Anatomical risk factors, surgical treatment, and clinical outcomes of left-sided pulmonary vein obstruction in single-ventricle patients

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ABSTRACT

Background: Patients with single-ventricle physiology frequently develop left-sided pulmonary vein obstruction (PVO), in which the pulmonary veins traverse the descending thoracic aorta. We hypothesized that a combination of cardiomegaly and an anteriorly positioned descending aorta is associated with PVO.

Methods: Among 494 consecutive single-ventricle patients, 15 were diagnosed with PVO by cardiac magnetic resonance, defined as anatomically localized narrowing of the pulmonary vein diameter. Using axial slices at the level of the left lower pulmonary vein, normalized dimensions were obtained to characterize the anatomic relationships of intrathoracic structures. Measurements were compared between patients with PVO and “control” patients (single-ventricle patients with normal pulmonary veins, n = 12).

Results: Patients with cardiac magnetic resonance–diagnosed PVO had larger cardiac size and more antero-laterally located descending aorta when compared with controls (normalized dimensions: cardiac/thoracic area ratio: 0.43 vs 0.38, $P = .035$, distance from vertebra to descending aorta normalized by the horizontal dimension of thoracic cavity: 0.09 vs 0.08, $P = .049$). Seven (47%) patients underwent PV sutureless repair, and 3 (of 7) failed to achieve Fontan. Patients who failed to achieve Fontan had a larger normalized cardiac size than those who achieved Fontan (cardiac/thoracic area ratio: 0.49 vs 0.39, $P = .001$).

Conclusions: The combination of relative cardiomegaly within the context of the thoracic cavity at the level of the pulmonary veins and antero-lateral displacement of the aorta is associated with left-sided PVO and subsequent failure to achieve Fontan completion. Further characterization of these unique geometric relationships may help inform both surveillance strategies and decision making in the timing of interventions, and guide the intraoperative objectives at the time of PVO repair. (J Thorac Cardiovasc Surg 2015;149:1332-8)

Combination of cardiomegaly and antero-laterally located descending aorta is a risk for left PVO.

Central Message

Characterization of both anatomic and physiologic feature of left PVO helps decision making in the timing and type of repair.

Perspective

In comparison with biventricular repair, the clinical significance of PVO is magnified in patients with single-ventricle physiology because PVO may prevent progression to second stage and Fontan procedures. This study showed that combination of cardiomegaly and antero-laterally displaced descending aorta was associated with left PVO and subsequent failure to achieve Fontan completion; hence, further characterization of this unique condition is important.

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Previous studies have noted that left-sided pulmonary vein obstruction (PVO) commonly occurs in close proximity to the descending thoracic aorta.¹⁻³ In comparison with biventricular repair, the clinical significance of PVO is magnified in patients with single-ventricle physiology

because PVO may prevent progression to second stage and Fontan procedures. Indeed, atrioventricular valve regurgitation (AVVR), which is commonly associated with single-ventricle volume loading, and PVO remain important risk factors for Fontan failure.^{4,5} We hypothesized that a combination of cardiomegaly and an anteriorly displaced descending aorta predisposes patients to left-sided PVO in the region where the pulmonary veins traverse the descending aorta (Figure 1). To test this hypothesis, we developed geometric models of intrathoracic anatomy using cardiac magnetic resonance (CMR) imaging obtained in patients with and without evidence of PVO. Furthermore, we correlated these anatomic models with progression to Fontan completion. Finally, we evaluated clinical outcomes in terms of Fontan candidacy of this unique pathology.

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Abbreviations and Acronyms

AVVR = atrioventricular valve regurgitation
 CMR = cardiac magnetic resonance
 IQR = interquartile range
 PVO = pulmonary vein obstruction

METHODS

A total of 494 single-ventricle patients who underwent staged Fontan palliation at The Hospital for Sick Children in Toronto between January 2000 and June 2012 were reviewed. Patients with total anomalous pulmonary venous connection were excluded. Twenty-four (4.9%) patients were diagnosed with left-sided PVO. Among the 24 single-ventricle patients with PVO, 15 had left-sided PVO, defined as anatomically localized narrowing of the PV diameter by CMR, and were included as the study group (Table 1); the remaining 9 patients were diagnosed on the basis of echocardiograms (ie, no CMR) and were excluded because the planned anatomic comparisons require CMR studies.

A control group was selected among single-ventricle patients who did not have a diagnosis of left-sided PVO and did have CMR studies available for comparison. Matching criteria included age, ventricular morphology, type of initial palliation, and stage of palliation. Twelve patients were selected for the control group based on matching criteria and the availability of CMR for comparison. Approval for the study was granted by The Hospital for Sick Children Research Ethics Board, which waived any requirement for patient consent.

Diagnostic Studies

The CMR protocol was performed on a 1.5-Tesla magnet (Avanto, Siemens Medical Solutions, Erlangen, Germany) and consisted of phase contrast flow velocity measurements of all great arteries and veins, as described elsewhere.⁶ Using an axial slice at the level of the left lower pulmonary vein, normalized dimensions were obtained to characterize the anatomic relationships of intrathoracic structures. The slice was selected to maximize the diameter of the most stenotic portion of the pulmonary veins in PVO patients and to go through the midpoint of the left pulmonary veins in the control patients (Figure 2). If the assessments of the 2 investigators differed, the image was reviewed until consensus was reached. Raw measurements were normalized using vertical and horizontal thoracic dimensions. The pulmonary to systemic flow ratio (Qp/Qs) was calculated by using CMR, as follows:

$$\frac{\text{Right pulmonary veins flow } \left(\frac{\text{L}}{\text{min}}\right) + \text{Left pulmonary veins flow } \left(\frac{\text{L}}{\text{min}}\right)}{\text{Descending aortic flow } \left(\frac{\text{L}}{\text{min}}\right) + \text{Superior vena cava flow } \left(\frac{\text{L}}{\text{min}}\right)}$$

The cardi thoracic ratio at the time of PVO was assessed by chest radiograph. Severity of AVVR was obtained from the echocardiographic assessment close to the time that left-sided PVO was diagnosed (in the study group) or that CMR was performed (in the control group). More than mild AVVR was considered to be significant AVVR. Patients who did not achieve Fontan completion or who required heart transplantation were categorized as being in the “failure group.”

Surgical Technique

Seven (47%) patients underwent pulmonary vein sutureless repair. A detailed surgical description has been provided elsewhere.⁷ In brief, moderate hypothermic cardiopulmonary bypass was used. The posterior pericardium and the anterior wall of the left pulmonary veins were opened and unroofed. The anterior wall of the individual pulmonary veins were cut back and unroofed if necessary. The anastomosis between the atrium

and the posterior pericardium was performed with a continuous suture technique, using 6-0 or 7-0 polypropylene sutures (Prolene, Ethicon, Inc, Somerville, NJ).

Statistical Analysis

Continuous data are presented as median (interquartile range [IQR]). Discrete data are presented as frequency (percentage). The level of statistical significance was set at $P \leq .05$. Differences between the groups were analyzed with the Mann-Whitney U test. Event frequencies were compared with χ^2 analysis.

RESULTS

Among patients with left-sided PVO, the left lower pulmonary vein was the most frequent site of obstruction in 13 of 15 patients (87%) (Table 1). PVO was diagnosed between stage I and II in 5 patients, between stage II and III in 9 patients, and after Fontan completion in 1 patient. Seven (47%) patients had corroborative echocardiographic evidence of physiologic PVO (flow acceleration with mean gradients of >3 mm Hg). Eight patients did not have any evidence of PVO on echocardiography, defined as a mean gradient of >3 mm Hg across the pulmonary vein. Seven patients (47%) with PVO had significant AVVR, and 2 patients (17%) in the control group had significant AVVR ($P = .10$).

Anatomic and Physiologic Relationship of Intrathoracic Structures

The CMR-derived intrathoracic dimensions are shown in Table 2. The normalized distance from vertebra to aorta in patients with left-sided PVO was greater than that in control patients (0.09, IQR [0.08-0.10] vs 0.08, IQR [0.07-0.09], $P = .049$). Patients with left-sided PVO had a larger cardiac cross-sectional area normalized to total thoracic cross-sectional area than control patients (cardiac/thoracic: 0.43, IQR [0.39-0.49] vs 0.38, IQR [0.36-0.38], $P = .035$). This difference was accentuated in the comparison of the portion of the cardiac area in the left portion of the thorax (left cardiac/left thoracic: 0.60, IQR [0.52-0.64] vs 0.52, IQR [0.47-0.54], $P = .014$). The comparison of physiologic data showed a non-statistically significant trend toward less left-pulmonary vein flow in patients with left-sided PVO compared with controls (1.02 vs 1.57 l/min/m², $P = .061$) (Table 3). In contrast, patients with left-sided PVO had larger right-pulmonary vein flows (2.25 vs 1.47 l/min/m², $P = .025$); therefore, the left-to-right PV flow ratio was lower in PVO patients compared with controls (0.52 vs 1.05 l/min/m², $P = .028$), indicating an unequal distribution of pulmonary blood flow. The ratio of pulmonary to systemic flow (Qp/Qs) was comparable between patients with left-sided PVO and those in the control group (0.97 vs 0.96, $P = .748$), reflecting compensatory flow in the right lung offsetting diminished flow in the left lung of PVO patients. The cardi thoracic ratio was comparable between the left-sided PVO and control groups (0.54 vs 0.54, $P = .111$) (Table 3).

FIGURE 1. Hypothesis of anatomic factor contributing left-sided pulmonary vein obstruction. Arrow shows narrowed left pulmonary vein between the heart and descending aorta.

Clinical Outcomes

After surgical repair of PVO using the sutureless technique, the confluence of the left pulmonary veins with the left atrium was shifted laterally with elimination of PVO

in 4 patients (Figure 3, A and B). Three patients, however, did not have improved PV augmentation with the sutureless technique and had residual PVO, determined by echocardiography.

Two patients died after left-sided PVO was diagnosed (Table 1). Patient no. 1 had a sutureless repair and was waiting for a Fontan operation, without any evidence of recurrent PVO or pulmonary hypertension, and subsequently died of mechanical valve thrombosis and cardiac arrest. Another patient (no. 9) had a sutureless repair at the time of a bidirectional cavopulmonary shunt procedure but developed low cardiac output and desaturation, owing to increased pulmonary artery pressure, resulting in multiorgan failure and subsequent death. Two patients had significant AVVR and ventricular dysfunction and required heart transplantation. In total, 7 (46%) patients with left-sided PVO were considered to have failure of single-ventricle physiology, or failure to achieve Fontan completion, owing to pulmonary hypertension (n = 4), need for heart transplantation (n = 2), or protein-losing enteropathy (n = 1); (failure group n = 7) (Table 1). Eight (54%) of 15 patients had no evidence of pulmonary hypertension and achieved or

TABLE 1. Clinical summary for patients with left-sided PVO

No.	Diagnosis	Timing of PVO diagnosis	Affected PV	PVO diagnosed by echocardiography?	Catheterization		Echocardiography			Reason for death	Fontan completion/candidate?	Reason for failure to achieve Fontan completion
					PAP (mm Hg)	PVR (Unit/m ²)	AVVR	Ventricular function	Last status			
1	TA, TGA	s/p BCPS	LL	No	15	NA	2	0	Dead	Mechanical valve thrombosis	Yes	Mechanical valve thrombosis
2	TA, TGA	s/p Fontan	LL	No	12	NA	1	0	Alive	—	Yes	—
3	DORV	s/p BCPS	LU, LL	Yes	9	3.13	2	0	Alive	—	Yes	—
4	TA	s/p BCPS	LU	Yes	21	6.70	0	0	Alive	—	No	Pulmonary hypertension
5	DORV	s/p BCPS	LU, LL	No	10	NA	2	0	Alive	—	Yes	—
6	DORV	s/p BCPS	LL, RL	No	9	NA	1	1	Alive	—	Yes	—
7	HLHS	s/p Norwood	LL	No	22	NA	2	3	Alive	—	No	Heart transplantation
8	HLHS	s/p Norwood	LU, LL	No	22	NA	2	2	Alive	—	No	Heart transplantation
9	DIRV	s/p Norwood	LU, LL	Yes	23	NA	2	1	Dead	Multi-organ failure	No	Pulmonary hypertension
10	HLHS	s/p Norwood	LU, LL	Yes	24	NA	1	0	Alive	—	No	Pulmonary hypertension
11	DIRV	s/p BCPS	LU	No	10	2.17	0	1	Alive	—	Yes	—
12	HLHS	s/p BCPS	LU, LL	No	20	NA	1	0	Alive	—	No	Protein-losing enteropathy
13	DIRV	s/p BCPS	LU, LL	Yes	12	NA	2	0	Alive	—	Yes	—
14	TA, TGA	s/p BCPS	LL	Yes	9	2.13	1	0	Alive	—	Yes	—
15	HLHS	s/p Norwood	LU, LL	Yes	18	8.10	1	0	Alive	—	No	Pulmonary hypertension

For AVVR, 0 = none, 1 = mild, 2 = moderate, 3 = severe; for ventricular function, 0 = normal, 1 = mildly decreased; 2 = moderately decreased; 3 = severely decreased. AVVR, Atrioventricular valve regurgitation; PVO, pulmonary vein obstruction; PV, pulmonary vein; PAP, pulmonary artery pressure; PVR, pulmonary vascular resistance; TA, tricuspid atresia; TGA, transposition of the great arteries; s/p, post status; BCPS, bidirectional cavopulmonary shunt; LL, left lower; NA, not assessed; DORV, double-outlet right ventricle; LU, left upper; RL, right lower; HLHS, hypoplastic left-heart syndrome; DIRV, double-inlet right ventricle.

FIGURE 2. Measurement of intrathoracic structures by cardiac magnetic resonance. Axial slice at the level of left lower pulmonary vein was chosen from cardiac magnetic resonance. All measurement was normalized by thoracic dimension. Antero-posterior dimension: (posterior sternal plate to anterior margin of vertebra)/(posterior sternal plate to posterior margin of the lungs). Horizontal dimension: (maximum horizontal dimension of thoracic cavity)/(posterior sternal plate to posterior margin of the lungs). Distance from vertebra to aorta: distance from anterior margin of vertebra to center of descending aorta normalized by using vertical and horizontal dimension of thoracic cavity.

were candidates for Fontan completion (success group $n = 8$). In contrast, 11 of 12 (92%) patients in the control group subsequently had Fontan completion or are considered satisfactory candidates awaiting a Fontan operation.

Relationship Between Clinical Outcomes and Anatomic Characteristics

Among patients with left-sided PVO, intrathoracic dimensions were compared between patients who achieved or were awaiting Fontan completion ($n = 8$) and those who failed to achieve Fontan completion ($n = 7$). Normalized distance from the vertebra to the descending aorta did not differ between the groups (0.09 vs 0.09, $P = .215$). Indexed cardiac area and cardiac/thoracic ratio were significantly larger in the failure group (indexed cardiac area: 9.71 vs 5.88 mm²/m², $P = .009$; cardiac/thoracic ratio: 0.49 vs 0.39, $P = .001$). The failure group had a larger heart in the left-

TABLE 2. Anatomic measurement in cardiac magnetic resonance			
	PVO (N = 15)	Control (N = 12)	P value
Anterior-posterior	0.70 (0.62-0.74)	0.64 (0.63-0.67)	.263
Horizontal	1.44 (1.31-1.51)	1.40 (1.37-1.50)	.660
Vertebra to descending aorta	0.09 (0.08-0.10)	0.08 (0.07-0.09)	.049
Cardiac (mm ²)	6278 (5828-9037)	6666 (5669-7189)	.186
Lung (mm ²)	10,020 (8741-10,645)	10,804 (9378-12,431)	.127
Total thoracic	16,376 (15,692-20,047)	17,193 (15,441-19,454)	.865
Cardiac/thoracic	0.43 (0.39-0.49)	0.38 (0.36-0.38)	.035
Left cardiac (mm ²)	5127 (4163-5841)	4383 (3968-5014)	.112
Left thoracic (mm ²)	9134 (8014-10,107)	8647 (8201-9810)	.879
Left/right cardiac	2.42 (1.99-2.90)	2.25 (1.78-3.02)	.730
C3/cardiac	0.40 (0.38-0.42)	0.38 (0.37-0.44)	.921
C4/cardiac	0.30 (0.26-0.34)	0.30 (0.25-0.32)	.748
C3/left thoracic	0.50 (0.44-0.57)	0.42 (0.33-0.48)	.047
Left cardiac/left thoracic	0.60 (0.52-0.64)	0.52 (0.47-0.54)	.014
C4/C3	0.71 (0.63-0.84)	0.67 (0.60-0.90)	.961

All measurement was normalized by thoracic dimension. Antero-posterior dimension: (posterior sternal plate to anterior margin of vertebra)/(posterior sternal plate to posterior margin of the lungs). Horizontal dimension: (maximum horizontal dimension of thoracic cavity)/(posterior sternal plate to posterior margin of the lungs). Distance from vertebra to aorta: distance from anterior margin of vertebra to center of descending aorta normalized by using vertical and horizontal dimension of thoracic cavity. Cardiac: C1 + C2 + C3 + C4; Lung: L1 + L2; Total Thoracic: Cardiac + lung; Left Cardiac: C3 + C4; Left Thoracic: C3 + C4 + L2; Right Cardiac: C1 + C2. PVO, Pulmonary vein obstruction.

sided portion of the thoracic cavity (C3/left thoracic area: 0.58 vs 0.49, $P = .035$). Figure 4 shows the relationship between heart size, location of descending aorta, and clinical outcome. Patients with left-sided PVO had more cardiomegaly and antero-laterally displaced descending aortae compared with patients without PVO. Furthermore, cardiomegaly was associated with failed Fontan completion (Figure 4).

DISCUSSION

Pulmonary vein obstruction is associated with increased pulmonary vascular resistance and is considered an adverse prognostic factor with regard to proceeding to a Fontan procedure.^{4,5} Previous studies have reported an anatomic association between left-sided PVO, the descending aorta, and the left atrium.¹⁻³ Although left-sided PVO is often seen in patients with single-ventricle physiology in clinical practice, little study has been done of the anatomic/geometric substrate for PVO in the single-ventricle population.

Anatomic and Physiologic Characteristics in Patients With Left-Sided Pulmonary Vein Obstruction

Previous studies have reported pulmonary vein compression between the atrium and descending aorta.¹⁻³ Kawahira

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TABLE 3. Physiologic data

	PVO (N = 15)	Control (N = 12)	P value
Cardiac magnetic resonance			
LPA size (mm ² /m ²)	40.2 (32.3-40.7)	53.2 (50.2-53.2)	.153
LPA flow (l/min/m ²)	0.27 (0.18-0.40)	0.86 (0.71-1.06)	.003
LPVs flow (l/min/m ²)	1.02 (0.80-1.39)	1.57 (1.54-1.73)	.061
Left upper PV flow	0.60 (0.40-0.92)	0.61 (0.44-0.72)	.657
Left lower PV flow	0.60 (0.38-0.70)	0.87 (0.80-1.03)	.084
RPA size (mm ² /m ²)	47.9 (43.8-51.4)	42.3 (42.3-42.8)	.145
RPA flow (l/min/m ²)	1.76 (1.02-2.17)	1.21 (0.75-1.24)	.162
RPVs flow (l/min/m ²)	2.25 (1.73-2.43)	1.47 (1.25-1.86)	.025
Right upper PV flow	1.18 (0.93-1.37)	0.85 (0.67-1.17)	.228
Right lower PV flow	0.97 (0.86-1.17)	0.71 (0.62-1.02)	.058
LPA/RPA size ratio	0.78 (0.76-0.85)	1.26 (1.17-1.26)	.106
LPA/RPA flow ratio	0.17 (0.10-0.19)	0.59 (0.58-1.32)	.007
LPVs/RPVs flow ratio	0.52 (0.47-0.72)	1.05 (0.86-1.36)	.028
VEDVi (ml/m ²)	129 (97-179)	117 (112-134)	.276
VESVi (ml/m ²)	56 (40-98)	47 (42-71)	.152
Qp/Qs	0.97 (0.91-1.07)	0.96 (0.91-0.98)	.748
Catheterization			
PAP (mm Hg)	15 (10-21)	11 (9-13)	.048
PVR (WU/m ²)	3.13 (2.17-6.70)	2.32 (1.48-2.65)	.040
VEDP (mm Hg)	8 (6-11)	7 (5-9)	.416
Radiograph			
Cardiothoracic ratio	0.54 (0.52-0.58)	0.54 (0.51-0.55)	.111

PVO, Pulmonary vein obstruction; LPV, left pulmonary vein; PV, pulmonary vein; LPA, left pulmonary artery; RPA, right pulmonary artery; RPV, right pulmonary vein; VEDVi, ventricular end-diastolic volume index; VESVi, ventricular end-systolic volume index; Qp/Qs, pulmonary to systemic flow ratio; PAP, pulmonary artery pressure; PVR, pulmonary vascular resistance; VEDP, ventricular end-diastolic pressure.

and colleagues² reported that a combination of the pulmonary venous connection to the atrium with a morphologically right appendage, with the descending aorta and the heart on the same side of the chest, poses a risk of pulmonary vein compression. Otsuki and colleagues³ defined 3 types of PVO by using computed tomography, in which 1 type of PVO was the result of compression by surrounding organs. O'Donnell and colleagues¹ examined 1995 patients undergoing catheterization and identified 26 (1.3%) patients who had left aortic arch in association with left-sided PVO. In the present study, we used axial CMR to show that the combination of cardiomegaly and a laterally displaced descending aorta was associated with development of left-sided PVO.

Cardiac Magnetic Resonance and Echocardiography as Diagnostic Tools for Left-Sided PVO

In the current study, left-sided PVO was associated with intrapulmonary flow redistribution with a higher ratio of right/left pulmonary vein flows. Interestingly, only half of the patients with left-sided PVO diagnosed by CMR had flow acceleration at the site of PVO documented by echocardiography. These data suggest that flow redistribution away from the site of PVO may diminish the potential for echocardiography to detect hemodynamically significant left-sided PVO

if flow acceleration or gradients are considered critical to the echocardiographically defined diagnosis. These findings suggest that CMR has greater sensitivity to detect PVO. Despite the apparently greater sensitivity of CMR, echocardiography is more commonly used to screen patients for PVO.⁸

Another disadvantage of echocardiography includes the difficulty in determining the dimensions of the pulmonary veins within the lung parenchyma. Computed tomography and magnetic resonance imaging are helpful in determining the pulmonary vein dimensions within the lung parenchyma.³ CMR can provide both anatomic (dimensions) and physiologic (flow) information for individual pulmonary veins within the lung parenchyma.^{9,10} Thus, we believe that CMR is a useful diagnostic tool to assess pulmonary vein anatomy and physiology and may have greater sensitivity than echocardiography in making the diagnosis of left-sided PVO, with the additional advantage of providing better visualization of the dimensions of the pulmonary veins "upstream" from the site of obstruction. In light of the sensitivity of CMR to detect lesions outside the boundaries of echocardiography, it may be important to utilize CMR in more liberal fashion. We speculate that compression of the pulmonary veins by the heart may be an early event in the pathogenesis of increased pulmonary vascular resistance, subsequent single-ventricle dysfunction, and progression to Fontan failure in a subset of patients with anatomic predisposition to PVO.

Surgical Treatment for Left-Sided PVO

In this study, about half of patients with left-sided PVO underwent surgical treatment. All operations utilized the "sutureless technique" that has been used in our institution for total anomalous pulmonary vein connection or recurrent PVO.^{7,11,12} The unproven rationale for the use of the sutureless technique is to avoid the direct anastomosis between the pulmonary veins and the left atrium, and thereby minimize postoperative inflammation and subsequent fibrosis formation, resulting in less recurrent obstruction. Despite this strategy, recurrent PVO was seen in 3 of 7 patients without improvement in the anatomic dimension of the pulmonary vein orifice. As we gain more insight into the role of the anatomic relationships between the heart mass and the descending aorta, our surgical strategy is shifting to the concomitant objectives of repairing the site of anatomic narrowing and shifting the site of the pulmonary vein-left atrial junction laterally beyond the descending aorta (Figure 3).

Clinical Relevance of Left-Sided PVO in Single-Ventricle Physiology

Our study suggests that patients with single-ventricle physiology and cardiomegaly are vulnerable for development of left-sided PVO. We speculate that ventricular volume overload, especially after stage I palliation with systemic-to-pulmonary shunt, with associated AVVR and

FIGURE 3. Left lower pulmonary vein before and after sutureless repair. A, Before sutureless repair, the left lower PV is narrowed by compression of the heart and descending aorta. B, After sutureless repair, the confluence of the left lower PV to the atrium is well augmented.

ventricular dysfunction contributes to anatomic relationships that promote potential for PVO, and this potential might be further increased in patients with an antero-laterally displaced thoracic aorta. Although comparison of the end-diastolic and end-systolic volumes between groups did not demonstrate a significant difference, the relative size of the cardiac mass within the context of the size of the thoracic cavity (cardiac/thoracic ratio, Table 2) was greater in the PVO group, suggesting that *relative* cardiomegaly may contribute to the development of PVO.

We did not, however, demonstrate a strong relationship between AVVR and left-sided PVO. The lack of a detectable relationship between AVVR and PVO may be related to the small number of patients in this study. If a relationship among AVVR, relative cardiomegaly, and PVO can be identified in a larger study, then conceivably, indications to intervene

earlier on single-ventricle patients with AVVR and antero-laterally displaced aorta to decrease ventricular size and thereby decrease the propensity to developing left-sided PVO might be contemplated. Furthermore, there may be a patient subset in whom a left thoracotomy can be used to move the descending aorta posteriorly to reduce compression on the left-sided pulmonary veins. We have not yet utilized this approach, but recognize that this technique has been described for tracheal stenosis.¹³ If utilized, we speculate that performing a descending aortopexy early in the PVO process will be important as a means to preempt development of stenotic fibrosis within the lumen of the pulmonary vein.

Limitations

The retrospective nature of this study poses significant limitations in the interpretation of results. Because we

FIGURE 4. Relationship between clinical outcomes and anatomic characteristics. Patients with left-sided PVO (*circle*) had more cardiomegaly and antero-laterally located descending aorta compared with patient without PVO (*diamond*). Cardiomegaly was associated with failed Fontan completion (*closed diamond*). PVO, Pulmonary vein obstruction.

were constrained to limit our analysis to patients who had CMR performed as part of the diagnostic work-up, the study cohort may not be representative of the more general population of single-ventricle patients with PVO. In our more recent practice, however, we nearly always obtain a CMR for patients with suspected PVO. A second limitation is that the control group is relatively small in comparison to the study group. The small size of the control group may have contributed to a failure to detect anatomic dimensions in the control patients without PVO that were similar to those in the PVO patients.

CONCLUSIONS

The combination of relative cardiomegaly within the context of the thoracic cavity at the level of the pulmonary veins and antero-lateral displacement of the aorta is associated with left-sided PVO and subsequent failure to achieve Fontan completion. Further characterization of this unique geometric association may help inform decision making in the timing of interventions (eg, treatment of AVVR and related cardiomegaly) and guide the intraoperative objectives at the time of PVO repair.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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