lower %DAo flow at rest correlated with both decreased exercise capacity and D_{AAo}/D_{DAo}. This is in agreement with our previous study[7] that also demonstrated changes in flow distribution with simulated exercise in an *in vitro* benchtop flow pump model. The altered flow distribution at rest may indicate that aorta size mismatch causes increased resistance to flow. %DAo flow may be a more reflective measurement of arch resistance and subsequent decreased exercise capacity.

Finally, our study showed that smaller aorta measurements, most significantly the distal transverse arch, impacted the development of hypertension. This finding is in line with previous reports suggesting that residual transverse arch hypoplasia is associated with an increased long-term risk of hypertension.[24] This further suggests that in the setting of transverse arch hypoplasia, more aggressive arch reconstruction during primary repair may impart benefits in exercise capacity long-term. Interestingly, exercise-induced hypertension was associated more with smaller proximal aorta measurements (root to proximal arch), and not distal measurements. Further study is needed to understand the mechanism of this finding.

This study is subject to the limitations inherent in a retrospective analysis. Our cohort was diverse with multiple types of repair and re-intervention occurring at different ages. The overall re-intervention rate in our study was slightly high (26%), typical of an imaging cohort, and overall similar to previous reports.[2,3] Only one patient had an interrupted aortic arch that was repaired with an interposition graft. This patient had normal exercise capacity of 104% and a D_{AAo}/D_{DAo} of 1.5, close to the study average of 1.4, suggesting that this patient was not an outlier. Additionally, both bicycle (66%) and treadmill (34%) ergometers were used with a known difference that maximum oxygen uptake is typically

higher with treadmills.[25] All exercise protocols were performed according to the clinical practice of the institution. Imaging was also performed according to clinical standards at each institution. The decision was made to use only flow measurements in the AAo and DAo at the level of the right pulmonary artery. This was done to include the most patients while maintaining a consistent technique. Finally, this study compared multiple measurements of the aortic arch, introducing the possibility of a Type I error. However, the very small *P* values found on most comparisons suggest that the chance of false positive results remains low. Future work should be done as a prospective study to standardize CPET and CMR techniques and include 4D flow CMR to explore important flow dynamics. Further, as discussed above, analysis of vascular stiffness would be additive as vascular function likely plays an important role in the haemodynamics of exercise.

Conclusion

Aorta size mismatch, due to AAo dilation or small DAo, and associated decreased %DAo flow, correlated significantly with decreased exercise capacity after aortic arch repair. These correlations were stronger in patients without residual arch obstruction and with a BAV. This study indicates that aorta size mismatch and %DAo flow are important measures for long-term outcomes, capturing multiple mechanisms of altered haemodynamics beyond blood pressure gradient or discrete obstruction. Cardiac magnetic resonance measurements of aorta geometry and flow can inform the definition of a successful repair beyond discrete recoarctation.

Lead author biography

Jason Mandell is an assistant professor of paediatric cardiology at the University of Rochester Medical Center in Rochester, NY. He completed paediatric cardiology and advanced cardiac imaging fellowships at Children's National Hospital in Washington, DC. His research interests focus on the use of advanced cardiac imaging to understand haemodynamic patterns to predict and improve outcomes in congenital heart disease.

Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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